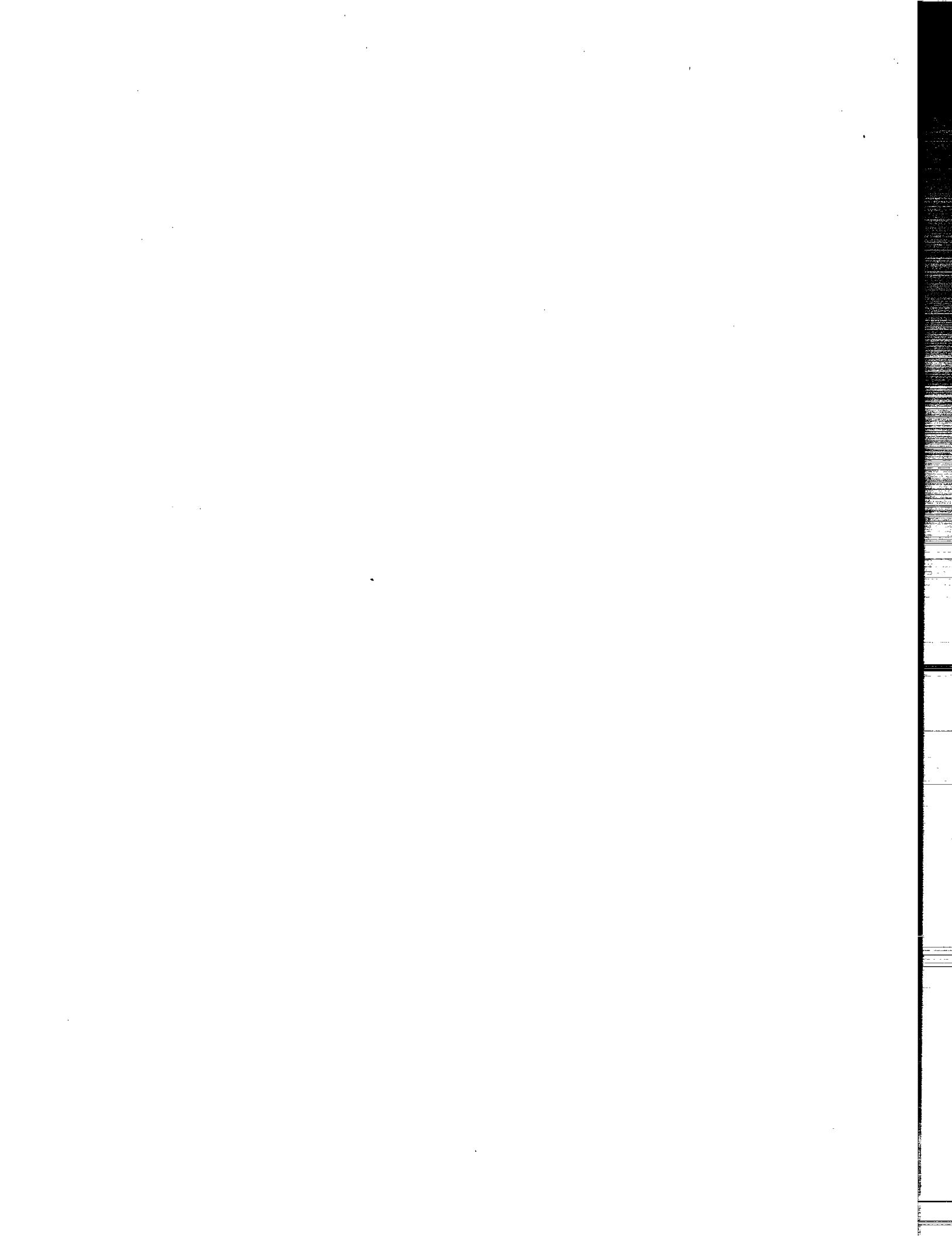


ESSENTIALS OF CARDIAC PHYSICAL DIAGNOSIS



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JONATHAN ABRAMS



ESSENTIALS OF CARDIAC PHYSICAL DIAGNOSIS

ABOUT THE COVER

The heart on the cover of this book is an original drawing by Jim Dine, one of America's outstanding contemporary artists. Since entering the American art scene in the Pop Art movement of the early 1960s, Mr. Dine has earned recognition worldwide. Several themes have reappeared throughout his work over the past two decades, including home and garden tools, bathrobes, trees, and hearts. The heart represents a very personal image for Mr. Dine, and he is perhaps best known for his heart paintings, assemblages, and sculptures.

ESSENTIALS OF CARDIAC PHYSICAL DIAGNOSIS

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Lea & Febiger



Philadelphia

1987

Lea & Febiger
600 Washington Square
Philadelphia, PA 19106-4198
USA
(215) 922-1330

Library of Congress Cataloging-in-Publication Data

Abrams, Jonathan.
Essentials of cardiac physical diagnosis.

Includes bibliographies and index.
1. Heart—Examination. 2. Heart—Diseases—
Diagnosis. 3. Physical diagnosis. I. Title.
[DNLM: 1. Heart Diseases—diagnosis. WG 141 A161e]
RC683.5.A9A27 1987 616.1'207'54 86-20175
ISBN 0-8121-1038-2

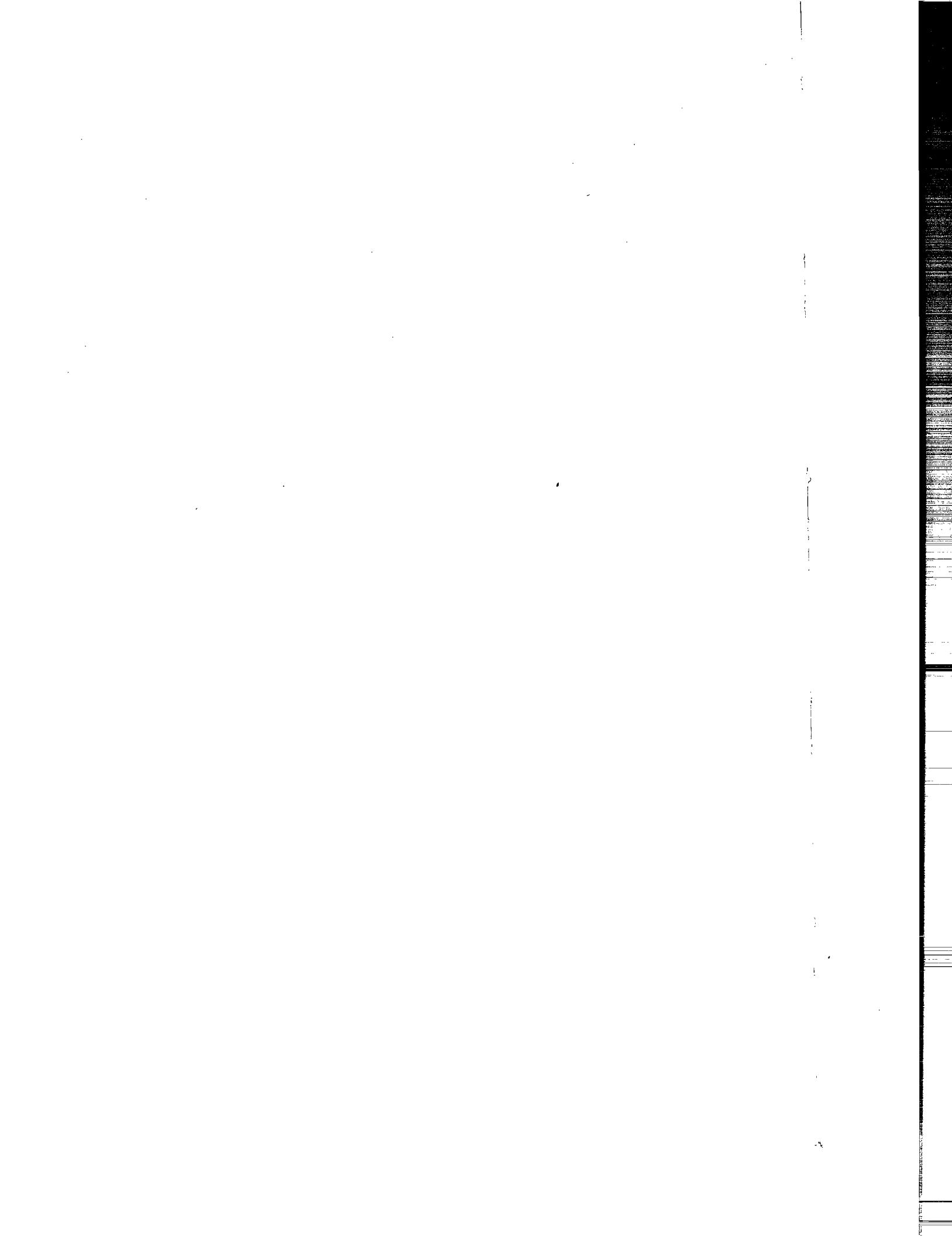
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Printed in the United States of America
Print Number: 5 4 3 2 1

Dedication

I dedicate this book to W. Proctor Harvey, M.D., and Joseph K. Perloff, M.D., two outstanding teachers whose commitment to teaching cardiovascular physical diagnosis is inspirational. They have stimulated and nurtured a devotion to careful bedside skills in hundreds of students. Some, including myself, have aspired to become disciples as well.

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Foreword

I am very pleased to be asked by Dr. Jonathan Abrams to write the foreword to his book "Essentials of Cardiac Physical Diagnosis." In this era of high technology in cardiovascular diagnosis, it is imperative that the clinician recognize that the cardiac diagnostic evaluation begins at the bedside with the cardiac physical examination. This basic diagnostic tool is both an art and a science. The art is based on practice, experience, and attention to detail, while the science has its foundation in the anatomic, hemodynamic, and physical principles that have been so clearly elucidated during the past three decades. In this text, dedicated entirely to cardiac physical diagnosis, Dr. Abrams incorporates both of these elements—the art being a product of years of careful personal observation and experience, and the science based on his thorough understanding of the mechanisms of production of cardiac physical findings.

In the first 12 chapters of this book, the specific findings of the examinations in normal and abnormal cardiac conditions are reviewed, with emphasis on their physiologic bases. In each chapter, the text is complemented by carefully selected phonocardiograms and pulse tracings from the author's personal file as well as from the cardiovascular literature. These recordings of the arterial, venous, and precordial pulsations, together with the auscultatory findings in various cardiac conditions, graphically demonstrate what is seen, felt, and heard at the bedside. The author has also made liberal use of pressure tracings and echocardiograms to give the reader a better understanding of the hemodynamic and anatomic abnormalities responsible for certain physical findings. In the remaining 11 chapters, the physical findings in common cardiac structural abnormalities are reviewed in detail, with the salient feature of each condition graphically displayed by pulse recordings and phonocardiograms.

This book should be a resource for students and house officers of general internal medicine and a definitive reference for the practicing cardiologist. Dr. Abrams' technique of highlighting important clinical points (practical points) is most helpful for the student. For the more seasoned clinician, the extensive correlation of physical findings in various cardiovascular conditions with recent hemodynamic and echocardiographic studies affords further insight into the underlying pathologic process, thereby making the detection of the specific physical finding even more meaningful in cardiac diagnosis. Included in each chapter are many clinical pearls. Such bedside subtleties as

the presystolic murmur of mitral stenosis and its association with short cycle lengths in atrial fibrillation, the absence of augmentation of the murmur of tricuspid regurgitation with inspiration when severe right ventricular failure is present, the technique of palpating a peripheral pulse after the assumption of the upright posture to exaggerate the finding of pulsus alternans, and the decrease in the pulse pressure following a long pause after a premature ventricular contraction in patients with hypertrophic cardiomyopathy are but a few examples of the gems presented in every chapter.

This book is a completely updated reference for cardiac physical diagnosis, in which all of the normal and abnormal cardiac findings of the physical examination are reviewed in great depth and put into appropriate perspective in the light of recent scientific data. The physician who fully appreciates and practices the principles of the cardiac physical examination as presented in this book will be identified as an individual who has truly mastered the discipline of clinical bedside cardiology.

James A. Shaver, M.D.

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Preface

Accurate bedside assessment of heart sounds and murmurs has been a long-standing goal of physicians. In the first part of the twentieth century, physical diagnosis represented the primary mode of evaluating patients with cardiac disorders. The advent of electrocardiography and roentgenography initially had an adverse effect on the importance and interest in auscultation in clinical medicine because these newer techniques seemed more scientific and quantitative to physicians. However, cardiac catheterization, developed in the 1950s, and echocardiography, the major diagnostic advance of the 1970s, have helped renew interest in the cardiac examination and have provided a means of precise correlation of various normal and abnormal cardiac acoustic phenomena with actual measurements of pressure, flow, and motion within the heart.

Echocardiography and angiography have allowed identification and quantification of structural defects and valve abnormalities, which provide an enlightened understanding of the genesis of cardiac sound and murmurs. In almost every instance, the meaning of various palpable and auscultatory cardiovascular events has been delineated by modern diagnostic techniques. The teachings of eminent modern day master clinicians such as Aubrey Leatham, W. Proctor Harvey, Jules Constant, Noble Fowler, Aldo Luisada, Ernest Craige, Morton Tavel, Abe Ravin, and Joseph Perloff have enabled countless students of medicine to utilize intelligently information obtained with the stethoscope. Yet there is a real danger that modern technology will again de-emphasize the role of the stethoscope.

The purpose of this book is to provide the clinician and student with a solid background in cardiac physical diagnosis so that the physical examination extracts the maximum diagnostic information before expensive and extensive tests are ordered. The text is written for medical students, house officers, and practicing physicians. Cardiovascular specialists should find some useful information as well.

Basic cardiovascular physiology is emphasized throughout. Underlying hemodynamic and structural conditions are correlated with abnormal sounds and murmurs heard at the bedside. Nonauscultatory physical diagnosis is highlighted, including examination of the arterial and venous pulses and assessment of precordial motion.

The book is organized into two major sections: Part I consists of a series of 12 chapters that individually deal with selected aspects of cardiac physical

diagnosis (e.g., the arterial pulse, murmurs). The nonauscultatory examination is reviewed first: the blood pressure, venous pulse, arterial pulse, precordial activity. Auscultation is then discussed in systematic fashion: normal and abnormal heart sounds and then cardiac murmurs.

The text opens with a discussion of the cardinal features of cardiac sound in relation to proper stethoscope technique (Chapter 1). The *arterial pulse* (Chapter 2) and *blood pressure* (Chapter 3) are usually evaluated simultaneously, followed by a careful assessment of the *jugular venous pulse* (Chapter 4). *Precordial palpation* (Chapter 5) is the next step, including a systematic evaluation of left and right ventricular areas.

At this juncture, the stethoscope is utilized for a comprehensive evaluation of heart sounds and murmurs. The physician must differentiate normal heart sounds from abnormal ones (Chapters 6 and 7) and should be able to detect the several varieties of extra sounds (Chapter 8) that point to specific intracardiac lesions. Finally, the clinician must detect and evaluate heart murmurs and determine whether a murmur is abnormal or organic (Chapter 9). Systolic murmurs are widespread in the general population and are usually benign (Chapter 10).

The expert diagnostician employs various maneuvers and occasional pharmacologic aids to enhance the information derived from the cardiac examination. These are discussed in Chapter 11.

Although the *general appearance* of the patient is usually only of limited usefulness in cardiac diagnosis, in a number of rare or uncommon conditions information obtained from careful inspection of the patient is most helpful. Dr. Irwin Hoffman thoroughly reviews this subject in Chapter 12.

Part II consists of a series of 11 chapters that deal with the common structural cardiovascular abnormalities that are seen in everyday practice. Chapters 13 through 19 discuss the classic abnormalities of the aortic, mitral, and tricuspid valves; Chapter 14 reviews the unique physical findings in hypertrophic cardiomyopathy. This book emphasizes adult cardiovascular medicine; the only material specifically related to congenital heart disease is in the chapters on atrial and ventricular septal defects (Chapters 20, 21). An entire chapter is devoted to the clinical examination of patients with acute myocardial infarction (Chapter 22) and another to the patient with a prosthetic cardiac valve (Chapter 23).

Thus, the interested reader can use material relating to one or more selected aspects of the general cardiac examination (Chapters 1 to 12), or can proceed directly to a specific valve or structural (Chapters 13 to 23) lesion. The book is intended to be practical, reasonably comprehensive, without burdensome esoteric details. Finally, the text is not meant to be a phonocardiographic atlas; many such excellent monographs are available for the specialist.

TABLE P-1. Common Diagnostic Methods in Cardiovascular Diseases

Electrocardiography
Chest x-ray
M-Mode echocardiography
Two-dimensional echocardiography
Doppler echocardiography
Ambulatory stress testing
Standard treadmill or bicycle exercise
Thallium imaging
Radionuclide angiography (gated blood pool studies)
Cardiac catheterization
Right heart catheterization
Coronary cineangiography
Left ventricular angiography
Electrophysiologic studies

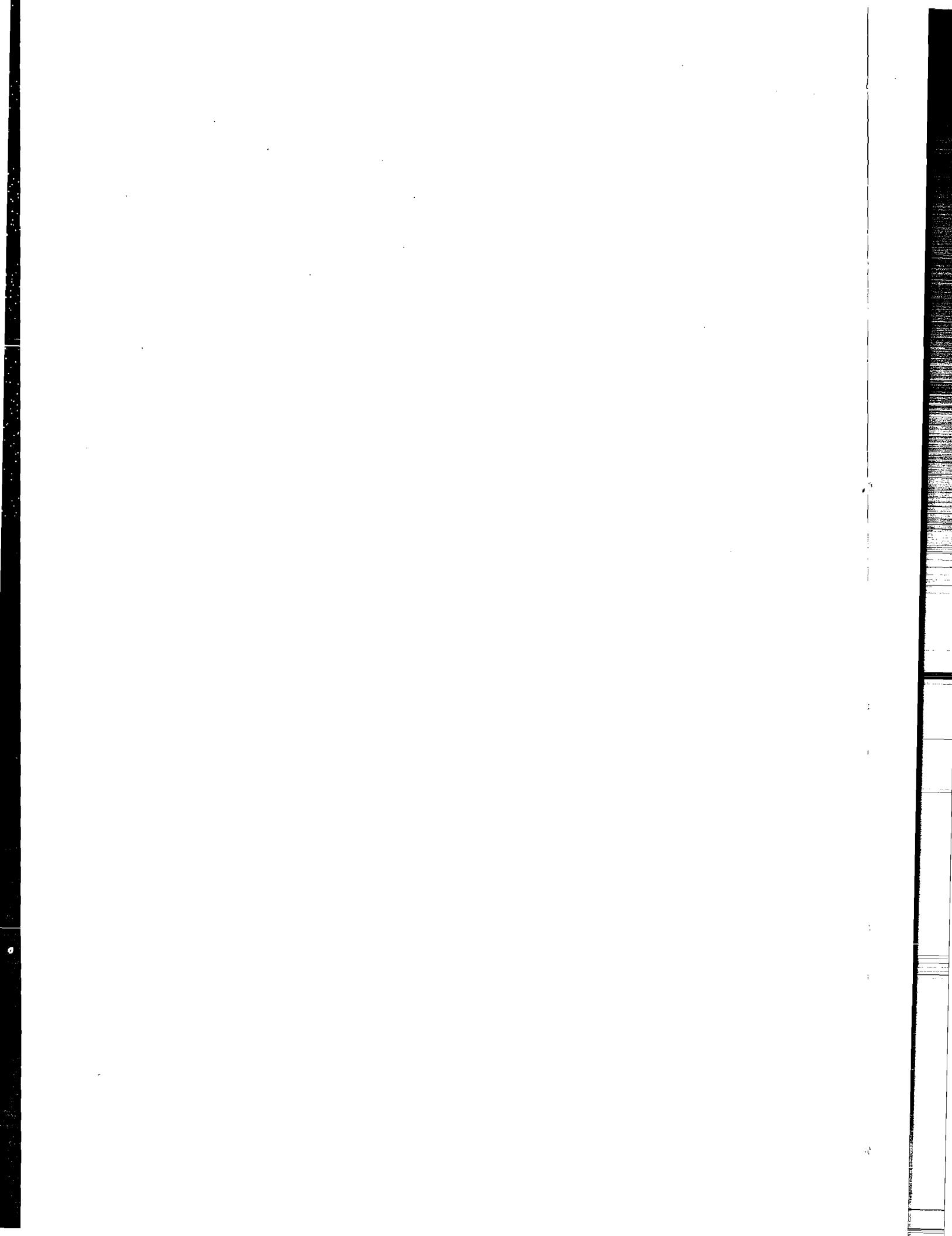
Through a careful and systematic approach to the cardiac physical examination, the physician will be rewarded time and time again with useful diagnostic information. The attentive reader will be able to approach normal persons or patients with structural cardiac disease and derive considerable information about cardiovascular function and anatomy following a careful and complete physical examination. The thorough cardiac examination should help determine appropriate selection of the wide variety of diagnostic tests available for the sophisticated (and often expensive) analysis of cardiovascular structure and function (Table P-1).

Proficiency in cardiac examination is also an end in itself, providing considerable personal fulfillment while enabling the physician to take optimal care of his patient.

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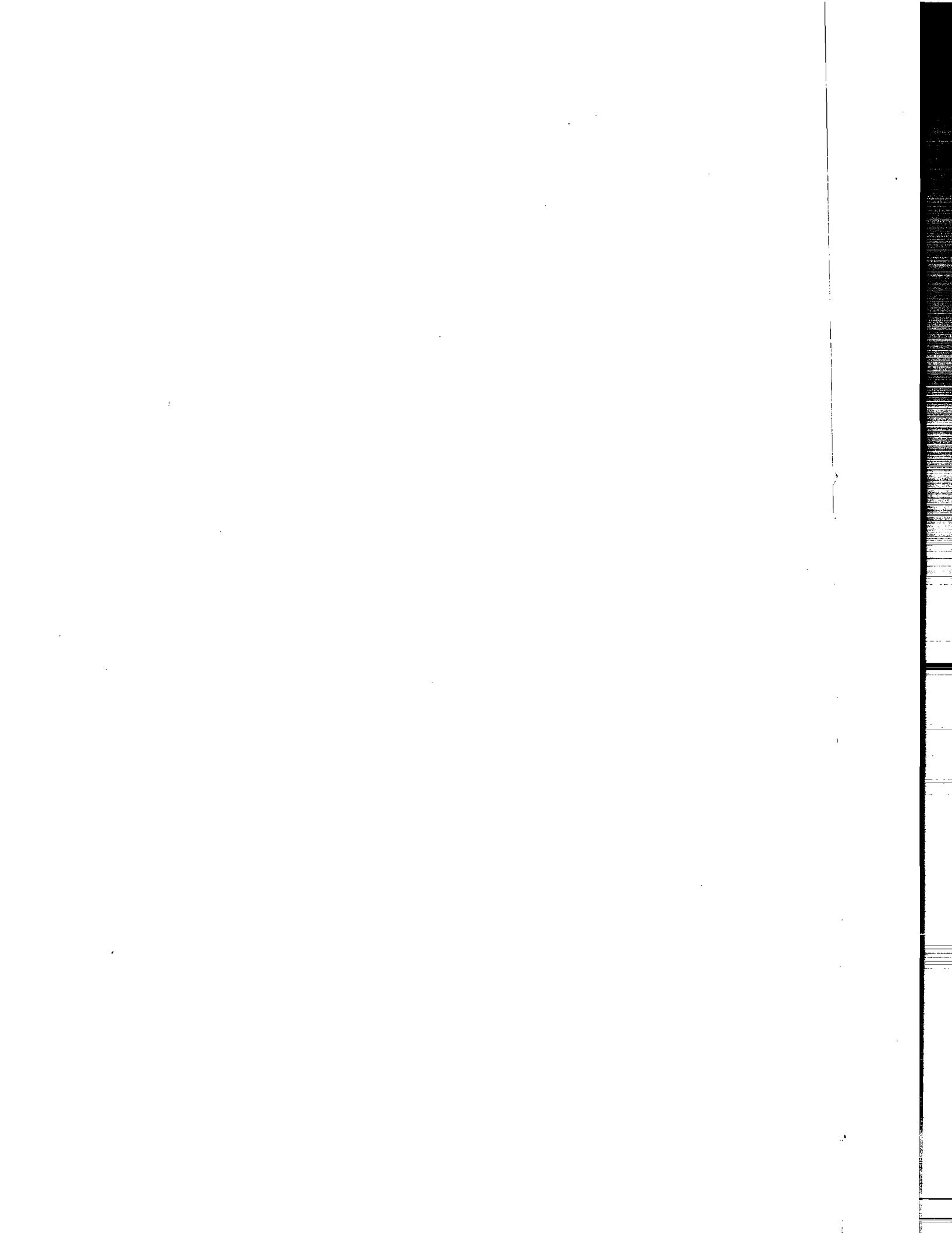
I am indebted to Lori Bennett, Adela Karliner, and Summers Kalishman for assistance in editing and typing the manuscript. Michael Norviel and the Medical Illustration Group at the University of New Mexico School of Medicine have provided invaluable assistance. My secretary, Nancy Jones, has been most helpful in assisting with many tasks related to preparation of the book.

I would particularly like to thank William Parmley, who kindly provided working space and moral support when I first began writing the book during a sabbatical leave. Jim Dine, the world renowned artist, has graciously agreed to provide an original drawing for the cover, thus allowing me to combine my two great passions of medicine and art.

Drs. David Berkson, William Berman, Michael Criley, Grant LaFarge, David Spodick, and Steven Yabek contributed their time in reviewing selective chapters, for which I am grateful. Finally, I acknowledge the assistance of Robert Spencer of *PW Communications*, who allowed me to include in this book modified versions from a series of articles that originally appeared in *Primary Cardiology* between 1981 and 1983. I received excellent medical illustration help from this publisher. Finally, apologies and thanks to all members of my family and colleagues who had to endure the rather long gestation of this project.

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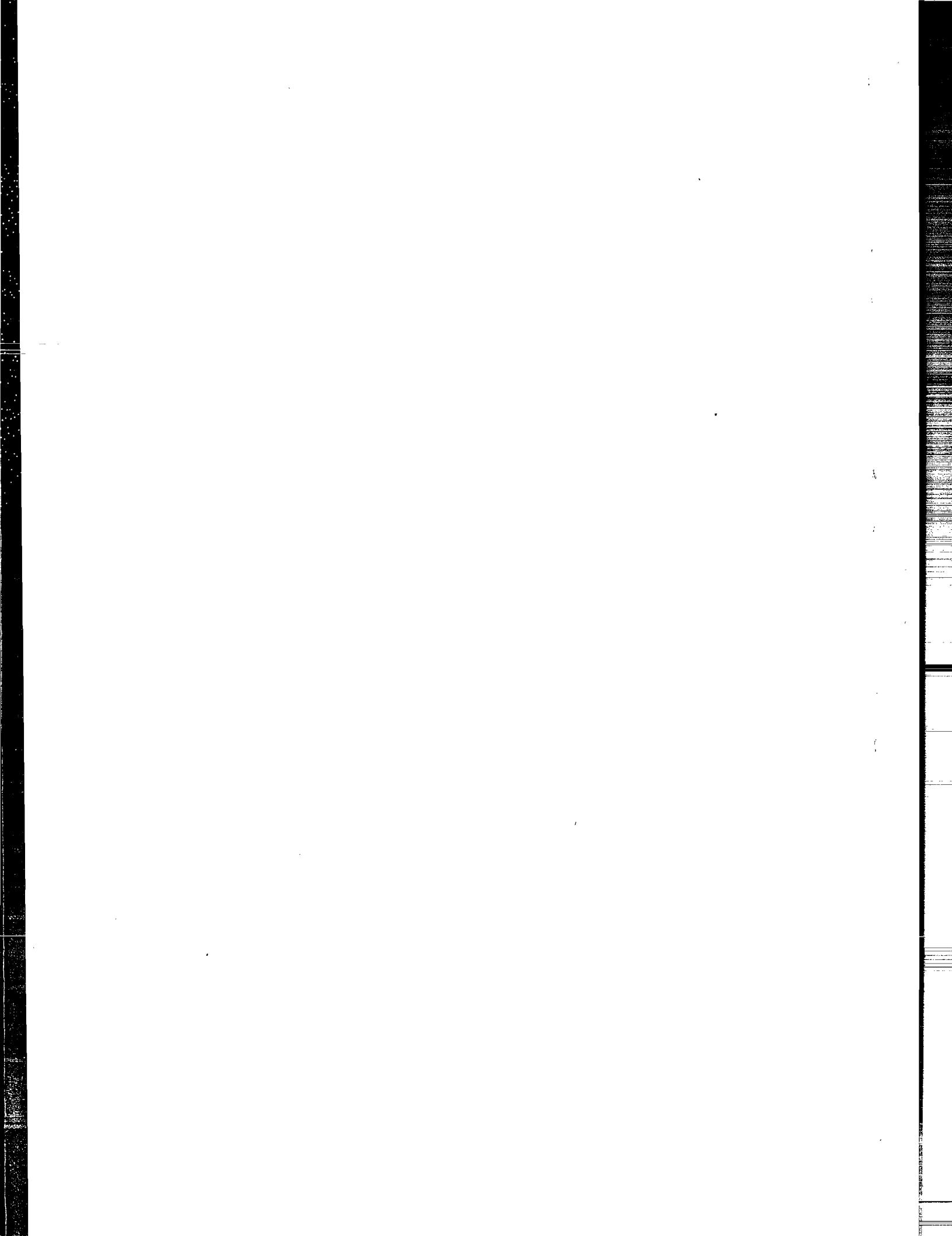
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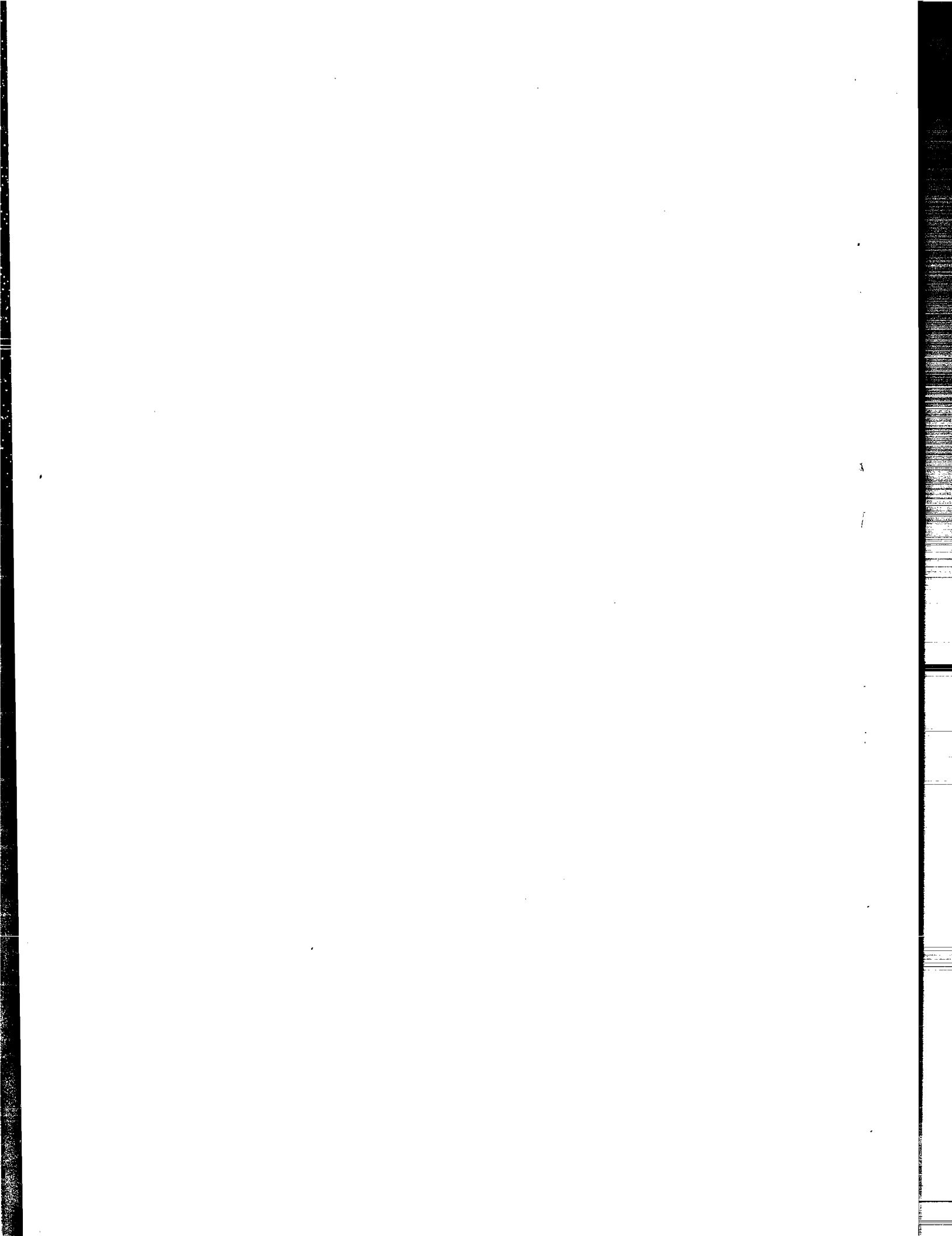
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**PART I BASIC PRINCIPLES OF
CARDIOVASCULAR PHYSICAL
DIAGNOSIS: THE APPROACH TO
THE PATIENT**

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Chapter 1

Cardiac Auscultation: The Stethoscope and Cardiac Sound

It is appropriate to begin this text with a brief discussion of the characteristics of cardiac sound as they relate to proper use of the stethoscope. To best utilize cardiac physical diagnosis, one must understand the basic cardiac cycle fully, and this knowledge must be integrated with the various aspects of the cardiac examination. Detailed discussions of the cardiac cycle and the pathogenesis of the various heart sounds and numbers are found in Chapters 6 through 10.

PERTINENT FEATURES OF CARDIAC SOUND

There are several important features of cardiac sound that require analysis during auscultation. *Loudness* of sound is a subjective judgement and is closely related to the amplitude or intensity of sound waves. The *pitch* of a heart sound or murmur relates to the underlying *frequency*. Soft, rough, low frequency sounds are low pitched (25 to 150 Hz), whereas high frequency sounds are high pitched. The *quality* or tone of a sound relates to the combination of wave form contours, harmonics, and overtones.

For most individuals, the audible range of cardiac sound is approximately 30 to 800 Hz, although the human ear is capable of detecting sound vibrations as high as 16 to 18,000 Hz. The optimal range of auditory acuity is 1000 to 2000 Hz, which is higher than the range of most cardiac sound. Most cardiac sound is low frequency (less than 150 Hz). Very low frequency events (less than 25 Hz) are inaudible but often may be palpable; this explains the occasional finding of palpable atrial and ventricular filling sounds (S4 and S3) in individuals in whom these components are inaudible. In humans, higher frequency (500 to 1000 Hz) events are better heard (e.g., louder) at any level of sound intensity than lower frequency sounds. Most murmurs usually have a frequency between 50 and 500 CPS (see Table 9-6).

Because of the normal ear's decreased acoustic sensitivity at frequencies below 1000 Hz, the examiner more commonly may miss low frequency sounds. On the other hand, low frequency sound can mask higher frequency events; this masking may explain the difficulty in distinguishing both components of S2 or an opening snap following a long rough systolic murmur. However,

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masking is more likely to occur when sounds or murmurs are of the same general pitch; higher frequencies do not usually mask lower frequencies.

Factors such as chest wall thickness, obesity, breast size, and emphysema will affect sound transmission from the heart. In general, anything between the stethoscope and the heart such as adipose tissue, breast, muscular tissue, or lung parenchyma with increased air will attenuate cardiac sound. Lean individuals with little excess body tissue tend to have louder heart sounds and murmurs.

Background Noise. Often unnoticed environmental noise can prevent the detection of heart murmurs, particularly those of high frequency. The level of noise in hospital rooms, clinics, or the typical examining room is 60 to 75 decibels, well above the optimal (but impractical) setting of an ideal soundproof auscultation room (35 decibels). Excessive noise levels in "routine" surroundings can have an even greater impact on cardiac auscultation. If there is an unusual amount of noise (auto or air traffic, construction, human conversation) in the examining area, higher frequency sounds and murmurs may be inaudible even to the most experienced clinicians.

Presbycusis. Young people normally hear high-frequency sounds better than older people. With aging, a selective high-frequency hearing loss is common. Fortunately, this does not have an important impact on auscultation because most cardiac sound is in the low-medium frequency range. Nevertheless, if the high tone deficit of aging drops down to 3000 Hz or less, detection of soft high frequency sounds and murmurs may be impossible. Physicians with such a problem should use a stethoscope that transmits high frequency sound especially well (see below).

THEORY OF HEART SOUND PRODUCTION

Interest in the mechanisms responsible for the production of heart sounds dates back to the beginning era of auscultation. Observant and thoughtful clinicians have debated the origins of various cardiac sounds over the years, and controversy still abounds concerning this fascinating subject. In spite of the technologic advances of the past 20 years (intracardiac phonocardiography, echocardiography, high resolution cineangiography), there are still unresolved questions regarding the genesis of the various cardiac sounds. This chapter will touch on only the most important of such controversies; in most instances, the actual clinical relevance is relatively unimportant.

It is generally accepted that cardiac sounds represent vibrations of cardiac structures and blood within the heart. These vibrations are produced by the acceleration or deceleration of the blood mass during the cardiac cycle. This view, formalized by Rushmer, emphasizes the effects of abrupt changes in pressure and flow within the heart. As a result of changes in momentum and velocity within the fluid-filled elastic system, reverberations of the ventricular

muscle mass, fibrous cardiac skeleton, valve leaflets and their supporting structures, and the blood itself produce *audible sound transients* in the mild to high frequency range. Corresponding low frequency vibrations result in *pressure transients* produced by the same hemodynamic events.

In the past, cardiac sounds were thought to originate from the valves themselves. Because normal valve tissue is not rigid or thick enough to produce sound solely due to leaflet apposition, this simplistic view is not completely tenable. Rather, it is the tensing of the closed valves resulting from rapidly increasing pressure gradients that produces sound vibrations. The heart sounds actually are generated by the leaflets, valve ring, ventricular mass, and blood mass. Sound related to closure of the A-V valves (S1) in part reflects tautening of the chordae tendinae and papillary muscles. Vibrations of the great vessels probably contribute to the opening and closing sound transients arising from the semilunar valves (ejection sounds, S2).

The pressure "crossover" initiating S1 or S2 (e.g., left atrial-left ventricular, aorta-left ventricular) always *precedes* the heart sound itself (Fig. 6-1). Blood flow continues in the face of a pressure "gradient" as a result of inertial forces. The valve leaflets are still in motion at the precise point of pressure crossover (Fig. 7-1).

All cardiac sound then, whether normal or abnormal, results from abrupt accelerating or decelerating forces that stretch the surrounding cardiac structures to their elastic limits. The more rapid these forces, the louder the sound and the higher the frequency. Low frequency sound (S3, S4) typically reflects large amounts of blood moving at low velocity.

THE CARDIAC CYCLE

The four heart sounds are intimately related to electromechanical events within the heart. To make any sense of auscultation, one must understand the basic sequence of the cardiac cycle, which is comprised of *systole*, the ventricular ejection phase, and *diastole*, the ventricular relaxation and filling phase.

Systole. Electrical activation of the ventricles is initiated by the QRS of the electrocardiogram. Depolarization of the left ventricle (LV) begins before that in the right ventricle (RV). The rapidly rising pressure in each ventricle helps close the two A-V valves, resulting in the first heart sound (see Chapter 6). However, because the LV has a far greater muscle mass and must develop a peak systolic pressure 4 to 5 times greater than the RV, it takes longer for the isovolumic pressure within the LV to open the aortic valve than for the RV pressure to open the pulmonary valve (isovolumic contraction time). Thus, although LV electrical activation begins before the RV, pulmonary valve opening and ejection of blood into the pulmonary artery begin before aortic valve opening and ejection into the aorta (Fig. 6-1).

Diastole. During inspiration, aortic valve closure (A2) occurs before pulmonary valve closure (P2), but during expiration the two valves normally close simultaneously (see Chapter 6). Pressure continues to fall in both ventricles after semilunar valve closure (isovolumic relaxation); the A-V valves open passively (and silently) in early diastole when ventricular pressure drops below that in the corresponding atrium. The mitral valve opens approximately 30 msec before the tricuspid valve does.

Rapid ventricular filling follows the maximal opening excursion of the mitral and tricuspid valves in diastole and results in the third heart sound (S3) (Chapter 7). Approximately 70% to 80% of the ventricular end-diastolic volume is achieved during early and mid-diastole, well before atrial contraction. The A-V valves drift back toward their respective atria with a closing motion in mid-diastole, expressed by the E-F slope on the echocardiogram (Fig. 7-1). Following the P wave, the atria contract and the A-V valves reopen, resulting in augmentation of blood flow into the ventricles in late diastole (S4). This atrial booster or atrial transport phenomenon is probably of greater magnitude in the left heart. It gains major importance when either ventricle is diseased; in this setting atrial contribution may represent a much greater portion of ventricular filling than it does in normal hearts. At end-diastole, the A-V valves again begin a closing movement, probably in part due to eddy currents behind the valve cusps (Bernoulli effect) (Fig. 7-1). The leaflets can close as a result of atrial contraction alone. The mitral and tricuspid valves close completely with the onset of ventricular contraction. Ventricular-atrial pressure crossover, however, precedes actual A-V valve closure by 25 to 50 msec (Fig. 6-1).

Heart Sound Summary. The first heart sound (S1) occurs during A-V valve closure and rising intraventricular pressure. The second heart sound (S2) is simultaneous with or occurs immediately following semilunar valve closure and deceleration of blood in the great vessels. The third heart sound (S3) occurs during rapid ventricular filling in early diastole. The fourth heart sound (S4) results from filling following LA contraction. Chapters 6 and 7 discuss the heart sounds in greater detail.

THE STETHOSCOPE

Knowledge of cardiac sound characteristics and their relationship to stethoscope design should be helpful to physicians motivated to obtain the maximum information from auscultation. Although the choice of stethoscope design and manufacturer is relatively limited, it is useful to examine the acoustic and practical characteristics of this indispensable instrument such as sound transmission, frequency filtration, masking, and interference. Many experts feel that the twentieth century stethoscope does not represent a major

advance over the wooden cylinder of Laennec. Electronic and magnetic stethoscopes are available but are unfamiliar to most physicians and will not be discussed.

The stethoscope is an instrument of straightforward design (Fig. 1-1). It consists of a dual *chest piece* with a valve that allows switching from bell to diaphragm, *binaural connectors*, and *earpieces*. The component parts should be well made, durable, without air leaks, and easy to use. A wide variety of satisfactory instruments are available. In selecting a stethoscope, certain important features should be sought. A good stethoscope does not distort cardiac sound. Nevertheless, analytic and integrating auscultation skills are far more important than the brand name of the stethoscope.

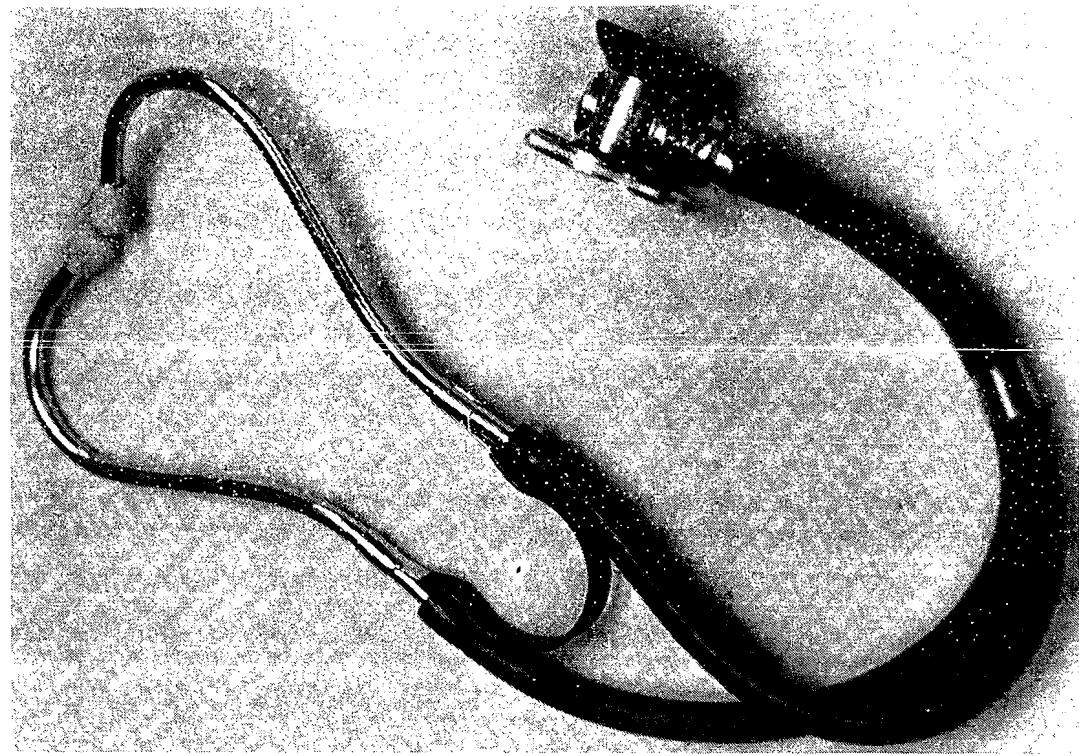
The Bell (Fig. 1-2). The bell is primarily used for detection of low frequency sound (30 to 150 Hz). Lower frequencies are attenuated, and higher frequency sound is accentuated as increased pressure is applied to the bell. Firm pressure with the bell stretches the skin of the chest wall, which then transmits higher frequency sound vibrations; the bell then performs similarly to the diaphragm of the stethoscope. *Practical Point:* To emphasize detection of low frequency sounds (e.g., S3, S4, mitral rumble), the lightest possible contact pressure should be used with the bell.

The bell should have a large diameter (at least 1 inch), a trumpet-shaped internal construction, and should be relatively shallow. A detachable rubber ring is highly desirable for use in conjunction with the bell (Fig. 1-2). This allows light skin contact pressure, minimizes production of an air leak at the skin-bell interface, increases the diameter of the bell to amplify sound, and does not get cold. These features result in optimal detection of low frequency cardiac sound.

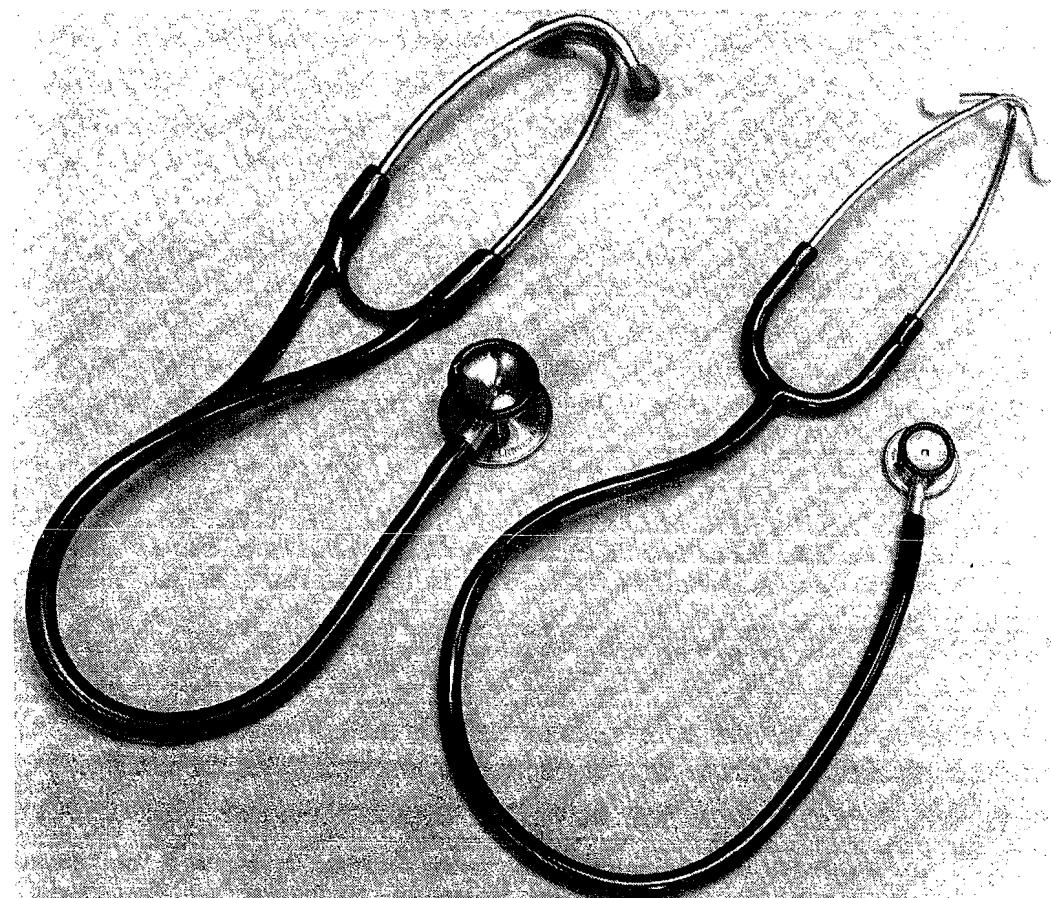
The Diaphragm. The diaphragm is the high frequency component of the chest piece (Fig. 1-3). (Some stethoscopes have two diaphragms, one for medium and one for high frequencies.) A proper diaphragm should filter out low frequencies (less than 300 Hz); often, it will appear to amplify high pitched cardiac sound. As with the bell, increasing pressure tightens the skin and allows transmission of higher frequency sound. The ideal diaphragm should be slightly bowed outward or may have a small central elevated ridge. These features allow for firmer stethoscope-chest wall compression without causing the diaphragm to bulge inward and interfere with sound transmission. This construction can prevent air leaks that result from incomplete stethoscope contact on a thin or bony thorax.

Tubing. The optimal tubing length is 10 to 12 inches; longer tubing may be easier to use but results in sound attenuation. Double tubing is best, as a single or Y-system results in attenuation of high frequency vibrations. This feature is of special importance for individuals with high-tone hearing deficits. Some manufacturers employ a double lumen within a single external tube (Fig. 1-1B). The tubing should be thick and durable to minimize buckling

8 Essentials of Cardiac Physical Diagnosis



A.



B.

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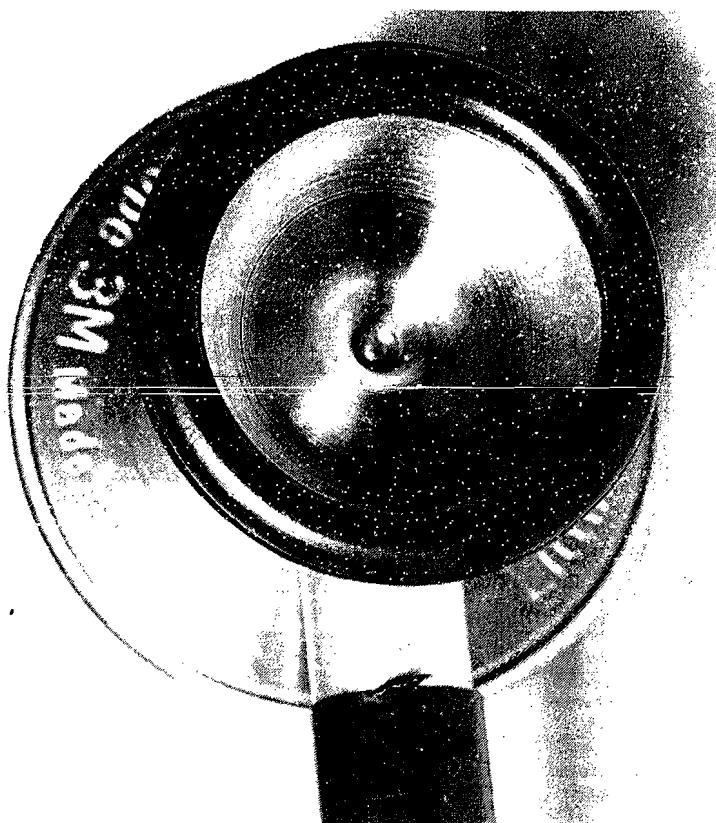


FIG. 1-2. Stethoscope bell. Note the smooth rubber rim attached to the metallic bell. It is advisable to use a rubber ring with the stethoscope bell in order to provide a skin seal with light stethoscope pressure, thus increasing the ability to hear low frequency cardiac sound.

and external noise contamination; plastic is better than rubber. The ideal inner diameter is 1/8 to 3/16 inch.

Earpieces and Binaurals. Probably the most common problem in stethoscope usage is improper fit of the binaurals and earpieces. Poor fit can result in decreased acuity in auscultation. Comfort is of primary importance. Large rather than small earpieces are best. Small earpieces can penetrate too far into the external ear canal and by impinging against the ear canal wall may even become occluded. The concha, or outer half of the ear passage, initially is directed anteriorly and superiorly and then turns posteriorly. The binaurals should be oriented slightly forward and up without creating undue pressure on the ear canal and producing any air leak and must be adjustable for the best fit. Since the conchae are variable in size from person to person and there may be small but potentially important variations between the right and left ear, earpieces of differing size should be tested until a good fit is

FIG. 1-1. The stethoscope. Three examples of commonly used models. The standard components include the chest piece, comprised of a bell and diaphragm; the binaural connectors; and the earpieces. A. The popular Hewlett-Packard double-tubed model. B. Two Littman stethoscopes, the standard adult model and a pediatric stethoscope with a small chest piece.

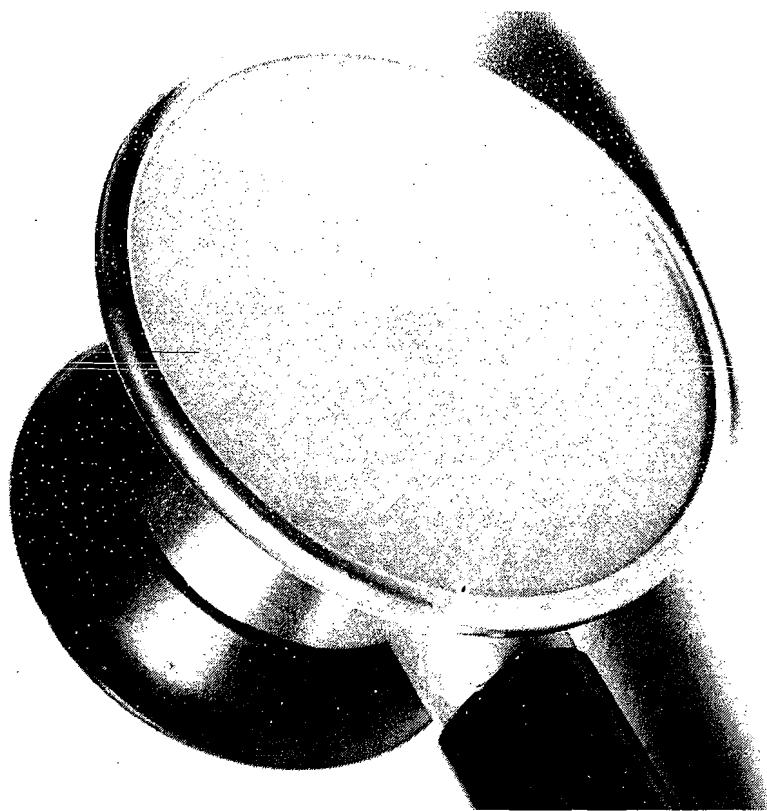


FIG. 1-3. Stethoscope diaphragm. Diaphragms are traditionally composed of a plastic material. Some have a central linear ridge and some are convex to enhance skin contact.

achieved. *Practical Point:* *The use of a poorly fitting stethoscope will result in decreased acuity in cardiac auscultation.*

Air Leaks. Air leaks can occur at the chest piece housing or at the earpiece-ear canal interface. Contamination of cardiac sound with background noise can decrease significantly the ability to hear low intensity sounds and murmurs. In addition, the loss of sound energy through the air leak may result in a decreased amplitude of audible events. Sound at the lower limits of the audible threshold (e.g., the faint diastolic blow of aortic regurgitation, the soft rumble of mitral stenosis) may be completely missed, even by experienced clinicians.

Design, Durability, and Ease of Transport. These factors are of lesser importance but should influence the choice of stethoscope. The chest piece valve mechanism should have a crisp and clean action. From a purely acoustic viewpoint, the best stethoscope is a triple headed device, although many clinicians find this type too heavy and bulky to use in everyday practice. Flexible, lightweight, single tube stethoscopes have become enormously popular in America, but the acoustic properties vary from model to model and should be tested before purchase. Individuals with a high-frequency hearing loss should use a double-tubed stethoscope with a diaphragm of proven capability that attenuates only low pitched sound.

HOW TO USE THE STETHOSCOPE

Identification of Systole and Diastole. Proper identification of the two phases of the cardiac cycle is usually not difficult. During tachycardia, however, diastole shortens more than systole and, with rapid heart rates, it may be difficult to distinguish between the two. When there are extra sounds (particularly an opening snap or midsystolic click), systole can easily be confused with diastole. If there are associated cardiac murmurs, the problem can be even more difficult. *Practical Point: Use the carotid pulse and apex impulse to identify the phases of the cardiac cycle. The carotid upstroke and the beginning outward thrust of the initial apex beat immediately follow S1. S2 occurs shortly after the carotid and apex impulses are felt (Fig. 1-4).* S2 is normally the loudest of the two heart sounds heard at the base of the heart, an area that includes the second right and left interspace adjacent to the sternum.

“Inching.” Levine and Harvey popularized this valuable technique in which the stethoscope is slowly moved or inched from a site on the chest where systole and diastole are identified clearly to other precordial locations where it is more difficult to determine which sound is systolic or diastolic. By focusing on the characteristics of S1 and S2, the cadence of the cardiac cycle enables the observer to identify correctly systole and diastole at other precordial locations. It is best to begin auscultation at the base where systole and diastole usually are most easily identified.

Selective Listening. Selective listening is the hallmark of accurate and meaningful cardiac auscultation. The importance of focusing on only one part of the cardiac cycle at a time during auscultation has been stressed by Dr. W. Proctor Harvey. For instance, when early systole is analyzed, only the acoustic events around S1 are auscultated. The rest of systole and all of diastole are consciously excluded. Systole and diastole should each be divided into thirds—early, mid, and late—and each segment focused upon in turn. Heart sounds should be assessed first, followed by attention to heart murmurs. With this approach, little will be missed. *Practical Point: Intense concentration is the key to competent cardiac auscultation.*

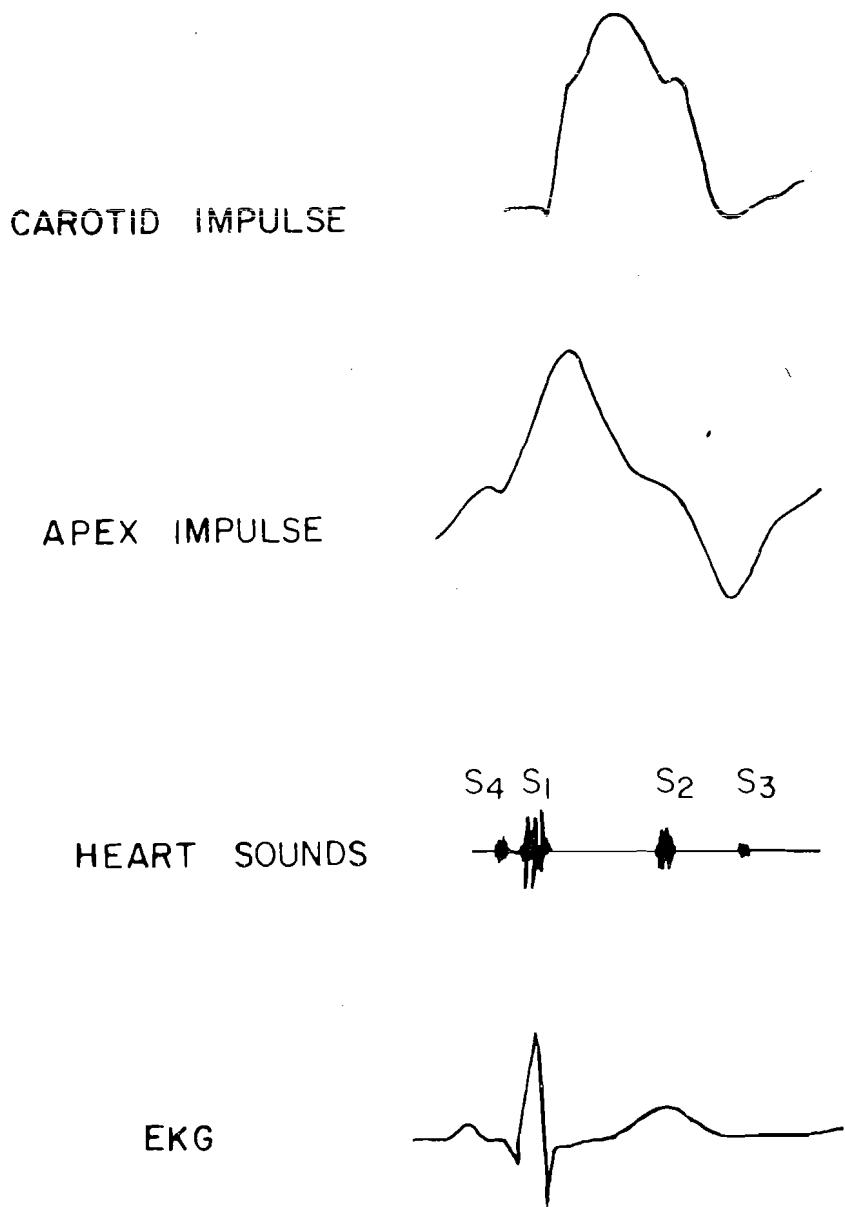


FIG. 1-4. Timing of heart sounds. It is important to use simultaneous palpation of the carotid arterial pulse or the apical impulse when listening to the heart sounds. Note that S₁ immediately precedes the upstroke of the carotid pulse and is virtually simultaneous with the earliest palpable precordial activity. S₂ marks the end of systole and follows the palpable carotid and apical impulse.

Chapter 2

The Blood Pressure

In general, taking the blood pressure is considered a mundane task, too often relegated to personnel with less interest and expertise than the physician. However, in order to obtain accurate blood pressure measurements, it is useful to have an understanding of the physiologic factors affecting arterial pressure, and it is essential to use proper technique. This chapter outlines important information about the physiologic determinants of arterial pressure in health and disease and reviews the appropriate technique for blood pressure determination.

PHYSIOLOGY OF ARTERIAL PRESSURE

Systolic arterial pressure is related to *cardiac factors* (e.g., stroke volume and velocity of left ventricular ejection) as well as to the characteristics of the *peripheral arterial system*, i.e., the elasticity or distensibility of the blood vessels and the volume of blood in the arteries at end-diastole. An increase in stroke volume and/or a decrease in arterial wall compliance (loss of elasticity, vasoconstriction) will result in elevation of systolic blood pressure.

Diastolic arterial pressure is influenced by the level of peripheral resistance to arterial runoff, cardiac cycle length, and the compliance of the arterial tree. Diastolic pressure is lower in conditions of decreased peripheral vascular resistance (i.e., aortic regurgitation, anemia, hyperkinetic states).

Pulse Wave Changes in the Peripheral Circulation. The arterial pulse wave contour changes as the distance from the aortic valve increases (see Figure 3-2). The systolic upstroke becomes steeper, and peak systolic pressure is greater in the distal arteries, although mean arterial pressure remains constant. In peripheral vessels, diastolic and mean arterial pressure decrease and the dicrotic notch disappears. *Practical Point:* *Systolic pressure taken in the legs is normally higher (up to 10 to 30 mmHg) than that in the arms.*

Korotkoff Sounds. These sounds represent arterial oscillations resulting from distention or tensing of the arterial wall with each cardiac impulse due to partial occlusion of the artery by the blood pressure cuff. The Korotkoff sounds are heard with a stethoscope placed directly over the brachial artery just below the lower edge of the cuff (Fig. 2-1). Five phases or categories of Korotkoff sounds are heard after the blood pressure cuff is inflated above systolic pressure and then steadily deflated (Table 2-1).

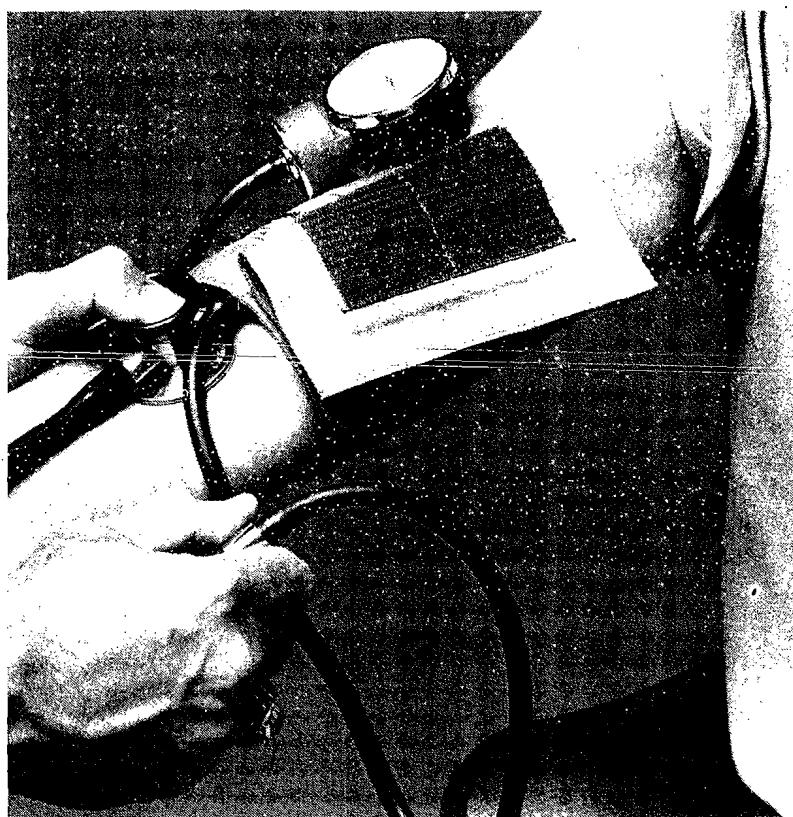


FIG. 2-1. Proper blood pressure recording technique. The cuff should be wrapped tightly around the upper arm just above the antecubital space. Firm pressure with the stethoscope diaphragm or bell must be applied directly over the brachial artery. The upper edge of the stethoscope diaphragm should touch or fit just under the distal edge of the cuff. The patient's arm should be positioned at midchest level.

Phase I. These are the first sounds detectable as the declining intra-bladder pressure approximates peak systolic arterial pressure. These initial sounds are relatively soft and rapidly increase in intensity. Phase I sounds persist for 10 to 15 mmHg as the cuff is deflated. The level at which phase I Korotkoff sounds first are heard is considered the systolic pressure.

Phase II. This phase is initiated by a murmur-like sound. Shortly after these Korotkoff sounds are heard, they may fade and transiently be undetectable as cuff pressure continues to fall. This results in a silent or *auscultatory gap*, which is important only if missed by the examiner who fails to inflate the cuff to sufficient pressure. Such an error results in a falsely low systolic blood pressure estimation. Phase II extends over a pressure range of 15 to 20 mmHg.

Phase III. Sounds in this phase are easily heard.

Phase IV. The Korotkoff sounds rather abruptly change pitch and intensity at the point of muffling (Phase IV) which typically occurs at a slightly higher arterial pressure than true diastolic blood pressure.

Phase V. As cuff pressure is further lowered, the Korotkoff sounds

disappear completely when arterial flow is no longer restricted by the surrounding cuff. This occurs at a level slightly below true intravascular diastolic pressure and best approximates true diastole in most instances.

When there is high velocity of blood flow, Korotkoff sounds can be heard even with the cuff completely deflated. Commonly this occurs in children, during or after exercise, in anemia, with aortic regurgitation, or in other hyperkinetic states. In such situations phase IV (muffling) best reflects true diastolic pressure.

Korotkoff sounds may be quite soft when the left ventricular ejection rate is reduced, e.g., poor left ventricular function or reduced limb blood flow. Thus, with patients in shock, with circulatory collapse, or with severe aortic stenosis, Korotkoff sounds may be very difficult to hear.

Postural Changes in Blood Pressure. Systolic blood pressure may fall by as much as 10 to 12 mmHg when a normal subject abruptly sits (legs dangling) or stands. In general, diastolic pressure remains the same or increases slightly. A decline in systolic pressure greater than 15 mmHg in the upright position is called *postural* or *orthostatic hypotension*. It is useful to check routinely for postural hypotension in patients with syncope, dizziness, or postural symptoms suggesting cerebral hypoperfusion, especially in elderly subjects. In addition, patients with endocrine causes of hypertension (pheochromocytoma, primary aldosteronism) may have an abnormally large orthostatic decrease in systolic blood pressure similar to some chronic diabetics with autonomic neuropathy. An excessive fall in systolic pressure in

TABLE 2-1 *Korotkoff Sounds*

Phase	Description	Comment
I	Onset of auditory sound	<ul style="list-style-type: none"> Represents peak systole Initially low intensity
II	Swishing sound or murmur	<ul style="list-style-type: none"> Occurs at least 10 to 15 mmHg below peak systolic pressure levels, or with a larger difference in hypertensives Auscultatory gap occurs when phase II sounds are soft, absent, or missed by the examiner
III	Crisp, easily heard sounds	<ul style="list-style-type: none"> 15 to 20 mmHg below phase II
IV	Abrupt dampening or muffling of tapping sounds	<ul style="list-style-type: none"> Although previously thought to be true diastole, typically occur 5 to 8 mmHg above intra-arterial diastolic pressure
V	Disappearance of sound	<ul style="list-style-type: none"> Current recommendations: Use as true diastolic pressure unless vasodilatation or aortic regurgitation is present

the upright position suggests relative hypovolemia or an abnormality in vasoconstrictor regulatory control such as that found in autonomic dysfunctional states.

MEASURING TECHNIQUE

Blood Pressure Cuff or Sphygmomanometer (Fig. 2-2). This is a pneumatic device for measuring the arterial pressure indirectly by allowing a controlled occlusion of the underlying artery. Either an aneroid or mercury manometer is connected to a distensible rubber bladder or balloon encased in a nondistensible outer cover (cuff). The length of this bladder should be at least 75 to 80% of the arm circumference. The cuff itself should be long enough to comfortably wrap around the arm. Table 2-2 lists the appropriate

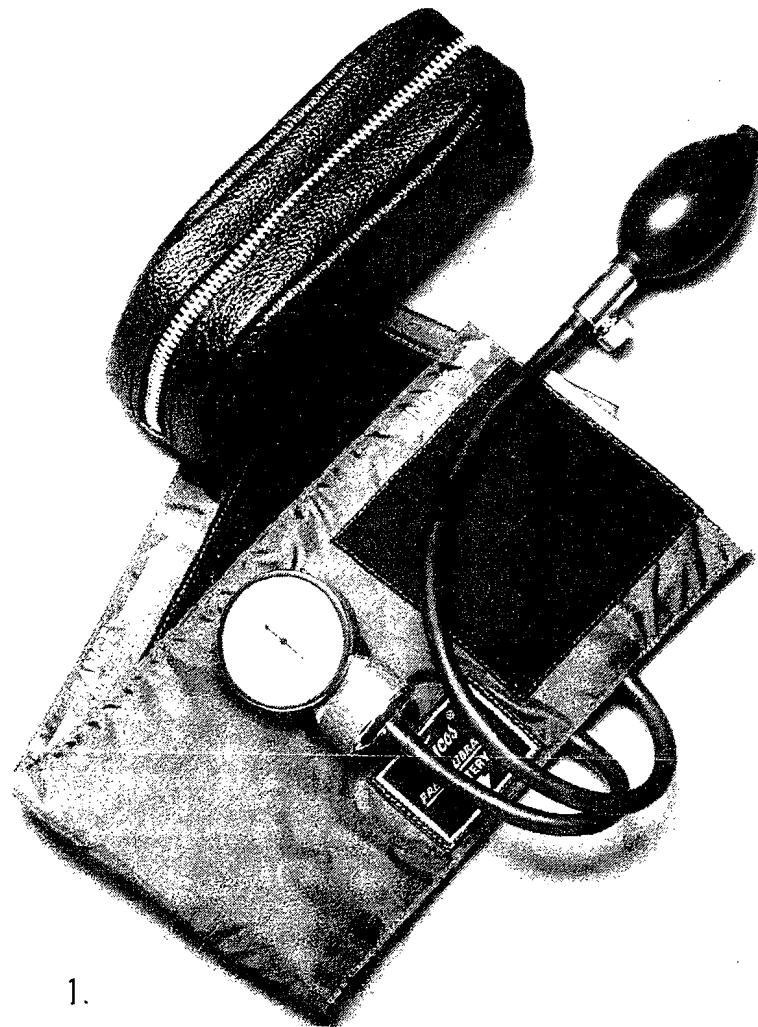


FIG. 2-2. Sphygmomanometer. A popular model that is easily transported in a small carrying case. Note the pressure gauge and inflatable bulb. (Courtesy of Ritter-Tycos.)

TABLE 2-2 *Optimal Sphygmomanometer Cuff Size (Centimeters)*
(Modified from American Heart Association's 1980 Recommendations)

Cuff Name	Arm Circumference (Midarm)	Bladder Width	Bladder Length
Child	13-20	8	13
Small adult	17-26	11	17
Standard adult	24-33	12-13	22-24
Large adult	33-42	15-17	32-33
Thigh	42-50	18-20	35-42

cuff selection for different arm sizes. The cuff width should be at least 20% greater than the arm diameter. Many commercial cuffs are too narrow and too short and may result in a falsely elevated blood pressure reading. A ratio of cuff width to length of 1:2 is optimal for most subjects, although the standard cuff (12 × 24 cm) is not long enough for some adults. Leg cuffs should be at least 15 cm in width and more than twice that in length (preferably 18 × 36 cm). Cuffs for infants and children come in variable widths ranging from 2.5 to 9 cm. Figure 2-3 demonstrates the variety of cuff sizes that are available.

The Patient. The preferred time to take the blood pressure is when the subject is relaxed. Patients' concerns about the blood pressure reading combined with fear of the physician or the office setting may cause considerable anxiety and often will result in a transiently elevated high blood pressure. The simultaneous heart rate can serve as a rough guide to the patient's emotional state. True basal blood pressure is difficult to obtain in an office or hospital. Recent cigarette smoking, exposure to cold, bladder distention, or tight clothing can affect the blood pressure. Patients should be instructed not to talk during the blood pressure examination.

Automated ambulatory blood pressure recordings have clearly documented that casual pressure determinations in the physician's office are considerably higher than multiple measurements taken at work or at home, in both normal and hypertensive subjects. Home pressure levels are lower than those at work, and sleeping blood pressures are much less than those while awake. On the other hand, people are daily subjected to stressful situations, and although elevations related to anxiety may indicate "labile" hypertension, it is likely that such blood pressure levels are representative for that individual. A transient elevation of blood pressure also may reflect the ease with which an individual's pressure becomes elevated; therefore, an elevated reading is a warning that hypertension is a potential medical problem. *Practical Point: It is useful to take the blood pressure both at the beginning and at the end of the examination, or only at the conclusion of the cardiac examination. More than likely, the subject will be calm and at ease by then and no longer preoccupied with the actual level of the blood pressure.*

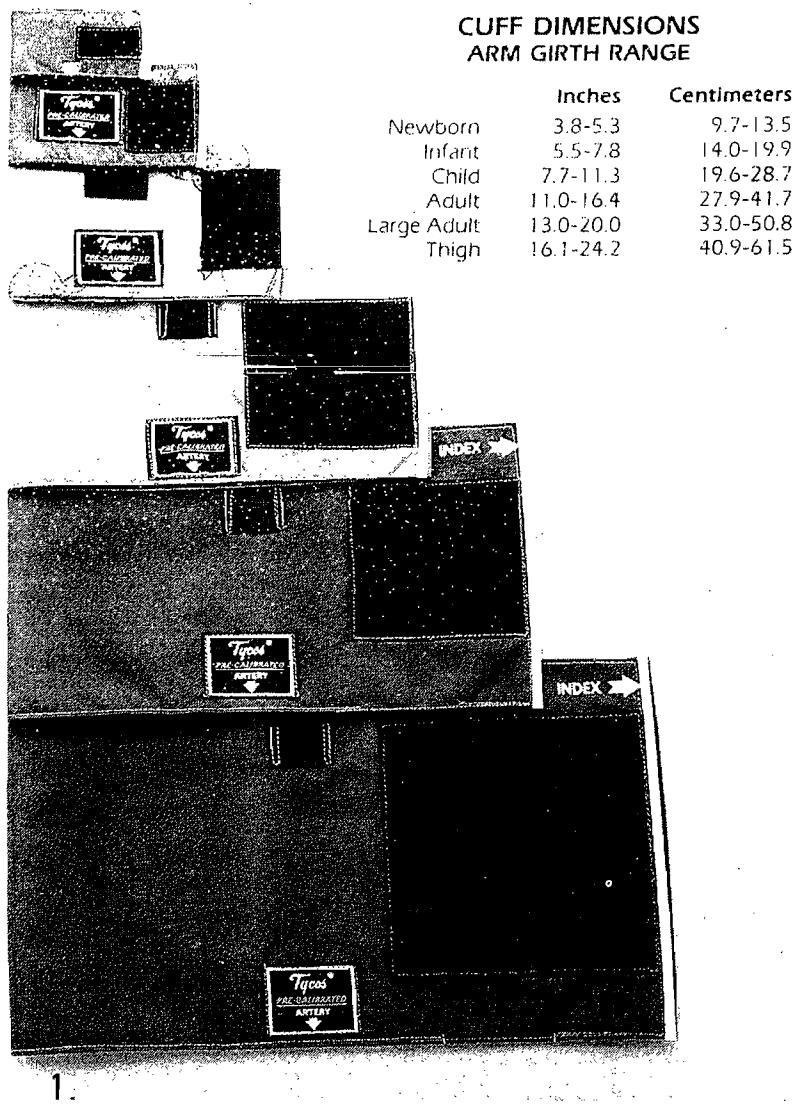


FIG. 2-3. Available models of blood pressure cuffs. A variety of cuffs are manufactured to obtain an optimal fit for differently sized arms. Note the large adult cuff, the thigh cuff for leg blood pressure determinations, and a series of cuff sizes for infants and children. There are standard recommendations for the sphygmomanometer cuff and bladder sizes, but manufacturers often ignore these guidelines. (Courtesy of Ritter-Tycos.)

In recording the blood pressure, the subject may be supine or sitting. The patient's arm must be at the level of the heart when nonrecumbent pressures are taken; gravity will significantly affect the blood pressure reading if the brachial artery is more than 10 cm lower or higher than the heart itself. The true pressure will be underestimated if the arm is above the level of the heart. For optimal stethoscope contact with the brachial artery, the arm should not be flexed. The cuff should be *firmly* wrapped around the distal upper arm with the edge of the cuff positioned just above the antecubital crease (see Figure 2-1). A loosely applied cuff will result in falsely high readings. The center of the inflatable bladder is placed squarely over the

artery, which first should be identified in its course by palpation just beneath the biceps muscle. For optimal auscultation of the low frequency Korotkoff sounds it may be preferable to use the bell of the stethoscope. This should be positioned close to or just under the cuff's distal edge.

How to Use the Sphygmomanometer. The cuff should be briskly inflated approximately 20 mmHg above peak systolic pressure; deflation should be somewhat less abrupt to avoid errors in detecting the actual maximum systolic pressure. The recommended decompression rate is 2 to 3 mmHg every heart beat or per second. The cuff pressure is slowly and constantly deflated until Korotkoff sounds are no longer audible. *Practical Point: After each cuff inflation, it is important to deflate the cuff completely and allow sufficient time for venous flow and congestion in the arm to be restored to normal.* Forearm venous congestion can falsely elevate the diastolic blood pressure. A tense arm with an elevated local venous pressure will decrease the amplitude of the Korotkoff sounds and accentuate the auscultatory gap, thus increasing the chance of error in identifying systolic pressure. Korotkoff sounds often are quite soft in congestive heart failure; careful technique is mandatory in such patients.

Systolic Pressure. Peak systolic pressure first may be estimated by palpation through placement of a finger or thumb on the ipsilateral brachial or radial artery as the cuff is inflated, noting the level when pulsations are no longer palpable. This method will ensure that the systolic blood pressure measured by auscultation will not be underestimated because of the auscultatory gap or too rapid deflation of the cuff, which could result in the earliest Korotkoff sounds being missed. This technique enables the examiner to avoid unnecessarily high cuff pressures that can cause arm discomfort during subsequent cuff reinflations.

The level of peak systolic pressure is identified by the auscultatory method when two consecutive Korotkoff sounds are heard. Normally, systolic pressure obtained by palpation is a few mmHg lower than the pressure measured by auscultation, but whichever is greater should be considered the actual systolic pressure. Awareness of the possibility of an auscultatory gap should preclude the examiner from missing the true maximal peak systolic pressure; this "silent" range is greater in hypertensive patients in whom such an error is clinically more important.

Diastolic Pressure. This has been the most problematic area in indirect blood pressure recording. The point of muffling (phase IV) previously had been recommended by the American Heart Association to be taken as true diastole for adults; this is normally higher (5 to 10 mmHg) than true intra-arterial diastolic pressure. The most recent (1980) AHA recommendations equate disappearance (phase V) of Korotkoff sounds as the true diastolic blood pressure. Usually, this is slightly lower than actual intra-arterial diastolic pressure. The point of muffling may be subtle and difficult to identify precisely.

In conditions with a high cardiac output and low peripheral resistance, the Korotkoff sounds may be well heard beyond true diastole even to zero pressure. This is common in exercise, anemia, and aortic regurgitation.

There is less interobserver disagreement when disappearance of sound is used as the indicator of diastolic pressure. Recent experimental studies support the use of disappearance of sound. Most experts and the AHA recommend recording the diastolic pressure at *both muffling and disappearance* of sound (e.g., 120/74/64). A good rule of thumb is to use muffling as the true diastolic pressure only when the discrepancy between muffling and disappearance is greater than 10 mmHg. In most people the point of muffling (phase IV) and disappearance (phase V) are close together.

In infants and children, phase IV (muffling) more closely approximates true arterial diastolic pressure. This parameter should be used for blood pressure measurements in all children.

Shock. Because of the low stroke volume and marked peripheral vasoconstriction found in most patients with shock, palpatory and auscultatory systolic pressure may be difficult or impossible to detect. True blood pressure in shock, measured by intra-arterial recordings, is often much higher than cuff pressure, particularly when there is a high systemic resistance and reduced blood volume. *Practical Point: Korotkoff sounds are very weak in shock. They can be enhanced by several maneuvers. Elevation of the arm immediately before cuff inflation will accentuate Korotkoff sounds, as will having the patient rapidly clench his fist 5 to 10 times after the cuff is blown up.* Both maneuvers will exaggerate the pressure gradient across the cuff and intensify the Korotkoff sounds. Rapid cuff inflation should be used to avoid trapping blood in the forearm. In hypotensive states, the stethoscope diaphragm or bell should be placed under the distal cuff edge prior to inflation for maximal sound transmission. In many cases of severe hypotension, however, it is best to insert an arterial line or needle, since arm cuff pressures are too unreliable.

The Fat or Large Arm. In patients who are particularly fleshy or who have arms with an excessive circumference (more than 35 cm), a normalized cuff may be too narrow to effectively occlude the brachial artery, resulting in a blood pressure reading that may be artificially high. This is a common source of error and results in "pseudohypertension." A number of studies have documented false elevations of systolic and diastolic pressure of 4 to 10 mmHg when a fat or large arm is undercuffed. A wider and longer than normal cuff (large adult cuff or thigh cuff) should be used in these cases (see Figure 2-3, Table 2-2). In such situations the systolic blood pressure should first be measured by palpating the radial pulse. Wrap a regular cuff around the forearm if a wide cuff is unavailable. Manning has suggested that one mark a vertical line on the sphygmomanometer 32 cm from the left border on the standard adult cuff, and 42 cm from the left border with the

large adult cuff in order to avoid "undercuffing" a large or fat arm. These values represent a maximum upper limit of 75% of the appropriate arm circumference.

Practical Point: If the blood pressure is normal in an overweight subject, hypertension is essentially excluded. When a hypertensive reading is found in a subject with a large upper arm (circumference greater than 33 to 35 cm), a large adult cuff must be used to be sure the pressure is truly elevated. In undersized arms (children, thin adults) a normal cuff may be too large, and blood pressure readings will be falsely low. Child and infant cuffs are available (Fig. 2-3). Common errors in routine blood pressure examination are listed in Table 2-3.

THE COMPLETE EVALUATION

In many clinical situations it is advisable to perform all or most of the following maneuvers for a thorough analysis of the arterial pressure and the detection of all possible important abnormalities. These additional approaches are not indicated in the routine examination of young or apparently healthy individuals without evidence of cardiovascular disease.

Postural Changes. The blood pressure should first be taken in the supine position after the subject has been resting quietly for at least 5 minutes. If orthostatic hypotension is suspected clinically and symptoms or a drop in pressure do not immediately occur in the upright position, wait 3 to 5 more minutes with the patient standing or sitting before rechecking the pressure. Occasionally, vasomotor or autonomic instability is manifest only after a protracted time. A drop of more than 12 to 15 mmHg in systolic pressure and any measurable fall in diastolic pressure in the standing position are

TABLE 2-3 *Common Sources of Error in Blood Pressure Determination*

-
- Failure to detect peak systolic pressure
 - insufficient level of initial inflation
 - too rapid initial cuff deflation
 - failure to recognize an "auscultatory gap"
 - Cuff applied too loosely
 - Fat or large arm: Cuff width too narrow or not long enough
 - Thin arm: Cuff width too large
 - Excessive venous pressure and congestion in the arm
 - decreased intensity of Korotkoff sounds
 - widened auscultatory gap
 - Arm above or below heart level
 - Variable intensity of Korotkoff sounds during arrhythmias
-

abnormal and suggest hypovolemia or autonomic nervous system dysfunction. The heart rate response to standing should also be noted.

Right and Left Arm. Bilateral arm blood pressures should be determined in the *initial* evaluation of most patients, particularly if there is hypertension or other evidence of vascular disease. Normally, there is not more than a 5 to 12 mmHg discrepancy in systolic pressure between the two arms and up to 5 mmHg difference in diastolic blood pressure. Obviously, the distal arm pulses must be carefully evaluated if a disparity in arm pressure is noted. Table 2-4 lists conditions that commonly give rise to significant differences in the right and left arm blood pressures. If there appears to be a large discrepancy between the two arms, multiple follow-up readings should be done in an attempt to quantitate this difference.

Leg. Measuring pressures in the leg is indicated in the evaluation of hypertensive subjects, particularly if there is any suspicion of coarctation of the aorta or in patients with possible peripheral vascular disease. The technique is basically the same as with the arm: an extra-wide cuff is wrapped around the lower third of the thigh and auscultation performed over the popliteal artery. Systolic blood pressure in the legs is normally higher than in the arms, whereas diastolic pressure is approximately the same.

Diurnal Variation. It has been shown in normal subjects and hypertensive individuals that circadian or diurnal changes in blood pressure occur in a predictable fashion (Fig. 2-4). Blood pressure is lowest during sleep and begins to rise abruptly between 7 and 8 AM. It remains higher during the day without major changes and begins to fall between 6 and 9 PM, reaching a nadir between 1 and 4 AM.

TABLE 2-4 *Differential Diagnosis of Discrepancy in Blood Pressure Between Right and Left Arm or Between the Arms and the Legs*

Arterial occlusion: embolus, atherosclerosis, Takayusu's arteritis, post-cardiac catheterization, aortic arch syndrome, subclavian steal syndrome

Dissecting aortic aneurysm

Thoracic outlet syndrome

Abdominal, iliac, or femoral aneurysm

Supravalvular aortic stenosis

Scalenus anticus syndrome

Coarctation of aorta

Patent ductus arteriosus

Error in technique

Normal or physiologic fluctuations of blood pressure

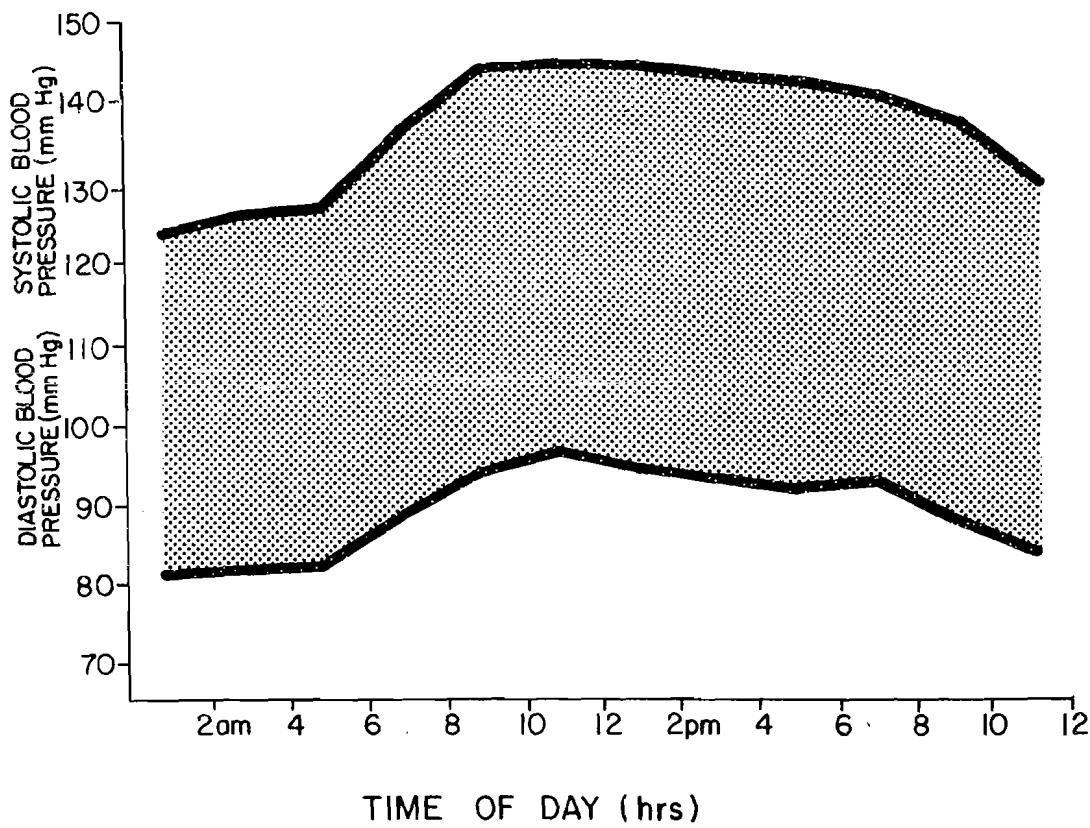


FIG. 2-4. Diurnal alterations in blood pressure. Both systolic and diastolic blood pressure rise from the early morning hours to reach maximal levels later in the day, declining in late afternoon or early evening. The nadir of blood pressure is achieved during sleep. While the data shown here were obtained from a hypertensive population, the diurnal pattern of blood pressure in normotensive subjects follows an identical course. (Adapted from Drayer J. et al.: Am J Med 73:497, 1982.)

SPECIFIC ABNORMALITIES

Hypertension. In order to make a diagnosis of essential or secondary forms of hypertension, both diastolic and systolic blood pressure must be elevated on *repeated* examinations (at least three basal determinations at different times). There is a normal rise of blood pressure with age, but too often physicians accept an elevated blood pressure in older subjects as "physiologic." The range for normal levels of blood pressure at various ages is listed in Table 2-5.

Isolated Increase in Systolic Pressure. In conditions associated with an increased left ventricular stroke volume, systolic pressure is often elevated. Examples include readings during or following exercise, in moderate to severe aortic regurgitation, and with very slow heart rates. Aging results in decreased compliance of the arterial tree due to loss of elasticity and/or arteriosclerosis; "stiffer" systemic arteries produce an increased pulse pressure with a rapid

TABLE 2-5 *Normal Upper Limits of Blood Pressures for Age**

Age	Pressure
14-18	130/90
over 18	140/90
over 65	160/95

* Diagnosis of hypertension should be made only after elevated readings under basal conditions on 2 or more separate occasions.

upstroke and elevated systolic pressure. Isolated systolic hypertension of the elderly is common. Systolic pressure may reach 160 to 180 mmHg in older individuals while diastolic pressure remains below 90 mmHg. Such patients are definitely at increased risk for stroke and myocardial infarctions.

Hypotension. In hypovolemia, shock, or vasomotor collapse, the blood pressure is low. In addition, the combination of weak Korotkoff sounds and generalized vasoconstriction of non-shock etiology may result in an apparently decreased cuff pressure that can simulate shock or a more severe hypotensive state than is actually present. Methods to enhance auscultation of Korotkoff sounds (see page 20) should be employed in "hypotensive" states. A direct arterial pressure determination often is necessary to resolve a conflicting or confusing clinical picture.

Pulsus Alternans and Pulsus Paradoxus. These conditions will be discussed more fully in Chapters 3 and 13. Careful and slow (2 to 3 mmHg/sec) deflation of the cuff is mandatory to detect these abnormalities. Deliberately slow cuff decompression should be used in the initial workup of any patient suspected of having cardiovascular disease. Detection of pulsus paradoxus or alternans by blood pressure measurement can be of vital diagnostic benefit. In *pulsus alternans*, diminution in the intensity of every other Korotkoff sound is noted at or near peak systolic pressure. The second (weaker) beat may not even be audible with sudden or gradual "doubling" of the pulse rate being noted as cuff pressure is lowered (see Figure 3-4). Pulsus alternans is seen with severe left ventricular dysfunction.

Pulsus paradoxus is an exaggeration of the normal decline in systolic pressure during inspiration (see Chapter 3). A paradoxical pulse is diagnosed by observing the level that Korotkoff sounds are heard only during expiration when peak systolic pressure is first identified. The level of systolic pressure at which Korotkoff sounds are heard both in inspiration *as well as* in expiration establishes the degree or magnitude of the paradoxical pulse. In normal subjects during quiet respiration, the difference between expiration and inspiration is no more than 6 to 8 mmHg. Pulsus paradoxus classically is found in pericardial tamponade, but may be present in severe congestive heart failure, asthma, or chronic obstructive lung disease.

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Chapter 3

The Arterial Pulse

Examination of the arterial pulse often reveals valuable information about the cardiovascular system. In certain conditions, such as aortic valve disease, hypertrophic cardiomyopathy, and pericardial tamponade, an accurate assessment of pulse contour and amplitude can be of great importance in making a proper diagnosis. This chapter will emphasize how to extract the most data from *careful bedside examination* of the arterial pulse. As with the venous pulse, carotid pulse recordings can provide more detailed information than that obtained from inspection and palpation. However, many physicians are intimidated by the detailed pulse wave analysis and terminology; high quality graphic records are increasingly difficult to obtain. This chapter will focus on what can be felt with the examining fingers and will not discuss pulse recordings.

NORMAL PHYSIOLOGY

The arterial pulse wave is related to many physiologic factors, including the left ventricular stroke volume and ejection velocity, as well as the relative compliance and capacity of the arterial system. The pulse contour is derived in part from a series of frequency waves that are produced by antegrade blood flow and from reflections of the arterial pulse returning from the peripheral circulation. The relative distensibility or "stiffness," of the arterial system affects both the contour and velocity of the arterial pulse.

The first portion of the central aortic pulse wave (Fig. 3-1) reflects peak velocity of blood ejected during early systole, which is "stored" in the central aorta. The mid to late systolic portion of the normal pulse wave is produced by blood moving from the central aorta to the periphery simultaneously with a reflection of the pulse wave returning *from* the arteries of the upper body. Arterial pressure is transmitted with a velocity 10 times greater than that of arterial blood flow.

The early systolic component of the arterial pulse often is called the percussion wave, and the later peak is known as the tidal wave. These terms have little physiologic or descriptive meaning. The arterial pulse contour is best described in terms of "early" and "late systolic" and "diastolic" components. With aging or decreased compliance of the arterial tree, the later portion of the pulse wave is accentuated, and the pulse contour becomes

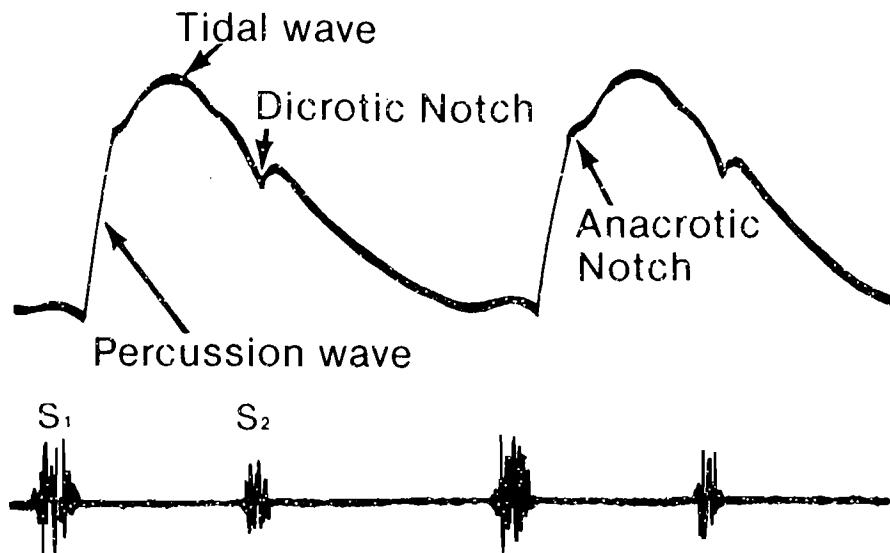


FIG. 3-1. Normal arterial pulse. Note the rapid upstroke, the rounded summit or peak, and the falloff in late systole. Normally, only the systolic peak is palpable; diastolic events are not felt. The dicrotic notch times precisely with S₂ and is coincident with aortic and pulmonic valve closure. The terms *percussion wave*, *tidal wave*, and *anacrotic notch* are discussed in the text. (From Abrams J: Prim Cardiol, 1982.)

somewhat more sustained. The normal change in pulse velocity and contour that occurs in early to midsystole often produces a notch or change in slope on the pulse contour, which is called the anacrotic notch or shoulder.

Diastole is initiated by an abrupt negative wave called the dicrotic notch (Fig. 3-1). The nadir coincides precisely with aortic leaflet closure and the aortic component of S₂ (A₂). *Practical Point:* *The anacrotic and dicrotic notches normally cannot be felt but are easily recorded. Do not expect to palpate them when examining the arterial pulse.* The positive wave following the dicrotic notch most likely represents reflection of the pressure pulse wave from the arteries in the lower half of the body. Normally, the arterial pulse is not palpable during diastole.

In certain cardiovascular conditions, a palpable double peaked or bifid arterial pulse is present with the two peaks occurring in systole; rarely, a "dicrotic pulse" is felt with the second peak occurring in diastole (see page 30).

Pulse Wave Alteration in the Periphery. The normal pulse contour is altered with increasing distance from the aortic valve. The reflecting transmitted waves in the peripheral arteries reverberate and result in a greater amplitude and velocity of the arterial pulse wave and a lower dicrotic notch; these changes become progressively more pronounced distally (Fig. 3-2). *Practical Point:* *The peripheral arteries (e.g., radial, femoral) should not be used to routinely assess the arterial pulse contour. Normal physiologic pulse wave alteration in distal vessels may mask important diagnostic information present in the proximal vessels.* On the other hand, the augmented rate of rise

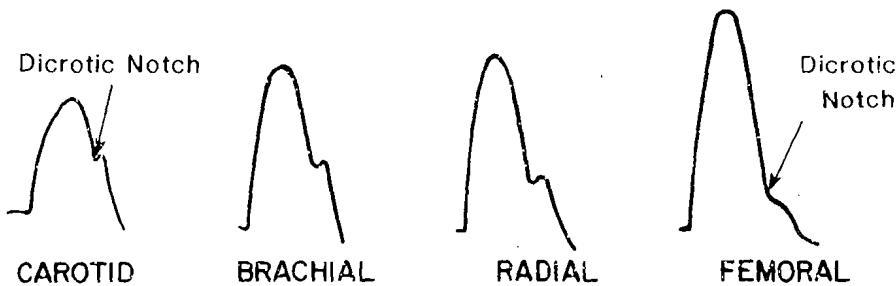


FIG. 3-2. Arterial pulse contour alteration in the peripheral circulation. As the distance increases from the peripheral artery to the central aorta, the amplitude and upstroke velocity of the pulse contour also increase and the dicrotic notch becomes lower. The higher systolic pressure in peripheral arteries is one reason why the arterial pulse is best evaluated at the carotid artery rather than distally. (From Abrams J: Prim Cardiol, 1982.)

and amplitude of the pulse in peripheral vessels increases the ability to detect the subtle abnormalities of pulsus paradoxus and pulsus alternans (see below).

Pulse Wave Alterations with Decreased Vascular Compliance. In states of increased vascular resistance or stiffness such as occur with hypertension and vasoconstriction, the relatively noncompliant arterial tree contributes to an increased pulse wave velocity. This increase results in a pulse contour with a rapid upstroke and greater amplitude. The thickened arterial walls of the aged or atherosclerotic subject may produce the same effect. Under such circumstances, a small or relatively normal LV stroke volume may be transmitted to the carotid or distal pulses with a brisk upstroke, giving a false impression of the actual stroke volume. *Practical Point: In older subjects, hypertensive patients, or individuals with diffuse vascular disease, information obtained from examination of the arterial pulse is less reliable. By way of contrast, in the presence of intense arterial vasoconstriction, the palpable pulse volume may appear to be reduced when in fact the ejected stroke volume is normal.*

THE EXAMINATION

Technique. Both the patient and the physician should be relaxed. The subject's head and thorax should be slightly elevated at 15 to 30 degrees. Either the thumb or the first two fingers are used to feel the arterial pulse; the finger pads usually are quite sensitive for optimal palpation. Gentle but firm pressure is necessary, with the thumb or finger placed directly on the summit of the vessel. To obtain maximal information, the palpating pressure should be varied. Always be certain that you are feeling the patient's pulse and not your own. Excessive pressure often reduces tactile sensation. It is best to press down and then slowly decrease the palpating pressure until the arterial pulse contour and amplitude are best appreciated. Surprisingly light

pressure may be needed once direct contact with the blood vessel has been established. Arteries may be quite tortuous in some patients.

It helps to create a "mental image" of the pulse wave while analyzing the quality of the early rise, peak, and drop-off. The normal pulse has a brief crest that is slightly sustained and somewhat rounded (Fig. 3-1). Clinical expertise in evaluating the normal and abnormal arterial pulse comes with experience and a conscious effort to analyze the pulse volume and contour.

Which Artery to Palpate? In a complete cardiovascular examination, all accessible arterial pulses should be assessed. It is important to compare bilateral vessels. A missing or markedly weak pulse may have major significance, suggesting atherosclerosis, embolic occlusion, dissection, vascular compression, or a congenital anomaly. The brachial artery is often buried beneath the biceps muscle and may be difficult to localize. The right carotid artery may appear dilated in older subjects, particularly if there is hypertension; this is usually due to an innocuous kinking or buckling of the innominate vessels. In any patient with hypertension or in infants with heart failure, the radial or brachial artery and the femoral artery *must be palpated simultaneously* to exclude coarctation of the aorta. Normally, the femoral pulsation immediately precedes the radial pulse; in coarctation, the femoral arterial pulse is distinctly delayed and reduced in volume when compared to the arm pulses.

Use the Carotid Artery. The carotid arterial pulse provides the most accurate evaluation of the arterial pulse volume and contour because it is the largest palpable proximal vessel, it is the closest accessible artery to the aortic valve, and its contour closely resembles the directly recorded central aortic pulse. On the other hand, distal vessels are affected by the characteristic waveform alterations that occur in peripheral arteries (Fig. 3-2). Thus, diagnostic abnormalities of the central arterial pulse may be attenuated or completely disappear in the peripheral circulation.

In examining the carotid artery, careful attention should be paid to the lower half of the neck. The adjacent sternocleidomastoid muscle should be under no tension, and pressure should not be placed on the carotid sinus itself.

Characteristics. In examining the arterial pulse, the following characteristics should be routinely noted:

- Cardiac rhythm
- Pulse volume (generally related to the size of the stroke volume in the absence of vasoconstriction)
- Pulse amplitude
- Pulse contour, with special attention to the crest or peak of the pulse
- Speed or rate of rise of ejection (early systole)
- Stiffness or distensibility of the vessel wall

Occasionally one can feel a shudder or thrill in the carotid pulse representing a palpable bruit or transmitted murmur. Localized atherosclerosis or abnormalities of the left ventricular outflow tract account for most carotid thrills. A hyperkinetic circulation, as is often found in normal children or patients with aortic regurgitation, may produce a slight "buzz" to the carotid pulse. A loud murmur of ventricular septal defect or pulmonic stenosis occasionally radiates to the neck and produces a carotid thrill.

In older subjects or those in whom there is a suspicion of vascular disease, both the carotid and femoral arteries should be routinely auscultated and palpated. Some cardiac murmurs that intensify in the upper chest or clavicular areas and are well heard along the sternal edge are easily heard over the carotid arteries. A venous hum can be heard in most younger subjects and should be differentiated from a true arterial bruit. Rotate the neck to the left and right and listen to the subject in both supine and upright positions to assess the possibility of a venous hum, which will characteristically wax and wane with these maneuvers (see Chapter 4).

ABNORMAL VARIANTS

Hyperkinetic Pulse

A hyperkinetic arterial pulse has a larger than normal amplitude and results from an increase in left ventricular ejection velocity, stroke volume, or arterial pressure. The pulse wave amplitude may also be augmented in persons with decreased arterial compliance, such as arterial wall thickening or systemic vasoconstriction. However, vasoconstriction may also result in a small pulse (see below). A hyperkinetic arterial pulse may result from cigarette smoking or any state of enhanced sympathetic activity; it is common in elderly subjects and usually implies an elevation in systolic blood pressure. Bounding or hyperkinetic pulses are typical of high output states, such as may be present with anxiety, anemia, thyrotoxicosis, exercise, a hot and humid environment, and alcohol intake.

Hypertension may cause a more forceful arterial pulse if the pulse wave velocity is increased, particularly if there is associated generalized arteriosclerosis. A hyperkinetic pulse is typical of aortic regurgitation where the runoff of the pulse may be very rapid, producing a bounding or collapsing pulse. Other high output states with increased distal arterial runoff, such as a patent ductus arteriosus, large A-V fistulae, Paget's disease, or severe cirrhosis, may simulate the classic pulse of aortic regurgitation. In severe aortic regurgitation, both the pulse amplitude and the volume may be greatly increased to the extent that the full and bounding pulsations are visible to the

clinician from the foot of the bed (Chapter 15). In hypertrophic cardiomyopathy an early brisk pulse wave may be felt in the carotid arteries. In severe mitral regurgitation the arterial pulse may also be brisk and tapping.

HYPOKINETIC PULSE

Small or diminished pulses are common in states of low cardiac output. The decreased left ventricular stroke volume results in a shorter left ventricular ejection time because there is less blood to eject. Impairment of LV function or overt congestive heart failure also is associated with a decreased velocity of ejection. Hypotensive states and LV outflow tract obstruction usually result in a small pulse volume. Intense vasoconstriction may reduce the pulse amplitude in the face of a normal stroke volume. *Practical Point:* *It is important to assess the rate of rise of the hypokinetic pulse. A normal upstroke (unsustained) indicates a decreased stroke volume without LV outflow obstruction, whereas a slow rising (sustained) pulse of small volume strongly suggests aortic stenosis.* The pulse in hypertrophic cardiomyopathy usually has a normal volume and is quick rising.

DOUBLE OR TWICE-BEATING PULSE (Table 3-1)

A double or twice-beating pulse occurs when there are two palpable arterial pulses per cardiac cycle. Usually, this double peak occurs in systole, such as the bisferiens pulse of aortic valve disease or the bifid pulse of hypertrophic cardiomyopathy. Much less commonly, the first palpable wave occurs in systole and the second in diastole: this is known as a dicrotic pulse. *Practical Point:* *Simultaneous auscultation and palpation are essential to the proper timing of the double arterial pulses.*

Bisferiens Pulse (Fig. 3-3A). This double-peaked systolic pulse wave is more easily detected in the brachial or radial artery than in the carotid

TABLE 3-1 *Causes of a Double Arterial Pulse*

Type of Pulse	Causes
BISFERIENS PULSE	aortic regurgitation combined aortic stenosis and regurgitation compensated tight aortic stenosis (uncommon) high output states
BIFID PULSE	hypertrophic cardiomyopathy ("spike and dome")
DICROTIC PULSE	cardiomyopathy or severe LV dysfunction pericardial tamponade febrile states in young subjects

artery. The bisferiens (or bisferious) pulse may be missed easily when strong palpating pressure is used. Light but firm compression on the vessel wall using a single finger or the thumb will aid in discovery of this important abnormality. The bisferiens pulse is found in pure aortic regurgitation or combined aortic stenosis and regurgitation with dominant regurgitation (see Chapter 15). In subjects with aortic regurgitation, the bisferiens pulse is more likely to be present with a large stroke volume and may disappear with the onset of congestive heart failure. A bisferiens contour occasionally may be detected in high output states in subjects with a normal heart.

Bifid Pulse. A bifid or "spike and dome" arterial impulse is commonly recorded but infrequently palpated in hypertrophic cardiomyopathy (Fig. 3-3B). Early systolic emptying of the LV is abnormally rapid in this condition and results in a brisk, tapping arterial pulse wave. A midsystolic pulse collapse may be followed by a second, late systolic impulse. The bifid pulse indicates significant outflow "obstruction" (see Chapter 14).

Dicrotic Pulse. The dicrotic pulse, an unusual finding, occurs when the *diastolic* pulse wave is conspicuously accentuated so that it becomes palpable (Fig. 3-3C). The detectable delay between the two palpable peaks is much longer than in the bisferiens pulse. On auscultation, S2 separates the two components of the dicrotic pulse. The systolic component of the dicrotic pulse is usually slow rising and small in volume, correlating with a reduced stroke volume.

A dicrotic pulse occurs most commonly in young persons with poor LV function (e.g., cardiomyopathy). It is typically associated with a low cardiac output, low blood pressure, high systemic vascular resistance, and tachycardia. A dicrotic pulse has been described as occurring during inspiration in pericardial tamponade and following valve replacement for aortic or mitral regurgitation; in these situations it appears to indicate poor LV function. It may be more noticeable in the beat following a premature ventricular contraction (PVC). Pulsus alternans (see below) and S3 and S4 gallops commonly occur in patients with a dicrotic pulse who have underlying pump dysfunction.

Some observers believe that a dicrotic pulse may be found occasionally in young healthy individuals with fever, although this is controversial. In general, a dicrotic pulse is not observed in subjects over the age of 45, probably because an elastic arterial tree is a prerequisite for its production.

PULSUS ALTERNANS

Pulsus alternans is present during sinus rhythm when the patient's peak systolic arterial pressure and pulse volume are alternately strong and weak; every other impulse is less forceful (Fig. 3-4). This phenomenon is related to a beat-to-beat alternation in developed LV pressure caused by changing ejec-

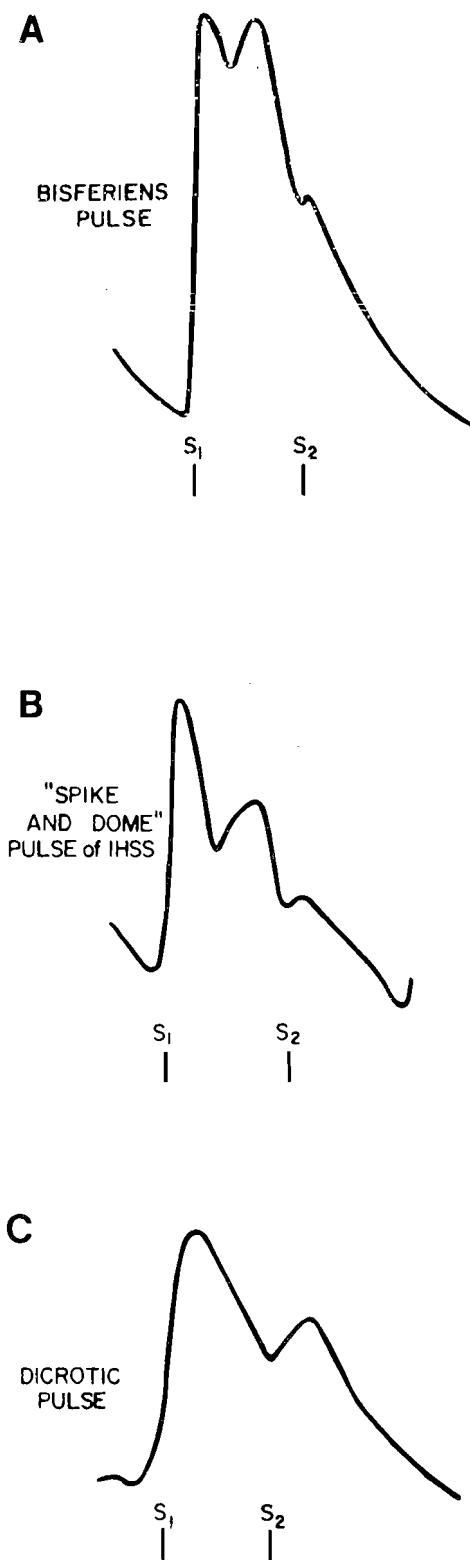


FIG. 3-3. Double-peaked or bifid arterial pulses. A. Bisferiens pulse, commonly found in severe aortic regurgitation or mild aortic stenosis with moderate aortic regurgitation (see also Chapter 15). B. "Spike and dome" arterial pulse of hypertrophic cardiomyopathy or IHSS. This unusual contour is recordable but not readily palpable (see Chapter 14). Note: The twice-beating bisferiens and spike and dome pulses are felt in systole. C. Dicrotic pulse. In this waveform the second palpable component is a diastolic reflection wave. This is frequency noted in the setting of young patients with low cardiac output and impaired left ventricular function. (From Abrams J: Prim Cardiol, 1982.)



FIG. 3-4. Pulsus alternans. Note that every other beat has a lower systolic pressure. The rate of rise of the second pulse is slower, relating to decreased contractile force in the alternate beats. Pulsus alternans is an important sign of severe left ventricular dysfunction. It is best detected in a peripheral pulse, such as the radial artery. Heart sounds and murmurs may also alternate in intensity (see also Fig. 13-3). (From Abrams J: Prim Cardiol, 1982.)

tion dynamics. *Practical Point:* *Pulsus alternans is a subtle abnormality that will not be detected unless carefully sought. Its presence always indicates severe left ventricular dysfunction.*

Pulsus alternans is best detected at the brachial or radial arteries by application of light pressure with the fingers; the carotid artery is not recommended for palpation in this setting. The use of two fingers to feel the arterial pulse is suggested, with variation of the pressure on the proximal finger in order to obliterate the second (weaker) beat. The rate of rise of the pulse wave, which is often decreased in the second beat of each pair, should receive careful attention. To help detect pulsus alternans, some clinicians use slow decompression of the blood pressure cuff while simultaneously listening for alteration in the intensity of the Korotkoff sounds. Because pulsus alternans indicates deranged LV function, an associated S3 gallop and other signs of LV failure commonly are found. Pulsus alternans usually is accentuated for several beats following a PVC. It is more easily detected during the Valsalva maneuver or with any sudden decrease in LV stroke volume. Having the patient abruptly assume the upright position or take a deep inspiration may be useful in "bringing out" pulsus alternans.

On occasion, heart sounds and murmurs may also be noted to alternate in intensity with every other beat. *Pulsus alternans* is not related to *electrical alternans*, a beat-to-beat variation in the amplitude of QRS complexes on the electrocardiogram that may occur in large pericardial effusions.

PULSUS PARADOXUS

Pulsus paradoxus refers to a marked and exaggerated inspiratory fall in systolic blood pressure in which the palpable peripheral arterial pulse and audible Korotkoff sounds *disappear during inspiration*. Paradoxus is a classic finding in pericardial tamponade (see below).

Because systolic arterial pressure normally falls slightly during inspiration, the word "paradoxus" is actually a misnomer, but its continued use is dictated by widespread use of this term. Normally, LV stroke volume decreases somewhat during inspiration because of diminished left heart filling;

the inspiratory decrease in intrathoracic pressure causes pooling of blood in the pulmonary bed. Systemic arterial pressure thus falls a few mmHg during inspiration. Right ventricular (RV) stroke volume, however, increases with inspiration, as the large inspiratory pressure gradient between the extra- and intrathoracic veins enhances right heart filling. During expiration, this increased RV stroke volume is transmitted across the pulmonary vascular bed to the left heart. This increases LV stroke volume and systolic arterial pressure, which are further augmented by the small expiratory rise in intrathoracic pressure. The combination of these complex physiologic events results in modest respiration-related alterations in systolic blood pressure in normal subjects.

Inspiration normally reduces systolic arterial pressure less than 6 to 8 mmHg; if the inspiratory fall is more than 8 mmHg, pulsus paradoxus is said to be present. In general, however, the fall in inspiratory pressure should approach 10 to 12 mmHg before a definite diagnosis of paradox is made. In severe cases, the fall in blood pressure during inspiration may be 30 to 40 mmHg or more.

Causes. Several mechanisms result in abnormally large phasic alterations in RV and LV stroke volume during the respiratory cycle which can produce a paradoxical pulse. *In all such situations, there is a marked inspiratory decrease in LV filling.* Conditions that may be associated with pulsus paradoxus are listed in Table 3-2. When paradox is detected, pericardial tamponade must be considered first and ruled out before other diagnoses are pursued.

Pericardial Tamponade. Pericardial tamponade is the most common cause of pulsus paradoxus. Detection of a paradoxical pulse may be the first or only clue to the underlying condition; recognition may be life-saving. In pericardial tamponade, there is a marked inspiratory rise in intrapericardial pressure which is instantaneously transmitted to the left ventricle, further restricting its filling and producing a fall in inspiratory LV stroke volume. Right ventricular stroke volume increases during inspiration; the additional RV blood reaches the LV during the next expiration and helps elevate systemic pressure. The RV may actually become compressed by the elevated pericardial pressure. Thus, systolic arterial pressure and both RV and LV stroke volume fluctuate markedly during the respiratory cycle in pericardial tamponade.

Pulsus paradoxus is not always found in severe tamponade. It may be absent if there is associated severe aortic regurgitation, which allows the LV

TABLE 3-2 *Causes of Pulsus Paradoxus*

pericardial tamponade
constrictive pericarditis
emphysema
asthma
severe congestive heart failure
marked obesity

to fill with blood during inspiration; this may occur in an aortic dissection resulting in both aortic insufficiency and pericardial tamponade. In circulatory hypovolemia and coexisting pericardial tamponade, a paradoxical pulse may not be present or will be modest. Large, noncompressive pericardial effusions do not usually produce a paradoxical pulse.

Asthma and Emphysema. A paradoxical pulse can be found in patients with severe obstructive lung disease as a result of exaggerated respiratory motion and wide swings in intrathoracic pressure. During forced or labored expiration arterial pressure typically rises; this phenomenon can also be seen in hyperventilating subjects without intrinsic lung disease, as after vigorous exercise. In such situations, pulsus paradoxus is typically modest (less than 15 mmHg) and is unlikely to be palpable.

Marked Obesity. Paradoxical pulse has been reported in very obese patients, probably as a result of the increased work of breathing and large fluctuations in intrathoracic pressure.

Severe Congestive Heart Failure. A paradoxical pulse occasionally can be noted in some patients in heart failure. In these cases, it appears to be related to tension on the heart and pericardium produced by downward movement of the diaphragm during inspiration. The enlarged heart is pulled inferiorly during inspiration, raising intrapericardial pressure and diminishing venous return.

Constrictive Pericarditis. A paradoxical pulse occasionally may be found in patients with pericardial constriction. It is a common misunderstanding that pulsus paradoxus is the rule in this condition. The obliterated pericardial space in constrictive pericarditis actually helps prevent marked respiratory alterations in ventricular filling from occurring.

DETECTION

Palpation Methods. Light pressure with the fingers or thumb should be applied to a peripheral artery (e.g., brachial or radial) with careful attention to alterations of the pulse amplitude as related to the phases of the respiratory cycle.

Sphygmomanometer Method. To measure the actual magnitude of the paradoxical pulse, the blood pressure cuff should be inflated beyond peak systolic arterial pressure and then decompressed slowly and evenly at increments of no more than 4 to 5 mmHg at a time. Note (1) the pressure at which the first Korotkoff sounds appear during expiration, (2) the level when all beats become audible, and (3) the moment when the inspiratory and expiratory Korotkoff sounds become equally loud.

The degree of paradoxus is the difference between *initial* systolic arterial pressure when the first Korotkoff sounds are heard during expiration and

the point at which *all* beats are well heard during *both* phases of respiration. Since a 4 to 6 mmHg fall in systemic arterial pressure is normal during inspiration, in subjects with a definite paradoxical pulse the Korotkoff sounds will be inaudible or barely detectable during inspiration at a level more than 6 to 8 mmHg below peak systolic pressure. *To assess the presence of pulsus paradoxus, it is important to be sure the patient is breathing quietly and not hyperventilating or performing an inadvertent Valsalva maneuver.*

ARTERIAL PULSE IN SPECIFIC CARDIAC DISORDERS

Examination of the arterial pulse can be helpful in establishing a diagnosis and assessing the severity of the underlying condition in a number of cardiovascular disorders.

AORTIC STENOSIS (see also Chapter 13)

In aortic stenosis, obstruction to LV ejection produces characteristic alterations of aortic pressure best detected in the carotid artery but which may also be reflected in the peripheral arterial pulses (Fig. 3-5, Table 13-1).

The carotid pulse in valvular aortic stenosis is slow rising (pulsus parvus) has a delayed peak (pulsus tardus or plateau pulse), is of small volume,

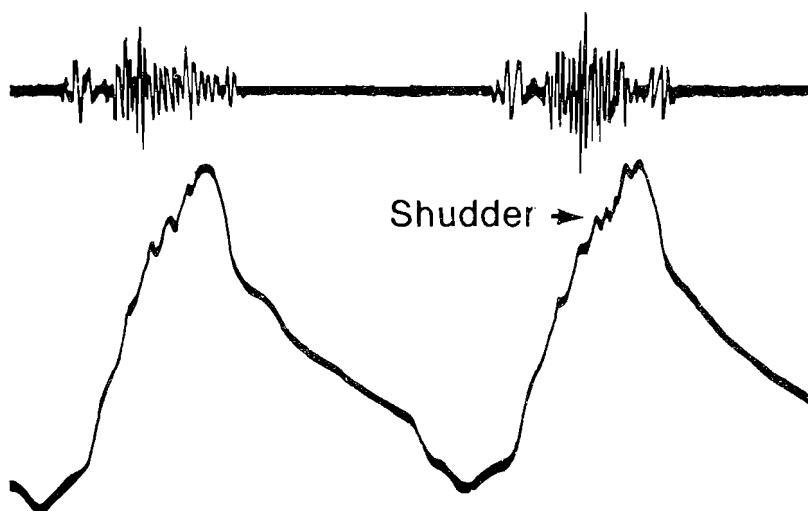


FIG. 3-5. Arterial pulse in aortic stenosis. Note the delayed upstroke and the jagged contour representing a palpable shudder or transmitted thrill. The pulse volume is usually decreased as well (pulsus parvus and tardus). See also Chapter 13. (From Abrams J: Prim Cardiol, 1982.)

exhibits a palpable thrill, and displays a prominent anacrotic notch. The decreased rate of rise is manifested by a "slow" or delayed upstroke. A sustained summit reflects the delayed peak. The small pulse volume is due to hemodynamic obstruction at valve level. A palpable thrill or shudder occurs near or on the arterial pulse upstroke and represents turbulence created by ejection of blood across a narrow and distorted valve orifice.

Anacrotic Pulse. The arterial pulse in aortic stenosis is often called anacrotic. This term means an "upbeating" pulse and refers to the exaggerated anacrotic shoulder or notch found in aortic stenosis. The late portion of the arterial upstroke is coincident with the systolic thrill or shudder (Fig. 3-5). The anacrotic notch or shoulder may be related to a jet effect produced by ejection across the abnormal valve or to decreased velocity of blood flow during early ejection. The anacrotic notch itself usually is not palpable but is easily recorded on carotid pulse tracings.

Pulsus Parvus et Tardus. The typical pulse in aortic stenosis is small in volume and slow-rising (Table 13-1). In general, the more abnormal the pulse contour and volume, the more likely that the aortic valve will be hemodynamically obstructive. *Practical Point: A rapid rate of rise and a normal pulse contour without a sustained peak excludes significant valvular aortic stenosis in many subjects.. Unfortunately, both false positive and false negative diagnoses are common.*

Chapter 13 discusses the problems in assessing the severity of aortic stenosis from the arterial pulse. In general, children and young adults may have hemodynamically significant aortic stenosis with little detectable abnormality of the arterial pulse, probably because of the considerable elasticity of the arterial walls and the relatively high cardiac output in this age group. In the aged, the arterial pulse is often accentuated due to a loss of distensibility of the vascular bed, resulting in a pulse wave with a rapid rate of rise and increased systolic pressure.

The diagnostic qualities of the arterial pulse can be attenuated if there is associated systemic hypertension or aortic regurgitation. In hypertension, the rate of rise and pulse amplitude is increased in late systole. With coexisting aortic regurgitation, the large stroke volume and more rapid ejection rate may "normalize" the typical small and obstructive pulse of aortic stenosis.

The arterial pulse can simulate aortic stenosis whenever the stroke volume is reduced and a prominent systolic murmur is present, as might be found in patients with congestive heart failure, cardiomyopathy, or mitral stenosis. In such conditions, the nonspecific systolic ejection murmur may be entirely functional. In a patient with mild aortic stenosis but significantly depressed LV systolic function, the carotid pulse may suggest more severe valve obstruction than is actually present.

Table 13-2 lists the common clinical situations in which the arterial pulse can result in a false positive or false negative assessment of aortic stenosis.

SUPRAVALVULAR AORTIC STENOSIS

This unusual congenital lesion frequently produces differential streaming of central aortic blood flow; the right carotid pulse is relatively normal, but the left carotid pulse has the characteristic features of aortic valve obstruction. This underscores the importance of routine palpation of *both* carotid pulses.

HYPERTROPHIC CARDIOMYOPATHY

In hypertrophic cardiomyopathy, the early rate of rise of the arterial pulse characteristically is brisk, giving a tapping quality to the pulse. These abnormalities become more pronounced with large left ventricular-aortic gradients. A second systolic peak occasionally may be recorded but less often palpated, producing a bifid or "spike and dome" configuration (Fig. 3-3B). However, many patients with hypertrophic cardiomyopathy have a normal carotid pulse (see also Chapter 14).

COARCTATION OF THE AORTA

In coarctation of the aorta and associated hypertension, the carotid pulses are increased in amplitude but have a normal contour. If there is associated aortic valve obstruction or stenosis due to a congenitally bicuspid aortic valve, the carotid pulse may have a slow upstroke and reduced volume. In all cases of hemodynamically significant coarctation the femoral pulses are small in volume (occasionally impalpable) and markedly delayed with respect to simultaneous palpation of the brachial or radial pulses.

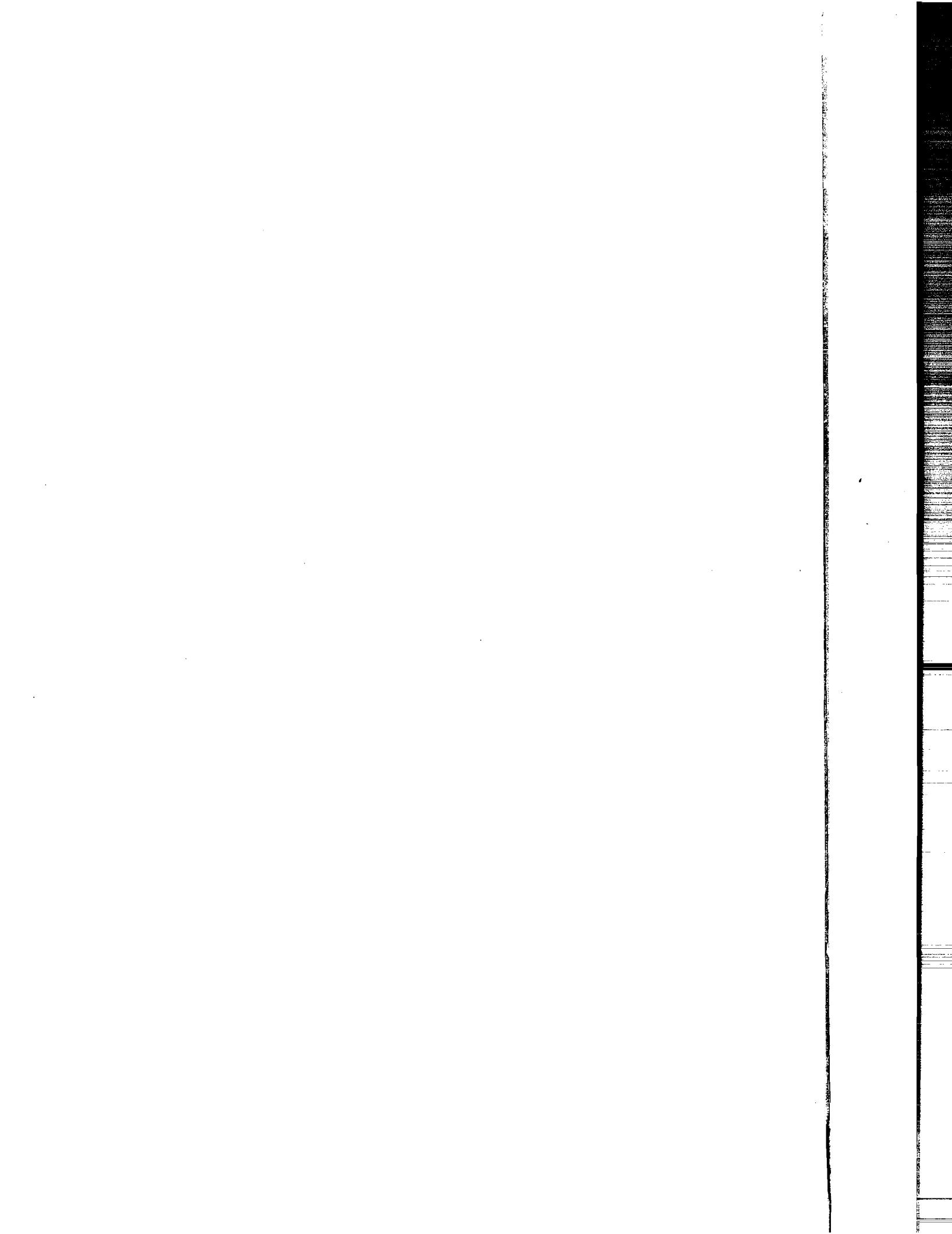
AORTIC REGURGITATION (see also Chapter 15)

In aortic regurgitation the increased stroke volume, rapid rate of ejection, and decreased peripheral resistance result in a high volume, bounding arterial pulse with a characteristic collapsing quality (Fig. 15-1). A systolic thrill may be present over the carotid arteries, even in the absence of aortic stenosis. With major aortic insufficiency, systolic blood pressure is increased, and diastolic blood pressure is abnormally low (Fig. 15-1). The systemic arteries may appear to swell or pulsate.

The alterations in pulse wave in moderate to severe aortic regurgitation are easily detected in all peripheral arteries. Elevation of the arm may accentuate the hyperkinetic, collapsing pulse in the brachial artery. In pure aortic regurgitation or mixed aortic stenosis and regurgitation, a bisferiens pulse is common, suggesting a hemodynamically significant degree of aortic regurgitation (Fig. 3-3A). With the onset of congestive heart failure or de-

creased left ventricular stroke volume, the bisferiens pulse may disappear, and the amplitude and pulse volume will decrease. *Practical Point:* *The typical changes in arterial pulse in severe aortic regurgitation may be minimal or even absent when left ventricular function is significantly depressed.*

Hyperkinetic States. Any condition associated with an increased stroke volume and decreased peripheral resistance may produce abnormalities of the arterial pulse that can simulate aortic regurgitation, such as a large A-V fistula, thyrotoxicosis, patent ductus arteriosus, or severe anemia.



Chapter 4

The Jugular Venous Pulse

Careful observation of the venous pulse is a basic component of the cardiac examination and can lead to valuable information about right-sided cardiac events. Examination of the venous pulse often is viewed with anxiety and dismay by medical students and physicians; the terminology appears arcane, the task obscure. Because graphic records of the jugular venous pulse provide more detailed information than that available on visual inspection, most physical diagnosis texts emphasize detailed analysis of venous pulse wave recordings. This chapter will stress what can be observed at the bedside.

NORMAL PHYSIOLOGY

Although the venous system contains about 70 to 80% of the circulating blood volume, it maintains a very low intravascular pressure (3 to 7 mmHg or 4 to 11 cm of water). Venous compliance therefore is very high; that is, veins are extremely distensible. The jugular venous pulse reflects the relationship between venous tone, the volume of blood in the venous system, and right heart hemodynamics. During diastole, the jugular veins directly reflect right ventricular filling pressure; during systole, they mirror right atrial pressure. Thus, analysis of the jugular venous pulse provides considerable information about right-sided cardiac physiology.

Waveforms. There are two visible peaks or waves and two visible descents or troughs in the normal jugular venous pulse (Fig. 4-1). The A wave is followed by the X descent, and V wave is followed by the Y descent. If the jugular pulse or the right atrial pressure is recorded, the wave analysis is more complicated. A C wave is usually present in these tracings, interrupting the X descent, and in a small number of persons, a late H wave or plateau terminates the Y descent. The physiologic basis of these waves and descents is as follows:

A Wave. This wave directly reflects right atrial (RA) contraction, which results in retrograde blood flow into the superior vena cava and jugular veins during RA systole. The jugular venous A wave follows the P wave of the EKG, precedes the upstroke of the carotid pulse, and is almost synchronous with S1.

X Descent. The early portion of the X descent results from RA relaxation during atrial diastole. The later and dominant portion (X') reflects the

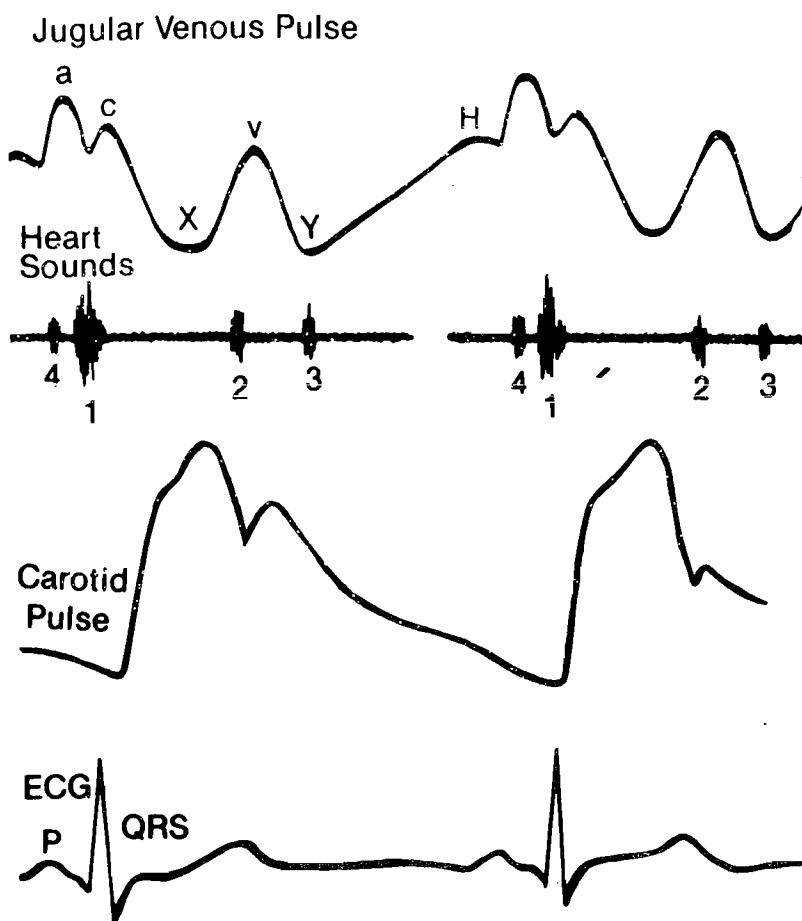


FIG. 4-1. Normal jugular venous pulse. Note the biphasic venous waveform with a large A wave immediately preceding the carotid upstroke and roughly coinciding with S1, and a smaller V wave that peaks almost coincident with S2. The jugular X descent occurs during systole and in some individuals may be quite prominent. The Y descent occurs during early diastole; the nadir of the Y descent times with S3. The C wave and H wave are not visible to the eye but are often recordable in venous pulse tracings. (From Abrams J: Prim Cardiol, 1982.)

fall in right atrial pressure during early right ventricular systole, as the tricuspid valve ring is pulled caudally by the contracting right ventricle ("descent of the base"). The right atrium actually may expand during systole as RV ejection helps to "suck" blood from the great veins into the right atrium.

The X descent is often the most prominent motion of the normal jugular venous pulse. It begins *during systole* and ends just before S2.

C Wave. This positive wave has caused enormous controversy and confusion over the years. Part of the problem relates to the fact that the C wave has two different causes. In the neck veins, the C wave is an artifact caused by transmitted carotid artery pulsations; however, in the right atrium the C wave reflects the upward bulging motion of the closed tricuspid valve during isovolumic systole. *Practical Point:* Because the C wave is not usually

visible as a separate wave form and has no importance in the examination of the jugular venous pulse, it should be disregarded.

V Wave. The V wave is the second major positive wave, begins in late systole, and ends in early diastole. The V wave results from continued venous inflow into the right atrium during ventricular systole while the tricuspid valve is closed. It is roughly synchronous with the carotid pulse and peaks just after S2.

Y Descent. The Y descent is the negative deflection of RA pressure that occurs when the tricuspid valve opens in early diastole. It begins and ends during diastole.

H Wave. The H wave follows the Y descent and results from passive right heart filling during diastole. It is not clearly visible but readily recorded at slow heart rates. Its only importance is that it may make recognition of the A wave very difficult.

Practical Point: In normal persons, the right atrial A wave is larger than the V wave, and the X descent is more prominent than the Y descent. When the neck veins are examined, in most conditions a larger A than V wave will be seen.

Respiratory Influences. Inspiration may result in increased visibility of the venous pulse. During inspiration, the velocity of venous flow and the return to the right heart increases; RA and RV contraction become more vigorous (Starling effect), exaggerating the X and Y descents. Although mean venous pressure falls slightly, the wave forms are accentuated during inspiration. During expiration, the A wave diminishes in size, and the V wave transiently may become the dominant positive deflection. The jugular venous pulse is evaluated by careful visual inspection of the neck veins. The physician must assess the level of the venous column and the timing and amplitude of the actual waveforms. Proper positioning of the patient is mandatory in order to derive information from the venous pulse analysis. Attention should be paid to the inspiratory behavior of the venous activity. On occasion, the use of the hepatojugular test is helpful as well.

THE EXAMINATION

Anatomy. The venous pulse is visible but not palpable. At the junction of the right internal jugular and subclavian veins, there is a slight dilatation known as the jugular bulb; this is found in the so-called internal jugular triangle just above the clavicle in the right supraclavicular fossa between the two heads of the sternocleidomastoid muscle. Usually, the internal jugular vein itself is not visible; indirect venous pulsations of the jugular bulb and internal jugular vein are transmitted to the overlying skin and soft tissues

with a subtle and undulant motion that must be carefully sought. The external jugular veins, coursing vertically over the sternocleidomastoid muscle posterior and lateral to the internal jugular vein, are usually more easily seen, and the discrete venous pulsations typically are more readily identified than in the internal jugular system (Fig. 4-2).

If visible, the internal jugular veins are *preferable* to the external jugular veins for the evaluation of venous pulses for the following reasons: (1) They take a more direct course to the superior vena cava and right atrium, whereas the external jugular veins empty into the innominate veins and follow a more circuitous route. (2) Anatomically, they are closer to the right atrium. (3) There are no valves in the internal jugular veins. Valves at the proximal portion of the external jugular veins may prevent transmission of pulsations from the right atrium; when distended, the external jugular veins should be “milked” by the finger in a caudal and cranial direction in order to determine the direction of venous filling. (4) The external jugular veins may become quite small and the pulsations barely visible in the presence of venoconstriction resulting from increased sympathetic tone (e.g., in congestive heart failure).

The right side of the neck is preferable to the left for examination of the jugular venous pulse. The left external jugular vein may have a higher

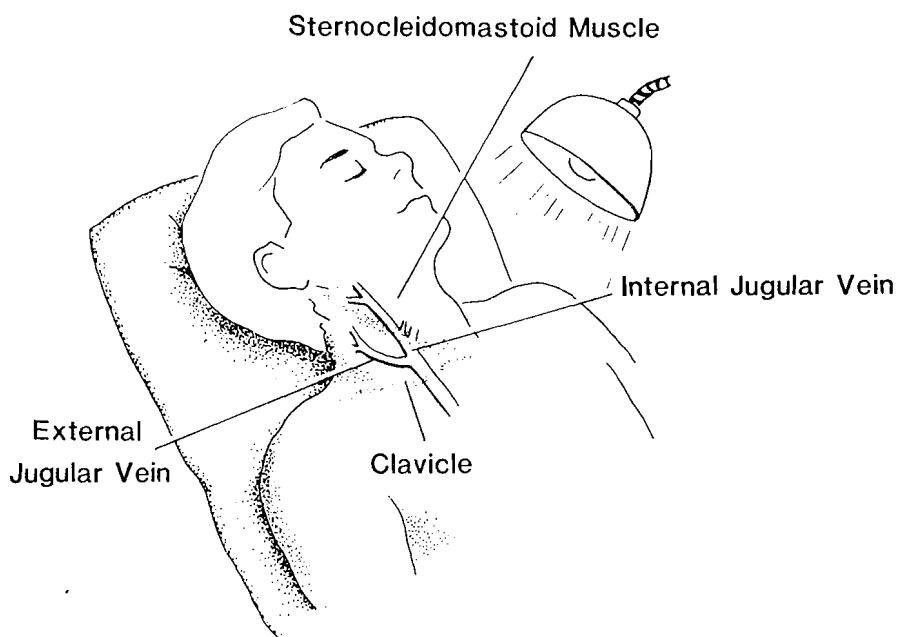


FIG. 4-2. Important landmarks of the venous pulse. The external jugular veins are easily seen lateral to the sternocleidomastoid muscles, extending vertically upward toward the back of the ear. The internal jugular veins are of small amplitude and undulant in nature. They are not usually well seen in individuals with a normal venous pulse but may be prominent when the jugular venous pressure is elevated or when prominent V waves are present. The ideal patient position consists of modest elevation of the thorax and head. It is important for the patient to be relaxed with no tension on the neck muscles. Tangential lighting is helpful to accentuate the jugular veins.

pressure than the right because of compression of the left innominate vein between the aortic arch and the sternum. Nevertheless, if there is any difficulty in seeing a clearly pulsatile jugular vein on the right, *both* sides of the neck should be carefully examined.

Position of Patient (Fig. 4-2). The patient should be reclining and comfortable and should not place excessive tension on the tissues of the neck. Often, it is helpful to elevate the chin and slightly rotate the head to the left, gently stretching the skin of the right lower neck and supraclavicular area. In hospitalized patients, removing the pillow often aids in proper positioning; however, the addition of a small pillow may sometimes be necessary for optimal visualization. Natural light is desirable for inspection of the venous pulsations, although a flashlight or bedside lamp can be used profitably to cast a shadow on the venous pulsations occurring in the lower neck. For this purpose, tangential lighting is best, as it can silhouette the neck veins to great advantage.

Estimating Venous Pressure. Optimal positioning of the neck and thorax is essential for analysis of the venous pressure and waveforms. The examiner first should estimate whether the venous pressure will be normal or elevated and then position the patient accordingly. Is it desirable to use an examining table or bed that can be easily adjusted over a range of 0 to 45 degrees; for evaluation of very high venous pressures, the sitting position (90 degrees) is necessary.

Normal Venous Pressure. In most patients without elevation of the venous pressure the supine or 15 to 30 degree position is best. The patient's head and neck must be positioned so that the venous waveforms are clearly identifiable. During inspiration clear-cut pulsations should be visible in order to ascertain that there is a patent, distortion-free venous column. The height of the venous column at the peak of the A and V waves generally is taken as an indication of the venous pressure, although the actual mean jugular venous pressure will be slightly lower.

The sternal angle (of Louis), found at the junction of the manubrium and sternum at the level of the second rib, is used as the standard reference point for determining venous pressure noninvasively (Fig. 4-3). The right atrium is 5 to 7 cm below this point, and the relative distance of the right atrium to the sternal landmark changes only a small variable degree in the supine, 45-degree, and 90-degree positions. The estimated height of the venous column should be related to the sternal angle, e.g., "1 cm above to . . .," "5 cm above . . ." The normal venous column should be no more than 2 to 3 cm above the sternal angle. When the mean jugular venous pressure is 4 to 5 cm higher than the sternal angle, the venous pressure is abnormally elevated. If the height of the venous column is equal to or only slightly higher than the sternal angle in the supine position, the venous pressure is normal. This will be found in most persons.

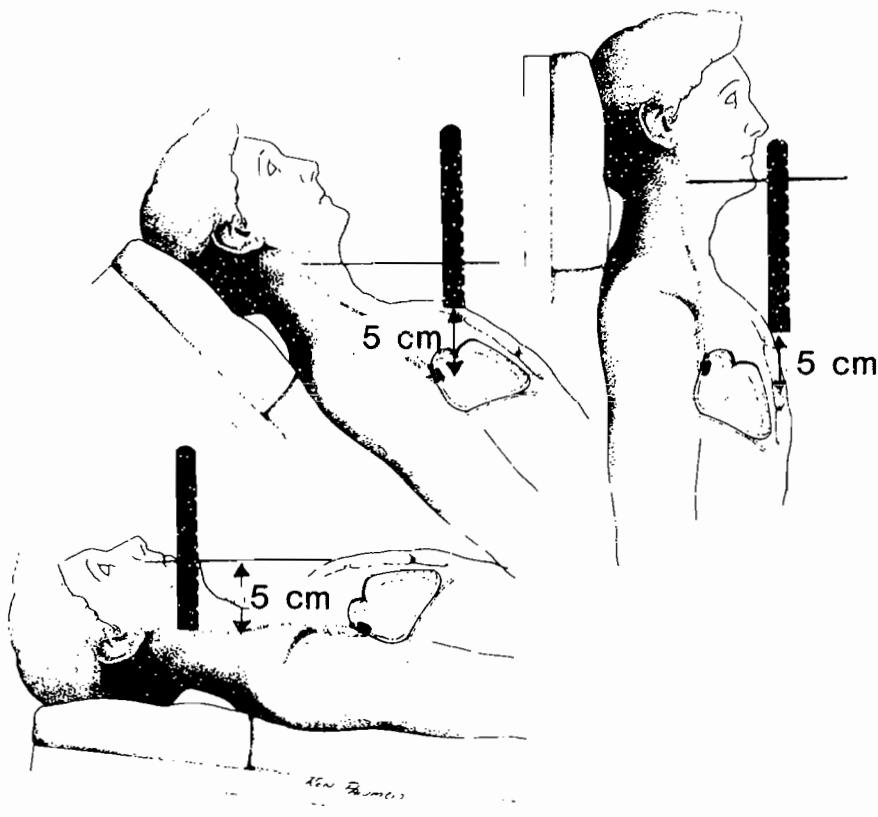


FIG. 4-3. Estimation of mean venous pressure. The right atrium is approximately 5 cm below the sternal angle of Louis with the subject in any body position. Thus with a patient supine or erect, the height of the venous pulsations from the sternal angle can be measured; by adding 5 cm one can estimate the actual venous pressure. The thorax and neck should be positioned until the peak of the venous column is readily identified. In subjects with a normal venous pressure, only the peaks of the A and V waves may be seen when the patient is sitting up at 45 degrees or greater. When the venous pressure is abnormal, the thorax and head must be elevated in order to accurately identify the true peak of the venous column. (From Abrams J: Prim Cardiol, 1982.)

Elevated Venous Pressure: In conditions where the venous pressure is very high, pulsations may not be seen until the patient is sitting up; even then the waves may be invisible. Careful inspection of the upper neck beneath the angle of the jaw is important; the external jugular veins should also be analyzed when increased levels of venous pressure are suggested. In severe tricuspid insufficiency, the ear lobes may move gently laterally with each V wave (Fig. 19-7).

Practical Point: Adequate visualization of the jugular venous pressure is difficult, if not impossible in some persons, particularly obese subjects or patients with short, thick necks. Nevertheless, with care and attention to detail, the venous pulse can be identified in the majority of subjects. Inspiration or modest elevation of the thorax frequently "brings out" the venous waveforms in patients with difficult-to-see jugular veins. If tachycardia is present, it may

be difficult to time the venous pulse properly; the A wave tends to fuse with the V wave and becomes relatively smaller. In such cases, carotid sinus pressure can be used to slow the heart rate. In order to analyze the waveform accurately, it is often better to re-examine the venous pulse at another time when the heart rate is slower.

Timing of Venous Waves. Timing the venous pulse may be assessed by two methods. In the first, the left carotid pulse is carefully palpated simultaneous with visual inspection of the right-sided jugular veins. In the second, the jugular venous pulse is analyzed in concert with auscultation of the heart sounds. Either or both techniques are useful.

Visual Technique. The first method relies on the dominance of the jugular A wave. Because RA contraction precedes LV contraction, the jugular A wave will be seen as a flickering pulsation just *before* the carotid artery pulse is felt. *Practical Point: Identification of the jugular A wave preceding the palpable carotid pulse usually is all that is necessary for accurate identification of the venous pulse waves.* The relative size of the A and V waves and X and Y descents may then be assessed.

Auscultation. If present, the venous A wave is coincident with an S4 and is approximately simultaneous with S1. The X descent occurs during systole and will be seen as a negative transient occurring between S1 and S2 with the trough occurring just before S2. The V wave begins in late systole and peaks just after S2; the Y descent or collapse begins in early diastole after the V wave. Dr. Jules Constant recommends that the examiner use the X and Y descents of the venous pulse rather than the A and V waves for timing and precise analysis of the venous pulse wave forms.

Hepatojugular Reflux (Abdominal Compression Test). This technique employs sustained pressure applied to the right upper quadrant of the abdomen, resulting in a transient elevation of the venous pressure that enables detection of abnormalities in circulatory function. The hepatojugular reflux test is a useful diagnostic maneuver when the jugular venous pulse is borderline-elevated or when latent RV failure or silent tricuspid regurgitation is suspected. Abdominal compression forces venous blood into the thorax. A failing or dilated right ventricle and vasoconstricted venous circulation may not be able to receive the augmented venous return to the right heart without a rise in mean venous pressure. In normal persons, prolonged (60 sec) abdominal pressure will not elevate the venous pressure or will cause only a slight (1 cm) elevation that is not sustained. In congestive heart failure or tricuspid regurgitation, this maneuver will result in an elevation of the venous pressure greater than 1 cm that persists throughout the time the pressure is applied. The hepatojugular reflux test is then said to be positive. In congestive heart failure, vigorous diuresis may normalize the venous pressure; the sustained abdominal compression maneuver still can be useful in demonstrating an abnormal response to increased RV venous inflow.

Technique. In the abdominal compression test, the patient is positioned so that the upper level of the venous column is at mid-neck level. Gentle but firm compression of the right upper quadrant with an open-fingered hand is applied for 30 to 60 seconds. Since it is critical that the patient remain comfortable and that he not perform an inadvertent Valsalva maneuver, the patient should be instructed to continue breathing with an open mouth. If there is significant tenderness or discomfort over the liver, the pressure can be applied elsewhere in the abdomen with similar results.

A positive test indicates incipient or actual RV failure. In isolated LV failure, the response will be normal. Patients with hypervolemia or fluid overload will have a positive test. Patients with chronic obstructive lung disease may have a false-positive test as a result of altered intrathoracic pressure relationships associated with the increased work of breathing. In conditions with increased generalized sympathetic tone, systemic venoconstriction (decreased distensibility of the venous bed) may result in a positive response.

Cervical Venous Hum. Although not specifically related to the jugular venous pulse analysis, the cervical venous hum deserves mention as an auscultatory phenomenon of the venous system. It is useful to auscultate the right supraclavicular area and base of the neck for a venous hum in children and young adults where it is a common finding. A venous hum can be detected in adults with high output states (Table 4-1). Careful examination of the neck with or without rotating the patient's head leftward will detect a surprising frequency of venous hums. The hum is a continuous murmur, loudest in diastole, produced by turbulence in the neck veins when the head is turned 30 to 60 degrees to the left. Very loud hums can be heard without the head being turned. Some patients can hear their own hum.

The venous hum apparently results from mild compression of the internal jugular vein by the transverse process of the atlas in subjects with a vigorous cardiac output. Typically heard only in the sitting position, it disappears when the subject is supine. The Valsalva maneuver or gentle compression of the neck veins with the thumb or fingers will attenuate or obliterate the venous hum.

The venous hum is a normal phenomenon of no intrinsic importance unless mistaken for a bruit or significant murmur. *Practical Point: The venous*

TABLE 4-1 *Hyperkinetic or High Output States Resulting in Augmented Venous Flow or Pressure*

Pregnancy
Thyrotoxicosis
Anemia
Cirrhosis
A-V fistula
Postexertion
Anxiety

hum may be quite loud and can be confused with murmurs of patent ductus arteriosus, A-V fistula, or aortic regurgitation, or it can mimic carotid arterial bruits. Cervical hums have recently been documented as common occurrences in patients undergoing hemodialysis. It is significant because its presence could lead to the potential misdiagnosis of a serious cardiovascular problem. Radiation into the upper thorax can simulate aortic regurgitation.

Venous Heart Sounds. Right heart gallops, S3 and S4, and murmurs occasionally may be audible over the internal jugular veins in patients with right ventricular hypertension and/or failure. In severe tricuspid insufficiency, the systolic murmur or systolic clicks occasionally can be heard in the neck veins.

Differentiation of Jugular and Carotid Pulses. Careful examination of the neck pulses, in most instances, should prevent confusing venous and carotid artery activity. In severe tricuspid regurgitation, however, venous pulsations may be palpable and visible from the foot of the bed and be confused with the arterial pulsations of severe aortic regurgitation. Table 4-2 lists the features that allow proper identification of the venous and arterial pulses in the neck.

ABNORMAL VARIANTS OF THE VENOUS PULSE

MEAN PRESSURE

Increased Mean Venous Pressure. Right ventricular failure is the most common cause of increased jugular venous pressure. Other important cardiovascular conditions associated with elevated mean venous pressure include constrictive pericarditis, pericardial tamponade, and tricuspid regurgitation.

TABLE 4-2 *Differentiation of Jugular Venous Pulsations from the Carotid Arterial Pulse*

	Internal Jugular Vein	Carotid Artery
Location	Low in neck; lateral	Deep in neck; medial
Contour	Double peaked	Single peaked
Character	Undulant, not palpable	Forceful, brisk, easily felt
Inspiration	A and V waves often more visible although mean pressure decreases	No change
Upright position	Decrease in mean pressure	No change
Compressibility	Readily obliterated by gentle pressure 3-4 cm above clavicle	Cannot compress easily
Abdominal compression	May see transient increase in pressure	No effect

due to RV hypertension. The venous pressure occasionally can also be elevated in persons with a normal cardiovascular system, typically in those with a high output or hyperkinetic state (Table 4-1). Fluid overload, obesity, or increased abdominal pressure from any cause can result in high venous pressure; and asthma, respiratory distress, and emphysema may produce distended neck veins as a result of expiratory increases in intrathoracic pressure. Obstruction of the superior vena cava obviously will elevate the jugular pressure. In this condition, however, no venous pulsations or waveforms are seen, and the hepatojugular test will be negative; frequently, venous collaterals will be visible in the upper chest.

RV Infarction. Recent recognition of the syndrome of right ventricular infarction has resulted in another indication for careful jugular venous pulse analysis. Right ventricular involvement is seen in conjunction with acute inferior or diaphragmatic myocardial infarction and may manifest in a wide variety of presentations from overt shock to an isolated elevation of the venous pulse. The extension of myocardial necrosis into the adjacent right ventricle and inferior wall of the left ventricle may "stiffen" the right ventricle and frequently causes an elevation of RV filling pressure. Thus, the mean jugular venous pressure will be elevated, often with an accentuated A wave even in the absence of an elevation of LV filling pressure. On occasion, the venous pulse in RV infarction can mimic that of constrictive pericarditis, with prominent X and Y descents producing two deep troughs in the venous pulse. Sustained abdominal compression (hepatojugular reflex test) will cause an elevation in the venous pulse even when the mean right atrial pressure is normal or borderline increased.

If the tricuspid valve has been damaged by the RV infarct, tricuspid regurgitation may produce large V waves in the venous pulse.

Decreased Mean Venous Pressure. Very low venous pressure may be a clue to the presence of hypovolemia or dehydration. When there is hypotension or overt shock, a flat or low-normal jugular venous pressure may be an invaluable clue to the underlying pathophysiology.

Increased A Wave Amplitude (Fig. 4-4A). Whenever the force of right atrial contraction is augmented, the A wave will increase and may become quite prominent. When it is very large, it is known as a "giant A wave." The most common cause of a large A wave is decreased RV compliance, usually associated with an increase in RV end-diastolic pressure. The altered compliance is seen in RV hypertrophy from any cause such as severe pulmonary hypertension, pulmonic stenosis, and pulmonary vascular disease.

In certain conditions resulting in marked left ventricular hypertrophy such as hypertrophic cardiomyopathy and even valvular aortic stenosis, the jugular A wave may be accentuated, presumably a result of the massively hypertrophied interventricular septum altering the pressure-volume characteristics of the right ventricle (Bernheim effect). Tricuspid stenosis, although

rare, is a classic cause of a large A wave and should be considered whenever unusually prominent A waves are seen in a patient being evaluated for rheumatic mitral valve disease.

Decreased A Wave Amplitude. In some unexpected situations, the A wave may be normal or small. In tetralogy of Fallot with systemic right ventricular pressure, the A wave is normal because the large ventricular septal defect "decompresses" the RV. Right atrial injury occurring from surgical cannulation during open-heart surgery can cause a reduction in A wave amplitude postoperatively.

X DESCENT

The systolic collapse of the jugular venous pulse becomes deeper whenever there is vigorous right ventricular contraction, as occurs in cardiac tamponade or RV overload states. The X descent may also be quite prominent in atrial septal defects. In mild tricuspid regurgitation, the X descent is attenuated and disappears with major degrees of regurgitation. In atrial fibrillation, the X trough is usually preserved, although it is often reduced in size.

V WAVE

The classic cause of an enlarged V wave is tricuspid regurgitation (Chapter 19), where the V becomes the dominant venous pulsation (Figs. 4-4B, 19-3). In mild tricuspid regurgitation the venous contour may be normal at rest; abdominal compression, exaggerated inspiration, or mild exercise will often bring out a diagnostic V wave. The V wave in tricuspid regurgitation (sometimes referred to as the C-V wave or S wave) begins earlier in systole than the usual V wave and, when prominent, can simulate carotid artery pulsations. They may even be palpable. Giant V waves frequently visibly distort the earlobes (Fig. 19-7), and the large undulant venous pulsations in the neck can often be seen from the foot of the bed.

The V wave may also increase in prominence in the setting of congestive heart failure without tricuspid regurgitation. In subjects with atrial fibrillation, the V wave can be quite large without tricuspid regurgitation being present. In atrial septal defect, the V wave is generally larger than normal, and the A and V waves are typically of equal amplitude (see Figure 20-2).

Y DESCENT

The Y descent or trough is exaggerated when the venous pressure is elevated from any cause. This prominent diastolic collapse is known as Friedreich's sign and is usually accompanied by a diastolic filling sound (S3 or

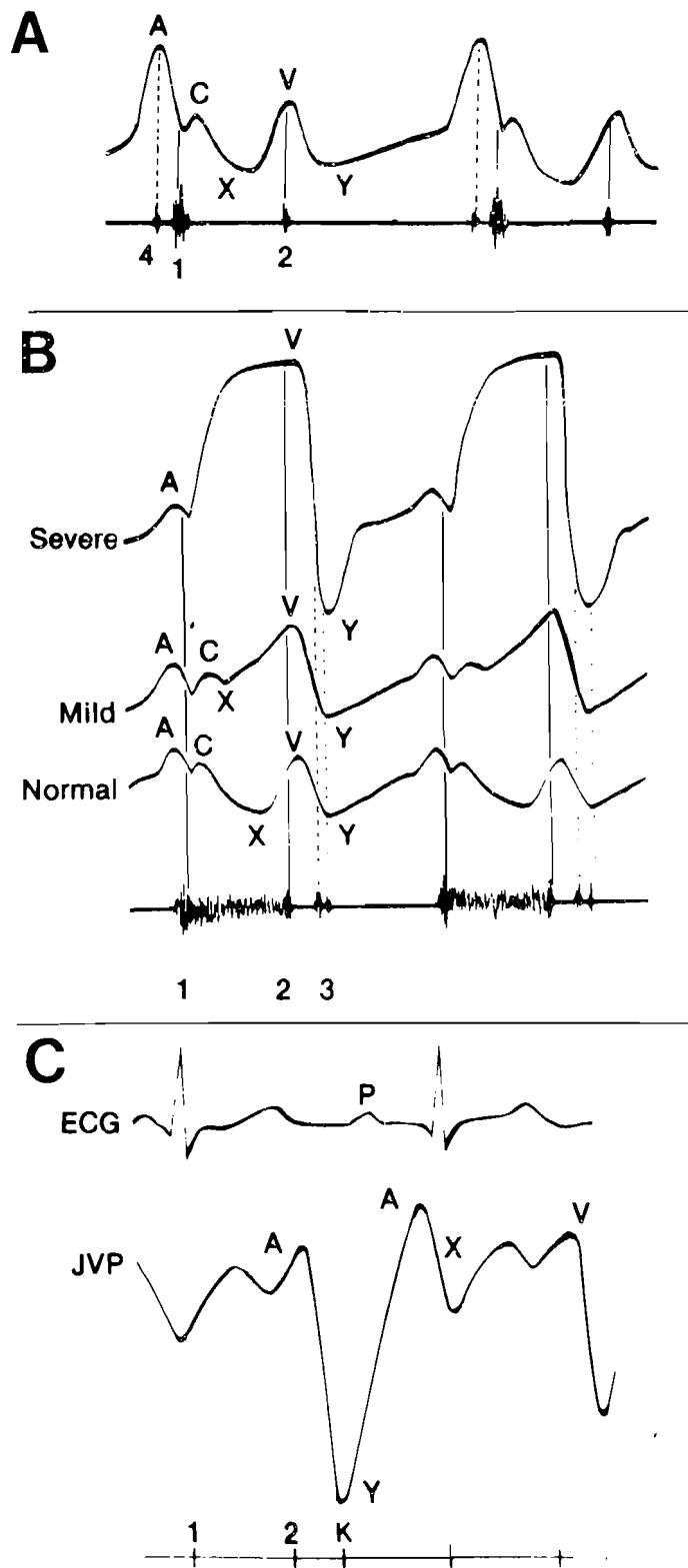


FIG. 4-4. Common abnormalities of the venous pulse. A. Large A waves associated with elevated right ventricular end diastolic pressure or decreased right ventricular compliance. Increased A wave size or giant A waves are seen when there is severe right ventricular hypertrophy, usually associated with right ventricular systolic hypertension. A right ventricular S4 is often present

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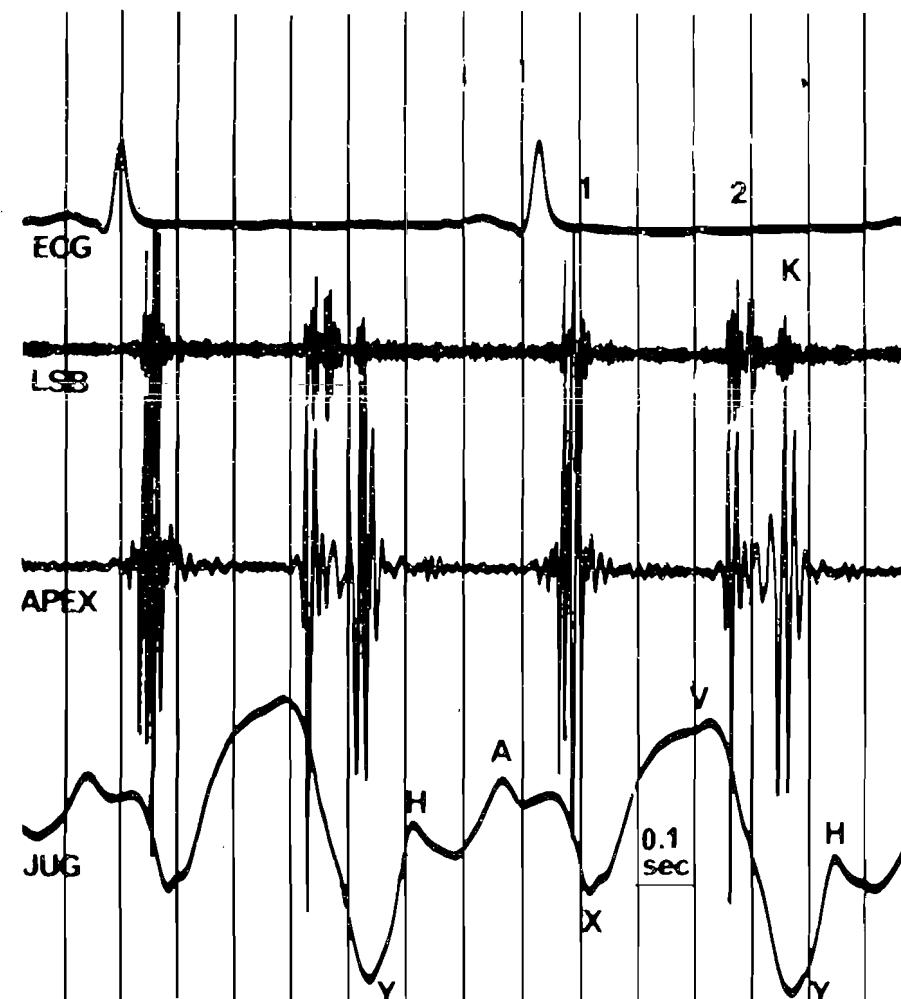


FIG. 4-5. Phonocardiogram and jugular venous pulse tracing from a patient with constrictive pericarditis. Note the prominent V wave and the steep Y descent which times with the loud pericardial knock (K). (From Tavel ME: Phonocardiography: clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 26:145, 1983.)

in such cases. B. Augmented V wave in tricuspid regurgitation. As reflux across the tricuspid valve increases in severity, the systolic V wave becomes higher as well as broader. The X descent disappears and the Y descent is progressively accentuated with increasing severity of tricuspid regurgitation. With severe tricuspid regurgitation the systolic wave may be so dominant as to mimic the carotid arterial pulsations; the entire lower neck will swell with each right ventricular systole (see also Chapter 19). C. Constrictive pericarditis. In this unusual cardiac condition, right ventricular diastolic pressure is markedly elevated. This elevation results in a prominent Y descent following tricuspid valve opening. The abrupt rise in venous pressure during right ventricular filling is due to the noncompliant right ventricular chamber encased in an unyielding pericardial shell. The venous pulse contour in constrictive pericarditis often takes on a "M" or "W" configuration. A pericardial knock (K), a high frequency early diastolic filling sound, is typically present. (From Abrams J: *Prim Cardiol*, 1982.)

pericardial knock) (Figs. 4-4C, 4-5). The Y descent may be diminutive in pericardial tamponade.

