Valvular Heart Disease

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1 Summary

Valvular heart disease is a common abnormality seen in the primary care setting. There are many causes of valvular heart disease including congenital, degenerative, infectious, traumatic, and many more. There is a wide variety of types of valvular heart disease with each valve having the ability to develop both regurgitation and stenosis by multiple mechanisms. All these complexities make diagnosis and management of valvular heart disease complicated, especially in the context of comorbidities. For this reason, it is important for primary care physicians to have a thorough understanding of how these diseases present and when interventions are indicated.

2 Key Points

a Disease staging of VHD is based on symptoms, valve anatomy, the severity of valve dysfunction, and the response of the ventricle and pulmonary circulation.

b A thorough physical exam should be used in all cases of suspected valvular heart disease to determine which valves are affected and whether they are stenotic or regurgitant.

c Transthoracic echocardiography should be used as the initial diagnostic study to evaluate the etiology and severity of valvular heart disease.

d Patients with valvular heart disease should be referred to a cardiologist if the disease is moderate to severe, symptomatic, rapidly progressing, or due to a concerning etiology.

e The decision to pursue intervention for valvular heart disease should be made with a multidisciplinary team approach.

3 Introduction

The aim of this review is to outline the diagnosis and management of commonly occurring valvular heart diseases (VHD) in general clinical practice. Disease staging of VHD is based on symptoms, valve anatomy, the severity of valve dysfunction, and the response of the ventricle and pulmonary circulation. Surveillance programs, management strategies, and recommendations for intervention are based on these differences. It is important for clinicians to have an understanding of pathologic murmurs to guide appropriate referral or the need for echocardiography, which is the gold standard for the diagnosis and grading of severity of VHD. In most cases, surgery or percutaneous intervention is recommended only when symptoms dictate or when ventricular dilation and systolic dysfunction have occurred. The goals of treatment are to maintain adequate hemodynamics (blood pressure and heart rate control), manage existing arrhythmias, and treat or prevent concomitant cardiac disease (coronary heart disease, arrhythmias, and stroke). Surgery or percutaneous intervention should only be performed after being discussed by a multidisciplinary heart team that includes both cardiologists and cardiac surgeons.

4 Pathophysiology

Stenosis

Valvular pathologic conditions lead to either stenosis or regurgitation. Stenotic valves restrict flow, whereas regurgitant valves allow blood to flow back across the closed valve into the preceding chamber. The aortic valve (AV) has no regurgitation in its physiologic state, whereas the other 3 valves have elements of physiologic regurgitation. Valvular pathologic conditions are further categorized into acquired or congenital. In general, the heart can withstand significant amounts of stenosis and regurgitation before clinical symptoms appear. These symptoms are unique to each valvular pathologic condition. The stages of VHD are grouped by severity. The end stage of most VHD is heart failure.

Regurgitation

Regurgitation leads to ventricle volume overload and increased stroke volume resulting in remodeling and increased myocardial work. Unlike the remodeling associated with increased afterload, regurgitation and the associated increased preload cause chamber dilation. These changes lead to a decrease in effective stroke volume. Additionally, regurgitation (but also stenosis) can lead to pulmonary congestion and low forward cardiac output, which can have multisystem implications.

5 Etiologic factors

Stenosis is primarily caused by congenital malformations of the valve, calcification of normal trileaflet valve, or rheumatic disease. Congenital malformations include unicuspid, bicuspid, and quadricuspid valves. A bicuspid AV is the most common congenital cardiac abnormality. Unicuspid valves can present with severe symptoms of stenosis as early as infancy. Bicuspid valves present with symptomatic stenosis later in life and when superimposed calcific changes result in clinically significant valvular obstruction. Calcific VHD disease, or degenerative valve disease, is the most common cause of VHD in adults. It is thought to be caused by proliferative and inflammatory changes that lead to calcium deposition and an associated reduction in the mobility of the valve cusps. Hypercholesterolemia, especially familial types, predispose for VHD and accelerate progression. Rheumatic VHD is caused by adhesions and fusion of the commissures and cusps, which results in reduction of the valve orifice size. With reduction in the incidence of rheumatic fever in developed countries, the incidence of rheumatic heart disease has declined. Rheumatic heart disease remains a problem in developing countries and is the leading cause of VHD worldwide.

Regurgitation is due to dilation of the vessels, and it is more common than primary valvular disease. Numerous conditions lead to regurgitation, including age-related degeneration, cystic medial necrosis (Marfan syndrome), syphilis, spondyloarthropathies, giant cell arteritis, or hypertension. Infective endocarditis is the common cause of regurgitation due to abnormalities of the valve. Regurgitation due to endocarditis can be caused by either valve leaflet perforation or vegetation that prevents leaflet coaptation.

Rheumatic heart disease remains the most common cause of mitral stenosis worldwide. As the orifice area decreases, a higher left atrium (LA) pressure is required to maintain LV filling and cardiac output. As LA pressures increase, there is an increase in pulmonary venous pressure leading to dyspnea. The LV is physiologically normal in MS though it may be small and underfilled. The filling of the LV depends on both the LA pressure and the diastolic filling time. With tachycardia (during exercise or with an atrial arrhythmia), diastolic filling time shortens, leading to underfilling of the LV and worsening symptoms. Other hemodynamic consequences include increased pulmonary arterial pressure and symptoms of right heart failure. With stasis in the LA, the risk of thrombus formation and systemic embolism increases significantly. Pregnancy is an important consideration in MS due to the associated increase in blood volume and relative tachycardia. Patients with MS who were previously asymptomatic can develop life-threatening pulmonary congestion during the second or third trimester. Other conditions that can lead to worsening MS symptoms due to increased hemodynamic load in the setting of stable valve disease include fever, anemia, and hyperthyroidism.

There are also defects or destruction in the mitral leaflets, chordae, and papillary muscles, caused by infective endocarditis or chordae tendineae rupture, but also infarction or hypertrophic cardiomyopathy, ventricular dilation.

6 Physical examination and presentation

All patients with VHD should be evaluated with history and examination annually. An trans-thoracic echocardiography should be performed annually.

The murmur of VHD is the main exploratory sign. The intensity of the murmur does not correlate directly with the severity of VHD. As the left ventricle fails and stroke volume decreases, the murmur becomes softer and can disappear entirely.

The carotid upstroke is a slow rising, low-amplitude pulse with a late peak known as *pulsus parvus et tardus*. These findings are also specific for severe aortic estenosis.

Regarding mitral regurgitation, the most common, abnormal physical examination findings are most often attributable to atrial fibrillation (AF) and heart failure.

7 Stages of valvular heart disease

At risk Patients with mild to moderate valve disease.

Severe asymptomatic Patients who meet criteria for severe valve disease based on imaging or invasive testing but who have not yet developed symptoms. Often divided into subgroups depending on whether the ventricles are compensating.

Severe symptomatic Patients with severe valvular disease who have symptoms that are most attributable to their valve disease. A multidisciplinary heart valve team should be used when intervention is considered.

8 Diagnosis

Transthoracic ecocardiography is used to confirm the diagnosis, evaluate the mechanism, assess ventricular function, estimate pulmonary artery pressure, and grade the severity. Echocardiographic findings will differ between rheumatic and calcific valvular heart disease, and can detect stenosis, regurgitation and severe features regarding the causes of the disease. Calculated valve area and mean trans-valve pressure gradient can help grade the severity.

9 Management and intervention

Severe and symptomatic cases require prompt surgical intervention (because of an acute heart failure). All symptomatic patients with severe rheumatic mitral stenosis should be evaluated for percutaneous mitral balloon commissurotomy.