Chapter 1

A Congenital Heart Disease Primer

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The ability to interpret cardiac data to determine an individual patient's cardiac anatomy and physiology is paramount in developing a safe plan for anesthesia or sedation. Although cardiac lesions can be placed into broad diagnostic categories, within each category and for each lesion significant variation can exist. For example, infants with tetralogy of Fallot (TOF) may have obstruction to pulmonary blood flow ranging from minimal to severe; if obstruction is minimal, they may exhibit signs and symptoms of pulmonary overcirculation or if severe, they may be overtly cyanotic. Patients who have been described as "pink tets" at home may, during the stress imposed by anesthesia and surgical manipulation, exhibit significant tet spells. Wide pathophysiologic variability exists even within a given lesion and each patient must be considered on an individual basis, rather than being defined by his or her diagnosis. Patients who have undergone corrective surgeries, although "repaired," often have important residua or sequelae that must be noted. It should also be emphasized that the effects of the patient's underlying cardiac disease on other organ systems must be taken into account as well.

The anesthesia practitioner must be able to efficiently assimilate data including the patient's current history, physical examination, pertinent imaging studies, and cardiac catheterization data in order to develop an accurate assessment of the individual patient's pathophysiology prior to surgery. Are there intracardiac shunts, elevated pressures in specific cardiac chambers and/or volume overload, or reduced ventricular function? Are there rhythm abnormalities that might lead to a reduction in cardiac output? These factors may all impact the perioperative and anesthetic plan. This chapter is dedicated to outlining major principles useful in guiding preoperative analysis of the patient with congenital heart disease (CHD).

Basic Concepts

When referring to cardiac chambers, the terms "right" and "left" refer to morphologic characteristics and the terms "right-sided," "left-sided," "anterior," and "posterior" give a spatial frame of reference. When referencing structures other than cardiac chambers, such as the vena cavae, the terms "right" and "left" refer to spatial positioning in the thorax.

- The ***segmental approach*** to analysis of congenital cardiac lesions offers a framework for assessment and analysis of the path of blood flow through the heart. The three major segments or building blocks considered are the **atria,** the **ventricles**, and the **great arterial trunks**, along with the connections between each of them. The segmental approach begins by determining the position of the heart in the thorax, the direction of the cardiac apex, and the situs of the thoracic and abdominal organs. Visceral situs, or sidedness, may be ***solitus*** (normal arrangement), ***inversus*** (liver on the left, stomach on the right), or ***ambiguous*** (indeterminate). Abnormal arrangements of the viscera, heart, and lungs are seen in heterotaxy syndromes and are associated with a high likelihood of CHD.
- The ***physiologic approach*** considers classification of lesions according to the presence of **shunts**, **obstruction**, or **combinations** of the two.

Shunting may be described as anatomic or physiologic. *Physiologic* shunting is defined as venous return from one circulatory system recirculating through the arterial outflow of the same circulatory system. *Anatomic* shunts are communications between two circulations, either at the atrial, ventricular, or great arterial level. Physiologic shunts are often the result of anatomic shunts, but they can also occur in the absence of an anatomic shunt. An example of physiologic shunting without anatomic shunting may be seen in transposition of the great arteries, where systemic venous return travels to the right atrium, the right ventricle, to the aorta, and then again returns to the right atrium.

Effective blood flow is defined as the quantity of venous blood from one circulation that reaches the arterial system of the other circulation. Therefore, effective pulmonary blood flow is the quantity of systemic venous return that reaches the pulmonary arterial system. Effective pulmonary blood flow and effective systemic blood flow are *always equal*. Effective blood flow is the flow necessary to maintain life.

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Total blood flow is the sum of both effective blood flow to a circulation and recirculated blood flow. Total systemic blood flow and total pulmonary blood flow are *not equal* even in normal patients, as a small amount of physiologic shunting always exists. Recirculated or physiologic shunt flow can be thought of as the extra noneffective blood flow added to effective blood flow, together yielding total blood flow to a circulation.