Inspiratory Increase in Venous Pressure (Kussmaul's Sign). This unusual phenomenon is seen in conditions where the right ventricle has such decreased compliance and/or capacity that the augmented venous return occurring during inspiration cannot be adequately "handled" by the right ventricle, and the venous pressure "backs up." Typically, it is seen in constrictive pericarditis (present in approximately 40% of cases) and severe congestive heart failure. It is distinctly uncommon in pericardial tamponade.

ARRHYTHMIAS

In atrial fibrillation, the jugular venous pulse may simulate tricuspid insufficiency, as the V wave is often prominent. The X descent persists but can be quite small; if the venous pressure is high, preservation of the X descent may be the best way to exclude associated tricuspid regurgitation, which characteristically obliterates the X collapse. Certain arrhythmias may result in intermittent giant A waves called "cannon waves" that occur when the right atrium contracts against a closed tricuspid valve. With ventricular, atrial, and/or A-V junctional extrasystoles, intermittent cannon A waves may be seen when the premature beat occurs early such as during the preceding QT segment. Complete A-V block and ventricular tachycardia with independent atrial activity result in periodic cannon waves and A waves of varying amplitude. These findings may be of major diagnostic value.

In general, however, inspection of the venous pulse (or arterial pulse) is not a reliable way to diagnose rhythm disturbances, and the electrocardiogram should always be used. Nevertheless, the experienced clinician may be rewarded at the bedside by careful observation of giant A waves or rapid atrial activity that may represent a supraventricular tachycardia or ventricular tachycardia with retrograde atrial capture.

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Chapter 5

The Precordial Impulse

Precordial palpation is that component of the cardiac physical examination that allows the physician to detect cardiac activity on the chest wall. In the normal individual cardiac motion is represented by the *apex beat* or *apex impulse*, which is produced by systolic contraction of the left ventricular free wall and septum. Right ventricular activity is usually not palpable. When cardiac hypertrophy or dilatation is present, abnormal systolic and diastolic events emanating from the left or right ventricle may be detected on palpation and, on unusual occasions, left and right atrial impulses may be felt.

For over 3500 years physicians have been aware that cardiac activity can be detected with the examining hand. Eminent scientists such as William Harvey, Rene Laennec, and Sir James MacKenzie have made important and surprisingly accurate observations about palpable cardiac movements. Modern medicine has added the scope of precision to this field, particularly with respect to the effects of specific chamber hypertrophy and dilatation on the characteristics of the cardiac impulse. Quantitative apex cardiography and, more recently, echocardiography have been useful techniques in elucidating the anatomic and pathophysiologic features of the abnormal precordial impulse.

Experienced clinicians can derive a great deal of information about intracardiac size and function from a careful analysis of precordial motion. The first clue to ventricular enlargement may be obtained, even before the electrocardiogram or chest roentgenogram shows diagnostic changes.

This chapter outlines a practical approach to precordial palpation. The focus is on *what can be felt* by the examiner. Careful application of these principles will enhance the utilization of this important physical diagnostic technique.

PHYSIOLOGY

NORMAL PRECORDIAL ACTIVITY

The palpable apical impulse in a normal subject is produced by an anterior movement of the left ventricle (LV) during early systole. As isovolumic, intraventricular pressure rises, the LV rotates in a counterclockwise direction on its long axis as the cardiac apex lifts and makes contact with the left anterior chest wall. Following aortic valve opening, the LV chamber moves

away from the chest wall after the first half of ejection, and the ventricle continues to decrease in size until systole is completed. Thus, the impulse felt or recorded on the precordium is comprised of an early outward thrust, followed by retraction during the last part of systole (Fig. 5-1). Normal palpable cardiac activity occurs only during the first half of systole.

Peak outward motion of the apex impulse occurs coincident with or just after aortic valve opening and the beginning of ejection. The impulse is sustained for a brief period (up to 0.08 seconds) and then the outward movement ceases as the LV apex moves inward (Figs. 5-1, 5-2). This negative retraction wave is due partly to recoil of the heart as it rotates clockwise on

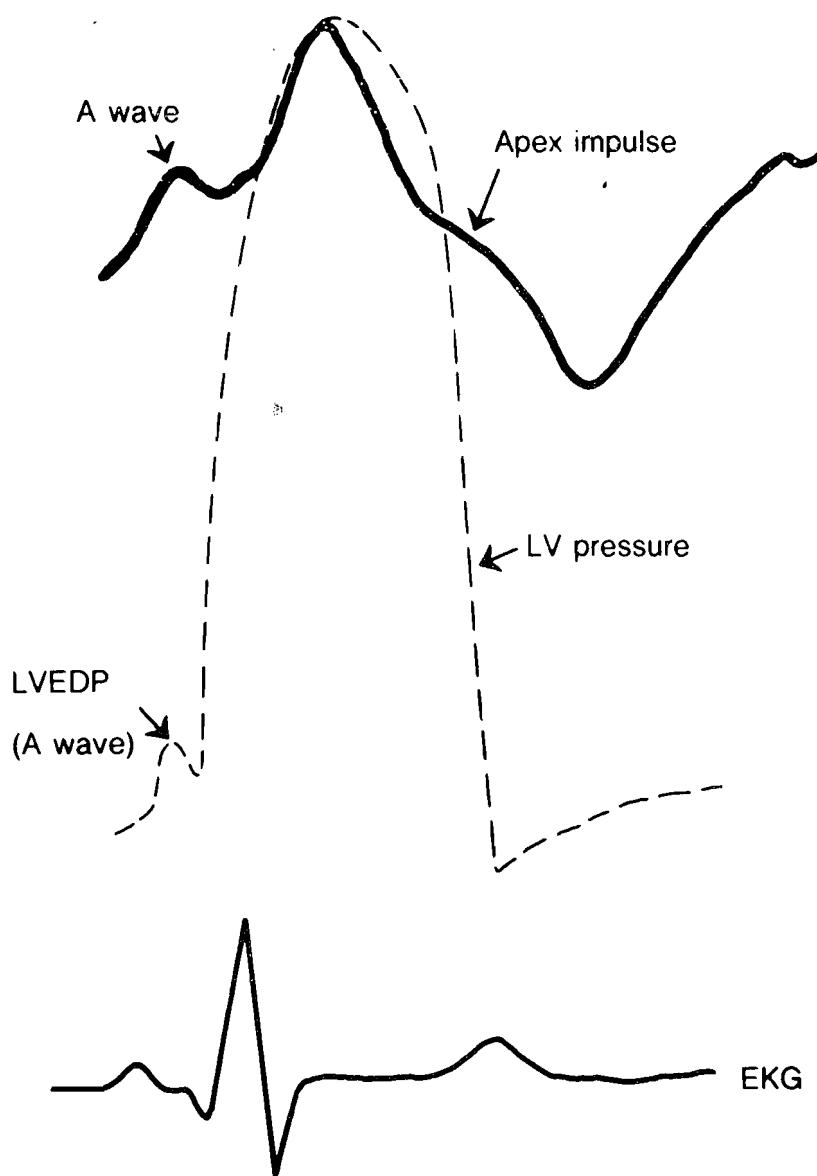


FIG. 5-1. Relationships of the normal apex impulse to left ventricular pressure. LVEDP = left ventricular end-diastolic pressure. (From Abrams J: Precordial motion. In Signs and Symptoms in Cardiology. Edited by LD Horwitz and BM Groves. Philadelphia, JB Lippincott Co, 1985.)

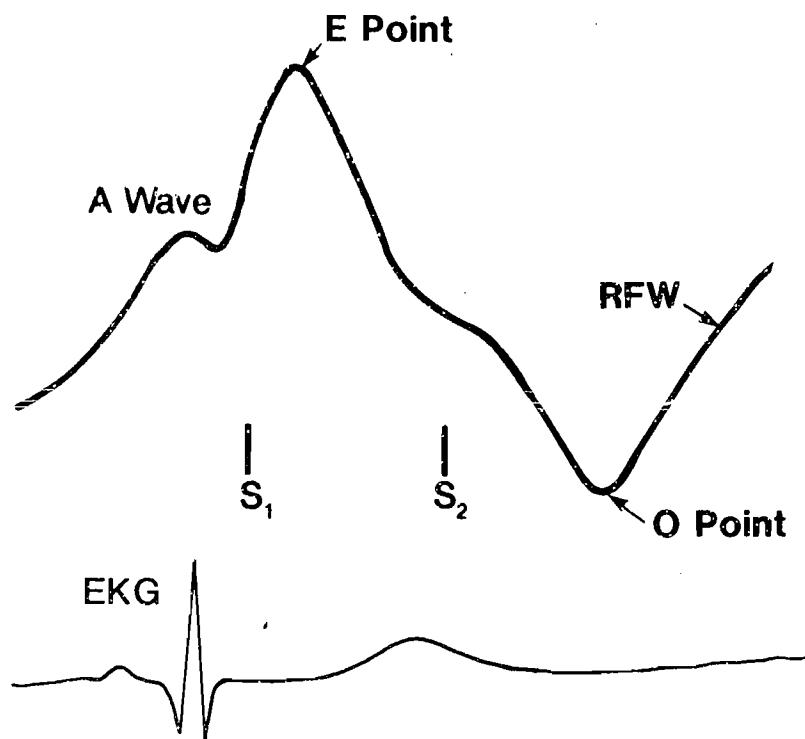


FIG. 5-2. Normal precordial impulse. Note that the outward motion occurs entirely within the first half of systole. The left ventricle retracts from the chest wall as it becomes smaller during late systole. The palpable apex impulse is brief. Normally, the A wave and diastolic events are not palpable. RFW = rapid filling wave.

its long axis during late systole. The retraction wave can be seen but is not felt. Diastolic events, such as those produced by rapid left ventricular filling (S3) or left atrial contraction (S4), are normally not palpable. The characteristics of the normal precordial impulse are discussed on page 69 and are listed in Table 5-1.

Angiographic studies suggest that the intraventricular septum (comprised mostly of left ventricular muscle) and the anteroseptal aspect of the LV make contact with the inner thoracic cage to form the palpable apex impulse. The "true" anatomic LV apex actually is slightly lower and more lateral than the palpable apical impulse.

TABLE 5-1 *The Normal Supine Apical Impulse*

-
- A gentle, nonsustained tap
 - Early systolic anterior motion that ends before the last third of systole
 - Located within 10 cm of the midsternal line in the 4th or 5th left intercostal space
 - A palpable area less than 2 to 2.5 cm² and detectable in only one intercostal space
 - Right ventricular motion normally not palpable
 - Diastolic events normally not palpable
 - May be completely absent in older persons
-

(From Abrams J: *In Signs and Symptoms in Cardiology*. Edited by LD Horwitz, and BM Groves. Philadelphia: JB Lippincott Co., 1985 with permission.)

Right Ventricular Activity. Although the right ventricle (RV), located just beneath the sternum and left 3rd to 5th ribs, is closer to the chest wall than the left ventricle, RV activity is not normally felt. This is probably because the RV is thin-walled (2 to 3 mm wall thickness), low pressure (20 to 25 mmHg peak systolic pressure), and it pulls away from the anterior chest as the heart rotates in a counterclockwise fashion in early systole. In normal children and young adults or thin subjects who have a narrow A-P thoracic diameter, gentle right ventricular activity occasionally may be felt. Careful subxyphoid palpation during held-inspiration in thin normal subjects may also occasionally detect RV motion.

Palpable Diastolic Events. The low pressure and volume transients of mid (S3) and late (S4) diastole are normally undetectable by precordial palpation. With alterations in ventricular diastolic volume, pressure, or compliance, these events (palpable S3 and S4) can be transmitted to the chest wall by the left or right ventricle and may be felt by the examining fingers.

ABNORMALITIES OF PRECORDIAL MOTION

When there are abnormalities in ventricular size, shape, or function, precordial activity is more easily detected (Tables 5-2, 5-3). The left and right ventricular precordial impulse may be altered in *location, amplitude, size, and contour*. Almost all diagnostic alterations in precordial motion occur in systole. Table 5-3 summarizes the most common abnormalities of the apical impulse.

Systolic Events

Left Ventricle. Characteristic changes may occur in the apical impulse with LV enlargement depending on whether there is dominant hypertrophy or dilatation. Such abnormalities often provide the clinician with the first clue of organic heart disease. Left ventricular wall motion abnormalities, focal or generalized, are often manifest by an abnormal apex motion pattern.

TABLE 5-2 *Causes of Palpable Precordial Abnormalities*

-
- LV hypertrophy and/or dilatation
 - LV wall motion abnormalities (fixed or transient)
 - Increased force of left atrial contraction (palpable S4)
 - Accentuated diastolic rapid filling (palpable S3)
 - Anterior thrust of the heart from severe mitral regurgitation
 - RV hypertrophy and/or dilatation
 - Loud murmurs (thrills)
 - Loud heart sounds (normal and abnormal)
 - Dilated or hyperkinetic pulmonary artery
 - Dilated aorta
-

TABLE 5-3 *Major Types of Precordial Impulses*

	Hyperkinetic	Sustained	Late Systolic
LEFT VENTRICULAR IMPULSE (apex beat)	Hyperkinetic circulatory states Thin chest wall Pectus excavatum Volume overload: aortic regurgitation mitral regurgitation ventricular septal defect	Pressure overload: hypertension aortic stenosis Chronic or severe volume overload states LV dilatation, especially with decreased ejection fraction LV dysfunction LV aneurysm	LV dyssnergy Hypertrophic cardiomyopathy Mitral valve prolapse (rare)
RIGHT VENTRICULAR IMPULSE (parasternal area)	Hyperkinetic circulatory state in young subjects Volume overload: atrial septal defect tricuspid regurgitation	Pressure overload: pulmonary stenosis pulmonary hypertension cor pulmonale mitral stenosis pulmonary emboli Cardiomyopathy	Severe mitral regurgitation

From Abrams J: Precordial motion in health and disease. Mod Con Cardiovasc Dis 49:55–60, 1980.

Volume Overload. The classic response to significant volume overload conditions is an *increase in the amplitude* (hyperkinetic impulse) of the LV impulse without a change in contour: early systolic outward and late systolic inward motion are preserved (Fig. 5-2B). However, early or mild degrees of mitral or aortic regurgitation, classic overload states, may not alter the amplitude or contour of the LV impulse. In severe volume overload states, particularly with depression of LV contractility and a decreased ejection fraction, the LV impulse may become *prolonged* or *sustained* into the second half of systole (Fig. 5-3C). This response may be due both to a longer duration of left ventricular ejection and to a more globular chamber configuration. With cardiac dilatation, the normal elliptical LV shape becomes more spherical, and the resulting apical impulse becomes sustained. Left ventricular dilatation with a major increase in LV end diastolic volume also results in *leftward and downward displacement* of the apex impulse, as well as an *increase in the size* of the actual contact area of the apex beat.

Pressure Overload. The initial response of the LV to increased outflow resistance (aortic stenosis, hypertension) is concentric hypertrophy without an increase in cavity size. Systolic function is well maintained. With time the LV impulse becomes *prolonged* in duration, reflecting an increased left ventricular ejection time (Fig. 5-3C). This produces a sustained left ventricular heave or thrust. The force of contraction is increased, but there is relatively little chamber dilatation. Thus, the apex impulse is not usually displaced but has an *increased force*. With long-standing disease and depression of cardiac function, the LV chamber dilates, and the impulse moves more laterally on the chest wall. The apex beat in pressure loaded hearts is not usually felt over as large an area of the chest wall as that in chronic severe volume overload states.

Abnormalities of Left Ventricular Systolic Function. When LV contractile performance is deranged, the apical impulse can be altered in several ways: (1) the normal brief outward contour may become sustained into the last half of systole (Fig. 5-3C). This may result from ischemic dysfunction due either to fibrosis or to depressed myocardial contractile performance or from LV dilatation and a globally depressed ejection fraction; (2) a mid or late systolic impulse (bulge) may be present, reflecting a disordered contraction pattern (coronary heart disease or cardiomyopathy) (Fig. 22-1); (3) an ectopic precordial impulse may appear, located at a site away from the normal apical impulse, usually superior and medial to it. This is most likely to occur with an LV aneurysm, but may also be seen with anterior wall dyskinesis in the absence of an aneurysm; (4) inferolateral displacement and enlargement of the area of the apex beat will be present whenever the LV chamber is substantially dilated, particularly if there is a depressed ejection fraction.

Hyperdynamic States. Anxiety, tachycardia, exertion, or catechol excess from any cause may increase cardiac contractility and systolic blood pressure,

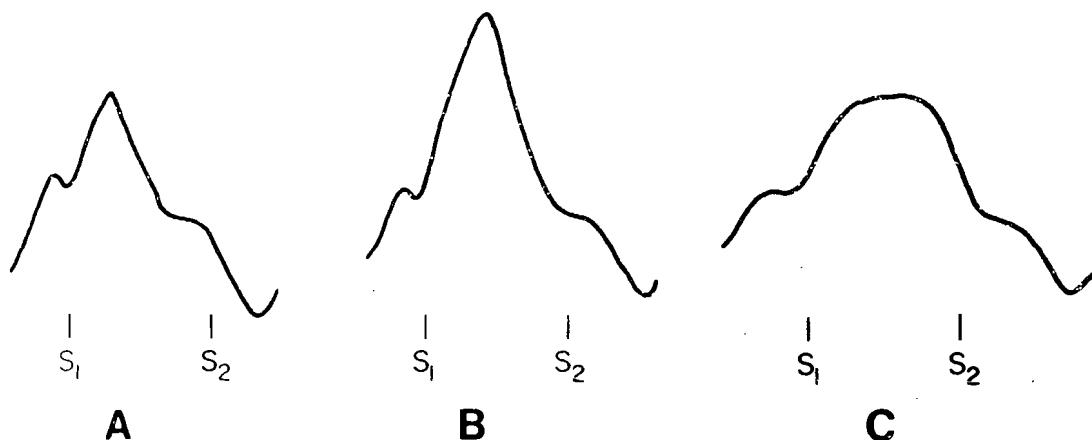


FIG. 5-3. Major types of left ventricular precordial motion. A. Normal. B. Hyperdynamic. C. Sustained. With a patient in the supine position, the presence of sustained left ventricular activity detectable in the latter half of systole is distinctly abnormal. Some experts believe that palpation of a sustained impulse when patients are in the left lateral decubitus position may have less specificity for underlying left ventricular enlargement. (From Abrams J: Precordial palpation. In Signs and Symptoms of Cardiology. Edited by LD Horwitz and BM Groves. Philadelphia, JB Lippincott Co, 1985.)

resulting in an increase in the force and amplitude of the apex impulse (Fig. 5-3B). In such situations, the location and contour of the precordial impulse is unchanged; the apex impulse increases in forcefulness but is not sustained (hyperkinetic).

Right Ventricle. The right ventricle (RV) responds similarly to the LV with respect to volume and pressure overload states. As the RV is not usually involved in ischemic dysfunction, there are fewer clinical variants of the abnormal right ventricular impulse.

Volume Overload. The volume loaded RV (atrial septal defect, tricuspid regurgitation without RV dysfunction) produces a hyperdynamic, high amplitude impulse that retains the pattern of mid-late systolic retraction (Fig. 20-3A). Thus, the motion is a brief anterior thrust. With a very large RV end-diastolic volume or depressed RV contractile function, this anterior parasternal motion may become sustained.

Pressure Overload. Pulmonary hypertension from any cause results in sustained systolic RV activity manifest as a palpable and/or visible anterior motion in the lower left sternal area (Figs. 17-4C, 20-3B). The RV inflow area is located at the left 4th to 5th intercostal space adjacent to the sternum, and the infundibular area is located more superiorly at the 3rd to 4th left intercostal space.

Mitral Regurgitation. In severe mitral regurgitation, there may be an "apparent" RV impulse that reflects systolic expansion of an enlarged left atrium with subsequent anterior displacement of the RV (Fig. 17-4B). Such a precordial movement is only found with a dilated left atrium and large degree of mitral reflux (Fig. 17-5). The palpable late systolic parasternal

impulse is out of synchrony with the earlier LV apex impulse and can be identified as a separate movement with careful examination technique (see below).

Diastolic Events

Left Ventricle. Left ventricular diastolic filling is comprised of two phases: initial rapid filling following mitral valve opening and late diastolic augmentation resulting from left atrial contraction (Fig. 7-1). In the normal heart, left ventricular pressure throughout diastole is low; these filling events (and their resultant heart sounds) are neither palpable nor audible.

When ischemia, fibrosis, hypertrophy, or dilatation are present, LV compliance, diastolic volume, and filling pressure are altered so that the two peaks of blood flow from the left atrium to the LV may result in large pressure transients and increased distending forces. In these situations, the rapid filling phase and left atrial systole may result in audible and palpable events (S3 and S4).

Palpable S3. An abnormal increase in transmitral flow, a large LV end-diastolic volume, and significant depression of LV function are all associated with the presence of an audible S3 which may also be palpable (Fig. 7-3). It is not clear why an S3 may be detectable by palpation in some individuals and not in others, nor is it known if there are any particular prognostic or physiologic implications associated with the presence of a palpable S3. It is likely that S3 audibility and palpability are closely related to factors associated with the "coupling" of the heart to the chest wall, i.e., the way in which the left ventricle makes contact with the inner left thoracic cavity.

A palpable S3 is found in patients with a major elevation in LV filling pressure and LV end-diastolic volume. In such individuals, early and mid LV diastolic pressure is usually elevated; left atrial contraction may not result in additional pressure augmentation at end-diastole. Typically, these hearts have a decreased ejection fraction. Patients with aortic valve disease, hypertensive heart disease, and coronary heart disease may develop a loud or palpable S3 when LV systolic function deteriorates, with or without overt congestive heart failure. Patients with congestive cardiomyopathy typically have a palpable LV filling sound. The pericardial knock of constrictive pericarditis is an early, exaggerated filling event, comparable to a loud S3, and is usually palpable (Fig. 5-4).

In subjects with an increased volume and rate of blood flow crossing the mitral valve, an S3 may be audible and palpable *in the presence of good left ventricular function*. Classically, severe mitral regurgitation is the cause of such filling events; a voluminous amount of blood returns to the LV from the left atrium during the rapid filling phase (Fig. 5-5). Left ventricular end-diastolic volume is usually elevated in such cases, but systolic function and ejection fraction are well preserved.

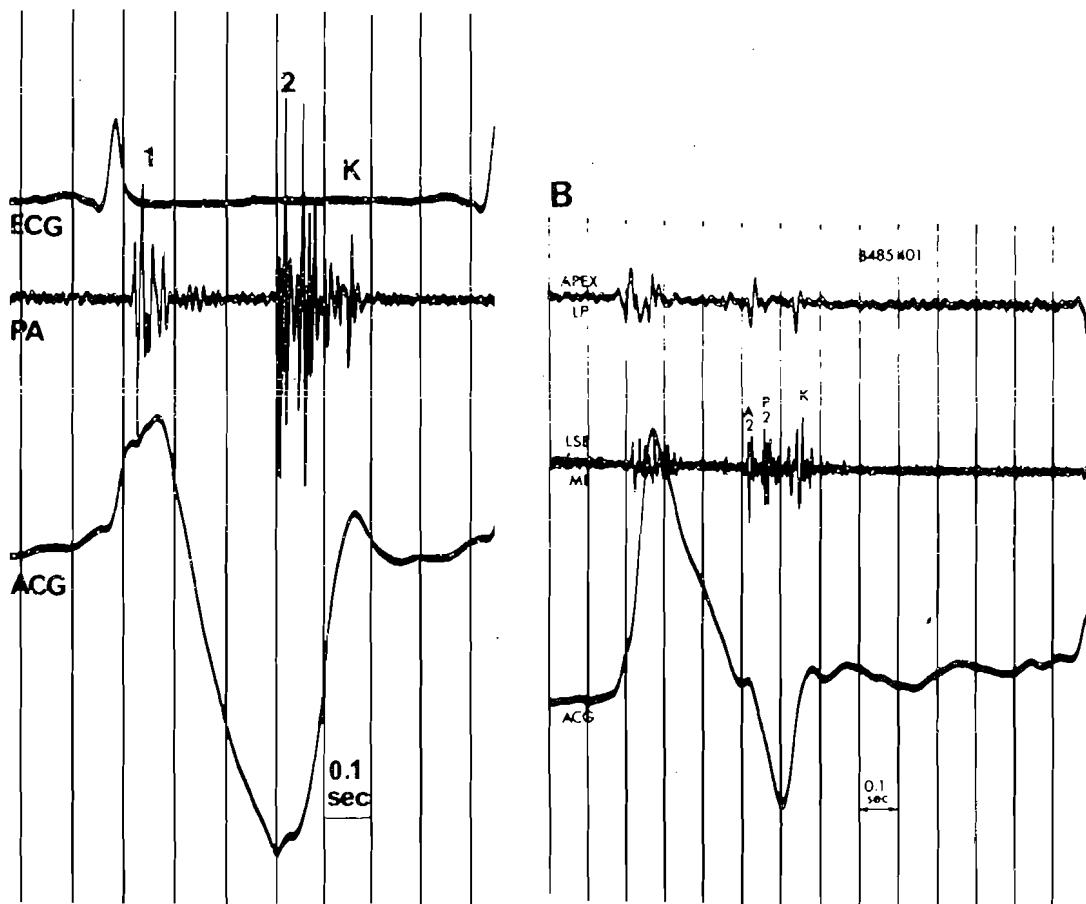


FIG. 5-4. Two examples of the precordial impulse in constrictive pericarditis. Note the prominent systolic retraction wave and early diastolic outward deflection, which are often palpable. This coincides with the pericardial knock (K), occurring at the peak of right ventricular filling (From Tavel ME: Phonocardiography: clinical use with and without combined echocardiography. Prog Cardiovasc Dis 26:145, 1983.)

Palpable S4 (Fig. 7-6). A palpable S4 is related to decreased LV compliance, usually a result of hypertrophy without dilatation or ischemia with increased diastolic stiffness. A palpable S4 always is associated with an elevated LV end-diastolic pressure, which may be strikingly high, although early mid-diastolic pressure is often normal. Thus, a palpable S4 is commonly found in aortic valve disease, hypertrophic cardiomyopathy, hypertensive heart disease, coronary artery disease, and occasional congestive cardiomyopathy. The implications of a palpable S4 are quite different from those of an S3. In the former case, a reduction in ventricular compliance is the key abnormality; LV systolic function is usually well preserved, and the ejection fraction is normal. An S3, however, when not associated with significant mitral regurgitation, is associated with high LV filling pressures, a dilated ventricle, and decreased systolic function.

Right Ventricle. The pathophysiology of a palpable S3 and S4 generated in the right ventricle is similar to that in the left heart. A decrease in RV compliance (e.g., RV hypertrophy secondary to pulmonary hypertension) may

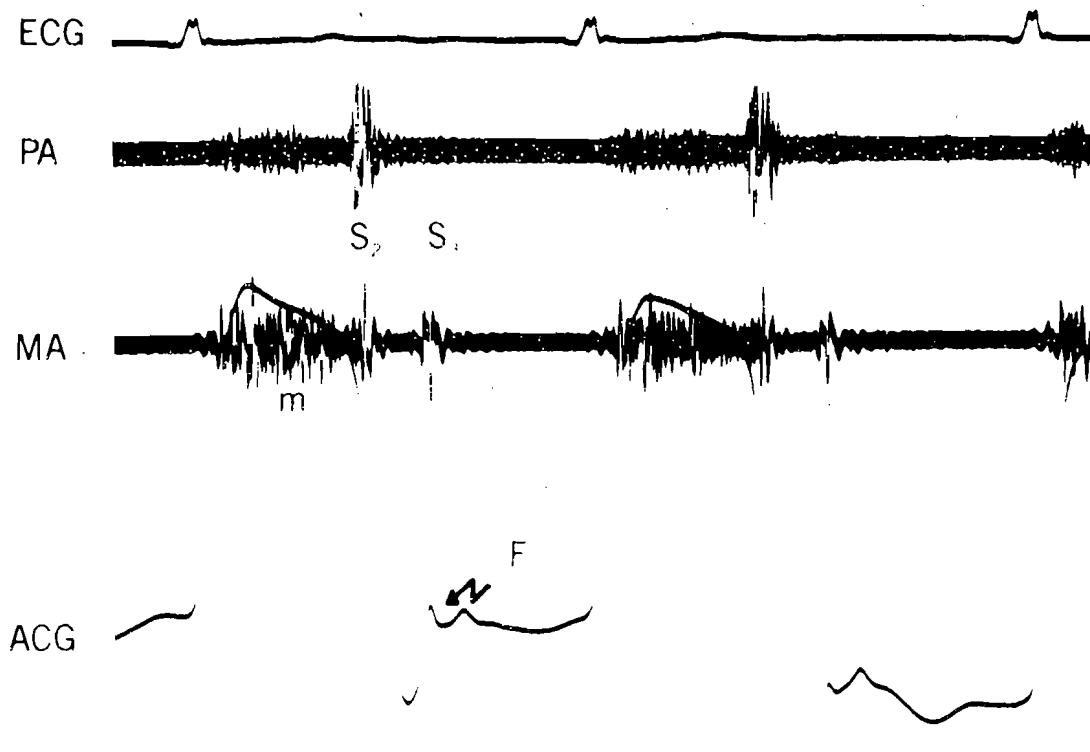


FIG. 5-5. Palpable and audible third heart sound in mitral regurgitation. A loud S3 may be transmitted as a palpable diastolic event and indicates moderate to severe mitral regurgitation. The sound is simultaneous with the peak of the rapid filling phase of the recorded apexcardiogram (F wave). Note the holosystolic murmur (m) at the apex. (From Stefadorous MA and Little RC: The cause and clinical significance of diastolic heart sounds. Arch Intern Med 140: 537, 1980.)

result in an RV S4. Increased blood flow crossing the tricuspid valve may produce a prominent palpable S3, such as may be associated with tricuspid regurgitation or a large left-to-right shunt (e.g., atrial septal defect). In severe RV dysfunction, an S3 may be heard as well as felt.

Classically, right ventricular events such as an S3 or S4 are characterized by inspiratory augmentation and expiratory attenuation or disappearance.

Other Causes of Palpable Cardiovascular Activity (Table 5-2)

In addition to the tactile precordial phenomena related to LV and RV systolic and diastolic events, palpable precordial activity occasionally is detectable as a result of vascular pulsations or ectopic left ventricular wall motion abnormalities.

Aorta. A palpable aortic impulse almost is never a normal finding. When the ascending aorta or aortic arch is enlarged or dilated, palpable systolic pulsations may be detected. Thus, with aortic aneurysms, diffuse aortic dilatation, or aortic dissection a systolic impulse may be felt in the right first or second intercostal space, the right or left sternoclavicular junc-

tion, or in the suprasternal areas. A tracheal tug may be present with a large ascending aortic aneurysm. Dilatation and tortuosity of the brachiocephalic vessels may also cause prominent vascular pulsations at the suprasternal notch or above the clavicles. A "normal" right-sided aorta occasionally can be palpated in the second right interspace in association with tetralogy of Fallot.

Pulmonary Artery. Dilatation or enlargement of the pulmonary artery usually is associated with pulmonary hypertension. This may result from hyperkinetic (high flow) states or from increased pulmonary vascular resistance. Idiopathic dilatation of the pulmonary artery results in an enlarged proximal pulmonary artery segment. All these conditions may produce a palpable systolic impulse in the second-third left interspace just to the left of the sternal edge. This is easily detectable in thin subjects.

Left Ventricular Ectopic Impulse. Focal wall motion abnormalities or an overt left ventricular aneurysm may produce systolic impulses that are detectable at sites away from the LV apex beat, usually medial and superior to the apical area. With anteroseptal scarring or dyskinesis the ectopic impulse may be found at the lower parasternal edge and thus may simulate right ventricular activity.

Palpable Heart Sound and Murmurs. Loud sound transients, such as an increased S1, A2, or P2, are often palpable and are known as precordial shocks. The opening snap and loud S1 of mitral stenosis are commonly palpable, and on occasion an alert physician can diagnose this valve lesion by palpation prior to using the stethoscope. A palpable S2 at the second left interspace suggests systemic or pulmonary hypertension. Ejection clicks may be easily felt; the aortic ejection sound is detected at the left ventricular apex, and a pulmonic ejection sound is palpable at the upper left sternal border.

Thrills. Any loud murmur may be transmitted to the chest wall and produce a vibratory sensation detectable by the examining hand. These palpable murmurs or thrills correlate with a murmur intensity of Grade IV/VI or greater. Such murmurs may be felt at the apex (mitral regurgitation, obstructive hypertrophic cardiomyopathy), lower left sternal border (ventricular septal defect), or cardiac base (pulmonic stenosis, aortic stenosis). In thin subjects the likelihood of a thrill is greater than in muscular or fleshy patients. A diastolic thrill occasionally may be felt at the apex in the lateral decubitus position in mitral stenosis or at the lower left sternal border in acute aortic regurgitation secondary to a perforated or ruptured aortic cusp.

CLINICAL PRESENTATION

EXAMINATION OF THE PRECORDIUM

For optimal precordial examination, it is important for the examiner and subject to be relaxed and to have maximum exposure of the patient's chest. The room should be comfortably warm. Clothing and undergarments must

be removed to allow unobstructed visualization and palpation of the chest; for example, the examiner must be prepared to count intercostal spaces and measure the distance of the precordial impulse from the sternum or axillary line. To obtain unobstructed contact between the precordial impulse and the examining hand in women with large breasts, gentle upward retraction of the left breast with the left hand is often necessary.

The subject should be lying comfortably in the supine position or with the thorax elevated no more than 30 degrees (Fig. 5-6). Perloff suggests that precordial examination in the 30-degree position is preferable and may bring out abnormalities more often than when the patient is supine. *Practical Point:* *Patients with suspect or definite cardiovascular disease should routinely be examined in the left lateral decubitus position; the subject should be instructed to turn on his or her left side at a 45- to 60-degree angle with the examining table and elevate the left arm over the head so that the physician may have unobstructed access to the left precordium (Fig. 5-7).*

Inspection. Careful visual observation of the chest is useful in the precordial examination; this may be more helpful after preliminary palpation has identified the site of the apex beat or other impulses, if present. Retraction movements may be more obvious to the eye than outward motion and can be quite prominent with severe degrees of left and right ventricular enlargement. Tangential lighting with the examining lamp or a pen light may accentuate visible movements on the chest wall.

In normal persons there may be a slight retraction of the thorax medial to the apex impulse. As the LV thrust becomes more vigorous (hyperdynamic states, left ventricular enlargement), this retraction becomes accentuated and assumes a rocking character. An extreme example of this phenomenon occurs



FIG. 5-6. Palpation of the apex, supine position. (From Abrams J: Precordial palpation. In Signs and Symptoms of Cardiology. Edited by LD Horwitz and BM Groves. Philadelphia, JB Lippincott Co, 1985.)



FIG. 5-7. Palpation of the apex impulse, left lateral decubitus position. This maneuver should be used in any patient with suspected left ventricular disease. Patient should be turned 45 to 60 degrees onto the left side with the left arm extended above the head. (From Abrams J: Precordial palpation. In Signs and Symptoms of Cardiology. Edited by LD Horwitz and BM Groves. Philadelphia, JB Lippincott Co, 1985.)

in constrictive pericarditis when a deep systolic retraction wave occurs over the entire left precordium and is followed by diastolic expansion (Fig. 5-4).

Pulmonary artery lifts commonly are visible as may be an accentuated rapid ventricular filling wave (S3), which is often more easily seen than felt.

Palpation. The examiner should be standing comfortably at the patient's right side. Both the palm of the hand and the ventral surface of the proximal metacarpals and fingers should be used for palpation (Fig. 5-8). One must learn which aspect of the hand is best for optimal tactile perception. It is advisable to use the pads of the fingers for precise localization and assessment of left and right ventricular activity; the palm and proximal metacarpals usually are best utilized for *initial* localization of palpable cardiac motion as well as the detection of precordial thrills.

Varying pressure with the hand should be applied once a precordial impulse is identified. High frequency sounds, such as an increased S1, opening snap, or transmitted thrill, are best detected with firm application of the hand to the chest. However, the subtle low frequency motion of a palpable S3 or S4 or double systolic apical impulse will be felt only with light pressure of the fingers and may be totally obscured if this examination is not performed correctly. If there is much precordial musculature or adipose tissue, it is often necessary to press quite firmly.

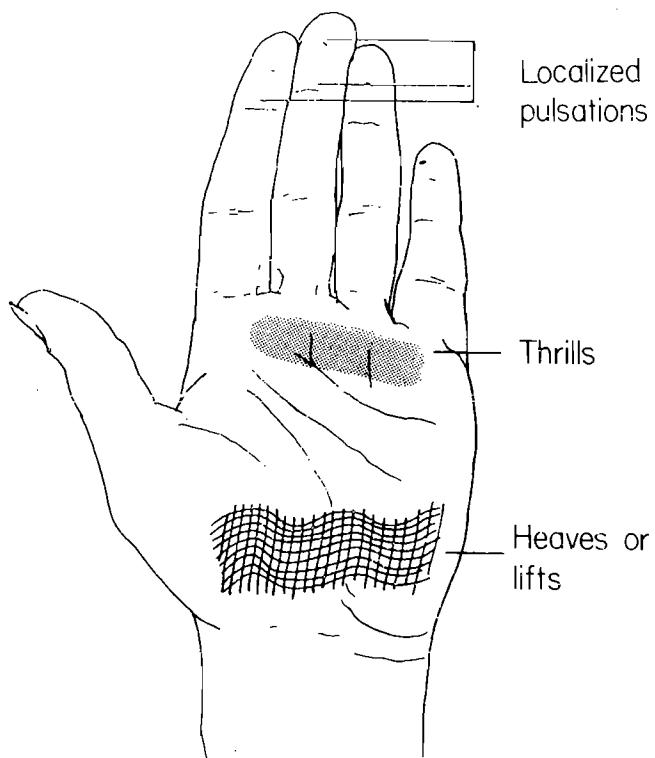


FIG. 5-8. Optimal areas on the examiner's hand for detecting precordial events. (From Constant J: Bedside Cardiology, 2nd ed. Boston, Little, Brown and Co, 1976.)

Timing of precordial events is best carried out using simultaneous palpation of the carotid arterial pulse with the left hand (Fig. 1-4). Some find that concomitant auscultation of S1 and S2 is useful for timing purposes. Actual observation of the stethoscope head positioned directly on the apex impulse can help identify systole; S1 and early ejection occur synchronously with anterior motion of the stethoscope head.

Right Ventricle. It is desirable to use held end expiration for the right ventricular examination. Firm pressure using the palm or heel of the hand with the wrist cocked upwards is advisable (Fig. 5-9). The lower sternum and adjacent 3rd through 5th ribs and left interspaces should be examined in this manner. Movement of the examining hand and fingers should be carefully observed, as the typical low amplitude RV activity is often better seen than felt.

Some experts also suggest exploring the subxiphoid or epigastric region with the extended fingers oriented superiorly; the patient should be instructed to hold his breath in end inspiration as careful palpation for the descending right ventricle is performed (Fig. 5-10). This technique is particularly useful in patients with an increased A-P diameter, COPD, obesity, or muscular chest when right ventricular enlargement is suspected but a parasternal impulse cannot be felt. The hypertrophied right ventricle may produce a caudad impulse that can be felt by extending several fingers or thumb beneath the costal margin toward the heart in the subxiphoid region. Sometimes epigastric palpation will also detect right ventricular activity. The examiner should be

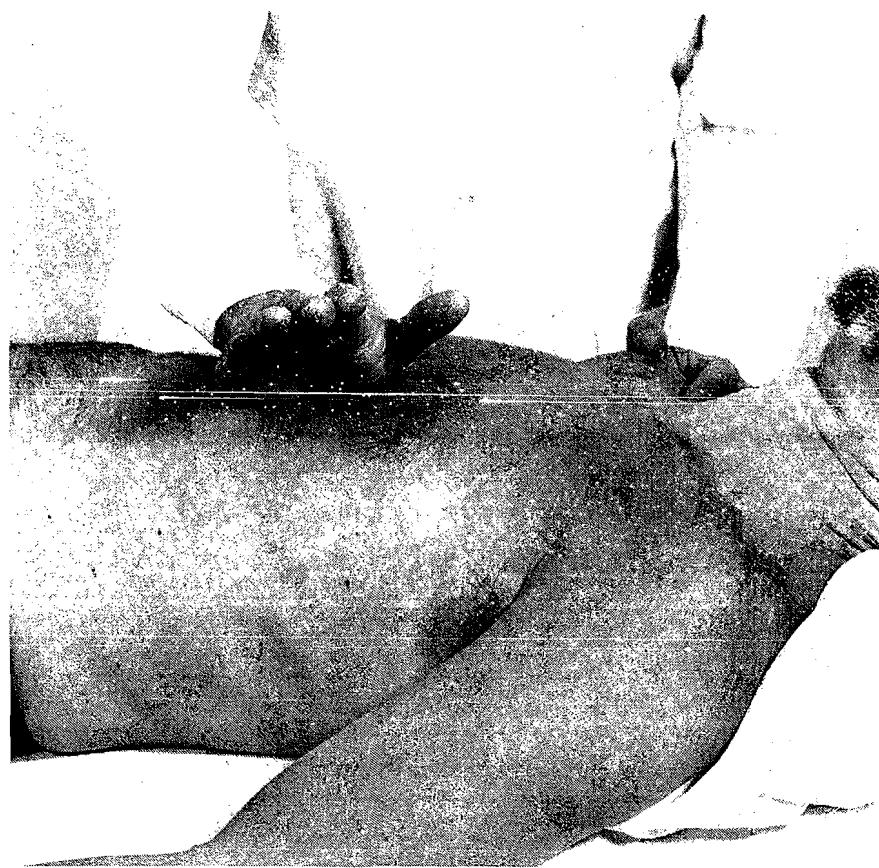


FIG. 5-9. Precordial palpation for detection of parasternal or right ventricular activity. Use firm downward pressure with the heel of the hand while the patient's breath is held in end expiration.

careful to differentiate the downward-directed motion of the enlarged right ventricle from the anterior pulsations of the abdominal aorta, which can often be felt in the epigastrium.

Characteristics of the Normal Apex Impulse (Table 5-1). In normal subjects, the apex impulse in the supine position or at 30-degree elevation produces a gentle outward motion that is usually felt in only one interspace (Fig. 5-2). This anterior movement is brief and nonsustained, pulling away from the examining fingers by midsystole. It occupies a maximal area of 2 to 2½ cm (no larger than a quarter) and is found in the 4th or 5th left interspace at or inside the midclavicular line. It is usually found within 7 to 8 cm from the left sternal edge and should not be located more than 10 cm to the left of the midsternal line. In tall, thin persons, the apex beat can be distal (6th interspace) and more medial than usual; when there is intrathoracic disease or a short stocky body habitus, the apex beat may be displaced leftward. There may be respiratory alteration in the amplitude of the apical beat; pay attention to end expiration if the impulse is hard to locate, although the peak amplitude may also occur during early inspiration.

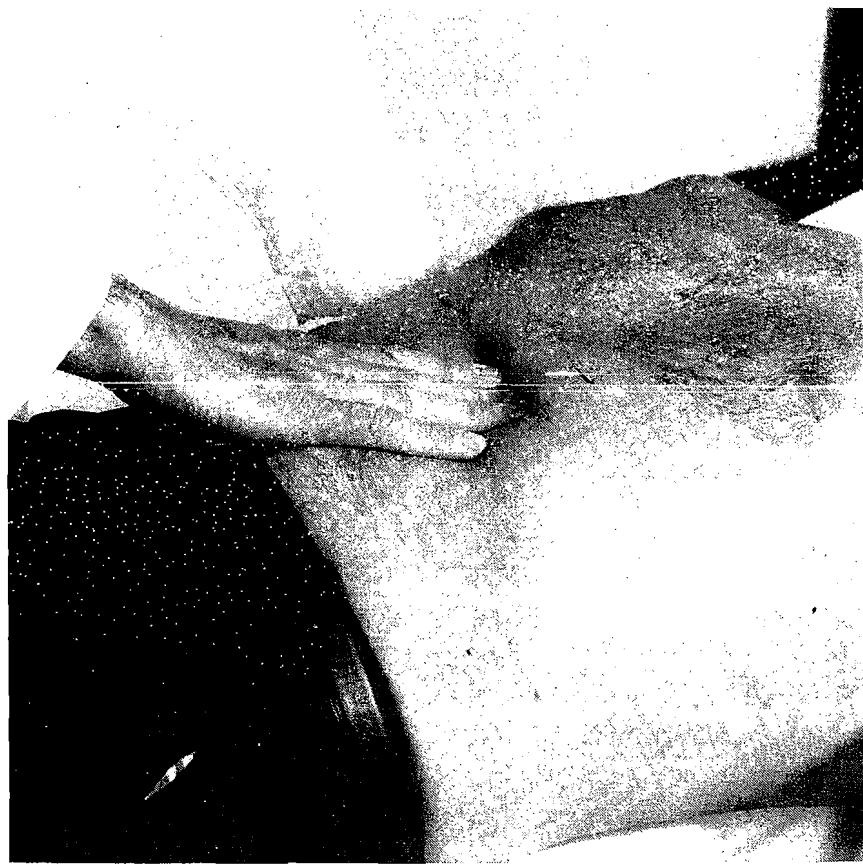


FIG. 5-10. Subxiphoid palpation. This technique is used to detect right ventricular activity in individuals with COPD, a large chest, or obesity. The patient should hold his breath in end expiration. Do not confuse anteriorly directed aortic pulsations for inferiorly directed cardiac motion. (From Abrams J: Precordial palpation. In *Signs and Symptoms in Cardiology*. Edited by LD Horwitz and BM Groves. Philadelphia, JB Lippincott Co, 1985.)

In the left lateral decubitus position the point of chest wall contact of the apex beat is usually slightly more lateral and inferior than in the supine position. There is considerable disagreement as to whether the contour of the apex impulse becomes altered when a subject assumes the left lateral position. A recent study suggested that an apical impulse of 3 cm in area or greater in the left lateral position is specific for left ventricular enlargement. In the supine position the palpable apex impulse should be no larger than a nickle (2 to 3 cm), and should be felt in only one intercostal space.

Right Ventricular Activity. In the normal subject, parasternal activity is usually not detectable except in young or thin individuals. In such cases, a gentle shock or tap at the lower left sternal border may be felt. Forceful, sustained, or high amplitude parasternal motion is always an abnormal finding (Figs. 17-4, 20-3). Occasionally a pulmonary artery impulse in the 2nd to 3rd left interspace adjacent to the sternum may be detected.

Point of Maximum Impulse (PMI). Many textbooks and articles in the literature use the term "PMI" to denote the apex beat. Although not ideal,

this expression is utilized so commonly that it has become acceptable. Nevertheless, it should be recognized that the terms "PMI" and "apical impulse" may not necessarily be synonymous. In some patients, the most prominent or "maximal" impulse may actually reflect ectopic LV motion, right ventricular activity, or a vascular impulse, all of which are located at sites distant from the true LV apex impulse.

Other Palpable Events. In a subject with suspected cardiac disease, the examiner should explore the entire precordium with firm pressure of the hand and proximal fingers, analyzing the aortic, pulmonic, lower sternal, and apical regions. In such a fashion, the unexpected vascular impulse, such as a dilated or aneurysmal ascending aorta or pulmonary artery, may be detected. Particular attention should be given to the upper sternal area, manubrium, and adjacent 1st and 2nd interspaces below the medial aspect of the clavicles. Aortic pulsations and systolic thrills are often found here.

If a patient has coronary artery disease, particularly with a previous myocardial infarction, careful examination for an ectopic impulse should be carried out. This typically occurs medial and superior to the apex impulse. The use of the entire palm and proximal metacarpals will help detect the diffuse lift of a very large left or right ventricle; on occasion, the entire anterior precordium will move in systole. This technique is also suited for the detection of thrills and palpable heart sounds.

Percussion. Under ordinary circumstances, percussion is not a useful and necessary procedure. When a PMI cannot be identified in the supine position, this technique may help establish the presence or absence of cardiomegaly and will identify the approximate left border of the heart. In patients with a large pericardial effusion, the apex beat may actually be detected inside or medial to the lateral border of the percussible cardiac silhouette.

VARIATIONS OF THE APEX BEAT

Absent Apical Impulse. It is not commonly realized that many older subjects (over age 50) do not have palpable cardiac activity when examined in the supine position. This may be due to an age-related increase in the A-P thoracic diameter, an increase of muscle or fat on the chest wall, or a physiologic decrease in the force of LV contraction with age. Whenever an apical impulse cannot be felt in the supine position, the left heart border should be percussed, and the patient should be carefully examined in the left lateral decubitus position (Fig. 5-7). The latter is a much more valuable maneuver than percussion. In most adults, LV activity can be detected when the subject is turned onto the left side, particularly in expiration; often the supine PMI becomes identifiable after the patient is again turned supine. In

some normal adults an apex impulse will not be detectable in either position. Causes of a nonpalpable apex impulse are listed in Table 5-4.

Lateral Displacement without Cardiomegaly. In occasional individuals, the apex beat or PMI may be displaced to the left, but the actual size of the cardiac and LV chamber is normal. This may occur with skeletal abnormalities such as scoliosis, the straight-back syndrome (narrow A-P diameter, loss of normal thoracic kyphosis, marked pectus excavatum), or in subjects who have intrathoracic pathology. A large right pleural effusion, pneumothorax, atelectasis, or extensive pulmonary fibrosis may distort the heart's position in the chest and result in a displaced cardiac impulse without actual cardiomegaly.

WHAT TO LOOK FOR

The assessment of the apical cardiac impulse should include analysis of the following parameters: (1) location, (2) duration, (3) size, (4) force, and (5) contour. In addition, visual inspection of the chest for the presence of prominent retraction waves, as well as systolic and diastolic events, should be carried out. As already mentioned, a complete precordial examination consists of a systematic evaluation of the lower sternal area, pulmonary and aortic regions, and sternoclavicular sites.

Characteristics of the Apex Impulse

Location. Identify the site of impulse on the thorax with respect to both the longitudinal and horizontal axes of the patient. Note in which intercostal space the PMI or apex beat is located; occasionally a large heart will result in detectable precordial activity in 2 or even 3 intercostal spaces. Localize the apical impulse with reference to the midclavicular line, distance from the midsternum, or relationship to the left anterior axillary line.

Duration. The duration of the systolic outward motion is probably the most important feature of the precordial exam. *Practical Point: Although cardiomegaly or hypertrophy can exist in the presence of a brief "normal" outward movement, or even when the PMI is absent, a truly sustained left*

TABLE 5-4 *Causes of Nonpalpable Apical Impulse*

-
- Obesity
 - Muscular chest wall
 - Barrel chest
 - Emphysema
 - Coronary artery disease with decreased apical motion
 - Pleural or pericardial effusion
 - Age 50 or over
-

ventricular impulse in the supine or 30-degree elevation positions is distinctly abnormal (Fig. 5-3C). Such findings suggest a pressure overloaded ventricle (e.g., aortic stenosis, hypertension), a depressed LV ejection fraction, or a substantially dilated LV cavity. A study by Conn demonstrated that a *sustained* apex impulse in the supine position correlates highly with an increase in angiographically-determined, left ventricular mass, and this observation was actually more sensitive than the EKG in the diagnosis of left ventricular hypertrophy.

The critical point to assess is whether or not the impulse "stays up" into the second half of systole. Proper timing of the apex beat using simultaneous auscultation of S1 and S2 is essential in making this observation. With practice, one can be quite accurate in assessing the actual duration of the apex impulse. Simple observation of movement of the head of the stethoscope resting on the PMI may be helpful.

Left Lateral Decubitus Position (Fig. 5-7). It is unclear whether an LV impulse that becomes sustained when the patient is turned into the left lateral position but is of normal duration in the supine position has the same specificity for left ventricular enlargement as a sustained impulse or LV heave that is present when the patient is lying flat. Because the left decubitus position causes the heart to more closely approximate the chest wall, many experts feel that the observation of a prolonged impulse in this position has little diagnostic value. I believe that a definite *sustained* impulse in the left lateral position is highly suggestive of true LV dilatation. False positives may occur, however, and echocardiography is recommended in equivocal cases.

Size. If the apex impulse is larger than normal, it is useful to note the area of contact with the chest. Any impulse greater than 2 to 2½ cm in the supine position, or more than 3 cm in the left decubitus position, represents cardiac enlargement.

Force or Amplitude. Is the apex beat a soft, unimpressive impulse or does it lift the examining fingers off the chest wall? Is the anterior or outward excursion greater than normal, consistent with a hyperdynamic or hyperkinetic PMI? An increase in force is consistent with LV hypertrophy and preserved systolic function. Assessment of the force of contraction is the most subjective and least quantifiable aspect of precordial examination.

Contour. The normal apical impulse consists of a brief, nonsustained anterior motion in early systole (Figs. 5-1, 5-2). A sustained LV beat is the commonest abnormality of contour, but occasionally other patterns are noted.

A double systolic impulse may be seen in hypertrophic obstructive cardiomyopathy and occasionally in some patients with severe left ventricular dyssynergy or an LV aneurysm in coronary artery disease (Fig. 22-1). Pre-systolic distention commonly is found in the left lateral position in patients who have decreased LV compliance, such as coronary artery disease, hypertensive heart disease, or aortic valve disease (Fig. 7-6) (see below). Palpable

diastolic events, such as an S3 or a pericardial knock, also fall into the general category of contour changes (Fig. 5-4).

Palpable A Wave (Presystolic Distention). (Table 5-5). This is usually detected in the left decubitus position. The apex beat is carefully identified, and varying pressure with the pads of the fingers is applied at the site of the LV impulse. A double, early systolic impulse is noted, which feels like a "shelf" or ridge on the upstroke of the beat (Fig. 7-6). Usually lighter pressure will maximize tactile perception of this low amplitude finding, whereas firm pressure with the fingers may make it more difficult or impossible to feel. On occasion this presystolic distention can be quite prominent.

The palpable A wave reflects a high LV end-diastolic pressure and decreased LV compliance. It is an important observation as it documents a definite abnormality and suggests the presence of LV hypertrophy and increased chamber stiffness. Audibility of the S4 does not correlate with palpability; presystolic distention may be detectable when the S4 is quite soft. Occasionally one is not able to hear an S4, although it is clearly palpable, because of the extremely low frequency vibrations of the diastolic filling event, usually less than 50 cycles per second.

Palpable S3. The third sound is less often palpable than an S4. It is most often found in severe mitral regurgitation or a markedly dilated cardiomyopathic ventricle. As with the S4, palpation of the LV S3 is greatly enhanced in the left decubitus position. The S3 will be noted as a brief outward motion occurring in early diastole which gently taps the examiner's finger pads (Figs. 5-5, 7-3).

Parasternal or Right Ventricular Impulses. All of the above also applies to the evaluation of RV or parasternal activity. However, the low amplitude impulse produced by right ventricular hypertrophy or dilatation usually is more difficult to evaluate than left ventricular apical activity. Nevertheless, an increase in amplitude or sustained parasternal motion usually is discernible with careful examination technique.

Right ventricular abnormalities are only detectable in the supine position. Firm downward pressure with the hand on the lower left sternal area is usually necessary; the patient should be alerted that the examiner will be pushing down on the sternum (Fig. 5-9). Since RV activity is usually low amplitude, it will not be detected without such firm compression. Held end expiration may be very useful in detecting a subtle or slight RV lift.

TABLE 5-5 *Palpable A Wave (Presystolic Distention)*

-
- Aortic stenosis
 - Hypertrophic cardiomyopathy (IHSS)
 - Coronary artery disease—acute and chronic
 - Acute mitral regurgitation
 - Aortic regurgitation (uncommon)
 - Longstanding hypertension
-

In adults with RV enlargement from acquired heart disease, pulmonary hypertension invariably is present. A palpable P2 and pulmonary artery impulse in the second or third interspace should be sought in these patients to provide valuable confirmatory evidence for RV hypertrophy.

Palpable RV diastolic events are often best detected with the subxyphoid or epigastric approach utilizing held inspiration with the fingers extended to identify these low frequency and low amplitude motion abnormalities (Fig. 5-10).

Precordial Motion Abnormalities in Specific Cardiovascular Disorders

The following section summarizes the major palpable findings in a variety of common adult cardiovascular conditions (see also Table 5-3).

Aortic Stenosis. In mild aortic stenosis the PMI may be normal. The apical impulse in hemodynamically significant valvular aortic stenosis is sustained with outward motion remaining palpable in late systole (Figs. 5-3C, 13-4). (Also see Chapter 13.) It is typically forceful and readily displaces the examining fingers. Unless LV function is impaired and/or LV dilatation has occurred, the apical impulse usually is not displaced laterally more than 1 to 2 cm from the normal location. The impulse may be felt in more than one interspace in severe aortic stenosis. Presystolic distention of the LV (palpable A wave) is common in such situations, particularly in the left lateral position (Figs. 7-6, 13-4). *Practical Point: A palpable S4 in a patient with aortic stenosis correlates with a large left ventricular-aortic pressure gradient (unless there is coexisting coronary or hypertensive heart disease).*

A systolic thrill at the 2nd right interspace is often present in valvular aortic stenosis. Palpable sound vibrations may radiate upward toward the clavicle, and a thrill may also be present over the manubrium or at the 2nd left interspace. In older patients or those with an increased thoracic diameter, a systolic thrill is occasionally detected at the apex. An aortic thrill is usually best felt with subject upright and leaning forward with the breath held in end-expiration.

A palpable aortic ejection sound is common in subjects with a congenitally bicuspid aortic valve (Fig. 13-6). This sound typically is felt only at the apex and is distinguishable from the more protracted LV impulse as a discrete high frequency transient. A palpable thrill or ejection sound correlates poorly with the hemodynamic severity of the valve obstruction, whereas a sustained apical impulse or presystolic distention of the LV usually indicates significant aortic stenosis.

Aortic Regurgitation (Also see Chapter 15). In mild to moderate aortic regurgitation the apex impulse may not be displaced but will typically be hyperdynamic and unsustained (Fig. 5-3B). As the left ventricular cavity enlarges, the PMI is displaced laterally and downward and often takes up

two or more interspaces. Marked medial retraction may be visible. With major volume overload secondary to aortic regurgitation, a sustained apical impulse is often present, indicating severe LV cavity dilatation. This may be present without significant depression of LV systolic function. In these patients the palpable surface area of the apex beat is increased, and in the left lateral position presystolic distention (palpable S4) may be felt. An audible or palpable S4 excludes coexisting mitral stenosis, which is often suggested by the presence of a diastolic rumbling murmur known as the Austin Flint murmur (see Chapters 13, 15). If coexisting aortic stenosis is present the apical impulse will be sustained, large, forceful, and displaced inferolaterally.

It is rare to detect a palpable diastolic thrill along the left sternal border unless there is perforation or eversion of an aortic cusp resulting in an extremely loud diastolic murmur.

Mitral Regurgitation (see Chapter 17). The apex impulse is normal to hyperdynamic in mild to moderate mitral regurgitation. In severe mitral regurgitation, particularly when chronic, the LV impulse is displaced laterally and has an increased force and amplitude. If the PMI is sustained, it suggests that LV systolic function has decreased and that the ejection fraction is abnormal or that major LV dilatation has occurred. The apical impulse may be quite large and detectable in two or more interspaces. It is common to feel a systolic apical thrill in severe mitral regurgitation; with the onset of congestive heart failure or marked depression of left ventricular function, the thrill may disappear as the murmur attenuates because of a decrease in systolic performance.

Mitral regurgitation may produce a *late* systolic impulse at the parasternal area or lower left sternal border (Fig. 17-4). This outward motion can be quite prominent, reflecting the systolic jet of a large volume of blood into a dilated left atrium that expands during LV ejection (Fig. 17-5). It is critical to time this parasternal activity to assess whether it is early or holosystolic as opposed to late systolic (Fig. 17-4). Simultaneous palpation of the LV and RV areas is mandatory; if the parasternal lift is due to left atrial expansion and not to pulmonary hypertension it will occur in the second half of systole following S1 and *after* the LV impulse is felt. If there is coexisting pulmonary hypertension, the parasternal impulse will be sustained throughout systole. In such cases, the independent effect of left atrial systole will not be detectable.

A visible and palpable S3 may occur in severe mitral regurgitation, particularly in the left lateral position (Fig. 5-5). In acute mitral regurgitation, a palpable S4 may be noted; the finding of presystolic distention is extremely important, as it indicates that the condition is of recent onset (see Chapter 17).

Mitral Stenosis. Precordial examination in mitral stenosis is often rewarding, and the diagnosis of this condition may be suggested even before the stethoscope is used (see Chapter 16). S1 is typically palpable at the apex or medially. S2 may be palpable if there is pulmonary hypertension (increased

P2). The opening snap commonly is palpable between the lower left sternal border and apex, especially in thin persons. In the left lateral position, a diastolic thrill may be manifest at the apex, but is usually palpable only over a small area.

Left ventricular activity in pure mitral stenosis is unimpressive. However, most patients with moderate to severe degrees of mitral stenosis will have a parasternal or right ventricular lift. Even when resting pulmonary artery pressure is not elevated, with exercise pulmonary artery pressure typically rises in mitral stenosis, and some degree of right ventricular hypertrophy is frequently present. A holosystolic parasternal lift is common in hemodynamically significant mitral stenosis. The large left atrium will thrust the RV more anteriorly, and this displacement may also contribute to the presence of detectable parasternal activity in mitral stenosis. The more vigorous and sustained the right ventricular lift, the more likely significant pulmonary hypertension will be present.

Hyperrophic Cardiomyopathy (IHSS) (see Chapter 14). In this unusual condition precordial palpation may be quite informative. Left ventricular compliance is markedly decreased. Thus, the A wave typically is very prominent and the left ventricular impulse is forceful and vigorous. The heart is usually not displaced to the left. A mid or late systolic secondary "bulge" may be present, resulting in a double or bifid precordial impulse (Fig. 14-5). When the A wave is palpable, the precordial motion actually will be trifid in nature ("triple ripple") (see Figure 14-6). A palpable A wave in hyperrophic cardiomyopathy does not necessarily correlate with a large gradient across the left ventricular outflow tract.

A systolic thrill, usually somewhat superior and medial to the apex, is often present. The murmur and thrill in hypertrophic cardiomyopathy typically do not radiate to the neck, in contradistinction to aortic stenosis. In occasional patients, obstruction of the RV outflow tract is also present; this may result in a parasternal heave as well as a systolic thrill in the 3rd to 4th interspace adjacent to the sternum.

Cardiomyopathy. The typical precordial finding in congestive cardiomyopathy is a diffuse anterior precordial motion; it is often difficult to be sure that this is entirely left ventricular in origin. The LV impulse is sustained and displaced inferolaterally, may or may not be forceful, and usually occupies more than one interspace. Presystolic distention (palpable A wave) and a palpable S3 are common. Parasternal activity is often present; this contributes to the diffuse heaving or rocking precordial motion typical in such patients. Careful observation may detect a retraction wave between the parasternal and apical regions. On rare occasions, a mid or late systolic bulge near or at the apex may be observed.

Coronary Artery Disease. In patients with angina pectoris who have no history of myocardial infarction, the apical impulse is usually normal,

although presystolic distention occasionally may be noted in the left lateral position (Fig. 7-6). A palpable S4 is probably the most common abnormality of precordial motion in subjects with coronary artery disease. It is important to examine all patients with suspected or proven coronary artery disease in the left lateral decubitus position for optimal palpation and auscultation. If one examines a patient during an episode of angina, the appearance of an ectopic ventricular pulsation, presystolic distention of the LV, or a sustained PMI may rarely be detected.

In subjects with prior myocardial infarction the apical impulse may be normal, sustained, or ectopic, or there may be late systolic motion suggesting LV dyssynergy (see Chapter 22; Fig. 22-1). Ectopic impulses are common in patients with left ventricular aneurysms or severe LV dyssynergy. If there is anteroseptal dyssynergy, a parasternal lift may be present simulating right ventricular hypertrophy. A sustained apex impulse suggests either LV hypertrophy or a wall motion abnormality.

Atrial Septal Defect (see Chapter 20). The uncomplicated atrial septal defect (ASD) causes volume overloading of the right ventricle, and this usually results in an easily detectable, nonsustained left lower sternal pulsation (hyperdynamic RV) (Fig. 20-3). A pulmonary artery lift is commonly felt in the 2nd and 3rd left interspace. If coexisting pulmonary hypertension is present, the parasternal or RV impulse is sustained and "heaving" in quality. Even without significant pulmonary hypertension, a high flow ASD with a large volume overload may produce a sustained right ventricular lift. P2 and a pulmonary artery impulse may be palpable in patients with an ASD and do not necessarily imply pulmonary hypertension.

Tricuspid Regurgitation (See Chapter 19). Tricuspid regurgitation in an adult almost always is associated with acquired pulmonary hypertension and right ventricular hypertrophy. In such instances, a right ventricular or parasternal heave is likely to be present. Systolic retraction is commonly seen medial to the apex (LV) beat. Remember that subxyphoid palpation may be necessary to detect RV activity in patients with chronic obstructive pulmonary disease (COPD) or obesity (Fig. 5-10). Severe tricuspid regurgitation rarely may produce right lower anterior chest pulsations that are palpable and visible, reflecting expansion of the right atrium during systole. Careful examination of the liver during held inspiration will usually reveal a hepatic pulsation with each cardiac systole (Fig. 19-5). This is an undulant, low pressure impulse and visual observation of the examining hand and fingers is often more helpful than palpation itself.

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Chapter 6

The First and Second Heart Sounds

FIRST HEART SOUND (S1)

In general, abnormalities of S1 do not provide a great number of important clues in cardiac physical diagnosis. Alterations in intensity of S1 are the most useful observations that can aid the clinician.

NORMAL PHYSIOLOGY

The first heart sound (S1) signals the onset of left ventricular contraction. However, electrical initiation of LV systole begins 50 to 60 msec before S1 is heard. Pressure within the LV begins to develop just prior to S1 (Fig. 6-1). Left ventricular pressure rises above LA pressure well *before* forward flow across the mitral valve ceases and the valve leaflets have reached their maximally closed position.

Two major, medium-high frequency components of S1 usually can be heard in normal people ("split S1") (Fig. 6-2). However, some high fidelity, filtered recordings have demonstrated a total of three or even four separate "components" to the normal S1; some vibrations are low frequency and low energy, and therefore inaudible. There are two schools of thought regarding the actual source of the first major vibrations of S1. Luisada asserts that the onset of isovolumic LV contraction results in a prominent tensing of the LV walls, septum, and mitral valve apparatus which produces a sound transient. This view, precluding any role for the mitral or tricuspid valve in the production of S1, assumes that mitral valve closure precedes the major sound vibrations of S1. In contrast, Craige, Leatham, and others have demonstrated convincingly that the first major component of S1 ("M1") is coincident with the maximal closing excursion of the mitral cusps. They believe that M1 reflects the sudden tensing of the closed mitral valve leaflets, which sets the surrounding cardiac structures and blood into vibration. Most, but not all, simultaneous echophonocardiographic studies indicate that mitral valve closure (M1) coincides precisely with the first major recordable sound transient of S1. Thus, despite Luisada's disagreement, it is correct to continue to use the terminology of "M1" for the first major vibration of S1.

The mechanism of the second major audible vibration of S1 is also controversial. Craige and Leatham have obtained echophonocardiograms con-

firmering the coincidence of tricuspid valve closure ("T1") with the second component of S1. However, other echocardiographic studies have demonstrated that tricuspid valve closure may coincide with either the first or the second component of S1. Luisada and Shaver believe that the second major audible vibration of S1 is an ejection component coincident with the opening

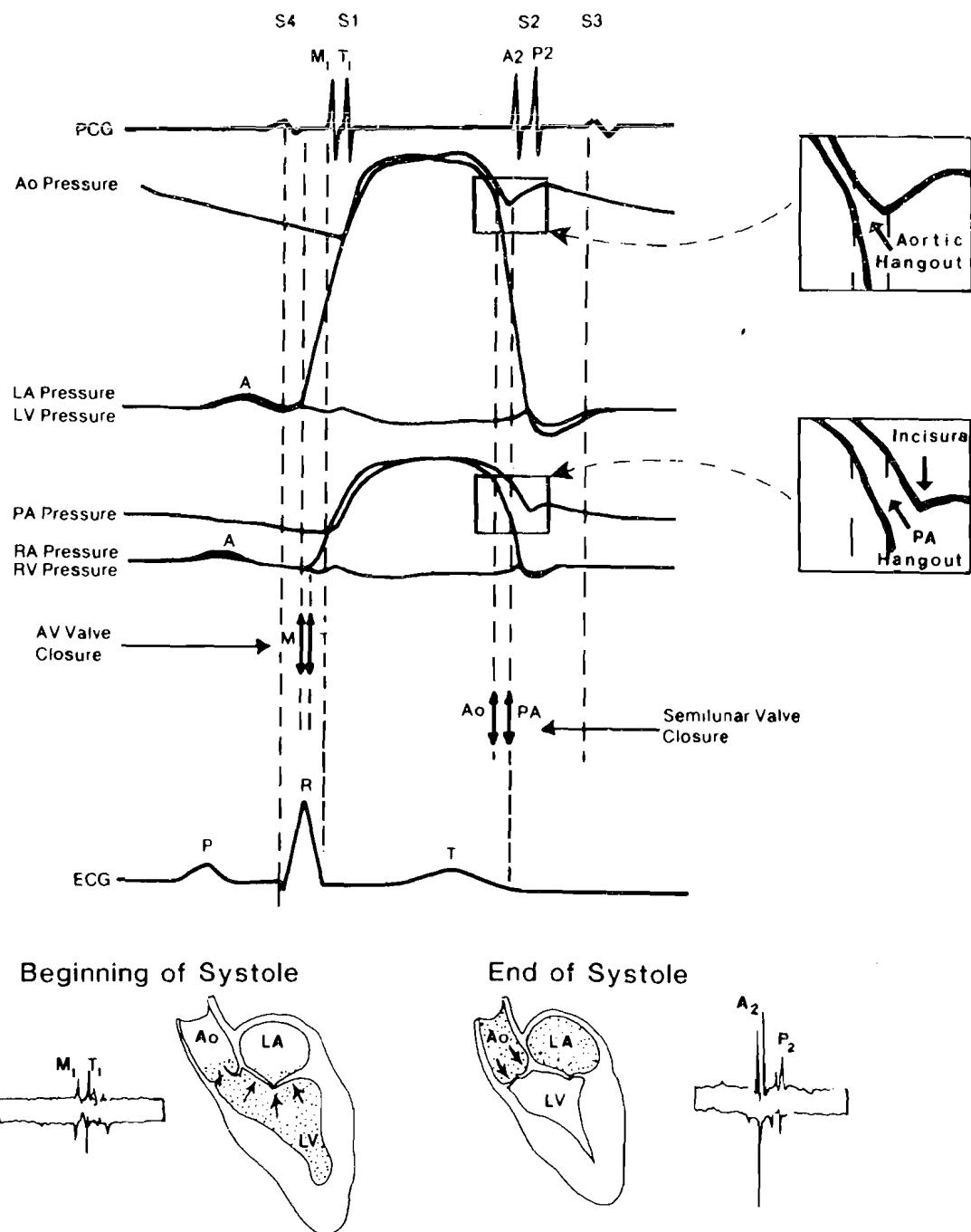


FIG. 6-1. Relationship of heart sounds to intracardiac pressures and valve motion. Note that pressure crossover between the atria and the ventricles as well as the ventricles and the great vessels always precedes the resultant cardiac sound. Actual valve closure is either synchronous with or immediately precedes the related heart sound. See text for further discussion. (Adapted in part from Luisada A and MacCanon DM and modified from Abrams J: Prim Cardiol, 1982.)

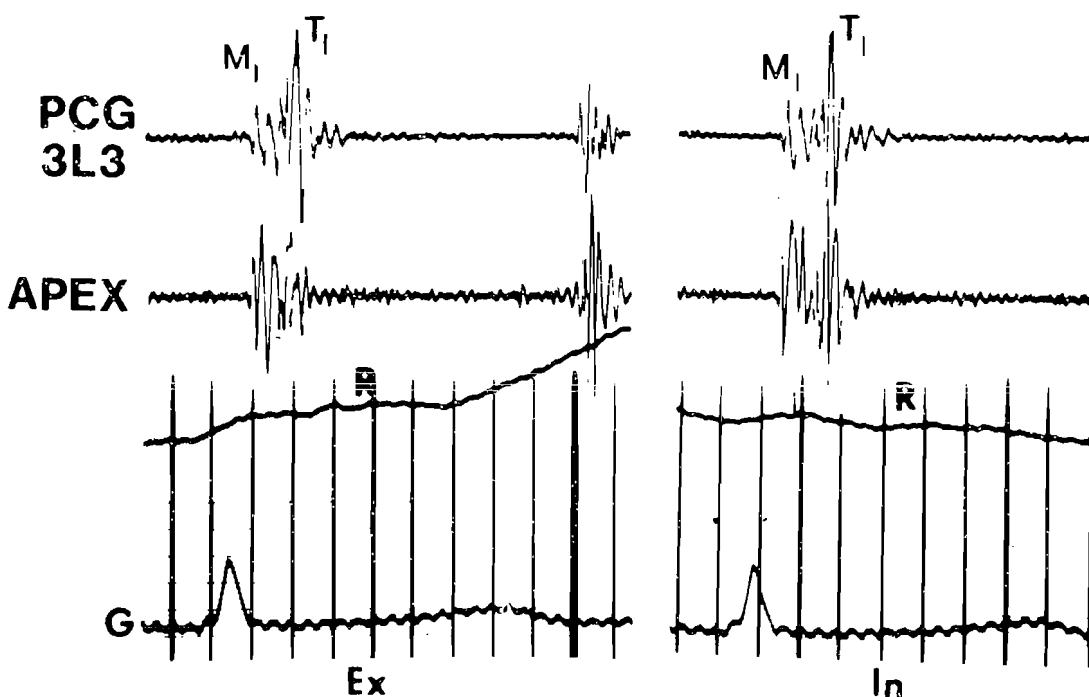


FIG. 6-2. Splitting of the first heart sound (S1). The components M1 and T1 represent mitral and tricuspid valve closure, respectively. There is still controversy as to whether the recordable and audible components of S1 reflect events relating to actual valve coaptation or are generated solely within the ventricles. Note the inspiratory increase in T1 in the recording at the apex. R = respiration, EX = expiration and IN = inspiration. (Adapted from Sainani GS et al: Cardiologia 52:252, 1968.)

of the aortic valve. Some echo studies demonstrate that tricuspid valve closure precedes aortic valve opening by 15 to 25 msec. One of the major difficulties in resolving this controversy is that so many intracardiac events occur within a brief period of time: rapidly rising LV pressure, mitral valve closure, tricuspid valve closure, aortic valve opening, and LV ejection. The temporal coincidence of several of these phenomena allows for varying interpretations and unresolved controversy. Most investigators concur that the last recordable component of S1 coincides with the onset of ejection into the aorta. Some believe that this is a normal ejection phenomenon that may become accentuated and delayed in disease states resulting in the ejection sound or click heard in certain pathologic conditions (see Chapter 8).

FACTORS AFFECTING INTENSITY OF S1 (Table 6-1)

PR Interval (Fig. 6-3). A shorter PR interval results in late mitral valve closure and a loud S1. When the PR is short, left ventricular isovolumic systolic pressure is higher at the time of LV-LA pressure crossover, causing a more rapid mitral valve closing motion and an increased intensity of S1. The relative distance between the two mitral valve leaflets at the beginning of mechanical systole is also important. If the mitral valve has already closed

TABLE 6-1 *Factors Affecting the Intensity of S1*

<i>Loud S1</i>
Short PR interval (< 160 msec)
Tachycardia or hyperkinetic states
"Stiff" left ventricle
Mitral stenosis
Left atrial myxoma
Holosystolic mitral valve prolapse
<i>Soft S1</i>
Long PR interval (> 200 msec)
Depressed left ventricular contractility
Premature closure of mitral valve (e.g., acute aortic regurgitation)
Left bundle branch block
Extracardiac factors (e.g., obesity, muscular chest, COPD, large breasts)
Flail mitral leaflet

or the leaflets are closing, the first sound is soft; if the leaflets remain open deep within the LV at the onset of systole, S1 will be loud.

Maximal (increased) intensity of S1 occurs at a PR interval range of 80 to 140 msec. PR intervals over 140 msec (0.15 sec) result in a normal sounding S1, and PR intervals greater than 200 msec produce an attenuated or absent S1 because the mitral valve has already closed prior to development of LV pressure. In complete heart block, a nonconducted P wave in early or mid diastole may result in closure of the mitral leaflets, which subsequently may drift open again; a ventricular contraction then may produce a loud S1.

Left Ventricular Contractility. In general, the more vigorous the LV contraction, the louder the S1. LV pressure during isovolumic systole at first rises slowly but then rapidly accelerates. Depressed LV contractility and a decreased rate of LV pressure development will result in a decreased intensity of S1. Hyperadrenergic states (excitement, fever, exercise) commonly increase S1 amplitude.

In general, the position of the mitral valve leaflets at end-diastole and their closing velocity are more important than the state of LV contractility in determining the intensity of S1.

Mitral Leaflets and Left Ventricle. Mobile but abnormally stiff mitral valve leaflets, such as those found in mitral stenosis, produce a loud S1 unless the cusps are severely distorted, fibrotic, or calcified. Inability of the LV to maintain isovolumic systole intact will reduce the intensity of S1 as in mitral regurgitation, ventricular septal defect, or a left ventricular aneurysm (where blood may be displaced within the ventricle prior to aortic valve opening).

HOW TO LISTEN TO S1

Timing. Experienced physicians rarely have difficulty identifying S1 unless there is a rapid heart rate and/or several murmurs are present. Tachy-

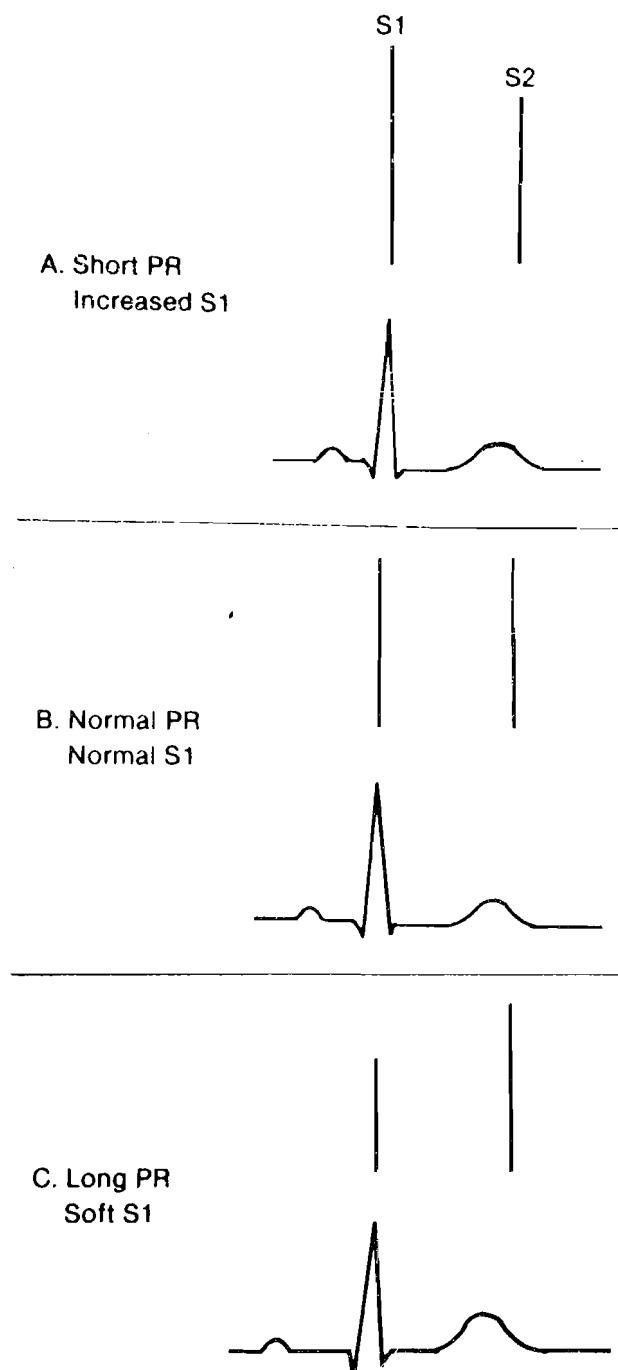


FIG. 6-3. Relationship of intensity of S1 to the PR interval. A. Short PR interval (< 0.15 seconds). S1 is accentuated. B. Normal PR and normal S1 intensity. C. With long PR intervals (> 0.20 seconds) the resultant S1 is reduced in intensity and may be very soft. (From Abrams J: Prim Cardiol, 1982.)

cardia shortens diastole relatively more than systole. At heart rates of 120 to 130, systole and diastole are of equal length. If there is any doubt of the proper identification of S1 and S2, S1 should be timed simultaneously with palpation of the carotid pulse or apex impulse (see Figure 1-4). S1 immediately precedes the palpable carotid arterial upstroke, and S2 occurs immediately after the peak of the carotid pulse. S1 will appear to initiate the outward LV thrust of the apex beat. If the stethoscope is placed directly on the apical impulse, it will begin to lift upward with the onset of S1.

Characteristics. Typically, S1 is of medium to high frequency, although occasionally it is low-pitched. It is almost always lower pitched than S2. Use of the diaphragm or increased pressure with the bell will bring out the crisp, high frequency vibrations of S1. Splitting of S1 is audible in many, but not all, normal subjects; in most individuals a split S1 can be recorded with a high quality phonocardiogram (Fig. 6-2). With narrow splitting or relatively low intensity vibrations of S1, both components will not be heard. S1 splitting often is better detected medial to the apex or at the lower sternal border. A loud first component (M1) at the apex may mask the second component of

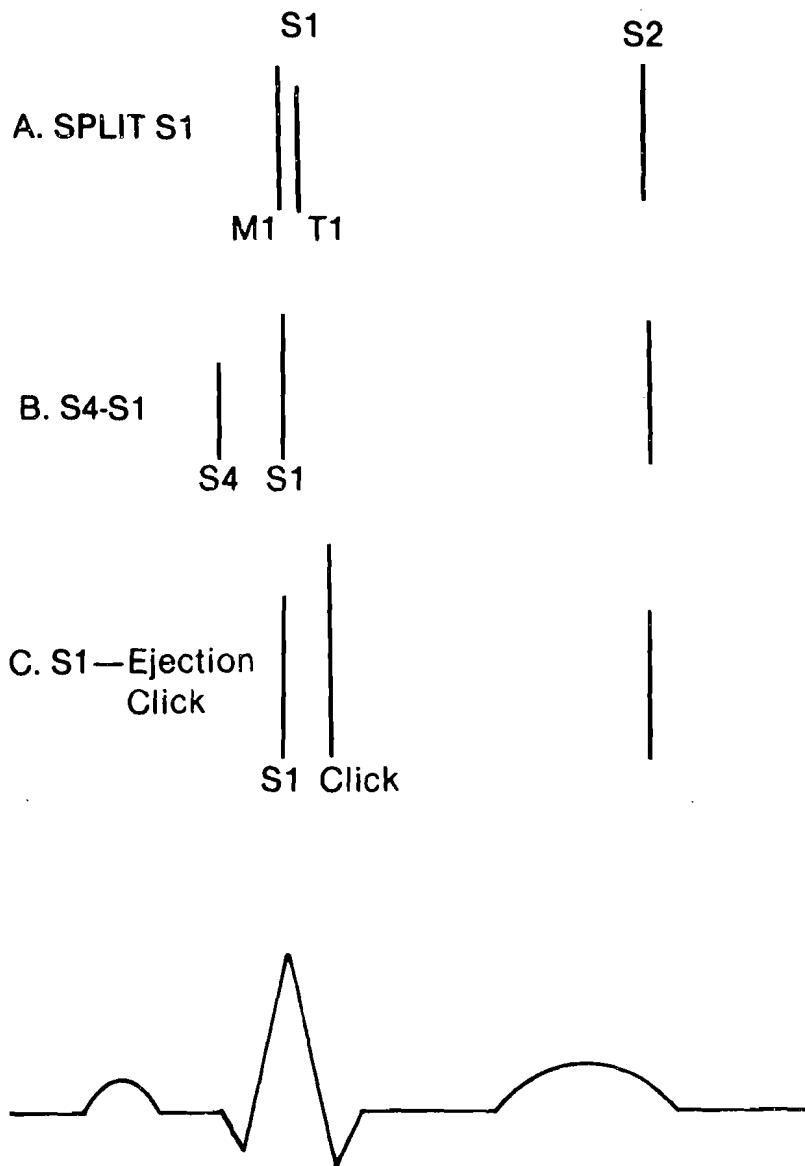


FIG. 6-4. Differential diagnosis of audible splitting of the first heart sound. A. M1 and T1 are both audible. B. S4-S1 complex. C. S1-ejection click complex. Techniques for differentiating these sound combinations are discussed in the text. (From Abrams J: Prim Cardiol, 1982.)

S1 (T1). T1 is usually softer at the apex and louder at the lower left sternal border (where splitting of S1 is typically best appreciated). S1 usually is not prominent at the base and should always be single at the 2nd and 3rd interspaces. *Practical Point: When apparent splitting of S1 is heard at the base, one should suspect the presence of an ejection sound or early midsystolic click.* Splitting of S1 is usually best detected in expiration, although the second component (T1) may be more prominent with inspiration (Fig. 6-2). The average splitting interval between the two components of S1 is 30 msec or 0.03 sec; M1 occurs 60 msec after the onset of the QRS and T1 occurs 90 msec after the QRS. Loud systolic murmurs may obscure the S1.

Differentiation of Split S1 from S4-S1 or S1-Ejection Click. Proper identification of two audible heart sounds close to S1 provides an important diagnostic challenge to the clinician (Fig. 6-4). An S4 may be confused with the first component of S1 (see Chapter 7; Fig. 6-4B). The S4 usually is audible only at the apex, and often only in the left lateral position. Fourth heart sounds of LV origin are not heard at the lower left sternal border where splitting of S1 is best detected. The S4 may vary in intensity with respiration, whereas S1 typically does not. The S4 is low pitched unless very loud and may be associated with palpable presystolic distension of the left ventricle. An S4 should attenuate when pressure on the bell of the stethoscope is increased, whereas the two components of S1 are best heard with the diaphragm.

An ejection click may be difficult to differentiate from a split S1 (see Chapter 8; Fig. 6-4C). The ejection sound usually is more intense than the second component of S1 (T1) and often will be heard well at the base. Splitting of the S1 is not heard in this area. A pulmonic ejection click will vary with respiration if caused by pulmonic stenosis; pulmonic clicks due to pulmonary hypertension will be accompanied by other signs of elevated pulmonary artery pressure (e.g., increased P2, RV lift). Aortic clicks typically are very discrete and snappy at the apex and occur later than the more closely split S1. The LV apex usually is the site of maximal intensity of the aortic ejection sound.

ABNORMALITIES OF FIRST HEART SOUND

The study of S1 and the way it changes in disease states has been useful in elucidating the mechanisms of heart sound production in normal subjects and in certain pathologic conditions such as mitral stenosis. Detectable abnormalities on auscultation are few. *Practical Point: The loudness of S1 provides the most important clinical information in auscultation.* The intensity should be noted and compared to the amplitude of S2 as well as to other cardiac sounds.

Increased Intensity of S1 (Table 6-1)

An accentuated S1 is always louder than A2 at the cardiac apex and may equal or exceed the intensity of S2 at the base. Commonly, S1 is loud in conditions with increased adrenergic activity or hyperkinetic states such as anemia, exercise, thyrotoxicosis, and anxiety. The common denominator is increased LV contractility due to enhanced sympathetic tone. Whenever tachycardia is present, shortened diastole and a decreased PR interval will produce a loud S1 as a result of atrial systole encroaching upon left ventricular isovolumic contraction.

Short PR intervals, such as in the Wolff-Parkinson-White and Lown-Ganong-Levine syndromes will produce a louder S1. An increased S1 often is associated with a hypertrophied, noncompliant, left ventricle. This results from an elevated LV end-diastolic pressure that causes a late and abrupt acceleration of mitral valve closure. In mitral valve prolapse with early or holosystolic prolapse S1 may be very loud (see Chapter 18).

Mitral Stenosis. The loud S1 in rheumatic mitral stenosis is an extremely valuable diagnostic clue (see Chapter 16). S1 in mitral stenosis is usually palpable at the apex or just inside the apex impulse. The loud S1 results from several factors: (1) persistently elevated LA pressure in late diastole that allows ventricular pressure to rise considerably higher than normal during isovolumic systole but before LV-LA crossover; (2) a wide closing excursion of the mitral valve leaflets, which are held open deep within the left ventricle during late diastole, with the result that the mitral component (M1) is delayed and may follow T1; (3) the stiff, noncompliant leaflet and chordal tissue probably resonate with an increased sound amplitude. *Practical Point: A loud S1 is almost always present in mitral stenosis, except when the mitral valve is so severely fibrosed and calcified that it is virtually immobile. In the absence of a loud S1, the diagnosis of mitral stenosis should be reconsidered.*

Left Atrial Myxoma. A related cause of an increased S1 is a left atrial myxoma. S1 may be loud and slightly delayed due to the obstructing tumor which results in elevated LA pressure as it keeps the mitral valve cusps apart in late diastole.

Decreased Intensity of S1 (Table 6-1)

A soft S1 is a clue to a long PR interval which may result in premature mitral valve closure (Fig. 6-3C). Audible reduction in S1 amplitude occurs at PR intervals greater than 0.16 sec, and S1 is markedly attenuated with a PR longer than 0.20 sec. Impaired LV function, accompanied by a depressed rate of rise of isovolumic pressure, can produce a decreased S1. Thus, S1 may be soft in congestive heart failure, severe aortic and mitral regurgitation,

as well as in patients with coronary disease and markedly decreased LV contractility. In aortic valve disease, particularly acute aortic regurgitation, a markedly elevated, left ventricular end-diastolic pressure may result in *premature closure* of the mitral valve, thus reducing the intensity of S1. S1 usually is decreased in left bundle branch block (LBBB), in part because of the decreased LV contractility often associated with LBBB and in part because of the delay in onset of LV contraction. M1 actually may follow T1 in patients with LBBB.

Attenuation in S1 may result from extra-cardiac factors such as thick muscular chests, obesity, large breasts, increased thoracic diameter, and chronic obstructive lung disease. As mentioned, severe calcific mitral stenosis may be associated with a diminished S1, and in such patients the opening snap is also quite soft or inaudible (see Chapter 16).

Abnormal Splitting of S1

Wide splitting of S1 can result from an electrical delay in ventricular activation, which results in asynchrony of contraction, or from an increase in LV isovolumic contraction time. Right bundle branch block (RBBB), premature ventricular contraction (PVC) with RBBB configuration, left ventricular pacing, and ectopic ventricular rhythms with RBBB pattern all have been associated with prominent splitting of S1. In these situations, S2 usually is widely split as well. Splitting of S1 may be more noticeable in older subjects due to slight prolongation of the LV preejection time that occurs with aging. In acute myocardial infarction, splitting may be prominent for the same reasons.

Certain abnormalities of the right heart producing an accentuated tricuspid closure sound (T1) have been documented. Prominent splitting of S1 is common in Ebstein's anomaly; the delayed closing motion of the septal leaflet of the tricuspid valve produces a loud, early systolic closing sound. The tricuspid closure sound is exaggerated in patients with atrial septal defect or anomalous pulmonary venous drainage. The exact mechanism of the loud second component of S1 in such cases remains controversial. In such cases, some believe that the loud T1 represents a pulmonic ejection sound (see Chapter 8) rather than accentuated tricuspid closure.

Variable S1

Whenever the relationship between the position of the mitral valve leaflets and LV pressure rise is inconstant, the intensity as well as splitting interval of S1 will vary. Thus, second or third degree heart block, AV dissociation, and ventricular rhythms with dissociated atrial rates all will result in an S1 of variable intensity.

SECOND HEART SOUND (S2)

Careful and intelligent evaluation of the intensity and splitting characteristics of the second heart sound (S2) represents one of the most valuable aspects of cardiac physical diagnosis. Clues to unsuspected or known cardiovascular abnormalities are frequently detected after accurate assessment of S2. The second heart sound is comprised of an aortic (A2) and a pulmonic (P2) component; both A2 and P2 should be separately sought, identified, and analyzed on auscultation.

NORMAL PHYSIOLOGY

The two components of S2 represent vibrations of cardiac structures and great vessels resulting from deceleration of the blood mass at the end of ventricular systole when the *semilunar valve cusps coapt* to prevent diastolic reflux of blood into the ventricles (Fig. 6-1). At end-systole forward flow into the great vessels is virtually complete, and deceleration of the blood mass toward the semilunar valves is increasing. Crossover of aortic-left ventricular and pulmonary artery-right ventricular pressures precedes semilunar coaptation and cessation of forward flow (Fig. 6-1). The aortic and pulmonic valves close completely only after forward flow ceases. A2 and P2 are coincidental with, or occur just after, the actual closure of the valve cusps. The sounds A2 and P2 represent the abrupt tensing of the closed leaflets, as the valves, great vessels, ventricular outflow tracts, and blood mass oscillate with an intense medium to high frequency vibration. Most investigators have documented aortic valve leaflet coaptation occurring synchronously with the major component of A2; other studies suggest that valve closure immediately precedes A2. In actuality these two events are likely to be synchronous. In the pulmonary artery, P2 follows right ventricular-pulmonary artery pressure crossover and pulmonic valve closure after an interval longer than in the left heart, particularly during inspiration.

A2 and P2 coincide precisely with the respective incisurae in the aorta and pulmonary artery pressure recordings (Figs. 6-1, 6-5). The incisura represents the peak deceleration of the blood mass and immediately is followed by a rebound of pressure. The relative distensibility or stiffness of the pulmonary and systemic vascular tree provides a partial explanation for differences in the timing of A2 and P2. In the central aorta, resistance is relatively high, compliance is low, and the recoil from the ejection of blood into the aorta is brisk. Consequently, A2 and its incisura closely follow the end of LV ejection. In the pulmonary artery the highly distensible, low resistance pulmonary vasculature allows for a late recoil following right ventricular ejection. Thus, P2 and the pulmonary artery incisura are somewhat delayed

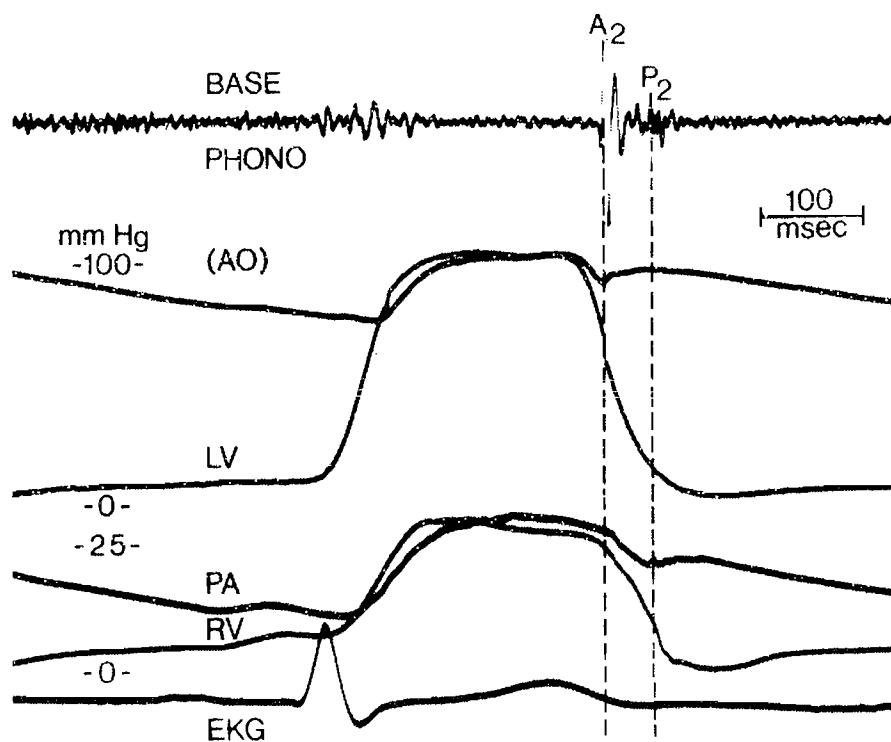


FIG. 6-5. Intracardiac pressures and their relationship to A₂ and P₂. Note that the aortic and pulmonic closure sounds (A₂ and P₂) are coincident with the incisurae of the aortic and pulmonary artery pressure trace, respectively. Right ventricular ejection ends after the left ventricle because of the prolonged right-sided hangout interval (see text). (From Shaver J and O'Toole J: Mod Concepts Cardiovasc Dis 46:7, 1977.)

after the end of RV systole. The distance between ventricular systolic pressure and its great vessel at the level of the incisura has been termed the *hangout interval* by Shaver and associates (Figs. 6-1, 6-5, 6-11). The pulmonary artery hangout interval typically is 2 to 5 times longer than aortic hangout interval.

Respiratory Effects of A₂ and P₂ (Fig. 6-6). Inspiration normally produces audible separation (inspiratory splitting) of A₂ and P₂. Recent investigations have helped to clarify this phenomenon. The differences in aortic and pulmonary artery vascular impedance and their respective hangout intervals are important to an understanding of the dynamic events that occur during respiration.

The classic view of the mechanism of inspiratory splitting of S₂ is that increased venous return during inspiration causes a delay in RV systole, whereas LV systole remains unchanged or is somewhat shortened; this difference results in an increase in the A₂-P₂ interval. As intrathoracic pressure decreases with inspiration, more blood enters the right ventricle, and its volume increases, prolonging RV ejection and the Q-P₂ interval. Several beats later during *expiration* this increased blood volume returns to the left ventricle. During expiration RV stroke volume diminishes as LV stroke volume increases; the Q-A₂ interval lengthens and Q-P₂ shortens. Inspiration also may

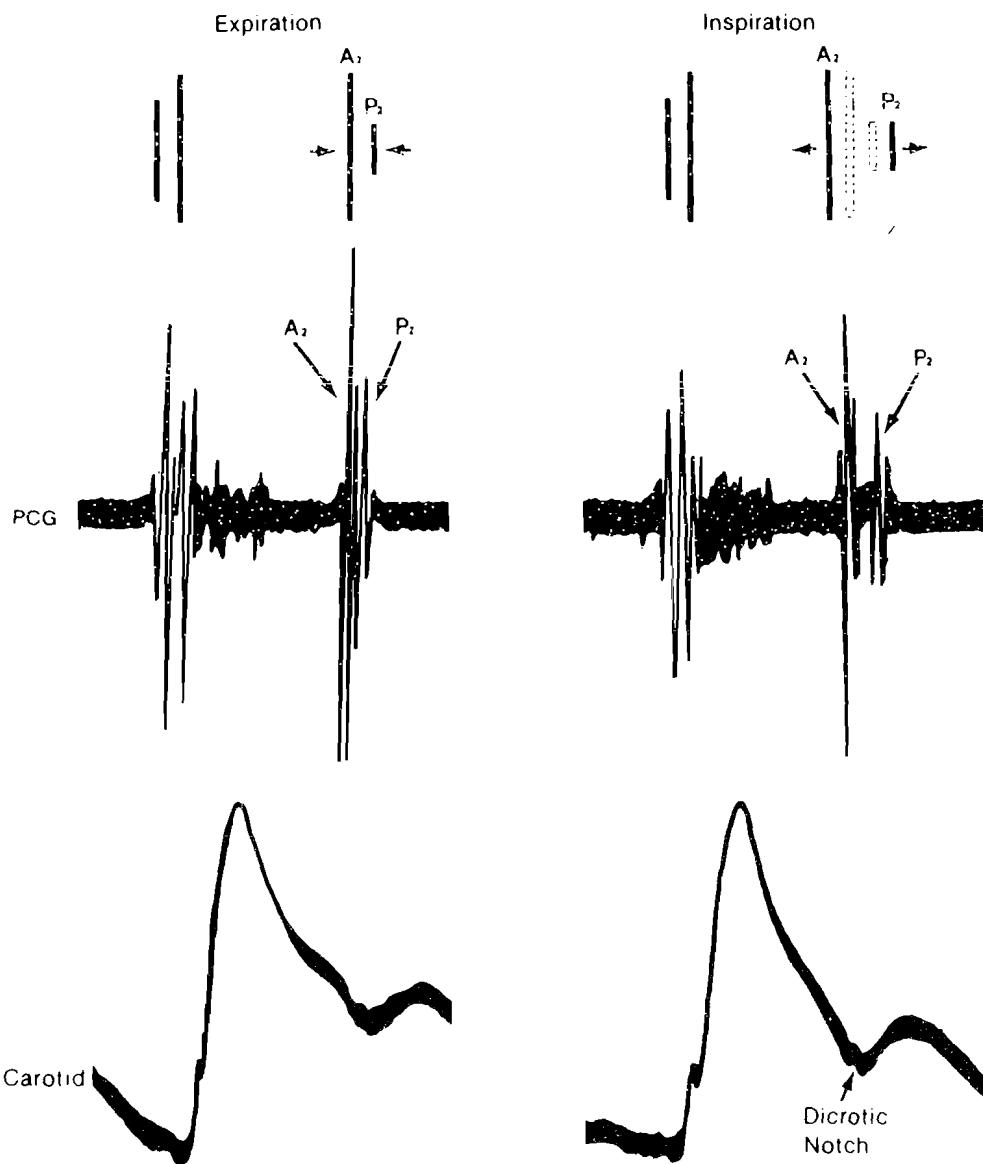


FIG. 6-6. Physiologic splitting of S2. Under normal circumstances S2 in expiration is heard as a single event, but in late inspiration audible splitting of A2 and P2 is noted. The normal range of expiratory splitting is from 10 to 60 msec averaging 40 msec. Most examiners cannot acoustically separate sounds that are 20 msec or less apart; thus a narrow splitting interval will be heard as a fused or single S2. The mechanisms of the inspiratory increase in the A2-P2 interval are discussed in the text; the inspiratory increase consists predominantly of an increase in the total Q-P2 duration. (From Abrams J: Prim Cardiol 1982.)

reduce pulmonary venous return to the left heart, shortening the Q-A2 interval. Alterations in LV filling during respiration are much less quantitatively important than the RV volume changes.

Importance of Hangout Interval. Alterations in the duration of the pulmonary hangout interval recently have been demonstrated to affect the splitting of S2. In general, the pulmonary artery hangout interval is inversely proportional to the pulmonary vascular resistance. Thus, the pulmonary artery hangout interval and recoil increase as pulmonary vascular resistance de-

creases during inspiration. The highly compliant pulmonary vascular bed is affected by respiratory intrathoracic pressure changes more than the systemic vascular tree. The normal pulmonary vascular resistance is 10 to 20% that of systemic resistance; forward flow during systole into the pulmonary artery thus continues over a longer time period than flow into the aorta. The incisura of the pulmonary artery is further delayed during inspiration, and the Q-P2 interval lengthens considerably. This phenomenon apparently is responsible for most of the inspiratory widening between A2 and P2.

The factors responsible for inspiratory delay in A2-P2 are listed in Table 6-2. The shortened LV systole (decreased Q-A2) plays a minor role in inspiratory splitting of the S2; in many normal persons, especially adults, the Q-A2 remains unchanged during respiration. Note that the change in the hangout interval accounts for approximately two thirds of the increased Q-P2 interval and that increased RV systole represents the other third.

COMMON FACTORS ALTERING S2

Inspiratory Splitting. Anything that results in either a delay or shortening of right or left ventricular systole or produces an alteration in great vessel impedance and distensibility will affect the respiratory relationships of A2 and P2. Thus, disturbances of ventricular electrical activation, abnormal contractile performance, outflow obstruction or changes in vascular resistance may affect S2 splitting patterns (see section on abnormalities later in this chapter).

Intensity. The components of S2 usually have greater acoustic energy and a higher frequency than those of S1. Elevation of pressure in either great vessel produces a more rapid and forceful deceleration of blood at end-systole and will result in an S2 of increased amplitude. Valvular disease may produce the opposite effect: a soft S2.

HOW TO LISTEN TO S2

The physician must carefully evaluate the characteristics of *both* A2 and P2 during the routine cardiac examination. It is insufficient to simply assess the loudness of S2 at various precordial locations.

TABLE 6-2 *Major Components of the Physiologic Inspiratory Delay in S2 Splitting (A2 - P2 Interval)*

Mechanism	Relative Contribution (%)
Increased Q-P2	73
Increased PA hangout interval	45
Increased RV ejection time	28
Decreased Q-A2 interval	27

Since the major vibrations of S2 are relatively high pitched, the diaphragm of the stethoscope should be used to auscultate S2. Firm or increased pressure on the diaphragm is helpful to amplify high frequency sound and filter out low-pitched components. During expiration, S2 is normally heard as a single sound. On inspiration, "splitting" or separation of the two components can be heard. Most people cannot differentiate two sounds that are only 20 to 25 msec apart. Therefore, if A2-P2 are separated by 20 msec or less, they will be heard as a single sound, although a phonocardiogram may record both components. Such a sound is described as *single* or *fused*. When one sound is abnormally loud (e.g., increased P2 in pulmonary hypertension), close splitting of A2 and P2 may be more readily detected. On the other hand, a very loud A2 or P2 may mask the softer component.

A2 is louder than P2 in normal subjects and in most abnormal states, unless pulmonary hypertension is present or the sound of aortic closure is diminished. A2 is well heard at the classic aortic area (second right interspace), the pulmonary area (second left interspace), and the cardiac apex. Normally, only A2 is audible at the apex; P2 normally does not radiate to the apex as a separate, audible sound except in young or thin subjects or with pulmonary hypertension. The normal P2 is somewhat softer than A2, even at the pulmonary area, and is usually heard over a relatively small area of the chest. P2 is readily detected at the 2nd to 4th left interspaces.

S2 During Respiration. During inspiration, clear-cut separation of A2 and P2 is normally observed. This splitting is prominent in young subjects but is found less predictably in older age groups. P2 usually becomes maximally delayed in mid to late inspiration; during expiration the two components of S2 are fused or single. If a patient takes a very deep breath, P2 may be attenuated because of interposing lung tissue and only a single component will be heard. When this is suspected, the subject should be instructed to breathe less deeply. Slow regular respirations are best for auscultation. The upright position often accentuates inspiratory separation of A2 and P2, as inspiratory changes in right heart filling are greater than in the supine position.

As the two components separate, a *ta-dup* cadence is heard in inspiration. The average A2-P2 splitting interval is 30 to 40 msec (0.03 to 0.04 sec), with a range of 10 to 60 msec; the interval can occasionally be as great as 70 to 80 msec. Any interval less than 30 msec is considered "narrow splitting;" in such cases the two components of S2 may not be detected, and S2 may seem impure or "dirty" during inspiration.

Detectable expiratory splitting of 40 msec or more is usually abnormal. Whenever a subject has *audible expiratory splitting* of S2 in the supine position, he should be examined sitting or standing (see below). Often S2 will be blurred in expiration but not discretely split. Asking the subject to hold his breath in end-expiration may be useful. Younger persons (children and young adults) are more likely to have normal expiratory asynchrony in the supine position

and almost always will have prominent inspiratory splitting of S2. By contrast, normal older subjects rarely have audible expiratory splitting but often have no detectable inspiratory splitting.

Body Habitus. In obese subjects, S2 may be best heard in the first left intercostal space. In tall, thin patients S2 may be best appreciated at the lower sternal edge and not the base. In chronic lung disease, the xiphoid area is often the best site for S2 analysis.

Proper Identification of Systole. During rapid tachycardias or with severe myocardial disease, it may be difficult to distinguish S1 from S2. Findings in patients with mitral stenosis or mitral valve prolapse often are confusing. An opening snap and midsystolic click can readily be mistaken for S1 or S2, particularly with rapid heart rates (see Figures 16-6, 16-12, 18-4). *Practical Point: It is important simultaneously to palpate the carotid artery or the apex impulse when identification of S1 and S2 is difficult* (see Figure 1-4).

It is best to begin auscultation at the base; the cadence of S1 and S2 is most characteristic at the 2nd to 3rd interspace. Murmurs and other heart sounds usually are more easily sorted out away from the cardiac apex. The inspiratory motion of A2 and P2 should identify definitely which sound is S2. S1 usually is louder at the apex unless the PR interval is long or LV function is deranged.

Is Splitting of S2 Present? In patients with rapid heart rates, particularly infants, the two components of S2 may be difficult or impossible to hear. In tachypneic patients, the rapid respirations may not allow sufficient time to hear the normal variation in S2, particularly if the respirations are shallow. During sustained arrhythmias, particularly atrial fibrillation or frequent PVCs, proper evaluation of respiratory splitting can be difficult. In atrial fibrillation, S2 splitting usually is wider after the shorter cycle lengths. PVCs may induce alterations in S2 synchrony and disruption in the basic rhythm by causing additional heart sounds during the ectopic beats.

ABNORMALITIES OF SECOND HEART SOUND

Intensity

Alterations in the intensity of the loudness of A2 or P2 can be valuable in detecting semilunar valvar abnormalities or elevated pressure in one of the great vessels. A particularly loud transient sound is often palpable. For example, a palpable S2 in the 2nd to 3rd left interspace adjacent to the sternum suggests pulmonary hypertension or, less commonly, systemic hypertension.

Increased A2. A loud A2 occurs when there is increased flow and/or pressure in the central aorta. Aortic root dilatation can also increase the amplitude of A2. Thickened aortic valve leaflets that retain good mobility

may be associated with increased A2 intensity, but more often dampen the amplitude of A2. In certain congenital lesions involving the great vessels, the aortic closure sound will be increased because the aorta arises more anteriorly than usual or the pulmonary artery is displaced posteriorly. Since A2 is normally the loudest component of S2, if P2 is also increased in amplitude in such patients, the ratio of A2 to P2 may be unaltered. A palpable or tambour A2 is unequivocally abnormal.

Increased P2. The sine qua non of an increased P2 is pulmonary hypertension. *Practical Point:* *The observation of an abnormally loud P2 is one of the most important findings in the cardiac examination and almost always indicates pulmonary hypertension. Since P2 is normally inaudible at the apex, the occurrence of two audible components of S2 at the apex strongly suggests pulmonary hypertension and should prompt a careful evaluation of the patient.* In a patient with mitral valve disease or suspected pulmonary vascular or parenchymal disease, a palpable P2 at the 2nd to 3rd left interspace is a marker for elevated pulmonary pressures. On occasion, a loud P2 may "backwards mask" detection of A2. When pulmonary hypertension is suspected and there is a loud single S2 at the base, careful auscultation for both components of S2 should be performed at the apex and lower left sternal border. In most patients with pulmonary hypertension, inspiratory splitting is narrower than normal. In an atrial septal defect (ASD) with a large left-to-right shunt, the RV is enlarged; P2 often can be heard at the apex in these subjects, even in the absence of elevated PA pressure. Thus, the diagnostic reliability of an increased P2 indicating pulmonary hypertension is reduced in patients with an atrial septal defect. However, if P2 is very loud or if it increases in intensity after mild exercise, the presence of an ASD with elevated pulmonary vascular resistance and pulmonary hypertension is suggested (see Chapter 20).

Decreased A2. In valvar aortic stenosis, aortic closure typically is diminished or absent because of extensive leaflet distortion, fibrosis, or calcification. A2 may be diminished in supravalvular aortic stenosis and usually is normal in hypertrophic cardiomyopathy and subvalvular aortic stenosis. A2 may be decreased in the presence of aortic valve sclerosis with obstruction, probably because the thickened valve tissue dampens oscillations of the leaflets. In aortic regurgitation, A2 often is diminished.

Decreased P2. Valvar pulmonic stenosis is associated with a soft or even absent P2. In addition, P2 often is delayed in pulmonic stenosis. Augmentation of RV filling by squatting, mild exercise, or leg elevation may increase the intensity of a soft P2. In patients with suspected pulmonic stenosis, it is important to characterize P2 accurately to help assess severity of the lesion as well as to differentiate pulmonic stenosis from an atrial septal defect (fixed and wide splitting of S2).

In severe tetralogy of Fallot, P2 is inaudible. However, in milder cases (acyanotic) a soft, delayed P2 may be recorded and occasionally is heard, especially if a pulmonic ejection click is present.

Masking. When P2 is soft or not audible, masking should be suspected. A loud A2 may mask P2. A loud, narrowly split P2 may backwards mask A2. A loud, early opening snap may backwards mask P2. Since prominent, long systolic murmurs obscure or mask the sounds of semilunar valve closure, A2 or P2 may be difficult or impossible to hear in subjects with a ventricular septal defect, pulmonic stenosis, aortic stenosis, or mitral regurgitation. In tetralogy of Fallot or transposition of the great vessels, the loud A2 readily masks the soft P2.

Extracardiac Factors. Increased chest wall thickness, significant chest deformity, chronic obstructive lung disease, and marked obesity can decrease the intensity of all heart sounds. Inspiration often attenuates P2 by insulation of the pulmonary valve sound due to the interposed lung tissue. Marked hypotension or low cardiac output from any cause can diminish both S1 and S2 as can pericardial effusion or tamponade. A2 may be soft and P2 loud if there is a high PA pressure in the setting of a low cardiac output (e.g., massive pulmonary embolus).

Splitting (Fig. 6-7)

Expiratory Splitting. Expiratory splitting refers to the presence of two audible components of S2 at end-expiration. Held expiration may accentuate this finding and induce greater asynchrony. S2 ordinarily will be heard as a single sound when A2 and P2 are separated by 20 msec or less; thus, audible expiratory splitting indicates an interval of at least 30 to 40 msec between the two sounds. *Practical Point: RV systole shortens more than LV systole in the sitting or standing position; thus, respiratory variation of S2 is enhanced when a subject is upright. In the majority of normal persons who demonstrate expiratory splitting of S2 in the supine position, S2 becomes single when the patient sits up (Fig. 6-8).*

Persistence of audible expiratory splitting in the upright position is a valuable clue to an underlying cardiovascular abnormality. Table 6-3 lists conditions that may cause asynchronous right and left ventricular systole and expiratory splitting. Most conditions associated with audible expiratory splitting will demonstrate a further increase in the A2-P2 interval with inspiration; if there is no detectable change in the S2 interval, the split is considered *fixed*.

Delayed P2 (Increased Q-P2). Right bundle branch block is the most common cause of expiratory splitting of S2. S1 also may be prominently split in RBBB; in this setting, P2 widens further with inspiration. Other "electrical" causes for prolonged RV systole and delayed P2 include PVCs of left ven-

tricular origin, LV epicardial pacing, and occasionally, certain patients with the Wolff-Parkinson-White syndrome.

Moderate to severe pulmonic stenosis produces delayed splitting of P₂ on expiration; although respiratory motion remains normal, P₂ is often late and may be barely detectable. Audible expiratory splitting often is the first clue to the fixed S₂ of an atrial septal defect. Mild pulmonic stenosis and an ASD are common causes of systolic murmurs in children. The presence of

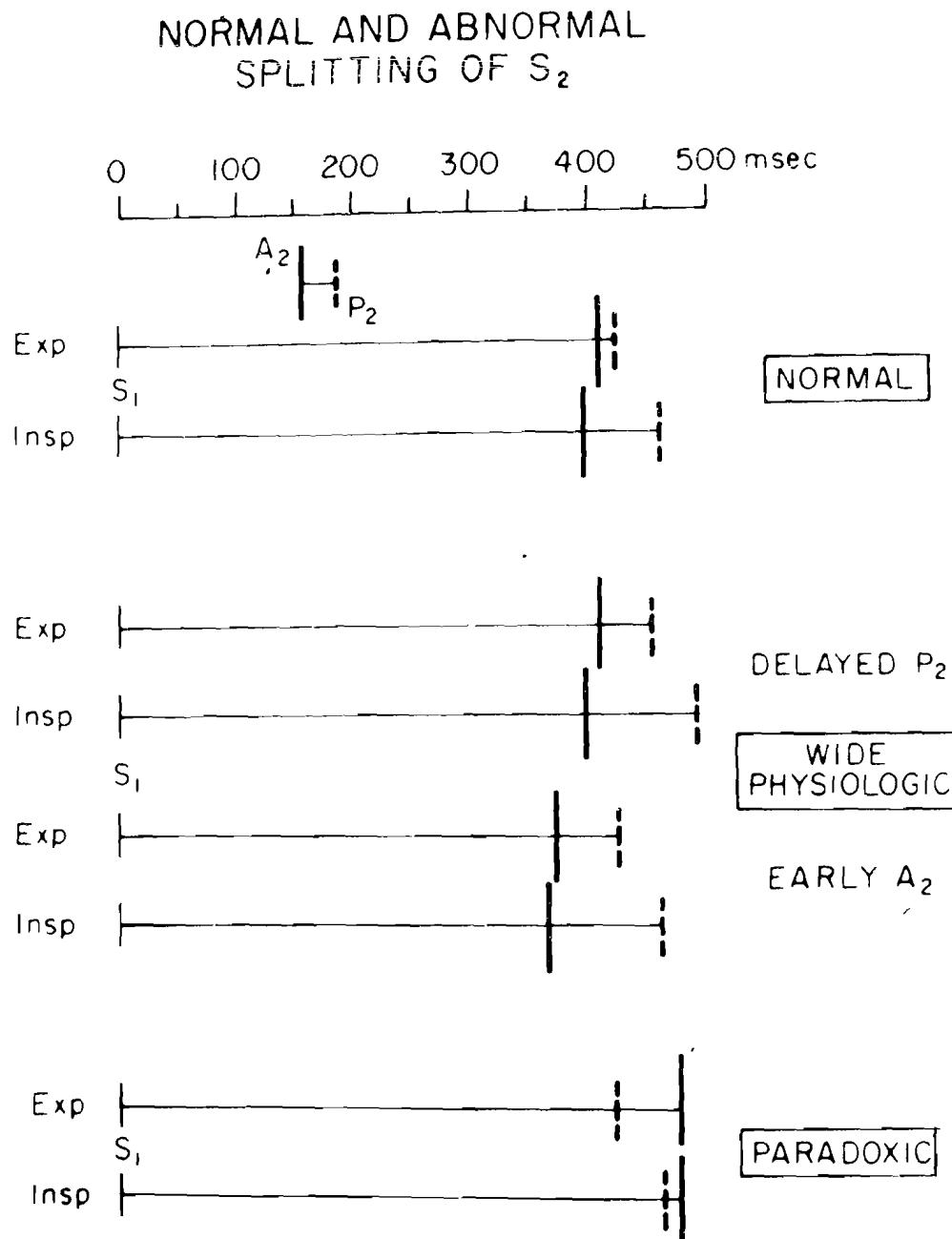


FIG. 6-7. Splitting patterns of S₂. EXP = expiration. INSP = inspiration. (From Curtiss EI, Reddy P: First and second heart sounds. In Signs and Symptoms of Cardiology. Edited by LD Horwitz, BM Groves, Philadelphia, JB Lippincott Co, 1985.)

Use of Sitting Position to Assess Audible Expiratory Splitting

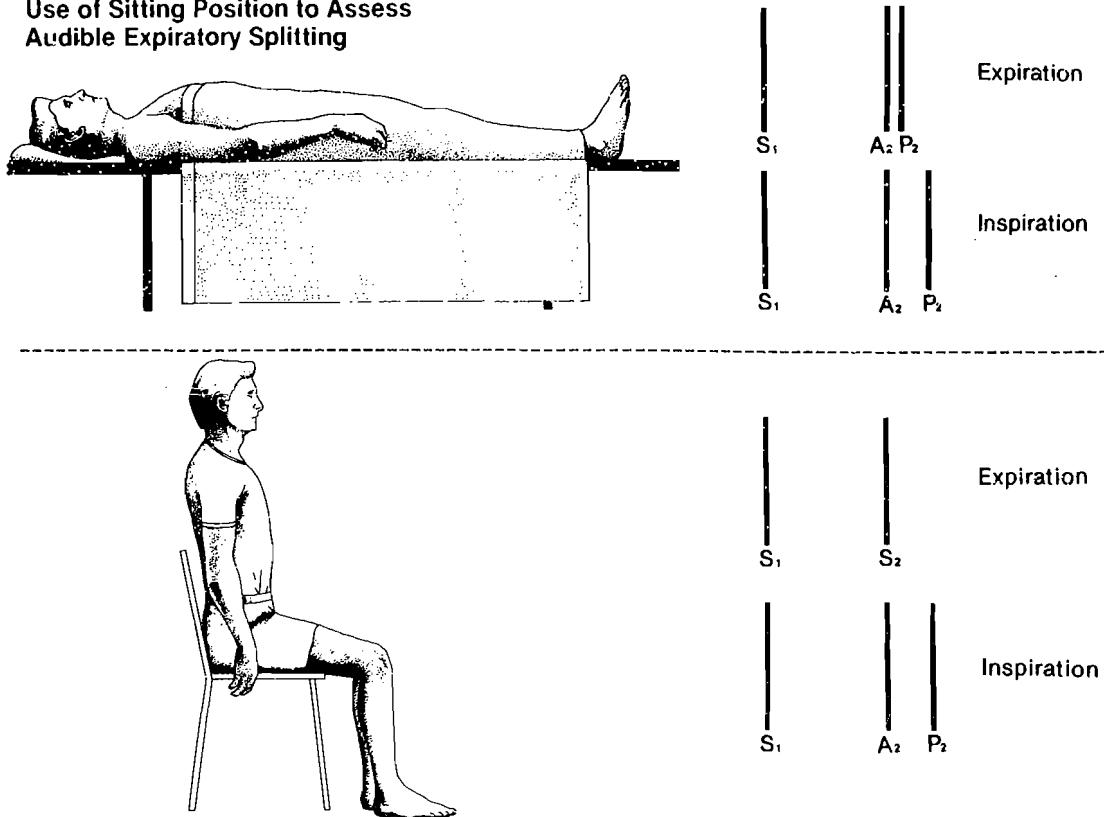


FIG. 6-8. Evaluation of audible expiratory splitting S2. The presence of expiratory splitting in the supine position is usually abnormal. Sometimes expiratory splitting of S2 in the supine position will disappear when the patient is upright and the S2 becomes single on expiration. This is a normal response. Subjects should be carefully examined in the sitting and standing positions whenever S2 appears to be abnormally split during expiration. (From Abrams J: Prim Cardiol, 1982.)

TABLE 6-3 Common Causes of Audible Expiratory Splitting of S2

Increased Q-P2

- Pulmonic stenosis
- Massive pulmonary embolus
- Pulmonary hypertension with RV failure
- Atrial septal defect
- Complete RBBB
- PVCs of LV origin
- Thoracic skeletal abnormalities
- Occasional normals

Decreased Q-A2

- Severe mitral regurgitation
- Ventricular septal defect

P₂-A₂ Reversal

- All causes of paradoxic or reversed splitting (see text)

expiratory splitting can be of diagnostic importance in the younger age groups and suggests the presence of an organic cardiac defect. Audible expiratory splitting in patients with an ASD often persists after surgical repair, presumably because of persistently abnormal pulmonary artery compliance and a persistently prolonged hangout interval.

Pulmonary hypertension is usually associated with a fixed Q-A2 and shortened Q-P2 (decreased hangout interval), resulting in narrow expiratory splitting. RV ejection is not prolonged unless associated RV failure occurs, and then wide splitting may occur. When RV failure or dysfunction accompanies pulmonary hypertension, S2 may be prominently split on expiration because of prolonged RV emptying.

Idiopathic dilatation of the pulmonary artery may produce prominent splitting of S2 on expiration and inspiration, presumably because of increased compliance in the dilated pulmonary artery that is composed of deficient elastic tissue. Congenital or pulmonic regurgitation may be associated with a prolonged RV systole and audible expiratory splitting. Occasionally, severe biventricular congestive heart failure may result in expiratory splitting of S2, which disappears with cardiac compensation.

Decreased Q-A2 Interval. Occasional cases of audible expiratory splitting may be explained by shortened LV systole. This may occur in severe mitral regurgitation, ventricular septal defects, or pericardial tamponade. Some have suggested that wide splitting of S2 in mitral regurgitation is found only with nonrheumatic etiologies. In ventricular septal defects, the Q-A2 interval may be short, and the increased volume of RV blood may prolong RV systole (increased Q-P2). In pericardial tamponade, the reduction in LV stroke volume may be greater than that of the RV, and expiratory splitting can be heard. The rare patient with left atrial myxoma may have a shortened LV systole and audible expiratory splitting. Some patients with severe pulmonary hypertension accompanying pulmonary embolism may have a shortened Q-A2, a normal or increased Q-P2, and prominent expiratory splitting.

Other Causes of Expiratory Splitting. Benign causes of expiratory splitting include a minor interventricular conduction delay, pectus excavatum, straight back syndrome, and other musculoskeletal abnormalities. Occasionally, normal children without any evidence of heart disease will display this finding.

Differential Diagnosis of Audible Expiratory Splitting. Any complex of sounds consisting of two components in late systole-early diastole may simulate a split S2 in expiration (Fig. 20-5). Such combinations include an A2-opening snap (Fig. 16-6), late systolic click-S2 (Fig. 18-4), S2-pericardial knock, and S2-S3 (particularly when the S3 has high frequency vibrations).

Wide Inspiratory Splitting. Most of the conditions that produce audible expiratory splitting may also result in a prominent *inspiratory* delay between A2 and P2. Occasionally, respiratory motion of A2-P2 is absent when S2 is

widely split; this is known as *fixed splitting* and will be discussed in the next section.

Increased Q-P2. When RV systole is prolonged by either electrical (RBBB, LV pacing) or hemodynamic factors, a prominent delay in P2 is common. Right ventricular failure from any cause will delay the Q-P2; in acute and chronic pulmonary embolization this may be dramatic, even in the absence of RBBB. A similar situation has been described in late pregnancy or advanced renal failure; the delayed Q-P2 presumably reflects hypervolemia.

In valvular pulmonic stenosis P2 is late, and the degree of A2-P2 delay correlates well with the severity of obstruction. Thus, an A2-P2 interval of 100 msec signifies severe stenosis with an RV-PA gradient greater than 100 mmHg. Because of the markedly reduced pulmonary artery diastolic pressure, the pulmonary closure sound is of low amplitude and may appear absent. When infundibular pulmonic stenosis is present, the correlation between the severity of obstruction and the length of the A2-P2 interval is no longer valid. The RV outflow tract contracts later than the RV inflow region, and in marked infundibular hypertrophy, a delay in RV outflow contraction may lengthen RV systole out of proportion to the actual measured gradient. Thus, the A2-P2 interval in patients with a combination of valve and infundibular pulmonic stenosis is not useful in assessing the degree of obstruction.

Patients with ventricular septal defects (VSD) who undergo pulmonary artery banding may exhibit a delayed P2. In atrial septal defects, the large RV stroke volume helps to increase the Q-P2 interval; an RV conduction defect usually contributes to the delay. The S2 in patients with an ASD is usually fixed (Fig. 20-4), although respiratory variation may occasionally occur, particularly in the upright position.

In patients with pulmonic stenosis, ASD, or idiopathic dilatation of the pulmonary artery, the Q-P2 interval is increased due to a delayed hangout interval. In these conditions, the dilated proximal pulmonary arteries contribute to an increased distensibility of the pulmonary vascular bed. This phenomenon probably accounts for the occasional prominently delayed P2 in mild valvular pulmonic stenosis with poststenotic dilatation of the pulmonary artery, and the observation that successful repair of an uncomplicated ASD does not always result in normalized splitting of S2.

Decreased Q-A2 (Table 6-4). A large VSD with hyperkinetic pulmonary circulation and normal pulmonary vascular resistance may demonstrate wide

TABLE 6-4 Wide Splitting of S2 Resulting from a Decreased Q-A2 Interval

-
- Mitral regurgitation (usually acute or severe)
 - Ventricular septal defect
 - Pericardial tamponade
 - Left atrial myxoma
 - Constrictive pericarditis
-

inspiratory splitting due to the combination of decreased Q-A2 and increased Q-P2 intervals. The long systolic murmur of the VSD may obscure A2. Thus, for the proper evaluation of S2, it is important to listen for A2 away from peak murmur intensity, e.g., at the apex and lower left sternal border. Patients with pericardial tamponade or left atrial myxoma may have wide inspiratory splitting because of shortened LV systole. In constrictive pericarditis, the RV stroke volume is relatively fixed and does not respond to respiratory alterations in venous return. However, LV filling may diminish markedly during inspiration with a resultant decreased Q-A2 interval and prominent wide inspiratory splitting. Shortened LV systole in severe mitral regurgitation is associated with an increased A2-P2 interval particularly if there is associated RV failure. If LV failure occurs, LV systole is prolonged and wide splitting disappears.

Fixed Splitting. When right or left ventricular stroke volume does not change during inspiration, or when there is a similar degree of respiratory alteration in both RV and LV filling, the splitting of S2 is fixed. Fixed splitting may occur with a relatively narrow A2-P2 interval, but is more common (and more noticeable) when the A2-P2 interval is wide. Apparent fixed splitting occasionally may be present in young normal patients, but usually disappears when the individual assumes the upright position (Fig. 6-8). *Practical Point:* *The diagnosis of fixed splitting should be made only after careful auscultation of S2 in both the recumbent and upright positions.* A minor delay of P2 on inspiration in a subject with expiratory splitting may not be readily appreciated. It is easier to detect inspiratory widening when A2 and P2 are fused or narrowly split in expiration. Although phonocardiograms will often document some respiratory variation of A2 and P2, changes in splitting intervals of 20 msec or less will not be detectable by most observers, and S2 will be considered to be fixed.

Failure to Increase Right Ventricular Stroke Volume with Inspiration. Inspiratory augmentation of RV filling may not be possible when there is a major pressure overload of the RV. This is seen in RV failure, acute and chronic pulmonary embolization, and severe pulmonary hypertension (uncommon). In such cases, left ventricular filling may also be subnormal, with a resultant decreased Q-A2 interval and prominent wide splitting. In all these conditions, clinical improvement may result in return of the normal respiratory excursion of P2.

When RV systole is substantially prolonged (e.g., RBBB with congestive heart failure), A2-P2 coupling will be both wide and fixed. Maneuvers that decrease venous return, such as tourniquets or assumption of the upright position (Fig. 6-8), may bring out respiratory motion; mild exercise may also produce respiratory variation of S2.

Simultaneous Increase in Right Ventricular and Left Ventricular Filling ATRIAL SEPTAL DEFECT (see Chapter 20). Fixed splitting is the

hallmark of ASD (see Figure 20-4). This diagnosis often is first suspected because of the presence of prominent expiratory separation of A2 and P2. The right and left atria act as a common venous reservoir; anything that influences RV or LV filling can alter the degree of left-to-right shunting across the atrial defect. During inspiration, the augmented venous return to the volume-overloaded RV causes a decrease in left-to-right shunting at atrial level and a resultant increase in LV filling. Some right atrial blood may transiently shunt right-to-left during inspiration. The net effect is to "balance out" the respective increases in right and left ventricular filling during inspiration: both Q-P2 and Q-A2 increase simultaneously without a change in the A2-P2 splitting interval. Typically, S2 is widely split; slight respiratory variation in A2-P2, when present, may not be appreciated by the ear. Respiratory motion may be detected, however, in the upright position, and in a small percentage of otherwise typical secundum atrial defects, respiratory splitting is readily detectable. In the pulmonary hypertensive ASD, fixed splitting of S2 is maintained, although P2 is markedly increased in intensity.

In patients suspected of having an ASD, the Valsalva maneuver can be used to assess S2 mobility. In normal subjects, S2 fuses during the strain phase as both right and left ventricular filling decreases; upon release of the Valsalva, the immediate surge in RV filling produces transient asynchrony of stroke volume, and prominent S2 splitting occurs for 6 to 8 beats. When the augmented blood volume reaches the left heart, splitting of A2 and P2 narrows as LV systole lengthens. In patients with an ASD, S2 splitting widens during the strain phase (due to increased left-to-right shunting). Following release of the strain, the expected increase in A2-P2 splitting fails to develop due to altered interatrial shunting.

In patients with an ASD and atrial fibrillation, the pattern of splitting after long cycle lengths may be diagnostically helpful. In contrast to subjects with atrial fibrillation without an intracardiac shunt, long cycle lengths in patients with an ASD result in increased A2-P2 separation due to greater left-to-right shunting during long diastoles.

VENTRICULAR SEPTAL DEFECT (see Chapter 21). S2 is normal in the VSD without pulmonary hypertension. In a VSD with markedly elevated pulmonary vascular resistance (Eisenmenger reaction) the ventricles act as a common chamber. Ejection of blood from both ventricles into the great vessels with an equivalent resistance results in a comparable inspiratory "delay" in each chamber, with absence of respiratory motion of S2. In most instances, S2 is fused or single on both inspiration and expiration. This finding is a valuable diagnostic clue, as it strongly suggests that the defect is inoperable. In some VSDs with a hyperkinetic pulmonary hypertension and normal vascular pulmonary resistance, splitting of S2 also may be fixed.

Differential Diagnosis of Wide Fixed Splitting. Occasionally, other conditions can simulate wide fixed splitting of S2. In patients with LBBB and

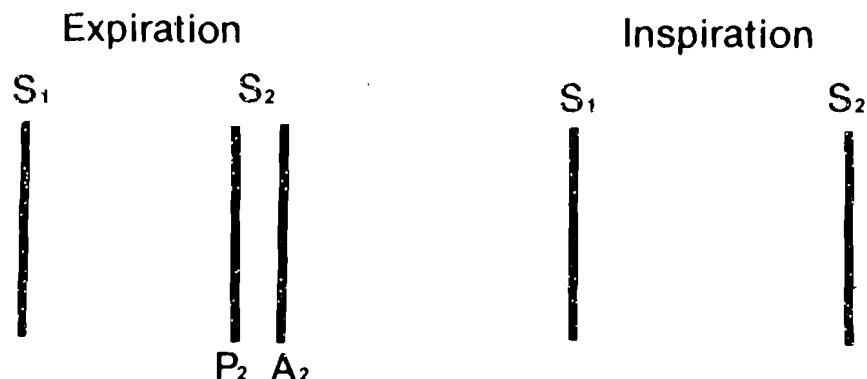
paradoxical splitting (see below) the presence of congestive heart failure may result in fixed but reversed splitting of S2 (P2-A2). An A2-opening snap complex in mitral stenosis also can be confused with a widely split S2, particularly when there is a close A2-OS interval (see Figure 16-6).

Reversed or Paradoxic Splitting. Reversed splitting occurs when S2 is maximally split on expiration and narrows or fuses on inspiration (Fig. 6-9). Because the directional changes of A2 and P2 during the respiratory cycle are reversed from normal, the term *paradoxic splitting* has been widely used for this phenomenon. In general, this type of splitting is found only when LV electromechanical systole is significantly delayed, i.e., increased Q-A2 interval. During expiration, prolonged LV systole causes A2 to follow P2 (audible expiratory splitting). With inspiration, the Q-P2 increases normally, but Q-A2 is unchanged or shortens. The two components of S2 coincide or occur so close together that the ear appreciates only a single second sound in inspiration. This is classic or Type I paradoxic splitting (Fig. 6-9). In lesser degrees of Q-A2 delay, or with a wide inspiratory lengthening of the Q-P2 interval, inspiration still may result in a normal A2-P2 relationship and audible inspiratory splitting, although S2 reversal occurs in expiration (Type II). If both expiratory and inspiratory separation of A2 and P2 are 20 msec or less apart, S2 will be heard as single or fused in both phases of respiration, even though A2 follows P2 in expiration (Type III paradox). Only Type I paradox can be detected by the physician as a definite abnormality of S2 motion; Type II and III can be diagnosed only with meticulous phonocardiographic techniques.

Causes of Increased Q-A2 Interval LEFT BUNDLE BRANCH BLOCK. The most common cause of S2 reversal is left bundle block (LBBB) (Fig. 6-9). Paradoxic splitting is detectable in most patients with LBBB. The prolonged systole (increased Q-A2 interval) has two causes: (1) an increased interval from the beginning of electrical activation (onset of the QRS on EKG) and the beginning of LV isovolumic contraction due to delayed and disordered LV depolarization; and (2) prolongation of left ventricular isovolumic contraction time. This is present in many patients with LBBB as a result of impaired LV contractility. The latter is related only indirectly to the electrical conduction disturbance and probably is a result of whatever underlying disease process that produced the LBBB.

S2 can be affected by other disorders of LV activation such as lesser degrees of LBBB (common in left ventricular hypertrophy) and, in some cases, type B Wolff-Parkinson-White syndrome (right ventricular pre-excitation from a right-sided bypass tract). In the latter case, Q-P2 actually may shorten with inspiration. These conduction defects commonly result in Type II paradox. Right ventricular pacing and PVCs of right ventricular origin will produce audible paradoxic splitting of S2.

INCREASED LEFT VENTRICULAR EJECTION TIME. Although relatively uncommon, a large stroke volume in LV volume overload conditions may



Paradoxic Splitting of S_2 in LBBB

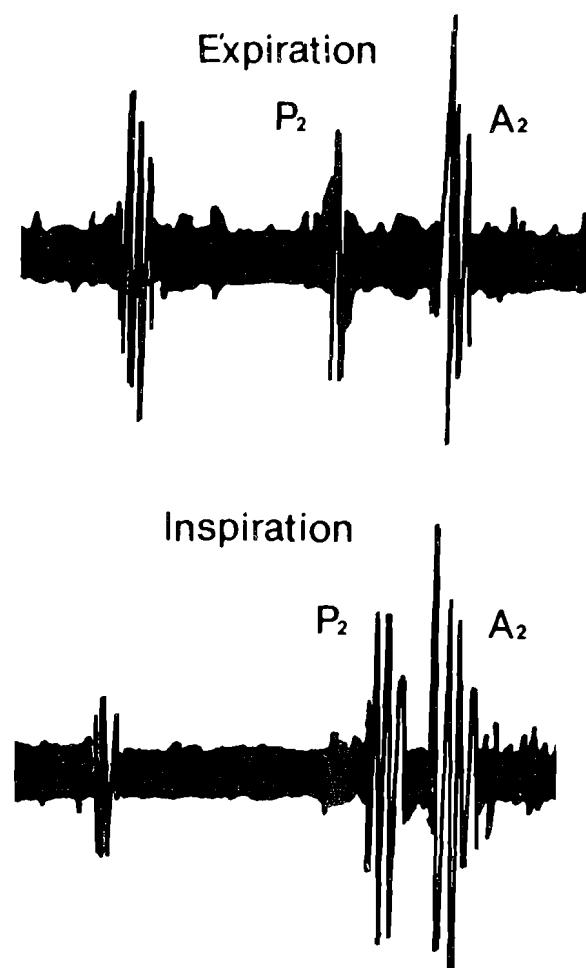


FIG. 6-9. Reversed or paradoxical splitting of the second heart sound. The usual cause of paradoxical splitting is abnormally delayed left ventricular ejection, such that aortic closure (A_2) follows pulmonic closure (P_2). This commonly results in audible expiratory splitting. During inspiration the normal delay the Q-P2 interval results in P2 "moving into" A2, and a single or narrower S2 occurs. Thus, the normal respiratory pattern is reversed. (From Abrams J: Prim Cardiol, 1982.)

result in a prolonged Q-A2 and reversed splitting. This has been demonstrated in some patients with a patent ductus arteriosus and a large left-to-right shunt and also in severe aortic regurgitation. In coronary artery disease or hypertension with acute or chronic LV dysfunction, reversed splitting of S2 occasionally is noted, although it is a rather uncommon finding.

LEFT VENTRICULAR OUTFLOW TRACT OBSTRUCTION. Reversed splitting may be observed in valvular aortic stenosis and hypertrophic cardiomyopathy. Splitting of S2 in hypertrophic cardiomyopathy may vary from normal to paradoxical in the same patient from day to day. In these conditions, the reversed splitting of S2 is not related to prolonged total LV ejection time, which typically is normal in aortic stenosis and abbreviated in hypertrophic cardiomyopathy. Instead, it is probably due to late aortic valve closure resulting from the delayed LV-aortic pressure crossover in patients with large gradients and slowed LV relaxation.

Poststenotic dilatation of the aorta in valvular aortic stenosis may contribute independently to the prolonged Q-A2 interval by delaying the aortic hangout interval as a result of increased compliance and distensibility of the dilated central aorta. This mechanism also may be a factor underlying paradoxical splitting in patent ductus arteriosus and aortic regurgitation with aortic root dilatation.

DECREASED Q-P2. Rarely, reversal of S2 has been observed as a result of shortened RV systole due to decreased filling of the RV such as in right atrial myxoma or tricuspid regurgitation.

Clue to Auscultation of Reversed Splitting. The physician should listen carefully over *many* respiratory cycles in order to be certain S2 moves paradoxically. In instances where the respiratory splitting pattern remains unclear, Constant suggests two approaches. First, it is important to carefully assess the amplitude of both components of S2 as the stethoscope is moved from base to apex. The component that softens as the stethoscope approaches the apex will be P2; this should clarify whether the S2 sequence is A2-P2 or P2-A2. Second, in patients with reversed splitting the Valsalva maneuver characteristically results in widening of S2 during the strain phase, narrowing of S2 upon release, and subsequent widening again. In contrast, a normal S2 narrows during the strain phase, widens upon release, and then narrows again (Fig. 6-10).

Pseudoparadoxical Splitting. Deep inspiration, particularly in subjects with large chests or chronic obstructive lung disease, may cause an artifactual muffling or disappearance of P2 due to interposition of the expanded lung between the stethoscope and the aorta. If S2 is audibly split in expiration, this phenomenon may result in the false diagnosis of reversed or paradoxical splitting as S2 appears to become "single" in inspiration.

Single (or Narrow) Splitting of S2. A single S2 will be heard on inspiration when either A2 or P2 is inaudible or when the inspiratory separation

USE OF VALSALVA MANEUVER TO DEFINE A₂-P₂
RELATIONSHIP IN SUSPECTED PARADOXIC SPLITTING OF S₂

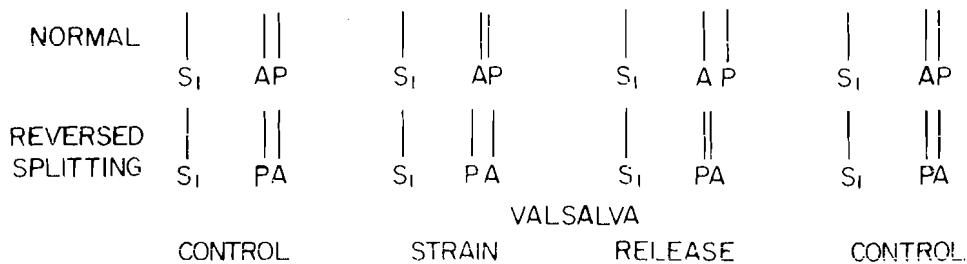


FIG. 6-10. The role of the Valsalva maneuver in the evaluation of possible reversed or paradoxical splitting of S₂. Normal splitting of S₂ narrows during the strain phase and widens markedly immediately during the release phase. Paradoxical splitting of S₂ widens during the strain phase and then becomes more narrow during the release phase.

of A₂ and P₂ is so narrow (less than 30 msec) that the ear cannot distinguish both components. The patient should be examined in both reclining and sitting positions to confirm the presence of a single S₂. Care must be taken to avoid irregular, rapid, and shallow respirations; ask the patient to breathe slowly and deeply. The following are common causes of a single S₂:

Aging. The incidence of audible inspiratory splitting of S₂ in normal subjects decreases with age. In up to 50% of subjects over 60 years of age, S₂ is single. This may be due either to an age-related lengthening of LV isovolumic contraction resulting in an increase in the Q-A₂ interval or to a decrease of the Q-P₂ interval accompanying a physiologic decrease in pulmonary vascular compliance and a shortening of the hangout interval in the elderly (Fig. 6-11).

Artifactual Muffling of P₂. This results from lung insulation of P₂ during inspiration and is more common in older people. It is more likely to occur in subjects with an increased thoracic A-P diameter and/or chronic obstructive lung disease. Patients who are obese or have a thick chest wall also may have poor transmission of P₂.

Reversed Splitting. In the clinical conditions reviewed in the previous discussion of reversed splitting, an equal duration of systole in both ventricles may cause S₂ to be heard as single in both phases of respiration. Reversed or paradoxical splitting of S₂ (Type I) should be readily distinguishable from a single S₂ by detection of expiratory separation of A₂ and P₂ in expiration.

Pulmonary Hypertension. In subjects with marked elevation of PA pressure and good RV function, a narrow A₂-P₂ interval is the rule, and S₂ may appear to be single in inspiration (Fig. 8-6). The loud P₂ may backwards mask A₂. When this is suspected, listen carefully at the apex and lower sternal area for both components of S₂.

Masking. P₂ can be masked in the presence of a high intensity and closely split sound of aortic closure or a loud mitral opening snap.

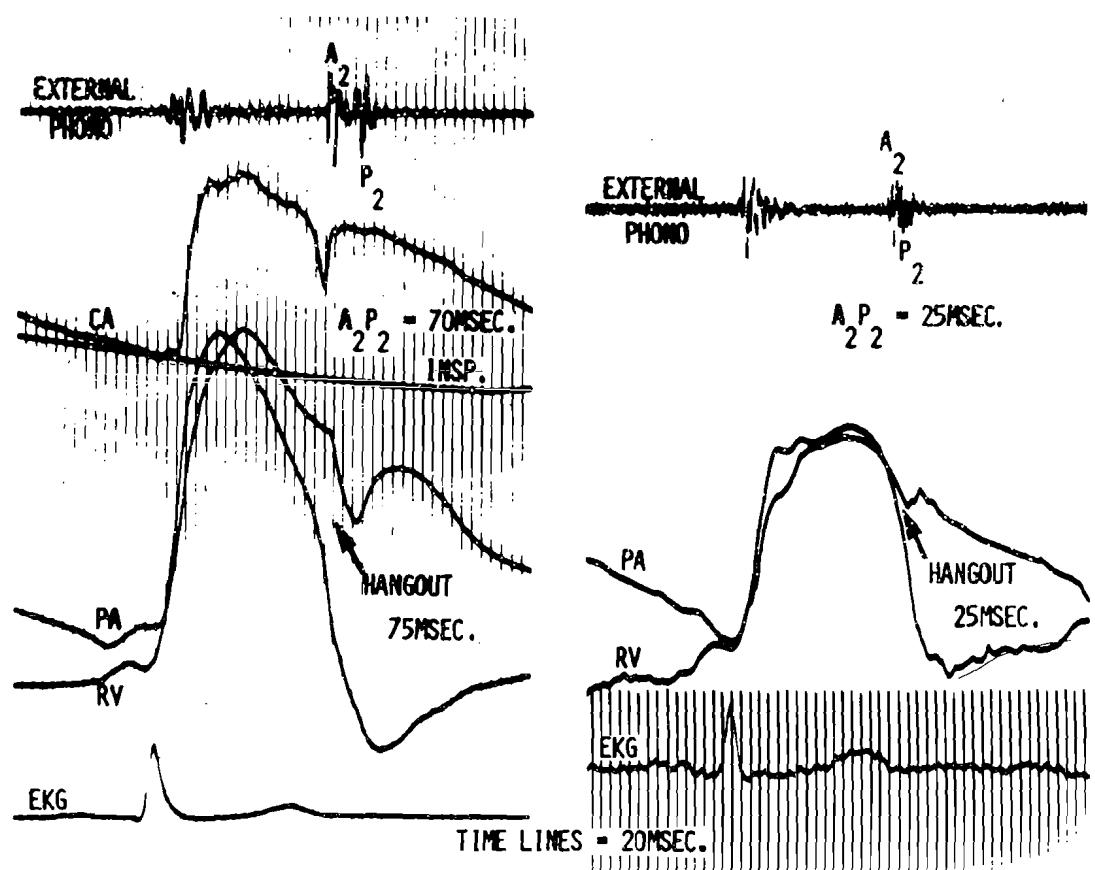


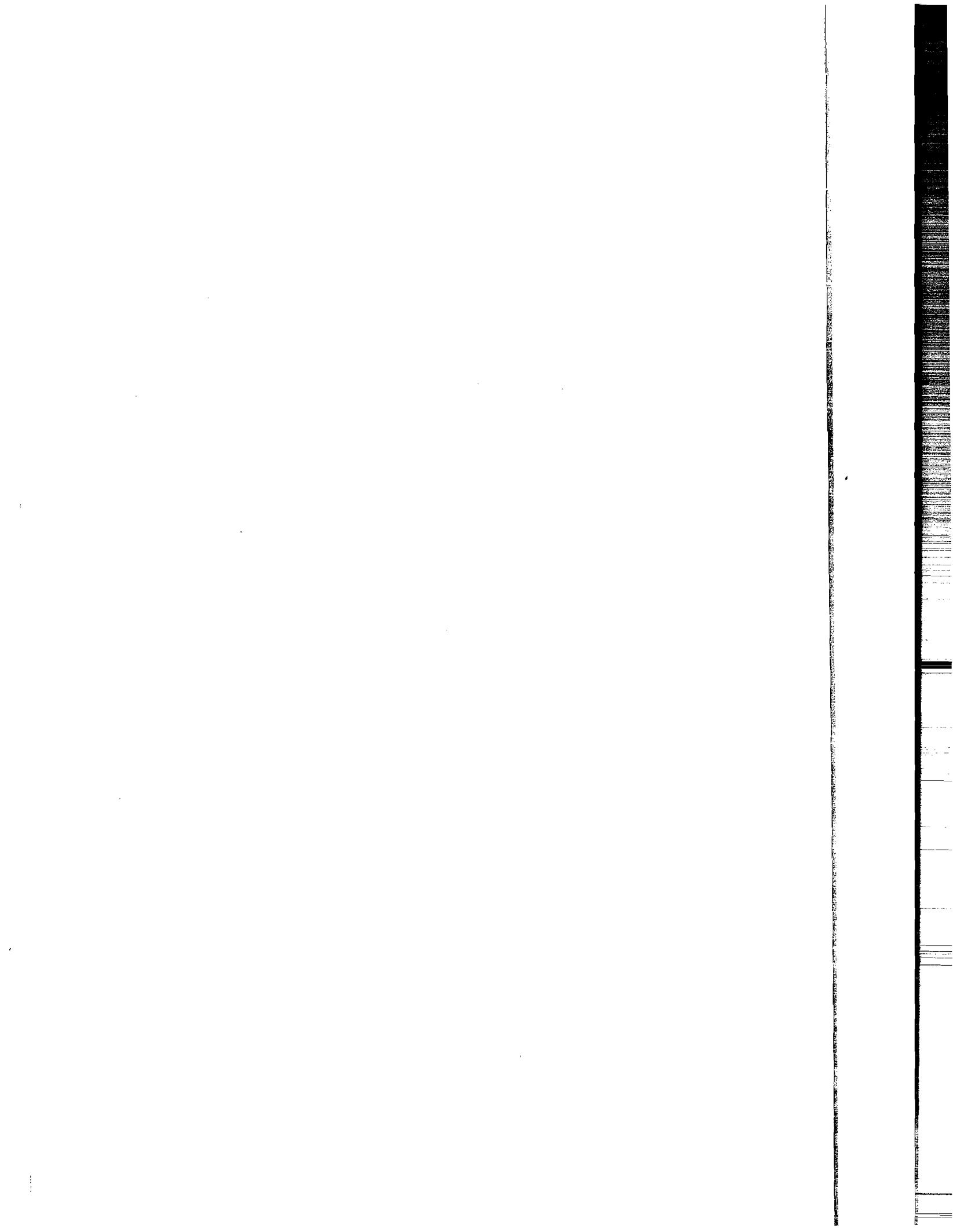
FIG. 6-11. Alterations of the right-sided hangout interval with aging. The left panel represents an inspiratory cycle from a 20-year-old normal individual. It demonstrates a wide hangout interval with prominent splitting of A₂ and P₂. The right panel shows the intracardiac pressures from a 61-year-old normal male who had narrow inspiratory splitting of S₂ and a narrow right-sided hangout interval. It is not abnormal to hear a single or fused S₂ inspiration in older individuals, possibly because of the physiologic decrease in the hangout interval because of age. (From Curtis SE et al: Newer concepts in physiologic splitting of the second heart sound. In *Physiologic Principles of Heart Sounds and Murmurs*. Edited by DF Leon and JA Shaver. American Heart Association Monograph No. 46, page 68, 1975.)

Murmur Obscuration. Long or holosystolic murmurs such as those of mitral regurgitation, ventricular septal defect, aortic stenosis, pulmonic stenosis, and patent ductus arteriosus can engulf or mask either A₂ or P₂. In these situations, careful listening at the apex and lower left sternal border for A₂ and P₂ is necessary. It is often useful to listen to the behavior of the S₂ splitting away from the site of maximal murmur intensity.

Pathologic Absence of P₂. Severe pulmonic valvular deformity with low or absent pulmonary blood flow and great vessel malposition accounts for the single A₂ heard in severe tetralogy of Fallot, pulmonary atresia, tricuspid atresia, truncus arteriosus, pseudotruncus, and transposition of the great vessels. P₂ can be inaudible in severe pulmonic stenosis. In these conditions a phonocardiogram will often record an inaudible P₂.

Semilunar Valvular Stenosis. In severe aortic valvular stenosis, a loud murmur can conceal or mask A2 (Fig. 13-6); if the valve is seriously deformed and calcific, A2 may be truly soft or absent (Fig. 13-8B). Delay in the LV-aortic pressure crossing may lengthen the Q-A2 interval to such a degree that the A2 and P2 are heard as a single sound. The murmur of severe pulmonic stenosis may engulf A2; P2 typically is late and soft and may be easily missed.

Eisenmenger VSD, Single Ventricle. When a "common" ventricular chamber ejects blood into two vascular beds with equivalent and high resistance, right and left ventricular systoles are identical in length, and S2 will be fused (single) in all phases of respiration (Fig. 8-6).



Chapter 7

The Third and Fourth Heart Sounds

The presence on auscultation of a third (S3) or fourth (S4) heart sound may be extremely important in the evaluation of adults with suspected or known cardiovascular disease. These low frequency, low amplitude diastolic sounds are heard at the cardiac apex. An S3 may be a normal or abnormal finding; the presence of an S3 in an adult over 40 may have serious implications about the status of left ventricular function, but in a young subject this finding is usually normal. An audible S4 is generally present only in abnormal hearts.

Both the S3 and S4 are discussed with respect to the underlying physiology and anatomy of the heart. The final section reviews the acoustic properties of these sounds and emphasizes the optimal technique for detection of the S3 and S4 during the cardiac examination.

THIRD HEART SOUND

PHYSIOLOGY OF S3

The S3 follows mitral valve opening and the onset of rapid ventricular filling (Figs. 7-1, 7-2A) as the mitral valve leaflets are moving toward the left atrium (E-F slope). It occurs near the end of the rapid filling wave. The initial vibrations of S3 generally coincide with the maximal excursion or "checking" of the LV septum and posterior free wall. This timing suggests that the S3 is related to forces active during early diastolic filling. Considerable data indicate that rapid deceleration of blood during inflow to the left ventricle is translated into vibratory energy, resulting in the S3. The relaxing ventricle and increasing diastolic blood volume set the ventricular walls, mitral valve apparatus, and blood mass into vibration. This theory is consistent with the common finding of an S3 in conditions of increased transmural blood volume and/or flow velocity, as well as those associated with an increased end-diastolic volume and viscous resistance. Recent studies suggest that the nature of "coupling" of the left ventricle with the inner chest wall is directly related to the presence or absence of an audible S3.

The S3 occurs as active ventricular relaxation ends and passive distension begins. Left ventricular relaxation often is abnormal in ischemic heart disease, as well as in other conditions affecting the myocardium. It is likely that abnormalities of LV relaxation, as yet poorly understood and difficult to evaluate at the bedside, may affect the occurrence, timing, and intensity of

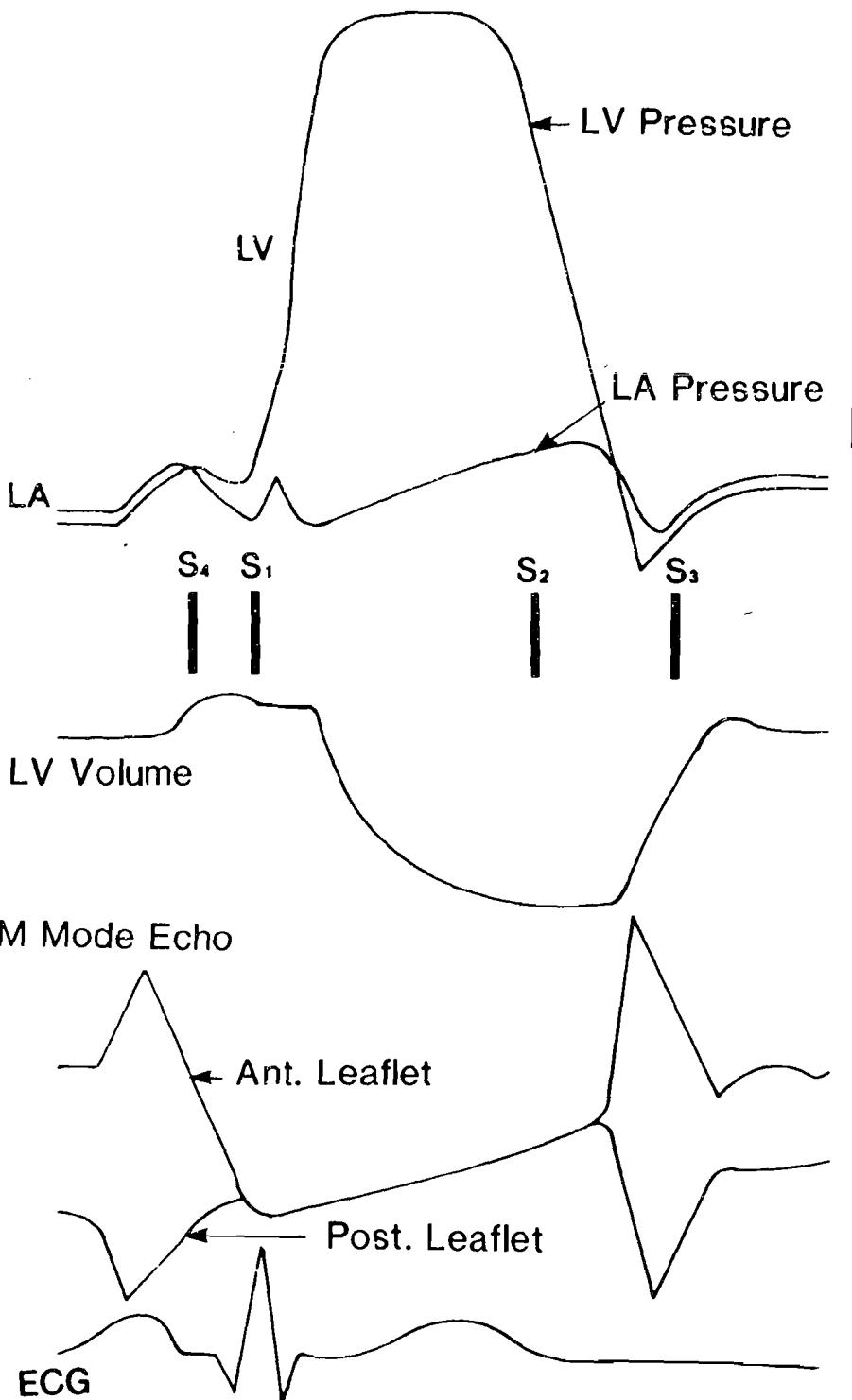


FIG. 7-1. Intracardiac pressure, mitral valve motion and left ventricular volume interrelationships in heart sound generation. Left ventricular and left atrial pressures are displayed with the simultaneous M-mode echo and left ventricular volume in relation to the four heart sounds. Note the timing of the heart sounds with respect to left atrial-left ventricular pressure crossover, mitral leaflet motion, and alterations in left ventricular volume. (From Abrams J: Prim Cardiol, 1982.)

the S3. Other factors, including the velocity of diastolic filling and ventricular compliance, play a role in the genesis of the third heart sound. The intensity of S3 is increased when early diastolic filling is rapid due to elevated left atrial pressure and/or blood volume, or when left ventricular distensibility in early diastole is increased. The presence of the S3 correlates with an increased amplitude and velocity of the rapid filling wave. When LV compliance is decreased, the rate of early diastolic filling is decreased; in these situations, S3 may not be present but an S4 may be audible (see page 118).

The physiologic explanation for the presence of an S3 in normal children and young adults but not in older subjects is unclear. Presumably, young subjects have more prominent, rapid diastolic filling as a result of higher cardiac output. Other related factors may include altered sound transmission characteristic in adults, a decrease in LV-chest wall coupling in older individuals, and physiologic changes in LV compliance that occur with aging. There is little objective evidence, however, to support any of these postulates.

An S3 may also be produced in the right ventricle when there is RV dysfunction or excessive flow across the tricuspid valve. Whether the right ventricle contributes to the normal or physiologic S3 is unknown.

Terminology. The S3 sometimes is called a ventricular gallop, early diastolic gallop or sound, S3 gallop, or protodiastolic gallop. Although the term *S3* or *third heart sound* is preferable, ventricular gallop is acceptable when there is definite underlying heart disease. The term *gallop* does not necessarily imply a rapid heart rate and is often used to signify an abnormal S3 or S4 heart sound at normal or even slow heart rates.

CLINICAL SIGNIFICANCE OF THE THIRD HEART SOUND

An S3 may be present in: (1) normal hearts; (2) diastolic overload states; and (3) patients who have LV dysfunction, with or without overt congestive heart failure (Table 7-1).

Normal Hearts

An S3 commonly is found in normal children and young adults (Fig. 7-2A, B). Although not usually present in healthy subjects over 40 years of age, an S3 occasionally may be found in individuals in their 30s (especially in women) without evidence of cardiac disease. In states such as anemia, thyrotoxicosis, anxiety, exercise, and pregnancy, a physiologic S3 may become more prominent or appear for the first time (Fig. 7-2C).

Normal Heart with Increased Cardiac Output. In the normal heart with an increased cardiac output and augmented velocity of diastolic filling, an audible S3 may be present, which is acoustically indistinguishable from a

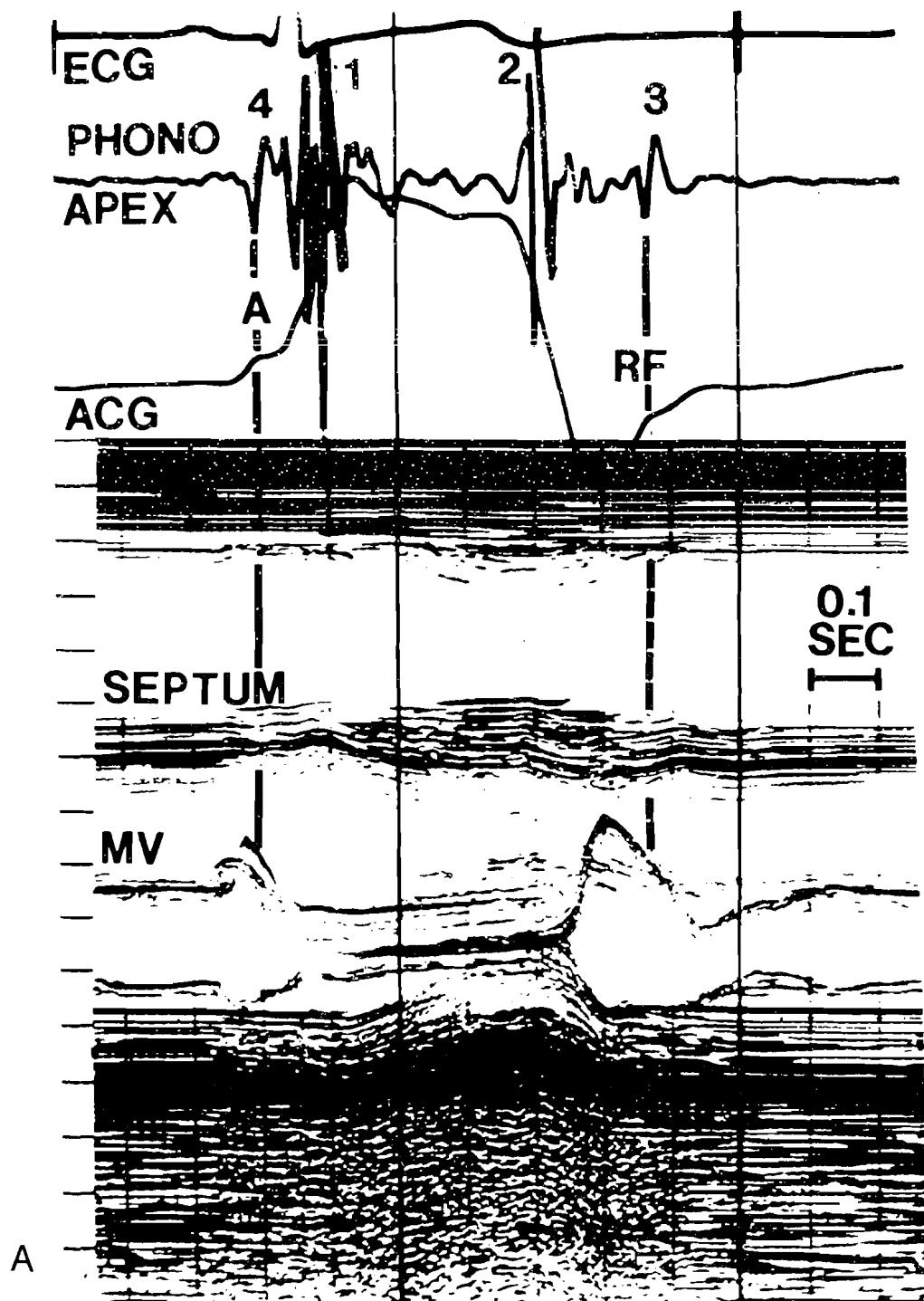


FIG. 7-2. A. The normal third and fourth heart sounds and their relationship to intracardiac events. This phono- and apex cardiogram was obtained simultaneously with an M-mode echocardiogram. The S3 coincides with the peak of the rapid filling wave (RF) and the S4 is synchronous with the A wave of the apex cardiogram, immediately following left atrial contraction and the reopening motion of the mitral valve at end-diastole. (From Tavel MC: Phonocardiography: clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 36:145, 1983.)

