Tricuspid atresia can be considered the prototype of the univentricular atrioventricular (AV) connection with atresia of the tricuspid valve and hypoplasia of the right ventricle. It belongs to a class of congenital malformations with a univentricular AV connection, with absent AV connection and hypoplasia of one of the ventricles. In most patients with functionally single-ventricle hearts, there are morphologically two ventricles, one of them being too small to sustain one of the circulations. Different terminologies have been used to describe functionally single-ventricle arrangements, causing a lot of confusion. Terms like univentricular heart, single ventricle, and common ventricle have all been used to describe the same anomalies. These terms can cause confusion because from a pathologic point of view, human hearts with a solitary ventricular chamber are extremely rare. In most cases, there is a dominant ventricle with a second hypoplastic or rudimentary chamber. From the physiologic point of view, this does not really matter, as there is only one chamber capable of sustaining systemic and pulmonary circulation.

Because the terminology used to define univentricular AV connections has been somewhat confusing, it is difficult to state the actual incidence. In different series of congenital heart disease patients, they represent about 1% to 2% of the total. From a therapeutic point of view, patients with a functionally single-ventricle heart continue to represent a challenging group; management is often complex and needs to be adapted to the specific anatomic variability of the individual patient.

**ANATOMY AND PATHOLOGY**

The anatomy of complex congenital heart lesions is best approached in a systematic way. The segmental approach has generally been accepted as the standard method for analysis and categorization of congenital heart malformations. This method is based on description of the four primary segments of the heart (veins, atria, ventricles, and arterial trunks) and the way they are joined together at the venoatrial, AV, and ventriculooarterial junctions. Central to the process is the use of constant “markers,” which are constant morphologic characteristics of a certain segment. So, for instance, a constant feature of left atrial morphology is the left atrial appendage with the characteristic triangle shape. For each chamber, a certain number of unique “identifiers” have been established. Once the morphology of each chamber is defined, the connections can be studied. To precisely describe the type of univentricular AV connection, the atrial arrangement, the pattern of AV connection, the ventricular morphology, the ventriculooarterial connections, and the associated lesions should be accounted for.

The atrial topologic arrangement can be solitus (the usual pattern), inversus (mirror-image pattern), or ambiguous, in which there is atrial isomerism with bilateral right or left atrial morphology. Either type of AV connection can be associated with any of the four forms of atrial arrangement. The AV connection is, by definition, univentricular (Fig. 118.1), but there may be a double inlet, a common inlet, or an absent right or an absent left AV connection. In a double-inlet connection, there are two separate AV valves, and it is usually not possible to define the valves as either mitral or tricuspid valves on the basis of their morphologic characteristics. Therefore, it is better to refer to them as left and right AV valves on the basis of the anatomic position within
the chest. In case of an absent AV connection, the absent valve can be on the right side or on the left side. The remaining AV connection will usually be concordant. This describes the classic forms of “tricuspid” and “mitral” atresia. In both the double-inlet connection and the absent AV connection, the dominant ventricle can be right, left, or indeterminate. 

**Overriding** refers to the commitment of an AV valve annulus to the ventricular chamber and results from malalignment of the atrial and ventricular septa. The percentage of annular commitment to a ventricular chamber relative to the position of the ventricular septum determines the AV valve connection to a ventricle. In the setting of an overriding AV valve, an atrium is considered to connect to the ventricle into which more than 50% of its valve empties (Figs. 118.2 and 118.3). A univentricular AV connection requires more than 50% of both AV valves to be committed to one ventricular chamber. The remaining chamber is considered an outlet or rudimentary chamber.

The identification of the ventricular myocardial morphology should be based on the most constant component of the ventricular anatomy, which is the trabecular component. A normal ventricle can be described as having an inlet, a trabecular part, and an outlet part. An abnormal ventricle can sometimes lack both inlet and outlet. In those cases, the morphology of the ventricle can still be identified on the basis of the structure of the apical trabecular component. Left ventricular myocardium typically has a relatively smooth appearance with numerous fine oblique trabeculations (Fig. 118.4), whereas right ventricular myocardium has an irregular surface with relatively few coarse trabeculations. On the basis of this, most ventricles can be recognized as being morphologically right or left. In most hearts, such left and right ventricles coexist, with the inlets and outlets being unequally shared between the apical trabecular components in those hearts with abnormal segmental connections. In rare situations, there is only one ventricular chamber in the ventricular mass, with a ventricle that has a trabecular pattern typical of neither left nor right ventricle (indeterminate or mixed ventricular morphology). Also, the topology (the relationship between the two ventricles in space) of the ventricle should be determined. Chirality can be used to describe ventricular topology. A ventricle is called a right-handed ventricle if the palmar surface of the right hand can be placed on the ventricular septum such that the thumb is pointing to the inlet and the fingers are pointing to the outlet, with the wrist being in the apical component. In a normal heart, the right ventricle is right-handed, reflecting the normal embryologic development with a D ventricular loop; whereas in AV and ventricular arterial discordance (also called congenitally corrected transposition), the right ventricle has left-handed topology consistent with embryologic development with an L ventricular loop (see Fig. 118.3).

