

# ESSENTIALS OF ECHOCARDIOGRAPHY #4

## Congenital Heart Disease

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### The Use of Echocardiography in Congenital Heart Disease

Two-dimensional echocardiography is ideally suited for the evaluation of congenital heart disease because of its ability to visualize cross-sections of complex cardiac anatomic structures. Visualizing heart walls, chambers, and valves, it is, in many ways, superior to angiography for revealing complex spatial morphologic information.

This unit discusses the use of two-dimensional echocardiography in the evaluation of congenital heart disease. It must be realized that currently, conventional Doppler methods and Doppler color flow imaging have added even more diagnostic power to cardiac ultrasound for evaluation of congenital heart disease. For many seemingly simple and some complex disorders, cardiac catheterization is no longer necessary when data can be reliably obtained by echo and Doppler methods. M-mode echocardiography has been almost totally supplanted by these newer modalities.

The purpose of this unit is to provide a *basic* understanding of congenital heart disease and how echocardiography is helpful in establishing diagnoses. As such, its aim is principally toward those with little background in this area. Not all disease entities can be covered and only the more common disorders will be described. Little discussion of patient management is possible.

*All* congenital heart disease is potentially complex. Such a statement should not be frightening as it only reflects the fact that the presence of one lesion increases the possibility for another. Multiple lesions are possible. For example, transposition of the great vessels may exist with or without ventricular septal defects or with or without right ventricular outflow tract obstruction. In addition, just like in adult acquired disease, congenital heart disorders represent a spectrum. There can be mild, moderate, or severe expressions of any disorder.

### Views for Congenital Heart Disease

As with acquired heart disease, the standard apical, parasternal, and subcostal views are used for the majority of recordings. In addition, emphasis is placed on certain views that are particularly rewarding. For example, the subcostal approaches (**Fig. 1**) identify the interatrial septum and the relationships of the atrial and ventricular septum to the atrioventricular valves. Suprasternal views are good for examination of the great vessels and the aortic arch. All views obviously must be utilized. In small children, lack of attenuation from the rib cage permits routine imaging with high frequency transducers such as 5MHz or higher.

## Classification of Congenital Heart Disease

Many physicians dealing with adults find congenital heart disease extremely complex. The terms and classifications used over the years by pediatric cardiologists may be largely to blame. Most previous nomenclature systems are largely based upon embryology. Thus, such terms as L-loop and D-loop were common and generally confused most individuals. In this unit, a simple descriptive nomenclature, known as the sequential segmental approach, introduced during the 1980's is employed. It avoids terms derived from embryology and, where possible, uses simple and uncomplicated descriptions. Its major goal is to convey information simply and accurately without regard to how the lesions came to be.

### The Sequential Segmental Approach

This newer nomenclature approach has remarkably simplified the classification of congenital heart disease. It is based on following the blood flow into the heart (systemic venous and pulmonary venous), through the heart (the atrioventricular valves and ventricles) and then out the great vessels (semilunar valves and great vessels). This nomenclature system is extraordinarily helpful to those conducting echocardiographic examinations as it forms a systematic guide for verification that all the pertinent chambers and valves and their relationships have been documented. The system is dependent on a few words that are very important in describing the various lesions:

**Connection** refers to the sequence of anatomic structures. Normally, the right atrium is connected to the right ventricle by means of the tricuspid valve. The right ventricle is then connected to the pulmonary artery by means of the pulmonic valve. Therefore, there are atrioventricular connections and ventriculo-great arterial connections.

**Concordance** describes the relationship between the various chambers, valves, and great vessels. In the normal heart all the connections and relationships in the anatomic sequence are concordant.

**Discordance** describes abnormal relationships between the various chambers and great vessels. For example, when the right atrium leads into the morphologic left ventricle and the left atrium into the morphologic right ventricle, the atrioventricular relationships are discordant, as seen in **Fig. 2**. Likewise, the atrioventricular relationships may be concordant (normal) but the ventriculo-great arterial relationships may be discordant where the aorta rises from the right ventricle and the pulmonary artery from the left ventricle. Known formerly as transposition of the great vessels, these abnormal relationships would now be termed ventriculo-great arterial discordance as seen in the right panel of **Fig. 2**.

**Absent or imperforate connections** – Valves normally form the connections between chambers. There are atrioventricular connections that lead from the right atrium to right ventricle or left atrium to vessels. When connections are not present, the term absent connection is used. Thus, when the tricuspid valve is absent, an absent right atrioventricular connection may not be totally absent, only severely malformed and does not allow blood to pass antegradely. In this setting the term imperforate connection may be used, also seen in **Fig. 3**.

**Commitment** further describes possible abnormalities of flow through valves into ventricles and great vessels. For example, in a patient with tetralogy of Fallot, the atria, atrioventricular valves,

and ventricles are positioned normally, and concordant. Since the aorta overrides a ventricular septal defect the aorta is doubly committed to both ventricles. Likewise, in cases where there is only one ventricle (univentricular heart), both atrioventricular valves are usually doubly committed to the single ventricle.

*Ambiguous* is used when precise identification of a ventricle or other structure cannot be made. For example, in a univentricular heart with a doubly committed atrioventricular connection it may not be possible to always identify clearly whether it is the right or left ventricle. Thus, the single ventricle would be *ambiguous*.

*Inlet* refers to anomalies of the structures and flow into the ventricle.

*Outlet* refers to anomalies of the structures and flow out of the ventricles into the great vessels.

## The System in More Detail

The system assumes that flow through the heart is normal and begins with properly identifying the atria, and their position in the chest. In a normal individual there are two atria, each with venous inflow. One must identify the inferior and superior vena cava inflows into the right atrium and, where possible, identify four pulmonary veins into the left atrium.

Following the normal sequence of flow, one then identifies the atrioventricular valves and ventricles. Normally there are two atrioventricular valves, tricuspid and mitral. The tricuspid valve is committed to the right ventricle and the mitral valve to the left ventricle. Normally both the atrial and ventricular septa are intact.

Again following the normal sequence of flow, blood should emerge out of the ventricles into the great vessels. The pulmonary artery, taking flow to the lungs, is normally committed to the right ventricle while the aorta, taking blood to the systemic circuit, is normally committed to the left ventricle.

The pulmonary artery emerges from the right ventricle and passes anterior to the aorta. The pulmonary artery then bifurcates and is differentiated from the aorta that forms an arch, giving off vessels to the head and neck. The pulmonary artery and aorta “criss-cross” as they arise from their respective ventricles.

Given these normal sequences and relationships the terms previously mentioned are used to describe abnormal hearts. Chambers, valves, or vessels may be absent (atretic) or small (hypoplastic). Relationships between chambers and valves may be concordant (normal) or discordant. In addition, chambers or valves may be doubly committed or normally committed. An outline of the disorders is presented in **Table 1**.

