Congenital Cardiac Anatomy and Operative Correction

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Abstract

There are numerous congenital defects that may present with human hearts, and many typically require surgical intervention. The primary goal of this chapter is to briefly define such abnormalities and introduce the reader to the various classification schemes that have been used to describe their relative anatomical and functional features. The chapter will also highlight the more common surgical procedures utilized to treat congenital cardiac lesions.

Keywords

Septal defect • Aortopulmonary window defect • Coarctation of the aorta • Interrupted aortic arch • Tetralogy of Fallot • Atresia • Ebstein's anomaly • Transposition of the great vessels • Total anomalous pulmonary venous connection • Persistent truncus arteriosus • Cardiopulmonary bypass

10.1 Introduction

The spectrum of congenital heart disease is enormously diverse, yet affects a relatively small portion of the human population [1]. Disease severity ranges from benign to lethal, and many lesions require intervention to allow survival or enhance life expectancy. Several nomenclature classifications have evolved to describe abnormalities of the cardiovascular system, with the Van Praagh and Anderson/Edwards systems being the most prominent.

Richard Van Praagh presented a system based on the segmental anatomy of the developing heart. His generalized theory states that, by understanding the anatomical position of the cardiac segments, the majority of cardiac defects may be accurately described. The three segments which Van Praagh described consist of the atria, the ventricle, and the great vessels. These segments may be described by delineating their relative positional relationships. The *visceroatrial situs* is defined as the relative position of the right and left atria to the sidedness of the abdominal (visceral) organs; the term situs means position or location. The *bulb ventricular loop* is described as the orientation of the right and left ventricles to the great vessels (pulmonary artery and aorta). These segments are referenced in sequence, with each being designated by a letter. The connection of the visceral venous vessels (the superior and inferior vena cavae) and the atrial body is termed the visceroatrial situs and is described by the letters: S, *situs solitus*; I, *situs inversus*; and A, *situs ambiguous*. When the visceral situs is normal (*situs solitus*), the stomach and spleen lie to the left, the right lobe of the liver is larger than the left, and the appendix is right sided. In situs inversus, the position of the abdominal organs is reversed, with the stomach and spleen lying on the right and the dominant lobe of the liver to the left; the appendix and inferior vena cava are to the left, with the left lung being typically trilobed. Situs ambiguous describes a group of anomalies in

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which the dominant characteristic is a lack of visceral sidedness. The abdominal organs may be positioned to the anatomical left or right, often with the liver lying in the midline. A unique characteristic of these individuals is the abnormal existence or a complete lack of a spleen. In general, polysplenic patients tend to have all atrial and pulmonary structures consistent with left-sided morphology, while asplenic patients tend to be right-side dominant.

The orientation of the right or left ventricular mass is described by how the embryonic cardiac tube loops during its development. In this system, the terms "right" and "left" are used to refer to the specific morphology of the ventricular mass rather than their spatial arrangement. The anatomical right ventricle has a very trabeculated endocardium, while the left ventricular endocardium is smoother, with finer trabeculations. The rightward (normal) orientation is given the term "D" for *D-loop*. This indicates that the morphologic right ventricle is oriented to the right and anterior to the morphologic left ventricle. If the cardiac tube undergoes looping in the leftward direction, the segment is given the reference letter "L" for *L-looping*. In this situation, the morphologic right ventricle lies posterior and to the left of the morphologic left ventricle. The final segmental orientation described by Van Praagh deals with the relationship of the great vessels and semilunar valves to the ventricles. The aorta is normally committed to the morphologic left ventricle with the aortic valve located leftward and posterior to the pulmonary valve. This normal relationship is given the reference letter "S." When the great vessels are transposed, with the aortic valve being rightward and anterior to the pulmonary valve, the convention used is "D." With *D-transposed* great vessels, the aorta is committed to the morphologic right ventricle and the pulmonary artery and valve to the morphologic left ventricle. When the orientation of these great vessels and semilunar valves is normal, but the aorta is committed to the right ventricle and the pulmonary artery to the left ventricle, the term *L-transposed* great vessels is used.

Efforts are underway to establish a common language in describing congenital cardiac defects, which will hopefully facilitate the tracking of outcomes and applications of procedures [2]. This chapter approaches the classification of these congenital heart lesions with a segmental anatomical method. Congenital heart abnormalities can also be categorized by whether or not they cause cyanosis (Table 10.1). Furthermore, cyanotic lesions refer to cardiac defects that typically result in systemic arterial desaturation. Finally, shunts refer to an anomalous pathway of blood flow resulting from anatomical defects and their relative resultant downstream pressures.

In the relative short history of surgical treatment for congenital heart defects, there has not been a more important technological breakthrough than the heart-lung bypass machine. Understanding of the various methods of its use is instrumental in understanding the logistics of surgical treatment of congenital heart disease.

Table 10.1 Anatomical lesions of the heart based on physiologic derangement

Acyanotic
• Left-to-right shunts Atrial septal defects Ventricular septal defects Atrioventricular septal defects Aortopulmonary window
• Left-sided obstructive lesions Aortic coarctation Congenital aortic stenosis Interrupted aortic arch
Cyanotic
• Right-to-left shunts Tetralogy of Fallot Pulmonary stenosis Pulmonary atresia – With intact ventricular septum – With ventricular septal defect Tricuspid atresia Ebstein's anomaly
• Complex mixing defects Transposition of the great vessels Total anomalous pulmonary venous connection Truncus arteriosus Hypoplastic left heart syndrome

10.2 Cardiopulmonary Bypass

Cardiac function is critical to the maintenance of ongoing hemostasis for any organism. Often, repair of congenital heart lesions requires interruption of this function for brief periods of time. The cardiopulmonary bypass circuit provides systemic support during these periods of cardiac arrest. A standard approach in cardiopulmonary bypass is draining blood from the right atrium and delivering it to the aorta past the aortic valve, with blood being oxygenated within the bypass machine. Blood not actively captured in the right atrium is sent through the lungs and out of the left ventricle when the heart is still beating. By directing the venous drains into the superior and inferior cavae and then snaring down on the cannulas, the right atrium can be safely entered as long as there is no communication with the left side of the heart (atrial septal defect, ventricular septal defect, etc.). Some lesions on the right side of the heart can therefore be fixed while the heart is still beating.