As a final part of the segmental approach, the ventricle–great artery connections should be defined. This connection can be concordant, with the pulmonary artery arising from the right ventricle and the aorta from the left ventricle, or discordant, with transposed great artery relationships. Double outlet (both great arteries arising from one ventricle) and single outlet (common arterial trunk or in pulmonary or aortic atresia) may also be present.

It is also important to describe associated anomalies: outflow obstructions like associated supravalvular, pulmonic, or supravalvular stenosis; subaortic or aortic valve stenosis; or associated coarctation of the aorta. Anomalies in pulmonary venous and systemic venous return commonly occur in the context of isomerism and are extremely important to identify in detail. Sidedness and branching pattern of the aorta and cranial vessels should also be recognized.

By use of the segmental approach, it is always possible to completely describe the anatomy of connections (Figs. 118.5 to 118.10).

**PATHOPHYSIOLOGY**

The pathophysiology of univentricular AV connection depends on the underlying anatomy and the associated lesions. In all lesions, there is a common mixing chamber in
Overriding atrioventricular valves.

In cases in which ductal patency can be prolonged with prostaglandins and afterward by ductal stenting if the arterial duct has favorable anatomy. Currently, more centers may approach this problem by a hybrid approach in the neonatal period by stenting of the aorta. Currently, more centers may approach this problem by a hybrid approach in the neonatal period by stenting of the arterial duct and bilateral banding of the pulmonary arteries.

Currently, more centers may approach this problem by a hybrid approach in the neonatal period by stenting of the arterial duct and bilateral banding of the pulmonary arteries. This will result in heart failure and pulmonary overflow. The pressures in the pulmonary artery will also be elevated, and obstructive pulmonary vascular disease is likely to develop if no surgical intervention is performed. In this case, the surgical strategy is to perform a banding of the pulmonary artery. Adequate banding is required to reduce pulmonary artery pressures and to prevent the development of pulmonary vascular disease. As discussed later, a low pulmonary vascular resistance is required for long-term palliation.

If there is obstruction to pulmonary flow caused by pulmonary stenosis or atresia or by high pulmonary vascular resistance, there will be desaturation and hypoxia. Early after birth, this may cause the pulmonary circulation to be duct dependent. In those cases, ductal patency can be prolonged with prostaglandins and afterward by ductal stenting if the arterial duct has favorable anatomy. In cases in which ductal stenting is not feasible, surgical palliation is used to increase pulmonary blood flow through different types of shunting operations. This can be either a modified Blalock-Taussig shunt or a central shunt. Shunting procedures improve the arterial oxygenation often at the expense of volume loading of the single ventricle. Because of this concern for ventricular volume loading, pulmonary blood flow may be altered to be more effective by use of a bidirectional cavopulmonary shunt, which can usually be performed at 3 to 4 months of age when the pulmonary vascular resistance has become normal. This shunt is also described in Chapter 113.

In some types of single ventricle, there may be obstruction of the outflow to the systemic circulation, which may potentially compromise systemic output and cause preferential flow to the pulmonary arteries. This can result in inadequate systemic perfusion, causing metabolic acidosis and shock. In those cases, the systemic circulation is duct dependent, and prostaglandin infusion is required in the neonatal period to stabilize those infants before surgery. The pressure load caused by the obstruction can lead to ventricular hypertrophy. As discussed later, the decreased compliance associated with hypertrophy causes unfavorable ventricular mechanics for surgical palliation later in life. Many of these hemodynamic variables are similar to those encountered in hypoplastic left heart syndrome. Different surgical options are possible to relieve the obstruction. In the past, one option has been the use of surgical procedures similar to those described in the Norwood procedure (aortopulmonary window, pulmonary artery detachment, aortic arch reconstruction, and shunting with a modified central shunt). More recently, the Sano operation has been performed to provide a more direct source of pulmonary blood supply. If there is subaortic obstruction within the heart, a Damus-Kaye-Stansel procedure is used to connect both outlets to the aorta. Currently, more centers may approach this problem by a hybrid approach in the neonatal period by stenting of the arterial duct and bilateral banding of the pulmonary arteries. This postpones other major surgery to later in life.