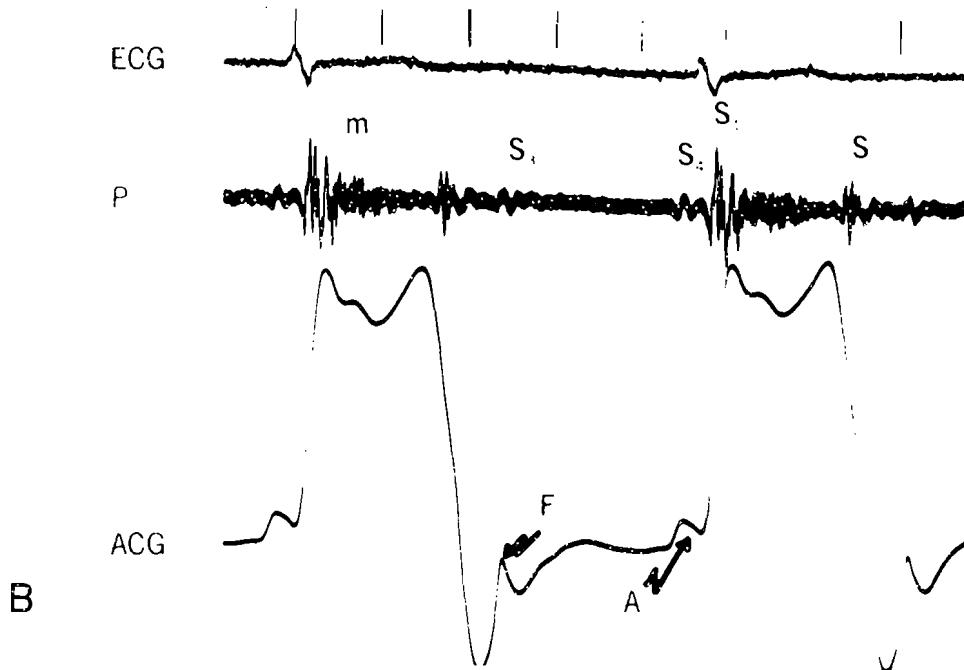


FIG. 7-2. (Continued). B. Relationship of the apex cardiogram to the physiologic third heart sound. The S₃ is synchronous with the prominent peak of the rapid filling wave (F) occurring in early diastole. A deflection representing an S₄ is simultaneous with the A wave of the apex cardiogram (ACG). A systolic flow murmur (m) is present. (From Stefadouros MA and Little R: The cause and clinical significance of diastolic heart sounds. Arch Intern Med 140:537, 1980.)

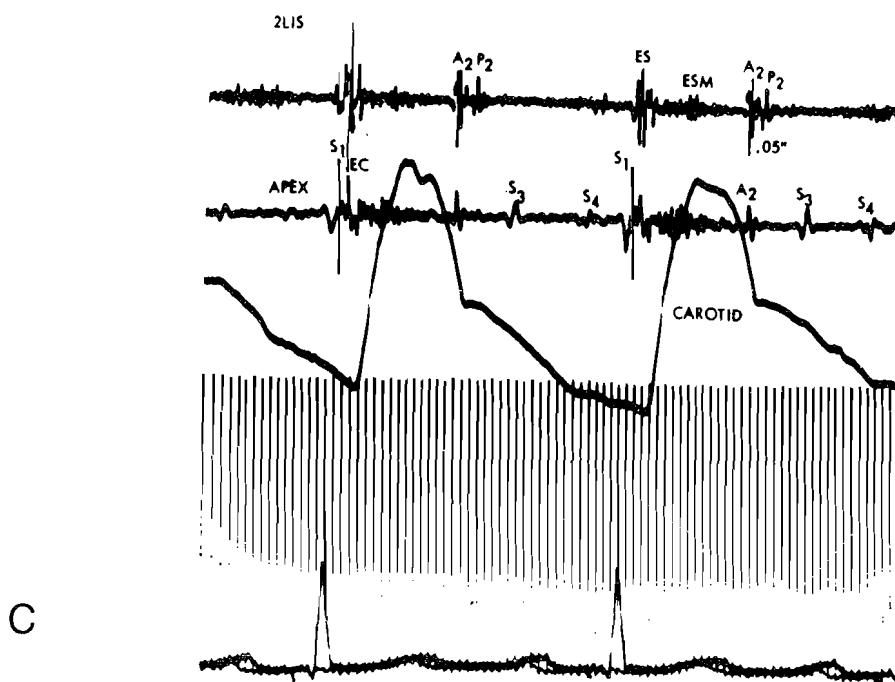


FIG. 7-2. (Continued). C. Third and fourth heart sounds associated with hyperkinetic circulation. This phonocardiogram was taken from a young person with thyrotoxicosis. Note the prominent S₃, systolic ejection (ESM) murmur, and the soft S₄. An ejection sound (ES,EC) and loud S₁ are present. Any condition associated with a high output state, such as anemia or thyrotoxicosis, can result in an S₃, which itself may be palpable. (From Reddy PS, Shaver JA, and Leonard JJ: Cardiac systolic murmurs: pathophysiology and differential diagnosis. Prog Cardiovasc Dis 14:1, 1971.)

TABLE 7-1 *Clinical Correlates of the Third Heart Sound*

<i>Normal or Physiologic S3</i>
Children, young adults
Hyperkinetic states in subjects with normal hearts
<i>Diastolic Overload States</i>
Left ventricle
Mitral regurgitation
Ventricular septal defect
Patent ductus arteriosus
Right ventricle
Tricuspid regurgitation
Atrial septal defect
(uncommon)
<i>Left Ventricular Dysfunction</i>
Congestive heart failure
Dilated left ventricle
Markedly depressed ejection fraction
<i>Constrictive Pericarditis</i>
Pericardial knock

pathologic ventricular gallop. The S3 may be soft or loud and will usually attenuate or disappear when the subject assumes the upright position. Most individuals with a physiologic S3 are young and may have associated systolic flow murmurs and/or a venous hum (Fig. 7-2B). *Practical Point: In children and young adults it is important to consider the possibility that an S3 is physiologic and related to increased cardiac output. This will avoid the false diagnosis of cardiac disease.* Occasionally, a physiologic S3 will be sustained in duration and can simulate a short diastolic murmur. In the thin subject or a child with a hyperactive heart, a normal S3 actually may be palpable.

Hemodynamics. Rapid rate of early LV diastolic filling with normal to slightly elevated left atrial pressure; normal or increased cardiac output.

Diastolic Overload States

An S3 may be produced whenever an increased volume of left atrial blood crosses the mitral valve in early diastole, especially if left atrial pressure is increased. Thus, an S3 is likely to be found in hemodynamically significant mitral regurgitation, ventricular septal defects, or a patent ductus arteriosus. In mitral regurgitation, the S3 is characteristically somewhat earlier than usual and may have dominant high-frequency vibrations simulating an opening snap (Fig. 5-5).

The S3 in diastolic overload states is acoustically similar to the normal S3. Left-to-right shunts with increased mitral valve flow (e.g., patent ductus

arteriosus, ventricular septal defect) and mitral regurgitation commonly produce loud diastolic filling sounds (S3), often with short after vibrations or a mid-diastolic flow rumble. These can be soft or quite prominent. If LV contractile function is normal in such patients, the S3 does not have an ominous prognosis but does indicate a major volume overload. In such cases the velocity and amplitude of early diastolic filling is increased, and the precordial LV impulse typically is hyperactive.

The development of significant pulmonary hypertension will soften the S3, although if RV failure subsequently occurs, a right-sided S3 may appear. Tricuspid regurgitation does not produce a prominent S3 unless there is an enormous RV volume overload. Atrial septal defects are not usually associated with a loud S3, although a mid-diastolic flow rumble is common in patients with a large left-to-right shunt. In both situations, there is increased blood flow across the tricuspid valve, usually with elevation of right atrial pressure. Perhaps because the RV is more compliant than the LV, the presence of an S3 with right heart overload is relatively uncommon.

Hemodynamics. Well-preserved cardiac output and ejection fraction with high peak diastolic filling rates; atrial pressure elevated, especially if there is a large V wave from mitral or tricuspid regurgitation.

Left Ventricular Dysfunction

An S3 is characteristic of global LV impairment. In a middle-aged or older adult without mitral regurgitation, the presence of an S3 suggests a significant decrease in myocardial contractility and a depressed ejection fraction. If the S3 is related to a new clinical event (e.g., accelerated hypertension, acute myocardial infarction), the S3 may be transient and will disappear when LV function improves.

Significant myocardial disease of any etiology may result in an abnormal S3 (Fig. 7-3). In the presence of left ventricular decompensation associated with secondary or functional mitral regurgitation, the S3 may be related more to the poor ventricular function than to the left atrial volume overload (Fig. 7-4). In such cases the systolic murmur tends to be soft and the heart larger than in patients with primary mitral regurgitation and good left ventricular function. With appropriate medical treatment the regurgitant murmur and S3 related to left ventricular dysfunction will become softer and may disappear; in the patient with a severely regurgitant valve with preserved LV function, however, the murmur and S3 become louder as cardiac compensation improves with therapy (Fig. 5-5).

A pathologic S3 caused by LV decompensation will persist when the patient assumes the upright position, although it may soften with this maneuver. The S3 may be very loud and is often associated with other signs of LV decompensation, such as pulsus alternans, a sustained LV apical impulse,

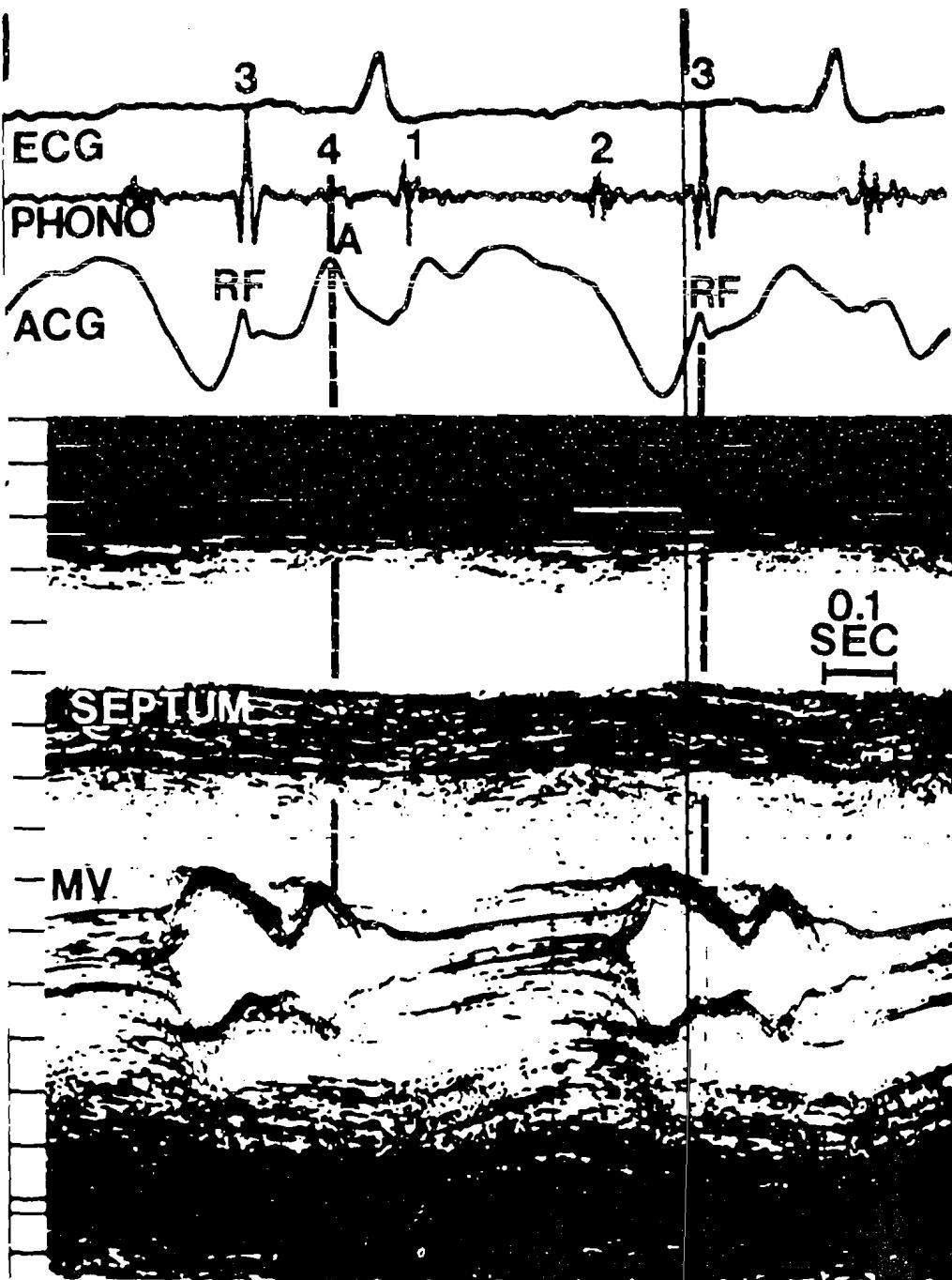


FIG. 7-3. Third and fourth heart sounds in congestive heart failure. This simultaneous phonocardiogram was obtained from a patient with heart failure due to an ischemic cardiomyopathy. Note the accentuated S3 and the prominent rapid filling wave as well as the S4 and A wave on the apex cardiogram. (From Tavel ME: Phonocardiography: clinical use with and without combined echocardiography. Prog Cardiovasc Dis 26:145, 1983.)



FIG. 7-4. Third heart sound produced by ischemic left ventricular dysfunction and mitral regurgitation. This phonocardiogram was obtained from a 67-year-old male with severe coronary artery disease and left ventricular dysfunction. Note the prominent S3. The systolic murmur of mitral regurgitation may be related to papillary muscle dysfunction. Wide splitting of S2 is caused by right bundle branch block. (From Cohen MV: Correlative atlas of adult cardiac disorders. Mount Kisco, NY, Futura Publishing Co., 1980.)

narrow pulse pressure, or signs of overt congestive heart failure. In general, a persistent and loud S3 heralds a poor prognosis.

An S3 in the presence of coronary artery disease strongly suggests major LV asynergy or an LV aneurysm (Figs. 7-3, 7-4). A sustained LV lift or ectopic impulse is a common associated finding. An S3 gallop is common in decompensated hypertensive and aortic valve disease. A loud S3 is typical in dilated cardiomyopathy but is not found in hypertrophic cardiomyopathy.

Hemodynamics. The cardiac index and ejection fraction are substantially decreased. Left atrial, pulmonary artery diastolic, and pulmonary capillary wedge pressures are elevated. Left ventricular early diastolic pressure is elevated; the late or end-diastolic ventricular pressure (A-wave) may or may not increase further with atrial contraction. The LV chamber is usually dilated with an increased end-diastolic volume; the cavity/wall thickness ratio is increased. Patients with coronary disease who are found to have an S3 are likely to have LV asynergy.

Pericardial Knock of Constrictive Pericarditis

The early, loud, and high-pitched diastolic sound that is often associated with constrictive pericarditis is actually an "S3" that occurs *earlier* than the typical S3 (0.08 to 0.10 seconds after A2) (Fig. 5-4). The pericardial knock usually has a clicking or snappy quality and is easily mistaken for an opening snap. However, this sound may be qualitatively identical to the typical S3, and one should not exclude the diagnosis of pericardial constriction because the diastolic sound is not high pitched or loud. The early and prominent pericardial knock or S3 is related to an elevated left atrial pressure and a rapid rise of early ventricular diastolic pressure caused by the nondistensible pericardial shell encasing the heart or the extremely stiff myocardium of restrictive cardiomyopathy. Often palpable, the S3 may be the loudest of all three heart sounds in such patients. As mentioned, in some subjects with constrictive pericarditis this sound is indistinguishable from a typical S3. This may be related to adhesions that are less dense than usual or to the presence of coexisting pericardial effusion.

In some patients with restrictive cardiomyopathy, an early S3 with distinct high frequency components can simulate a pericardial knock.

Hemodynamics. Markedly elevated left and right atrial and ventricular diastolic pressures. Increased velocity of diastolic filling. Ventricular systolic function usually remains normal.

FOURTH HEART SOUND

Although there is no disagreement about the mechanism of production of the S4, there is considerable controversy regarding its true incidence and clinical implications.

PHYSIOLOGY OF S4

The S4 is a low frequency sound following left atrial systole (Figs. 7-1, 7-2A). Atrial contraction results in an abrupt increase in LV end-diastolic fiber length. Although the S4 is also known as the atrial sound, the S4 is ventricular in origin. It follows the P wave by 0.14 to 0.20 sec and precedes S1. The actual contraction of the atria produces no audible sound. Rather, audible vibrations of the S4 result from sudden tensing of the LV mass, the mitral valve apparatus, and the blood within the ventricle. Coupling of the LV with the chest wall is an important but poorly understood factor in S4 audibility.

The *hemodynamic correlates* of the abnormal S4 include: an LV chamber that often is hypertrophied but with little or no increase in LV diameter;

normal LV pressure in early diastole with an elevation of *end-diastolic* pressure; a low compliance ventricular pressure-loop; impaired velocity of early LV diastolic filling; and a well-preserved cardiac output. The LV cavity/wall thickness ratio is low; the LV chamber is stiff and noncompliant.

Normal S4. An S4 occasionally may be heard when there are no detectable abnormalities of ventricular function other than increased blood flow, as in young subjects or patients with thyrotoxicosis (Fig. 7-2C), or in the presumed physiologic loss of left ventricular compliance with aging. Whether a distinct audible S4 is truly normal in older adults is the subject of considerable controversy (see below).

Abnormal S4. In the normal heart, the LV passively receives 70 to 80% of its diastolic filling volume from the left atrium during early to mid-diastole, with a smaller contribution from atrial contraction in late diastole. In myocardial disease such as hypertrophy and/or dilatation, left atrial contraction becomes responsible for a greater proportion of LV filling. This LA "booster pump" function takes on increasing importance as LV distensibility decreases; left atrial contraction may provide up to 30 to 40% of LV diastolic filling in some circumstances. Under these conditions, the S4 or atrial gallop may be quite prominent (analogous to P wave enlargement on the EKG). Early to mid LV diastolic pressure usually is normal, but LV end-diastolic pressure is elevated.

As LV dysfunction progresses, early and mid ventricular diastolic pressure rises; in general, left atrial contraction produces a relatively small further increase in pressure once the LV cavity has enlarged, particularly if LV failure has appeared. At this stage the S4 becomes softer or is inaudible. This sequence also is reflected in the LV apex impulse. Initially, the increased A wave amplitude may be palpable due to reduced LV compliance (see Chapter 5). As the LV chamber dilates, the palpable atrial sound may disappear, even though mean left atrial pressure is distinctly abnormal and higher than when the A wave and S4 were present.

Importance of Left Atrial Function. Vigorous left atrial contraction is necessary to provide sufficient blood flow to abruptly distend the LV in late diastole and produce an audible S4. When atrial contractile forces are reduced by fibrosis, ischemia, or dilatation, the S4 may be soft or absent. For this reason, patients with intermittent atrial arrhythmias frequently do not manifest an S4 when in sinus rhythm, presumably because of atrial dysfunction. In chronic severe mitral regurgitation, an S4 is rare in spite of elevated left atrial pressure; although the left atrium is very large and distensible, atrial contractile force is diminished. When the onset of mitral regurgitation is acute or recent, however, an S4 is common; the normal-sized atrium contracts with a markedly increased, hyperdynamic LA systole (Chapter 17; Fig. 17-8).

In complete heart block, random atrial sounds can be heard; these may be quite prominent whenever the atrium contracts against closed AV valves.

In this condition, the atrial sounds are not generated from the LV walls but probably reflect tensing of the atrium and mitral valve apparatus.

PREVALENCE OF S4: AN UNRESOLVED CONTROVERSY

In recent years two major unresolved issues regarding the S4 have been the subject of considerable debate. These relate to (1) the *audibility* of the S4 and (2) the *normality* of finding an atrial sound in adults. These questions are relevant because of numerous observations that phonocardiograms can record an S4 at the cardiac apex in many adults with no evidence of overt heart disease (Figs. 7-2B, 7-5). Some studies have documented a recordable S4 in the supine or left lateral positions in 35 to 70% of normal middle-aged adults. Thus, a *recordable* low frequency sound following atrial contraction can be demonstrated in many healthy people. However, these sound transients often are of very low intensity and may not be audible (Fig. 7-5). Most experts believe that a recordable but inaudible S4 vibration is clinically inconsequential but agree that a *distinctly audible S4 is abnormal*. Some believe that atrial sounds are so common in normal older people as to have no clinical significance. One recent study of 100 "normal" men with a recordable S4 demonstrated a threefold increase in the incidence of coronary events during a five-year follow-up. This suggests that many subjects with an apparently normal heart who have a recordable S4 may actually have latent heart disease.

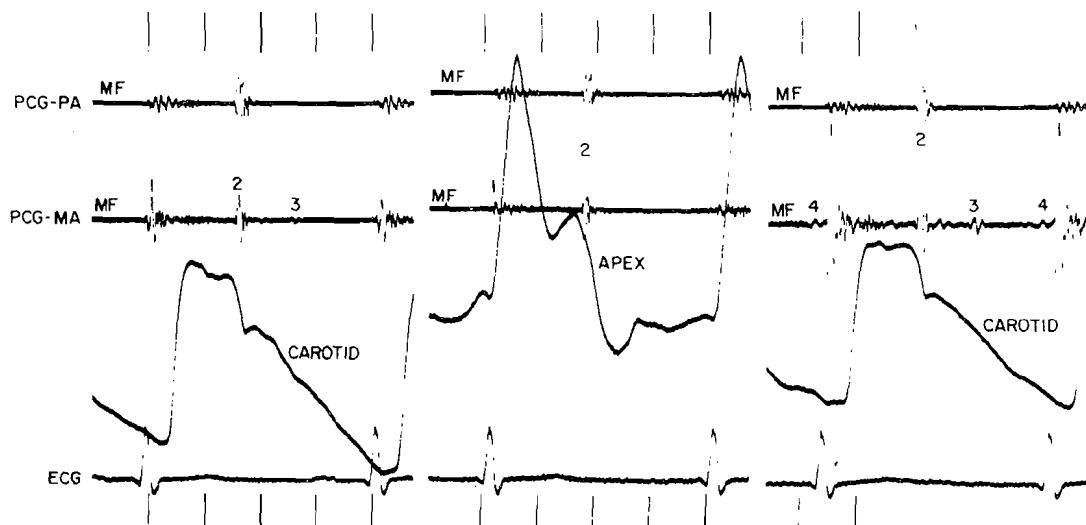


FIG. 7-5. Effect of phonocardiogram frequency settings on recordability of the fourth heart sound. In the left and middle panels the medium frequency setting is used, and an S4 is not recorded. In the right panel, a low frequency filter is used, and an S4 is recorded. Note also the accentuated S3. It is relatively easy to record an atrial sound or S4 in adults, but this does not necessarily imply audibility. (From Craige E: The fourth heart sound. In *Physiologic Principles of Heart Sounds and Murmurs*. Edited by DF Leon and JA Shaver. American Heart Association Monograph No. 46, 1975.)

There is a consensus among all investigators that a *very loud* or *palpable S4* is always abnormal and signifies decreased LV compliance (Figs. 7-3, 7-6). Clearly, the examiner's subjective evaluation of the intensity of the S4 plays a large role in this argument. *Practical Point: The majority of clinicians and investigators believe that the presence of a distinctly audible atrial sound or S4 in an older subject is almost always abnormal and, conversely, that normal persons infrequently have a distinctly audible S4.*

Palpable Atrial Sound

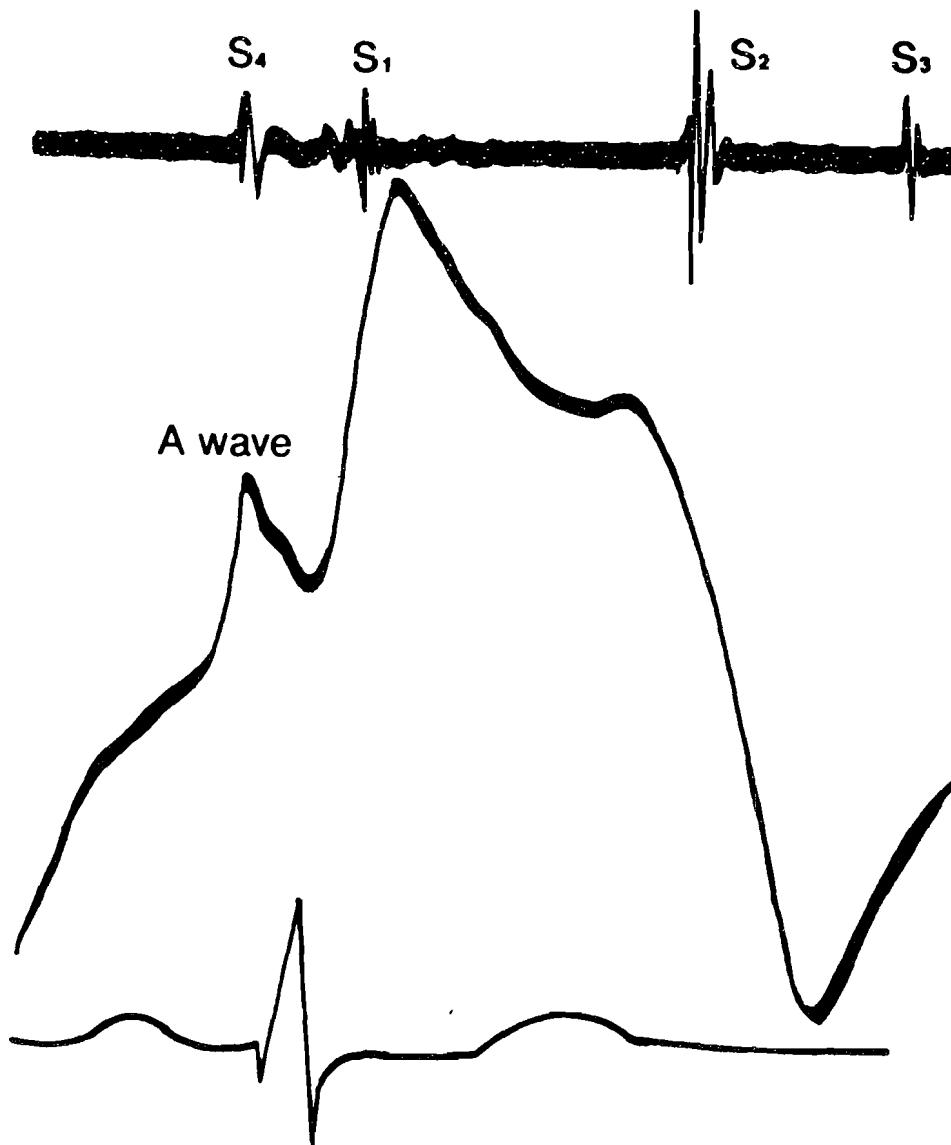


FIG. 7-6. Palpable atrial sound. When an S4 is very loud it may be associated with a palpable presystolic impulse on careful examination of the left ventricular apex beat. This is much more likely to be found when patients are turned into the left lateral decubitus position. The presence of the palpable S4 indicates that the atrial sound is pathologic. (From Abrams J: Prim Cardiol, 1982.)

CLINICAL SIGNIFICANCE

A prominent S4, often with an increased precordial A wave amplitude, is commonly found in patients with underlying coronary disease, hypertension, aortic stenosis, and hypertrophic cardiomyopathy (Fig. 7-7). The intensity of an S4 varies considerably; when prominent, the S4 may be the loudest heart sound heard at the apex. The common denominator in individuals with a prominent S4 is left ventricular hypertrophy with increased LV end-diastolic pressure and restricted early diastolic filling produced by a stiff left ventricle. An S4 is a far more sensitive indicator of decreased LV compliance than an S3. The clinical implication of an audible S4 is, however, far different from that of the S3, which often signifies cardiac decompensation, incipient or overt congestive heart failure, and a poor long-term outlook. In general, an S4 implies a less serious alteration in left ventricular function and carries a more benign prognosis than the S3.

CLINICAL CORRELATES

Normal Hearts. An atrial sound occasionally may be heard in normal children and young adults. Hyperkinetic states, such as anemia or thyrotox-

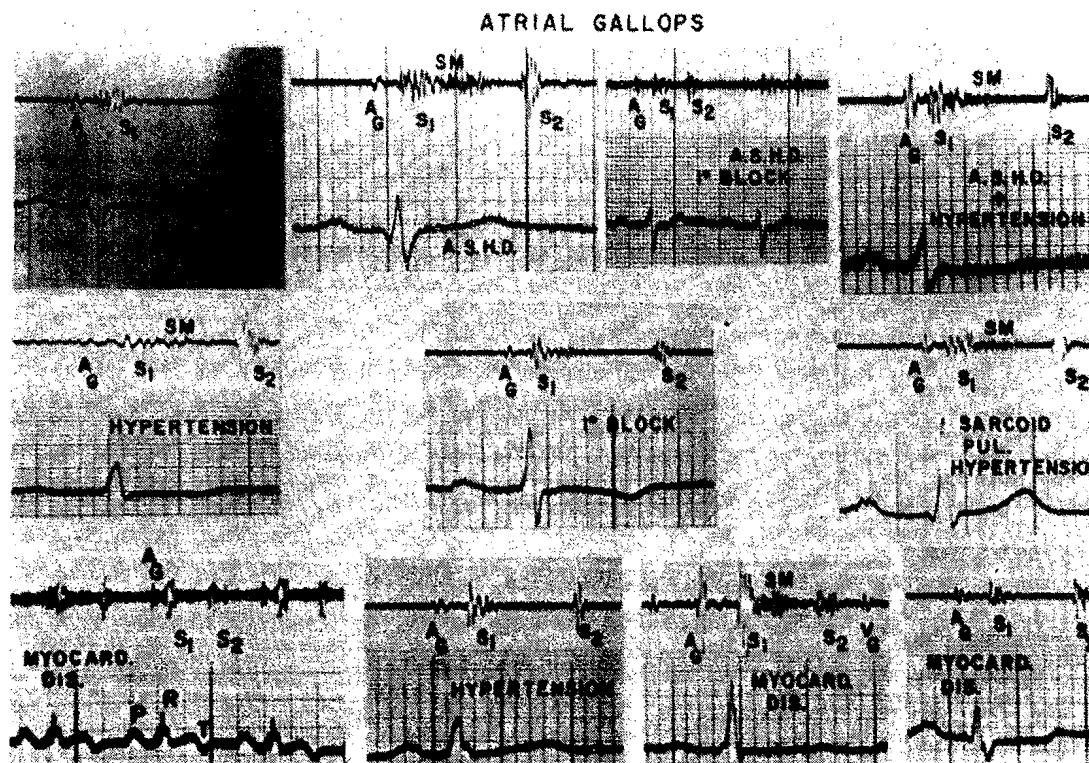


FIG. 7-7. S4 or atrial gallop in cardiovascular disease. This composite of 11 patients demonstrates atrial gallops (AG) in various cardiac conditions. SM = systolic murmur. (From Harvey WP: Heart sounds and murmurs. Circulation 30:262, 1964.)

icosis, are likely to produce an audible S4 (Fig. 7-2C). As already discussed, there is considerable controversy over the meaning of an audible S4 in a healthy adult.

PR Interval. Subjects with a long PR interval (first degree AV block) may have an audible S4 in the absence of other evidence of cardiovascular disease (Fig. 7-7). In second or third degree A-V block, independent atrial contractions can produce "atrial sounds" throughout diastole when the mitral valve is open. Atrial contraction against a closed mitral valve may also produce audible sound vibrations.

A short PR interval may or may not prevent detection of an S4; when the S4 is close to S1, the ear may not be able to detect two separate sounds. S1 is loud with short PR intervals (see Chapter 6). Some experts believe that a short PR may accentuate the audibility of the S4.

Coronary Heart Disease. An S4 is a hallmark of ischemic heart disease (Figs. 7-3, 7-7). For instance, the reduced LV compliance resulting from a prior myocardial infarction frequently results in an audible S4. An audible S4 has been demonstrated in 80 to 90% patients during acute myocardial infarction (Chapter 22). The S4 initially may be inaudible in the acute infarct patient, but usually will be manifest after one or two days. In patients with intermittent atrial arrhythmias, the S4 may be absent, perhaps because of less effective atrial contraction during sinus rhythm. It has been suggested that an S4 that persists into the post-infarction period may indicate that the patient is at higher risk for subsequent cardiac events, although some investigators believe that an S4 is almost always detectable in patients with overt coronary heart disease. Experienced clinicians commonly hear atrial gallops in patients with angina or prior myocardial infarction. An S4 may appear during anginal pain and disappear after administration of nitroglycerin or rest. In patients with LV asynergy or an overt LV aneurysm, an S4 is common and is typically palpable (Fig. 7-3). Similarly, an atrial sound commonly is heard immediately after exercise testing. Many believe that an isolated atrial sound in an apparently healthy adult may be a harbinger of a future coronary event.

Hypertension. An isolated S4 is common in chronic hypertension, suggesting LV hypertrophy and reduced compliance even in the absence of definite EKG evidence of LV. In this setting, the S4 does not indicate serious left ventricular dysfunction (Fig. 7-7).

Aortic Stenosis. An audible S4 in patients with valvar aortic stenosis may be a valuable sign indicating a significant left ventricular-aortic gradient. Concentric LV hypertrophy and a stiff ventricle provide the necessary substrate for an atrial sound in such patients. The finding of an S4 in patients over 50, however, is less specific in predicting severe aortic stenosis, as coronary disease or hypertension frequently may cause an S4 in this age group. A palpable or extremely loud atrial sound in aortic stenosis usually indicates severe aortic valve obstruction in any age group (Fig. 13-4).

Hypertrophic Cardiomyopathy. Audible and palpable atrial sounds are almost always present in this condition; if an S4 is absent, the diagnosis should be seriously questioned. Often a huge precordial A wave is the dominant outward impulse (Fig. 14-6). An S4 is characteristic of both obstructive and nonobstructive varieties of hypertrophic cardiomyopathy.

Cardiomyopathy. An S4 is common and usually palpable in subjects with significant myocardial disease (Fig. 7-7). An S3 is also typically present, and in late stages is more likely to be present than an S4 (Figs. 7-3, 7-4).

Acute Mitral Regurgitation. An S4 is characteristic of patients with mitral regurgitation of recent onset (Fig. 17-14). The acute left atrial volume overload results in a forceful atrial contraction that expels blood into a normal-sized left ventricle and in a resultant atrial sound. In chronic mitral regurgitation, an atrial sound usually is absent, but an S3 is common. Thus, the presence of an S4 in a patient with mitral regurgitation strongly suggests that the mitral lesion is of relatively recent onset.

Arrhythmias. In atrial fibrillation, the S4 is absent, but on reversion to sinus rhythm, a dilated atrium may have a decreased force of contraction that cannot produce an S4, even when there is underlying left ventricular dysfunction. Sinus tachycardia may accentuate or bring out an S4; if, however, the PR is short, the atrial sound may not be distinguishable from a loud S1. In second and third degree A-V block, atrial sounds can be heard variably throughout diastole; asynchronous atrial contraction can open a closed mitral valve. During systole, atrial contraction against a closed mitral valve may also produce audible sound in patients with high-grade heart block.

Right Ventricular S4. In pulmonary hypertension and pulmonary stenosis, a right-sided S4 is common. The presence of an S4 in a patient suspected of pulmonary embolization is an important finding, suggesting that there is a significant RV pressure overload. A split S1 often is best heard at the lower left sternal border and may be confused with a right-sided S4. Inspiratory augmentation of the S4 and maximal location at the lower left sternal border are helpful in the differential diagnosis of a right ventricular S4.

Summation Gallop and Quadruple Rhythm. Whenever the PR interval is long and/or the heart rate is rapid, atrial contraction will "move into" the rapid filling phase of diastole. Since diastole shortens more than systole when heart rate increases, passive ventricular filling is superimposed on the augmented flow across the mitral valve due to LA systole. This combination may cause a very loud *summation gallop* (S3 plus S4). The summation gallop often has some duration and can mimic a mid-diastolic rumble (Fig. 7-8). When the heart rate slows as a result of therapy or carotid sinus pressure, the individual S3 and S4 sounds may each be less prominent or even inaudible. A very soft S3 may summate with an S4 during tachycardia, producing an extremely loud single sound. Inducement of cardiac slowing by carotid sinus massage is a useful bedside technique to evaluate gallop rhythm during tachy-

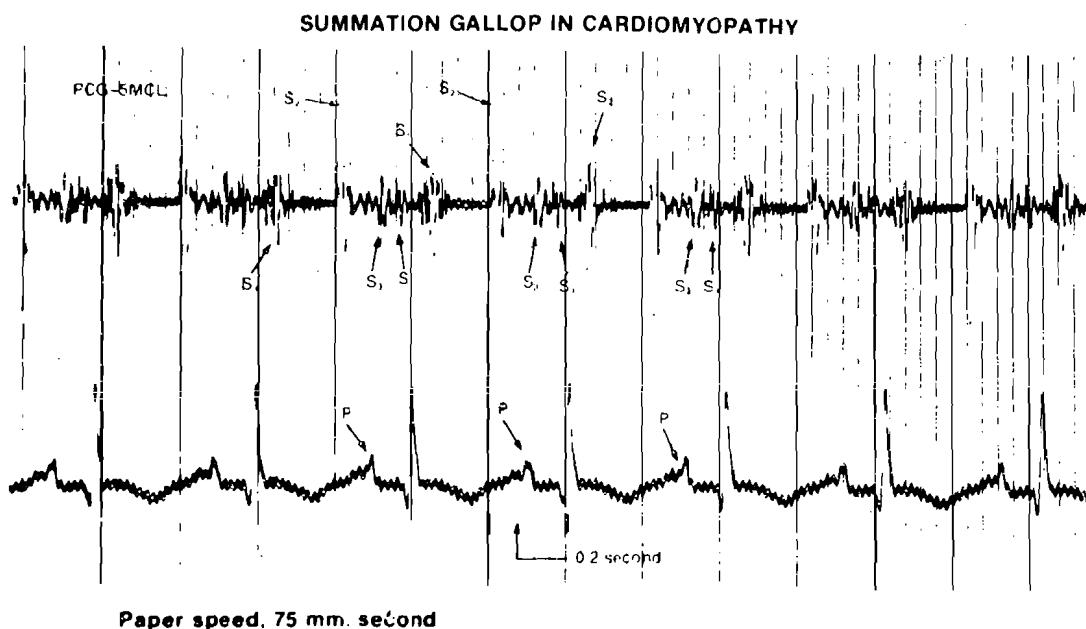


FIG. 7-8. The summation gallop. When an S3 and S4 are both present and the heart rate is rapid, the sounds move close together and may be heard as a single loud diastolic sound. This combination can also simulate a mid-diastolic or flow rumble. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co., 1979.)

cardia or to help analyze other confusing cardiac findings when there is a rapid heart rate.

When both an S3 and S4 are clearly audible, the term *quadruple rhythm* is used. Increasing the heart rate by forced coughing or mild exercise may cause the two sounds to summate and produce a loud single filling sound. Quadruple rhythm can be confusing, as the series of audible low frequency vibrations during diastole may simulate a mitral stenosis murmur. Quadruple rhythm commonly is found in patients with LV aneurysm, cardiomyopathy, or severe LV failure and dilatation from any cause.

Factors Relating to S4 Audibility. In cases of acute or chronic left ventricular decompensation, the S4 closely follows the P wave of the EKG and is more widely separated from S1 at any given PR interval. Thus, it may more readily be heard. As LV pressure and/or volume decreases during therapy for heart failure or hypertension, the P wave-S4 interval lengthens, the S4 occurs later and "moves" closer to S1, decreasing audibility. An S4 may seem to disappear following the acute phase of myocardial infarction or after treatment of significant hypertension, but in fact the sound is simply more difficult to appreciate as it moves closer to S1. On the other hand, some believe that the S4 may be more easily distinguished if it is close to S1.

With long PR intervals an S4 is more readily heard and S1 is soft; this may mimic a split S1 if both sounds are of equal intensity. Thus, an audible S4 in a patient with first degree AV block does not have the same implication

as in a subject with a normal PR interval. A low frequency, split S1 can be confused with S4. Very loud or "long" atrial sounds occasionally can simulate a presystolic murmur, particularly if the first heart sound also has some duration.

HOW TO LISTEN FOR S3 AND S4 (Table 7-2)

The third and fourth heart sounds are low pitched (20 to 60 cps) and usually of low intensity. They occur at the lower range of human audibility at a frequency range below which most people are accustomed to hearing cardiac sound. The S3 often has some "duration" or after-vibrations; the S4 is slightly higher pitched and usually is louder.

Both sounds must be actively sought in order to be detected. They typically sound like a distant thud. The sounds may be palpable, particularly the S4 (Figs. 7-3, 7-6). In order to hear the S3 and S4 best a noise-free room without background vibrations from air conditioning or heating systems is desirable. One should avoid sound artifacts from muscle tremor or interfering noises from clothing or stethoscope tubing.

The S3 and S4 often are best heard immediately upon beginning auscultation and may appear to fade away after a short period of time. Frequently, gallop sounds are heard only immediately after the patient assumes a new

TABLE 7-2 *Proper Approach to Auscultation of S3 and S4*

Stethoscope Technique

- Have a quiet room and surroundings
- Routinely use left lateral position
- Identify LV apex impulse
- Use the bell with light pressure
- Use rubber outer ring on bell

Helpful Physiologic Maneuvers

Alterations in venous return	Response
Increase: leg elevation coughing sit-ups abdominal compression Valsalva release phase	Increase intensity
Decrease: sitting standing Valsalva strain phase	Decrease intensity
Sustained handgrip (isometric) If heart rate is rapid, use carotid sinus massage	Increase intensity

position, such as getting onto the examining table or turning over into the left lateral position.

Timing and Respiratory Variation. The S3 is linked to S2 in timing, whereas the S4 occurs just before S1 (Fig. 7-1); these relationships hold for very slow or very rapid heart rates. The S3 occurs in early diastole, usually 0.14 to 0.16 sec after A2 (range 0.10 to 0.20 sec). The S4 is presystolic; it follows the P wave by 0.12 to 0.20 sec, and immediately precedes S1 (Figs. 7-1, 7-2A). The S3 may vary with respiration and usually is loudest during expiration, fading at end inspiration. The S4 also may wax and wane with the respiratory cycle but not as prominently as the S3. Usually it is loudest in early expiration.

Helpful Maneuvers in Auscultation of S3 and S4 (See also Chapter 11). Maneuvers that alter venous return are useful during auscultation of the low frequency diastolic filling sounds (Table 7-2). They will affect both the S3 and S4. Anything that increases venous return and intracardiac blood volume accentuates the loudness of the S3 and S4; conversely, a decrease in cardiac filling will attenuate these sounds. Maneuvers that increase venous return include elevation of the legs, coughing, mild exercise, abdominal compression, and the release phase of the Valsalva maneuver. A right-sided S3 or S4 will augment with inspiration. Decreased venous return occurs with sitting, standing, tourniquets, and the strain phase of the Valsalva maneuver. Having the patient assume the upright posture is often helpful; both an S3 and an S4 typically attenuate or disappear. Some experts stress that a physiologic S3 or S4 usually will disappear when the patient becomes upright but that pathologic gallops persist, although they may become softer.

Positional changes in sound intensity may help the physician distinguish the S3 and S4 from other sounds or combinations, such as S3 versus opening snap, split S1 versus S4-S1, S1-ejection click versus S4-S1. Carotid sinus pressure may be effective in slowing the heart rate when there is difficulty in timing the gallop rhythm. An S4 may increase or decrease in intensity following this vagal maneuver without any change in the PR interval.

Handgrip exercise may increase the intensity of both the S3 and S4. This isometric maneuver has been used as a diagnostic aid in patients with suspected coronary artery disease. The increased myocardial oxygen demands following sustained handgrip may evoke left ventricular ischemia and increase the stiffness of the ventricle, thus precipitating a ventricular or atrial gallop (Chapter 11). Not all workers believe that the handgrip maneuver reliably augments the intensity of S4.

PROPER USE OF THE STETHOSCOPE

The S3 and S4 are low-frequency transients that are best and often *only* heard with the bell of the stethoscope. A large diameter bell with a rubber

outer ring is best (Fig. 1-2). The lightest possible pressure to make a skin seal should be used (Fig. 7-9). As stethoscope pressure is increased to filter out low frequency vibrations, the S3 and S4 will attenuate (Fig. 7-5). A loud S3 or S4 usually has medium to high frequency components and may sound more high-pitched than usual. A particularly loud S3 or S4 will remain audible even when firm stethoscope pressure is applied. When very prominent, these sounds may also be heard at sites away from the cardiac apex.

Location. The left ventricular S3 and S4 are maximal at the apex impulse. The apical impulse should be carefully identified with the examining finger (Fig. 5-6), and then the bell should be placed directly on the apex using light pressure. Occasionally, an S3 or S4 is best heard slightly medial or just superior to the apical impulse. Loud gallops may be audible medial to the apex or even at the lower left sternal border but are typically softer

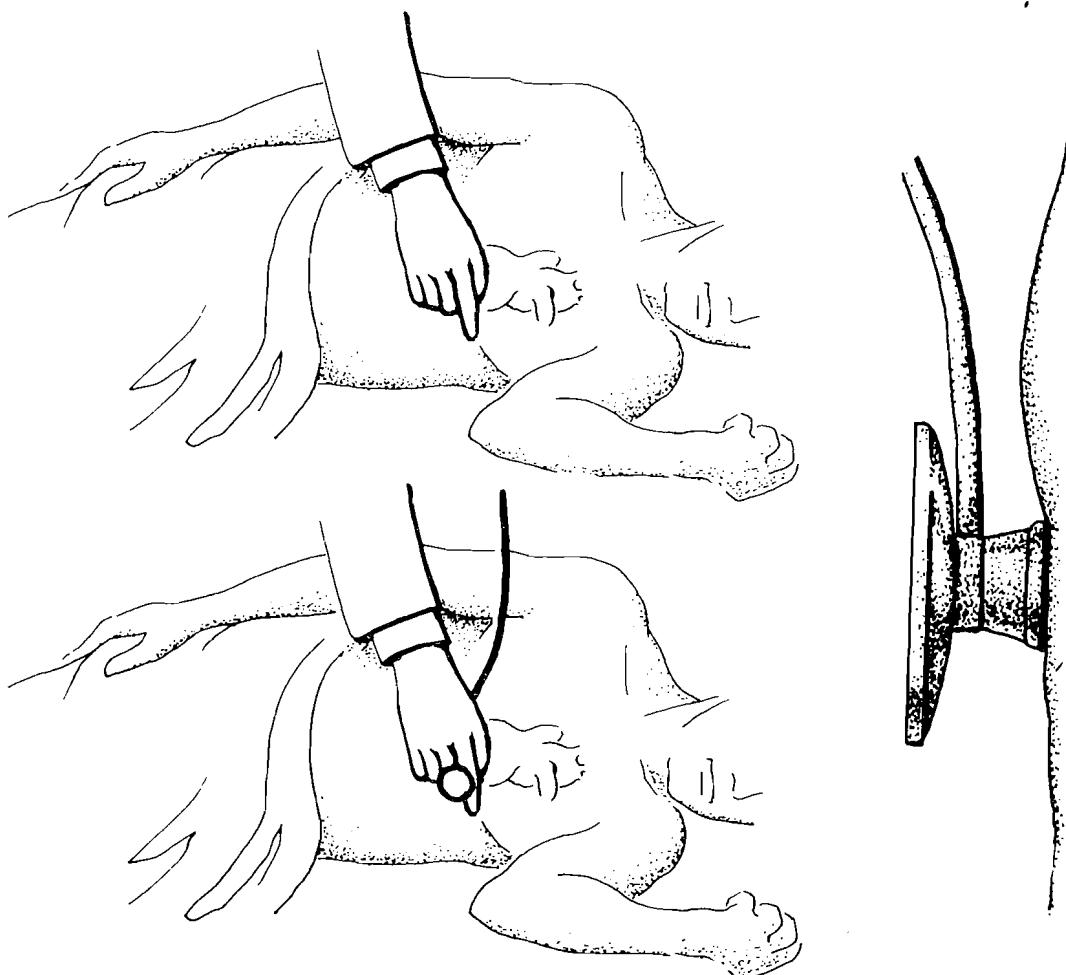


FIG. 7-9. Use of the left lateral decubitus position in detection of the S3 and S4. Upper: The left ventricular apex is first identified by careful palpation (above). Lower: The bell of the stethoscope is then applied directly over the apical impulse, using the lightest pressure possible that will create a skin seal. This technique enhances audibility of low frequency cardiac sounds (e.g., S3, S4, mitral diastolic murmurs). (Adapted from Abrams J: Prim Cardiol, 1982.)

at these locations than at the apex. On occasion, gallop sounds, particularly the S4, can be heard in the right supraclavicular area; this location may be helpful in patients with lung disease or obesity. In patients with emphysema, the LV S3 or S4 may be best heard at the subxiphoid area or lower left sternal border. Differentiation from gallops of RV origin in these patients may be difficult.

Left Lateral Decubitus Position. It is essential to examine any patient suspected of having left ventricular disease in the left oblique or left lateral decubitus position (Figs. 5-7, 7-9). *Practical Point: The S3 and S4 often are inaudible when the patient is supine and may be heard only in the left lateral position.* This maneuver thrusts the apex of the left ventricle close to the chest wall and accentuates audibility of low pitched diastolic sounds, causing the amplitude of both the S3 and S4 to increase dramatically. This maneuver may also cause these low frequency events to become palpable; light pressure on the pads of the fingers should be used, with careful attention for a pre-systolic bulge or an early systolic thrust. Palpable presystolic distention (S4) is much more common than a palpable S3 and is a most important confirmatory finding in patients with a suspected S4 (Figs. 7-3, 7-6).

Right Ventricular Origin of S3 and S4. Any condition resulting in right ventricular hypertrophy or dilatation may produce a right-sided diastolic heart sound. The cardinal features of an S3 and S4 generated in the right heart are: (1) maximal intensity at the lower left sternal border (tricuspid area); (2) inspiratory augmentation; and (3) evidence for associated right ventricular disease (e.g., parasternal lift, increased P2, elevated jugular venous pressure with large A or V waves).

A right ventricular S3 may be associated with tricuspid regurgitation. In a patient with a large right ventricle extending into the left chest, a right-sided S3 may be difficult to distinguish from an LV S3. The RV S3 usually is slightly higher pitched and increases with inspiration.

A right-sided S4 is common in the presence of pulmonary hypertension, which results in right ventricular hypertrophy with decreased compliance. Differentiation of a right-sided S4 from a split S1 can be difficult. Both the S4 and the second component of a split S1 (T1) usually increase in intensity with inspiration, and this may be a helpful distinguishing clue. Right-sided atrial sounds occasionally can be heard in the right supraclavicular fossa, probably representing vibrations of vigorous right atrial contraction transmitted retrogradely into the neck. Patients with a right-sided S4 usually have a large and prominent A wave in the jugular venous pulse; this finding may help to identify accurately the ventricular origin of the S4.

Auscultation of S4. Many important aspects of proper auscultation of the S4 have already been discussed. Several points require emphasis:

1. The S4 frequently is audible *only* in the left lateral position and must be carefully sought in any patient with suspected LV disease (Fig. 7-9).

2. Confusion between a split S1 or S1-ejection click complex is common. Most double sounds heard at the apex in the adult represent a split S1 and not an S4-S1 complex (Fig. 6-4).
3. The S4 is often palpable in the left decubitus position; this finding indisputably confirms that the S4 is an abnormal event in a given patient (Fig. 7-6).
4. Other signs of cardiovascular disease (e.g., increased venous A wave, LV heave) may influence the examiner's impression regarding the presence or absence of S4. Complete objectivity is important as one assesses all clues that suggest underlying cardiovascular disease.

Differential Diagnosis of S4. When two sounds in close proximity are heard at the apex at the time of S1, the differential diagnosis includes a prominently split S1, an S4-S1 complex, and an S1-early click complex (Fig. 6-4, Table 8-1).

S4-S1 Complex. The S4 is the first of the two sounds; it is low pitched and may vary with respiration. The S4, but not the S1, will attenuate or disappear with firm pressure on the stethoscope bell; it becomes softer in the upright position. The S4 will increase with handgrip, following sit-ups, or coughing. Maneuvers that increase the intensity of S4 also tend to separate S4 from S1, further enhancing audibility. The S4 may be heard in the left lateral position. It is usually a duller sound than S1, but when the S4 is loud it may become medium-high pitched. An S4 often is palpable.

Split S1. Splitting of S1 is audible at the apex, but the two components of S1 usually are more discrete at the lower left sternal border. Only a single S1 will be heard at the base. The first component of S1 (M1) does not alter its intensity with respiration, whereas both the intensity and splitting interval of T1 increase during inspiration. The two components of S1 are best heard with the diaphragm. The intensity of a split S1 does not vary when the subject sits or stands, whereas an S4 softens with the upright position.

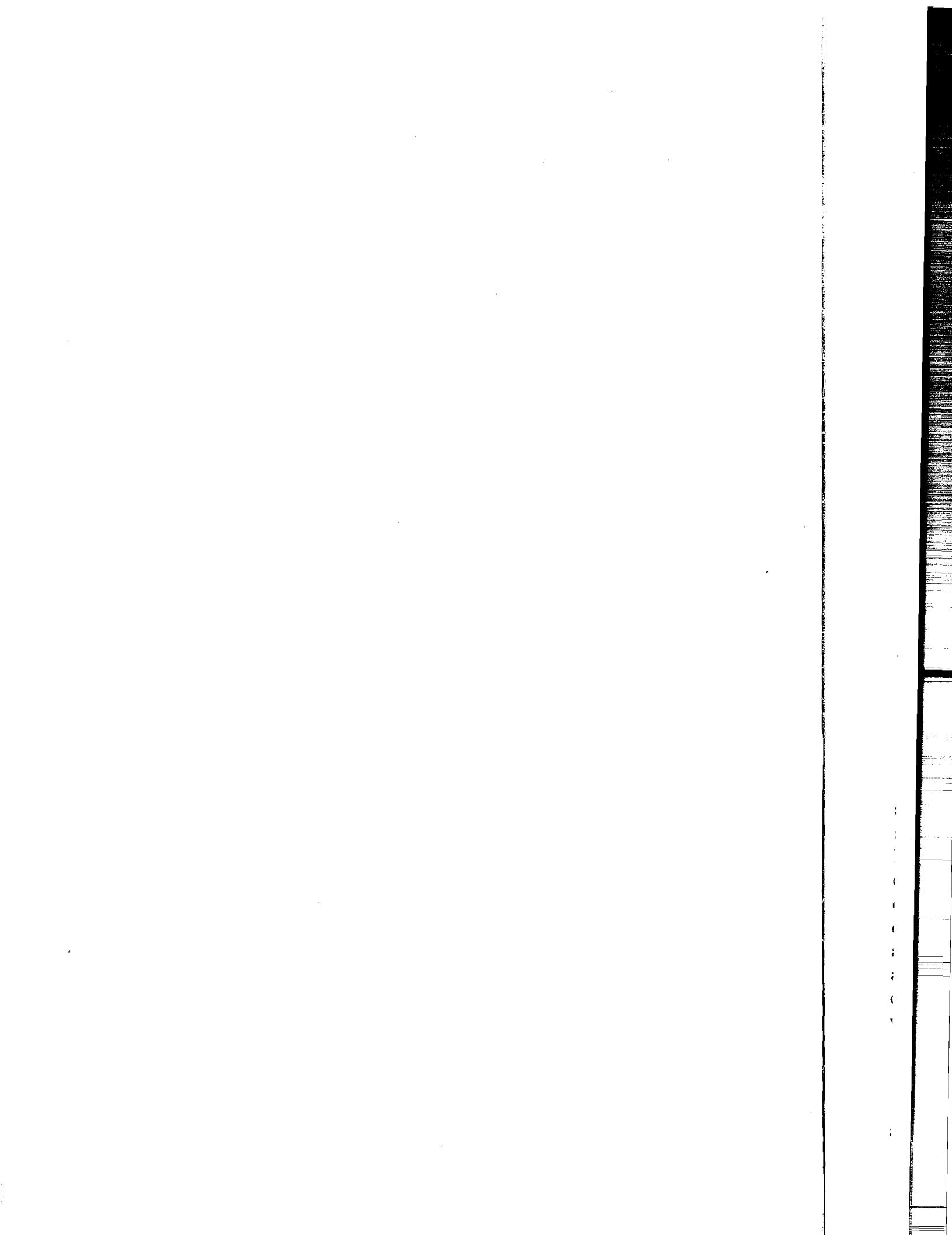
S1-Ejection Click. An S1-ejection click complex is more easily confused with a split S1 than the S4-S1 combination, and these two sounds will infrequently simulate an S4-S1 complex (Table 8-1). Both S1 and ejection clicks are medium- to high-pitched sounds. A pulmonic ejection sound will vary prominently with respiration. Aortic ejection clicks usually are best heard at the apex; the pulmonic click is loudest at the upper sternal edge and is not audible at the apex. Assuming the upright position does not alter the intensity of ejection sounds.

Palpable S4. Careful palpation of the apical impulse in the left decubitus position may be rewarded by detection of an outward presystolic thrust in patients with a palpable S4 (see Chapter 5; Figs. 5-7, 7-3, 7-6). This motion often is quite subtle; commonly, it feels as if there is a shelf or notch on the upstroke of the apex impulse. Constant calls this the "atrial hump." In general, the larger the apical A wave, the higher the LV end-diastolic pressure.

The palpable A wave occurs just before the maximal outward apical excursion that occurs during isovolumic systole. Two fingers should be used for detection of a palpable S4. It is important to feel the apex beat over many cardiac cycles, as the site of maximum impulse may alter slightly with respiration and the low amplitude A wave may not be palpable with each heart beat. When the A wave is very large it is felt as a forceful thrust and occasionally may be even larger than the outward LV impulse. *Practical Point: The finding of a palpable S4 indicates that the associated atrial sound is definitely abnormal.*

However, some pathologic atrial sounds that are definitely audible may not be palpable, particularly in obese, thick-walled, or large-chested patients. Conversely, a palpable S4 occasionally is not accompanied by an audible S4, particularly when the atrial transient is very low frequency.

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Chapter 8

Ejection Sounds

Ejection sounds are high frequency transients that occur in early systole immediately following the first heart sound (S1) (Fig. 8-1). They are also known as ejection clicks; either term is acceptable. Ejection sounds (ES) occur more frequently than is commonly realized. Unless the observer is aware of the possibility of such acoustic events, the ES easily can be assumed to be a part of S1 and will be completely missed. This is not an academic problem; the finding of an ejection click may be the critical clue that a systolic murmur is truly organic. The presence of a definite ES almost always implies underlying cardiovascular disease.

PATHOPHYSIOLOGY OF EJECTION SOUNDS

Most experts agree that two different mechanisms produce an ejection click: (1) the snapping open or doming of a stenotic thickened or malformed *pulmonary* or *aortic valve* (Fig. 8-2A), or (2) a sound transient produced by sudden tensing or reverberation of the proximal *aorta* or *pulmonary artery* at the time of early ejection coincident with the upstroke of pressure rise in the responsible great vessel (vascular or root origin) (Fig. 8-2B). Ejection clicks of valve origin classically are found in congenital aortic and pulmonary artery valve stenosis, and their mechanism is well understood. However, there is controversy as to whether an ES that occurs in the absence of an obvious semilunar valve abnormality is produced by the proximal great vessels themselves or whether the "root" ES is also of valve origin. Some investigators have timed such ejection sounds to coincide with the maximal opening excursion of the pulmonic or aortic valves. Thus, it is possible that in some clinical settings an apparently normal semilunar valve may produce an audible click when its leaflets reach their maximal opening position. In any event, aortic and pulmonary ejection sounds of the nonstenotic variety invariably are associated with either a dilated aorta or pulmonary artery or a systemic or pulmonary vascular tree with an increased systolic pressure and reduced vascular compliance.

SEMITILUNAR VALVE STENOSIS

When commissural fusion of a semilunar valve and leaflet thickening are present, the rapidly rising ventricular pressure in early systole drives the

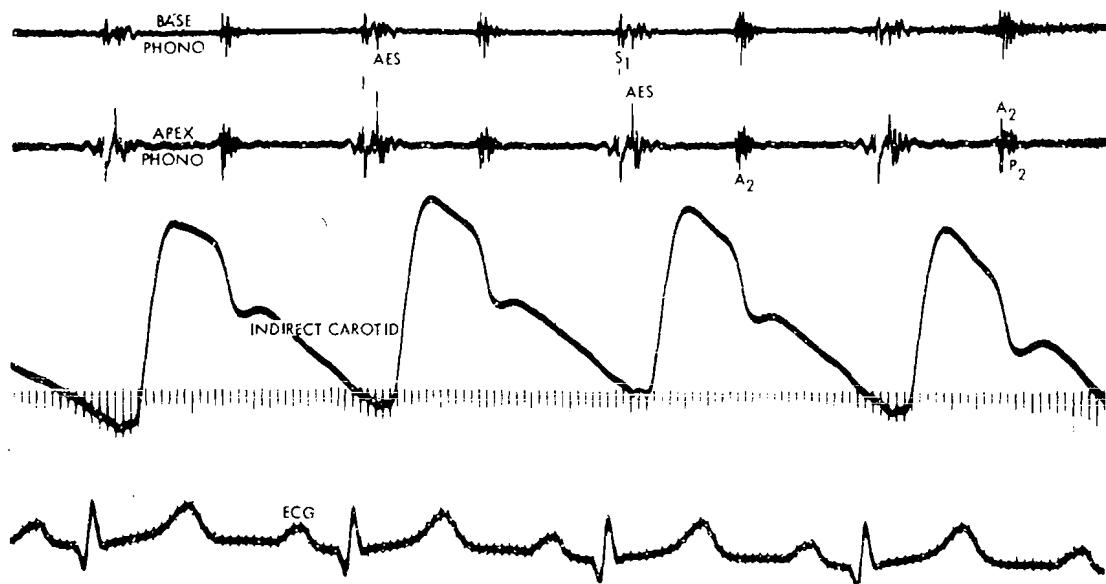
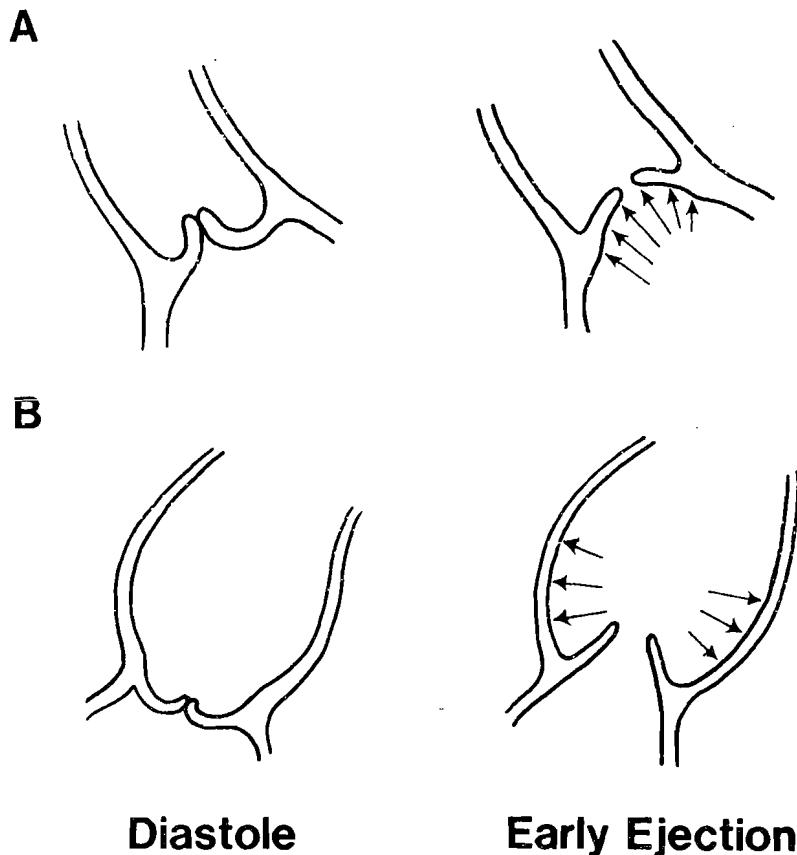


FIG. 8-1. Aortic ejection sound. This phonocardiogram and carotid arterial pulse tracing demonstrates a prominent, discrete aortic ejection sound that is better heard and recorded at the apex than at the base. This is characteristic of aortic ejection sounds or clicks. Note the prominent separation of the ejection sound from S1 by approximately 40 to 50 msec. (From Shaver JA, Griff FW, and Leonard JJ: Ejection sounds of left-sided origin. In *Physiologic Principles of Heart Sounds and Murmurs*. Edited by DF Leon and JA Shaver. American Heart Association Monograph No. 46, 1975.)

abnormal valve leaflets upward. At the precise moment of maximum ascent, the taut valve cusps bulge or dome into their respective great vessel, producing a high frequency sound (Fig. 8-2A). Flow across the stenotic valve occurs only after maximal valve opening has been reached, and the resultant systolic ejection murmur immediately follows the ejection click. The closing movement of the stenotic valve leaflets normally produces a reciprocal high frequency sound, i.e., A2 or P2. When the valve stenosis is extremely severe and the valve cusps are relatively immobile, both the ES and the respective valve component of S2 are diminished or even absent. Extensive calcification, as is frequently found in aortic stenosis, is another major cause of inaudibility of both an ejection click and its S2 counterpart. By contrast, a loud ejection click in the presence of semilunar valve stenosis implies that the leaflets are mobile and remain somewhat flexible.

Aortic Stenosis. An ejection click is almost always present in congenital abnormalities of the aortic valve (Figs. 8-3, 13-1, 13-6). Most often an aortic ES indicates a bicuspid aortic valve. This is probably the most common congenital cardiac abnormality. Often, an isolated ejection click may be heard in the absence of an ejection murmur (Fig. 8-1); echocardiography is mandatory in these situations to assess the possibility of a bicuspid or otherwise thickened aortic valve. In *acquired* aortic valve stenosis, usually of rheumatic or degenerative origin, ejection clicks are much less common, and A2 typically



Diastole

Early Ejection

FIG. 8-2. Origin of ejection sounds. A. Ejection sound or click produced by the opening motion of a thickened, often stenotic aortic or pulmonary valve. B. Ejection sound produced by sudden tensing of the proximal aorta or pulmonary artery during early ejection. This is usually associated with a dilated and/or hypertensive great vessel (see text).

is poorly heard, because the severe anatomic deformity, calcification, and rigid leaflets preclude substantial valve leaflet excursion.

Pulmonic Stenosis. Ejection sounds are common in valvular pulmonic stenosis. As with the aortic valve, the presence of an ES localizes the obstruction to valve level. Infundibular pulmonic stenosis does not produce an ejection click. In very mild or very severe pulmonic stenosis, the pulmonic ejection click may not be heard. *Practical Point: The most characteristic attribute of the pulmonic valve ejection sound is its marked variability with respiration.* In contradistinction to almost all other right-sided acoustic phenomena, which become louder with inspiration, the pulmonic valve ejection click typically softens or disappears with inspiration (Figs. 8-4, 8-7).

The mechanism of an inspiratory decrease in ES amplitude deserves a thorough explanation (Fig. 8-5). Pulmonary artery diastolic pressure is very low in pulmonic stenosis; right ventricular end diastolic pressure is normal or elevated due to decreased right ventricular compliance and right ventricular hypertrophy. With inspiration, augmented venous return to the right heart results in a more forceful right atrial contraction, which in turn produces a

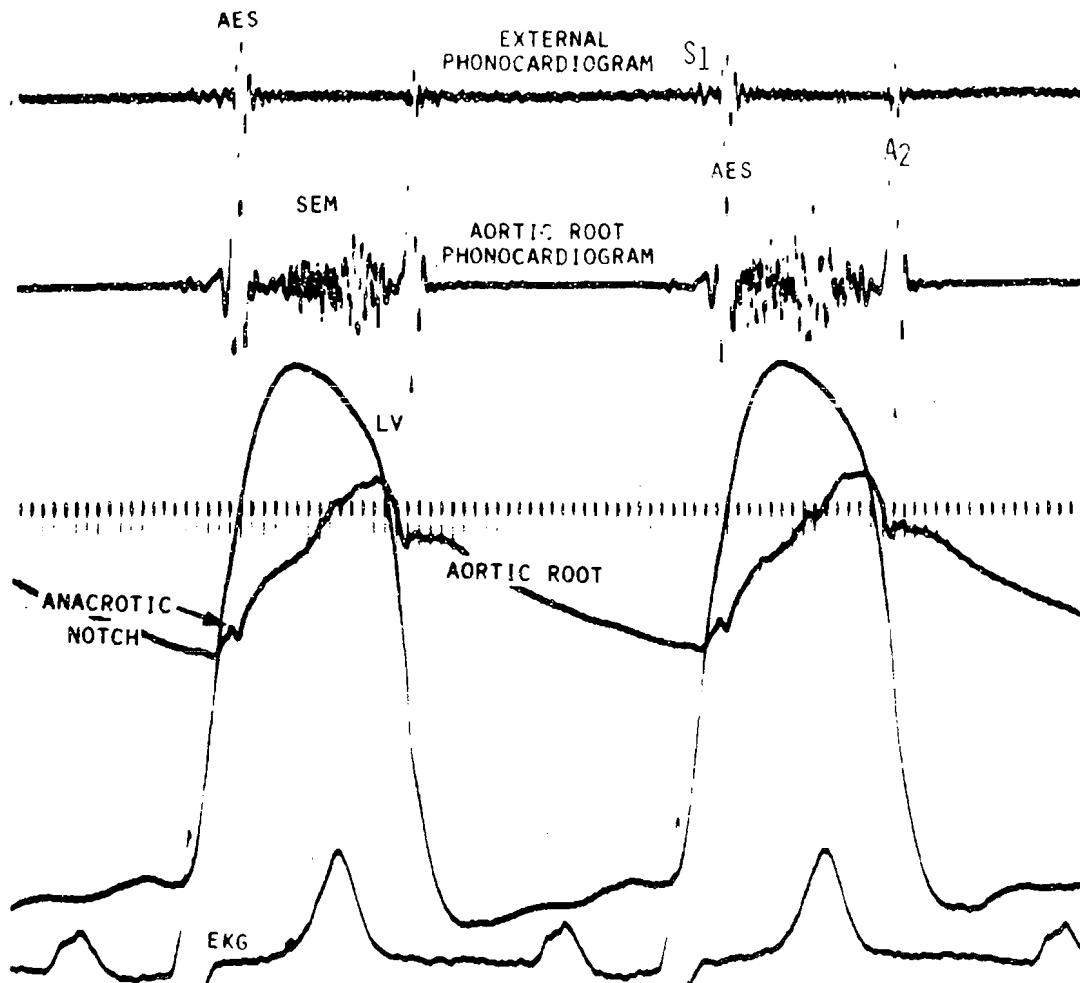


FIG. 8-3. Aortic ejection sound in congenital aortic stenosis. A prominent ejection sound (AES) is recorded on the chest wall as well as directly within the aortic root in this patient with mild congenital aortic stenosis, presumably due to a bicuspid aortic valve. The ejection sound coincides precisely with the maximal doming motion of the abnormal valve, as documented by simultaneous angiography. (From Shaver JA, Griff FW, and Leonard JJ: Ejection sounds of left-sided origin. In *Physiologic Principles of Heart Sounds and Murmurs*. Edited by DF Leon and JA Shaver, American Heart Association Monograph No. 46, 1975.)

rise in RV end-diastolic pressure. RV late diastolic pressure exceeds the simultaneous pulmonary artery diastolic pressure, and the pulmonary valve cusps literally are pushed towards an open position during late diastole. Thus, the actual opening excursion of the pulmonic valve cusps is minimal or even absent, and the opening click is soft or not present.

In severe pulmonic stenosis, the pulmonary valve may actually not open fully until mid or late systole, and the click will be later in timing. In some instances, RV diastolic pressure may exceed pulmonary artery diastolic pressure throughout the respiratory cycle, thus precluding any abrupt doming of the pulmonary valve with ejection. Thus, an ES will not be audible. In very mild pulmonic stenosis, there may not be respiratory alteration in the intensity

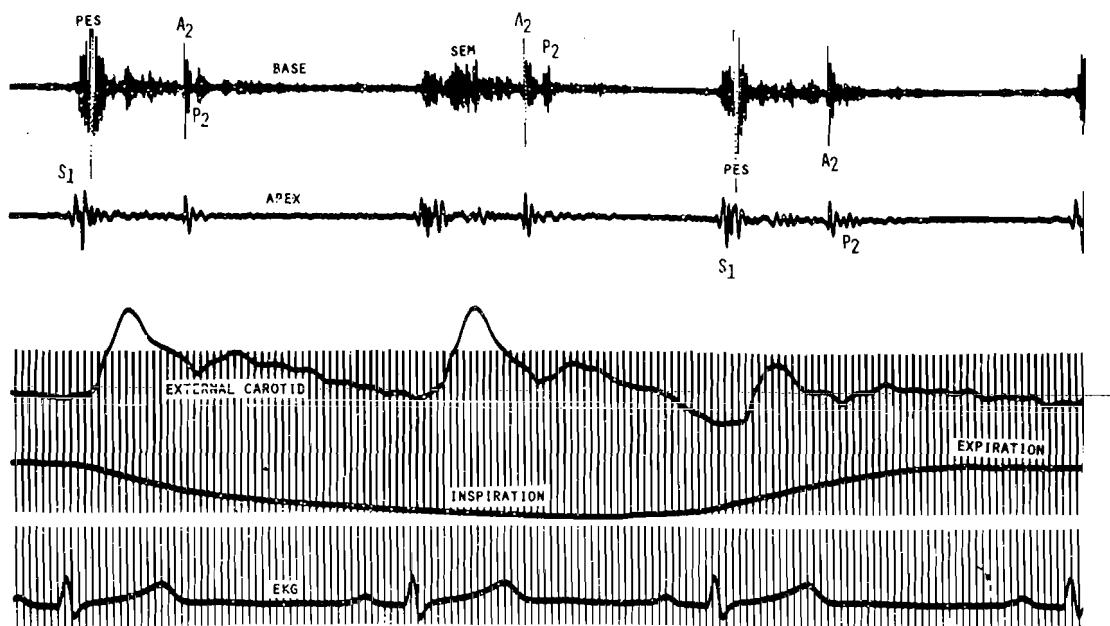


FIG. 8-4. Pulmonic ejection sound in congenital pulmonic stenosis. Note the inspiratory disappearance of the pulmonic ejection sound or click (PES), which is only identifiable during expiration. This patient has moderately severe pulmonic stenosis at valve level. Inspiration results in accentuation of the systolic murmur as well as a disappearance of the PES (see text, Fig. 8-5). (From Martin CE et al: Ejection sounds of right-sided origin. In Physiologic Principles of Heart Sounds and Murmurs. Edited by DF Leon and JA Shaver. American Heart Association Monograph No. 46, 1975.)

of the click. In valvular pulmonic stenosis, P2 may be very soft or inaudible, even with a well-preserved ejection click, because of the very low pulmonary artery pressures. In addition, P2 usually is delayed, which results in further difficulty in hearing this sound.

NONVALVULAR EJECTION SOUNDS

An ES with a normal semilunar valve typically is associated with dilatation of the aorta or pulmonary artery, which occurs in conditions of an increased stroke volume and/or increased force of ejection. Some have suggested that dilatation of either great vessel may predispose an individual to an audible ES, presumably because of alterations in the compliance characteristics of the aorta or pulmonary artery (Fig. 8-2B).

Aortic Root Ejection Sounds. These are believed by some to be an accentuation of the normal ejection component of S1, which occurs at the onset of left ventricular ejection into the aorta. Such sounds occasionally are found in subjects with aortic arteriosclerosis, aortic aneurysm, aortic regurgitation, or systemic hypertension. Very occasionally, high output states (anemia, exercise, thyrotoxicosis) will be associated with an exaggerated aortic ejection sound (Fig. 7-2C).

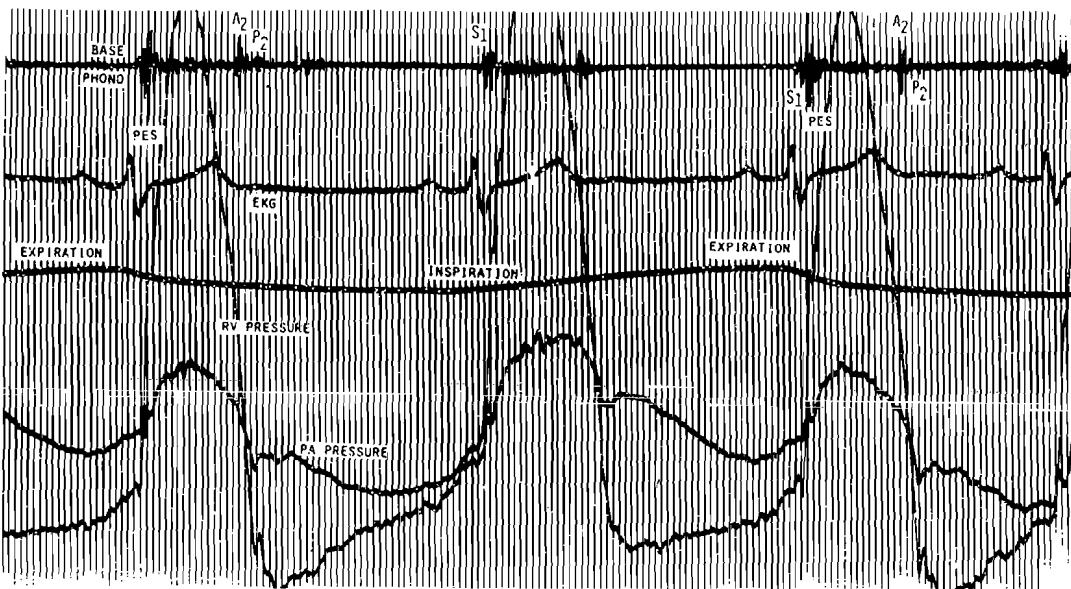


FIG. 8-5. Mechanism of the pulmonic ejection sound in pulmonic stenosis. During inspiration right ventricular (RV) end-diastolic pressure exceeds pulmonary artery diastolic pressure, producing an upward opening movement of the pulmonic valve cusps prior to actual ejection. Thus, the valve already assumes a partially "domed" position and undergoes less upward excursion during early systole; the ejection sound attenuates or completely disappears. These intracardiac pressures are from the same patient whose phonocardiogram is shown in Figure 8-4. (From Martin CE et al: Ejection sounds of right-sided origin. In *Physiologic Principles of Heart Sounds and Murmurs*. Edited by DF Leon and JA Shaver. American Heart Association Monograph No. 46, 1975.)

Pulmonary Ejection Sounds. They may occur in any condition resulting in pulmonary hypertension (Fig. 8-6). In such situations, respiratory variation of the pulmonic ES is not common. Early ejection of blood into a tense, noncompliant pulmonary artery is thought to be the cause of the ES, although echocardiography has shown that some of these clicks time precisely with the maximal opening excursion of the pulmonic valve leaflets, suggesting a valve origin for the pulmonary vascular ejection click. The higher the pulmonary artery diastolic pressure, the later the ejection sound; this is a result of an increased right ventricular isovolumic contraction time and "delayed" opening of the pulmonic valve.

Pulmonary ejection clicks also are heard in idiopathic dilatation of the pulmonary artery where the sound is produced in the dilated pulmonary artery itself. Inspiratory diminution is common in these cases, perhaps due to an increase in pulmonary artery distensibility during inspiration. Atrial septal defects typically do not have associated pulmonic ejection sounds unless there is coexisting pulmonary hypertension. The appearance of a pulmonic ejection click after closure of a high flow atrial septal defect has been noted and may relate to chronic changes in pulmonary artery compliance.

CLUES TO AUSCULTATION OF EJECTION SOUNDS

Quality and Timing. Both aortic and pulmonic ejection clicks typically are high frequency, sharp, discrete sounds that are at least equal in intensity to S1 (Figs. 8-1, 8-3, 8-4, 8-5; 13-6). They are best heard with the diaphragm of the stethoscope. The later the ejection sound, the more audible it is. Ejection sounds that are distinctly separate acoustically from S1 follow the first heart sound by at least 0.05 seconds. The opening clicks of stenotic valves occur later in systole than those of root origin and are more easily heard. An early

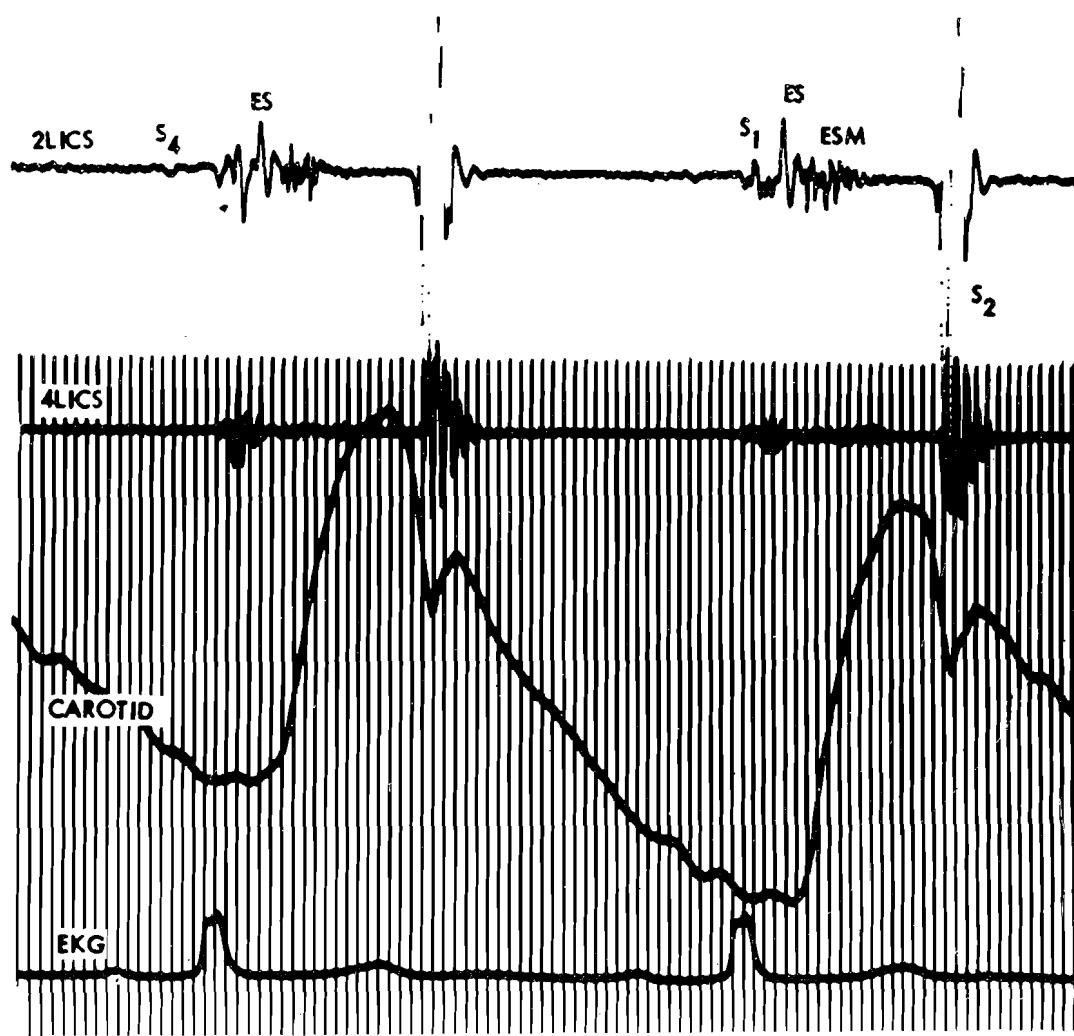


FIG. 8-6. Pulmonary ejection sound in pulmonary hypertension. This phonocardiogram was taken from a patient with pulmonary hypertension due to an Eisenmenger ventricular septal defect. S₂ is single and accentuated. In this situation, the pulmonic ejection click is believed to originate from the dilated and tense pulmonary artery and not from the pulmonary valve itself. (See Fig. 8-2B) (From Reddy PS, Shaver JA, and Leonard JJ: Cardiac systolic murmurs: pathophysiology and differential diagnosis. Prog Cardiovasc Dis 14:1, 1971.)

ES may merge with S1 or, more likely, is thought by the examiner to be part of the S1 complex (split S1) (Fig. 6-4). In pulmonic stenosis, the more severe the valve obstruction, the earlier the click; in these cases, the ES is easily confused with S1.

Location. Aortic ejection sounds are audible at the cardiac base and the aortic area, and, in particular, are well heard at the apex. In many individuals with an aortic ejection click, the sound is heard *only* at the apex. This is particularly true in the elderly or in patients with chronic lung disease. The ES of valvular aortic stenosis typically is loudest at the apex, whereas the ejection sound of aortic root origin is better heard at the base and radiates poorly to the apex. The systolic murmur of both aortic and pulmonary stenosis begins with the click, and this may obscure separate identification of the click at the base.

Pulmonic ejection sounds are best heard at the second and third left interspace and are poorly heard or inaudible at the apex (Fig. 8-7). The pulmonic click of pulmonary hypertension also may be heard lower down the sternum. In patients with a huge right ventricle, the pulmonic click may be well heard in the midprecordium. Respiratory variation, typical of valvular pulmonic stenosis, must be carefully sought; in more severe cases of pulmonic stenosis, the click occurs close to S1 and may not be easily distinguished from it. Whenever S1 appears to vary in intensity during the respiratory cycle, the possibility of a pulmonic click should be considered. A pulmonic click at the pulmonic area may easily be mistaken for S1, which is usually quite soft at the left second or third interspace (Fig. 8-6). In patients suspected of having pulmonic stenosis, listen with the subject in the upright position; the resultant decrease in right ventricular venous return often results in the click becoming more audible both in inspiration and expiration.

Pseudo-ejection Sounds. In atrial septal defects and Ebstein's anomaly, augmented (and delayed) tricuspid closure sound (T1) can simulate a pulmonic ejection sound. In hypertrophic cardiomyopathy, early 'explosive' ejection sounds have been described (Fig. 14-4). An early systolic sound has been reported in membranous ventricular septal defect with a septal aneurysm.

DIFFERENTIAL DIAGNOSIS

Several possibilities may account for two discrete sounds heard in and about S1. There are an S4-S1 combination; a split S1; and an S1-ejection click complex (Fig. 6-4). Obviously in some individuals, two or even three of these sounds may be present, but the human ear usually cannot discriminate well enough to hear three consecutive sounds in rapid succession. In addition, a loud heart sound of any etiology acoustically may mask other sounds. Loud systolic murmurs may further contribute to the difficulty of identifying an ejection click by enveloping the discrete sound.

Table 8-1 lists differentiating features of the various diagnostic possibilities to be considered when two sounds are heard in rapid succession in the beginning of systole. In severe mitral valve prolapse, the abnormal systolic motion of the mitral valve leaflet(s) may occur early in systole, resulting in "holosystolic" prolapse. Usually, this is in the setting of mitral regurgitation. In such cases, the nonejection click of MVP may move close to S1 and simulate a true *ejection* sound. Rarely, an early systolic click can occur in mitral valve prolapse (MVP) in the absence of the murmur of mitral regurgitation; obviously, this will be difficult or impossible to distinguish from an aortic or pulmonic ES without an echocardiogram.

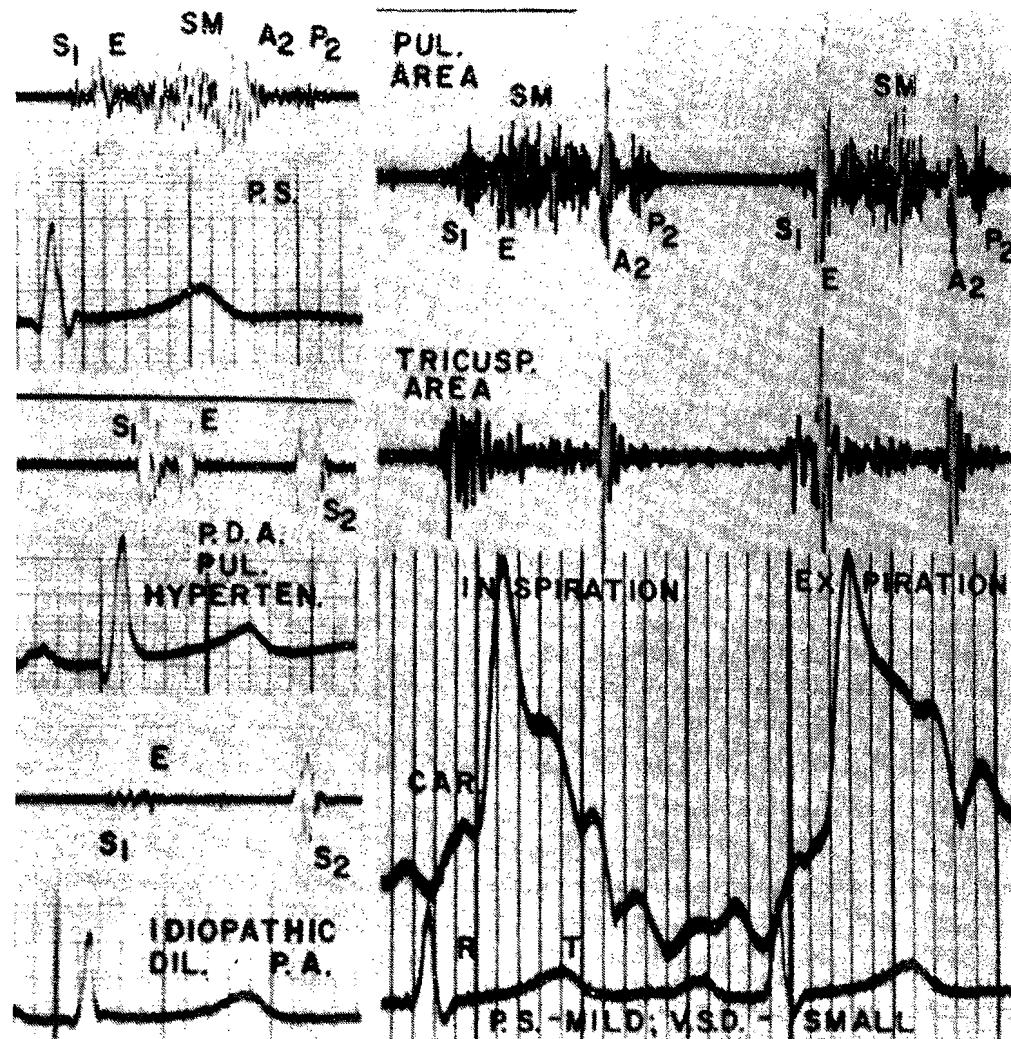


FIG. 8-7. Examples of pulmonary ejection sounds. This composite represents 4 different patients: Upper left: valvar pulmonic stenosis. Note the long systolic ejection murmur and delayed P2. Middle left: pulmonary hypertensive patent ductus arteriosus. Lower left: idiopathic dilatation of the pulmonary artery. The ejection sound is produced within the dilated proximal pulmonary artery. Right: mild pulmonic stenosis with ventricular septal defect. Note the attenuation of the ejection sound with inspiration. E = ejection sound, SM = systolic murmur. (From Harvey WP: Heart sounds and murmurs. Circulation 30:263, 1964.)

TABLE 8-1 *Differential Diagnosis of Systolic Ejection Sounds*

Audible Sound Complex	Useful Differentiating Features
Aortic or pulmonic sound or click	<ul style="list-style-type: none"> • Aortic: usually best heard at apex • Pulmonic: inspiratory decrease common • Evidence of associated aortic/pulmonary valve or root abnormality • Firm pressure with stethoscope diaphragm increases audibility
S4-S1 complex	<ul style="list-style-type: none"> • S4 audibility enhanced with use of light pressure on stethoscope bell • S4 attenuates with firm pressure or use of stethoscope diaphragm • S4 not heard at cardiac base • Often associated evidence of LV hypertrophy or dilatation • S4 waxes and wanes with changes in venous return • RV S4: will increase with inspiration; associated evidence for pulmonary hypertension or RV enlargement
Split S1	<ul style="list-style-type: none"> • Poor radiation of both components (M1 and T1) away from apex area • Second component (T1) usually not snappy or loud • Second component (T1) not audible at cardiac base • Both components better heard with firm pressure on stethoscope diaphragm
Early click in mitral prolapse	<ul style="list-style-type: none"> • Variable behavior of click timing with respect to body position, maneuvers • Associated murmur of late systolic mitral regurgitation may be present, occasionally a holosystolic murmur

SPECIFIC CONDITIONS ASSOCIATED WITH EJECTION SOUNDS (See Table 8-2)

Aortic Stenosis. The presence of an ejection click strongly suggests the presence of a congenitally bicuspid aortic valve and, in general, excludes supra- or subvalvular stenosis or hypertrophic cardiomyopathy. Although an audible click indicates a valve with a significant degree of leaflet mobility, the presence or absence of an ES has no correlation with severity in aortic stenosis. The intensity of A2 usually parallels that of the aortic ejection click (Figs. 8-3; 13-5).

Pulmonic Stenosis. Most patients with valvular pulmonic stenosis maintain a well-preserved pulmonary ES (Figs. 8-4, 8-5, 8-7). In very mild or very severe pulmonic stenosis, the click may be inaudible or, if present, inspiratory variation may be absent. The presence of a pulmonic click in a

TABLE 8-2 *Conditions Associated with Ejection Sound or Click*

<i>Aortic</i>	Congenital valvular aortic stenosis Bicuspid aortic valve Aortic regurgitation Aortic aneurysm Aortic root dilatation Systemic hypertension Severe tetralogy of Fallot
<i>Pulmonic</i>	Pulmonary valve stenosis Idiopathic dilatation of the pulmonary artery Atrial septal defect Chronic pulmonary hypertension Tetralogy of Fallot (with pulmonic valve stenosis)
<i>Pseudo-ejection sound</i>	Prominent splitting of S1 Increased T1 (Ebstein's Anomaly; ASD) Hypertrophic cardiomyopathy Early non-ejection click of holosystolic mitral valve prolapse High pitched S4 (S1 confused for ES)

young patient with a prominent pulmonic outflow murmur and wide splitting of S2 helps identify pulmonic stenosis; patients with an atrial septal defect usually do not have prominent ejection sounds.

Aortic Root Dilatation. Ejection sounds may be heard whenever the central aorta is dilated, but the ES has no particular clinical significance in this setting. These sounds may also be detected in hypertensive patients. In the presence of an aortic ES, it may be difficult to exclude an associated aortic valve abnormality. Aortic root ES do not radiate well from the aortic area, whereas the ES of aortic valve disease is often best heard at the cardiac apex.

Pulmonary Hypertension. The presence of a pulmonic ejection click is common with chronic elevation of pulmonary artery pressure. A pulmonic ES may be a useful ancillary diagnostic sign of pulmonary hypertension, along with an increased P2 and a right ventricular lift (Fig. 8-6). The pulmonary artery is typically dilated in chronic pulmonary hypertension.

Coarctation of Aorta. An aortic ejection click heard in a patient with aortic coarctation implies a coexisting, congenitally bicuspid aortic valve, a common associated abnormality (Fig. 8-1). The ES may also result from aortic dilatation due to sustained upper body hypertension.

ROLE OF NONINVASIVE TECHNIQUES

Although this monograph minimizes discussion of pulse wave and sound recordings, the problem of accurately identifying ejection sounds deserves

further comment. In many situations, it is simply not clear if a certain sound complex around S1 represents an ejection click or an unimportant splitting of S1. Simultaneous phono- and echocardiography may be invaluable in timing the sounds in question so that precise identification of the ES can be made to clarify the clinical diagnosis. Details of such investigations go beyond the scope of this text; the interested reader is referred to the monographs by Tavel, Constant, Leatham, and Ravin.

Chapter 9

Heart Murmurs

Physicians must be able to assess heart murmurs accurately in order to properly evaluate patients with congenital or valvular heart disease. In the general population, systolic murmurs are widespread but reflect abnormal cardiac structure in only a small percentage of persons. The presence of a heart murmur raises questions regarding prophylaxis for endocarditis, restriction from athletics, eligibility for life insurance or employment, risk of pregnancy to the mother or fetus, and the safety of noncardiac surgery. In the practice of medicine, physicians confront these issues daily and must be able to distinguish heart murmurs that have no intrinsic cardiac abnormalities from those that indicate organic disease. If the practitioner has acquired fundamental skills in cardiac auscultation, capable physician should not need to refer all suspicious murmurs to a specialist for evaluation.

SIGNIFICANCE OF A MURMUR

A pragmatic approach to the evaluation of heart murmurs should be taken. When someone is found to have a murmur, several issues require resolution. The first question to ask is whether the murmur is organic or pathologic, or does it represent a normal variant. A "yes" answer indicates the presence of structural cardiovascular disease. Based on the information derived from the cardiac physical examination, one can then determine the likeliest hemodynamic cause of the murmur. Once the anatomic origin of an organic murmur is characterized, the possible etiologies of the heart disease should be investigated, and assessment of the severity of the underlying condition can be made.

Usually, a capable clinician can resolve these questions after a careful physical examination. While ancillary diagnostic testing (e.g., EKG, chest roentgenogram, M-mode or two-dimensional echocardiogram, cardiac catheterization) is often important, typically these studies provide additional data about the severity of a problem that already has been identified and roughly quantitated by the physical examination.

When a heart murmur is felt to be "innocent" (see Chapter 10), the diagnosis of *non-disease* should be made firmly and without equivocation. *Practical Point: Individuals with heart murmurs with no intrinsic cardiac abnormality are far more common than those with organic cardiovascular*

disease. Thus, "ruling out" significant heart disease is an important aspect of everyday clinical practice.

PHYSIOLOGY

Turbulence is thought to be the primary factor in the genesis of heart murmurs, although there are many other theories about the production of cardiovascular sound, including vortex shedding, collision and cavitation phenomena, and eddy production. Turbulent blood flow produces both physiologic murmurs and organic murmurs. Turbulence is directly related to intracardiac sound energy that reaches an audible threshold when it is of sufficient amplitude. An important determinant of turbulence is the velocity of blood flow; increases in velocity result in marked increases in turbulence, often producing audible sound. An obstruction to blood flow, like stenosis, produces abnormal turbulence, which is related both to the velocity of blood flow and to alterations in accelerating forces; turbulence increases as the gradient across the valve becomes larger. Because acceleration of blood flow is maximal during the early ejection phase of systole, turbulence and cardiac sound are usually maximal in early systole.

For any given size of valve orifice, an increase in blood flow results in increased flow velocity; conversely, for any given flow rate, a smaller or narrowing orifice results in a greater velocity of blood flow and a louder murmur. Thus, changes in cardiac output will affect the intensity of ejection-related murmurs, and a narrowing orifice (e.g., the closing mitral valve during diastole) may increase the loudness of a murmur related to flow across an obstructed orifice. Turbulence is enhanced when blood moves from a smaller chamber to a larger one. Low blood viscosity (e.g., anemia) enhances turbulence.

NORMAL EJECTION DYNAMICS

During the first part of systole, there is a small pressure gradient between each ventricle and its respective great vessel (Fig. 9-1). These normal "impulse gradients" may result in the production of cardiac sound in many normal subjects at rest (innocent murmur). With exercise and a larger cardiac output, the normal impulse gradient increases along with an acceleration of early systolic flow and enhancement of murmur intensity.

Sound Frequency. In general, turbulent flow produces random sound that has many frequencies and results in mixed frequency murmurs. High flow rates or large gradients produce more high pitched sound; low flow rates and/or small gradients result in low pitched sound.

Transmission. Transmission or radiation of cardiac sound results from vibration of cardiac structures and blood vessels set up by turbulence or eddy formation. In part, the intensity of a given murmur is related to its transmission characteristics. High frequency sound does not transmit well downstream but may be prominent closer to the site of maximal murmur production. Low frequency vibrations are better heard downstream and thus transmit more widely.

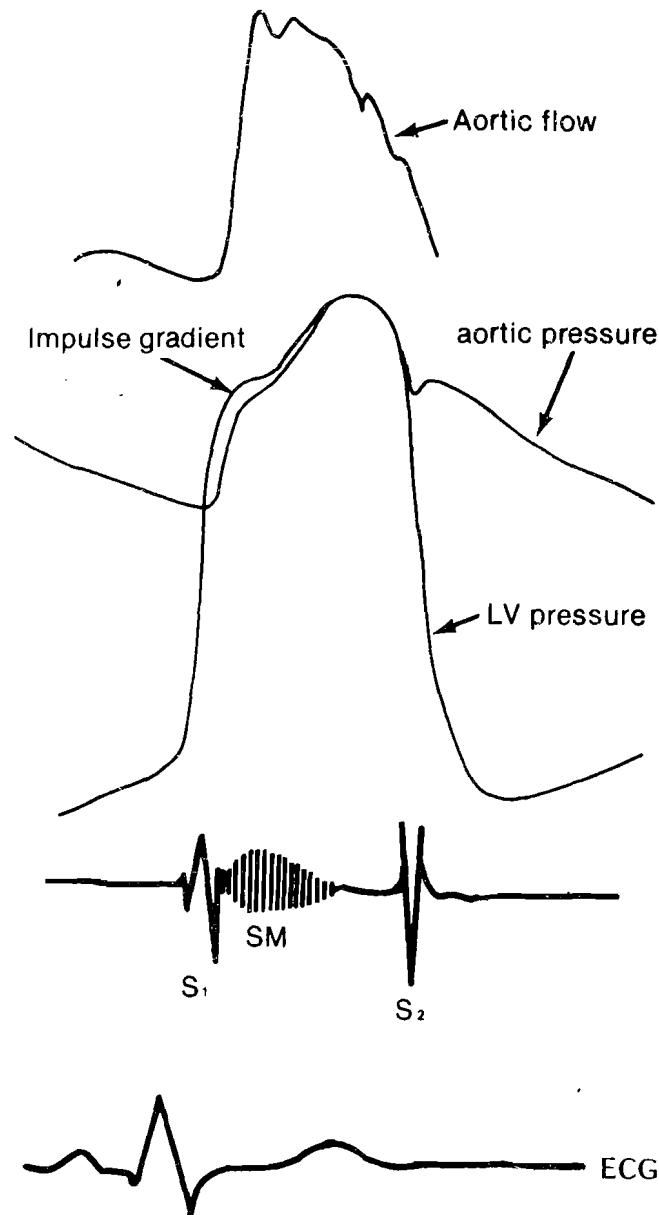


FIG. 9-1. Pressure flow relationships of the systolic ejection or flow murmur. During early systole peak blood flow and velocity are maximal, producing sufficient turbulence for the production of audible sound. A typical systolic ejection murmur ends within the first 2/3 of systole. Note the small "impulse gradient" in early systole. (From Abrams J: Auscultation of heart murmurs. Prim Cardiol, 1981.)

GENERAL PRINCIPLES OF CARDIAC AUSCULTATION

Use of the Stethoscope. The acoustic properties of cardiac sound and the stethoscope are reviewed in Chapter 1. The bell of the stethoscope is ideal for low frequency murmurs (25 to 125 cps) (Fig. 1-2); light skin contact is desirable when the bell is used in order to maximize detection of low pitched sound. The diaphragm is best for high frequency or mixed frequency murmurs; increasing pressure on the rigid diaphragm will attenuate lower pitched sounds and accentuate the higher frequencies. One should routinely alter the stethoscope pressure during auscultation in order to assess optimally the frequency characteristics of heart sounds and murmurs.

Grading Murmurs. The 1 through 6 grading protocol was created to provide a systematic and consistent method of evaluating heart murmurs. Although individual observers may vary in describing the loudness of a particular murmur, careful application of the grading system should not result in inter-observer differences of two or more grades, and consistent and reliable descriptions of murmur intensity during long-term follow-up of heart murmurs by physicians are quite feasible.

Grade 1: The faintest murmur that can be heard under optimal conditions (quiet room, relaxed patient and physician). It may take several seconds for a grade 1 murmur to be clearly heard by the observer. Skeptics have labeled this a "murmur that only cardiologists hear."

Grade 2: A soft but readily audible murmur.

Grade 3: A prominent murmur. Murmurs of grade 3 intensity should always stimulate a careful search for cardiac disease.

Grade 4: A very loud murmur that is palpable (thrill).

Grade 5: Louder still (thrill).

Grade 6: A murmur audible with the stethoscope held off the chest wall (thrill).

The intensity of a murmur is graded on a 1 to 6 basis (e.g., a 3/6 ejection murmur, a 2/6 decrescendo diastolic murmur). It is acceptable to assign two grades to a murmur (e.g., 1 to 2/6, 2 to 3/6) to designate an intermediate degree of loudness. Most innocent or functional murmurs will be grade 1 to 2/6; some will be grade 3/6. Clinically, there is no significant difference between a grade 4/6 and grade 6/6 murmur; both are extremely loud. Grades of 5/6 or 6/6 murmurs are rare.

Valve Areas. Customarily, specific locations on the chest have been designated as relating to a particular cardiac valve. This practice stems from observations regarding the site where a specific murmur (e.g., aortic stenosis) usually is best heard. Thus, the second right interspace is known as the "aortic area"; the second to third left interspace is the "pulmonary area"; the lower

left sternal border is the "tricuspid area"; and the apex is the "mitral area." However, there are many exceptions to this oversimplified approach. Radiation patterns of various organic murmurs often are discordant with the precise anatomic location of the valves that produce the murmur making the specific precordial "area" less reliable in determining the site of origin of a murmur.

For these reasons, Shah and Luisada have suggested substitution of a more anatomic and physiologic schema to classify precordial areas of auscultation (Fig. 9-2). This taxonomy relates more to the cardiac chambers than to the heart valves and is influenced by specific cardiac sounds and murmurs that are best detected by intracardiac phonocardiography. These auscultatory sites are called the left ventricular area, the right ventricular

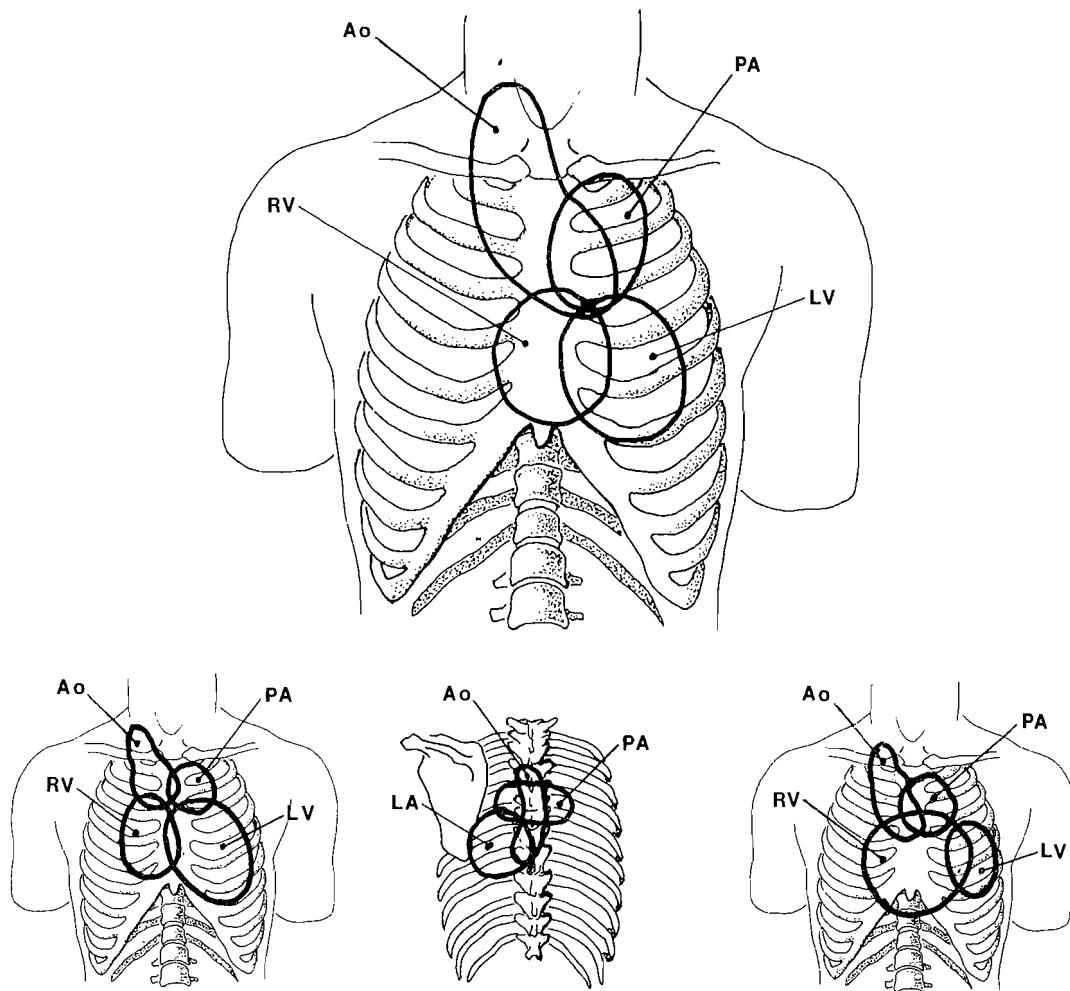


FIG. 9-2. Radiation patterns and precordial areas of specific valve abnormalities. Upper: classic precordial sites for auscultation. Lower left: projection of auscultatory areas when the left ventricle is enlarged. Lower middle: radiation sites in the back. Lower right: projection of auscultatory areas when the right ventricle is enlarged. AO = aorta, PA = pulmonary artery, RV = right ventricle, LV = left ventricle, LA = left atrium. (Modified from Shah, Slodki, and Luisada).

area, the left and right atrial areas, and the aortic and pulmonary areas. The actual locations correlating with these areas are listed in Table 9-1.

Diagramming Heart Sounds and Murmurs. Some authorities have urged the routine use of charts or diagrams to document a patient's precise

TABLE 9-1 *Locations of Auscultatory Sites*

Prior Area	Designation	Location	Murmurs Heard Best	Sounds Heard
Left ventricular	"Mitral area"	At apex impulse: extends to 3-5 LICS, 2 cm medially and laterally to left anterior axillary line. Isolated LVE: extends medially; isolated RVE: may be displaced to left axilla	Mitral stenosis Mitral regurgitation Aortic stenosis Aortic insufficiency IHSS Functional middiastolic rumble	LV S ₃ LV S ₄ A ₂
Right ventricular	"Tricuspid area"	Lower sternum and 3-5 LICS 2 cm to left and right isolated RVE: can extend laterally and occupy the apex	Tricuspid stenosis Tricuspid regurgitation Pulmonary regurgitation Ventricular septal defect	RV S ₃ RV S ₄
Left atrial		Left posterior thorax between axillary line and spine at level of scapular tip	Mitral regurgitation	
Right atrial		Lower sternum and 4-5 RICS, 2 cm to right of sternum	Tricuspid regurgitation	
Aortic area	"Erbs point" (3rd left interspace)	3 LICS near sternal edge across manubrium to 1-3 RICS, may include 2 LICS, suprasternal notch, right sternoclavicular joint	Aortic stenosis Aortic insufficiency Aortic flow murmurs	A ₂ Aortic ejection click
Pulmonary area		1-3 LICS adjacent to sternum, medial left intraclavicular area; posterior thorax: T _{4s} , 2-3 cm to either side of spine	Pulmonary stenosis Pulmonary regurgitation Pulmonary flow murmurs PDA murmur	Pulmonary ejection click P ₂
Descending thoracic aorta		Posterior thorax: T ₂ -T ₁₆ , 2-3 cm to either side of the spine	Coarctation of the aorta Aortic aneurysms Aortic stenosis	

(From Abrams, J: Prim Cardiol, 1981)

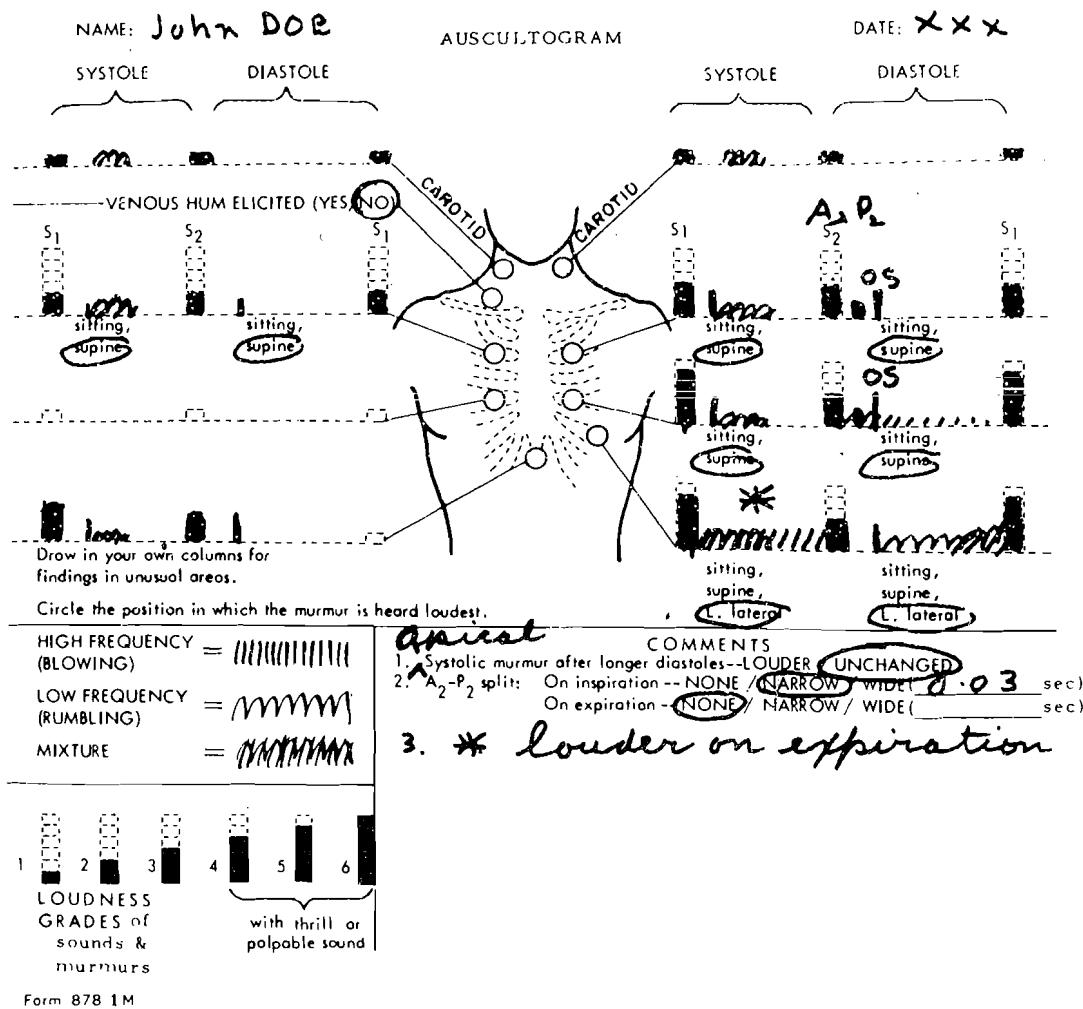


FIG. 9-3. Diagramming heart sounds and murmurs. This auscultogram is an example of a method for illustration of a patient's cardiac findings suitable for teaching purposes as well as for record keeping. (From Constant J: Bedside Cardiology, 2nd ed. Boston, Little, Brown and Co, 1976.)

cardiac physical findings. These incorporate information regarding such characteristics as amplitude, pitch, timing, and the shape of murmurs as well as an assessment of normal and abnormal heart sounds. Figure 9-3 demonstrates such a chart designed by Jules Constant. This is a valuable tool to help obtain a precise and thorough analysis of the cardiac physical findings.

CLASSIFICATION OF HEART MURMURS

The general classification of murmurs proposed by Aubrey Leatham in 1958 has been widely adopted (Table 9-2). Leatham divided systolic murmurs into two major types, *midsystolic ejection* and *pansystolic* or *regurgitant*. *Diastolic* and *continuous* murmurs are the other two major types of heart murmurs. Ejection murmurs not caused by valvular obstruction or narrowing are known as flow murmurs, functional murmurs, or innocent murmurs.

TABLE 9-2 *Classification of Heart Murmurs***SYSTOLIC**

- Ejection murmurs
 - Flow or functional murmurs
 - Innocent murmur
 - Physiologic murmur (related to increased cardiac output), e.g., anemia, thyrotoxicosis, postexercise
 - Pathologic or significant murmurs
 - Abnormal but nonstenotic aortic or pulmonary valve
 - Aortic or pulmonary valve stenosis
 - Dilatation of aorta or pulmonary artery
 - Left or right ventricular outflow tract obstruction (nonvalvular)
- Regurgitant murmurs
 - Mitral regurgitation
 - Tricuspid regurgitation
 - Ventricular septal defect

DIASTOLIC

- Semilunar valve incompetence
 - Aortic or pulmonary regurgitation
- Ventricular filling murmurs
 - Mitral or tricuspid stenosis
 - Augmented AV valve flow (e.g., mitral regurgitation, VSD, ASD)
 - Presystolic murmur due to atrial contraction (e.g., mitral or tricuspid stenosis)

CONTINUOUS

- Communication between high pressure chamber or artery with low pressure chamber or vein (e.g., patent ductus arteriosus, coronary AV fistula, sinus of Valsalva to right atrial communication)

When defining or classifying a murmur, it is desirable to use a physiologic descriptor as well as an indication of the timing of the murmur (e.g., late systolic regurgitant murmur, or early systolic ejection murmur).

Some classifications of murmurs describe only the timing (early, mid, late) of systolic and diastolic murmurs, whereas others refer to the inflow or outflow tract origin of the murmur. The classification and terminology recommended in this book reflects a useful amalgam of terms and relies heavily on the work of Leatham. The specific types of murmurs listed below are discussed more extensively in the chapters dealing with the various cardiac lesions (Table 9-2). Figure 9-4 is a graphic representation of the intracardiac pressure and sound relationships of most types of cardiac murmur.

SYSTOLIC MURMURS

Ejection Murmurs

The most common murmur heard in everyday practice is the ejection or flow murmur produced by rapid ejection of blood during the first part of systole. Peak acceleration of blood flow occurs in early systole just after aortic valve opening, and the impulse gradient and peak systolic pressure are max-

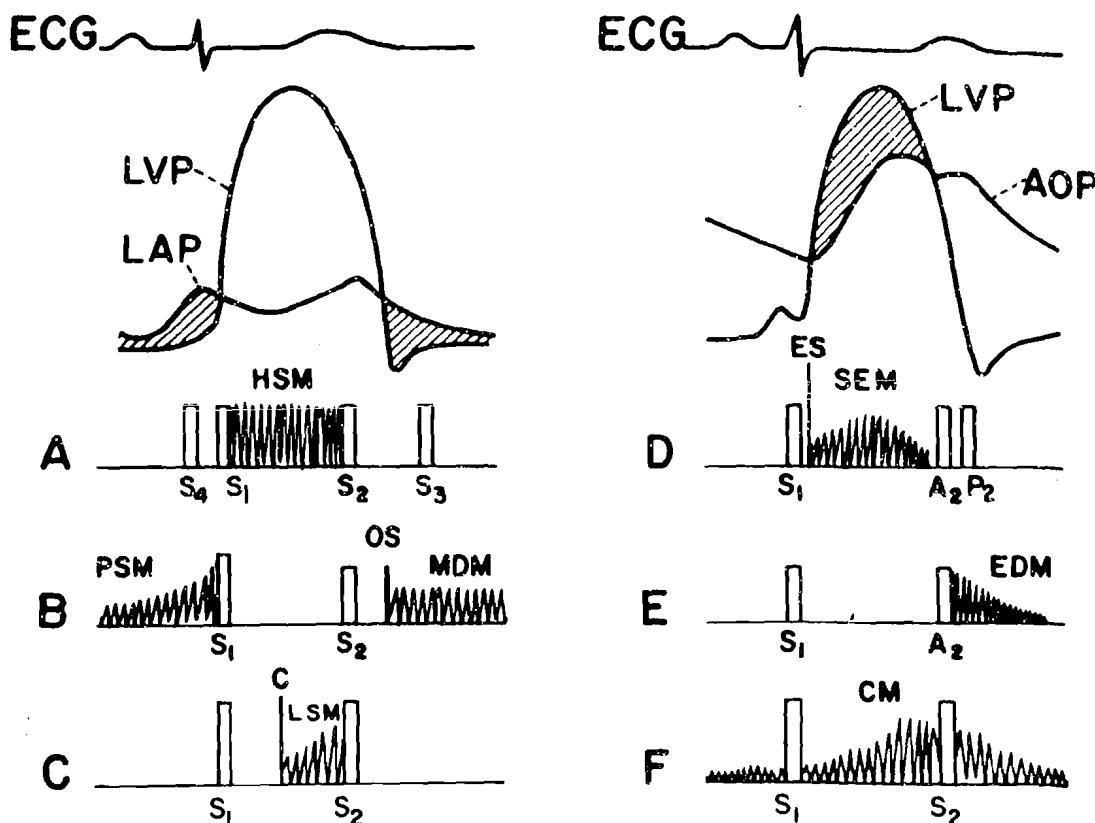


FIG. 9-4. Intracardiac pressures and heart murmurs in the major cardiac valve abnormalities. See text for discussion of specific murmurs. LVP = left ventricular pressure, LAP = left atrial pressure, AOP = aortic pressure, HSM = holosystolic murmur, PSM = presystolic murmur, OS = opening snap, MDM = mid-diastolic murmur, C = mid-systolic click, LSM = late systolic murmur, ES = ejection sound, SEM = systolic ejection murmur, EDM = early diastolic murmur, CM = continuous murmur. (From Crawford MH and O'Rourke RA: A systematic approach to the bedside differentiation of cardiac murmurs and abnormal sounds. *Curr Prob Cardiol* 1:1, 1979.)

imal during the first half of ejection. During the last third of systole very little forward flow occurs across the semilunar valves (Fig. 9-1). The rapid velocity of blood produces turbulence, which in turn results in audible sound. *Early systolic murmurs are common, even in the presence of normal valves and a basal cardiac output.* Irregular, thickened, or stenotic semilunar valves leaflets result in greater turbulence and increased murmur intensity.

The typical systolic ejection murmur begins after S1 at the completion of isovolumic contraction. (Figs. 9-1, 9-4D, 9-5A, 9-6). Although phonocardiograms may document a small gap between S1 and the onset of the murmur, the ear usually cannot discriminate this delay, and the murmur appears to begin coincident with S1. The classic flow murmur peaks early (crescendo) and then falls away (decrescendo) as the velocity and the ejected volume of blood diminish in mid to late systole. *The systolic ejection murmur usually ends before S2.* However, with semilunar valve stenosis, prolonged ventricular emptying may produce a late peaking murmur that can extend to or beyond A2 or P2 (Fig. 10-3). The intensity of the ejection murmur is related to the

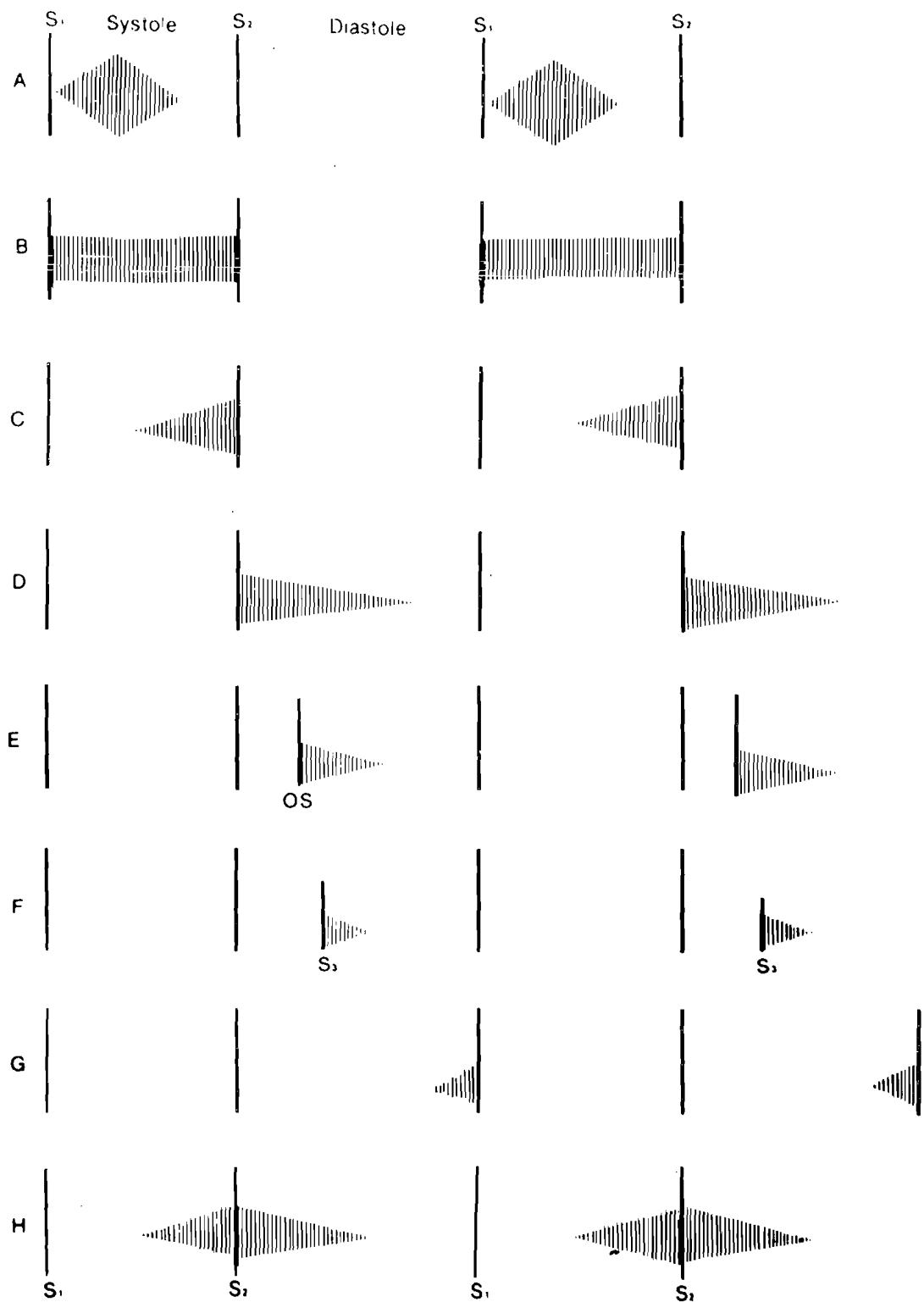


FIG. 9-5. Major types of cardiac murmurs. See text for description. (From Abrams J: Prim Cardiol, 1981.)

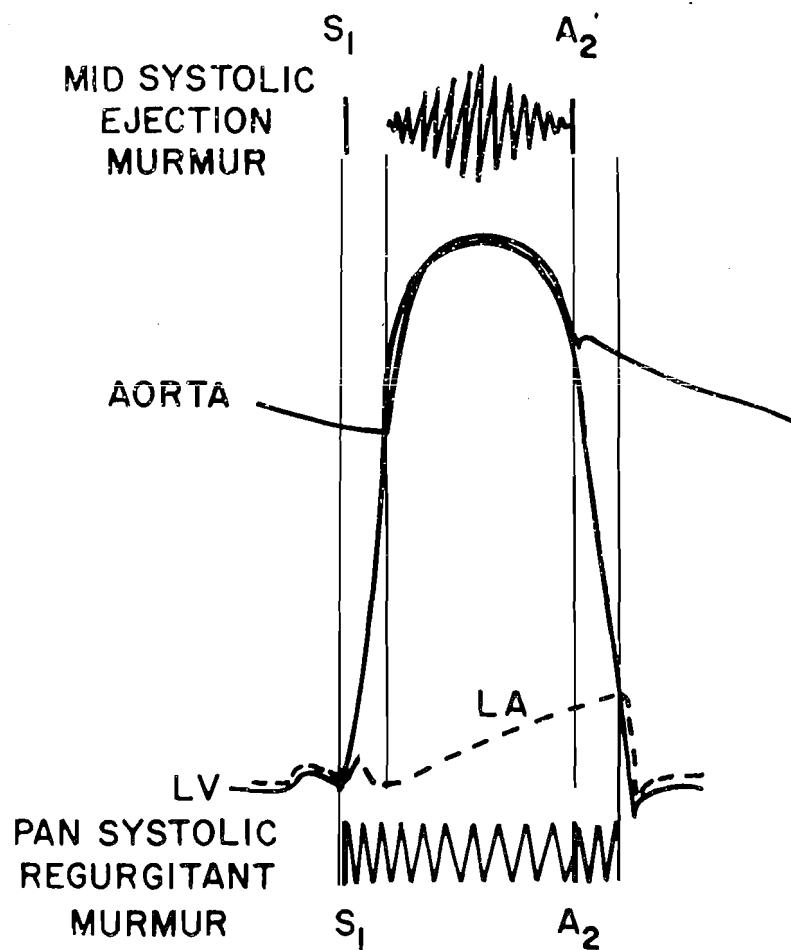


FIG. 9-6. Intracardiac pressure relationships of the midsystolic ejection and pansystolic regurgitation murmur. The crescendo-shaped ejection murmur begins during early to midsystole and ends before closure of the semilunar valves. The holo- or pansystolic murmur of mitral regurgitation begins with S₁ and continues without a decrease in amplitude to A₂ or beyond. Note that left ventricular pressure exceeds left atrial pressure throughout this period. (From Reddy PS, Shaver JA, and Leonard JJ: Cardiac systolic murmurs: pathophysiology and differential diagnosis, Prog Cardiovasc Dis 14:1, 1971.)

velocity of blood flow, which in turn is dependent upon stroke volume, valve orifice area and accelerating forces. A large stroke volume results in a louder and longer murmur.

Types of Ejection Murmurs

Murmurs related to the ejection of blood across the semilunar valves are either *flow-related* or *pathologic* in origin (Table 9-3).

Flow or Functional Murmur. These are by far the most common heart murmurs and are found in many normal persons (Chapter 10). In the presence of a normal resting cardiac output the murmur is known as an *innocent* or *functional murmur*. If the murmur is related to a hyperkinetic state with increased cardiac output and stroke volume, the murmur is known as a *physiologic murmur*. Some authors object to the term "functional murmur"

TABLE 9-3 *Causes of Abnormal Ejection Murmurs*

Abnormal semilunar valve—no gradient
Abnormal semilunar valve—with gradient (stenosis)
Increased diameter or dilatation of a great vessel
Abnormal sub- or supravalvular narrowing of the left or right ventricular outflow tract
An abnormal increase in blood flow across a structurally normal outflow tract where the augmented blood volume results from an underlying organic cardiac lesion (e.g., the systolic murmur associated with an atrial septal defect or pure aortic regurgitation)

as it is vague and subject to varying interpretations. Nevertheless, the term enjoys widespread popularity; specifically, it refers to a nonpathologic, systolic ejection murmur.

Pathologic, Significant, or Organic Ejection Murmurs. These murmurs result from underlying structural abnormalities of the semilunar valves, the cardiac chambers, or great vessels. The anatomic abnormality may be minimal, but these murmurs imply an underlying structural cardiac abnormality. Several anatomic and pathophysiologic conditions can cause such murmurs (Table 9-3).

SYSTOLIC REGURGITANT MURMURS

Systolic regurgitant murmurs are generated by a continuous systolic pressure gradient produced by an abnormal structural or functional communication between two chambers of the heart. A typical regurgitant murmur begins with the development of ventricular pressure (isovolumic contraction) and continues until S2 (Fig. 9-4A). If a large pressure gradient between two cardiac chambers persists into late systole, blood flow and cardiac sound will continue until the onset of diastole (Fig. 9-6). The classic regurgitant murmur is holo- or pansystolic with a constant amplitude and shape throughout systole (Figs. 9-4A, 9-5B, 9-6). In some patients, however, such murmurs may have decrescendo or crescendo characteristics. Systolic regurgitant murmurs are present in mitral and tricuspid regurgitation and ventricular septal defects. Late systolic murmurs, beginning after S1 and extending to S2, typically reflect mild degrees of mitral regurgitation (Figs. 9-4C, 9-5C).

DIASTOLIC MURMURS

Semilunar Valve Regurgitation

Semilunar valve regurgitation murmurs begin with semilunar valve closure (S2) and are usually decrescendo in configuration (Figs. 9-4E, 9-5D). The shape and length of the murmur reflect the diastolic pressure gradient between aorta or pulmonary artery and the respective ventricle. Because of

the high velocity of regurgitant blood flow from the great vessels, the murmurs of semilunar valve incompetence typically are of high frequency.

Ventricular Filling Murmurs

A-V Valve Stenosis. These murmurs are caused by obstruction to inflow of blood to the left or right ventricle in patients with mitral or tricuspid valve stenosis (Figs. 9-4B, 9-5E). The onset of the murmur follows complete opening of the A-V valve, and the murmur is mid-diastolic in timing. Typically, these murmurs are low-pitched because of the relatively small pressure gradients between the atrium and ventricles, which result in low flow velocity even with severe stenosis.

Increased A-V Valve Flow Without Valvular Stenosis. Under conditions of excessive atrial blood volume and augmented flow across the mitral or tricuspid valve, a short, mid-diastolic "filling murmur" often can be heard. Such a murmur may begin with a ventricular filling sound (S3) (Fig. 9-5F). Mid-diastolic flow murmurs are common when there is increased flow across an A-V valve due to a left-to-right shunt (atrial septal defect, ventricular septal defect, or patent ductus arteriosus). Although these flow murmur's do not indicate diseased A-V valves, they do indicate organic cardiac disease as cause of the increased flow. The Austin Flint murmur of aortic regurgitation is another variant of the "functional" diastolic filling murmur (see Chapter 15). It results from a large volume of blood crossing the mitral valve during diastole while the anterior leaflet of the valve is displaced toward the valve orifice.

Presystolic Murmur. A late diastolic or presystolic murmur may be heard in patients with mild to moderate mitral or tricuspid stenosis; this results from augmentation of A-V flow following atrial contraction. Increased velocity of blood flow and turbulence are produced as the closing A-V leaflets further narrow the AV valve orifice after atrial systole. This causes a presystolic murmur that extends to S1 (Figs. 9-4B, 9-5G) and is often associated with an earlier mid-diastolic murmur.

Continuous Murmurs

Continuous murmurs result from a persistent gradient between a high pressure site (arterial or ventricular) and a low pressure site (vein or right heart chamber). These murmurs typically begin in systole and "spill over" into early diastole, peaking in mid to late systole. Rarely, they are audible in late diastole (Figs. 9-4F, 9-5H). The shape of a continuous murmur depends upon the magnitude of the pressure gradient at any specific time in the cardiac cycle.

DIAGNOSIS OF ORGANIC MURMURS

Because nonpathologic or innocent flow murmurs are so common in normal persons, the clinician has a great responsibility to determine whether or not a heart murmur is pathologic. Chapter 10 discusses the characteristics of the innocent or physiologic ejection murmur. Unfortunately, there are only a few criteria that absolutely identify the organic or pathologic cardiac murmur.

1. *All diastolic murmurs are pathologic.* There should be no cardiac sound during diastole. Occasionally, the physiologic S3 of children or young adults will produce short, after-vibrations and is heard as a brief, mid-diastolic murmur. A prominent, pulmonic closure sound (P2) can reverberate and produce a brief diastolic trill, simulating a short decrescendo diastolic murmur. Mid-diastolic flow murmurs across the mitral (VSD, PDA, mitral regurgitation) or tricuspid (ASD, tricuspid regurgitation) valves do not imply organic disease of the A-V valves, but reflect the marked diastolic volume overload traversing the A-V valve during rapid ventricular filling.
2. *All pansystolic and late systolic murmurs are pathologic.* Because the volume of flow and the velocity of ejection in normal hearts is markedly decreased in late systole, cardiac sound typically is absent during the last third of ejection. Murmurs that extend to S2 are organic and indicate continuing turbulent blood flow at a time in the cardiac cycle when silence is expected. It is essential to selectively listen to late systole for the presence or absence of sound vibrations. In some varieties of mitral regurgitation (typically with lesions due to abnormalities of the mitral supporting apparatus), mitral regurgitant flow begins well after ejection, and abnormal blood flow into the left atrium occurs only during mid to late systole (Chapters 17, 18). Thus, a murmur beginning in the last third of systole usually reflects "late" mitral regurgitation.
3. *Continuous murmurs always indicate organic heart disease.* A murmur that continues up to S2 and spills over into diastole must reflect a continuing pressure differential between two cardiovascular structures and is, therefore, abnormal.
4. *Very loud murmurs are usually pathologic.* Any murmur associated with a thrill (grade 4 or greater) has a pathologic basis. In rare cases of pure aortic regurgitation without aortic stenosis, the huge stroke volume traversing the diseased aortic valve will result in a very loud ejection murmur and a thrill. In such situations the presence of the systolic ejection murmur may not indicate actual aortic stenosis. Table 9-4 lists common noncardiac conditions associated with an

TABLE 9-4 *Factors Affecting the Loudness of Heart Murmurs*

Increased Intensity
High cardiac output (hyperdynamic) states
Thin chest wall
Narrow thoracic diameter, e.g., "straight back," pectus excavatum
Anemia (decreased blood viscosity)
Tortuous aorta (close to chest wall)
Decreased Intensity
Obesity
Muscular or thick chest wall
Obstructive lung disease
Barrel chest (increased A-P diameter)
Pericardial thickening or fluid
Decreased cardiac output (CHF, low ejection fraction)

increase or decrease in murmur intensity. These factors should always be kept in mind during auscultation of the patient.

5. *Associated cardiac abnormalities.* Evidence of underlying cardiac disease raises the likelihood that a given heart murmur is organic. For instance, the presence of left ventricular hypertrophy on precordial palpation, an ejection or midsystolic click, or an opening snap would suggest that the heart murmur represents a cardiac structural abnormality. However, these phenomena may be unrelated to the production of the murmur itself.
6. *Frequency, shape or contour, and radiation* characteristics of heart murmurs are too nonspecific to definitively establish the organic etiology of a murmur.

ADDITIONAL AIDS TO IDENTIFICATION OF MURMURS

Maneuvers and Pharmacologic Agents. Clinicians can utilize a number of ancillary approaches to help sort out the problems of differential diagnosis in cardiac auscultation. Chapter 11 discusses the maneuvers and drugs that are helpful in evaluation of heart murmurs (Table 9-5).

EVALUATION OF HEART MURMURS

The clinician should take a systematic approach to the accurate assessment of heart murmurs. The following features of murmurs should be consciously analyzed in each patient:

- Whether the murmur is *systolic* or *diastolic*

TABLE 9-5 *Pharmacologic Aids and Physical Maneuvers Used in Heart Murmur Evaluation*

Squatting	Pressors: phenylephrine
Handgrip	aramine
Dynamic exercise	angiotension
Valsalva maneuver	Amyl nitrite
Muller maneuver	Nitroglycerin
Upright posture	
Respiration	
Changes in cardiac rhythm	
Post-PVC response	
Long cycle lengths (atrial fibrillation, carotid sinus massage)	

- The *timing* of the murmur within systole or diastole—early, mid, or late
- The *duration* of the murmur
- The *intensity* or *loudness* of the murmur, e.g., its grade
- The transmission of the murmur
- The *frequency* and *shape* of the murmur
- Other cardiac abnormalities present on examination that may relate to the etiology of the murmur

Some of these parameters will be discussed in further detail. The reader is also referred to Chapter 1 for additional information on cardiac sound and the use of the stethoscope.

TIMING

Once it is clear that a given murmur is systolic or diastolic (see Chapter 1), it is necessary to clarify whether the murmur is early, mid, or late in systole or diastole.

Systole. Ejection murmurs, whether pathologic or not, are all early to midsystolic in timing. When there is a severe degree of obstruction of the outflow tract, ejection murmurs will lengthen and peak later in systole. Not all early systolic murmurs are ejection murmurs. In some patients, the murmur of a ventricular septal defect or A-V valve regurgitation begins in early systole and tapers off in late systole, ending before A2.

A “late systolic murmur” is not heard in the first third of systole and should not be confused with an ejection murmur; late systolic murmurs usually represent mitral regurgitation (Figs. 9-4C, 9-5C; Chapter 17). These murmurs begin in mid systole with sound vibrations heard up to A2.

Diastole. Early diastolic murmurs result from semilunar valve incompetence (aortic or pulmonary regurgitation). Mid-diastolic murmurs are produced by flow across either the mitral or tricuspid valve during passive filling of the ventricles. In stenosis of either A-V valve, the diastolic murmur is caused by a narrowed and obstructive orifice. Such murmurs lengthen in

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proportion to the severity of valvular obstruction, and cardiac sound may persist up to S1. Although these murmurs are often called "pandiastolic," this characterization is incorrect as the murmur typically begins following a mitral or tricuspid opening snap. Mid-diastolic murmurs also can occur without narrowing of the A-V valve if there is markedly augmented blood flow with rapid ventricular filling. Here, the murmur is brief and usually follows an S3. Late diastolic and presystolic murmurs are produced by atrial contraction in the presence of mitral or tricuspid stenosis.

Continuous murmurs represent cardiac sound that spills over into diastole. Care must be taken to differentiate these murmurs from the combination of a long systolic and early diastolic murmur that can be heard in mixed semilunar valve stenosis and regurgitation.

DURATION

Systole. The most important characteristic of systolic murmurs relates to their length. *Practical Point: Whether a systolic murmur extends to S2 and is, therefore, holosystolic is the most valuable part of murmur analysis.* Any murmur that is truly holo- or pansystolic, regardless of shape, is caused by continuous pressure differential between two cardiac chambers (Figs. 9-4A, 9-6). Thus, the classic murmurs of a ventricular septal defect (Chapter 21) or mitral regurgitation (Chapter 17) begin with S1 and extend through S2. The abnormal intracardiac communication results in the onset of blood flow before the aortic valve opens and continues after aortic valve closes. Severe obstruction of the left or right ventricular outflow tract may cause a prolonged systolic murmur that is actually pansystolic; this is particularly true with "tight" pulmonic stenosis where sound vibrations may extend beyond S2 (Fig. 10-3).

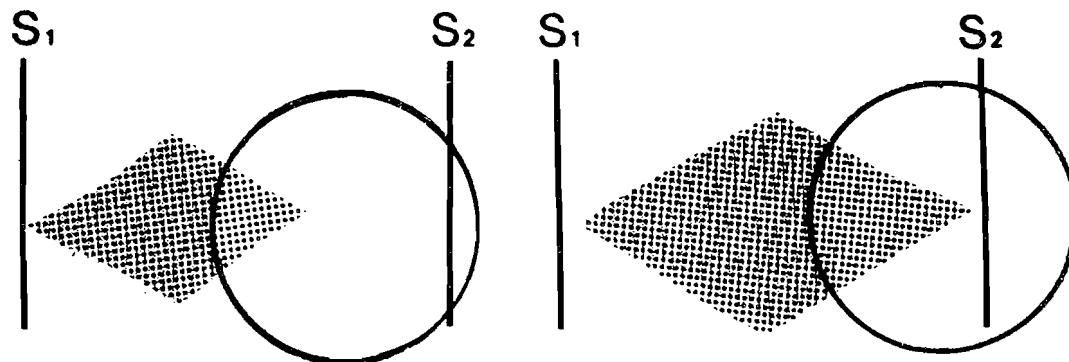


FIG. 9-7. Importance of late systole in evaluation of systolic murmurs. It is essential to assess the last part of systole to determine whether a murmur is ejection in nature or is holosystolic. On the left, an early peaking murmur ends before the last third of systole. This is the rule in functional murmurs or with mild semilunar valve stenosis. On the right, a long ejection murmur is shown which peaks later in systole. Sound vibrations extend to S2 suggesting severe obstruction to ventricular outflow. In severe semilunar valve stenosis, the vibrations may extend beyond S2.

(From Abrams J: Prim Cardiol, 1981.)

The presence of a long systolic murmur should stimulate a careful evaluation of late systole through the use of selective listening. A careful focus on the last 25% of systole is important (Fig. 9-7). If sound vibrations end before S₂, the murmur is usually an ejection murmur. If the murmur extends to S₂ (pansystolic), the differential diagnosis should include regurgitant lesions as well as severe, semilunar valvular stenosis.

Diastole. The duration of diastolic murmurs is important, particularly in the assessment of left ventricular filling murmurs. As already emphasized, flow across a nonobstructive or mildly stenotic A-V valve produces a short mid-diastolic murmur. A long diastolic murmur following an opening snap is indicative of a persistent A-V gradient and significant mitral stenosis. With short cycle lengths, these murmurs will extend to S₁. With bradycardia or long R-R cycles during atrial fibrillation, the mitral diastolic murmur usually ends in mid to late diastole.

Murmurs of semilunar valve regurgitation usually are quite long and may be pandiastolic, continuing to S₁. Usually, these high frequency murmurs are of low intensity and may be very faint in late diastole. The persistent, late diastolic gradient between the great vessel and the ventricle produces sound vibrations that last into late diastole. With severe aortic regurgitation, particularly in acute valvular incompetence, the exceedingly high, left-ventricular end-diastolic pressure may produce a small or absent aortic left-ventricular gradient in late diastole resulting in a relatively short, diastolic regurgitant murmur (see Chapter 15).

INTENSITY

The intensity or loudness of a murmur is directly related to the degree of turbulence. Increased volume and/or velocity of flow results in enhanced turbulence and louder murmurs. First, the site of maximum intensity should be localized and attention should be paid to where else on the precordium the murmur is heard. In general, the site of maximal loudness is related to the anatomic site of the cardiovascular lesion. This explains the emphasis on specific "valve areas" for cardiac auscultation. However, there are many exceptions to these relationships, and the informed physician must be aware of the radiation patterns of specific valve lesions (Fig. 9-2). The examiner should utilize the 1 to 6 murmur grading system in a consistent fashion.

Often, it is assumed that the intensity of a murmur is directly related to the severity of the underlying condition. Although this is frequently true, there are many exceptions. The careful auscultator should be aware of the various factors that may modify the intensity of murmurs (Table 9-4). The common causes of decreased intensity of heart murmurs include poor cardiac function that results in a reduced stroke volume and ejection velocity and "buffering" of the heart from the stethoscope by excessive body tissue (e.g., obstructive lung disease, obesity). For example, in congestive heart failure

resulting from aortic stenosis or mitral regurgitation, the systolic murmur may be soft and unimpressive, possibly causing a false assessment of the underlying problem. On the other hand, occasional innocent murmurs may be prominent (but never grade 4/6 intensity). The classic, small ventricular septal defect can produce an extremely loud murmur.

TRANSMISSION

Radiation and transmission patterns of murmurs are directly related to the intensity or loudness of the murmur. Loud murmurs transmit widely; soft ones do not. Radiation patterns are probably related to turbulent blood flow that sets cardiac chambers and vascular walls into vibration; in turn, these vibrations are transmitted to the chest wall. The intensity of the murmur diminishes rapidly away from the site of sound production. High frequency sound is best heard proximal or "upstream" from the origin of the murmur; low frequency vibrations transmit best "downstream" or distal to the murmur's origin. This accounts for the common observation that the harsh components of the aortic stenosis murmur transmit to the base and neck, while higher frequency vibrations are best heard at the apex. Low frequency sound transmits through thoracic tissues better than does high frequency sound which explains the common finding of a thrill in aortic stenosis, but not in aortic regurgitation.

The cardiac auscultatory regions (Fig. 9-2; Table 9-1) are related to the typical radiation patterns of classic valvular defects.

SHAPE

Assessing the contour or shape of heart murmurs may help the clinician to distinguish systolic ejection murmurs (crescendo-decrescendo) from regurgitant (holosystolic) ones. The contour of a murmur is also known as the "murmur envelope" (Fig. 9-8). Recordings of murmurs using selected frequency filters may be different from what is actually heard by the human ear. The typical kite-shaped or diamond-shaped ejection murmur often looks far more classic on a phonocardiogram than it appears to the ear, which may only perceive a rough, slightly tapering murmur ending before S1. The true shape of a murmur is more difficult to determine than is commonly appreciated.

There are important exceptions to the classic contour of murmurs. For example, the pansystolic murmur of mitral regurgitation may have late systolic accentuation or can taper off in late systole. It is useful to diagram the shape of the murmur in the clinical record.

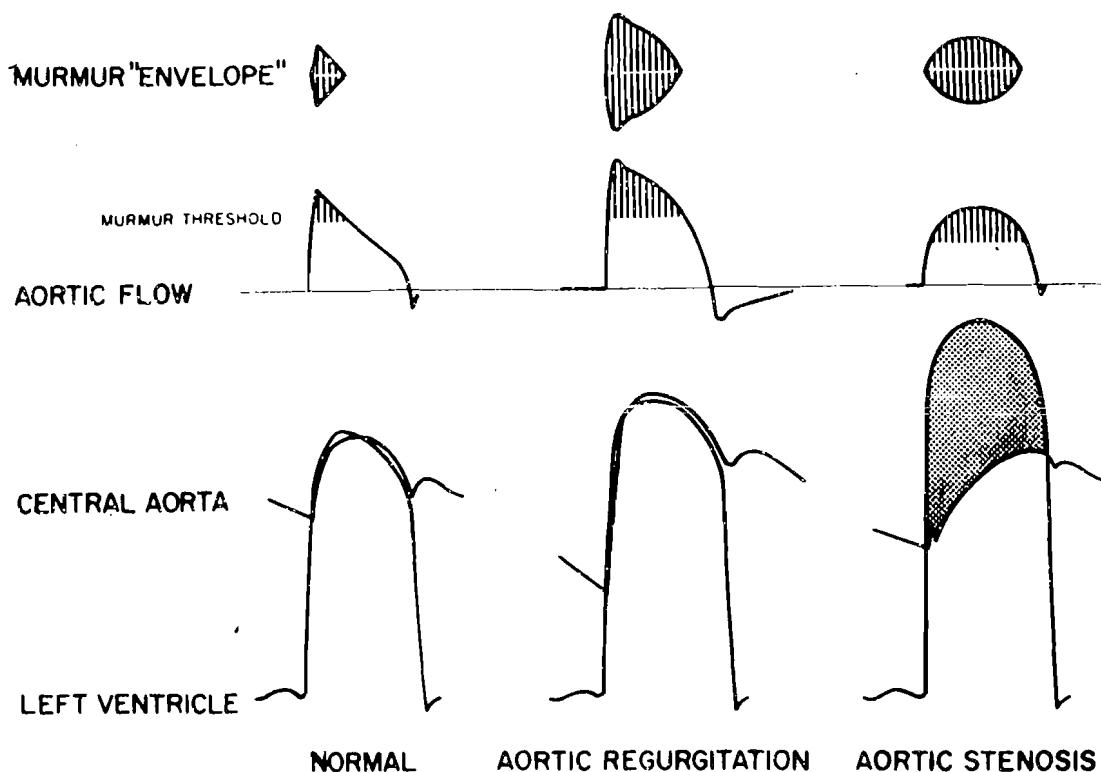


FIG. 9-8. The murmur envelope. This is a diagram of aortic flow, murmur intensity, and simultaneous intracardiac pressures, which in combination determine the murmur envelope. During normal left ventricular ejection (left), peak flow occurs early in systole. The center panel shows the exaggeration of the normal pattern of left ventricular ejection during high output states. With severe aortic stenosis (right), the flow contour is rounded and prolonged, as rapid early ejection is not possible; thus, a diamond-shaped murmur of aortic stenosis is seen. (From Reddy PS, Shaver JA, and Leonard JJ: Cardiac systolic murmurs: pathophysiology and differential diagnosis. *Prog Cardiovasc Dis* 14:1, 1971.)

FREQUENCY

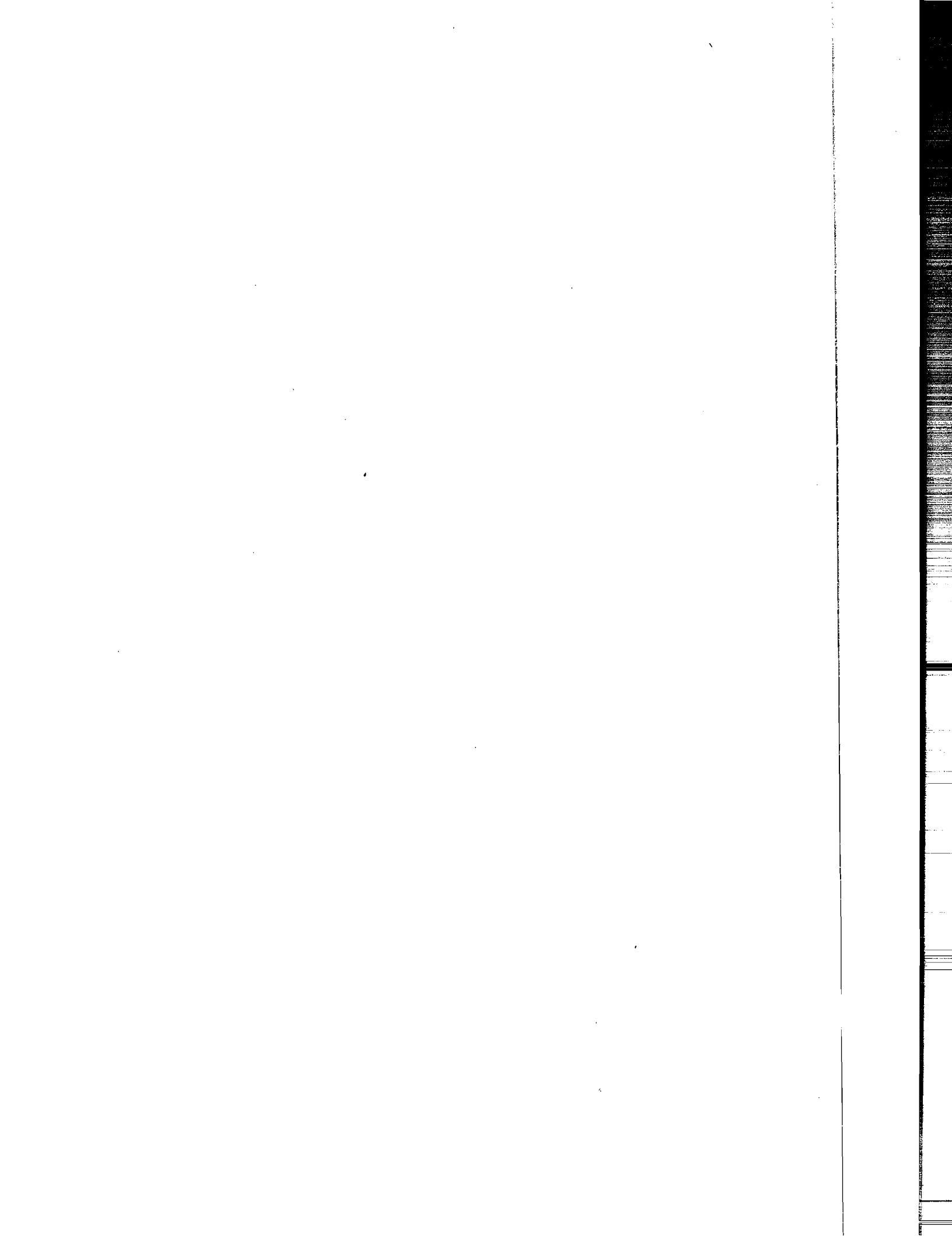
The pitch or frequency of a heart murmur has some diagnostic value and should be noted. Table 9-6 lists the range of frequencies for most cardiac murmurs. High-pitched cardiac sound results from high velocities of flow and/or large gradients between chambers. Thus, the murmurs of aortic and mitral regurgitation or of pulmonic regurgitation secondary to pulmonary

TABLE 9-6 Frequency and Pitch of Common Cardiac Murmurs

	CPS	Common Descriptive Terms	Examples
Low frequency	25–125	Rumbling, low-pitched	Diastolic murmur of mitral stenosis
Medium frequency	120–300	Rough "flow" murmur	Innocent murmur, physiologic flow murmur
High frequency	> 300	Blowing, whirring, high-pitched	Aortic regurgitation Mitral regurgitation

hypertension are classically high frequency. Conversely, low blood flow and small gradients result in lower pitched sound; such murmurs often are called "rumbling." Because of a large proportion of dominant higher frequencies, higher pitched murmurs are more musical or pure in tone than lower frequencies. These often are called "blowing" murmurs. Very harsh murmurs (e.g., aortic valve stenosis) are mixtures of medium and high frequencies; the ear best perceives the lower tones, which tend to mask the higher frequencies. Murmurs with a preponderance of a particular frequency often have a certain resonance that is called "musical" or "vibratory." Murmurs with a relatively clear pitch and pure fundamental tone are often called "cooing" or "seagull" murmurs.

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Chapter 10

The Systolic Ejection Murmur: Innocent, Physiologic and Pathologic

A wide variety of normal and abnormal conditions can produce the relatively nonspecific "ejection" murmur. Therefore, it is important to focus on characteristics that help define a normal or flow murmur from one that indicates organic heart disease. Most systolic murmurs heard in everyday practice are related to physiologic blood flow and are of no clinical importance. It is imperative for the physician to be able to properly identify these murmurs, thus avoiding the need for referral to a cardiologist, additional diagnostic tests, prophylaxis against acute rheumatic fever and bacterial endocarditis, and restriction of activities or employment.

This chapter will focus on the distinguishing features of systolic ejection murmurs. The differential diagnosis of regurgitant or pansystolic murmurs, continuous murmurs, and diastolic murmurs is found in the individual chapters dealing with specific valve or anatomic conditions.

INNOCENT OR FUNCTIONAL MURMURS

The innocent murmur, by far the most common murmur heard in clinical practice, has certain characteristics that should enable the careful examiner to identify it more accurately than 90% of the time. If ancillary techniques are also employed (ECG, chest roentgenogram, and, occasionally, echocardiography), virtually all functional systolic ejection murmurs can be properly classified.

Terminology. There are literally dozens of names for the innocent systolic murmur. Cacares and Perry have written an entire book on the subject and compiled well over one hundred names from the literature. Some examples include Still's murmur, normal murmur, harmless murmur, innocuous murmur, basal ejection murmur, twanging string murmur, nonorganic murmur, and vibratory murmur. The sine qua non is that the underlying cardiovascular system is entirely normal and that the murmur is audible at rest.

The use of such descriptive terms as "flow," "innocent," "physiologic," or "functional" murmur is very important as it clearly implies that a systolic ejection murmur is not the result of an organic valve lesion or an abnormal communication within the heart or great vessels. Some experts object to the nonspecific terminology of a "flow" or "functional" murmur, but common

usage and common sense dictate continued utilization of these descriptors. Another commonly used term for such murmurs is "physiologic," implying that the murmur results from blood flow and turbulence produced by normal cardiac structures. In this text, the preferred definition of a *physiologic murmur* is a systolic ejection murmur resulting from increased blood flow due to an augmented velocity and/or volume of blood. "Innocent" or "benign" are commonly employed terms indicating a normal functional heart murmur audible under basal conditions. When such murmurs become louder with exercise, excitement, anemia, or tachycardia, they then may be labeled as physiologic murmurs.

Etiology. The innocent murmur is the result of normal turbulence occurring during the ejection of blood from the heart. The actual anatomic site of origin is probably within the great vessels rather than the ventricles, but some murmurs may be generated in the outflow tract of either ventricle. Classic teaching has been that the typical innocent flow murmur arises in the pulmonary artery. This belief has been supported by the site of peak murmur intensity (2nd to 3rd left interspace), the proximity of the pulmonary artery to the chest wall, and older intracardiac phonocatheter studies. However, recent work of Stein and Sabbah suggests that most, if not all, innocent murmurs actually arise in the aorta as a result of the normal turbulence generated by left ventricular ejection. Their meticulous intracardiac investigations of both the left and right heart of normal subjects have clearly documented that noise energy, turbulence, and recordable sound are far greater in the aorta than the pulmonary artery in individuals with no structural cardiovascular abnormality. It is likely that in some instances the right heart and pulmonary artery may be responsible for the innocent systolic ejection murmur. Studies in children suggest that enhanced cardiac contractility is an important common denominator in the genesis of an innocent murmur, particularly if the murmur persists through childhood. The etiology of such increases in cardiac contractile function is obscure. In summary, most available evidence supports the concept that innocent or physiologic murmurs are due to rapid early ejection of blood into the aorta resulting in sufficient turbulence to produce audible sound.

Age and Incidence. Under optimal acoustic conditions, experienced clinicians hear sound vibrations in systole in as many as 80 to 90% of children and young adults. The majority of these subjects will have an audible S3 as well (Fig. 10-1). High quality phonocardiograms can record sound in systole in virtually all subjects. With increasing age, the innocent murmur tends to attenuate or disappear; nevertheless, it is quite common (30 to 40% of individuals) to hear a soft systolic ejection murmur in normal adults.

Systolic Murmur of Aging. Many older subjects in their 6th through 9th decades have an audible systolic ejection murmur that may have a different etiology than the innocent murmur of youth. These murmurs typically radiate

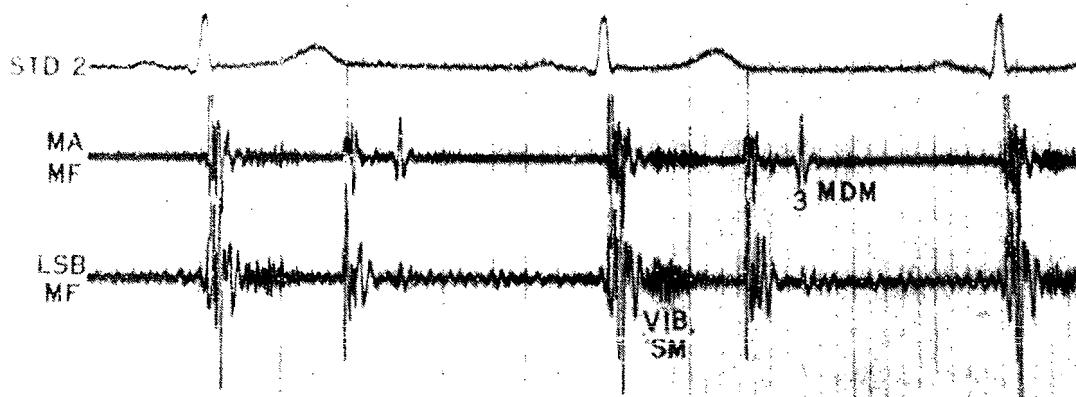


FIG. 10-1. Functional systolic ejection murmur and third heart sound in a normal child. Note the short vibratory systolic murmur and the prominent S3 (3) followed by a brief low intensity mid-diastolic murmur (MDM). MA = mitral area, MF = medium frequency, LSB = left sternal border. (From McLaren MJ et al: Innocent murmurs and third heart sounds in black school children. Br Heart J 43:67, 1980.)

well to the aortic area (2nd right interspace) and neck and may also be well heard at the apex. They often have a rougher quality than the typical innocent murmur of childhood and are particularly common in patients with hypertension. These systolic murmurs are felt to reflect underlying aortic valve sclerosis; they can simulate valvular aortic stenosis and, on occasion, may present a difficult differential problem (see Chapter 13). Most observers believe that this murmur is related to specific changes that accompany aging: thickening of the aortic valve leaflets, slight dilatation or tortuosity of the aorta, and calcification or fibrosis of the aortic ring or leaflets. When there are accompanying ECG changes or cardiovascular symptoms, it is easy to confuse these murmurs with those reflecting organic heart disease. Particular attention should be paid to the carotid pulse for signs of aortic stenosis, although this can be misleading because of age-related compliance changes in the peripheral vessels (see Chapter 3). The length of the systolic murmur is also useful in assessing whether the murmur is significant. If the murmur ends well before S2, important aortic valve stenosis is unlikely. Echocardiography should be employed in doubtful cases to assess the thickness and excursion of the aortic valve cusps.

Location, Loudness, and Radiation. The innocent murmur is best heard along the left sternal border at the 2nd to 4th interspace, most often between the apex and lower sternal border. Both the pulmonic and aortic valves are located in this region; the pulmonic valve is slightly superior to the aortic valve. It is important to assess the site of maximal intensity of the murmur. Innocent murmurs are uncommonly loudest at the apex, although they may be well heard at that site. As a rule, they do not radiate well into the neck, although loud flow murmurs may be easily heard throughout the precordium.