Alternatively, lesions that communicate with or are within the left side of the heart need an alternative strategy to prevent air being passed into the cranial vessels. To isolate the left heart, a clamp is placed between the aortic valve and the outflow of the cardiopulmonary bypass cannula. The heart is placed in a metabolically protected state by both physically cooling the heart and chemically inactivating it with a solution of cardioplegia. The length of time that the clamp can stay on depends on the solution used and the baseline function of the heart.

A third method in operating on the heart is to metabolically inactivate the entire body through cooling, therefore not needing any cardiopulmonary function for a short period of time. Cardioplegic arrest is used in procedures where maintaining the outflow of the bypass machine is not possible. In this method, the bypass machine is used to cool the entire body to a level where the period of decreased flow will not lead to permanent damage. The outflow of the bypass machine is turned off once the body has been sufficiently cooled and work is done expeditiously. Through these various bypass methods, the many varied congenital heart defects can be surgically treated. The remaining portions of the chapter will describe treatment methods used for specific lesions. For additional information, see Chap. 33.

10.3 Systemic Venous Anomalies

Persistence of the left-sided superior vena cava (LSVC) in isolation can be a variant of normal, when it drains to the coronary sinus, which is usually of no physiologic consequence. When an LSVC drains directly to the left atrium, or to an unroofed coronary sinus, systemic venous blood mixes with the pulmonary venous return, causing systemic desaturation. A persistent LSVC may herald additional anatomical issues or be part of a complex of lesions seen in heterotaxy syndromes. When an LSVC is paired with congenital heart lesions, this may alter the repair and must be identified and taken into consideration in preoperative planning [3].

The term interrupted inferior vena cava (IVC) describes the absence of the intrahepatic IVC, and the IVC blood is diverted to the superior vena cava (SVC) via the azygos vein. This finding is of physiologic significance in patients who need to undergo single ventricle palliative procedures in which the SVC blood is diverted to the pulmonary arteries (cavo-pulmonary or Glenn anastomosis), followed years later by diversion of the IVC blood to the pulmonary arteries (Fontan procedure). In this situation, the large majority of the systemic venous blood will be channeled to the pulmonary arteries with the Glenn anastomosis, which is not well tolerated at a young age, and different strategies must be employed. In the setting of an interrupted IVC, a full evaluation for other indicators of a heterotaxy syndrome must be performed [3].

10.4 Atrial Septal Defects

Fetal circulation requires the presence of a patent foramen ovale (PFO), or communication between the right and left atria, for the more highly saturated umbilical venous blood to bypass the lungs and be directed to the brain. After birth, pulmonary blood flow increases, closing the flap of the foramen ovale in approximately 75 % of all individuals, with the remainder maintaining at least probe patency of this communication [4]. This small communication is most commonly of no hemodynamic significance in early life.

Secundum-type atrial septal defects (ASDs) are due to deficiencies in the septum primum portion of the atrial septum, which allows a portion of the pulmonary venous return to cross the atrial septum into the right atrium, then into right ventricle, and finally recirculate to the lungs. When large enough, this leads to dilation of the right atrium and ventricle, and over time, this overcirculation of the lungs can lead to permanent damage of the pulmonary arterial vasculature and/or lung disease [5]. Primum type ASDs will be discussed in the atrioventricular septal defect section. Sinus venosus-type ASDs include defects in the septum separating the cavae from the pulmonary venous return. This allows for drainage of the associated pulmonary venous return to the right atrium, with the same hemodynamic consequences as a secundum-type ASD. This will be further discussed in the section on pulmonary venous anomalies.

Coronary sinus ASDs present variably, including as a partial or completely unroofed coronary sinus, i.e., allowing desaturated blood to drain to the left atrium and left atrial blood to cross the os of the coronary sinus into the right atrium. This can associated with a persistent LSVC (effectively draining directly to the left atrium), with additional atrial septal communications, or with atrioventricular valve abnormalities [6]. The physiologic consequences of this communication and shunting are similar to a secundum-type ASD (Fig. 10.1).

The correction of an ASD involves closing off the communication between the right and left atria. Correction is usually performed as a young child prior to long-term sequelae of a left-to-right shunt development. Surgical correction requires an arrested heart with a bicaval cannulation. As there are many types of ASDs, there are different surgical corrections. The most standard defect, the secundum defect, can be closed by simply closing the defect with suture or, if larger, using a patch material to close the defect. Traditionally the patient's own pericardial tissue has been used as patch material, although pericardium from animals or completely artificial materials such as GORE-TEX can be used. Closure of sinus venosus generally requires more extensive redirection of blood flow to allow proper drainage of the pulmonary veins to the left atrium. In these cases a patch must baffle the blood flow of the pulmonary veins to the left atrium. Pulmonary venous connection with the SVC requires disconnecting the portion of SVC communicating with the veins and using the right atrial appendage as a graft to the remaining SVC in a so-called *Warden procedure*.

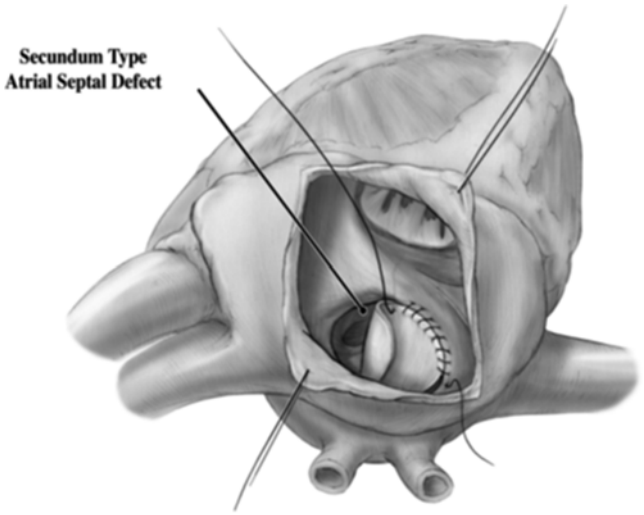

Fig. 10.1 Secundum-type atrial septal defect seen via an incision in the right atrial appendage

10.5 Anomalies of the Tricuspid Valve

Tricuspid atresia refers to a complete lack of communication between the right atrium and right ventricle and can present in multiple forms. The inlet portion of the right ventricle is underdeveloped, with the remainder of the ventricle dependent on the presence and size of a ventricular septal defect and the ventriculo-arterial connection. Table 10.2 describes the various subtypes of tricuspid atresia, organized by relationship of the great vessels, with type I referring to normally related great vessels, type II describing d-transposed great vessels, and type III as l-transposed great vessels; subtypes describe the amount of pulmonary stenosis (or atresia) [7].