Figure 118.2 Overriding atrioventricular valves. The basic concept of univentricular AV connection necessarily includes a determination of the degree of AV connection based on the 50% rule. If 50% or more of an AV annulus overrides or is committed to a ventricle, the atrium is said to connect to that ventricle. Univentricular AV connection requires predominant connection to a single ventricular chamber. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

which systemic venous return and pulmonary venous return come together. This results in arterial desaturation, the level of which depends on the relative amount of oxygenated and desaturated blood mixing in the single ventricle, provided there is no significant systemic obstruction (coarctation or interrupted aortic arch). Because the systemic flow index is relatively similar in different lesions (between 2 and 3 L/min $\cdot$ m$^2$), the systemic saturation will depend on the amount of blood returning from the lungs. The output from a functional single ventricle is divided between the systemic and pulmonary circulations, so the relative proportion of blood flow to both circulations depends on the relative resistances to flow in both circuits.
Figure 118.3 Embryologic development of univentricular atrioventricular connection as a result of increasing override of an AV valve into the contralateral ventricle. The concept, proposed by Elliot and Gedgaudas, is applicable to virtually all forms of double-inlet ventricle whether the ventricular looping is right-handed or left-handed. LV, left ventricle; RV, right ventricle.

Figure 118.4 Comparative morphologic features of the right and left ventricular myocardium. Note the coarse trabeculations filling the inferior and apical portions of the right ventricle (RV) and the fine, delicate trabeculations and smooth wall of the left ventricle (LV). (Courtesy of William D. Edwards, MD.)

Figure 118.5 Single-inlet left ventricle with right AV valve atresia in AV concordance or classic tricuspid valve atresia. Note the atretic plate for the right AV valve (asterisk). IVC, inferior vena cava; LA, left atrium; LV, left ventricle; RA, right atrium.
CLINICAL PRESENTATIONS

The clinical features of univentricular AV connections are primarily determined by the presence or absence of pulmonary or systemic outflow obstruction. Patients without obstruction to the pulmonary artery blood flow present with signs and symptoms of other lesions with large left-to-right shunting (like large ventricular septal defects). The large pulmonary blood flow together with volume loading of the single ventricle causes congestive heart failure, which develops during the first 3 to 6 weeks of life, when pulmonary vascular resistance falls. The findings at that time are tachypnea, subcostal retraction, tachycardia, diaphoresis, failure to thrive, and hepatomegaly. If symptoms of congestive heart failure develop during the neonatal period, this is often secondary to associated lesions, such as AV valve abnormalities, obstruction to systemic outflow, or obstruction of pulmonary venous return (also causing more pronounced cyanosis). As pulmonary flow is high, the common mixing chamber will be associated with only mild cyanosis. A soft systolic ejection murmur secondary to flow-related relative pulmonary outflow obstruction or flow through the ventricular septal defect may often be heard. Because of the large pulmonary blood flow, a diastolic AV valve inflow murmur and a third heart sound may be present. In older patients with unobstructive pulmonary blood flow, pulmonary vascular obstructive disease may develop, resulting in progressive reduction in pulmonary blood flow and cyanosis. The second heart sound...
may then be loud and single. A diastolic murmur of pulmonary valve regurgitation may be present.

If pulmonary outflow obstruction or pulmonary atresia is present, hypoxemia and cyanosis are the presenting symptoms during the neonatal period. Patients with severe pulmonary stenosis or pulmonary atresia present with profound cyanosis and acidosis during the first days of life when the ductus spontaneously closes. A systolic thrill may be present with pulmonary outflow obstruction. A harsh systolic ejection murmur and single heart sound may be heard. The pulmonary valve closure sound may be audible with milder degrees of pulmonary stenosis. No murmur may be audible or there may be only a soft continuous murmur secondary to a patent ductus arteriosus or systemic-pulmonary collaterals if pulmonary atresia is present. In this case, the pulmonary circulation may be duct dependent.

If severe obstruction is present to systemic outflow (restrictive ventricular septal defect often associated with a hypoplastic aortic arch and coarctation), signs of severe organ hypoperfusion may be evident during the first days of life when the ductus arteriosus spontaneously closes. This may be manifested as metabolic acidemia, oliguria, cold-constricted extremities, and gut ischemia (necrotizing enterocolitis). Prompt initiation of intravenous prostaglandins is required to improve systemic cardiac output.

There may also be obstruction to the pulmonary venous return. In patients with left AV valve atresia and nearly intact interatrial septum, inadequate interatrial shunting can cause pulmonary venous hypertension because the outflow of the pulmonary venous return is severely obstructed. Symptoms may develop progressively during the first days of life as pulmonary arteriolar resistance falls and pulmonary blood flow increases. This “restrictive atrial septum” will be manifested with cyanosis and respiratory distress caused by pulmonary venous hypertension. In those cases, balloon atrial septostomy is required in the neonatal period. In rare cases, pulmonary venous hypertension may be caused by obstructed total abnormal pulmonary venous return. This is often associated with right atrial isomerism.

Figure 118.10 Common-inlet ventricle with ventricular inversion as observed in a left-handed ventricular loop.

In this example, the two ventricles are of nearly equal size. CA, common atrium; LV, left ventricle; RV, right ventricle.

DIAGNOSTIC TECHNIQUES

The complete evaluation of complex congenital heart disease depends on the association of different techniques that all provide complementary information. During recent years, echocardiography has become the cornerstone in establishing the diagnosis of univentricular AV connection. It allows definition of the abnormalities in great anatomic detail and provides valuable hemodynamic information. The input of the other diagnostic modalities is discussed.