Anatomic shunts may additionally be characterized as simple or complex. In ***simple shunts*** the degree of shunting is determined by the size of the orifice. With a small orifice, the size of the opening determines the amount of shunting. For large or nonrestrictive orifices (***dependent shunt***), the quantity and direction of shunting is determined by the outflow resistances, or the ratio between the pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR). This ratio is known as the $\boldsymbol{Q_p:Q_s}$. As the direction and magnitude of shunting are determined by the relationship between PVR and SVR, the effects of hemodynamic and ventilatory manipulations on these resistances during an anesthetic assume great importance.

Obstruction(s) may exist to either systemic or pulmonary blood flow at one or multiple levels. Infants with critical obstruction to either circulatory system frequently require prostaglandin E_1 infusions to maintain ductal patency and flow to the obstructed circulation until a surgical or catheter-based therapeutic intervention can take place. ***Complex shunts*** occur when obstruction exists along with a shunt. The degree of shunting in a complex shunt is determined by the degree of obstruction along with the PVR or SVR; the more significant the obstruction, the less the PVR and/or SVR will impact shunting. Obstructions may be either *fixed* or *dynamic* in nature. In a lesion such as tetralogy of Fallot it is common for elements of both fixed and dynamic obstruction to be present and to impact the direction and magnitude of shunting through the ventricular septal defect. For example, a child with TOF has a large ventricular septal defect (VSD) frequently accompanied by significant fixed and/or dynamic subvalvular right ventricular outflow tract (RVOT) obstruction as well. For this child, the degree and direction of shunting at the level of the VSD is determined primarily by the degree of RVOT obstruction; therefore left-to-right shunting decreases as RVOT obstruction increases with a tet spell, resulting in a decrease in shunting or even reversal of shunting. Although increases in PVR will contribute to the total right ventricular outflow resistance for the patient with TOF, the role of PVR is not as significant in this scenario compared to a child who has an isolated large or nonrestrictive VSD and no RVOT obstruction.

Clinical Pearl

Shunting occurs when venous blood from one circulatory system (either pulmonary or systemic) returns or recirculates through the arterial outflow of the same circulatory system, completely bypassing the other circulation. In a nonrestrictive or dependent shunt, as shunting is determined by the relationship between pulmonary and systemic vascular resistances, the effects of hemodynamic and ventilatory manipulations during an anesthetic assume great importance.

Another important concept in understanding complex CHD involves the concept of ***series*** versus ***parallel circulations***. In general, the normal systemic and pulmonary circulations are in series, with blood traveling through each circulation once, without mixing of deoxygenated and oxygenated blood (excepting the bronchial veins). An example of parallel circulations is unrepaired dextro (d)-transposition of the great arteries, where blood travels only to the pulmonary or systemic circulation. Mixing at the atrial, ventricular, or great arterial level (patent ductus arteriosus) is essential to allow mixing of oxygenated and deoxygenated blood and maintain life. (See Figure 1.1.) Another example of a parallel circulation is the child with single ventricle physiology, with both pulmonary and systemic circulations dependent on the same pump. Manipulations affecting resistance or flow in either circulation will therefore affect the performance of the other circuit. This is often referred to as "balancing" circulations and will be discussed in several chapters.

Imaging

Cardiac Catheterization

Cardiac catheterization in patients with CHD may be performed for diagnostic or therapeutic/interventional indications. Echocardiography has become the gold standard for initial diagnosis and ongoing assessment, particularly because in most patients it avoids the need for sedation or general anesthesia and for invasive vascular access. Fewer cardiac catheterizations are now performed solely for diagnostic indications. Conversely, interventional applications for cardiac catheterization have continued to grow in importance and include percutaneous implantation of valves and hybrid procedures utilizing both surgical and catheterization techniques. Most children require general anesthesia for cardiac catheterization, particularly when interventional procedures are anticipated. Unless extenuating circumstances exist, all efforts are made to utilize an FiO_2 of 0.21 and to maintain

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Table 1.1 Calculations and Normal Values