Ebstein's anomaly is a spectrum of anomalies of both the tricuspid valve and right ventricle, with displacement of the tricuspid valve annulus toward the apex of the heart. The tricuspid valve is adherent to the right ventricle, prohibiting complete coaptation or closure of the tricuspid valve leaflets. This leads to regurgitation of the tricuspid valve and resultant dilation of the right atrium and ventricle. An ASD, pulmonary stenosis or atresia, and/or Wolf-Parkinson-White syndrome may often be associated with this anomaly [8]. In 1988, Carpentier and colleagues attempted to classify Ebstein's anomalies based on the relative volumes of the right ventricle. More specifically, these authors proposed four subtypes: (1) type A, the volume of the right ventricle is adequate; (2) type B, there is a large atrialized component of the right ventricle, but the anterior leaflet moves freely; (3) type C, the anterior leaflet is severely restrictive in its movement and may cause significant obstruction of the right ventricular outflow tract; and (4) type D in which there is almost complete atrialization of the ventricle, with the exception of a small infundibular component.

Table 10.2 Tricuspid atresia

Normally related great vessels
• Type I (a) pulmonary atresia
• Type I (b) pulmonary hypoplasia, small VSD
• Type I (c) no pulmonary hypoplasia, large VSD
D-transposed great vessels
• Type II (a) pulmonary atresia
• Type II (b) pulmonary or subpulmonary stenosis
• Type II (c) large pulmonary artery
L-transposed great vessels
• Type III (a) pulmonary or subpulmonary stenosis
• Type III (b) subaortic stenosis

VSD ventricular septal defect

Surgical repair of an Ebstein's anomaly is dependent on the degree and timing of presentation of tricuspid insufficiency. Neonates with severe tricuspid insufficiencies that inhibit forward flow through the right heart will need early interventions due to severe cyanosis; this approach typically leads neonates to a *Fontan circulation*. The first stage is performed as a neonate with an arrested heart. In addition to the creation of an aortopulmonary shunt, the tricuspid valve is patched closed, and the atrial septum is resected.

In patients that have less severe tricuspid regurgitation, repair is delayed until symptoms develop. Surgical goals are to form a well-functioning tricuspid valve and close any atrial septal communication. Many techniques have been developed, most of which involve plicating and/or resecting the atrialized portion of the right ventricle.

10.6 Ventricular Septal Defects

Ventricular septal defects (VSDs) are channels that permit interventricular shunting and can be found in isolation or in association with other congenital malformations. These channels may be described by their locations within the interventricular septum that is deficient, or by their anatomical nature [9], and also assigned a size relative to other anatomical structures of the heart (most often the aortic valve). The physiologic sequelae of VSDs depend on the size and location of the defects, with unrestricted VSDs leading to elevations of the right ventricular and pulmonary arterial pressures. Further, over time, the pulmonary vasculature develops muscular thickening which, if left unchecked, can lead to irreversibly high pulmonary vasculature resistance, or *Eisenmenger's syndrome*.

Redundant nomenclature exists for VSDs in the literature (Table 10.3). The intraventricular septum can be divided into four quadrants as seen in Fig. 10.2 [10] utilizing the *septomarginal trabecularis* as a landmark. For each region of the IVS, there are unique concerns and probabilities of spontaneous closure of a defect. Defects posterior to the posterior arm of the *septomarginal trabecularis*, inferior to

Table 10.3 Classification of ventricular septal defects

Type 1	Subarterial Supracristal Conal Infundibular
Type 2	Perimembranous Paramembranous Conoventricular
Type 3	Inlet Atrioventricular canal type
Type 4	Muscular • Anterior • Midventricular • Posterior • Apical

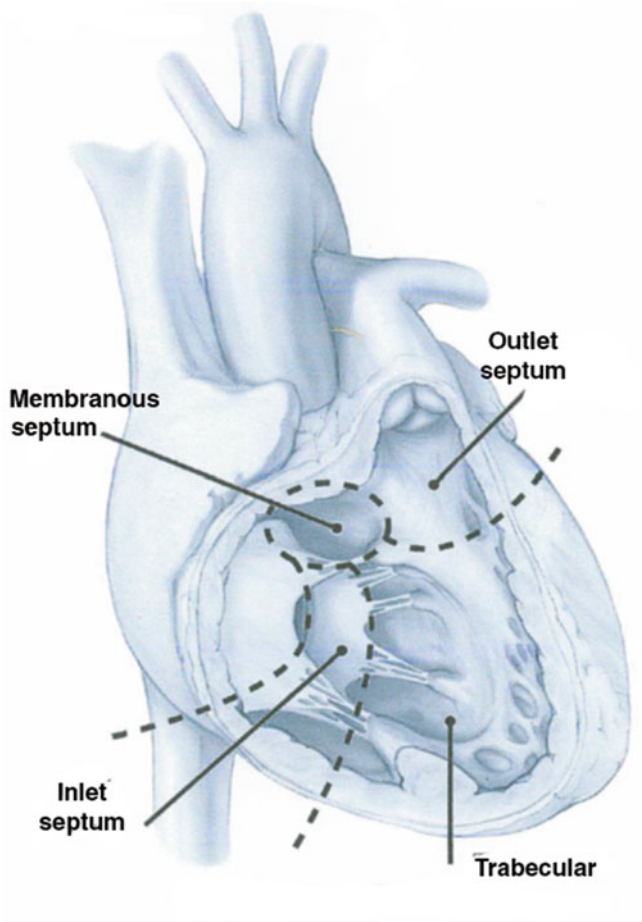

Fig. 10.2 Ventricular septum visualized through the right ventricular free wall. Note the three regions, including the inlet region supporting the tricuspid valve, the trabeculated muscular septum, and the outlet septum forming the pulmonary annulus

the atrioventricular valve, are termed inlet or atrioventricular canal-type VSDs and are further discussed in the atrioventricular septal defect section. Defects of the trabecular or muscular septum are the most likely to close spontaneously and can be subcategorized into anterior, posterior, mid-ventricular, or apical [10]. Those defects within or in close proximity to the membranous septum are termed perimembranous, paramembranous, or conoventricular VSDs. Defects in the outlet regions of the right ventricle (superior to the anterior portion of the septomarginal trabecularis) carry the names subarterial, supracristal, conal, or infundibular. These defects can lead to prolapse of the aortic valve and thus aortic regurgitation [11]. When closing these defects surgically or with a device, one must remain cognizant of the conductive tissues that pass in close proximity to many of these defects.