**TABLE 1**

Outline of Congenital Heart Disorders Discussed in This Book

- I. When chambers and valves are in normal sequence and position
  - A. When shunting is predominant
    1. Atrial septal defects (secundum, primum, sinus venosus, and coronary sinus)

2. Ventricular septal defects (subarterial, muscular, inlet, and perimembranous)
  3. Atrioventricular septal defects (AV canal defects)
  4. Patent ductus arteriosus
- B. When stenosis or obstruction is predominant
1. Absent atrioventricular connections (tricuspid and mitral atresia)
  2. Absent or obstructed ventriculo-great arterial connections (pulmonary atresia, aortic)
  3. Obstructed great arteries (coarctation of the aorta, aortic atresia)
  4. Obstructed venous inflow (total anomalous pulmonary venous return)
- C. Anomalous valve position (Ebsteins's anomaly)
- II. When chambers and valves are not in normal sequence or relationship
- A. Anomalies of relationships between atria and ventricles
1. Double-inlet or right ventricle (with univentricular heart)
  2. Atrioventricular discordance (corrected transposition)
- B. Anomalies or relationships between ventricles and great vessels
1. Tetralogy of Fallot
  2. Double-outlet right and left ventricles
  3. Truncus arteriosus
  4. Ventriculo-great arterial discordance (transposition of the great vessels)

### **Atrial Situs**

In the complex myriad of possible anomalies, some of the most difficult to determine are those involving abnormalities of atrial situs. Mention of atrial situs is made only to point out that any complex malposition of the atria may occur.

Four possible atrial arrangements are encountered in congenital heart disease: the normal arrangement of the right atrium and left atrium (situs solitus), the direct opposite to the normal where the right atrium is on the left and the left atrium on the right (situs inversus), bilateral right atria (right atrial isomerism associated with the asplenia syndrome) or bilateral left atria (left atrial isomerism associated with the polysplenia syndrome). For the purposes of this volume, we shall assume atrial situs to be normal in all patients. The reader is referred to more detailed texts for determination of this important facet of evaluating complex congenital heart disease.

### **When Chambers and Valves Are in Normal Sequence and Position**

It is easiest to begin with disorders where chambers and valves are in the normal, or relatively normal position. Such disorders comprise those of atrial and/or ventricular septal defects and those disorders where chambers and valves are absent or small, causing obstruction. As a consequence, these types of defects will be divided into those when shunting (abnormal blood flow between the left and right circulations), obstruction, or regurgitation are predominant.

### **When Shunting is Predominant**

In the structurally normal heart, right and left sides are divided and are without communication. Shunting refers to those flow anomalies where there is an abnormal communication, such as an atrial or ventricular septal defect, that allows abnormal flow between the right and left sides. Since right-sided pressures are normally lower than those on the left side, when such defects are encountered abnormal flow is usually left-to-right and increased flow into the lungs results. Normally, the lungs can accommodate the increased flow without significant symptoms if the degree of shunting is small or moderate.

When significant shunting is present and exceeds the ability of the lungs to accept the increase, the lungs are literally flooded and symptoms of cardiac failure ensue. Where no failure is clinically evident, such shunting, over time, may result in reactive changes in the pulmonary vasculature where normally low pressures then rise within the right heart and exceed those on the left side. Depending on the clinical situation, such changes may be permanent and cause right-to-left shunting (Eisenmenger's Syndrome). Without early recognition and correction, these simple problems may result in permanent, inoperative damage to the lungs. In addition, any communication between right and left sides of the heart may allow for the possibility of venous emboli entering the arterial circuit.

### **Atrial septal defect**

Defects in the atrial septum are traditionally divided into four types according to their location: secundum, primum, sinus venosus, and coronary sinus defects. The general location of these defects is shown in **Fig. 4**. It is important to note the type of defect or defects present as each type is associated with other anomalies and may require different surgical approaches. As shall be noted later, primum atrial septal defects are very complex and argument exists whether they should continue to be classified as atrial septal defects alone.

Any atrial septal defect results in shunting of blood from the left atrium into the right atrium. When the defects are large and the left-to-right atrial level shunting is significant, the right ventricle and right atrium enlarge significantly because of the increased volume.

Frequently, the actual defects may be identified. Ostium secundum atrial septal defects are best visualized from the subcostal position. As seen in **Fig. 5**, secundum defects are seen to lie centrally within the atrial septum, and are bound on all sides by atrial septal tissue. Note in **Fig. 6** that the normal position of the atrioventricular valves is characteristic in that the tricuspid valve is always located closer to the ventricular apex when compared to the normal location of the mitral valve. In this normal position of atrioventricular valves, an area of septal tissue is seen between the left ventricle and right atrium. This is known as the atrioventricular septum.

In this entity, the atrial septum close to the atrioventricular valves (known as the primum septum) is usually intact. Similarly, the septum shared by the left ventricle and right atrium is also intact. Thus, adequate interrogation of the atrial septum requires visualization of its midportion and also portions adjacent to the atrioventricular valves.

Secundum defects are areas adjacent to the atrioventricular valves are best imaged from the subcostal approach where all these structures in the center of the heart are readily identified. Using the subcostal approach, most, but not all, secundum defects may be imaged. The apical four-chamber view is usually unreliable for imaging of secundum defects as the atrial septum lies parallel to the transducer beam and an absence of septum in this view may be due

to a “drop-out” of targets. Thus, the presence of an atrial septal defect should never be determined only from the apical four-chamber view.

Ostium primum defects involve areas of the atrial septum adjacent to the atrioventricular valves. It is important to recognize that primum defects are not only defects in the atrial septum. Rather, the defect also involves the common atrioventricular septum, and the result is deformity of the alignment of the atrioventricular valves and may be associated with defects of the adjacent interventricular septum.

Such defects are also best examined from the subcostal approach as seen in **Fig. 7**. One diagnostic hallmark of the disorder is the absence of the atrial septum adjacent to the atrioventricular valves. In addition, since the defect extends into the atrioventricular septum, the level of insertion of both the tricuspid and mitral valves onto the crest of the ventricular septum are equal. Thus, in this disorder there is invariably a defect of the shared septum and some possible or real malformation of the atrioventricular valves. In contrast to secundum defects, primum defects are reliably visualized from the apical as well as the subcostal approach, with atrioventricular valve tissue forming their lower margin, and the secundum septum forming their upper margin.

It is important to note that a primum defect may extend into one of the other atrioventricular valve leaflet. **Fig. 8** shows a parasternal short axis where the defect extends into the anterior mitral leaflet giving the appearance of two anterior mitral valve leaflets. Such a mitral anomaly is commonly known as a “cleft” mitral valve. Such a cleft may, or may not, result in mitral regurgitation. Multiple arguments exist whether this anomaly should properly be referred to as a “cleft” and are beyond the scope of this discussion. In short, the mere presence of an impressive cleft does not always imply the presence of severe mitral regurgitation. As a consequence, not all such cleft valves are deserving of repair.

Sinus venosus defects are surprisingly difficult to detect as they are located superiorly on the interatrial septum near its junction with the superior vena cava. Traditional subcostal views are frequently unrewarding and other nonconventional views must be attempted. **Fig. 9** demonstrates a subcostal long axis of the inferior and superior venae cavae where the defect is readily recognized. To reliably obtain this view requires considerable practice and experience. Supraclavicular and suprasternal views of this defect are also possible, but less reliable. Because these defects are in an area so difficult to access, they are frequently missed except by the most experienced examiners.