	Calculation	Normal Value
Ejection fraction (EF) (%)	(SV/EDV) × 100	54%–75%
Shortening fraction (SF) (%)	(EDD – ESD)/EDD × 100	30%–40%
Cardiac output (CO)	SV × HR	0.8–1.3 L/m (neonate/infant) 1.3–3.0 L/m (child) 4–8 L/m (adolescent/adult)
Cardiac index (CI)	CO/BSA	4.0–5.0 L/m/m^2 (neonate/infant) 3.0–4.5 L/m/m^2 (child) 2.5–4.0 L/m/m^2 (adolescent/adult)
Oxygen content	(O_2 sat × 1.36 × 10 × hemoglobin concentration)	
Pulmonary blood flow (Q_p)	VO_2 (mL/min)/PV O_2 conc – PA O_2 conc	
Systemic blood flow (Q_s)	VO_2 (mL/min)/ SA O_2 conc – MV O_2 conc	
Q_p:Q_s (simplified)	SA sat – MV sat/PV sat – PA sat	
SVR	(MAP – CVP) × 80/CO	10–15 iWu (infants) 15–20 iWu (1–2 years) 15–30 iWu (child)
PVR	(mPAP – mLAP) × 80/CO	8–10 iWu (<8 weeks) 1–3 iWu (>8 weeks)
TPG	mPAP – LAP	

BSA, body surface area; CVP, central venous pressure; EDD, end-diastolic diameter; EDV, end-diastolic volume; ESD, end-systolic diameter; HR, heart rate; iWu, indexed Wood units; MAP, mean arterial pressure; mLAP, mean left atrial pressure; mPAP, mean pulmonary artery pressure; MV, mixed venous; MV sat, mixed venous saturation; PA sat, pulmonary artery saturation; PV sat, pulmonary venous saturation; PVR, pulmonary vascular resistance; Q_p:Q_s, ratio of total pulmonary blood flow to total systemic blood flow; SA, systemic artery; SA sat, systemic arterial saturation; SV, stroke volume; SVR, systemic vascular resistance; TPG, transpulmonary gradient; VO_2, oxygen consumption.

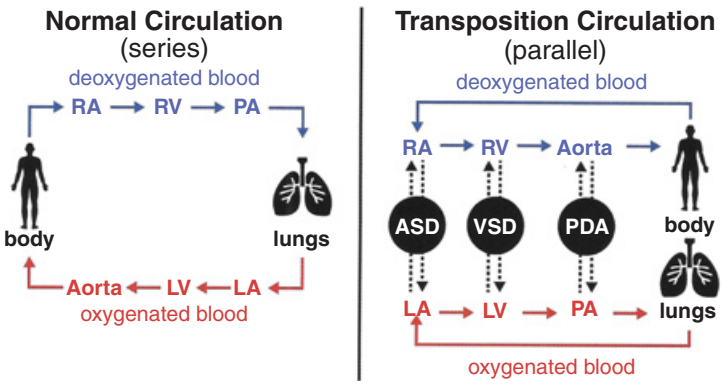

Figure 1.1 Comparison of the normal circulation, which is in series, with that in transposition of the great arteries (TGA), which is in parallel. In the normal circulation (left), deoxygenated blood from the body enters the right-sided circulation, then is oxygenated by the lungs and then distributed to the body via the left side of the heart. In the transposition circulation (right), the potential sites of mixing are shown: atrial septal defect (ASD), ventricular septal defect (VSD), and patent ductus arteriosus (PDA). The circulation is in parallel, so deoxygenated blood from the body enters the right side of the heart, but because of the anomalous connection, it is recirculated to the body via the aorta. Similarly, the oxygenated blood from the lungs enters the left heart but is recirculated to the lungs via the PA (pulmonary artery). RA, right atrium; RV, right ventricle. From McEwan A. and Manolis M. *Anesthesia for Transposition of the Great Arteries*. In Andropoulos D. B., Stayer S., Mossad E. B., et al. eds. *Anesthesia for Congenital Heart Surgery*, 3rd ed. John Wiley & Sons; 2015: 542–66. With permission.

ventilatory and hemodynamic parameters that parallel awake conditions, at least while initial hemodynamic parameters are measured. Any changes should be discussed with the cardiologist, as these changes will impact both the measured values that are obtained as well as calculated values.

The following data may be appreciated from a catheterization report. (See Table 1.1.)