Echocardiography

Two-dimensional Doppler echocardiography has become the most important tool to establish the diagnosis of univentricular AV connections. It allows definition of the anatomic details of the abnormal heart and the associated extracardiac abnormalities (Figs. 118.11 to 118.13). A complete description of the anatomically abnormal heart requires a careful segmental approach. First, the basic anatomy of atrial and visceral situs and location of the cardiac apex must be defined. For this purpose, subcostal views and abdominal scans are particularly useful. The same subcostal scans can be used to define the systemic and pulmonary venous connections to the atri. Abnormalities in systemic venous return, like bilateral superior caval veins or interruption of the inferior caval vein withazygos continuation, can be diagnosed echocardiographically. Abnormal pulmonary venous connections can also be identified. This is very important, especially if obstruction of pulmonary venous connection is present. Effort should be made to identify all four pulmonary veins because partial abnormal pulmonary venous connections can be present.

The diagnostic echocardiographic features of the univentricular AV connection are best viewed from an apical view delineating the crux of the heart, such as the apical four-chamber view. It can be used to describe the AV connection as being double inlet, atresia of one of the inlets, or common AV valve. In addition, parasternal and subcostal views are particularly helpful to detail the morphology of the dominant ventricle (right, left, or indeterminate), the location of the rudimentary ventricles or outlet chambers (anterior or posterior, to the left or right), the location of the papillary muscles and chordal attachments within the ventricular chambers (straddling or overriding), the status of the AV valve leaflets, and the location and commitment of the great arteries and their relationships (concordant or discordant). From the suprasternal notch, the sidedness of the aortic arch and the presence and location of a patent ductus arteriosus can be imaged. During the two-dimensional scan, it is also important to assess the size of the interatrial and interventricular communications. A restrictive interatrial communication may produce pulmonary venous obstruction, especially in left AV valve atresia. In case of tricuspid atresia, this will result in reduced cardiac output. The size of the interatrial communication should be defined by two-dimensional echocardiography, and the degree of restriction should be assessed. The size of the interventricular communication should also be evaluated. The size of the intraventricular communication between the dominant and the rudimentary ventricle can be or can become restrictive. If the aorta is originating from a small outlet right ventricle, as in double-inlet left
ventricle or right AV valve atresia with transposition, a restrictive ventricular septal defect can cause severe systemic outlet obstruction. Especially the initial size of the defect is predictive of subsequent development of subaortic stenosis. Patients with an initial ventricular septal area of 2 cm²/m² or less are at high risk of having associated coarctation of the aorta or even interruption of the aortic arch. They are also at risk for subsequent development of subaortic stenosis.²

Color-flow imaging is particularly helpful for determination of the exact location of pulmonary or systemic outflow tract obstruction. Subvalvular, valvular, or supravalvular stenosis can be defined. Ductal, interventricular, and interatrial shunting can also be evaluated by color-flow imaging.
Both the location and the degree of shunting can be viewed. Finally, color-flow imaging is extremely helpful in assessing AV valve function; stenosis and AV valve regurgitation can be recognized and evaluated with color-flow imaging. Pulsed and continuous Doppler study of the outflow tracts can be added to quantify the degree of outflow obstruction.

Two-dimensional echocardiography can also be used to assess global ventricular function. Wall motion abnormalities are common in single ventricle, limiting the usefulness of M-mode–derived measurements of shortening fraction or ejection fraction. Volumetric techniques such as biplane Simpson’s volumes can provide useful estimates of ventricular volume and systolic function (ejection fraction), but these techniques are all based on geometric assumptions of a normal left ventricle. This may limit their applicability to more complex single ventricles. Possibly the further development of three-dimensional echocardiography will provide better tools to evaluate systolic function of the single ventricle, but this needs further validation.13

Chest Radiography
In the era of modern ultrasonography, the diagnostic value of chest radiography in patients with univentricular AV connection is probably limited. The chest film can provide information on the position of the heart within the chest (levocardia, mesocardia, or dextrocardia). The sidedness of the stomach bubble also provides information about visceral situs. Further findings on chest radiography are variable and depend on the anatomic type of single ventricle as well as on the presence or absence of pulmonary or systemic outflow obstruction. When pulmonary blood flow is unobstructed, there will be cardiomegaly with increased pulmonary vascular markings. When pulmonary flow is moderately obstructed, there may be a normal heart size and normal or slightly decreased pulmonary vascular markings. When there is severe obstruction of the pulmonary outflow or pulmonary atresia, pulmonary vascular markings are reduced or asymmetric because of systemic–pulmonary artery collaterals.

If pulmonary venous obstruction is present, the lung fields may show a reticular pattern characteristic of pulmonary venous engorgement, although associated pulmonary outflow obstruction may conceal this typical picture.