Importantly these defects are frequently associated with anomalous drainage of the right upper pulmonary vein to the right side of the atrial septum. Careful examination of all views is required to detect this complex anatomy.

Coronary sinus defects can be recognized by finding an interatrial communication at the anticipated site for the coronary sinus. The fossa ovalis may be intact, or there may be a secundum defect. Small coronary sinus defects are easily missed, with the atrial septum appearing intact in every view recorded. These defects are exceedingly rare, difficult to detect with certainty and associated with anomalous insertion of a left-sided superior vena cava into the coronary sinus.

It is important to precisely locate the position of the atrial septal defect. Secundum defects are usually easy to repair. Primum defects frequently include other structural anomalies of the atrioventricular junction and valves, and therefore may require very complex preoperative planning. Sinus venosus defects, located very high, require different venous cannulation

techniques than normally employed. The presence of one atrial defect should alert the examiner to look carefully for another, as two different types may be found in any one patient. Indeed, echocardiographic location of atrial defects provides the surgeon with detailed data by which to plan the operative approach.

The abnormal flow through the defects may be readily detected using Doppler methods, particularly Doppler color flow imaging. If such methods are not available, an echo contrast study can usually confirm the diagnosis of atrial septal defect. To perform a contrast study, 3 to 10 ml of fluid (usually saline) is injected rapidly into an antecubital vein. The amount of fluid injected depends on patient size, with 10 ml the full adult volume. Mild agitation of the fluid before injection may enhance the contrast. Rapid infusion through a narrow canula lowers the pressure of the injectate so that some dissolved gas comes out of the solution in the form of microbubbles. These are carried to the heart by the bloodstream where they act as strong ultrasound reflectors, causing a normally sonolucent, blood-filled cavity to become opacified. This improves cavity delineation and allows intracardiac shunts to be detected. The microbubbles are fully absorbed in the lungs, so a peripheral venous injection shows only a shunt with a right-to-left component. **Fig. 10** shows the presence of bi-directional shunting in a patient with an atrial level communication.

The use of saline contrast for the identification of atrial level shunting depends on the small degree of right-to-left shunting present in all patients with interatrial flow communications. This shunting occurs due to a short period of elevation of right atrial pressure over left atrial pressure that occurs just after the onset of ventricular systole. Saline contrast techniques are the most sensitive method available for the detection of interatrial shunting and are more reliable than angiography, green-dye, or oximetry. In fact, small degrees of interatrial shunting, through presumed patent foramen ovale may be seen in 12-to-17 percent of the otherwise normal population.

Continued experience with echocardiography indicates that other anomalies of the interatrial septum may be identified. Normally, the interatrial septum is relatively fixed in position and moves passively with the movement of the entire heart. Occasionally, a hypermobile septum may be seen (**Fig. 11**) and has been anatomically linked with the presence of an aneurysm of the interatrial septum. In this setting the septum is usually thin and may be fenestrated, resulting in variable degrees of interatrial flow. In fact, when such a finding is noted by echo, the majority of patients show small degrees of interatrial shunting by saline microcavitation techniques. The degree of shunting is rarely hemodynamically significant. It should be noted, however, that recent evidence has indicated some link between this disorder and the presence of embolic stroke in older age population. The evidence is not strong enough as yet to warrant operative intervention.

### **Ventricular septal defects**

The interventricular septum is a highly complex, three-dimensional structure formed from a number of morphologically distinct subunits. It can be divided into subarterial, perimembranous and muscular components, the subarterial and perimembranous septa being tiny in comparison to the vast bulk of the muscular septum (**Fig. 12**). As with atrial septal defects, it is important to note the precise location of these defects as each may have a different clinical prognosis for ultimate spontaneous closure and each may require a different surgical approach when operative intervention is indicated.

When shunting is significant, the right ventricle invariably enlarges depending on the degree of excess flow into the right side. If the defect is large and the shunt unrestricted, the

pressures within the right heart will be identical to those in the left. In this case the right-sided pressures are referred to as “systemic”. On occasion of long-standing high flow into the lungs, right-sided pressures may become “suprasystemic”.

Compared to atrial septal defects, where it is common for the anomaly to be restricted to one or the other portion of the atrial septum, there is frequent crossover between ventricular septal defects. For example, those in the muscular septum may extend into the perimembranous septum. Any combination is possible.

Two-dimensional echocardiography is highly rewarding in identifying the presence of moderate-to-large ventricular septal defects, particularly in small children. When the defects are small, they may be quite difficult to visualize. Doppler methods are very helpful in identifying the defects, whether large or small. Contrast injection is less helpful, except when right ventricular pressures are in excess of those in the left ventricle and result in right-to-left shunting. In most cases where right ventricular pressures are lower than on the left side, a negative “wash-out” of uncontrasted blood must be identified. Such approaches are only rarely helpful.

Using two-dimensional echocardiography, each type of ventricular septal defect can be identified and classified on the basis of a specific echocardiographic pattern. Each view must be used for proper spatial orientation of the defect.

Classic muscular defects are those bound entirely by areas of the muscular septum. **Fig. 13** demonstrates ventricular short axes from two patients with very large midmuscular ventricular septal defects with virtually free communication between the ventricles. **Fig. 14** shows a parasternal long axis from a patient with a somewhat smaller ventricular septal defect in the midmuscular septum. Such ventricular septal defects, when located toward the apex in the trabecular septum may be difficult to identify by echo alone. In these cases, Doppler methods are required.

Outlet (or subarterial) defects are noted in subarterial regions. **Fig. 15** demonstrates a notably large subarterial defect from the parasternal long axis. The right ventricle is dilated indicating significant shunting.

Any ventricular septal defect, regardless of its location, may be covered by an aneurysm, as seen in **Fig. 16**. Such aneurysms are thought to occur as a result of a spontaneous attempt of closure and are quite variable in size.

Experienced examiners are usually able to image almost all hemodynamically significant ventricular septal defects in infants, children, and adults (the exception being those with small, multiple trabecular defects). Precise location of these defects is very important, as their location gives some indication as to the likelihood for spontaneous closure. For example, small perimembranous and muscular defects may spontaneously close, and in the proper clinical setting may be followed by echocardiography and Doppler.

It is also important to note the precise location of the defects when surgical intervention is indicated. Muscular defects may be multiple, and depending on their size, may be quite difficult to see through the tricuspid valve as they may be obscured by the multiple trabeculations of the right ventricle. Such multiple muscular defects are referred to as a “Swiss cheese” septum.