- ***Anatomic diagnosis*:** The child's anatomy, including the effect of any interventions, is documented and assessed.
- ***Saturation data*:** Saturation data can be used to calculate Q_p:Q_s ratios and to document shunts via "step-ups" or "step-downs" in saturation between vessels and chambers. They may also help differentiate intracardiac shunting from ventilation/perfusion mismatch or intrapulmonary shunting.

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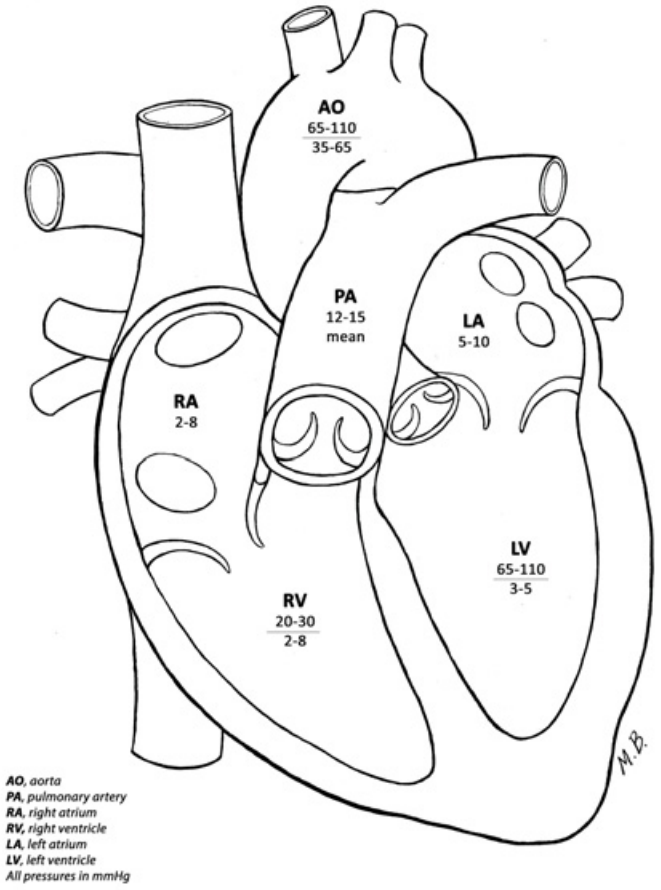

Figure 1.2 Normal cardiac pressures.

- ***Angiography*:** Cineangiography may be used to demonstrate blood flow patterns and ventricular function.
- ***Pressures*:** Right and left intracardiac pressures as well as vascular pressures are measured, along with pressure gradients. (See Figure 1.2.) Pressure gradients may be reported as either peak or mean gradients. Gradients may vary according to cardiac output: the higher the output, the higher the gradient. Thus a gradient can appear decreased with decreased cardiac output.
- ***Shunts*:** The direction and magnitude of shunts are recorded. The ratio of pulmonary to systemic blood flow is calculated and is an important piece of information to obtain prior to anesthetizing a child with CHD. (See Chapter 2, Q_p:Q_s.)
- ***Resistances*:** Resistance is measured as change in pressure divided by flow and is reported in Wood units (Wu). Systemic and pulmonary vascular resistances may be calculated and are most often normalized or indexed to body surface area. Notations for indexed pulmonary vascular resistance (PVR) can thus appear as "PRVI," "PVRi," or "iWu." In children with pulmonary hypertension it is important to note not only the initial PVR but also the response after any vasoreactivity testing or initiation of drug therapies.
- ***Cardiac output*:** Cardiac output (CO) for children is often indexed to body surface area and reported as cardiac index (CI). In measuring CO, either thermodilution or the Fick determination can be used; when utilizing the Fick determination, estimates of oxygen consumption (VO_2) based on body surface area or heart rate and age may be referenced for children. Maintaining the patient's FiO_2 at room air during cardiac catheterization is important while gathering this information in order to allow the dissolved oxygen component of the equation to be ignored.

Echocardiography

Echocardiography is the gold standard of diagnosis and ongoing assessment for most patients with CHD. Whenever possible, echo reports should be compared to previous reports

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to assess the progression of pathophysiology, and it is also useful to note in the cardiology evaluation the frequency of echocardiographic assessment for a particular patient. In patients with complex CHD echocardiographic reports may list a multitude of findings, and echocardiographic findings often prove useful to roughly sketch the heart, beginning with systemic venous return to the heart and adding each diagnosis as listed in the report. This heightens appreciation of the sources of pulmonary and systemic blood flow, as well as the presence of any shunts or obstructions, and aids immeasurably in understanding the patient's physiology and developing an appropriate anesthetic plan.