Some innocent murmurs do radiate into the carotid arteries. Normal flow murmurs typically are of low or moderate intensity (usually 1-2/6) and are never grade 4/6 intensity. Thin persons tend to have louder innocent murmurs; a particularly loud systolic murmur in a subject who is very muscular, barrel-chested, or obese should prompt a careful search for organic disease.

Length and Shape. The typical innocent murmur occupies only one half to two thirds of systole and is crescendo-decrescendo in configuration (Figs. 10-1, 10-2). In general, ejection murmurs unrelated to outflow tract obstruction peak early and end well before S2. At times the innocent murmur will appear to be entirely decrescendo in shape; this occurs if the brief crescendo portion is not detectable as it blends in with the vibrations of S1.

Quality. The innocent murmur is usually of low to medium frequency (60 to 180 Hz) and often has a preponderance of relatively pure frequencies. It may appear to be vibratory to the ear; terms such as "whirring," "buzzing," and "humming" are not uncommon descriptors. Still's murmur is a term that denotes a particularly vibratory murmur of a dominant harmonic, usually low to medium frequency (Fig. 10-2A). There is no particular connotation to this murmur and the term is not helpful, although it is entrenched in the literature.

Response to Maneuvers. The intensity or loudness of an innocent or flow murmur will be enhanced by anything that increases the velocity of blood flow. Exertion, anxiety, or excitement will exaggerate this murmur. Amyl nitrite initially will decrease and then increase loudness of the murmur. The innocent murmur is louder in a post-PVC beat or after a long R-R cycle. It decreases with the strain phase of the Valsalva maneuver and increases with prompt squatting. Sitting or standing usually attenuates the innocent murmur by reducing the cardiac output. The typical functional murmur becomes much softer or even disappears in the upright position. Respiration and handgrip do not produce diagnostic alterations in the typical flow murmur.

Associated Findings. The presence of a definite accompanying cardiovascular abnormality makes the diagnosis of an innocent murmur more tenuous (Table 10-1). For example, a coexisting systolic ejection or nonejection click, diastolic murmur, opening snap, or left ventricular heave would indicate that there is something wrong with the heart. In such cases, one must be careful before dismissing an associated systolic ejection murmur as physiologic or innocent. Nevertheless, cardiac abnormalities may exist in association with an unrelated innocent murmur. For instance, middle-aged hypertensive patients with ECG evidence of an old myocardial infarct or LVH frequently have systolic ejection murmurs with no clinical significance. Patients with pure mitral stenosis can have an innocent systolic flow murmur that must be differentiated from mitral regurgitation. On the other hand, the association of an ejection murmur with an ejection click strongly implies a congenital

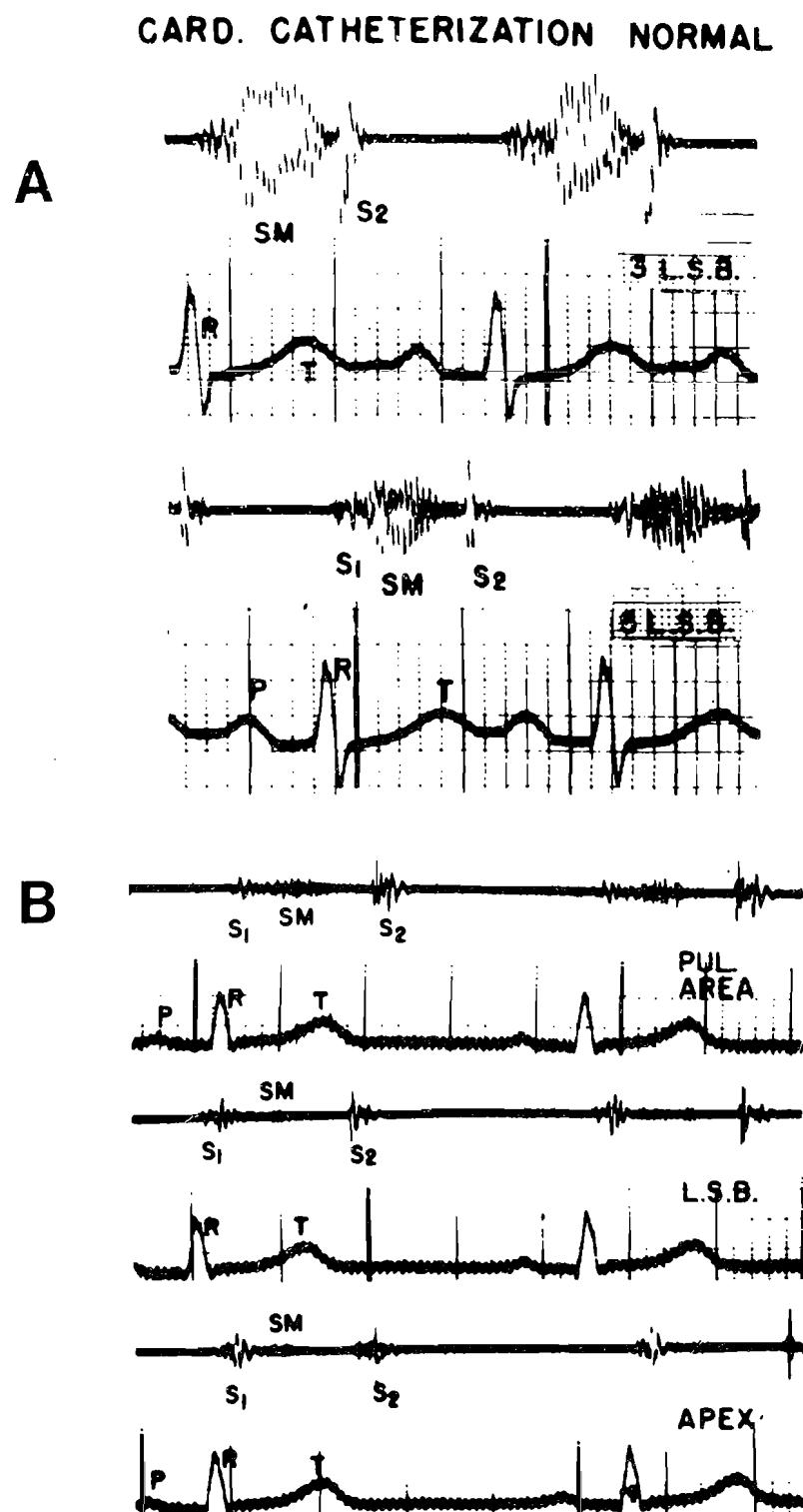


FIG. 10-2. Examples of normal systolic ejection murmurs. A. Musical vibratory murmur in a 13-year-old girl without heart disease. This "innocent" systolic murmur is somewhat longer in duration than usual. The relatively pure frequency vibrations are consistent with a Still's murmur. B. Short systolic murmur in a normal 23-year-old. (From Harvey WP: Innocent vs significant murmurs. *Curr Prob Cardiol* 1:1, 1976.)

TABLE 10-1 *Associated Physical Findings That Suggest a Systolic Ejection Murmur May Not Be Innocent*

Ejection sound
Audible expiratory splitting of S2
Fixed splitting of S2
Mid or late systolic click
Opening snap
Diastolic murmur
Very loud S1
Hyperdynamic or sustained left ventricular impulse
Right ventricular heave
Very loud A2 or P2

aortic valve deformity or great vessel dilatation. In such instances, the systolic ejection murmur is likely to be related to the underlying condition.

The characteristics of S2 may be useful in the evaluation of a systolic ejection murmur. Completely normal splitting of S2 makes an ASD or pulmonic stenosis most unlikely, though there are rare exceptions to this rule. In suspicious cases a full cardiac evaluation is necessary, including an EKG, chest film, and an echocardiogram.

PHYSIOLOGIC MURMURS

The only distinction between an innocent murmur and a physiologic flow murmur is that the latter is caused by a transient increase in blood volume and/or velocity of ejection (Fig. 7-2C). When the causative factor for the augmented blood flow is removed (e.g., correction of anemia, lysis of fever, childbirth), the murmur is no longer audible or there is only a faint (grade 1-2/6) innocent murmur. The features of such murmurs are identical in every way to those of the innocent murmur, although there may be detectable clues as to the underlying cause of the increased turbulence of blood. In essence, anything that can result in increased blood flow or augmented cardiac contractility may result in a physiologic systolic murmur, which then disappears when the basal state is restored (Table 10-2).

PATHOLOGIC EJECTION MURMURS

SYSTOLIC EJECTION MURMURS CAUSED BY ABNORMALITIES OF THE CARDIOVASCULAR SYSTEM

In certain situations, underlying cardiac disease results in an increased cardiac stroke volume which produces a flow murmur; in these cases, the structural abnormality is only indirectly related to the murmur. For example,

TABLE 10-2 Common Causes of a Physiologic Systolic Murmur

Anxiety	Pregnancy
Tachycardia	Thyrotoxicosis
Fever	Postexertion
Anemia	

in atrial septal defects or anomalous pulmonary venous drainage, the augmented blood flow produced by the large left-to-right shunt produces a systolic ejection murmur in the 2nd to 3rd left interspace (see Chapter 20). This is an example of pulmonary artery origin of a flow murmur. In aortic or pulmonary regurgitation, the ventricles eject an abnormally large stroke volume into their respective great vessels, and a systolic flow murmur is characteristically present. In severe aortic regurgitation, the associated systolic murmur can be very loud (grade 3 to 4) without any stenosis; the deformed aortic valve leaflets often are abnormal and contribute to the turbulent blood flow resulting in a systolic murmur.

Occasionally, the cardiac stroke volume is normal, but a systolic ejection murmur results from the ejection of blood into a dilated great vessel with resultant eddy formation and increased turbulence. Systolic murmurs commonly are associated with proximal ascending aortic dilatation or aneurysm, idiopathic dilatation of the pulmonary artery, or the enlarged main pulmonary artery accompanying pulmonary hypertension. In some patients with a systolic ejection murmur, the semilunar valves may be thickened, stiff, or fibrotic without any obvious clinical reason.

ABNORMAL SEMILUNAR VALVES

The classic organic systolic ejection murmur is that of aortic or pulmonic valve stenosis (see Chapter 13). A reduction of valve orifice area to at least 70% of normal is required before there is pressure difference across the valve in the resting state; mildly fused or rigid cusps may not produce sufficient obstruction to result in a pressure gradient. It may be impossible to diagnose early or mild aortic valve stenosis at the bedside. The presence or absence of an ejection click, abnormal S2, delayed or small volume carotid pulse, left ventricular hypertrophy, or echocardiographic abnormalities all help in the differential diagnosis of an innocent flow murmur versus mild valvular stenosis.

In general, the more significant the valve stenosis, the later peaking and the longer the murmur (see Chapter 13). The gap between the end of the sound vibrations and S2 narrows with increasing obstruction of either the aortic or pulmonary valve (Fig. 10-3). Other associated abnormalities (ab-

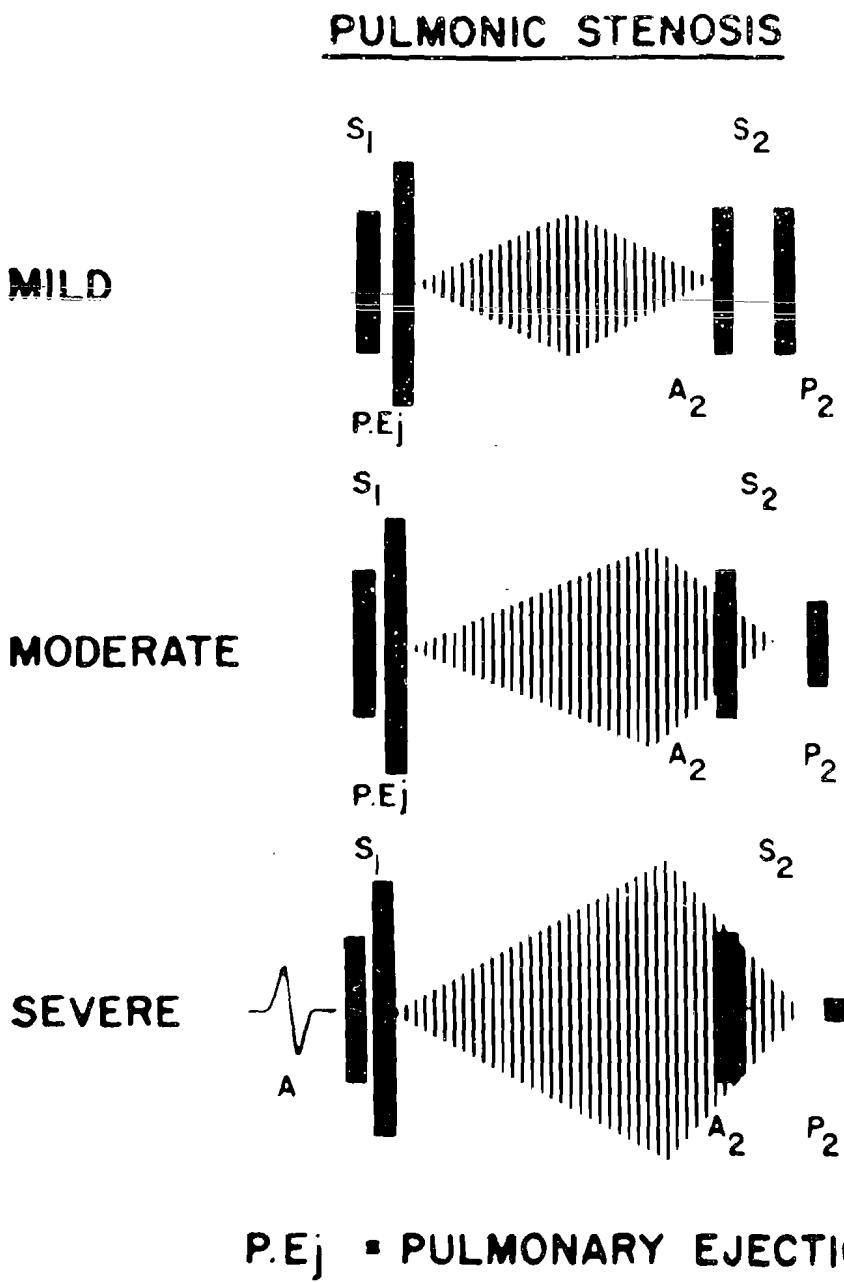


FIG. 10-3. Systolic ejection murmur in pulmonic valve stenosis. This diagram demonstrates the effect of increasing obstruction to ventricular outflow when a semilunar valve is narrowed. The murmur of valvular pulmonic stenosis increases both in duration as well as in time to peak intensity as obstruction to flow becomes more severe. Note that the murmur may actually become pansystolic, although it retains its crescendo shape even with severe valve stenosis. A right ventricular S4 (A) and a delayed and diminished pulmonic closure (P2) sound are also found in pulmonic stenosis. A pulmonic ejection click (PEj) is also shown (see Chapter 8). (From Reddy PS, Shaver JA, and Leonard JJ: Cardiac systolic murmurs: Pathophysiology and differential diagnosis. *Prog Cardiovasc Dis* 14:1, 1971.) See also Figure 13-7.

normal precordial motion, S4, ejection click, abnormal splitting of S2) are likely to be prominent with increasing grades of obstruction.

Hypertrophic Cardiomyopathy. Hypertrophic cardiomyopathy, a condition in which there is abnormal muscular thickening of the interventricular septum and an abnormal relation of the anterior leaflet of the mitral valve, produces variable obstruction to left ventricular ejection (see Chapter 14). A long systolic ejection murmur is the hallmark of hypertrophic cardiomyopathy; the murmur is produced in part by turbulence relating to rapid early systolic flow passing through the narrowed subvalvular area. Even in subjects without a measurable gradient, a murmur usually is present. In many patients, there is associated mitral regurgitation that produces a murmur superimposed on the ejection murmur but also radiates to the apex.

In hypertrophic cardiomyopathy, the maximal intensity of the murmur is usually heard lower on the chest than in valvar aortic stenosis, and its location is more medial than the typical mitral regurgitation murmur. The presence of a brisk, carotid pulse, a double precordial LV impulse, and a loud S4 is strong evidence for hypertrophic cardiomyopathy (see Chapter 14).

OTHER CAUSES OF SYSTOLIC EJECTION MURMURS

Coarctation of Aorta. A systolic ejection murmur usually is present in patients with coarctation. Its site of maximal intensity (left posterior thorax) and timing (beginning well after S1, often spilling over into diastole) are important clues. Many of these patients have an associated congenital bicuspid aortic valve, which results in a systolic ejection click and an early systolic ejection murmur at the base (see Chapters 8, 13). The cardinal diagnostic feature of coarctation is the finding of delayed and diminished femoral artery pulsations.

Mitral Regurgitation. The spectrum of murmurs found in mitral regurgitation is wide (see Chapters 17, 18). On occasion, the murmur may taper off in late systole and, in others, it takes on a crescendo-decrescendo configuration. This is an uncommon finding in rheumatic mitral regurgitation and is more likely to be found in the unusual etiologies of mitral regurgitation, such as hypertrophic cardiomyopathy, mitral prosthetic valve dysfunction, ruptured chordae tendinae, and papillary muscle dysfunction. In the latter two conditions, the development of acute mitral regurgitation often results in a large high pressure (V) wave in the left atrium. In turn, this produces a decreasing left ventricular-left atrial gradient with a concomitant decrease in the degree of mitral regurgitation in late systole (Fig. 17-8). The murmur of acute mitral regurgitation simulates an ejection murmur, as it is attenuated in late systole, and is decrescendo in shape (see Chapter 17).

Extracardiac Functional Murmurs. There are several varieties of systolic murmurs that do not arise within the heart and proximal great vessels.

These murmurs generally have a benign implication, but may cause diagnostic confusion if their etiology is unrecognized.

Venous Hum. The cervical venous hum is discussed in detail in Chapter 4. This continuous murmur heard in young children may have diastolic accentuation. The venous hum has its maximal intensity in the neck, especially on the right. When loud, the venous hum can radiate to the base of the heart and mimic a systolic ejection murmur. Light compression of the internal jugular veins or turning the head to the right will attenuate this bruit (see Chapter 4).

Cervical or SuprACLAVICULAR Arterial Bruits. In children or young adults, short, early ejection bruits can be heard at the cardiac base, clavicles, and lower neck due to the rapid velocity of ejected blood traversing the brachiocephalic arteries. These murmurs usually are loudest on the right. The bruits truly are physiologic and are not associated with vascular obstruction. Probably, they are caused by turbulence produced by the relatively small caliber vessels in the upper thorax and neck. In older subjects, such a murmur should raise the question of atherosclerotic disease in the arterial circulation.

Obviously, all such bruits must be carefully differentiated from transmitted cardiac murmurs; the technique of inching (see Chapter 1) is invaluable here. Careful auscultation over the suprACLAVICULAR area is important. Use of various positions of the arms or hand exercise may help distinguish between intra- and extracardiac origin of these bruits.

Cardiorespiratory Murmurs. On occasion, faint, high-pitched "murmurs" can be heard over the anterior thorax during inspiration, markedly diminishing or disappearing with expiration. These sounds are characterized as blowing or swishing, and can be heard in the region of the cardiac apex. They are probably breath sounds that are affected by intrathoracic changes of the cardiac anatomy and the heart's position during respiration.

Chapter 11

Auscultatory Clues Elicited By Physical Maneuvers and Pharmacologic Agents

Paul T. Cochran, M.D.

Considerable information during cardiac auscultation may be obtained by paying attention to the response of heart sounds and murmurs to physical maneuvers and occasionally to the administration of pharmacologic agents. During the examination of each patient with suspected or proven cardiac disease, the physician should pay close attention to the auscultatory responses to alterations in cardiac cycle length, normal respiration, postural changes (sitting, standing, squatting), isometric hand grip, and the Valsalva maneuver (Table 9-5). The administration of a vasoactive drug such as amyl nitrite or phenylephrine may elicit rather marked changes of the physical findings to aid in differential diagnosis. Figure 11-1 illustrates how common diagnostic problems in identification of a systolic maneuver can be resolved with a classic maneuver or the administration of amyl nitrite.

RESPIRATION

Potain in 1866 was first to note the normal respiratory variation in splitting of the second heart sound. In 1954, Leatham brought Potain's observations to clinical attention through emphasis of their significance. With normal respiration, on inspiration the intrathoracic pressure decreases and results in an augmentation of right heart blood flow with a commensurate decrease in left heart flow (Chapter 6). Murmurs generated within the right heart, therefore, generally become louder on inspiration, whereas those originating in the left heart chambers decrease as a result of reduced left heart flow as well as increased "insulation" by the air-filled lung (Table 11-1). Right ventricular S3 and S4 sounds as well as the murmurs of tricuspid regurgitation, tricuspid stenosis, pulmonic stenosis, and congenital pulmonary insufficiency increase in intensity with inspiration. Conversely, left heart murmurs are best heard in expiration when left heart filling is maximal and the amount of air-filled lung interposed between the heart and the chest wall

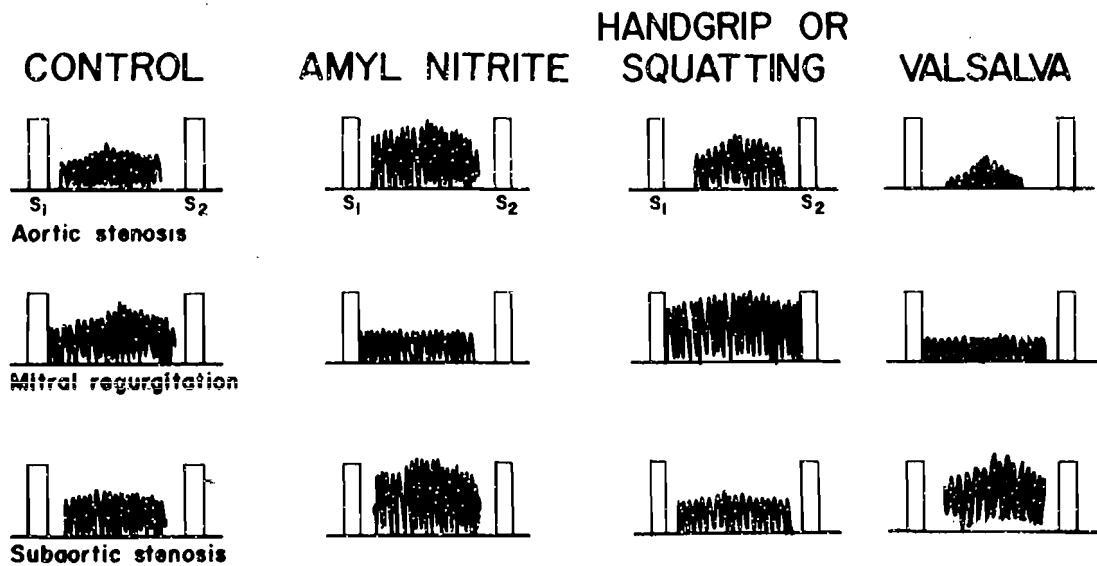


FIG. 11-1. Maneuvers helpful in differentiating the murmurs of aortic stenosis, mitral regurgitation, and hypertrophic cardiomyopathy (subaortic stenosis). See test for explanation. (From Crawford MH and O'Rourke RA: A systematic approach to the bedside differentiation of cardiac murmurs and abnormal sounds. *Curr Prob Cardiol*, 1:1, 1979.)

is least. Figure 19-4 demonstrates the classic inspiratory augmentation of the murmur of tricuspid regurgitation, and Figures 8-4 and 8-5 document the behavior of the pulmonic ejection click in patients with pulmonic valve stenosis.

In general, it is best to assess respiratory variation in heart sounds and murmurs during normal respiration. Occasionally, it will be helpful to demonstrate to the patient the depth of respiration required in order to derive the maximum benefit from auscultation. Patients who have pulmonary hypertension and severe right heart failure may not demonstrate inspiratory augmentation of right heart murmurs and gallops due to little or no increase in venous return to the right heart with inspiration because right ventricular filling pressure is so high. By requesting that these patients stand (thus reducing the resting right heart filling) and by repeating cardiac auscultation, the examiner may appreciate the expected respiratory changes.

POSTURAL CHANGES

Attention to auscultatory changes that may develop when the patient changes position is a simple, though often neglected, aid to auscultation (Table 11-2). As patients move from a *recumbent* to a *sitting* or a *standing* position, systemic venous return is abruptly reduced. Stroke volume decreases, and the ensuing reflex increases in cardiac rate and systemic vascular resistance occur immediately. There are coincident alterations in heart sounds and murmurs.

TABLE 11-1 Effects of Respiration on Filling Sounds and Pathologic Murmurs

Loudest on Expiration	Loudest on Inspiration
Left ventricular S3, S4	Right ventricular S3, S4
Mitral regurgitation	Tricuspid regurgitation
Mitral stenosis	Tricuspid stenosis
Aortic stenosis	Pulmonic stenosis
Aortic regurgitation	Pulmonic regurgitation

While listening as the patient goes from a recumbent to a sitting or standing position, one notes a diminution in intensity of all murmurs originating from the right as well as the left heart, except for those systolic murmurs of hypertrophic obstructive cardiomyopathy (HC) and mitral valve prolapse (MVP). Physiologic changes associated with the standing position that cause increased "obstruction" in the left ventricular outflow tract in patients with HC include a smaller left ventricular cavity and the reflex inotropic stimulus from increased catecholamines (Chapter 14). In patients with MVP, the systolic click and murmur occur earlier in systole as a result of the smaller left ventricle that develops upon assumption of the upright position (Chapter 18, Fig. 18-6).

Additional information can be obtained by requesting that the patient go from a *standing to a squatting* position as one auscults the heart (Fig. 11-1). With squatting, there is an initial large increase in venous return in addition to augmentation of peripheral arterial resistance secondary to the "kinking" of the femoral arteries. There is a secondary increase in systemic arterial pressure and reflex reduction of heart rate. The systolic murmur of HC frequently disappears during squatting only to become quite loud with resumption of the upright position. Right heart systolic murmurs, as well as the murmurs of mitral regurgitation and valvular aortic stenosis, also increase

TABLE 11-2 Effects of Posture on Common Pathologic Murmurs

Louder on Upright Posture	Louder on Squatting
Hypertrophic cardiomyopathy Mitral valve prolapse (early onset of click and murmur)	Aortic regurgitation Mitral regurgitation Ventricular septal defect Pulmonic stenosis Aortic stenosis Tricuspid regurgitation
Softer on Upright Posture	Softer on Squatting
Aortic stenosis Pulmonic stenosis	Hypertrophic cardiomyopathy Mitral valve prolapse— murmur may increase or decrease but begins later in systole

in intensity with squatting. The murmur of aortic regurgitation that might otherwise be missed may be easily heard with the patient in the squatting position during held expiration. Similarly, left ventricular S3 and S4 sounds may be present only during the squatting maneuver.

CHANGES IN CARDIAC CYCLE LENGTH (Table 11-3)

In 1960, Hencke called attention to the augmentation of the murmur of valvular aortic stenosis following longer cardiac cycle lengths and the importance of this observation in distinguishing the murmur of aortic stenosis from that of mitral regurgitation (Chapters 13, 17). During the compensatory pause that follows a ventricular premature contraction or following longer cycle lengths in atrial fibrillation, there is an increase in ventricular filling and secondary augmentation of ventricular contractility of the next beat. This combination increases the intensity of murmurs that are caused by obstruction to either left or right ventricular ejection. This is in marked contrast to the behavior of regurgitant murmurs of the AV valves, which do not change appreciably following longer cycle lengths. The same observations may be made while producing transient slowing of the sinus rate during light carotid sinus pressure and simultaneous auscultation.

VALSALVA MANEUVER (Table 11-3)

In order to get maximum information from auscultatory change during a Valsalva maneuver, it is often necessary to demonstrate to the patient how to perform the Valsalva maneuver and to allow the patient to practice this maneuver before trying to make assessments about the behavior of cardiac auscultatory events. The Valsalva maneuver is helpful in differentiating right-sided systolic murmurs from those originating from the left heart. It is also of considerable importance in identifying the systolic murmur of HC. During the held or strain phase of the Valsalva maneuver, there is marked reduction of systolic venous return, with a resultant decrease in cardiac output, systemic arterial pressure, pulse pressure, and a reflex increase in heart rate. Most heart sounds and murmurs are decreased during the strain phase of the Valsalva (Fig. 11-1). Upon release of the Valsalva maneuver, murmurs from the right heart generally return to baseline intensity within two to three cardiac cycles, whereas murmurs originating within the left heart do not generally return to baseline until five to ten heartbeats have occurred. Contrary to most LV outflow murmurs, during the strain phase of the Valsalva maneuver, the reduced cardiac output and smaller size of the left ventricular chamber augment the systolic murmur of HC, often dramatically (Figs.

TABLE 11-3 Effects of Valsalva Maneuver, Isometric Handgrip, and Changes in Cardiac Cycle Length on Pathologic Murmurs

	Increased	No Change	Decreased
Valsalva strain phase	Hypertrophic cardiomyopathy		Aortic stenosis Pulmonary stenosis
	Mitral valve prolapse—earlier onset		
Handgrip	Aortic regurgitation Mitral regurgitation Ventricular septal defect, mitral stenosis		Hypertrophic cardiomyopathy
Long cycle lengths	Aortic stenosis Pulmonic stenosis Hypertrophic cardiomyopathy	Mitral regurgitation Aortic regurgitation	

11-1, 14-8). Figure 6-10 diagrams how the Valsalva maneuver can help in identification of paradoxical splitting of the second heart sound.

ISOMETRIC HANDGRIP (Table 11-3)

Isometric handgrip can provide helpful diagnostic information in some patients with cardiac disorders. It is important to insure that the patient not perform a coincident Valsalva maneuver during isometric handgrip. Isometric effort increases cardiac contractility, cardiac output, and arterial pressure without significant change in size of the ventricular chamber. Left ventricular S3 and S4 sounds often are augmented during this maneuver or may even appear *de novo*. Mitral regurgitation murmurs, particularly if related to papillary muscle dysfunction, may be augmented or first recognized during isometric handgrip (Fig. 11-1). The murmurs of aortic insufficiency, as well as those of ventricular septal defect, similarly increase. Murmurs generated within the right heart structures, as well as the systolic murmurs of valvular aortic stenosis and HC, generally are unchanged or decrease during isometric handgrip. This maneuver is also helpful in augmenting the murmur of mitral stenosis.

PHARMACOLOGIC AGENTS (Table 11-4)

Amyl nitrite has been available for many years. When used during the bedside examination it can be an easy, safe, and helpful aid in cardiac aus-

TABLE 11-4 *Effect of Amyl Nitrite and Vasopressors on Various Murmurs*

Diagnosis	Amyl Nitrite	Methoxamine or Phenylephrine
<i>Systolic Murmurs</i>		
Mitral regurgitation	Decrease	Increase
Ventricular septal defect	Decrease	Increase
Patent ductus arteriosus	Decrease	Increase
Fallot's tetralogy	Decrease	Increase
Atrial septal defect	Increase	Increase or no change
Hypertrophic cardiomyopathy (IHSS)	Increase	Decrease
Aortic stenosis (valvular)	Increase	No change
Pulmonic stenosis (valvular and muscular)	Increase	No change
Tricuspid regurgitation	Increase	No change
Systolic ejection murmur (innocent)	Increase	Decrease
<i>Diastolic murmurs</i>		
Aortic regurgitation	Decrease	Increase
Austin Flint murmur	Decrease	Increase
Mitral stenosis	Increase	Decrease
Pulmonic regurgitation	Increase	No change
Pulmonic regurgitation secondary to Eisenmenger's syndrome	Decrease	Increase
Tricuspid stenosis	Increase	No change

From Tavel M: Phonocardiography: Clinical use with and without combined echocardiography. Prog Cardiovasc Dis, 26:145-175, 1983, with permission.

cultation. Inhalation of amyl nitrite may result in dramatic reduction in systemic blood pressure due to systemic vasodilatation. There is a reflex increase in heart rate and cardiac output. Amyl nitrite should be administered only to patients who are recumbent because of the drug's pronounced hypotensive effects. The fall in blood pressure is usually present within 30 to 40 seconds of inhalation and may be very transient, the reflex increase in cardiac output occurring 30 to 60 seconds later. The effects of amyl nitrite during cardiac auscultation is to increase the intensity of systolic ejection murmurs (except for that of Fallot's tetralogy), as well as to enhance the diastolic murmurs of mitral and tricuspid stenosis.

The fall in systemic arterial pressure diminishes the intensity of the murmurs of aortic regurgitation, mitral regurgitation, the Austin-Flint rumble, ventricular septal defect, patent ductus arteriosus, and systemic arteriovenous fistula (Fig. 15-9). Since the systolic murmur of tetralogy of Fallot originates across the stenotic pulmonary orifice, amyl nitrite will attenuate this murmur by decreasing systemic arterial resistance; as the impedance to ejection of blood from the right ventricle into the aorta is reduced, pulmonary blood flow is diminished. In patients with ventricular septal defect, there is an occasional paradoxical increase in the systolic murmur following adminis-

tration of amyl nitrite. Vogelpoel suggests that in some patients this is the result of a pulmonary vascular bed that is more vasoreactive than the systemic arterial bed, thus increasing left-to-right shunting and augmenting the VSD murmur.

Amyl nitrite has been particularly helpful in the differential diagnosis of murmurs due to left ventricular outflow tract obstruction (which become louder following administration of amyl nitrite) from those of mitral regurgitation (which decrease) (Fig. 11-1). Another useful area is differentiation of the apical diastolic rumble of mitral stenosis, which increases after administration of amyl nitrite, in contradistinction to the Austin-Flint rumble, which decreases (Fig. 15-9). The murmur of ventricular septal defect decreases as distinguished from the murmur of valvular pulmonic stenosis, which increases. In acyanotic tetralogy of Fallot, the murmur decreases, and this lesion may be distinguished from isolated valvular pulmonary stenosis where the systolic murmur increases in response to amyl nitrite.

VASOPRESSOR DRUGS (Table 11-4)

Vasopressor drugs such as phenylephrine hydrochloride, methoxamine, and angiotensin are less commonly used largely because of the inconvenience of intravenous administration and the greater care needed in titrating these drugs to achieve the desired effect. They are, however, occasionally beneficial. All vasopressors result in an increased systemic vascular resistance and blood pressure, with reflex reduction in heart rate and cardiac output. The physiologic responses are opposite to those of amyl nitrite. Vasopressors are helpful when trying to elicit a low intensity murmur of aortic insufficiency or mitral regurgitation. Murmurs of ventricular septal defect and the Austin-Flint rumble are also increased when a pressor is administered. The systolic murmur of HC may dramatically diminish during vasopressor infusion, whereas the murmur due to fixed obstruction of the left ventricular outflow tract is unchanged.

In summary, attention to the behavior of heart sounds and murmurs in response to respiration, cardiac cycle length, Valsalva maneuver, isometric handgrip, and postural change can be rewarding. These maneuvers seldom add more than two minutes to the cardiac examination, yet the potential benefit to the patient makes this time well spent.

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Chapter 12

Clues to Specific Cardiovascular Disorders Detectable From the General Examination of the Patient

Irwin Hoffman, M.D.

The general appearance of patients with most types of acquired adult cardiovascular disease is normal. Thus, clues to underlying cardiac disease are unlikely to be noted unless a patient is acutely ill. Tachypnea, dyspnea, orthopnea, anxiety, pallor, sweating, or central cyanosis are all nonspecific signs of an acute process, such as myocardial infarction or congestive heart failure.

A number of rare chromosomal and familial disorders are detectable in infants and children. Some of these are briefly discussed in this chapter in conjunction with their associated cardiovascular anomalies. The interested reader is referred to textbooks of pediatric cardiology for a detailed discussion of these unusual presentations.

“Inspection” of a patient may be divided into the following sequential steps, as modified from Perloff: (1) general appearance and body conformation; (2) gestures and gait; (3) detailed appearance of the face and ears; the eyes; the extremities with attention to hands and feet, muscles, tendons, and joints; the skin, the thorax, and the abdomen.

BODY CONFORMATION: POSTURE, GESTURES, AND GAIT

The most common abnormality within this category of general appearance is the Marfan syndrome (Fig. 12-1). The unusually increased stature and long extremities in which span exceeds height of the Marfan patient may be accompanied by mitral valve prolapse with myxomatous degeneration, dilatation of the aorta and sinuses of Valsalva, and even aortic dissection. Patients with Klinefelter’s syndrome have tall stature and long extremities,

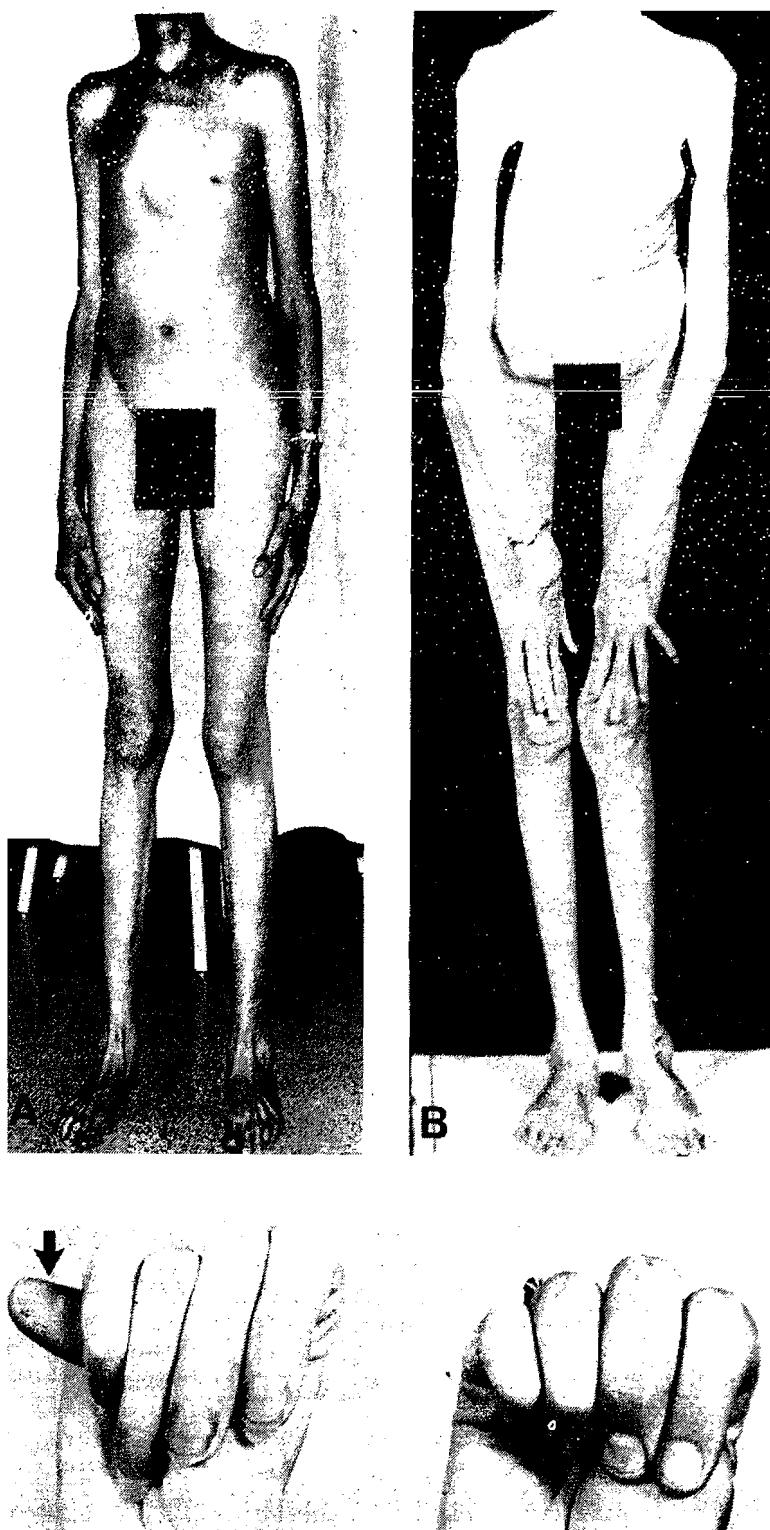


FIG. 12-1. Marfan's syndrome. This patient displays the typical body habitus of adults with the Marfan's syndrome. Note the remarkably long limbs, particularly the arms, and the thoracic pectus deformities. The clenched hand of a patient with Marfan's syndrome (left) is contrasted to a normal hand on the right. Note the arachnodactyly and excessive joint laxity. (From De Leon AC: Musculoskeletal abnormalities and heart disease. In *Signs and Symptoms in Cardiology*. Edited by LD Horwitz and BM Groves. Philadelphia, JB Lippincott Co, 1985.)

as well as a eunuchoid configuration and gynecomastia. Congenital heart diseases associated with Klinefelter's syndrome include atrial or ventricular septal defects, patent ductus arteriosus, and, in some cases, tetralogy of Fallot.

Homocystinuric patients may suffer thrombosis of intermediate sized arteries. Their body habitus includes long extremities, kyphoscoliosis, and pectus carinatum.

Increased stature may also be seen in acromegaly associated with "spade" hands and enlargement of the ears and nose. (Fig. 12-2) The associated cardiac disorders include hypertension, cardiac hypertrophy, cardiomyopathy, and conduction defects.

Abnormally short stature (dwarfism or nanism) may be seen in Turner's syndrome, associated in females with coarctation of the aorta and in males with valvular pulmonic stenosis (Fig. 12-3). The dwarfism of the Ellis-Van Creveld syndrome is associated with large atrial septal defects or common atrium.

Obesity, especially when extreme (body weights in excess of 300 pounds), may be associated with biventricular hypertrophy and heart failure. The combination of somnolence and obesity with hypoventilation resulting in hypoxemia, hypercapnia, increased pulmonary resistance, and, ultimately, pulmonary hypertension is widely known as the "Pickwickian syndrome."

In Cushing's syndrome, obesity is confined to the trunk, with a "buffalo hump" evident behind the neck and shoulders. Coronary artery disease is the associated cardiac disorder, especially if the syndrome is due to the long-term administration of adrenocortical steroids.

Although persons with any bodily habitus may acquire coronary artery disease, an obese, mesomorphic build should alert the physician to this possibility, especially if it is accompanied by premature baldness and a transverse earlobe crease (Fig. 12-4).

Generalized muscular underdevelopment may be seen in left to right shunts (as in atrial septal defects) and, especially in infancy and childhood, may be the reason an infant is brought for medical examination. Underdeveloped musculature of the legs, associated with normal or overdevelopment of the thoracic and upper extremity muscles, may be a clue to the presence of coarctation of the aorta.

In Friedreich's ataxia, the characteristic lurching gait is associated with hammer toe and pes cavus. Cardiac abnormalities seen in Friedreich's ataxia include hypertrophic cardiomyopathy (Chapter 14), anginal syndrome, and sick sinus syndrome.

A typical, waddling walk is a clue to the presence of the Duchenne type of pseudohypertrophic muscular dystrophy. These patients may have a prominent lumbar lordosis and striking pseudohypertrophy of the calves. Short Achilles tendons cause the patient to stand on his toes. The cardiac involvement in Duchenne muscular dystrophy is a posterobasal cardiomyopathy,

typically affecting the posterolateral wall of the left ventricle, resulting in an ECG picture resembling true posterior myocardial infarction.

A straight back and stiff gait are characteristic of ankylosing spondylitis, which is commonly associated with aortic valvular regurgitation and occasionally with complete heart block. Aortic regurgitation (Chapter 15) occurs in approximately 10% of these patients who survive with spondylitis for thirty years or longer.



FIG. 12-2. Acromegaly. This patient demonstrates the classic features of late-stage acromegaly, which include coarsening of facial features and swelling of the hands and fingers. The mandible is increased in size and thickness; the skull is thickened with an exaggeration of the bony ridges and muscle attachments and remarkable enlargement of the sinuses. The head, nose, lips, and ears are enlarged. The jaw protrudes and the ends of the fingers become spadelike. (From Clinico-pathologic Conference: Acromegaly, diabetes, hypermetabolism, proteinuria and heart failure.

Am J Med 20:133, 1956.)

An ataxic gait may result from syphilitic disease of the spinal cord, which may accompany tertiary luetic disease of the aorta and aortic valve, resulting in aortic valve regurgitation and aneurysm of the ascending thoracic aorta.

Squatting is typical of the child with tetralogy of Fallot. This body position increases systemic afterload, reduces the right to left shunt through the ventricular septal defect, and facilitates flow across the stenotic pulmonic outflow tract or valve. Cyanosis is part of this syndrome.

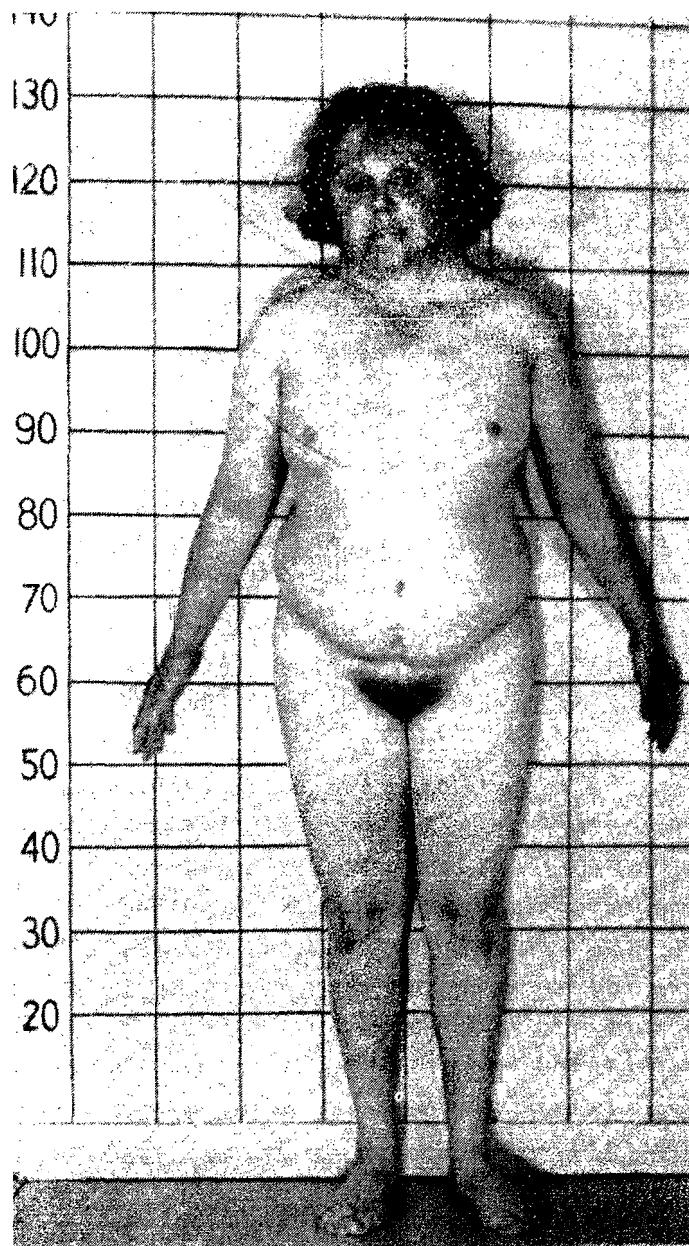


FIG. 12-3. Turner's syndrome. This patient demonstrates classic features, including short stature, lowset ears, a hypoplastic jaw, web neck, and a shield chest with wide-set nipples. These patients have only an X-sex chromosome and are referred to as XO. (From McKusick VA: A genetical view of cardiovascular disease. *Circulation* 30:326, 1964.)



FIG. 12-4. Earlobe crease. It has been suggested that the presence of a diagonal earlobe crease may identify individuals at risk for coronary artery disease. There is some controversy about validity of the correlation. (From Elliot WJ: Earlobe crease and coronary artery disease. Am J Med 75:1024, 1983.)

Perhaps the most characteristic gesture accompanying cardiac disease is the clenched fist formed by the patient with acute myocardial infarction to indicate graphically the squeezing character of his chest pain (Levine's sign).

EYES

Blue sclerae are a well-recognized manifestation of osteogenesis imperfecta, but may also be seen in Marfan's syndrome and in the Ehlers-Danlos syndrome. Examination of the iris may reveal tremulousness (iridodonesis) in Marfan's syndrome and gray-white (Brushfield's) spots in Down's syndrome (ostium primum defect). Iritis may be a manifestation of sarcoidosis, often associated with cardiomyopathy and heart block. A circumferential inferior ring around the iris is known as the arcus senilis, often associated with hypercholesterolemia and premature atherosclerosis. (Fig. 12-6A)

Cataracts commonly accompany a large variety of congenital and other cardiac diseases, of which diabetes mellitus and associated coronary atherosclerosis are the most common. Various syndromes and disease states involving the heart and associated with cataracts include polychondritis, homocystinuria, myotonia dystrophica, the maternal rubella syndrome, and syndromes named after Marfan, Down, Friedreich, and Werner. The associated cardiac disorders are discussed elsewhere in this section.

Exophthalmos is commonly seen in hyperthyroidism. However, severe right-sided heart failure of any cause, including tricuspid regurgitation, may cause ocular protrusion as a result of marked elevation of venous pressure. In the apathetic variety of hyperthyroidism, which may present with atrial fibrillation and wasting of the temporal muscles, the eyes may be sunken rather than protruded.

Bacterial endocarditis may present with petechiae in the upper or lower conjunctival sacs, or with subconjunctival hemorrhage. Retinal occlusion and hemorrhages may also be seen.

Yellow sclera are typical of jaundice, which frequently accompanies right-sided heart failure with congestive hepatomegaly. Opacities of the cornea may be seen in Hurler's syndrome, associated with mitral regurgitation and coronary stenosis.

An unusual abnormality of the iris (cat eye) results from fissuring or coloboma. This is sometimes associated with total anomalous pulmonary venous return. A small, irregular pupil, responding to accommodation but not to light, is a feature of neurosyphilis (Argyll Robertson sign), sometimes accompanied by tertiary lues of the aortic root and aortic valve. This may result in coronary ostial stenosis or valvular aortic insufficiency.

SKIN

Yellowing of the skin or jaundice can be seen in patients with right-sided heart failure, congestive hepatomegaly, and, occasionally, after pulmonary infarction. Rarely, jaundice may occur in the persistent hemolytic state secondary to a prosthetic cardiac valve, as well as in sickle cell anemia. In the latter case, the associated cardiovascular abnormalities may be pulmonary hypertension (arteriolar occlusion by sickled cells) or myocardial disease (myocardial vascular occlusion of similar nature). Large cutaneous ulcers on the legs may also be seen in sickle cell anemia. In congenital heart disease with right-to-left shunting or in patients with chronic arterial desaturation from pulmonary disorders (cor pulmonale), cyanosis and clubbing of the distal phalanges may be seen. (Fig. 12-5)

Lentigines (leopard syndrome, multiple lentigines syndrome) are brown macular skin lesions occurring on the neck and trunk, which do not increase



FIG. 12-5. Clubbing of distal phalanges. Note the swollen distal digits with filling in of the angle of the nailbed and dark discoloration of the tips of the distal fingers. (Courtesy of Dr. Jonathan Samet.)

with sunlight and, thus, are distinguished from freckles. The associated cardiac disease is hypertrophic obstructive cardiomyopathy or pulmonic stenosis. Erythema marginatum is a transitory, truncal rash commonly seen in acute rheumatic fever.

Petechiae of the skin are commonly seen with infective endocarditis.

Cutaneous scarring along the course of the antecubital veins is an important clue to intravenous drug abuse and is commonly a clue to acute infectious endocarditis.

Hereditary telangiectasias (Osler-Rendu-Weber disease) are small capillary hemangiomas widely distributed in the respiratory and gastrointestinal tract, as well as skin, tongue, and nasal mucosa. Sometimes they are associated with pulmonary arteriovenous fistulae, which are a cause of central cyanosis (right-to-left shunting).

Neurofibromatosis of the skin and café-au-lait spots may be seen in patients with pheochromocytoma of the adrenals. Another cutaneous sign of pheochromocytoma is axillary freckling (Crowe's sign).

Primary amyloidosis, which may affect the myocardium and result in a restrictive cardiomyopathy, can manifest with a thick yellowish infiltration of the skin, most common in the regions of the head and neck. These elevated,

waxy cutaneous nodules may become hemorrhagic when massaged lightly. In lupus erythematosus, a variety of cutaneous manifestations may provide diagnostic clues. A typical butterfly rash and flushing occurs on the face. The fingertips may be erythematous; a brownish-red palmar rash may be present, and, in addition, Raynaud's phenomenon of the hands, urticaria, or vitiligo may be present. The cardiac involvement in these patients includes endocarditis of the verrucous type, myocarditis, or pericarditis. The diagnosis of maternal lupus may be a clue to the development of complete heart block in the newborn.

In dermatomyositis, a typical lavender discoloration of the eyelids and dermatitis of the dorsum of the hands may be seen. The cardiac involvement includes myocardial disease, heart block, and pericarditis. In the carcinoid syndrome, chronic reddish cyanosis of the face, as well as telangiectasia, may be complicated by periodic episodes of intense flushing. The associated lesions are stenosis or regurgitation of the right-sided cardiac valves.

Sarcoidosis, which may cause a cardiomyopathy and heart block, often involves the skin, with nodules, erythema nodosum, or lupus pernio.

In Fabry's disease, purplish, tiny angiokeratomata affect the lips, trunk, and genitalia. The heart and blood vessels are infiltrated with glycolipid, with subsequent valve regurgitation, heart failure, arrhythmias, and myocardial infarction.

Livedo reticularis is a reddish-blue, mottled network resulting from interference with cutaneous blood supply. It may be seen in a variety of conditions, including lupus erythematosus (Libman-Sacks endocarditis and pericarditis) and periarteritis nodosa (arteritis of coronary and peripheral vessels). Recently, a case of livedo reticularis associated with bacterial endocarditis was described. Adenoma sebaceum (more accurately angiofibromata) may be seen in tuberous sclerosis, a condition also associated with rhabdomyoma of the heart. These are yellow or orange and are symmetrically distributed on the cheeks and nose.

Xanthomata may be seen in type II or type III hyperlipoproteinemia (Fig. 12-6). Tendinous xanthomas occur on the extensor surface of the hands, Achilles tendons, and elbows. In some cases, xanthelasma of the eyelids are markers of the type II variety and are associated with elevated blood cholesterol, premature atherosclerosis, and coronary artery disease. Palmar and tuberoeruptive xanthomata are markers of type III (broad beta) hyperlipoproteinemia and are also associated with atherosclerosis. Cutaneous eruptive xanthomata are small (up to 2 mm in size), yellowish nodules, which may occur anywhere on the body and are markers of elevated levels of chylomicrons. They are common in patients with type I or type V hyperlipoproteinemia and are not markers of vascular disease.

The general appearance of nonerupted skin may provide clues to underlying disease states associated with cardiac abnormalities. Yellow, thickened skin, especially in the antecubital and neck regions, is seen in

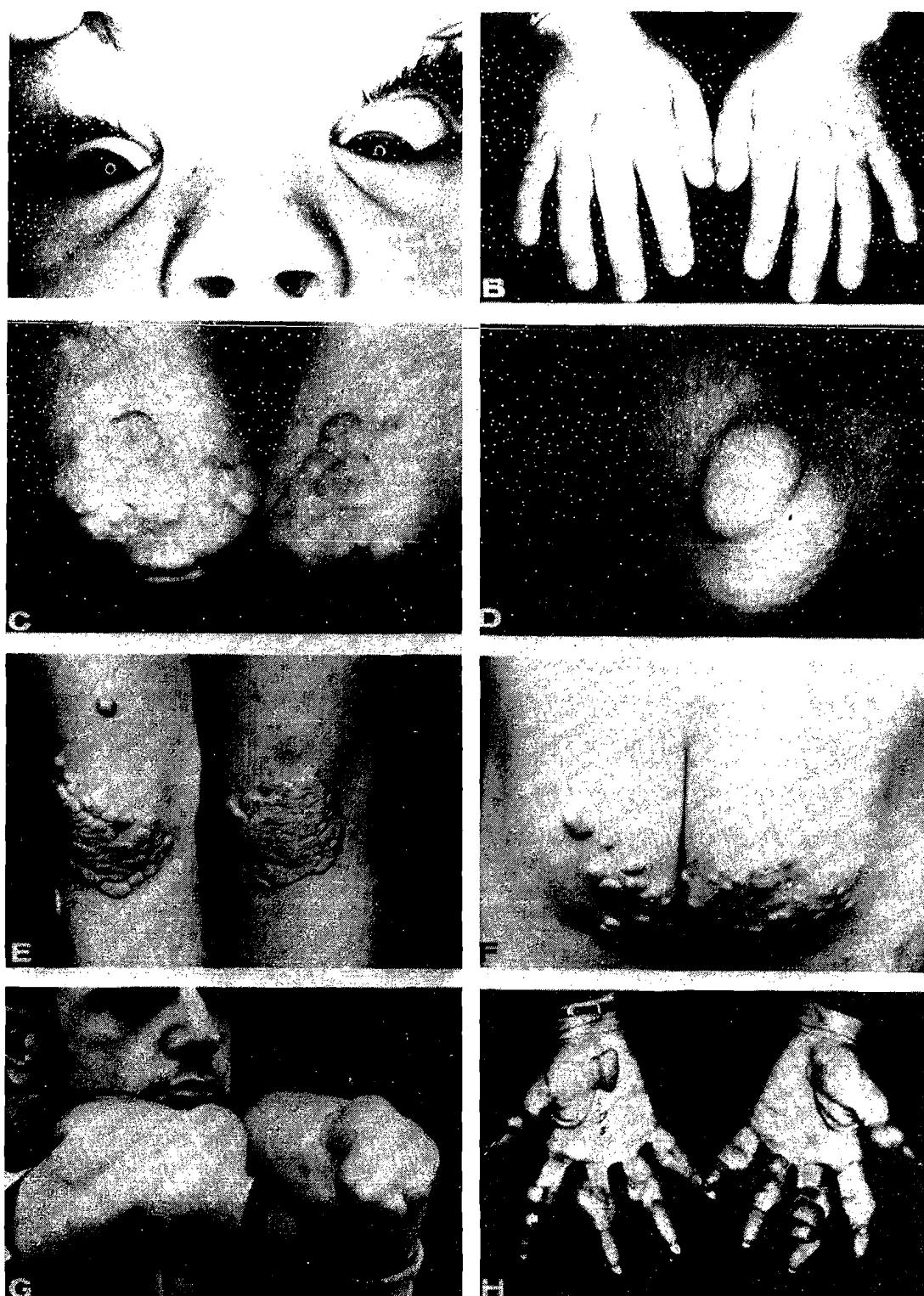


FIG. 12-6. Xanthomata and lipid deposits associated with familial hypercholesterolemia (homozygotes). A. Arcus cornea. B,C,E,F. Cutaneous planar xanthomas. D,G: Tuberous xanthomas. H: Tendon and tuberous xanthomas. (From Goldstein JL and Brown MS: Familial hypercholesterolemia. In *The Metabolic Basis of Inherited Disease*, 5th ed. Edited by JB Stanbury et al, New York, McGraw-Hill Book Co, 1983.)

pseudoxanthoma elasticum. In this condition, angina and claudication are secondary to calcified arteries. In Raynaud's syndrome, the skin of the hands is smooth, hairless, and glossy (lupus erythematosus, scleroderma) in the absence of ischemic attacks.

Myxedematous skin is coarse and dry (pericardial effusion), whereas thyrotoxic skin (atrial fibrillation, congestive heart failure) is usually fine, moist, and silky. Pretibial myxedema is a localized, nonpitting edema of the shins, occasionally seen in hyperthyroidism. In Cushing's syndrome, the skin is ruddy (polycythemia), but more easily permits needle puncture (loss of subcutaneous elastic tissue). Atherosclerosis and hypertension are common cardiovascular complications.

In the Name syndrome, profuse, cutaneous pigmented lesions, as well as subcutaneous myxoid neurofibromata, are associated with atrial myxoma. In this syndrome, the cutaneous lesions are variously blue nevi and pigmented macules, but the subcutaneous nodules are myxoliposarcomas.

Easily hyperextensible skin with charnois-like texture is seen in the Ehlers-Danlos syndrome. The fragility and poor healing of the skin results in scarring that resembles cigarette paper. These patients may have mitral or tricuspid prolapse and dilatation of the aorta. In trisomy 16-18, the skin may be lax and wrinkled. The associated cardiac defects are patent ductus, right ventricular origin of both great arteries, or ventricular septal defect.

EXTREMITIES

Cyanosis and clubbing of the distal phalanges of both hands and feet indicate a right to left intracardiac shunt (Fig. 12-5). Central mixing of this type also occurs with pulmonary arteriovenous fistula and with drainage of the inferior vena cava into the left atrium. Diagnostic clues to specific shunts may be present when differential cyanosis and clubbing are present. When clubbing and cyanosis are found in the feet and left hand, the likely diagnosis is reversed shunting through a patent ductus arteriosus in the presence of pulmonary hypertension. The shunt occurs distal to the left subclavian artery and, consequently, the right hand is spared.

"Reversed differential cyanosis" (and clubbing) means that the hands are cyanotic and clubbed, but the feet are not. This condition occurs when the aorta originates from the right ventricle (transposition), arising above a ventricular septal defect. The pulmonary trunk receives oxygenated blood. If pulmonary resistance is high, oxygenated pulmonary artery blood may flow through a patent ductus arteriosus to the descending aorta and to the feet, which thus will be less cyanotic and clubbed than the hands.

Clubbing of the distal phalanges may also be seen in bacterial endocarditis (especially subacute), in bronchogenic carcinoma (a clue to pericardial met-

astatic disease, pericardial effusion, and atrial arrhythmias), and in sarcoidosis (cardiomyopathy, heart block).

"Tuft erythema" refers to a bright red coloration of the fingertips, which are painless and not clubbed, in the presence of small or intermittent right to left shunts. Persistent cyanosis may result from an increase in the size or a protracted duration of the right-to-left shunt.

The thumb may be malformed in several syndromes that are associated with congenital heart disease. Rubinstein-Taybi's syndrome, also known as the broad thumb-hallux syndrome, consists of broad terminal phalanges of the thumb and big toe with kyphoscoliosis and associated congenital heart disease; pulmonic stenosis is the most common lesion, but patent ductus arteriosus and double aortic arch have also been described. In the ventricular-radial dysplasia syndrome, hypoplasia or aplasia of the radius, with residual or absent thumb, is associated with ventricular septal defect. In the Holt-Oram syndrome, the thumb may be absent, rudimentary, or finger-like. The common associated cardiac defect is ostium secundum atrial septal defect. Commonly, simple polydactyly is associated with a ventricular septal defect. However, if the defect is in the atrial septum, it is usually of the ostium primum type.

In the Ellis-Van Creveld syndrome, the nails may be dysplastic, and polydactyly may be present. The cardiac association is an atrial septal defect.

In the Smith-Lemli-Opitz syndrome, cutaneous syndactyly of the second and third toes may be seen. Twenty percent of these patients have a ventricular septal defect, patent ductus arteriosus, or endocardial cushion defect.

In Down's syndrome (ostium primum defect), in addition to the well-known simian crease in the palm, the fourth and fifth fingers are abnormally separated, with the fifth finger being short and curved inward. In the Hurler syndrome, the hands have a clawlike appearance and are wider than long.

In the trisomy 16-18 syndrome, the fingers bend and overlap (clinodactyly) and promptly return to the overlapped position after opening. Protruding heels produce a "rocker-bottom" appearance to the feet. The associated cardiac anomalies are ventricular septal defect, right ventricular origin of the great arteries, or patent ductus arteriosus.

Short fingers and hyperconvex fingernails are seen in Turner's syndrome, in which short stature, medial deviation of the extended forearm, and cubitus valgus also occur (Fig. 12-3). The associated cardiac lesion is coarctation of the aorta.

In constrictive pericarditis with protein-losing enteropathy, chronic hypoalbuminemia may result in parallel striping of the nails. In acromegaly, spadelike hands and sausage-shaped fingers are commonly seen. Myocardial hypertrophy and hypertension may be present clinically (Fig. 12-2).

Tight, shiny skin with contraction of the fingers and ulcerations of the knuckles and fingertips may be seen in scleroderma, often associated with

pulmonary hypertension as well as myocardial, pericardial and valvular disease (Fig. 12-7).

In Marfan's syndrome, arm span typically exceeds the excessive height, and the joints are lax (Fig. 12-1). Spidery fingers and toes may be present. Mitral valve prolapse, often with regurgitation, and a dilated proximal aorta are common; aortic dissection may occur in these patients. Hypermobile joints may also be seen in the Ehlers-Danlos syndrome (large ASD). In osteogenesis imperfecta, extremity and bony deformity result from the recurrent fractures, especially of the long bones. The associated cardiac lesion is mitral regurgitation.

In myotonic dystrophy, distal limb weakness in the hands and feet precedes weakness in the shoulder and pelvis. Neurologic examination reveals abnormalities of muscular relaxation. Cardiac abnormalities include bundle branch block and abnormal axis as well as atrioventricular block. Mitral valve prolapse may also be seen.

"Rockerbottom" feet may be seen in De Lange's syndrome, associated in 15 to 20% of cases with ventricular septal defect, patent ductus arteriosus, or pulmonic stenosis.

Sudden pallor or cyanosis of toes or fingers associated with small, tender areas on the skin of the fingers are clues to the presence of systemic emboli.



FIG. 12-7. Scleroderma. This photograph shows classic changes of scleroderma or the CREST syndrome. The skin is smooth, shiny, and tight. Telangiectasias are present. Note the loss of small wrinkles on the distal interphalangeal joints. (Courtesy of Dr. Tedd Goldfinger.)

These may originate from various sources, including atherosclerotic lesions of the aorta, cardiac myxomata, clots or infection on prosthetic heart valves, atrial myxomas, or intracardiac thrombi.

In bacterial endocarditis, two rather specific types of embolic lesions occur. Osler's nodes are 3 to 4 mm raised swollen areas on the pads of the fingers or toes. These are tender, in contrast to the much less common Janeway lesions that occur on the palms or soles.

Rheumatoid arthritis, with typical hand deformity resulting from subluxation of the metacarpophalangeal joints and ulnar deviation of the digits, may be associated with subclinical pericardial, valvular, or myocardial disease in up to 60% of patients.

A rare chronic postrheumatic fever arthritis (Jaccoud's arthritis) may also result in ulnar deviation. It is associated with valvular stenosis and insufficiency of the mitral and aortic valves.

FACE

In Turner's syndrome, hypertelorism (wide spacing between the eyes) may be associated with ocular epicanthal folds as well as pigmented moles. A webbed neck may also be present. (Fig. 12-3) The associated cardiac abnormality is coarctation of the aorta.

The expression "cardiofacial syndrome" has been applied in several quite different cardiologic situations. Patients with one well known "cardiofacial syndrome" manifest hypertelorism, elfin facies, and low set ears. These facies commonly are associated with stenotic lesions of the pulmonic or aortic valves.

Williams' syndrome combines the above facies with supravalvular aortic stenosis and hypercalcemia. In Noonan's syndrome, similar facies are associated with webbing of the neck (pterygium colli) and a low posterior hairline. These patients manifest valvular pulmonic stenosis, as well as the Turner "phenotype," inasmuch as their chromosomal studies do not show typical Turner's syndrome XO chromosomal characteristics.

In congenital valvular pulmonic stenosis with a domed valve, the patient may present with a round chubby face and healthy appearance. Another "cardiofacial syndrome" has been described, in which a unilateral, partial lower facial weakness is apparent only when the patient cries. This finding may be elicited in infants with congenital heart disease, particularly a ventricular septal defect.

In Werner's syndrome the patient has grayed hair, frontal baldness, ocular proptosis, and rapidly progressing cataracts. Cardiac manifestations include coronary and systemic atherosclerosis.

The typical manifestations of the Klippel-Feil syndrome are low set ears, a low hairline, and a short neck. The associated cardiac disease is congenital ventricular septal defects.

Atrial or ventricular septal defect may be seen in the children of heavy drinkers (fetal alcohol syndrome). These infants present a characteristic appearance, which includes a hypoplastic upper lip, midfacial and mandibular growth deficiency, and short palpebral fissures.

In Cornelia de Lange's syndrome, the patient presents with long eyelashes; a small mandible; a broad, flat upturned nose; and bushy, confluent eyebrows. The associated congenital abnormality is a ventricular septal defect.

In Hurler's syndrome, the common cardiac abnormalities are stenosis or regurgitation of the mitral and/or aortic valves, sometimes associated with coronary artery stenoses, arterial hypertension, and thickened endocardium. The facial appearance of these patients is best described as "gargoylism." The facial features are bizarre and grotesque, with malformations of the skull and supraorbital ridges. The pharynx is deformed, with secondary nasal discharge, the tongue is large and protrudes, the neck is short, and the bridge of the nose is depressed. The basic abnormality is an underlying mucopolysaccharidosis.

In Down's syndrome (trisomy 21, mongolism), the circumference of the skull is reduced and there is anteroposterior flattening. An epicanthic fold is also characteristic. The tongue protrudes; the nose is short with a flat bridge. The neck is short and broad and the lower jaw hypoplastic. The associated cardiac condition is an endocardial cushion defect.

In Rubinstein-Taybi's syndrome, the forehead is prominent, the nose beaked, the ears large and low set. The slant of the eyes is anti-mongoloid. The congenital heart anomaly is usually ventricular septal defect (see "Extremities").

In myotonia dystrophica, the patient presents with a myopathic facies (drooping eyelids and masklike expression). A receding hairline and cataracts may also be present. The associated cardiac disease includes conduction disorders, arrhythmias, and mitral prolapse.

Polychondritis of the facial cartilage may result in saddle-shaped collapse of the nose and in cauliflower ears. The associated cardiac disease is aneurysm or dissection of the aorta and aortic valve insufficiency.

The typical hypothyroid facies (scanty, dry hair; loss of the lateral eyebrows; dry, thickened skin; and macroglossia) may be associated with pericardial effusion, hypercholesterolemia, and coronary atherosclerosis with myocardial infarction. Recently recognized, a lateral earlobe crease has been associated with coronary artery disease (Fig. 12-4).

Systolic pulsations of the earlobe are common in the presence of severe tricuspid valve regurgitation (Fig. 19-7).

In the leopard syndrome (multiple lentigines), hypertelorism is associated with pulmonic stenosis, as well as idiopathic hypertrophic subaortic stenosis, also known as hypertrophic cardiomyopathy (see Chapter 14).

In scleroderma, the facial clues are a thickened, leathery skin tightly bound to the underlying tissue. Distinctive immobility about the mouth is

evident. The cardiovascular pathology typically is pulmonary hypertension. Systemic hypertension may also be present.

In the Kearns-Sayres syndrome, drooping of the lids and external ophthalmoplegia are due to muscular dystrophy of the extraocular muscles. The associated heart diseases are complete heart block and heart failure.

In severe rheumatic mitral stenosis, the typical facies include flushed cheeks and cyanotic lips. Telangiectasis of the face are part of the carcinoid syndrome, which also can include a chronic reddish, cyanotic coloration and episodes of intense flushing. The usual cardiac lesions are stenosis and regurgitation of the tricuspid and/or pulmonic valves and are due to the release of a variety of vasoactive substances from metastatic tumor deposits to the liver.

In the Larsen syndrome, the face appears flattened and has frontal bossing, a depressed nasal bridge, and a flattened facial appearance with hypertelorism. These patients may present with aortic root dilatation, aortic insufficiency, and aneurysm of the ductus arteriosus.

In the Pierre Robin syndrome, a hypoplastic mandible allows the tongue to retract and obstruct the airway. The congenital heart disease present may be ventricular or atrial septal defect.

THORAX AND ABDOMEN

The characteristics of a thoracic bulge may provide a clue to an underlying cardiac disorder. For example, ventricular septal defect (Chapter 21) in an adult commonly will result in a left parasternal bulge, located in the fourth left intercostal space, and the same defect in a child often results in bilateral bulging of the upper anterior chest including the upper two thirds of the sternum. Atrial septal defects (Chapter 20) may result in left parasternal bulges located in the second and third interspaces.

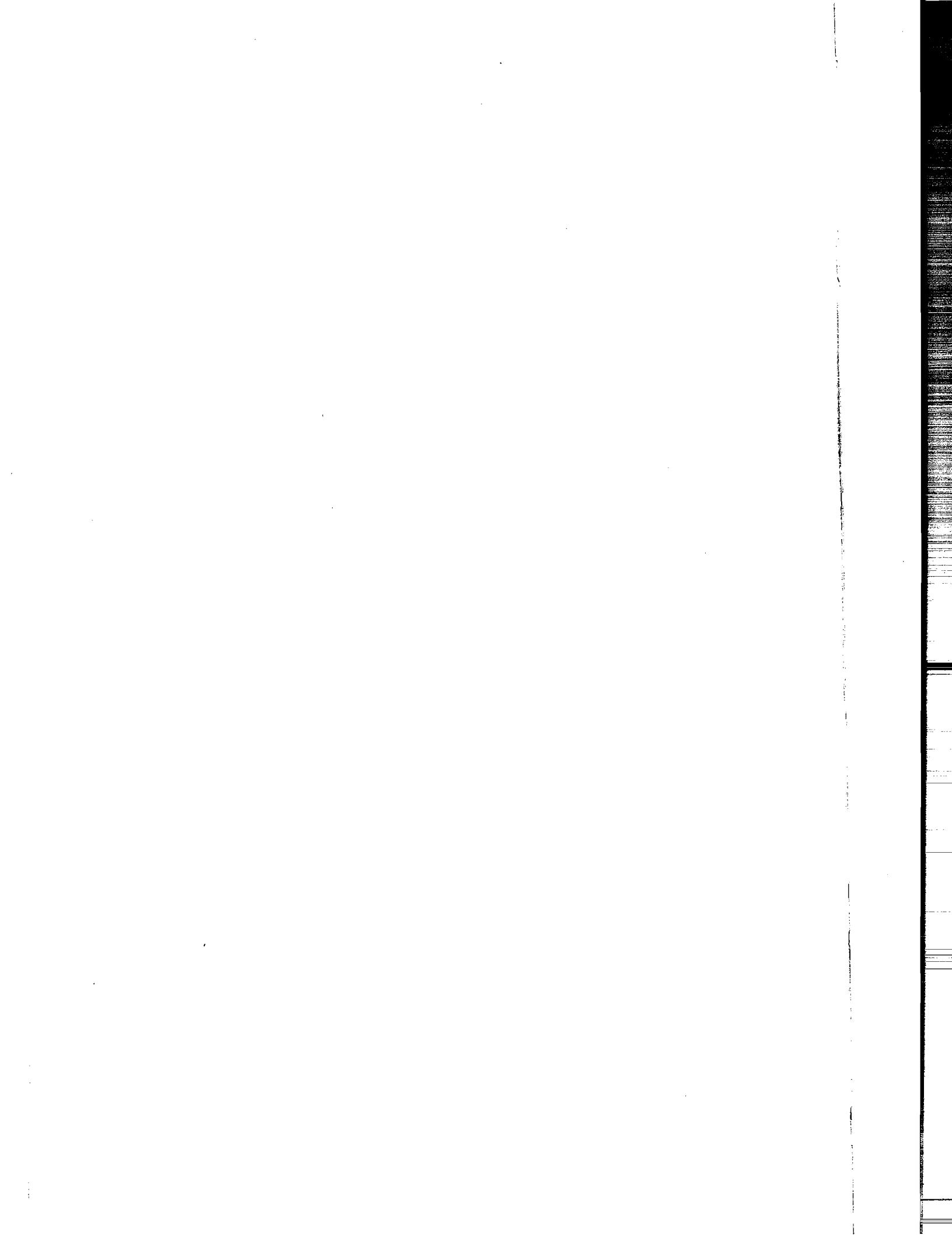
Obvious tortuous vessels pulsating around the scapula or along the lateral chest wall may represent the collateral arterial circulation in coarctation of the aorta. Bilateral distension of the jugular venous system plus obvious cutaneous venous distension of the chest wall is seen in superior vena cava obstruction.

Structural deformities and neuromuscular disorders of the thorax may alter normal respiration and lead to hypoventilation, hypoxia, vasoconstriction, chronic pulmonary hypertension, and right heart failure. A variety of disorders in addition to congenital kyphoscoliosis may initiate this chain of events. Some of these underlying conditions include rheumatoid spondylitis, thoracoplasty, poliomyelitis, the muscular dystrophies, and osteogenesis imperfecta.

The straight back syndrome and pectus excavatus reduce the anteroposterior thoracic dimension and displace the heart to the left (pancake heart). Systolic and, rarely, diastolic murmurs may be present, but true organic heart disease is uncommon. However, there is an increased incidence of straight back syndrome in patients with atrial septal defect (Chapter 20). Pectus excavatum and straight back syndrome are also seen in patients with mitral valve prolapse (Chapter 18).

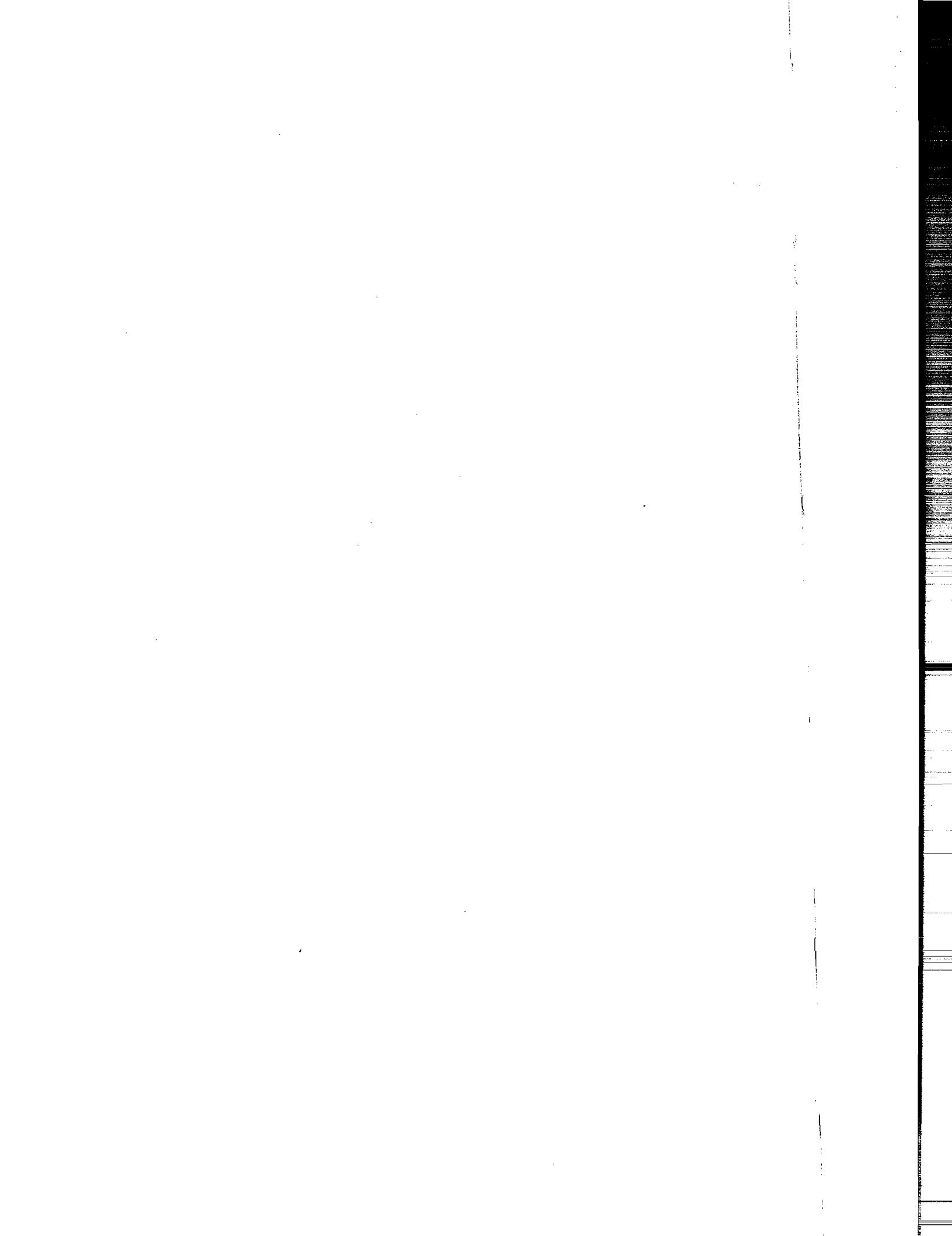
Pulsation of the sternoclavicular joint may be a manifestation of aneurysm of the ascending aorta. In Turner's syndrome (aortic coarctation), inspection of the chest may reveal widely spaced nipples (shield chest) and hypoplastic breast tissue (Fig. 12-3). Gynecomastia, unilateral or bilateral, may be seen in patients receiving digitalis preparations or the steroid aldosterone inhibitor, spironolactone.

In chronic obstructive pulmonary disease with pulmonary emphysema, thoracic kyphosis is common, as well as an increased anteroposterior chest diameter and sternomanubrial bowing. Right upper quadrant pulsations are usually due to cyclic hepatic expansion most commonly seen in tricuspid insufficiency (systolic expansion) or tricuspid stenosis (diastolic expansion). Abdominal ascites may sometimes be evident on inspection of the patient and is a clue to the presence of right-sided heart failure or constrictive pericarditis.



PART II PHYSICAL FINDINGS IN SPECIFIC CARDIOVASCULAR CONDITIONS

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Chapter 13

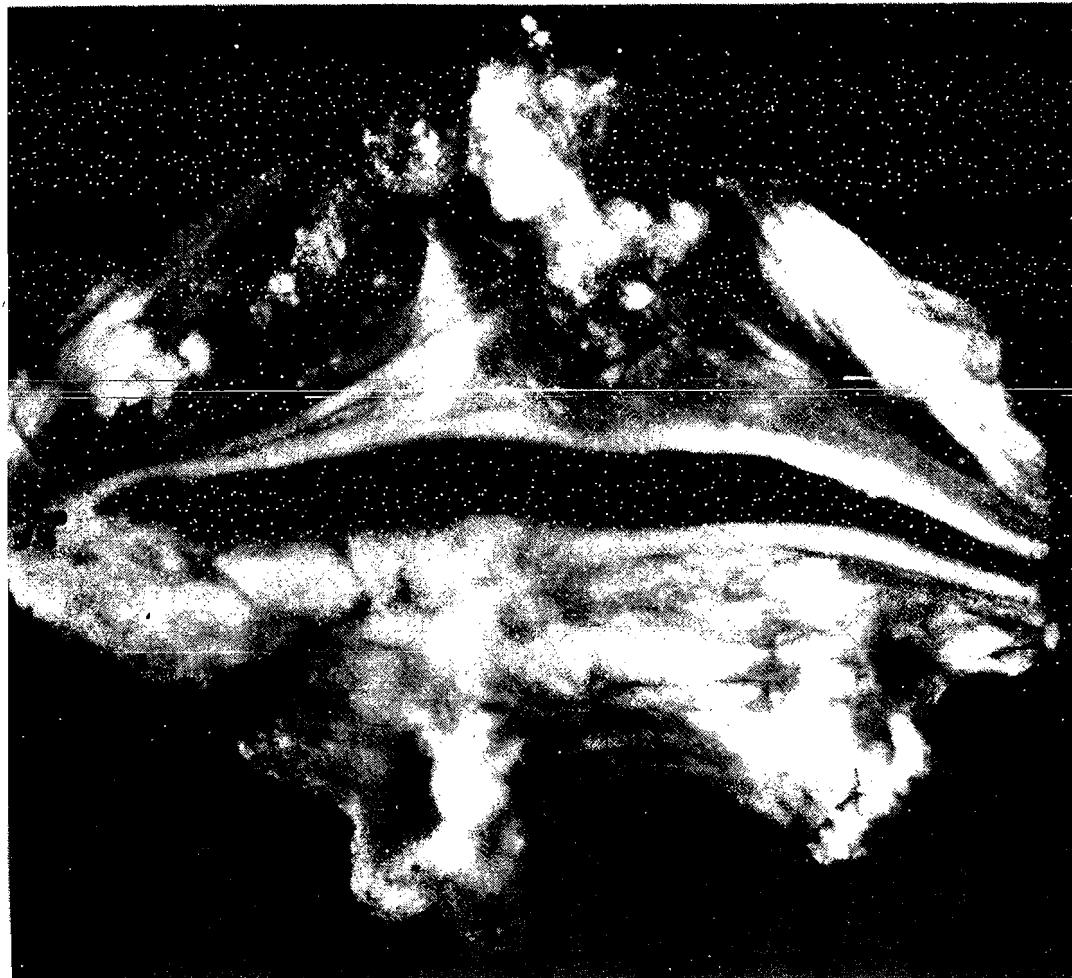
Aortic Stenosis

Aortic stenosis represents a valve lesion in which the physical findings are directly related to the underlying hemodynamic burden. Actually, several types of obstruction to left ventricular outflow fall into the general category of aortic stenosis. These include stenosis of the aortic valve itself, as well as the rare varieties of supravalvular and subvalvular obstruction. This chapter will focus on cardiac physical diagnosis in *valvular aortic stenosis*. The reader is referred to textbooks or reviews of congenital heart disease for details about congenital supravalvular and subvalvular stenosis. Hypertrophic cardiomyopathy (IHSS), a far more common entity, will be discussed in Chapter 14.

ETIOLOGY

Valvular aortic stenosis has three major causes: congenital, rheumatic, and degenerative. The majority of cases of isolated aortic stenosis in patients under 50 years of age are due to a congenital defect, usually a bicuspid aortic valve. This anomaly is perhaps the most common congenital cardiac abnormality, estimated to occur in 1 to 2% of the general population. It is not known how many of these functionally normal bicuspid valves will ultimately thicken and/or calcify and become stenotic (Fig. 13-1A, B). Many persons with this lesion have a structural abnormality of the valve (audible ejection click with or without a short systolic murmur), but show no evidence of obstruction to blood flow. In patients with coexistent mitral valve disease with or without a prior history of acute rheumatic fever, rheumatic heart disease is the likely cause of aortic stenosis (Fig. 13-1C). In older adults with isolated aortic stenosis but without evidence for other valve involvement, by far the most common etiology is degenerative stiffening and thickening of the leaflets of an inherently normal tricuspid aortic valve (Fig. 13-1D). Calcification often occurs, leading to a further reduction in orifice area (Fig. 13-1A, C, D).

Frequently, older persons with hypertension develop nonobstructive thickening of the base of the aortic valve cusps. This is known as *aortic sclerosis* and is the most common cause of a systolic murmur in the elderly population. Aortic sclerosis may also be present in the absence of hypertension. This murmur can simulate obstructive aortic stenosis, and it is important to differentiate the two entities (see below). It is not known whether aortic



A.

B.

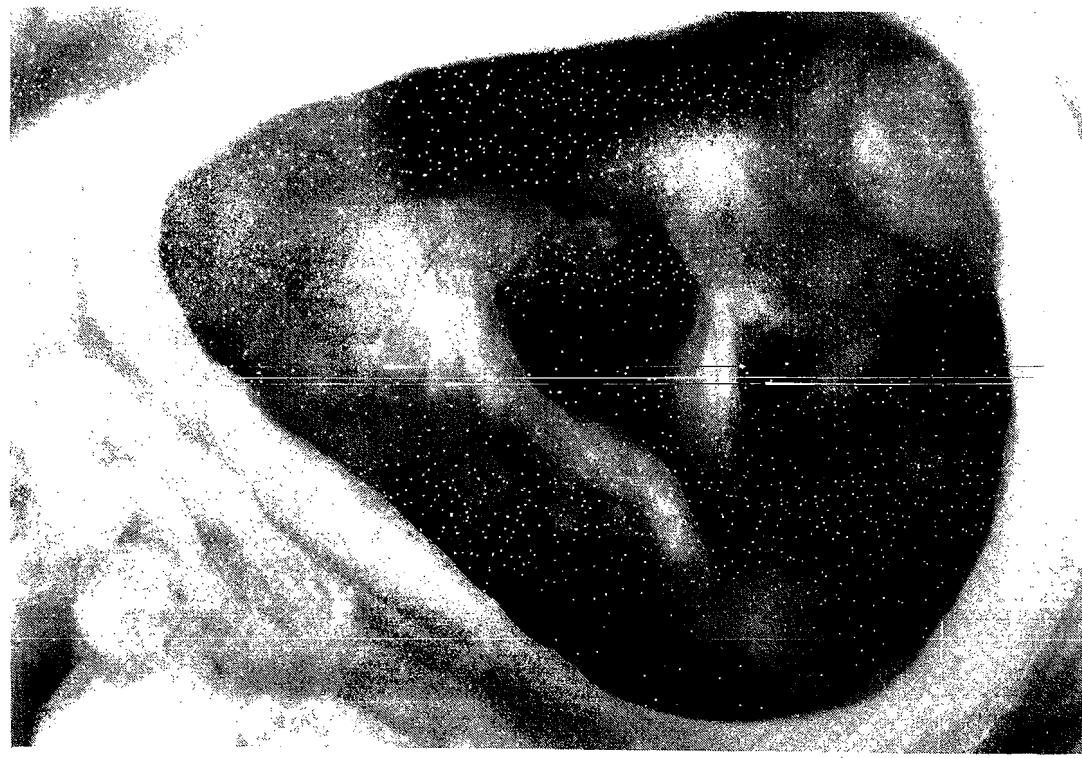
FIG. 13-1. Pathologic specimens of stenotic aortic valves. A. Congenitally bicuspid valve with calcification in valve pockets and mild fusion of the right commissure (*). B. Postrheumatic aortic stenosis. There is an eccentric commissural lesion of all three commissures with heavy calcification. C. Postrheumatic valvulitis. Note fusion of all three commissures, resulting in a small triangular valve orifice. Extensive calcification is present, a common finding in postinflammatory aortic stenosis. D. Bicuspid aortic valve. This valve does not have a raphe. The leaflets are rigid. Calcification is present, an extremely common finding in all bicuspid aortic valves in the adult population. (A and C, from Subramanian R, Olson LJ, and Edwards WD: Surgical pathology of pure aortic stenosis: a study of 374 cases. Mayo Clinic Proc 59:683, 1984. B and D, from Becker AE and Anderson RH: Cardiac Pathology. New York, Raven Press, 1983.)

sclerosis represents an early stage in the development of degenerative aortic valve stenosis, is a forme fruste of this lesion, or is totally unrelated.

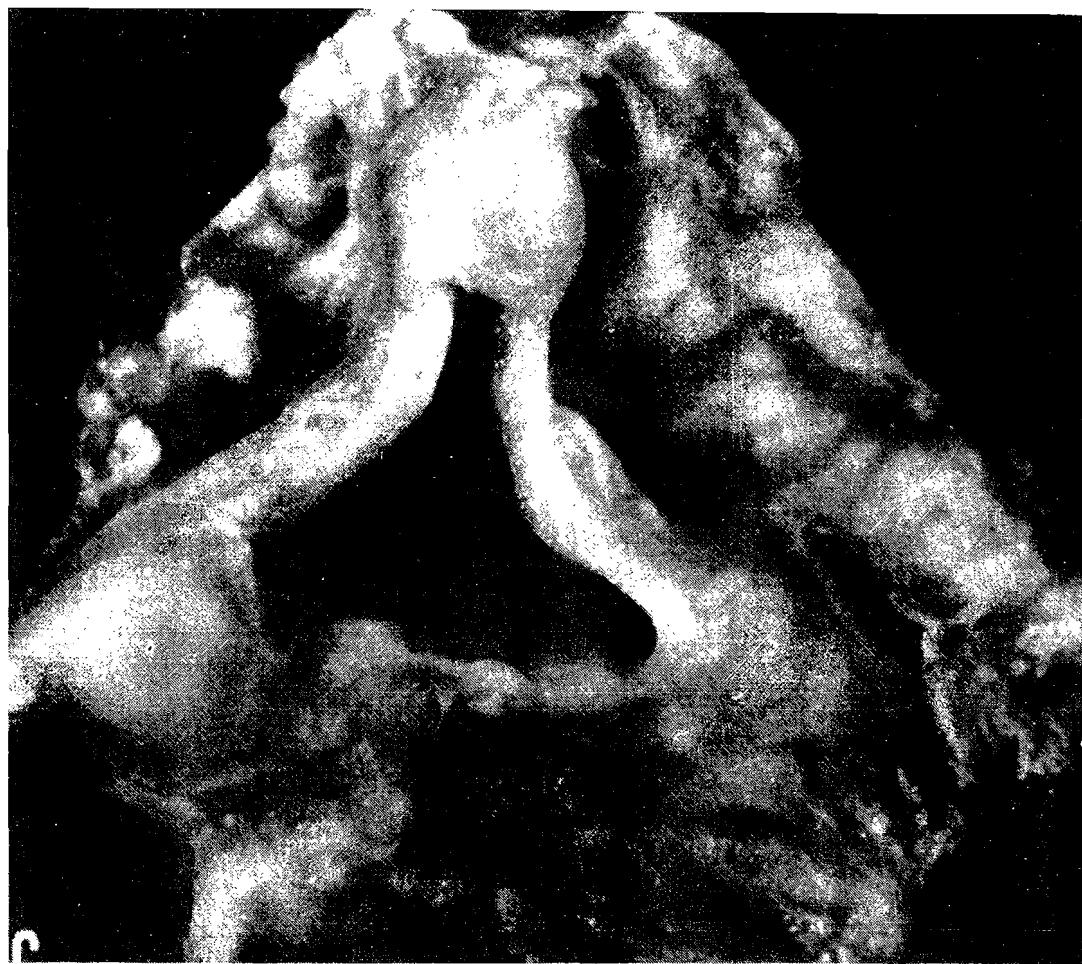
PATHOPHYSIOLOGY

Most of the physical findings in aortic stenosis can be explained by an understanding of its pathophysiology. It is possible to assess the severity of the aortic obstruction by integration of information obtained from palpation of the carotid and apex impulses with careful auscultation. The normal aortic

C.



B.



C.

(Legend on Facing Page)



D

(Legend appears on page 206.)

valve area in adults is quite large, ranging between $2\frac{1}{2}$ to $3\frac{1}{2}\text{ cm}^2$. The valve has three cusps or leaflets of equal size. Reduction of the size of the aortic orifice does not result in a pressure drop across the valve at rest until at least 50% of the valve area is narrowed (i.e., aortic valve area of approximately 1 cm^2). However, a prominent systolic murmur may be present even in mild degrees of aortic stenosis. As the aortic orifice further narrows, left ventricular pressure becomes elevated in order to maintain forward blood flow and a normal systemic arterial pressure. Hemodynamically significant aortic stenosis occurs when the aortic valve area is reduced by 60 to 75%, resulting in a calculated aortic valve area of 0.7 to 0.8 cm^2 . Symptoms are likely to appear at this stage, and definite evidence of left ventricular hypertrophy is usually present. Moderate aortic stenosis is present when the left ventricular-aortic gradient is 50 to 60 mmHg. Severe aortic stenosis is present when the gradient is 75 to 80 mmHg or higher, assuming a normal stroke volume. The onset of left ventricular dysfunction and congestive heart failure may lower substantially the peak-to-peak systolic gradient, and the characteristic physical findings of severe aortic stenosis may be masked in the setting of left ventricular failure.

In aortic stenosis the left ventricle undergoes concentric hypertrophy to compensate for the increased resistance to outflow; this process usually produces a very thick-walled chamber with no dilatation of the LV cavity. Thus, LV mass is substantially increased, but left ventricular volume remains normal

or only slightly increased. *Practical Point:* Severe left ventricular dilatation and enlargement, as detected by palpation, chest roentgenogram, or echocardiography, is not a characteristic of compensated, pure aortic stenosis. Left ventricular hypertrophy resulting from obstruction to LV outflow results in a noncompliant chamber with an elevated LV filling pressure. The force of left atrial contraction increases; this may be manifest clinically by a palpable and/or audible S4, large P waves on the electrocardiogram, and an increased left atrial size on the echocardiogram.

Left ventricular systolic pressure must increase markedly to maintain adequate flow across a severely obstructed valve. In time the velocity of cardiac fiber shortening decreases; in major outflow obstruction, the left ventricular ejection time is prolonged and the rate of rise of central aortic pressure is markedly decreased. With long-standing aortic stenosis, left ventricular contractile force is reduced. When this occurs, the heart dilates and symptoms of congestive heart failure, angina, or syncope develop. Paradoxically, the physical findings of aortic stenosis may become *less impressive* with the onset of left ventricular dilatation and congestive heart failure.

PHYSICAL EXAMINATION

Blood Pressure

In the absence of hypertension or associated aortic regurgitation, the systolic blood pressure is normal in most subjects with aortic stenosis. With advanced degrees of aortic obstruction, the pulse pressure narrows as systolic arterial pressure decreases. A pulse pressure of 30 mmHg or less may be found in severe aortic stenosis, but this is a nonspecific finding. Even in severe aortic stenosis only a minority of patients will have a systolic blood pressure of 100 mmHg or less.

Contrary to popular belief, systemic arterial *hypertension* can be associated with significant valvular aortic stenosis, particularly in the older population. Nevertheless, it is most unusual for the systolic blood pressure to be greater than 170 to 180 mmHg in a patient with moderately severe aortic stenosis. Systolic blood pressures greater than 140 mmHg often are found in older subjects with aortic stenosis. When aortic regurgitation is also present, the blood pressure may be altered if the degree of aortic "leak" is substantial. In this setting, systolic arterial pressure will be higher and diastolic pressure lower.

Carotid Arterial Pulse

A hallmark of aortic stenosis is the typical slow-rising, small volume arterial pulse (Figs. 3-5, 13-2, 13-6, Table 13-1). Many experts have empha-

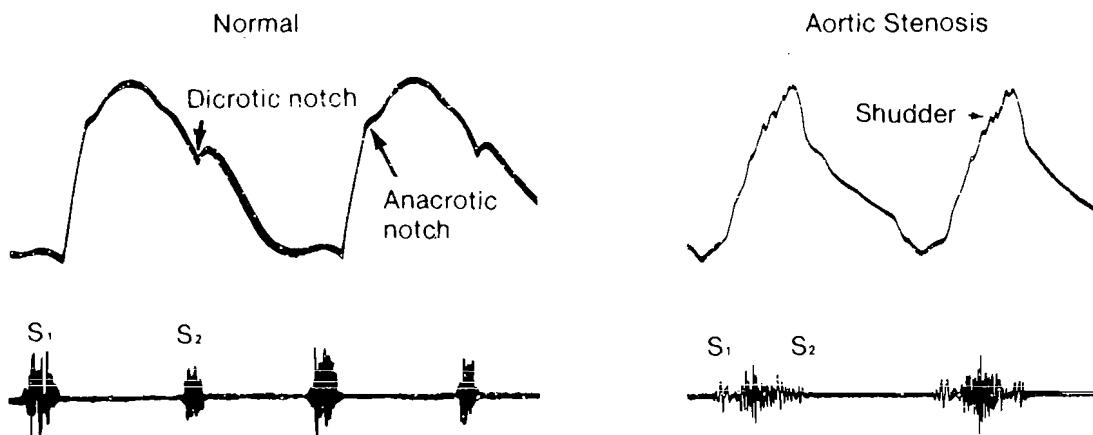


FIG. 13-2. Carotid arterial pulse in aortic stenosis. On the left, a diagram of the normal carotid contour is shown. On the right is a carotid pulse tracing from a patient with significant valvular aortic stenosis. Note the delayed upstroke and shudder that represents the transmitted murmur. The pulse volume is usually diminished in aortic stenosis (*pulsus parvus et tardus*). (From Abrams J: Prim Cardiol, 1983).

sized the plateau quality of the pulse in aortic stenosis, i.e., a smooth, prolonged peak with a more gradual drop-off. Usually, there is an associated systolic thrill or shudder on the upstroke of the pulse, but this is not invariable. The classic pulse in aortic stenosis is often called *pulsus parvus et tardus* (slow and late) or an *anacrotic* pulse. The latter term refers to the lowered anacrotic notch or shoulder that can be seen on graphic recordings of the carotid pulse. Other recordable phenomena include a lower and later dicrotic notch and a prolonged left ventricular ejection time. Palpation alone cannot detect the anacrotic or dicrotic notch.

In efforts to assess the severity of the aortic valve obstruction, many investigators have derived various formulae and indices utilizing the duration of the carotid upstroke and left ventricular ejection time. It is easy to be confused by these reports, which are dependent on high quality phonocardiograms. In general, these approaches have not been widely applied, and bedside evaluation remains important. *Practical Point: The classic carotid pulse in aortic stenosis has a slowed upstroke, reduced amplitude, and sustained contour, with or without an accompanying palpable thrill (Figs. 3-5, 13-2, 13-6, Table 13-1).*

In detecting aortic stenosis, the focus of the examination of the arterial system should be on the carotid arteries and not on the peripheral pulses.

TABLE 13-1 *Cardinal Features of the Carotid Pulse in Valvular Aortic Stenosis*

-
- Slow-rising (*pulsus parvus*)
 - Delayed peak (*pulsus tardus* or *plateau pulse*)
 - Small volume
 - Palpable thrill
 - Prominent anacrotic notch (not usually palpable)
-

The normal alterations of increased amplitude and rate of rise of the arterial pulse wave contour in the peripheral circulation minimizes the diagnostic usefulness obtainable from palpation of the brachial or radial arteries (Fig. 3-2).

Severity. The severity of the valvular obstruction is roughly proportional to the degree of abnormality of the carotid pulse. Unfortunately, many factors may also affect the pulse contour, and it is not uncommon to seriously under- or overestimate the severity of aortic stenosis by carotid artery palpation (Table 13-2). *Practical Point:* In an adult under 60 years of age with clinically normal left ventricular function (no congestive heart failure or gross cardiomegaly), a completely normal carotid pulse contour and arterial pulse pressure generally excludes moderate to severe aortic stenosis. A distinctly delayed carotid upstroke in the same setting is consistent with moderate to severe valve obstruction. Occasionally, the carotid pulse may be abnormal with a small volume or slow rate of rise, yet the left ventricular-aortic gradient is small. This condition is usually due to an aortic valve deformity without major obstruction coupled with a low cardiac output, thus simulating a small volume, delayed contour (Table 13-2). The converse may also occur with a relatively unremarkable pulse contour and amplitude in the presence of significant aortic valve stenosis. Carotid pulsations will not be visible when there is significant obstruction to the left ventricular outflow unless there is associated aortic regurgitation.

Factors Leading to Underestimation of Severity of Aortic Stenosis from Carotid Pulse Analysis. Table 13-2 lists factors or associated conditions that may mask the severity of aortic stenosis (see also Chapter 3). In children with congenital aortic stenosis, the arterial pulse may be only mildly deranged or even normal in the face of a large aortic valve gradient. Presumably, this is due to the highly elastic and compliant peripheral vasculature of children. A far greater problem in older adults is the loss of arterial elasticity that occurs with aging with or without associated systolic hypertension. In this setting, a normal or reduced stroke volume ejected across the stenotic valve may not produce the typical slow upstroke, small volume pulse of aortic

TABLE 13-2 Pitfalls in Evaluating the Arterial Pulse in Aortic Stenosis

Factors that can "normalize" the arterial pulse and mask the apparent severity

- High cardiac output and elastic vessels in children and young patients
- Increased stiffness of vessels in the elderly
- Associated aortic regurgitation
- Systemic hypertension
- Low stroke volume of congestive heart failure

Factors that can exaggerate the apparent severity of aortic stenosis

- Decreased left ventricular function
- Hypovolemia
- Mitral stenosis

stenosis because of the decreased compliance of the systemic arteries. Thus, a "normal" rate of rise and pulse amplitude will be misleading.

The presence of a low cardiac output and stroke volume, with or without overt congestive heart failure, may also mask the typical carotid pulse of severe aortic stenosis. The reduced pulse volume may render the delayed upstroke and shudder impalpable. Thus, in the setting of a low stroke volume, the arterial pulse can be quite misleading in aortic stenosis. Associated mitral stenosis can also attenuate the classic pulse contour of aortic stenosis.

Aortic regurgitation, a frequent accompaniment of aortic stenosis, may have an important effect on the arterial pulse of aortic stenosis. The increased stroke volume and rate of ejection in aortic regurgitation may result in a carotid upstroke with a fuller pulse volume and a greater rate of rise than found in an equivalent degree of aortic stenosis without regurgitation. The greater the valvular leak, the greater the "normalization" of the small volume, slow-rising carotid pulse. Associated aortic regurgitation typically will result in bisferiens or palpable double systolic impulse (see Chapters 3, 15). One must be sure that this pulse abnormality is not due to a prominent carotid shudder. A bisferiens pulse in a patient with aortic stenosis implies moderate to severe aortic regurgitation and well-preserved left ventricular function, as this finding may disappear with deterioration of left ventricular contractility.

Pulsus Alternans. In "tight" aortic stenosis with left ventricular dysfunction, subtle alternation of the peak systolic pressure and rate of rise is common. This is known as pulsus alternans. Every second beat is stronger than the preceding impulse (Figs. 3-4, 13-3). The heart sounds and systolic murmur may also alternate in intensity. This alternation in pulse volume is best detected using very light finger pressure; most experts suggest using the radial artery to identify this important abnormality. Pulsus alternans is caused by mechanical alternans of the left ventricle and is usually associated with an S3. It is more easily observed for several beats following a PVC. Pulsus alternans is an uncommon finding in subjects under 40 years of age.

Congenital Supravalvular Aortic Stenosis. Patients with this abnormality usually have selected "jet streaming" of blood into the right innominate vessels, resulting in a greater pulse amplitude in the right subclavian, brachial, and carotid arteries. The systolic blood pressure may be 10 to 20 mmHg higher in the right arm than in the left.

Jugular Venous Pulse

The venous pulse configuration and pressure are unremarkable in aortic stenosis. Occasionally, the jugular A wave will be prominent in the absence of an elevation in mean jugular pressure; this effect probably results from decreased right ventricular compliance in the setting of a hypertrophic left

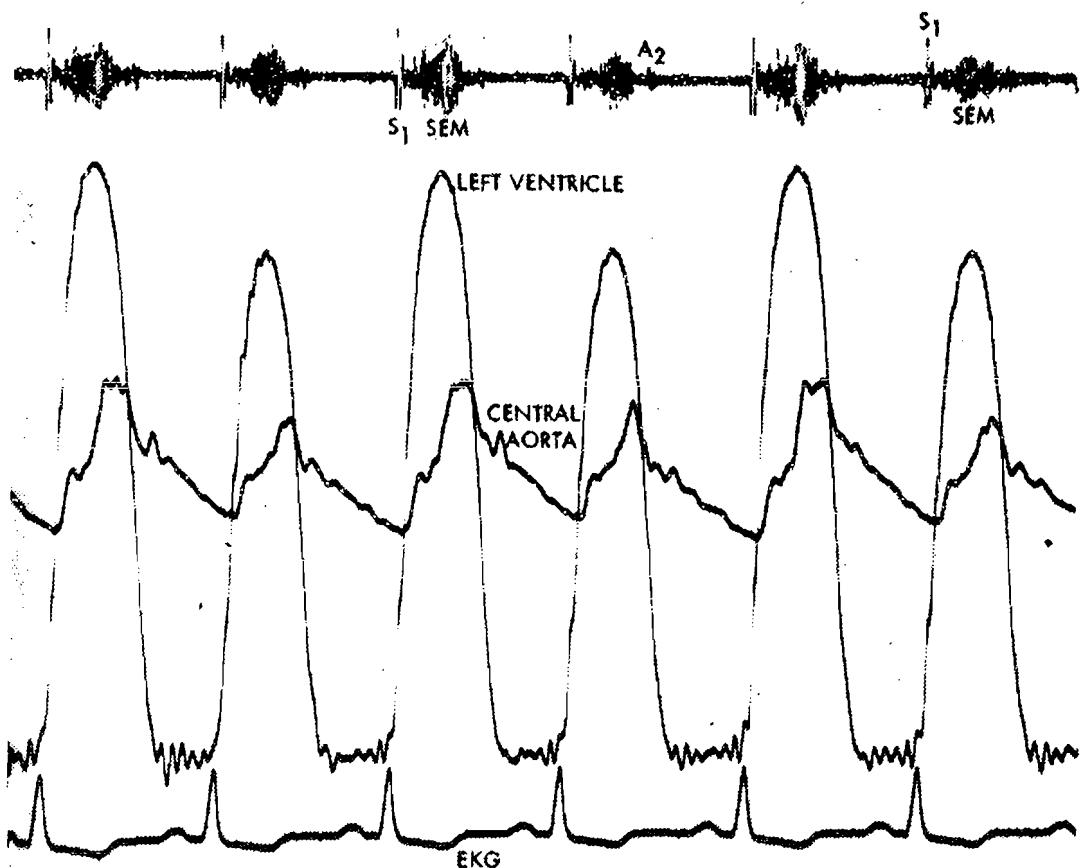


FIG. 13-3. Pulsus alternans. A phonocardiogram is recorded simultaneously with left ventricular and central aortic pressure tracings in a 55-year-old male with severe calcific aortic stenosis. Note the beat-to-beat alteration in the intensity of the murmur, correlating with beat-to-beat changes in peak systolic left ventricular and aortic pressure. The presence of pulsus alternans indicates incipient or actual left ventricular dysfunction. (From Reddy PS, Shaver JA, and Leonard JJ: Cardiac systolic murmurs: Pathophysiology and differential diagnosis. Prog Cardiovasc Dis 14:1, 1971.)

ventricular chamber and septum (Bernheim effect). If biventricular heart failure is present, the mean jugular pressure will be elevated.

Precordial Motion

The apex impulse is hemodynamically important, compensated aortic stenosis is typically a *sustained* left ventricular lift with little or no leftward displacement of the point of maximal impulse (PMI) (Fig. 13-4). The duration and force of the left ventricular impulse is increased due to the increased left ventricular mass, high intraventricular pressure, and obstruction to ventricular outflow.

A normal LV impulse is characteristic of mild aortic stenosis; in subjects with lung disease, obesity, deep chests, or large breasts, the apex beat may be diminutive or impalpable even in the presence of severe aortic stenosis.

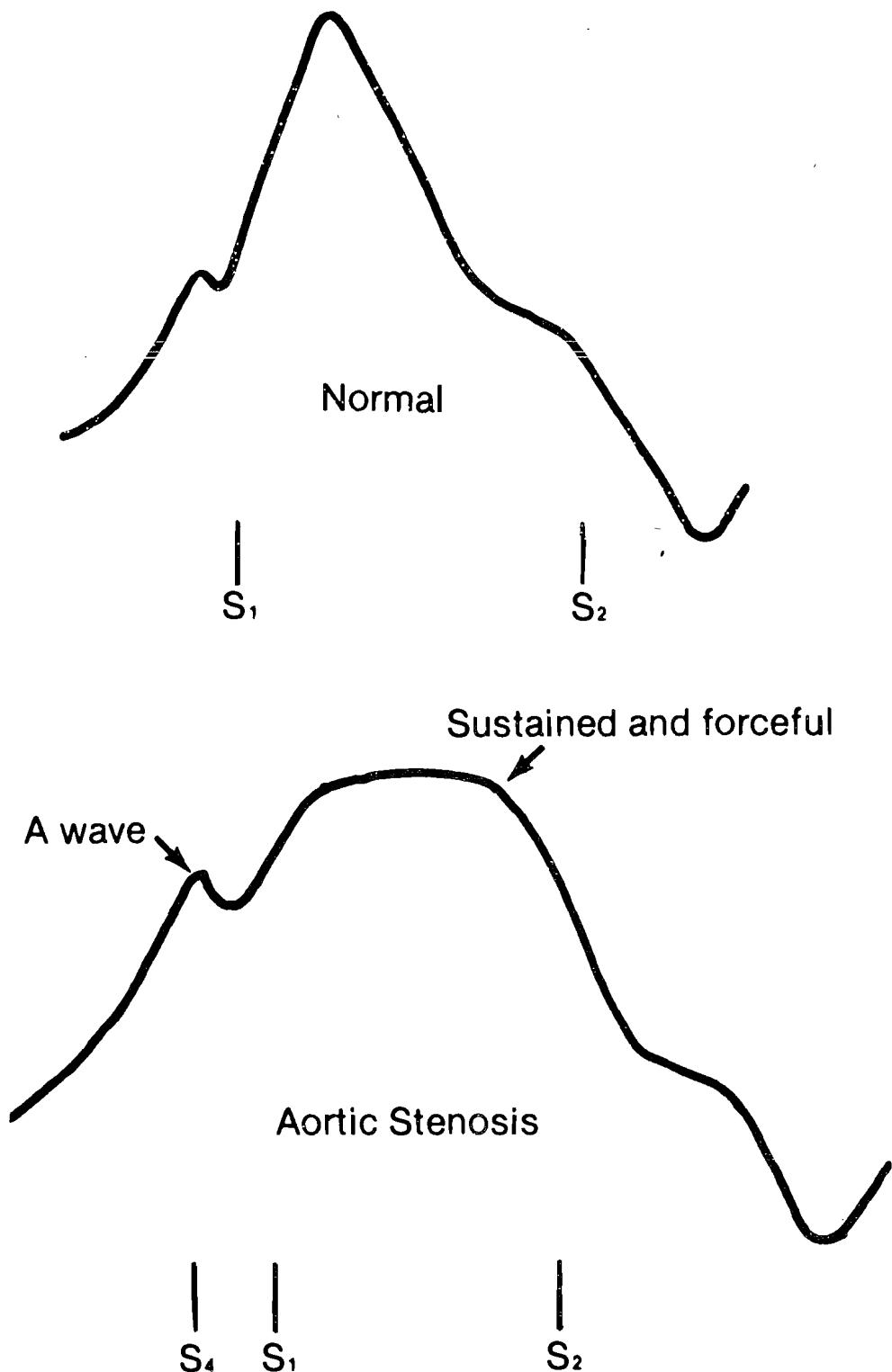


FIG. 13-4. Precordial motion in aortic stenosis. The classic left ventricular impulse in left ventricular pressure overload due to aortic stenosis is concentric hypertrophy without left ventricular dilatation, producing a sustained impulse that remains palpable into late systole. The impulse is typically more forceful than normal. Careful palpation will frequently detect a palpable A wave (S_4). (From Abrams J: Prim Cardiol, 1983.)

The presence of a sustained but otherwise unimpressive left ventricular impulse in an older subject with a large chest and a long systolic murmur suggests important aortic obstruction. *Practical Point: The concentrically hypertrophied left ventricle of isolated aortic stenosis does not produce significant lateral or downward displacement unless cardiac dilatation has occurred or there is associated aortic regurgitation.* During precordial palpation, pay close attention to the *duration* of the apex impulse, which will be sustained into the second half of systole in significant aortic valve obstruction (see Chapter 5).

A palpable A wave (presystolic distension of the left ventricle) in the supine or left decubitus position is a most valuable observation (Figs. 7-6, 13-4). This finding indicates elevation of left ventricular end diastolic pressure and suggests a thick, noncompliant chamber. In the absence of other cardiovascular conditions (e.g., coronary or hypertensive heart disease), a palpable A wave correlates with a left ventricular-aortic gradient equal to or greater than 70 mmHg. The palpable atrial thrust can be found in the absence of an audible S4.

Systolic Thrill. The loud, harsh (grade III to IV/VI) murmur of aortic stenosis often is accompanied by a systolic thrill. The most common location of the thrill is the first or second right intercostal space, with radiation upward and rightward towards the neck and right shoulder (Fig. 9-2). The thrill is often detectable at the suprasternal notch and right supraclavicular area. In some patients, a thrill can be felt at the second or third left interspace or cardiac apex. *Practical Point: For optimal detection of the systolic thrill, ask the subject to sit up and lean forward while holding his breath in expiration.* This is particularly true for obese patients or those with emphysema or deep chests.

A systolic thrill is the rule, not the exception, in aortic stenosis. Detection of a thrill indicates that aortic stenosis is present, but does not necessarily indicate severe obstruction. Systolic thrills are more easily felt in thin subjects, and are particularly common in subjects with associated aortic regurgitation.

First Heart Sound

The first heart sound is usually unremarkable in isolated aortic stenosis. It may be decreased in intensity, but is never accentuated. A loud "S1" in a patient with suspected or proven aortic stenosis suggests the presence of an aortic ejection sound or associated mitral stenosis. S1 may be decreased in amplitude when left ventricular diastolic pressure is high or if left ventricular contractility is impaired. In the former case, the high ventricular filling pressure will result in premature closure of the mitral valve. The status of left ventricular contractility directly affects the amplitude of S1 (see Chapter

- 6). In patients with aortic stenosis and congestive heart failure, S1 is quite soft as a result of both mechanisms.

Second Heart Sound

Because abnormalities in the amplitude of A2 and in the inspiratory behavior of S2 are common in aortic stenosis, careful assessment of S2 is particularly useful in the evaluation of patients with aortic stenosis.

Intensity of A2. In patients with pliable and relatively thin aortic leaflets, A2 may be normal or even increased in amplitude. This is typical of congenital aortic stenosis (bicuspid valve) without calcification and occasionally may be found in young subjects with severe aortic obstruction. However, as thickening and rigidity of the aortic leaflets ensues, the amplitude of A2 decreases. The leaflets no longer act as a vibrating membrane during aortic valve closure. Characteristically, older patients with degenerative or rheumatic aortic stenosis have a damped A2 due to the marked distortion and change in physical characteristics of the valve tissue. Calcification of the valve usually contributes to the diminished loudness of A2. A2 may be totally inaudible in severe aortic stenosis. *Practical Point: The amplitude of the aortic ejection click and that of A2 are closely related. Both are prominent in a subject with a pliable, noncalcified bicuspid valve. Both are decreased in intensity in the presence of calcium or significant valvular thickening.*

Because A2 is commonly soft or absent in aortic stenosis, a normal or increased P2 (e.g., as might occur with associated pulmonary hypertension or congestive heart failure) can readily be mistaken for A2. In older patients with aortic calcification and coexisting arterial hypertension, A2 may be normal or increased, which gives a false impression of the valve quality.

Splitting of S2 in Aortic Stenosis. Hemodynamically important aortic stenosis produces abnormalities in the sequence of S2 splitting (Fig. 13-5). The characteristic alteration of S2 is an increase in the Q-A2 interval with A2 "moving into P2" and a tendency for S2 to become single. Two factors play a role in delaying A2: (1) an increased duration of LV ejection and (2) the prolonged time for LV pressure to drop below aortic pressure at end-systole in patients with a large left ventricular-aortic gradient. Typically, these factors result in a single S2 during expiration and inspiration. If LV ejection time is substantially delayed, reversed or paradoxical splitting of S2 occurs (see Chapter 6; Figs. 6-9, 6-10, 13-5). *Practical Point: Detection of paradoxical splitting of S2 in aortic stenosis in the absence of left bundle branch block or impaired left ventricular function is an important observation, which implies that the left ventricular-aortic gradient is 75 mmHg or greater, e.g., severe aortic stenosis.*

Normal splitting of S2 is generally found in mild aortic stenosis in adults (Fig. 13-5), and is common in congenital aortic stenosis, even when severe. A single S2 may result from a decreased intensity of A2 due to fibrocalcific

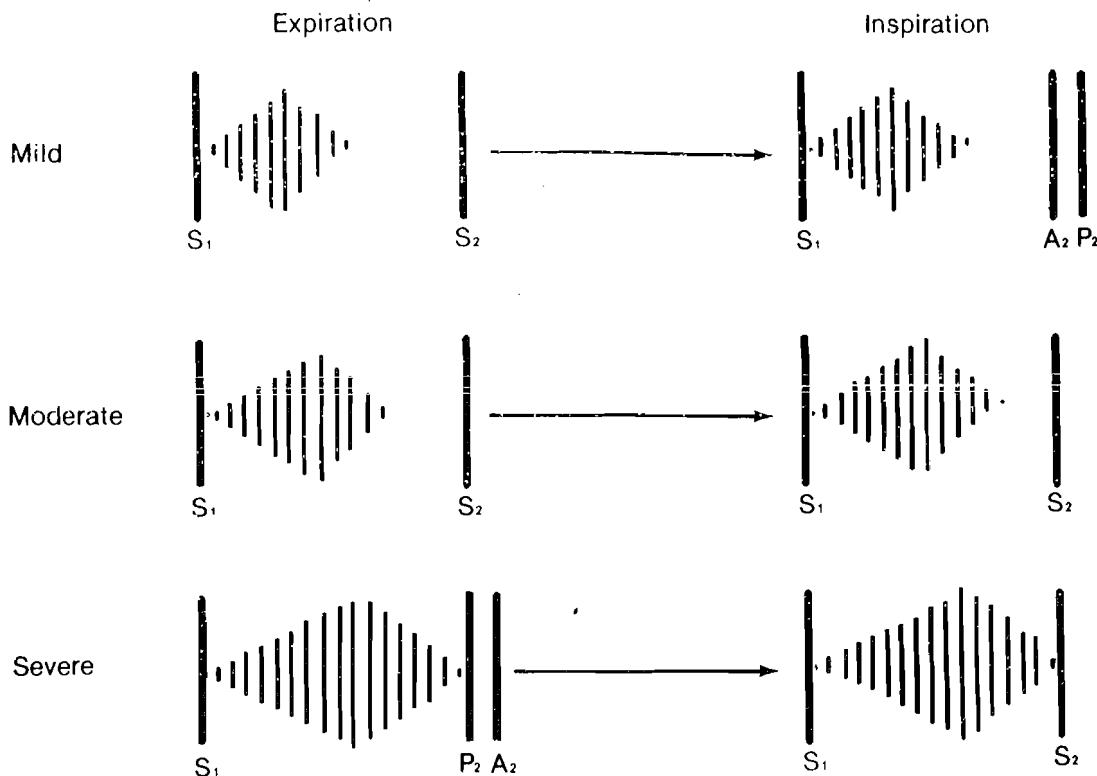


FIG. 13-5. Patterns of S2 splitting in aortic stenosis. Upper: In mild degrees of left ventricular obstruction splitting of S2 remains physiologic. Middle: Frequently, a single S2 is heard in aortic stenosis, reflecting moderate obstruction; the delayed A2 results in fusion of the two components of S2. Marked attenuation in the intensity of A2 from severe fibrocalcific changes may cause A2 to be inaudible and result in a single S2 (which is actually P2). Lower: With advanced aortic stenosis, left ventricular ejection time is prolonged sufficiently such that in expiration aortic valve closure follows pulmonic closure. During inspiration P2 moves away from S1 and may become synchronous with A2, resulting in paradoxical or reversed splitting of S2. (From Abrams J: Prim Cardiol, 1983). (See also Fig. 6-9.)

changes, which are probably a major cause of the single S2 in many patients with acquired aortic stenosis. *Practical Point:* In approximately two thirds of older patients with moderate to severe aortic stenosis, S2 will be single. S2 will be reversed or paradoxical in another 20 to 25% and will be completely normal in a small number of patients.

Problems in Listening to S2 in Aortic Stenosis. A prominent, long systolic murmur can mask audible splitting of S2; therefore, one must listen carefully for the characteristics of splitting at the apex, second right, and second left interspace. Typically, P2 is not audible at the apex unless there is pulmonary hypertension. Thus, a single apical second sound usually represents A2. If the systolic murmur of aortic stenosis is loud and prolonged, A2 or P2 may be "buried" within the murmur and be inaudible. It is therefore best to focus on the characteristics of S2 at sites away from the maximal murmur intensity. In subjects with deep chests or COPD, P2 may be quite soft or inaudible during inspiration, giving a false impression of a single or

even absent S2. In subjects with prolonged right ventricular ejection (e.g., RBBB) and audible expiratory splitting of S2, the inspiratory decrease in the intensity of P2 can produce apparent paradoxical splitting of S2, falsely suggesting severe aortic stenosis.

Third Heart Sound

An S3 is not a normal or expected finding in adults with aortic stenosis. Its presence suggests significant left ventricular dysfunction or overt congestive heart failure. When an S3 is heard, look for evidence of left ventricular dilatation as well as pulsus alternans. In such cases, the left ventricular impulse often will be displaced laterally. The S3 may be palpable in early diastole. Remember that an S3 is a normal finding in children and young adults and has no diagnostic significance if found in a patient with congenital aortic stenosis who is under the age of 35 or 40.

Fourth Heart Sound

An S4 is valuable clue to the presence of LV hypertrophy and decreased LV compliance associated with severe aortic stenosis, but only in younger patients (e.g., age 12 to 40). The presence of an audible or palpable S4 correlates with a large left ventricular aortic-gradient (greater than 70 mmHg) and an abnormally elevated left ventricular end-diastolic pressure (LVEDP). Because an S4 is normal in young children and is commonly found in older patients with coexisting atherosclerotic heart disease or hypertension, its significance is considerably reduced in these age groups. *Practical Point: The presence of an audible S4 in an older patient does not necessarily mean the aortic stenosis is severe.* The finding of a palpable S4 (presystolic distention) at any age (Fig. 13-4) in a patient with aortic stenosis implies that the valve obstruction is of major hemodynamic importance.

Aortic Ejection Click (see also Chapter 8)

The detection of an aortic ejection click or sound is an important observation (Fig. 13-6). (1) It confirms the diagnosis of structural heart disease in a patient with a systolic ejection murmur. (2) It localizes the abnormality to the aortic valve (in the absence of a dilated aorta or chronic hypertension). (3) It suggests that the etiology of the valve abnormality is a congenitally deformed aortic valve, usually a bicuspid valve. *Practical Point: Although an ejection click indicates that the deformity is at aortic valve level, its presence does not correlate with the severity of the lesion.* The aortic valve may be functionally normal without any reduction in size of the orifice, particularly when the systolic murmur is soft, short, or absent.

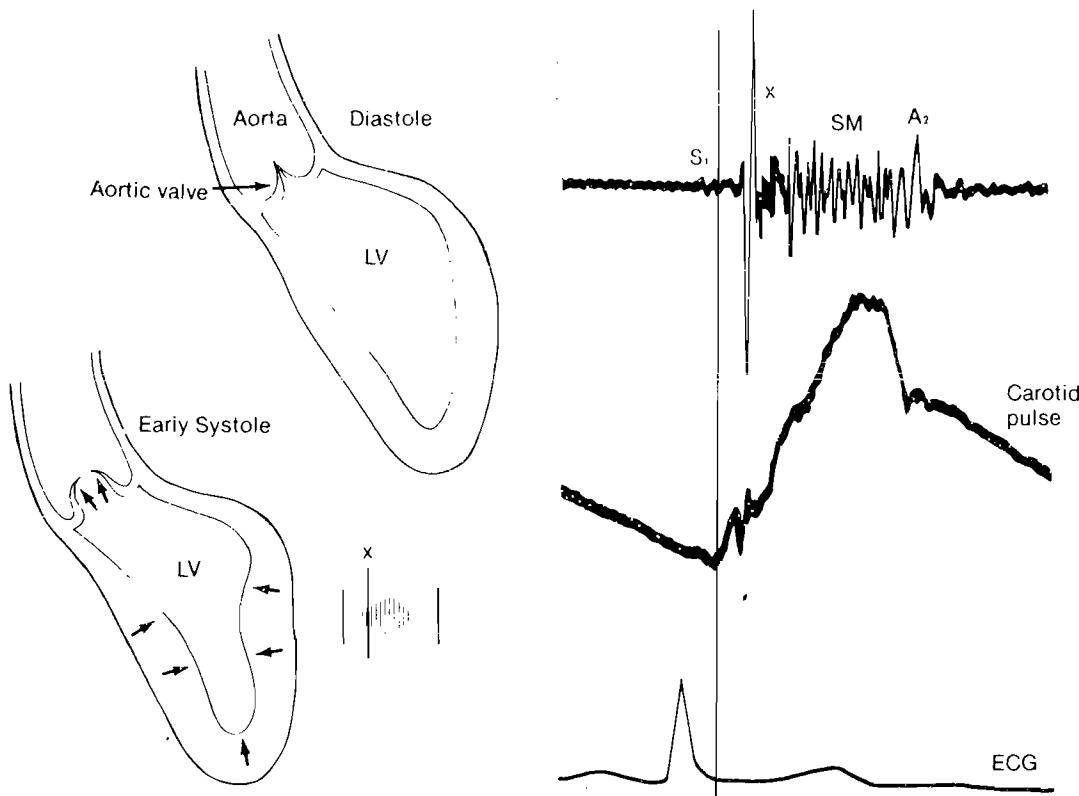


FIG. 13-6. Ejection click associated with aortic stenosis due to a congenitally bicuspid valve. Note the high frequency, high amplitude sound that follows S₁ and is coincident with the onset of ejection into the aorta. The aortic ejection sound is formed by sudden cessation of the opening motion of the abnormal valve leaflets (doming). Note also the delayed carotid upstroke and long systolic murmur. (From Abrams J: Prim Cardiol 1983.)

The ejection click is related to the maximal upward excursion of an abnormal bicuspid or unicuspid valve (Fig. 13-1A, B), occurring precisely when the piston-like upward motion of the valve leaflets abruptly halts (Figs. 8-2A; 13-6). The leaflets must be sufficiently pliable to produce an ejection sound. With increasing calcification and thickening, the valve cusps lose mobility and their excursion lessens; the click softens and ultimately may disappear.

An ejection sound is uncommonly found in *acquired* aortic stenosis on a tricuspid aortic valve. When there is rheumatic scarring and distortion of the valve or with thickening and calcification of the basal leaf typical of degenerative aortic stenosis of the elderly, the valve cusps are too thick and rigid to open rapidly during early ejection and a click does not occur. As already discussed, the intensity of aortic closure (A₂) mirrors that of the aortic ejection sound. A₂ is well preserved, even loud, in young patients with congenital aortic stenosis, but is attenuated and often absent in the fibrocalcific varieties of rheumatic and degenerative aortic stenosis.

Practical Point: Systolic clicks are present in less than one third of older

patients with aortic stenosis (> 50 years of age), but are the rule in congenital aortic stenosis. Clicks are less commonly heard in severe congenital aortic stenosis.

How to Listen to the Ejection Click or Sound. The ejection click is a high frequency, crisp sound (Figs. 8-1, 8-3, 13-6). It occurs 40 to 80 msec after S1, a longer interval than that separating the normal mitral and tricuspid components of S1. *Contrary to what one might expect, the aortic ejection sound is usually best heard at the cardiac apex, where it is often louder than both S1 and S2.* A typical click has a “snappy” quality and is heard throughout the precordium. It is readily mistaken for S1, particularly at the base of the heart. A loud “discrete S1” heard at the first or second interspace on either side of the sternum should raise the possibility that this is actually an ejection sound, as S1 is not usually prominent at the base.

Ejection sounds or clicks are not commonly considered or sought in routine cardiac auscultation. One should remember that a “split” S1 at the apex and lower left sternal border may represent an S1-ejection click complex. Occasionally, a click is best identified when listening at a site away from the apex (e.g., at the second right interspace), particularly if the systolic murmur is softer at the base than apex. If an S4 is present, there may be three audible sounds around the occurrence of S1; firm pressure with the bell or diaphragm will attenuate an S4 and will “bring out” the S1 and click. A discussion of the differentiation of these auscultatory findings is found in Chapter 8. Remember that the aortic ejection click does not vary with inspiration and therefore is readily distinguishable from a pulmonic ejection sound.

Murmur

Aortic stenosis produces the prototype of the classic systolic ejection murmur: a crescendo-decrescendo murmur that begins after S1 and ends before S2. The murmur typically is harsh, rough, or grunting and maximal at the second left interspace. It has been likened to the sound of someone clearing his throat. Although the classic aortic stenosis murmur is diagnostic, a number of factors may alter the usual findings. The characteristics of the murmur do not help differentiate congenital from acquired aortic stenosis, nor valvular from discrete supra- or subvalvular stenosis.

Site of Maximal Intensity. In most subjects, the murmur will be loudest at the second right interspace, the so-called aortic area. However, it is not uncommon for the murmur to be maximal at the second or third left interspace (Erb’s point), and in older patients with large chests or obstructive lung disease, the murmur may be loudest at the apex. Thus, it is best to think of distribution and radiation of the aortic stenosis murmur as a “sash pattern” that extends from the aortic area downwards to the left and to the cardiac apex (Fig. 9-2). The murmur at the cardiac base typically radiates upwards

and to the right and is often well heard over both carotid arteries. There may be a waning of intensity of the murmur between the second right interspace and the neck; the murmur may also decrease in intensity when the stethoscope is moved from the aortic area towards the apex where it may become louder.

Length of Murmur. In general, the length of the murmur is proportional to the severity of the valvular obstruction in the absence of other factors that modify stroke volume or the rate of ejection (Figs. 13-5, 13-7, 13-8). The presence of associated aortic insufficiency usually produces a louder murmur because of an increased stroke volume. In the presence of decreased left ventricular function or overt congestive heart failure, the murmur of aortic stenosis may shorten or completely disappear.

Some studies have shown that the time to the peak intensity of the murmur is a better indicator of severity of aortic stenosis than its overall length (Fig. 13-7). *Practical Point: It is important to assess the duration to the maximal murmur intensity in the evaluation of the severity of aortic stenosis. A murmur that peaks within the first third or half of systole suggests mild obstruction. A murmur that peaks within the second half of systole indicates severe disease (Figs. 13-5, 13-7, 13-8).*

Murmur Frequency and Pitch. The typical rough, grunting quality of the murmur of aortic stenosis is best heard at the aortic area. It has long been observed that in adults with acquired aortic stenosis, the systolic murmur at the apex can be quite musical in its pure frequency and high-pitched sound (the Gallavardin murmur). This is often so dramatic that the systolic murmurs at the aortic area and the apex are thought to be two separate murmurs, the latter typically confused with mitral regurgitation. It has been suggested that the harsh basal murmur reflects the effect of a jet or spray effect of turbulent blood in the ascending aorta, producing rough, multiple frequency sound. The high frequency apical murmur is believed to reflect vibrations emanating from aortic leaflets with intact commissures and stiffening and thickening of the basal cusps. Whatever the case, it is important to recognize that in older patients, particularly those with an increased chest diameter, the systolic murmur of aortic stenosis may be markedly different in tone at the base and apex, although the shape and length of the murmur is similar (ejection). In such patients, the apical murmur has been described as a seagull, cooing, or musical murmur.

Amplitude. In general, the louder the murmur of aortic stenosis, the more severe the valve obstruction. This observation is probably more true in children or young adults with bicuspid valves in whom a loud murmur almost always indicates a large aortic gradient. This relationship is less reliable in adults, as evidenced by the soft or unimpressive murmur that occasionally is found in patients with severe aortic stenosis. In the presence of chronic obstructive lung disease, obesity, or a big chest it is important to auscultate at the base with the patient upright and leaning forward. The murmur of

The Systolic Murmur of Aortic Stenosis: Assessment of Severity

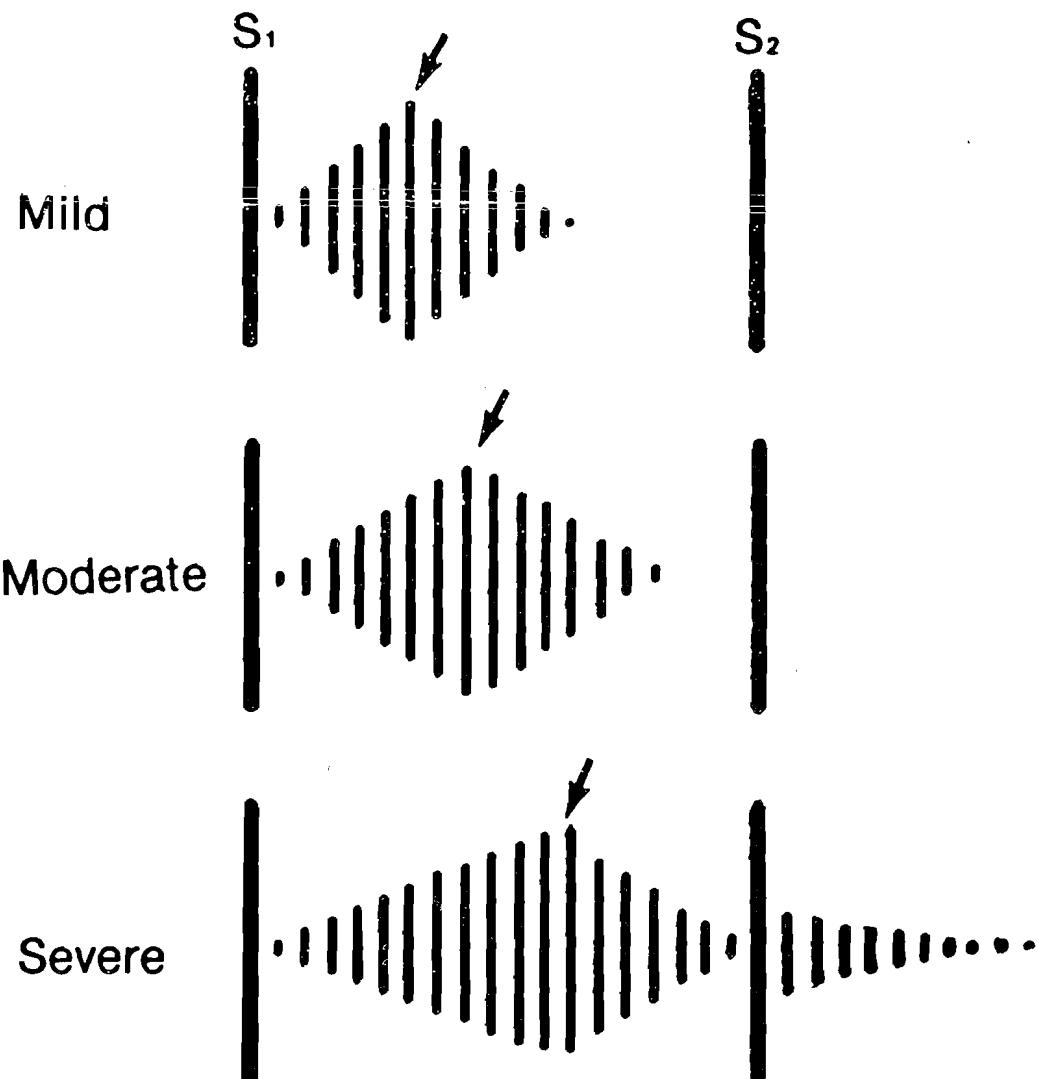


FIG. 13-7. Relationship of the shape of the systolic murmur to severity of the underlying aortic stenosis. As obstruction to left ventricular outflow becomes more severe, the murmur becomes longer and the time to peak intensity (arrow) is delayed. Physicians should assess the duration to maximal intensity as well as the total length of the murmur; both are useful in determining severity. (From Abrams J: Prim Cardiol, 1983). See also Figure 10-3.

aortic stenosis in such individuals often is very well heard over and above the clavicles (bone is a good sound conductor) and in the neck. Conversely, in such patients or in the presence of congestive heart failure or those with coexisting mitral stenosis, the finding of a prominent grade 3 to 4 murmur of aortic stenosis is a most important observation that suggests severe aortic valvular obstruction. The systolic murmur is usually quite soft and unim-

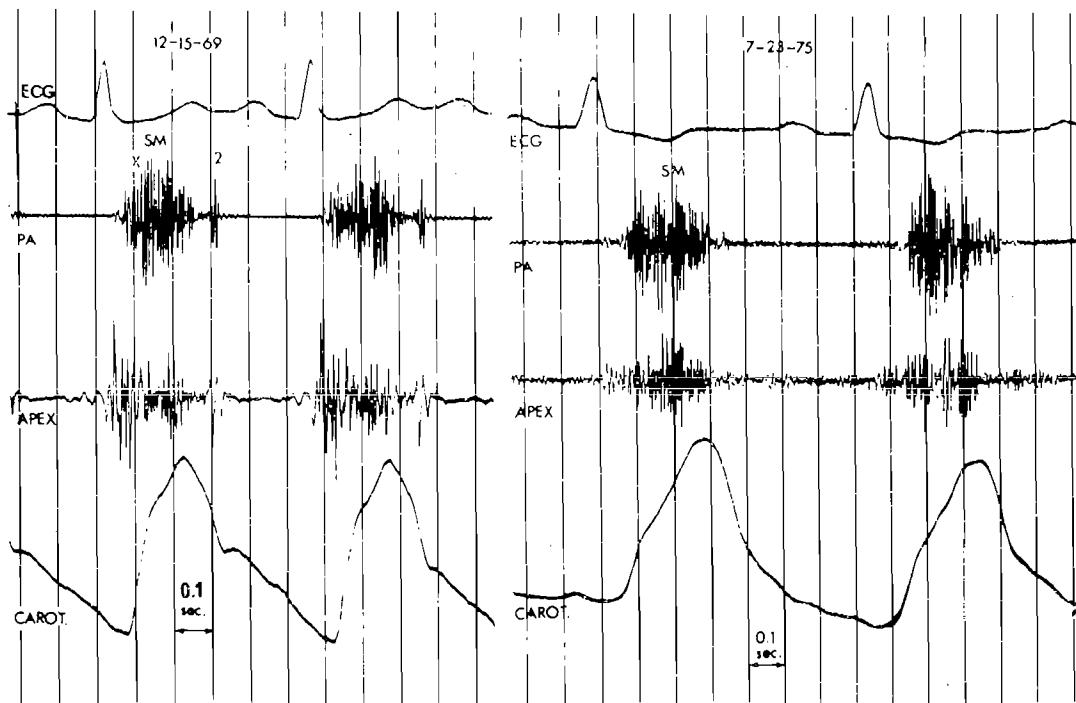


FIG. 13-8. Progression in severity of aortic stenosis with time. The phonocardiogram on the left was taken in a middle-aged man with mild aortic stenosis. Note the early peak of the murmur (SM) and sound vibrations ending well before S₂. The carotid upstroke is not clearly abnormal. On the right is a phonocardiogram taken six years later when the patient had become symptomatic. Note that duration of the murmur is longer and the time to peak intensity is more delayed. The carotid upstroke is now abnormal. (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. Prog Cardiovasc Dis 26:145, 1983.)

pressive in these situations, giving the false impression that aortic stenosis is mild or absent. In some patients with aortic stenosis, the maximal intensity of the systolic murmur may be at the left second or third interspace or apex.

Associated Aortic Regurgitation. Many patients, particularly those with rheumatic heart disease, have some degree of aortic regurgitation even in the presence of severe aortic stenosis (Fig. 13-7). The rigid, contracted, and often calcified valve may be truly immobile, unable to adequately open or close (Fig. 1-C, D). It is quite common for a trivial degree of aortic regurgitation to coexist with severe aortic stenosis (LV-aortic gradient greater than 75 mmHg), resulting in a grade 1-2 / 6 high frequency aortic regurgitation blow. *Practical Point:* Specifically listen for a blowing diastolic murmur in all patients with suspected aortic stenosis by having the subject sit up and lean forward with the breath held in expiration while using firm pressure with the diaphragm of the stethoscope. The presence of aortic regurgitation of any magnitude will tend to augment the stroke volume and thus the intensity and length of the systolic murmur for any degree of valve obstruction. Aortic regurgitation may be present in both valvular and discrete subvalvular aortic stenosis, but as a rule it is not found with supravalvular aortic stenosis or hypertrophic cardiomyopathy.

DIFFERENTIAL DIAGNOSIS

Aortic Sclerosis or the Systolic Murmur of the Elderly. This common condition in older persons results from stiffening of the basal aspects of the aortic valve leaflets without commissural fusion; thickening of the leaflet tissue and even calcification can be present. There is no actual valve obstruction or aortic gradient. The murmur of aortic sclerosis is that of a classic systolic ejection murmur of less than grade 4/6 intensity. A2 is typically well preserved. The carotids have a brisk upstroke. There is no evidence of left ventricular enlargement on precordial palpation. Most important, the overall length and peak intensity of the murmur fall within the first half of systole. Nevertheless, the differential diagnosis can be most difficult at times.

Mitral Regurgitation. When an aortic stenosis murmur radiates to the apex, or is louder at the apex, the differential diagnosis should include mitral regurgitation. This is a particular problem in the older patient, or in the subject with a large chest where the apical aortic murmur is likely to be of a higher frequency and have a more musical tone. To make the distinction between a murmur of aortic and mitral origin, it is critical to assess the length of the murmur with close attention paid to late systole. In the setting of a long systolic murmur, if A2 is soft or inaudible, precise timing of overall length may be impossible. Careful evaluation at the base of the heart is helpful in such cases to see if the murmur ends before S2. The murmur of aortic stenosis is often heard above the clavicles; the murmur of mitral regurgitation usually radiates well into the axilla. A normal carotid pulse favors mitral regurgitation.

Pharmacologic agents, such as amyl nitrite (see Chapter 11), may be of help in this differentiation. If an arrhythmia is present, an accurate diagnosis may be made more easily. The intensity of the aortic stenosis murmur varies directly with the magnitude of the LV stroke volume, whereas a mitral regurgitation murmur tends to be equally loud as left ventricular volume changes on a beat to beat basis. Thus, in a post-PVC beat or following long R-R cycle in atrial fibrillation, the murmur of aortic stenosis will increase in loudness, but there will be no change in intensity of the mitral regurgitation murmur.

Some patients, particularly those with rheumatic heart disease, have both aortic stenosis and mitral regurgitation. In addition, the murmur resulting from selective protrusion and incompetence of the posterior leaflet of the mitral valve (an unusual condition usually found with ruptured chordae tendinae), may have an ejection quality and radiate to the base. This murmur readily mimics that of aortic stenosis (see Chapter 17, Figs. 17-13, 17-15).

Hypertrophic Cardiomyopathy. See discussion beginning page 243.

Chapter 14

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy, also known in the United States as idiopathic hypertrophic subaortic stenosis (IHSS) and hypertrophic obstructive cardiomyopathy (HOCM), is one of the most unusual cardiac disorders. Its precise etiology and pathophysiology remain to be fully elucidated. This condition has been recognized for only a little over 20 years and has been the subject of much attention in the medical literature.

The hallmark of hypertrophic cardiomyopathy (HC) is the presence of massive interventricular septal thickening, typically in the superior portion of the muscular septum (Fig. 14-1). There usually is associated distortion of the mitral valve apparatus resulting in an abnormally displaced anterolateral papillary muscle and anterior leaflet of the mitral valve. The large anterior mitral cusp normally comprises the posterolateral aspect of the left ventricular outflow tract. In HC, this leaflet is distorted by such vigorous left ventricular contraction that it may be displaced towards the interventricular septum during systole; some believe that the mitral valve contributes to left ventricular blood flow "obstruction" during mid to late ejection. Recent evidence suggests that the posterior leaflet also moves abnormally in systole. There is considerable controversy as to whether complete or partial coaptation of the anterior mitral leaflet with the interventricular septum is a primary or secondary event in producing the deranged hemodynamics and physical findings in the obstructive form of HC.

Hypertrophic cardiomyopathy has two major variants. The first variant is an *obstructive* form with a measurable pressure gradient at rest or with provocative maneuvers between the body of the left ventricle and the distal left ventricular outflow tract. The second variant is a *nonobstructive* form characterized by left ventricular hypertrophy involving the interventricular septum as well as a portion of the left ventricular free wall, no resting or provokable intraventricular gradient, and relatively little abnormality of mitral valve structure or function. Both the obstructive and nonobstructive variants of HC demonstrate hyperdynamic ejection characteristics with extremely rapid and forceful left ventricular contraction, especially in early systole. Although the presence of a resting or provokable gradient in the obstructive variety of HC is associated with characteristic physical findings and a typical echocardiographic profile, there is little clinical evidence that a fundamental difference exists in the long-term prognosis of HC patients with or without obstruction. Some authorities (Criley, Goodwin, Murgo) firmly believe that

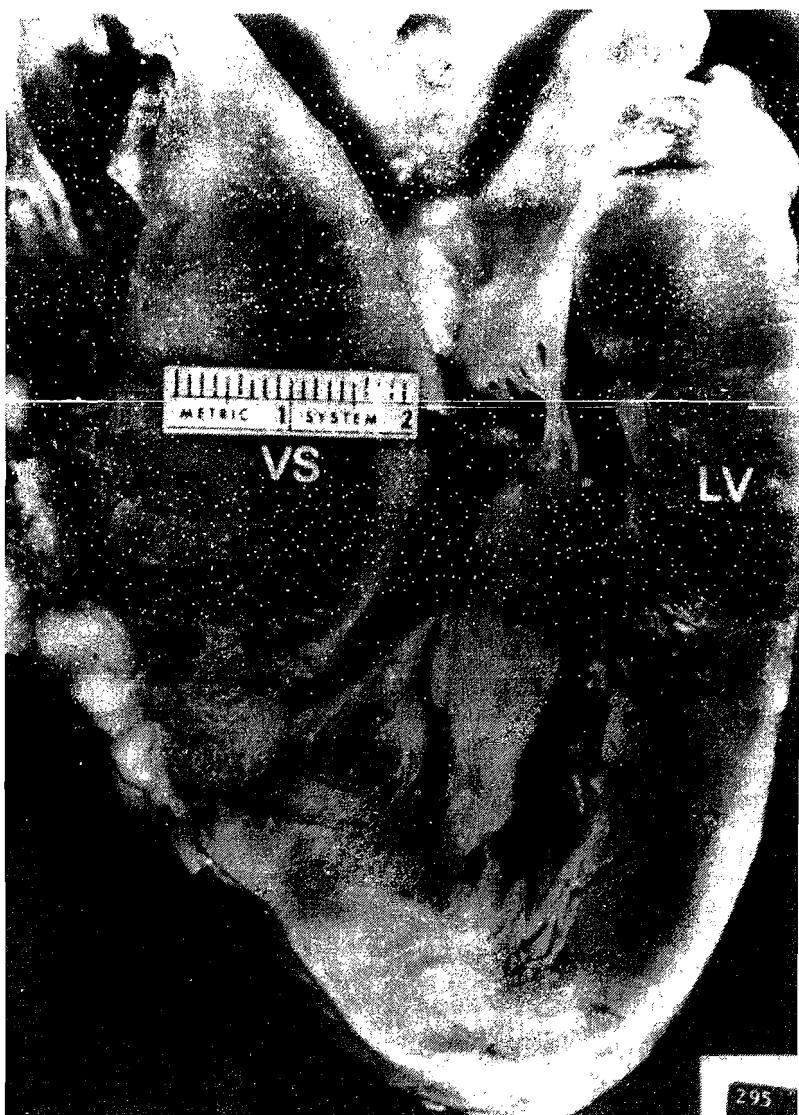


FIG. 14-1. Massive ventricular septal thickening in hypertrophic cardiomyopathy. The septum bulges into the left ventricular cavity. The left ventricular free wall (LV) is hypertrophied and has a small left ventricular cavity. Asymmetric septal hypertrophy (VS) is prominent in most but not all cases. (From Maron BJ: Cardiomyopathies. In Moss' Heart Disease in Infants, Children and Adolescents, 3rd ed. Edited by FH Adams and GC Emmanouilides. Baltimore, Williams & Wilkins, 1983.)

true obstruction does not exist and that the measured pressure gradient between the left ventricular body and the outflow tract is produced by excessively forceful and complete emptying of the left ventricle.

Hypertrophic cardiomyopathy exists in both familial and "sporadic" varieties. Genetic transmission is consistent with an autosomal dominant mode of inheritance and a high degree of penetrance. Large scale screening of family relatives using echocardiography suggests that even isolated or sporadic cases of HC have a genetic link. The manifestations of HC in family members range from symptomatic obstructive or nonobstructive cardiomyopathy to

asymptomatic individuals with asymmetric septal hypertrophy (ASH) on echo but without any clinical evidence of cardiac dysfunction. Equal sex distribution is found in the familial forms of HC, although males dominate in the sporadic cases.

PATHOLOGY

The typical heart in obstructive HC has dramatic asymmetric hypertrophy of the interventricular septum, with normal or somewhat hypertrophied left ventricular free walls (Fig. 14-1). The basal aspect of the posterior LV free wall is thickened. Left ventricular hypertrophy of the free walls in part may be a secondary phenomenon resulting from chronically increased intraventricular systolic pressure. The septal cardiac muscle is composed of striking myofiber disarray with loss of normal cellular architecture (Fig. 14-2). Cellular abnormalities include shorter, thicker myofibrils with marked disorganization and crisscrossing of muscle cells. Increased connective tissue is present. The myofibrils are hypertrophied and bizarre in appearance. In

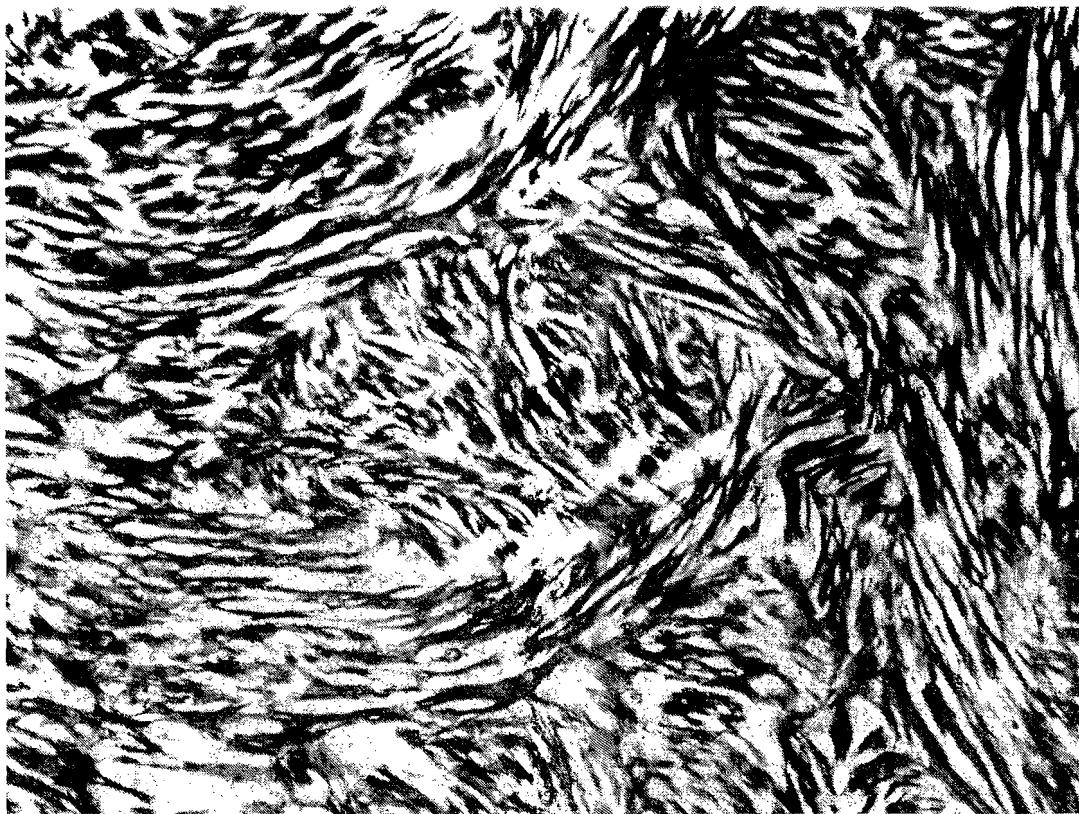


FIG. 14-2. Myocardial fiber disarray in hypertrophic cardiomyopathy. This photomicrograph is from the ventricular septum in a 19-year-old boy. Evidence for typical myocardial fiber disorganization is found in the vast majority of patients with hypertrophic cardiomyopathy. (From Roberts WC: Congenital cardiovascular abnormalities usually "silent" until adulthood. In *Congenital Heart Disease in Adults*. Edited by WC Roberts. Philadelphia, FA Davis Co, 1979.)

patients with a measurable pressure gradient across the left ventricular outflow tract, the peculiar cellular pattern is confined to the massively hypertrophied septum with normal cardiac muscle in the left ventricular free walls. Recent 2-D echo studies have demonstrated focal or diffuse LV hypertrophy in eccentric locations throughout the ventricles, unrelated to the degree of septal thickening.

While still a matter of controversy, some have suggested that in the nonobstructive variant of HC, typical cellular disarray is found throughout the left ventricular free wall. The unusual and markedly abnormal cellular architecture occasionally can be found to a far lesser degree in hearts of patients without HC who have other abnormalities of cardiac structure such as pulmonary atresia.

The anterior leaflet of the mitral valve typically is thickened in HC. An area of fibrosis on the left side of the interventricular septum is common, possibly related to the mitral leaflet impinging on the septal muscle, or perhaps due to a jet effect from the disordered blood flow profile during ejection. The aortic valve is normal in HC. At autopsy, approximately 10% of subjects have variable amounts of focal scarring or fibrosis in the left ventricular walls or septum.

PATHOPHYSIOLOGY

The dynamics of LV ejection in symptomatic patients with HC are similar (and unique to this condition) and unrelated to the presence or absence of a left ventricular-aortic pressure gradient. Ejection of blood is extremely rapid and forceful, resulting in premature cavity emptying and an abnormally small LV end-systolic cavity. Whether or not there is "obstruction" to outflow, the left ventricle ejects 85 to 90% of its end-diastolic contents by the middle of systole. Ejection is virtually complete by two thirds of left ventricular systole. Normal persons, by contrast, eject 60% of the LV end-diastolic volume during the first half of systole and do not effectively complete emptying until 85 to 90% of systole has been completed. Blood flow profiles in HC show little difference between patients with resting obstruction (i.e., a detectable LV-aortic gradient), those with only a provokable gradient, and subjects with no evidence of LV outflow tract obstruction under any conditions.

These observations raise questions about the physiologic implications of a gradient in "obstructive" HC. Indeed, some experts doubt the theory that obstruction is physiologic and clinically harmful. These scientists (Criley, Murgo) believe that actual obstruction to LV ejection does not exist and that there are no clinically important differences between patients with and without a gradient. Nevertheless, subjects with a large left ventricular-aortic pressure difference at rest or with provocation have the most prominent physical findings and are almost always symptomatic. Most workers believe that out-

flow obstruction is a valid concept with clear-cut clinical implications (Braunwald, Epstein, Wigle). The nonobstructionists discount the meaning of the pressure gradient, and the controversy continues.

Anterior Leaflet of the Mitral Valve. The role of the anterior leaflet of the mitral valve in HC remains controversial. A peculiar *systolic anterior motion* (SAM) is readily identified on echocardiography and angiography in obstructive HC (Fig. 14-3). The anterior leaflet may make contact with the interventricular septum during mid to late systole. Abutment of the valve cusp against the septum may be transient or may persist. Most experts believe that SAM is responsible for the pressure differences between the body of the LV and the more distal LV outflow tract in patients with a resting or provokable gradient. SAM is found most commonly in patients with HC who have significant "obstruction" to outflow, but may also be found in occasional patients with nonobstructive HC and even those with mitral valve prolapse. Some workers believe the posterior leaflet also may move abnormally in systole. In obstructive HC the left ventricular outflow tract is abnormally narrowed both in systole as well as diastole, even though the hypertrophied septal muscle does not contract as vigorously as it does in normal patients.

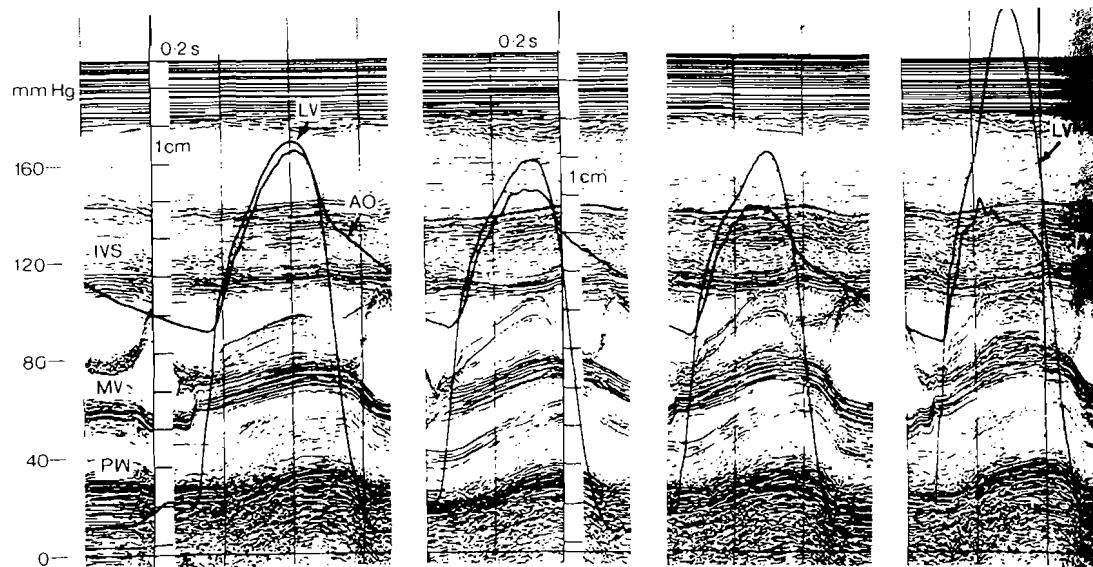


FIG. 14-3. Relationship of systolic anterior motion (SAM) of the anterior mitral leaflet and left ventricular-aortic pressure gradient in hypertrophic cardiomyopathy. In the left panel SAM is present without leaflet-septal contact. There is no pressure gradient between the left ventricle and aorta in the basal state. Late systolic SAM-septal contact is seen in the second panel; the contact is brief and the pressure gradient is small. During isoproterenol infusion, a left ventricular-aortic gradient appears (panels 3 and 4) as SAM-septal contact develops earlier in systole and becomes more prolonged. The changes correlate with the occurrence of a large pressure gradient between the body of the left ventricle and the aorta (panel 4). It has been postulated that "obstruction" in hypertrophic cardiomyopathy is related to both the timing and duration of contact of the anterior leaflet of the mitral valve with the interventricular septum. IVS = septum; MV = mitral valve; PW = posterior wall; LV = left ventricle; AO = aorta. (From Pollick C, Rakowski H, and Wigle ED: Muscular subaortic stenosis: the quantitative relationship between systolic anterior motion and the pressure gradient. Circulation 69:43, 1984.)

Left Ventricular Compliance in HC. John Goodwin of England has championed the view that the predominant clinical effects of HC are not related to left ventricular outflow tract obstruction but are due to the profound alteration in compliance characteristics of the left ventricle in both the obstructive and nonobstructive variants. Thus, the extremely stiff, massively hypertrophied LV muscle resists diastolic inflow from the left atrium. Left ventricular filling pressure is abnormally high, with resultant elevation of pulmonary capillary pressure and enlargement of the left atrium. Systolic function of the noncompliant left ventricle is hyperdynamic, although interventricular septal motion may be hypokinetic. The left ventricular ejection fraction is normal to elevated, and the end-systolic volume is abnormally reduced.

Mitral Regurgitation. Mitral regurgitation occurs in 30 to 50% of affected subjects and can contribute to and complicate the hemodynamic picture if the regurgitant fraction is large. The mitral valve may become incompetent because of abnormal anterior angulation of the papillary muscles and chordae tendineae (a variant of papillary muscle dysfunction). Mitral regurgitation typically is found only in patients with a resting or provokable gradient, and its presence correlates with echocardiographic systolic anterior motion of the mitral valve. Mitral reflux is greatest in subjects with large LV-aortic pressure gradients.

Influences on the Pressure Gradient. A variety of agents and maneuvers can profoundly affect the hemodynamics in hypertrophic cardiomyopathy (see section on physical maneuvers and pharmacologic agents; Table 14-1), underlining the fact the physiology of HC is extremely labile and variable. Dr. Michael Criley has used the term "ventriculovalvular disproportion" to emphasize the fact that the left ventricular cavity is literally too small for the mitral valve in subjects with HC. Any maneuver or pharmacologic in-

TABLE 14-1 *Effects of Physiologic and Pharmacologic Maneuvers in Hypertrophic Cardiomyopathy*

Intervention	Left Ventricular Outflow Obstruction	Murmur
Valsalva		
Phase 2-3 (strain)	Increase	Increase
Phase 4 (release)	Decrease	Decrease
Squatting	Decrease	Decrease
Upright posture	Increase	Increase
Exercise	Increase	Increase
Amyl nitrite	Increase	Increase
Methoxamine	Decrease	Decrease
Isoproterenol	Increase	Increase
Propranolol	Decrease or unchanged	Decrease or unchanged

From Shah PM: Newer concepts in hypertrophic obstructive cardiomyopathy. JAMA, 242:1773, 1979, with permission.

tervention that reduces left ventricular dimensions tends to increase or bring out a pressure gradient. The converse is also true. Increasing afterload, i.e., the forces tending to distend the left ventricular outflow tract and aorta distal to the site of obstruction or increasing end-diastolic volume (preload), reduces the LV-aortic gradient, when present. An agent or maneuver that augments cardiac contractility will increase the outflow gradient and accentuate the systolic murmur and other physical signs of HC.

CLINICAL PRESENTATION

Hypertrophic cardiomyopathy represents a cardiac syndrome with markedly variable course. Unique aspects of this condition are its unpredictability and the lack of an obvious correlation between the clinical manifestations of HC and underlying hemodynamic profile as assessed by echocardiography or cardiac catheterization. Patients may die suddenly without having had any prior symptoms; there is no apparent relationship between these tragic sudden deaths and the presence or absence of obstruction in HC. The controversy over the significance of a resting or provokable gradient is still unresolved. The concept that the gradient is a "secondary" phenomenon, related to the premature and virtually complete ejection, would help explain the failure of any clinical or hemodynamic parameter to predict late morbidity or mortality in HC.

Asymptomatic individuals with HC are usually detected by accident, such as noting prominent LV activity, an S4, or a loud systolic murmur. Some cases are recognized through screening of affected family members. HC appears to develop over time in susceptible individuals. The abnormal LV hypertrophy is not usually detected at birth or in early childhood, although it has been described in infancy. Hypertrophic cardiomyopathy has been documented in the elderly. The diagnosis is easy to miss in a patient in the 7th or 8th decade of life because of a similarity of the clinical presentation to that of coronary or hypertensive heart disease (e.g., prominent LV, S4, systolic murmur). Of particular interest is the evidence that the majority of patients over the age of 60 with documented HC are women. It has been suggested that a condition similar to HC may result from long-standing hypertension or aortic valve stenosis.

Presenting symptoms of HC include dyspnea on exertion, dizziness, syncope, angina, or palpitations. Shortness of breath is the most common symptom and is a result of high LV filling pressures at rest or exercise due to the noncompliant, hypertrophied LV chamber. Dizziness and syncope may result from severe LV outflow tract obstruction or may be caused by arrhythmias. Angina may occur as a result of the markedly increased myocardial oxygen demands of the hypertrophied ventricle without a commensurate increase in coronary blood flow. Sudden death, often occurring in young adults, is usually caused by malignant ventricular dysrhythmia.