The correction of VSDs involves closing off the communications between the right and left ventricles. Corrections are usually performed in infants, as heart failure symptoms can develop that then, in turn, limit growth.

Surgical correction of VSDs typically requires an arrested heart with a bicaval cannulation. A correction almost always requires a patch, as the stresses on the repair site are much higher than that on an ASD (Fig. 10.3). The patch often will abut the conduction system which, if damaged, may lead to the lifelong need for an implanted pacemaker, i.e., secondary to third-degree heart block. The approach to a VSD repair depends on its specific location within the ventricular septum. Defects under the pulmonary valve can be approached through an incision in the right ventricular outflow tract. Muscular VSDs, although more likely to close on their own, can be more difficult to repair surgically due to their location within trabeculations of the heart. Surgical closure can often require a ventricular incision. Many muscular defects are now closed with an endovascular device either through a percutaneous or periventricular approach. The most commonly surgically closed VSD is a perimembranous defect and is approached through the right atrium.

10.7 Atrioventricular Septal Defect

Atrioventricular septal (canal type or endocardial cushion) defects (AVSDs) are a spectrum of defects that involve an ostium primum septal defect, the atrioventricular valves, and/or the inlet portions of the interventricular septum. Depending on the position of the atrioventricular valves, one ventricle may be favored over the other leading to an unbalanced size of the ventricles. The conduction system is often abnormal, with the usual pathways disturbed. A *complete* AVSD includes a large primum-type ASD, a common atrioventricular valve with varying number and distribution of leaflets, and an inlet (canal-type) VSD. A *transitional* AVSD has two separate atrioventricular valves, with an inlet VSD, and primum-type

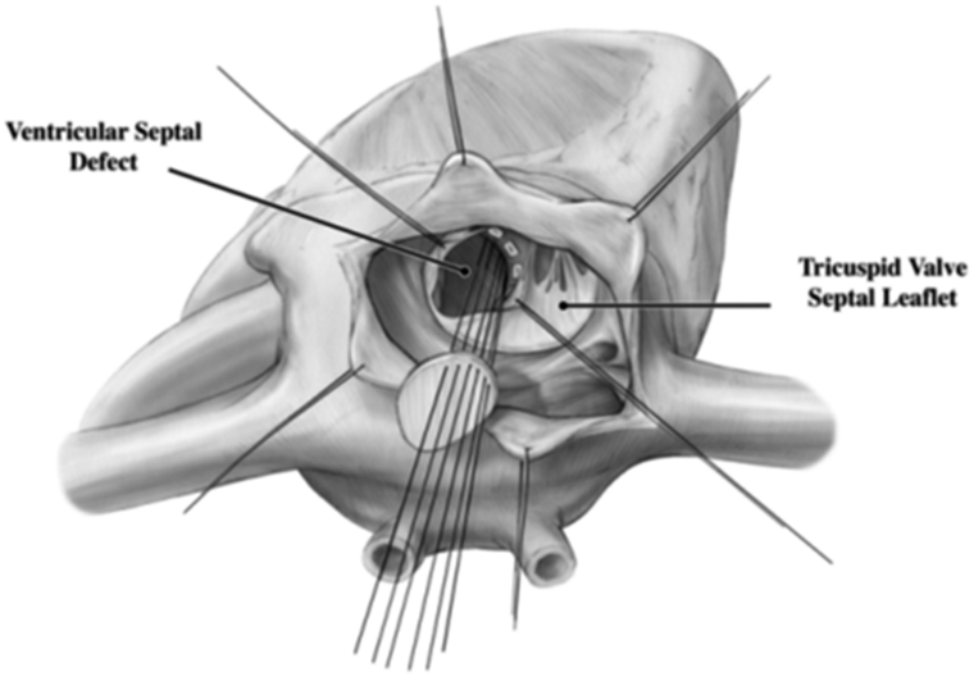

Fig. 10.3 Patch closure of a perimembranous septal defect working through the tricuspid valve

ASD. A *partial or incomplete* AVSD has a primum-type ASD and generally two separate atrioventricular valves, with a cleft in the anterior leaflet of the left-sided valve. An AVSD can be found in isolation, but there is a higher prevalence in trisomy 21, in heterotaxy syndrome with additional associated anomalies, or with variations including tetralogy of Fallot or double outlet right ventricle (DORV) [12].

Currently, repairs of these defects typically require bicaval cannulation and an arrested heart, and they are usually treated when the child is an infant. The goal of these repairs is to separate the ventricles and atrium into left and right sides; during this bipartition, the atrioventricular valves are reconstructed. There are many ways an atrioventricular canal can be repaired. In a traditional one patch repair, a single patch is first sewn to the crest of the VSD. The common atrioventricular valve is then divided into a left and right side. The two sides of the valve are then attached to the patch at its midpoint. The patch is brought up to the crest of the VSD completing the repair. Other groups have used two patches, one for the VSD and the atrioventricular valve and another for the ASD. Finally, a modified single patch technique uses a primary closure of the VSD with pledgeted suture; the sutures are then brought up through the atrioventricular valve and then through a patch which closes the ASD. Notably, the closure of the atrial and VSDs can lead to heart block requiring a permanent pacemaker.