The following important parameters may be appreciated from an echocardiographic report:

- ***Ventricular function*:** The ***ejection fraction*** (EF) is defined as the fraction of blood ejected by the ventricle relative to its end-diastolic volume. It is important to note that EF assesses systolic function of the ventricle, and that diastolic dysfunction may exist in the face of a normal EF. The ***shortening fraction*** is calculated utilizing the percentage of change in ventricular diameter during the cardiac cycle and is dependent on preload and afterload.
- ***Gradients*:** The degree of obstruction across semilunar valves and outflow tracts may be estimated using the peak and/or mean instantaneous gradients.
- ***Regurgitant lesions*:** These are generally classified as mild, moderate, or severe and can best be appreciated by evaluating trends in the reported data.
- ***Pressure data*:** Information regarding right ventricular systolic pressure can be estimated by utilizing peak velocity of a tricuspid regurgitant jet.
- ***Measurement of chamber sizes*:** The size and thickness of the interventricular septum, the chamber dimensions of the ventricles, and information regarding valve annular sizes are reported. The "***z-score***" reported along with the values establishes a reference representing standard deviations of the measured value from the mean in a comparative population.

Other Imaging Modalities

Cardiac magnetic resonance imaging (CMRI) is noninvasive and avoids the use of ionizing radiation. It can provide excellent assessment of ventricular volumes, intracardiac anatomy, valvular regurgitation, blood flow through the heart, and extracardiac vascular anatomy. It is frequently utilized for ongoing assessment of right ventricular volume, ejection fraction, and pulmonary regurgitant fraction in patients who have undergone repair of TOF, and for serial assessment of coronary artery aneurysms in patients with Kawasaki disease. Use of CMRI often requires sedation or general anesthesia for pediatric populations, particularly for patients under the age of 8 years.

Computed tomography (CT), although it involves ionizing radiation exposure, is an important modality for assessment of CHD, particularly extracardiac vasculature, and is the gold standard for assessment of coronary artery disease.

High-Risk Patient Populations

Within the wide spectrum of patients with CHD who require noncardiac surgery, several groups have been specifically defined as having higher risk during the perioperative period. As studies have documented increased risk, they have also aided in delineating those patients and diagnostic categories at higher risk for adverse events, cardiac arrest, and/or mortality. Younger age, higher American Society of Anesthesiologists physical status, and need for emergent surgery have been shown to contribute to increased risk. Not surprisingly, analysis of outcome data for noncardiac surgeries in adult CHD patients also reveals increased mortality when compared with a cohort of patients without CHD. Significantly, Maxwell et al. identified that the number of procedures being performed in this patient population was increasing over time, and many were performed outside of teaching hospitals [1].

Specific categories or lesions have also been identified as follows:

- ***Pulmonary hypertension***, particularly with systemic or suprasystemic right ventricular pressures
- ***Single ventricle physiology***, particularly patients with shunt-dependent physiology, significant atrioventricular valve regurgitation, and the failing Fontan
- ***Severe ventricular dysfunction*,** including cardiomyopathies
- ***Severe left-sided obstructive lesions*** (aortic stenosis (gradient >60 mm Hg), subaortic stenosis, mitral stenosis)
- ***Williams syndrome***

Mortality in children with and without heart disease has recently been evaluated using the American College of Surgeons National Surgical Quality Improvement Program database. Children with heart disease were assessed and divided into groups with minor, major, or severe residual lesion burden and functional status. Using this strategy, children with minor CHD were found to have no greater risk than the general population for overall mortality or adverse events, while children with major or severe CHD had a higher mortality [2]. Therefore, utilizing

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the expertise of colleagues with specific training in cardiovascular anesthesia, as well as consultation with colleagues in pediatric cardiology, is always recommended when caring for patients who meet the above criteria or when other specific high-risk factors are identified.

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Suggested Reading

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