PHYSICAL EXAMINATION

There is nothing unusual about the overall appearance of the patient with HC. Some have commented on the increased prevalence of this disorder in young persons with a muscular body habitus. Rarely, HC is found in children with diffuse lentiginosis, a dermatologic syndrome consisting of numerous darkly pigmented spots on the trunk and arms.

Jugular Venous Pulse

The jugular venous A wave is characteristically prominent in HC; a "flicking motion" in the neck veins appearing just before the carotid upstroke is commonly present. The large A wave is a result of the stiff RV myocardium and powerful right atrial contraction; the A wave may be even more noticeable in inspiration. When especially prominent, an augmented jugular venous A wave suggests an RV outflow tract gradient. Typically, the mean jugular venous pressure in HC is normal; right heart failure is distinctly uncommon except in end-stage patients.

Arterial Pulse

The typical carotid pulse in HC reflects the early, exaggerated systolic emptying of the LV. It has a brisk or sharp upstroke that literally taps against the palpating fingers. This rapid upstroke can be reminiscent of aortic regurgitation, although the overall pulse volume in HC is not increased. *Practical Point:* The presence of a brisk carotid arterial pulse contour in a patient being evaluated for possible aortic valve stenosis immediately should raise the suspicion of hypertrophic cardiomyopathy, particularly if there is a prominent systolic murmur.

Much attention has been given to the presence of a bifid carotid arterial pulse in HC, particularly in the obstructive variety. This phenomenon is not easily appreciated by the examiner's fingers but, when present, is readily recorded on an arterial pulse tracing. This characteristic pulse abnormality is associated with so-called obstruction to LV outflow and rarely is found in subjects without an LV-aortic gradient. The classic pulse contour in HC consists of a rapid upstroke, followed by a midsystolic dip or collapse, which in turn is followed by a second late systolic wave (Fig. 14-4). The first peak is the equivalent of a sharp percussion wave, and the second peak represents a prominent, delayed tidal wave (see Chapter 3). The mechanism for the secondary wave is unclear; the second peak occurs at the time when LV ejection is minimal, and it probably is a reflected wave or rebound phenomenon. The magnitude of the midsystolic dip correlates with the size of the LV-aortic pressure gradient. The typical bifid pulse contour in HC has been called the "spike and dome" or "pointed finger" pulse.

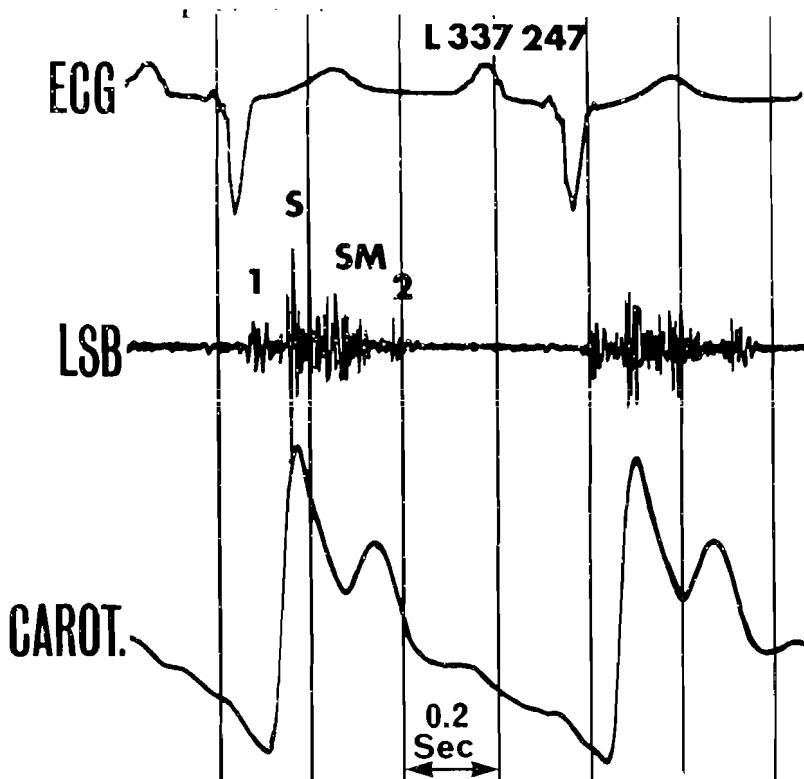


FIG. 14-4. Carotid arterial pulse in hypertrophic cardiomyopathy. Note the brisk upstroke with a midsystolic dip and secondary wave. There is a high frequency ejection sounds (S) coincident with the peak of the carotid upstroke. This is consistent with the "pseudoejection sound" that has been previously described in hypertrophic cardiomyopathy, possibly related to rapid reversal of central aortic pressure outflow "obstruction" becomes more significant. (From DeJoseph RL, Seibert V, and Tavel ME: Systolic ejection sound in idiopathic hypertrophic subaortic stenosis. Chest 66:382, 1974.)

The degree of the arterial pulse contour abnormality is directly related to the size of the outflow tract gradient; the carotid wave form may vary as the degree of obstruction changes on a beat-to-beat basis. Patients without a resting gradient do not usually have the spike and dome pulse contour, but may transiently develop a bifid pulse in the beat following a PVC or with any maneuver that induces or accentuates LV outflow tract obstruction. Although this phenomenon has received considerable attention in the medical literature, the spike and dome pulse is rarely palpable and therefore of little clinical utility unless a graphic tracing of the carotid pulse is obtained. *Practical Point: The cardinal arterial pulse alteration in hypertrophic cardiomyopathy is a rapid, jerky, or sharp carotid pulse. The arterial pulse wave contour often is normal to palpation, especially in the absence of a resting gradient.*

The use of the brachial arterial pulse has been suggested as a way to better differentiate valvular aortic stenosis from obstructive hypertrophic cardiomyopathy. The carotid pulse of HC may simulate aortic regurgitation if a brisk upstroke is present. In many patients the arterial pulse following a PVC will be diminutive, reflecting the decreased arterial pulse pressure that

is usually (but not always) found in a post-PVC beat (the Brockenbrough phenomenon).

A thrill or shudder rarely is felt over the carotid arteries in nonobstructive or obstructive HC, in contradistinction to valvular aortic stenosis. There is no characteristic abnormality of the systemic blood pressure in HC.

Precordial Motion

Abnormal LV activity is common in subjects with HC, and this may be an important first clue that "something is not quite right" in the evaluation of a young patient with a systolic ejection murmur. The hallmark of precordial examination is the ubiquitous presence of a loud and typically palpable S4 (presystolic distension of the LV) accompanied by a late systolic apical heave, thrust, or bulge (Fig. 14-5). Augmented left atrial contraction is common

ABNORMAL ACG

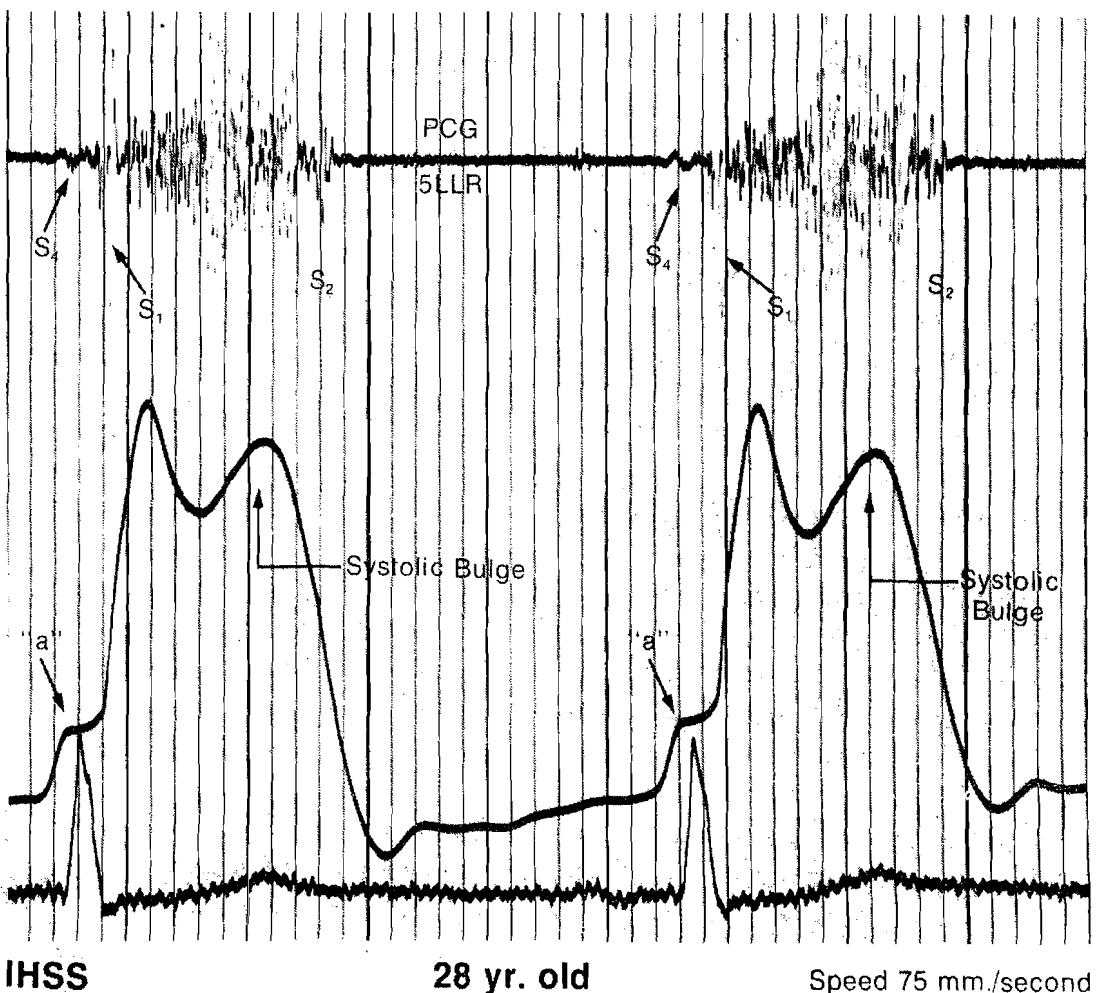


FIG. 14-5. Apical impulse in hypertrophic cardiomyopathy. Note the large A wave or palpable S4 and the prominent late systolic bulge. The systolic murmur is long and late peaking, suggesting significant obstruction to left ventricular outflow. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co, 1979.)

and is detectable at the cardiac apex irrespective of whether there is a resting gradient. In contradistinction to valvar aortic stenosis, increased left atrial activity in HC does not correlate with the pressure gradient.

There may be an early systolic dip or retraction of the apical impulse, followed by an anterior midsystolic bulge or thrust. The combined presence of a palpable S4, (normal) early systolic impulse and a late systolic bulge may produce a trifid contour to the apex impulse; this has been called the "triple ripple" (Fig. 14-6). As with the midsystolic dip and late systolic wave in the carotid artery, the second outward movement of the LV may not be palpable, although the abnormal motion is readily recorded on an apex cardiogram.

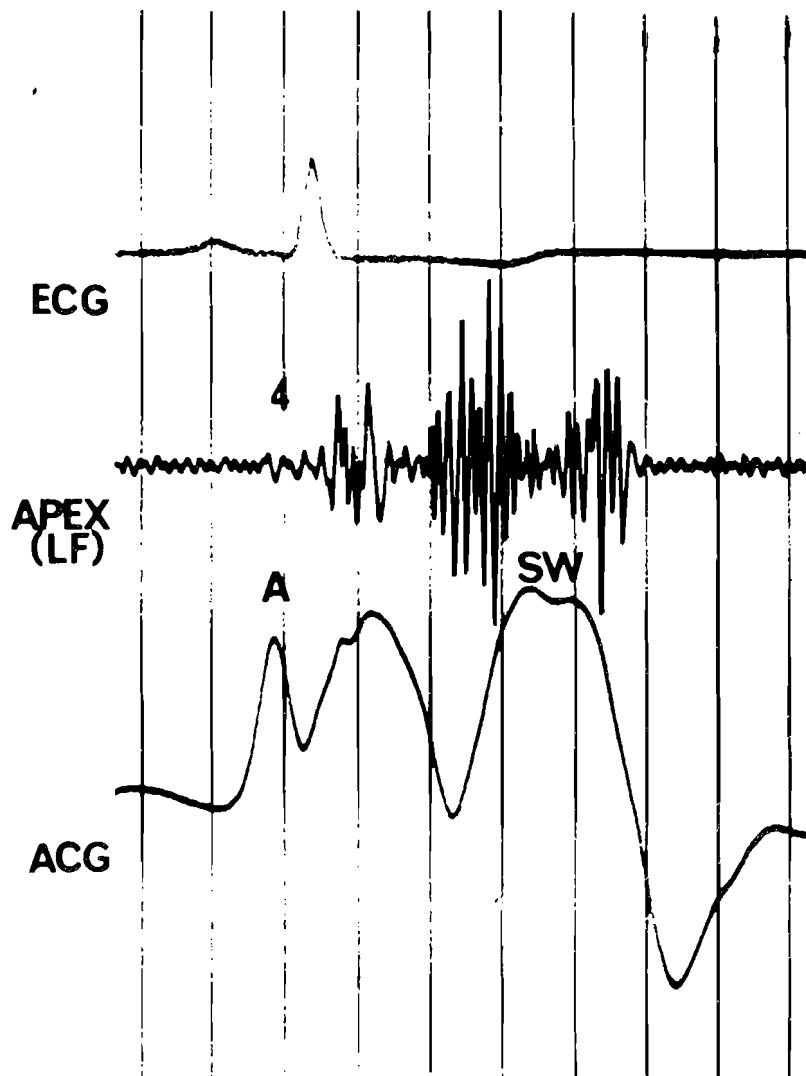


FIG. 14-6. Apical impulse in hypertrophic cardiomyopathy. In this apex recording the huge presystolic A wave is followed by a normal systolic impulse and subsequently by a late systolic wave or bulge (SW). These events produce the characteristic "triple ripple"; all three waves may be palpable on careful examination. (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 26:145, 1983.)

The typical LV impulse in HC is sustained, forceful, and palpable in more than one interspace. It is particularly prominent in the left decubitus position. A systolic thrill is often palpable and is directly related to the presence and severity of a resting gradient. The thrill may be maximal at the lower left sternal border or at the apex. It is rarely felt at the base, in contrast to valvular aortic stenosis. *Practical Point: The presence of a systolic thrill in hypertrophic cardiomyopathy suggests that the patient has obstruction to left ventricular outflow.*

Parasternal Impulse. On occasion, abnormal parasternal activity can be palpated in HC. While this suggests the presence of "obstruction" to right ventricular outflow, an RV lift may be found in relatively thin subjects with asymmetric septal hypertrophy and does not necessarily mean that right ventricular hypertrophy is present.

Heart Sounds

First Heart Sound. The first heart sound is normal or accentuated in HC. An increased intensity of S1 is probably related to the abnormally rapid rate of rise of LV pressure during isovolumic systole.

Second Heart Sound. Splitting of S2 may be disordered in HC, particularly when there is a pressure gradient across the LV outflow tract. Left ventricular ejection is usually prolonged, and as a result S2 is often narrowly split with inspiration. When significant obstruction is present, A2 may be delayed beyond P2 and reversed or paradoxical splitting of S2 appears (Fig. 14-7). *Practical Point: Usually there is no diagnostic abnormality of the second heart sound in hypertrophic cardiomyopathy. Reversed splitting of S2, when present, suggests a large left ventricular-aortic gradient.*

Third Heart Sound. An S3 in HC is not uncommon. When present, the S3 does not indicate poor left ventricular function, but may reflect the profound alteration in left ventricular diastolic compliance. An S3 is more likely to be present when there is associated mitral regurgitation. In one large study an S3 was recorded in over half the patients with HC, most of whom had a resting pressure gradient.

Fourth Heart Sound. An S4 is commonly heard or palpated in obstructive or nonobstructive HC (Fig. 14-5, 14-6). The presence of a prominent atrial sound in a young subject with a systolic murmur should initiate a careful examination for hypertrophic cardiomyopathy. A loud S4 is related to augmented left atrial contraction and the typically stiff and hypertrophied LV. *Practical Point: Be wary of making the diagnosis of hypertrophic cardiomyopathy in any person who does not have an S4. A loud atrial sound is the rule in this disorder.*

Often a prominent S1 in HC is mistaken for an S4; this gives the false impression that the systolic murmur begins very early in systole.

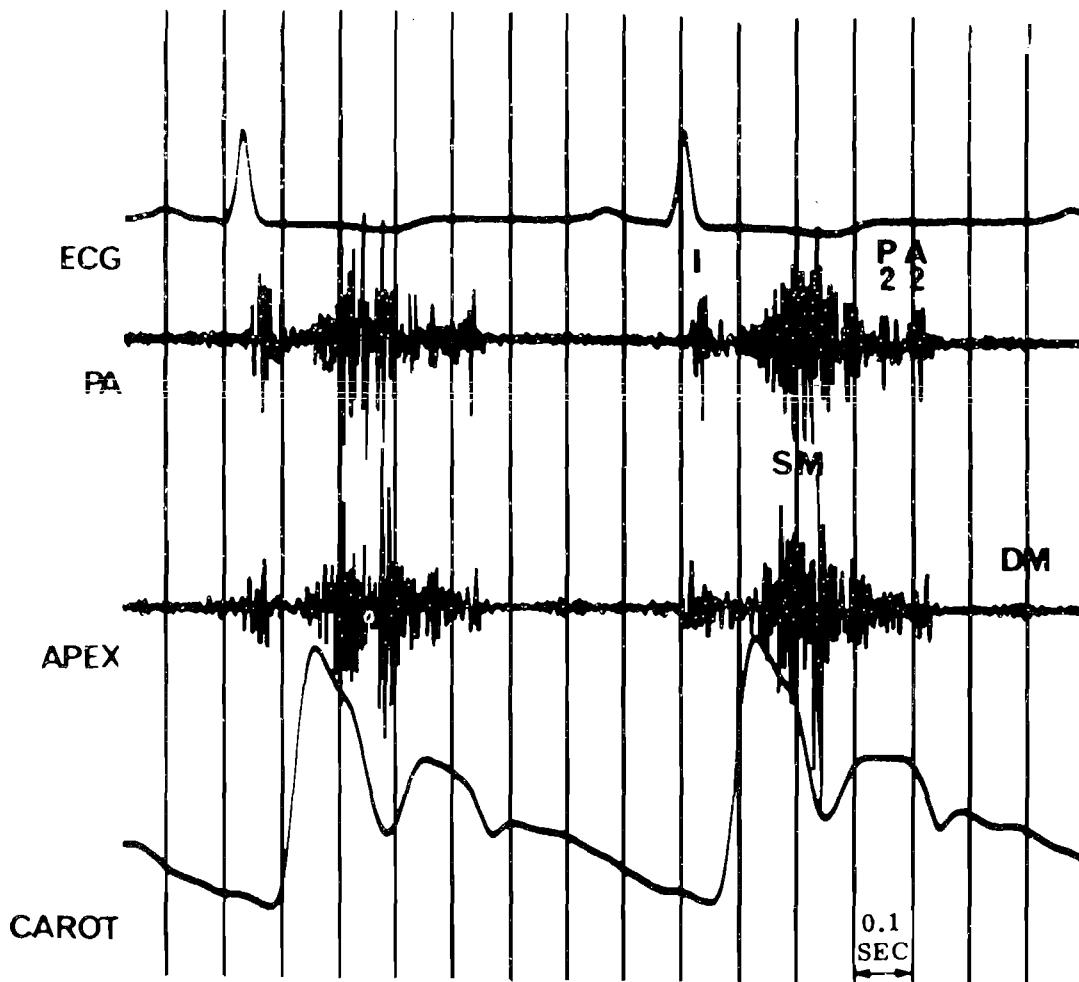


FIG. 14-7. Typical murmur and arterial pulse in hypertrophic cardiomyopathy. Note the systolic ejection murmur that begins after S1 and ends just before A2. The carotid pulse shows the characteristic "spike and dome" configuration. S2 demonstrates paradoxical splitting following a long, late peaking murmur. These findings are suggestive of significant "obstruction" to left ventricular outflow. (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 26:145, 1983.)

Pseudoejection Sound. There is disagreement as to whether an ejection sound can be present in HC. Phonocardiograms often display a large amplitude vibration at or near the onset of the systolic murmur (Fig. 14-4). However, such a sound transient is rarely audible. Shah and others have documented that in certain patients with HC an ejection-like sound is commonly recorded, perhaps in as many as 50% of affected individuals. In general, these subjects have the obstructive variety of the HC. The ejection transient is closely associated with the sudden halting motion of the anterior leaflet of the mitral valve at the moment of its maximal systolic anterior motion as seen on the echocardiogram. This sound can be augmented by maneuvers that accentuate the outflow tract obstruction.

The ejection-like sound also coincides with the peak of the carotid arterial pulse, which suggests that it may originate in the aortic root at time of rapid

early systolic pressure rise. Whether the sound is related to mitral valve motion or to an increased velocity of ejection, it is clearly a phenomenon associated with obstruction to left ventricular outflow. Because of its uncertain etiology and the fact that its timing is later than the typical ejection sound in valvular aortic stenosis, the term "pseudoejection sound" has been used. The sound is of low to medium frequency, is usually inaudible, and on phonocardiography initiates the systolic murmur (Fig. 14-4). In some persons, this sound takes on higher frequency vibrations; in such situations, it is more likely to be audible and may simulate an aortic ejection click. This could mislead the physician as to the etiology of the left ventricular outflow tract obstruction (e.g., valve versus muscular).

Practical Point: The pseudoejection sound in hypertrophic cardiomyopathy is rarely heard, easily recorded, and correlates with a large left ventricular-aortic gradient.

Opening Snap. Rarely, an opening snap can be recorded in patients with obstructive HC. This is probably related to the thickened anterior leaflet of the mitral valve, which generates a sound as it swings open in early diastole. This sound transient is not audible.

The Systolic Murmur

The literature on the systolic murmur in HC is confusing, and the murmur continues to be described variably. Some authorities state that typically it is an early systolic murmur, but others emphasize that it is a late systolic murmur. There are also conflicting opinions about a separate holosystolic murmur present in those patients with coexistent mitral regurgitation.

The characteristic murmur of obstructive hypertrophic cardiomyopathy is a long, somewhat harsh systolic ejection murmur (Figs. 14-4 to 14-7). It is well heard at the apex and lower left sternal border and may be extremely variable in intensity. Because of the rapid ejection velocity and "premature" emptying of the left ventricle, there often is an early systolic "flow" component to the murmur, even in the absence of a LV-aortic gradient. In patients with a large pressure gradient across the left ventricular outflow tract, the murmur tends to peak in late systole. Usually, there are sound vibrations in early systole that precede the onset of detectable obstruction. In mild degrees of outflow tract obstruction, a soft, late peaking systolic murmur may be heard. In general, with increasing severity of the gradient, the murmur becomes louder and longer.

Timing. The murmur of HC typically has a crescendo-decrescendo shape, peaking late in systole in the obstructive variant and ending before A2 (Figs. 14-4 to 14-7). When there is associated mitral regurgitation, the murmur may seem longer at the apex than at the base; a silent gap between

the end of the murmur and A2 is usually recorded. Several carefully performed phonoangiographic correlative studies have been unable to demonstrate a difference in the systolic murmur of HC through the use of phonocardiography in patients with and without mitral regurgitation. On auscultation, such murmurs appear to be holosystolic, often with a tapering configuration in late systole. It is unusual to hear the classic whirring pansystolic murmur of mitral regurgitation in HC. In addition, the systolic murmur of HC typically begins *after S1*, even in subjects with documented mitral incompetence. The physician should focus intently on early and late systole to assess the duration of the murmur.

In patients without obstruction, or those with a small gradient, the systolic murmur is shorter and may even be absent. Asymptomatic subjects with echo-proven ASH but no other evidence of HC often have no murmur suggestive of this disorder.

Intensity. The amplitude of the HC murmur is extremely variable. It may change in volume as much as 1 to 2 grades with a change in body position or exaggerated respiration. Often the murmur is louder on initial examination of the patient and softens considerably after a period of time, probably due to a decrease in anxiety-related sympathetic activity. The murmur may vary in intensity and length from day to day or even hour to hour. Various maneuvers or agents may be utilized to augment the loudness of the murmur in patients suspected of having HC (see section on physical maneuvers and pharmacologic agents; Table 14-1).

Loud murmurs typically are associated with a measurable outflow gradient and may have an accompanying systolic thrill. A thrill suggests a large gradient.

Location and Radiation. The systolic murmur in HC is best heard in the lower, midprecordial area between the apex and lower left sternal border. Usually, it is maximal close to the sternum in the 3rd to 4th left interspace at a site considerably lower than the location of the typical murmur of valvular aortic stenosis. Less commonly, the murmur is loudest at or just inside the apex. The murmur does not usually radiate well to the aortic area and neck and rarely is prominent over the carotid arteries or in the suprasternal area. Although the murmur is not very prominent at the base of the heart (second left and right interspaces), on occasion it may be loudest at this site. This suggests obstruction to right ventricular ejection and a coexisting right ventricular-pulmonary artery gradient.

When mitral regurgitation is associated with obstructive HC, the systolic murmur often appears to be longer and may be best heard at the cardiac apex. It may radiate into the axilla. *Practical Point: Hypertrophic cardiomyopathy should be considered whenever a systolic murmur appears to have both ejection and regurgitant characteristics with a maximal intensity lower than the site of a typical ejection murmur.*

PHYSICAL MANEUVERS AND PHARMACOLOGIC AGENTS USED IN DIAGNOSIS

Transient hemodynamic alterations produced by altered physiology can produce striking changes in the physical findings in hypertrophic cardiomyopathy (Table 14-1, Figs. 14-8, 14-9). The use of various maneuvers and drugs have been most informative in our understanding of the basic pathophysiology of this fascinating condition (see Chapter 11). Any intervention that increases the resting outflow tract gradient in a patient with "obstructive" HC or that can initiate a pressure gradient when there is no resting gradient will accentuate or produce the systolic murmur. The murmur will typically become louder and longer and peaks later in systole (Figs. 14-8, 14-9). Other changes in the physical examination that may occur with increased obstruction include (1) the development or accentuation of the systolic dip or spike and dome configuration of the carotid pulse (Fig. 14-8); (2) similar enhancement of midsystolic retraction of the apex impulse; and (3) the appearance of a pseudoejection sound initiating the systolic murmur. The converse is also true: maneuvers that decrease the left ventricular outflow tract gradient will reduce the amplitude and length of the systolic murmur and minimize other features of the cardiac examination that are associated with obstruction. On occasion, however, provocation of a markedly increased gradient (Valsalva

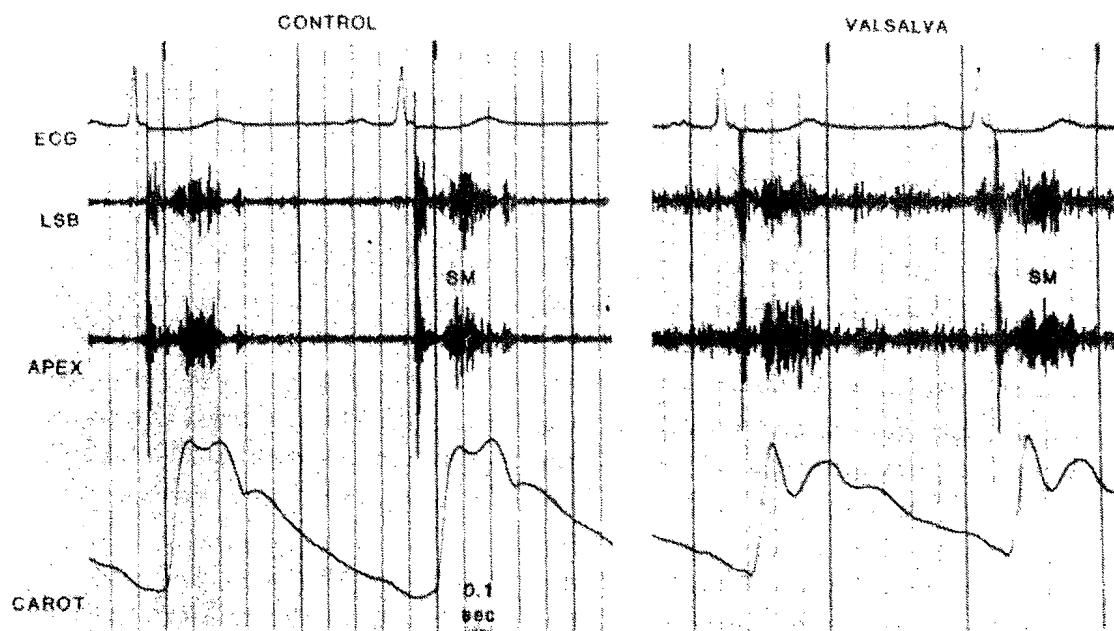


FIG. 14-8. Effects of Valsalva maneuver in hypertrophic cardiomyopathy. In the baseline study on the left an ejection murmur (SM) is seen and the carotid arterial pulse is normal. During the strain phase of the Valsalva maneuver (right), the characteristic "spike and dome" configuration of the arterial pulse appears and the systolic murmur becomes louder, longer, and later peaking as left ventricular obstruction becomes more pronounced. (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 26:145, 1983.)

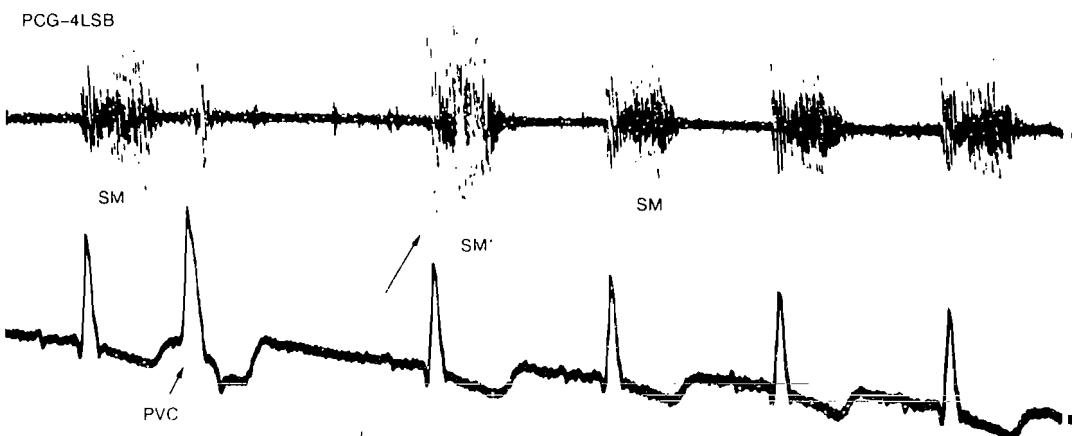


FIG. 14-9. Post-PVC augmentation of the systolic murmur (SM) in hypertrophic cardiomyopathy. Note the accentuation of the murmur that occurs in the beat following the PVC. Augmentation of the murmur is related to increased "obstruction" to left ventricular outflow in the post-PVC beat. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co, 1979.)

maneuver, post-PVC beat) will result in no change or even a decrease in the murmur.

One can predict what will happen to the pressure gradient and the quality of the murmur in a patient suspected or known to have obstructive hypertrophic cardiomyopathy by remembering the following guidelines:

The magnitude of the gradient and the systolic murmur are accentuated by anything that causes:

1. A REDUCTION IN LEFT VENTRICULAR CAVITY VOLUME OR PRESSURE (DECREASED PRELOAD).
2. A DECREASE IN SYSTEMIC VASCULAR RESISTANCE AND/OR ARTERIAL PRESSURE (DECREASED AFTERLOAD.)
3. AN INCREASE IN THE LEFT VENTRICULAR CONTRACTILE STATE.

Conversely, apparent obstruction to left ventricular outflow is reduced and the systolic murmur will soften as a result of:

1. AN INCREASE IN LEFT VENTRICULAR CAVITY VOLUME AND/OR FILLING PRESSURE.
2. AN INCREASE IN SYSTEMIC ARTERIAL PRESSURE OR RESISTANCE.
3. A DRUG OR INTERVENTION THAT DECREASES LEFT VENTRICULAR CONTRACTILITY.

Changes in left ventricular cavity size affect the spatial relationships between the circumference of the left ventricular outflow tract and the encroachment of the anterior mitral leaflet towards the massive interventricular septum. A smaller left ventricular outflow tract and small left ventricular cavity in systole accentuates the ventricular-valvular disproportion and exaggerates the pressure gradient, whether present or latent. Such murmur variations are similar to the behavior of the systolic murmur of mitral valve prolapse.

Changes in the distending forces exerted on the distal left ventricular outflow tract and central aorta beyond the site of intracavitory obstruction will alter the gradient across the left ventricular outflow tract. An increase in aortic pressure will "open up" the distal outflow tract, help push the anterior leaflet of the mitral valve posteriorly towards the left atrium, and reduce any existing or latent obstruction. Conversely, a decrease in aortic pressure or vascular resistance enhances anterior displacement of the mitral leaflet towards the septum and increases the gradient.

A more vigorous left ventricular contraction will result in a smaller end-systolic cavity and accentuates the gradient between the body of the left ventricle and the aorta.

The maneuvers and pharmacologic agents listed in Table 14-1 are particularly helpful when the diagnosis of hypertrophic cardiomyopathy is being considered. Use of these interventions often clarifies the clinical situation; making the diagnosis of HC may be influenced by behavior of the murmur with various interventions.

The use of amyl nitrite and the Valsalva maneuver are recommended for optimal diagnostic utility (Fig. 14-8). An equivocal or soft murmur of HC should become much louder following these maneuvers. *Practical Point:* *A I-II/VI systolic murmur that does not augment by one to two grades after amyl nitrite inhalation and/or the strain phase of the Valsalva maneuver is unlikely to be caused by obstructive hypertrophic cardiomyopathy. However, in the nonobstructive variant (ASH) without a provokable gradient, the failure of the murmur to increase in intensity with such maneuvers does not rule out HC.*

RIGHT VENTRICULAR INVOLVEMENT

The huge septal mass in patients with HC may affect right ventricular diastolic and systolic performance, although this effect is not common. In approximately 10% of patients with a left ventricular outflow tract gradient, there is a measurable gradient across the right ventricular outflow tract as well; this can be quite large and, in part, may be a result of infundibular hypertrophy. In addition, decreased right ventricular compliance can result in delayed filling of the right ventricle and an elevation of right atrial pressure; the venous A-wave is often very large.

The jugular A-wave is particularly prominent and increased in amplitude. There may be a parasternal or right ventricular lift. S2 is widely split on inspiration (delayed P2). The maximal intensity of the systolic murmur typically is higher on the sternum and may be best heard at the second or third left interspace. Although the murmur augments with inotropic interventions (isoproterenol, digitalis), the Valsalva maneuver causes a *decrease* in the murmur intensity because right ventricular filling is substantially reduced;

this is in contradistinction to the effects of the Valsalva maneuver in left-sided HC. *Practical Point:* *The majority of patients with right ventricular involvement have associated left ventricular obstruction. The physical findings are related to the presence of both right and left ventricular outflow tract gradients.* Coexisting left ventricular hypertrophy is always present, and evidence of left ventricular outflow tract obstruction may still dominate the clinical picture even with a detectable right-sided gradient.

DIASTOLIC MURMUR

On occasion a short apical diastolic murmur may be detected in HC. This peculiar murmur frequently has a crescendo-decrescendo shape on phonocardiography and may occur in up to 10% of affected individuals. It is early to mid-diastolic in timing and occurs during the filling phase of diastole. The marked ventricular hypertrophy and decreased LV compliance results in a decreased rate of diastolic inflow from the left atrium; presumably, sufficient turbulence is created in the LV cavity to produce the murmur.

This murmur can be found in patients with and without mitral regurgitation, but is more likely to be present with severe mitral insufficiency. Nevertheless, increased diastolic flow cannot account for this murmur as severe mitral regurgitation is distinctly uncommon in hypertrophic cardiomyopathy. This murmur can simulate the diastolic rumble of mitral stenosis. If S1 is accentuated and S4 is mistaken for a presystolic murmur, the acoustic similarity to mitral stenosis can be striking. However, an audible opening snap is rare in HC, and the presence of left ventricular hypertrophy should initiate a careful re-evaluation if mitral stenosis is felt to be the primary diagnosis.

DIFFERENTIAL DIAGNOSIS

The systolic murmur of HC is commonly confused with valvular aortic stenosis, mitral regurgitation, or a ventricular septal defect. Careful synthesis of all the physical findings can usually differentiate these entities, although echocardiography may be required as the final arbiter. The sequelae of coronary artery disease occasionally can be confused with HC.

Valvular Aortic Stenosis. The cardinal differential feature between HC and aortic stenosis is the quality of the carotid pulse. In obstructive HC the carotid upstroke will be quick and the pulse volume normal (Figs. 14-4, 14-8). In aortic stenosis the upstroke is delayed, often slurred and the pulse volume is reduced (Figs. 13-6, 13-8). However, in the elderly population, the stiff systemic arterial tree may produce a brisk or high amplitude arterial pulse in the presence of significant valvular stenosis (see Chapter 3). The

murmur of aortic valve stenosis is usually well heard at the base and radiates into the neck, whereas in HC the murmur is optimally detected lower in the precordium, and as a rule radiates poorly to the base. Post-PVC augmentation is typically much more prominent in HC than in valvular stenosis (Fig. 14-9). The classic response to Valsalva strain is opposite in the two conditions (20 to 30% of HC patients do not show an increase in murmur intensity with Valsalva). The presence of a high frequency aortic ejection click or the murmur of aortic regurgitation is good evidence against HC as being the primary diagnosis. S1 is normal to decreased in valvular aortic stenosis and normal to increased in HC.

Discrete subaortic stenosis can mimic HC. The absence of murmur variability is a useful differentiating feature.

Mitral Regurgitation. Chronic mitral regurgitation may simulate HC. A holosystolic murmur at the apex indicates the presence of mitral regurgitation, with or without a left ventricular outflow tract gradient. An audible or palpable S4 is distinctly unusual in chronic mitral regurgitation but is common in acute onset mitral reflux (see Chapter 17). Evidence for left ventricular hypertrophy usually is much more prominent in HC than in chronic mitral regurgitation. The quality of the carotid pulse may be similar in both conditions but usually has a quicker rise in HC. Reversed splitting of S2 is not a feature of mitral regurgitation. The response to amyl nitrite, Valsalva maneuver, and the post-PVC beat should be useful in the differential diagnosis; the murmur of pure mitral regurgitation behaves in an opposite direction to the responses in HC (Tables 11-3, 11-4, 14-1). Squatting decreases the HC murmur and increases that of mitral regurgitation; standing results in directionally opposite changes.

Mitral Valve Prolapse. The murmur of mitral valve prolapse behaves similarly to HC with many maneuvers (Valsalva, standing, squatting), but the absence of prominent LV hypertrophy and an S4, and the presence of a midsystolic click should easily differentiate the late systolic murmur of mitral prolapse from HC.

Coronary Artery Disease. Chronic coronary artery disease may be a considerable problem in the differential diagnosis, particularly in older subjects. A history of chest pain, ST and T wave changes, or Q waves on the electrocardiogram and a mid late systolic murmur would suggest coronary atherosclerosis in a middle-aged or older patient. A prominent S4 can be seen in either condition. In the elderly subject, one is more likely to think of coronary artery disease when HC may actually be the underlying problem. Echocardiography can readily resolve this dilemma.

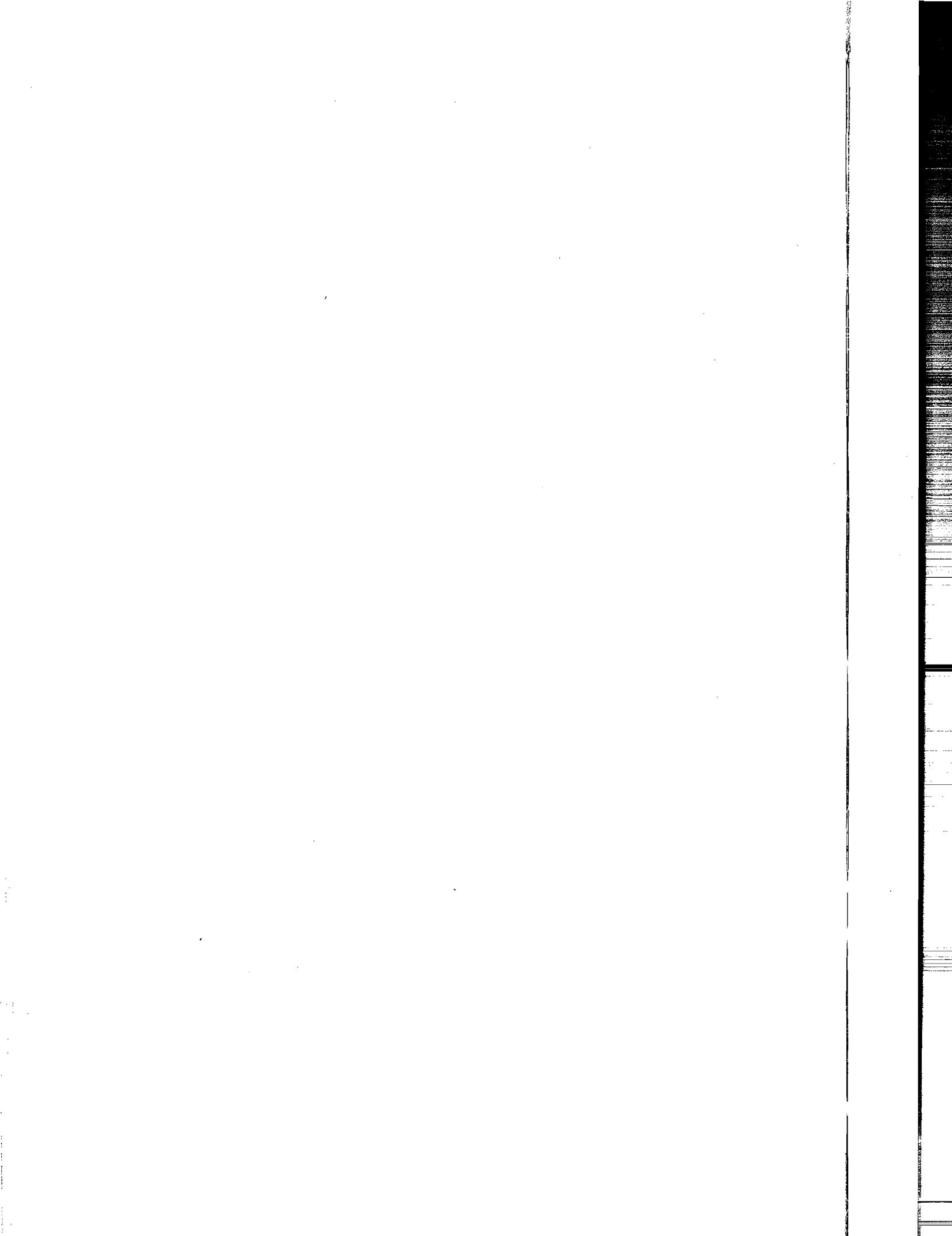
Ventricular Septal Defect. A long systolic or holosystolic murmur at the parasternal areas coupled with an abnormal electrocardiogram (deep Q waves in the lateral leads, RVH pattern) in a young patient with a ventricular septal defect can be confused with HC. Conversely, the findings of HC may

be mistaken for those of a VSD. Careful search for murmur vibrations in late systole and detection of an S4 help resolve this question in a young patient with a confusing murmur. Other differentiating features are the quality of the carotid pulse (brisk in HC) and the response to various maneuvers. The Valsalva maneuver will usually obliterate the murmur of a VSD.

Hypertensive Heart Disease. In older patients with long-standing hypertension and a prominent left ventricular impulse, an ejection murmur in the presence of an S4 may suggest hypertrophic cardiomyopathy. This can be a difficult diagnostic dilemma. Attention to the carotid pulse contour and response to maneuvers and amyl nitrite will aid in this differential diagnosis.

Mitral Stenosis. The increased intensity of S1, prominent S4, and early diastolic murmur of obstructive HC occasionally can simulate mitral stenosis. The S4 is taken for the presystolic component of the diastolic murmur. Some of the acoustic changes simulating mitral stenosis may be related to the thickened anterior leaflet of the mitral valve that commonly is found in patients with obstructive HC. The key to the proper diagnosis of HC is the brisk carotid pulse and the evidence for left ventricular hypertrophy.

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Chapter 15

Aortic Regurgitation

Aortic regurgitation (AR) is an interesting valvular lesion that is associated with a wide variety of underlying disorders. As with mitral regurgitation, there is an acute variant of aortic regurgitation that can be life threatening to the patient. The physical findings in severe chronic aortic regurgitation can be striking and have interested physicians for several centuries.

ETIOLOGY

Rheumatic heart disease remains the most common cause of aortic regurgitation, but many other conditions can produce an incompetent aortic valve such as endocarditis, syphilis, cystic medial necrosis of the aorta, congenital deformities (bicuspid aortic valve is the most common; associated VSD), and diseases of the aortic root. The latter category includes aneurysms of the aorta or sinus of Valsalva but usually is idiopathic. Acute aortic regurgitation can be produced by blunt or penetrating trauma, endocarditis, or dissection. Occasionally, aortic regurgitation is associated with such unusual conditions as ankylosing spondylitis, Reiter's syndrome, and osteogenesis imperfecta.

PATHOPHYSIOLOGY

Aortic regurgitation results from failure of the aortic valve leaflets to coapt completely and remain competent during diastole, thus allowing reflux of blood into the left ventricle. An acquired or congenital structural abnormality is present in the majority of patients, but "functional" aortic regurgitation occasionally has been demonstrated in patients without an intrinsic valve defect. In such situations (e.g., severe hypertension, chronic anemia, patients on renal dialysis) it is likely that vascular volume and/or pressure overload results in aortic ring dilatation to produce the aortic leak. Aortic regurgitation may be caused by a tear or perforation of a single aortic valve cusp or may result from a more diffuse process that affects the valve leaflets, the aortic ring, or both. Intrinsic disease of the aortic root (e.g., aneurysm, dissection) can produce aortic regurgitation in the presence of normal valve leaflets when the proximal aorta is sufficiently dilated or distorted.

The severity of aortic regurgitation is directly related to the amount of ejected stroke volume that refluxes back into the left ventricle during diastole. The regurgitant fraction (RF)* is a useful concept in assessing the severity of the condition. When the regurgitant fraction (RF) is less than one third, the aortic regurgitation is mild. Moderate aortic regurgitation occurs with a RF of 0.30 to 0.60, and in severe or "free" aortic regurgitation, the RF is greater than 0.70. In the latter situation, the actual amount of blood available for perfusing body tissues is less than one third of the ejected total stroke volume.

Normally there is a large pressure gradient across the aortic valve in early diastole, and therefore a small diastolic orifice area may result in a large amount of reflux. In addition to the diastolic surface area of the abnormal aortic valve orifice, other factors play an important role in determining the quantity of aortic regurgitation, including the compliance of the left ventricle, the level of diastolic blood pressure, and the systemic vascular resistance. The duration of diastole (bradycardia vs. tachycardia) and the actual pressure difference across the aortic valve throughout diastole are additional determinants affecting the amount of aortic regurgitation.

Aortic regurgitation produces a classic *volume overload* state. The left ventricle initially dilates and ejects more blood per beat: stroke volume remains proportional to the regurgitant fraction until left ventricular decompensation occurs. Eccentric left ventricular hypertrophy results with prominent chamber enlargement and a proportionately smaller increase in wall thickness. There is an increase in left ventricular compliance in chronic aortic regurgitation; left ventricular filling pressure remains relatively normal in the presence of considerable ventricular dilatation. Cardiac output is maintained, and the ejection rate and ejection fraction are actually increased in early or mild aortic regurgitation. In time, however, major aortic incompetence results in a decrease in the compliance of the ventricle with subsequent elevation of left ventricular filling pressure. Left ventricular contractile reserve may be abnormal in the presence of "normal" systolic function or ejection fraction. At this point, patients typically become symptomatic; congestive heart failure may result when left ventricular contractility becomes sufficiently impaired.

Relationship to Physical Findings. The increase in left ventricular ejection velocity and stroke volume is clinically evident as the ventricle ejects most of its diastolic volume in early systole. Systolic blood pressure rises, systemic vascular resistance falls, and the arterial tree dilates. Peripheral arterial runoff increases and diastolic blood pressure decreases. The regurgitant flow across the aortic valve further contributes to the fall in diastolic blood pressure. The combination of increased systolic blood pressure and decreased diastolic pressure results in a widening of the arterial pulse pressure which can be striking. The chronic decrease in systemic vascular resistance

* regurgitant fraction = $\frac{\text{regurgitant volume}}{\text{stroke volume}}$

and increased pulse pressure produces a broad spectrum of vascular phenomena that have long been associated with severe aortic regurgitation (Table 15-1). The large, rapidly ejected stroke volume contributes to these "peripheral" manifestations. Such alterations are not specific to aortic regurgitation but can occur in any hyperkinetic state associated with a decreased systemic vascular resistance (e.g., thyrotoxicosis, severe anemia, A-V fistula).

Typically, chronic severe aortic regurgitation is well tolerated. In time, the enlarged, low-compliance left ventricle deteriorates; distensibility decreases; left ventricular filling pressure increases; and the ejection fraction falls. As left ventricular failure ensues, reflex peripheral arterial vasoconstriction may actually increase the degree of aortic regurgitation; the low diastolic blood pressure and other signs of peripheral arterial dilatation may disappear. The characteristic diastolic murmur of aortic regurgitation may shorten and become less impressive as the high left ventricular diastolic pressure reduces the aortic-left ventricular gradient and diminishes the degree of reflux in late diastole.

ACUTE AORTIC REGURGITATION

In acute aortic incompetence, the physical findings may be substantially different from those of chronic regurgitation (see page 269). When the left ventricle is challenged abruptly with a severe diastolic overload, normal

TABLE 15-1 *Peripheral or Non-Auscultatory Signs of Severe Aortic Regurgitation: A Glossary of Terms*

Bisferiens pulse	A double or bifid systolic impulse felt in the arterial pulse.
Corrigan's sign	Visible pulsations of the supraclavicular and carotid arteries.
Pistol shot of Traube	A loud systolic sound heard with the stethoscope lightly placed over the femoral arteries.
Palmar click	A palpable, abrupt flushing of the palms in systole.
Quincke's pulse	Exaggerated sequential reddening and blanching of the fingernail beds when light pressure is applied to the tip of the fingernail. A similar observation can be made by pressing a glass slide to the lips.
Duroziez's sign	A to-and-fro bruit heard over the femoral artery when light pressure is applied to the artery by the edge of the stethoscope head. This bruit is caused by the exaggerated reversal of flow in diastole.
De Musset's sign	Visible oscillation or bobbing of the head with each heart beat.
Hill's sign	Abnormal accentuation of leg systolic blood pressure, with popliteal pressure 40 mmHg or higher than brachial artery pressure.
Water-hammer pulse	The high amplitude, abruptly collapsing pulse of aortic regurgitation. This term refers to a popular Victorian toy comprised of a glass vessel partially filled with water which produced a slapping impact on being turned over.
Müller's sign	Visible pulsations of the uvula.

compensatory mechanisms do not have time to develop. The LV is unable to adequately handle the large regurgitant flow, and the left ventricular filling pressure becomes markedly elevated. The effective cardiac output falls. Peripheral vascular signs of severe aortic regurgitation are attenuated or may never appear due to marked systemic vasoconstriction. The heart rate is usually increased. Left ventricular failure in this clinical context carries a grave prognosis. Therefore, early recognition of acute aortic regurgitation is imperative.

ROLE OF THE CLINICIAN

Detection of a diastolic decrescendo murmur localized to the left sternal border indicates a probable abnormality of the aortic valve or ascending aorta and should prompt a search for an appropriate etiology. Pulmonic regurgitation can produce an identical murmur, but this valve lesion is uncommon in adults in the absence of pulmonary hypertension. Because acute aortic regurgitation is a potentially lethal condition, the duration of existence of aortic incompetence in any given patient is a critical consideration. Is the aortic reflux minimal with only a modest volume overload, or is there evidence of a severe degree of regurgitation with a large regurgitant fraction resulting in major cardiac enlargement? In most instances, the astute clinician can accurately estimate the severity of aortic regurgitation by a careful assessment of the cardiac and peripheral vascular findings on physical examination.

PHYSICAL EXAMINATION

Blood Pressure

The systemic blood pressure is a valuable clue to the degree of aortic regurgitation. With progressive severity of aortic reflux, the systolic blood pressure increases and the aortic diastolic pressure declines (Fig. 15-1). *Practical Point:* *The presence of a completely normal blood pressure in a patient with aortic regurgitation and excellent left ventricular function virtually excludes moderate to severe aortic incompetence.* In hemodynamically significant aortic regurgitation, diastolic pressure typically falls below 70 mmHg and systolic pressure may rise to 140 to 150 mmHg. Systolic pressure will not increase more than 150 to 160 mmHg in the absence of associated systemic hypertension. An aortic regurgitation murmur and associated blood pressure of 180/90 does not imply severe aortic reflux even though the pulse pressure is wide. More likely this represents the presence of hypertension and coexisting aortic incompetence.

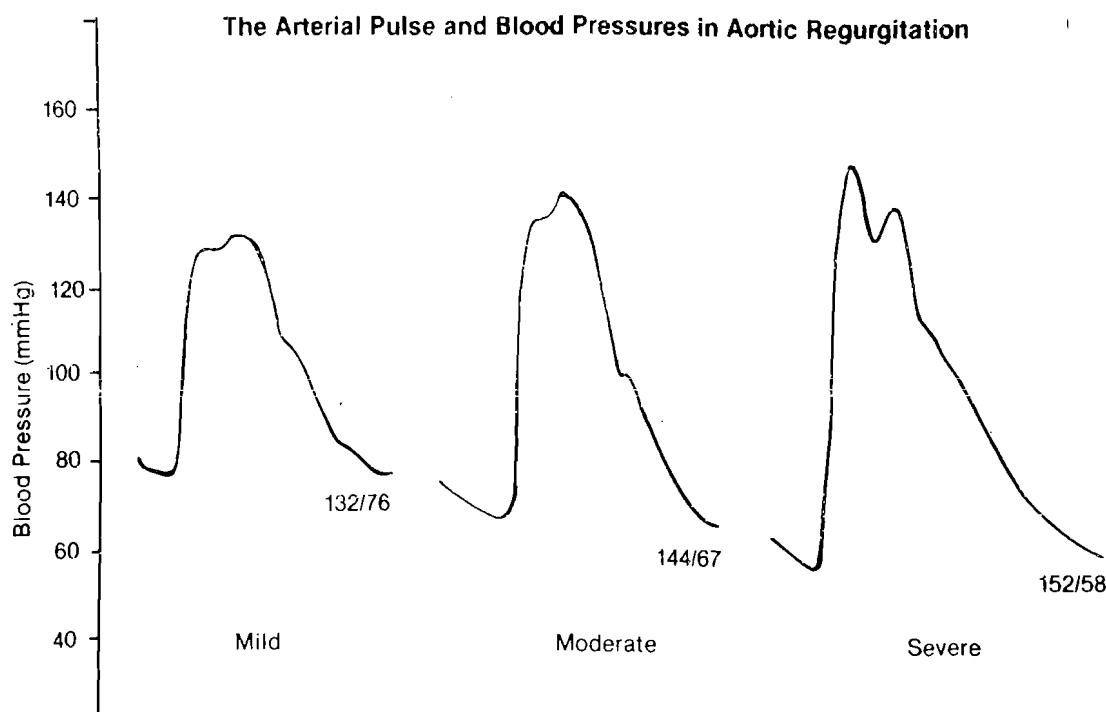


FIG. 15-1. The arterial pulse and blood pressure in aortic regurgitation. There is little alteration in the arterial pulse and pressure in mild aortic regurgitation. With increasing reflux across the aortic valve, systolic blood pressure increases and diastolic blood pressure decreases, resulting in widening of the pulse pressure. The increased left ventricular stroke volume results in a high amplitude, palpable arterial pulsation with rapid falloff in late systole. This produces a collapsing quality to the pulse. In severe aortic regurgitation, arterial pulsations are prominent and often visible in the peripheral circulation as well. (From Abrams J: Prim Cardiol, 1983.)

The degree of decrease in diastolic blood pressure is a better benchmark for assessing the severity of aortic regurgitation than the increase in systolic pressure. In severe or "free" aortic incompetence, the diastolic blood pressure is in the range 40 to 50 mmHg, approaching or equalling the markedly elevated LVEDP. True aortic diastolic blood pressure is never lower than maximal ventricular diastolic pressure, although cuff blood pressure determinations in patients with severe aortic regurgitation often indicate readings of zero. The Korotkoff sounds in such patients are often audible down to 0 to 10 mmHg. It is best to use the point of *muffling* of Korotkoff sounds as the best indication of diastolic blood pressure in subjects with aortic regurgitation. When possible, record the diastolic pressure at both the level of muffling and disappearance of sound. Direct intraarterial pressure measurements can be very discordant from cuff pressures in aortic regurgitation, particularly with respect to the "true" diastolic pressure.

Attempts have been made to assess the severity of aortic regurgitation using the systemic blood pressure or a ratio of the pulse pressure to the systolic blood pressure. In general, a wide pulse pressure indicates a major degree of reflux; however, on occasion, patients with mild aortic incompetence

will have an increased pulse pressure representing more than 50% of the systolic blood pressure. Because there are so many false-positive and false-negative values, the ratio of pulse pressure to systolic pressure is not a reliable index of the magnitude of regurgitation. For instance, elderly patients may have an elevated systolic blood pressure due to the "physiologic" decrease in elasticity of the arterial tree (see Chapter 3), which may result in a wide pulse pressure irrespective of the degree of aortic regurgitation. Conversely, the blood pressure may "normalize" in the presence of decreased left ventricular function or congestive heart failure resulting from severe aortic regurgitation. In these situations, peripheral arterial vasoconstriction causes an elevation of diastolic blood pressure. Systolic pressure may fall due to a depressed ejection fraction and smaller stroke volume, and the pulse pressure-systolic pressure ratio will be falsely normal. *Practical Point:* Severe isolated aortic regurgitation uncommonly will be present in a patient with a diastolic blood pressure greater than 60 mmHg if there is no evidence of reduced left ventricular pump function or systemic hypertension. Conversely, a diastolic blood pressure of 50 mmHg or less almost always indicates a major degree of aortic incompetence, irrespective of the level of systolic pressure.

Heart Rate. Tachycardia shortens the time for diastolic reflux and thus may increase the diastolic blood pressure in aortic regurgitation. Conversely, bradycardia will accentuate the decrease in diastolic blood pressure because of the increased duration of aortic regurgitation.

Leg Blood Pressure. The normal increase in systolic blood pressure found in arteries distal to the proximal aorta is exaggerated in aortic regurgitation. In severe aortic incompetence, systolic pressure in the popliteal artery is much higher than in the brachial artery (Hill's sign). A difference in systolic blood pressure between the arms and the legs of greater than 40 to 50 mmHg typically is seen in major degrees of aortic regurgitation.

Arterial and Carotid Pulses

The large stroke volume and enhanced rate of ejection produced by hemodynamically significant aortic regurgitation results in a characteristic *high amplitude arterial pulse* (Fig. 15-1). The pulse has a *collapsing* quality caused by the low systemic vascular resistance and early diastolic reflux of blood into the left ventricle which result in rapid "unloading" into the aorta. Thus, the classic carotid pulse of aortic regurgitation is one of an increased force and amplitude of ejection followed by an abrupt falling away or collapse. With a very large stroke volume, the pulse is quite full, literally swelling under the examining finger, then rapidly abating. In advanced disease, the dilated brachiocephalic arterial tree dilates, and arterial pulsations in the sternal notch and supraclavicular area are easily seen and often are palpable. Pulsation of proximal carotid arteries may be visible (Corrigan's sign). Patients

may be conscious of this arterial throbbing. Arterial pulses throughout the body are increased in force and amplitude and display a typical bounding quality. These changes are less reliable in elderly subjects.

Bisferiens Pulse. One of the hallmarks of aortic regurgitation is the bisferiens or double systolic arterial pulse (Fig. 15-2). This bifid pulsation is best felt with light finger pressure over the carotid arteries. On occasion, a transmitted systolic thrill or bruit is also felt, making precise identification of the bisferiens contour difficult. In severe aortic regurgitation, a systolic shudder of the carotid artery may be noted with or without associated aortic

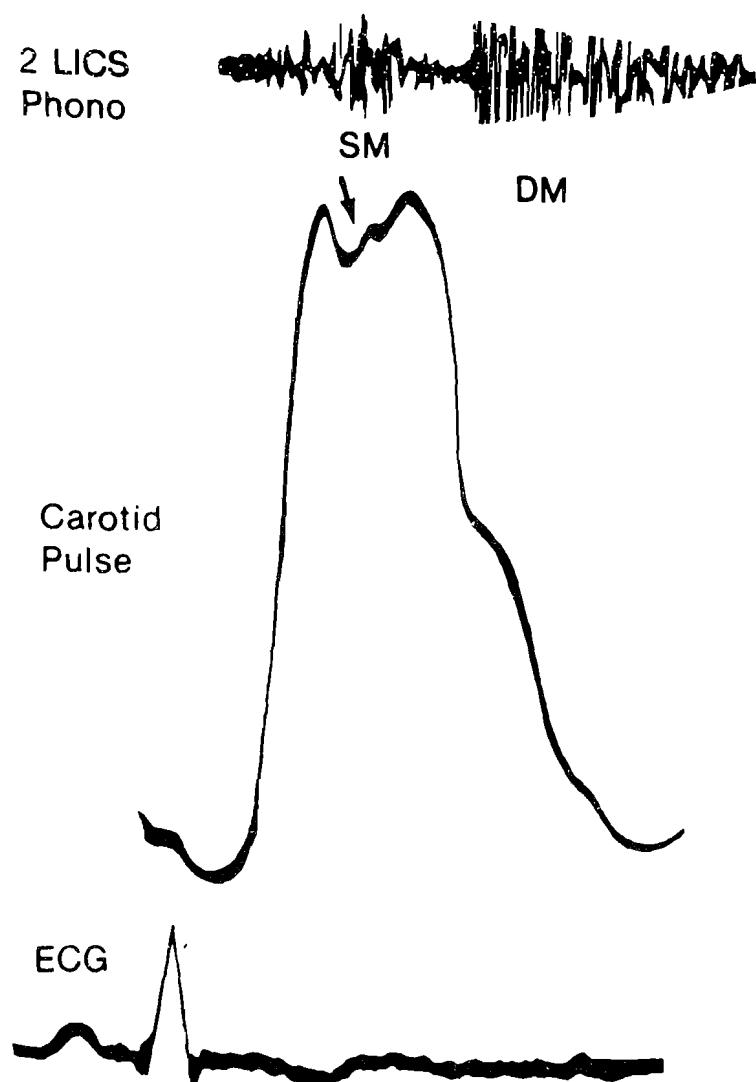


FIG. 15-2. Bisferiens pulse of aortic regurgitation. Note the bifid systolic pulse wave, which is best detected using light finger pressure over the carotid arteries. This contour is associated with an increased pulse volume. The bisferiens pulse must be differentiated from a transmitted systolic murmur or palpable thrill. Note the soft S1 and S2. SM = systolic murmur; DM = diastolic murmur; 2 LIC = 2nd left intercostal space. (From Abrams J: Prim Cardiol, 1983.)

stenosis. Often this double-beating pulse is exaggerated in the periphery and, therefore, it should be sought in the brachial, radial, and femoral arteries. The bisferiens pulse usually is present only in moderate to severe aortic regurgitation or in patients with aortic regurgitation and associated mild aortic stenosis. In the latter case, the aortic regurgitation is always dominant. The precise mechanism underlying the bisferiens pulse is unclear. A bisferiens pulse is more likely to be found in younger patients; perhaps the arterial tree is more distensible than in the elderly.

Peripheral Signs (Table 15-1)

Major degrees of aortic incompetence produce a wide variety of abnormalities directly related to forceful ejection of an increased stroke volume into a dilated arterial bed. The abrupt rise and fall of the arterial pulse wave causes a distinctive pounding or collapsing quality that is accentuated in the peripheral arteries. Marked systemic vasodilation may produce noncardiac phenomena such as increased sweating, warm flushed skin, and accentuated retinal vein pulsations.

Many well-known eponyms have been given to these *nonauscultatory signs of aortic regurgitation*. These phenomena are not pathognomonic for aortic incompetence and, occasionally, can be noted patients with a hyperkinetic circulation and marked arterial vasodilation from other causes.

In general, the presence of such peripheral abnormalities correlates well with the increased systolic and pulse pressure and decreased diastolic pressure common to advanced aortic regurgitation. *Practical Point: Nonauscultatory signs are never seen in mild aortic regurgitation but are the rule in chronic severe aortic incompetence in the absence of congestive heart failure.* Low cardiac output or heart failure may attenuate these signs. Arterial vasoconstriction occurs as stroke volume falls. An increased heart rate in congestive heart failure (CHF) may also play a role in blunting the signs of aortic regurgitation. Table 15-1 lists most of the peripheral abnormalities that have been described in aortic regurgitation.

Precordial Motion

The quality of the left ventricular impulse, in general, parallels the severity of the aortic regurgitation. In *mild to moderate* aortic regurgitation, the left ventricular impulse is *normal in size but is often hyperdynamic*. Thus, the amplitude is exaggerated, but there is no leftward displacement of the apex beat; the apical impulse falls away from the palpating finger by midsystole (Fig. 15-3B). With greater degrees of aortic regurgitation, the hyperkinetic

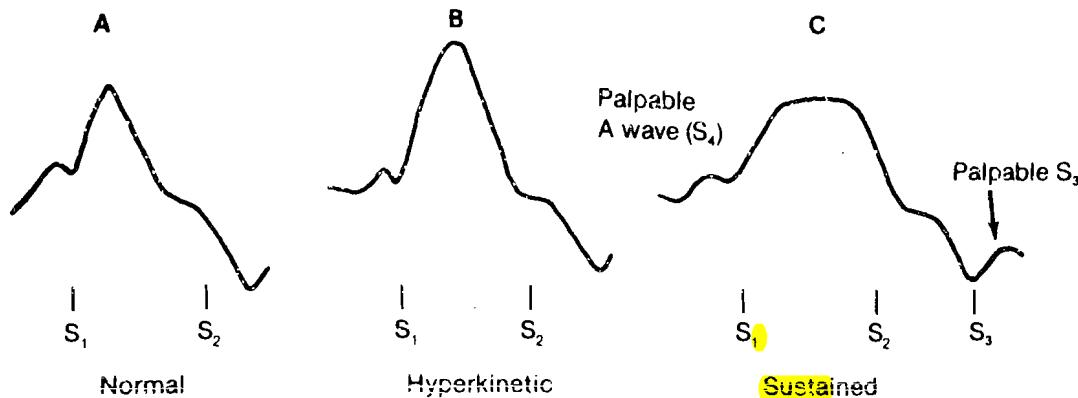


FIG. 15-3. Precordial motion patterns in aortic regurgitation. A. In mild regurgitation, the left ventricular impulse is normal. B. As the left ventricle dilates with increasing volume overload, the apical impulse becomes more forceful with a higher amplitude (hyperkinetic impulse). Outward precordial motion is still felt only in early systole. The site of the PMI may be displaced laterally and downward. C. A sustained left ventricular impulse will be felt when the ventricular cavity is substantially enlarged. At this stage, left ventricular contractility and ejection fraction may be preserved or may be abnormally depressed. A sustained impulse or left ventricular heave results in palpable systolic activity into the second half of systole. The PMI is always displaced to the left and inferiorly at this stage, and the force of the apical impulse is usually increased. The impulse takes up a larger area than normal on the chest wall. An A wave (S₄) and rapid filling wave (S₃) are often palpable in the left lateral position. (From Abrams J: Prim Cardiol, 1983.)

impulse becomes more prominent, and the cardiac apex is displaced inferolaterally. When LV dilatation or a decrease in the ejection fraction occurs, resulting in an increased end-systolic volume, the apex beat becomes *sustained* (Fig. 15-3C). In patients with very large hearts or significant depression of left ventricular function, a prolonged and forceful LV lift or heave is predictable, and this finding may be quite impressive.

Downward and lateral displacement of the apex impulse reflects the major volume overload that develops in chronic, severe aortic regurgitation. The normal elliptical LV cavity becomes spherical as chamber dilatation progresses. In severe aortic regurgitation, the apex impulse typically is found in the left anterior axillary line at the 5th or 6th interspace, usually occupies at least two interspaces, and is sustained into late systole. The area medial to the left ventricular apex may demonstrate rather prominent retraction. Not uncommonly, a palpable S₄ (presystolic distension) may be noted in the left decubitus position (Figs. 15-3C, 7-6). This finding correlates with an elevated LEVDP and suggests decreased left ventricular compliance. However, the LVEDP may be quite high without a palpable or audible S₄. A visible and palpable rapid LV filling wave or palpable S₃ may be detected in severe aortic regurgitation, reflecting increased end-diastolic blood volume and not necessarily implicating severe left ventricular dysfunction (Fig. 15-3C).

HEART SOUNDS

First Heart Sound. S1 has a normal intensity in mild to moderate cases but is often decreased in severe aortic regurgitation (Fig. 15-4). The decrease in amplitude of S1 in moderate to severe aortic regurgitation has several possible explanations. (1) The PR interval is commonly prolonged in aortic regurgitation. (2) Left ventricular contractility is frequently depressed in chronic, severe aortic regurgitation. (3) In severe aortic regurgitation with diminished left ventricular compliance or overt left ventricular failure, diastolic pressure within the ventricle is markedly elevated. The increased LVEDP can result in premature closure of the mitral valve, causing the mitral cusps to coapt earlier than normal in late diastole. S1 will be soft whenever the mitral valve is closed at the onset of left ventricular contraction (see Chapter 6).

An aortic ejection sound is easily mistaken for S1 in aortic regurgitation; this can give a false impression of a normal or even loud S1. Whenever "S1" is particularly prominent at the base in patients with aortic incompetence, an aortic ejection click instead of S1 probably is being heard.

Second Heart Sound Intensity. The aortic component of S2 is quite variable in intensity in patients with aortic regurgitation. Although many textbooks have emphasized an increased amplitude of A2, recent work indicates the A2 may be softer than usual in aortic regurgitation because of a decreased ability of the valve leaflets to vibrate after aortic valve closure. A2 may be accentuated or tambour in quality in patients with syphilitic aortitis or other types of aortic root disease. In young persons, particularly those with only mild to moderate aortic regurgitation, A2 usually is slightly decreased.

Splitting of S2. In patients with moderate to severe aortic regurgitation, S2 is often single. This finding may be a result of a prolonged left ventricular ejection time (A2 moves into P2) or to inaudibility of A2, or both. P2 may become "lost" in a prominent diastolic murmur, giving the impression of a single second sound. Reversed or paradoxical splitting of S2 is uncommon unless there is underlying left bundle branch block.

Third Heart Sound. An S3 is not a feature of mild to moderate aortic incompetence. However, in severe aortic regurgitation an S3 is common (Fig. 15-4). The increase in LV diastolic blood volume is in part responsible for the prominent S3; left ventricular dilatation and decreased contractility may be significant additive factors. The S3 is frequently a visible and palpable event, coinciding with the rapid filling phase of left ventricular diastole. An S3 is more likely to be heard in younger patients with aortic regurgitation. In general, an audible S3 correlates well with the presence of peripheral manifestations of aortic regurgitation and a large left ventricular end-diastolic

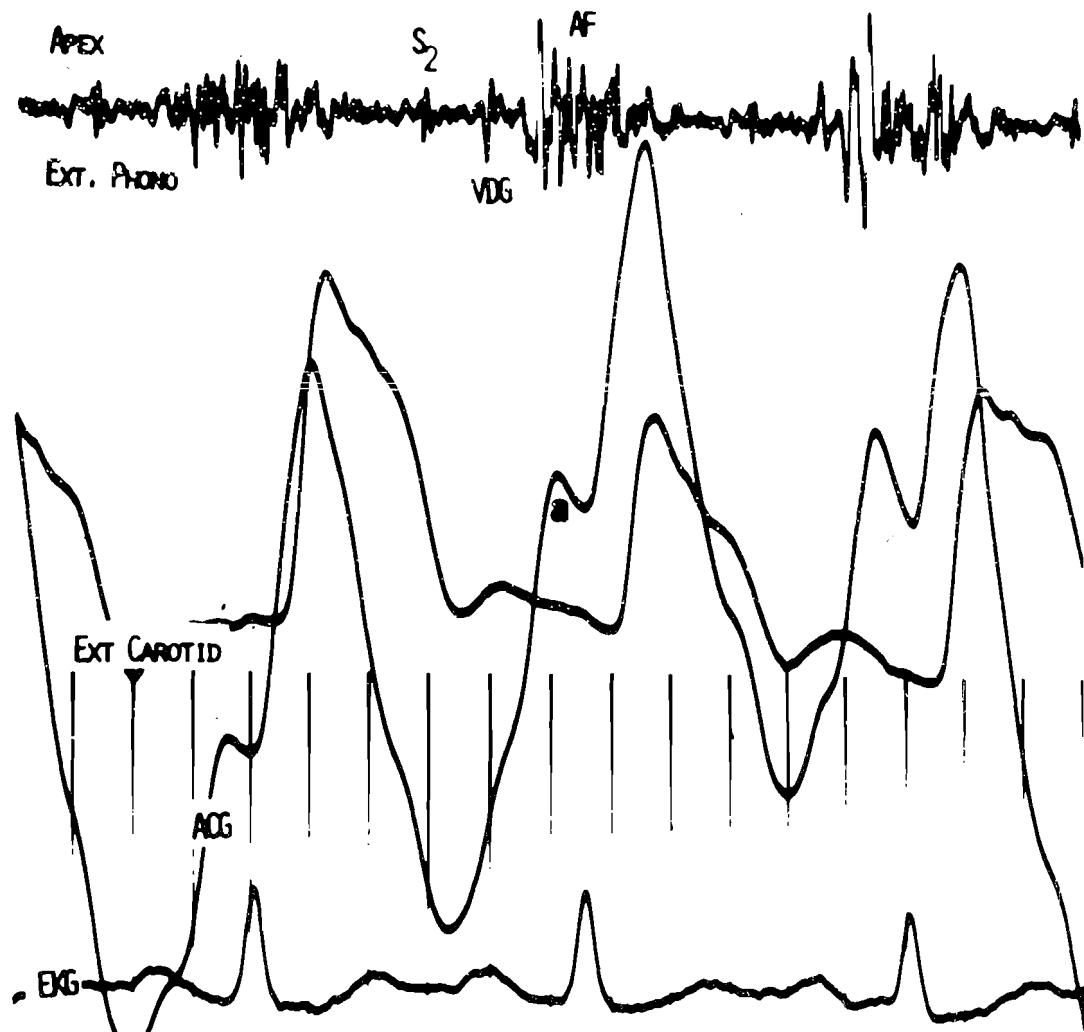


FIG. 15-4. Third heart sound and Austin Flint murmur in acute aortic regurgitation. This phonocardiogram demonstrates an S3 or ventricular diastolic gallop (VDG) followed by a loud Austin Flint murmur (AF) extending into late diastole. Note the accentuation of this murmur with atrial systole. The apex cardiogram (ACG) reveals a markedly augmented A wave and a hyperdynamic LV impulse. S1 is virtually absent. The auscultatory findings in such a patient may be extremely difficult to interpret. The presence of an S3 and an Austin Flint murmur in acute or chronic aortic regurgitation indicates a major degree of aortic reflux (From Reddy PS, et al: Syndrome of acute aortic regurgitation. In Physiologic Principles of Heart Sounds and Murmurs. Edited by DF Leon and JA Shaver. American Heart Association Monograph No. 46, 1975.)

volume. An S3 often initiates an Austin Flint rumble when present (see page 265; Fig. 15-4).

Fourth Heart Sound. While an atrial sound is an uncommon finding in mild aortic regurgitation, it may occasionally be found in moderate aortic incompetence and is common in severe disease. The long PR interval common in many patients with aortic regurgitation increases audibility of an S4. An S4 suggests increased left ventricular stiffness and an elevated LVEDP. The S4 may be manifest at the apex as palpable presystolic distension when the

patient is turned onto his left side (Figs., 7-6, 15-3C). In some persons, the S4 is of such low frequency and intensity as to be inaudible, although it is readily palpated.

Ejection Sound. An aortic ejection click or sound is detectable in many subjects with aortic regurgitation. More commonly it is heard in mild disease when there is good left ventricular function. The ejection click may be of valve or root origin (see Chapter 8). When there is a bicuspid valve, the ejection click is produced by the maximal opening excursion of the abnormal valve cusps. In subjects with an enlarged, abnormal aortic root, the ejection sound is most likely produced by sudden systolic expansion of the ascending aorta itself during early ejection (Fig. 8-2B).

In severe aortic regurgitation, a low aortic diastolic pressure may cause the ejection sound to occur earlier as the subsequent shortening of LV isovolumic systole results in earlier aortic valve opening. The ejection click may merge with S1 and no longer be audible as a separate auditory event. A snappy or prominently split S1 or an S1 that is well heard at the base in a patient with aortic regurgitation should suggest to the clinician that the S1 is actually an aortic ejection click.

Cardiac Murmurs

Three different murmurs may be found in patients with aortic regurgitation. (1) The classic decrescendo diastolic murmur results from reflux of blood into the left ventricle. (2) A systolic ejection murmur is produced by the large stroke volume, increased rate of ejection, and abnormal valve anatomy. (3) The Austin Flint murmur is a low-pitched diastolic murmur beginning in mid-diastole and found only in major degrees of aortic regurgitation.

Diastolic Murmur. The typical diastolic murmur of aortic regurgitation has a decrescendo shape (Figs., 15-2, 15-5, 15-6). The volume and velocity of refluxing blood across the incompetent aortic valve tapers off in mid to late diastole as the aortic-left ventricular pressure gradient decreases. *In general, the length of the AR murmur is dependent on the severity of the leak except in very severe aortic regurgitation (see below; Fig. 15-6).* The classic finding in AR is a diastolic murmur that tapers during diastole and may or may not extend to S1. The maximal diastolic pressure difference between the aorta and left ventricle occurs immediately following left ventricular isovolumic relaxation when left ventricular diastolic pressure falls to its lowest point in early diastole (Fig. 15-6). For this reason the murmur of aortic regurgitation may have a brief early crescendo contour (Fig. 15-5), with its peak intensity occurring synchronously with the maximal early diastolic pressure gradient. This phenomenon is more readily recorded than heard. On occasion, it may produce a "gap" between A2 and the apparent onset of the diastolic murmur. Astute observers may be able to appreciate an early, short

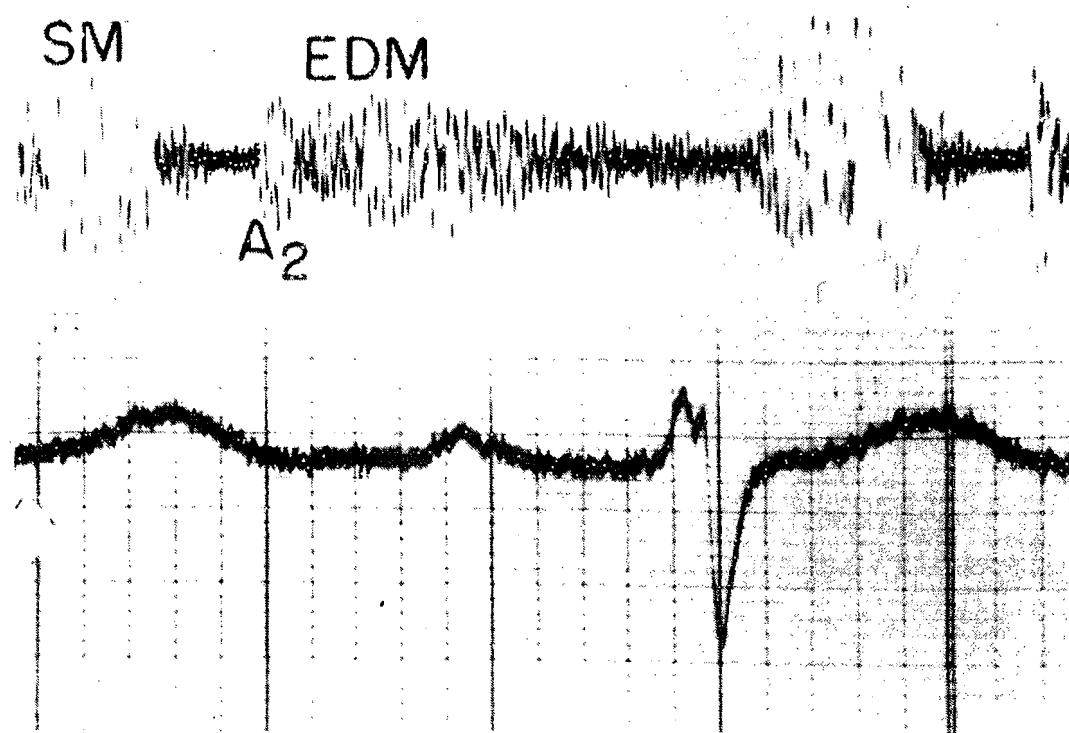


FIG. 15-5. Diastolic murmur of severe aortic regurgitation. This phonocardiogram, taken at the second right intercostal space in a patient with chronic severe aortic regurgitation, reveals a classic long diastolic murmur (EDM) that has a brief crescendo configuration with a long decrescendo portion. Note the extremely soft S1 (not labeled) and the prominent midsystolic ejection murmur (SM). The latter is related to a very large stroke volume ejected rapidly across the aortic valve and does not necessarily imply associated aortic stenosis. (From Perloff JK: Murmurs. In Signs and Symptoms in Cardiology. Edited by LD Horwitz and BM Groves. Philadelphia, JB Lippincott Co, 1985.)

crescendo component initiating a longer decrescendo diastolic murmur, particularly in mild degrees of AR.

Frequency. The diastolic murmur typically is high frequency or ‘blowing’ in quality in mild to moderate aortic regurgitation. The high frequency vibrations are caused by a high velocity of flow and a relatively small regurgitant volume. The classic murmur may be mimicked by whispering “ah” or “R” or by exhaling with the lips pursed. This murmur can easily simulate breath sounds. Therefore, it is important to auscultate as the patient holds his breath in held-expiration so as not to confuse the AR murmur with the noise of breathing. *Practical Point: The high frequency murmur of mild aortic regurgitation is frequently soft. As most physicians are unaccustomed to hearing sounds of such high pitch, this faint AR murmur is easily missed by the unsuspecting examiner.* The murmur in trivial degrees of aortic incompetence may be quite short, another factor that can impair audibility (see below).

With more severe degrees of AR, low to medium frequency sound vibrations occur, and the murmur may be surprisingly low pitched, probably as a result of excessive turbulence in the left ventricular outflow tract. The

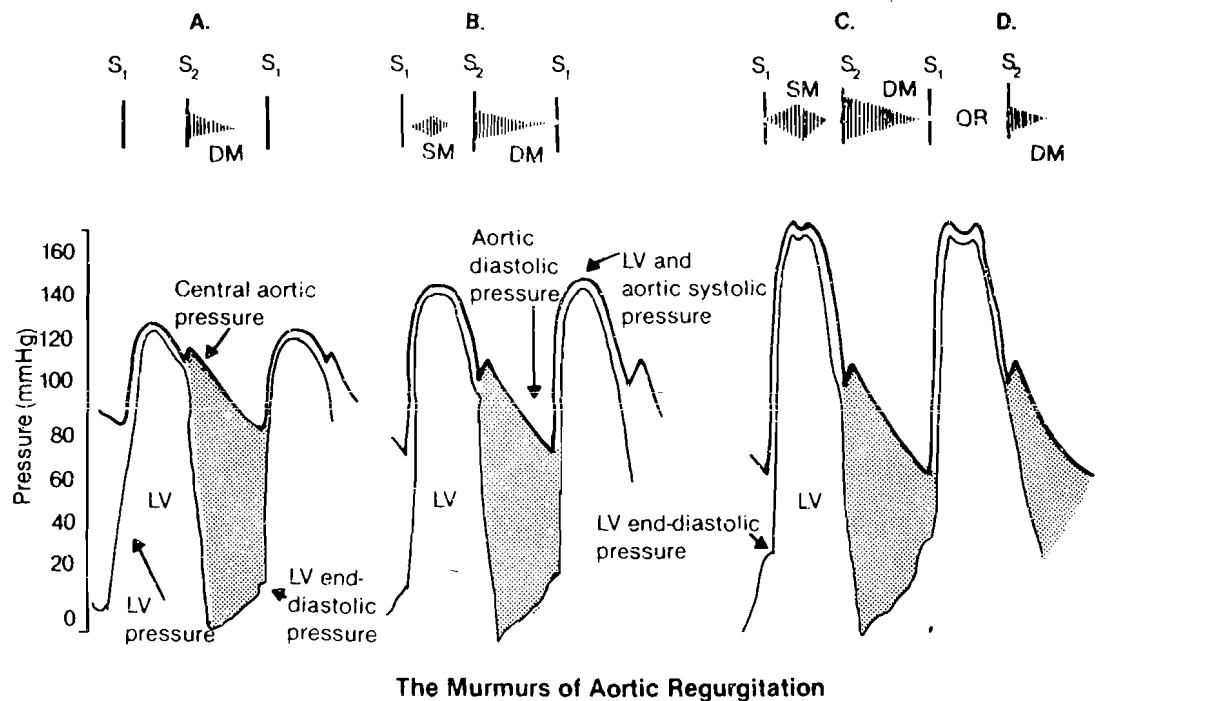
**The Murmurs of Aortic Regurgitation**

FIG. 15-6. Diastolic and systolic murmurs of aortic regurgitation. This diagram shows the relationship between the severity of aortic regurgitation, hemodynamic findings, and the characteristic murmurs of mild, moderate, and severe aortic regurgitation. A. Mild aortic regurgitation. The diastolic gradient between the aorta and left ventricle is large; reflux of blood is high in velocity and small in volume. The resultant diastolic murmur (DM) is of variable length and high frequency. A systolic ejection murmur (SM) may not be present. B. Moderate aortic insufficiency. The diastolic murmur is decrescendo and invariably long. The murmur may have medium or high frequency vibrations. A systolic ejection (flow) murmur is common. C,D. Severe aortic regurgitation. The length of the diastolic murmur is variable. With preserved left ventricular function (C), the murmur is usually pandiastolic and medium frequency (high velocity, large volume reflux). The systolic murmur may be very prominent and long in duration, even in the absence of aortic stenosis. If there is left ventricular failure or the late end-diastolic pressure is markedly elevated, resulting in a narrow gradient between the aorta and left ventricle during the last third of diastole, the diastolic murmur may be short and unimpressive (D). The systolic murmur may also be unremarkable in this setting; this combination can result in a serious underestimation of the severity of aortic regurgitation. (From Abrams J: Prim Cardiol, 1983.)

pitch of the regurgitant murmur is probably influenced by the large volume of refluxing blood. In addition, as the diastolic aortic-left ventricular gradient decreases, the velocity of reflux may be less rapid than with lesser degrees of AR, and the murmur becomes more low pitched.

Duration. In mild degrees of aortic regurgitation (regurgitant fraction of less than 0.3 to 0.4), the diastolic murmur may not be audible in late diastole, although a large gradient between the aorta and left ventricle persists throughout diastole (Fig. 15-6A). In fact, the murmur of trivial or very mild AR may be so short as to be little more than a blurring sound off A2. Such murmurs typically are low amplitude and high frequency and are very hard to hear without selective focus on early diastole. With increasing degrees of

aortic reflux, the murmur may become truly holodiastolic but remains decrescendo in quality (Fig. 15-6B, C). Thus, in a patient with a diastolic blood pressure of 60 mmHg or less, a pandiastolic murmur would be anticipated. In less severe aortic incompetence, the murmur may taper off more gradually than in subjects with a bigger leak, where the AR murmur often loses substantial intensity by mid-diastole. Thus, some patients with *severe* aortic regurgitation may actually have a *shorter* decrescendo diastolic murmur than those with milder disease (Fig. 15-6D). "Free" aortic regurgitation commonly is associated with a very high late left diastolic ventricular filling pressure and a low aortic diastolic pressure. This results in a small aortic-left ventricular gradient in late diastole so that the amount of reflux may be quite small or nil at end-diastole (Fig. 15-6D).

With the onset of congestive heart failure, it is common for the aortic diastolic murmur to shorten and decrease in intensity. This can give the false impression of less severe aortic regurgitation than is actually present.

Intensity. The aortic regurgitation murmur ranges in loudness between vary faint to quite prominent, but is rarely of grade 4-5/6 intensity and only occasionally has an associated thrill. Trivial degrees of aortic regurgitation produce murmurs that are typically soft. It is important not to miss such a faint murmur, as the presence of any degree of AR documents a pathologic condition of the aortic valve or root and supports the diagnosis of organic heart disease. The high-pitched aortic regurgitant blow is often "close to the ear" and demands a conscious "tuning in" to early diastole by the auscultator. Maneuvers such as mild exercise, handgrip, squatting, or administration of a pressor, such as phenylephrine, will enhance the audibility of these murmurs by increasing their intensity.

Although the amplitude of the diastolic murmur roughly correlates with severity, some persons with a small degree of aortic leak will have a loud murmur. Patients in heart failure resulting from severe aortic regurgitation occasionally may have no more than a faint, short murmur. Severe AR can actually be silent in rare cases.

Optimal Sites for Auscultation. The murmur of aortic regurgitation usually is best heard adjacent to the sternum between the 2nd and 4th left interspace. In mild or trivial AR, the murmur may be localized only to the 2nd left interspace at the left sternal edge. With increasing severity, the maximal intensity of the murmur may be as low as the 4th or 5th interspace. The murmur is often audible at the apex when its intensity is grade 2-3/6 or more, and in some subjects it actually may be best heard at the apex. This finding is more likely to occur in elderly patients, perhaps because of tortuosity of the proximal aorta with streaming of the regurgitant jet towards the LV apex. The AR diastolic murmur typically fades away and may become inaudible between the site of maximal audibility at the left sternal border and the cardiac apex, where it again becomes recognizable.

Aortic Regurgitation Heard to the Right of the Sternum (Table 15-2).

The typical aortic regurgitation murmur may be heard easily at the 2nd to 3rd right interspace where it can be quite prominent. However, the murmur is usually not detectable lower down the right side of the sternum. Many years ago, Levine and Harvey observed that, on occasion, an aortic regurgitation murmur is very well heard at the *lower right sternal border* (3rd to 5th right interspace). They noted that this phenomenon occurred commonly in patients who had *disease of the aortic root* rather than at the valve level. They postulated that a rightward and superior displacement of the dilated proximal aorta could cause the murmur to radiate down the right side of the sternum. Thus, in *aortic dissection, aortic aneurysm, sinus of Valsalva aneurysm, or any patient with marked tortuosity of the aorta, the murmur of AR may be louder to the right of the sternum than to the left.* The recognition of such a murmur should stimulate a search for a *nonrheumatic cause* for the aortic regurgitation (Table 15-2).

Harvey has emphasized that this observation is only valid if the murmur is *best heard at the right 3rd-4th interspace or lower.* Conversely, the AR murmur in patients with an enlarged aortic root may be maximally heard down the left sternal edge or have equal intensity on both sides of the sternum.

Aids to Auscultation. Special techniques are not necessary to increase audibility of a loud regurgitant murmur. However, the elusive, faint, high-pitched blowing murmur of AR can be made more audible by certain maneuvers. In some cases, these maneuvers may be necessary in order to hear the murmur. The optimal method of auscultation is as follows: The examination should take place in a quiet room with the patient sitting, standing, or leaning forward with the breath held in mid or full expiration. Use firm pressure with the diaphragm of the stethoscope (enough to leave an imprint on the skin). Background noise such as that from air conditioning or the patient's own breath sounds can readily mask detection of a high-pitched, grade 1-2/6 aortic murmur. When there is a question or no murmur is heard but there is reason to believe that aortic regurgitation is present, the use of sustained handgrip or squatting may "bring out" a faint murmur. Both maneuvers increase systemic resistance and central aortic pressure and, consequently, can augment the degree of aortic reflux making the murmur more

TABLE 15-2 *Conditions Resulting in an Aortic Regurgitation Murmur Heard Best on the Right Side of the Sternum*

Aortic aneurysm
Cystic medial necrosis
Syphilis
Idiopathic
Sinus of Valsalva aneurysm
Aortic dissection—acute or chronic
Selective perforation or eversion of the right coronary cusp

audible. Simultaneous application of blood pressure cuffs to both arms for 20 seconds inflated to a level 20 mmHg above systolic pressure has recently been reported to be very useful in bringing out the diastolic murmur of AR.

In some patients, particularly younger persons, the AR murmur may be more easily heard in the supine position. Remember that in older adults, especially those with chronic lung disease or congestive heart failure, the AR murmur may be maximal or heard only at the LV apex; this can be misleading. Some authors refer to this phenomenon as "silent aortic regurgitation." In an older patient with heart failure, the diagnosis of aortic valve disease may be missed completely if this apical diastolic murmur is not appreciated. Occasionally, the AR murmur is best heard in the axilla (Cole-Cecil murmur). In some subjects, the murmur of aortic regurgitation also may be heard in the supraclavicular area.

The Musical Murmur of Aortic Regurgitation. In rare individuals, the regurgitant jet produces a resonating, vibratory diastolic murmur. This murmur consists of relatively pure frequency sound that may wax and wane throughout diastole. This is often called the "cooing-dove" or "seagull" murmur. It is more likely to be heard with perforation or rupture of an aortic cusp or retroversion of a leaflet (e.g., following chest trauma). This murmur may be very loud, may be accompanied by a thrill, and may radiate to the neck or down the right side of the sternum. When present, it strongly suggests a nonrheumatic etiology for the aortic incompetence.

Systolic Murmur. A systolic ejection murmur is common in moderate to severe aortic regurgitation. It results from an abnormally large stroke volume that is ejected with rapid force often across an anatomically deformed aortic valve into an enlarged proximal aorta. *Practical Point: A loud systolic murmur in a patient with severe aortic regurgitation does not necessarily imply coexisting aortic stenosis.* Typically, this systolic murmur is short and peaks before the second half of systole if there is no aortic valve obstruction (Figs. 15-5, 15-6B, C, 15-7A). However, with a very large ejected stroke volume the systolic murmur lengthens in proportion to the increase in left ventricular ejection time. Thickened aortic valve leaflets with minimal reduction in area of the aortic orifice in the absence of a systolic left ventricular-aortic gradient may accentuate the intensity and duration of the systolic murmur. In such instances, the systolic ejection murmur may be both loud and long without true aortic stenosis being present. Severe aortic regurgitation without stenosis rarely may be associated with a grade 4/6 systolic murmur and an accompanying thrill. In these difficult cases, the focus should be on any nonauscultatory signs of aortic regurgitation, the blood pressure, and the quality of the carotid pulse to assess the possibility of associated aortic stenosis. If the diastolic blood pressure is low, the pulse volume is full and bounding, and there is evidence of peripheral vasodilation, aortic stenosis can usually be readily excluded.

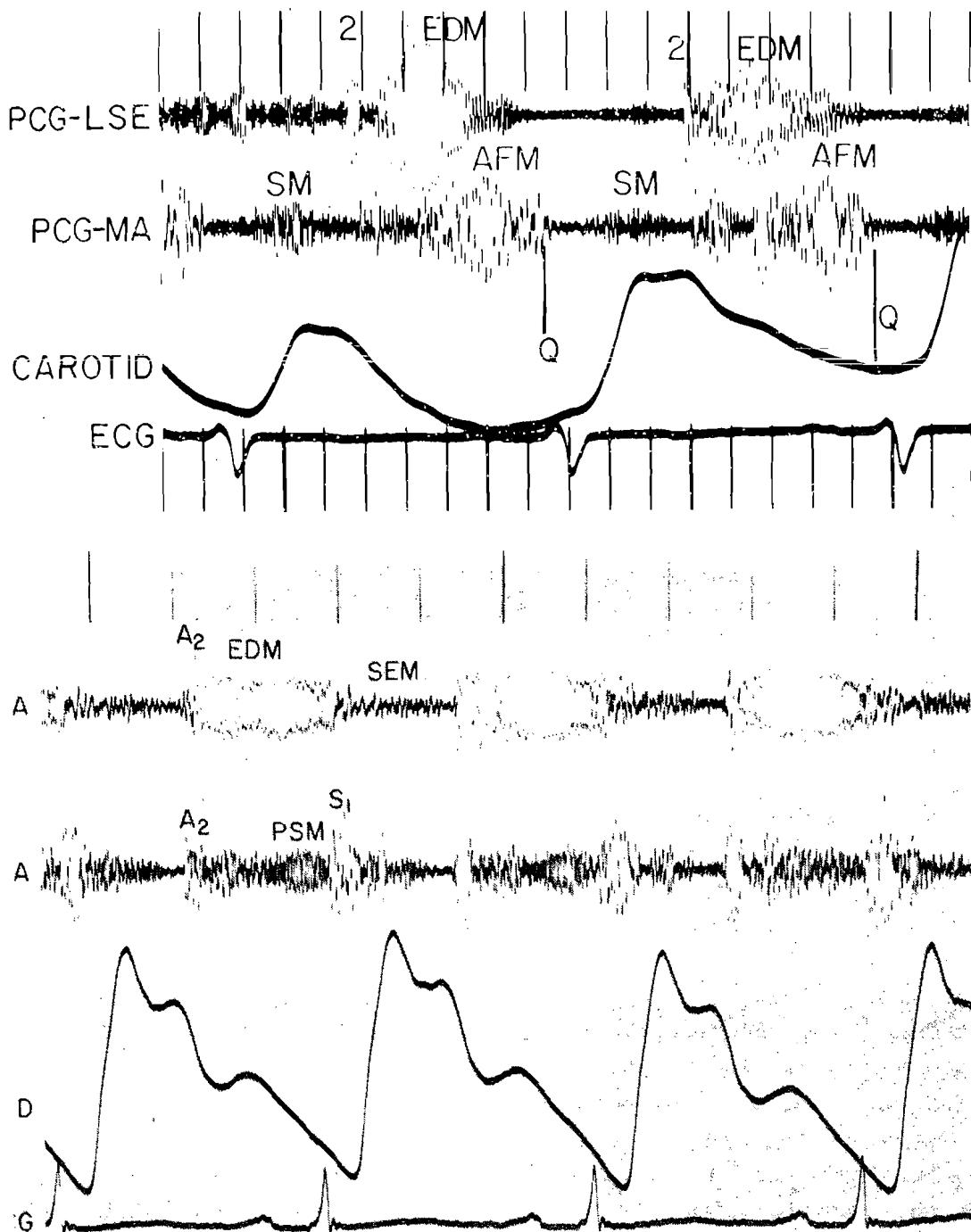


FIG. 15-7. The Austin Flint murmur (AFM) in severe aortic regurgitation. These phonocardiograms from two different patients demonstrate the two different types of diastolic murmur found in aortic regurgitation: the typical decrescendo murmur directly produced by the aortic reflux and the low-pitched apical Austin Flint murmur. A. A loud Austin Flint murmur (AFM) is recorded at the apex in a patient with severe aortic regurgitation. Note the difference in timing from the early diastolic murmur (EDM). The AFM occurs relatively late in diastole; there is no presystolic accentuation. LSE = left sternal border; MA = mitral area; SM = systolic murmur. (From Craige E and Millward DK: Diastolic and continuous murmurs. Prog Cardiovasc Dis 14:38, 1971.) B. Moderately severe aortic regurgitation of recent onset resulting in an early diastolic murmur (EDM) and a presystolic Austin Flint murmur at the apex (PSM). The latter

(Legend Continues on Facing Page)

Associated Valve Lesions. Patients can have both aortic stenosis and regurgitation, particularly in chronic rheumatic valvulitis. In these instances, the systolic murmur will also reflect the presence of aortic valve obstruction. Typically, this results in a prominent systolic *and* diastolic murmur, the so-called bellows murmur. When both the systolic and diastolic murmurs are long, this combination may mimic a continuous murmur along the left sternal border. If there is little evidence for severe aortic regurgitation on physical examination, aortic stenosis is likely to be the dominant lesion. The quality of the carotid pulsations and precordial impulse will be a hybrid of the findings of pure aortic valve obstruction and pure insufficiency.

Patients with severe aortic regurgitation often have long apical systolic murmurs resembling the murmur of mitral regurgitation. As mentioned, the systolic flow murmur in patients with a large stroke volume may be quite long and radiate well to the apex, mimicking mitral regurgitation. Nevertheless, functional mitral regurgitation may occur in some patients with a very dilated left ventricular chamber, possibly in part due to distortion of the papillary muscle relationships (see Chapter 17; Fig. 17-9)). Angiographic studies have failed to confirm a high incidence of mitral regurgitation in such patients.

When mitral stenosis coexists with AR, the obstruction to inflow to left ventricular filling may attenuate the full expression of the aortic regurgitation on clinical examination. The diastolic murmur of AR may be less prominent, and evidence for LV dilatation will be less impressive than that usually produced by a comparable degree of aortic regurgitation.

Austin Flint Murmur. The Austin Flint murmur (AFM), a well-known auscultatory phenomenon, is a low-pitched, rumbling apical diastolic murmur that sounds exactly like the murmur of mitral stenosis (Figs. 15-4, 15-7, 15-8). The mechanism for the genesis of the AFM has been the focus of many investigators. Its presence generally indicates a large diastolic leak with a regurgitant fraction of over 50%. The AFM is usually found in association with the peripheral signs of severe AR (Table 15-1). It is important to correctly identify the AFM for the following reasons: (1) its presence indicates that the aortic reflux is severe, and (2) accurate identification of the AFM on physical examination usually means that associated mitral stenosis is not present.

Pathogenesis. Many theories have been advanced to explain the Austin Flint murmur. The mechanism appears to be related to an incomplete opening of the anterior leaflet of the mitral valve during diastole as a result of the impact of the regurgitant stream of blood into the left ventricular cavity. The

has low frequency vibrations and terminates with a soft S1. Note that the loudest vibrations of this murmur follow atrial contraction. The EDM is unusually loud and high pitched. (From Craige E: The Austin Flint murmur. In Physiologic Principles of Heart Sounds and Murmurs. Edited by DF Leon and JA Shaver. American Heart Association Monograph, No. 46, 1975.)

**The Austin Flint Murmur vs
the Diastolic Rumble of Mitral Stenosis**

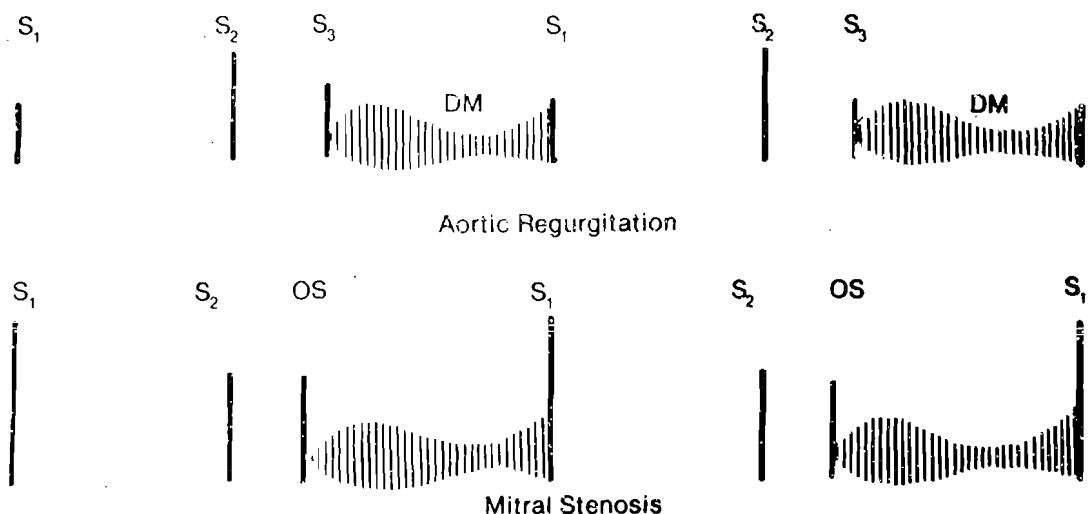


FIG. 15-8. Similarity between the Austin Flint murmur of aortic regurgitation and the diastolic rumble of mitral stenosis. Both diastolic murmurs (DM) are low frequency and sound similar to each other. They are best heard using the bell of the stethoscope with light pressure at the left ventricular apex with the patient in left lateral position (see Fig. 7-9). The Austin Flint murmur is usually accompanied by a soft S₁ and prominent S₃. The latter may be palpable, as may be presystolic distention of the ventricle (S₄). The rumbling murmur of mitral stenosis is classically associated with an accentuated S₁ and a high frequency opening snap (OS), which occurs closer to S₂ than does the S₃. In spite of these associated points, it may be exceedingly difficult to differentiate one murmur from the other. (From Abrams J: Prim Cardiol, 1983.)

mitral valve closing motion in mid-diastole is more rapid than usual in patients with an AFM. The first vibrations of the Austin Flint murmur occur while the mitral valve leaflets are rapidly moving towards the left atrium as antegrade blood from the atrium transverses the narrowing but still open mitral valve orifice. The resultant low-pitched murmur may persist into late diastole as a result of turbulence within the left ventricular cavity. A late, presystolic murmur occasionally is part of the Austin Flint complex (Fig. 15-7B) and may actually be the dominant diastolic component (Fig. 15-4). Its timing is related to the closing motion of the mitral valve leaflets that immediately follows left atrial contraction. Antegrade blood flow across the mitral valve produces turbulent eddies within the left ventricle as the mitral valve orifice is narrowing in late diastole. The presystolic component of the AFM is often not present when there is severe aortic reflux resulting in a major rise in late LV filling pressure that literally "forces" the mitral valve cusps to prematurely slam shut. This *premature closure of the mitral valve* in mid-late diastole prevents left atrial contraction from reopening the valve, and therefore, the presystolic component of the AFM is absent.

Typically, the AFM is found in patients with a markedly increased LV end-diastolic volume and a very large aortic regurgitant fraction. On occasion, patients with a noncompliant left ventricle without excessive ventricular dilatation may have an AFM. These subjects have a very high LVEDP.

Other postulated mechanisms for the AFM include "relative or functional mitral stenosis" (small left atrium, large left ventricle), diastolic mitral regurgitation, fluttering or vibration of the anterior leaflet of the mitral valve resulting from the aortic diastolic jet, and selective transmission of low to medium frequency vibrations of the aortic regurgitation murmur to the apex. None of these phenomena adequately explain the AFM.

Severity of AR and AFM. In mild AR with a small regurgitation volume, LVEDP is low and there is no AFM. In moderate degrees of reflux an AFM may occur; it is more likely to be heard solely in late diastole as a result of an increased LV end diastolic volume in a noncompliant chamber. With greater degrees of AR the AFM begins earlier in diastole, coincident with LV rapid filling; this murmur is mid-diastolic in timing (Fig. 15-7A). A presystolic component may also be heard (Fig. 15-7B). In very severe aortic regurgitation, the mitral valve closes prematurely due to an extremely high, left ventricular filling pressure, and the presystolic component disappears leaving only a mid diastolic murmur.

Auscultatory Features. The Austin-Flint bruit is a rumbling diastolic murmur that usually begins in early to mid-diastole (Figs. 15-4, 15-7, 15-8). If a presystolic component is present, one will hear a crescendo low frequency murmur in late diastole that extends to the S1 (Fig. 15-7B). The pitch of the AFM is identical to the rumble of mitral stenosis. Typically, the murmur begins with a prominent S3, and in many instances the S3 and the AFM fuse to produce an explosive initiation of a mid-diastolic rumble (Fig. 15-4).

Although the AFM is typically low frequency, in some patients it has a higher pitch and may even simulate a transmitted murmur of AR. Careful attention to timing should prevent this misinterpretation. The AFM may be quite loud, radiating to the lower left sternal border and axilla, but it rarely is accompanied by a thrill. In most instances, the murmur is well localized to the left ventricular apex or immediately medial to it and is best heard using the bell of the stethoscope while the patient is in the left decubitus position. This technique is identical to that used when listening to the mitral stenosis rumble (Fig. 16-3).

Because the AFM is directly related to the degree of aortic regurgitation, maneuvers that increase diastolic reflux will augment the murmur, and those that decrease the degree of aortic regurgitation will soften it. Thus, handgrip, mild exercise, application of a bilateral blood pressure cuff, and squatting all intensify the amplitude of the Austin Flint rumble, but amyl nitrite diminishes the murmur. The latter is an important diagnostic clue in differentiating the AFM from the rumble of the mitral stenosis (Fig. 15-9).

Austin Flint or the Diastolic Murmur of Mitral Stenosis? Patients with rheumatic heart disease frequently have combined aortic regurgitation and mitral stenosis. Therefore, it is of great importance to accurately identify the etiology of a low frequency, apical diastolic murmur in any patient with

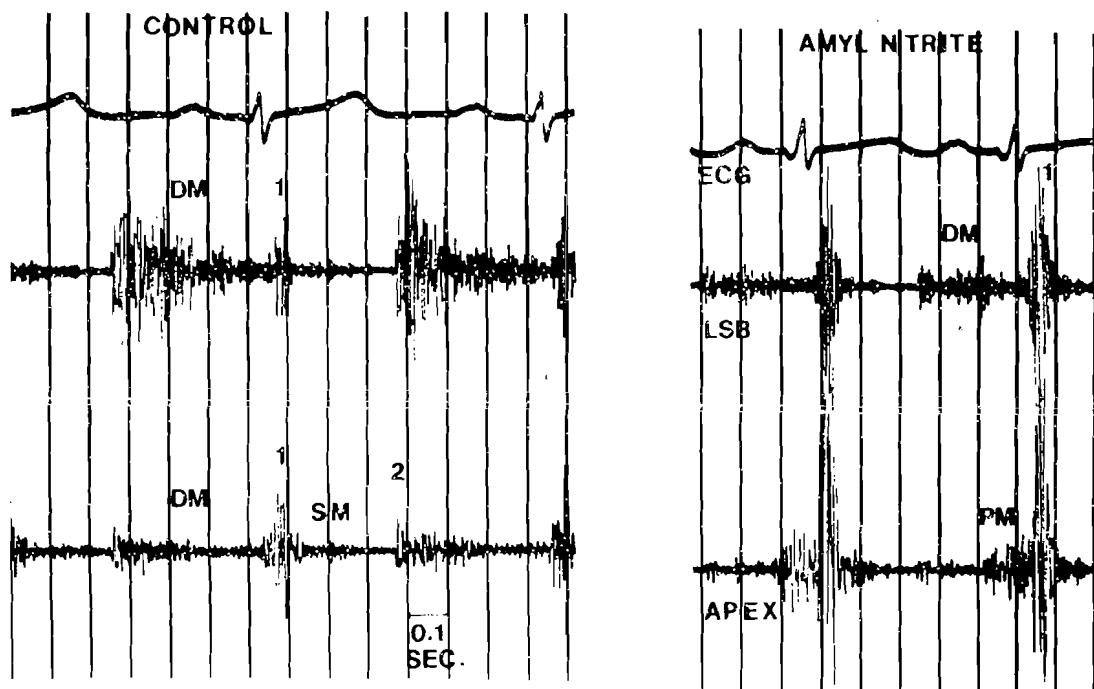


FIG. 15-9. Use of amyl nitrite in differentiation of the murmur of mitral stenosis from an Austin Flint murmur in severe aortic regurgitation. Note that the diastolic (DM) at the left sternal border (LSB) decreases after administration of amyl nitrite compared to control intensity, whereas a presystolic murmur (PM) appears at the apex. The latter finding supports the presence of organic mitral stenosis. If the apical diastolic murmur represented an Austin Flint murmur, it would have diminished in intensity rather than increased during amyl nitrite administration. Conversely, the diastolic murmur at the sternal border decreased in loudness, consistent with aortic regurgitation. This patient had mild mitral stenosis and moderate aortic regurgitation. (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 26:145, 1983.)

evidence of moderate to severe aortic regurgitation (Fig. 15-8). Evidence of associated mitral valve disease implies that the rumble is due to organic mitral stenosis and not secondary to severe aortic incompetence. Table 15-3 lists a number of differentiating features. A case is built for or against associated mitral stenosis through the use of all of the clinical information at hand. Amyl nitrite can be employed to resolve this question. This potent arterial vasodilator causes a marked decline in systolic blood pressure and systemic vascular resistance; the degree of aortic regurgitation acutely diminishes, and the aortic regurgitation diastolic murmur and the Austin Flint murmur soften (Fig. 15-9). If the low-pitched apical diastolic rumble is caused by mitral stenosis, the administration of amyl nitrite results in an increase in murmur intensity at the apex, as the reflex tachycardia causes an increase in the left atrial-left ventricular gradient. Figure 15-9 demonstrates the simultaneous attenuation of the aortic regurgitation blowing murmur with the appearance of an apical presystolic murmur after amyl nitrite administration in a patient with aortic regurgitation and mild mitral stenosis.

In some patients, an M-mode echo may be required to resolve this dilemma. In the presence of mild mitral stenosis, S1 may be relatively normal.

TABLE 15-3 *Helpful Differentiating Features of an Apical Diastolic Murmur in Aortic Regurgitation: Austin-Flint Murmur Versus Mitral Stenosis*

	Austin-Flint	Mitral Stenosis
Rhythm	Normal sinus	Atrial fibrillation
Left ventricular heave	Common	Absent
Right ventricular lift	Absent	Present
S1	Normal to decreased	Loud
Opening snap	Absent	Present
S3	Present	Absent

Therefore, the clinical impression could favor pure aortic regurgitation with an Austin Flint rumble if an opening snap was not identified. On the other hand, in a patient with pure AR, an aortic ejection sound can be confused with a loud S1, and a prominent S3 can be mistaken for an opening snap. Here the diastolic rumble is readily mistaken for the murmur of mitral stenosis.

The AFM may make auscultation confusing when both a loud systolic and a regurgitant diastolic murmur radiate well to the apex. The presence of three separate murmurs can create a serious auscultatory problem for the examiner; this combination simulates a continuous murmur when heard at the apex or left anterior axillary line. With rapid heart rates, differentiation of the AFM from apical radiation of the AR murmur is difficult. The systolic murmur during tachycardia may appear to be holosystolic, further contributing to diagnostic chaos. The murmur of pure AR itself also can simulate mitral stenosis or an AFM if it transmits well to the apex with selective enhancement of the low frequency vibrations.

Proper identification of an apical AFM in a patient with only a soft AR murmur in the setting of congestive heart failure can be of great value because it directs attention to the real problem, i.e., severe AR as the cause of congestive heart failure.

ACUTE AORTIC REGURGITATION

In recent years, the unusual syndrome of acute massive aortic regurgitation has become increasingly recognized. Often this is a dramatic, life-threatening condition resulting from the sudden influx of an excessive amount of blood into a left ventricle unable to accommodate the large regurgitant volume. In such cases, the aortic valve usually is normal or only mildly abnormal prior to the onset of the sudden regurgitation. Acute or subacute bacterial endocarditis, aortic dissection, aortic valve perforation or rupture secondary to trauma, or myxomatous degeneration are the most common causes of this syndrome.

A prompt and accurate diagnosis of acute AR is of great importance, as urgent surgical intervention usually is mandatory. However, the typical physical signs of severe chronic aortic regurgitation often are absent in such patients, and the aortic regurgitation murmur itself may be quite unimpressive in these severely ill patients. Why is this so? (1) The voluminous regurgitant volume often precipitates acute left ventricular failure, with secondary systemic vasoconstriction and tachycardia. The arterial tree "clamps down," preventing or attenuating the classic blood pressure alterations and peripheral signs of chronic severe aortic regurgitation. (2) Sinus tachycardia is common in acute AR. The increase in heart rate results in a relative shortening of diastole, which hampers accurate auscultation by making identification of systole and diastole difficult. (3) Because the left ventricle is usually of normal thickness and diameter prior to the onset of the acute AR it is nondistensible and unable to dilate. Left ventricular filling pressure becomes markedly elevated, and early to mid LV diastolic pressure rises excessively. This rise results in premature closure of the mitral valve. S1 is soft and there is no S4 or presystolic component to the Austin Flint murmur. In these patients, left ventricular function may often be acutely depressed. Ejection fraction decreases and LV end-diastolic volume increases further due to the reduction of systolic function. The tremendously elevated left ventricular pressure in late diastole may approximate or even surpass aortic diastolic pressure. This causes a shortening of the AR murmur, which is often surprisingly soft and short because of a low cardiac output and decreased late diastolic aortic-LV gradient.

PHYSICAL FINDINGS (Table 15-4)

General Appearance

Patients with acute aortic regurgitation frequently are quite ill and may be in acute heart failure. Resting tachycardia and orthopnea are common. The skin may be pale, cool, and moist, reflecting intense sympathetic vasoconstriction.

Blood Pressure and Pulses

The low diastolic blood pressure common to severe, chronic aortic regurgitation may be absent. The small forward stroke volume and decreased rate of ejection in these patients prevents the expected increase in systolic blood pressure. Thus, the pulse pressure may be normal, slightly increased, or wide. The nonauscultatory signs of severe AR in the peripheral circulation may be unimpressive or totally absent in contradistinction to chronic AR of equivalent degree. A bisferiens pulse is not usually present.

TABLE 15-4 Features of Acute Versus Chronic Severe Aortic Regurgitation

	Chronic	Acute
Resting heart rate	Normal	Sinus tachycardia common; easy to confuse systole for diastole
Blood pressure	Increased systolic BP (> 140 mmHg) Decreased diastolic BP (< 70 mmHg) Increased pulse pressure	Normal or slight reduction in systolic BP. Diastolic BP may or may not be low Pulsus alternans
Peripheral pulses	Bisferiens contour Increased amplitude and volume Peripheral signs of severe AR	Can have unremarkable contour with little to no evidence for peripheral vasodilatation
Jugular venous pulse	Normal	Mean pressure may be elevated. V wave if functional tricuspid regurgitation
Precordial motion	LV impulse at 5/6th ICS, left anterior axillary line—hyperdynamic or heaving contour. Palpable S3 or S4 common.	Normal to slight LV enlargement. Bifid diastolic impulse with palpable S3, sustained late diastolic motion. RV impulse if severe pulmonary hypertension
Heart sounds	S1 normal to decreased S2 often unremarkable; increased to decreased A2 S3 very common S1 uncommon Ejection click possible	S1 decreased to absent S2 single, soft to absent A2, increased P2 S3 “always” No S4 Ejection sound common Mid-diastolic mitral valve closure sound
Aortic regurgitation murmur	Medium frequency Usually holodiastolic May be short, with rapid decrescendo	Medicumb frequently, often harsh Musical if ruptured cusp Usually not holodiastolic, may be very short, rapidly decrescendo
Austin Flint murmur	Grade 3 unless CHF present Common mid-diastolic component with or without presystolic murmur	Can be quite soft “Always” mid-diastolic component with or without presystolic murmur
Systolic murmur	Typically present Can simulate aortic stenosis or mitral regurgitation	Typically present Mitral regurgitation murmur common

Jugular Venous Pulse

Right ventricular failure resulting from pulmonary hypertension is common in severe acute AR. The jugular venous mean pressure may be elevated and tricuspid regurgitation may be present, resulting in large V waves in the neck veins (see Chapter 19).

Precordial Motion

The left ventricular impulse may be unimpressive in recent onset aortic incompetence or may be displaced laterally with a thrusting contour. The markedly enlarged left ventricular impulse typical of chronic aortic regurgitation is absent if there has been no prior cardiac involvement. A bifid *diastolic* apical impulse has been described in such patients. The first component reflects early diastolic filling (palpable S3), whereas the second outward motion reflects late diastolic expansion due to continued diastolic filling of the ventricle. A right ventricular or parasternal impulse may be present if pulmonary artery pressure is elevated because of acute left ventricular failure.

Heart Sounds

S1 is soft or even absent due to the premature closure of the mitral valve (Fig. 15-4). S2 may be normal or single. Either or both an S3 and S4 are commonly noted.

Aortic Regurgitation Murmur

The murmur of acute AR may be unimpressive and clinically misleading. If congestive heart failure or severe depression of LV function is present, the diastolic AR murmur may be very soft. Frequently, severe aortic regurgitation produces a murmur with medium to low frequency vibrations, and the length of the murmur may be surprisingly short. If the acute AR is related to preexisting aortic root disease or isolated rupture of the right coronary cusp, the diastolic murmur may radiate best down the right sternal edge (see page 262).

Austin Flint Murmur

The AFM is commonly found in acute aortic regurgitation where it is likely to be mid-diastolic with or without a presystolic component (Fig. 15-4). In some patients, early closure of the mitral valve in mid-diastole occurs as left ventricular pressure rapidly rises and precludes antegrade flow across the mitral valve in late diastole. It has been suggested that this early closure of the mitral valve "protects" the left atrium and pulmonary capillary bed

from the marked elevation of left ventricular end-diastolic pressure, which may exceed 40 to 50 mmHg.

Systolic Murmur

The systolic flow murmur commonly heard in severe chronic AR may also be present in acute AR. If left ventricular function is depressed, this murmur is likely to be soft and short. Frequently, a long systolic murmur suggestive of mitral regurgitation is heard at the apex. Sinus tachycardia, commonly present, may cause an apparent "lengthening" of the systolic and diastolic murmurs, which may be difficult to time. A loud S3 may be mistaken for S1 or S2 if systole and diastole are confused with each other; careful palpation of the carotid arteries with simultaneous auscultation should resolve this dilemma. In any patient with aortic regurgitation, slowing of the heart rate (e.g., carotid sinus pressure) will aid accurate auscultation.

DIFFERENTIAL DIAGNOSIS

Pulmonary Regurgitation. In severe pulmonary hypertension, a high pitched blowing murmur is common (the Graham Steell murmur). Its acoustic characteristics are identical to those of mild aortic regurgitation. The Graham Steell murmur is invariably associated with other signs of pulmonary hypertension, such as a right ventricular lift and increased P2. Often, there is an obvious cause for the pulmonary hypertension (e.g., severe mitral stenosis). Integration of data from the cardiac examination, chest roentgenogram, and ECG usually resolves this issue. One recent angiographic study of patients with mitral stenosis and a blowing diastolic murmur along the left sternal border and coexisting pulmonary hypertension documented that most of these murmurs were associated with aortic regurgitation. On the other hand, the apparent association of an AR murmur in patients undergoing renal dialysis may be incorrect, as a recent study suggests that these murmurs are due to pulmonic regurgitation.

Congenital pulmonic regurgitation is rare. This murmur is low to medium pitched, and usually begins after an audible pause or gap after P2. It is associated with low pulmonary and right ventricular pressures and does not mimic the high frequency AR "blow" or a Graham Steell murmur. As a rule, isolated, congenital pulmonic regurgitation should not provide confusion with aortic regurgitation.

Mitral Stenosis. Occasionally, the diastolic murmur of mitral stenosis is medium to high frequency and may simulate aortic regurgitation if it radiates well to the lower, left sternal border. Careful auscultation should identify a clear-cut interval between A2 and the onset of the mitral stenosis

murmur; an opening snap and an increased S1 typically will be present. If the opening snap is close to A2 and the mitral stenosis murmur long and loud, one can be readily misled as to the murmur origin. Tachycardia accentuates this confusion.

Other Conditions. In children, a prominent venous hum can be confused with AR. Appropriate positioning of the head and neck with careful compression of the jugular veins should clarify the situation (see Chapter 4). Rarely, a proximal obstruction of the left anterior, descending coronary artery produces a diastolic murmur over the midprecordium. Other continuous murmurs (e.g., patent ductus arteriosus, AV fistula, coronary sinus aneurysm) may mimic aortic regurgitation if the systolic component is not well heard.

Chapter 16

Mitral Stenosis

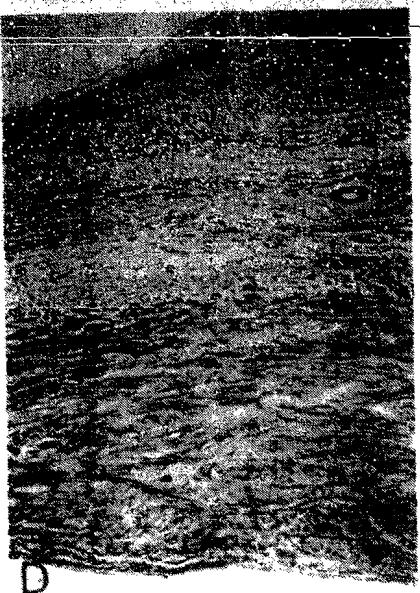
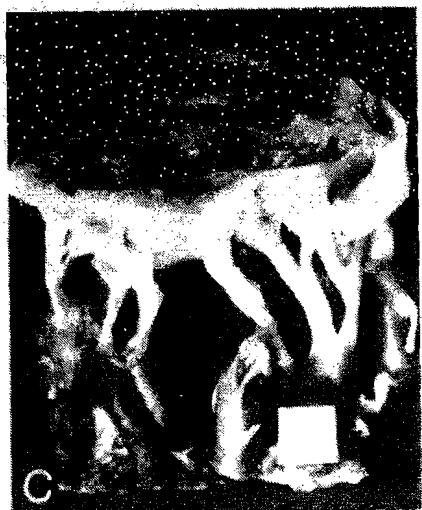
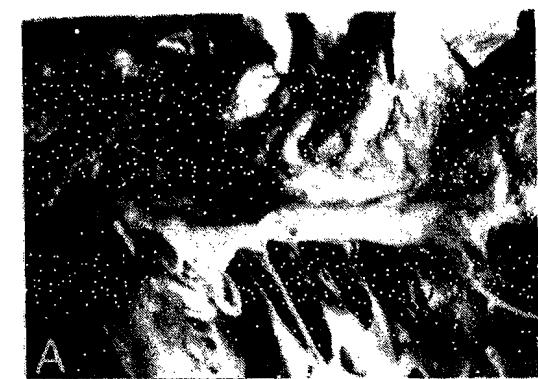
Among major cardiac valve abnormalities mitral stenosis is the only one that has a single etiology—rheumatic fever. A rare congenital form of mitral stenosis has been recognized (parachute mitral valve) in infants. In adults, left atrial myxoma can simulate the findings of mitral stenosis if it produces obstruction to left ventricular inflow. Elderly subjects with a heavily calcified mitral annulus occasionally can have some narrowing of the mitral orifice that can produce abnormal physical findings.

Mitral stenosis is a common sequela of acute rheumatic fever and is found in as many as 40% of individuals with chronic rheumatic heart disease. Only half of these patients can recall a prior history of an illness compatible with acute rheumatic fever. In India, Ceylon, and other developing countries, acute rheumatic fever and mitral stenosis commonly are seen in children and teenagers. In the western hemisphere, clinical rheumatic mitral stenosis usually manifests itself 15 to 30 years after the acute attack of rheumatic fever.

FUNCTIONAL ANATOMY AND PATHOPHYSIOLOGY

Chronic rheumatic valvulitis culminating in mitral stenosis is a result of thickening and distortion of the valve leaflets and chordae tendinae along with fusion and dense adherence of the mitral valve structures (Fig. 16-1). Commonly, the process begins at the mitral valve commissures and may be limited to commissural fusion resulting in a pliable valve that swings freely open although the central orifice is narrowed. Typically, the leaflets are thickened and have blunted and rolled edges (Fig. 16-1A-D). The chordae are foreshortened and often matted together so that the normal spaces between the chordae may be obliterated. The entire valve apparatus becomes altered into a funnel-like sleeve; when severe, the mitral valve takes on a “fish mouth” appearance and loses the ability to open during diastole (Fig. 16-1E). In many instances the stiff, distorted mitral valve complex is both unable to open adequately (stenosis) or close completely (regurgitation). Dense calcification is a frequent late accompaniment that may further impede function of the mitral valve.

Relationship of Physical Findings to Pathologic Anatomy. The thickened and stiff mitral apparatus produces prominent sound transients as the mitral leaflets hinge open and then close during the cardiac cycle (Fig. 16-2). The mitral closure sound (S1) is accentuated, and the mitral valve opening



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motion, which is normally silent, results in a loud sound (opening snap) occurring as the fused valve cusps open abruptly into the LV cavity. The large pressure gradient between the left atrium (LA) and left ventricle (LV) contributes to loudness of the S1 opening snap. The valve stenosis delays mitral closure into early LV isovolumic systole as rapidly rising LV pressure helps "slam" the valve shut, accentuating S1. The opening snap (OS) follows LA-LV crossover in early diastole; the abnormally high LA pressure causes the mitral valve opening motion to begin earlier than normal.

The dominant murmur of mitral stenosis consists of an early diastolic murmur. In addition, there may be a late diastolic or presystolic murmur that occurs during left atrial contraction and results in a late diastolic increase in atrial pressure and an accentuation of the end-diastolic LA-LV pressure gradient.

CLINICAL SPECTRUM

The normal mitral valve area is approximately 5 cm^2 (range 4 to 6 cm^2). Although physical signs may be detectable in very mild mitral stenosis (opening snap, short diastolic mumble) symptoms do not usually appear until the mitral valve area has been reduced to at least 50% of normal (e.g., to 2 to 2.5 cm^2). When the mitral valve area is reduced to 1.5 to 2.0 cm^2 , the mitral stenosis is mild to moderate and symptoms of fatigue and dyspnea are common.

Major hemodynamic sequelae occur when the valve area is reduced to 1.5 cm^2 or less; resting cardiac output is depressed and LA pressure is elevated. Often, relatively little effort may cause severe symptomatic limitation. When the mitral valve area is reduced to 1.0 cm^2 or less, severe or "tight" mitral stenosis is present. In order to maintain adequate flow at rest, the LA pressure must be elevated to 20 to 25 mmHg or greater. In some individuals with massive LA dilatation, LA pressure may be surprisingly low for any given severity of mitral obstruction because of the voluminous and distensible atrial chamber. In patients with a less compliant left atrium, the high pulmonary venous pressures result in marked shortness of breath and orthopnea.

FIG. 16-1. Upper. Anatomy of mitral stenosis. A. Effacement of scallops of the posterior leaflet due to fusion of leaflet clefts. B,C. Two views of a stenosed mitral valve removed at surgery. Note the commissural and cleft fusion in leaflets (B) and chordal changes (C). D. Photomicrograph of mitral leaflet demonstrating sclerosis, vascularization, and nonspecific chronic inflammation. (From Silver MD: Cardiovascular Pathology, Vol. 1. New York, Churchill Livingstone, 1983.) E. Atrial view of a crescent-shaped stenotic orifice. Note the loss of individual scallop identity of the posterior leaflet. The anterior leaflet usually retains its shape and mobility until late in the disease process. This valve is transilluminated from the ventricular side; note the relative transparency of the anterior leaflet. (From Becker AE and Anderson RW: Cardiac Pathology. New York, Raven Press, 1983.)

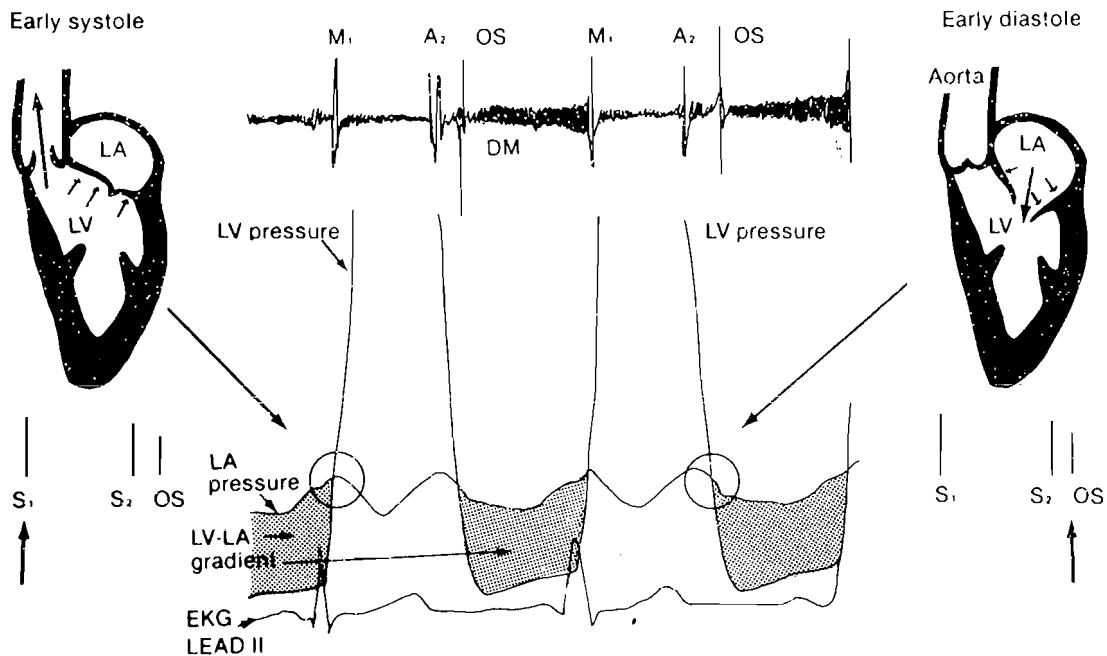


FIG. 16-2. Intracardiac pressure and sound relationships in mitral stenosis. Pressure crossover between the left atrium (LA) and the left ventricle (LV) always precedes the cardiac sounds generated by mitral valve closure (M1) and opening (opening snap). The persistence of a late diastolic gradient between the left atrium and left ventricle in combination with the thickened mitral valve apparatus results in an accentuated S1. Similarly, the maximal opening excursion of the rigid and fibrotic valve generates an opening snap (OS), which immediately precedes early diastolic filling of the left ventricle and the resultant diastolic murmur (DM). (From Abrams J: Prim Cardiol, 1983).

IMPORTANT VARIABLES AFFECTING THE COURSE OF MITRAL STENOSIS

Response of the Left Atrium. A thickened and relatively small, non-compliant left atrium results in severe elevation of LA pressure and “congestive” symptoms, whereas a dilated and compliant LA chamber may decompress LA pressure. When atrial fibrillation occurs, either paroxysmal or sustained, clinical decompensation often follows.

Pulmonary Hypertension. The elevation of LA pressure in mitral stenosis causes a passive rise in pulmonary artery pressure that is directly related to the level of mean atrial pressure. In occasional patients, there may be an additional pulmonary vasoconstrictive component causing a major increase in pulmonary arteriolar resistance and pulmonary artery pressure that may be striking during exercise. This will result in right ventricular hypertrophy and dilatation and, possibly, right ventricular failure. In such subjects the physical findings of pulmonary hypertension, right ventricular hypertrophy and tricuspid regurgitation may dominate or even mask the clinical evidence for mitral stenosis. The findings of overt right ventricular failure (elevated

venous pressure, peripheral edema, and possible ascites) are only seen in individuals with mitral stenosis and severe pulmonary hypertension (unless coexistent organic tricuspid valve disease is present).

PHYSICAL EXAMINATION

General Appearance

There are no distinguishing features in most patients with mitral stenosis. The majority of subjects are women. In individuals with severe mitral stenosis, especially with major pulmonary hypertension, "mitral facies" may be observed, i.e., a patchy, pinkish-purple appearance of the cheeks resulting from dilated venules. Such subjects often manifest peripheral cyanosis as well. Patients with advanced mitral valve disease and right ventricular failure typically are thin and often have acrocyanosis and peripheral edema. Patients with severe mitral stenosis may be tachypneic and/or orthopneic at rest. The lung fields may be surprisingly clear unless the patient is in overt pulmonary edema.

Jugular Venous Pulse

The venous pulse is normal in mitral stenosis unless there is associated atrial fibrillation, pulmonary hypertension, or right ventricular (RV) failure. Patients in sinus rhythm with elevated pulmonary artery pressure may show a prominent A wave in the venous pulse as a result of right ventricular hypertrophy and decreased RV compliance. Once right ventricular compensation ensues, the mean venous pressure will be elevated. Tricuspid regurgitation, a common complication, will result in a large systolic CV wave (see Chapter 19). In the presence of atrial fibrillation, the attenuated X descent and the venous pulsations, consisting of irregular V waves, may simulate tricuspid regurgitation.

Carotid and Arterial Pulses

The arterial pulse in mitral stenosis has a normal or decreased pulse volume and a normal contour. If the cardiac stroke volume is decreased, the carotid impulse will be diminutive, and the finding of a small arterial pulse is common in patients with hemodynamically significant mitral stenosis. The presence of associated mitral regurgitation or aortic regurgitation increases the carotid pulse amplitude and rate of rise. In atrial fibrillation, the arterial pulse will be irregular with a variable pulse volume.

Precordial Examination

Considerable clinical information can be derived from precordial examination in mitral stenosis, and often the diagnosis is suspected before auscultation. This is possible because of the loud sound transients (S1, P2, and opening snap), which may be identified by palpation. Typically, S1 is felt at or inside the site of the LV apex beat, which itself usually is very small or impalpable. An increased P2 and, less commonly, a pulmonary artery lift may be felt at the 2nd to 3rd left interspace. The opening snap often is palpable in the region between the lower left sternal border and the cardiac apex. Careful timing of these palpable shocks is essential. The carotid and apical impulses are essential to aid in the identification of systole and diastole. A diastolic thrill at the apex, produced by a very loud mitral rumble, occasionally may be detected in the left decubitus position.

Left Ventricular Impulse. The left ventricle in pure mitral stenosis typically is underfilled and, therefore, does not produce a forceful apical impulse. It is essential to locate the LV apex beat to assist in optimal auscultation of the mitral diastolic rumble. This is best done in the left recumbent position (Figs. 16-3A, 5-7). The PMI is normal or of decreased amplitude, nonsustained, and never associated with a palpable S4 or S3 unless an additional valve abnormality is present, such as mitral regurgitation or aortic stenosis.

Right Ventricular Impulse. It is common to detect parasternal activity in pure mitral stenosis. Often this is manifest as a gentle, low amplitude RV lift detectable at the 3rd to 5th left interspace adjacent to the sternum. *Practical Point:* To detect parasternal motion, it is desirable to use firm pressure with the heel of the hand during held-expiration (see Figs. 16-3B, 5-9). A right ventricular impulse will often be detected in mitral stenosis even when resting pulmonary pressure is high normal or only moderately elevated. In individuals with more severe pulmonary hypertension, the RV impulse can be very prominent; careful visual inspection may detect lateral retraction of the chest wall between the normal right and left ventricular areas. The parasternal heave or major RV enlargement often extends leftward, resulting in the entire central precordium being palpable in a rocking motion. In severe degrees of RV enlargement, the apex beat may be formed by the right ventricle. Consideration of this possibility is important. The typical rumble of mitral stenosis may not be heard unless auscultation occurs when one listens precisely over the LV impulse, which may be displaced laterally in the anterior or midaxillary line by a huge RV.

When pulmonary hypertension is severe, a right ventricular A wave (S4) may be palpable, (even less often an RV filling wave or S3 may be detected). It is useful to employ gentle, but firm, subxiphoid palpation during held inspiration to detect these palpable diastolic events (Fig. 5-10). Commonly, an accentuated pulmonic second sound will be palpable in the pulmonic area;

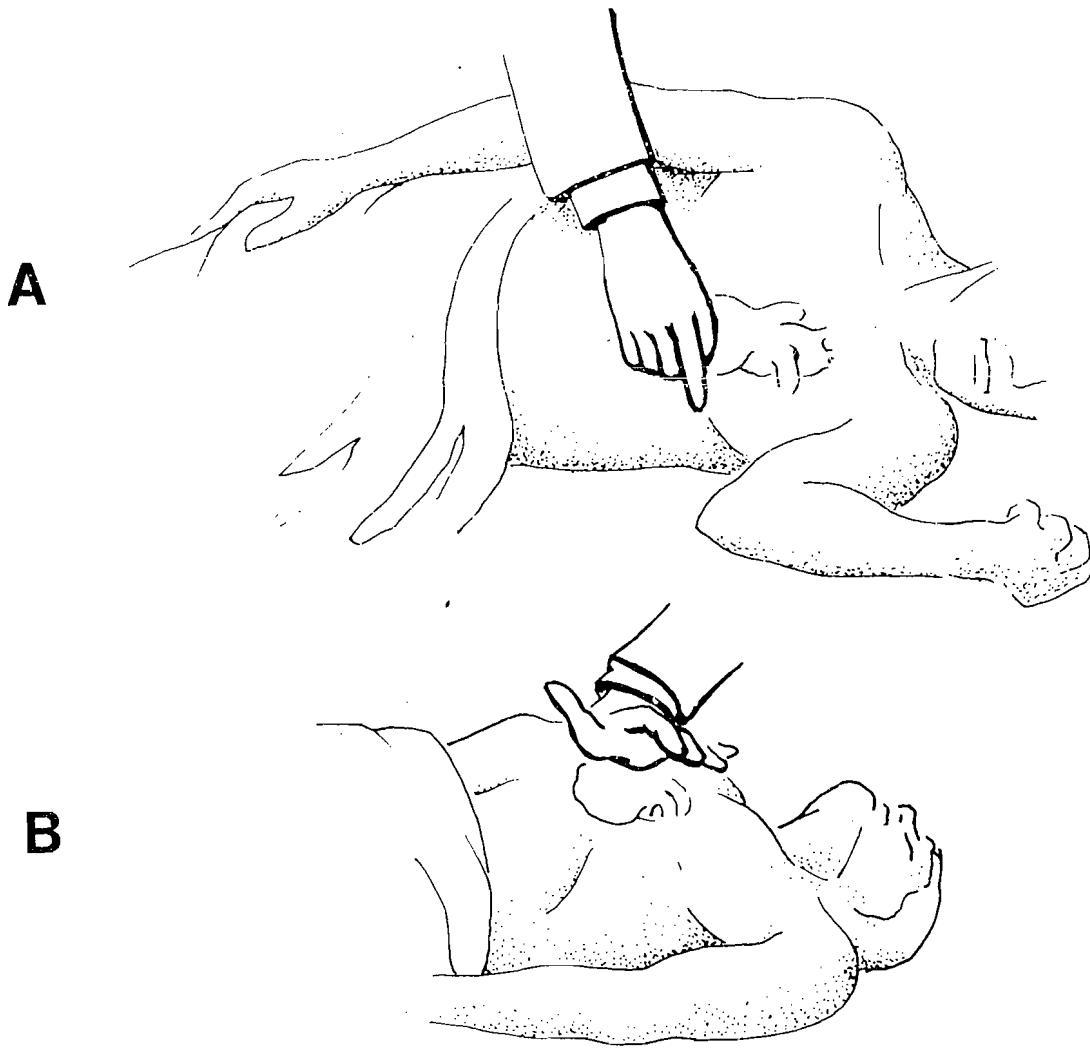


FIG. 16-3. Precordial examination in mitral stenosis. A. Use of the left lateral decubitus recumbent position is mandatory for optimal detection of the murmur of mitral stenosis. The apex impulse is first identified, and one finger positioned at this site. The bell of the stethoscope is placed at the apex, using the lightest pressure necessary to make a skin seal. Frequently the diastolic murmur is audible only over a small precordial area and radiates poorly (see Fig. 16-12). B. Right ventricular activity is best felt with the patient supine and the breath held in mid or end expiration. Firm pressure with the heel of the hand should be used over the third to fifth interspace at the lower left sternal border. A parasternal lift indicates increased right ventricular hypertrophy or dilatation, possibly augmented by the enlarged left atrium displacing the right ventricle in an anterior direction.

its presence in conjunction with a prominent RV lift confirms the diagnosis of pulmonary hypertension. Rarely, a loud pulmonic ejection click will be felt, and the pulmonary artery itself also may be palpable.

On occasion, the physical findings in the patient with severe mitral stenosis are those of isolated right heart disease. Evidence for pulmonary hypertension dominates the clinical picture, and mitral valve obstruction may be difficult to diagnose on physical examination. *Practical Point: In adults, severe pulmonary hypertension is always an acquired phenomena. Mitral stenosis should*

be considered in such cases, although the classic physical findings may not be present.

Heart Sounds

A loud first heart sound and an opening snap are consistently observed in mitral stenosis. An accentuated P2 suggests associated pulmonary hypertension.

First Heart Sound. A loud, snapping S1 is one of the hallmarks of mitral stenosis. Whenever S1 is unusually prominent on cardiac examination, evidence of mitral valve disease should be carefully examined. The loud S1 results from several factors:

(1) Mitral valve closure normally occurs following left atrial systole *prior* to the onset of LV contraction. S1 occurs simultaneously with the maximal bulging of the mitral leaflets into the left atrium and takes place after initial coaptation of the leaflets. In mitral stenosis, mitral valve closure is late due to the elevated LA pressure. LA-LV pressure crossover actually occurs after LV pressure has begun to rise (Fig. 16-2), and closure of the mitral valve is delayed into early systole. The rapidly increasing LV pressure in early systole contributes to the closing motion of the already narrowing mitral valve.

In order to produce a loud S1, the mitral leaflets must be sufficiently pliable to close rapidly. Extensive fibrosis and calcification of the cusps may result in a decreased S1 amplitude.

(2) The persistent LA-LV gradient in late diastole keeps the mitral valve leaflets open deep in the LV cavity when, normally, they would be closing immediately following LA contraction. The increased distance the cusps must travel during their closing motion contributes to the increased amplitude of S1.

(3) The actual quality of the valve tissue in mitral stenosis may affect the amplitude of both the closing and opening sounds. The leaflets and chordae are thickened and less compliant than normal. Thus, the entire mitral valve apparatus may resonate with a higher amplitude than normal valve tissue.

Amplitude of the S1 and Opening Snap. There is a direct relationship between audibility and intensity of the first heart sound and opening snap. A loud S1 and loud OS can occur only when the mitral leaflets have sufficient mobility and pliability to move rapidly into an open or closed position. It is believed that the more capacious and mobile anterior (septal) cusp is responsible for most of the intensity of S1 and opening snap. When there is calcification, fibrosis, or distortion sufficient to impede leaflet motion, the velocity of leaflet excursion is decreased, and the amplitude of S1 and the opening snap are diminished accordingly (Table 16-1). When cardiac output is reduced, as in congestive heart failure, severe pulmonary hypertension,

TABLE 16-1 *Conditions Frequently Associated with a Decreased Loudness of the Opening Snap and/or the First Heart Sound*

Severe pulmonary hypertension
Extensive mitral valve calcification (especially anterior leaflet)
Congestive heart failure or very low cardiac output state
Large right ventricle
Very mild mitral stenosis
Mixed mitral valve disease, with dominant mitral regurgitation
Aortic stenosis (decreased left ventricular compliance)
Aortic regurgitation

associated aortic valve disease, or left ventricular dysfunction, both S1 and the opening snap will have a decreased amplitude.

Characteristics of S1. The first heart sound is typically discrete and loud and has a slapping quality (Fig. 16-4). There may be a series of vibrations that can simulate a brief presystolic or early systolic murmur. The increased S1 is audible throughout the precordium and is heard maximally between the lower sternal edge and the apex. The loud S1 is often palpable. It is important to recognize the palpable shock of S1 and differentiate it from the left ventricular apex impulse, which usually is feeble in mitral stenosis. S1 is often well heard at the base and may be equal to or louder than S2 at the aortic and pulmonic areas in contradistinction to a normal situation, in which S2 is louder than S1 at the cardiac base.

In subjects with a long PR interval, S1 may be softer than expected, and in atrial fibrillation there will be variation in the intensity of S1. Nevertheless, the amplitude of S1 is increased in atrial fibrillation even after long cycle lengths.

Opening Snap. The opening snap (OS) is one of the classic findings in cardiac physical diagnosis. *Practical Point.* For practical purposes a loud opening snap indicates the diagnosis of mitral stenosis. This sound results from the maximal opening excursion of the mitral valve cusps into the left ventricular cavity in early diastole after LV pressure falls below that in the left atrium. The OS does not represent the onset of mitral valve opening. It follows the LA-LV ventricular pressure crossover by 20 to 40 msec (Fig. 16-2). Flow across the mitral valve into the LV begins just before the opening snap, although peak transmitral flow occurs after the OS. The OS coincides precisely with the maximal opening movement of the anterior leaflet of the mitral valve (Fig. 16-4).

Timing (A2-OS Interval). The simple presence of an OS provides no information as to the severity of the mitral stenosis, nor does its intensity have clinical significance except as a marker of leaflet pliability. However, the timing of the OS with respect to S2 and, specifically, its relationship to the aortic component of S2 (A2) is useful in assessing the severity of the

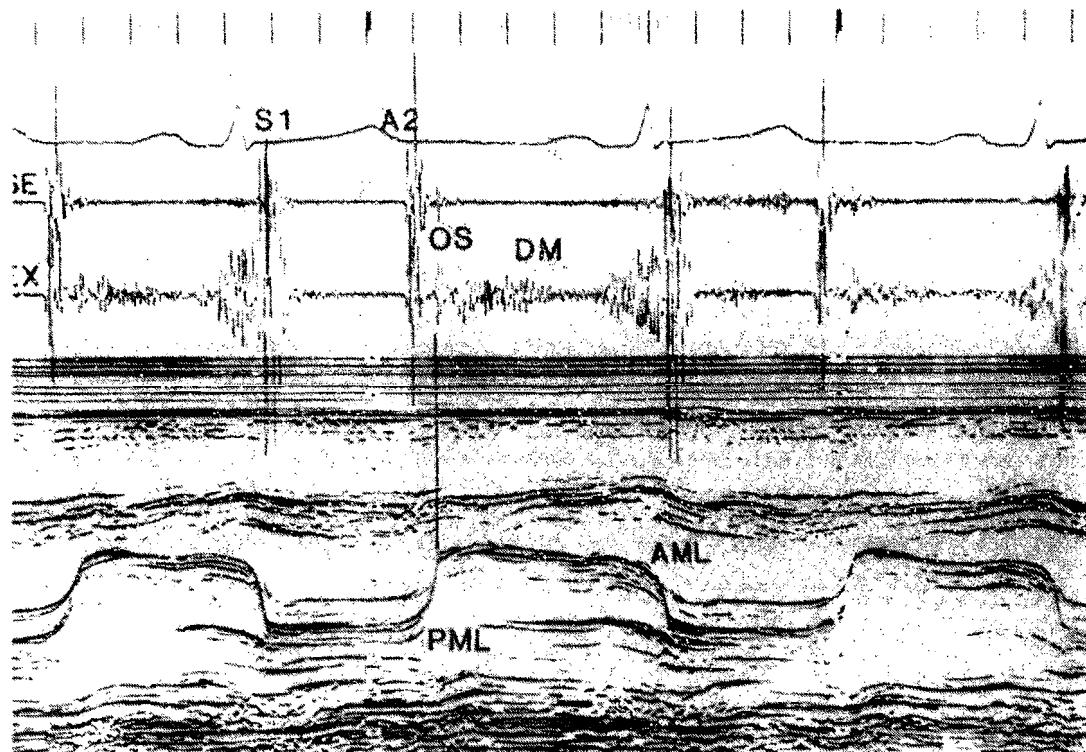


FIG. 16-4. Echocardiographic correlates of the loud first sound and opening snap in mitral stenosis. S1 is produced by mitral valve closure and is accentuated and delayed due to elevation of left atrial pressure and the loss of valve compliance. A prominent presystolic diastolic murmur merges with S1; this represents augmented transmural flow with left atrial contraction. The opening snap (OS) times precisely with the maximum opening excursion of the anterior leaflet of the mitral valve and is produced by tensing of the valve cusps during early diastole. Left ventricular filling and the resultant early to mid-diastolic murmur (DM) follows the OS. (From Reddy PS, Salerni R, and Shaver JA: Normal and abnormal heart sounds in cardiac diagnosis. Part II. Diastolic sounds. *Curr Prog Cardiol* 10:1, 1985.)

mitral obstruction (Fig. 16-5). The A2-OS interval, commonly represented as the II/OS or 2-OS interval, has received an inordinate amount of attention in the literature as an indicator of the severity of mitral stenosis. In general a narrow A2-OS interval indicates severe stenosis (Fig. 16-5A), and a long interval suggests mild disease (Fig. 16-5C). The timing of these two sounds is related to the level of left atrial pressure. With a high LA pressure, as is found in severe mitral stenosis, the mitral valve opens much earlier than normal. In mild to moderate mitral stenosis, LA pressure is less elevated and the LV-LA pressure crossover, as well as mitral valve opening, occurs somewhat later in diastole. In addition, the A2-OS interval is relatively longer (Fig. 16-5A).

The duration of the A2-OS interval in mitral stenosis ranges between 0.04 to 0.14 seconds, usually between 0.06 and 0.11 seconds. A narrow interval, indicative of "tight" mitral stenosis, is less than 0.08 seconds; a long A2-OS interval is over 0.10 seconds. With some practice, the timing of the A2-OS interval can be predicted. For instance, a prominent splitting of S2

during inspiration represents an A2-P2 interval of 0.04 to 0.05 seconds. Constant suggests repeating the words "Pa-Pa" to approximate 0.10 seconds and "Pa-Da" to simulate an interval 0.05 to 0.07 seconds.

Unfortunately, the A2-OS relationship is affected by many factors in addition to left atrial pressure. These include peak systolic pressure, the rate of decline of LV pressure during isovolumic relaxation, and the presence of any condition that independently affects the velocity of mitral valve opening such as aortic regurgitation, decreased LV compliance, and mitral valve calcification. If the left atrial pressure is high because of associated mitral regurgitation, the mitral valve will open earlier for any given degree of mitral obstruction. Table 16-2 lists common associated conditions which, when present, may decrease the predictive value of the A2-OS interval in assessing the severity of mitral stenosis. Note that most of these factors tend to prolong the A2-OS interval. *Practical Point: A short A2-OS interval (less than 0.08 seconds) almost always indicates relatively severe mitral stenosis, although the converse is not true. In some patients, a long A2-OS interval significantly underestimates the severity of mitral stenosis.*

In atrial fibrillation, the A2-OS intervals are directly related to cycle length. During short cardiac cycles, LA pressure remains elevated; thus, A2-OS shortens. With long R-R intervals, LA pressure has more time to decrease, thus prolonging the A2-OS timing. With exercise, the A2-OS intervals shorten; LA pressure rises as the tachycardia decreases the time available for LA decompression. After mild exertion, careful attention to the A2-OS interval may be helpful in assessing the severity of mitral stenosis. In moderately severe disease, this interval should shorten to less than 0.06 seconds.

A large number of indices and formulae have been devised using various heart sound measurements in addition to the A2-OS interval to quantitate mitral stenosis. The Q-S1 interval and apex cardiographic measurements often have been utilized. The Davies or Craige formulae and the Wells index have been widely tested. These techniques require careful and precise graphic recordings and have little to offer the clinician. A careful history, examination,

TABLE 16-2 Factors That May Affect the A2-OS Interval in Mitral Stenosis

<i>Associated Condition</i>	<i>Effect on A2-OS Interval</i>
Systemic hypertension	Increase
Mitral regurgitation	Decrease
Aortic regurgitation	Decrease or increase
Aortic stenosis	Increase
Calcified mitral valve	Increase
LV dysfunction	Increase
Bradycardia	Increase
Tachycardia	Decrease
Old age	Increase

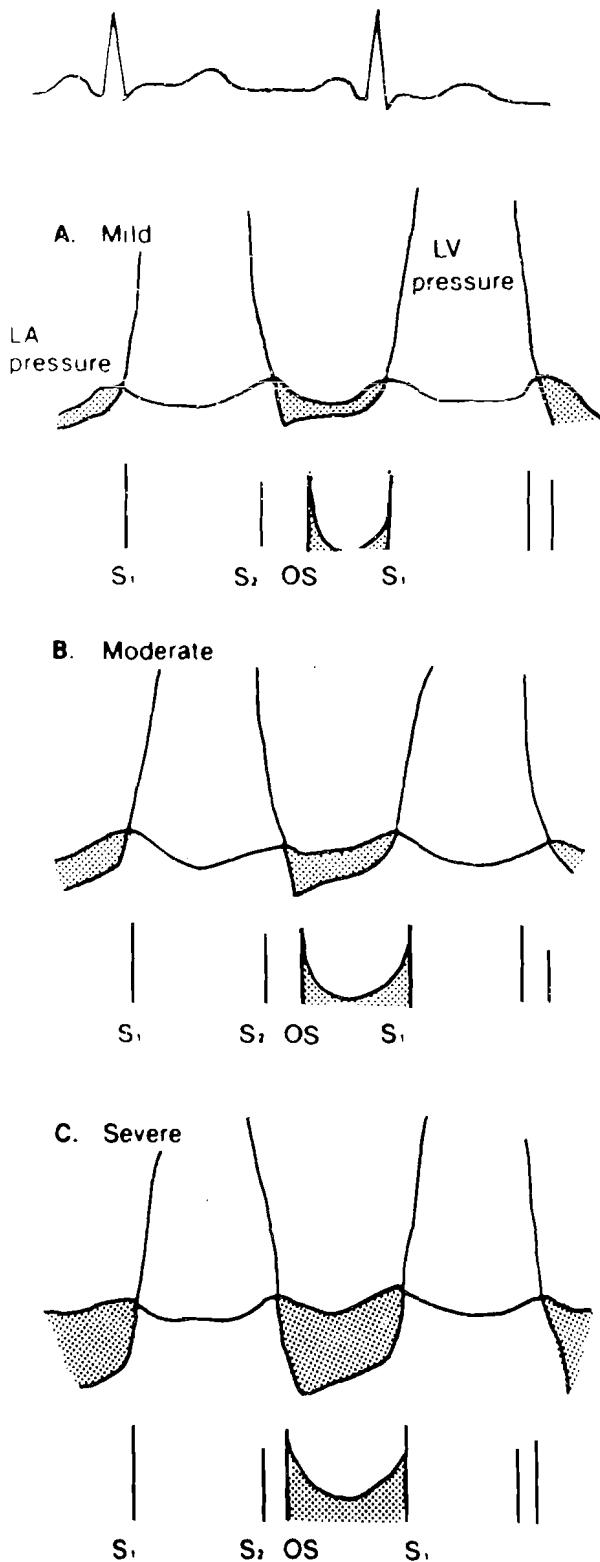


FIG. 16-5. Relationship of severity of mitral stenosis to the timing of the diastolic murmur and opening snap. There are two separate components to the murmur of mitral stenosis. The first occurs in early diastole during the rapid filling phase and immediately follows the opening snap. The second follows left atrial contraction and represents late diastolic augmentation of blood flow across the mitral valve. It is often difficult to distinguish two completely separate murmurs,

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and an echocardiogram will enable the skilled physician to characterize accurately the severity in the large majority of patients with pure mitral stenosis.

Acoustic Characteristics. The opening snap is typically a medium to high frequency sound, quite distinct and sharp, which initiates the mitral diastolic rumble. When the OS is low frequency, it may be easily confused with an S3. In general, the OS has a similar acoustic quality to S1 and S2. In patients with a mobile anterior leaflet and preserved cardiac output, the OS is quite prominent and frequently palpable. The OS is best heard inside or medial to the LV apex in the mid-precordial area. It may be best appreciated at the left sternal border and, occasionally, is maximal at the cardiac apex. In some patients, the OS is better heard in the left decubitus position. When the OS is loud, it tends to radiate widely and will be audible throughout the precordium. Often, the opening snap is extremely well heard at the base and second right interspace (aortic area).

Aids to Detection. The opening snap should be sought by using firm pressure with the diaphragm of the stethoscope in both the supine and left lateral recumbent positions. Mild exercise or handgrip will augment the intensity of the snap. Pharmacologic agents, such as amyl nitrite or pressors, or the Valsalva maneuver should not be necessary to identify the OS under ordinary clinical circumstances.

It may be difficult during auscultation to separate an opening snap from a prominently split S2 (Fig. 16-6), and it is usually hard to clearly distinguish A2, P2, and an OS as three separate sounds during inspiration. Often, the OS is best or only appreciated during expiration. Commonly, S2 becomes blurred or "dirty" during inspiration, but the OS remains crisp. An inspiratory thrill may be heard with the three sounds occurring in rapid sequence. *One must concentrate on detecting all three sound transients by auscultation during many respiratory cycles.* Having the subject sit or stand is useful: The A2-P2 interval will narrow during inspiration in the upright posture, whereas the A2-OS interval will widen as LA pressure falls due to decreased right heart

although clinicians should attempt to detect both an early and late component. The later presystolic murmur usually disappears when atrial fibrillation occurs. A. Mild mitral stenosis. When the mitral obstruction is not severe, a murmur may be heard in early or late diastole or both. There may be a silent gap between the two murmurs in mid to late diastole (see also Fig. 16-4). The opening snap (OS) is relatively late. B. Moderately severe mitral stenosis. There is a pressure gradient across the mitral valve that persists into late diastole. The mitral murmur typically begins with a loud rush, tapers somewhat in mid-diastole and then becomes louder again in late diastole as it crescendoes into the loud S1 (presystolic accentuation). The OS occurs closer to S2. C. Severe mitral stenosis. The large holodiastolic gradient and markedly elevated left atrial pressure produces blood flow across the mitral valve that is accompanied by considerable turbulence. A prominent holodiastolic murmur follows an early OS and extends to S1. There may or may not be presystolic accentuation, depending upon whether the patient remains in sinus rhythm. When the murmur has dominant medium frequency vibrations and is heard throughout diastole, it can mimic the murmur of aortic regurgitation. (From Abrams J: Prim Cardiol, 1983).

DIFFERENTIAL DIAGNOSIS OF THE OPENING SNAP

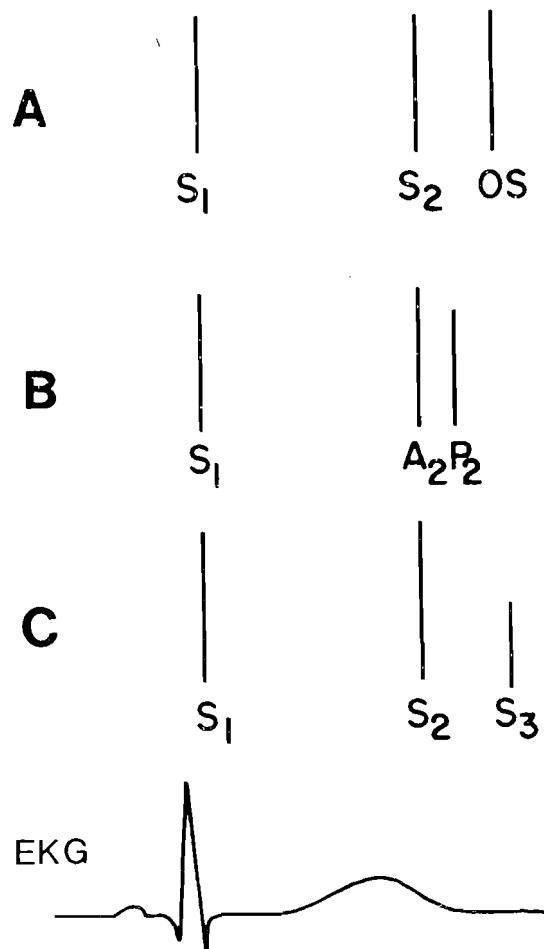


FIG. 16-6. Differential diagnosis of the opening snap (see text).

return, resulting in delayed mitral valve opening. The OS typically moves away from S₂ in the upright position, enhancing its audibility.

A loud opening snap may mask P₂, interfering with proper assessment of S₂ behavior during respiration. If the A₂-OS interval is narrow and the A₂-P₂ interval is wide, considerable difficulty may exist in differentiating the various heart sounds. The presence of an OS may simulate reversed or paradoxical splitting of S₂; in expiration, A₂ and P₂ fuse, leaving a prominently split sound complex (S₂-OS) that can simulate "A₂-P₂." Inspiration may decrease the intensity of the OS but does not change the timing of the A₂-OS interval.

It is useful to employ the inching technique for optimal auscultation and identification of the OS, beginning at the pulmonary area and carefully progressing towards the lower left sternal border and apex. Strict attention must

be given to the timing and intensity of the various heart sounds during the respiratory cycle at each precordial site of auscultation in both the supine and upright position.

Decreased Amplitude. One should be alert to conditions or situations where the OS may be quite soft or even absent in the presence of mitral stenosis (Table 16-1). The same factors that decrease OS intensity tend to decrease the loudness of the mitral diastolic rumble and S1. Most important is dense calcification or extensive fibrosis of the mitral valve apparatus resulting in marked restriction of leaflet mobility.

Low blood flow velocity, common in patients with a markedly decreased cardiac output (e.g., congestive heart failure or severe pulmonary hypertension), will attenuate the opening snap by decreasing the velocity of mitral valve opening as well as transvalvular flow. A very loud P2 may mask detection of a soft opening snap.

Absent Opening Snap. In the presence of dense mitral valve calcification an opening snap may be present in only one third to one half of subjects; the snap may be recorded on phonocardiography and not be audible in some of these individuals. When the right ventricle is dilated, it may occupy the cardiac apex and displace the left ventricle posterolaterally, decreasing the likelihood of detection of the OS.

A recent two dimensional echocardiographic study demonstrates that extensive mitral calcification can be present with a well-preserved opening snap if only the tip, or distal portion, of the anterior leaflet of the valve is heavily calcified with the belly or body of the valve remaining relatively thin. This allows the anterior leaflet to bow forward into the ventricle during its maximal opening excursion. However, when heavy calcification extends into the *body* of the valve leaflet, its mobility is impaired sufficiently to preclude arching of the anterior leaflet into the left ventricle, and the OS is not audible. Table 16-1 lists conditions associated with decreased intensity of the opening snap.

Absence of Mitral Stenosis. It is not commonly appreciated that an opening snap occasionally may be present in the absence of rheumatic mitral valve stenosis, particularly in mitral regurgitation (Table 16-3). The physician should be aware of this possibility, as it may give rise to considerable diagnostic

TABLE 16-3 Cardiac Conditions That May Result in an Opening Snap in the Absence of Mitral Stenosis

Mitral Valve Origin	Tricuspid Valve Origin
Mitral regurgitation	Atrial septal defect
Patent ductus arteriosus	Tricuspid regurgitation
Ventricular septal defect	Ebstein's anomaly
Thyrotoxicosis	Tetralogy of Fallot
	Hypertrophic cardiomyopathy

confusion if not recognized. In such instances, the opening sound is typically soft. It is usually better recorded than heard in such situations (Fig. 16-7).