10.8 Anomalies of the Great Arteries

10.8.1 Transposition of the Great Arteries

Transposition of the great arteries refers to the abnormal relationship and position of the great vessels at the base of the heart. Normally the pulmonary artery arises from the right ventricle, anterior to the aorta; the aorta arises from the left ventricle. In congenitally corrected transposition of the great arteries (cc-TGA), the aorta is anterior and leftward of the pulmonary artery and often arises from the morphologic right ventricle situated as the leftward ventricle (ventricular inversion). If there are no associated additional cardiac defects, this is often not found until later in life (if ever), as there are no physiologic embarrassments, outside of a right ventricle working as the systemic ventricle.

In dextro-transposition of the great arteries (d-TGA), the aorta arises from the right ventricle and is anterior and rightward to the pulmonary artery. The pulmonary artery arises from the left ventricle. Without additional cardiac defects, all of the desaturated systemic venous blood returns to the body without being refreshed in the lungs, and all of the fully oxygenated pulmonary venous blood returns to the lungs, acting as two parallel circuits and never intermixing. Newborns are able to survive initially with mixture from a patent ductus arteriosus (PDA) and a PFO, but as the PDA closes, the infant becomes progressively more cyanotic. Most frequently in

these children, in the first few days after birth, a balloon atrial septostomy (Rashkind) procedure is performed to tear open the atrial septum, followed within several days by an arterial switch operation, where the main pulmonary artery and ascending aorta are transected and the coronary arteries are excised from the aorta and transferred to the new aorta (native pulmonary artery) along with the ascending aorta. The branch pulmonary arteries are stretched over top of the newly reconstructed ascending aorta with the *LeCompte maneuver*, and the atrial septum is closed. This procedure restores the flow of desaturated blood to the lungs and the flow of fully saturated blood to the body and coronary arteries [13].

Transposition of the great arteries is now normally repaired shortly after birth. Repair is performed most often with an arrested heart and bicaval cannulation. Technically much of the repair depends on the location and insertion of the coronary arteries. Problems with the dissection and implantation of the coronaries lead to the majority of the mortalities associated with these procedures. In order to align the coronaries, often a LeCompte procedure is performed which involves bringing the pulmonary arteries anterior to the reconstructed aorta. Closure of the ASD finishes the repair.

10.8.2 Persistent Truncus Arteriosus

Persistent truncus arteriosus is the lack of separation of the pulmonary trunk and aorta, resulting in a common semilunar valve and artery arising from the ventricles, overriding an associated VSD. This common (truncal) artery supplies the coronary arteries, lungs, and aorta and was initially described by Collett and Edwards [14]; using the origin of the pulmonary blood supply, they differentiated subtypes (Fig. 10.4). In this categorization, type I refers to early bifurcation of the aorta and the pulmonary trunk from the arterial trunk. In type II, the branch pulmonary arteries arise in close proximity to each other but separately from the posterior aspect of the truncal artery. In type III, there is more separation of the origins of the branch pulmonary arteries. Type IV (also known as pseudotruncus) refers to the complete blood supply of the lungs from aortopulmonary collateral vessels, though such a defect can be argued to be pulmonary atresia with VSD. The Van Praagh classification system is also shown in Fig. 10.4 and varies slightly from the Collett/Edwards classification [15]. The anatomy of truncus arteriosus allows for the complete mixture of saturated and desaturated blood in the truncal artery and thus to the body; this will result in early heart failure, due to the often unprotected pulmonary circulation. The common surgical approach for the correction of these lesions varies depending on type but most often includes creation of an RV to pulmonary artery conduit and closure of the VSD.

In general, repair of truncus arteriosus involves separating the pulmonary artery or branches of the pulmonary arteries from the root of the aorta. Such surgical repair requires cardiopulmonary bypass with an arrested heart. The resulting aortic defect is normally patched. The VSD is normally closed through the pulmonary artery root or right ventricular

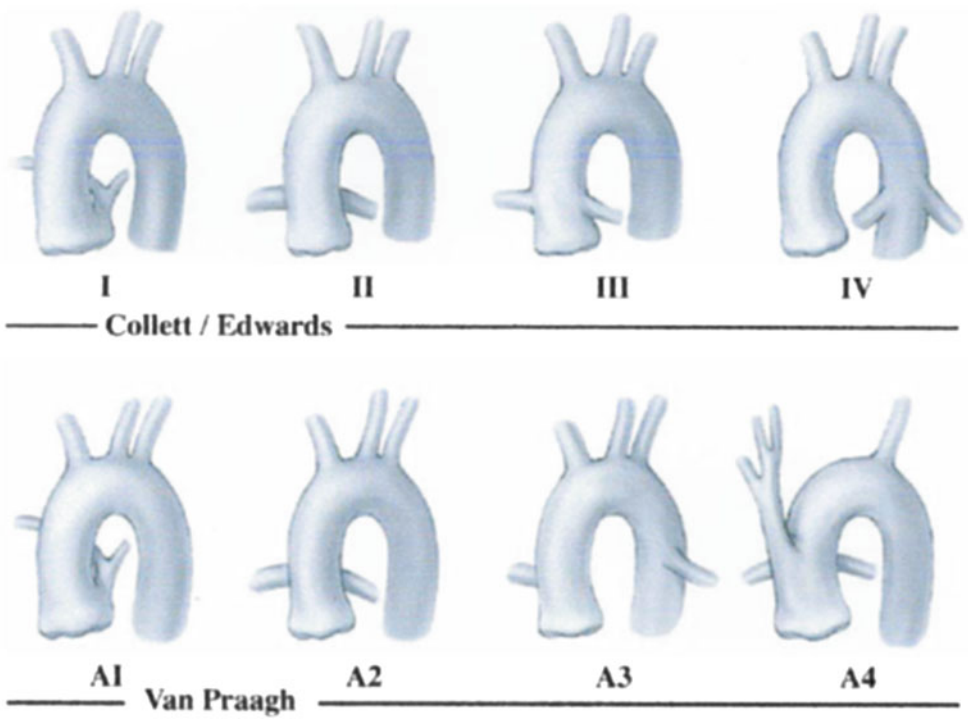

Fig. 10.4 Classification schemes for persistent truncus arteriosus

outflow; a valved conduit is used to create right ventricle to pulmonary artery continuity.