Similarly, the position of the defects guide the surgeon's approach. **Fig. 17 (left)** shows a short-axis view of a subarterial outlet defect entering the right ventricle just below the pulmonic valve. Such a defect would be very difficult to approach through the tricuspid valve as it is a great distance away and requires a ventriculotomy for proper closure. In addition, these types of defects may, over time, allow for prolapse of the right coronary cusp of the aortic valve into the defect resulting in aortic insufficiency. **Fig. 17 (right)**, in contrast, shows a short axis view of a perimembranous defect. Its proximity just under the tricuspid valve indicates its easy surgical closure through the tricuspid valve and obviates an unwarranted ventriculotomy.

### **Atrioventricular septal defects**

Formerly known as "canal defects" or "endocardial cushion defects" atrioventricular septal defects (AVSD) are present when any abnormality exists of the shared septum between the left ventricle and the right atrium (the atrioventricular septum). As previously discussed, ostium primum atrial septal defects extend into this common septal area and are really a mild form of an atrioventricular septal defect.

In its most severe form (**Fig. 18**) the primum septum is absent with the defect extending to the muscular septum leaving the entire central portion of the heart without dividing septal tissue, either atrial or ventricular. In this setting, a common atrioventricular valve orifice is present with a double-committed single atrioventricular valve (i.e., the single atrioventricular valve enters into both ventricles). In this case the AVSD is referred to as a complete defect.

Those inexperienced with congenital heart disease frequently have difficulty understanding these defects. This is principally brought about because of the failure to recognize that AVSDs are comprised of a family of defects with ostium primum as a mild form and a complete AVSDs as the most severe. Many possibilities exist between the two extremes.

**Fig. 19** shows a parasternal diastolic four-chamber view of an infant with a complete AVSD. Only a small portion of the interatrial septum is present. There is total communication and mixing through the primum defect, ventricular septal defect, and common atrioventricular orifice. **Fig. 7** demonstrated the other extreme of a primum defect alone.

**Fig. 20** shows the diastolic and systolic appearance of a patient with a complete AVSD and common atrium. Note the insertion of some of the chordal structures onto the crest of the ventricular septum. AVSDs include a broad spectrum of atrioventricular junction abnormalities, all of which have two common features: an absent atrioventricular septum and abnormally formed atrioventricular valves. **Fig. 21** shows an unusual subcostal short-axis view through the common atrioventricular valve orifice from a patient with a total AVSD. It is important to identify all the leaflets present and trace their insertion into the left or right ventricle. Occasionally, an atrioventricular leaflet may bridge the central defect and have chordal insertion into both ventricles. In such cases, the leaflet is known as a "bridging leaflet" and must be surgically divided into left and right portions. It then is resuspended from a central patch to create separate atrioventricular orifices at the time of correction.

Certain "transitional" forms of AVSD exist. In these, a primum defect is seen in the atrial septum and a small ventricular septal defect is noted near the atrioventricular junction. Two separate atrioventricular valve orifices may be seen. Notably, patients thought to have merely a primum defect may, indeed, have a ventricular component.

## **Patent ductus arteriosus**

The patency of the ductus arteriosus, a small tube connecting the pulmonary artery to the aorta, is necessary during fetal life. Soon after birth, however, this connection spontaneously closes in most infants. If it does not close, and pulmonary pressures fall in the antenatal period, shunting between the aorta and the pulmonary artery occurs. In this setting, the shunting occurs through a patent ductus arteriosus.

Identification of the ductus arteriosus is an important part of pediatric echocardiography. The ductus is encountered in a number of clinical conditions, commonly in premature infants or full term infants with other forms of congenital heart disease.

It is possible to identify a large patent ductus arteriosus by two-dimensional echocardiography. A moderate-size ductus arteriosus is very difficult to identify with imaging alone except in the most skilled hands. Currently, the method of choice involves the adjunct use of conventional Doppler and/or Doppler color flow methods.

Without Doppler, only indirect signs of a patent ductus arteriosus are evident. With marked increase in pulmonary blood flow, the left atrium dilates significantly, together with an increase in left ventricular end-diastolic dimension. In the normal infant, the ratio of maximal left atrial dimension to aortic root dimension is less than 0.9:1. An increase in this ratio to greater than 1.1:1, combined with an abnormal increase in left ventricular end-diastolic dimension, is strongly suggestive of a patent ductus. Such findings, however, lack specificity as identical echocardiographic findings are also associated with both mitral incompetence and ventricular septal defect. Also, the left atrial enlargement associated with a persistent ductus is dependent on an intact atrial septum. Such indirect indices are not reliable in the hypovolemic neonate. Furthermore, a ductus associated with complex congenital heart disease cannot be excluded on the basis of normal left dimensions.

## **When Obstruction Is Predominant**

It is also possible that all the cardiac chambers and most valves are in normal sequence but some valves are not well formed, leading to absent valvular connections. When such valves are absent, the term “atresia” is also used, implying that no antegrade (or forward) flow is possible across the valve. Thus, atresia of any valve may occur. Valvular atresia on the right side prevents blood from reaching the lungs; valvular atresia on the left side prevents blood from reaching the systemic circuit. For any infant with valvular atresia to survive past the first few hours of life, a shunt lesion must be present to allow blood to progress antegradely through the heart.

Not all obstructive lesions prevent the total forward flow of blood. Some, such as subvalvular aortic stenosis, offer partial obstruction to flow. The degree of obstruction relates to the anatomic severity of the lesion.

## **Absent atrioventricular connections**

Absent atrioventricular valve connections (tricuspid or mitral atresia) are less common than those lesions previously discussed. Atresia of either of these valves may have one of two underlying causes: an absence of the atrioventricular connection, or an imperforate membrane blocking the valve orifice as noted in **Fig. 3**. Gradations between the two extremes also exist.

Where an imperforate membrane has caused atrioventricular valve atresia, there is a formed (but usually hypoplastic) atrioventricular valve ring blocked by an imperforate membrane. Atrioventricular communication is potentially possible by excision of the membrane.

More commonly, atretic valves have completely absent tissue. Hypoplasia of proximal or distal chambers to the atretic valve is also possible. **Fig. 22** demonstrates an absent right atrioventricular connection (tricuspid atresia) from the subcostal approach. The right ventricle is hypoplastic and difficult to visualize. The left-sided atrioventricular valve is present.

In this patient, survival would be impossible because no route for antegrade blood flow to the lungs is present. Note, however, that there is a large secundum atrial septal defect that allows blood to immediately mix with oxygenated blood in the left atrium and then transit the left ventricle into the system circuit. A central shunt (a synthetic tube between the aorta and the pulmonary artery) was surgically placed to allow some mixed blood to then indirectly travel back to the lungs for oxygenation.

**Fig. 23** shows an apical four-chamber view from another patient with tricuspid atresia. A thick ridge of tissue replaces the tricuspid valve. The right ventricle is poorly formed. The left-sided atrioventricular valve is present and the left ventricle is normal. Note that the atrial septum is bowed from the right atrium into the left atrium.