The sine qua non of the nonstenotic opening snap is high-volume, high-velocity flow across one of the A-V valves. The OS is generated by excessive excursion of a rapidly moving mitral or tricuspid valve during a high flow state. It is believed that the anterior or septal leaflet of the mitral valve is responsible for the production of the opening snap in the setting of pure mitral regurgitation. Both an S3 and an opening snap may be present in some patients with pure mitral regurgitation or in those with mixed mitral stenosis and regurgitation, even with dominant mitral insufficiency.

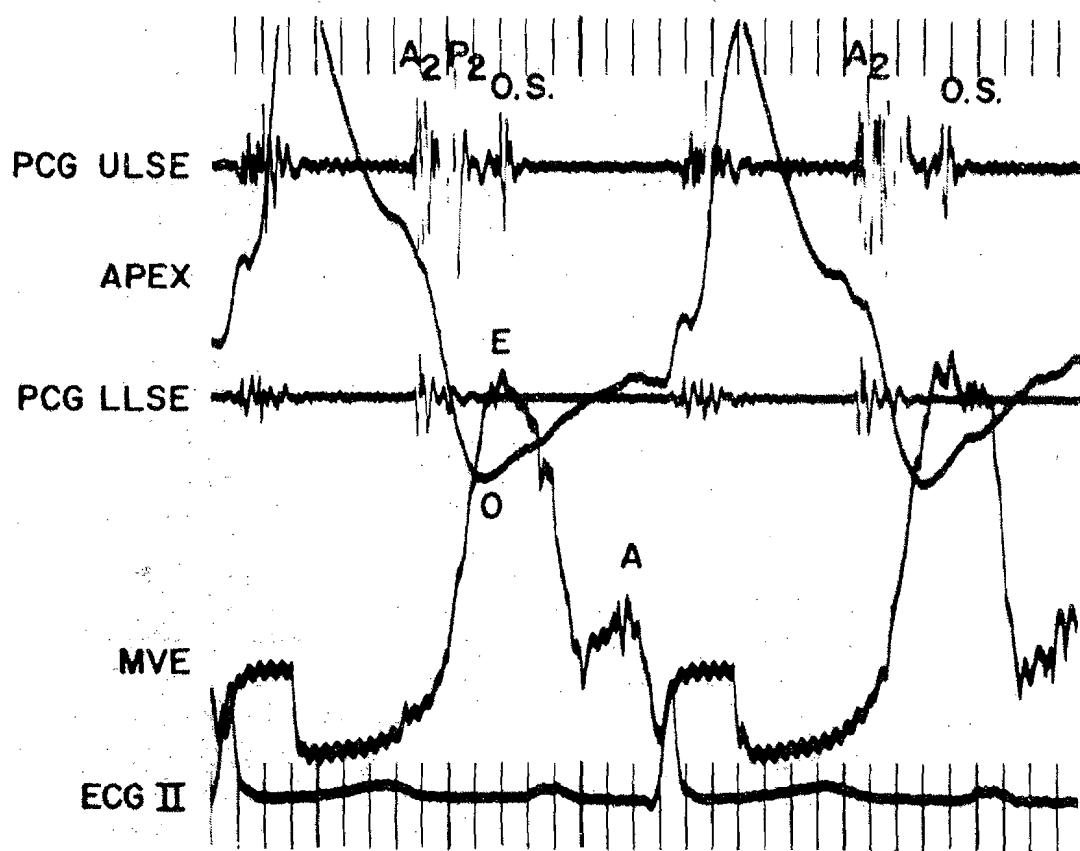


FIG. 16-7. Presence of an opening snap in the absence of mitral stenosis. Diastolic sounds produced by the maximal opening excursion of a normal anterior leaflet of the mitral valve may be heard in high flow states as the excessive blood flow crosses a normal mitral valve. This simultaneous phonoechoes was obtained from a patient with thyrotoxicosis who did not have rheumatic heart disease. Note that the opening snap (OS) is coincident with the maximum opening excursion of the anterior leaflet (E) of the mitral valve and occurs near the O point of the apex cardiogram. A = A wave of the anterior leaflet; MVE = mitral valve echo; ULSE = upper left sternal border. (From Millward DK, McLaurin LP, and Craige E: Echocardiographic studies to explain opening snaps in presence of nonstenotic mitral valves. Am J Cardiol 31:64, 1973.)

Differential Diagnosis (Fig. 16-6). SPLIT S2. A prominently and widely split S2 may simulate an opening snap, and P2 is confused for the OS. A loud P2 can mask the presence of the OS. P2 is best heard at the pulmonic area without radiation unless there is pulmonary hypertension. The opening snap optimally is detected between the left sternal border (fourth interspace), and the apex and, when loud, can be well heard at the base. With inspiration, P2 may increase in intensity; the loudness of the OS may decrease with inspiration. A2-P2 splitting increases with inspiration; the A2-OS interval remains constant. As mentioned, the presence of an opening snap may mimic paradoxical splitting of S2 if P2 fades during inspiration, and only A2 and the opening snap are heard during expiration.

Asking the patient to assume the upright position may help differentiate P2 from the opening snap: the A2-P2 interval decreases and the A2-OS interval lengthens. Usually, the opening snap has a more slapping or sharper sound than P2, but on occasion it may be dull and of low frequency. It is important to try to identify the three separate components (A2, P2, OS) during inspiration.

With severe mitral stenosis and extremely high left atrial pressure, the opening snap may occur very early, only 0.04 to 0.05 seconds after A2. In such circumstances, it may be extremely difficult, if not impossible, to separate the OS from P2.

S3. A third heart sound may be confused for an opening snap. This is particularly true during tachycardia; efforts should be made to slow the heart rate with carotid massage or drug therapy when rapid rates make auscultation difficult. When an S3 is loud, it often has high frequency vibrations and is more easily mistaken for an OS. Conversely, the combination of a low-medium frequency OS and a long A2-OS interval can simulate an S3. The A2-S3 interval range (0.10 to 0.20 seconds) is much greater than that of the A2-OS (0.04 to 0.14 seconds), but there is potential overlap at intervals of 0.10 to 0.15 seconds. During tachycardia, it may be impossible to time these sounds with accuracy. Compared to the higher pitched, discrete OS, the S3 usually is a dull sound of low frequency and low intensity. An S3 usually is heard only at the apex; it is accentuated in the left lateral decubitus position and softens in the upright position. The OS radiates more widely and is usually equally loud in all positions.

TRICUSPID OPENING SNAP. In tricuspid stenosis the opening snap is generated by the tricuspid valve. Characteristically, this sound is best heard at the lower left sternal border and increases in intensity with inspiration. There is usually a wider A2-OS interval than in mitral stenosis. The problem in the clinical detection of the tricuspid OS is that virtually all patients with tricuspid stenosis have coexisting mitral stenosis. Therefore, two opening snaps may be present, usually superimposed. Phonocardiography may record two

separate opening snaps. Clues to tricuspid origin include respiratory variation in OS intensity and maximal loudness at the lower sternal border. A tricuspid OS can be found in Ebstein's anomaly, tricuspid regurgitation, and large flow atrial septal defects. Often, the OS is recordable but barely audible.

ATRIAL MYXOMA. The well known "tumor plop" of a left atrial myxoma can simulate an opening snap. Usually, it is later, of lower frequency, and more variable in its presence and intensity (Fig. 16-8).

PERICARDIAL KNOCK. A loud, medium-high frequency early diastolic sound may be present in constrictive pericarditis. This is the equivalent of

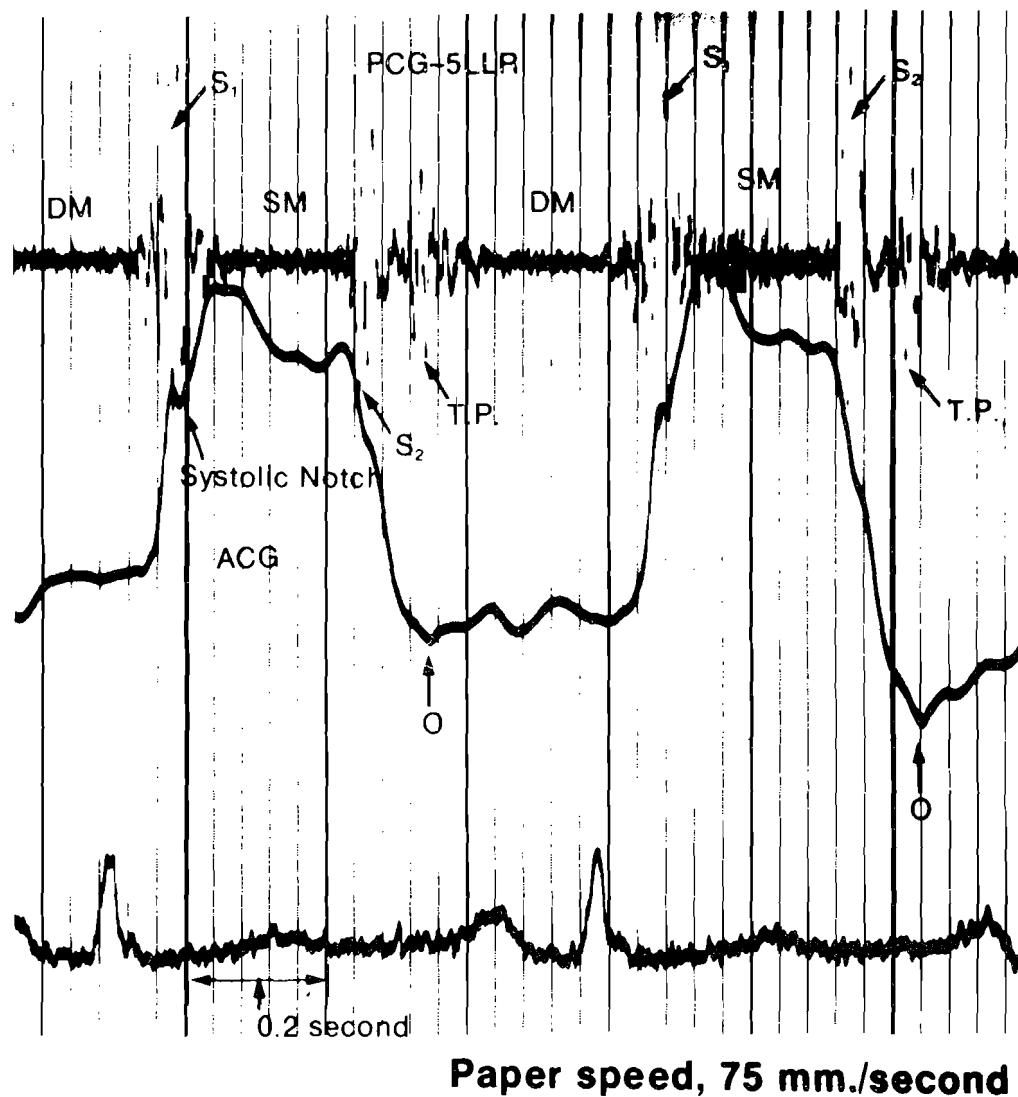


FIG. 16-8. Tumor plop of a left atrial myxoma simulating an opening snap. Note the early diastolic sound (TP), which reflects movement of the atrial myxoma towards the left ventricular cavity. Diastolic murmurs (DM) may be produced if the tumor partially obstructs blood flow into the left ventricle. ACG = apex cardiogram. SM = systolic murmur. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co, 1979.)

an early S-3 and is caused by sudden expansion of the noncompliant left ventricle and pericardium in early diastole. This often produces an audible sound called a pericardial knock; it is easily confused with a mitral OS (Figs. 4-4C, 4-5).

EARLY DIASTOLIC SOUND. Early high frequency diastolic sound transients have been documented in an occasional patient with mitral prolapse (see Chapter 18). These can be confused with an opening snap (Fig. 18-8).

Murmur of Mitral Stenosis

The typical diastolic rumbling murmur of mitral stenosis has two components: an early diastolic murmur that begins with or just after the opening snap and mirrors the rapid filling phase of diastole, and a *late* diastolic or presystolic murmur that follows left atrial contraction and represents a sudden accentuation of transvalvular flow and velocity just prior to mitral valve closure. In patients with mitral stenosis, either or both murmurs may be heard (Figs. 16-4, 16-5, 16-9). In the second (and more common) situation the murmur begins after the OS and then wanes in intensity, only to increase again at the end of diastole (presystolic accentuation). *Practical Point:* It is important to analyze both the early and late portions of diastole for an accurate assessment of the acoustic events in mitral stenosis.

Early Diastolic Murmur. Blood flow across the stenotic mitral valve begins shortly after isovolumic LV pressure falls below LA pressure (Fig. 16-2). The OS occurs just after antegrade mitral valve flow begins, and the first vibrations of the diastolic murmur are coincident with the OS. The mitral valve leaflets normally float to a closed position immediately after their initial opening motion in early diastole (Fig. 7-1). In mitral stenosis the closing motion of the valve cusps is attenuated (Fig. 16-4). The murmur is loudest in mid-diastole. As flow diminishes in late diastole, the murmur tapers in intensity and may disappear. The presence or absence of the murmur in late diastole is dependent on the magnitude of the residual left atrial-left ventricular pressure gradient. In severe mitral stenosis and/or with rapid heart rates, there is a continuous late diastolic gradient and a long diastolic murmur (Fig. 16-5C). If the mitral obstruction is only mild to moderate, the left atrium may be decompressed, and no gradient or murmur will be present in late diastole. The absence of a late diastolic component is most noticeable following long cycle lengths during atrial fibrillation or with slow heart rates in normal sinus rhythm (Fig. 16-10).

Presystolic Murmur. In patients who are in sinus rhythm, late diastolic enhancement of the earlier diastolic murmur or an isolated short presystolic murmur occurs after LA systole and the sudden increase in the LA-LV pressure gradient (Fig. 16-11). Mitral valve closure is delayed and may not occur until left ventricular pressure has begun to rise during isovolumic

systole. Thus, the murmur literally crescendos into S1, producing a presystolic murmur-S1 complex that may be surprisingly loud (Figs. 16-4, 16-9).

When atrial fibrillation is present, the presystolic murmur related to LA contraction disappears. However, it has been documented that in patients with moderately severe mitral stenosis who are in atrial fibrillation a presystolic murmur or accentuation of the long diastolic murmur may be heard, particularly during short R-R intervals in spite of the absence of left atrial

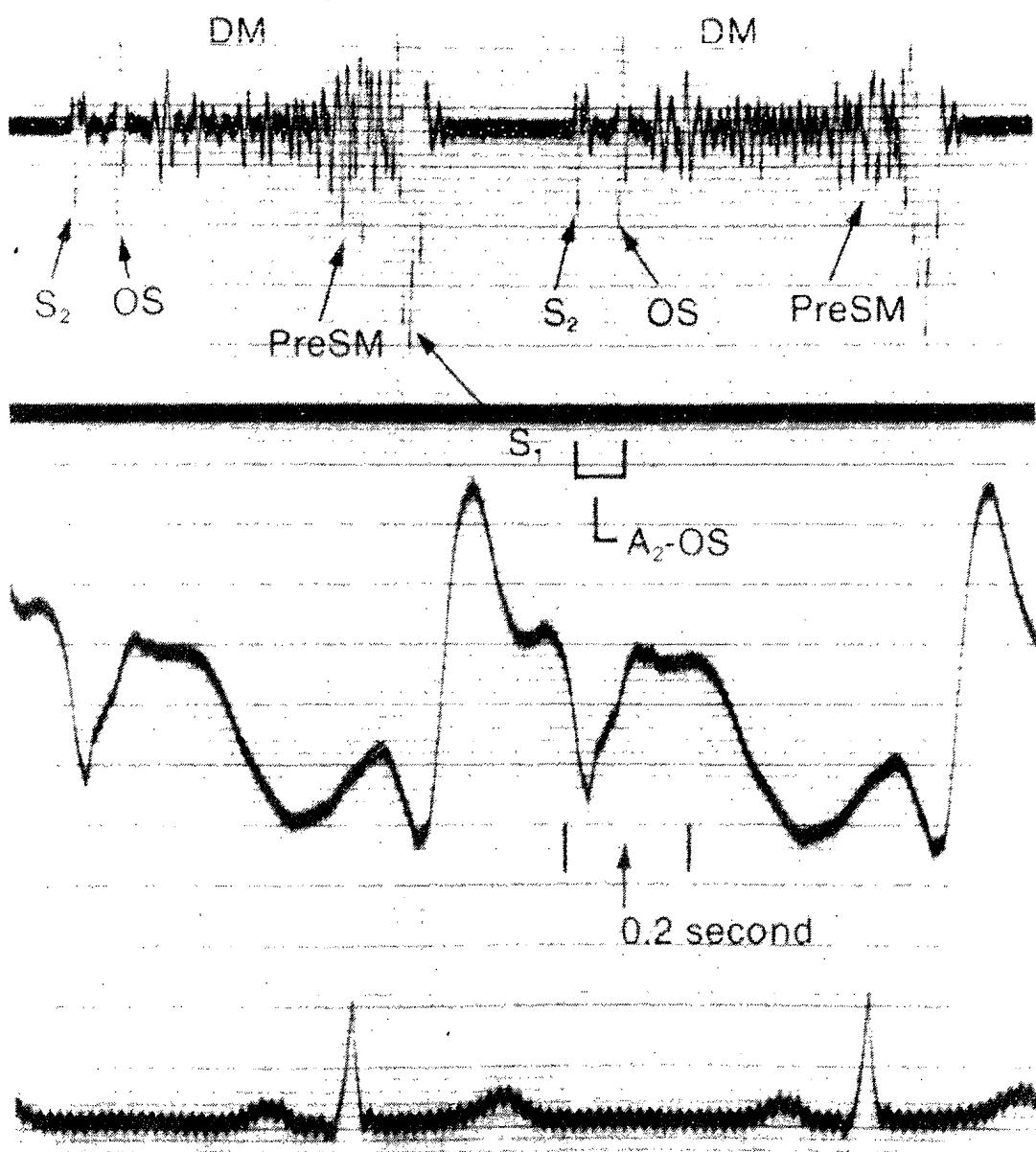


FIG. 16-9. Presystolic murmur of mitral stenosis. In this phonocardiogram, the most prominent component of the diastolic murmur is a crescendo-like augmentation that follows left atrial contraction. On auscultation, the presystolic murmur appears to "explode" into a loud S1. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co., 1979.)

contraction (Fig. 16-10). The mechanism has been nicely explained by Criley and associates: if there is a persistent end-diastolic pressure gradient, the closing of the mitral valve provides a decreasing orifice area for left atrial blood to flow into left ventricle. Although the volume of transmural blood is diminished in late diastole, the actual *velocity* of mitral blood flow increases as the valve orifice narrows. This can cause sufficient turbulence to produce a presystolic murmur. *Practical Point:* In patients with atrial fibrillation, the presence of presystolic accentuation of the diastolic rumble during short cycle lengths indicates mitral stenosis of major hemodynamic importance. This phenomenon is not seen in mild mitral stenosis (Fig. 16-10).

Relationship to Severity. *Mild Mitral Stenosis.* In patients with a small resting LA-LV gradient, there may either be a short, early diastolic murmur,

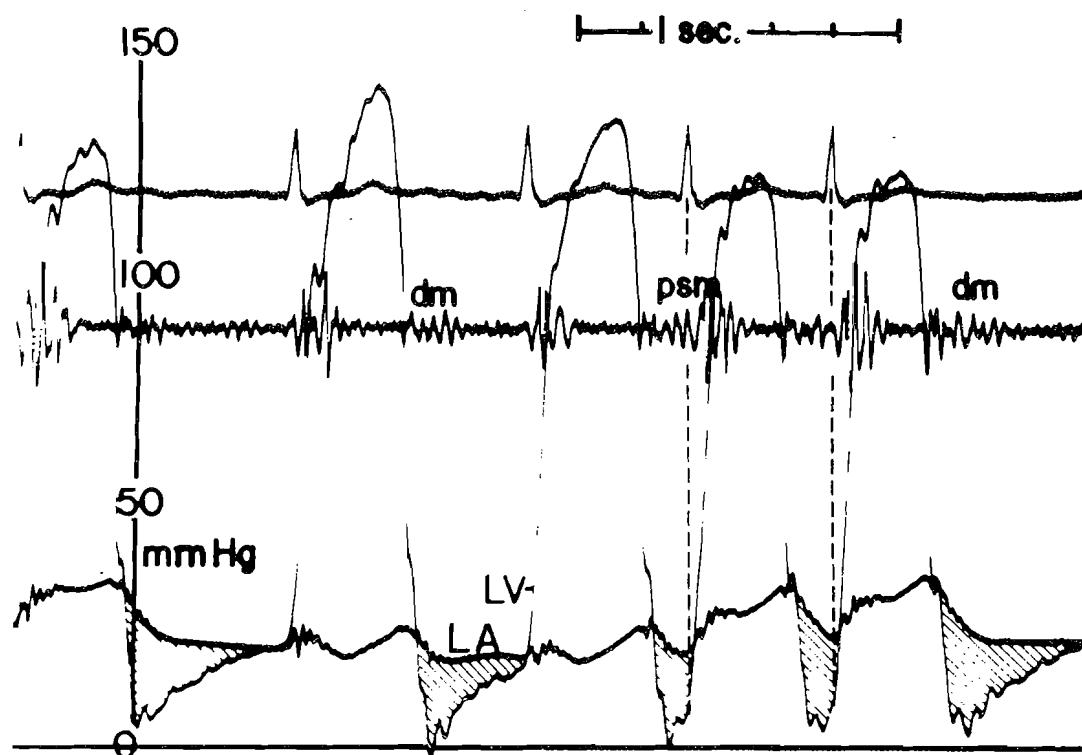


FIG. 16-10. Presystolic murmur of mitral stenosis in the presence of atrial fibrillation. Note that during the short diastolic cycles (third and fourth beats from the left) there is a crescendo presystolic murmur that becomes more accentuated with the onset of left ventricular systole (PSM). During longer cycles there is an early diastolic murmur but no presystolic component. Thus, a presystolic murmur may still be present in the absence of synchronous left atrial contraction. It is produced by a persisting diastolic gradient during short cycles, reflecting continuous diastolic flow across the mitral valve. As the mitral orifice continues to narrow in late diastole, transmural velocity is augmented and a presystolic murmur immediately precedes S1. During longer cycle lengths there is adequate time for the left atrium to decompress prior to the onset of left ventricular contraction; mitral valve closure is completed and there is no presystolic murmur. (From Criley JM, et al: Mitral stenosis: mechanico-acoustical events. In Physiologic Principles of Heart Sounds and Murmurs. Edited by DF Leon and JF Shaver. American Heart Association Monograph No. 46, 1975.)

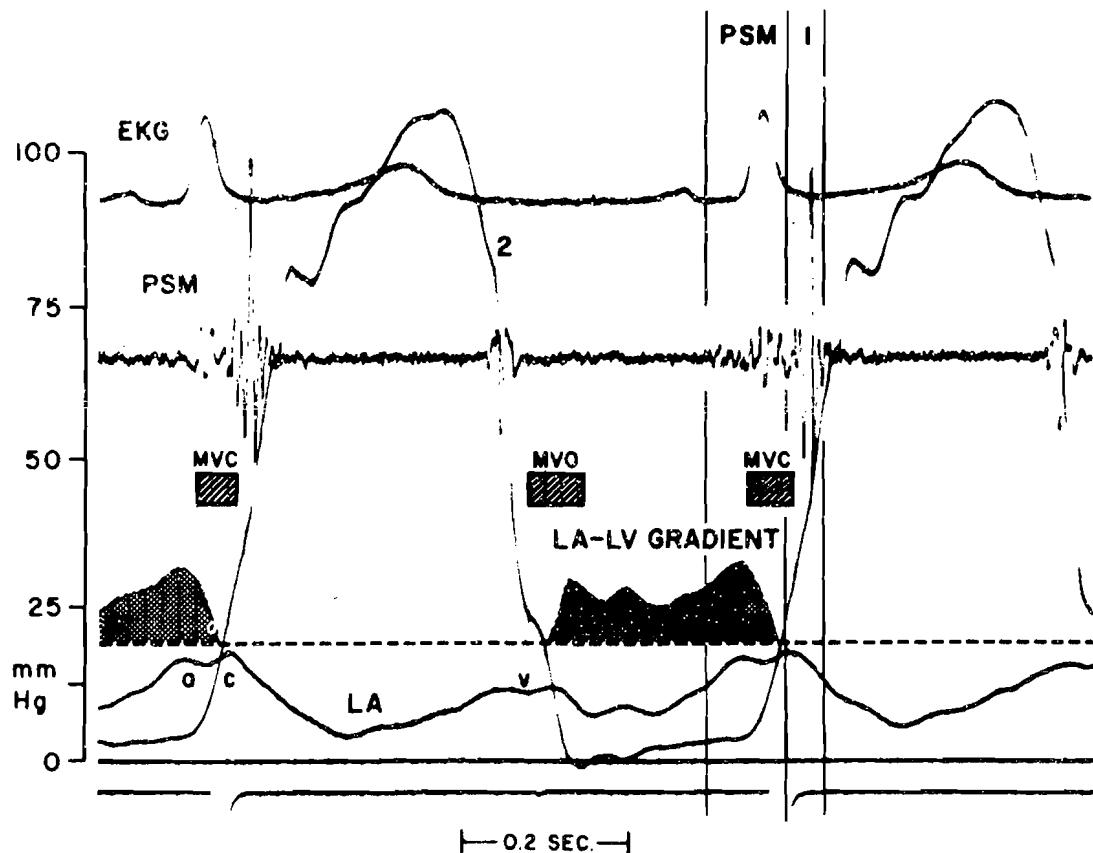


FIG. 16-11. Intracardiac pressures and the presystolic murmur of mitral stenosis. The dark area indicates the magnitude of the left atrial-left ventricular gradient. The presystolic murmur reflects augmented blood flow across the mitral valve in late diastole following left atrial contraction. The closing motion of the mitral valve contributes to the crescendo shape of the presystolic murmur by narrowing the mitral orifice during continuous flow of blood into the ventricle. The delayed closure of the valve contributes to the accentuated S1. MVC = mitral valve closure, MVO = mitral valve opening. PSM = presystolic murmur. LA = left atrial pressure. (From Criley JM, Feldman IM, and Meredith T: Mitral valve closure and the crescendo presystolic murmur. *Am J Med* 51:456, 1971.)

or, on occasion, only a presystolic crescendo murmur (Fig. 16-5A). If both murmurs are present, there is usually a silent gap in mid to late diastole between the two audible murmur components.

Mild to Moderate Mitral Stenosis. Both the early and late murmurs are readily heard, but a distinct tapering or silence is observed after the mid diastolic component (Fig. 16-5B).

Moderate to Severe Stenosis. In patients with a substantial and persistent LA-LV gradient, the diastolic rumble is truly pandiastolic; sound vibrations start with the opening snap and extend to S1, typically with an increase in intensity at end-diastole (Figs. 16-5C, 16-9). In subjects with slow heart rates or variable cycle lengths during atrial fibrillation, it is important to carefully assess the length of the murmur. *Practical Point: A holodiastolic murmur*

following the OS during long R-R intervals in sinus rhythm or atrial fibrillation is consistent with major obstruction at mitral valve level.

Heart Rate. The effects of tachycardia on the length of the murmur of mitral stenosis are important to consider. As diastole shortens during rapid heart rates, both LA pressure and the LA-LV gradient increase. This will cause an apparent increase in the duration of the murmur and accentuate the late diastolic or presystolic component. If tachycardia causes the mitral diastolic murmur to become more prominent, it may result in an overestimation of the severity of the mitral stenosis by prolonging the length of the murmur. Thus, it is important to assess the duration of the mitral rumble at normal or even slow heart rates to best evaluate the severity of obstruction.

Acoustic Characteristics. *Contour.* The typical murmur is decrescendo in early diastole and crescendo in late diastole. Long pandiastolic murmurs tend to be even in intensity with or without presystolic accentuation.

Frequency. Because of the relatively small pressure gradient developed across the mitral valve, blood flow velocity and turbulence within the LV are usually of a low order of magnitude, and the frequency or pitch of the mitral stenosis murmur is quite low. Analogies between the characteristic rumble of mitral stenosis and distant thunder, the sound of a bowling ball slithering down the alley, or the noise of a distant subway train are fanciful but valid. When mitral flow is relatively rapid, as after mild exercise or in subjects with a good cardiac output, the murmur may be higher pitched. The bell of the stethoscope should be used for auscultation of all but the very loudest murmurs of mitral stenosis. Use only the lightest pressure necessary to make a skin seal. The left recumbent position is mandatory to enhance detection of the low frequency vibrations that become much more audible with this maneuver (Figs. 16-3A, 16-12). Often the diastolic rumble can be heard only with the patient in the left decubitus position. One can readily attenuate or obliterate most mitral stenosis murmurs with firm pressure on the bell or through the use of the diaphragm of the stethoscope.

Location. One of the difficulties in detection of the murmur of mitral stenosis is the limited precordial area over which the murmur is heard. With murmurs of low to medium intensity, the zone of auscultation is often quite small, localized to the apex impulse in the left recumbent position. The murmur may be inaudible in the supine position. It is useful to listen carefully over the site where S1 is loudest as well as at the cardiac apex. *Practical Point:* Careful identification of the apical impulse with one finger and simultaneous application of the bell of the stethoscope in the left lateral position is mandatory for auscultation of the mitral stenosis murmur in many patients (Fig. 16-3A). The murmur may be inaudible even 1 to 2 cm away from this area.

Radiation. The low intensity mitral stenosis murmur is well localized and does not radiate appreciably. When the murmur is very loud, it may be

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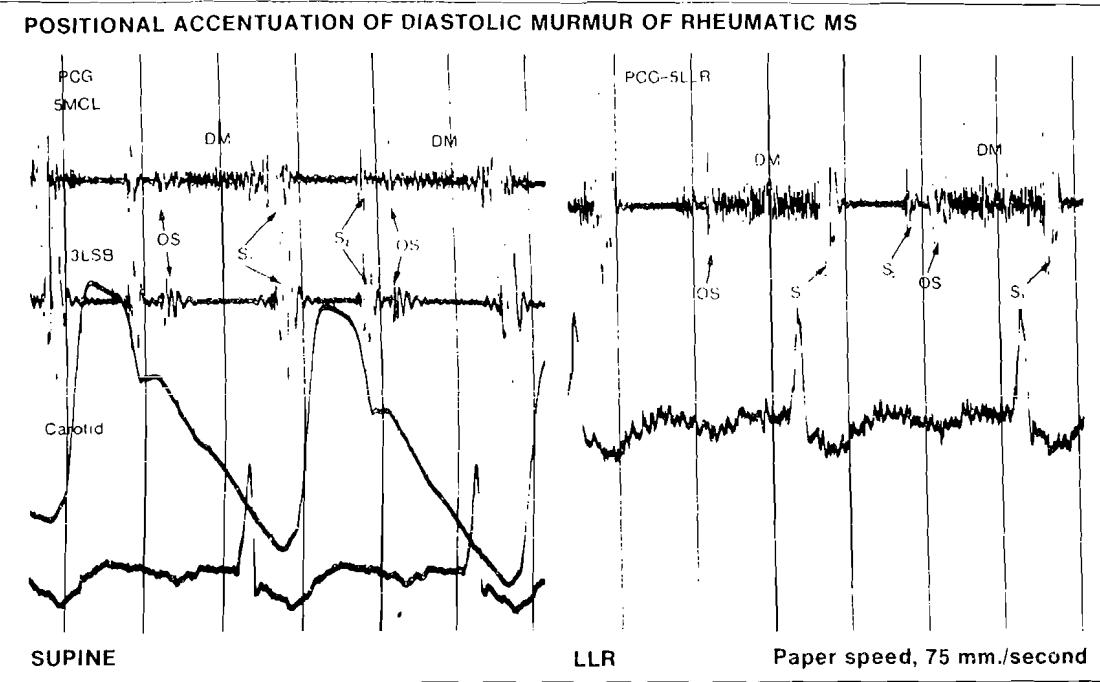


FIG. 16-12. Augmentation of the diastolic murmur of mitral stenosis in the left lateral position. This sequence of phonocardiograms demonstrates the increase in the intensity of the diastolic murmur that occurs when the patient is turned to the left lateral position. The gain settings of the phonocardiogram are the same. DM = diastolic murmur; OS = opening snap. (From Delman AJ and Stein E: Dynamic Cardiac Auscultation and Phonocardiography. Philadelphia, WB Saunders Co, 1979.)

heard toward the lower left sternal border. In patients with a small heart and an apical impulse that is located inside the midclavicular line, the murmur may be audible just to the left of the left sternal edge. When the murmur is of medium frequency and medially located, it can readily simulate the diastolic murmur of aortic regurgitation. Careful attention to the presence of an OS and a sound delay or gap between A₂ and onset of murmur usually will distinguish these two possibilities.

Intensity. The loudness of the mitral rumble does not bear as consistent a relationship to the severity of the obstruction as does the length of the murmur. The intensity of the murmur of mitral stenosis is directly related to the velocity of blood flow across the mitral valve and the severity of the stenosis. High flows result in loud murmurs; a narrow valve orifice produces an increased velocity of blood flow if stroke volume is maintained. However, when mitral stenosis becomes severe, flow velocity may diminish as stroke volume falls. Mitral stenosis murmurs may be so loud as to produce an apical thrill in the left decubitus position, particularly in thin patients. Increased flow results in increased turbulence and a louder murmur; thus one should utilize various maneuvers to accentuate mitral blood velocity for optimal auscultation. Many subjects with mitral stenosis have a murmur of such

softness as to make detection difficult. This is discussed in the section, "Aids to Detection of Mitral Stenosis."

Factors Decreasing the Intensity. Severe pulmonary hypertension may minimize audibility of the mitral rumble if RV hypertrophy and dilatation occur; the optimal anatomy for auscultation will be distorted as the RV takes over the cardiac apex. A depressed cardiac output in this setting may also be a problem. Calcification of the mitral valve is more likely to be associated with a rigid, limited orifice and low flow state resulting in a soft murmur. Extracardiac factors such as body habitus and emphysema can attenuate the discretely localized murmur of mitral stenosis. Associated cardiac conditions can mask the murmur of mitral stenosis. In patients with atrial septal defect and coexisting mitral stenosis, left to right shunting of blood across the atrial defect may diminish transmural flow. In aortic stenosis or aortic regurgitation, the mitral diastolic murmur may be attenuated because of left ventricular factors (hypertrophy, decreased compliance), resulting in a decreased opening motion of the anterior leaflet of the mitral valve. It is common to completely miss the diagnosis of mitral stenosis when there is associated aortic stenosis, as the rumble and OS are often inaudible.

Table 16-4 lists the causes of the decreased murmur intensity of mitral stenosis.

TABLE 16-4 *Factors Associated with Decreased Intensity of the Diastolic Rumble of Mitral Stenosis*

Low Flow States

- Severe mitral stenosis
- Severe pulmonary hypertension
- Congestive heart failure
- Atrial fibrillation, especially with rapid ventricular rate

Characteristics of the Mitral Valve

- Extensive calcification of the mitral apparatus
- Subvalvular chordal obliteration
- Left atrial thrombus protruding into mitral orifice
- Anatomic distortion of the mitral valve with posteromedial deviation of the orifice

Associated Cardiac Lesions

- Aortic stenosis
- Aortic regurgitation
- Atrial septal defect
- Pulmonary hypertension with marked RV enlargement

Other

- Cardiac apex formed by right ventricle
- Inability to localize cardiac apex:
 - Obesity
 - COPD
 - Muscular chest
 - Large breasts

Silent Mitral Stenosis. It is uncommon for mitral stenosis to be truly undetectable or silent, but this situation certainly does occur. In very mild mitral stenosis with a large mitral valve orifice and a small gradient, the murmur may be absent. The most common cause of silent mitral stenosis is poor auscultatory technique such as improper application of the stethoscope (failure to use the left decubitus position; failure to use light pressure with the bell), erroneous identification of the cardiac apex, lack of the use of murmur enhancement techniques (see below), or misinterpretation of the cardiac findings. Nevertheless, mitral stenosis can be truly inaudible when there is low blood flow regardless of cause. Left atrial thrombus may further narrow the mitral valve orifice. Marked distortion of the mitral valve from fibrocalcific changes may direct the stream of blood away from the left precordial area. With right ventricular dilatation, the apex impulse may not be left ventricular in origin and is occupied by the right ventricle. In such cases, the murmur may be audible only in the left or midaxillary line, and should be carefully sought in this area. Very severe mitral stenosis, associated pulmonary hypertension, and combination lesions (e.g., aortic valve disease) commonly result in silent mitral stenosis on auscultation.

When the clinical setting suggests possible mitral stenosis (e.g., atrial fibrillation, "lone" mitral regurgitation, systemic embolization, unexplained pulmonary hypertension, unexplained left atrial enlargement on ECG or chest roentgenogram), and a diastolic murmur is not audible, a careful search for the elusive mitral rumble should be undertaken with use of special techniques, particularly if S1 is prominent or a diastolic filling sound is heard.

Factors Increasing the Intensity. In combined mitral regurgitation and stenosis, there is an increased volume of left atrial blood that accentuates both the intensity and duration of the mitral rumble. In general, males tend to have louder murmurs than females. Tachycardia or short cycle lengths during atrial fibrillation will also increase murmur intensity.

AIDS TO DETECTION OF MITRAL STENOSIS (Table 16-5)

The proper use of the stethoscope (light pressure with bell at the cardiac apex in the left decubitus position) has been emphasized. Auscultation should be performed in mid- or end-expiration; the faint mitral rumble can be easily masked by respiratory sounds. It is surprising how easily the rumble is heard when the patient is auscultated in the process of turning onto the left side or immediately after assuming the left recumbent position (Fig. 16-12); this probably is related to a brief increase in heart rate and transvalvular flow occurring during the turning maneuver. *Practical Point: Anything that increases the heart rate and/or the left atrial-left ventricular gradient will enhance audibility of the mitral rumble.* The most practical maneuvers to accomplish this include (1) asking the patient to cough deeply several times in succession, (2) using mild exercise, such as sit-ups or deep knee bends, or

TABLE 16-5 *Essentials for Optimal Auscultation of the Mitral Stenosis Murmur*

Identify left ventricular apex
Listen with the bell of the stethoscope, using light pressure with the bell placed directly on the apex impulse
Employ the left lateral decubitus position (listen while patient is turning)
Use maneuvers that will increase heart rate and left atrial pressure: coughing, deep respirations, mild exercise, sudden squatting
Accurately identify S1, S2, and the opening snap—begin by inching towards the apex from the cardiac base or pulmonic area
Listen during held expiration

(3) listening while the patient changes from the squatting to the supine or left lateral position. Isometric effort in the form of a sustained handgrip may also be useful. Some advocate having the patient breathe deeply several times in rapid succession to accelerate the heart rate.

The intensity of the murmur tends to increase during expiration and to decrease or remain unchanged during inspiration. Amyl nitrite has been used effectively in the diagnosis of mitral stenosis, as the intense reflex tachycardia markedly accentuates the mitral rumble. This drug also can be used to differentiate the mitral murmur of mitral stenosis from the Austin Flint murmur of aortic regurgitation (see Chapter 15, Fig. 15-9). The latter classically will diminish after inhalation of amyl nitrite as the volume of regurgitant aortic flow decreases. The Valsalva maneuver may be used in bringing out the mitral rumble. The strain phase dampens the mitral stenosis murmur, but 6 to 8 beats after the Valsalva release, the murmur may increase in intensity as the heart rate and cardiac output increase.

These various techniques and maneuvers generally have the same effect on both the early and late components of the mitral rumble. The presystolic murmur will definitely be augmented whenever diastole is shortened and left atrial pressure increases. Because the frequency of this murmur is usually higher than the earlier diastolic rumble, the bell or diaphragm may be used; the presystolic component should be specifically sought during auscultation, as it may be more easily detected than the lower pitched vibrations of the early diastolic murmur.

Identification of Systole and Diastole. Diastole is easily confused for systole in patients with mitral stenosis, particularly when there are no other associated murmurs and then the heart rate is relatively fast. A prominent opening snap can be readily mistaken for S1, with the diastolic murmur appearing to be systolic in timing. The loud S1 is confused for S2. Because most cardiac sounds are heard during systole and are associated with a long silent diastole, the subtle diastolic acoustic events in pure mitral stenosis and the silent systole often cause confusion (Fig. 16-13). Rapid heart rates accentuate this problem.

CONFUSION OF DIASTOLE FOR SYSTOLE IN PURE MITRAL STENOSIS

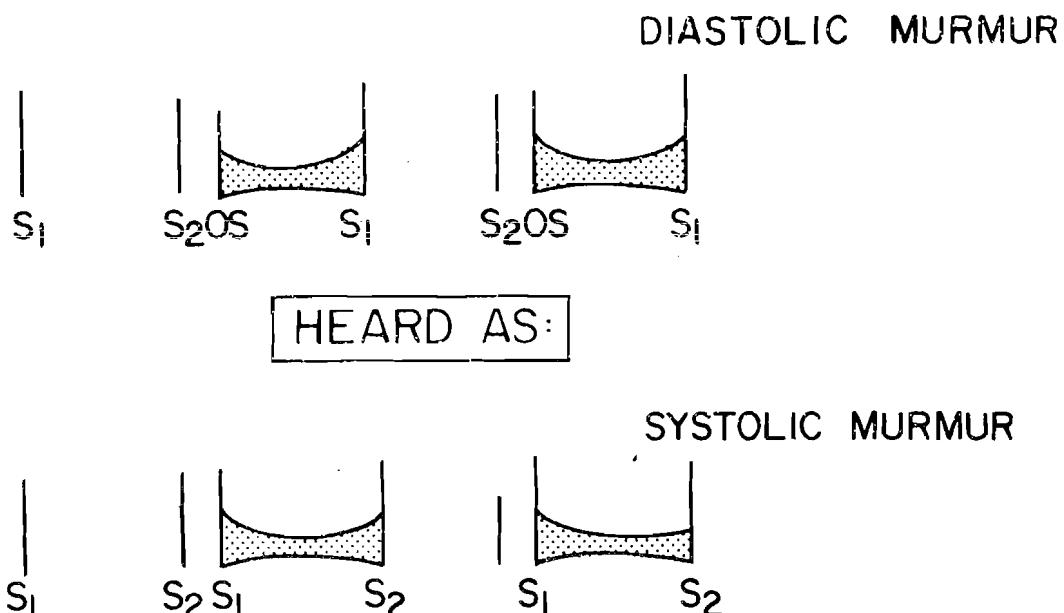


FIG. 16-13. Auscultatory confusion of diastole for systole in mitral stenosis. This diagram indicates how an observer can mistake the diastolic murmur of mitral stenosis for a systolic murmur. The opening snap is readily heard as an "S1," and systole is confused with diastole. Because physicians are used to hearing the majority of murmurs in systole, the acoustic findings in pure mitral stenosis can be misleading. This is particularly true if the underlying heart rate is rapid. Careful attention to the proper identification of S1 and S2 is essential to the correct interpretation of cardiac events.

Practical Point: Always utilize careful simultaneous palpation of the carotid artery and/or apex impulse to accurately identify S1 and S2 and to define the OS and the proper sequence of systole and diastole. The "inchng technique" is helpful in preventing this confusion; S1 and S2 are more easily identified at the base where the OS may be heard but the diastolic murmur will be absent. Careful inching of the stethoscope towards the apex after definite identification of these cardiac sounds will avoid confusing systole with diastole.

PULMONARY HYPERTENSION AND MITRAL STENOSIS

In certain patients with mitral stenosis, typically younger women, there is a severe pulmonary vascular reaction consisting of intense pulmonary vascular vasoconstriction and sustained pulmonary hypertension. The degree of elevation of pulmonary artery pressure is out of proportion to the level of pulmonary capillary wedge or LA pressure, and the pulmonary arteriolar

resistance is very high. In such individuals, right heart hemodynamics are substantially affected, and marked right ventricular enlargement and right ventricular failure are common. Such patients are likely to have a low, fixed cardiac output without symptoms of a high left atrial pressure.

The physical examination in such individuals can be very misleading. First, the clinical evidence for mitral stenosis may be minimal; the opening snap and mitral rumble are often attenuated as a result of the severe pulmonary hypertension and low flow state. Second, prominent right ventricular enlargement may lead the unsuspecting clinician on a search for intrinsic pulmonary vascular or parenchymal disease. Table 16-6 lists the differences in the physical examination in patients with typical mitral stenosis and normal pulmonary artery pressures compared to those with severe pulmonary hypertension. In such situations, an astute observer may be alerted to the possible presence of mitral stenosis by a loud or prominent S₁ or an unexpected opening snap, but often echocardiography is necessary to establish the definitive diagnosis of mitral stenosis.

DIFFERENTIAL DIAGNOSIS

A number of conditions can simulate mitral stenosis. Any cause of enhanced A-V valve flow may result in an S₃ and short mid-diastolic murmur, a sound complex mistaken for an opening snap and mitral stenosis rumble (see Table 16-7).

TABLE 16-6 *Physical Findings in Mitral Stenosis with and Without Pulmonary Hypertension*

	No Pulmonary Hypertension	Severe Pulmonary Hypertension
General appearance	Normal	Malar flush common, often with cyanosis
Rhythm	NSR or atrial fibrillation	NSR more common
Venous pulse	Normal	Increased mean pressure, large A waves if NSR. Large V waves common (tricuspid regurgitation)
Precordial motion	Small LV impulse 1-2+ RV impulse	LV not palpable; apex formed by RV 3-4+ RV impulse
S ₁	Increased	Increased
S ₂	Normal; P ₂ may be increased	Booming P ₂ , often palpable. PA ejection sound, RV S ₃ or S ₄ may be palpable
Opening snap	Prominent	Soft
Easily heard	Easily heard	May be faint or totally absent
Associated murmurs	Faint mitral or aortic regurgitation murmurs	Tricuspid regurgitation, pulmonary regurgitation (Graham Steele) murmurs

TABLE 16-7 *Conditions Associated with Prominent Diastolic Filling Sounds and/or Murmurs Simulating Mitral Stenosis*

Thyrotoxicosis	Atrial septal defect
Anemia	Ventricular septal defect
Third degree A-V block	Patent ductus arteriosus
Mitral regurgitation (pure)	Ebstein's anomaly
Large left ventricle (cardiomyopathy)	

Tricuspid Stenosis. Patients with rheumatic tricuspid stenosis will have a tricuspid opening snap and diastolic rumble on examination. Because tricuspid stenosis invariably is associated with mitral stenosis, it is difficult to separate the acoustic features of each lesion. Inspiratory augmentation of the diastolic murmur and maximal intensity at the lower sternal border favor tricuspid stenosis. The jugular A wave should be very prominent (in the absence of atrial fibrillation). The tricuspid stenosis rumble may be somewhat more high pitched and tends to occur earlier than the mitral rumble, and even can simulate semilunar valve insufficiency. S1 may not be particularly loud in tricuspid stenosis.

Carey Coombs Murmur of Acute Rheumatic Fever. This is a diastolic rumbling murmur, often higher pitched than the mitral stenosis murmur, that may vary from day to day. This murmur probably is caused by mitral leaflet thickening due to acute valvulitis exacerbated by the high flow state from fever and anemia. An opening snap and increased S1 usually are not present unless there has been preexisting mitral valve damage. An apical diastolic murmur in a patient with acute rheumatic fever is not likely to be caused by organic mitral stenosis unless there have been previous episodes of rheumatic inflammation and scarring.

Left Atrial Myxoma. Obstruction of the mitral valve orifice by tumor can simulate mitral stenosis. The resultant murmur often is solely presystolic. S1 may be increased and an opening snap may be present (tumor plop) (Fig. 16-8). There may be variability in the acoustic findings from day to day. Echocardiography has caused embarrassment to many clinicians who previously had diagnosed mitral stenosis when, subsequently, the patient was found to have a left atrial myxoma.

Constrictive Pericarditis. The early, loud pericardial knock can mimic an opening snap (Fig. 4-5). However, S1 typically is not increased, and a diastolic murmur will not be present. The jugular venous pulse will manifest prominent X and Y troughs and invariably will have an elevated mean pressure.

Austin Flint Murmur. An apical rumbling murmur in the presence of aortic regurgitation represents mitral stenosis or an Austin-Flint murmur. This differential is discussed in Chapter 15. The lack of a loud S1 and an opening snap are the most valuable criteria for exclusion of the diagnosis of

mitral stenosis in patients with aortic regurgitation. The combination of rheumatic aortic regurgitation and mitral stenosis is common. However, the Austin-Flint murmur is found only with severe aortic regurgitation (see Fig. 15-7).

Augmented A-V Valve Flow. Table 16-7 lists the conditions in which a diastolic murmur may be heard in the absence of the mitral valve obstruction as a result of increased flow across the mitral or tricuspid valve. An S3 is common in such cases, often followed by after vibrations that simulate a mid-diastolic murmur (see Chapter 9). It is the S3-after vibration complex that produces a short-mid-diastolic murmur that can mimic mitral stenosis. These murmurs will not have presystolic accentuation, and a loud S1 and opening snap are usually not present. However, in hyperdynamic states, S1 may be increased, similar to that in mitral stenosis, and lead to further confusion. Careful timing of the diastolic sound usually will differentiate an S3 from an opening snap with mitral stenosis. The diastolic rumble is short in such cases. This differential is also discussed in Chapter 9.

COMBINED MITRAL STENOSIS AND MITRAL REGURGITATION

Although many patients with rheumatic mitral valve disease have either pure mitral stenosis or regurgitation, others have an admixture of mitral valve obstruction and incompetence. In such cases, leaflet distortion, thickening, and commissural fusion prevent normal valve function. Because abnormalities of the chordae (foreshortening, thickening, matting) will further affect the distorted leaflet architectures, many valves can neither open nor close adequately.

Usually, it is possible to come to an accurate determination at the bedside as to which is the dominant lesion. The following guidelines should be helpful.

Rhythm. The presence of atrial fibrillation favors dominant mitral stenosis, but chronic mitral regurgitation associated with a very large left atrium can produce this dysrhythmia.

Carotid Pulses. Unless the arterial pulse is quick rising, suggesting a large mitral leak, examination of the carotid arterial pulse is not helpful.

Jugular Venus Pulse. This provides no useful clues.

Left Ventricular Impulse. A prominent hyperdynamic or sustained left ventricular apical thrust strongly supports dominant mitral regurgitation (in the absence of aortic valve disease or any obvious process affecting the left ventricle). A small, unimpressive apex beat with a palpable S1 suggests mitral stenosis as the major hemodynamic problem.

Parasternal Impulse. Definite right ventricular hypertrophy is more common in dominant mitral stenosis with or without major pulmonary hy-

pertension. A large amount of mitral regurgitation may produce a late systolic parasternal lift simulating right ventricular hypertrophy (Figs. 17-4, 17-5, Chapters 5 and 17).

S1. A loud S1 is consistent with dominant stenosis, although a freely mobile anterior leaflet with major mitral regurgitation can produce a snapping S1. A soft S1 favors dominant mitral regurgitation.

S2. S2 provides no real help unless split widely, a finding that favors high volume mitral regurgitation (shortened LV systole).

S3. The presence of an S3 excludes all but the mildest degree of mitral stenosis. Be sure that the S3 is not right ventricular in origin in a patient with mitral stenosis and severe pulmonary hypertension. The louder the S3, the more hemodynamically important is the mitral regurgitation.

Opening Snap. Occasional patients with mitral regurgitation may have an opening snap (see Table 16-3, Chapter 17). Nevertheless, a prominent opening snap suggests a significant degree of mitral stenosis.

Holosystolic Murmur. Dominant mitral stenosis with mild or trivial mitral regurgitation typically produces a soft mitral regurgitation murmur. A very loud, apical pansystolic murmur (grade 3 or more) suggests that the mitral regurgitation is of hemodynamic importance, if not the dominant lesion.

Diastolic Rumble. Because of severe mitral regurgitation itself can produce a short, mid-diastolic murmur, one must pay close attention to the *length* of the diastolic murmur in assessing the degree of mitral stenosis versus mitral regurgitation. A long diastolic murmur with presystolic accentuation is consistent with dominant stenosis, whereas a short or diminutive diastolic murmur suggests dominant mitral regurgitation. The intensity of the murmur may be misleading, as any degree of mitral incompetence will augment the loudness of a coexisting mitral stenosis murmur by increasing the volume and velocity of diastolic flow across the mitral valve.

Chapter 17

Mitral Regurgitation

In its various forms, mitral regurgitation is the most diverse of all acquired valvular lesions. The functional anatomy of mitral valve closure is complex; abnormal valve function may result from disease or distortion of the mitral leaflets, the valvular suspensory or supporting "apparatus," or the left ventricle itself. In addition, a number of pathologic conditions can affect the mitral valve, making mitral regurgitation the most common valve lesion in adults. In contradistinction to the other major valve disorders, the presence of mitral regurgitation may be acute, transient, or chronic, and the subsequent hemodynamic sequelae may wax and wane in severity.

NORMAL ANATOMY AND FUNCTION OF THE MITRAL VALVE

The mitral valve has two major leaflets or cusps, a large anterior or aortic leaflet, and a smaller posterior or mural leaflet (Fig. 17-1). The anterior leaflet is much broader in depth than the posterior and is very mobile. The triscalloped posterior cusp comprises two thirds of the circumference of the mitral valve annulus; it is relatively narrow and has a much more limited motion than the anterior leaflet. During valve closure, coaptation of the extensive anterior and posterior leaflet tissue results in an interdigitating apposition of the two cusps. The total surface area of leaflet tissue is two to three times greater than the area of the functional mitral orifice, thus, providing ample margin for effective systolic coaptation that prevents reflux of blood into the left atrium.

The two mitral cusps are anchored by a complex network of chordae tendinae, which insert at or near the free edges of the leaflets and also attach to the commissural aspects of the valve (Figs. 17-1, 17-2). Most of the chordae arise from either of the two major papillary muscles in a cascading network of individual chords. The chordae are smooth, delicate strands of connective tissue and are vulnerable to stretching, thickening, foreshortening, and rupture. The papillary muscles essentially are specialized extensions of the LV muscular trabeculae: there is an anterolateral and a posteromedial papillary muscle. The anterior muscle originates in the lateral free wall of the left ventricle; the posterior muscle rises from the junction of the posterior interventricular septum and the inferior-posterior, left ventricular wall. *Practical Point: The posteromedial papillary muscle is more vulnerable to fibrosis, con-*

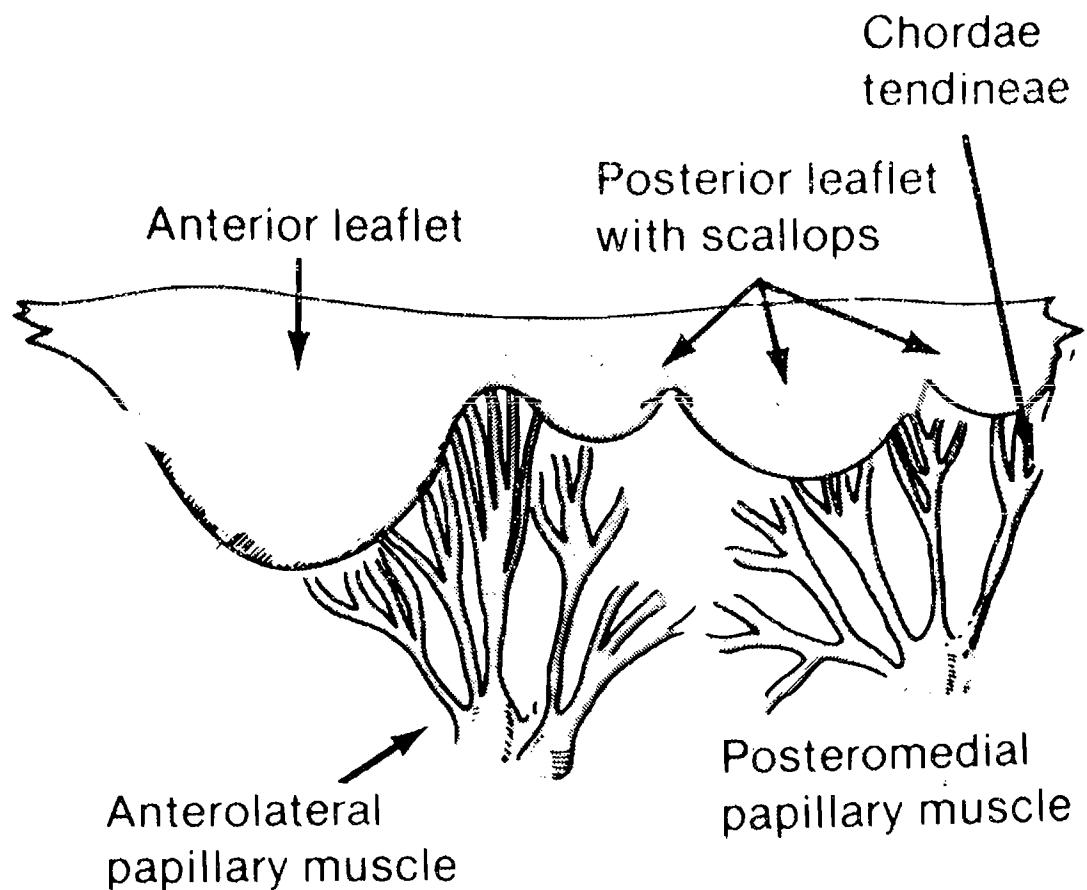


FIG. 17-1. Anatomy of the mitral valve and its supporting structures. The anterior leaflet is much broader than the posterior and is also more mobile. The posterior leaflet or cusp comprises two thirds of the entire annular circumference and is usually formed by three individual scallops. The width of the posterior leaflet is relatively narrow and its motion more limited than that of the anterior leaflet. The anterolateral and posteromedial papillary muscles supply chordae tendineae to both leaflets and are much more extensive than the drawing indicates. (From Abrams J: Prim Cardiol, 1983.)

traction, or rupture, probably because it has a less well-developed coronary blood supply than the anterolateral papillary muscle. The two main papillary muscles have two to four separate heads, each giving rise to large first order chordae tendinae. Each papillary muscle and its respective chordal attachments provide support to *both* the anterior and posterior valve leaflets.

During left ventricular isovolumic systole, the papillary muscles begin to develop tension as soon as intraventricular pressure rises and the mitral valve cusps move towards their final closing position. The apposing mitral leaflets bulge convexly towards the left atrium as the papillary muscles and chordae tauten during the remainder of systole. The leaflets themselves do not normally extend above the plane of the mitral valve annulus during ejection. Tension on the papillary muscles is greatest in mid to late systole when LV size has diminished substantially and the leaflets are prevented from excessive protrusion into the left atrium.

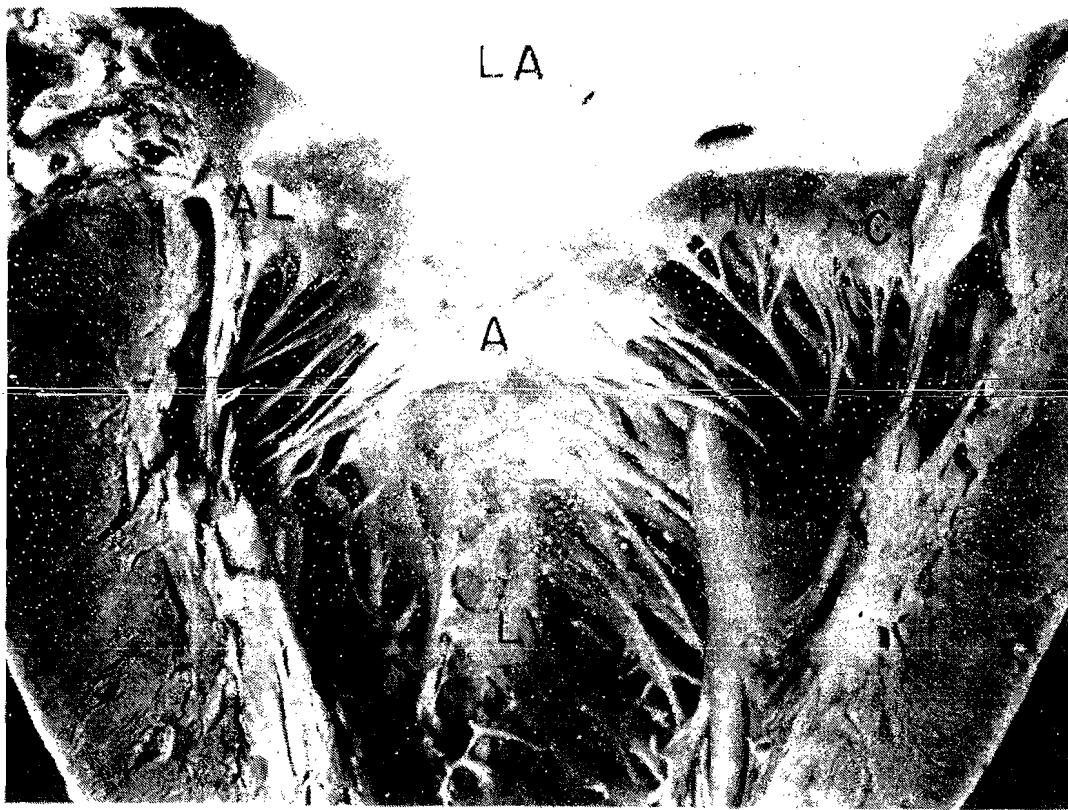


FIG. 17-2. Normal mitral valve. This is an autopsy specimen of a patient with a normal heart. Note the complexity of the chordae tendineae and their fine structure. AL, PM, and C are the scallops of the posterior leaflet. AL = anterolateral, PM = posteromedial, C = central, A = anterior mitral leaflet, LA = left atrium, and LV = left ventricle. (From Lucas RV and Edwards JE: The floppy mitral valve. *Curr Prob Cardiol* 7:1, 1982.)

In early diastole the papillary muscles relax as the mitral valve leaflets swing down into the LV during ventricular filling. The leaflets quickly return to a closed position in mid-diastole after rapid filling and are then reopened by left atrial contraction at the end of diastole. The mitral valve leaflets again move rapidly to their final closed position following left atrial contraction. Eddy currents in the left ventricle and a possible Bernoulli effect result in the final closing motion of the mitral valve cusps. The normal mitral valve ring or annulus becomes considerably smaller during ejection as a result of muscular contraction of the basal portion of the LV. This sphincter-like action aids in maintaining mitral valve closure by decreasing the area necessary for leaflet coaptation.

FUNCTIONAL AND ANATOMIC CAUSES OF MITRAL REGURGITATION (Table 17-1)

There are a variety of causes of mitral regurgitation, each relating to specific derangements of valvular and/or ventricular anatomy. The major

etiologies of mitral regurgitation are discussed in greater detail later in this chapter.

Disorders of the Mitral Leaflets

The classic cause of mitral regurgitation is *chronic rheumatic valvulitis*, which produces scarring and distortion of the valve tissue resulting in failure of the valve cusps to adequately coapt during systole. The chordae are also abnormal, and associated mitral stenosis is often present.

Mitral valve prolapse occurs when mitral leaflet tissue protrudes excessively into the left atrium during ejection. Mitral regurgitation may or may not be present, depending upon the degree of apposition of the prolapsing mitral tissue. There may be a specific morphologic alteration of the valve tissue as is found in Marfan's syndrome or primary mitral prolapse. A number of conditions result in "secondary" mitral valve prolapse (see Chapter 18); in such cases the underlying abnormality may involve the chordae tendinae or the left ventricular muscle and the valve cusps may remain normal.

Endocarditis can directly affect the mitral valve leaflets. Leaflet erosion or perforation, abscess, or sinus tract formation can profoundly alter mitral valve function and structure.

Disorders of the Mitral Valve Supporting Apparatus

The most common etiology of mitral regurgitation due to abnormalities in the supporting apparatus is *papillary muscle dysfunction*, typically caused by acute or chronic manifestations of coronary artery disease. The papillary muscles may be functionally deranged due to transient subendocardial ischemia, or they may be permanently scarred and foreshortened. Rarely, a papillary muscle or one of its heads may rupture during an acute myocardial infarction and produce profound acute mitral regurgitation. Papillary muscle fibrosis is found at autopsy in a wide variety of conditions that involve the left ventricle such as hypertension, aortic stenosis, or cardiomyopathy.

Mitral regurgitation related to disordered papillary muscle function may occur whenever there is *left ventricular dilatation* or severe left ventricular dysfunction of any etiology. In these patients, the papillary muscles may be "pulled" laterally by globular cardiac enlargement. The mitral cusps can become incompetent during systole because the papillary muscles are malaligned, and a prolapsed mitral valve may result. This hypothesis is not accepted by all.

Abnormalities of the *chordae tendinae* may be the cause of mitral regurgitation in a small percentage of patients. Usually one or more of the chordae rupture and allow unsupported mitral leaflet tissue to protrude or hood into the left atrium. In myxomatous degeneration of the mitral valve,

TABLE 17-1 Major Causes of Mitral Regurgitation

<i>Disorders of Mitral Valve Leaflets</i>
Rheumatic valvulitis—acute or chronic
Floppy valve syndrome
Mitral valve prolapse—some variants
Bacterial endocarditis
<i>Disorders of Papillary Muscles</i>
Ischemic heart disease—dysfunction or rupture
Fibrosis
Cardiomyopathy
Aortic valve disease
Hypertensive heart disease
Lateral migration
Gross cardiac dilatation
<i>Disorders of Chordae Tendinae</i>
Floppy valve syndrome
Rupture
idiopathic
endocarditis
<i>Disorders of Mitral Annulus</i>
Calcification
Marked dilatation—e.g., floppy valve syndrome

the chordae are often excessively long, thin, and delicate, which predisposes them both to rupture and prolapse.

Disorders of the Mitral Annulus. Contraction of the mitral valve annulus during systole helps narrow the area of the valve orifice and assists in complete coaptation of the leaflets. When tissue of the annular ring is deranged, mitral regurgitation may result from failure of the mitral valve to close. Typically this occurs with calcification of the annulus, which usually is found in older women. When LV dilatation is marked, the annulus may stretch or dilate; this derangement can contribute to mitral regurgitation in some patients, particularly those with the floppy mitral valve syndrome or severe degrees of LV enlargement.

PATHOPHYSIOLOGY OF MITRAL REGURGITATION

The effects of an incompetent mitral valve on cardiac size and function are modulated by two factors: the *severity* of the mitral leak itself and the *duration* of the underlying hemodynamic disturbance. Mild mitral regurgitation produces relatively little derangement in left ventricular function. Moderately severe mitral regurgitation, when chronic, is surprisingly well tolerated for years. Major mitral regurgitation, particularly when acute or of recent onset, is poorly tolerated and frequently results in profound clinical deterioration. In general, the pathophysiology and compensatory changes are re-

lated more closely to the severity and duration of the valvular leak than to the underlying etiologic process.

The Holosystolic Murmur. In patients with a classic holosystolic murmur of mitral regurgitation, reflux of left ventricular blood into the left atrium begins immediately with the rise in LV intracavitory pressure (Fig. 17-3). Some studies have shown that up to 50% of the entire regurgitant volume may reflux into the left atrium *before* the aortic valve opens. *Thus, the systolic murmur begins with the first heart sound.* The large, left ventricular-left atrial pressure gradient during systole causes regurgitation to continue throughout ejection; the resultant murmur vibrations are heard up to and occasionally even beyond the aortic closure sound (A2).

Left Ventricular Alterations. In severe mitral regurgitation, the regurgitant fraction* is over 50%, i.e., more than half of the LV stroke volume regurgitates back into the left atrium. The LV enlarges in response to the volume overload. During diastole the ventricle receives the blood that had refluxed into the left atrium *in addition to* the normal venous return from the right heart. Initially, the LV chamber dilates; wall hypertrophy is a later development.

Most mitral regurgitant flow occurs before the middle of systole, as the LV "unloads" its large end-diastolic volume more rapidly than normal. Although maximal forward flow may be limited by a large regurgitant fraction, stroke volume usually is well preserved until late in the course of chronic, severe mitral regurgitation. The LV ejection fraction is well maintained for a long period of time, perhaps because there is a relative sparing of energy costs in late systole as LV pressure and tension rapidly decrease. Early systolic aortic blood flow and velocity are abnormally high, but late systolic flow is diminished. In chronic mitral regurgitation, left ventricular and left atrial compliance is increased to cause relatively low intracardiac filling pressures in spite of large intracardiac volumes. In long-standing rheumatic mitral regurgitation, severe elevation of left atrial pressure or significant pulmonary hypertension is unusual. Left atrial dilatation can be enormous and predisposes to atrial fibrillation.

LV function deteriorates relatively late in the course of severe mitral regurgitation. Once LV function becomes depressed, LV ejection fraction falls and end-diastolic and end-systolic volumes increase further; ventricular compliance decreases and LV filling pressure rises. Usually, the result is pulmonary congestion at rest or with relatively mild effort.

Acute Mitral Regurgitation. When acute mitral regurgitation results in abrupt or rapid onset volume overload of the left heart, the pre-existing

* regurgitant fraction = $\frac{\text{regurgitant volume}}{\text{stroke volume}}$

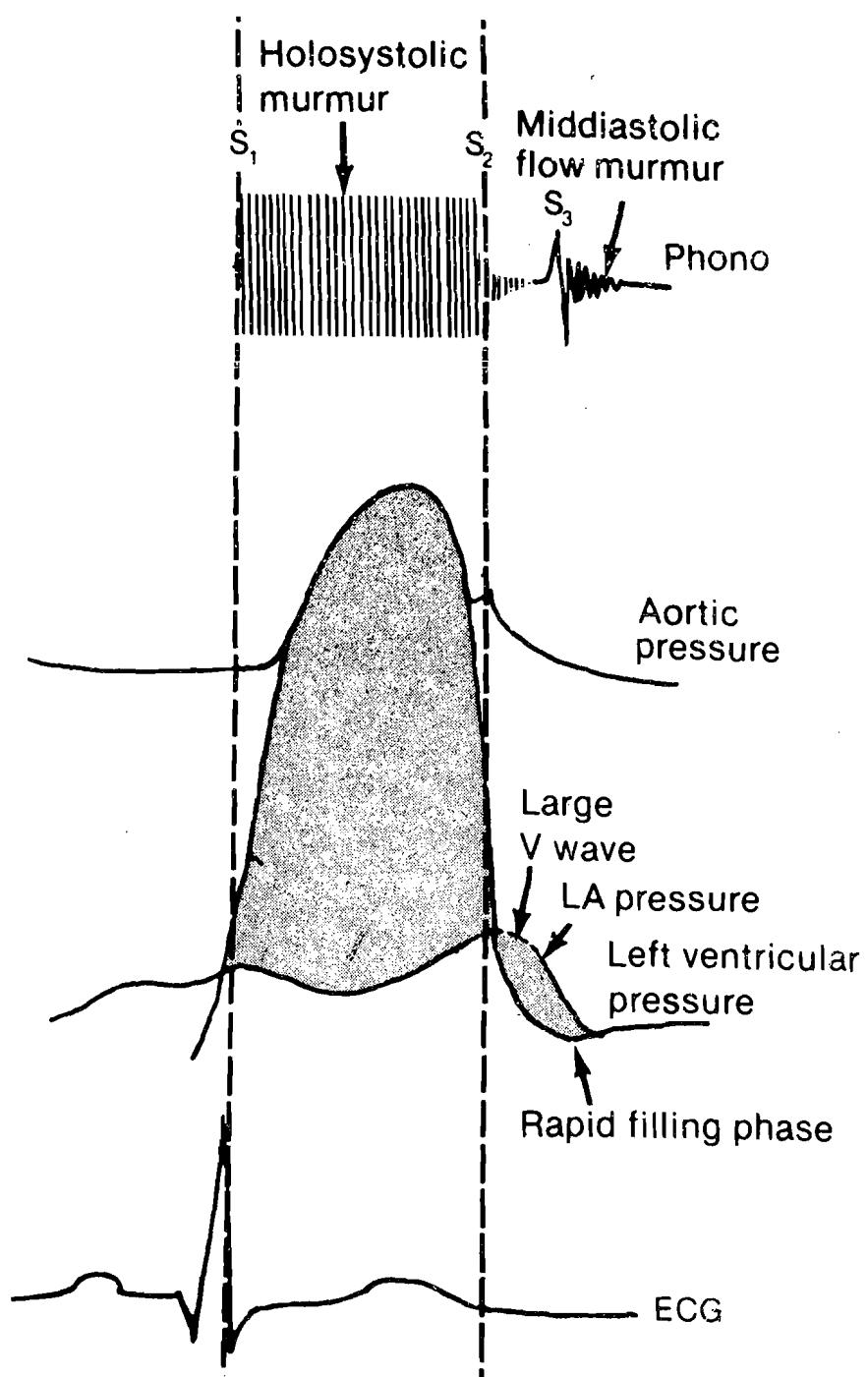


FIG. 17-3. Pressure-sound correlations in mitral regurgitation. There is a large pressure gradient between the left ventricle and the left atrium that begins before the aortic valve opens and ends during isovolumic relaxation. This pressure difference results in a holosystolic murmur with sound vibrations beginning with S₁ and extending to S₂. The murmur is classically even or plateau-like in configuration, although many variants exist. An S₃ is frequently present when there is a significant degree of mitral regurgitation; the S₃ reflects the excessive blood volume traversing the mitral valve in early diastole. Such voluminous left ventricular filling may produce a short mid-diastolic flow murmur in patients with severe mitral regurgitation. (See Fig. 17-6B.)

(From Abrams J: Prim Cardiol, 1983.)

size and compliance of the left atrium are paramount in determining the resultant signs and symptoms. A large volume of regurgitation into a normal, noncompliant atrium results in high left atrial pressures. The LV does not tolerate well an acute volume load when compensatory mechanisms of dilatation and hypertrophy do not have time to develop; left ventricular diastolic and left atrial pressures increase markedly. The clinical course of such patients may rapidly decline downhill. The physical findings often are considerably altered from those in chronic mitral regurgitation (see page 348; Table 17-6).

PHYSICAL FINDINGS

Practical Point: Abnormalities of the carotid and jugular venous pulse, as well as precordial motion, are similar in all types of mitral regurgitation and are related directly to the severity of the valvular leak. Thus, the following discussion of these physical findings is pertinent to *all causes* of mitral regurgitation. However, characteristics of the systolic murmur and associated heart sounds vary considerably among the different types of mitral regurgitation.

Chronic rheumatic mitral valve disease is the prototype for all varieties of mitral regurgitation. In rheumatic mitral valvulitis, fibrosis and distortion of the valve tissue result in thickened and deformed cusps and chordae tendinae that may not function normally. Frequently, there is an element of commissural fusion resulting in associated mitral valve stenosis. Dilatation of the cardiac chambers (both the left atrium and left ventricle) develops over a *prolonged* time; left ventricular and left atrial compliance is increased, and intracardiac pressures may be low at rest and exercise. Cardiac output is well maintained unless the regurgitant fraction increases enormously (e.g., regurgitant volume significantly larger than the forward stroke volume) and/or left ventricular function deteriorates.

Carotid Pulse

In mild mitral regurgitation of any etiology, the arterial pulse is normal. However, with moderate to severe regurgitation the carotid pulse may be brisk or jerky, often with a decreased pulse volume. This results from a normal to decreased forward stroke volume that is ejected more rapidly than normal during early systole. The carotid pulse in significant regurgitation is quick rising, poorly sustained, and low amplitude; it has been called a "small waterhammer" pulse, reminiscent of aortic regurgitation. *Practical Point: A small, quick arterial pulse in a patient with pure mitral regurgitation suggests that the valvular leak is hemodynamically important.* There is no alteration of the systemic blood pressure in mitral regurgitation.

Jugular Venous Pulse

There are no characteristic abnormalities of the venous pulse in mitral regurgitation unless right heart failure has occurred. Right heart decompensation results in an elevation of mean venous pressure and usually is a consequence of major pulmonary hypertension. If the patient is in sinus rhythm, an augmented jugular A wave may be noted, reflecting elevated right ventricular end-diastolic pressure. Patients with severe mitral regurgitation and pulmonary hypertension often have functional (and occasionally organic) tricuspid regurgitation. In such instances, large jugular venous V waves that increase with inspiration will be seen.

Patients with chronic rheumatic mitral valve disease frequently have atrial fibrillation; in this case, the venous A wave disappears and the V wave becomes more prominent, even in the absence of tricuspid regurgitation.

Precordial Motion

Left Ventricular Impulse. Mitral regurgitation typically produces a hyperdynamic apical impulse (see Chapter 5) with an increased amplitude and relatively brief and normal outward motion (Fig. 17-4A). In chronic mitral regurgitation, the left ventricular apex beat will be displaced laterally and downward as the LV enlarges. Pure mitral regurgitation rarely causes massive cardiac enlargement. With the development of abnormal LV function, the dilated LV takes on a more spherical or globular shape, and the apical impulse may become sustained (Fig. 17-6A).

A systolic apical thrill will be felt when the systolic murmur is loud (grade 4/6 or greater). An early diastolic outward impulse is commonly detectable in severe mitral regurgitation; this can be subtle and may be best appreciated when the patient holds the breath in mid-expiration (Fig. 17-4A). The motion is the tactile component of the LV rapid filling wave, i.e., a palpable S3. A palpable A wave (S4) is never found in chronic rheumatic mitral regurgitation.

Parasternal Impulse. There are several causes of a palpable lower sternal lift or impulse in patients with mitral regurgitation (Table 17-2). In severe mitral incompetence without significant pulmonary hypertension, the large regurgitant jet may reflux into the left atrium and produce a palpable outward recoil of the anterior cardiac structures (Fig. 17-5). In such cases, careful palpation will reveal a brief, *late systolic impulse* beneath the lower sternum.

TABLE 17-2 Causes of Systolic Parasternal Lift in Mitral Regurgitation

Late systolic	Large regurgitant jet into dilated left atrium
Holosystolic	Pulmonary hypertension with right ventricular hypertrophy
Holosystolic	Associated mitral stenosis

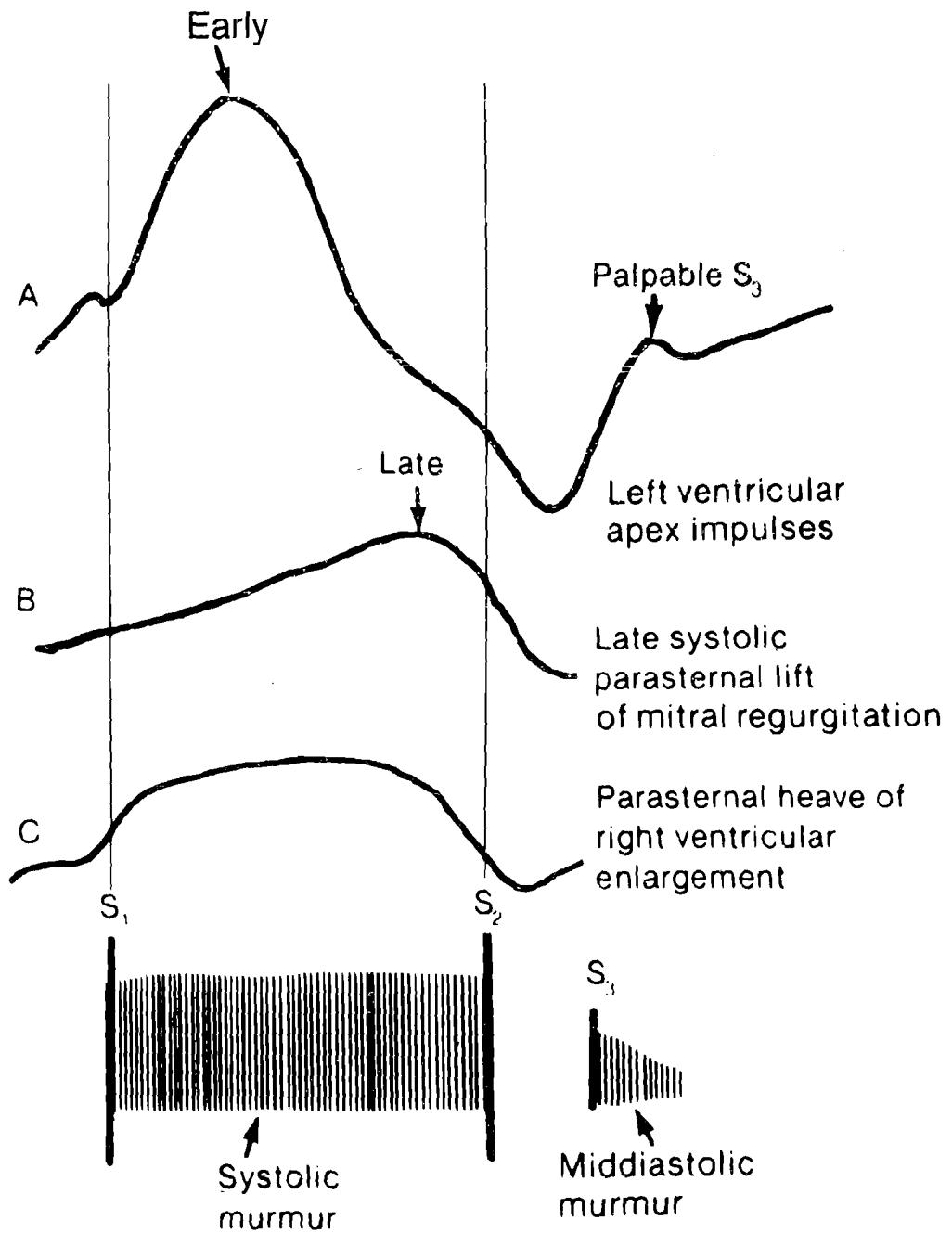


FIG. 17-4. Precordial motion patterns in mitral regurgitation. A. The left ventricular impulse is hyperdynamic with a normal contour and increased amplitude of the early systolic outward motion. A palpable third heart sound may be felt in the left decubitus position. In severe chronic mitral regurgitation the left ventricular impulse may be sustained and heaving in quality (not diagrammed). B and C represent parasternal (right heart) activity. B. A late systolic parasternal lift. This reflects an anterior thrusting motion of the heart that occurs in late systole. It is produced by regurgitation of a large volume of blood into an enlarged left atrium (see Fig. 17-5). This late systolic lift is asynchronous with the left ventricular apical impulse, which is early systolic (A). C. Sustained parasternal lift. The most common finding in severe mitral regurgitation is a gentle, holosystolic outward impulse palpable at the lower left sternal region. This usually is the result of pulmonary hypertension and right ventricular enlargement. (From Abrams J: Prim Cardiol, 1983.)

This outward movement is *asynchronous* with the apical LV impulse occurring just *after* the apex beat is felt. The outward movement of the lower sternum and adjacent interspaces mirrors the large left atrial "V" wave. The typical chest wall motion is a slow rising, late peaking, systolic impulse that collapses in early diastole (Fig. 17-4B).

More commonly, a parasternal impulse in the presence of mitral regurgitation is related to pulmonary hypertension that causes a *sustained* right ventricular lift (Fig. 17-4C). High pulmonary artery pressures result in right ventricular hypertrophy and dilatation. In rheumatic heart disease, severe pulmonary hypertension is most commonly found in *combined* mitral regurgitation and stenosis and is less frequently present in pure mitral regurgitation. The presence of even mild mitral stenosis may result in substantial elevation of left atrial pressure in patients with an incompetent mitral valve, producing further increases in the pulmonary artery and right ventricular systolic pressures. Whenever there is associated mitral stenosis, the isolated *late* systolic sternal impulse usually is absent and a *holosystolic* parasternal heave is felt (Fig. 17-4C). Other clues to pulmonary hypertension are usually present, including a palpable pulmonic ejection sound, loud P2, and a right ventricular S4. The murmur of tricuspid and pulmonary regurgitation may also be present, particularly the former. As already mentioned, many patients with chronic rheumatic mitral regurgitation have a large, compliant left atrium with a relatively low pressure and only moderate elevation in pulmonary artery pressure. A small subset will have severe pulmonary hypertension on the basis of pulmonary arteriolar vasoconstriction, even in the absence of coincident mitral valve obstruction.

First Heart Sound

Alterations in the intensity of S1 are common in mitral regurgitation but are not of diagnostic significance. The most frequent abnormality is a *decrease* in S1 amplitude, probably related to the early onset of regurgitation during isovolumic LV contraction. A loud systolic murmur beginning with the rise of LV pressure may mask or blur identification of S1. The first heart sound is rarely increased in pure rheumatic mitral regurgitation; however, if there is associated mitral stenosis, S1 may be normal or increased in intensity. In nonrheumatic etiologies of mitral incompetence, a loud S1 is common, presumably due to a freely mobile anterior leaflet. This is common in patients with holosystolic prolapse of the mitral valve.

Second Heart Sound

S2 is unremarkable in mild to moderate mitral incompetence. In more severe cases, S2 becomes audibly split in expiration and widely split during

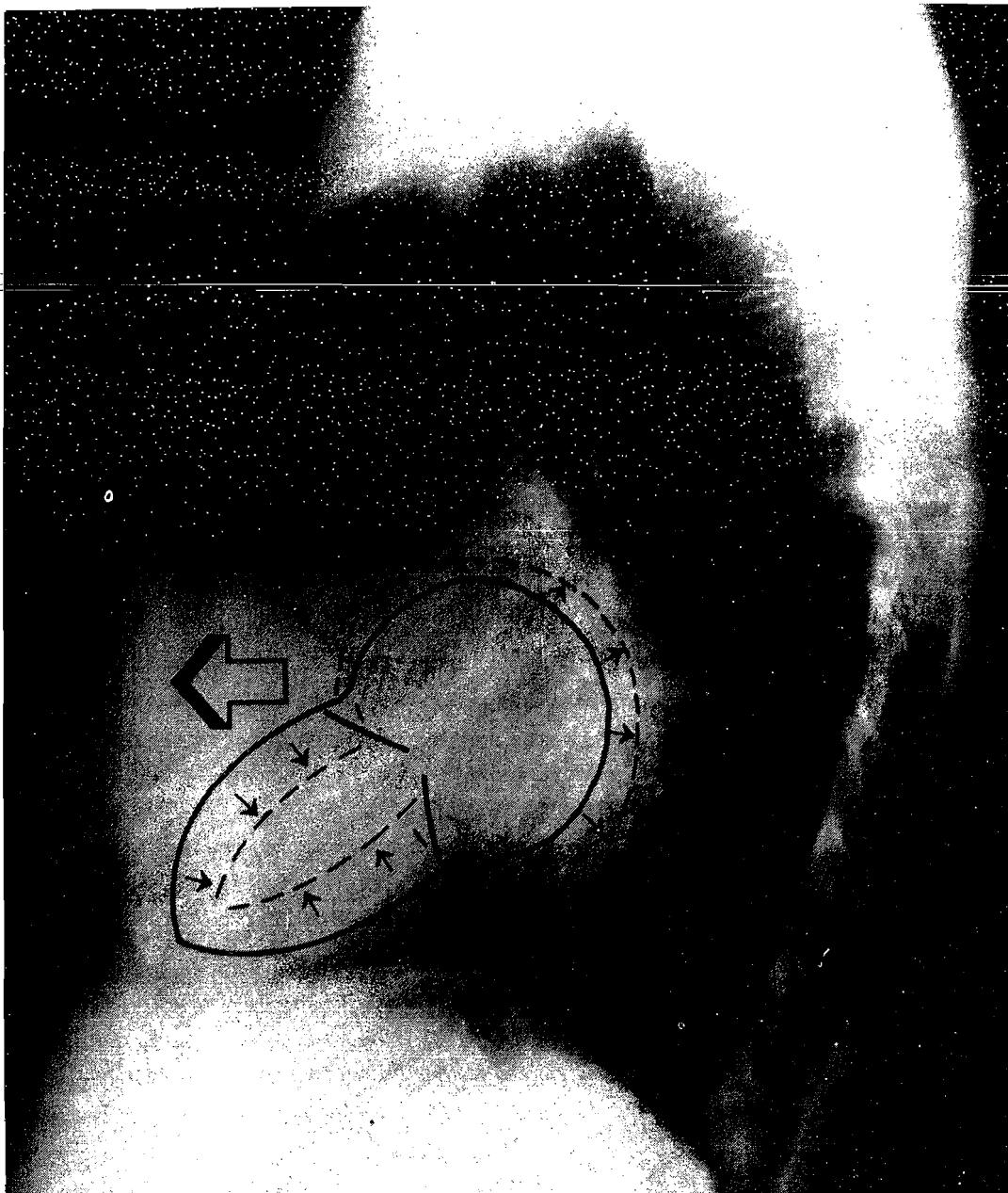


FIG. 17-5. Mechanism of the anterior parasternal thrill in severe mitral regurgitation. During systole a large regurgitant volume fills a dilated left atrium, thrusting the right ventricle forward. This results in palpable anterior displacement of the lower left sternal border, which is late systolic in timing and can be felt as a separate and delayed impulse when compared with simultaneous palpation of the left ventricular apex beat (see Fig. 17-4). (From Abrams J: Precordial Palpation. In *Signs and Symptoms of Cardiology*. Edited by LD Horwitz and BM Groves. Philadelphia, JB Lippincott Co, 1985.)

inspiration. The explanation for the wide S2 is that patients with moderately severe regurgitation have a *shortened* LV ejection time because the LV "unloads" its contents more quickly than normal. This results in early closure of the aortic valve, which in turn causes a widely split S2. Respiratory motion is normal and A2-P2 splitting widens during inspiration. The interval between A2 and P2 increases with the severity of mitral regurgitation and parallels the decrease in LV ejection time. *Practical Point: Prominent expiratory and inspiratory splitting of S2 suggests hemodynamically significant mitral regurgitation and a large regurgitant fraction.* When severe mitral regurgitation and right ventricular failure coexist, additional inspiratory widening of S2 may not be noted. Occasionally, S2 can be single in chronic mitral regurgitation.

Frequently, P2 is accentuated in moderate to severe mitral regurgitation. Although this accentuation typically indicates pulmonary hypertension, occasionally P2 may be louder than normal without significant elevation of pulmonary artery pressure. This has been attributed to abnormal anterior displacement of the pulmonary artery as a consequence of a very dilated left atrium.

Third Heart Sound

A third heart sound is common in mitral regurgitation of hemodynamic importance and implies a large regurgitant fraction (see Chapter 7). *Practical Point: An S3 in severe mitral regurgitation is related to the large volume of blood returning to the LV in early diastole and does not necessarily indicate left ventricular failure.* The S3 occurs 0.12 to 0.20 seconds after A2 and may be quite loud (Figs. 5-5, 17-6). Its presence excludes mitral stenosis of more than an insignificant degree. Frequently, the S3 can be felt in early diastole as a palpable, rapid filling wave, particularly when the patient is turned into the left lateral recumbent position.

If congestive heart failure or LV dysfunction is present, the S3 may reflect impaired cardiac function with LV dilatation. In this setting, the presence of an S3 does not necessarily indicate a large regurgitant fraction. Thus, detection of an S3 in a subject with mitral regurgitation means one of two things: (1) a major mitral leak with a large regurgitant fraction and good ventricular function or (2) a diseased LV with a depressed ejection fraction. One must use all available clinical information to make this critical distinction.

Mid-diastolic Murmur

The S3 in mitral regurgitation often is followed by a short mid-diastolic rumble or murmur (MDM) that represents reverberations from rapid and excessive filling of a distended left ventricle (Figs. 17-4C, 17-6B). *Practical*

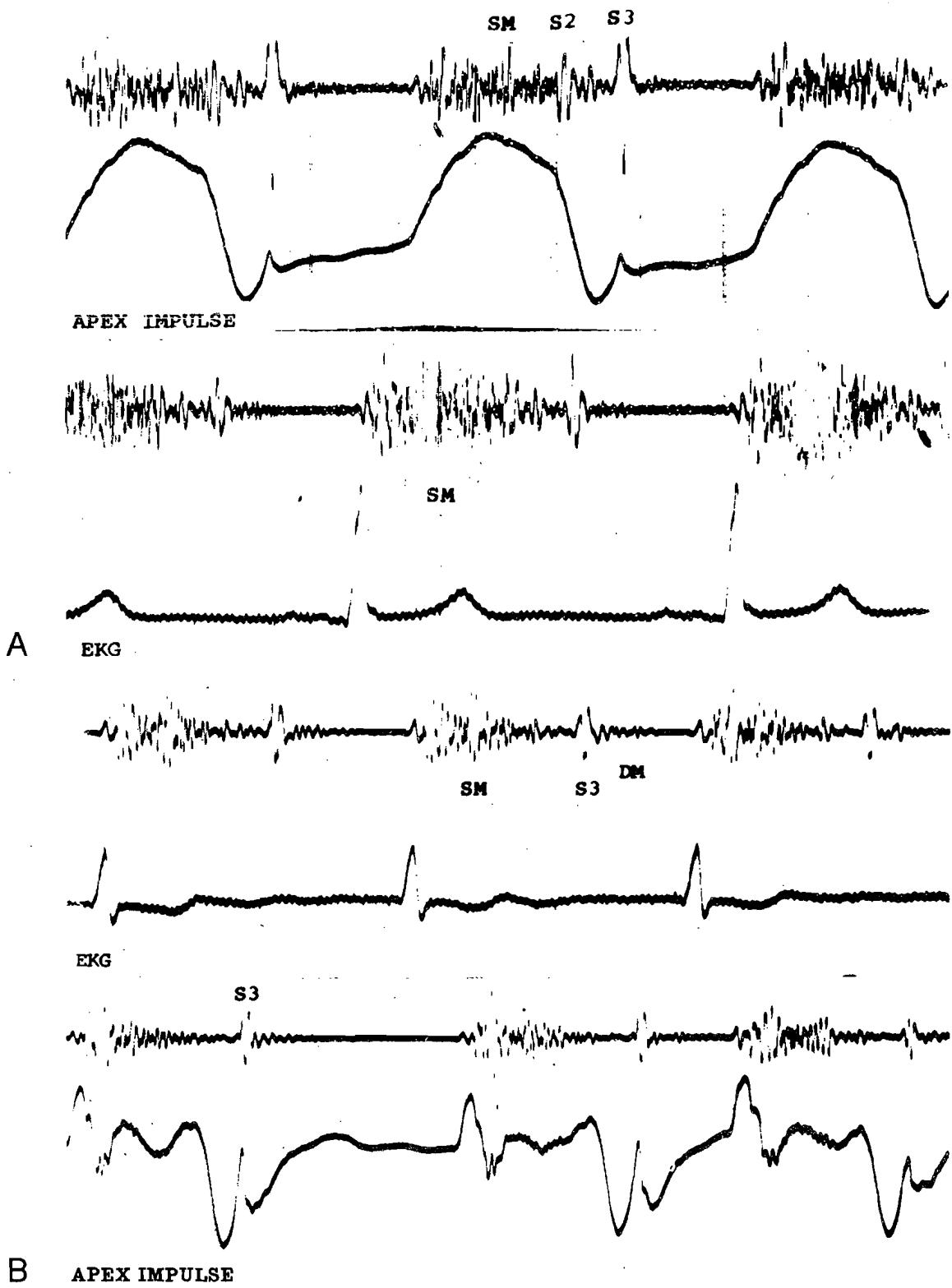


FIG. 17-6. Third heart sound in mitral regurgitation. A. A very loud S3 is recorded in early diastole, coinciding with the peak of the rapid filling wave on the apex cardiogram. The holosystolic murmur (SM) of mitral regurgitation is present. B. Medium frequency vibrations of a short mid-diastolic murmur (DM) are noted following a loud S3 in this patient with mitral regurgitation. The peak of the rapid filling wave of the apex cardiogram coincides with the S3.

Point: The presence of an S3 and mid-diastolic murmur indicates severe mitral regurgitation in the absence of coexisting mitral stenosis.

The MDM is a brief, low-medium pitched apical diastolic rumble best heard with the bell of the stethoscope when the patient is in the left decubitus position. It should be carefully sought in all patients with mitral regurgitation. It is critical to focus on the timing and the length of the MDM as it is readily confused with the diastolic murmur of mitral stenosis. In pure mitral regurgitation, the MDM does not extend into late diastole. When the heart rate is rapid, the S3 and MDM readily stimulate the physical findings of mitral stenosis; the S3 is confused with an opening snap, and the murmur is mistaken for the diastolic rumble across the stenotic valve. In resolving this differential, the quality of the S3 and the length of the murmur are important. If related to mitral regurgitation, the S3 will be a low-pitched thud and the MDM will end well before S1. Carotid message may be used to slow the heart rate to help make a more accurate auscultatory judgment.

Fourth Heart Sound

An S4 of LV origin is never a feature of rheumatic mitral valve disease. The left atrium in chronic mitral regurgitation of any etiology is dilated and compliant and is unable to generate an atrial sound or S4. This is in contradistinction to acute mitral regurgitation where the normal sized atrial chamber has a very high pressure and increased volume; the left atrium contracts more forcefully than normal, typically producing an audible S4 (see pages 344, 350). A right ventricular S4 may be present in patients with severe pulmonary hypertension who are in sinus rhythm; inspiratory augmentation and maximal intensity at the lower sternal edge suggest an S4 of right-sided origin.

Opening Snap

Contrary to popular belief, an opening snap occasionally may be present in patients with pure rheumatic mitral regurgitation. This probably relates to thickened, stiff, and distorted mitral leaflets. The opening snap coincides with the opening motion of the anterior leaflet. In such cases, presumably the anterior cusp produces the opening sound as it rapidly swings open in early diastole. Phonocardiographic studies have documented a recordable opening snap in patients with mitral regurgitation, although the sound itself is difficult or impossible to hear. Obviously, the presence of an audible opening

Note that the systolic murmur (SM) tapers in late systole, suggesting severe mitral regurgitation. (From Hultgren HN, Hancock EW, and Cohn KE: Auscultation in mitral and tricuspid valvular disease, Prog Cardiovasc Dis, 10:298, 1968.)

snap raises the question of coexisting mitral stenosis. In such cases, the intensity of S1 and the presence and length of the diastolic rumble require special attention. Auscultation in these patients may be confusing, as a separate S3 is often audible as well. Characteristically, the opening snap is best appreciated at or near the lower left sternal border, is a crisper sound than the S3, and is medium to high frequency. As the stethoscope is moved closer to the apex, the S3 is heard. It occurs later in diastole and is lower pitched. The S3 optimally (and often only) will be heard with the bell of the stethoscope; the diaphragm is best for hearing an opening snap. The presence of both sounds is a definite possibility, and the clinician must be able to identify each separately.

Murmur of Rheumatic Mitral Regurgitation

The typical murmur of mitral regurgitation is a constant amplitude, systolic murmur beginning immediately with S1 and extending to S2. It is usually of medium-high frequency, best heard at the LV apex, and radiates clearly into the left axilla. While there are frequent variations of the murmur, this classic description has withstood the test of time.

Length and Shape. In rheumatic mitral regurgitation, there is usually a central jet of blood refluxing into the left atrium and producing a sound vector that extends posteriorly and laterally. Because of the large pressure gradient between the left ventricle and left atrium throughout systole, regurgitation begins during isovolumic systole and extends into the isovolumic relaxation phase. Thus, the murmur is typically holo- or pansystolic with an even intensity throughout systole (Figs. 17-3, 17-7A). Alterations in the length of the cardiac cycle, such as occurs in atrial fibrillation or with PVCs, typically do not alter the murmur characteristics of length or loudness, in contradistinction to the murmur of aortic stenosis.

In auscultation of systolic murmurs, concentration on the last third of systole is of great importance. Does the murmur extend to S2? Is there a sound gap or murmur dropout before A2? *Practical Point: In almost all cases of mitral regurgitation, sound vibrations in late systole continue until S2, although the murmur may have a variable contour.* Occasionally, the murmur of mitral regurgitation appears to begin just after S1. This may be a result of masking of audible sound by S1 or very low amplitude murmur vibrations in early systole. Phonocardiography can be utilized effectively in questionable cases to document the presence or absence of sound vibrations immediately following S1 and immediately preceding S2.

The murmur of rheumatic mitral regurgitation is not always plateau in intensity and may taper or augment in late systole. The most common variant is mid-to-late systolic accentuation of the murmur. There may be a fanning out of sound vibrations during the last third of systole (Fig. 17-7B). Such a

murmur, similar to that of papillary muscle dysfunction or mitral valve prolapse, is typical of mild mitral reflux. In more severe mitral regurgitation, the murmur is more likely to have midsystolic accentuation, giving it a spindle-shaped or ejection quality (Fig. 17-7C). In all these situations, careful auscultation usually will identify sound vibrations at the *beginning* and at the *end* of systole, which confirm that the murmur is truly regurgitant in quality as opposed to a more common systolic ejection murmur.

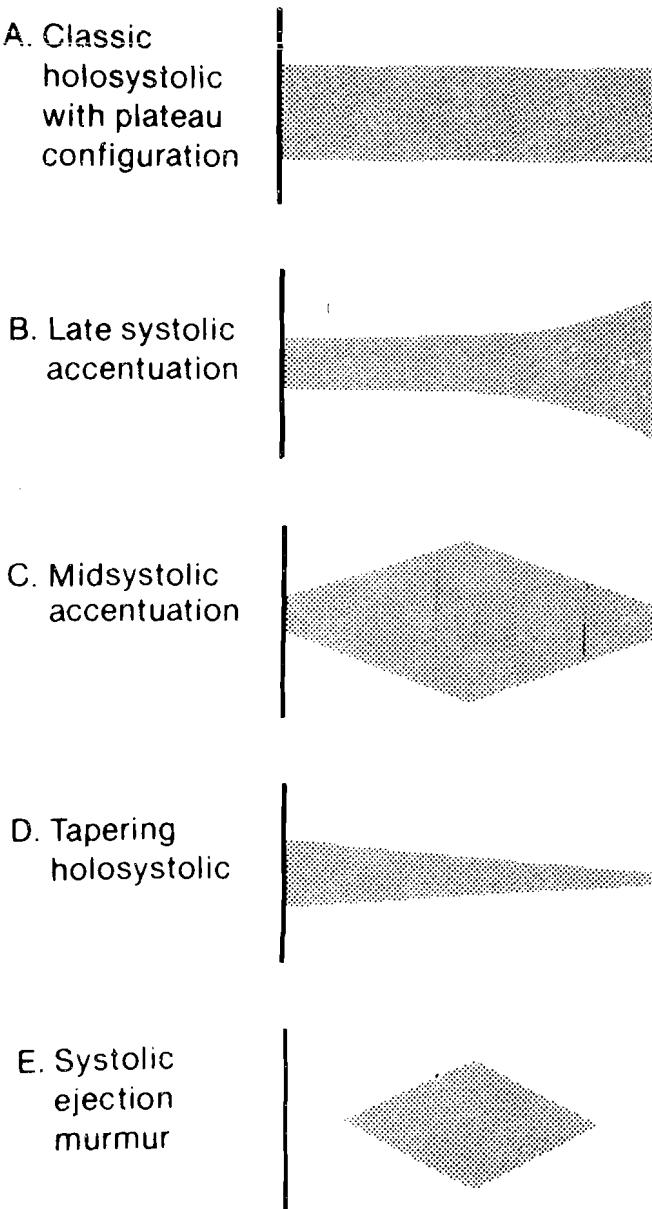


FIG. 17-7. Variable contours of the murmur of mitral regurgitation. A through D represent holosystolic murmurs with differing configuration (see text); note that sound vibrations extend to S2 in each example. E. A typical systolic ejection murmur is shown for comparison purposes. Note the sound-free interval immediately following S1 and, most importantly, at the end of the murmur. (From Abrams J: Prim Cardiol, 1983.)

The least common variant of the mitral regurgitation murmur is one that tapers or decreases in intensity in late systole (Fig. 17-7D, Table 17-3). This configuration is also more likely to be found in trivial degrees of mitral regurgitation; this murmur is usually very soft. In such cases, the major question for the observer is whether the murmur is truly holosystolic. This may be exceedingly difficult to resolve. In severe, recent onset, mitral regurgitation the murmur often is decrescendo in late systole due to the enormous left atrial V wave and a decrease in late systolic reflux into the atrium (Fig. 17-8). This murmur may take on ejection characteristics (Fig. 17-7E). Such attenuation in late systole does not occur in chronic rheumatic mitral regurgitation. The murmur of mitral regurgitation may wane in late systole if the degree of the reflux decreases during the last third of systole due to a large left ventricle with foreshortened chordae tendinae. The smaller cavity size in late systole allows more complete coaptation of the mitral leaflets, and the degree of late systolic regurgitation is diminished.

Intensity. In general, the louder the murmur of rheumatic mitral regurgitation, the greater the degree of reflux. Patients with a moderate to severe mitral leak and a large regurgitant fraction usually have grade 3-4/6 systolic murmurs. Thus, systolic apical thrill is common in severe mitral regurgitation. However, there are important exceptions to this rule; certain conditions predispose to a murmur of decreased amplitude even when the regurgitation is major. On occasion, the murmur may be barely audible in the setting of severe regurgitation. *Practical Point:* *The most important factor affecting murmur intensity is the state of left ventricular function. With preserved LV contractility and vigorous systolic function, the velocity and volume of blood flow is high and the murmur is loud. If LV dysfunction occurs and the LV ejection fraction falls, the murmur will become softer even though the degree of mitral incompetence remains unchanged.* Many patients with severe mitral regurgitation and a soft murmur are in overt congestive heart failure. With appropriate medical therapy, LV function may improve; end-diastolic volume decreases, ejection fraction increases, and the murmur becomes louder. When there is profound pump failure and end-state mitral regurgitation, the systolic murmur may be virtually inaudible. This situation is more likely to occur in acute, (nonrheumatic) mitral regurgitation, such as severe papillary muscle dysfunction or rupture in acute myocardial infarction in association with marked depression of myocardial performance.

TABLE 17-3 *Causes of Late Systolic Tapering of the Murmur of Mitral Regurgitation*

-
- Mild or trivial degree of mitral regurgitation
 - Recent or acute onset mitral regurgitation (giant V wave)
 - Severe mitral regurgitation with relatively small left atrium (giant V wave)
 - Improved mitral valve coaptation resulting from decreased LV cavity size in late systole
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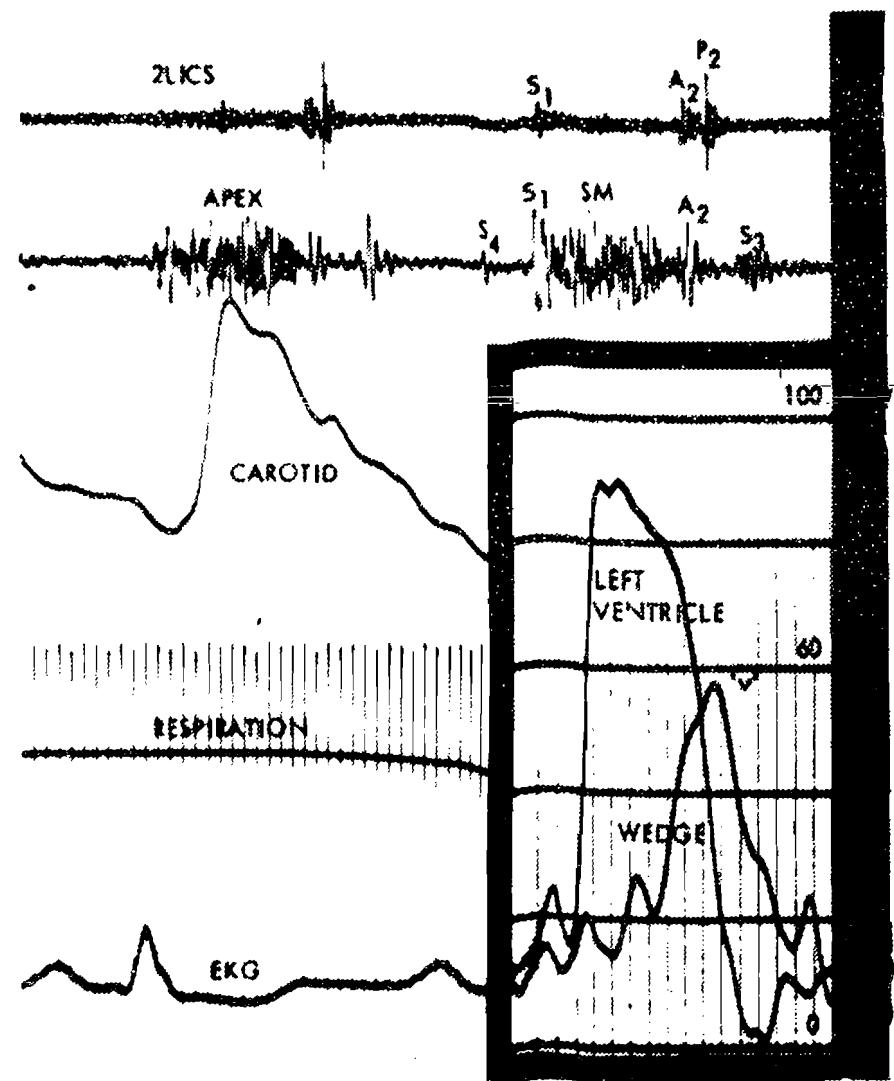


FIG. 17-8. Decrescendo configuration of the systolic murmur in severe mitral regurgitation. This postinfarction patient has mitral regurgitation and congestive heart failure. Cardiac catheterization (lower right) reveals an enormous V wave in the pulmonary capillary wedge tracing, consistent with severe mitral regurgitation. Because of the rapidly diminishing pressure gradient between the left ventricle and the left atrium in late systole, the murmur decreases in intensity during the last part of systole. Thus, the murmur of severe mitral regurgitation, particularly when of recent onset, may take on ejection characteristics. Note the presence of an S3 and S4, common in severe acute mitral regurgitation. (From Reddy PS, Shaver JA, and Leonard JJ: Cardiac systolic murmurs: pathophysiology and differential diagnosis. *Prog Cardiovasc Dis* 14:1, 1971.)

Table 17-4 lists conditions in which the intensity or loudness of the mitral regurgitation murmur will be decreased out of proportion to the degree of reflux. Concomitant mitral stenosis is often associated with a huge left atrium that dilutes the regurgitant volume. In addition, there may be selective streaming of the regurgitant jet away from the posterior-lateral aspect of the left atrium due to marked distortion of the mitral valve orifice.

TABLE 17-4 *Associated Conditions That May Decrease the Intensity of Mitral Regurgitation Murmur*

Congestive heart failure or left ventricular dysfunction
Low cardiac output states
Associated mitral stenosis
Huge left atrium
Large right ventricle
Obesity*
Chronic obstructive lung disease*
Thick muscular chest*

*Tendency for all intracardiac sound to be diminished.

The most common cause of a soft murmur is a trivial degree of mitral regurgitation. When there is a faint murmur, a decision must be made as to whether *all* cardiac sounds are decreased in intensity or just the murmur. The intensity of the murmur of mitral incompetence does not vary with changes in cycle length (e.g., in atrial fibrillation, post-PVC beats). Typically, the murmur retains its baseline loudness, irrespective of cycle length. This is in contradistinction to ejection or flow murmurs that augment in intensity following a long diastolic filling period.

Frequency or Pitch. In mild degrees of mitral reflux, the left ventricular-left atrial gradient is high, the amount of regurgitant blood is relatively small, and the flow velocity is high; the resultant murmur is high pitched (greater than 200 cycles per second). Firm pressure with the diaphragm of the stethoscope routinely should be used in auscultation for mitral regurgitation. An apical, high-pitched, holosystolic murmur is a common finding in young persons with mild mitral regurgitation who have a prior history of acute rheumatic fever. Typically, on physical examination no other abnormalities will be present.

With increasing degrees of reflux the left atrial "V wave" increases in size as the gradient between the left ventricle and the left atrium decreases. This change produces a murmur with lower frequency vibrations. In many patients with a substantial degree of mitral regurgitation, the murmur has mixed frequencies, reflecting both a large gradient and high flow. The typical mitral regurgitation murmur has a whirring, somewhat musical pitch. In severe mitral regurgitation, the murmur may be harsh in quality.

Location and Radiation. Mitral regurgitation of any etiology is best appreciated at the left ventricular apex, although on unusual occasions the murmur will be better heard elsewhere over the chest. In some individuals, the murmur may be louder just inside the apex impulse; in tall thin subjects with small hearts, the murmur and apical impulse can be quite medial, near or adjacent to the left sternal edge. The murmur of chronic, rheumatic mitral regurgitation rarely is best heard away from the PMI. However, in some cases of isolated rupture of chordae tendinae with selective or dominant

incompetence of the posterior leaflet, the murmur may actually be maximal at the first or second left interspace (see page 345). The classic rheumatic murmur radiates leftward into the axilla and often is well heard at or beneath the left scapula in the posterior chest. When the left atrium is large, the murmur may be audible over the thoracic vertebral column.

Left Decubitus Position: *Practical Point: Auscultation in the left recumbent position (Fig. 16-3) will usually augment the murmur of mitral regurgitation and often increase its intensity by one to two grades. In addition, this maneuver occasionally accentuates the holosystolic nature of a late tapering murmur. Use this position routinely when an apical systolic murmur is of low amplitude and long duration.* Differentiation of a functional or ejection systolic murmur that radiates well to the apex from a truly holosystolic murmur often can be resolved easily by use of the left decubitus position.

Occasionally, loud mitral regurgitant murmurs radiate rightward and are heard readily at the lower sternal border. These murmurs may occur in patients with large left ventricles; in such cases, one must be sure there is not coexisting tricuspid regurgitation. With marked enlargement of the right ventricle, the palpable apex beat may not be formed by the LV and the mitral regurgitation murmur may be heard best in the left axilla *lateral* to the apparent apical impulse. With a markedly dilated LV, the murmur also may be louder in the axilla than at the apex. In patients with small hearts, the murmur of mitral regurgitation may be quite medial, with poor radiation into the axilla due to interpositioning of the lungs. Unless there is prominent posterior leaflet incompetence, the murmur of mitral regurgitation does not radiate well to the base of the heart.

The murmur of mild mitral regurgitation is not only soft but often well localized to a small area of the precordium at or near the apex. Selective listening or tuning in for this faint, high-pitched murmur is important. Auscultation while the patient's breath is held in mid- or end-expiration is helpful in identifying these soft murmurs. Often, it is useful to have the patient sit up and lean forward while the examiner listens at the apex during held-expiration. This maneuver is also used for detection of the faint, high-pitched murmur of aortic regurgitation.

Mid-diastolic Rumble

A brief diastolic, low frequency rumble at the apex, following a loud S3, is common in severe mitral regurgitation (Figs. 17-3, 17-4, 17-6B), and has already been discussed (page 319). It is important to remember that such a murmur does not necessarily indicate associated mitral stenosis.

This murmur is best heard using light pressure with the bell of the stethoscope precisely positioned on the apex beat. The left recumbent position is essential, as often the mid-diastolic murmur cannot be detected at any

other site or position. This murmur may begin with an intense accentuation representing the S3 occurring at the peak of rapid filling of the ventricle. The genesis of this murmur is similar to that produced by an excessive blood flow crossing the A-V valves in patients with large left-to-right shunts.

Assessment of Severity

Table 17-5 lists various findings on physical examination that suggest hemodynamically significant chronic mitral regurgitation.

Differential Diagnosis of the Systolic Murmur of Mitral Regurgitation

The murmur of mild mitral regurgitation can be mistaken readily for an *ejection murmur* with apical radiation, particularly if the mitral murmur is decrescendo in quality. The critical question relates to the *length* of the murmur in late systole. An apical murmur that is crescendo or fans out to S2 is invariably some form of mitral regurgitation. Attention to post-PVC beats may be of aid in this differential diagnosis; ejection murmurs will augment in intensity, whereas the loudness of the mitral regurgitation murmur will remain unchanged.

Tricuspid regurgitation may be a difficult problem to separate from mitral regurgitation when there is significant right ventricular enlargement. In this setting, the apical murmur may actually arise from the right heart and reflect tricuspid regurgitation. Careful attention to respiratory variation is essential. The tricuspid murmur usually, but not always, increases during inspiration, but the murmur of mitral regurgitation typically softens with inspiration. Unfortunately, some tricuspid regurgitation murmurs do not vary with respiration. If there is associated atrial fibrillation, respiratory changes in murmur intensity may be virtually indetectable.

A ventricular septal defect murmur can simulate mitral regurgitation, although its maximal location at the lower sternal border should readily differentiate the two. However, in patients with small hearts or long chests,

TABLE 17-5 Bedside Clues to Hemodynamically Severe Chronic Mitral Regurgitation

-
- Rhythm: Many patients are in atrial fibrillation
 - Carotid pulse: Small volume, quick-rising
 - Left ventricular impulse: Hyperdynamic, forceful, often displaced to left. May be sustained.
Systolic apical thrill common
 - Parasternal impulse: Early or late systolic (in absence of mitral stenosis)
 - S2: Widely split; increased P2
 - S3: Present—may be palpable. Mid-diastolic “flow” rumble often audible
 - Murmur: Usually very loud. If CHF present, murmur often becomes louder after successful therapy.
-

the mitral regurgitation murmur can be quite medial. In all three causes of a holosystolic murmur (mitral regurgitation, tricuspid regurgitation, ventricular septal defect), the murmur may be decrescendo in late systole and can simulate a systolic ejection murmur. Mitral regurgitation, as opposed to the other lesions, typically is *best* heard at the apex and usually will have some radiation to the axilla.

In *hypertrophic cardiomyopathy*, mitral regurgitation is commonly present. A long systolic ejection murmur is typical in these patients, and it is often difficult to be sure that there is a *separate* murmur of mitral regurgitation. The typical radiation pattern of the regurgitant murmur often is absent in hypertrophic cardiomyopathy. Careful assessment of the response of the systolic murmur to positional changes, maneuvers, and pharmacologic agents may be helpful (see Chapter 11, Table 14-1).

PAPILLARY MUSCLE DYSFUNCTION

In the early 1960's, Dr. George Burch first focused attention on the role of the papillary muscles in mitral valve closure and pointed out that a wide variety of conditions leading to fibrosis, ischemia, or contractile dysfunction of the papillary muscles and the surrounding left ventricular muscle can result in mitral regurgitation. Subsequently, papillary muscle dysfunction has been recognized to be responsible for most cases of mitral regurgitation associated with coronary artery disease.

PATHOPHYSIOLOGY

Normal function of the papillary muscles helps prevent the mitral valve leaflets from protruding into the left atrium during systole. As the size of the left ventricular cavity decreases during ejection, the mitral valve cusps and chordae are held firmly in place by the two contracting papillary muscles (Fig. 17-9A). Tension generated by the papillary muscles is maximal in *late systole* when LV dimensions are smallest, as these specialized muscle bodies maximally tauten and stabilize the mitral supporting apparatus during leaflet coaptation. When abnormal structure or function alters papillary muscle contraction, the mitral leaflets may prolapse, resulting in reflux of blood into the left atrium typically during the last half of systole (Figs. 17-9B, C). Thus, the classic murmur of papillary muscle dysfunction is a *late systolic crescendo murmur* extending up to S2 (Fig. 17-10A). When there is severe contractile dysfunction and/or marked fibrosis and shortening of the papillary muscles, the resultant mitral regurgitation may be *holosystolic*; however, in such situations the murmur often retains late systolic accentuation (Fig. 17-10B). Papillary muscle murmurs often have midsystolic accentuation and take on

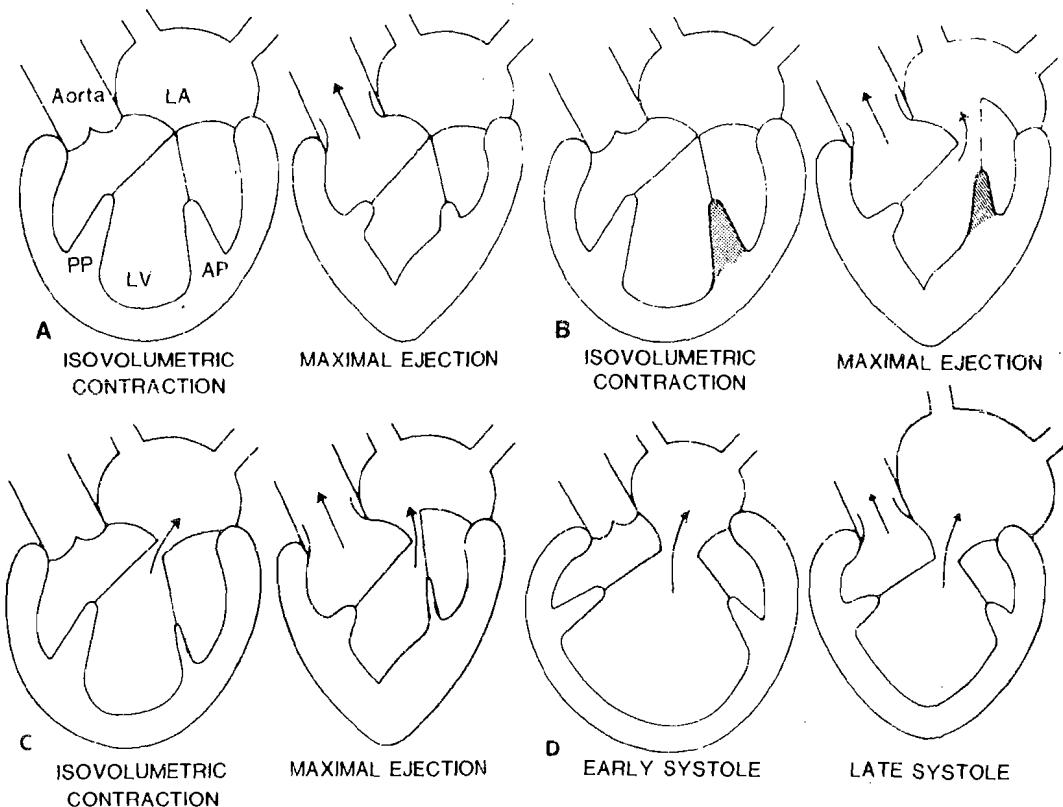


FIG. 17-9. Mechanisms of mitral regurgitation caused by papillary muscle dysfunction. A. Normal heart. The papillary muscles contract during systole as left ventricular cavity size diminishes. An intact mitral apparatus prevents reflux of blood from the ventricle into the left atrium. B. Papillary muscle dysfunction. In this diagram, the posterolateral papillary muscle is functionally abnormal (ischemia or infarction) and is unable to maintain valvular apposition during peak systole when left ventricular cavity size is at its nadir. The mitral leaflet(s) protrudes into left atrium and mid-late mitral regurgitation occurs. The mitral valve typically remains competent in early systole; therefore, there is no murmur during the first third of systole. C. Papillary muscle scarring. In this example, the papillary muscle is fibrotic and/or atrophic, such that even during isovolumic systole it is unable to prevent reflux of blood into the left atrium because of an inadequate tethering effect on the chordae tendineae. The mitral regurgitation may worsen during ejection. D. In this example of a markedly dilated left ventricle, the papillary muscles are displaced laterally and lose their normal alignment within the left ventricular cavity. This may result in inability of the mitral valve to adequately coapt because of abnormal tension on and displacement of the mitral valve apparatus. The resultant murmur of mitral regurgitation is believed to be related to distortion of papillary muscle-ventricular anatomy rather than true dysfunction. (From Burch GE, DePasquale MP, and Phillips JH: The syndrome of papillary muscle dysfunction. *Am Heart J* 75:399, 1968.)

ejection characteristics (Fig. 17-10C). This was the configuration that Burch first described.

Myocardial ischemia is the most common cause of papillary muscle dysfunction. Because these structures are further from the coronary ostia than any other portion of the left ventricle, coronary blood flow to the papillary muscles is at increased jeopardy. The posteromedial papillary muscle is especially vulnerable because its blood supply usually is scant and its collateralization is poorer than that of the anterolateral papillary muscle. Animal

THE SYSTOLIC MURMUR OF PAPILLARY MUSCLE DYSFUNCTION

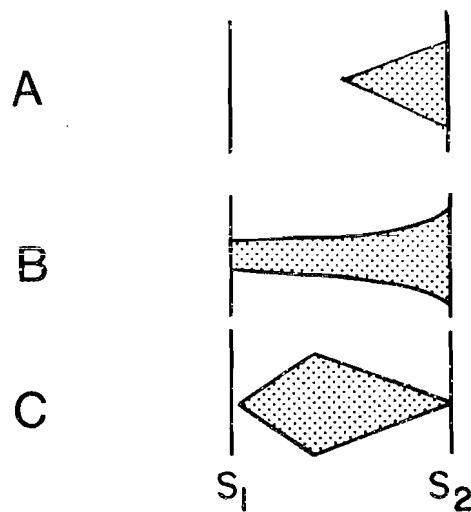


FIG. 17-10. Configuration of papillary muscle murmurs. (See also Fig. 22-2.) A. The classic late systolic contour. The mitral valve leaflets remain competent in early systole. B. A holosystolic murmur with late systolic peaking or accentuation. In this example, the papillary muscle is unable to sustain valve competency throughout systole; when the ventricle reaches its minimal size, mitral reflux increases and the murmur becomes louder. C. An ejection-shaped murmur may occur due to a very large left atrial V wave resulting in a late systolic decrease in mitral regurgitation (see Fig. 17-8). Another proposed mechanism for the decrescendo murmur of papillary muscle dysfunction is improvement in late systolic leaflet coaptation due to the decrease in late systolic left ventricular cavity size.

experiments suggest that the left ventricular muscle *surrounding* the papillary muscles must be diseased or ischemic for significant mitral regurgitation to occur. In humans, the papillary muscles *and* adjacent left ventricle are often found to be scarred or necrotic at autopsy. The murmur of papillary muscle dysfunction commonly may be heard during acute myocardial infarction and rarely may be observed during attacks of angina pectoris.

Patients with considerable fibrosis in the left ventricle from any cause frequently have a mitral regurgitation murmur that may result from abnormal papillary muscle function. Severe LV wall motion abnormalities or an overt ventricular aneurysm is likely to involve one of the papillary muscles (the posteromedial most commonly), resulting in mitral regurgitation.

Many different disease processes can cause scarring or damage to the papillary muscles (Table 17-6). Myocardial fibrosis can result from severe left ventricular hypertrophy of any cause. Moderately severe left ventricular dilatation also may result in mitral regurgitation related to the papillary muscles. In such patients, the papillary muscles are literally pulled laterally

TABLE 17-6 *Etiologies of Papillary Muscle Dysfunction*

Ischemic heart disease
Acute myocardial infarction
Angina pectoris (transient)
Healed myocardial infarction
Left ventricular aneurysm
Left ventricular fibrosis
Left ventricular dilatation
Congestive cardiomyopathy
Hypertrophic cardiomyopathy
Miscellaneous
Carcinoid heart disease
Vasculitis
Myocarditis
Endocardial fibroelastosis
Abscess
Trauma
Amyloid
Endocarditis

away from their normal alignment in the left ventricular cavity and lose the optimal angle for generation of wall tension, producing prolapse of the mitral valve leaflets (Fig. 17-9D). This proposed mechanism has been disputed by some experts. Mitral regurgitation is common in hypertrophic cardiomyopathy; in this condition, the papillary muscles probably contribute to the mitral regurgitation as a result of the disturbed spatial relationships of the left ventricular architecture.

Papillary Muscle Rupture. While acute transmural myocardial infarction rarely causes complete rupture of the body or tip of a papillary muscle, when this does happen, it typically involves the posteromedial muscle. In this situation or with severe combined necrosis and ischemia of the papillary muscle without actual rupture, the degree of mitral regurgitation is massive and usually fatal. A flail mitral leaflet results from this profound disruption of the mitral supporting apparatus. The resultant syndrome of acute mitral regurgitation (see page 348) is a true emergency and these patients can be saved only by surgical treatment.

PHYSICAL EXAMINATION IN PAPILLARY MUSCLE DYSFUNCTION

Carotid Pulse

There are no specific abnormalities of the carotid pulse. If papillary muscle dysfunction is associated with severely depressed left ventricular function and a low cardiac output, the amplitude of the pulse wave will be small and mechanical alternans may be detectable (see Chapter 3). If the mitral

regurgitation is severe, the carotid pulse may be quick-rising and of low amplitude producing a soft tapping impulse under the palpating finger.

Jugular Venous Pulse

No abnormalities of the venous pulse are present in papillary muscle dysfunction unless there is biventricular congestive failure in which case the mean venous pressure will be elevated.

Precordial Motion

Because the majority of patients with a papillary muscle dysfunction murmur have left ventricular dysfunction or dilatation due to underlying coronary artery disease, the apex impulse is often abnormal (Table 17-7). An ectopic precordial impulse may be present, suggesting left ventricular aneurysm or a severe localized abnormality of wall motion. More commonly, a palpable S4 (presystolic distension of the left ventricle) will be noted (Figs. 7-3, 7-6). If the mitral leak is large and left ventricular diastolic volume is excessive, a palpable rapid filling wave (S3) may be detected. A bifid apical thrust or a sustained or late systolic lift consistent with deranged LV function may be associated with the murmur of papillary muscle dysfunction. The left ventricular impulse commonly is sustained into the second half of systole in the supine or left recumbent position.

When there is severe mitral regurgitation, a late systolic parasternal impulse reflecting left atrial expansion in systole occasionally can be felt (Figs. 17-4B, 17-5). If there is significant pulmonary hypertension, there may be a sustained right ventricular lift, but major elevation of pulmonary artery pressure is uncommon unless chronic, severe, left ventricular dysfunction or a left ventricular aneurysm is present.

First Heart Sound

The first heart sound is soft, normal, or increased in papillary muscle dysfunction and therefore is of little diagnostic help. Cheng has emphasized

TABLE 17-7 *Precordial Motion Abnormalities Commonly Associated with Papillary Muscle Dysfunction*

-
- LV heave (sustained)
 - Bifid or double LV apical impulse
 - LV impulse displaced laterally
 - Ectopic LV impulse
 - Palpable S4
 - Palpable S3
 - Late parasternal lift
-

the frequent finding of an S1 of increased intensity in such patients, which is perhaps related to abrupt tautening of abnormally lax, elongated chordae tendineae as the mitral valve closes in late systole.

Second Heart Sound

The second sound is unremarkable in the majority of cases of papillary muscle dysfunction. S2 may show abnormally wide splitting if there is severe mitral regurgitation resulting in early aortic valve closure (see Chapter 6). On the other hand, ischemic dysfunction of the left ventricle or left bundle branch block may produce reversed splitting of S2 in the setting of a papillary muscle dysfunction murmur. If there is any degree of pulmonary hypertension, P2 will be increased in intensity.

Third Heart Sound

An audible and even palpable S3 is a common accompaniment of papillary muscle dysfunction. This sound may result from a large volume of blood crossing the mitral valve in diastole if the mitral regurgitant fraction is large. The S3 may also be related to left ventricular dysfunction and increased left ventricular diastolic pressure and volume; other evidence of heart failure is common in this situation.

Fourth Heart Sound

A prominent S4 or atrial gallop typically is found in association with papillary muscle dysfunction (Fig. 17-11). Underlying left ventricular dysfunction is associated with a high left ventricular end-diastolic pressure and increased myocardial stiffness. In this case, the force of left atrial contraction is enhanced. The S4 may be extremely loud and is often palpable in the left decubitus position. A soft or absent S4 may be brought out by isometric handgrip, which will also intensify the murmur of mitral regurgitation. *Practical Point:* *The absence of an S4 in the setting of a late systolic murmur suggests the murmur is not due to papillary muscle dysfunction.* An S4 rarely is audible in chronic mitral insufficiency due to a primary disorder involving the valve leaflets (e.g., rheumatic heart disease, floppy mitral valve).

Systolic Murmur

The hallmark of papillary muscle dysfunction is an apical, medium-high frequency systolic murmur that begins well after S1, usually in midsystole, and fans outward to S2 (Figs. 17-10A, 17-11A). The murmur is typically soft, usually of grade II/VI intensity, and may wax and wane with serial observation.

Shape and Duration. Papillary muscle murmurs have been documented by phonocardiography to have many configurations, which include early decrescendo, midsystolic, pansystolic, and late systolic (Fig. 17-11). This variability probably relates to differing degrees of mitral reflux, left ventricular cavity size, and ventricular function. *Practical Point: The classic murmur configuration of papillary dysfunction has its onset in early to midsystole, with sound vibrations extending up to S2. The murmur may or may not appear to begin with S1. If left ventricular contractility is preserved, the murmur actually*

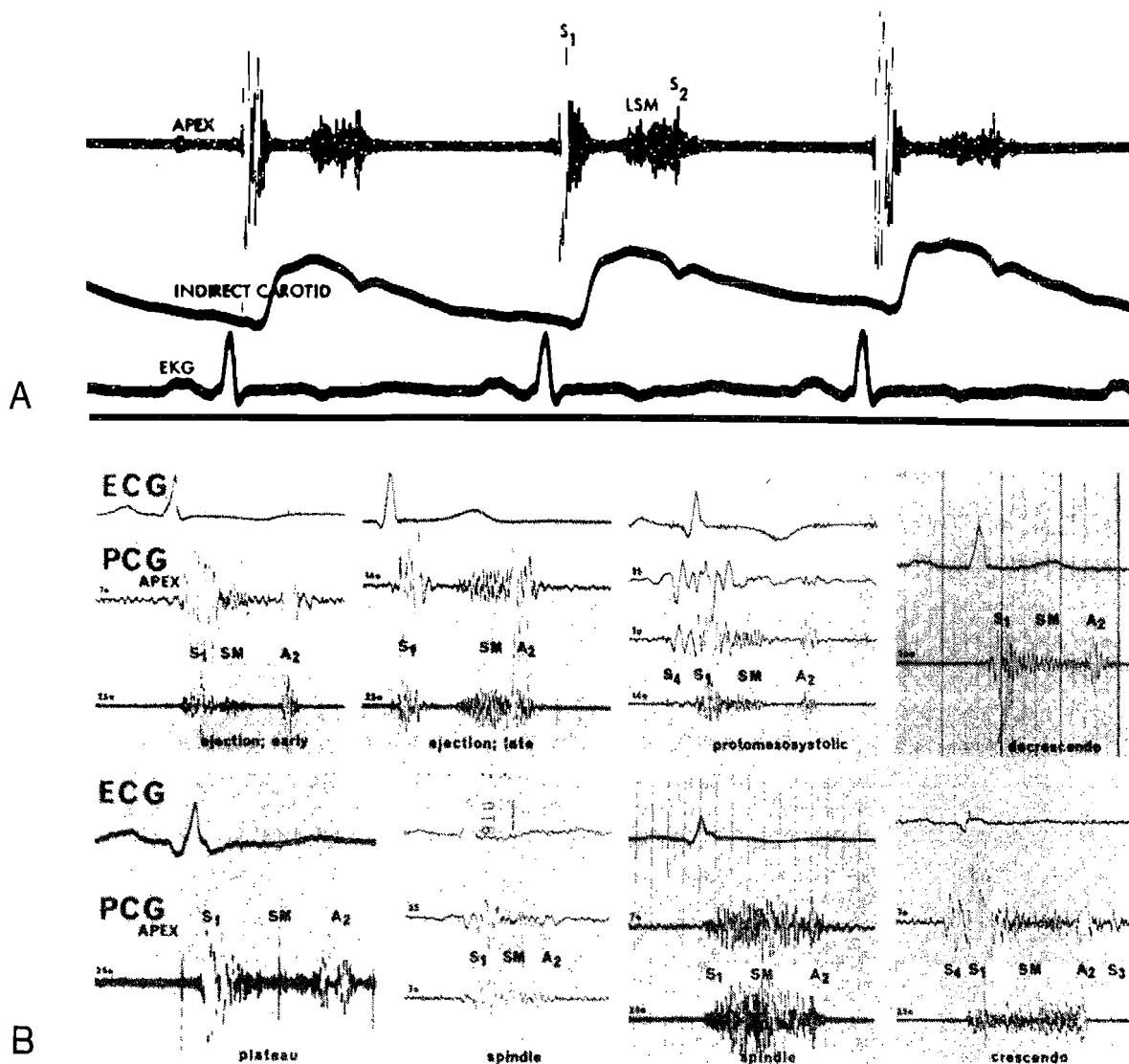


FIG. 17-11. Examples of murmurs of papillary muscle dysfunction. A. The classic late systolic murmur in a patient with coronary artery disease. This subject had mild mitral regurgitation at angiography. (From Reddy PS, Shaver JA, Leonard JJ: Cardiac systolic murmurs: pathophysiology and differential diagnosis. Prog Cardiovasc Dis 14:1, 1971.) B. Papillary muscle dysfunction murmurs in acute myocardial infarction. Note that most murmurs are holosystolic although their contour varies. Some have an early ejection contour, but others peak in late systole. In general, the more severe the papillary muscle dysfunction, the greater the degree of mitral regurgitation and the longer the murmur. The murmur of papillary muscle dysfunction in acute myocardial dysfunction often varies from day to day. (From Heikkila J: The fate of mitral valve complex in acute myocardial infarction. Ann Clin Res 3:386, 1971.)

can be decrescendo in late systole as LV cavity size decreases and the degree of late systolic mitral reflux diminishes. Frequently, the configuration of the murmur varies, being ejection quality one day and holosystolic the next. If the degree of mitral regurgitation is moderate to severe, the murmur will be holosystolic. Major mitral regurgitation due to papillary muscle dysfunction produces a pansystolic murmur that often has late systolic accentuation (Fig. 17-10B) but may be diamond shaped (Figs. 17-7C, 17-10C). Some authors have emphasized an apparent gap between S1 and the onset of the systolic murmur of papillary muscle dysfunction; nevertheless, careful auscultation in the supine and left lateral positions often demonstrates the holosystolic nature of such a murmur. It is important to concentrate on both the *beginning* and the *end of systole* to accurately assess the murmur's onset and cessation.

Intensity and Frequency. The murmur of papillary muscle dysfunction is usually not prominent, although with severe mitral regurgitation (as with papillary muscle rupture) the murmur can be quite loud. Considering the actual degree of the valvular leak, commonly the murmur is quite soft, particularly in acute myocardial infarction where the pump function of the ventricle may be profoundly depressed. In papillary muscle dysfunction, the relationship between murmur intensity and the severity of mitral regurgitation typically is poor. Most likely this is due to the coexistence of serious abnormalities of left ventricular contraction. Cases of truly silent, severe papillary muscle dysfunction or rupture with major mitral regurgitation have been documented. *Practical Point: Be careful not to underestimate the potential hemodynamic significance of a soft papillary muscle dysfunction murmur in acute myocardial infarction. On occasion, profound mitral regurgitation can exist with a very faint (Grade I-II/VI) systolic murmur.*

As left ventricular function improves, the murmur of papillary muscle dysfunction either may disappear or may get louder as contractile force and ejection fraction increase. During a PVC, usually the murmur intensity increases but softens in the post-PVC beat, a variation that is contrary to that of the systolic murmur of aortic stenosis or hypertrophic cardiomyopathy with a PVC. *Practical Point: Variability in the amplitude of the murmur is a common feature of papillary muscle dysfunction.*

The murmur of papillary muscle dysfunction usually is medium pitched, pure frequency in tone, and often has high frequency components. Because these murmurs typically are soft, firm pressure with the diaphragm of the stethoscope is essential for optimal auscultation.

Location and Radiation. Usually, the murmur of papillary muscle dysfunction or rupture is maximal at the apex. In a small percentage of patients, the murmur is best heard at the lower left sternal border. Radiation patterns of the low intensity papillary muscle murmurs are usually unremarkable.

Papillary Muscle Dysfunction in Acute Myocardial Infarction. Careful, repetitive auscultation of patients with acute, transmural myocardial

infarction will reveal an apical systolic murmur in 30 to 50% of patients (Fig. 17-11B). Both the intensity and shape are likely to change during the course of hospitalization. While often this murmur is ejection in nature at first, repeated listening will reveal holosystolic and/or late systolic vibrations. As a result of the decreased stroke volume and depressed ejection velocity that occurs in most patients with acute myocardial necrosis, typically a low frequency and low amplitude murmur is heard. *It is of great importance to listen for this murmur in a very quiet setting or it will be easily missed.* In the case of a patient with an acute infarction, development of a new systolic murmur is common and is likely to be heard within the first few days after admission. Infrequently the murmur will retain its ejection quality, or it may have a purely late systolic accentuation that implies mild mitral regurgitation. The murmur of severe mitral regurgitation can be loud but is rarely associated with a thrill. More commonly the murmur of severe papillary muscle dysfunction is relatively soft.

Papillary Muscle Rupture. This devastating but rare complication of acute myocardial infarction produces acute, severe mitral regurgitation. The murmur may be silent or very soft or can be quite loud and harsh. An S4 and S3 are common, and the patient is likely to be in acute pulmonary edema with or without hypotension. Late systolic tapering of the murmur may be present as a result of a decreasing left ventricular-left atrial pressure gradient in end-systole produced by a huge left atrial V wave (Figs. 17-7D, 17-8).

Mitral Regurgitation Second to Left Ventricular Dilatation. Patients with cardiac decompensation or massive cardiomegaly often have a murmur of mitral regurgitation in the absence of intrinsic disease of the mitral valve. Frequently, the holosystolic murmur disappears or softens after treatment of congestive heart failure and subsequent clinical improvement. This murmur has been described as *functional* mitral regurgitation. Many older textbooks and articles state that in such patients the incompetent mitral valve results from dilatation of the mitral annulus. More recent descriptions recognize that the mitral annulus does not always enlarge its circumference even in severe left ventricular enlargement; actually the maximal left ventricular dilatation is usually at midchamber level. The voluminous surface area available for mitral leaflet coaptation seems to preclude regurgitation produced solely from modest dilatation of the mitral annulus. In a patient with myxomatous degeneration or the floppy valve syndrome, the mitral ring or annular size consistently is found to be dilated. Recent studies of patients with cardiomegaly are conflicting, and some data suggest that mitral annular dilatation is a factor in some patients with mitral regurgitation due to a dilated left ventricle.

The precise mechanisms for mitral regurgitation in dilated hearts remain controversial. Initially, Burch suggested that mitral regurgitation may result from malalignment and lateral migration of the two papillary muscles in

patients with large ventricles, with inadequate ability to tether the chordae leaflets at the appropriate tangent to the mitral valve orifice (Fig. 17-9D). The papillary muscles themselves may remain normal. In many patients with significant left ventricular enlargement, the left ventricular cavity becomes more spherical or globular; the circumference of the dilated left ventricle at midventricular level usually is larger than the annular circumference. The subsequent loss of normal papillary muscle geometry with respect to the long axis of the left ventricle could result in failure of the mitral valve apparatus to cause complete valve closure.

In these patients, the physical examination is similar to that of classic papillary muscle dysfunction; in addition to the holosystolic murmur, abnormalities of left ventricular precordial motion are likely to be found. Although this murmur may have late systolic accentuation, it typically fills systole.

ABNORMALITIES OF THE CHORDAE TENDINEAE

The multiple thin chordae tendineae supporting the mitral valve become abnormally stretched and attenuated in various conditions, and rarely a chord may actually rupture. The resultant mitral regurgitation ranges from mild to severe depending on the number and location of affected chordae, the pre-existing status of the mitral valve, and the size and function of the left ventricle. The chordae to one or both leaflets may be involved. Often the supporting struts to one or two scallops of the posterior leaflet will rupture. If the ruptured chord is distal to the main truncal chordae and is attached directly to the valve, the resultant valve deformity will be minimal. However, rupture of a proximal first or second order chord (Fig. 17-12) will cause major instability of the mitral cusp and significant mitral regurgitation.

Table 17-8 lists conditions that have been associated with ruptured chordae tendineae. In patients with the floppy mitral valve syndrome, mitral regurgitation may result from severe prolapse of the mitral leaflets caused by

TABLE 17-8 Causes of Ruptured or Severely Stretched Chordae Tendineae

Idiopathic (spontaneous)
Infective endocarditis—acute and healed
Blunt chest or cardiac trauma
Myxomatous degeneration of the mitral valve ("floppy valve syndrome")
Marfan's syndrome
Previous myocardial infarction
Acute myocardial infarction with rupture of tip of the papillary muscle and chordae tendineae (rare)
Rheumatic mitral valvulitis
Vigorous physical effort (?)

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The Normal Left Ventricular Papillary Muscle and its Chordae

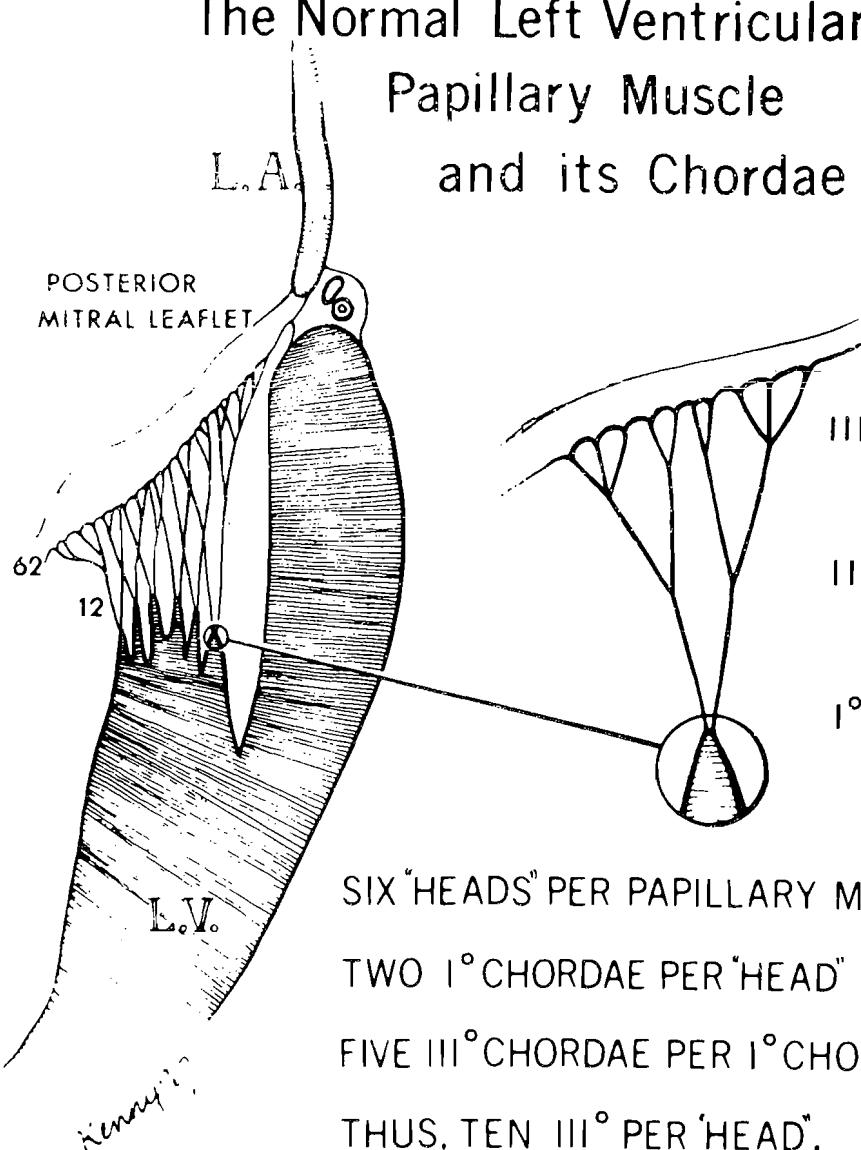


FIG. 17-12. Normal papillary muscle and its attachments. Rupture of a distal papillary muscle head results in mitral regurgitation because of the large number of chordae that arise from one papillary muscle head. In rupture of a chordae tendineae, the degree of mitral reflux is directly related to the site of rupture. If a first order cord (1°) is ruptured, the mitral regurgitation will be far more severe than when second or third order chordae (II° and III°) are torn. The average papillary muscle gives rise to an average of six "heads." (From Roberts WC and Cohen LS: Left ventricular papillary muscles. Description of the normal and a survey of conditions causing them to be abnormal. Circulation 46:138, 1972.)

markedly elongated but intact chordae or snapping of the abnormally thin chordae, producing sudden mitral incompetence. In the majority of such cases the posterior leaflet is involved. In such patients, the mitral valve annulus often is extremely dilated, and the mitral leaflets are composed of excessive and redundant tissue. Recent surgical experience indicates that chordal rupture in subjects with myxomatous degeneration of the mitral valve is a common cause of severe mitral regurgitation.

Patients with rheumatic mitral disease usually do not undergo chordal rupture, which may be related to their thickened, foreshortened, subvalvular structures. Nevertheless, occasionally a chord can snap in subjects with pre-existing rheumatic mitral regurgitation. In such cases, the degree of mitral reflux (and murmur intensity) will suddenly increase. Endocarditis can disrupt chordal function either in the setting of acute valve infection or months to years after bacterial eradication. Pre-existing mitral or aortic regurgitation with left ventricular dilatation may predispose the patient to chordal rupture because of increased tension on the supporting apparatus of the mitral valve, which results in abnormal stretching of the chordae.

The most severe sequelae of ruptured chordae tendinae are experienced by patients with a previously normal heart, in whom a major proximal chord ruptures suddenly, with resultant severe acute mitral regurgitation (flail mitral leaflet). The normal left ventricle and left atrium are suddenly presented with an acute, massive volume overload resulting in excessively high intracardiac pressures. The left atrium and ventricle respond with vigorous and hyperactive contraction (Starling effect), which is often detectable on physical examination. Huge, left atrial V waves and increased left atrial pressure can result in severe pulmonary hypertension. Initially well-tolerated, the mitral regurgitation typically results in dilatation and hypertrophy of the left atrium and ventricle over time; chamber compliance increases and eventually the mitral regurgitation converts to a more chronic hemodynamic state. This process of adaptation may take from 6 to 12 months, at which time the physical findings become indistinguishable from those of chronic mitral regurgitation.

If the chordae to only one valve cusp are disrupted, only one of the mitral leaflets or scallops may protrude into the left atrium. The posterior leaflet is affected far more often than the anterior; an individual posterior scallop may prolapse by itself. On occasion, the portion of the valve that prolapses into the atrium produces a hoodlike deformity, which directs the regurgitant stream opposite to the site of valve prolapse (Fig. 17-13). This "ectopic" jet of blood may produce unusual patterns of murmur radiation. Myxomatous alterations of the leaflet tissue and mitral valve prolapse usually underlie these cases of the floppy valve syndrome.

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PHYSICAL FINDINGS IN RUPTURED CHORDAE TENDINEAE

In patients with ruptured chordae, the physical examination is strongly influenced by the presence or absence of pre-existing structural heart disease. The heart that had been normal will present the picture of *acute mitral regurgitation* without evidence of significant enlargement of the cardiac chamber. On the other hand, a patient with evidence of left ventricular dilatation

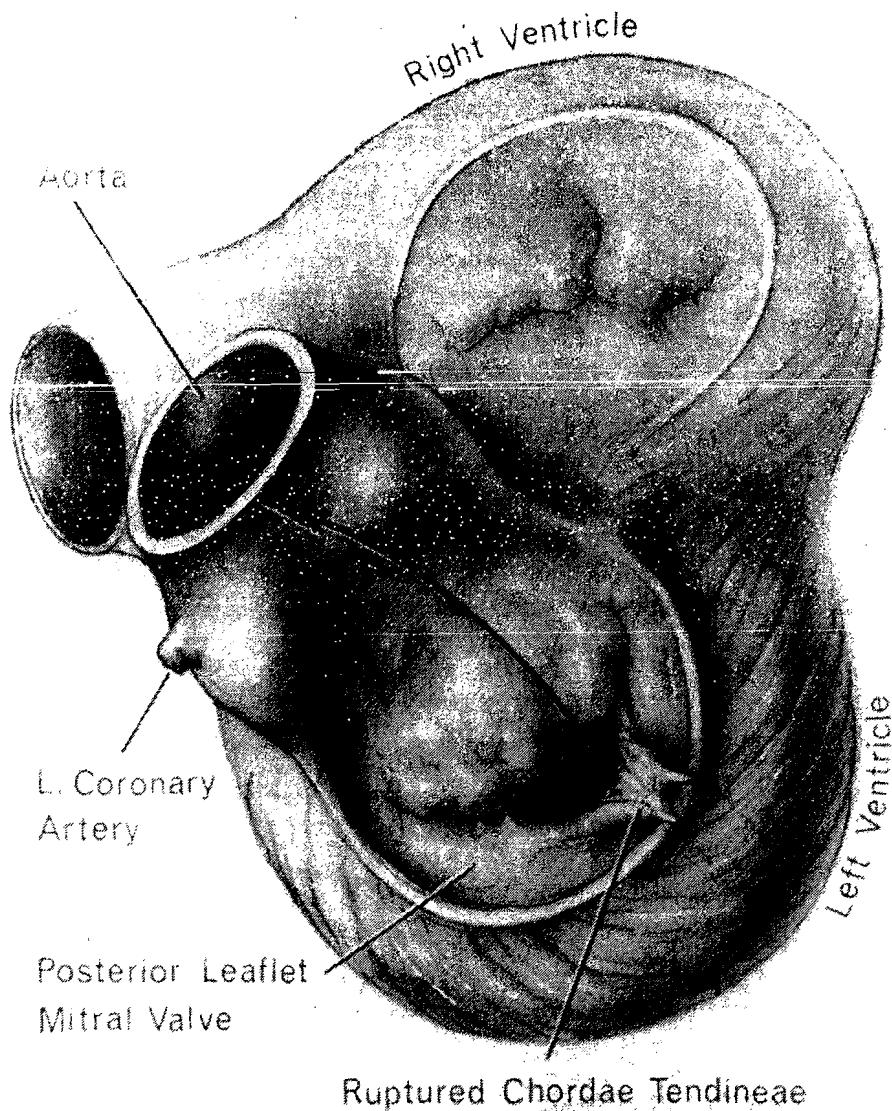


FIG. 17-13. Ruptured chordae tendineae to the posterior mitral leaflet with subsequent selective posterior cusp insufficiency. The mitral regurgitant jet impinges on the medial aspect of the left ventricular outflow tract. This can produce a systolic murmur that radiates to the cardiac base. Posterior leaflet insufficiency is more common in isolated rupture of the chordae tendineae, probably because the chordae to the posterior leaflet are thinner and less sturdy than those supplying the anterior leaflet. (From Selzer A et al: The syndrome of mitral insufficiency due to isolated rupture of the chordae tendineae. Am J Med 43:822, 1967.)

or atrial fibrillation is likely to have had *prior* mitral regurgitation. In this case, the diagnosis of a ruptured chordae tendineae may be based on the abrupt deterioration of the clinical picture and/or a change in the intensity of the pre-existing murmur. Patients with ruptured chordae tendineae and a flail mitral leaflet have severely deranged hemodynamics and often are in overt congestive heart failure.

Carotid Pulse

If the degree of mitral regurgitation is large, the arterial pulses are likely to be quick-rising with a rapid falloff. The pulse volume is normal to decreased. The rhythm is almost always regular (sinus) in patients with ruptured chordae unless there have been long-standing, mitral regurgitation and chronic atrial fibrillation.

Venous Pulse

There are no abnormalities of the jugular venous pulse unless pulmonary hypertension and right ventricular failure occur as a result of the acute massive volume overload. In such instances, the mean venous pressure will be elevated, and large V waves of tricuspid regurgitation may be noted.

Precordial Motion

In a patient without prior cardiovascular disease, the apex impulse will not be displaced laterally but will be exaggerated in amplitude (hyperkinetic) as a result of the acute volume overload. A palpable S4 in the left recumbent position is common and its presence should be sought. *Practical Point: Detection of a palpable or audible S4 (Fig. 17-14) confirms the presence of acute or recent onset mitral regurgitation and virtually excludes a chronic rheumatic etiology.* With massive mitral reflux, a sustained right ventricular lift may be present at the lower left sternal border as a result of pulmonary artery hypertension. The recoil from the large regurgitant jet (huge left atrial V wave) may produce an anterior parasternal impulse confirmed solely to late systole (Figs. 17-4, 17-5).

In patients with pre-existing cardiac disease, left ventricular enlargement is likely to have been present prior to the onset of chordal rupture. A sustained and displaced apex beat indicates that there has been previous cardiac enlargement and/or depression of left ventricular function.

A visible and palpable S3 is often present. Because the typical murmur of mitral regurgitation from ruptured chordae is quite loud, an apical systolic thrill may be present. If an eccentric regurgitant jet occurs as a result of posterior leaflet hooding, the transmitted thrill may be at the base or at the second and third left interspace (Figs. 17-13, 17-15). Of course, this would be an unusual site for the typical mitral regurgitation murmur.

First Heart Sound

There are no specific alterations in the first heart sound in ruptured chordae tendinae. S1 may be soft if the P-R interval is long or loud if the anterior mitral leaflet is excessively mobile.

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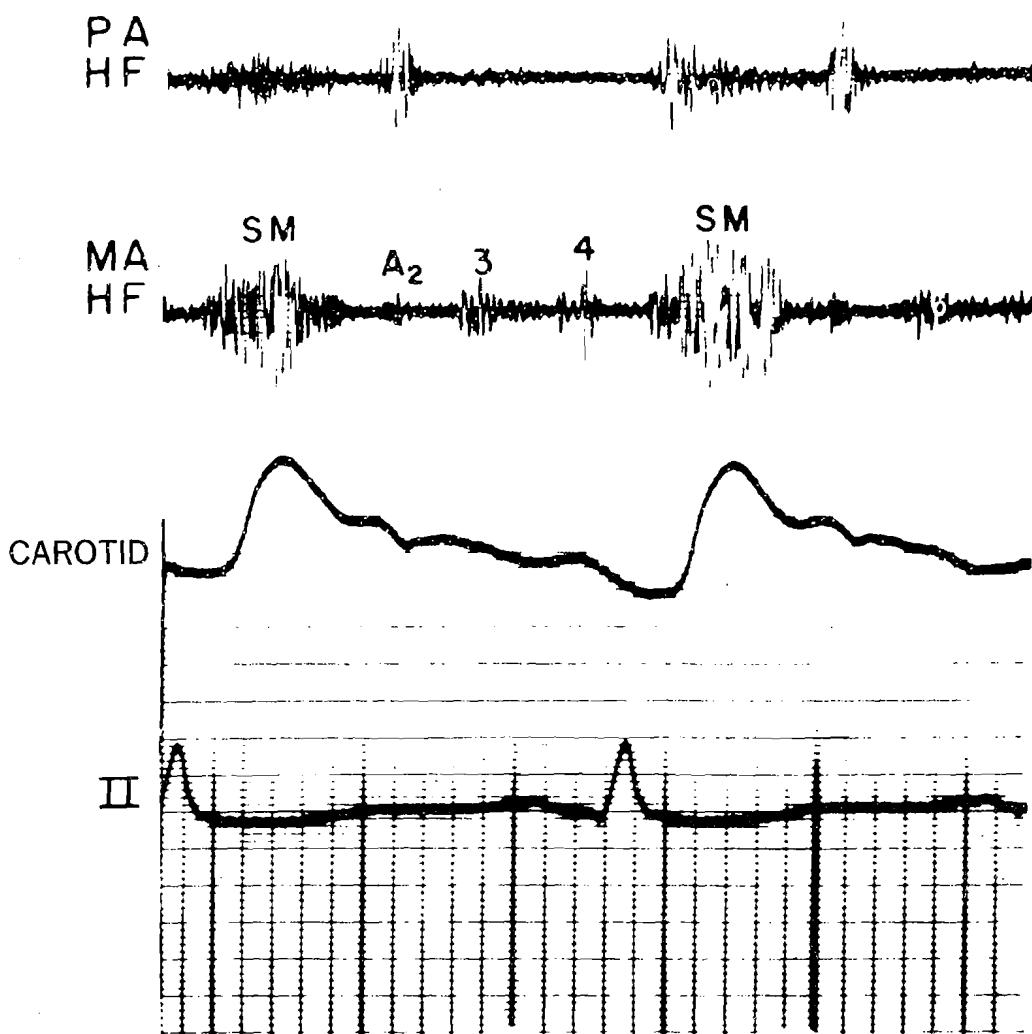


FIG. 17-14. Physical findings in acute mitral regurgitation. Note the prominent S₃ and S₄ as well as the crescendo-decrescendo shape of the systolic murmur (SM). Murmur vibrations do not extend to A₂ because of a huge left atrial V wave and virtual equilibration of left ventricular and left atrial pressure in late systole (see Fig. 17-8). It is easy to mistake this type of murmur for that of aortic stenosis. This patient also had a prominent A wave and rapid filling wave recorded on an apex cardiogram (not shown). (From Sutton GC and Craige E: Clinical findings of severe acute mitral regurgitation. Am J Cardiol 20:141, 1967.)

Second Heart Sound

Changes in S₂ in patients with ruptured chordae are similar to those in severe mitral regurgitation of any etiology. These include wide splitting of S₂ in expiration (shortened left ventricular systole) and an increased P₂ due to pulmonary hypertension, if present. A₂ may be lost in the loud holosystolic murmur and give the appearance of a single second sound.

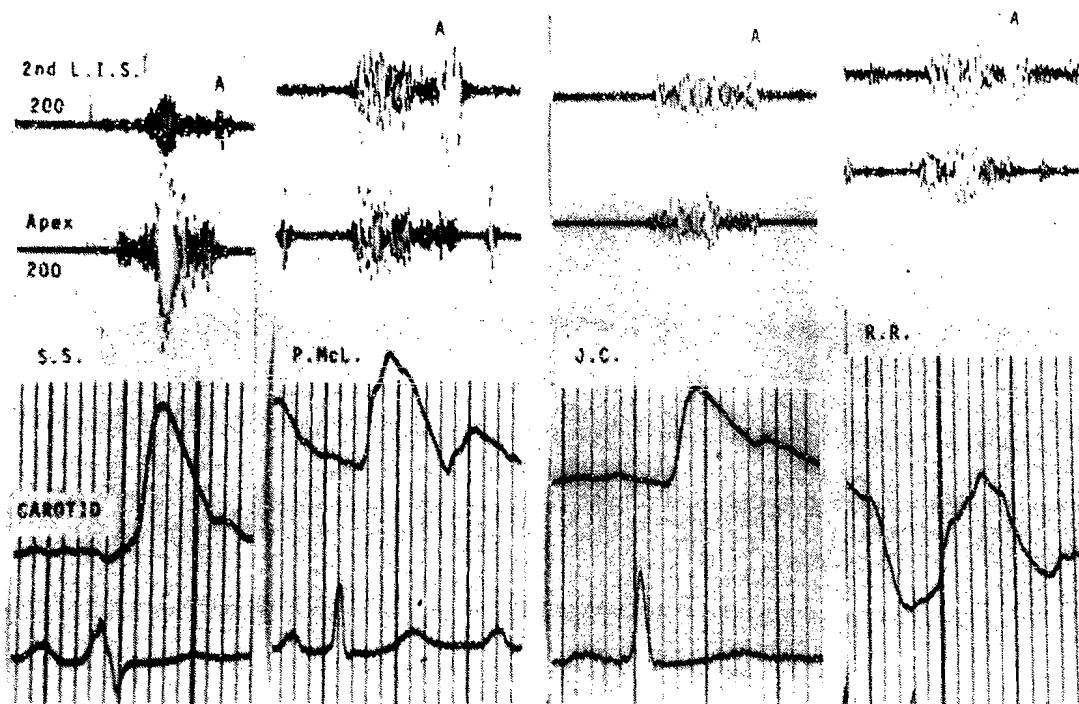


FIG. 17-15. Murmur of ruptured chordae tendineae. Phonocardiograms from 4 patients with ruptured chordae. Note that late systolic murmur vibrations are decreased in all patients, giving an ejection quality to the murmur. When selective posterior leaflet insufficiency is present, the murmur frequently radiates to the aortic area, simulating an aortic stenosis murmur (see Fig. 17-13). (From Ronan JA et al: The clinical diagnosis of acute severe mitral insufficiency. Am J Cardiol 27:284, 1971.)

Third Heart Sound

Frequently, an S3 is found in patients with ruptured chordae tendineae. The S3 may be palpable and is coincident with the rapid filling wave that may usher in a short, mid-diastolic rumble.

Fourth Heart Sound

An atrial sound (S4) coincident with a loud mitral regurgitation murmur is of great diagnostic importance, as it suggests the presence of a normal to modestly enlarged left atrium that is forcefully contracting (Fig. 17-14). This finding precludes the presence of a dilated, baggy left atrium typically found in rheumatic heart disease and other chronic causes of severe mitral regurgitation. The low frequency S4 is often better felt than heard; careful palpation and auscultation in the left recumbent position is mandatory. In such cases, the presence of an S4 correlates with an elevated left ventricular end-diastolic pressure.

Opening Snap

An opening snap occurs in some patients with a ruptured chord. This may be audible or may only be recordable. Loss of chordal support to the posterior leaflet of the mitral valve has been associated with the presence of an opening snap.

Mitral Regurgitation Murmur

Configuration and Duration. Because the mitral reflux caused by ruptured chordae tendinae usually occurs in the presence of a normal, noncompliant left atrium, a moderate to severe degree of regurgitation will result in giant left atrial V waves and substantial elevation of atrial pressure. The left ventricular-left atrial pressure gradient in late systole is often decreased, because the actual amount of reflux into the left atrium decreases considerably in the latter third of systole (Fig. 17-8). This results in a tapering of sound vibrations and a decrescendo configuration to the murmur (Figs. 17-14, 17-15). Often, peak murmur intensity occurring in midsystole results in a crescendo-decrescendo systolic murmur. *Practical Point:* Although the murmur of ruptured chordae tendineae commonly simulates a systolic ejection murmur, careful auscultation will invariably detect sound vibrations extending to S2 (Fig. 17-15). In fact, such a murmur often obliterates A2.

Frequency and Intensity. Most patients with ruptured chordae initially have normal left ventricular function unless severe, pre-existing heart disease is present. As the amount of the regurgitant volume is likely to be large, the murmur is typically loud (Grade III-IV/VI intensity), often with an accompanying thrill. Because of high volume and high velocity mitral reflux, the murmur is composed of mixed frequencies and is somewhat harsh, as compared to the more musical, whirring murmur of rheumatic mitral regurgitation. In general, this distinction is true, but one should not exclude ruptured chordae tendinae solely on the basis of the pitch of the systolic murmur.