10.8.3 Aortopulmonary Window

An *aortopulmonary window* is the incomplete separation of the aorta and pulmonary trunk due to impaired or improper development of the truncal cushions during development (see also Chap. 3). In this case, there are two separate semilunar valves, and the communication can exist in the proximal portion of the aortopulmonary septum (type I), the distal septum (type II), or a combination of types I and II (also often noted as type III) [16, 17].

These defects are repaired again through the use of an arrested heart with either bicaval or single atrial cannulation. Isolation of the pulmonary arteries is important at the commencement of cardiopulmonary bypass so as not to create a circular circuit from the aortopulmonary-artery continuity. Once the pulmonary artery and aorta are separated, the resulting defects are closed usually with patches on both the pulmonary artery and aorta (Fig. 10.5).

10.8.4 Coarctation of Aorta

Coarctation of the aorta is a narrowing of the blood flow through the descending thoracic aorta in both varying degrees and locations. In the early 1990s, Amato and colleagues proposed a classification system based on the degree of aortic hypoplasia and the existences of associated cardiac lesions (Table 10.4). Type I describes obstruction in the juxta-ductal region, and type II is hypoplasia of the aortic isthmus in addition to type I narrowing. More specifically, type III includes severe (tubular) hypoplasia in much of the aortic arch, often including the distal transverse aorta [18]. During fetal life and immediately after birth, a PDA allows for blood to bypass the narrow areas and/or the right ventricle to supplement cardiac output to the lower half of the body. As the ductus arteriosus ampulla closes, the narrowing is worsened, and the infant will develop hypoperfusion of the lower body commensurate with the degree of narrowing.

Surgical repairs are normally performed on these patients as neonates, utilizing mild hypothermia without bypass. They are conducted through a posterolateral thoracotomy, which allows excellent exposure to the great vessels (Fig. 10.6). In older children, left heart bypass can be employed, diverting blood from the left atrium to the distal aorta, allowing continuous perfusion to the lower body. Most often the aortic lesion is isolated between two vascular clamps and then repaired expeditiously.

Table 10.4 Classification of coarctation of the aorta

Type I	Primary coarctation
Type II	Coarctation with isthmus hypoplasia
Type III	Coarctation with tubular hypoplasia of the distal arch • Coarctation associated with ventricular septal defects • Coarctation associated with other major cardiac defect

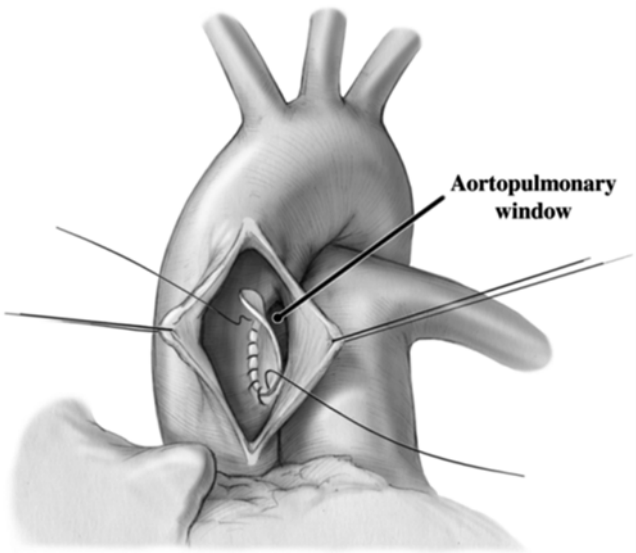

Fig. 10.5 An aortopulmonary window is created by a deficiency in the separation of the truncus. This defect is visualized through an incision in the ascending aorta and closed with a patch

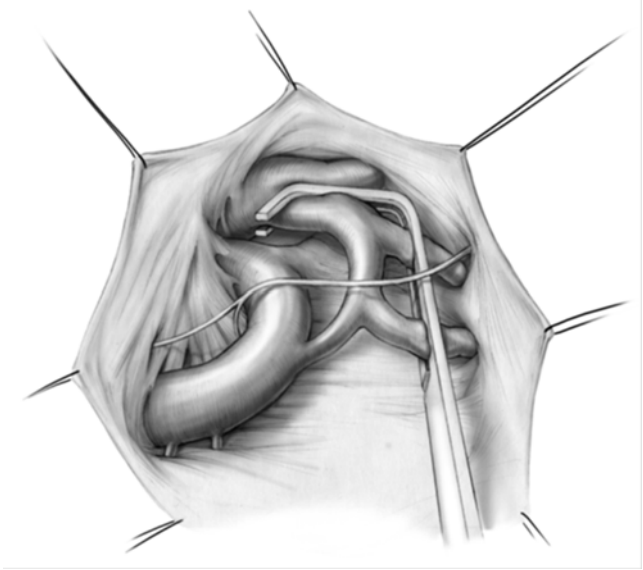

Fig. 10.6 Coarctation of the newborn is often associated with severe hypoplasia of the transverse aortic arch. A proximal vascular clamp has been placed in preparation for coarctation resection. Notice the recurrent laryngeal nerve rapping around the ductus arteriosus

Historically isolated lesions were opened by placing a patch of material across the narrowed segment. These defects now are most commonly repaired by resecting the narrowed sections and then performing end-to-end anastomoses. An alternative or additive option is using the subclavian artery as a patch of tissue to augment the narrowed section. Risks of these repairs include: damage to the recurrent laryngeal nerve, potential restenosis, scoliosis, and/or paralysis. Residual or recurrent narrowing of a coarcted site is normally treated by endovascular balloon arterioplasty.

10.8.5 Interrupted Aortic Arch

Interrupted aortic arch (IAA) is a congenital defect with a complete discontinuity in the transverse aortic arch, which may occur in the region of the aortic isthmus (type A), between the left common carotid artery and the left subclavian artery (type B, most common), or between the right innominate artery and the left common carotid artery (type C, least common), as described by Celoria and Patton [19]. Figure 10.7 demonstrates these different types, with a PDA supplying the vessels distal to the interruption, as well as the lower body. This defect is associated with a conoventricular septal defect with the conal septal tissue crowding the subaortic region. Surgical repair of this defect includes restoring continuity between the ascending and descending aorta and closure of the VSD.