Electrocardiography
An electrocardiogram will give important information about the cardiac rhythm and AV conduction.14 In most patients, the rhythm is sinus, except in case of isomerism, when junctional or left atrial type of rhythm may be present. The PR interval is normal, although first-degree AV block has been reported in up to 30% of all cases. The frontal axis of the QRS vector is variable. Most cases with a common AV valve of the AV septal defect type have left axis deviation (−40 to 120 degrees) as commonly observed in the biventricular forms of AV septal defects. Left axis deviation, however, can also be present in other forms of single ventricle, especially right AV valve atresia (tricuspid atresia). Thus, the status of AV valves cannot be predicted from the frontal QRS axis. Depending on ventricular morphology, different patterns of right, left, and biventricular hypertrophy can be present on the electrocardiogram. The ventricular morphology (right or left) cannot be predicted on the basis of electrocardiographic characteristics.

Magnetic Resonance Imaging
In the management and preparation of patients for the different stages of palliation, magnetic resonance imaging has taken on an important role in different centers. In most infants, a detailed echocardiographic examination allows the cardiac malformation to be reliably diagnosed, and only for interventional reasons like a balloon septostomy or ductal stenting is a cardiac catheterization still required. If there is some doubt about certain connections or pulmonary blood supply, magnetic resonance imaging provides a good alternative for better description of the morphology of complex congenital heart disease before consideration and planning of a cardiac catheterization. Magnetic resonance imaging gives information not only on morphology but also on blood flow and ventricular function. Three-dimensional reconstruction techniques with use of magnetic resonance imaging have now become the “gold standard” for volume and mass assessment of unusually shaped ventricles.15 Magnetic resonance tagging techniques also allow quantification of regional myocardial mechanics, although the clinical value of this experimental technique is still uncertain.16 More and more, cardiac magnetic resonance imaging is replacing routine cardiac catheterization. A recent prospective randomized trial has shown that in the preoperative evaluation before a bidirectional Glenn shunt, cardiac magnetic resonance imaging can safely replace routine cardiac catheterization.17 The same might be true in the preoperative evaluation for the total cavopulmonary connection.18

Cardiac Catheterization and Angiography
Because of the development of the other imaging techniques, the role of cardiac catheterization in the evaluation of univentricular AV connection has changed importantly. As the anatomy will in most cases be completely described by echocardiography or by cardiac magnetic resonance imaging, the current indications for a diagnostic cardiac catheterization are limited. Only if certain clinically relevant information cannot be reliably obtained by the echocardiographic examination or one of the other imaging modalities should a catheterization be considered. Currently, the majority of children with functionally single ventricles will undergo the first operation (like banding of the pulmonary artery, a modified Blalock-Taussig shunt, or even a Norwood-type repair) and even subsequent palliative procedures without a cardiac catheterization. If children are referred for cardiac catheterization, the indication will be interventional in most cases.

A possible indication for catheterization is the identification of systemic and pulmonary venous connections if there is any doubt on echocardiography. In addition, catheterization may be performed to determine

- adequacy of the interatrial communication;
- AV connections;
- basic ventricular morphology, ventricular volume, and systolic function;
- ventricle–great artery connections;
- pulmonary artery size, pressure, and resistance;
- systemic circulation; and
- abnormal venous or aorta to pulmonary collaterals.

Adequacy of the Interatrial Communication
A large interatrial communication is often present; but in some cases, the interatrial communication may be restrictive and obstructing pulmonary or systemic venous drainage.
If necessary, atrial balloon or blade septostomy or stenting of the intra-atrial septum should be performed.

**AV Connections**
Although echocardiography will generally provide excellent demonstration of the status of the AV connections, angiography can demonstrate AV valve commitment, determine annulus size, and assess the presence and severity of AV valve regurgitation.

**Ventricular Morphology, Ventricular Volume, and Systolic Function**
The basic ventricular morphology, ventricular volume, and systolic function can be evaluated by angiography. Despite the irregular shape of the ventricle, a good estimate of the ejection fraction can be obtained from ventricular volume calculations. Ventricular diastolic function can be assessed by measuring end-diastolic volume, ventricular mass, and atrial filling pressures and end-diastolic pressures.

**Ventricle–Great Artery Connections**
By use of angiography, the connection of the great arteries to the ventricles can be assessed. The patency of the outflow tracts can also be evaluated by careful catheter pressure measurements and angiography (Fig. 118.14).

**Pulmonary Artery Size, Pressure, and Resistance**
This is probably still one of the very important indications for cardiac catheterization as pulmonary artery size and distribution are critical components in planning palliative surgical procedures. Central pulmonary artery size can be measured, and distortion or stenoses of the central or distal pulmonary arteries can be excluded. A pulmonary artery index was proposed by Nakata and associates, a normal value being more than 330 mm²/m². Previously placed systemic-to-pulmonary or cavopulmonary shunts must be angiographically demonstrated. Preoperative assessment should also include measurement of pulmonary artery pressure and calculation of the pulmonary arteriolar resistance. Mair and colleagues derived a Fontan index, which includes pulmonary arteriolar resistance ($R_{pa}$), ventricular end-diastolic pressure (VEDP), and systemic and pulmonary flow volumes (Fontan index = $R_{pa}$ + VEDP/Qs + Qp). Surgical mortality after a modified Fontan procedure was lower in patients with a Fontan index below 4. If any significant stenosis of the pulmonary arteries is present, balloon angioplasty or stent implantation can be considered.