Location and Radiation. Often chordal rupture causes eccentric localization and radiation of the murmur as the protrusion or hooding of one of the leaflets or scallops directs the regurgitant jet in a well-localized stream. Selective incompetence of the posterior leaflet or scallop resulting from chordal stretching or rupture tends to deflect the refluxing torrent of blood towards the medial left atrial wall and left ventricular outflow tract (Fig. 17-13). The sound vibrations radiate toward the proximal aorta, and the murmur may have its maximal intensity at the second left or right interspace. A preponderant deformity of the anterior leaflet directs the blood posterolaterally to produce a murmur that is loudest at the apex and left posterior thorax. The posterior leaflet is more likely to be involved in chordal rupture because its

supporting chordae are normally thinner than those to the anterior cusp; in addition, the overall cuspal length is far greater than the anterior. The size and shape of the left atrium, as well as the direction, volume, and velocity of the refluxing blood, also will influence the site and amplitude of murmur intensity.

Mitral Regurgitation Masquerading as Aortic Stenosis. Posterior chordal rupture repeatedly has been documented to result in a loud murmur at the cardiac base (second or third interspace) simulating an aortic ejection murmur (Fig. 17-15). This murmur may be maximal either at the aortic area or apex, but even in the latter case it still may be very prominent at the base. Often the murmur appears to have a shorter duration at the aortic area. *Practical Point: When the murmur of a posterior ruptured chordae tendineae has a crescendo-decrescendo shape, it is easily mistaken for aortic stenosis. To avoid the error of confusing mitral regurgitation with aortic stenosis, one must pay attention to the length of the systolic murmur at both the aortic area and apex as well as to the quality of the carotid upstroke.*

Selective anterior mitral leaflet incompetence due to chordal rupture is much less likely to cause confusion. In this case, the regurgitant jet is directed posteriorly; when loud, this murmur can radiate to the vertebral column and left posterior chest. Astute clinicians even have detected this systolic murmur over the midthoracic spine as well as at the top of the head!

In spite of the above observations, several large series of patients with ruptured chordae tendineae have demonstrated that there is no predictable or consistent relationship between the site of maximal murmur intensity and the specific leaflet involved. Clinicians should be aware of the vagaries of the clinical presentation of chordal rupture, particularly with respect to the false diagnosis of aortic stenosis.

Mid-diastolic Murmur

The large volume leak found in many of these patients will result in a mid-diastolic flow rumble (Figs. 17-3, 17-4, 17-6B), usually initiated by an S3. Its presence indicates involvement of a major degree of mitral regurgitation.

CALCIFICATION OF THE MITRAL ANNULUS

Calcification of the mitral annulus is a relatively uncommon cause of mitral regurgitation usually found in older patients. The increased use of echocardiography has resulted in the "accidental" discovery of annulus calcifications in many older patients with mitral insufficiency. More than 10% of subjects over the age of 50 are estimated to have detectable calcification

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of the mitral annulus or A-V cusps. The cardiac fibrous skeleton, which includes the mitral valve ring or annulus, is susceptible to degenerative calcification. Because the posterior mitral leaflet has an annulus but the anterior leaflet does not, the calcium deposits typically are in proximity to the posterior cusp.

The mitral annulus is partially made up of smooth muscle and thus is able to contract. The mitral valve ring itself narrows in systole as left ventricular cavity size decreases during ejection. This normal sphincter-like action may become impaired when there is calcification of the mitral annulus. Frequently, calcium deposits also are found on the ventral surface of the mitral valve leaflets with spicules of calcium projecting into the surrounding myocardium. The calcified annulus may remain relatively dilated during systole with respect to the contracting left ventricle, thus impairing adequate mitral leaflet coaptation. In such cases, the mitral regurgitation produced is usually of mild to moderate degree, and the size of the left atrium is often increased.

The diagnosis of calcification of the mitral annulus is made either on a lateral roentgenogram of the chest, which reveals a dense curvilinear calcific band of O, U, C, or J configuration in the region of the mitral valve or, more commonly, by the presence of densely reflecting echoes on M-mode or two-dimensional echocardiography (Fig. 17-16).

Calcification of the mitral annulus is most likely to be found in elderly women. There is an increased prevalence in patients with hypertension, cor-

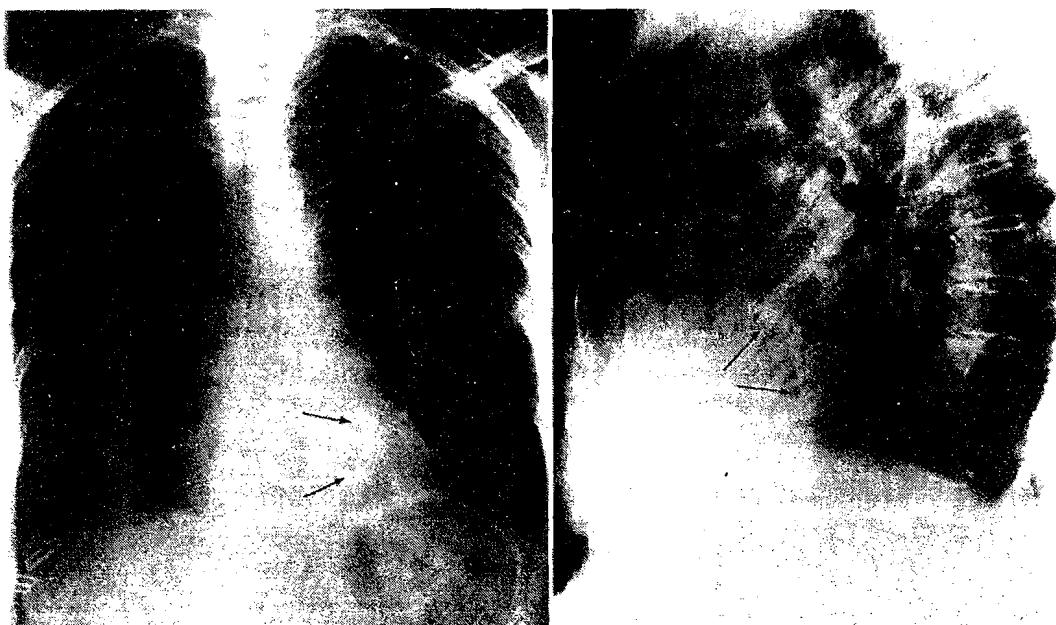


FIG. 17-16. Mitral annular calcification. Notice the heavy calcific density in both the PA and lateral projections. A classic C or J rim of calcium is seen on the border of the heart in the presence of severe mitral annular calcification. Calcium typically extends down into the left ventricular myocardium below the posterior mitral valve attachment. (From Silverman ME and Hurst JW: The mitral complex. Am Heart J 76:399, 1968.)

onary artery disease, and diabetes. Patients with congestive heart failure appear to have an increased incidence of calcification of the mitral annulus. In younger age groups, patients undergoing long-term dialysis for renal failure frequently have extensive calcification of the annulus and mitral valve.

When the mitral annulus is calcified, the aortic valve leaflets (but not the annulus) may be sclerotic with or without calcific deposits. Calcification of the mitral annulus may increase the severity of mitral regurgitation in subjects with mitral valve disease of other etiologies. Mitral annular calcification occasionally may be associated with mitral valve prolapse and has an increased incidence in Marfan's syndrome and hypertrophic cardiomyopathy. One recent report suggested that heavy calcium deposits in the annulus, particularly in the posterior position, can result in an unusual variant of mitral stenosis.

PHYSICAL FINDINGS

The cardiac examination in patients with calcification of the mitral annulus is similar to that in chronic rheumatic mitral regurgitation. There are no distinguishing clinical features on palpation or auscultation. Although some authors believe that systolic ejection murmurs may be related to annular calcification, this is likely to be a chance association. Some have observed that the apical systolic murmur may be more low pitched than the classic MR murmur.

ACUTE MITRAL REGURGITATION

Acute mitral regurgitation is potentially a life-threatening condition. It is therefore important to understand the unique hemodynamics that produce a clinical picture different from that of chronic mitral regurgitation. Physicians should be alert to the important differences between the presentation of acute and chronic mitral insufficiency (Table 17-10).

ETIOLOGY

Severe *papillary muscle dysfunction* or *rupture* and *ruptured chordae tendineae* typically produce mitral regurgitation of acute or recent onset. *Endocarditis* is another cause of sudden mitral regurgitation; in this condition, the mitral valve leaflets can erode or perforate, or infection can involve the supporting apparatus and result in rupture of a chord. Rarely, a myocardial abscess can interfere with mitral valve function by tunneling into the mitral valve annulus or by direct involvement of the papillary muscle itself. Blunt

or perforating *trauma* can result in acute mitral regurgitation if the mitral leaflets or supporting apparatus are damaged or disrupted.

PATHOPHYSIOLOGY

In all cases of acute mitral regurgitation, the sine qua non is the sudden onset of severe reflux into a normal sized left atrium. The full force of left ventricular systolic pressure is directly transmitted into a nonexpansile atrium, producing huge V waves. Left ventricular end-diastolic pressure increases and left atrial contractile force is enhanced. Left ventricular ejection rate and the LV ejection fraction may actually be higher than normal. Pulmonary capillary wedge and pulmonary arterial pressures increase, often dramatically. Severe pulmonary hypertension may ensue, and signs of right ventricular failure may appear.

As long as left ventricular systolic function is preserved, the marked increase in LV diastolic volume is vigorously ejected into both the aorta and left atrium; forward cardiac output typically is maintained despite a large regurgitant fraction. With time, LV mass and size increase, and this acute hemodynamic state may convert to the pathophysiology of chronic mitral regurgitation: left ventricular and left atrial size and compliance increase, left atrial and pulmonary venous pressures drop, and atrial fibrillation may ensue. This process can take from 6 to 18 months to evolve.

In some patients with severe acute mitral regurgitation, the clinical picture may be dramatic. The clinical course is far shorter, and early surgical intervention may be mandatory if left ventricular function becomes deranged and/or pulmonary congestion is severe. With the onset of overt congestive heart failure, the elevated systemic vascular resistance produced from reflex arterial vasoconstriction may worsen the hemodynamic situation by increasing the afterload of the left ventricle, thus increasing the degree of mitral regurgitation. In general, acute mitral regurgitation produces less stress on the left ventricle than acute aortic regurgitation, probably because of a decrease in late systolic tension that occurs as the left ventricle unloads itself during the last half of systole.

PHYSICAL FINDINGS IN ACUTE MITRAL REGURGITATION

(Tables 17-9, 17-10)

At the time of examination, many patients are in mild to severe congestive heart failures and therefore may manifest orthopnea, tachypnea, and audible pulmonary crepitations. The heart rate will usually be increased. The arterial pulse is quick to rise and collapse as a consequence of severe mitral regurgitation, although the presence of congestive heart failure may sufficiently

TABLE 17-9 *Bedside Clues to Hemodynamically Severe Acute Mitral Regurgitation*

Hyperdynamic LV impulse (not usually displaced)
Parasternal lift may be present
Widely split S2 with loud P2
Prominent S4—may be palpable
S3 ± flow rumble. S3 may be palpable.
Murmur may be surprisingly soft if associated with acute MI
Murmur may simulate aortic stenosis (ejection quality with radiation to base) if due to chordal rupture

attenuate the pulse volume as to preclude a meaningful assessment. The mean jugular venous pressure is often elevated; large A and V waves may be present. The latter will be dominant if there is associated tricuspid regurgitation caused by a high pulmonary artery pressures.

Precordial examination will usually reveal a hyperdynamic left ventricular impulse, often with a palpable S3. Unless there is pre-existing cardiomegaly, the apex impulse will not be displaced laterally. Patients with papillary muscle dysfunction may demonstrate an ectopic left ventricular impulse or a sustained or bifid apical thrust. One of the hallmarks of acute mitral regurgitation is the increased force of left atrial contraction producing an audible and often palpable S4 (Fig. 7-14). Palpation in the left recumbent position is mandatory and may be more sensitive than auscultation in detecting the low frequency S4. An S3 is commonly heard in severe acute mitral regurgitation (Fig. 17-14). Most subjects with acute or recent onset mitral regurgitation have a large volume leak; the resultant vigorous left atrial expansion may produce an anterior thrust of the entire heart. This produces a late systolic parasternal lift (see pages 315-317, Figs. 17-4, 17-5).

TABLE 17-10 *Differentiating Features of Severe Acute and Chronic Mitral Regurgitation*

	Chronic	Acute
Rhythm	Often atrial fibrillation	Sinus rhythm or sinus tachycardia
Precordial examination	Hyperkinetic LV with lateral downward displacement LV heave possible. Palpable S3 often. Apical thrill. Can have parasternal or LA lift	Hyperkinetic LV without lateral downward displacement Palpable S3. Palpable S4 Apical thrill frequently common Parasternal LA lift
Heart sounds	S3 common. No S4. OS occasionally noted Increased P2 if pulmonary hypertension	S3 and S4 common. Occasional OS (if posterior leaflet, especially with RCT) Increased P2
Systolic murmur	Typically holosystolic, even, whirring. May have mid or late systolic accentuation, maximum at apex	Typically loud, harsh murmur with late systolic decrescendo. May appear to be ejection in quality. May be very prominent at aortic area or 2-LICS

As previously indicated, the murmur of acute mitral regurgitation may taper markedly in late systole and have a crescendo-decrescendo contour (Figs. 17-7D, 17-14, 17-15) because the huge, left atrial V wave causes near equilibration of the left ventricular-left atrial pressure in mid- to late systole (Fig. 17-8). If left ventricular function remains well preserved, the murmur will be quite loud, typically accompanied by a thrill.

Eccentric radiation patterns, especially with posterior leaflet incompetence, may produce a systolic murmur that is loudest at the second left interspace or aortic area. The resultant basal systolic murmur with a decrescendo configuration readily mimics aortic stenosis (Figs. 17-14, 17-15). The majority of murmurs of acute mitral regurgitation from any cause will be loudest or equally loud at the apex; careful auscultation will demonstrate that the murmur is truly holosystolic. Some murmurs may end before S₂, which is consistent with an enormous left atrial V wave (Fig. 17-8). Typically, the murmur of a flail anterior mitral leaflet is loudest in the left posterior chest and may be heard along the vertebral column as well as at the top of the skull. In fact, many murmurs of acute mitral regurgitation can be heard over the entire thoracic vertebral column.

A short, mid-diastolic murmur that immediately follows S₃ is common in patients with severe mitral regurgitation of acute or chronic etiology (Figs. 17-6B, 17-16B). Its presence indicates a large regurgitant fraction. A separate holosystolic murmur along the lower sternal border, which increases with inspiration, indicates associated tricuspid regurgitation that may occur as a result of marked elevation of pulmonary artery pressure. Tables 17-5, 17-9 and 17-10 compare and contrast some of the important diagnostic features of acute and chronic mitral regurgitation.

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Chapter 18

Mitral Valve Prolapse

Mitral valve prolapse (MVP), first identified and characterized in the 1960s, is the most common abnormality of the mitral valve (whether or not mitral regurgitation is present) and has been estimated to occur in between 4 to 8% of the population. This condition and its various manifestations have been the subject of intense scrutiny over the past decade.

The spectrum of MVP ranges from an isolated systolic click in an asymptomatic individual to full-blown, severe mitral regurgitation necessitating mitral valve replacement (floppy valve syndrome). Many cases are discovered accidentally during routine physical examination. Affected individuals may have a variety of symptoms such as palpitations, atypical chest pain, or dyspnea. Anxiety, fatigue, and occasionally syncope may also occur in symptomatic patients. Rare sudden deaths have been reported, presumably caused by malignant ventricular dysrhythmias.

Mitral valve prolapse initially was documented angiographically by Cribley in 1966. The mitral origin of a nonejection click and late systolic murmur was postulated independently by Barlow and Reid in South Africa in the early 1960s. MVP has a multitude of names and eponyms. Some of the most common of these are listed in Table 18-1. The term *mitral valve prolapse* is most desirable, although many authors still refer to MVP as Barlow's syndrome.

A spectrum of noncardiac abnormalities have been associated with the mitral valve prolapse syndrome. These include evidence for a hyperadrenergic state such as resting tachycardia, increased plasma catecholamines, and diffuse anxiety, as well as an increased sensitivity to infusion of isoproterenol. Enhanced vagal activity has also been suggested. One study found an association of MVP with thyrotoxicosis. Individuals with MVP have been shown to have a wide variety of supraventricular and ventricular tachyarrhythmias as well as bradyarrhythmias.

Mitral valve prolapse has been identified in some individuals as a heritable disorder with an autosomal dominance pattern. MVP is common in such connective tissue disorders as the Ehlers-Danlos syndrome, Marfan's syndrome, pseudoxanthoma elasticum, osteogenesis imperfecta, and the von Willebrand syndrome. These, and other associated abnormalities, have led to speculation that MVP may well be a syndrome related to mesenchymal dysplasia and disordered embryologic development.

TABLE 18-1 *Partial List of Proposed Names for Mitral Valve Prolapse Syndrome*

Click-murmur syndrome
Click-late systolic murmur syndrome
Systolic click syndrome
Mitral click-murmur syndrome
Systolic click-late systolic murmur syndrome
Midsystolic click-late systolic murmur syndrome
Syndrome of mid-systolic extra sound and late systolic murmur
Billowing mitral leaflet syndrome
Billowing posterior mitral valve leaflet syndrome
Prolapsing mitral valve leaflet syndrome
Ballooning posterior leaflet myocardiopathy
Mitral valve prolapse-click syndrome
Mitral valve prolapse syndrome
Floppy valve syndrome
Barlow's syndrome
Syndrome of apical systolic click, late systolic murmur, and abnormal T-waves
The auscultatory-electrocardiographic syndrome

From Abrams, J: Am Heart J, 92:414, 1976, with permission.

PATHOPHYSIOLOGY

The basic defect of MVP appears to be an alteration in the composition of the mitral valve tissue and chordae. There is a loss or dissolution of the normal dense collagen fibers (fibrosa) and replacement and invasion by a less sturdy type of connective tissue (spongiosa) that has increased amounts of uronic acid mucopolysaccharide material. Increased amounts of collagen precursor are present, suggesting a related abnormality of collagen synthesis. The resultant myxomatous transformation may affect individual scallops or an entire mitral cusp. One or both leaflets may be involved. The chordae tendineae may also be abnormal and are often elongated and attenuated. Whether the chordae are primarily involved in prolapse or are secondarily distorted by the abnormal mitral valve anatomy is unknown. The mitral leaflet tissue itself is often thickened, redundant, and excessive (Fig. 18-1). During systole individual scallops or an entire leaflet billow excessively into the left atrium.

Criley aptly has depicted the mitral valve in MVP as being literally being "too big" for the left ventricle; the valve leaflets protrude or prolapse abnormally above the mitral annulus during ventricular contraction (ventriculovalvular disproportion). Mitral regurgitation, if present, occurs when there is loss of normal coaptation resulting from true prolapse of the mitral leaflet due either to protrusion of the redundant valve tissue or to abnormally long and lax chordae. The greater the distortion of leaflet apposition, the greater the severity of the mitral leak.

Mitral regurgitation when present is typically confined to *late systole* and occurs at the time that the size of the left ventricular chamber has

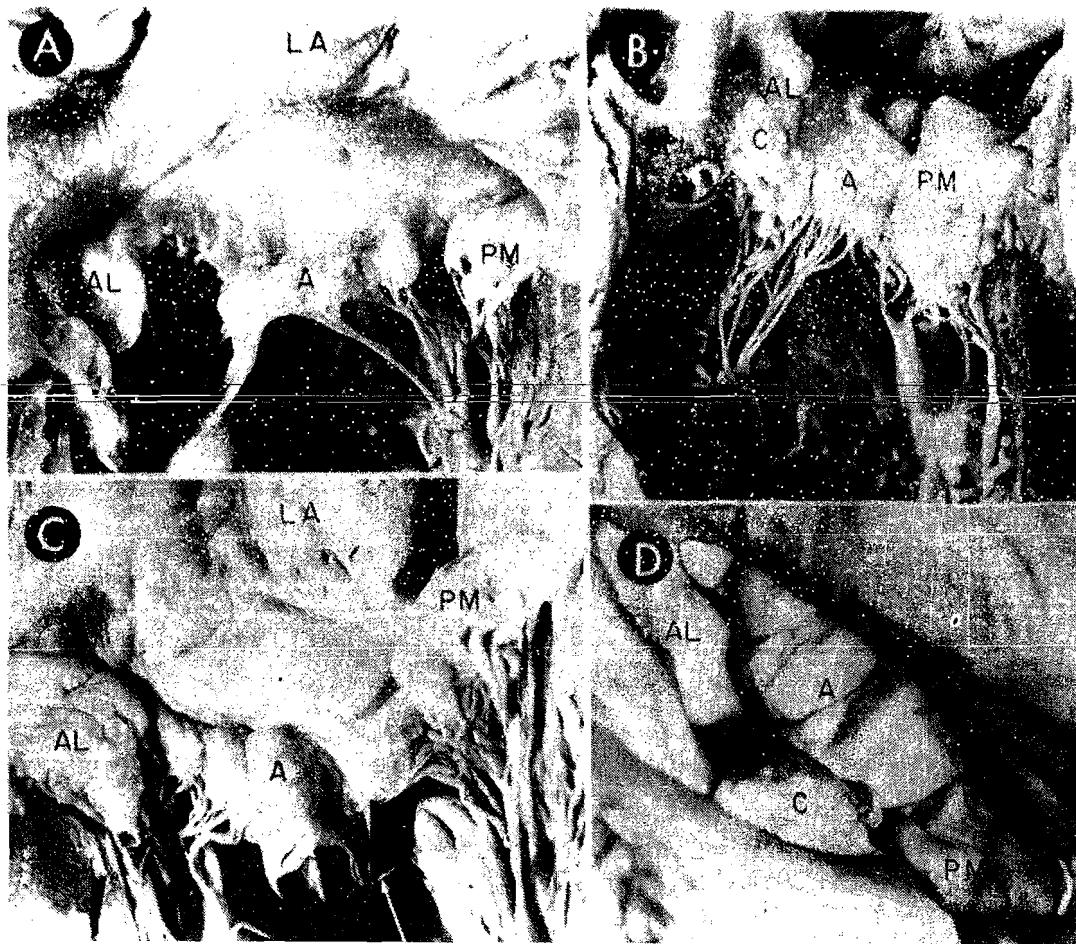


FIG. 18-1. Anatomy of mitral valve prolapse. A. Note bulging of the outer portion of the anterior leaflet and hooding of the posteromedial and anterolateral scallops of the posterior leaflet. B. More advanced degree of prolapse with hooding of the anterior leaflet as well as hooding of all three scallops of the posterior leaflet. C. Hooding of the anterior leaflet, the posteromedial, and anterolateral scallops. Two ruptured chordae are attached to the anterior leaflet, with elongated and thinned chordae elsewhere. D. Same specimen as in B, with the mitral valve viewed from the atrium. AL = anterolateral scallop; PM = posteromedial scallop; C = central scallop; A = anterior leaflet; LA = left atrium. (From Lucas RV and Edwards JE: The floppy mitral valve. *Curr Prob Cardiol* 7:1, 1982.)

decreased maximally and the anatomic extent of prolapse is greatest. In subjects with severe prolapse, the murmur and the mitral regurgitation may be holosystolic. There appears to be a "prolapse threshold" during systole for each patient related to the size of the left ventricular cavity in systole and the propensity of the redundant valve tissue to bulge into the left atrium. Although valve prolapse appears to occur at the same end-systolic volume, this threshold may be highly variable from day to day or even moment to moment, depending on ventricular volume, posture, heart rate, as well as other factors.

Both mitral leaflets can be affected in MVP, but the posterior (mural) cusp is more commonly involved. The posterior leaflet has a greater potential

for prolapse, probably because of the multiple scallops and less adequate chordal support than that of the anterior cusp. For severe mitral regurgitation to be present, both leaflets must be affected or one cusp may be flail, e.g., a ruptured chordae tendinae. In such individuals, the mitral valve tissue is predictably voluminous and eccentrically thickened. It has been suggested that isolated posterior leaflet prolapse is more likely to produce mitral regurgitation than isolated anterior cusp prolapse. Anterior prolapse without posterior cusp abnormalities appears likely to produce systolic clicks but no murmur of mitral regurgitation. In severe mitral valve prolapse, the mitral valve annulus is often quite dilated, which probably contributes to the mitral regurgitation. The stress on the ballooning leaflets during ejection may result in additional stretching of the valve tissue and chordae, producing increasing disruption and slippage of the collagen fibers within the leaflets and further elongation of the overly stretched chordae themselves. Thus, prolapse may beget greater prolapse.

Peculiar left ventricular contraction abnormalities have been documented in MVP patients, and some investigators believe mitral prolapse is a primary disorder of cardiac muscle, i.e., a cardiomyopathy. Systolic contraction bulges have been noted in the left ventricle, particularly in the inferoposterior region, and dyskinetic patterns of left ventricular shortening have been demonstrated. In such cases, the papillary muscles may become distorted and malpositioned, with prolapse occurring as a papillary muscle protrudes excessively into the left ventricular cavity during systole. Such contractile abnormalities and papillary muscle displacement may result from excessive tension produced by the prolapsed valve and its stretched chordae during systole.

ETIOLOGY

Many conditions have been associated with mitral valve prolapse (Table 18-2). It is best to categorize prolapse as either a *primary* or *secondary* disorder. By far the most common, primary cases represent an idiopathic abnormality of mitral valve tissue that appears to have its onset after childhood. MVP is uncommon in children. The increased incidence of mitral valve prolapse in older persons may relate in part to the decrease in the size of the left ventricular chamber that occurs with aging, thus accentuating the disparity between the volume of mitral valve tissue and the left ventricular cavity. In addition, the collagen content of the valve leaflets appears to be decreased in older subjects.

Primary mitral valve prolapse is familial and is transmitted as an autosomal dominant trait with incomplete penetrance. The rare systemic connective tissue disorders such as Marfan's syndrome, Erdheim's cystic medial necrosis, von Willebrand syndrome, and Ehlers-Danlos syndrome have an extremely high incidence of mitral valve prolapse, but primary or idiopathic mitral prolapse is unrelated to these unusual conditions in the vast majority of affected individuals.

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TABLE 18-2 Conditions That Have Been Associated with Mitral Valve Prolapse

Floppy valve syndrome (myxomatous degeneration)
Mafran's syndrome
Ehlers-Danlos syndrome
Von Willebrand syndrome
Atrial septal defect
Rheumatic mitral valve disease
Hypertrophic cardiomyopathy
Hyperthyroidism
Straight back syndrome
Muscular dystrophy
Wolff-Parkinson-White syndrome
Lupus erythematosus
Relapsing polychondritis
Pseudoxanthoma elasticum

Mitral valve prolapse has been found to occur in patients with clinically manifest coronary artery disease and has been documented to appear following myocardial infarction. It may also occur with rheumatic mitral valve disease and occasionally is noted following mitral valve commissurotomy for mitral stenosis. Patients with secondum atrial septal defects and hypertrophic cardiomyopathy appear to have an increased prevalence of MVP on echo, particularly the patient with atrial septal defect (up to 30% in some series). The association of these relatively common disorders and mitral prolapse may be fortuitous, or the MVP may relate to an abnormality of the mitral valve supporting structures. *Practical Point: Mitral valve prolapse is the final common pathway for a number of conditions affecting the mitral valve apparatus. It most commonly results from an abnormality in the composition of mitral leaflet collagen but may occur as a result of independent abnormalities of the left ventricle, papillary muscles, or chordae tendineae.*

A number of serious complications may occur in patients with MVP (Table 18-3). Therefore, it is of more than academic importance to accurately detect this condition on physical examination. For instance, this diagnosis suggests consideration of lifelong antibiotic prophylaxis to prevent bacterial endocarditis. MVP is a clinical entity that is best delineated by auscultation and echocardiography. In the absence of a click and/or late systolic murmur on repeated examination, an individual should not be labelled as having the MVP syndrome solely because of echocardiographic findings. Patients with

TABLE 18-3 Serious Complications of Mitral Valve Prolapse

Bacterial endocarditis
Transient cerebral ischemic attacks
Cerebral (or systemic) emboli
Severe mitral regurgitation (floppy mitral valve syndrome)
Congestive heart failure
Necessity for mitral valve replacement
Sudden death (rare)

prolapse on echo who are asymptomatic or who have symptoms of atypical chest pain or palpitations but a consistently normal cardiac examination fall into a most difficult diagnostic category. Some clinicians consider these subjects to have true MVP, but others do not.

PHYSICAL EXAMINATION

General Appearance

Mitral valve prolapse is one of the few cardiac conditions in adults where the outward appearance of the subject may suggest the diagnosis. Individuals with the familial variety of prolapse are often red-haired and fair-skinned. Affected subjects have an increased prevalence of nonspecific skeletal abnormalities in both the nonfamilial and familial varieties. Many patients are thin or asthenic with long extremities. Some degree of pectus excavatum and, less commonly, pectus carinatum is common. Scoliosis and loss of the normal dorsal thoracic kyphosis is frequently observed; a narrow A-P chest diameter (straight back) may be present. While such subjects are not true Marfan variants, some will be found to have hyperextensible fingers and high-arched palates. Flat or hollowed feet and incurring fifth fingers have been noted in occasional patients with mitral valve prolapse. Recently, MVP has been associated with very small breasts in women (hypomastia) in addition to the thoracoskeletal abnormalities.

The incidence of such minor skeletal abnormalities in perhaps 20 to 30% of patients with primary mitral valve prolapse suggests an embryonic defect in mesenchymal formation that has been called a linked mesenchymal dysplasia. Formation of the atrioventricular valves, mitral annulus, and atrial septum secundum, as well as chondrification and ossification of the skeletal cage, all occur around the sixth to seventh week of gestation. Breast differentiation begins at this time. Thus, a common but unidentified embryologic defect of connective tissue formation and/or structure may be a link to our ultimate understanding of mitral valve prolapse.

Carotid and Jugular Venous Pulse

No detectable abnormalities of the arterial or venous pulse are present in the large majority of individuals with mitral prolapse. In the unusual case with severe mitral regurgitation, the arterial pulse may be brisk and collapsing in quality. A retraction notch coincident with the midsystolic click has been recorded in the carotid pulse in some patients with MVP, but this is not usually palpable. The venous pulse is unremarkable. If severe mitral regurgitation (floppy mitral valve syndrome) has resulted in right ventricular failure, the mean venous pressure will be elevated and V waves of tricuspid regurgitation may be present.

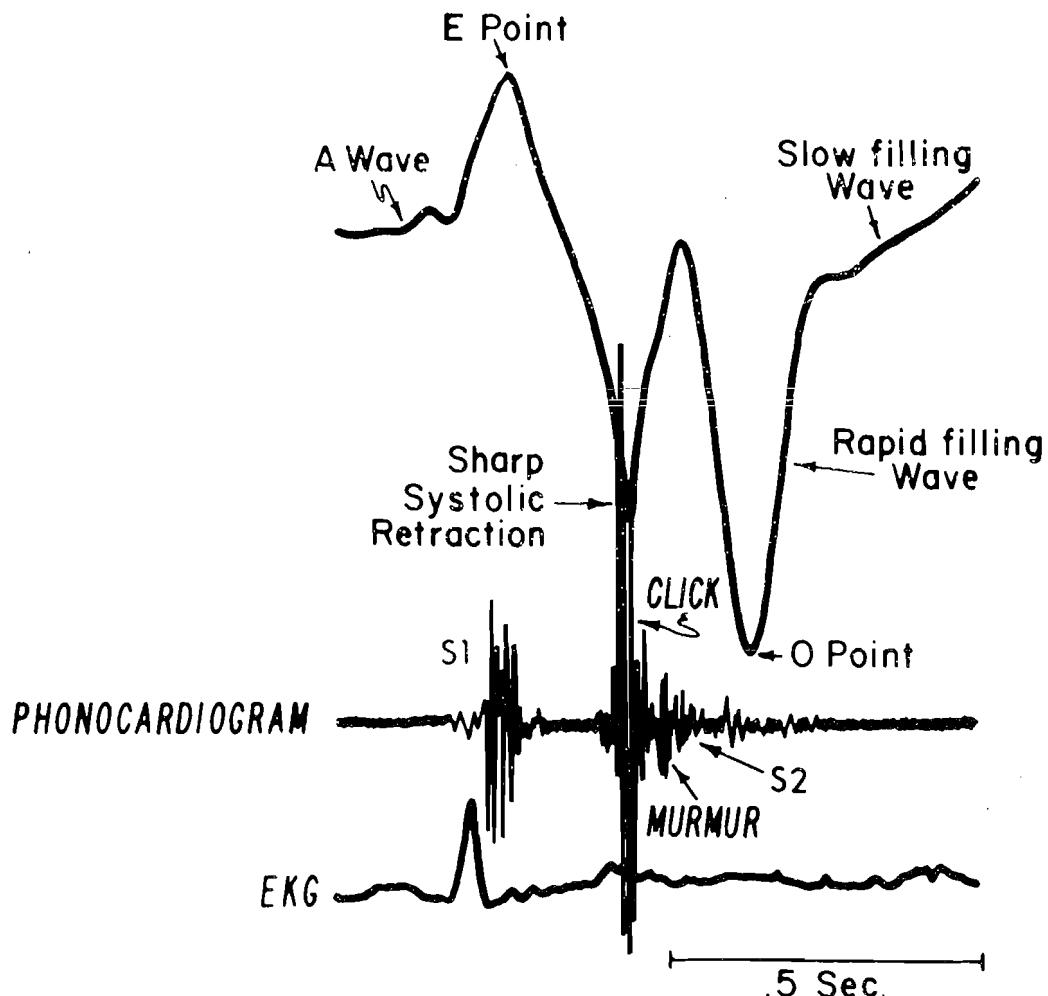


FIG. 18-2. Apical midsystolic retraction wave in mitral valve prolapse. Note that the sharp inward systolic motion on the apex cardiogram coincides with a loud midsystolic click on the phonocardiogram. This variant of a double apical impulse may or may not be detectable on palpation. (From Spencer WH, Behar VS, and Orgain ES: Apex cardiogram in patients with prolapsing mitral valve. Am J Cardiol 32:276, 1973.)

Precordial Motion

The apex impulse in MVP is normal on palpation. Because the associated mitral regurgitation, when present, is typically mild, left ventricular enlargement does not occur and the left ventricular impulse is unremarkable. Apex cardiology has documented a recordable midsystolic retraction wave coinciding exactly with the nonejection click at the point of maximal excursion of the prolapsing leaflet (Fig. 18-2). Occasionally, this may be detected as a bifid apical impulse by careful palpation with the subject in the left lateral decubitus position, but the retraction motion usually is too rapid and subtle to be of diagnostic help. If MVP is associated with another cardiovascular process (e.g., coronary artery disease, atrial septal defect), the precordial impulse may be abnormal because of the coexisting condition. When severe mitral regurgitation is present, the apical impulse will be hyperdynamic, and

may be displaced leftward if there is LV dilatation (see Chapters 5, 17, Fig. 17-4).

Heart Sounds

There are no predictable abnormalities of the first and second heart sounds in patients with MVP. The S1 is usually normal in intensity in the common variant of MVP with a mid or late systolic click, with or without a late systolic murmur. In the unusual case of a flail mitral leaflet and gross mitral regurgitation, S1 may be attenuated or even absent.

An increase in intensity of S1 may occur if the click is close to or coincides with S1. A loud S1 highly correlates with early or holosystolic prolapse and a holosystolic murmur, although early systolic prolapse itself is not always associated with mitral regurgitation.

When present, the degree of mitral reflux in MVP usually is only mild to moderate; therefore, the widely split S2 common to severe mitral regurgitation is not found in most patients with prolapse. An S3 may be noted in a patient with severe mitral regurgitation. An S4 is not a feature of mitral valve prolapse, although it is a possible finding in a patient with coronary artery disease and a midsystolic click.

The Click(s)

Practical Point: *The presence of a single or multiple systolic clicks in mid to late systole is the most specific auscultatory feature of mitral valve prolapse (Figs. 18-2, 18-3).* The clicks in MVP are brief, high frequency, popping, or snapping sounds that are produced by systolic billowing of the mitral valve leaflets or individual scallops and are synchronous with the maximal prolapsing motion of the involved valve tissue. It is possible that these sounds are also generated by abrupt tautening of the chordae supplying the prolapsing portion of the valve leaflets; some have used the term "chordal snap" for these clicks. When multiple clicks are present, one click is usually more prominent than others (Fig. 18-3). The clicks are often variable in timing and may change their position within systole (see below). Multiple clicks are frequently present but are often missed by the inexperienced examiner. A series of high frequency clicks typically sounds like a faint crackling or rustling noise. The clicks often appear to be disassociated from the murmur and may have a metallic quality. Occasionally, a series of high-pitched clicks can simulate a pericardial friction rub. When several clicks are present in succession, the resultant sound may be mistaken for a scratchy systolic murmur. Multiple clicks do not appear to have a prognostic or diagnostic implication different from that of a single midsystolic click.

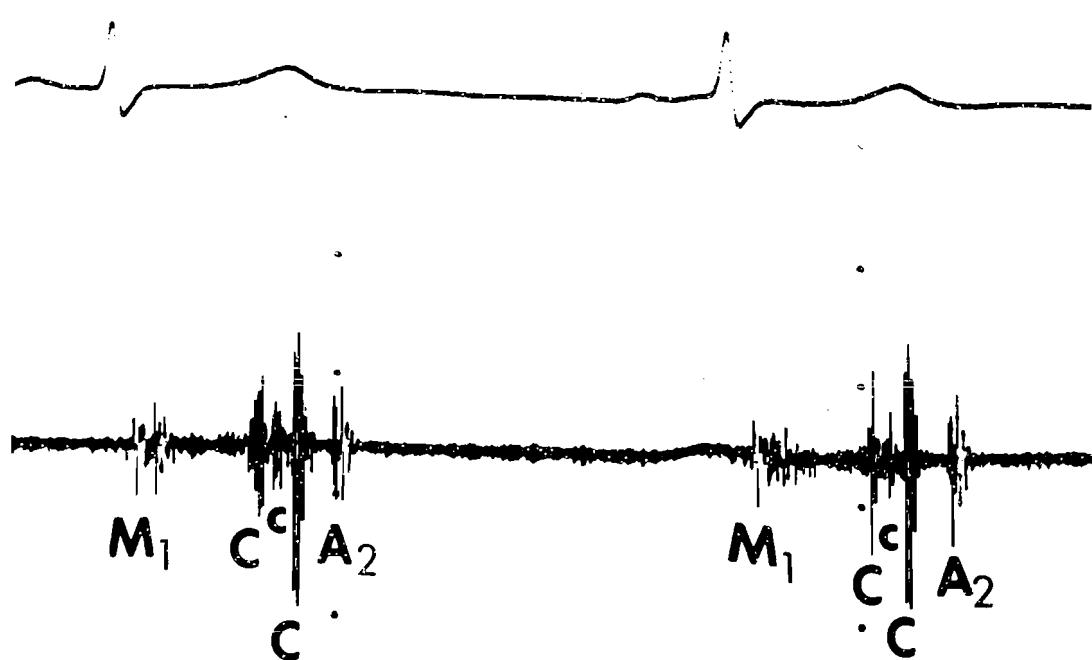


FIG. 18-3. Multiple systolic clicks in a patient with mitral valve prolapse. There are at least three dominant clicks, with the loudest in late systole. (From Cheitlin MD and Byrd RC: Prolapsed mitral valve: The commonest valve disease? *Curr Prob Cardiol* 8:1, 1984.)

MVP clicks typically appear to be close to the ear. They often simulate adventitial sounds, such as those produced by rubbing the stethoscope with a sheet or clothing. In fact, for many years midsystolic clicks were not recognized as coming from the heart itself but were thought to be of pericardial, pulmonary, or mediastinal origin. It is important to concentrate or tune in to the high frequency range to best detect midsystolic clicks; holding one's breath is helpful for optimal auscultation. Clicks are often very faint. Acoustically, they are dissimilar to the far more common low frequency cardiac sounds (e.g., S3, S4). Most physicians are not accustomed to listening to sounds in the high frequency range. The clicks (as well as the murmur) may vary in intensity and number over time and may be inconsistently present even on a beat-to-beat basis. A prominent systolic murmur can easily obscure the presence of clicks unless the examiner concentrates on different frequency ranges during auscultation. *Practical Point:* Many patients with mitral valve prolapse have only a midsystolic click or multiple clicks without a late or holosystolic murmur. The absence of a murmur indicates the absence of mitral regurgitation.

Timing of the Click. The midsystolic click of mitral valve prolapse, in contradistinction to an ejection click, bears no relationship to ejection of blood from the heart. The clicks coincide with the precise moment of prolapse of

the leaflet or a scallop, which usually occurs after left ventricular systolic size has considerably decreased. Thus, the click is normally found in *mid- to late systole* and not during early systole (Fig. 18-4A). Some authors favor the term "nonejection click" to emphasize the difference between an ejection click or sound produced by a stiffened or stenotic semilunar valve. Nevertheless, the click of MVP may occur just after S1 (Fig. 18-4B), or it can be simultaneous with S1 when the onset of the prolapse occurs very early in systole (Fig. 18-4C). In such situations the billowing motion of the voluminous and redundant mitral leaflet begins at the onset of systole.

An early click may simulate a loud S1, a split S1, or an S1-ejection sound complex (Fig. 18-5). Similarly, when maneuvers that accentuate the discrepancy between the size of the mitral valve and left ventricular cavity size are carried out, the prolapse will appear earlier and the click will move closer to S1 (Fig. 18-6). When very early prolapse occurs, the click can move into S1 and a separate sound may not be audible; S1 will appear to be increased in intensity (Fig. 18-4C). Whenever one hears a loud "S1" in conjunction with a holosystolic murmur, it should suggest the diagnosis of holosystolic mitral leaflet prolapse.

POSITION OF THE SYSTOLIC CLICK OF MVP

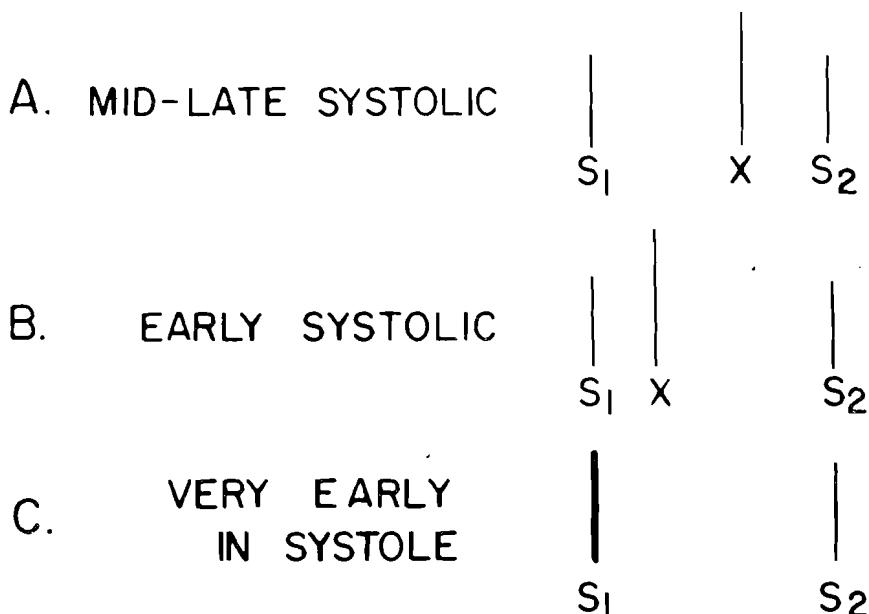


FIG. 18-4. Variable timing of the systolic click in mitral valve prolapse. While the click (X) is typically heard in mid to late systole (A), it may be heard in early systole if prolapse of one or more leaflets begins shortly after ejection (B). In holosystolic prolapse the first heart sound is often accentuated, suggesting superimposition of the click on the normal S1 (C).

DIFFERENTIAL DIAGNOSIS OF
AN EARLY SYSTOLIC CLICK
IN MITRAL VALVE PROLAPSE

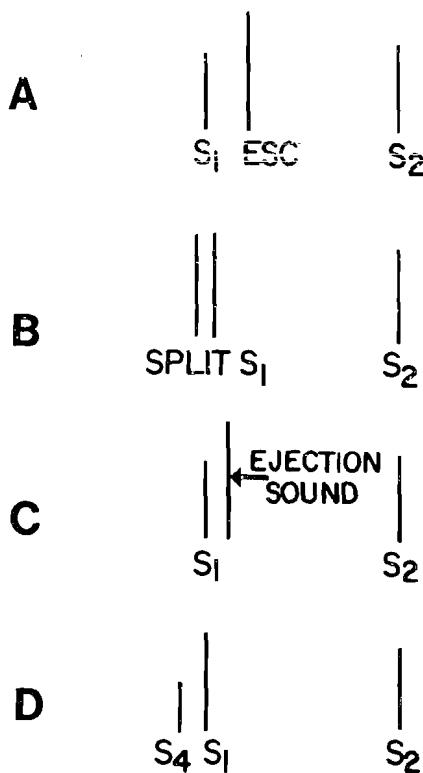


FIG. 18-5. Differential diagnosis of an early systolic click (see text).

Alterations in the timing of a nonejection click with pharmacologic agents or other maneuvers correlate closely with the behavior of the systolic murmur (Fig. 18-6, Table 18-4). Thus, the click moves closer to S_1 when the subject assumes the upright posture; the systolic murmur lengthens and the murmur vibrations extend towards S_1 . Squatting decreases the duration and extent of mitral prolapse by increasing left ventricular volume; both the click and murmur are heard later in systole (Fig. 18-6).

The Isolated Click. The diagnosis of MVP should be strongly suspected whenever a midsystolic click is heard. Many asymptomatic subjects will demonstrate a click or several clicks only and no murmur at all (Fig. 18-3). Occasionally, the presence of a click heralds the subsequent development of a systolic murmur or the findings of MVP months or years later. The murmur may be variable in its presence on a day-to-day basis. Several well-documented cases of bacterial endocarditis have been described occurring in persons who had a midsystolic click and *no* systolic murmur.

Optimal Auscultation of Systolic Clicks. Because clicks are often difficult to detect, one should be prepared to bring out these sounds during

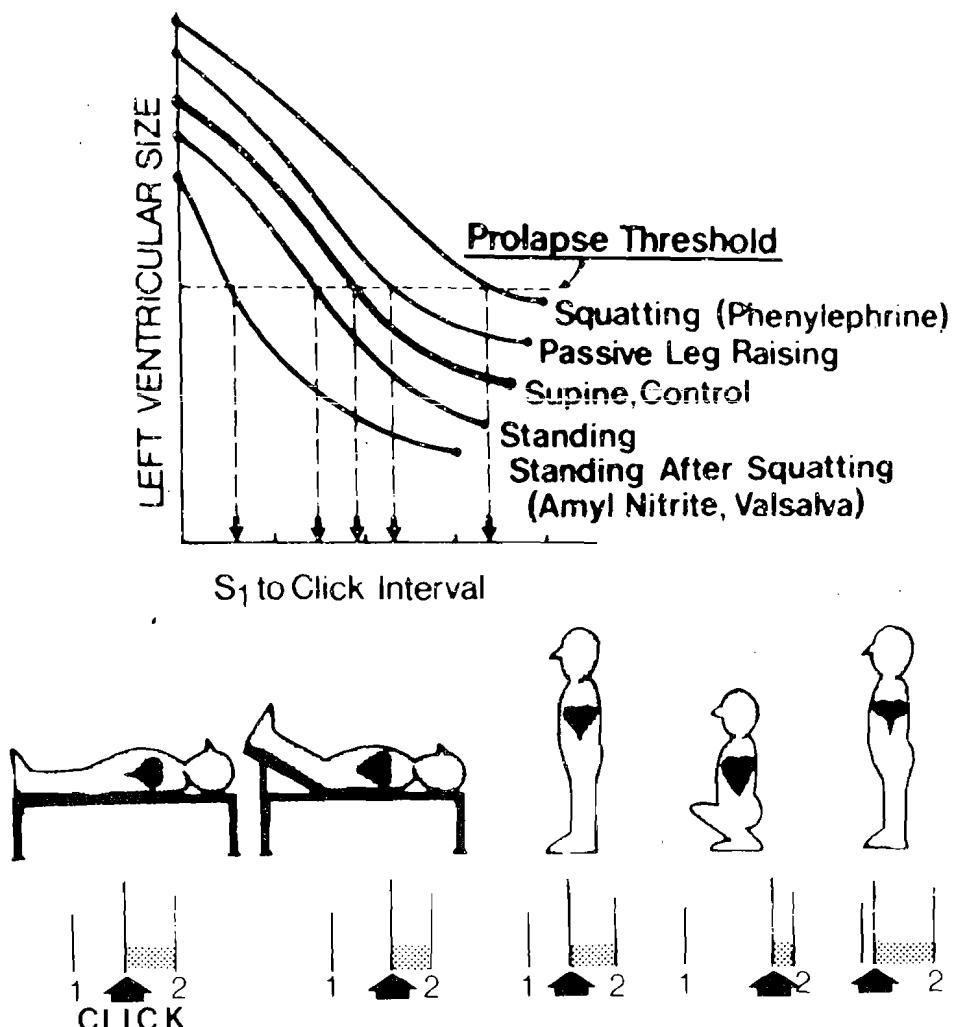


FIG. 18-6. Variation of the timing of the click and systolic murmur in mitral valve prolapse with changes in body position. The diagram indicates the presence of an arbitrary "prolapse threshold" and suggests a relationship between left ventricular size and the mitral valve that affects the timing and extent of leaflet prolapse. Whenever the prolapse threshold is reached during left ventricular systole, the leaflets prolapse, resulting in a click and systolic murmur. If the threshold is achieved in early systole as a result of reduced preload and/or afterload (e.g., standing position) the click moves closer to S1 and the murmur becomes longer and louder. Conversely, an increase in preload and afterload, such as occurs during squatting, results in the click and murmur appearing later in systole because the prolapse threshold is not reached until later during ejection. Pharmacologic agents may also affect left ventricular filling and outflow resistance and will alter ventricular geometry to produce predictable changes in the timing of the click and murmur. (From Criley JM and Heger J: Prolapsed mitral leaflet syndrome. In *Congenital Heart Disease in Adults*. Edited by WC Roberts. Philadelphia, FA Davis Co, 1979.)

auscultation. Firm pressure with the stethoscope diaphragm is optimal for detection of these high pitched, brief snapping sounds. Clicks are usually best heard at the apex or more medially towards the lower sternum. They do not radiate well unless they are very loud. Because a prominent systolic murmur may obscure or mask the presence of the midsystolic click, it is often best to listen away from the site of maximum murmur intensity.

TABLE 18-4 *Characteristic Variations with Different Maneuvers of Click and Murmur Produced by Mitral Valve Prolapse*

Intervention	Resultant Functional Change	Acoustic Consequences*
Upright posture, sitting or standing	Decrease LV volume, earlier prolapse	C moves closer to S1, often louder LSM M becomes longer, may become holosystolic; peaks earlier in systole M may become louder
Long cycle lengths (post-PVC beats, during atrial fibrillation, bradycardia)	Larger LV volume, later prolapse	No change or decrease in M intensity, shorter M, later C
Left decubitus position	Heart closer to stethoscope ? Other mechanisms	C and M may be louder
Inspiration	Decrease LV EDV volume, increase HR	C and M move toward S1 M unchanged or louder; may decrease in intensity C variably decreased in intensity
Valsalva maneuver		
Strain	Decrease LV EDV, increase heart rate	C earlier, softer M may transiently decrease in early phase (first 30 sec) M earlier, often louder later during strain M can become holosystolic, musical
Release	Increase LV EDV, bradycardia	Increase intensity of M and C to baseline by 6-8 beats after release M & C can occur later (if overshoot)
Isometric handgrip	Increase systemic vascular resistance, increase BP	Increase intensity of C and M C and M may occur earlier in systole
Squatting	Increase systemic vascular resistance Increased abdominal pressure Increase venous return Increase systolic BP Increase LVEDV Reflex bradycardia	Delay in onset of C and M (prolapse may disappear) Decrease M intensity; variable response

*M = murmur; C = click; LSM = late systolic murmur; EDV = end-diastolic volume; HR = heart rate; BP = blood pressure.

The maneuvers used to enhance detection of mitral valve prolapse are discussed later. Careful auscultation in the supine, left recumbent, and sitting or standing positions is important for optimal detection of the click and any accompanying murmur.

Differential Diagnosis of the Mid-Systolic Click (Fig. 18-7). A mid- or late systolic click can readily be mistaken for part of a widely split S2 complex if the click is assumed to be A2. An opening snap is another acoustic event simulated by mitral prolapse. In this case, the click is mistaken for S2 and the actual S2 is believed to be the opening snap (see Fig. 18-7). The presence of a late systolic murmur can add to this acoustic problem, as it may easily be mistaken for an early diastolic murmur if the click is confused for S2 (Fig. 18-8).

Diastolic Sound and Murmur

It is not generally appreciated that a high frequency diastolic sound and murmur may be found in some patients with MVP. This sound may also be associated with an early diastolic murmur simulating aortic regurgitation (see below). This murmur begins with A2 and, in fact, is continuous with the late

ACOUSTIC CONFUSION IN DETECTING THE CLICK AND MURMURS OF MVP

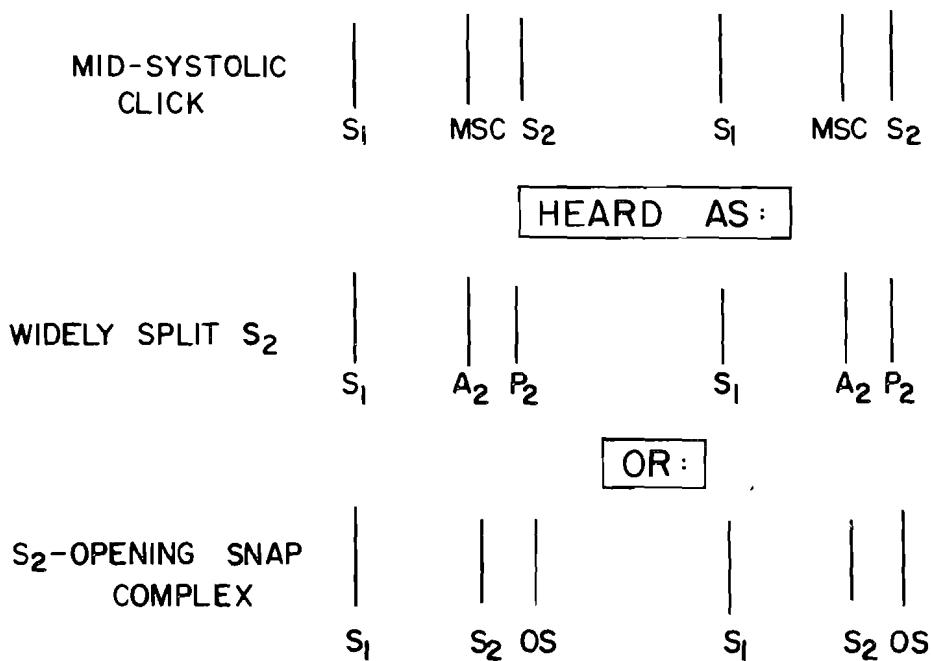


FIG. 18-7. Differential diagnosis of the midsystolic click. A midsystolic click (MSC)-S2 complex is frequently confused for wide splitting of the second heart sound and less often for an S2-opening snap sound complex. It is easy for the inexperienced physician to mistake a mid or late systolic click for S2 or one of its components.

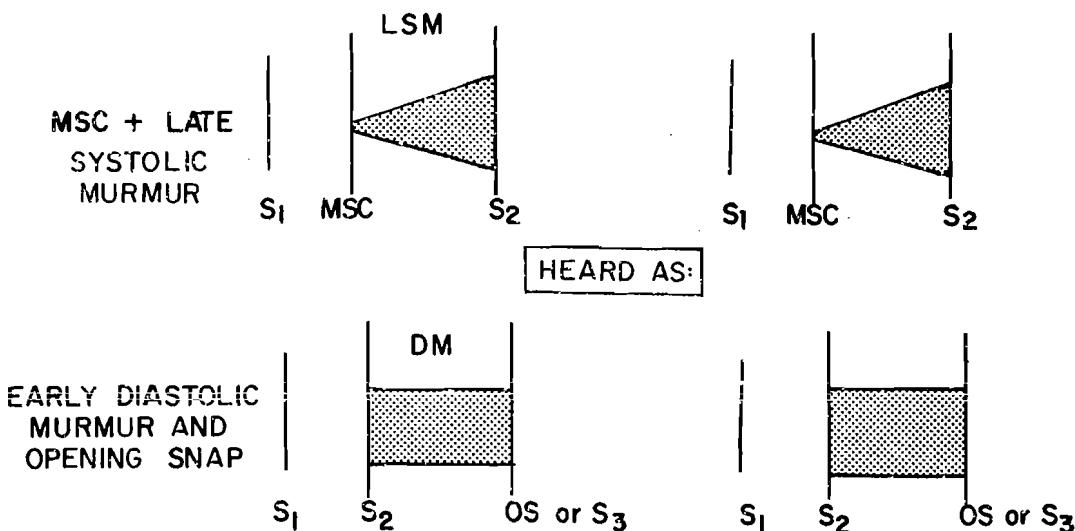


FIG. 18-8. Confusion of the midsystolic click-late systolic murmur for a diastolic murmur and opening snap. Because physicians are used to hearing the majority of murmurs in early to mid-systole, a late systolic murmur is frequently confused for a diastolic murmur, particularly when it follows a prominent click that is mistaken for S2. It is not uncommon for patients with classic findings of the click-murmur syndrome to be referred with a diagnosis of mitral stenosis.

systolic murmur of mitral regurgitation. This murmur has been described only in subjects with a diastolic sound. The sound, when present, occurs approximately 100 msec after A2; thus, its timing is identical to the opening snap found in mitral stenosis (Fig. 18-9). However, on simultaneous echophonocardiography the sound occurs *before* the maximal opening of the mitral leaflets and is coincident with the return of the prolapsing posterior mitral leaflet as it briefly recoaps with the anterior leaflet in early diastole. It has been postulated that early relaxation of the left ventricle sucks the prolapsed posterior leaflet back towards the ventricle, transiently bringing the two leaflets together.

This sound is very well heard at the base and apex and readily simulates an opening snap. Echocardiography is necessary to exclude mitral stenosis. The incidence of this unusual event is unclear, but it probably is found in no more than 5 to 10% of all patients with primary mitral valve prolapse. It is important for the astute clinician to be aware that in a small percentage of subjects with MVP, diastolic sounds and murmurs can be heard. The possible confusion caused by such diastolic events is obvious, and the differential diagnosis includes aortic regurgitation as well as mitral stenosis.

Murmur of Mitral Valve Prolapse

Characteristically, the murmur of mitral valve prolapse begins in mid-to late systole (Fig. 18-10). It typically fans out to S2 similar to all murmurs of late mitral regurgitation. The prolapse murmur sounds like the systolic

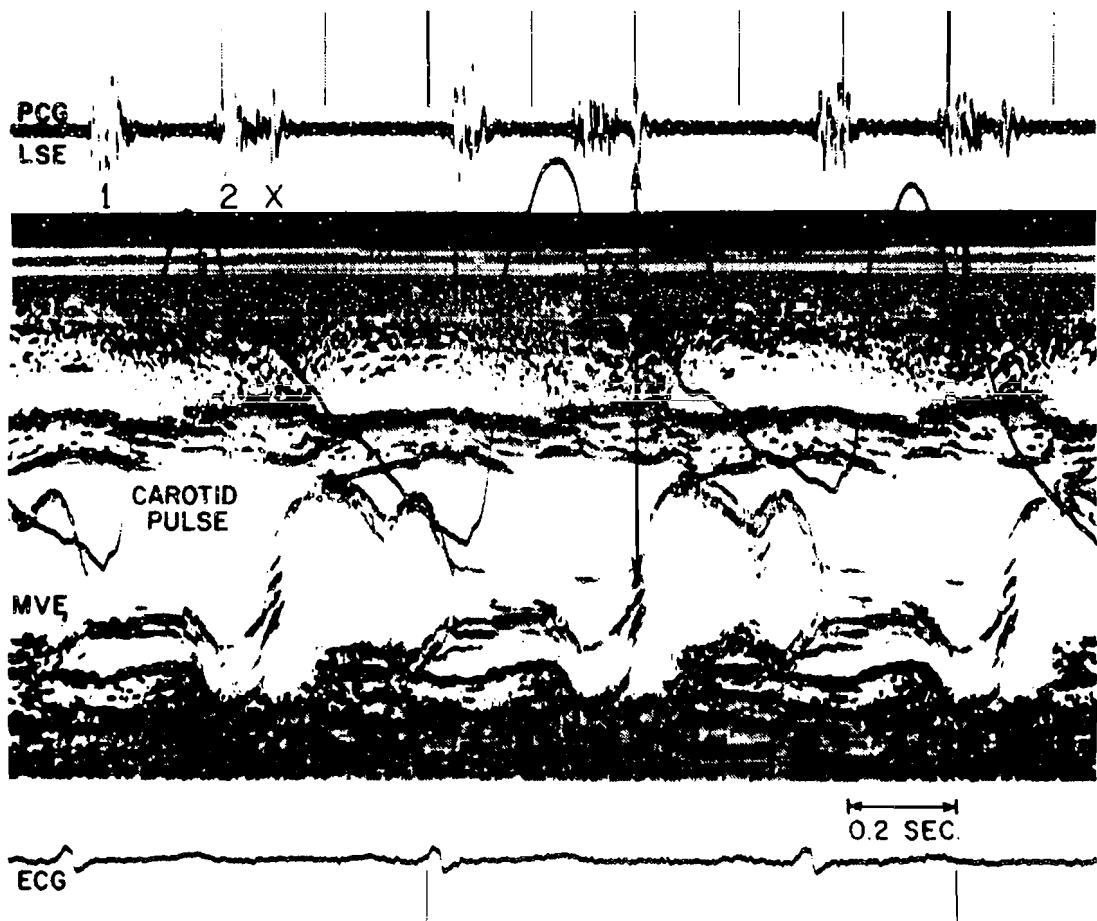


FIG. 18-9. Early diastolic sound associated with mitral valve prolapse. Note the high frequency sound in early diastole (X) that occurs just before the opening motion of the anterior mitral leaflet on echo (MVE). It has been postulated that this sound may relate to an early closing motion of the prolapsed posterior leaflet that moves toward the left ventricle and briefly coapts with the anterior leaflet of the valve. In this simultaneous echo and phonocardiogram, marked midsystolic prolapse is seen. The diastolic sound (X) coincides with the point of re-coaptation of the two leaflets on the echocardiogram. The sound occurs well before the E point, which represents maximal mitral valve opening, at the moment when the mitral opening snap would occur. Early diastolic murmurs have also been associated with mitral valve prolapse and a diastolic sound; their origin is also thought to be related to motion of the prolapsed leaflet in early diastole. (From Wei JY and Fortuin NJ: Diastolic sounds and murmurs associated with mitral valve prolapse. Circulation 63:559, 1981.)

murmur of papillary muscle dysfunction and, in fact, may be acoustically indistinguishable if there is no associated systolic click. Perhaps 30 to 40% of identified subjects with mitral valve prolapse have both a click and a murmur. A majority will have a click only, and a minority will have only a late systolic murmur. Definite MVP has been documented by M-mode or 2-D echocardiography or left ventricular angiography in individuals *without any auscultatory clues* for MVP, i.e., no murmur and no click. It is unclear whether these "silent" cases of prolapse fit into the broader MVP syndrome.

LATE SYSTOLIC MURMUR OF MVP

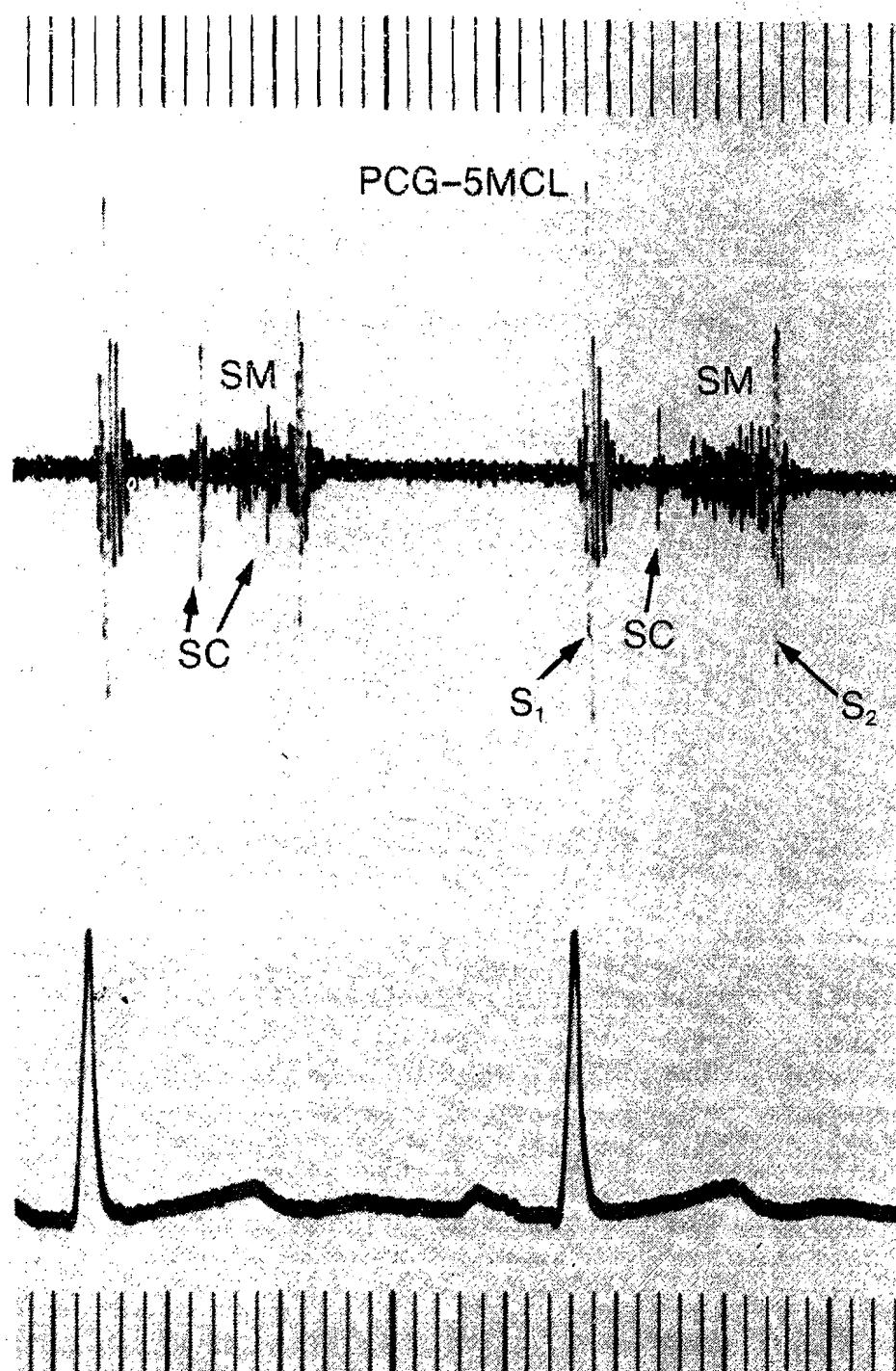


FIG. 18-10. The classic late systolic murmur of mitral valve prolapse. Note the crescendo configuration of the murmur, which begins in midsystole following the first systolic click. The frequency of this murmur is usually relatively pure. SM = systolic murmur. SC = systolic click. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co, 1979.)

Intensity. Because the degree of mitral regurgitation when present is usually mild, the systolic murmur typically is soft. However, the late systolic component occasionally displays considerable accentuation and may be quite prominent. If a systolic whoop or honk is present (see below), these auscultatory phenomena can be very loud indeed. Various maneuvers can either accentuate or diminish the amplitude of the murmur. In general, the loudness of the murmur of mitral regurgitation is related to the velocity and amount of refluxing blood into the left atrium. *Practical Point: The intensity of the MVP murmur is extremely variable. In some persons, the murmur may be inaudible in one position but grade 2-4/6 intensity in another position. The murmur may also vary several grades with various maneuvers.*

Frequency. Murmurs of MVP are usually medium to high pitched. They are typically of relatively pure frequency and have a whirring or regurgitant quality.

Timing. The mid- to late systolic nature of the murmur of mitral prolapse is characteristic, although there is considerable variability in the timing of murmur onset related to different body positions and various physical maneuvers. A prolapse murmur that begins in early systole or is clearly holosystolic suggests moderate to severe mitral regurgitation and indicates as well that the abnormal prolapsing or billowing leaflet motion begins with or near the onset of ejection. A pansystolic murmur of MVP occurs in approximately 10% of the subjects with MVP. In these patients, the degree of mitral regurgitation may result in left atrial and left ventricular enlargement. The term "floppy valve syndrome" has been loosely applied to such patients with MVP and major mitral regurgitation. Associated midsystolic clicks are not likely to be present in such individuals, although an increased S1 intensity is common (see pages 360, 362). The mitral annulus is often quite dilated in these patients. The diagnosis of a floppy mitral valve related to the myxomatous leaflet changes that are characteristic of primary MVP is best made by echocardiography, during left ventricular angiography, or at operation. This condition is probably the most common cause of severe chronic mitral regurgitation in adults.

Contour. The murmur of MVP is typically crescendo in late systole (Fig. 18-10). Less commonly, it may be crescendo-decrescendo in shape. In such cases, the peak intensity of the murmur coincides with the maximal degree of leaflet prolapse. Early systolic murmurs that taper in late systole (ejection quality) do not usually represent MVP. A pansystolic murmur, with or without associated clicks, can be caused by holosystolic prolapse. If there is late systolic accentuation to a pansystolic murmur, the underlying etiologic possibilities also include papillary muscle dysfunction, a calcified mitral annulus, or rheumatic mitral regurgitation.

Figure 18-11, prepared by Dr. W. Proctor Harvey, indicates most of the possible combinations of click, murmur, and whoop that may be found in

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S1

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FIG.
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cendo
WP:

MVP. Remember that these auscultatory phenomena may be quite variable from day to day, or even during the same examination.

Systolic Whoops and Honks. Occasionally, patients with mitral valve prolapse develop peculiar loud vibratory or musical systolic murmurs known as whoops or honks (Figs. 18-11, 18-12). Usually, these come and go in an intermittent fashion and may suddenly appear and disappear during auscultation. Changes in body position such as standing or sitting forward often initiate these murmurs. These peculiar murmurs probably are caused by an intense resonance or fluttering of mitral valve leaflet tissue or chordae tendineae at a uniform and periodic frequency, producing medium-to-high pitched, pure frequency musical vibrations (Fig. 18-12). Typically, the patient will have the classic acoustic findings of midsystolic MVP, which abruptly becomes more pronounced, initiating a whoop. These murmurs can be so loud that the patient may hear them. A careful observer may detect unusual murmurs without a stethoscope.

Tricuspid Valve Prolapse. Prolapse of both the mitral and tricuspid leaflets has been frequently documented by echocardiography, and isolated

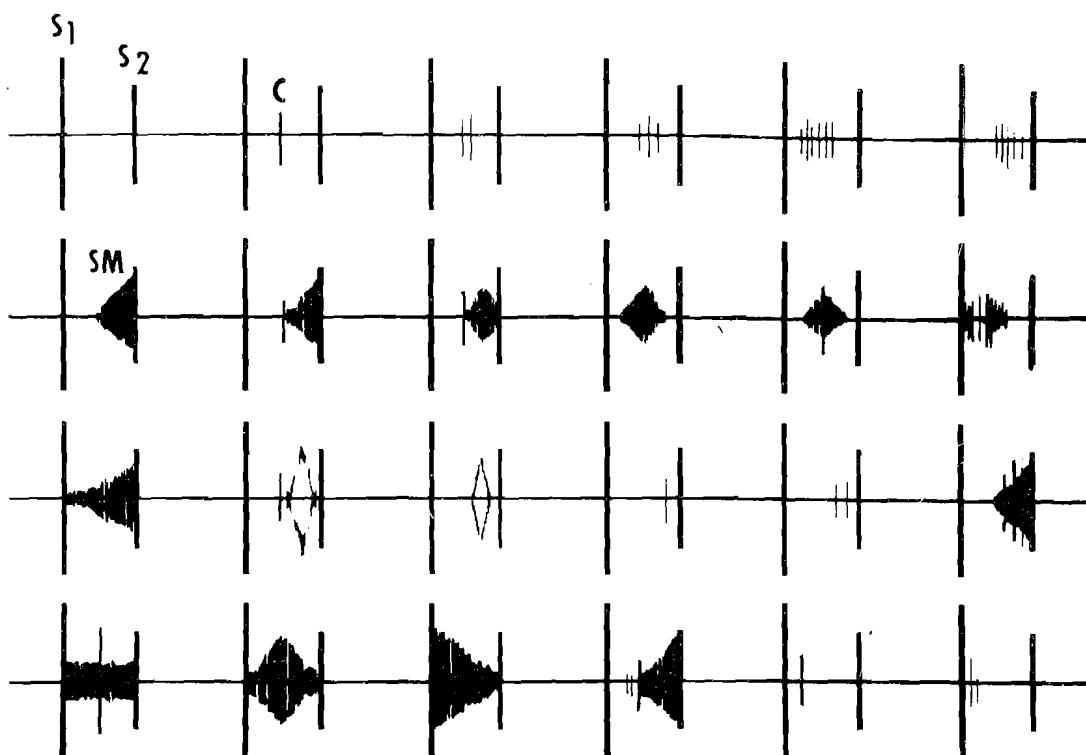


FIG. 18-11. Spectrum of physical findings in mitral valve prolapse. This diagram demonstrates a wide variety of clicks, murmurs, and click-murmur complexes that may be found in patients with mitral valve prolapse. The second and third sketches on the bottom line represent decrescendo tapering of a holosystolic murmur, typical of a ruptured chordae tendineae. (From Harvey WP: In Prolapsed mitral valve: The commonest valve disease? MD Cheitlin and RC Byrd. Curr Prob Cardiol 8:1, 1984.)

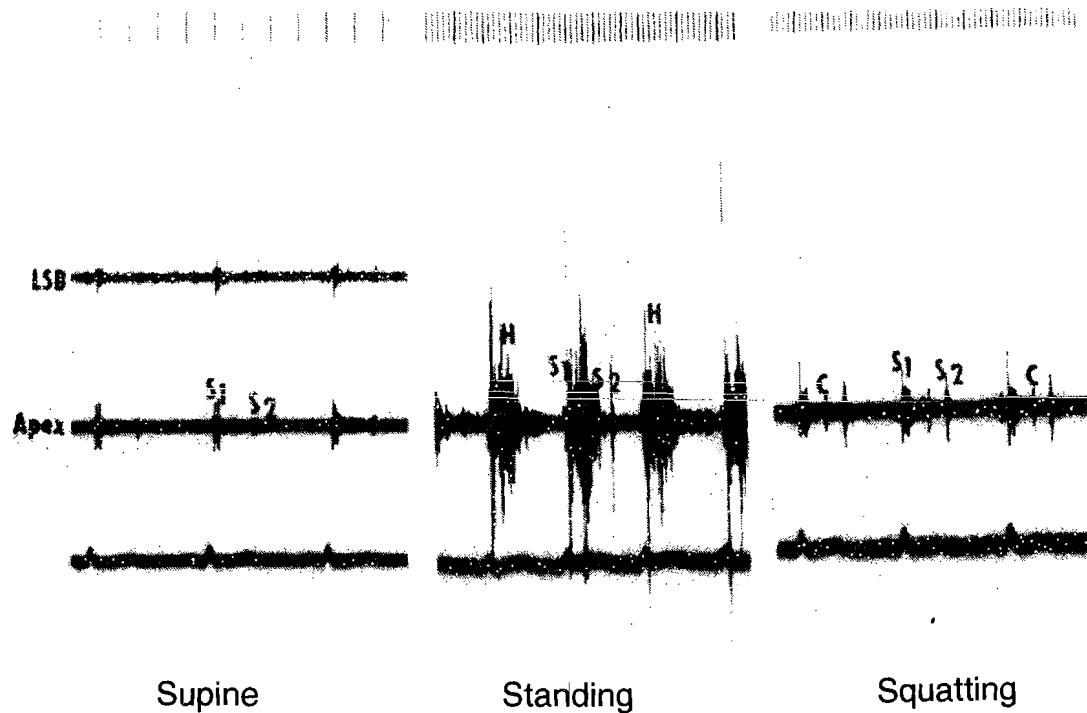


FIG. 18-12. Systolic whoop in mitral valve prolapse. In this phonocardiogram, a loud systolic whoop or honk appears when the patient is standing but disappears when the patient squats. The "prolapse threshold" is quickly reached as left ventricular volume decreases in the upright position, resulting in sudden prolapse of one or both leaflets and a peculiar resonating vibration of the leaflet tissue that produces the pure frequency musical sound of the systolic whoop. Note that the patient has a midsystolic click in the squatting position. Systolic whoops and honks may be very loud and may vary from day to day and with changes of body position. H = honklike musical murmur or whoop. C = midsystolic click. (From Sheikh MU, et al: Musical murmurs: Clinical implications, long-term prognosis, and echo-phonocardiographic features. Am Heart J 108:377, 1984.)

tricuspid valve prolapse may occasionally occur (Fig. 19-2). The physical findings of tricuspid prolapse are almost identical to those of mitral valve prolapse. Consideration should be given to this diagnosis when a click and/or murmur is especially well heard at the lower left sternal border and/or if the late systolic murmur increases with inspiration.

EFFECTS OF BODY POSITION, DRUGS, AND PHYSICAL MANEUVERS (Table 18-4, Fig. 18-6)

The use of ancillary maneuvers and pharmacologic agents in auscultation is most rewarding in several cardiac conditions, such as hypertrophic cardiomyopathy and mitral valve prolapse (Chapter 11). The murmurs of both these disorders behave in a similar fashion with *most* but *not all* interventions: in both conditions the underlying pathophysiologic abnormality is accentuated by any alteration that produces a *smaller left ventricular cavity* or an *increase in the force of contraction*. In MVP, such maneuvers cause the prolapse to

occur earlier in systole, moving the click(s) and murmur closer to S1 and increasing the duration of the mitral regurgitation, if present. The opposite occurs when left ventricular cavity size abruptly increases. Alterations in left ventricular preload and afterload also produce predictable changes in the timing of the onset of the click and murmur. As a rule, the *intensity* of the murmur of MVP responds to interventions in the same manner as other types of mitral regurgitation (e.g., an increase in murmur amplitude and degree of mitral reflux with elevation of systolic blood pressure).

Table 18-4 summarizes the typical responses of the click and murmur to a variety of altered physiologic states and body positions. *Practical Point:* *The behavior of the timing of the click and murmur is of greater diagnostic usefulness than changes in intensity.* Often the acoustic abnormalities may be minimal or even absent at rest in the supine position, but are obvious when the patient is upright or turned onto his left side. *Practical Point:* *In any patient suspected of or known to have MVP, auscultation should be carried out in the supine, left lateral, sitting forward, and standing positions. It is helpful to listen while the subject squats and then abruptly returns to the standing position (Fig. 18-6).*

DIFFERENTIAL DIAGNOSIS

For the experienced clinician, the classic finding of a prominent non-ejection click and a late systolic murmur should not be confused for other cardiovascular abnormalities. A holosystolic murmur, if present, must be differentiated from other causes of mitral regurgitation, such as rheumatic heart disease; the diagnosis of MVP in such cases is suggested if there is a prominent click (rare) or a loud S1, but the etiology must be confirmed by an echocardiogram. Prominent alteration of murmur intensity with varying body positions suggests prolapse as the etiology of the holosystolic murmur. *Postmitral commissurotomy valves* may prolapse; such patients tend to have low frequency clicks that may be quite loud. The click and murmur tend to change less with alterations or various maneuvers in such subjects. *Hypertrophic cardiomyopathy* may be confused with mitral valve prolapse in patients who have only a mid to late systolic murmur; the presence of an S4, the absence of a click, and exaggeration of the murmur following a PVC strongly favor hypertrophic cardiomyopathy. The murmur in this condition usually has prominent ejection characteristics (Chapter 14).

The late systolic murmur of *papillary muscle dysfunction* typically is accompanied by an S3 or S4, along with some evidence of left ventricular dysfunction (Fig. 17-11). Papillary muscle murmurs do not increase in the upright posture. A midsystolic click is not heard. However, auscultatory evidence of MVP has been documented in patients with coronary artery disease following myocardial infarction. Thus, a late systolic murmur that

fans out to S2 can be a manifestation of either MVP or papillary muscle dysfunction. Patients with *coarctation of the aorta* may have a late systolic murmur and an early (ejection) click. The maximal intensity of the murmur away from the cardiac apex (high left chest and interscapular) should exclude mitral valve prolapse. The systolic murmur of coarctation also may spill into diastole.

In severe MVP the click may occur early in systole and simulate an ejection sound or produce a loud S1. This is uncommon and, when present, is typically associated with a holosystolic murmur.

Confusion of the late systolic murmur with a diastolic murmur and a mistaken identification of a midsystolic click-S2 complex for a wide splitting of S2 or an opening snap have been discussed (Figs. 18-7, 18-8). Careful inching with the stethoscope away from the base of the heart is important in uncertain cases. One first identifies S2 and then remains focused on the sequence of S1 and S2 at different precordial sites for the proper identification of diastole and systole.

Chapter 19

Tricuspid Regurgitation

Tricuspid regurgitation is not usually viewed as an important valve lesion; yet this abnormality is far more common than generally appreciated. While isolated tricuspid regurgitation is rare, tricuspid regurgitation associated with existing cardiac disease often can be detected on careful physical examination. A common denominator of tricuspid regurgitation in adults is the presence of severe pulmonary hypertension. Signs of elevated pulmonary artery pressure (increased P₂, right ventricular lift) always should stimulate a careful search for tricuspid regurgitation; conversely, any suggestion of tricuspid regurgitation on clinical examination should provoke a meticulous evaluation for evidence of an increased pulmonary artery pressure.

FUNCTIONAL ANATOMY

The tricuspid valve consists of a septal, an anterior, and a posterior leaflet, each anchored via the chordae tendineae by a series of papillary muscles located in the right ventricle (Fig. 19-1). There are several varieties of chordae with an average of 25 chordae per leaflet. The septal leaflet is anchored to the right ventricular septal wall by short, obliquely positioned chordae. The anterior cusp is the most mobile leaflet. The posterior leaflet consists of a series of scallops.

As with the mitral valve it is useful to think of a *tricuspid valve complex*. Tricuspid valve incompetence may be due to disease or abnormalities of the tricuspid leaflets, chordae, papillary muscles, right ventricular myocardium, annulus, or a combination of the above. While a large number of conditions can effect tricuspid valve function, acquired isolated tricuspid incompetence is rarely found without associated cardiovascular abnormalities except following infective endocarditis or cardiac trauma.

Many authors have commented that the tricuspid valve is not optimally designed for conditions of increased right ventricular volume or pressure. Although the valve functions well when the right ventricular is normal, elevations in RV diastolic volume or of pulmonary artery systolic pressure often result in incompetence of the tricuspid valve. This may be a transient or variable phenomenon, most likely related to intermittent dilatation of the tricuspid annulus. The threshold for functional or secondary tricuspid regurgitation is low; reflux of blood into the right atrium occurs readily in the absence of organic tricuspid valve disease, particularly in the presence of an



FIG. 19-1. Normal tricuspid valve. The three leaflets are shown, although the posterior leaflet is incomplete because of the dissection technique. S = septal leaflet; P = posterior leaflet; A = anterior leaflet. The fan-shaped chordae (F) define the commissures between the leaflets. Note that there are several large papillary muscles, each giving rise to chordae to more than one leaflet. Chordae also arise from smaller papillary muscles. (From Wooley CF: Rediscovery of the tricuspid valve. *Curr Prob Cardiol* 6:1, 1981.)

RV pressure overload. In general, pulmonary artery systolic pressure must be at least 50 mmHg to produce tricuspid annular dilatation in the presence of a normal tricuspid valve. When there is intrinsic disease of the tricuspid valve (e.g., rheumatic valvulitis), lesser degrees of pulmonary hypertension may result in tricuspid regurgitation. Biventricular congestive heart failure with a high right ventricular end-diastolic pressure may cause tricuspid incompetence even in the absence of severe pulmonary hypertension. Hypervolemic states can induce tricuspid regurgitation. Right ventricular dysfunction from any cause (e.g., RV infarction, cardiomyopathy) may produce tricuspid regurgitation.

ETIOLOGY (Table 19-1)

Organic or Primary Disease of the Tricuspid Valve. Rheumatic fever affects the tricuspid valve, and autopsy evidence of tricuspid valvulitis is not infrequent. Clinically apparent tricuspid disease, manifest as tricuspid regurgitation, tricuspid stenosis, or a combination of both lesions, is present in less than 20% of patients with overt rheumatic heart disease. Infective endocarditis

TABLE 19-1 *Etiology of Tricuspid Regurgitation*

ORGANIC INVOLVEMENT OF THE TRICUSPID VALVE	
Rheumatic valvulitis	
Infective endocarditis	
Trauma to the heart	
Acute right ventricular infarction	
Carcinoid syndrome	
Tricuspid valve prolapse	
Ebstein's anomaly	
FUNCTIONAL ABNORMALITIES OF THE TRICUSPID VALVE	
Right ventricular dilatation—acute or chronic	
Pulmonary hypertension	
Combination of RV volume and pressure overload	

is the most common cause of isolated tricuspid regurgitation in adults and is found almost exclusively in drug abusers. Right-sided endocarditis is common in heroin addicts (present in 5% of this population); involvement of the tricuspid valve is far more frequent than that of the pulmonic valve. Cardiac trauma can produce tricuspid regurgitation by disruption of the RV papillary muscles, chordae tendineae, or the individual tricuspid valve leaflets. Although penetrating injuries can injure the valve, blunt chest trauma is a more common cause of tricuspid incompetence. A motor vehicle accident that produces sternal compression during diastole with simultaneous obstruction to right ventricular outflow is one mechanism that can result in traumatic tricuspid regurgitation. The actual episode of trauma may have occurred months to years earlier. Steering wheel and motorcycle accidents are particularly suspect.

Right ventricular infarction resulting from right coronary artery occlusion and acute inferior wall myocardial infarction is a recently identified cause of tricuspid regurgitation. Tricuspid regurgitation may result from RV papillary muscle fibrosis or rupture, as well as from severe septal and right ventricular wall dyskinesis. An iatrogenic cause of tricuspid regurgitation is transvenous placement of a pacemaker in which the electrode catheter is positioned across the tricuspid valve.

A variety of unusual systemic diseases can result in tricuspid regurgitation. Carcinoid syndrome resulting from a rare serotonin-producing tumor of the intestine may result in a peculiar right heart fibrosis with the under-surface of the tricuspid and pulmonary valve leaflets becoming adherent to the RV septal wall. Scleroderma and systemic lupus erythematosus have also been uncommonly associated with tricuspid regurgitation. Right heart tumors (myxoma) and endocardial fibroelastosis are other rare causes of tricuspid incompetence.

Tricuspid valve prolapse is another potential cause of tricuspid regurgitation (see Chapter 18). In most subjects, this condition is found in association with mitral valve prolapse and is difficult to recognize clinically. High quality echocardiographic tracings are necessary to make this diagnosis (Fig.

19-2). The incidence of tricuspid regurgitation in tricuspid prolapse is unclear, but the condition has been reported in up to 48% of affected subjects. The prevalence of tricuspid valve prolapse in subjects with mitral valve prolapse varies markedly in the literature.

Congenital abnormalities of the right heart can result in tricuspid regurgitation. By far the most common is Ebstein's anomaly in which the tricuspid valve is abnormally displaced into the RV cavity resulting in a small RV chamber and a very large right atrium, which is comprised of both right atrial and right ventricular muscle. Often, this lesion is associated with an atrial septal defect or patent foramen ovale with right to left shunting.

Functional or Secondary Tricuspid Regurgitation. Incompetence of the tricuspid valve resulting from RV dilatation or elevation of RV systolic and diastolic pressures is the most common cause of tricuspid regurgitation in adults. Often this condition is present in patients with moderate to severe pulmonary hypertension. Rheumatic mitral valve disease, particularly mitral stenosis, is a common cause of secondary tricuspid regurgitation. It is often

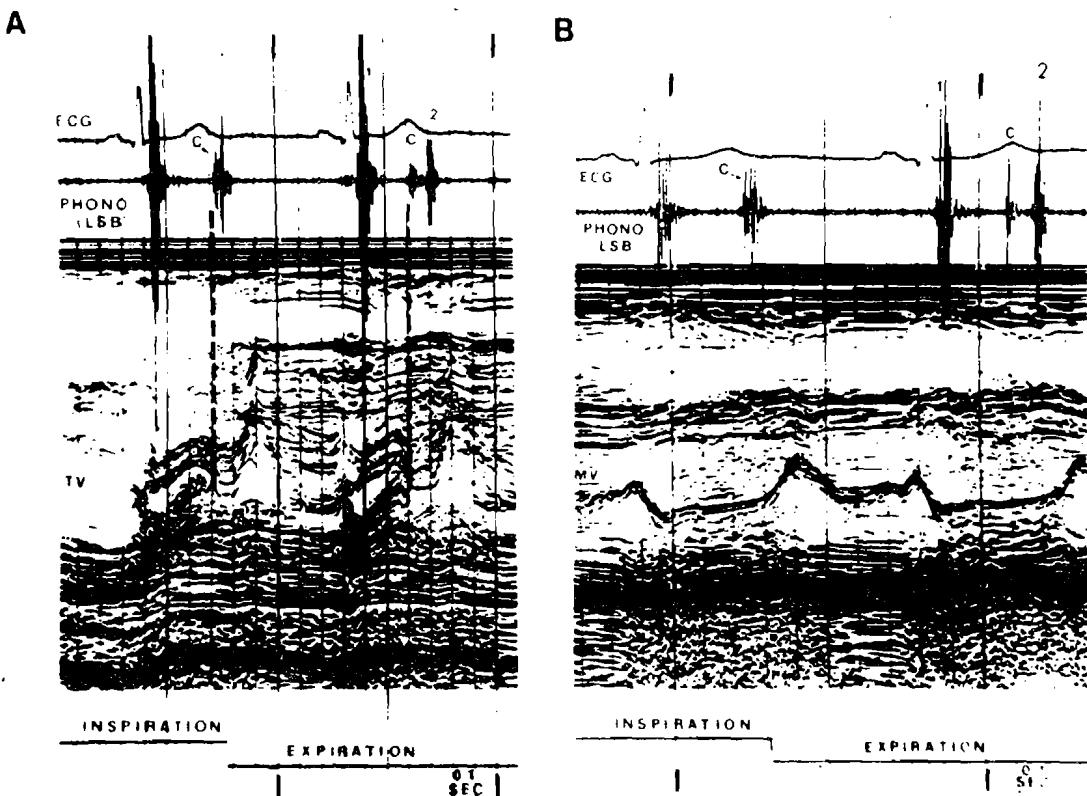


FIG. 19-2. Tricuspid valve prolapse. A. This echophonocardiogram reveals a late systolic click (C) that moves into S2 with inspiration and is widely separated from S2 during expiration. The echo demonstrates tricuspid valve prolapse. The timing of prolapse changes with the respiratory cycle. B. The mitral valve echocardiogram in the same patient shows no evidence of prolapse. (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 26:145, 1983.)

impossible to exclude organic rheumatic involvement of the tricuspid valve in patients with bivalvular abnormalities and pulmonary hypertension.

Tricuspid regurgitation may be produced by pulmonary hypertension of other etiologies such as primary pulmonary hypertension, major pulmonary emboli, and severe cor pulmonale. Severe left ventricular failure from cardiomyopathy or other causes can be associated with an incompetent tricuspid valve, especially during periods of cardiac decompensation.

Tricuspid regurgitation is a common accompaniment of pulmonary hypertensive atrial septal defects and may be present in any patient with Eisenmenger reaction.

PATHOPHYSIOLOGY

Importance of Pulmonary Artery Pressure

When discussing the consequences of tricuspid regurgitation, it is useful to categorize patients into two groups: those with a normal pulmonary artery pressure and those with pulmonary hypertension. While the resultant clinical sequelae ultimately may be similar in subjects with a severely incompetent tricuspid valve, there are significant differences on physical examination between these two groups.

Normal Pulmonary Artery Pressure. In the absence of elevation of pulmonary artery pressure, the RV faces a pure volume overload in tricuspid regurgitation. In general, the magnitude of the valvular incompetence is less than when pulmonary hypertension is present. The normal right ventricle and the right atrium are relatively compliant; systemic venous distension is not seen in "low pressure" tricuspid regurgitation unless the tricuspid leak is enormous or the severe RV volume overload results in RV dysfunction. The volume and velocity of blood refluxing from the RV into the right atrium and great veins are generally modest; thus, the classic tricuspid murmur and elevated jugular venous pressure with V waves may be attenuated or absent. The right atrial pressure contour shows a loss of the X descent and a large V or regurgitant wave, but these findings are usually modest (Fig. 4-4). *Practical Point: An inspiratory increase in the intensity of the systolic murmur and the amplitude of venous V wave are of great diagnostic importance in patients with low pressure tricuspid regurgitation.* Such subjects often have no abnormal findings during expiration. In these patients, the murmur of tricuspid regurgitation is heard only in inspiration. Systemic sequelae, such as hepatomegaly and edema, are uncommon in chronic low pressure tricuspid regurgitation unless the volume overload is very large.

Elevated Pulmonary Artery Pressure. Tricuspid regurgitation resulting from or complicated by pulmonary hypertension tends to be severe. Decreased compliance of the hypertrophied right ventricle will augment the magnitude

of the tricuspid regurgitation. In patients with rheumatic mitral valve disease or chronic congestive heart failure, tricuspid regurgitation is seen only in subjects with mean right atrial pressures greater than 8 to 10 mmHg, suggesting that a high RV diastolic pressure is important in the production of functional tricuspid regurgitation. The right ventricle in chronic severe pulmonary hypertension has an increased diastolic pressure and volume (chamber dilatation); ultimately, the tricuspid valve ring or annulus dilates.

Effects on Right Atrial Dynamics

The right atrial X descent is attenuated in mild tricuspid regurgitation and is completely obliterated with moderate to severe tricuspid reflux (Fig. 4-4). A systolic regurgitant wave replaces the normal decrease in right atrial and jugular venous pressure during systole. Although the right atrial A wave may be increased early in the course of right ventricular hypertrophy, eventually a dominant V (or "CV") wave appears. In severe tricuspid reflux the systolic wave may be sustained and have a plateau configuration (Figs. 4-4, 19-3, 19-6).

During systole and following right atrial contraction, the tricuspid annulus normally contracts or narrows by 20 to 30% of its circumference. This sphincter-like action probably aids in keeping the tricuspid valve competent during RV systole. In tricuspid regurgitation, there is a reduction in this physiologic annular narrowing, which could contribute to the tricuspid insufficiency. Similarly, loss of RA contraction by atrial fibrillation might adversely affect optimal RV coaptation and enhance the appearance of tricuspid regurgitation. Severe tricuspid regurgitation causes the RV to dilate further; thus, tricuspid regurgitation begets more tricuspid regurgitation. The tricuspid valve orifice may be enormous. The right ventricle and tricuspid annulus dilate in response to major A-V valve regurgitation.

Severe tricuspid regurgitation causes retrograde systolic flow with reversal of flow in the great veins. Filling of the right heart occurs only in diastole. The right atrial Y descent becomes sharp and steep, reflecting rapid and voluminous right ventricular filling that takes place in the presence of an elevated right atrial pressure (Figs. 19-3, 19-6).

Systemic Effects

Chronic systemic venous pressure elevation may cause congestion of the liver and bowel. Hepatomegaly with parenchymal dysfunction of the liver and ascites are not unusual in chronic severe tricuspid regurgitation. Decreased renal perfusion and low cardiac output lead to further fluid retention and edema.

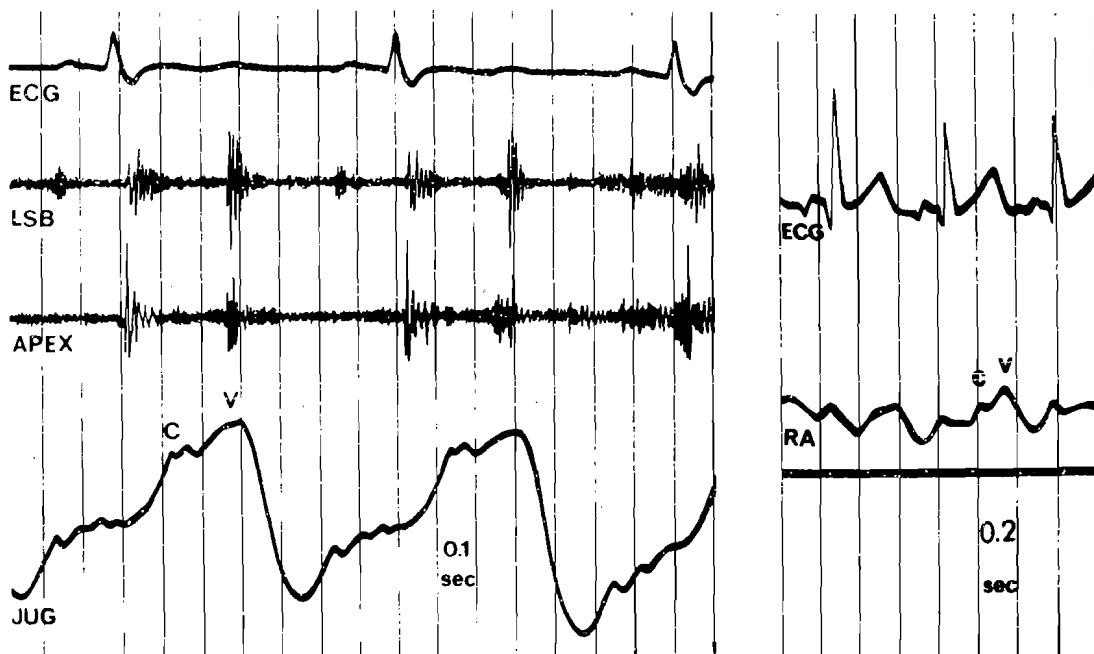


FIG. 19-3. Jugular venous contour in tricuspid regurgitation. In the left panel, a jugular venous tracing (JUG) is shown demonstrating an enormous C-V wave with loss of the X descent. On the right, a direct right atrial (RA) tracing is shown but with a different pressure scale. Notice the similarity in the waveform contour between the right atrium and the jugular venous pulse. (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 26:145, 1983.)

Severe tricuspid regurgitation may unload the left heart when the RV regurgitant fraction is very large; a smaller proportion of the RV stroke volume reaches the pulmonary capillary circulation and left atrium. The signs and symptoms of backwards failure (orthopnea, dyspnea on exertion, rales) may diminish when severe tricuspid regurgitation occurs, but at the expense of a further decrease in cardiac output.

Effects of Respiration

The normal physiologic inspiratory increase of venous return to the right heart is of great importance in cardiac physical diagnosis when evaluating abnormalities of the tricuspid valve. If the tricuspid valve is incompetent, the large inspiratory RV stroke volume refluxes into the right atrium. For any given RV regurgitant fraction, the actual volume and velocity of RV blood refluxing across the tricuspid valve into the right atrium will increase during inspiration. Thus, inspiration increases the degree of tricuspid regurgitation. The inspiratory augmentation of regurgitant flow results in a greater amplitude (and often duration) of the jugular venous V wave as well as the murmur of tricuspid regurgitation. The behavior of the murmur during the respiratory cycle provides a basis for the definitive diagnosis of tricuspid regurgitation.

Practical Point: *Inspiratory augmentation of the systolic murmur is a classic physical finding in tricuspid regurgitation and is known as Carvallo's sign (Fig. 19-4).*

Failure of Murmur to Increase in Inspiration

If the right ventricle is unable to increase its end-diastolic volume further during inspiration, there can be no additional inspiratory augmentation of regurgitant flow or murmur enhancement. This situation occurs when the degree of tricuspid regurgitation is massive and RV diastolic volume is maximal and/or the right ventricle has failed and can no longer alter its stroke volume during the respiratory cycle. In such cases, there will not be respiratory alteration in murmur intensity.

Diastolic events such as an RV S3 and RV S4, a tricuspid opening snap, and a tricuspid mid-diastolic flow rumble are of greater intensity during inspiration in patients with tricuspid valve disease. The presystolic and mid-diastolic murmurs of tricuspid stenosis will also demonstrate inspiratory augmentation.

Atrial Fibrillation

In chronic tricuspid regurgitation, particularly when moderately severe, atrial fibrillation is a common finding. In some series, this is present in greater than 85% of patients, most of whom have chronic valvular heart disease.

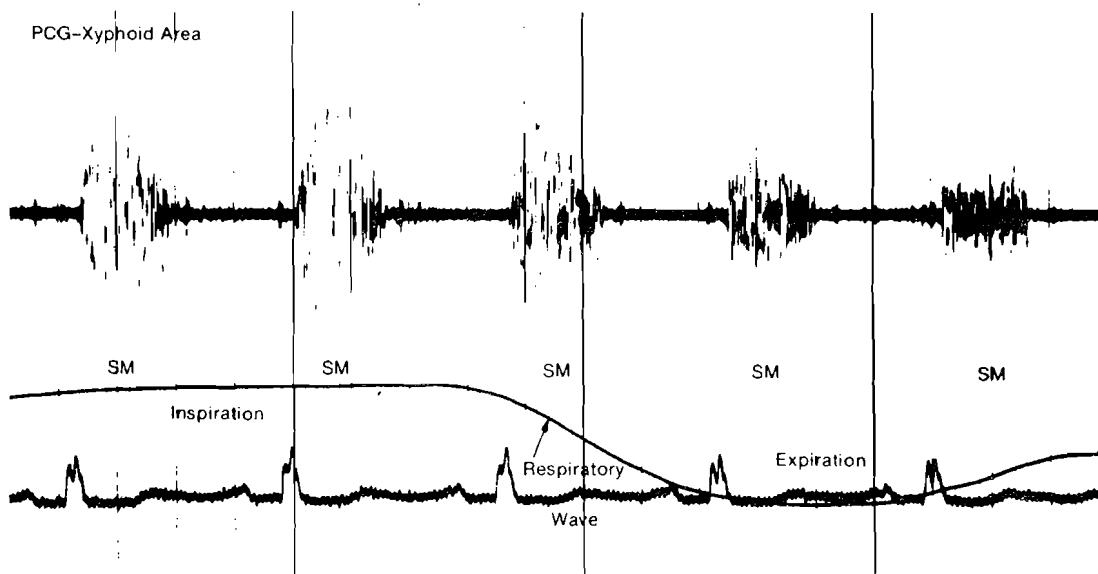


FIG. 19-4. Inspiratory augmentation of the murmur of tricuspid regurgitation. Note the dramatic increase in amplitude of the systolic murmur (SM) during inspiration. Augmented right ventricular filling and a greater tricuspid regurgitant volume during inspiration produces a louder murmur. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co, 1979.)

PHYSICAL EXAMINATION

General Appearance

In mild tricuspid regurgitation there are no alterations in the physical appearance. Chronic severe tricuspid regurgitation, usually in association with pulmonary hypertension, frequently results in sustained elevation of systemic venous pressure, which can result in hepatic and renal dysfunction. Some authors have observed that severe tricuspid regurgitation is a systemic process. Such patients may demonstrate indirect evidence of long-standing tricuspid valve disease. Noncardiac physical findings associated with tricuspid regurgitation are listed in Table 19-2. All are related to marked elevation of the systemic venous pressure and large transmitted V waves.

The Liver. Typically, the liver is enlarged in long-standing tricuspid regurgitation. The liver may be tender or hard and insensitive to palpation (cardiac cirrhosis). The liver usually is *pulsatile*, a finding frequently missed at the bedside (Fig. 19-5). The low amplitude hepatic pulsations directly reflect reversed systolic blood flow in the great veins. A pulsatile liver must be distinguished from the pulsations of a normal, a dilated, or a tortuous abdominal aorta. The latter are more medial, and the pulsations are directed anteriorly. Bimanual palpation of the liver can be useful in this differentiation; hepatic pulsations tend to be expansile in all directions. The more rightward and lateral in the abdomen the liver can be felt, the easier it is to distinguish a hepatic impulse from that of the abdominal aorta. The right lobe of the liver and the right atrium can become quite enlarged in tricuspid regurgitation and may produce a rightward and anterior motion to the lower right chest and right upper quadrant.

Practical Point: *The low-amplitude hepatic pulsations of tricuspid regurgitation often are better seen than felt. The patient should be asked to hold his breath (fixing the diaphragm) in deep inspiration while the clinician carefully observes the examining fingers and hand for motion with each cardiac cycle.* Close attention should be given to the hepatic motion during held inspiration. In patients with atrial fibrillation, it is more difficult to detect

TABLE 19-2 Noncardiac Manifestations of Severe Tricuspid Regurgitation

Head bob
Proptosis
Anterior motion of eyes
Engorged neck
Hepatomegaly
Pulsatile liver
Splenomegaly
Ascites
Peripheral edema
Mild icterus
Acrocyanosis
Cachexia—weight loss, muscle wasting

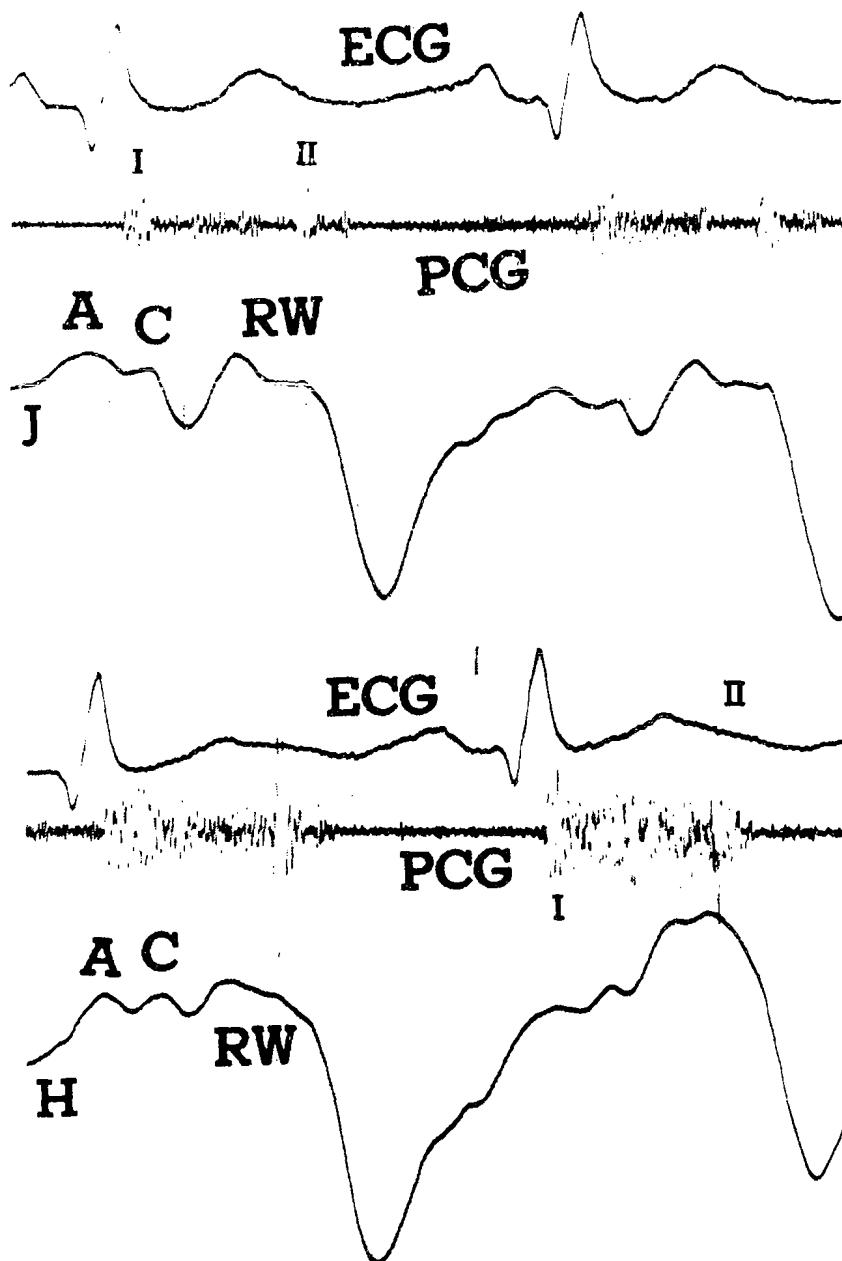


FIG. 19-5. Hepatic pulsations in tricuspid regurgitation. This figure displays recordings from the jugular venous pulse (J) and liver (H) in a middle-aged male with alcoholic cardiomyopathy, heart failure, and tricuspid regurgitation. Note the prominent regurgitant wave (RW), the loss of the X descent, and the steep Y descent (unlabeled). A = A wave; C = C wave. The hepatic and jugular venous tracings are almost identical in contour. (From Luisada AA, Singhal IA, and Robles R: Diagnosis of tricuspid insufficiency by non-invasive methods. *Angiology* 35:139, 1984.)

inspiratory augmentation because of the variability in regurgitant volume from beat to beat; nevertheless, liver pulsations should be readily detectable.

Chronic passive hepatic distention may produce low grade jaundice and scleral icterus. Liver function tests will be deranged. A protein-losing enteropathy has been described in chronic severe tricuspid regurgitation as a result of lymphangectasia and direct seepage of protein into the gut. The resultant

depression of serum proteins further potentiates formation of edema. Splenomegaly has been observed in these patients and is a direct consequence of chronic passive congestion of the systemic venous bed. The low cardiac output typical of patients with chronic tricuspid regurgitation may cause mild cyanosis of the lips, fingers, and toes. The combination of cyanosis and icterus can give some patients a sickly, sallow hue.

Fluid Retention. Most subjects with chronic tricuspid regurgitation have chronic edema, which may be marked. Ascites is also common, reflecting hepatic venous and lymphatic congestion as well as renal salt and water retention. A low serum albumin may accentuate this problem.

In end-stage rheumatic heart disease or severe long-standing pulmonary hypertension, patients with tricuspid regurgitation will demonstrate muscle wasting and weight loss (cardiac cachexia) often associated with edema and ascites. Patients with severe tricuspid regurgitation and associated cardiac lesions typically have little evidence of pulmonary congestion. Such subjects can lie relatively flat without orthopnea or paroxysmal nocturnal dyspnea.

Arterial Pulse

There are no characteristic alterations of the carotid pulse in tricuspid regurgitation. The arterial pulse amplitude may be diminutive if left ventricular stroke volume is abnormally small. This is a result of the hemodynamic effects of tricuspid regurgitation and associated cardiac abnormalities. Many patients with chronic severe tricuspid regurgitation will be in atrial fibrillation, whereas subjects with tricuspid regurgitation of recent onset or mild degrees of valve incompetence are likely to remain in normal sinus rhythm.

It is easy to confuse the dramatic swelling of the jugular veins found in patients with severe tricuspid regurgitation (see below) with the arterial pulse. These pulsations (V or regurgitant waves) are systolic in timing and may be visible as well as palpable (Figs. 19-3, 19-6). Chronic elevation of venous pressure may result in thickened jugular veins with decreased compliance; the forcefulness of the venous V wave may be surprisingly great. Such pulsations may simulate the arterial pulse of aortic regurgitation. As a general rule, large systolic jugular *venous* pulsations are more lateral in the neck and generally have a more gradual or rounded contour. The venous pulse can be obliterated by firm pressure at the base of the neck. A problem of differentiation of venous from arterial pulsations occurs only in patients with severe tricuspid regurgitation and a very high, mean venous pressure.

Jugular Venous Pulse

Neck vein pulsations in tricuspid regurgitation are of extreme diagnostic importance. The diagnosis of an incompetent tricuspid valve can often be suggested prior to auscultation after a careful examination of the jugular

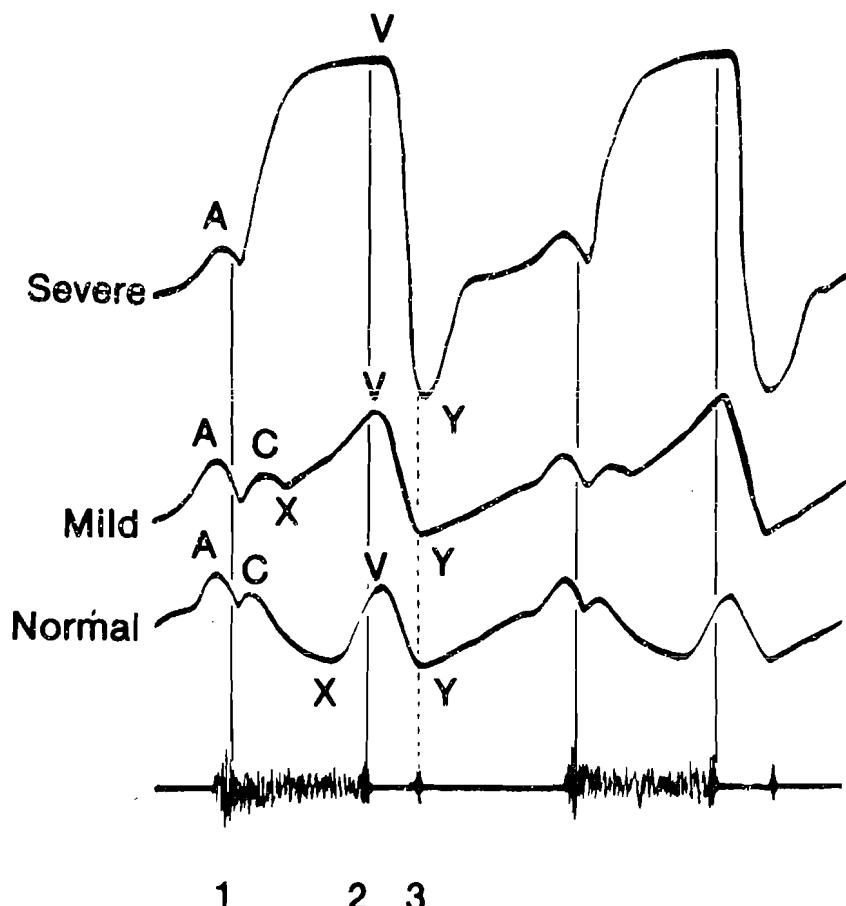


FIG. 19-6. Jugular venous pulse in tricuspid regurgitation. Alterations of the venous contour in mild tricuspid regurgitation are depicted in the middle tracing. The V wave is augmented and the Y descent is more prominent; the X descent is markedly attenuated. With severe tricuspid regurgitation (top tracing) there is a plateau-like systolic regurgitant C-V wave, which in part represents "ventricularization" of the right atrial and jugular venous pulses. Note the right ventricular S3 coinciding with the nadir of the Y descent. A normal venous pulse is depicted in the bottom drawing. (From Abrams J: Prim Cardiol, 1982.)

pulse. In patients with nonspecific or atypical systolic murmurs without classic inspiratory augmentation, the venous pulse may be more specific and sensitive than the systolic murmur in establishing the presence of tricuspid regurgitation.

As the severity of tricuspid regurgitation increases in magnitude, the X descent becomes attenuated and ultimately disappears (Fig. 19-6). With further degrees of tricuspid reflux, a positive venous pulse wave appears during systole. This has been called S wave, C-V, V wave, or regurgitant wave. It is often quite prominent. The regurgitant wave is systolic and approximately simultaneous with the palpable carotid arterial pulse, although the precise timing, peak, and contour are somewhat different from the arterial pulsation. The venous C-V wave has a more rounded contour and is somewhat sustained or plateau-like, in contrast to the brisk up-and-down motion of the arterial pulse (Fig. 19-3, 19-5, 19-6).

With increasing degrees of tricuspid regurgitation, the venous pulsation begins earlier in systole. The large systolic V or regurgitant wave is terminated by a sharp steep trough, the Y descent, which represents the decompression of the right atrium as the tricuspid valve opens fully in early diastole (Fig. 19-6). The right atrial blood is under increased pressure and rapidly traverses the tricuspid valve during the rapid filling phase of diastole. The prominent Y descent is easily seen and may actually be more prominent than the swelling of the venous pulse (V wave) preceding the Y collapse. Some patients with severe tricuspid regurgitation have an enormous regurgitant or V wave with a dramatic Y collapse (Figs. 19-3, 19-5, 19-6).

In most adult patients with acquired tricuspid regurgitation due to pulmonary hypertension, the mean venous pressure will be elevated. Thus, the peak of the venous systolic wave may be high; it is often necessary to examine such subjects in the semirecumbent or sitting position. With very high venous pressures, the meniscus of the venous blood column may be invisible, as the venous system is tense and distended with blood. Patients may have to be examined sitting or even standing up in order to see the peak of the venous pressure. *Practical Point:* *Do not conclude that the venous pulse is normal unless the venous wave forms are readily identified and the mean venous pressure can be estimated in the 45-degree, recumbent or upright position.*

Effects of Respiration. The magnitude of tricuspid reflux increases as RV inflow increases during inspiration; as the right atrium distends, more blood regurgitates into the superior vena cava. This is manifest as a larger V wave, which has a higher peak and mean pressure and a more prominent Y descent during inspiration. This inspiratory swelling of the venous pulse is a variant of Carvallo's sign (see below) and is an important observation. In patients with mild tricuspid regurgitation or a large compliant right atrium, the inspiratory increase in the V wave may be a more reliable clue to the presence of tricuspid regurgitation than auscultatory changes in the systolic murmur. If severe right ventricular failure or massive tricuspid regurgitation is present, the right ventricular may be unable to fill any further, and an inspiratory increase in venous pressure will not be noted.

Atrial Fibrillation. This arrhythmia may create difficulty in diagnosing tricuspid regurgitation, as the varying cycle lengths alter the degree of tricuspid reflux on a beat-to-beat basis. Respiratory alterations in the level of the venous pressure and V wave height may be impossible to detect.

The loss of the A wave in patients with atrial fibrillation means that only a V wave will be seen in the venous pulse. In the absence of tricuspid regurgitation this dominant jugular V wave may simulate the abnormal V wave of tricuspid regurgitation, whether the systemic venous pressure is normal or elevated. When the ventricular rate is rapid in atrial fibrillation, it may be difficult to time the venous pulsations.

Ancillary Signs. On occasion, severe chronic tricuspid regurgitation

causes some remarkable findings related to the huge venous V waves and high systemic venous pressure. Audible sounds or pistol shot transients have been heard in the region of the internal jugular veins; these may even be palpable as systolic shocks. Pulsations of the head (right-to-left head bob with each cardiac cycle), eyeballs, and earlobes have been observed in major tricuspid regurgitation. The eyes may also be slightly proptotic from chronically elevated venous pressure. *Practical Point: Subtle systolic motion of the earlobes, and/or lateral swelling of the neck with each heart beat are common clues to the presence of severe tricuspid regurgitation (Fig. 19-7).*

For an optimal assessment of the venous pressure and contour one must inspect the neck carefully, using tangential lighting and various degrees of elevation of the head (Fig. 4-2). Chronic distention of the systemic veins may render the venous valves incompetent, allowing transmission of the systolic V wave to the periphery of the venous tree. This may result in pulsations of the veins on the backs of the hands, arms, or legs that are seen when the extremity is elevated to the level of actual mean venous pressure. Pulsations of varicose veins have also been reported.

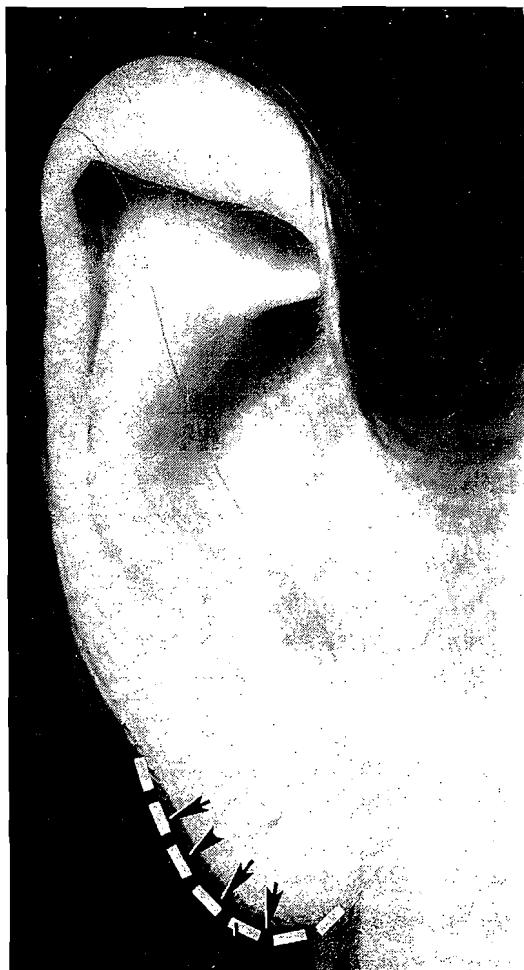


FIG. 19-7. Earlobe pulsations in tricuspid regurgitation. In severe tricuspid regurgitation, the jugular venous pressure is markedly elevated and the large V waves may be difficult to discern. Careful inspection of the ear lobe with the patient in the 30 to 90 degree position may reveal subtle lateral pulsations of the large systolic V waves in the right atrium throughout the proximal venous system.

A "Normal" Venous Pulse. On occasion, the jugular venous pressure and pulse wave contour may be normal in tricuspid regurgitation, particularly in patients in sinus rhythm who have a mild degree of tricuspid reflux. The right atrium normally is more compliant than the left atrium. With slowly increasing tricuspid regurgitation, the right atrial chamber may dilate and retain its distensibility. The additional blood refluxing into the atrium may be absorbed in the distensible right atrial chamber. On rare occasions, even severe tricuspid regurgitation can be associated with a minimal abnormality of right atrial pressure.

One can augment or bring out an abnormal V wave in equivocal cases. Anything that increases return to the right heart will augment the magnitude of tricuspid regurgitation such as leg elevation, slow deep inspiration, or mild exercise (e.g., sit-ups). The hepatojugular reflex maneuver (see pages 47, 395) may also be helpful in the detection of tricuspid regurgitation; sustained abdominal pressure results in selective augmentation of the venous V wave and the systolic murmur in tricuspid regurgitation.

Other Causes of a Large Venous V Wave. Congestive heart failure without overt tricuspid regurgitation can be associated with a prominent V wave in the venous pulse; the elevated mean venous pressure contributes to potential confusion. Cor pulmonale can also produce a large V wave. In both conditions the mean venous pressure and the V wave itself decrease rather than increase with inspiration. Atrial fibrillation, as already mentioned, is a common problem because the only visible venous wave is a V wave. Recordings of the venous pulse in actual fibrillation demonstrate preservation of the X descent, but this is difficult to detect on visual inspection. Patients with atrial septal defects often have prominent venous V waves, equal in height to the A wave in the absence of tricuspid regurgitation (see Fig. 20-23). In subjects who have previously undergone right atrial and great vein cannulation during open heart surgery, the V wave may be prominent in the absence of overt tricuspid reflux.

Precordial Motion

Abnormalities of precordial motion are detectable in many patients with tricuspid regurgitation. The presence or absence of pulmonary hypertension will greatly influence the physical findings.

In the absence of pulmonary hypertension, tricuspid regurgitation causes prominent systolic unloading of the RV. Typical parasternal activity shows a retraction wave during late systole. A brisk early systolic outward motion occurs; during the remainder of systole, the RV pulls away from the chest wall. An exaggerated early diastolic RV filling wave may be palpable (RV S3). In subjects with pulmonary hypertension, a systolic RV impulse typically

will be present; this is a low amplitude, sustained parasternal lift or heave (see Chapter 5).

Palpable right ventricular activity in tricuspid regurgitation thus reflects the level of pulmonary artery pressure, the RV size, and the severity of tricuspid regurgitation. An outward systolic impulse will be felt in most instances; it will be brief and early systolic in the nonpressure overloaded right ventricle, and is likely to be sustained when there is major pulmonary hypertension. When the RV is quite large, as is often the case with chronic tricuspid regurgitation, the precordial activity may produce a rocking or seesaw motion. The left anterior chest and LV apex retract as the medial RV area, adjacent to lower left sternal border, moves anteriorly and then retracts during mid to late systole. A markedly enlarged RV may occupy the usual apex area, literally pushing the LV posterolaterally in the thorax, where it is no longer palpable on the chest wall.

Right ventricular and even right atrial activity occasionally may be felt or percussed at the lower right sternal border in patients with severe major tricuspid regurgitation. A palpable RV A wave (S4) may be felt, as can the rapid filling wave (S3). The epigastric approach is best for detection of diastolic RV events (Chapter 5, Fig. 5-10). When tricuspid regurgitation is severe, right precordial activity may be visible as hyperkinetic cardiac motion. Palpable hepatic pulsations in systole are virtually pathognomonic for tricuspid regurgitation (Fig. 19-5). A pulsatile liver may be detected even in the absence of a systolic murmur.

Heart Sounds

S1. There are no important alternations of S1 in tricuspid regurgitation. The intensity of S1 may be diminished, normal, or increased in amplitude. An accentuated tricuspid component (T1) may be audible in the presence of tricuspid stenosis or in the rare case of a right atrial myxoma.

S2. Abnormalities of S2 in tricuspid regurgitation are common, but are caused by the underlying cardiac lesion rather than the tricuspid valve incompetence. As severe pulmonary hypertension is a common cause of tricuspid regurgitation, an accentuated P2 is often present and may be widely transmitted over the precordium. If severe RV failure is present, RV ejection will be prolonged and the stroke volume relatively fixed during respiration; S2 will be widely split both in inspiration and expiration. This can simulate the fixed splitting of an atrial septal defect (see Chapter 20).

S3. An audible RV S3 can be heard in tricuspid regurgitation. The S3 may reflect the excessive volume of blood crossing the tricuspid valve in early diastole and/or RV decompensation associated with a large RV end-diastolic volume and decreased ejection fraction. Right-sided ventricular filling sounds

(S3 and S4) are typically louder during inspiration and wane with expiration. Such sounds may be audible *only* in inspiration. When the tricuspid regurgitation is hemodynamically important, the S3 may be palpable, particularly from the subxiphoid approach.

A right ventricular S3 is best heard at the lower left sternal border and will be inaudible at the LV apex. However, if the RV is very large and occupies the apical region in the left thorax, an RV S3 may be readily mistaken for an S3 of left ventricular origin.

The RV S3 is often associated with very high RV diastolic pressures and may be quite high-pitched, simulating a pericardial knock or a tricuspid opening snap. The pericardial knock of constrictive pericarditis is an unusually prominent, early, and high-pitched RV filling sound (i.e., an S3). As is the case with mitral regurgitation, severe tricuspid regurgitation with a large regurgitant fraction may produce a brief diastolic flow rumble; this murmur will increase with inspiration and can simulate tricuspid stenosis (Figs. 19-8, 19-9B).

Opening Snap. An opening snap may be generated by the tricuspid valve, even in the absence of tricuspid stenosis, but is a relatively uncommon finding in tricuspid regurgitation (Figs. 19-8, 19-9B). Tricuspid opening snaps have been documented to occur with high velocity tricuspid valve flow in the presence of freely mobile and nonobstructive valve leaflets such as in atrial septal defect or Ebstein's anomaly. The opening snap is generated at the time of abrupt halting of the tricuspid leaflets during the opening movement in early diastole. The snap is usually slightly later than the mitral opening snap, occurring 0.09 to 0.15 seconds after A2. It cannot be easily distinguished from an opening snap of mitral valve origin except by indirect clues such as maximal intensity at the lower left sternal border, augmentation with inspiration, and association with other right-sided acoustic abnormalities.

S4. A right-sided atrial gallop may be heard in patients with acute onset tricuspid regurgitation. This is similar to the left-sided S4 found in patients with acute mitral regurgitation (see Chapter 17). In such cases, the right ventricle and the right atrium are usually of normal size; the regurgitant volume load results in vigorous right atrial contraction and produces a large increase in end-diastolic flow and pressure in the right ventricle. Acute tricuspid regurgitation is likely to occur in a drug addict with tricuspid valve endocarditis or a patient with traumatic rupture of a tricuspid valve cusp.

The right ventricle S4 may be very loud. It is best heard at the lower left sternal border at the 4th to 5th interspace and augments with inspiration.

Systolic Clicks. Early. Patients with severe pulmonary hypertension may have a pulmonary artery ejection sound, often decreasing with inspiration and best heard at the upper left sternal border (Fig. 8-6). If tricuspid regurgitation is present, the ejection click will still be audible unless the systolic



FIG. 19-8. Diastolic flow murmur in tricuspid regurgitation. This phonocardiogram demonstrates a mid-diastolic flow murmur in a patient with tricuspid regurgitation. The diastolic murmur or rumble represents the large volume of blood traversing the tricuspid valve during the rapid filling phase of diastole. SM = systolic murmur; DM = diastolic murmur; ACG = apex cardiogram. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co, 1979.)

murmur is extremely loud and obliterates the ejection sound. Respiratory variation of the click (decrease with inspiration) should differentiate this sound transient from an aortic ejection click or prominently split S1.

Mid-Late. Tricuspid valve prolapse (see Chapter 18) is usually not diagnosed on auscultation, although it may be detected on echocardiography. Nevertheless, the prominence of a mid to late systolic click at the lower left sternal border that becomes louder and later during inspiration should raise the suspicion of tricuspid valve prolapse (Fig. 19-2).

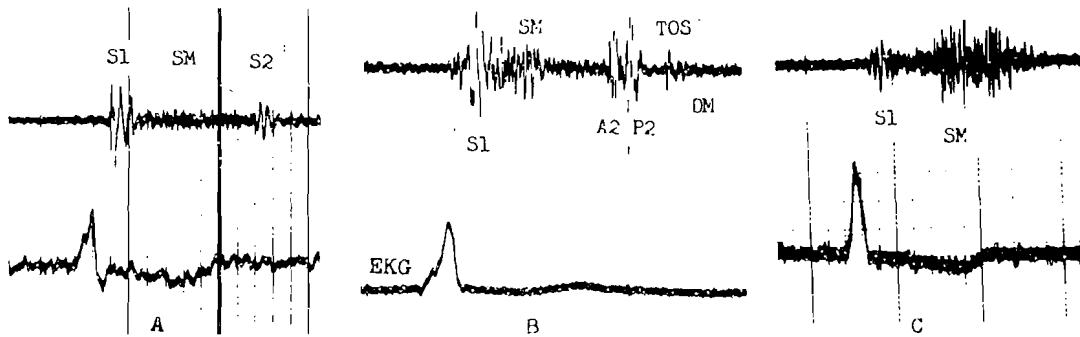


FIG. 19-9. Murmurs of tricuspid regurgitation. These phonocardiograms are taken at the lower left sternal border in three individuals with tricuspid insufficiency. A. The soft holosystolic murmur has a plateau-like configuration. B. The murmur peaks in early systole, although sound vibrations appear to extend to A₂. Note the tricuspid opening snap, followed by a diastolic flow murmur. C. This tricuspid murmur has late systolic accentuation. SM = systolic murmur; TOS = tricuspid opening snap; DM = diastolic murmur. (From Hultgren HN, Hancock EW, and Cohn KE: Auscultation in mitral and tricuspid valvular disease. Prog Cardiovasc Dis 10:298, 1968.)

Murmurs

The classic murmur of tricuspid regurgitation is holosystolic, increases with inspiration, and is best heard at the 4th and 5th interspaces at the lower left sternal border. The tricuspid regurgitation murmur is usually not very loud and may be extremely variable in character. Frequently, it is missed by physicians who do not see many patients with cardiac disease, and it is often not recognized by experienced cardiologists. *Practical Point: The correct diagnosis often will be made only if the examiner specifically looks for the subtle clues of tricuspid regurgitation.* The possibility of tricuspid regurgitation should be actively considered in any patient with rheumatic heart disease or other conditions who has evidence for pulmonary hypertension.

Contour. The usual tricuspid regurgitation murmur has an even, pansystolic configuration extending to S₂ (Figs. 19-4, 19-6, 19-9A)). However, it is common for the murmur to wane in late systole, often with early to midsystolic accentuation (Fig. 19-9B). Occasionally, the murmur may have late systolic accentuation, particularly in tricuspid valve prolapse (Fig. 19-9C).

On occasion, a tricuspid regurgitation murmur that tapers off in late systole is not pansystolic; this type of murmur is most likely to occur in mild tricuspid regurgitation, in the absence of pulmonary hypertension, or in acute-onset tricuspid regurgitation. When the tricuspid valve is incompetent and the RV has a normal systolic pressure, the volume and velocity and blood refluxing into the right atrium in late systole is small. In such cases, the tricuspid murmur is commonly soft and short, and the correct diagnosis is easily missed. One may have to rely on *respiratory variation* in the murmur intensity and the *ancillary signs* of tricuspid regurgitation (jugular venous pulse, pulsatile liver) to make the appropriate diagnosis.

Intensity. The murmur of tricuspid regurgitation is seldom very loud and usually is of Grade II or II-III intensity. Tricuspid regurgitation has been documented to be acoustically silent in some patients, particularly in subjects with a markedly reduced RV stroke volume and a huge right atrium, a combination that allows for insufficient turbulence to produce audible sound. Silent tricuspid regurgitation also is more likely to occur with normal RV systolic pressure such as is found with traumatic or infectious damage to a previously normal valve. Making the proper diagnosis in such cases is not easy. In some patients the murmur is heard only during inspiration, which can be misleading, as it may simulate a pulmonary or pericardial process. Variation in inspiratory intensity may be difficult to detect when the tricuspid murmur is very loud.

In general, a prominent tricuspid regurgitation murmur indicates large volume reflux, but the converse is not true; severe tricuspid regurgitation can be completely soundless. With atrial fibrillation, the systolic murmur intensifies after long cycle lengths. This can be a most important clue in differentiation from mitral regurgitation. The murmur of mitral regurgitation remains unchanged during alterations of cycle length. Similarly, the tricuspid regurgitation murmur typically augments in the beat following a PVC in contradistinction to mitral regurgitation.

Frequency. The typical tricuspid regurgitation murmur is medium frequency, but on occasion can be rough and raspy. High frequency murmurs of tricuspid regurgitation are not common, probably due to the relatively low velocity of regurgitant flow into the right atrium, even with severe right ventricular hypertension. The murmur may appear to be more high-pitched at sites away from the area of maximal intensity. It is best to use both the bell and diaphragm when listening for tricuspid regurgitation.

Tricuspid Honk or Whoop. Tricuspid regurgitation rarely can be associated with a vibratory, high-pitched whoop that usually demonstrates a prominent inspiratory increase. The tricuspid honk usually is intermittent and has been associated with severe pulmonary hypertension. Its etiology is obscure, but it may be similar to that of mitral valve honks and whoops. Tricuspid valve prolapse is likely to be an etiologic factor.

Location and Radiation. The tricuspid regurgitation murmur is best heard over the right ventricular area at the lower left sternal edge at the 4th to 5th interspace (Fig. 9-2). The murmur may radiate into both the right and left chest for a short distance. Transmission to the lower right sternal area is common, and one should listen carefully at the lower right sternal border whenever tricuspid regurgitation is suspected. When loud, the murmur may also be heard at the upper left sternal edge. Soft tricuspid murmurs may be well localized to a small area of the lower left sternal border.

When the RV is very large, the murmur may be heard at both the left lower sternal edge and the cardiac apex, which may be formed by the dilated

right ventricular chamber. In such cases, the murmur may be equally loud at the mid left chest and sternal edge, but it will never be loudest at the apex. It is easy to mistake the apical murmur of tricuspid regurgitation for mitral regurgitation. If the apical murmur is solely tricuspid in origin, it will not radiate into the axilla. Inspiratory augmentation should be carefully assessed.

Tricuspid regurgitation murmurs often are easily detected at the xiphoid and subxiphoid areas, particularly in patients with large chests or chronic obstructive pulmonary disease. The murmur may be audible below the right costal margin and over the liver. In rare cases, the murmur may be faint or even inaudible at the lower sternal edge, but readily heard at the subxiphoid and right subcostal regions. *Practical Point: Whenever tricuspid regurgitation is suspected, listen carefully for murmur radiation to the right lower sternal border, xiphoid region, and over the superior aspect of the liver (Fig. 9-2).*

Respiratory Alteration. The inspiratory increase in the intensity of the murmur of tricuspid regurgitation is a well-known phenomenon, reported by Rivero Carvallo in 1946 and known as Carvallo's sign (Fig. 19-4). Respiratory alteration, however, is frequently missed by clinicians. The increase in murmur loudness with inspiration often is only $\frac{1}{2}$ to 1 grade and may be a subtle finding. For optimal detection of respiratory variation, it may be useful to auscult several centimeters away from the point of peak murmur intensity. With very loud murmurs minor respiratory alterations in intensity may be difficult to detect.

One must be sure the patient is breathing deeply and smoothly without forced respirations or performance of an inadvertent Valsalva maneuver. Noisy breath sounds can interfere with detection of subtle respiratory changes. The subject should breathe somewhat more deeply than usual in a continuous fashion. Some authorities recommend that the patient hold his breath during inspiration with the physician listening for an increase in murmur intensity during the period of postinspiratory apnea. However, this maneuver often results in a decrease in loudness of the murmur.

In patients with a soft murmur, inspiration may increase the grade dramatically for only one or two beats. Rarely, the systolic murmur will be heard only during inspiration while diastole remains silent. *Practical Point: For optimal auscultation, listen for respiratory variation over many respiratory cycles and focus on phasic alterations in peak intensity of the systolic murmur.* The enhancement of RV stroke volume may be difficult to detect and may only last for a few beats at a time. The examiner should demonstrate considerable patience when listening for the subtle murmur of tricuspid regurgitation.

Hepatojugular Reflux. The use of the hepatojugular reflux maneuver recently has been shown to be helpful in identifying tricuspid murmurs. Firm upward pressure with the hand over the right upper quadrant of the abdomen for 10 to 15 seconds will accentuate the murmur of tricuspid regurgitation

(see page 47). The maneuver is especially useful in patients with only mild to moderate tricuspid regurgitation and in the absence of pulmonary hypertension where Carvallo's sign is equivocal or not present. The tricuspid murmur typically increases in loudness for 4 to 6 beats.

Atrial Fibrillation. The changing cycle lengths in atrial fibrillation cause the intensity of the tricuspid murmur to vary from beat to beat, being loudest after long cycles. Respiratory variation is extremely difficult or even impossible to detect in patients with this arrhythmia. *Practical Point: Atrial fibrillation is common in patients with chronic rheumatic heart disease and tricuspid regurgitation. Do not conclude tricuspid regurgitation is absent because respiratory variation in the systolic murmur cannot be demonstrated in such patients.* In general, it is not productive to focus on detection of inspiratory augmentation in patients with atrial fibrillation suspected of having tricuspid regurgitation.

Failure to Change with Respiration. In some instances, there is no detectable or consistent respiratory alteration in intensity of the tricuspid regurgitation murmur (Table 19-3). The hepatojugular reflux maneuver should be tried in such cases. This situation is more likely to be true with mild tricuspid regurgitation and normal RV systolic pressure or, conversely, with tricuspid regurgitation associated with severe RV dysfunction and right heart failure. When RV failure is present, the distended ventricle may be unable to accept more blood from the right atrium and great veins during inspiration. RV stroke volume is relatively fixed during the respiratory cycle. *Practical Point: Always have the patient sit or stand up during auscultation when tricuspid regurgitation is suspected but respiratory variation is absent.* The decrease in right heart volume and diastolic pressure caused by the diminished return in the upright posture may allow the RV to vary its stroke volume somewhat during respiration with a resultant increase in murmur intensity during inspiration.

Changes with Various Maneuvers. The murmur of tricuspid regurgitation can be affected by factors other than respiration (Table 19-4). Anything that increases right heart return such as elevation of the legs, squatting, the hepatojugular reflux maneuver, Mueller's maneuver (inhalation with the glottis closed), or exercise will augment the systolic murmur. The tricuspid murmur usually increases during long R-R cycles (atrial fibrillation, post-PVC beat) because there is increased RV filling during such beats.

TABLE 19-3 *Conditions Associated with Failure of the Tricuspid Regurgitation Murmur to Augment with Inspiration*

Mild or trivial tricuspid regurgitation (usually normal right ventricular pressure)
Severe right ventricular dilatation and/or failure
Atrial fibrillation
Very loud murmur

TABLE 19-4 Factors Affecting the Loudness of the Murmur of Tricuspid Regurgitation

<i>Increased Murmur Intensity Caused by Increased Right Heart Filling</i>
Inpiration
Hepatojugular reflux maneuver
Leg elevation
Squatting
Mueller's maneuver
Mild exercise
Volume overload or decompensated RV failure
Valsalva maneuver—release phase (immediate)
<i>Decreased Murmur Intensity Caused by Decreased Right Heart Filling</i>
Expiration
Standing
Sitting
Valsalva maneuver—strain phase
Nitroglycerin
Diuresis or hypovolemia

The murmur of tricuspid regurgitation will decrease in intensity whenever RV volume diminishes, e.g., the upright position or the strain phase of the Valsalva maneuver. However, following release of the Valsalva maneuver the normal intensity returns immediately within 1 to 2 seconds, is in contrast to the typical left heart murmur response to the Valsalva maneuver, when there is a delay in the return to baseline murmur intensity after 3 to 4 seconds or 6 to 8 beats.

Variability. The clinical picture of tricuspid regurgitation may vary from day to day, particularly in the "secondary" or functional variants. The variation of signs of tricuspid regurgitation are due to alterations in cardiovascular hemodynamics; the level of pulmonary artery pressure and the degree of RV dilatation and dysfunction act as important variables that may change from day to day. In patients with pulmonary hypertension and RV failure, intracardiac and pulmonary pressures may fluctuate considerably, depending on the status of intra- and extravascular volume, the level of physical activity, and cardiac rate and rhythm. Tricuspid regurgitation commonly is present when such patients present in a decompensated state. Severe RV failure may cause the tricuspid regurgitation to be completely missed on initial examination due to marked distention of the venous system (failure to observe V wave) and absence of respiratory change in the murmur (presence of atrial fibrillation and/or severe right ventricular failure). After therapy, the tricuspid regurgitation may then become clinically evident. When treatment is maximal (e.g., conversion to normal sinus rhythm, following substantial diuresis, or rate control in atrial fibrillation), the tricuspid regurgitation may completely resolve.

Patients with chronic severe congestive heart failure and tricuspid regurgitation usually will demonstrate fluctuation during the physical examination; the signs of tricuspid regurgitation parallel the functional status of the patient.

Practical Point: If tricuspid regurgitation is no longer evident in patients who have had previously documented tricuspid regurgitation, the use of various provocative maneuvers to bring out this abnormality is indicated (Table 19-4).

Diastolic Murmur. An early mid-diastolic tricuspid murmur may be noted in patients with substantial tricuspid regurgitation (Figs. 19-8, 19-9B). This murmur represents diastolic flow across the open tricuspid valve. The murmur usually is medium to low frequency. It is best heard at the lower left sternal border (4th to 5th interspace), and follows the tricuspid opening snap or S3, if present. It commonly augments with inspiration.

The diastolic murmur may be confused with the murmur of pulmonic or aortic regurgitation. Careful attention to the timing of onset is critical. The tricuspid diastolic murmur begins well after P2 in early diastole, whereas the murmurs of semilunar valve insufficiency begin with S2.

ACUTE TRICUSPID REGURGITATION

The sudden onset of tricuspid regurgitation often results in deviations from the classic clinical findings of chronic tricuspid regurgitation. These alterations in the physical findings are similar to those found in acute mitral regurgitation (see Chapter 17) and result from a precipitous alteration in anatomic integrity and cardiac function. Most, but not all, patients with acute tricuspid regurgitation have had a normal cardiovascular system *prior to onset* as, for example, individuals who suffer blunt or penetrating cardiac trauma or a narcotic addict with tricuspid valve endocarditis. When tricuspid regurgitation occurs in the setting of acute inferior wall myocardial infarction and RV papillary muscle dysfunction, the right heart chambers typically are normal in size and function prior to the infarct. Right ventricular and right atrial volume overload may result from the acute tricuspid incompetence. As the right heart chambers have no time to hypertrophy and dilate, the tricuspid regurgitant wave may produce a very high right atrial and mean atrial V wave.

PHYSICAL FINDINGS

The venous pulse has a larger A wave in acute tricuspid regurgitation than in chronic tricuspid disease; the X descent is attenuated or absent and an abnormally large V wave is present. Such patients usually are in normal sinus rhythm (in the absence of pre-existing cardiac disease). Precordial examination may demonstrate a rocking motion, but little RV prominence will be noted, as the right ventricle has had no time to hypertrophy.

A prominent (and often booming) right ventricular S4 is present at the lower left sternal edge in acute tricuspid regurgitation. This right-sided atrial

gallop augments markedly with inspiration. The S4 may be palpable. An RV S3 may also be present. The murmur of acute tricuspid regurgitation is similar to the murmur in acute mitral regurgitation in regard to an often absent holosystolic contour with silence in late systole. Such murmurs are typically short and have an ejection quality. The murmur of acute tricuspid insufficiency will augment in intensity and become longer during inspiration. The cause for the abbreviated systolic murmur is the large V wave in the right atrium that occurs from "ventricularization" of the right atrial pressure pulse, which results from the RV emptying a large portion of its stroke volume into a small, stiff right atrium. The murmur may be so short as to be completely inconspicuous, and its intensity may be unimpressive. In patients in whom trauma or endocarditis is the cause of tricuspid regurgitation, the normal, low pressure right ventricle does not produce a high velocity, turbulent jet, and thus the murmur intensity is soft.

Because the findings in acute tricuspid regurgitation may be very atypical, it is advisable to use various maneuvers (Valsalva, sitting, passive leg elevation, squatting, hepatojugular reflux) in an effort to bring out respiratory variation or alterations in the length and the duration of the murmur by changing of the degree of RV filling.

DIFFERENTIAL DIAGNOSIS

The murmur of tricuspid regurgitation often is missed, particularly when there are other obvious causes of a heart murmur. The murmur may be thought to be innocent or it may be confused with mitral regurgitation. Associated cardiac lesions may confuse the issue; many clinicians fail to think of tricuspid regurgitation as a diagnostic possibility. The frequent, atypical presentation of the tricuspid regurgitation murmur (not holosystolic, no increase with inspiration) is also a problem. Tricuspid regurgitation will be diagnosed appropriately far more often if the physician has an expectant attitude, particularly in any patient suspected of having pulmonary hypertension.

Mitral Regurgitation. The holosystolic murmur of mitral regurgitation occasionally may radiate towards the sternum. If the heart is not large, the murmur may be prominent in the tricuspid area. Respiratory variation and augmented venous V waves indicate tricuspid regurgitation; in the absence of overt congestive heart failure, the inability of the holosystolic murmur to increase with inspiration favors mitral regurgitation. In atrial fibrillation respiratory variation of the murmur may be easily missed. Patients with RV dilatation present a different problem because tricuspid regurgitation may be readily mistaken for mitral regurgitation when the right ventricle forms the cardiac apex. The absence of murmur radiation into the axillary region and

the presence of respiratory variation are crucial diagnostic points favoring tricuspid origin of the murmur.

It is not uncommon for a patient to have both mitral and tricuspid regurgitation. In such instances, one of the valve lesions may be missed on auscultation. Typically, the holosystolic murmur will wane and then increase in intensity as the stethoscope is inched from the lower left sternal border towards the cardiac apex, or vice versa. Respiratory variation will be audible medially but not laterally in the left chest. When the tricuspid regurgitation murmur is loud, it may be extremely difficult to be sure whether or not coexisting mitral regurgitation is present.

Ventricular Septal Defect. Both a ventricular septal defect and tricuspid regurgitation produce a holosystolic murmur at the lower left sternal edge, but there is rarely a problem in differentiating these two conditions. Ventricular septal defects are found exclusively in children and only infrequently in young adults. There is no inspiratory augmentation of the murmur, and the jugular venous pulse is normal in patients with VSD. In children with evidence of pulmonary hypertension, the differential diagnosis may be difficult if there is associated RV failure, particularly if the tricuspid regurgitation murmur fails to augment with inspiration. This is a rare diagnostic problem. In acute myocardial infarction complicated by a ventricular septal defect, the differential between the two lesions may be difficult or even impossible if there is no respiratory change in the murmur.

Functional Murmurs. When the tricuspid regurgitation murmur is short (normal pulmonary artery systolic pressure, acute tricuspid regurgitation) and does not extend to S2, it may readily simulate an ejection murmur of nonspecific nature. The location is lower (4th to 5th interspace at left sternal edge) than the usual flow murmur. Respiratory variation is the most important discriminator. Functional murmurs tend to soften in inspiration, if at all.

Chapter 20

Atrial Septal Defect

An atrial septal defect (ASD) is a relatively common abnormality in adults. It is common in children and represents 7 to 15% of all congenital heart lesions in young patients over the age of one year. Atrial septal defects are more frequent in females, with estimates ranging between 1-1/2 and 4 times the prevalence in males. Because symptoms, if present, are relatively mild in younger patients with ASDs, in many adults with ASDs the correct diagnosis often has not been made at the time of discovery. An individual may be seen by physicians for years without suspicion with the finding of a "functional" systolic murmur. Thus, it is important for clinicians to keep the diagnosis of an ASD in mind when evaluating any adult with an ejection murmur.

ANATOMY (Table 20-1, Fig. 20-1)

The vast majority of ASDs in the older age group are *ostium secundum* defects. This is an abnormal communication in the upper atrial septum generally uncomplicated by other serious cardiac abnormalities. It is also known as a fossa ovalis defect. A small number of patients (5 to 10%) have a defect located at the junction of the superior vena cava and lateral right atrium, the so-called sinus venosus ASD. Usually, this defect is associated with anomalous drainage of the right superior pulmonary vein into the superior vena cava or directly into the high right atrium.

Ostium primum defects account for the remaining 25% of ASDs, but are rare in older children and adults. They are part of a group of endocardial cushion (atrioventricular canal) disorders and typically may be accompanied by abnormalities of the A-V valves as well as of the ventricular septum. The hemodynamics and physical findings of the *secundum ASD* are the subjects of this chapter.

Associated Defects. The two most common abnormalities associated with secundum ASD are prolapse of one or both leaflets of the mitral valve and partial anomalous pulmonary venous return. There is a definite association of auscultatory and echocardiographic findings of mitral valve prolapse in patients with a secundum ASD. Drainage of one or more of the pulmonary veins into the right atrium, superior vena cava, or inferior vena cava occurs in about 15% of the secundum ASDs and may be found in 85% of the sinus

TABLE 20-1 *Atrial Septal Defects: Anatomic Variants*

Type	Incidence
Septum secundum (fossa ovalis)	70%
Septum primum	20–25%
Sinus venosus	8–10%

venosus variety. Generally, this results in little additional hemodynamic burden to the patient.

PATOPHYSIOLOGY

In an uncomplicated ASD, the most consistent hemodynamic feature is left-to-right shunting across the defect with subsequent volume overload of the right ventricle and pulmonary vascular bed. The magnitude of the shunt depends less on the size of the defect than on the relative compliance of the right and left ventricles and the pulmonary vascular resistance.

Size of Defect. Openings in the atrial septum may be tiny to very large, single, or less commonly, multiple. If the defect is less than 1 cm^2 in diameter, it is unlikely to permit significant shunting, and left atrial pressure remains greater than right atrial pressure. Atrial defects of 2 to 3 cm^2 or larger result in a relatively low resistance shunt from left atrium to right atrium with near

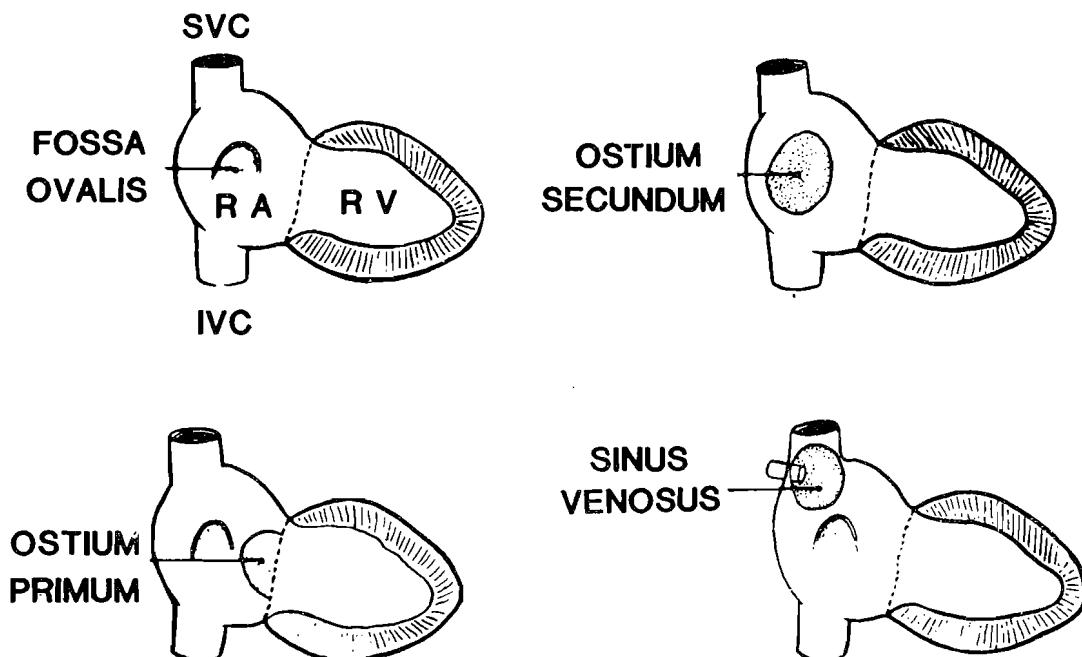


FIG. 20-1. Anatomic sites of atrial septal defects. Upper left: location of the normal fossa ovalis (ostium secundum); upper right: site of the ostium secundum defect, the commonest variant of ASD; lower left: ostium primum defects are found in the lower portion of the interatrial septum; lower right: the sinus venosus ASD (note drainage of the right superior pulmonary vein communicating with the right atrium through the defect). (From Perloff JK: Atrial septal defect. In *The Clinical Recognition of Congenital Heart Disease*, 2nd ed. Philadelphia, WB Saunders Co, 1978.)

equalization of mean atrial pressures. It is now recognized that small atrial defects rarely may close with time (usually by age 2). Thus, the anomaly may literally heal itself (analogous to the far more common phenomenon with ventricular septal defects in young children).

Determinants of Shunt Volume. At birth, the normal right ventricle is thick and relatively stiff or noncompliant: pulmonary and right ventricular pressures are high. The pulmonary arteries and arterioles are thick-walled and noncompliant, and pulmonary arteriolar resistance is high. Thus, the relative inflow resistance of the right heart is similar to that of the left heart, and little left-to-right atrial shunting occurs. In time, resolution of the high physiologic pulmonary vascular resistance takes place, and thinning of the right ventricular wall occurs concomitantly. In the first few months of life, the normal increase in compliance of the right ventricle allows for the development of left-to-right shunting. Right ventricular and right atrial pressures fall, and the shunting of blood flow across the defect from the left atrium to the right atrium increases significantly. This explains the frequent initial appearance of the systolic murmur of the ASD and wide fixed splitting of S₂ later in childhood, with minimal or absent physical findings of an ASD in infancy and at a very young age.

As right ventricular distensibility increases concomitant with the continuing physiologic decrease in pulmonary arteriolar wall thickness and fall in pulmonary vascular resistance, the degree of left-to-right shunting across the ASD increases. This will result in excess volume loading of the right ventricle and dilatation of the right ventricle as well as the pulmonary arteries. Augmented left-to-right flow with shunt ratios of greater than 3:1 can occur, resulting in a massive increase in right ventricular work. For instance, the right ventricle may pump 10 to 20 liters/minute through the pulmonary circulation compared to the 4 to 5 liters/minute output of the left ventricle, as in a 4 to 1 pulmonic to systemic flow ratio. This rate may produce hyperkinetic or flow-related pulmonary hypertension, although usually pulmonary artery systolic pressure does not rise beyond 35 to 40 mmHg in children. The torrential flow may result in a modest gradient (up to 40 mmHg) across the right ventricular outflow tract in the absence of anatomic outflow obstruction and in prolonged right ventricular ejection time.

Timing of Left-To-Right Shunting. Most of the flow across the atrial defect occurs during early ventricular diastole. Left-to-right shunting increases with left atrial contraction and in late systole. The instantaneous magnitude of the shunt decreases and may even reverse (right-to-left) during early ventricular contraction.

Pulmonary Vascular Disease. Although the pulmonary arteriolar circulation in patients with large ASDs undergoes medial hypertrophy over time, severe pulmonary vascular changes (marked intimal proliferation, small vessel occlusion) are uncommon and usually occur after the second decade of life. When these do occur, pulmonary resistance becomes markedly in-

creased, and the right ventricle then is confronted with an increasing pressure load that gradually replaces the previous volume load. The degree of left-to-right shunting decreases, and a dominant right-to-left shunt across the defect eventually may occur. At this stage, typically, there are irreversible morphologic changes in the pulmonary vessels, with pulmonary hypertension being at or above systemic arterial pressure. This is a variant of the Eisenmenger syndrome; the presenting clinical picture is one of cyanosis and severe pulmonary hypertension similar to other end-stage conditions that lead to malignant pulmonary vascular disease. Fortunately, this sequence is rare in patients with ASDs (see also page 433).

Atrial Septal Defects in Adults

Typically, symptoms first appear in late adolescence or later; it is unusual to find a truly asymptomatic subject with an ASD after the age of 40 unless the shunt is very small (less than 1.2 to 1). Fatigue and dyspnea on exertion are early complaints and usually are not severe. Atrial arrhythmias, particularly atrial fibrillation, may develop over time, probably as a result of the chronic right atrial volume overload with atrial dilatation and fibrosis.

The onset of left ventricular disease of any cause (e.g., hypertension, coronary artery disease) may cause a secondary rise in left atrial pressure with a resultant increase in left-to-right shunting. Furthermore, many adults with chronic left-to-right shunting from a large ASD display abnormalities of left ventricular function with decreased left ventricular stroke volume and decreased ejection fraction. Right atrial pressure usually rises only after left atrial pressure has reached an abnormal level; the increased right atrial pressure then may produce signs of systemic venous congestion. If the right ventricle finally fails, right atrial pressure paradoxically will be relatively low, as right-to-left shunting across the defect "decompresses" the right heart.

Mitral Valve Prolapse. In recent years, an estimated 8-37% of patients with secundum ASDs have been demonstrated to have a mid-systolic click and late systolic murmur, with or without echocardiographic evidence of mitral valve prolapse. While the etiology is unclear, it may be a reflection of a common embryologic defect, as the mitral valve leaflets and atrial septum are both formed about the seventh week of gestation. Others believe that prolapse reflects reduced left ventricular size and subnormal stroke volume that results from chronic left-to-right shunting at atrial level.

PHYSICAL EXAMINATION

General Appearance

Some authorities have emphasized a typical body habitus in patients with ASDs, described as gracile (thin, small, and frail). Most believe that there is no pattern to the general appearance of subjects with an ASD. In patients

who have had a large left-to-right shunt since early life, the left chest may bulge anteriorly due to the chronically dilated and hypertrophied right ventricle. Atrial septal defects have been associated with rare disorders such as the Holt-Oram syndrome (absence of the thumbs and other bony defects of the hand and arm), Ehlers-Danlos syndrome (hyperelastic skin, kyphoscoliosis, hyperextensible joints), and Marfan's syndrome. Patients with severe pulmonary hypertension and Eisenmenger's syndrome will be cyanotic, and the distal phalanges will be clubbed; the onset of pulmonary vascular disease and arterial desaturation in late life is known as cyanosis tardive.

Arterial Pulse and Blood Pressure

There are no diagnostic abnormalities of the pulse contour or systemic blood pressure in patients with an ASD. In older subjects, the prevalence of atrial fibrillation is common; 20 to 30% of patients over the age of 40 will have chronic atrial fibrillation.

Jugular Venous Pulse

Careful inspection of the venous pulse in subjects suspected of having an atrial septal defect can be rewarding (Table 20-2). Although the mean venous pressure is rarely elevated, the right atrial V wave characteristically is accentuated in subjects with a large left-to-right shunt. When the atrial defect is of sufficient size, the left and right atria function almost as a common chamber with close approximation of mean atrial pressures. In normal individuals with an intact atrial septum, the left atrial V wave is larger than the A wave; in contrast, in the right atrium the opposite is seen with a dominant A wave. In subjects with a large ASD, the larger left atrial V wave is transmitted to the right atrium, increasing the amplitude of the right atrial V wave, which becomes equal to or exceeds the height of the A wave. This alteration in waveform can be detected in the venous pulse with careful observation (Fig. 20-2).

The height of the right atrial V-wave peak is roughly proportionate to the size of the left-to-right shunt. The X descent is well preserved, and the jugular venous contour exhibits a prominent early systolic collapse (X + X')

TABLE 20-2 *Jugular Venous Pulse Patterns in Atrial Septal Defect*

Waveform Pattern	Clinical Implication
V wave equal to or larger than A wave	Suggests large left-to-right shunt
A wave greater than V wave	Small shunts Occasional moderate to large shunts Associated pulmonary hypertension LV failure
Elevated mean venous pressure	Usually indicates onset of left ventricular dysfunction

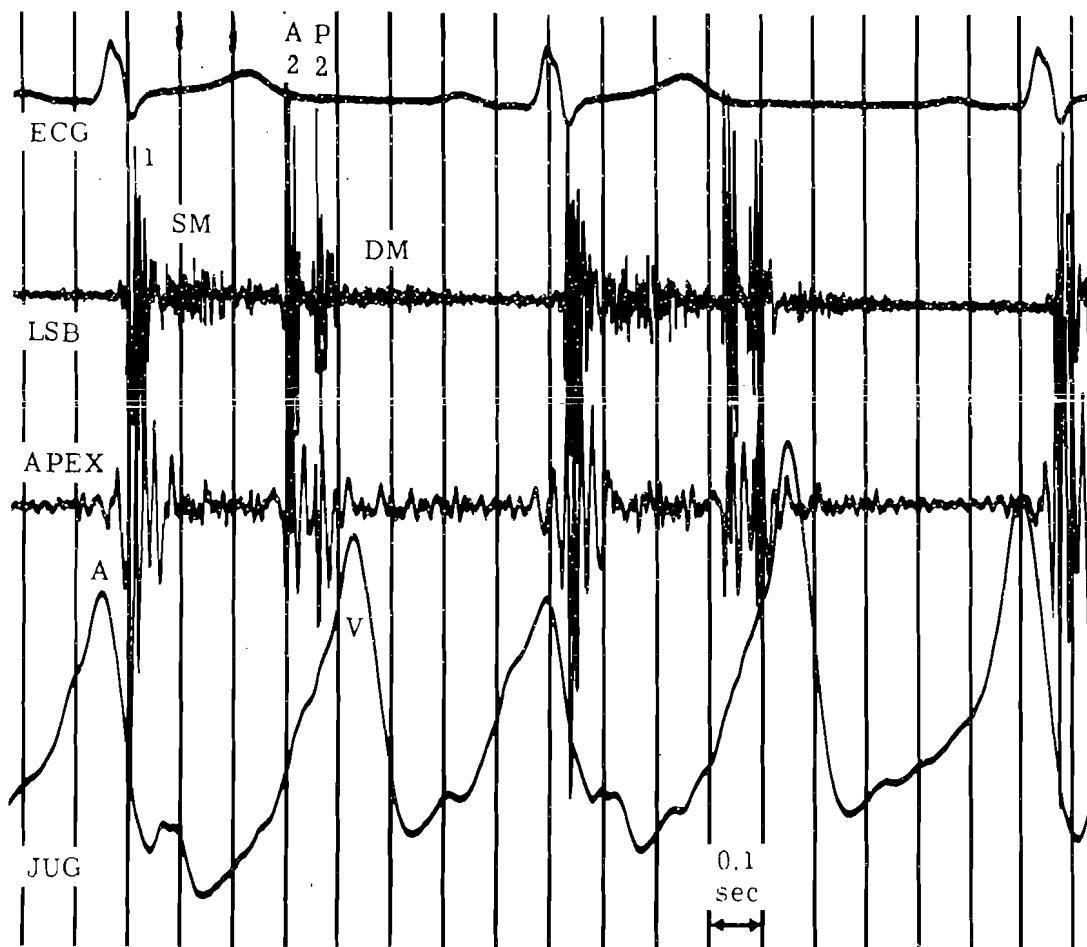


FIG. 20-2. Jugular venous pulse in atrial septal defect. The venous pulse in this patient with a moderately large ASD demonstrates an A and V wave of roughly equal amplitude. The V wave is actually larger than the A wave, which is the reverse of the normal situation. Note also the prominent splitting of S₂ and systolic ejection murmur (SM), as well as a diastolic flow murmur (DM). LSB = lower left sternal border. (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. Prog Cardiovasc Dis 26:145, 1983.)

descent followed by a large V wave). The Y descent is also easily visible. The JVP tracing may have an "M" configuration. *Practical Point: In subjects being evaluated for an ASD the presence of a prominent V wave (equal to or greater in amplitude than the A wave) is suggestive of an atrial communication with a large left-to-right shunt.* The absence of a large V wave, however, does not rule out an ASD. In some patients the A wave retains its normal dominance, usually in those with small left-to-right shunts. If the pulmonary artery pressure is elevated, the right atrial A wave may increase because of an increase in right ventricular end-diastolic pressure and myocardial stiffness.

When left ventricular failure is present, right atrial pressure becomes elevated as the increased left atrial pressure is transmitted to the right atrium. Rarely, the venous pressure in the left jugular vein is higher than that on the right. This finding suggests the presence of partial anomalous pulmonary

venous return draining into the right atrium by way of a persistent left-sided superior vena cava and the coronary sinus.