Repair of IAA traditionally has been treated with the use of hypothermic arrest due to the inability to flow through an aortic cannula while working on the aortic reconstruction. A surgical alternative is to direct a lower volume of flow to the cranial circulation, through the innominate artery; some surgeons have included this low flow approach to the distal aorta as well. The interrupted segment can be reconstructed end to end, as in a coarctation repair or augmented with a patch. Occasionally an interposition graft is used to bridge the gap.

10.9 Pulmonary Atresia

Pulmonary atresia is the lack of blood flow across the pulmonic valve. This may happen in the setting of incomplete development of the pulmonic valve, or it may be functional atresia where the flow jet from a PDA holds the pulmonic valve shut. In such patients, the main and branch pulmonary arteries can be formed to varying degrees from normal size and branching pattern, to discontinuous pulmonary arteries with multiple collateral arteries supplying blood to the lungs [20]. Pulmonary atresia can also present either with a VSD or with an intact ventricular septum. When a large VSD is present, some argue it to be within the spectrum of a tetralogy of Fallot congenital abnormality (see below).

Patients with pulmonary atresia with an intact ventricular septum are associated with varying degrees of tricuspid valve hypoplasia, right ventricular hypoplasia, and/or fistulous ventriculocoronary connections. In this situation, there is often shunting of desaturated blood to the left side of the heart at the atrial level. Importantly, the relative extent of fistulous connections of the right ventricle to the coronary arteries must be understood prior to attempted repair or palliation, as the coronary blood flow may be dependent on the right ventricle.

10.10 Tetralogy of Fallot/Double Outlet Right Ventricle

Tetralogy of Fallot is a collection of lesions including: pulmonary stenosis (below, at, or above the level of the pulmonary valve), a large conoventricular VSD with anterior malalignment (override of the aorta), and right ventricular

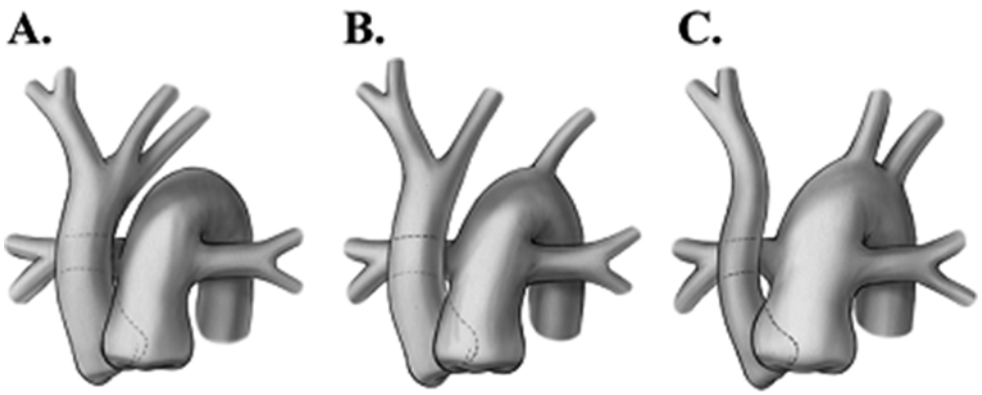

Fig. 10.7 Classification scheme for interrupted aortic arch. Type A occurs when all the arch vessels originate proximal to the interruption. Type B occurs when the left subclavian artery originates distal to the discontinuous segment

hypertrophy (Fig. 10.8). In DORV, both arterial trunks are dominantly associated with the right ventricular outflow tract [21]. The amount of right to left shunting of desaturated blood across the VSD depends on the degree of pulmonary outflow obstruction, which ranges from minimal stenosis to pulmonary atresia. Surgical repair includes closure of the VSD and relief of the pulmonary outflow obstruction. DORV can also be associated with malposition of the great arteries.

The timing of repair of tetralogy of Fallot is dependent on the severity of pulmonary arterial and/or infundibular narrowings. Severe narrowing as a neonate often requires augmentation of the pulmonary arterial flow with an aortopulmonary shunt. Most often the shunt is created with a GORE-TEX graft between the innominate artery and the pulmonary artery. Depending on pulmonary artery blood flow and hemodynamic stability, the procedure can be performed with or without cardiopulmonary bypass. Some groups are using a ductal stent to augment pulmonary arterial flow. Nevertheless, typically the full repair of tetralogy of Fallot requires an arrested heart with bicaval cannulation. The augmentation of the pulmonary arterial system can be either at the pulmonary valve or subvalvar level; repair of the VSD is performed with a patch.

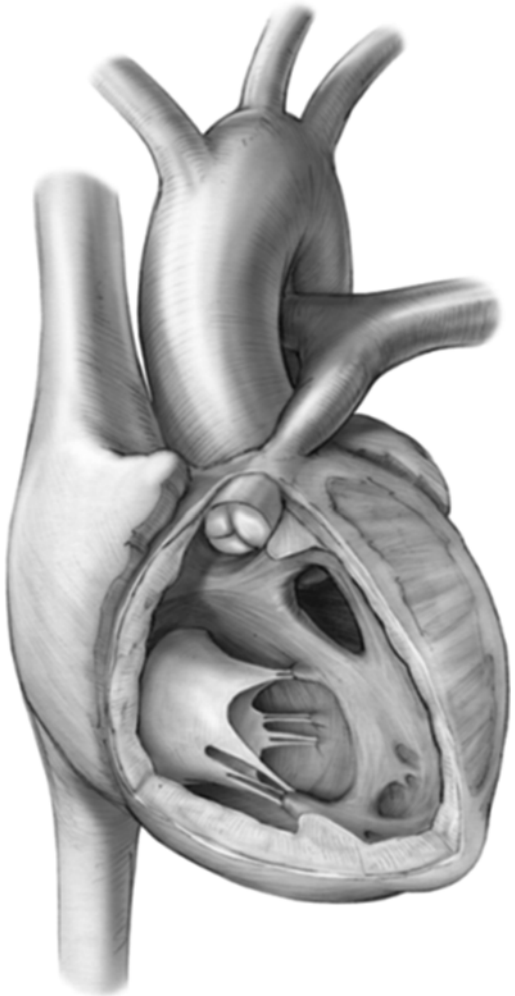

Fig. 10.8 Four components of tetralogy of Fallot include: pulmonary annular hypoplasia, a malaligned ventricular septal defect, right ventricular hypertrophy, and an overriding aortic annulus. The defect is viewed by removal of the right ventricular free wall