**Systemic Circulation**
The aortic arch anatomy should clearly be defined by angiography. The location of the aortic arch and brachiocephalic branches, the presence of a ductus arteriosus, and systemic

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**Figure 118.14** Angiographic studies in the diagnosis of functionally single-ventricle hearts. **A**, Typical anteroposterior and lateral angiographic features of single-inlet left ventricle with right AV valve atresia. Note the hypoplastic stenotic subpulmonary right ventricle (arrow). **B**, Anteroposterior view of typical double-inlet left ventricle with a hypoplastic subaortic right ventricle (short arrow). A pulmonary artery band is also evident (long arrow). **C**, Anteroposterior view of common-inlet ventricle of right ventricular morphology. Note the large common AV valve orifice (arrow).
to pulmonary artery collaterals should all be determined. Coarctation of the aorta should be excluded because the increased afterload would be poorly tolerated after the modified Fontan procedure.

Abnormal Venovenous or Aorta to Pulmonary Collaterals

Venovenous collaterals can be present after a bidirectional cavopulmonary anastomosis. These often originate on the innominate vein and connect to the atrium or pulmonary veins. These collaterals can be closed by coiling or use of a vascular plug. Aorta to pulmonary artery collaterals can also be present, causing additional flow to the pulmonary arteries. If significant, they need to be closed before the Fontan operation.

MANAGEMENT

A normal cardiovascular system consists of a double pulmonary and systemic circuit, connected in series, powered by two ventricular pumps—the “right” and “left” heart. A functioning single ventricle has to maintain both the systemic and pulmonary circulations, which are not connected in series but in parallel (Fig. 118.15). Such a circuit has two major disadvantages: arterial desaturation, both at rest and increasing during exercise, and a chronic volume overload to the single ventricle. Chronic volume overload will in time impair ventricular function, causing from the third decade onward congestive heart failure and death. In 1971, Francis Fontan reported a new approach to the surgical palliation of these malformations, separating the systemic and pulmonary circulations. In a “Fontan circulation,” the systemic venous return

Figure 118.15 A, Schematic representation of the normal cardiovascular circulation. The pulmonary circulation (P) is connected in series with the systemic circulation (S). The right ventricle maintains the right atrial pressure lower than the left atrial pressure and provides enough energy to the blood to pass the pulmonary resistance. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RV, right ventricle. B, Schematic representation of a patient with a univentricular heart. The systemic (S) and pulmonary (P) circuits are connected in parallel, with a considerable volume overload to the single ventricle (V). The width of the line reflects the degree of volume load. There is complete admixture of systemic and pulmonary venous blood, causing arterial oxygen desaturation. C, Schematic representation of a Fontan circulation. The systemic (S) and pulmonary (P) circulations are connected in series. The right atrium (RA) or systemic veins are connected to the pulmonary artery (PA). The volume overload to the single ventricle is now less than expected for body surface area. In the absence of a fenestration, there is no more admixture of systemic and pulmonary venous blood, but the systemic venous pressure is markedly elevated.
is connected directly to the pulmonary arteries without the interposition of a ventricular pump (see Fig. 118.15). Advantages of a Fontan circuit are that the arterial saturations nearly normalize and that the ventricular volume overload is relieved. This occurs at the expense of a chronic increase in systemic venous pressures and a decreased cardiac output both at rest and during exercise. The most important determinant for cardiac output in this circulation is the transpulmonary gradient, which is mainly determined by pulmonary vascular resistance and atrial pressures. Since its original description, the Fontan circuit has known numerous modifications. Initially, surgeons used valves and created various connections between the right atrium and the pulmonary artery (anteroatrio-pulmonary connection, with or without inclusion of a small hypoplastic right ventricle; postero-atrio-pulmonary connection), with different materials (valved conduits, homografts, patches, direct anastomosis). The very high incidence of late reoperations, up to 40% in some series, did reflect the suboptimal design of the first Fontan circuits. These older circuits are no longer created and are considered obsolete; however, many patients still survive after this type of surgery. Therefore, it is important to know which type of surgery these older Fontan patients underwent before evaluating them. These initial connections were replaced by the total cavopulmonary connection. In this approach, both caval veins are connected to the pulmonary artery, bypassing not only the right ventricle but also the right atrium. The superior vena cava is connected directly to the pulmonary artery (cavopulmonary anastomosis, also referred to as bidirectional Glenn shunt). There are currently two variants to connect the inferior vena cava: the lateral tunnel and the extracardiac conduit. Introduced in the mid-1980s, the lateral tunnel is a prosthetic baffle in the atrium between the inferior vena cava and the pulmonary artery. Because of inclusion of native atrial tissue, this connection has growth potential and can therefore be created in children as young as 1 year. The disadvantage is that a minimal amount of atrial tissue remains exposed to high pressure, and there is a large suture line within the atrium that in time may be a substrate for atrial arrhythmia. Therefore, the extracardiac conduit was introduced in the 1990s; it is a tube graft between the transected inferior vena cava and the pulmonary artery. This circuit leaves the entire atrium at low pressure and avoids suture lines within the atrium. It can be performed without aortic cross-clamping or even cardiopulmonary bypass. As the extracardiac graft has no growth potential, the operation can be offered only to patients large enough to allow placement of an adult size conduit (16 to 18 mm). This influenced the current strategy to achieve the Fontan circulation and resulted in a staged approach whereby the Fontan operation is prepared through different palliative steps tailored to the patient’s individual needs.