This patient was treated in infancy, similar to the previous patient. Later, the atrial septum was closed and the central shunt removed. Blood flow to the pulmonary artery was established by placing a conduit from the right atrium directly to the proximal pulmonary artery (the Fontan procedure). With no intervening right ventricle to pump blood into the lungs, hydrostatic pressure rises on the venous side. This is frequently sufficient to provide adequate blood for oxygenation. An absent left atrioventricular connection (mitral atresia) is somewhat less common. **Fig. 24** shows an infant with a normally formed tricuspid valve from the apical four-chamber view. The mitral valve has been replaced by a thick band of tissue. Survival in this case is impossible unless some route of blood flow returning to the lungs from the left atrium is provided. In this case, the atrial septum was emergently removed to allow for mixing of oxygenated blood with venous blood in the right atrium. Note that there is only one ventricle. The ventricle gave rise to both the pulmonary artery and aorta, assuring some blood flow to both the lungs and periphery.

### **Absent or stenotic ventriculo-great arterial connections**

In its most severe forms, atresia of the pulmonary valve prevents any antegrade blood from exiting the right ventricle to the lungs. In these cases the right ventricle may be poorly formed (hypoplastic right heart syndrome). Without an atrial septal defect to provide some mixing of blood and a route (such as a patent ductus arteriosus or surgically placed central shunt) to provide some flow to the lungs, survival is impossible.

In less severe forms, where an adequate right ventricle and tricuspid valve are present, it may be possible to surgically place a conduit between the right ventricle and pulmonary artery. Such a procedure is termed a “Rastelli operation”, and it establishes the normal sequence of flow by bypassing the atretic pulmonary valve.

Aortic atresia has a somewhat more dismal prognosis. If all the other chambers and valves are in normal sequence and position, aortic atresia in its most severe form is associated with severe hypoplasia of the left ventricle. No adequate route for oxygenated blood to reach the

systemic circuit is, therefore, available. In addition, with a hypoplastic left ventricle, no adequate ventricle is present to pump blood into the aortic arch. This extreme form is termed “hypoplastic left heart syndrome”.

Other, less severe forms of ventriculo-great arterial connections also occur. Outflow obstruction of the left heart in congenital heart disease can be at subvalvular, valvular, or supra-valvular levels, or may be more distally situated (as in coarctation of the aorta or interruption of the aortic arch).

Aortic stenosis originating from a congenital abnormality of the aortic valve is probably the most common form of aortic valve disease. There is usually symmetric left ventricular hypertrophy, although asymmetric septal thickening has been described. The aortic valve is frequently bicuspid (or virtually so, with two large cusps and one very hypoplastic cusp.) Examples of congenital aortic stenosis are presented in another unit. In critical cases the left ventricle is often considerable hypertrophied. Mixed stenosis and regurgitation is rarer in childhood than in adult life.

The features of subvalvular obstruction are similar to those seen in adults. The most common form of subvalvular aortic stenosis in childhood is a discrete fixed subvalvular narrowing frequently associated with a membranous ring. **Fig. 25** demonstrates a patient with a subvalvular membrane preoperatively, and then after the membrane was surgically removed. The subvalvular membrane is easily recognized on the two-dimensional long-axis view. Symmetric or asymmetric septal hypertrophy may be present and the aortic valve shows abnormal systolic closure and fluttering.

Hypertrophic obstructive cardiomyopathy with asymmetric septal hypertrophy does occur in childhood, though more rarely than in adults. The asymmetric septal hypertrophy is variable in degree and position and may be secondary to other lesions, such as coarctation, systemic hypertension, right ventricular hypertrophy, or valvular aortic stenosis. The whole trabecular septum may be involved or only a segment. Outflow tract obstruction is usually associated with systolic anterior movement (SAM) of the anterior mitral leaflet and partial midsystolic closure of the aortic valve. Examples of this entity have been presented in another unit.

### **Obstruction of the great arteries**

Obstruction to flow within the great arteries and distal to the semilunar valves is also possible. Most commonly, coarctation of the aorta is encountered as an area of discrete narrowing near the junction of the transverse and descending aorta (**Fig. 26**). While this diagnosis can be made frequently from the two-dimensional echocardiogram, Doppler methods have enhanced the reliability of ultrasound to detect this entity. Suprasternal views are necessary for proper visualization of the entire aortic arch.

Supra-valvular aortic stenosis is a rare form of left heart obstruction, commonly associated with idiopathic hypercalcemia in infancy. The aortic valve is frequently within normal echocardiographic limits. Narrowing of the aortic root can be observed just above the sinuses of Valsalva in the parasternal long-axis and short-axis views. The severity of left ventricular hypertrophy gives some indication of the degree of aortic narrowing.

Other anomalies of the aortic arch are also possible. Interruption of the aortic arch is a rare and highly lethal condition of infancy, which can be diagnosed by two-dimensional echocardiography from the suprasternal approach. Tubular hypoplasia of the arch as well as

various aortic rings may also be suspected from two-dimensional echo but frequently require angiography for more detailed delineation.

Distal obstructions in the pulmonary arteries may also be identified. Stenoses at the origin of the main stem pulmonary arteries may be recognized (peripheral pulmonary stenosis). Obstructive lesions beyond this level are difficult to detect with two-dimensional echocardiography.

### **Obstruction of venous inflow**

Many different anomalies of venous return to the heart also exist and some may be associated with obstruction. One important entity, total anomalous pulmonary venous return, exists when all four pulmonary veins fail to drain into the left atrium. Rather, the four veins enter a common pulmonary venous chamber that lies posterior to the left atrium but fails to connect with it.

When total anomalous veins are present there must be some mechanism for the returning oxygenated blood to enter back into the bloodstream. This common chamber usually has some entry into the systemic venous circuit and is accomplished by one of the following means: by connecting superiorly to a vertical vein and thence to the superior vena cava (**Fig. 27**); laterally to the right atrium, right superior vena cava, or coronary sinus; or inferiorly to the portal venous system. Severe obstruction to pulmonary venous drainage may be present in any configuration, but is most common where the confluence drains inferiorly to the hepatic system. **Fig. 28** shows a parasternal short axis from an infant where the pulmonary venous confluence is seen posterior to the left atrium forming a vertical vein that was noted to drain inferiorly. In this infant, immediate surgery was performed to connect the vertical vein to the left atrium to establish a normal flow sequence.

As with all congenital lesions, other variations also exist. All four veins need not drain to a common confluence and such mixed forms may produce extremely complex pulmonary venous drainage patterns. In most cases, two-dimensional echocardiography is very reliable for the identification of total anomalous pulmonary venous drainage and in identifying its ultimate drainage into the venous system.

A myriad of other venous anomalies are also possible, usually without obstruction. **Fig. 29** shows a dilated coronary sinus in a child with a large ventricular septal defect. When such a large coronary sinus is detected it results from two likely possibilities. Most commonly, two superior venae cavae are present with the right superior vena cava draining normally into the right atrium and the aberrant left superior vena cava draining into the coronary sinus. A saline microcavitation examination performed from the left arm readily identifies those patients with an aberrant left superior cava as contrast readily fills the coronary sinus.