Precordial Motion

Right Ventricular Impulse. Chronic right ventricular diastolic volume overload caused by an ASD produces an enlarged and hyperdynamic right ventricle. Typically, this results in a prominent parasternal systolic impulse that is easily palpable. The most common parasternal impulse has increased amplitude and normal duration (hyperdynamic motion, see Chapter 5) with retraction during late systole (Fig. 20-3A). This brief outward impulse may be subtle and felt only in held expiration, or it may be quite prominent, producing a forceful parasternal motion, depending on the magnitude of the atrial shunt. If no RV impulse is felt on examination that includes end expiration, ask the patient to stop breathing at maximal inspiration and then palpate the subxiphoid area with the fingers directed upwards (Fig. 5-10). When the right ventricle is massively enlarged, it extends laterally, and the cardiac apex may be formed largely by the right ventricle. More typically, however, the apical area retracts in systole as the more medial midprecordium expands, imparting a rocking motion to the left chest.

Although most large ASDs without pulmonary hypertension produce only a brief hyperdynamic parasternal impulse, large shunts on occasion will result in a *sustained* right ventricular lift or heave, even in the absence of pulmonary hypertension (Fig. 20-2B, Chapter 5). This is likely to be associated with a very large RV chamber that produces palpable activity extending toward the left midchest. Nevertheless, whenever a sustained parasternal heave is detected, the presence of pulmonary hypertension should be strongly

PATTERNS OF PARASTERNAL (RV) MOTION IN ATRIAL SEPTAL DEFECT



FIG. 20-3. Parasternal or right ventricular activity in atrial septal defects. A. A hyperkinetic right ventricular impulse is the initial response to volume overload of the right ventricle. The low amplitude parasternal pulse contour is accentuated and becomes palpable; there is systolic retraction in the second half of the systole. B. Sustained right ventricular lift or parasternal heave. This contour is present when there is a large right ventricular volume overload. When a sustained impulse is present, it indicates a very large right ventricular chamber and/or the presence of pulmonary hypertension.

suspected. Occasionally, a large left-to-right shunt will be associated with normal (impalpable) right ventricular activity. In general, the more prominent the left parasternal activity (amplitude and duration), the larger the left-to-right shunt.

The hyperdynamic parasternal rocking motion often is visible as well as palpable, and the prominent retraction wave may be the most obvious aspect of precordial activity in an ASD. A palpable right ventricular A wave (S4) occasionally may be noted, as well as a palpable rapid filling wave (S3) at the lower left sternal border.

Left Ventricular Impulse. Because the left ventricle is normal in the uncomplicated ASD, palpable left ventricular activity usually is minimal or absent, even with large ASDs. The right ventricle may displace the left ventricle posteriorly, further decreasing the likelihood of a palpable left ventricular apex.

Right Ventricular Outflow Tract and Pulmonary Artery. Careful palpation of the upper left sternal region often will reveal a palpable lift or impulse in the second or third left intercostal space. This is caused by an enlarged right ventricular outflow tract and dilated central pulmonary artery transmitting a large volume of blood during systole. This is particularly noticeable in thin patients. A systolic thrill rarely may be felt over the pulmonary artery area. Gentle palpation with the central part of the hand is best for the detection of this subtle buzzing sensation. Such a finding suggests pulmonic stenosis that may be either functional or anatomic.

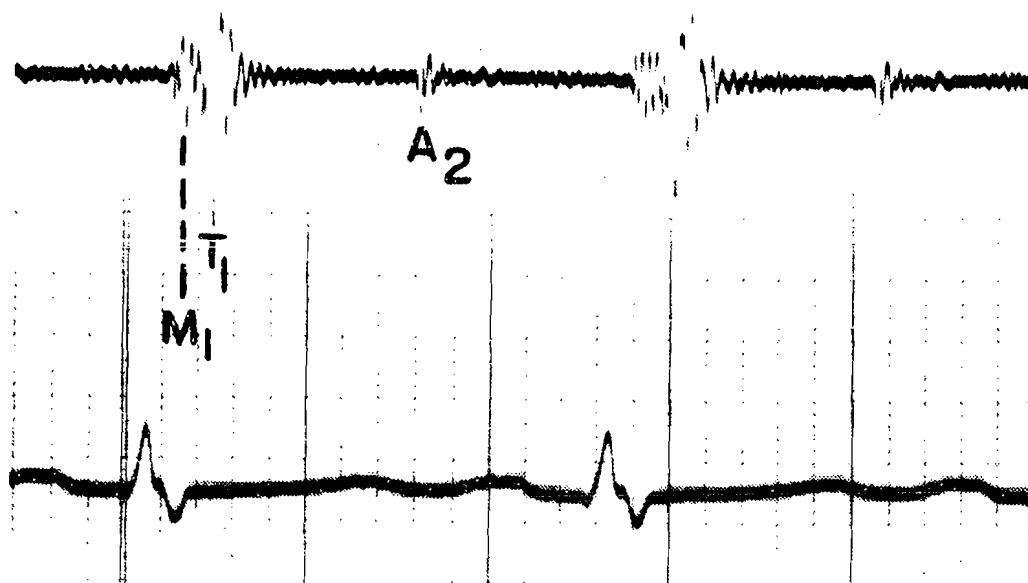
A palpable P2 is another common finding in subjects with ASDs and should suggest the presence of some degree of pulmonary hypertension. Nevertheless, palpable pulmonary artery activity and an increased P2 in the second or third left interspace can be seen with large left-to-right shunts in the presence of normal pulmonary artery pressure.

First Heart Sound

The major characteristic of S1 in a subject with an ASD is increased amplitude of the second major component (T_1) (Fig. 20-4). Audible splitting of S1 is common, resulting from the loud T_1 which also may be delayed because of prolonged RV ejection. On phonocardiography, T_1 is typically discrete and louder than M_1 . However, on auscultation prominent splitting may not always be obvious, and some authors believe that the emphasis on accentuated splitting of S1 in patients with an ASD has been exaggerated.

The mechanism of the increased T_1 amplitude is believed to be rapid and forceful tricuspid valve closure during early right ventricular contraction. This sound coincides with the maximal closing motion of the tricuspid valve and tends to be louder (and later) with larger left-to-right shunts. There is some controversy about the origin of the sound. Some investigators suggest

LSE



2 ICS

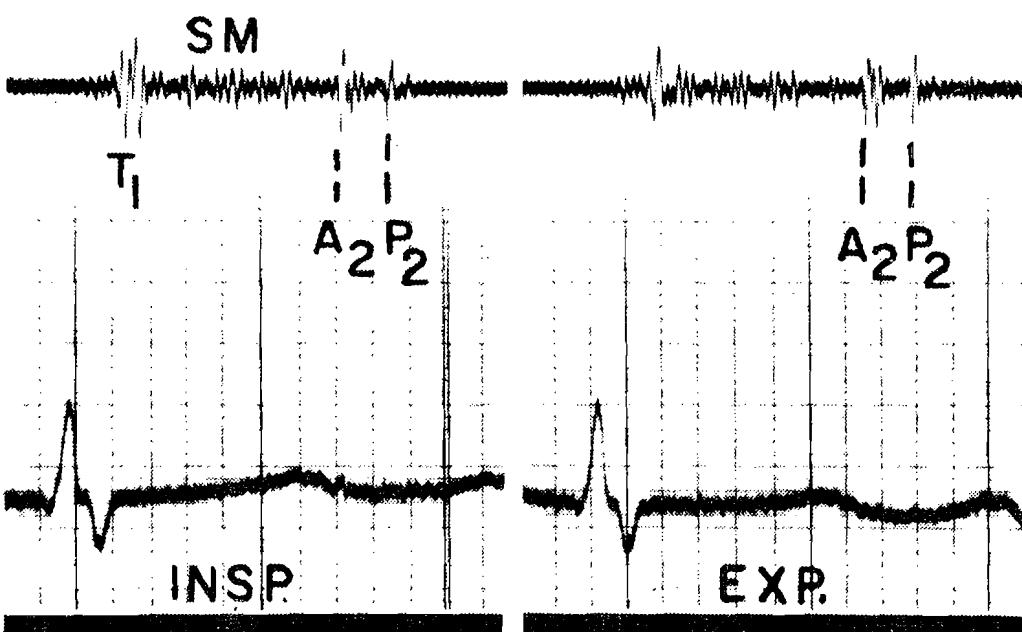


FIG. 20-4. Wide splitting of S₁ and S₂ in atrial septal defect. This phonocardiogram was taken from a young girl with an ostium secundum ASD and normal pulmonary artery pressure. T₁ is prominent and does not change with respiration. At the pulmonic area there is a long pulmonic systolic ejection murmur and wide fixed splitting of S₂. LSE = lower external edge; SM = systolic murmur. (From Perloff JK: Atrial septal defect: In The Clinical Recognition of Congenital Heart Disease, 2nd ed. Philadelphia, WB Saunders Co, 1978.)

that this augmented second component of S1 actually coincides with the onset of pressure rise in the pulmonary artery and is not synchronous with tricuspid valve closure; thus, "T," may actually be a pulmonary artery ejection sound. Both theories may be valid.

Second Heart Sound

The classic S2 in a secundum ASD is widely split in both expiration and inspiration, with little or no detectable respiratory variation (Fig. 20-4). *Practical Point:* *The sine qua non of an atrial septal defect is wide, fixed splitting of S2. This observation is the most specific abnormality detectable on physical examination and whenever found should immediately raise the suspicion of a left-to-right shunt at atrial level, specifically in the absence of a right ventricular conduction delay.* The average interval between A2 and P2 during expiration is 50 msec (range 40 to 70 msec), with an inspiratory variation of 0 to 20 msec. Such small variations are often not detectable by the untrained ear, although Dr. W. Proctor Harvey estimates that slight motion of S2 can be detected in as many as one third of patients with an ASD.

Mechanism of Wide and Fixed Splitting. The combination of a large right ventricular stroke volume and increased capacitance of the pulmonary arterial bed accounts for the wide splitting of S2. The dilated pulmonary artery has a low impedance and long hangout interval (see Chapter 6); thus, the pulmonary incisura is delayed, and P2 follows A2 by a prolonged interval. In addition, the voluminous shunt flow of blood returning to the right ventricle results in a prolonged RV ejection time. In contrast to the normal heart, changes in the degree of right and left ventricular filling during the respiratory cycle are approximately equal in subjects with an ASD; thus, P2 does not move away from A2 in inspiration. Any increase in inspiratory right ventricular return also tends to decrease the degree of left-to-right shunting at atrial level. Right ventricular filling thus stays the same or increases slightly during inspiration, and left ventricular filling remains constant or increases slightly. Therefore, the duration of right and left ventricular ejection is roughly equivalent, and A2 and P2 maintain a constant relationship to each other. Since the normal respiratory alteration in right and left heart filling is balanced by reciprocal changes in shunting across the ASD, respiratory changes in A2 and P2 splitting are blunted.

Other Factors Affecting S2 Splitting. For unclear reasons, splitting of S2 in an ASD tends to widen with age and typically is more prominent in adults than children. In fact, infants and young children with ASDs commonly have narrow splitting of S2, although audible expiratory splitting usually is detectable. Bradycardia results in a wider A2-P2 interval, whereas rapid heart rates shorten this interval. The width of the splitting interval thus is only generally related to the size of the left-to-right shunt.

In normal persons, the Valsalva maneuver causes fusion of A2 and P2 during the strain phase, with wide splitting occurring immediately after release (Fig. 6-10). The converse occurs with an ASD; A2 and P2 move apart during the strain phase, as LV filling diminishes but RV filling remains constant. After release of the Valsalva maneuver, there is no further widening, compared to the normal situation. This is a useful maneuver to perform during auscultation of patients suspected of having an ASD.

In some patients deep inspiration will result in detectable motion of P2 and further widening of the A2-P2 interval. In contrast to normal subjects, assumption of the sitting or standing position does not cause A2 and P2 to fuse in the patient with an ASD.

Atrial Septal Defect with Normal Motion of S2. Occasional patients with documented ASDs have completely normal respiratory motion of A2 and P2. This is more likely to occur with small defects and pulmonic to systemic flow ratios under 2:1. Up to 8 to 10% of patients with ASDs may display audible respiratory motion, and normal splitting is more common in sinus venosus defects. Rarely, normal respiratory variation of S2 may be documented by phonocardiography in pulmonary hypertensive ASDs.

Optimal Evaluation of S2 in Patients Suspected of Having an ASD. Careful auscultation necessitates listening to S2 over many respiratory cycles during normal (nonexaggerated) breathing. It is critical to identify both A2 and P2 and to focus on the interval between them and whether it changes with respiration. After long R-R intervals, the splitting interval tends to increase, often quite prominently. Many normal children and young adults have audible expiratory splitting of S2 in the supine position but normal motion (e.g., expiratory fusion) in the sitting position. Audible expiratory splitting of S2 in the upright position is abnormal and demands further evaluation (see Chapter 6).

When P2 is very loud, auscultation away from the pulmonary area may be helpful in the assessment of respiratory motion. The typical S2 splitting interval is 40 to 60 msec; longer intervals suggest associated pulmonic stenosis. Pulmonary hypertension may or may not cause a decrease in the A2-P2 interval.

Pulmonic Second Sound. Because the pulmonary artery is dilated and blood flow through the right heart is increased in velocity and volume, the pulmonary closure sound (P2) is often accentuated even in the absence of significant pulmonary hypertension. Only a minority (approximately 25%) of patients with ASDs have pulmonary hypertension, although many individuals with ASDs and normal pulmonary artery pressure will have a prominent and snappy P2. If the right ventricle is considerably enlarged, P2 may be well heard throughout the precordium, simulating significant pulmonary hypertension. In some subjects, P2 actually may be audible at the apex in the absence of increased pulmonary artery pressure. In patients with COPD,

obesity, or very large chests, P2 is often attenuated or even absent. If associated pulmonic stenosis is present, P2 may be softer and more delayed than usual.

Differential Diagnosis of the Widely Split S2. The delayed, loud P2 of an ASD can readily simulate an early diastolic sound, such as an opening snap, pericardial knock, or accentuated S3 (Fig. 20-5). Failure of A2 and P2 to move during inspiration may contribute to the confusion. Other causes of wide splitting of S2 may mimic an ASD, such as right bundle branch block, pulmonic stenosis, or idiopathic dilatation of the pulmonary artery. However, in these conditions, careful auscultation should reveal respiratory motion. Patients with pulmonary hypertension and right ventricular failure from any cause may have wide and fixed splitting of S2.

Third and Fourth Heart Sounds

There are conflicting views as to whether an S3 or an S4 may be commonly associated with an ASD in the absence of congestive heart failure; many experienced clinicians do not believe an S3 is a frequent finding in adults with an ASD, although a physiologic S3 can often be heard in normal young children (see Chapter 7). Because an opening snap occasionally may be found on phonocardiography in subjects with an ASD (see below), it is likely that some opening snaps have been mistaken for an S3. An S4 has been documented to occur in some patients with an ASD.

Opening Snap. Recordable early diastolic sounds consistent with an opening snap may be found on high quality phonocardiograms in as many

DIFFERENTIAL DIAGNOSIS

OF WIDE FIXED SPLITTING OF S₂

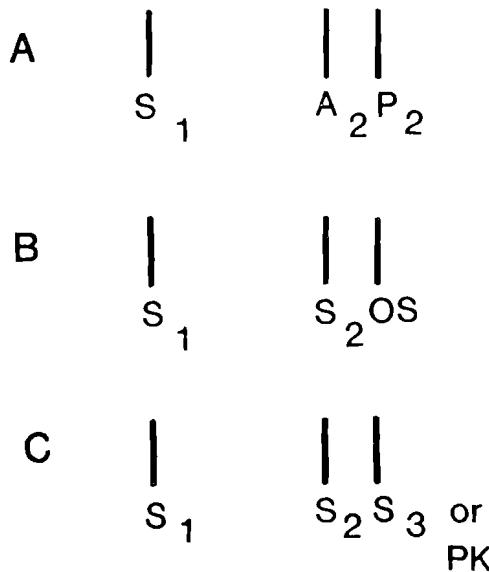


FIG. 20-5. Differential diagnosis of wide fixed splitting of S2. A. Wide splitting of A2 and P2, characteristic of an ASD. There is little to no detectable respiratory variation. B. An S2-opening snap complex can simulate the typical S2 of an ASD, particularly if the opening snap is well heard at the pulmonic area. C. A relatively high pitched S3 or pericardial knock (PK) can also masquerade as wide splitting of S2. In all three examples respiratory fluctuation in the interval between the two audible components is absent.

as one third of patients with an atrial septal defect. The timing of this sound is similar to that of an A-V valve opening snap, occurring 30 to 70 msec after P2 or 110 to 120 msec after A2. These sounds are *usually inaudible* when the stethoscope is used, although they are easily recorded by phonocardiography. The opening snap appears to relate to atrial hypertension and rapid, torrential flow across the tricuspid valve and occurs at the point of maximum opening of the tricuspid leaflets. An opening snap, when present, suggests a pulmonary to systemic flow of 2:1 or greater. When heard, the opening snap of the tricuspid valve in an ASD is soft and lower pitched than the typical mitral opening snap. It is located at the lower left sternal border and may increase in intensity during inspiration.

Pulmonic Ejection Sound. An ejection click or sound is not uncommon in patients with atrial septal defect (Chapter 8). As with the opening snap, it is more readily recorded than heard. The sound is best heard at the upper left sternal border (2nd to 3rd left interspace) and is more easily detected during inspiration. It is softer, and not as high in frequency as the pulmonic ejection sound of pulmonic stenosis. The pulmonic ejection click occurs 40 to 90 msec after S1 and slightly later when pulmonary hypertension is present (delayed right ventricular isovolumic contraction). It is more likely to be present in patients with pulmonary hypertension. An ejection click may be present in as many as 50% of subjects with ASDs. Some experts suggest a lower prevalence and believe a click is present only in subjects with associated pulmonic stenosis or marked dilatation of the main pulmonary artery.

It is important to try to differentiate this sound from an accentuated T₁; both events occur at approximately the same point in the cardiac cycle. A late T₁ and pulmonic ejection sound frequently are confused for one another in patients with ASDs, largely because of superimposition of sounds.

Systolic Murmur

The characteristic murmur in patients with an ASD is a *pulmonic ejection murmur* related to increased flow across the right ventricular outflow tract and pulmonic valve (Figs. 20-2, 20-6). No audible sound is created by the low-pressure shunt flow across the atrial defect itself. Turbulence is created by the often torrential RV stroke volume crossing a relatively narrow right ventricular infundibulum and pulmonic valve into a dilated pulmonary artery. The resulting murmur is often described as scratchy or superficial. It is rarely greater than grade III/VI intensity and typically is of grade II-III/VI intensity. It is best heard at the upper left sternal border, in the 2nd to 3rd left intercostal space, although it may also be prominent at the 4th left intercostal space adjacent to the sternum.

The murmur has a crescendo-decrescendo shape. Typically, there is a short, silent period after S1, followed by the murmur, which ends well before

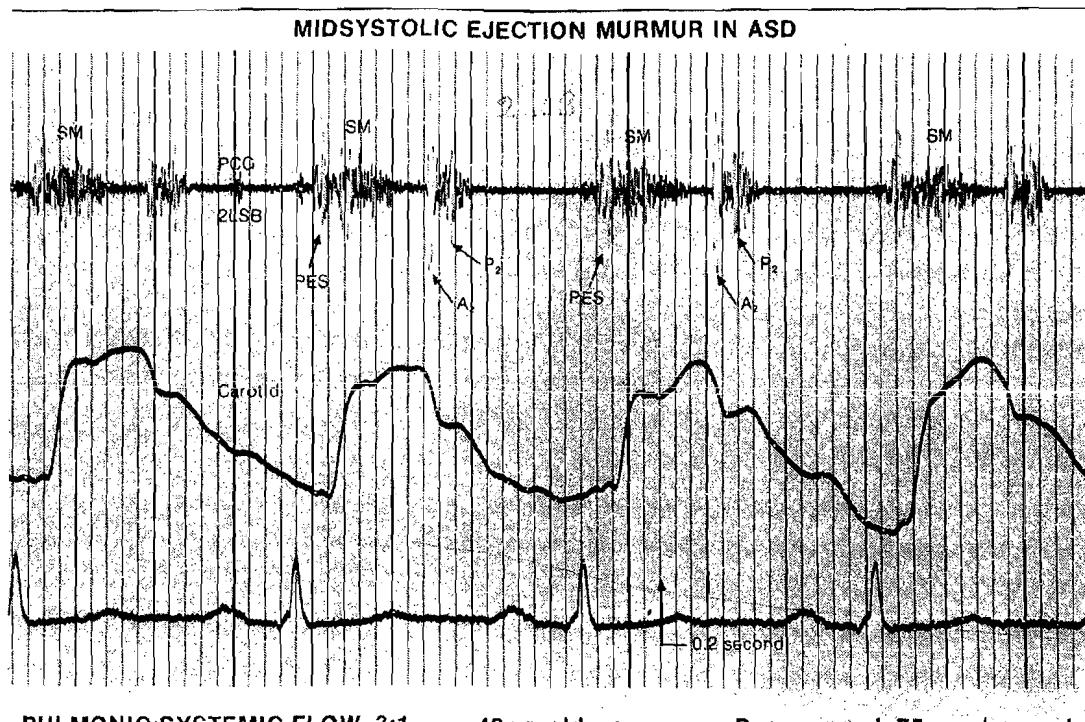


FIG. 20-6. Ejection murmur and wide splitting of S2 in atrial septal defect. This phonocardiogram is taken from a patient with a large left-to-right shunt. A prominent systolic ejection murmur is recorded at the pulmonic area. S2 is widely split and a pulmonic ejection sound is present. SM = systolic murmur; PES = pulmonic ejection sound. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co, 1979.)

S2. Generally, the shorter the murmur, the smaller the shunt. A longer murmur suggests a large shunt and significant right ventricular volume overload or possible associated pulmonic stenosis. As a rule, if the ASD murmur is of IV/VI intensity, one should suspect associated pulmonic stenosis. In such cases, the murmur should be prominent high along the sternal edge and associated with an ejection sound. The systolic murmur may be loud, long, and high-pitched in infants.

Radiation. Careful auscultation often will detect an audible systolic murmur all over the precordium. The murmur may be heard at the apex if the right ventricle is very large. It is often audible in the posterior thorax, interscapular area, and axillae. A prominent peripheral systolic murmur suggests branch pulmonic stenosis, especially if heard in the right chest, axilla, and back; such abnormal narrowing of distal pulmonary arteries is necessary to produce peripheral systolic murmurs unless the left-to-right ASD shunt is very large.

Associated Pulmonic Stenosis. A common dilemma in young patients is the possible association of an ASD with valvular pulmonic stenosis. If there is an abnormal gradient across the pulmonary outflow tract, the systolic murmur tends to be longer and louder. A systolic thrill is far more likely to be present. A pulmonic ejection sound can be heard without pulmonary

stenosis in the presence of an ASD, although a loud click normally favors pulmonic stenosis. P2 may be normal or prominent in spite of pulmonary valve narrowing in an ASD combined with pulmonic stenosis.

Other Systolic Murmurs

Mitral Valve Prolapse. This is a common associated abnormality. When present, a midsystolic click and late systolic murmur may be detected. These abnormalities are best heard at the cardiac apex and should be sought in any patient evaluated for an atrial septal defect. If the prolapse is severe and occurs in early systole, it may produce a holosystolic murmur of mitral regurgitation. The physical findings of mitral valve prolapse are discussed in Chapter 18.

Tricuspid Regurgitation. In patients with an atrial septal defect and severe pulmonary hypertension, the consequent right ventricular dilatation may result in functional tricuspid regurgitation. In such cases, a holosystolic murmur will be present, maximal at the lower left sternal border and increasing with inspiration. Patients with pulmonary hypertension severe enough to produce tricuspid regurgitation will not have the typical pulmonic ejection murmur of an ASD. As the left-to-right shunt disappears, dominant right-to-left shunting occurs, and the pulmonic murmur may disappear entirely, leaving only the tricuspid regurgitation murmur. The typical physical findings of tricuspid regurgitation are reviewed in Chapter 19.

Diastolic Murmurs

Tricuspid Valve Flow Murmur. In patients with large shunts and torrential flow across the tricuspid valve, a functional right ventricular filling murmur may be heard, similar in etiology to the mid-diastolic murmur of severe mitral and tricuspid regurgitation (Figs. 20-2, 20-7). This murmur is usually found in patients with large pulmonary to systemic blood flow ratios greater than 2:1 and begins early in diastole, approximately 40 to 70 msec after P2. It is best heard at the lower left sternal border, in the 4th to 5th interspace; however, when the right ventricle is very large, the murmur may be detected laterally and may be heard at the apex when the latter is formed by the right ventricle.

The diastolic flow murmur is short and may have a crescendo-decrescendo shape. It has been described by some as a scratchy, superficial, rublike sound, higher pitched than most other diastolic filling murmurs; the murmur can be low frequency as well. This murmur can be variable and hard to appreciate. Some have described it as "an absence of silence." It may be augmented by maneuvers that increase venous return to the right heart, such as deep inspiration or leg elevation. It may simulate a loud S3 when brief.

TRICUSPID DIASTOLIC "FLOW" MURMUR IN CONGENITAL ASD

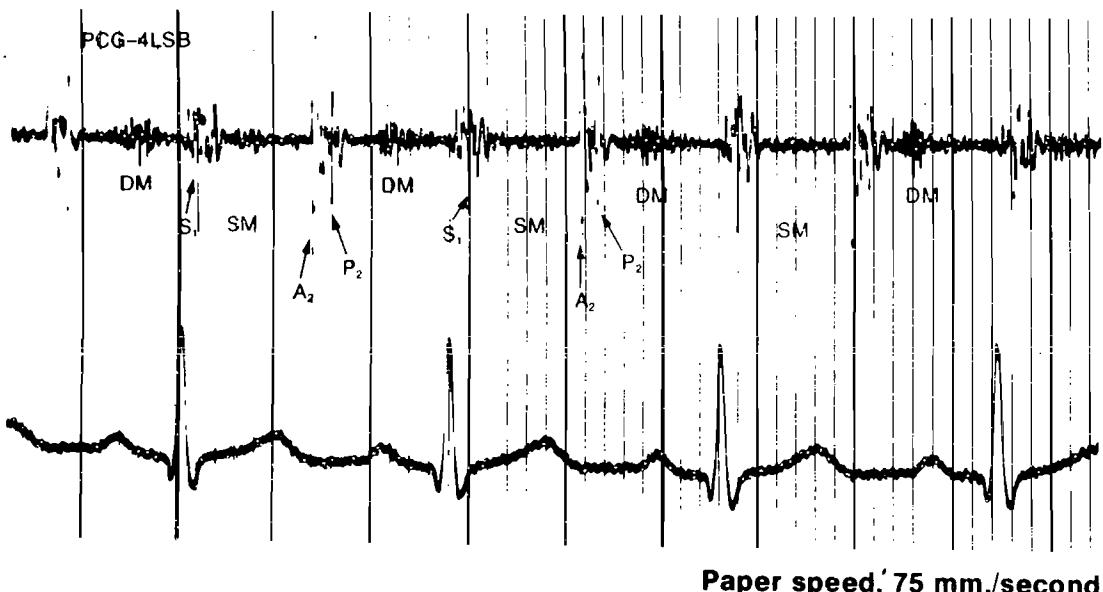


FIG. 20-7. Tricuspid flow murmur in atrial septal defect. A prominent diastolic murmur (DM) is recorded at the lower left sternal border. This represents augmented flow across the tricuspid valve during early right ventricular filling in a patient with a large left-to-right shunt. Note the wide splitting of S₂ and prominent systolic ejection murmur (SM). The presence of a tricuspid flow murmur indicates significant left to right shunting and suggests that the ASD is of surgical importance. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co, 1979.)

Practical Point: The presence of an early to mid-diastolic murmur at the lower left sternal border in a patient with an atrial septal defect suggests a hemodynamically important left-to-right shunt.

Differential Diagnosis. The tricuspid valve flow murmur may be readily confused with the diastolic rumble of mitral stenosis; on occasion, the ASD diastolic murmur is of much higher frequency. The delayed, fixed P₂ in an ASD can simulate an opening snap, and if the diastolic murmur is audible at the apex, the confusion is compounded. Many adults have been referred for cardiac catheterization or cardiac surgery with the diagnosis of rheumatic mitral stenosis, only to be found to have an unexpectedly large atrial septal defect and no rheumatic mitral disease.

Diastolic Murmur Generated Across the Atrial Defect. When left atrial pressure is abnormally elevated, a large pressure differential across the atrial septum can exist, and the resultant flow across the defect may cause sufficient turbulence to produce a murmur. This murmur actually is a continuous murmur, beginning in late systole simultaneous with the left atrial V wave and extending into diastole. It is found when a relatively small atrial septal defect coexists with either mitral stenosis (*Lutembacher's syndrome*) or mitral regurgitation with an elevated left atrial pressure. This diastolic murmur can also be scratchy or crackling. It is audible at the 2nd or 3rd left interspace

and may radiate well to the apex. It may be well heard to the right of the sternum. The murmur can be very soft or quite prominent and may augment with inspiration. When associated with the typical systolic murmur of an atrial septal defect, mitral regurgitation, or the diastolic murmur of mitral stenosis, the physical findings can be confusing. In these unusual situations, the ASD typically is small and may not be easily detected on auscultation.

Pulmonic Insufficiency. Although uncommon, pulmonic insufficiency is another cause for an early diastolic murmur at the left sternal border in subjects with an ASD. Presumably, it results from a very dilated pulmonary artery found in large shunts and is likely to be present when there is significant pulmonary hypertension. The murmur is decrescendo and begins with or shortly after P2; it is best heard along the left sternal border. If pulmonary artery diastolic pressure is high, the murmur is high frequency and is known as a Graham-Steell murmur. If the pulmonary artery diastolic pressure is low, the murmur may be of medium to low frequency.

PULMONARY HYPERTENSIVE ATRIAL SEPTAL DEFECT

Although uncommon, some patients with an ASD develop pulmonary vascular changes that may be severe and result in pulmonary hypertension. In these individuals, the pulmonary vasculature loses its normal distensibility; the increased capacitance of the pulmonary bed associated with a low pressure ASD converts to a stiff, noncompliant, high resistance circulation. Calculated pulmonary arteriolar and total pulmonary vascular resistance are high. In time, the right ventricle may fail. With increasing pulmonary resistance and pressure, the alterations in right ventricular diastolic compliance and pressure result in elevated right ventricular diastolic and right atrial pressures. Left-to-right shunting across the atrial defect diminishes and may disappear. In the early stages, right-to-left flow occurs phasically during the cardiac cycle, and in late pulmonary hypertension the dominant pattern of shunting is right-to-left. The pulmonary arteries demonstrate marked medial hypertrophy and intimal proliferation. These changes, once established, will be progressive throughout adult life.

The resultant severe pulmonary hypertension and high pulmonary vascular resistance typically alter the classic physical findings of an ASD and may make the diagnosis of a shunt at the atrial level impossible to determine at the bedside. This situation is known as the Eisenmenger syndrome, which also occurs with ventricular septal defects and patent ductus arteriosus (both far more commonly than in atrial septal defects). Estimates of the incidence of significant pulmonary hypertension in ASDs range between 20 and 30% of patients, of which a smaller number will progress to the classic Eisenmenger reaction.

General Appearance (Table 20-3)

Typically, the pulmonary hypertensive ASD patient shows central cyanosis of the lips and mucous membranes as well as clubbing of the distal phalanges and cyanosis of the nail beds (Fig. 12-5). Such patients may become easily fatigued and weak.

Jugular Venous Pulse

An abnormally large jugular A wave generally is present. It reflects the increased right ventricular end diastolic pressure. The prominent V wave found in nonhypertensive ASDs is not seen. If tricuspid regurgitation ensues, there may be a large C-V wave in the venous pulse, which can be equal to or greater than the A wave and typically will augment with inspiration.

Precordial Motion

Evidence of marked right ventricular hypertrophy and dilatation will be present. In addition to a sustained and diffuse parasternal lift (Fig. 20-3B), the right ventricle may occupy the entire left medial thorax, including the apex, imparting a rocking motion to the left chest. Both a palpable P2 and a pulmonary artery lift in the 2nd to 3rd left interspace are typical findings. Often, careful examination reveals presystolic distension of the right ventricle, i.e., a palpable right ventricular S4. This is best felt with subxiphoid palpation, using the fingertips directed in the cranial direction (Fig. 5-10).

S2

The sine qua non of the Eisenmenger ASD is a very loud P2, which is typically tambour in quality, palpable, and audible throughout the precor-

TABLE 20-3 The Physical Examination in the Pulmonary Hypertensive Atrial Septal Defect

General Appearance	Cyanosis and clubbing
JVP	Increased A wave amplitude Large V wave if tricuspid regurgitation present
Precordial Motion	Accentuated RV activity—sustained RV lift Palpable P2 and pulmonary artery impulse Palpable RV S4
S2	P2 very loud Splitting can be wide or narrow, but little motion
Pulmonic Ejection Click	
RV S4	
Murmurs	Absence of ASD systolic murmur Absence of diastolic flow murmurs Tricuspid insufficiency murmur common Pulmonic insufficiency murmur common

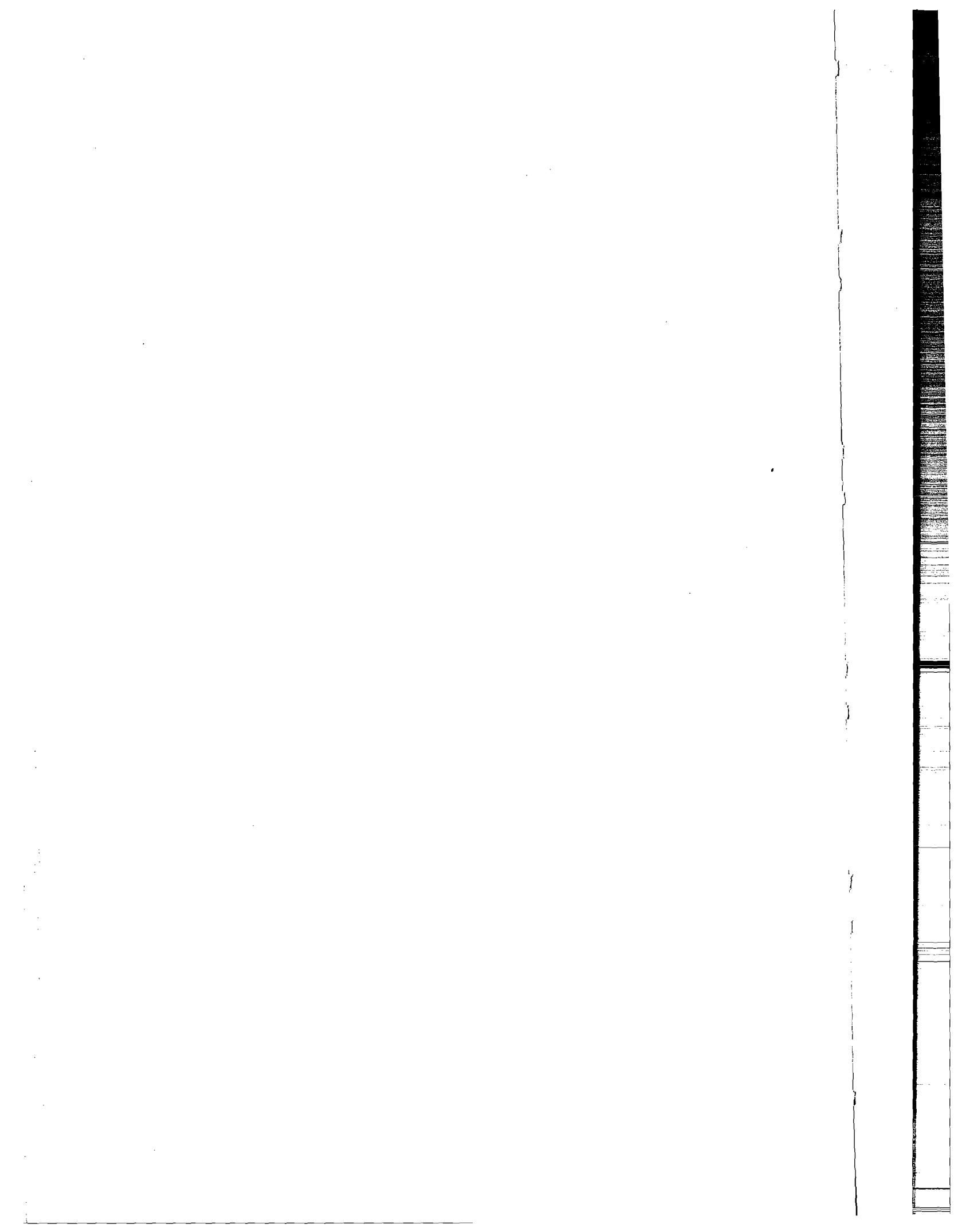
dium. Splitting of S2 is variable in these cases; A2 and P2 may remain wide and fixed, or the splitting interval may narrow considerably. Occasionally, S2 is single, particularly in severe pulmonary hypertension with complete loss of left-to-right shunting. However, when pulmonary artery pressure is elevated but the resistance is not maximally increased, left to right shunting persists and S2 splitting will be little different from usual.

Pulmonic Ejection Sound

The presence of a pulmonary ejection click is variable and, when present, usually unimpressive when the ASD is not accompanied by pulmonary hypertension. When pulmonary artery pressure is significantly elevated, an ejection click is usually heard, produced by abrupt tensing of the stiff, dilated, proximal main pulmonary artery walls during early right ventricular ejection (Figs. 8-2B, 8-6). The ejection sound may soften during inspiration, but this softening is not nearly as prominent as in valvular pulmonic stenosis. The pulmonic ejection sound is best heard at the 2nd to 3rd left intercostal space. *Practical Point: Often it is best to listen to P2 and the pulmonic ejection click away from the pulmonic area in patients with pulmonary hypertension, as respiratory variation may be easier to characterize at precordial sites where the heart sounds are of less amplitude.*

Murmurs

The ASD flow murmur shortens and disappears as pulmonary hypertension becomes more severe, although an ejection murmur can persist because of ejection of blood into a very dilated main pulmonary arterial trunk. Tricuspid regurgitation is a common, late finding in patients with severe pulmonary hypertension. If the right ventricle occupies the entire precordium, including the cardiac apex, the tricuspid regurgitation murmur may be mistaken for mitral regurgitation or a VSD. Careful attention to respiratory behavior of the murmur and inspection of the jugular venous pulse is essential in making the correct diagnosis. The Graham-Steell murmur of pulmonary regurgitation may be present; this is a nonspecific finding in any individual with severe pulmonary hypertension. Presumably, it reflects functional pulmonic valve incompetence related to dilatation of the main pulmonary artery.



Chapter 21

Ventricular Septal Defect

A ventricular septal defect (VSD) is the most common congenital cardiac anomaly, representing 20 to 25% of all *isolated* congenital cardiovascular defects. VSDs are found in 1.5 to 2.5 per thousand live births, are slightly more common in females than males, and are more likely to be present in premature infants. VSDs also are found in association with more complex congenital lesions. When isolated and complex forms are considered together, the total incidence of a VSD approaches 50% of all congenital cardiac lesions. This chapter will deal only with the isolated VSD. Endocardial cushion or A-V canal defects will not be discussed; the reader is referred to pediatric cardiology textbooks for descriptions of these less common cardiovascular abnormalities.

ANATOMY

There are three major varieties of an isolated VSD as classified by location in the heart (Table 21-1). Important identifying anatomic landmarks include the crista supraventricularis in the right ventricular outflow tract, the papillary muscle of the conus, the membranous and muscular ventricular septa, and the medial (septal) leaflet of the tricuspid valve (Fig. 21-1). The most common location for a VSD is in the infracristal region situated beneath and posterior to the crista supraventricularis in the membranous interventricular septum (Fig. 21-1). Typically these *membranous* or *infracristal* defects are located 1 to 2 cm below the aortic valve cusps and occur just above the insertion of the septal leaflet of the tricuspid valve. Frequently there is an associated deficit of the muscular septum. Rarely, a high membranous infracristal VSD results in a communication between the left ventricular outflow tract and the right atrium.

Muscular VSDs (Fig. 21-1D,E) are also infracristal but are much less common, occurring in 5 to 20% of cases. These may be multiple and often appear as ovoid or slitlike fenestrations in the mid or posterior aspect of the trabeculated interventricular septum, anterior to the papillary muscle of the conus.

The least common variety is the *supracristal* VSD (also known as infundibular or subpulmonic) located high in the membranous septum above the crista supraventricularis (Fig. 21-1A). These defects are anterior to and just beneath the pulmonic valve, which forms the superior margin of the

TABLE 21-1 Anatomic Types of Ventricular Septal Defect

	Frequency %
Infracristal or membranous—isolated membranous septum	80
Muscular	5–20
Supracristal	5–7

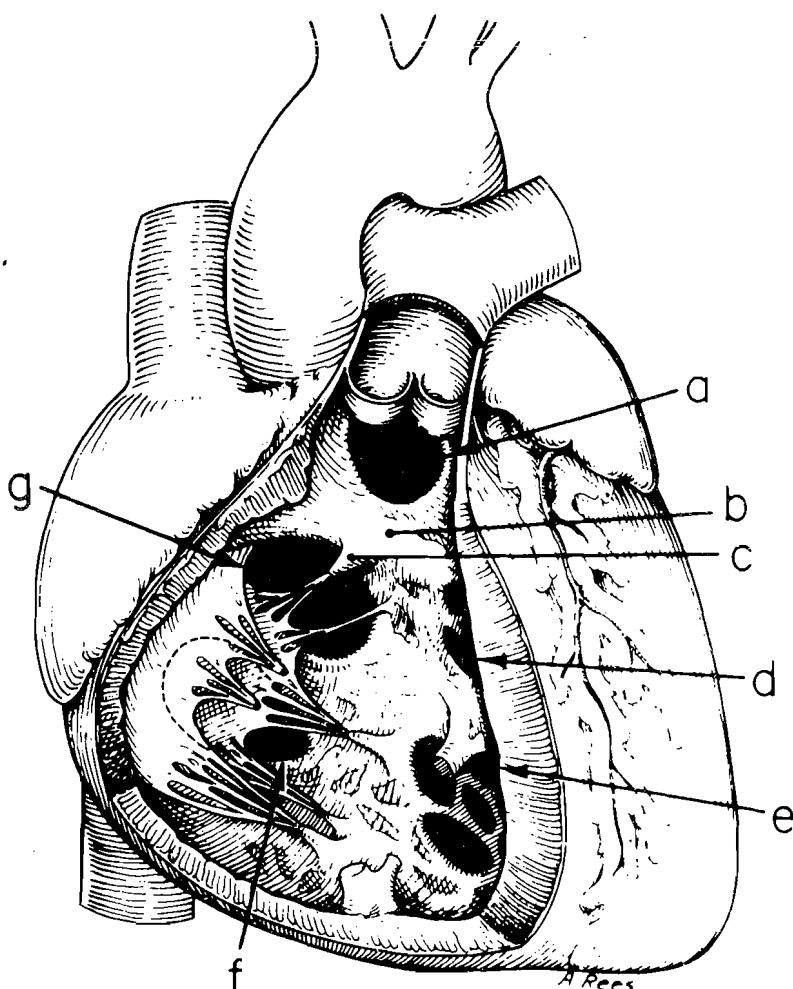


FIG. 21-1. Anatomic locations of ventricular septal defects. The right septal surface of the heart is seen with the right ventricular free wall removed. Important landmarks include the crista supraventricularis (b) and the papillary muscle of the conus (c). Supracristal VSDs are located in the right ventricular outflow tract below the pulmonic valve and above the crista supraventricularis (a). Infracristal or membranous VSDs are located below the crista supraventricularis (g). Muscular VSDs are found in the anterior aspect of the trabeculated portion of the muscular interventricular septum. They may be multiple (d and e). Atrioventricular canal defects are usually part of an endocardial cushion complex anomaly and are typically found near the septal leaflet of the tricuspid valve in the posterior portion of the muscular septum (f). See test for detailed discussion. (From Graham TP, Bender HW, and Spach MS: Ventricular septal defect. In Moss' Heart Disease in Infants, Children, and Adolescence, 3rd ed. Edited by FH Adams and GC Emmanouilides. Baltimore, Williams & Wilkins, 1983.)

defect itself. For unknown reasons, supracristal VSDs are more common in Japan.

Important Variants and Associations. Aortic regurgitation is a well-recognized but uncommon complication of ventricular septal defect (see Chapter 15). It is considered to be an *acquired* abnormality related to inadequate support of the semilunar valve ring or of a sinus of Valsalva because of the contiguous VSD. Aortic regurgitation is found mostly in association with supracristal defects but also with high infracristal defects.

Aneurysms of the membranous septum can occur in association with high ventricular septal defects. These are recognized on angiography as an outpouching into the right ventricular outflow tract from the septum.

Infundibular right ventricular hypertrophy can develop in high flow VSDs producing obstruction to right ventricular outflow. In very large VSDs with torrential left-to-right shunting, severe pulmonary vascular changes can result in markedly elevated pulmonary vascular resistance and subsequent reversal of the left-to-right shunt (Eisenmenger syndrome) (Fig. 21-2C). These individuals have a markedly shortened life expectancy (see also Chapter 20).

PATHOPHYSIOLOGY

The hemodynamic consequences of a VSD relate to both the *size* of the defect and the *status of the pulmonary vasculature*; the location of the defect is of less importance in influencing the degree of left-to-right shunting. Defects may be tiny or huge. Large VSDs are often compared to the size of the proximal aorta; when equal to or bigger than the aortic diameter, the VSD is very large, and torrential left-to-right flow may occur. Extremely large communications effectively create a physiologic single ventricle, with equalization of pressures in the right and left ventricles (Fig. 21-2). Such defects are typically greater than $1 \text{ cm}^2/\text{m}^2$ body surface area.

Severe pulmonary vascular changes are rarely seen early in life, although thick, hypertrophied pulmonary arteries may be persistent from infancy in large VSDs, the pulmonary vascular resistance never falling to normal. Such infants and young children may develop the Eisenmenger syndrome in the first years of life. Severe medial and intimal proliferative changes in the pulmonary bed are generally irreversible, although a period of pulmonary vascular reactivity may exist prior to the later fixed changes.

Left-to-Right Shunting. If the VSD is small (less than $1/2 \text{ cm}$), there is sufficient resistance to left-to-right shunting across it to prevent equalization of ventricular pressures; flow across the defect is dominantly systolic, and the hemodynamic consequences are minimal (Fig. 21-2). Larger anatomic communications allow greater volumes of left-to-right blood flow, which may cause hyperkinetic (high flow) pulmonary hypertension and transmission of left ventricular systolic pressure to the right ventricle and pulmonary artery

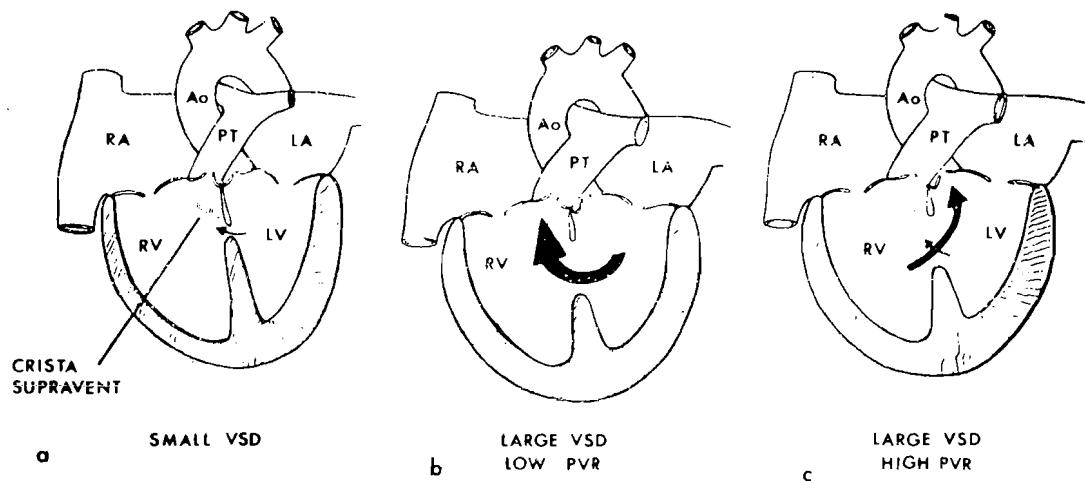


FIG. 21-2. Physiology of ventricular septal defects. A. Small infracristal VSD with minimal to moderate left-to-right shunting, no volume overload, and normal pulmonary artery pressure. B. Large infracristal VSD with low pulmonary vascular resistance and major left-to-right shunting. C. Large infracristal VSD with elevated pulmonary vascular resistance and dominant reversed (right-to-left) shunting. (From Perloff JK: Ventricular septal defect. In *The Clinical Recognition of Congenital Heart Disease*. Philadelphia, W. B. Saunders Co., 1978.)

(Fig. 21-2). Shunting occurs both in systole and in late diastole as long as right ventricular pressures are below that in the LV, and pulmonary vascular resistance remains relatively low. Essentially, the hemodynamic burdens are a pure LV volume overload and pure RV pressure overload at this stage.

With very large unrestrictive defects there is virtual equalization of right and left ventricular pressures; shunt flow occurs in systole as well as in diastole, and some right-to-left shunting may be present as well. The enormous blood flow across the defect into the right heart exposes both the right and left ventricles to the hemodynamic burden. If pulmonary vascular resistance increases, the patient is at risk for developing serious pulmonary arteriolar changes (Eisenmenger syndrome) with subsequent right-to-left shunting (Fig. 21-2). Occasionally, pulmonary artery pressure and resistance become greatly elevated only over a long span of years, but usually the Eisenmenger reaction occurs in children who have had huge left-to-right shunts and hyperkinetic pulmonary hypertension from birth.

In infants and children with very large left-to-right shunts, left ventricular failure may ensue. In older children growth may be slow, and lower respiratory infections are common. Disappearance of left ventricular failure and signs of improvement may occur with spontaneous closure of the defect, the development of right ventricular infundibular stenosis, or the appearance of severe pulmonary vascular disease. Physiologic adaptations of the left ventricle (hypertrophy, dilatation) often result in a compensated hemodynamic state.

Spontaneous Closure of the VSD. It is now believed that the majority of small to moderate defects close completely or reduce their size over time. Estimates of closure rates range between 30 and 40% in the first year of life to 50 to 60% by age 8 to 10. Almost all VSDs that close do so by late

childhood. While it is true that smaller defects are far more likely to disappear, large communications with huge left-to-right shunts and elevated pulmonary artery pressures have been documented to close in about 5 to 10% of cases. Several explanations have been given to explain this phenomenon, including adherence and reduplication of tricuspid valve tissue, endocardial proliferation/fibrosis, and increasing overall cardiac growth without a commensurate change in the defect size.

PHYSICAL EXAMINATION

General Appearance

Small to moderate VSDs produce no change in the general appearance. In infants or children with big defects and large left-to-right shunts, congestive heart failure may be present, and the child may develop slowly with a decreased body weight. A bulge in the left chest area reflects cardiomegaly produced by the large left ventricular volume load that was present in infancy.

In children with the Eisenmenger complex, the lips, fingers and nail beds may be dusky or overtly cyanotic. Clubbing of the distal phalanges may be present (Fig. 12-5). End-stage patients will be weak and easily fatigued; pedal edema and ascites may be present if severe right ventricular failure occurs.

Arterial Pulse

The arterial waveform is usually normal. If the left-to-right shunt is large, there may be a brisk upstroke resulting in a carotid pulse with a rapid rate of rise and normal pulse pressure. In the presence of congestive heart failure or the Eisenmenger complex, the pulse volume is reduced and the contour remains normal.

Jugular Venous Pulse

The venous pulse pressure and contour is within normal limits in the vast majority of subjects with a VSD. If there is congestive heart failure, the mean pressure may be elevated and the A and V waves may be accentuated. In the Eisenmenger complex, the A wave typically is prominent. If tricuspid regurgitation from right ventricular dilation is present, dominant V waves may be seen in the neck veins; this is a late event in the course of the patients.

Precordial Motion

The uncomplicated small to moderate VSD results in a pure left atrial and ventricular volume overload. Thus, the LV impulse is normal to hyperdynamic, similar to that of aortic regurgitation or patent ductus arteriosus;

the apical impulse may be increased in amplitude but is not sustained. In the unusual cases with coexistent aortic regurgitation, the apical impulse may be broad and prolonged. Even with large, left-to-right shunts the LV is not typically displaced laterally, although the impulse may become more forceful and sustained, particularly if left ventricular failure occurs. The actual area of the LV impulse may be bigger than normal if there is significant left ventricular hypertrophy and dilatation.

When pulmonary artery pressure is elevated in large shunts and pulmonary vascular resistance is increased, a palpable pulmonary artery impulse (second left interspace) and lower sternal or parasternal lift may be present (Chapter 5). With marked elevation in pulmonary vascular resistance, signs of right ventricular hypertrophy and elevated pulmonary artery pressures may dominate the precordial examination: right ventricular and pulmonary artery lift, palpable P₂, and even a palpable pulmonic ejection click. In some individuals with a supracristal or membranous VSD, a pulmonary artery lift may be felt in the absence of an abnormal right ventricular impulse, as most of the left-to-right shunting may flow directly into the pulmonary artery.

Systolic Thrill. A systolic thrill at the lower left sternal border (3rd to 4th left interspace) is common in VSDs with loud murmurs. Supracristal defects produce a more superior thrill, which may be maximal at the pulmonic area and radiate toward the left clavicle and neck and the suprasternal notch.

Heart Sounds

First Heart Sound. No significant abnormalities of S₁ occur in patients with ventricular septal defects, although S₁ often is increased in large left-to-right shunts. Very loud systolic VSD murmurs frequently obscure S₁.

Second Heart Sound. Auscultation of S₂ in a patient with a VSD can be difficult. The loud (grade 3 to 4) holosystolic murmur may obscure detection of A₂ and even P₂. A₂ is commonly "lost" in the murmur and only P₂ can be heard, simulating a single S₂. In such cases, phonocardiograms invariably record both components of S₂. Careful auscultation at sites away from maximal murmur intensity may be helpful in identifying both components of S₂.

Respiratory motion of S₂ is normal, although the maximal A₂-P₂ interval during inspiration may be quite wide if the shunt is large. Prominent splitting of S₂ is common, often with audible expiratory separation of A₂ and P₂. The failure of expiratory fusion of the S₂ components is related both to "early" left ventricular emptying (decreased Q-A₂) and delayed right ventricular systole (increased Q-P₂). The expiratory split ranges between 0.02 to 0.06 seconds, but typically is 0.02 to 0.03 seconds. The degree of S₂ splitting is unrelated to size of the left-to-right shunt. However, large flow left-to-right shunts tend to have relatively fixed splitting. Right ventricular dysfunction, if present, will also result in wide S₂ splitting.

Pulmonary Hypertension. P2 is accentuated in patients with elevated pulmonary artery diastolic pressure, whether due to hyperkinetic pulmonary flow or to fixed pulmonary hypertension associated with elevated pulmonary resistance. In the Eisenmenger complex, S2 may be closely split or single, with no respiratory variation (Fig. 8-6). When P2 is very loud it may be helpful to listen several centimeters away from the pulmonic area in an effort to hear both components of S2; backward masking of A2 by a loud P2 is common. A *normal* splitting interval with a loud P2 in the absence of a conduction delay on ECG suggests that pulmonary vascular resistance is not yet maximally elevated and is a clue suggesting that the patient is operable.

Third Heart Sound. The presence of an S3 roughly correlates with the magnitude of left-to-right shunting. It is a sign of a large flow VSD with an excessive volume of blood returning to the left heart across the mitral valve. A mid-diastolic flow rumble following the S3 is common in such situations (Fig. 21-3C); this murmur is an important indicator of substantial left-to-right shunting and is typically present with flow ratios of greater than 2 to 1 (see below). An S3 also may be present in infants with congestive heart failure, but is often a normal finding in children and adolescents (see Chapter 7).

A left ventricular S3 in conjunction with signs of pulmonary hypertension (increased P2, PA, or RV lift) suggests hyperkinetic pulmonary hypertension and implies that the defect is still operable. As pulmonary vascular resistance increases and left-to-right flow decreases, the S3 will disappear.

Opening Snap. On rare occasions, an opening snap may be recorded in high flow shunts due to increased venous return to the left atrium that subsequently crosses the mitral valve (Table 16-3). This sound usually is not audible.

Ejection Sound. With long-standing pulmonary hypertension and a large, tense pulmonary artery, a pulmonic ejection click or sound may be heard (Fig. 8-6). This is a high frequency sound just after S1, best heard at the 2nd to 3rd left interspace. It may decrease somewhat with inspiration. The presence of a pulmonic ejection click is common in patients with high pulmonary artery pressures and the Eisenmenger complex (Fig. 21-3E).

Nonejection Sounds. Peculiar early systolic sounds have been recorded in patients with membranous aneurysm of the interventricular septum. These are often associated with supracristal or high infracristal defects. Presumably such sounds result from tensing of the aneurysm wall.

Murmurs

Systolic Murmur. The hallmark of the VSD is a loud holo- or pansystolic murmur at the lower left sternal border (Fig. 21-3A,C). Classically described as a mixed frequency, even-shaped murmur extending from S1 to S2, the VSD murmur may have midsystolic accentuation and appear to be

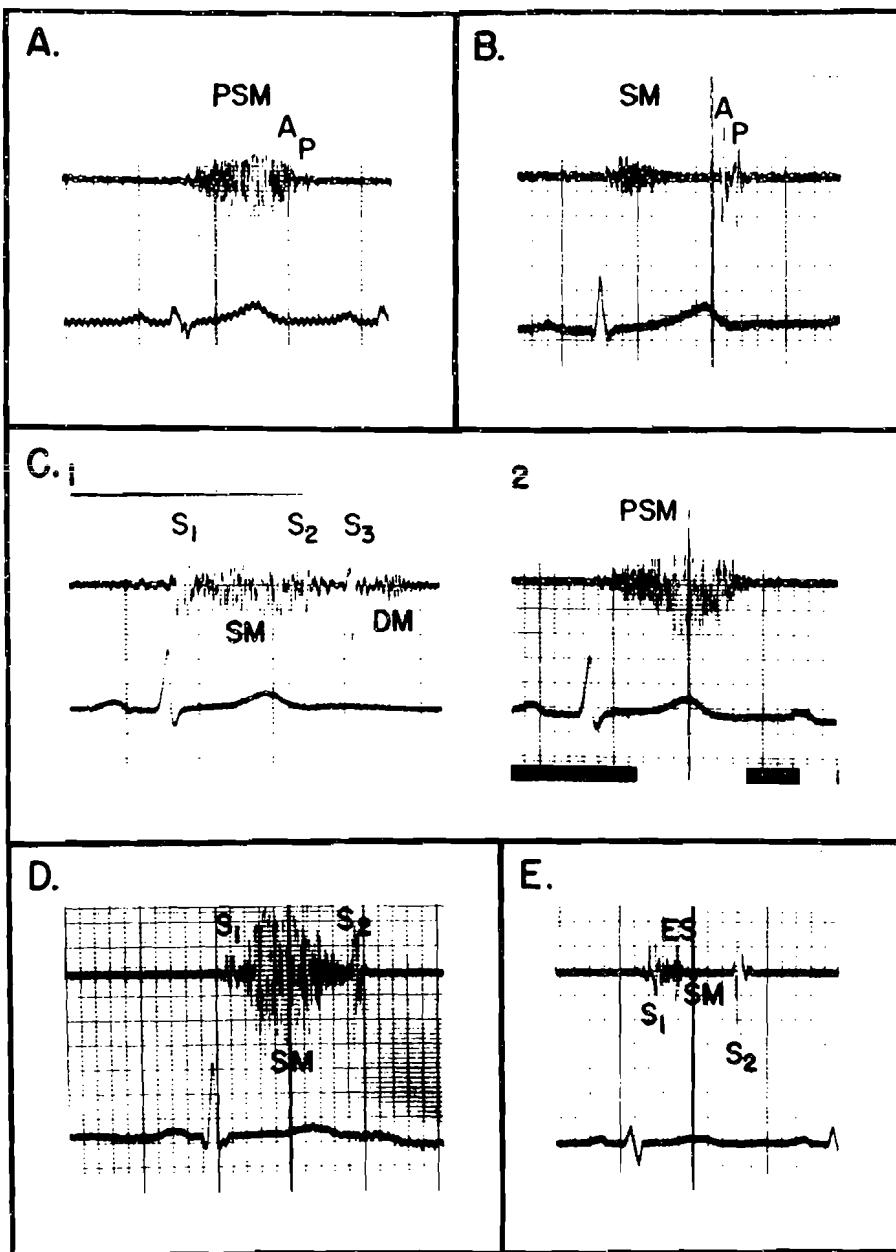


FIG. 21-3. Heart sounds and murmurs in ventricular septal defect. **A.** Small VSD with minimal left-to-right shunt and normal right-sided pressures. Note the prominent systolic murmur (PSM) and wide splitting of S₂. **B.** Small VSD with early systolic murmur (SM). Left-to-right shunting terminates well before the end of systole in certain small defects, particularly those of the muscular variety. **C.** VSD with significant left-to-right shunt and mild pulmonary hypertension. (1.) A prominent S₃ and mitral flow rumble (DM) at the apex. These findings suggest the presence of a hemodynamically important left to right shunt. (2.) Phonocardiogram from the same patient taken at the third left intercostal space where the murmur was most prominent. This murmur is very loud and harsh. **D.** Large VSD with pulmonary hypertension, equivalent right and left ventricular systolic pressure, and significant left-to-right shunting. Note the decrescendo shape of the murmur, which clearly terminates before S₂. This suggests pulmonary hypertension and a decrease in left-to-right shunting in late systole. **E.** Large VSD with marked elevation of pulmonary vascular resistance and bidirectional shunting. Note the pulmonic ejection sound (ES), narrow splitting of S₂, and a short systolic ejection murmur. These findings suggest Eisenmenger physiology and indicate significant pulmonary hypertension. (From Graham TP, Bender HW, and Spach MS: Ventricular septal defect. In Moss' Heart Disease in Infants, Children, and Adolescence, 3rd ed. Edited by FH Adams and GC Emmanouilides. Baltimore, Williams & Wilkins, 1983.)

somewhat crescendo-decrescendo in shape (Fig. 21-3D). When pulmonary vascular resistance is elevated and major pulmonary hypertension is present, the typical holosystolic murmur decreases in length and may ultimately disappear when the left-to-right shunting is no longer present (Fig. 21-3E).

Duration. The murmur of a nonpulmonary hypertensive ventricular septal defect begins with S1 and extends to S2 (Fig. 21-3A,C). As mentioned, A2 and even P2 can be lost in a very loud murmur. In small defects, particularly of the muscular septum, the murmur may be decrescendo and end well before S2, as left-to-right shunting ceases in late systole. This may indicate spontaneous closure is likely (Figs. 21-3B, 21-4). In the presence of congestive heart failure, the systolic murmur may be short.

A systolic murmur that is not holosystolic may create diagnostic difficulties; in such cases the clinician should be alerted to the possibility of a VSD by the location, acoustic qualities, and ancillary findings. Typically,

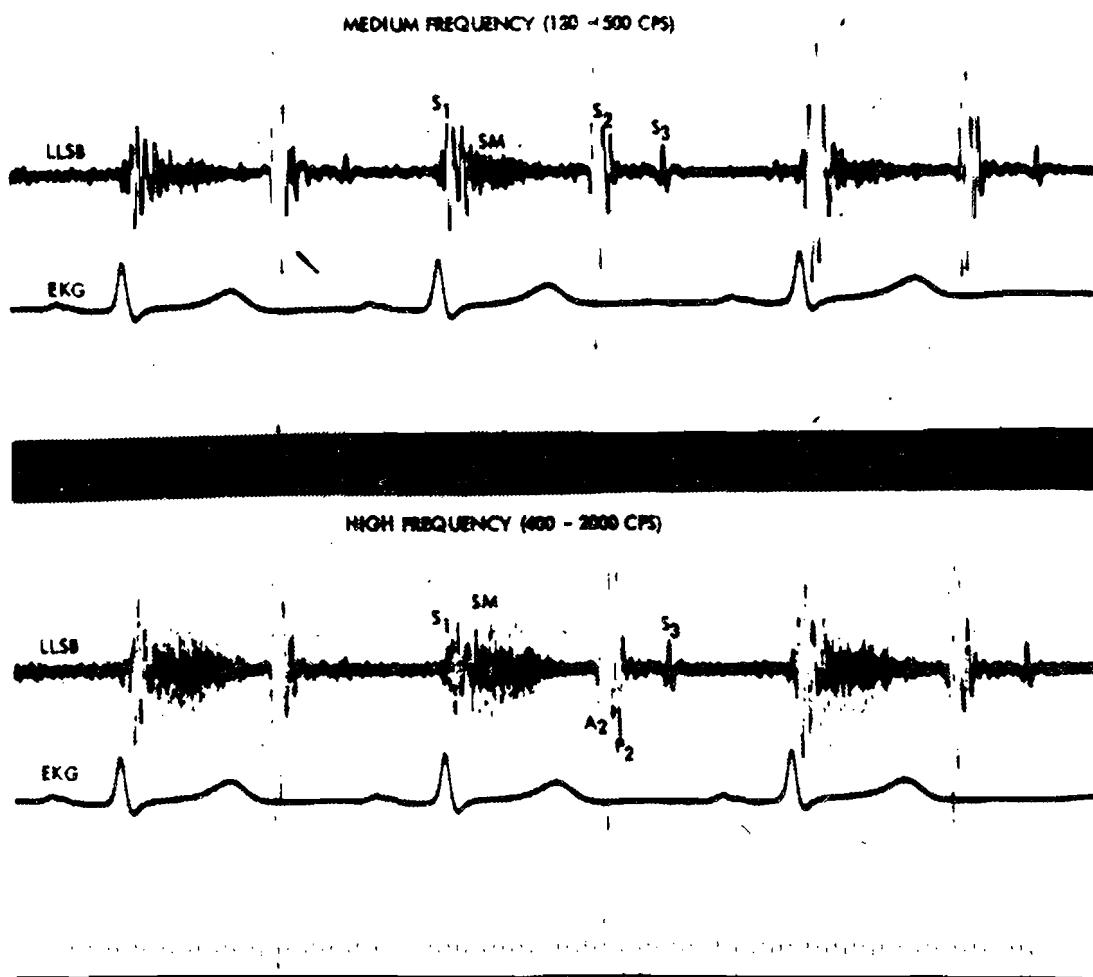


FIG. 21-4. Muscular VSD. The phonocardiogram reveals the typical findings of a small muscular septal defect, with a high frequency early systolic murmur beginning with S1 and ending well before S2. Left to right shunting ceases in mid to late systole as the muscular septum functionally closes off the communication. (From Reddy PS, Shaver JA, and Leonard JJ: Cardiac systolic murmurs: Pathophysiology and differential diagnosis. Prog Cardiovasc Dis 14:1, 1971.)

nonholosystolic VSD murmurs get longer during PVC beats and shorter in the post-PVC beat. In the presence of coexisting valvular or infundibular pulmonic stenosis, the systolic murmur always is long and may extend beyond A2.

When significant pulmonary hypertension is present, the systolic murmur shortens and may become decrescendo in shape (Fig. 8-6, 21-3D,E). Both hyperkinetic pulmonary hypertension (hugh flow) and a nonreactive pulmonary vascular bed can result in a shortened systolic murmur. On the other hand, a holosystolic murmur at the lower sternal edge in the presence of evidence for severe pulmonary hypertension may represent tricuspid regurgitation. This suggests a dilated right ventricle related to the Eisenmenger physiology. Careful attention to respiratory variation will resolve this differential.

If the defect begins to close, usually in early childhood, the astute observer will note a decreasing length of the systolic murmur, which may eventually disappear.

Location and Radiation Patterns. Typically, the maximal intensity of the VSD murmur is at the 3rd to 4th left interspace adjacent to the sternum. This area lies over the right ventricular outflow tract. The murmur may also be well heard at the xiphoid area. Usually, it radiates widely and may be heard to the right of the sternum. Often, the VSD murmur may be best heard just lateral to the left sternal edge, midway between the sternum and the apex, especially if the right ventricle is enlarged.

Usually, a supracristal VSD should be suspected when the maximal murmur intensity is situated more superiorly, e.g., at the 1st to 2nd interspace. In such cases, a thrill may be palpable at the suprasternal notch. These murmurs may radiate into the neck and beneath the left clavicle and do not radiate well to the lower sternal edge.

The rare LV-RA communication causes a superior and rightward murmur orientation, with a peak intensity at the 2nd to 3rd right interspace and radiation to the right clavicle.

Intensity. While the VSD murmur can range from very soft to very loud, it is usually very prominent. *Practical Point: Intensity or loudness does not correlate with the magnitude of left-to-right shunting.* Very small defects may produce the loudest murmurs, and the classic "maladie de Roger" is typically a small shunt with an extremely loud murmur (Fig. 21-3A). These murmurs are usually of grade 4/6 intensity or louder and typically are accompanied by a systolic thrill.

Soft (grade 1-2/6) murmurs are found occasionally in very small defects ("pinhole") or rarely with hugh shunts across very large septal communications. In the presence of congestive failure, the murmur may be soft. The advent of pulmonary hypertension causes the systolic murmur to become softer as well as shorter. *Practical Point: Decreasing amplitude of the VSD*

murmur in an individual over a period of months to years suggests either spontaneous closure of the defect or the development of a major increase in pulmonary vascular resistance. As left-to-right shunting decreases and right-to-left flow dominates, the murmur may completely disappear. Note that the associated conditions producing a shorter than usual murmur often result in a softer than usual VSD murmur (Table 21-2).

Frequency. There is considerable variety to the tone or frequency of the VSD murmur. The loud Roger murmur may be harsh, but high frequency vibrations are also audible; auscultation is best carried out using the diaphragm of the stethoscope. Softer murmurs are often quite musical, whirring, and regurgitant in quality (relatively pure frequency). Small defects may produce a relatively high frequency murmur. Some authors have commented that muscular defects may be quite superficial and high pitched in quality.

Shape. While the classic VSD murmur is relatively even in intensity throughout systole, variations are common. Decrescendo systolic murmurs are common as the defect closes in very small (often muscular) defects (Fig. 21-4), as well as with the development of elevated pulmonary vascular resistance (Fig. 21-3B, D, E). On the other hand, midsystolic accentuation may be present with large left-to-right shunts. In such instances a pulmonic flow murmur is superimposed upon the VSD murmur (Fig. 21-3D). These patients may have a higher than usual point of maximum intensity, e.g., third left interspace. This configuration is also seen when there is pulmonary stenosis at valvular or infundibular level. The murmur of the supracristal VSD may be crescendo-decrescendo in shape and may peak later in systole.

VSD murmurs that simulate ejection systolic murmurs may be found in large shunts and in pulmonary hypertensive patients. In such cases, careful auscultation usually will detect sound vibrations extending to S2; however, the peak intensity is in midsystole.

Mid-Diastolic Murmur. An early to mid-diastolic murmur or "flow rumble" is an important finding in subjects with a VSD (Fig. 21-3C). This murmur is similar in etiology to the mid-diastolic murmur of severe mitral regurgitation or the tricuspid flow murmur of large atrial septal defects. Such a murmur signifies a large volume of blood traversing the mitral valve and indicates both increased pulmonary flow and a large pulmonary to systolic flow ratio (greater than 2 to 1). *Practical Point: The presence of a mid-diastolic murmur in a patient with an isolated ventricular septal defect implies that the*

TABLE 21-2 Causes of a Nonholosystolic Murmur in Subjects with a Ventricular Septal Defect

Very small defect
Muscular defect
A closing VSD
Pulmonary hypertension
Hyperkinetic due to huge L to R shunt
Pulmonary vascular disease (Eisenmenger syndrome)

degree of left-to-right shunting is hemodynamically significant and suggests that the patient should undergo catheterization and possible surgical correction, especially if indicated by other signs or symptoms.

Acoustical Qualities. This murmur typically follows a loud S3 (also related to large flow into the left ventricle) and has a brief duration, ending well before late diastole. It is best heard at the apex and may be audible only when light pressure is used in placing the bell of the stethoscope in the left lateral decubitus position. The murmur is low frequency, rumbling in quality, and similar in characteristic to the diastolic murmur of mitral stenosis; however, no opening snap is present, and the murmur is not audible in late diastole.

Mild exercise (sit-ups, running in place) may increase the intensity of the mid-diastolic murmur. In the upright position, this murmur may not be heard or may become softer. The murmur does not increase with inspiration. If there is evidence for congestive heart failure, a mid-diastolic murmur indicates that the cause of decompensation is torrential left-to-right shunting with a severe, left ventricular volume overload.

The disappearance of a mid-diastolic murmur is an indication of decreasing left-to-right shunting due either to development of increased pulmonary vascular resistance and pulmonary hypertension or to the occurrence of spontaneous closure of the defect.

AORTIC REGURGITATION

An unusual but important complication of VSDs is aortic regurgitation, occurring in up to 5% of patients. This is generally an acquired lesion related to inadequate support of the aortic valve cusps and sinuses of Valsalva. Aortic regurgitation may be seen in either supracristal or high infracristal defects. For unknown reasons, supracristal VSDs occur with greater frequency in Japan. Typically, the right coronary cusp is involved. There may be thickening and distortion of the valve leaflets or overt prolapse of an entire cusp. Occasionally, the aortic leaflet can herniate into the left ventricular outflow tract and obliterate the ventricular septal defect. Rarely, the cusp can herniate through the VSD and cause right ventricular outflow tract obstruction.

Associated aortic regurgitation may not be discovered until late childhood. It is important to realize that the degree of aortic leak is likely to be progressive. The regurgitation can be quite severe, and its hemodynamic importance ultimately may far outweigh the consequences of the ventricular defect itself.

The murmur of aortic regurgitation may be a faint blowing diastolic murmur or a loud, medium-frequency rough regurgitant murmur (see Chapter 15). The combination of a prominent systolic and diastolic murmur may simulate the machinery murmur of a patent ductus arteriosus, especially if

the VSD murmur is harsh and loud. One should listen on both the left and right side of the sternum for an aortic regurgitation murmur.

RIGHT VENTRICULAR OUTFLOW TRACT OBSTRUCTION

Occasionally, VSDs are complicated by obstruction to right ventricular outflow, either at the pulmonic valve or at the sub- or supravalvular level. A VSD can coexist with congenital valvular pulmonic stenosis. When the obstruction to right ventricular outflow is due to hypoplasia of the crista supraventricularis, this combination is called the tetralogy of Fallot. When mild, the pulmonary obstruction results in a loud, long systolic murmur, often with midsystolic accentuation, which is as prominent at the 2nd to 3rd left interspace as at the 3rd to 4th interspace. On careful auscultation, this murmur may be found to augment with inspiration. P2 may be more widely split, but only if there is an unusual delay in right ventricular ejection will this be of diagnostic importance. A pulmonic ejection click that decreases with inspiration may be heard (see Chapter 8). P2 also may be softer than expected in an isolated ventricular septal defect.

Some children with large VSDs actually develop subvalvular, right ventricular infundibular hypertrophy that can be obstructive. In such instances, the right ventricle is faced with an increasing afterload and may hypertrophy out of proportion to the VSD. A midsystolic murmur is superimposed on the VSD murmur, and if the RV-PA gradient is large, left-to-right shunting into the right ventricle will decrease and the volume overload of the VSD will diminish as well. The signs of pulmonic stenosis may come to dominate the clinical picture, although a pulmonic ejection click is not heard when pure infundibular stenosis is present.

SEVERE PULMONARY HYPERTENSION AND THE EISENMENGER COMPLEX

The most feared complication of a VSD is the development of structural alterations in the pulmonary vasculature, with intimal proliferation, marked medial hypertrophy, and fibrosis of the pulmonary arterioles leading to irreversible and marked elevation of pulmonary vascular resistance. As resistance to right ventricular ejection increases, the left-to-right shunt diminishes and finally becomes a right-to-left shunt (Fig. 21-2). The presence of severe pulmonary hypertension with right-to-left shunting is known as the Eisenmenger syndrome, complex, or reaction. The term "Eisenmenger complex" is usually related to a VSD, whereas "Eisenmenger syndrome" or "reaction" relates to other shunt lesions where elevated pulmonary vascular resistance results in dominant right to left shunting.

The Eisenmenger complex is rare in infants and very young children and usually occurs later in childhood or adolescence. Typically, it results from a very large shunt; pulmonary artery pressure never decreases from systemic levels while pulmonary vascular resistance steadily increases. Alternatively and less commonly, a large left-to-right shunt with initially normal or mildly increased pulmonary artery pressures may evolve into an Eisenmenger reaction over time. Pulmonary vascular resistance increases progressively, and the left-to-right shunt decreases concomitantly. In either case, the sequelae are the same: continuous, severe right ventricular pressure overload and gradually decreasing left ventricular volume overload as the left-to-right flow diminishes. Ultimately, the clinical picture is that of severe pulmonary hypertension and marked cyanosis. It may be impossible to differentiate the original site of the left-to-right shunt in patients with Eisenmenger physiology: patients with an ASD, VSD, or patent ductus arteriosus, alone or in combination, may be clinically indistinguishable (see Table 21-3).

TABLE 21-3 Physical Findings of Ventricular Septal Defects in Relation to the Magnitude of the Left-to-Right Shunt

Magnitude	Physical Findings
Small	Looks healthy Precordial exam normal Usually loud, long grade 3-4 holosystolic murmur, often associated with thrill S2 prominently split with normal motion; S3 may be present L-to-R shunt less than 1.5 to 1.0
Medium	Looks healthy May be somewhat thin Hyperdynamic LV impulse Possible RV, PA lift Loud grade 3-4/6 holosystolic murmur, usually with thrill; S3 followed by middiastolic murmur at apex S2 widely split; P2 increased slightly L-to-R shunt 1.6-3.0
Large	May be in overt CHF; often poor growth JVP: may have a large A wave Precordial motion: LV hyperdynamic, RV impulse, PA lift, palpable P2 VSD murmur may be holosystolic; S3 likely P2 increased with narrow split (S2 may appear single)
Pulmonary hypertension with Eisenmenger reaction (Right to left shunting)	Cyanosis and clubbing common JVP: Big A or V wave Carotids small Precordium: Sustained RV lift, PA impulse, and P2 often palpable; LV impulse small P2 single, loud RV S3 may be heard Murmurs of pulmonic regurgitation or tricuspid regurgitation may be present L-to-R shunt less than 1.5; R-to-L shunt greater than 1.0

Physical Examination. Cyanosis is usually present, although in the early stages it may be noticeable only with exertion. Clubbing of the distal phalanges may develop (Fig. 12-5). The jugular venous pulse generally does not suggest an increased mean pressure because the right ventricle is able to decompress via the systemic circulation. However, a prominent jugular A wave is typical. If the right ventricle dilates, tricuspid regurgitation may result, which in turn can produce a jugular C-V wave with an increase in mean venous pressure; this occurs late in the course of the Eisenmenger complex.

The precordial examination is altered considerably with a "quiet" or even impalpable left ventricle and a prominent right ventricular heave. The right ventricle may occupy the cardiac apex if the chamber enlarges sufficiently. A palpable, pulmonary artery lift in the 2nd to 3rd left interspace, as well as a palpable P2, may be present.

On auscultation, the classic findings of severe pulmonary hypertension are heard (Fig. 21-3E): a loud or booming P2, typically with a single or narrow S2 and relatively fixed splitting; a pulmonary ejection click; and often a right ventricular S3 (Fig. 8-6). The systolic murmur of the VSD disappears, although a pulmonary flow murmur may be present as a result of flow into the dilated main pulmonary artery. The murmurs of pulmonic insufficiency (Graham Steell murmur) and tricuspid regurgitation may appear once pulmonary vascular resistance exceeds systemic. The pulmonic regurgitation murmur simulates aortic regurgitation and is detectable as a high frequency blowing diastolic murmur along the left sternal border. If tricuspid regurgitation appears, inspiratory augmentation of a new holosystolic murmur is likely, along with prominent jugular V waves and a pulsatile liver (see Chapter 19). All of these findings are similar to those of an Eisenmenger ASD (see Chapter 20).

DIFFERENTIAL DIAGNOSIS

The classic case of a VSD is easily recognized at the bedside. Small defects, especially in the muscular septum, may produce soft systolic murmurs that do not extend to S2 and can simulate a functional or innocent systolic ejection murmur. The murmur's high frequency characteristics, early onset (coincident with S1), and location at the lower left sternal border should suggest a VSD. On occasion, the ejection murmur of mild aortic and pulmonic stenosis may be confused with a small VSD, but in general this should not be a diagnostic problem. Congenital semilunar valve stenosis usually is associated with an ejection click not found in the uncomplicated VSD (although early systolic sounds are associated with supracristal defects). The location

of the VSD murmur is lower down the left sternal edge than stenotic valve murmurs.

Mitral regurgitation, the cause of the most common holosystolic murmur, should not be confused easily with a ventricular septal defect. The maximal location at the apex with radiation to the axilla are the best clues to mitral origin.

The to-and-fro murmurs of a VSD complicated by aortic regurgitation can mimic a number of defects having continuous murmurs, such as patent ductus arteriosus, sinus of Valsalva aneurysm, coronary AV fistula, as well as combined aortic stenosis and regurgitation.

When the VSD is very large in infants, the typical murmur may not be present. Any congenital defect (e.g., patent ductus, truncus arteriosus) that results in congestive heart failure and a large left-to-right shunt may be hard to distinguish from the VSD in such situations.

The pulmonary hypertensive sequelae of the Eisenmenger reaction produce findings that are similar to all causes of severe pulmonary hypertension. A patent ductus arteriosus and an atrial septal defect can also result in Eisenmenger physiology and on physical examination may be impossible to differentiate from an Eisenmenger VSD. The presence of left ventricular hypertrophy on precordial examination favors either a VSD or PDA, whereas a widely split S₂ suggests an atrial defect. When cyanosis of the toes is present without cyanotic fingers or lips, the diagnosis is likely to be a patent ductus arteriosus with an Eisenmenger reaction.

Chapter 22

Acute Myocardial Infarction

Michael A. Chizner, M.D.*

Important physical findings are often detectable in patients with acute myocardial infarction. Through careful, repeated examination in an orderly, organized manner (utilizing the "five-finger" approach advocated by W. Proctor Harvey, M.D.), information regarding the hemodynamic state may be readily obtained.

General Appearance

The physical appearance of the patient during the acute onset of myocardial infarction may vary considerably, depending upon the severity of the chest pain and the presence of any associated hemodynamic changes. The patient's facial expression, gestures, and bodily position can reveal information about the degree of discomfort and overall clinical status. While chest pain is still present, the patient may appear restless, anxious, and agitated, often thrashing about in bed in an effort to find a more comfortable position. He may have an anguished facial expression, clenching his fist against the sternum, while complaining of pain. This gesture is referred to as Levine's sign, a type of "language of the hands." The patient may exhibit evidence of marked sympathetic discharge, appearing ashen, pale, and profusely diaphoretic with cool and clammy skin, even in the absence of shock. Most of these signs are caused by vasoconstriction resulting from an intense reaction to increased catecholamine levels.

Respiratory distress may dominate the clinical picture when left ventricular failure is present. The patient may be sitting upright in bed gasping for breath. If cardiogenic shock is the predominant feature, signs of peripheral circulatory collapse become evident, e.g., cold and clammy skin, marked facial pallor, cyanosis of the lips and nail beds, and bluish-red mottling of

* This chapter is modified with permission from the monograph, *Bedside Diagnosis of the Acute Myocardial Infarction and its Complications*, by Michael A. Chizner, M.D., *Current Problems in Cardiology*, Vol. 7 No. 9, 1982, Year Book Medical Publishers, Chicago.

the extremities. The patient may become confused, disoriented, or obtunded, reflecting an altered sensorium resulting from cerebral hypoperfusion.

Such dramatic and severe clinical presentations are not usually present. Many individuals appear in good health with no visible abnormality in appearance or behavior, even while complaining of chest discomfort, shortness of breath, nausea, or weakness. Occasionally, patients may appear slightly feverish, as low-grade temperature elevation may be present for several days following the infarction. This is a nonspecific response to tissue necrosis, usually occurring after the first 24 hours and reaching its peak on the second or third hospital day.

Arterial Pulse

Initially, the systemic blood pressure is elevated in many patients in the early stages of acute myocardial infarction, even in the absence of pre-existing hypertension; this is part of the marked reaction to endogenous catecholamine release resulting from pain, agitation, and apprehension. The majority of such patients demonstrate a gradual decline in blood pressure during the first 1 to 2 days following the myocardial infarction; as recovery occurs, the arterial blood pressure tends to return toward preinfarction levels. Hypotension may result from many factors, including the venodilating effects of morphine and nitrate administration, reduced cardiac output due to impaired left ventricular function, or hypovolemia. Some patients with inferior wall infarction may be hypotensive in the absence of cardiogenic shock due to intense parasympathetic discharge. This vagus-mediated reflex presumably originates in chemoreceptors in the heart, resulting in bradycardia, hypotension, and peripheral vasodilation.

Careful palpation of the radial arterial pulse will reveal information about the heart rate and rhythm. The radial pulse is palpated with the tips of the first two or three fingers, varying the amount of pressure until a maximal impulse is felt. The heart rate is often within normal limits in uncomplicated myocardial infarction. During the first few days, however, especially in anterior wall infarctions, sinus tachycardia is often present. If persistent, it may reflect an inadequate cardiac output and in general suggests a less favorable prognosis. A number of underlying etiologies should be considered, including congestive heart failure, pulmonary embolism, pericarditis, and borderline cardiogenic shock. Occasionally, bradycardia is present, especially with inferoposterior wall myocardial infarctions. Arrhythmias of some type occur in almost all patients with an acute myocardial infarction.

Information about left ventricular ejection and cardiac function may be derived by careful palpation of the carotid arterial pulse (see Chapter 3). A small volume, hypokinetic pulse suggests the presence of a diminished cardiac

output. In the presence of left ventricular failure, one may also appreciate a slow upstroke and a small volume pulse, reflecting a reduced rate of left ventricular ejection. A brisk, hyperkinetic carotid upstroke may be present in patients with severe mitral regurgitation due to papillary muscle dysfunction or rupture or in patients with an acute left-to-right shunt resulting from a ventricular septal rupture. Gentle palpation of the radial (or femoral) arteries may detect pulsus alternans, a valuable early sign of left ventricular decompensation (Fig. 3-4). In pulsus alternans, each cardiac impulse occurs at a regular interval, but there is a variation in the force or amplitude of the pulse with alternate beats, i.e., stronger arterial pulses alternating with weaker arterial pulses. Pulsus alternans, even when barely detectable, presents an immediate clue to the presence of compromised left ventricular function and should alert the examiner to search carefully for ventricular diastolic (S3) gallop (see also page 447).

Jugular Venous Pulse

Inspection of the internal jugular veins may reveal a normal venous pulse, an increased mean pressure, or abnormal venous pulsations. For optimal examination the patient should be comfortably positioned without a pillow, with the neck relaxed and the head turned slightly away from the examiner to allow maximal visibility of the neck veins. The head of the CCU bed should be adjusted until the venous pulsations of the right internal jugular vein are easily appreciated (Fig. 4-2).

The height of the jugular venous pulse is usually normal or only slightly elevated in patients with an acute myocardial infarction, even in the presence of mild to moderate left ventricular failure. Marked elevation of the neck veins occurs with right ventricular failure, in right ventricular infarction, or with the rare case of tricuspid insufficiency due to right ventricular papillary muscle necrosis. Large or "giant A" waves may be seen when there is vigorous right atrial contraction due to an increased resistance to emptying, resulting from pulmonary hypertension associated with left ventricular failure or pulmonary emboli complicating an acute myocardial infarction (Fig. 4-4A).

Arrhythmias. When the atrium contracts against a closed tricuspid valve large "cannon" A waves are present; these occur in complete heart block, during ventricular tachycardia, and with premature ventricular contractions.

When first degree A-V block is present, common in acute inferior wall myocardial infarction, the time from the jugular venous A wave to the upstroke of the carotid arterial pulse is prolonged, corresponding to a long PR interval on the electrocardiogram. To observe this phenomenon, it is often easier to feel the left carotid artery while inspecting the right internal jugular vein. In atrial flutter, rapid flutter waves occasionally may be visualized in

the neck veins at a rate much greater than that of the ventricular rate. If atrial fibrillation occurs, A waves are no longer visualized, and only V waves remain. The venous V wave, normally smaller than the A wave, may become accentuated (pathologic C-V wave) when the tricuspid valve does not remain completely closed during ventricular systole. This may occur with tricuspid regurgitation from right ventricular papillary muscle necrosis or in the presence of a right ventricular infarction.

Precordial Palpation

In the patient hospitalized with a first myocardial infarction and in the absence of prior heart disease, the heart size is normal and the apical impulse or point of maximal impulse (PMI) will be palpable in its normal location at the fifth left intercostal space within the midclavicular line (see Chapter 5). The normal impulse is appreciated as a brief outward movement in early systole without palpable diastolic movements (Fig. 5-1).

Abnormal impulses, however, may be commonly palpated during acute myocardial infarction. Analysis of the apical impulse is best assessed with the pulps of the fingertips with the patient turned to the left lateral decubitus position (Fig. 5-7). Palpable presystolic distention of the left ventricle, corresponding to the S4 gallop on auscultation, occurs as a result of vigorous atrial contraction into a ventricle with reduced compliance from the stiffened area of infarcted myocardium (Fig. 7-6). If left ventricular decompensation occurs, a palpable outward movement of the left ventricle in early diastole may develop, corresponding to the S3 gallop on auscultation. An S3 also may be palpable when early diastolic filling of the left ventricle is accentuated, such as may occur with ventricular septal rupture or mitral regurgitation due to papillary muscle rupture. In these situations, a markedly increased volume of blood flows into the left ventricle across the mitral valve immediately after mitral valve opening. At times, evidence of a gallop rhythm can be seen and felt, even when not well heard.

An *ectopic systolic impulse* (bulge) may be palpated in the third to fifth interspaces between the lower left sternal border and the cardiac apex ("ectopic" area); this represents the infarcted segment of myocardium expanding paradoxically during systole. This may be a clue to the presence of a ventricular aneurysm (Fig. 22-1). In some patients, the ectopic impulse is palpable medial and superior to the apical impulse, clearly separable from the apical left ventricular impulse. In others, the abnormal LV impulse is located at the actual PMI. Careful simultaneous palpation of the apex and ectopic area with two hands will help reveal the asynchrony between the two impulses. The abnormal outward movement may be diffuse and not clearly separable from the apical impulse. Such generalized heaves or bulges also can be observed during an episode of angina pectoris, or in the acute phase of myocardial

infarction, prior to anatomic thinning of the myocardium. These may be considered "physiologic" aneurysms, since they are present when the heart is ischemic or in its early stages of infarction.

If a murmur of acute ventricular septal defect or mitral regurgitation is present and is sufficiently intense (Grade IV or more), the vibrations may be transmitted to the surface of the chest and be perceived as a palpable thrill. These sensations are best detected with light palpation using the palmar surface of the hand at the junction of the fingers and the palm, rather than with the fingertips.

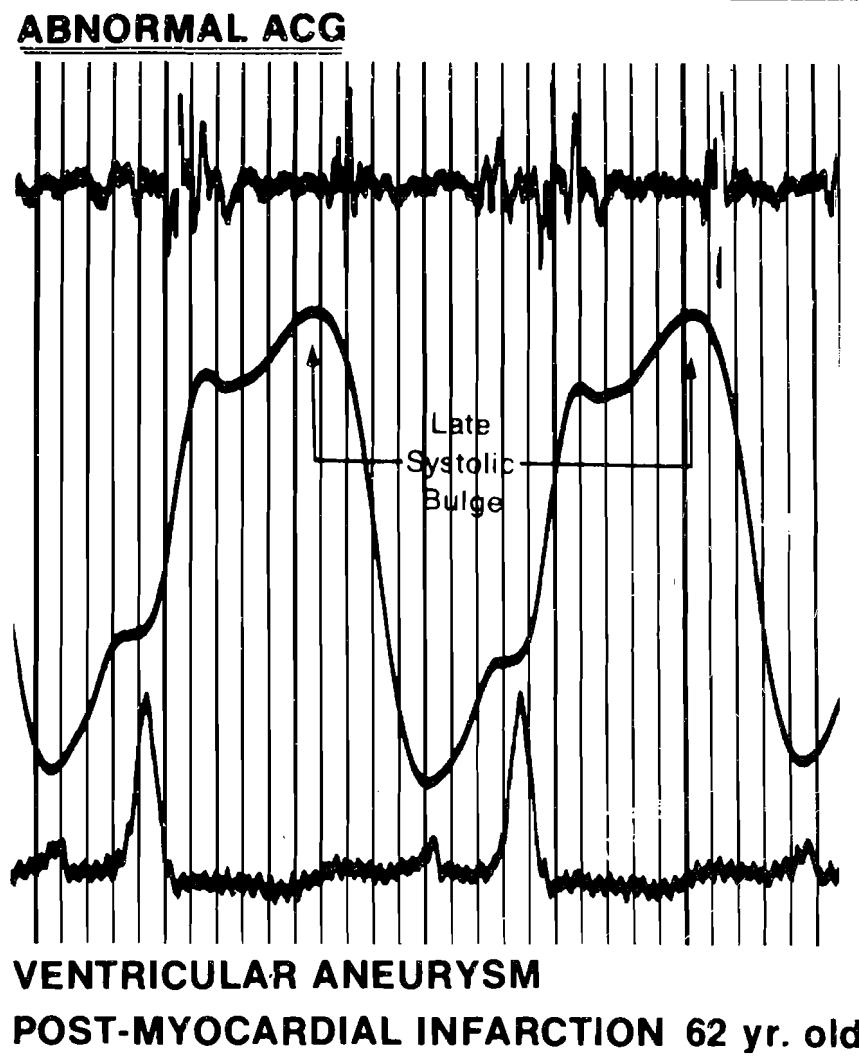


FIG. 22-1. Abnormal precordial systolic impulse or apical bulge associated with a left ventricular aneurysm. Note the late systolic expansion of this sustained left ventricular impulse. The A wave is also accentuated and would probably be palpable. This pattern can also be seen with a dyskinetic left ventricular impulse in the absence of a true left ventricular aneurysm. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co., 1979.)

First and Second Heart Sounds

In many cases of acute myocardial infarction, the first heart sound (S1) and the second heart sound (S2) are faint in intensity and muffled in quality because of diminished myocardial contractility. S1, in particular, usually is quite soft and may be indistinguishable in quality from S2, resulting in a tic-tac rhythm. A soft S1 may be present when there is a prolonged P-R interval such as occurs in first degree AV block associated with inferior infarction; here the atrioventricular valves have had more time to "float" to a partially closed position after atrial contraction but before the onset of left ventricular contraction. On the contrary, when the P-R interval is short, as in sinus tachycardia, S1 is typically loud, since the atrioventricular valves are wide open and their excursion is large (Fig. 6-3). In acute infarction, impaired LV contractile performance will independently reduce S1 amplitude even in the presence of sinus tachycardia. The examiner will occasionally appreciate variation in the intensity of S1 if the P-R interval varies, such as in complete heart block or some cases of ventricular tachycardia; S1 also varies with changing cycle lengths during atrial fibrillation.

Close attention to the inspiratory motion of S2 may reveal reversed or paradoxical respiratory splitting, which can rarely occur in the presence of severe left ventricular dysfunction in the absence of left bundle branch block. In complete left bundle branch block, the sequence of aortic and pulmonic valve closure may be reversed due to delayed activation and contraction of the left ventricle (Fig. 6-9). Abnormally wide or exaggerated "physiologic" respiratory splitting of S2 may occur in such conditions as right bundle branch block (because of delayed activation of the right ventricle) or an acute ventricular septal defect because of the large RV stroke volume and delayed pulmonic valve closure. Severe mitral insufficiency, usually due to papillary muscle involvement, may also result in wide physiologic splitting of S2 due to earlier closure of the aortic valve caused by shortened left ventricular ejection time as blood rapidly egresses from the left ventricle into both the aorta and left atrium.

If pulmonary hypertension is present (during left ventricular failure or from pulmonary emboli complicating an acute myocardial infarction), P2 may be accentuated in intensity.

Third and Fourth Heart Sounds

Careful auscultation at the cardiac apex may reveal atrial (S4) or ventricular (S3) gallop sounds in many instances of acute infarction. Their presence, particularly with rapid heart rates, may produce a cadence referred to as gallop rhythm. The atrial (S4) gallop, or fourth heart sound (Fig. 7-7), is essentially universal in patients with acute myocardial infarction, reflecting a reduction in left ventricular compliance due to stiffening of the infarcted

myocardium; this results in a more powerful left atrial contraction. The left-sided S4 gallop, like all low-pitched events, is best elicited by turning the patient to the left lateral decubitus position. Identify the apex beat with the palpating fingers of one hand, placing the bell of the stethoscope precisely on the apical impulse with the other hand and applying light pressure while barely making an air seal (Fig. 7-9). Visible and palpable presystolic distention of the left ventricle at the cardiac apex frequently accompanies the audible S4 gallop (Figs. 7-3, 7-6). Coupling of the left ventricle with the chest wall is an important but poorly understood factor influencing S4 audibility.

The appearance of a ventricular (S3) gallop, or third heart sound, denotes more severe changes in left ventricular compliance and usually implies more extensive and advanced left ventricular damage and dysfunction. The S3 gallop is frequently detectable but is not nearly as common as the S4 gallop in acute myocardial infarction. A pathologic S3 is an auscultatory feature of left ventricular failure, but an S4 has a much less ominous implication. The left-sided S3 gallop is also best heard with the patient turned in the left lateral decubitus position, with the bell of the stethoscope positioned lightly over the cardiac apex (Fig. 7-9). It is a low frequency sound heard during the early diastolic rapid filling phase, occurring approximately 0.15 seconds after S2 (Fig. 7-3). It is more common with transmural anterior wall myocardial infarctions than with inferior or nontransmural infarctions. In patients with an acute myocardial infarction, the presence of an S3 gallop is associated with an abnormally elevated left ventricular filling pressure, and if louder and persistent, the S3 carries a poor prognosis.

The presence of an S3 does not always indicate left ventricular failure, as it may also occur as a flow phenomenon related to increased mitral blood flow associated with mitral regurgitation or a ventricular septal defect (see Chapter 7). The S3 gallop occasionally is accompanied by the palpitory sensation of a sharp outward precordial movement in early diastole, associated with the rapid inflow of blood to the LV. Because the S3 is of low frequency, it may be seen or palpated more readily than heard.

Third and fourth heart sounds can originate in either the left or the right ventricle. As the right ventricle underlies the anterior precordium adjacent to the lower sternal edge, right-sided gallop sounds are therefore heard best at the lower left sternal border, increase in intensity with inspiration, and are often associated with other features of right ventricular involvement such as a parasternal lift, accentuated P2, or elevated jugular venous pressure with large A or V waves. These findings suggest pulmonary hypertension, pulmonary emboli, or right ventricular infarction.

Murmurs

Systolic murmurs, transient or persistent, are commonly audible in patients with acute myocardial infarction. A new systolic murmur in the

setting of an acute myocardial infarction may represent mitral regurgitation resulting from papillary muscle dysfunction (due to ischemia, left ventricular dilatation, or ventricular aneurysm formation) or papillary muscle rupture (see Chapter 17). The murmur of papillary muscle dysfunction may be transient or persistent and can be early, mid, late, or holosystolic in timing (Figs. 17-10, 17-11, 22-2). It is best heard at the cardiac apex and may radiate to the axilla or base. More severe involvement of the papillary muscles, as in left ventricular papillary muscle rupture, is catastrophic, resulting in pulmonary edema and cardiogenic shock. This murmur typically is loudest at the cardiac apex; it is holosystolic, but decrescendo in the latter part of systole due to high pressure V waves in the left atrium with a reduced regurgitant flow in late systole (Fig. 17-8). If cardiogenic shock is present, a surprisingly soft murmur may be heard even in the presence of hemodynamically severe mitral regurgitation or the murmur may be absent. Rupture of the interventricular septum may rarely occur, producing a harsh, loud, grade IV or greater, holosystolic murmur, loudest at the lower left sternal border with a frequently associated palpable thrill. Although a thrill is less commonly associated with papillary muscle rupture, the maximal location of the murmur and thrill (i.e., left sternal border for ventricular septal rupture and apex for papillary muscle rupture) should help differentiate between the two conditions at the bedside, but this can be difficult at times.

Tricuspid regurgitation may result from acute right ventricular failure complicating left ventricular failure, right ventricular papillary muscle dysfunction, or from right ventricular infarction. The holosystolic murmur of tricuspid regurgitation is heard best at the lower left sternal border; it increases on inspiration due to the increase in venous return to the right side of the heart resulting from the inspiratory fall in intrapleural pressure (see Chapter 19, Fig. 19-4).

Pericardial Friction Rub

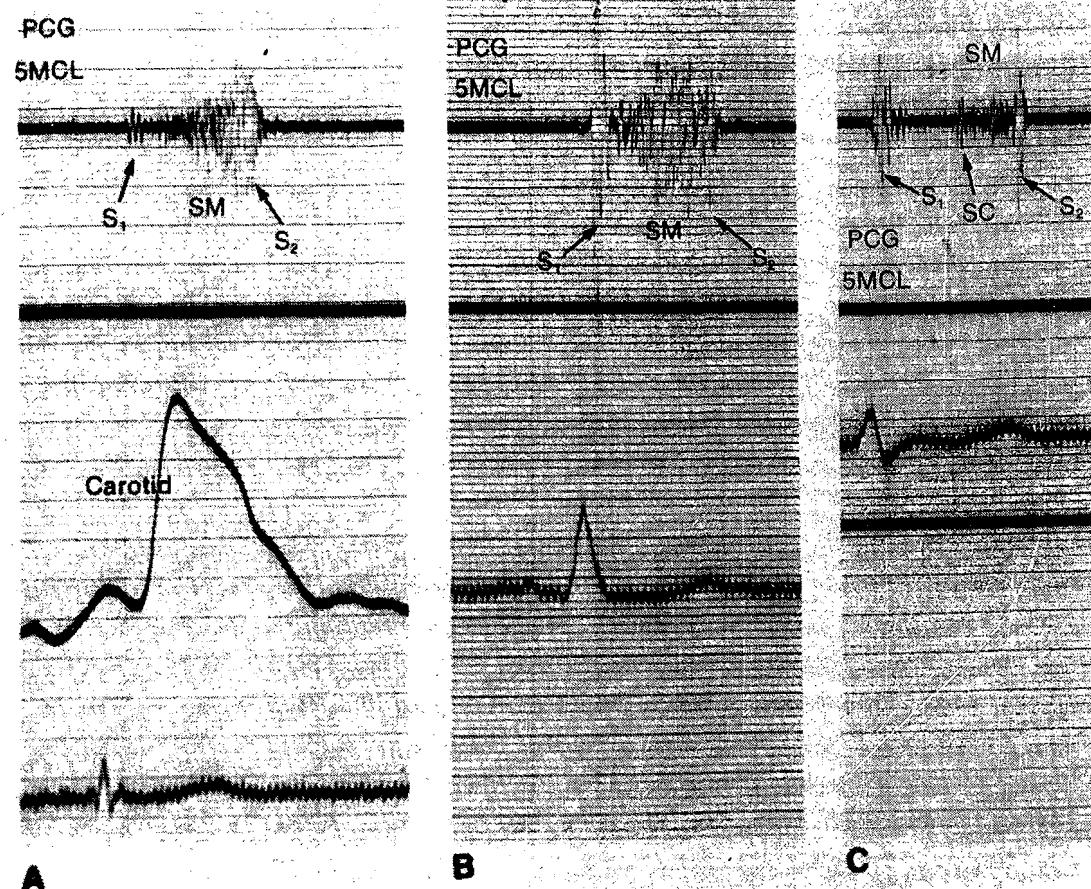
Pericardial friction rubs may be present, usually occurring several days into the course of acute myocardial infarction. Rubs are usually absent within the first 24 hours of infarction, but occasionally may be present when the patient is first seen if there has been a delay in hospitalization. Friction rubs are audible in up to 20% of patients with acute myocardial infarction and are notorious for their evanescence. Frequent, repeated cardiac auscultation is necessary for their identification. Rubs are characterized by a scratchy, superficial, leathery quality, coinciding with the three phases of the cardiac cycle: atrial systole, ventricular systole, and early ventricular diastole (Fig. 22-3). They are heard at the lower left sternal border, over the sternum, or, occasionally, at the cardiac apex. Use firm pressure on the diaphragm of the stethoscope (try to leave an impression in the skin on removal). Friction rubs

may be influenced by phases of respiration; as a rule they increase in intensity and thus are most readily detected during held inspiration. If loud enough, they too may be palpable, (friction fremitus).

Congestive Heart Failure

The most common mechanical complication and the most frequent cause of death in patients hospitalized with acute myocardial infarction today is "pump failure." This occurs at least transiently in one third to one half of all patients with myocardial infarction. Myocardial infarction compromises ventricular function by reducing contractility, producing wall motion abnormalities, and altering chamber compliance. Because anterior infarctions, par-

LATE SM OF PAPILLARY MUSCLE DYSFUNCTION



CORONARY ARTERY DISEASE

FIG. 22-2. Papillary muscle dysfunction. Three examples of papillary murmurs in patients with coronary artery disease. A. The classic late systolic murmur which begins in early-mid systole and crescendoes to S₂. B. A louder and longer systolic murmur that is markedly accentuated in mid-late systole. Note the increased S₁. C. Late systolic murmur with mid-systolic click. The presence of a non-ejection click suggests mitral valve prolapse secondary to coronary artery disease due to papillary muscle fibrosis or dysfunction. (From Delman AJ and Stein E: Dynamic cardiac auscultation and phonocardiography. Philadelphia, WB Saunders Co., 1979.)

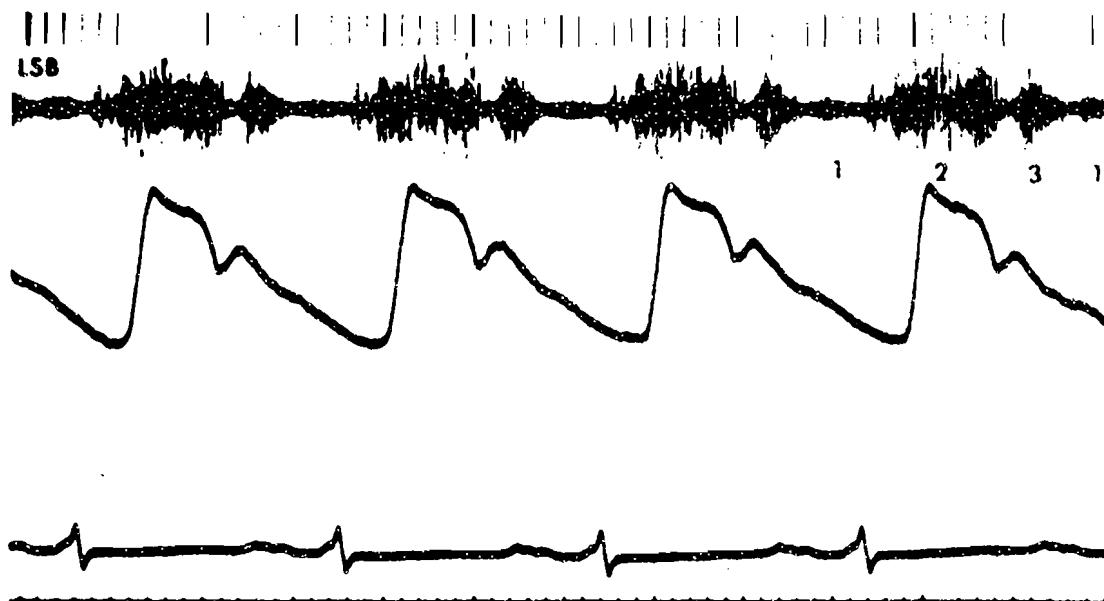


FIG. 22-3. Acute pericarditis. Note the three component friction rub characteristic of pericarditis. This is a relatively infrequent finding following acute myocardial infarction. The pericardial rub is usually heard after the first hospital day and may persist for several days. (From Vanden Belt RJ, Ronan JA, Bedynek JL: *Cardiology*. Chicago: Yearbook Medical Publishers, Inc., 1979.)

ticularly transmural ones, are typically larger than inferior wall infarctions, congestive heart failure is a much more common complication of the former than the latter. Combined infarctions, whether new or old, reduce left ventricular performance even more than does a single infarction alone. Therefore, the degree of left ventricular decompensation is a function of the extent of myocardial damage incurred from past (fibrosis) and present necrosis; greater than 25% of left ventricular myocardial damage results in left ventricular failure.

Increased left atrial pressure, common in heart failure, is a direct consequence of the increase in left ventricular end-diastolic pressure and/or a reduction in left ventricular compliance. In a minority of patients with acute myocardial infarction, heart failure is related to mechanical complications, such as dysfunction or rupture of a papillary muscle or, more rarely, rupture of the interventricular septum. When heart failure develops in a patient with an inferior wall infarction, the astute clinician should always consider the possibility of coexistent mitral regurgitation due to the involvement of the posteromedial papillary muscle, an acute ventricular septal rupture, or an overlooked, remote anterior wall myocardial infarction. If heart failure is predominantly right-sided, the possibility of a coexistent right ventricular infarction should also be entertained. In general, the full clinical picture of biventricular failure does not occur within the first twenty-four hours of an acute myocardial infarction, unless the patient has had previous myocardial

damage. Clues to congestive heart failure include pulsus alternans, the presence of an S3 gallop, basilar rales, or overt pulmonary edema.

Pulsus Alternans. One of the earliest yet frequently overlooked signs of myocardial decompensation is the presence of pulsus alternans (Fig. 3-4). Pulsus alternans is characterized by an alternating strong and weak peripheral arterial pulse in the setting of a regular rhythm, which is unrelated to respiration. The mechanisms responsible for its production remain controversial, but it is generally accepted that the varying pulse strength reflects beat by beat changes in stroke volume, left ventricular myocardial contractility, and/or end-diastolic volume or fiber lengths. Pulsus alternans is best appreciated by gentle palpation of a more distal peripheral artery, such as the radial or femoral, which has a slightly wider pulse pressure than a more central artery, e.g., the carotid. The clinician attempting to elicit pulsus alternans should apply light pressure with the index and middle fingers of the palpating hand to the radial or femoral artery, while paying careful attention to the character of the pulse; the patient's respiration should be held in order to avoid the small changes in arterial pulse that occur from respiration alone.

Pulsus alternans may also be documented by taking the blood pressure with a sphygmomanometer. One may be able to discern a detectable difference in peak systolic blood pressure as the blood pressure cuff is slowly deflated, to the point that every other beat is heard, or an alteration in the character of the Korotkoff sounds is evident (see Chapter 2). The difference in mmHg between the alternation in pressure levels allows one to quantitate the degree of alternans present. Alternation of the force of myocardial contraction also may be associated with an alternation in the intensity of heart sounds and heart murmurs (referred to as auscultatory or auditory alternans). In some patients, alternans is apparent only in the first few beats following a premature ventricular contraction, referred to as postextrasystolic or latent pulsus alternans. Persistent or sustained pulsus alternans is present in association with severe myocardial decompensation. In rare instances, the weak pulse is too small to feel, resulting in total alternans. Pulsus alternans usually is associated with a ventricular filling sound or S3.

Third Heart Sound (see Chapter 7). The ventricular diastolic gallop, a low frequency sound (thud) in diastole, occurs 0.14 to 0.16 seconds after S2 during the early rapid filling phase of the ventricle (Fig. 7-3). In the setting of an acute infarction, the presence of an S3 gallop indicates an elevation of left ventricular filling pressure in greater than 90% of cases. When the S3 gallop is loud and persistent, the prognosis is worse and the mortality rate is higher.

Chest Examination (rales, pleural effusion, acute pulmonary edema). Examination of the lungs may indirectly reveal evidence of a high left atrial pressure. Rales are the classic auscultatory sign of left heart failure. They result from the transudation of fluid into the pulmonary interstitial or alveolar

spaces secondary to elevation of pulmonary venous and capillary pressures resulting from left ventricular dysfunction. In the presence of mild to moderate left heart failure, the rales are fine and crepitant in character and are confined to the lung bases. With more progressive left ventricular dysfunction, they become audible throughout the lung fields. In acute pulmonary edema, rales are coarse and moist and may be extensive, extending up to both apices of the lungs, often associated with wheezes and rhonchi scattered throughout. Rales of cardiac origin are postural in nature, migrating to the most dependent portion of the lungs with changes in body position. Congestive rales do not usually disappear following a few deep breaths or coughs. This contrasts with rales of pulmonary origin, which are independent of posture and frequently clear with coughing. The presence of rales, common in congestive heart failure, indicates a poor long-term prognosis.

Pleural effusions are common in chronic congestive heart failure, occurring more often on the right side than on the left. When pleural fluid accumulates, examination of the lung bases while the patient is in the upright position reveals the presence of dullness to percussion, with diminished or absent breath sounds on auscultation and decreased tactile fremitus.

Acute pulmonary edema may be found in severe, ischemic left ventricular dysfunction; it often implies massive myocardial necrosis or a profound mechanical defect and can present with a most dramatic and terrifying picture. The patient suddenly develops extreme shortness of breath, becoming agitated, anxious, and frightened; he or she sits bolt upright in bed, unable to talk due to overwhelming respiratory distress and intense air hunger. The respirations are rapid and shallow, and the pulse is often rapid and thready. The blood pressure is elevated in the absence of shock, and the skin is cold, clammy, pale, diaphoretic and peripherally cyanotic, reflecting the presence of heightened sympathetic discharge and associated vasoconstriction. The alae nasi are dilated, and there is inspiratory retraction of the intercostal spaces and the supraclavicular fossa, with extensive use of the accessory muscles of respiration. Moist rales are present in both lung fields extending up to the apices, with diffusely scattered wheezes and rhonchi. When extreme, the patient may be coughing up pink, frothy sputum. Cardiac auscultation in this setting is often hampered by the respiratory noise present. Often, an S3 gallop, pulsus alternans, alternation of heart sounds and murmurs, and an accentuated pulmonic component of the second heart sound may be recognized. Inadequate systemic blood flow and a diminished stroke volume may be manifest at the bedside by the presence of a small amplitude hypokinetic arterial pulse on palpation, along with a diminished pulse pressure.

Cheyne-Stokes Respirations. Some patients with left ventricular failure, especially older persons with cerebrovascular disease, will manifest a cyclic, irregular breathing pattern characterized by noisy hyperpnea alternating with silent apnea, referred to as Cheyne-Stokes respiration. Cheyne-Stokes breath-

ing, although relatively common in the setting of left heart failure, is often overlooked on clinical examination. It is commonly observed during sleep and sedation and in the presence of reduced cerebral blood flow due to poor cardiac output. Fluctuations in systemic oxygen saturation and carbon dioxide concentration in the blood alternately stimulate and depress the respiratory center, resulting in cyclic increases and decreases in the rate and depth of respiration. During the period of apnea, the oxygen saturation reaches its lowest level and the carbon dioxide tension its highest, causing the cycle to be repeated again.

Killip Classification

Killip and Kimball have described four subgroups of patients with acute myocardial infarction, categorized according to their hemodynamic status and the degree of left ventricular decompensation and pulmonary congestion as detected at the bedside. Class I represents an uncomplicated infarction, without the presence of an S3 gallop or pulmonary rales. Class II patients have mild to moderate congestive heart failure, manifest by an S3 gallop and bibasilar rales that persist after coughing. Pulmonary edema, with rales extending over one half of the lung fields bilaterally is classified as Class III. Severe pulmonary congestion associated with hypotension and signs of reduced peripheral perfusion represents cardiogenic shock or Class IV. These four general subgroups of patients have different prognoses, with mortality rates ranging between very low for Class I to very high for Class IV subjects.

Right Ventricular Decompensation

In a patient with an acute myocardial infarction, right ventricular decompensation may occur as a consequence of left heart failure, associated pulmonary hypertension, or from selective damage to the right ventricle in the setting of a right ventricular infarction.

Venous Pulse. Analysis of the jugular venous pulse will provide the clinician with much helpful information about right-sided cardiac physiology. The venous pulse reflects the relationship between systemic venous tone, venous blood volume, and right heart hemodynamics (see Chapter 4). Normal venous pressure should be no more than 2 to 3 cm above the sternal angle; i.e., no more than 7 to 8 cm of water. In right heart failure, the jugular venous pressure is usually elevated more than 2 to 3 cm above the angle of Louis. Elevation of right ventricular diastolic pressure results in increased right atrial pressure, salt and water retention, and increased adrenergic tone; all contribute to an elevation in jugular venous pressure. When the venous pressure is very high, A and V wave pulsations may not be seen until the patient is sitting upright. Therefore, the examiner should always carefully

inspect the upper portion of the neck, beneath the angle of the jaw, in order not to miss an elevated venous pulsation.

In patients suspected of having latent right ventricular dysfunction, but who have a normal resting jugular venous pressure, the hepatojugular reflux test is useful (see Chapter 3). This test is performed by applying sustained, firm pressure with the palm of the outstretched hand to the right upper quadrant of the abdomen for 30 to 60 seconds. It is important that the patient is comfortable, trying to maintain a normal rate and depth of respiration, and is not performing an inadvertent Valsalva maneuver. A compromised right ventricle will not be able to handle the increase in blood volume resulting from the enhanced venous return to the right side of the heart caused by this maneuver without causing a rise in its filling pressure, which, in turn, is transmitted to the neck veins. A positive test consists of an elevation of venous pressure greater than 1 cm that persists during the entire time of manual abdominal compression. Kussmaul's sign is another early, subtle sign of right ventricular decompensation. It is characterized by an inspiratory increase in venous pressure due to the decreased compliance and/or capacity of the right ventricle.

Large or giant A venous waves may be seen when right ventricular compliance is diminished and end diastolic pressure in the right ventricle is elevated.

If the tricuspid valve is incompetent, as occurs in tricuspid regurgitation secondary to right ventricular papillary muscle dysfunction, (e.g., right ventricular infarction) blood is transmitted back to the neck veins in systole as a large, pathologic C-V wave (Fig. 19-3). In mild tricuspid regurgitation, the V wave may be normal during the resting state, but deep inspiration (Kussmaul's sign) or abdominal compression (hepatojugular reflux) may bring out a large C-V wave. When severe tricuspid regurgitation occurs, the large C-V waves may be visible from a distance, causing the earlobes to be displaced laterally by the large undulant venous pulsation (Fig. 19-7). In severe tricuspid regurgitation, the venous waves, if forceful enough, may be transmitted, resulting in palpable pulsations of the liver during systole (Fig. 19-5). Although right ventricular failure most often is caused by left ventricular failure with associated pulmonary hypertension, it should be emphasized that there can be a profound disturbance in left ventricular function without any abnormality of the neck veins whatsoever.

S2. An accentuated P2 and wide splitting which remains physiologic are common in RV failure.

S3 and S4. In right-sided heart failure, right-sided S4 and S3 gallops are often heard (and felt) along the lower left sternal border, increasing in intensity with inspiration as the resulting drop in intrathoracic pressure enhances right ventricular filling. A right-sided atrial S4 gallop sound is not unusual in the setting of pulmonary hypertension and diminished right ventricular compliance.

Tricuspid Regurgitation Murmur. A systolic murmur of "functional" tricuspid regurgitation correlates with the large C-V waves in the neck veins and may be audible in the presence of right ventricular decompensation resulting from right ventricular papillary muscle dysfunction. The murmur of tricuspid insufficiency is usually holosystolic, high-pitched, although sometimes harsh, and is best heard along the left sternal border, increasing in intensity with inspiration (Fig. 19-4). It is best elicited by having the patient take slow deep and continuous respirations. In eliciting the effects of respiration on tricuspid murmurs, meticulous attention should be paid to the patient's manner of breathing. The patient should be instructed to breathe slowly, quietly, and deeply, and the physician should concentrate on the peak of inspiration and the nadir of expiration. If needed, the physician can set the tempo of respiration by instructing the patient to breathe in and out in unison with the examiner's upward and downward movement of his free hand (see also Chapter 19).

Cardiogenic Shock

Cardiogenic shock is the most extreme and ominous form of left ventricular pump failure and is now the major cause of death in patients hospitalized with acute myocardial infarction. It complicates approximately 10 to 15% of myocardial infarctions and carries an exceptionally high in-hospital mortality. The fundamental physiologic defect is severe widespread depression of myocardial contractility due to loss of functioning heart muscle. Post-mortem studies demonstrate irreversible necrosis of at least 40% of the left ventricular myocardium sustained by either the present acute myocardial infarction or the cumulative loss of myocardium resulting from past and present necrosis. Many of the patients develop the syndrome early after myocardial infarction, usually within the first two days. It may, however, occur immediately or be delayed one week or more as a consequence of infarct extension or the development of mechanical complications, e.g., ruptured papillary muscle, ruptured ventricular septum, dyskinesis, or aneurysm formation of the left ventricle, which can exacerbate the depression of cardiac function resulting from the infarction. The incidence of cardiogenic shock is higher in patients with anterior wall infarcts than in those with inferior wall myocardial infarction, since the extent of myocardial damage and derangement of pump function is greater due to occlusive disease of the left anterior descending coronary artery involving a large quantity of left ventricular myocardium in patients with anterior wall infarcts. The physical findings of cardiogenic shock that can be detected at the bedside relate to the drastic reduction in regional blood flow, leading to progressive deterioration of tissue perfusion and organ function as a direct result of myocardial dysfunction, i.e., inadequate cardiac output.

The general appearance of the patient is characterized by weakness, apathy, confusion, agitation, or restlessness at first and progresses to obtundation, stupor, and unconsciousness, reflecting the disturbance in mentation resulting from cerebral hypoperfusion.

The skin may be pale, diaphoretic, cold, and clammy, with evidence of peripheral cyanosis due to compensatory reflex, sympathetic vasoconstriction. Skin pallor is an early manifestation of peripheral vasoconstriction and diminished cutaneous perfusion due to the shunting of a part of the cutaneous circulatory volume into the central circulation of the vital organs. Diaphoresis often is an early sign of cardiogenic shock. Beads of sweat appear on the forehead and below the lower lip at first, subsequently progressing to generalized profuse perspiration. The skin temperature diminishes accordingly as a result of the reduction in cutaneous flow, and the associated diaphoresis that is present. Cyanosis begins distally in the extremities and progresses centrally as shock worsens. The skin appears mottled due to the uneven distribution of the reduction in skin flow. Palpation of the arterial pulse reveals it to be weak, small in volume, hypokinetic, rapid, and thready, reflecting the reduction in cardiac output and stroke volume, and the heightened sympathetic drive leading to intense peripheral vasoconstriction. Hypotension, i.e., a systolic arterial blood pressure of less than 90 mmHg, or a drop of more than 60 mmHg in previously hypertensive patients is characteristic. The decreased cardiac output and systemic arterial blood pressure that results from impairment of myocardial pumping function, diminishes coronary perfusion even further, thereby compounding myocardial ischemia and dysfunction and creating a vicious cycle. The pulse pressure may narrow and the blood pressure may even become inaudible as shock deepens. Oliguria, i.e., a marked diminution in urinary output (less than 30 cc of urine per hour), may result due to the decrease in renal perfusion.

All of the physical signs of severe left ventricular failure previously discussed, as well as any or all of the other bedside findings of an acute myocardial infarction and its mechanical complications, such as dysfunction or rupture of the papillary muscles or ventricular septum, may be present.

Mechanical Complications. Although transient papillary muscle dysfunction is relatively common in acute myocardial infarction, rupture of a papillary muscle is a rare, life-threatening event. An acute VSD, typically occurring in anteroseptal or inferoseptal infarcts, can also be a dramatic hemodynamic event. Finally, a rupture or tear of the LV free wall, usually results in rapid demise without suggestive physical findings. This is classically associated with electromechanical dissociation immediately prior to death. The physical findings of papillary muscle dysfunction or rupture and VSD are discussed in Chapter 17 and those of a ventricular septal defect in Chapter 21. The presence of coexisting severe LV dysfunction may decrease both the loudness and length of the classic systolic murmurs.

Chapter 23

Prosthetic Cardiac Valves

Neale D. Smith, M.D., Veena Raizada, M.D., and
Jonathan Abrams, M.D.*

Surgery for valvular heart disease has been available for twenty-five years, and many thousands of individuals are alive today with functioning prosthetic heart valves. A wide variety of cardiac prosthetic devices have been utilized since the initial efforts at valve implantation in the 1950s and the first successful direct valve replacement in 1960, simultaneously undertaken by Starr and Harken. The types of valves used vary considerably. Prosthetic valves are composed of differing materials, including composite metals, pyrolytic carbon, silastic rubber, as well as biologic tissues from animals or humans. Over 50 different valves have been created over the last several decades, and at least 15 to 20 have been employed widely in one center or another. This chapter will not discuss in any detail the wide array of valves that are currently available but will focus on the auscultatory phenomenon associated with the commonly implanted valves.

It is important for the clinician to have a clear understanding of the four major types of valves that are currently in use: ball valves, tilting disc valves, the bivalve prosthesis, and the porcine bioprostheses (Fig. 23-1).

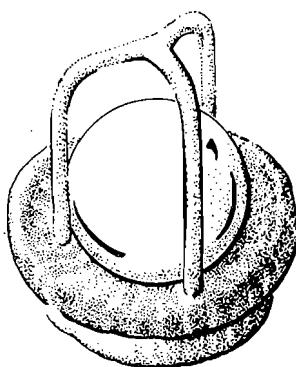
Valve Sounds. Prosthetic valves are associated with distinct auscultatory events produced by component motion and/or alterations in blood flow. The intensity, timing, and character of valve sounds differ among the various types of prostheses and may also be affected by the patient's rhythm and hemodynamic status. Auscultation can provide valuable information about prosthetic valve malfunction, such as thrombosis, incompetence, or obstruction. Thus, physicians should be familiar with the normal acoustic phenomena associated with these devices. Most published reports of prosthetic valve sounds emphasize careful phonocardiographic recordings. These are not always equivalent to clinical auscultation, are often unavailable to the clinician, and require special interpretive skills.

* This material has been modified with permission from Smith ND, Raizada V, and Abrams J: Auscultation of the normally functioning prosthetic valve. Ann Intern Med 95:594-598, 1981.

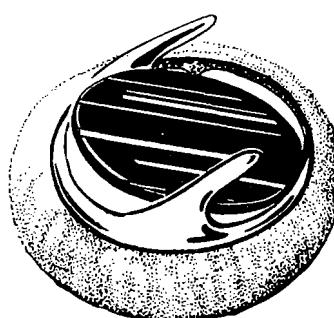
Factors Affecting the Physical Findings in Patients with Implanted Cardiac Valves. Prosthetic valves produce alterations in the normal laminar pattern of blood flow. All valves are inherently obstructive and have a significantly smaller orifice area than the normal native valves. The average effective orifice area of prosthetic valves ranges between 1.5 and 2.5 cm², considerably less than that of the normal anatomic valve area. Thus, a pressure gradient of variable magnitude exists across virtually all prosthetic valves at rest, which typically becomes larger during exercise (Table 23-1). Smaller sizes of prosthetic valves are more likely to be obstructive than larger sized devices. Aortic gradients are larger than those across the prosthetic mitral valve.

Prosthetic valves are implanted in individuals with underlying derangements of the cardiovascular system who often demonstrate significant residual abnormalities of cardiac function, such as pulmonary hypertension, right or

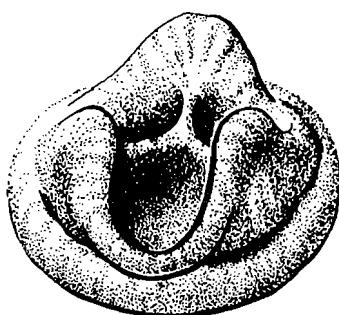
Prosthetic Valve Types



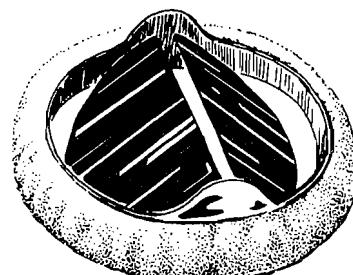
A Ball



B Disc



C Porcine



D Bivalve

FIG. 23-1. Examples of common prosthetic valves. A. Ball and cage valve. B. Tilting disc valve. C. Porcine bioprosthetic. D. St. Jude bivalve prosthesis. (From Smith MD, Raizada V, and Abrams JA: Auscultation of the normally functioning prosthetic valve. Ann Intern Med 96:594, 1981.)

TABLE 23-1 Approximate Basal Pressure Gradients Across Normally Functioning Prosthetic Valves

Prosthetic Valve	Aortic	Mitral
I. <i>Caged Ball</i> Starr-Edwards	10-25 mmHg	4-9 mmHg
II. <i>Tilting Disc</i> Bjork-Shiley, Lillehei-Kaster	5-20 mmHg	4-8 mmHg
III. <i>Porcine Heterograft</i> Hancock, Carpentier-Edwards	8-20 mmHg*	3-7 mmHg
IV. <i>Bivalve</i> St. Jude	5 mmHg*	5 mmHg

*Gradients can be larger with the smaller aortic valve sizes. These are average mean gradients taken from the literature. The range among individual patients is much greater, with some subjects having virtually no gradient and others having very large transvalvular pressure differences. The prosthetic valve size is important; smaller prostheses are more likely to be obstructive, particularly at the aortic position. The pressure gradient across a normal prosthetic valve will increase with exercise.

left ventricular hypertrophy, and myocardial dysfunction. Concomitant abnormalities of cardiac rate and rhythm are often present. Loud and prominent prosthetic sounds may camouflage or even mask normal acoustic events.

Unfortunately, significant prosthetic valvular dysfunction may occur without audible changes in the valve opening or closing sounds or without pathognomonic murmurs. Nonetheless, careful auscultation and familiarity with the characteristics of the normal prosthesis can help the physician make a useful clinical assessment of prosthetic function in these complex patients. An understanding of the typical acoustic findings associated with normally functioning prosthetic valves should provide a background for the clinician in evaluating suspected abnormalities of prosthetic valve function. Careful serial observations of the auscultatory findings is most important in the long-term follow-up of the patient who has undergone valve replacement.

ACOUSTIC CHARACTERISTICS OF THE COMMON PROSTHETIC VALVES

The four major prosthetic valve types are depicted in Figure 23-1. The sounds and murmurs produced by these valves are diagrammed in Figure 23-2.

Ball Valves (Fig. 23-1A)

Ball valves (of which the prototype is the familiar Starr-Edwards valve) have easily audible *opening* and *closing* sounds, which coincide with the maximum excursion of the ball and its subsequent seating, respectively (Fig.

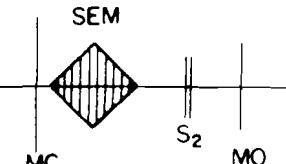
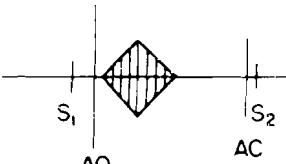
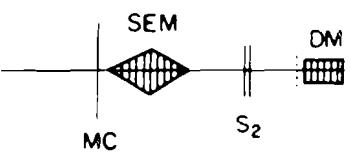
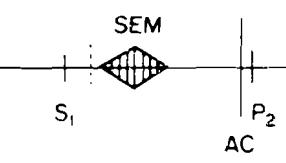
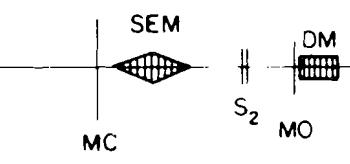
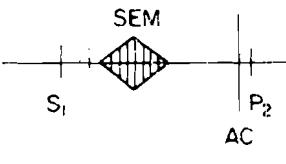
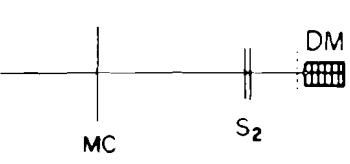
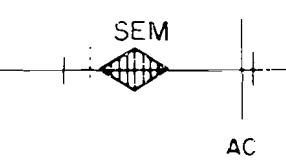
Prosthesis type	Mitral Prosthesis	Acoustic Characteristics	Aortic Prosthesis	Acoustic Characteristics
1) Ball Valves		1) A ₂ -MO interval 0.07-0.11 sec. 2) MO > MC 3) II-III / VI Systolic ejection murmur (SEM) 4) No diastolic murmur		1) S ₁ -AO interval 0.07 sec 2) AO > AC 3) II / VI harsh SEM 4) No diastolic murmur
2) Disc Valves		1) A ₂ -MO interval 0.05-0.09 sec. 2) MO is rarely heard 3) II / VI SEM is usually heard 4) I-II / VI diastolic rumble is usually heard		1) S ₁ -AO interval 0.04 sec. 2) AO is uncommonly heard, AC is usually heard 3) II / VI SEM is usually heard 4) Occasional diastolic murmur
3) Porcine Valves		1) A ₂ -MO interval 0.1 sec. 2) MO is audible 50% 3) I-II / VI apical SEM 50% 4) Diastolic rumble 1/2 - 2/3		1) S ₁ -AO interval 0.03-0.08 sec. 2) AO is uncommonly heard, AC is usually heard 3) II / VI SEM in most 4) No diastolic murmur
4) Bileaflet Valve (St. Jude)		1) MC well heard 2) MO usually not audible 3) Diastolic rumbling murmur may be heard		1) AO not well heard readily recorded 2) AC prominent 3) Soft SEM on occasion 4) No diastolic murmur

FIG. 23-2. Auscultatory findings of common prosthetic valves. MO = mitral opening; MC = mitral closure; SEM = systolic ejection murmur; DM = diastolic murmur; AO = aortic opening; AC = aortic closure. (Modified from Smith MD, Raizada V, and Abrams JA: Auscultation of the normally functioning prosthetic valve. Ann Intern Med 96:594, 1981.)

23-3A, B). These are high pitched, metallic, prominent, and easily distinguishable from normal cardiac sounds. These sounds can be quite loud, depending on the prosthetic model and the composition of the ball.

Mitral Ball Valve. A very prominent opening click (MO) of the mitral ball prosthesis follows A2 by 0.07 to 0.13 seconds (Figs. 23-2, 23-3A). The interval between A2 and the opening sound (A2-MO) of the prosthetic valve is similar in duration to the A2-opening snap interval of moderate mitral stenosis. MO is best appreciated between the cardiac apex and the lower sternal edge. The mitral valve opening transient usually is louder than the mitral closure sound (MC), which coincides with and typically obscures S1. MC is heard best at the lower left sternal border.

The interval from A2 to MO differs only minimally from beat to beat, even during atrial fibrillation, but a long preceding R-R interval may decrease the intensity of the prosthetic opening sound. Shortening of the A2-MO interval to 0.06 second or less suggests prosthetic obstruction (Fig. 23-4A), severe mitral regurgitation (Fig. 23-4B), or left ventricular dysfunction, whereas prolongation of A2-MO to greater than 0.17 second or readily detectable beat-to-beat variation of this interval suggests abnormal interference with poppet excursion. A decrease of the A2-MO interval during long-term follow-up suggests an elevated left atrial pressure due to valve dysfunction, usually progressive obstruction.

Mitral closure may diminish in intensity in first degree heart block or following long R-R intervals when the poppet has already reached its closed position before the onset of LV isovolumic systole. This is a similar phenomenon to the relationship between the intensity of S1 and the preceding PR interval in subjects without mitral valve disease (Chapter 6, Fig. 6-3). MC may be less loud in the presence of a high left ventricular filling pressure. A single prosthetic MC sound without an opening sound may be heard when the poppet fails to open after a premature beat; the poppet subsequently may drift silently open during the next diastole (absent MO) and close forcibly and audibly with the next ventricular contraction (MC).

The second heart sound (S2) generally is unchanged in subjects with a mitral ball prosthesis that is functioning normally, although P2 may be increased if pulmonary hypertension is present. A sound that may be recorded (but not often heard) 0.04 to 0.13 second after MO that coincides with a rapid filling wave probably represents a ventricular gallop or S3 and is not of prosthetic valve origin. Diastolic sounds in presystole have been recorded with Starr-Edwards mitral valves following the P wave of the EKG. These sounds probably relate to atrial systole and subsequent motion of the ball against the cage.

An apical early to midsystolic murmur commonly is heard at the lower left sternal border in patients with a mitral Starr-Edwards prosthesis. This systolic murmur does not represent mitral regurgitation and is probably

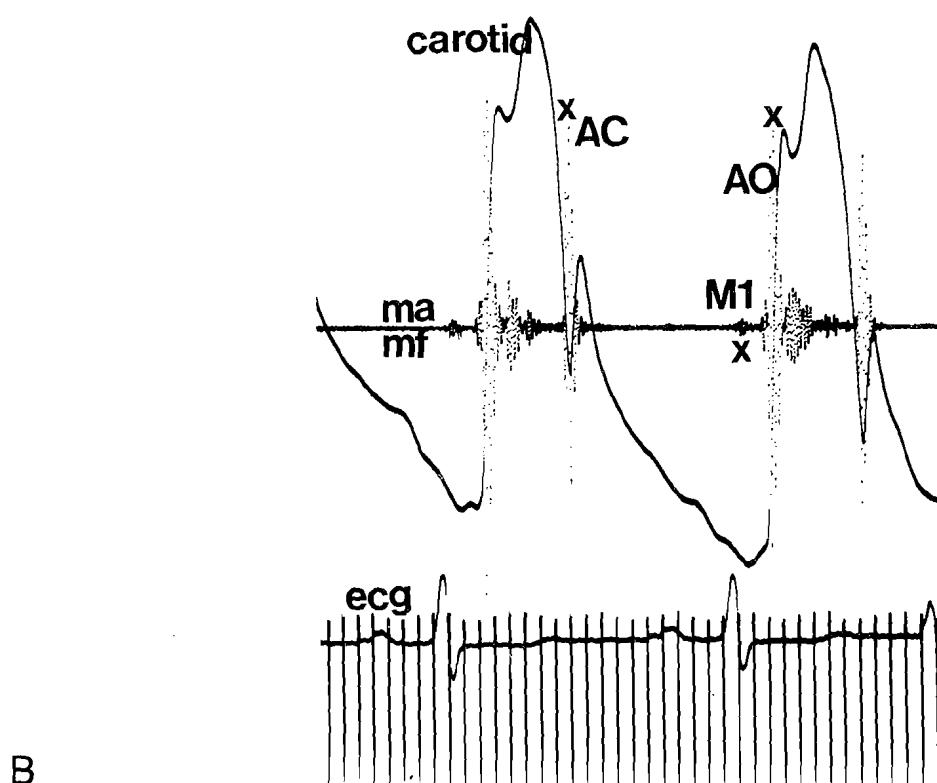
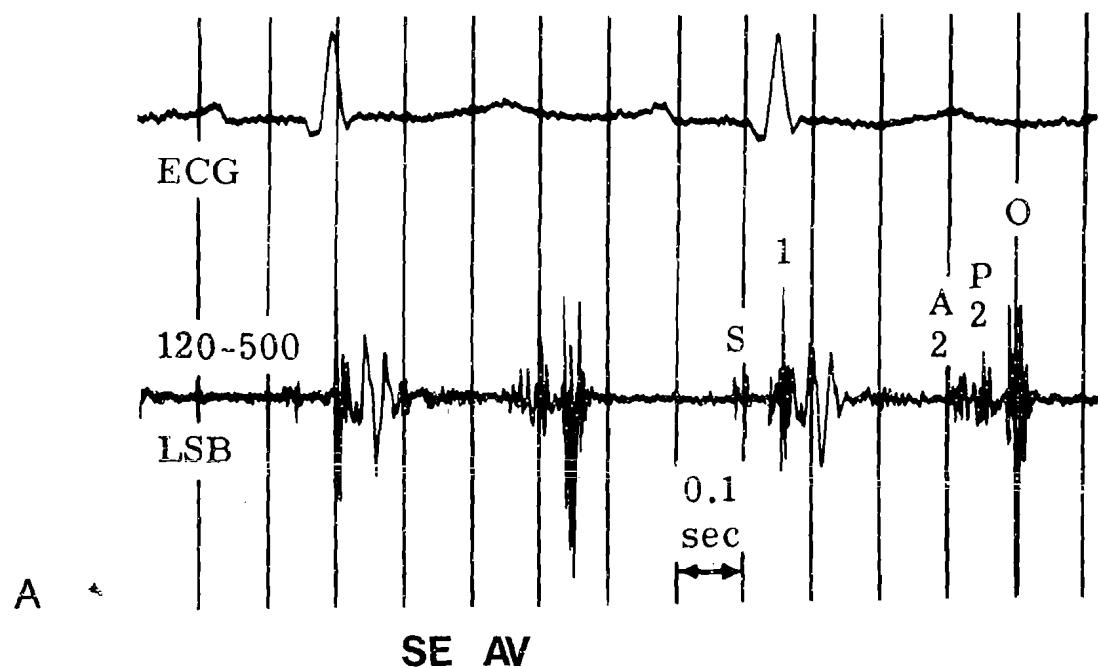


FIG. 23-3. Auscultatory findings with Starr-Edwards ball and cage valves. Prosthetic valve sounds from ball and cage valves are loud, high pitched, and metallic. The ball often oscillates during the cardiac cycle, resulting in multiple prosthetic clicks. A. Mitral position. Closing sound (1) and opening sound (O). There is an additional late diastolic click (S) resulting from movement of the ball in the cage following atrial systole. (From Tavel ME: Clinical phonocardiography and external pulse recording, 3rd ed. Chicago, Year Book Medical Publishers, 1978.). B. Aortic position. Note the systolic ejection murmur and the very prominent aortic prosthesis closing (AC) and opening sound (AO), identified by the symbol X. (From Smith MD, Raizada V, and Abrams JA: Auscultation of the normally functioning prosthetic valve. Ann Intern Med 96:594, 1981.)

caused by turbulence produced by projection of the rigid prosthesis cage into the left ventricular outflow tract. *Practical Point:* The presence of an apical mid-diastolic murmur in a patient with a ball valve prosthesis is abnormal and suggests prosthetic valve dysfunction or obstruction. Some have reported a mitral flow fumble in the absence of valve obstruction, but this is quite unusual.

Aortic Ball Valve. The aortic ball valve produces loud opening click (AO) that is distinct from the first component of S1 and separated from it by an average of 0.07 second (Fig. 23-3B). Poppet closure (AC) is prominent but can be less loud than AO. The normal amplitude ratio of AO to AC as measured by phonocardiography is greater than 0.5; a clinically useful sign of ball valve dysfunction is a reduction of this ratio with a decreased intensity or even absence of the AO sound. Preservation of the relationship of AC to AO, however, does not exclude serious valve malfunction.

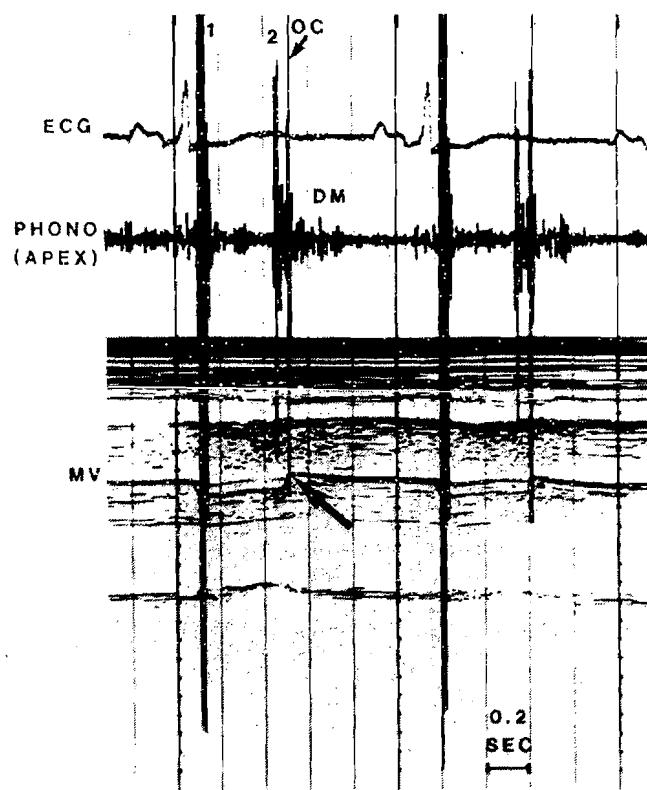
When left ventricular pressure exceeds aortic pressure, the ball rapidly moves to the apex of the cage. It may remain there or assume a midcage position for the duration of systole, where it may produce multiple systolic clicks related to oscillations of the poppet within the cage. An early ventricular ectopic beat may not open the aortic valve, whereas a late ventricular ectopic beat can be associated with both an AO and AC sound. A low cardiac output or severe left ventricular dysfunction may result in a reduction of the prosthetic sound intensity, whereas tachycardia or anemia tend to increase the intensity of the prosthetic valve sounds.

AO is best appreciated at the apex or lower left sternal border and usually transmits widely. The somewhat less prominent AC also radiates widely. It precedes P2, and the normal inspiratory relationship of these two sounds is usually maintained; P2 may be masked by a loud AC.

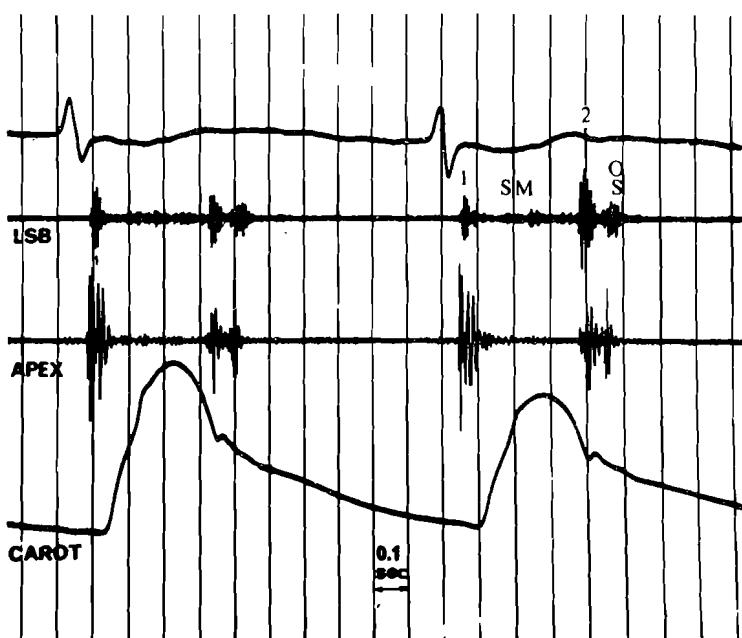
A prominent grade 2 to 3/6 early decrescendo or systolic ejection murmur usually is present, typically with radiation into the carotids where a shudder may be palpated (Fig. 23-3B). This systolic murmur can be loud and harsh and may increase in intensity when the stroke volume is augmented, such as during anxiety, tachycardia, or anemia. The murmur is caused by blood flow turbulence and a "normal" aortic transvalvular gradient. *Practical Point:* A diastolic murmur is not normally present in the setting of an aortic ball valve prosthesis and, when heard, suggests prosthetic valve dysfunction or a perivalvular leak.

Tilting Disc Valves (Fig. 23-1B)

The commonly used tilting disc valves (Lillehei-Kaster or Omniscience valve, Bjork-Shirley) do not typically produce audible opening sounds, although an aortic or mitral OC is recordable on phonocardiography. The relatively lightweight discs do not strike any resonant structure as they swing open; therefore, a prominent opening sound is not heard. However, a distinct



A



B

FIG. 23-4. Dysfunction of Starr-Edwards prosthetic mitral valves. A. Marked shortening of the S2-opening click (OC) interval. Abnormally high left atrial pressure results in early or premature opening of the mitral valve. In this patient, an excessively small prosthesis was implanted three years earlier, resulting in prosthetic valve obstruction. In addition to the short 2-OC interval, a prominent diastolic rumble (DM) is present. The simultaneous echocardiogram shows the OC coinciding with cessation of the opening movement of the ball (arrow). MV = mitral valve echogram. (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 26:145, 1983.) B. Early prosthetic mitral opening sound

(Legend Continues on Facing Page)

closure sound is always heard, and absence of a closing sound is always abnormal. Disc valve sounds tend to be clicking, distinct, and high frequency in nature. They are much less prominent than the sounds produced by a ball valve.

Mitral Disc Valve. Although phonocardiograms consistently record a prosthetic opening sound (MO) at the apex or lower left sternal border in patients with tilting disc valves (Fig. 23-5A), an MO sound is usually not well heard on auscultation. Because this sound is associated with the onset of disc motion, the interval between A2 and the opening of the disc mitral valve is short (0.05 to 0.09 second). A prominent mitral closure sound (MC) is present at the apex. With left ventricular dysfunction or first degree heart block, the disc may seat early, resulting in a softer or even absent MC. The closure sound may also decrease in intensity or may be absent if disc movement is hindered by fibrosis or thrombosis.

A grade 2/6 early to midsystolic low frequency ejection murmur usually is heard. In contradistinction to the mitral ball valve, apical diastolic murmurs are common in patients with mitral disc valve prostheses. In one study, 14 of 20 patients with normal valve function had a recordable diastolic murmur. As with other normally functioning prosthetic valves, disc valves typically have an orifice area that is one half to one third the area of a normal *in situ* valve, resulting in a resting left atrial-left ventricular pressure gradient (Table 23-1). The resultant turbulent blood flow across the mitral valve may be responsible for the audible diastolic murmur, which is often rumbling in quality. Nevertheless, the presence of a *prominent* diastolic murmur, especially if new or changing, should initiate a meticulous search for valve-patient mismatch, valve thrombosis, tissue ingrowth, or other mechanical difficulties.

Aortic Disc Valve. In contrast to ball valves, the opening sound of a disc valve in the aortic position is not commonly heard, although a soft AO may be audible and can always be recorded on a phonocardiogram. It follows M1 by 0.04 second (Fig. 23-5B). Disc valves produce distinct, audible closing sounds (AC) that typically have a clicking quality and are louder than S1. Absence or diminution in the intensity of AC indicates inhibition of disc motion by thrombus or tissue ingrowth; it rarely can be caused by poor left ventricular function.

(OS) with soft systolic murmur (SM) recorded at the lower left sternal border (LSB). The 2-OS interval is approximately 0.06 seconds, suggesting elevation of left atrial pressure. This patient had a mitral valve ball and cage prosthesis implanted five years earlier and complained of progressive dyspnea. Angiography revealed massive mitral regurgitation across the prosthesis with marked elevation of the pulmonary wedge pressure. In this case, the short 2-OS interval was not due to prosthetic valve obstruction but to the marked left atrial pressure elevation resulting from severe mitral regurgitation. Note that the systolic murmur (SM) is not prominent. (From Tavel ME: Clinical phonocardiography and external pulse recording, 3rd ed. Chicago, Year Book Medical Publishers, 1978.)

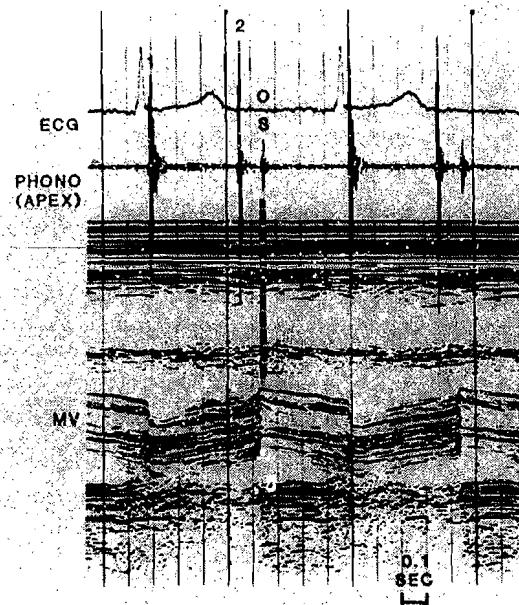
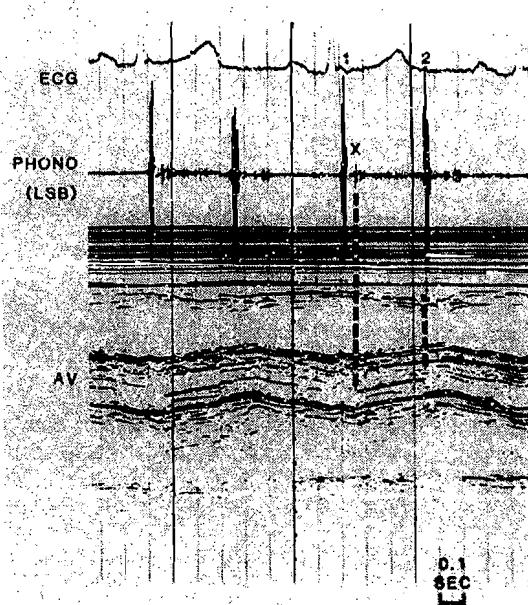
A**B**

FIG. 23-5. Normal Bjork-Shiley tilting disc mitral (A) and aortic (B) valves. A. Note the prominent opening sound (OS) that coincides with the abrupt termination of anterior disc motion of the mitral prosthesis. B. The opening and closing of the aortic prosthesis is indicated by the dashed lines. Valve opening produces a soft opening sound (X). (From Tavel ME: Phonocardiography: Clinical use with and without combined echocardiography. *Prog Cardiovasc Dis* 26:145, 1983.)

An early to midsystolic ejection murmur at the base commonly is heard in patients with an aortic disc prosthesis; smaller sized prostheses result in larger gradients and louder murmurs. Occasionally, a soft aortic diastolic murmur also is noted, which may not necessarily reflect a paravalvular leak. Significant valvular insufficiency may exist with this or other prosthetic devices, even in the absence of an audible murmur.

Tissue Valves (Fig. 23-1C)

The opening sounds of the porcine valve (Hancock, Carpentier-Edwards) originates from an abrupt halting of the opening motion of the prosthetic leaflets. Audible closing sounds of porcine valves occur simultaneously with leaflet coaptation and may be due to deceleration of the blood column and/or contact between the leaflets themselves. Although porcine opening and closing sounds are discrete and relatively high pitched, they are typically

much less prominent and "clicking" than those produced by mechanical devices, and auscultation of these valves does not produce valve sounds very different from normal.

Porcine Mitral Valve. A mitral opening sound transient (MO) can be detected by auscultation in approximately one half of the patients (Fig. 23-2). The interval from A2 to opening of the valve ranges from 0.07 to 0.11 second and, when heard, is best appreciated at the apex. The porcine mitral prosthetic closing sound (MC) is audible in most patients at the lower left sternal border; it is less prominent than sounds produced by metallic valves and tends to blend into the S1 complex. Soft apical mid-diastolic rumbling murmurs are present in one half to two thirds of patients and bear no clear-cut relation to the valve size or hemodynamic state. This murmur is heard best when the subject is turned to the left lateral decubitus position. The murmur is a physiologic variant of mitral stenosis, as these valves normally have a transmural gradient at rest (Table 23-1). Apical midsystolic murmurs can be heard in most patients with porcine mitral valves and are probably due to turbulence produced by the rigid prosthesis extending into the left ventricular outflow tract. A new diastolic murmur, a change in the character of a previously described murmur, or an increase in intensity of the systolic or diastolic murmur may indicate prosthetic dysfunction due to tissue degeneration, infection, thrombosis, or suture disruption.

Porcine Aortic Valve. Aortic closing sounds are audible in most patients with porcine aortic valves, although an opening sound (AO) is not usually heard (Fig. 23-2). The aortic closure sound is prominent, discrete, and best heard at the aortic and pulmonic areas. A high frequency, grade 2/6 early to midsystolic murmur commonly is heard at the left sternal border. An aortic diastolic murmur should not be heard in a normally functioning bioprosthesis in the aortic position.

Bivalve or St. Jude Prosthesis (Fig. 23-1D)

Experience is relatively limited regarding the acoustic characteristics of the bivalve or prosthesis (St. Jude valve) (Fig. 23-2). In general, this valve produces very small transvalvular gradients (Table 23-1).

Aortic Valve. The St. Jude valve in the aortic position does not produce a distinctly audible opening sound, although an opening click in patients with an aortic bivalve prosthesis is easily recorded on phonocardiography. A prominent high-pitched, metallic closing sound coincides with the bivalve leaflet closure and the dicrotic notch of the carotid pulse tracing. An aortic diastolic murmur is not audible, and only an occasional soft early systolic murmur can be detected in patients with an aortic prosthesis.

Mitral Valve. In the mitral position, a distinct opening sound is easily recorded but is usually not audible (Fig. 23-6). As at the aortic position, a

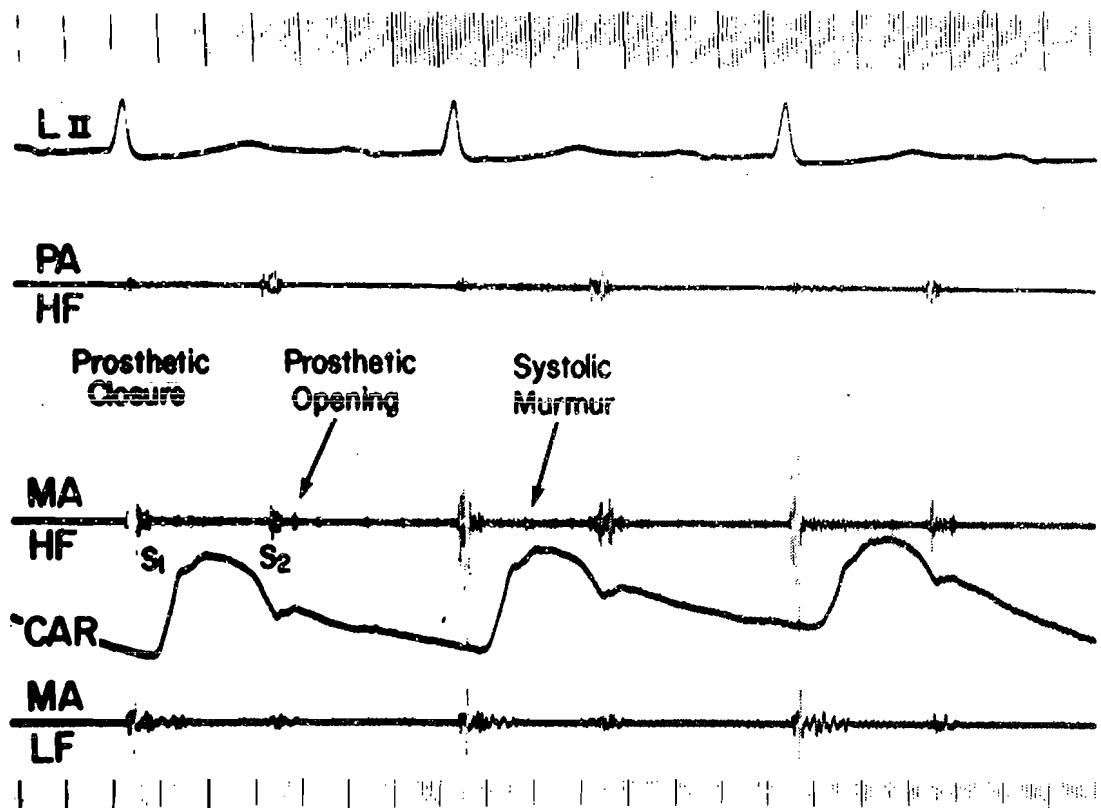


FIG. 23-6. Normal St. Jude mitral prosthesis. Note the soft prosthetic valve opening and closing sounds and the nondescript systolic ejection murmur. PA = pulmonary area; MA = mitral area; HF = high frequency; LF = low frequency; CAR = carotid arterial tracing. (From Feldman HJ, et al: Noninvasive *in vivo* and *in vitro* study of the St. Jude mitral valve prosthesis. Am J Cardiol 49:1101, 1982.)

prominent, high frequency closing click has been consistently described. A mid-diastolic apical rumbling murmur has been reported as a normal finding in some subjects and may not indicate prosthetic valve obstruction, although on auscultation it is more usual not to hear a diastolic murmur. Occasional mid-diastolic valve clicks have been described, presumably resulting from reclosure of the valve leaflets in late diastole.

Prosthetic Valve Dysfunction

Abnormalities of prosthetic valve function include primary failure (degeneration, calcification, and stenosis) of tissue or biologic valves; thrombosis; ingrowth of connective tissue or pannus; paravalvular leaks; frank dehiscence; ball or disc variance; and infective endocarditis. Some of these phenomena may affect acoustic findings, but these are unpredictable, and usually auditory changes are subtle, even for the experienced clinician. The use of M-mode and two-dimensional echocardiography, pulse Doppler echo, and cinefluoroscopy is important in the evaluation of the suspected dysfunctional pros-

TABLE 23-2 Abnormalities on Cardiac Auscultation Suggesting Prosthetic Valve Dysfunction

Absence or clear-cut attenuation of a prosthetic valve opening (OC) or closing click (CC) that had previously been heard on auscultation and/or recorded on a phonocardiogram
Change in the relationship of intensity of the opening to closing click
Starr-Edwards valves: A decrease in the aortic OC:CC ratio to less than 0.5
Tilting disc valves: Decrease in amplitude or disappearance of the closing click
Beat to beat variability in the amplitude of prosthetic valve opening and/or closing sounds or random alterations in the timing relationships of valve sounds
Presence of an aortic regurgitation murmur in the setting of an aortic valve prosthesis (with possible exception of the tilting disc valves)
Presence of a mitral regurgitation murmur in the setting of a mitral valve prosthesis
A loud diastolic "flow" murmur at the apex in the mitral position in patients with the tilting disc and ball and cage valves
A diastolic flow murmur or "mitral rumble" can be a normal finding in many patients (50-70%) with porcine mitral bioprostheses and in subjects with a St. Jude's mitral valve
Development of a long, late peaking systolic "ejection" murmur in the presence of a prosthetic aortic valve
A short A2-MO interval (<0.08 sec) implies mitral valve obstruction or paravalvular regurgitation
A long A2-MO interval may occur when the prosthetic mitral valve ball or disc "sticks" to the valve seat because of thrombus or tissue ingrowth

thetic valve. Recently, spectral phonocardiography has been employed to help differentiate normal from abnormal valve function.

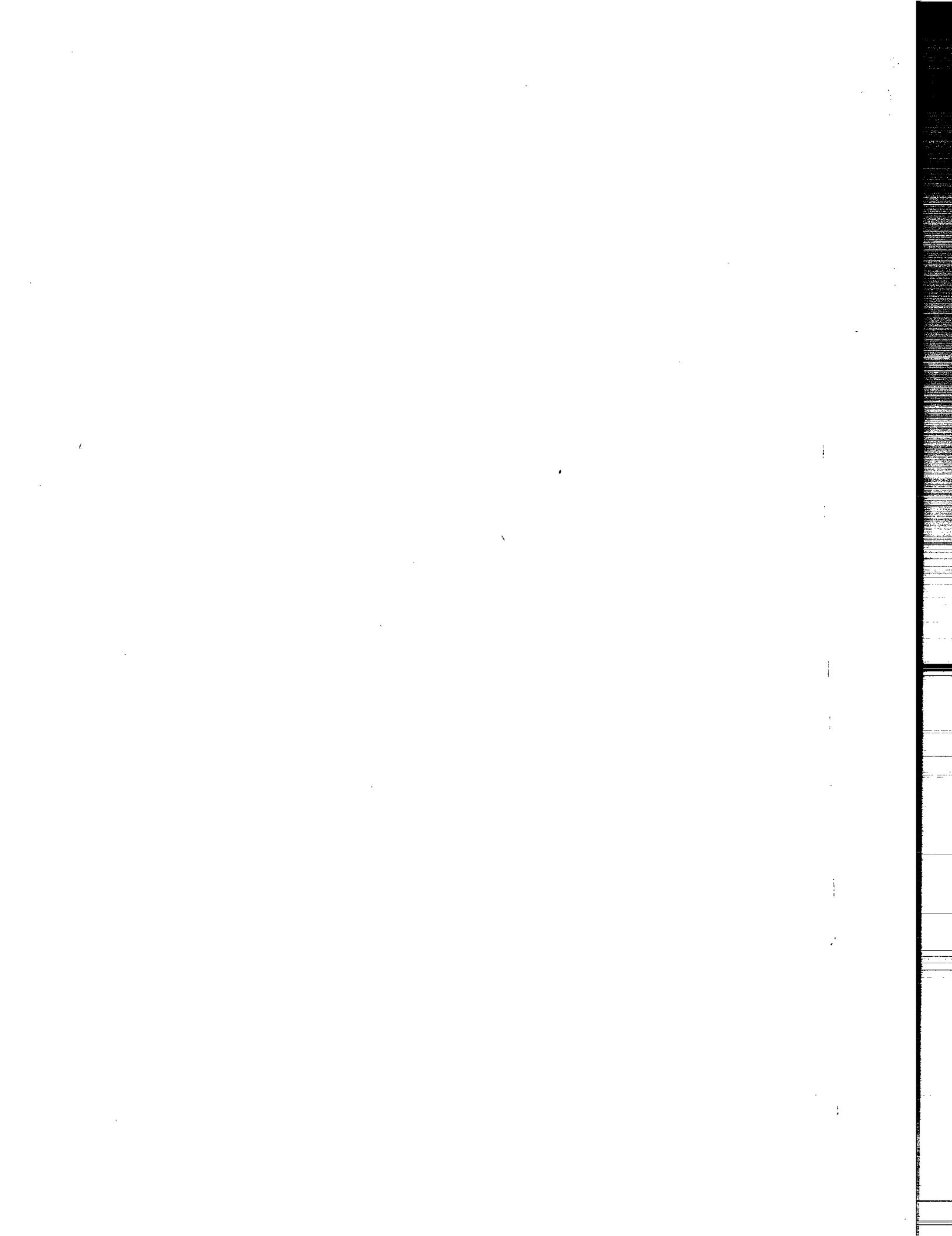
It is very important to establish a careful baseline examination in all patients after valve replacement. Ideally, this should be combined with phonocardiography and echocardiography for documentation of the actual and relative intensities of the opening and closing valve sounds. Serial assessment is extremely valuable. Some experts believe that serial measurements of the A2-mitral valve opening interval, as measured by echophonocardiography, is important to assess in the long-term evaluation of these patients. Shortening of the A2-MO interval to less than 0.08 sec. implies an elevated left atrial pressure that can result from prosthesis obstruction, paravalvular regurgitation, or increased ventricular filling pressure from left ventricular dysfunction (Fig. 23-4).

Table 23-2 lists the most important abnormalities of prosthetic valve function that can be detected on cardiac physical examination.

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