The Staged Approach to an Extracardiac Fontan Circulation

During the first 2 to 3 months of life, it is not possible to create a Fontan circulation because of the neonatal increased pulmonary vascular resistance and the small size of the vessels. During this period, there is a progressive fall in pulmonary artery resistance, making a cavopulmonary connection possible after 3 to 4 months of life. Currently, a staged approach is preferred, connecting the superior and inferior caval veins during two different steps. Such staged approach allows the cardiac condition to be better optimized for the Fontan circulation and reduces the overall operative morbidity and mortality.

Initially, in the neonatal period, management must aim to provide unrestricted flow from the heart to the aorta (if required: coarctectomy, Damus-Kaye-Stansel procedure, Norwood repair), a well-balanced limited flow to the lungs (if required: pulmonary artery band, shunt [modified Blalock-Taussig shunt, central shunt, or stent in duct]), and unrestricted return of blood to the ventricle (if required: Rashkind balloon septostomy). The infant is then allowed to grow for several months. During this time, the heart is submitted to a chronic volume overload, which is good for development of the pulmonary vasculature; but if it is excessive, negative influences are exerted on ventricular function. The infant will have mild desaturation, inversely related to mild cardiac failure. At the age of 4 to 12 months, most centers will perform a cavopulmonary connection or bidirectional Glenn shunt. This includes anastomosis of the cardiac end of the superior vena cava to the ipsilateral pulmonary artery, leaving the pulmonary arteries confluent. Through this type of shunt, the systemic venous return from the upper part of the body is directed toward the pulmonary artery, bypassing the heart. This provides pulmonary blood flow while at the same time reducing the volume load on the single ventricle if other shunts are eliminated or reduced. The creation of this type of shunt is a preparation for the later Fontan procedure, in which the inferior vena cava flow is also redirected to the pulmonary artery. The disadvantage of the bidirectional cavopulmonary anastomosis is that when the pulmonary arteries are small at the time the shunt is created, the absence of pulsatile flow might provide insufficient growth stimulus for the pulmonary arteries. This can result in small pulmonary arteries at the time of Fontan completion, which can cause an unsatisfactory outcome. The bidirectional cavopulmonary shunt provides insufficient pulmonary blood flow for adequate palliation of the univentricular AV connection beyond 6 to 7 years of age. The presence of a bidirectional cavopulmonary shunt as the only source of pulmonary blood flow may also be a potent stimulus for the development of multiple systemic to pulmonary artery collateral vessels to provide more pulmonary flow. Unfortunately, these collaterals do not stimulate pulmonary artery growth and are hemodynamically unfavorable after a Fontan procedure.

Most patients will undergo Fontan completion at the age of 1 to 5 years, depending on preference of the center, growth of vascular structures, and cyanosis at rest and during exercise. The Fontan circuit is completed by connection of the inferior caval vein to the pulmonary artery, currently by putting in an extracardiac conduit or lateral tunnel. Some centers advocate the use of a residual fenestration between the conduit and the atrium as an intermediate step toward further Fontan completion. In some studies, it has been suggested that this further reduces early mortality and morbidity, especially in those patients who are considered to be at high risk for the Fontan procedure. The fenestration decompresses the systemic veins by allowing right-to-left
shunting and increases preload of the single ventricle. It thus increases cardiac output at the expense of some degree of desaturation. It is possibly helpful in adapting to the body to the new circulation. This fenestration can later be closed percutaneously by different devices.

In 1996, Hausdorf and colleagues23 introduced the concept of percutaneous Fontan completion. Instead of a classic bidirectional Glenn operation, a hemi-Fontan operation is performed. In this approach, the upper part of the transected superior vena cava is connected to the right pulmonary artery; at the same time, the lower part connecting to the right atrium is connected to the pulmonary artery, but this connection is sealed off by a fenestrated patch or banding. This connection from the atrium to the pulmonary artery is used later to connect the inferior caval vein to the pulmonary artery by putting in a covered stent, opening up the restrictive communication.