## **Anomalies of Valve Position**

### **Ebstein's anomaly**

There are very few anomalies where the sequence and relationships of chambers and valves are normal, but a valve may be improperly positioned in the sequence. One such abnormality is Ebstein's anomaly where the tricuspid valve is displaced apically. **Fig. 30** demonstrates a severe expression of Ebstein's anomaly where the marked apical position of the tricuspid

valve “atrialized” much of what should have been the right ventricle. Two-dimensional echocardiography is the most reliable method available for the detection of this abnormality. Such patients usually have variable degrees of tricuspid regurgitation. Ebstein’s anomaly is frequently associated with an atrial septal defect and the Wolff-Parkinson-White syndrome.

### **When Chambers and Valves are Not in Normal Sequence or Relationship**

Some of the most complex anatomic defects obviously occur when chambers and valves are not in their normal sequence or position. Fuller understanding of these types of anomaly is obtained with an appreciation of the vocabulary mentioned previously.

### **Anomalies of the Relationship Between Atria and Ventricles**

Anomalies of the relationship between the atria and the ventricles frequently result in the so-called univentricular hearts. Such hearts may have both atrioventricular valves entering a single ventricle or there may be an absent left or right atrioventricular connection.

#### **Double-inlet ventricles**

Both atria can enter the same ventricle through separate atrioventricular valves. Both atrioventricular valves can be dedicated to either the left or right ventricle. In most cases of double inlet ventricle, the ventricle receiving the flow from the atria is very large. The size of the other ventricle may be very small, or indeed, rudimentary. This is the common setting for so-called univentricular heart.

The term univentricular heart is somewhat misleading since the other ventricle is frequently present but rudimentary. Included under this heading is the heart previously described as single ventricle, primitive ventricle, and common ventricle, as well as the heart with tricuspid or mitral atresia when these are due to absence of the atrioventricular connection. The common feature of these conditions is that the atria communicate or are in potential communication with only one ventricular chamber. This chamber is usually of the left ventricular type, but may rarely have right ventricular or indeterminate morphology. When the main chamber is of left ventricular type, a rudimentary hypoplastic right ventricle is usually present; when the main chamber is of right ventricular type the small chamber has left ventricular morphology.

The two most common types of univentricular atrioventricular connection are double-inlet ventricle (both atrioventricular valves can be seen in one ventricular chamber) and absent right or left ventricular connection. **Fig. 31** demonstrates an apical four-chamber view from a patient with a double-inlet left ventricle with a single ventricle of left ventricular morphology. **Fig. 32** shows an anatomic specimen of a univentricular heart with absent right atrioventricular connection.

#### **Atrioventricular discordance**

Another anomaly of the relationship of structures at the atrioventricular junction occurs when the ventricles are reversed. In this condition, the flow from the right atrium enters a morphologic left ventricle and flow from the left atrium enters a morphologic right ventricle. Almost invariably the relationship between the ventricles and the great vessels is also discordant with the left ventricle giving rise to the pulmonary artery and the right ventricle giving rise to the aorta.

Thus, the left ventricle becomes the venous ventricle, receiving blood from the right atrium and pumping it out through the pulmonary artery. Likewise, the right ventricle becomes the systemic ventricle, receiving oxygenated blood from the left atrium and pumping it out through the aorta. In each case the sequence of blood remains proper, only the ventricles are switched.

This entity is readily diagnosed by two-dimensional echo. Normally, the tricuspid valve is closest to the apex of the heart and the mitral more distal. With atrioventricular discordance, the mitral valve is located on the venous side and is identified farthest from the apex, while the tricuspid side is in the systemic position but is closest to the apex. It is additionally helpful to note the presence of a normal moderator band located at the apex of the systemic (but right) ventricle (**Fig. 33**). In this case, recognition of ventricular morphology from the insertion of the atrioventricular valves on the septum is of value.

This entity is also known by many other names. Among them are: congenitally corrected transposition, ventricular inversion, and L-transposition. Whatever the name, the hemodynamics are normal except for the fact the right ventricle, being in the systemic position, must pump against systemic pressures. Other anomalies may be associated with this condition and consist of Ebstein's anomaly of the tricuspid valve (which leads to systemic atrioventricular valve regurgitation since it is located in the mitral position) and ventricular septal defect. Such patients may also have congenital heart block. Frequently patients present only as an echo oddity with no associated anomalies.

### **Anomalies of the Relationships Between the Ventricles and Great Vessels**

These anomalies may also be quite confusing and it is important to recognize that mastering and understanding the abnormal morphology is dependent on determining which ventricle is committed to which great vessel. Proper echocardiographic technique requires the examiner to trace the great vessels distally in all cases to determine which vessel bifurcates (the pulmonary artery) and which great vessel gives rise to an arch (the aorta). Then the examiner traces backward to determine to which ventricle each great vessel connects.

Such malformations are comprised of many entities. Among them are: tetralogy of Fallot, double-outlet right ventricle, double-outlet left ventricle, ventriculo-great arterial discordance (or transposition of the great vessels), and truncus arteriosus. Except for transposition of the great vessels, these lesions require that a subarterial ventricular septal defect be present.

### **Tetralogy of Fallot**

In Fallot's tetralogy, the aorta overrides the septum (**Fig. 34**). The aorta is, therefore, doubly committed to both ventricles to a variable degree. By definition, however, the aorta is committed to the left ventricle by at least 50 percent.

A narrowed right ventricular outflow tract is also seen. If a pulmonary valve is seen, pulmonary atresia is excluded, but in severe tetralogy this valve may easily be lost in the mass of echoes arising from the hypertrophied outflow tract.

**Fig. 35** shows a subcostal view of the right ventricle and right ventricular outflow tract in a patient suspected of having tetralogy of Fallot. The aorta arises partly from the right ventricle, and the right ventricular outflow tract is severely narrowed by both the infundibular septum (between the aorta and pulmonary artery) and thickening on the free wall of the right ventricle. In this patient, the areas of narrowing essentially formed two

chambers, one below and one above the area of narrowing to form a “double-chambered right ventricle”. While such discrete narrowing is uncommon, such an image does help to understand the location of obstruction in these patients.

Surgical correction of this anomaly requires closure of the ventricular septal defect. The mass of anomalous muscle in the right ventricular outflow tract must also be removed, and the right ventricular outflow tract opened, by placing a patch graft on the outer wall of the right ventricular outflow tract.

### **Double-outlet ventricle**

Double-outlet right ventricle may be thought of as a severe form of tetralogy of Fallot, except for the fact that most, if not all, of the aorta is committed to the right ventricle (**Fig. 36**). In this setting oxygenated blood crosses from the left ventricle to the aorta across the subarterial ventricular septal defect. Right ventricular outflow tract obstruction may or may not be present. The pulmonary artery also arises from the right ventricle.

Clinically, double-outlet right ventricle can mimic several other lesions. With pulmonary stenosis, an anterior pulmonary artery, and overriding of the aorta, the clinical findings indicate tetralogy of Fallot. Indeed, the dividing line between the two lesions can be difficult to determine. Absence of fibrous continuity between the mitral and posterior semilunar valves, once thought to be the definitive feature of the condition is only sometimes present.