10.11 Pulmonary Venous Anomalies

Pulmonary venous anomalies are variable and include partial anomalous pulmonary venous connection (PAPVC) where some, but not all, of the pulmonary veins connect or drain to somewhere other than the left atrium. Total anomalous pulmonary venous connection (TAPVC) occurs when none of the pulmonary veins connect to the left atrium, or cor triatriatum, where the pulmonary venous return to the left atrium is separated by a wall of tissue (effectively forming three atria). TAPVC can connect to the right atrium or vessels above or below the heart or diaphragm, with the latter being more concerning for obstruction to blood flow. TAPVC can be classified into the following categories: (1) in type I, the pulmonary veins drain above the heart, to the SVC or innominate vein; (2) in type II, they connect directly to the heart in the right atrium or coronary sinus; (3) in type III, the connection is below the diaphragm; and (4) type IV is a mixture of several of the other types [22]. TAPVC is a mixing lesion and often presents with cyanosis reflective of the type and stability of the connections. Cor triatriatum can present with variable levels of obstructions of pulmonary venous return to the left atrium and additional connections to the heart and surrounding vessels. The physiologic consequences of these anomalies depend on the origin of the connections and any obstructions to blood flow. PAPVC is hemodynamically similar to an ASD and is often found later in life, if at all.

Repair of TAPVCs depends on the anatomical arrangement of either supracardiac-, intracardiac-, and/or infracardiac-type pulmonary venous connections. Infracardiac arrangements are most often associated with obstructions of pulmonary venous return; these can lead to pulmonary artery hypertension and cardiovascular collapse.

Repair of a TAPVC will necessitate cardiopulmonary bypass and an arrested heart. Adequate visualization often necessitates brief hypothermic arrest to allow precise anastomoses of the left atrium with the pulmonary venous confluences. Once anastomoses of the left atrium are complete, reconstruction of the atrial septum is performed, i.e., to enlarge the small left atrium.

10.12 Obstructive Left Heart Lesions

10.12.1 Mitral Valve Anomalies

Obstructive malformations of the mitral valve and its apparatus can include: (1) hypoplasia or atresia of the mitral valve, (2) abnormalities of the papillary muscles and chordae supporting the mitral valve leading to parachute or arcade mitral valve, and/or (3) left ventricular outflow tract obstructions [23]. As mitral obstruction worsens, left atrial pressure, and ultimately pulmonary arterial pressure, will increase.

10.12.2 Hypoplastic Left Ventricle

Hypoplastic left heart syndrome is characterized by varying degrees of underdevelopment of the left ventricle which can be due to a constellation of anomalies that lead to inadequate blood flow to the ascending aorta. For example, this abnormality may be secondary to mitral valve stenosis and/or atresia or aortic stenosis and/or atresia. In any case, these individuals are left with one functional ventricle (the right ventricle) and are dependent on a PDA and an atrial communication to supply saturated blood to the body. These patients typically require either cardiac transplantation or a series of palliative procedures to survive past early infancy [24].

10.12.3 Subaortic Ridge

A *subaortic ridge* is a defect with a fibromuscular ridge of tissue found below the aortic valve in the ventricular outflow tract, and it may restrict blood flow from the ventricle to the aorta. This can range from mild to severe and over time can lead to damage of the aortic valve from abnormally high turbulent blood flow. Further, in some cases, subaortic narrowing can be discrete or tunnellike and can be found in isolation or in association with other defects, primarily other left-sided obstructive lesions and VSDs [25].

10.12.4 Bicuspid Aortic Valve

A *bicuspid aortic valve* (BAV) is the most commonly presented congenital heart defect and generally consists of the lack of separation between two leaflets of the valve, effectively creating two leaflets rather than three. This leads to varying degrees of outflow obstruction and is often associated with regurgitation or leakage back across the valve back into the ventricle. Interestingly, mild cases may not be found until later in life, if ever, whereas severe cases need early intervention [26]. Note that a BAV may be found in isolation or in combination with other congenital cardiac malformations, especially coarctation of the aorta.

10.13 Coronary Artery Anomalies

Coronary artery anomalies may be found in isolation or in combination with other cardiac lesions. When the left coronary artery (LCA) arises, in part or in whole, from the right aortic sinus and travels between the aorta and pulmonary artery, there exists the potential for intermittent compression of the LCA, especially during exercise, thus resulting in inadequate blood supply to the myocardium. Multiple case series exist where this anatomy was thought to be the cause of sudden cardiac death [27]. Similarly, when the right coronary artery (RCA) originates from the left aortic sinus and courses between the aorta and pulmonary artery, a similar situation exists, especially in the setting of a right dominant coronary system; however, the actual risk to the individual remains debated in the literature.

In cases with an anomalous left coronary artery from the pulmonary artery (ALCAPA), the LCA arises from the pulmonary trunk, most often from the left sinus, or rarely from the pulmonary trunk or branch pulmonary arteries. In many patients, as the newborn pulmonary vascular resistance drops, the blood begins to flow from the coronary artery into the pulmonary artery causing a coronary steal phenomenon and resulting in underperfusion of the myocardium. These children may also present with: (1) depressed ventricular function and/or left ventricular dilation and (2) mitral valve regurgitation [28]. Surgical correction may include excision of the coronary arteries from the pulmonary artery (with a rim of surrounding tissue) and implantation onto the aorta or the creation of a tunnel to channel the blood away from the pulmonary artery.

10.14 Summary

There are numerous congenital defects that may present at birth within the human heart, and many typically require surgical intervention. In this chapter, we defined these abnormalities and introduced the reader to several classification schemes that have been used to describe their relative anatomical and functional features. This chapter also highlighted the more common surgical procedures utilized to treat congenital cardiac lesions. For additional technologies used to repair defects, the reader is referred to Chap. 37. Furthermore, Chap. 11 provides important information on mechanical circulatory support devices in the pediatric patient.

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