The relationship of the great vessels may be normal or reversed. **Fig. 37** shows a subcostal view of a double-outlet right ventricle where the aorta and the pulmonary artery are reversed. Here, the aorta is anterior to the pulmonary artery. Less commonly encountered is a double-outlet left ventricle where both great vessels emerge from the left ventricle. A ventricular septal defect is usually seen in this entity.

### **Truncus arteriosus**

Truncus arteriosus is a general diagnostic term used when there is a common origin of the pulmonary artery and aorta from the ventricles. A ventricular septal defect is present. Thus, both ventricles have a single outlet. Blood flow to the lungs is supplied from any number of possibilities: the main pulmonary artery may arise directly from the aorta, main stem pulmonary arteries may arise from the sides or back of the aorta, or pulmonary blood flow is supplied only by collaterals. Obviously, prognosis in the latter case is dismal.

The echocardiographic features of persistent truncus arteriosus are remarkably similar to those of Fallot’s tetralogy despite the marked clinical differences between the two lesions. Absence of the right ventricular outflow tract and pulmonary valve are the essential features. It may also be possible to identify pulmonary arteries arising from the truncus, if present.

### **Ventriculo-great arterial discordance**

This entity is also known as transposition of the great vessels. In pure transposition, there are two atria, two atrioventricular valves and two ventricles – all positioned normally. The pulmonary artery, however, arises from the left ventricle and the aorta (with coronary arteries) arises from the right ventricle (**Fig. 2**). If there is no arterial or ventricular septal defect, the venous blood returns to the heart and immediately passes through the right ventricle into the aorta without passing through the lungs. Likewise, the oxygenated blood

returns to the left atrium and transits through the left ventricle to the pulmonary artery and back into the lungs.

Thus, venous and systemic circuits are entirely separate rather than in tandem. This situation is incompatible with life. In such a critical situation, a balloon-tipped catheter is passed across the atrial septum from right to left. The balloon is then inflated and pulled back across the atrial septum creating a large atrial septal defect where venous and oxygenated blood can mix. Although such a measure does not correct the problem, it can be life-saving until more definitive correction can be performed. In the presence of an atrial and/or ventricular septal defect, such emergent measures are usually not required.

The echocardiographic diagnosis of ventriculo-great arterial discordance is dependent on proper identification of the great vessels and their commitments. **Fig. 38** shows a parasternal long axis with the aortic arch seen arising from the anterior ventricle (right) and the pulmonary artery (characterized by its bifurcation) arising from the posterior ventricle (left). A modified parasternal short axis is shown in **Fig. 39** where the pulmonary bifurcation in the posterior great vessel is readily recognized.

**Fig. 40** shows inverted (but anatomically correct) images from the subcostal area in an infant with transposition. Posterior angulation of the transducer shows the left ventricle to connect to the pulmonary artery while anterior angulation shows the right ventricle to connect to the aorta.

Surgical correction of ventriculo-great arterial discordance is based on restoration of the proper sequence of blood flow. The atrial septum may be surgically removed and complex baffles may be placed in the atria that redirect flow. The baffles allow returning venous flow to pass through the mitral valve into the left ventricle and out the pulmonary artery to the lungs. Returning oxygenated blood then passes on another side of the baffles through the tricuspid valve into the right ventricle and out the aorta. Such an “atrial switch” is known as a Mustard or Senning procedure. The long-term problem with such procedures is that the right ventricle is forced to perform at systemic pressures for many years, resulting in dilatation and, ultimately, failure in many patients.

More recently, success has been achieved with a great arterial switch where the pulmonary artery and aorta are sectioned just above the valvular levels and surgically reconnected to the proper ventricles. The coronary arteries are also moved from the anterior great vessel to the newly created aorta. The advantage of this operation is that it recommitments the proper ventricle to the proper circuit. The disadvantage is that the initial operative risk is higher since the very small coronary arteries must be manually moved and reimplanted without jeopardizing coronary blood flow.

Ventriculo-great arterial discordance may also occur with right ventricular outflow tract obstruction. The presence of such obstruction complicates patient management and surgical correction.

One unusual disease of the coronary arteries, Kawasaki disease, results in inflammatory changes of the coronaries that cause dilatation of the vessels, or localized aneurysm formation and thrombosis. Such coronary abnormalities may be detected with two-dimensional echocardiography. **Fig. 41** shows serial short axes of the left main and left anterior descending coronary arteries with marked dilatation.

## Surgical Procedures for Congenital Heart Disease

The repair of congenital heart disease has three major goals. Most ideal is total correction of the given disorder. This results in the heart being anatomically normal in the sequence of blood flow. Obviously, it is not possible to reestablish such a normal sequence of chambers and valves for many forms of complex congenital heart disease, and partial correction may be the only achievable goal.

The third major intent of surgery is palliation. Palliative surgery does not correct the problem, but rather minimizes the problems that result from any given disorder. In some instances, palliation may be elected until the child becomes a candidate for total correction. A variety of different shunts are possible between the aorta and the pulmonary arteries to supply ample blood to the lungs for oxygenation. Although there is a strong tendency in many institutions to totally correct this problem in infancy, there are some situations where early total correction is not possible and palliative procedures must be performed.

Many procedures are listed under the proper name of the surgeon who initially devised the operative approach. **Table 2** has been included as a brief general reference for those unfamiliar with cardiac surgical approaches.

**TABLE 2**  
Common Surgical Procedures in Surgery for Congenital Heart Disease

<b>Procedure</b>	<b>Description</b>	<b>Intent</b>	<b>Result</b>
Blalock-Hanlen*	Surgical removal of the atrial septum	PAL	Increases mixing of blood
Blalock-Taussig	Subclavian artery to PA anastamosis	PAL	Increase PULM flow
Brock's*	Closed PVotomy and infundibulectomy	PAL	Increase PULM flow
Central Shunt	Conduit or anastamosis	PAL	Increase PULM flow
Damus-Kaye-Stansel	PA end to side anastamosis to AO, valved conduit between RV-MPA	COR	Increase flow to AO and PA when there is AO stenosis and two VENT. Reestablishes RV to PA continuity
Fontan	Anastamosis or conduit between RA and PA	PC	Increase PULM flow in cases of univentricular morphology or TA
Glenn's	SVC to PA anastamosis	PAL	Increase PULM flow
Great Arterial Switch	AO and PA moved to proper ventricles, coronaries reimplemented	COR	Creates normal relationship between the VENT and GA
Konno	Replacement of AV with AV annular	COR	Alleviates sub-AO obstruction and replaces abnormal

Mustard's	Atrial switch using intra-atrial baffle made of pericardium	COR	AV Reestablishes proper flow sequence to PA and AO in TGA
<b>Procedure</b>	<b>Description</b>	<b>Intent</b>	<b>Result</b>
Norwood	PA anastomosis to AO, conduit from AO to MPA	PAL	Increase flow to AO for sub AO obstruction and only one VENT
Park	Atrial septostomy with catheter blade	PAL	Increases mixing of blood to TGA
Patch	Closes an opening or surgical incision	COR	Closes a shunt
PDA Ligation	Ties off PDA	COR	Closes a shunt
Potts-Smith-Gibson*	Descend AO to PA shunt	PAL	Increases PULM flow
PA Band	Constrictive band around MPA	PAL	Decreases PULM flow
Rashkind	Atrial septostomy with catheter balloon	PAL	Increases mixing of blood for TGA or TA
Rastelli's	Valved conduit from RV to PA, VSD closure	COR	Increase PULM flow, may reestablish proper sequence of flow to AO and PA
Senning	Atrial switch using intra-atrial baffle made of atrial wall flaps	COR	Reestablishes proper flow sequence to PA and AO in TGA
Valvectomy	PV excision	PAL	Relieve PV obstruction
Valvotomy	Surgical opening of obstructed valve	COR	Open obstructed valve
Valve replacement	Replaces any valve	COR	Relieve obstruction or regurgitation
Valvuloplasty	Repair of any valve	COR	Relieve regurgitation
Waterston*	Ascending AO to RPA	PAL	Increase PULM flow

## Congenital Heart Disease (Figure Legends)

Fig. 1 Photograph of an echocardiographic examination of a neonate from the subcostal position.

Fig. 2 Discordant atrioventricular and ventriculo-arterial connections.

Fig. 3 Diagrammatic representation of the two types of tricuspid atresia.

Fig. 4 Diagrammatic representation of the four types of atrial septal defect.

Fig. 5 Above: Pathologic specimen of a secundum atrial septal defect cut in the four-chamber plane. (Courtesy of Professor R.H. Anderson, the National Heart and Lung Institute, London). Below: subcostal four-chamber view showing a secundum atrial septal defect. The defect lies in the central region of the atrial septum, and is separated from both the atrioventricular valves and the atrial roof by septal tissue.

Fig. 6 2-D subcostal four-chamber view showing a secundum atrial septal defect.

Fig. 7 2-D subcostal four-chamber view showing a primum atrial septal defect (arrow).

Fig. 8 2-D parasternal short-axis view of the left ventricle at the mitral valve level.

Fig. 9 2-D subcostal axis view with superior angulation showing a sinus venosus defect (arrow).

Fig. 10 2-D apical four-chamber view with saline contrast in the right side demonstrating a left-to-right shunt (left) (arrow). A right-to-left shunt following contrast saline injection is demonstrated (right) (arrow).

Fig. 11 2-D subcostal four-chamber view showing a hyper mobile interatrial septum. This septum is bulging toward the right atrium (left) (arrow) and toward the left atrium (right) (arrow).

Fig. 12 The three main stages of ventricular septal defects.

Fig. 13 2-D parasternal short-axis views of the left ventricle from two different patients. Both views demonstrate large midmuscular ventricular septal defects (arrows).

Fig. 14 2-D parasternal long-axis view of the left ventricle showing a muscular ventricular septal defect.

Fig. 15 2-D parasternal long-axis view of the left ventricle showing a subarterial ventricular septal defect (arrow).

Fig. 16 2-D parasternal long-axis view of the left ventricle showing a ventricular septal defect covered by an aneurysm.

Fig. 17 2-D parasternal short-axis view in two patients with ventricular septal defects. The echo on the right shows a perimembranous defect under the tricuspid valve while that on the left shows a subarterial outlet defect just below the pulmonic valve (arrows).

Fig. 18 Pathologic specimen of an atrioventricular septal defect cut in the four-chamber plane. The posterior bridging leaflet of the common atrioventricular valve is clearly seen. (Courtesy of Professor R.H. Anderson, the National Heart and Lung Institute, London).

Fig. 19 2-D parasternal four-chamber view from an infant with a complete atrioventricular septal defect (AVSD).

Fig. 20 2-D parasternal long-axis view in a patient with a complete AVSD during diastole (left) and systole (right). Note during systole the insertion of some of the chordae onto the crest of the ventricular septum (arrow).

Fig. 21 2-D subcostal short-axis view of the common atrioventricular valve orifice in a patient with a total atrioventricular septal defect (AVSD).

Fig. 22 2-D subcostal view of an absent right atrioventricular connection (tricuspid atresia).

Fig. 23 2-D apical four-chamber view from another patient with tricuspid atresia.

*Fig. 24 2-D apical four-chamber view in a patient with an absent left atrioventricular connection (mitral atresia).*

*Fig. 25 2-D parasternal left ventricular long-axis view in a patient with a subvalvular membrane preoperatively (left) and postoperatively (right) (arrow).*

*Fig. 26 2-D suprasternal view of the aortic arch in a patient with coarctation (arrow).*

*Fig. 27 Pathologic specimen of total anomalous pulmonary venous drainage (TAPVD). The heart has been removed to show the common pulmonary venous chamber (CPVC) which drains into vertical vein that terminates in the superior vena cava. (Courtesy of Professor R.H. Anderson, the National Heart and Lung Institute, London).*

*Fig. 28 2-D parasternal short-axis view from a patient with total anomalous pulmonary venous drainage (TAPVD). A vertical vein (VV) is seen posterior to the left atrium.*

*Fig. 29 2-D parasternal short-axis view in a patient with a persistent left superior vena cava (PLSVC) draining into the coronary sinus (arrow). A large ventricular septal defect (VSD) is also present.*

*Fig. 30 2-D apical four-chamber view in a patient with Ebstein's anomaly.*

*Fig. 31 2-D apical four-chamber view from a patient with a double-inlet left ventricle.*

*Fig. 32 Pathologic specimen of an absent right atrioventricular connection cut in the four-chamber plane.*

*Fig. 33 2-D apical four-chamber view in a patient with atrioventricular discordance (corrected transposition).*

*Fig. 34 2-D parasternal left ventricular long-axis view in a patient with tetralogy of Fallot.*

*Fig. 35 2-D subcostal view of the right ventricle and great vessels in a patient with a double-chamber right ventricle.*

*Fig. 36 2-D parasternal ventricular long-axis view in a patient with a double-outlet right ventricle.*

*Fig. 37 2-D subcostal view of a double-outlet right ventricle with reversal of the great vessels.*

*Fig. 38 2-D parasternal left ventricular long-axis view with the aorta (Ao) arising from the right ventricle and the pulmonary artery (PA) arising from the left ventricle (transposition of the great vessels).*

*Fig. 39 2-D parasternal short-axis view modified so that the pulmonary artery bifurcation is easily seen. In this patient with transposition, the pulmonary artery (PA) is identified as the posterior vessel.*

*Fig. 40 Subcostal views orientated to correspond to those obtained by angiography, in a case of complete transposition of the great arteries. Above: the elongated left ventricle connects to the pulmonary artery, identified by its early bifurcation. Below: the triangular, rough-walled right ventricle joins to the nonbifurcating aorta.*

*Fig. 41 2-D parasternal short-axis view at the level of the aorta (left) showing marked dilatation of the coronary arteries. A dilated left anterior descending coronary (LAD) in cross section is shown (right) arrow.*