The overall early mortality after Fontan-type surgery has decreased significantly during the past 10 years. Comparing early mortality in a very early cohort of patients (operation between 1973 and 1986) with that in a more recent cohort (operation between 1987 and 1992) in the Mayo Clinic, early mortality decreased from 16% to 9%.24,25 This decline occurred despite increased anatomic complexity of patients. The very recent outcomes with the staged approach suggest a further reduction in Fontan mortality. d'Udekem and colleagues26 reported the Melbourne experience with a hospital mortality of 6% between 1980 and 1990 and a 0% mortality of the Fontan procedure after 1990. Risk factors for early mortality include a common AV valve, elevated mean preoperative pulmonary artery resistance, and ventricular dysfunction. Early problems after surgery include low output state, prolonged pleural effusions, and atrial arrhythmias.

PROGNOSIS

Since the Fontan operation was first introduced in 1973, it is still uncertain how this type of palliation affects the long-term survival of these patients. The data are encouraging, however, as survival into adulthood and even successful outcome of pregnancy have been reported.24,25 Current survival data show a 20-year survival of up to 84%, with a better survival for the more recent techniques (15-year survival of 94% for the lateral tunnel technique; only 81% for the atriopulmonary connection).26 Patients report a subjective improvement in their quality of life after the Fontan operation, and exercise tolerance improves compared with their preoperative condition. Most patients are reported to be in New York Heart Association functional class I and class II. Exercise capacity continues to be subnormal in most patients, as indicated by a decreased maximal oxygen uptake and a decreased ventilatory anaerobic threshold. Exercise tolerance remains relatively stable during the range of 13 years after the Fontan operation. It seems, however, that poor functional outcome becomes more frequent with increasing duration of follow-up. Possibly, failure of the Fontan circulation related to ventricular dysfunction and the effects of chronically elevated systemic venous pressures may become a common problem beyond the third to fourth decade of life. One of the major concerns is the long-term preservation of single ventricular function. Improved management early in life aimed at maximal preservation of ventricular function by reducing volume overload and hypoxia and optimizing the pulmonary circulation by preventing pulmonary vascular disease and pulmonary hypoplasia and distortion could improve the long-term functional outcome of the Fontan operation.

Long-term problems include ventricular dysfunction, AV valve insufficiency, atrial arrhythmia, thromboembolic complications, and protein-losing enteropathy. Often, but not always, these complications appear to be secondary to excessively elevated right atrial and systemic venous pressures and chronic reduced cardiac output. This can be caused by mechanical problems in the Fontan connections, like obstruction to the atriopulmonary or cavopulmonary connections, pulmonary artery distortion or hypoplasia, pulmonary vascular disease, pulmonary venous obstruction, significant AV or semilunar valve regurgitation, and residual left-to-right shunts. Especially the occurrence of protein-losing enteropathy is a bothersome problem that often is very difficult to treat.27 It occurs in about 1% to 5% of all Fontan patients and is associated with a very high morbidity and mortality.

The Failing Fontan Circulation

A Fontan circulation may fail, causing persistent systemic venous congestion with edema, low cardiac output, very limited exercise tolerance, tachycardia, protein-losing enteropathy, or plastic bronchitis. Immediate complete evaluation of these patients including cardiac catheterization is required. Underlying causes, such as conduit stenosis, stenosis of the pulmonary arteries, thrombosis in the conduit, extensive aorta-pulmonary collaterals, severe AV valve insufficiency, increased pulmonary vascular resistance, and ventricular dysfunction, need to be identified. Prevention of these problems is important in the preparation before the Fontan operation and also in the management afterward. Depending on the underlying cause, a treatment strategy can be developed. In some patients, no good underlying cause can be found. Classic heart failure treatment consisting of inotropes, vasodilators, and diuretics will usually show little effect. If possible, optimizing the circuit can improve the patient’s condition. This can consist of dilatation or stenting of stenosis, embolization of collaterals, and conversion of older circuits to total cavopulmonary connection. For those patients in whom ventricular failure is the main problem, cardiac transplantation is often the only alternative. This can be performed with good results even for patients with complex anatomy.28 Another important problem that might develop is a small but progressive rise in pulmonary vascular resistance by microthrombi or endothelial dysfunction in the pulmonary circulation. This often becomes obvious only after cardiac transplantation,29 as the evaluation of pulmonary vascular resistance in this nonpulsatile lung perfusion is not evident.

SUMMARY

In patients with univentricular AV connection, the segmental approach should be used to give a complete description of the anatomy. The pathophysiology depends on the underlying anatomy and always includes common mixing physiology.
The clinical presentation can be diverse, ranging from severe cyanosis to severe congestive heart failure. Echocardiography has become the standard technique for determination of the anatomy and hemodynamics of the lesion. Cardiac catheterization often provides additional information, especially on pulmonary artery anatomy, size, and pressures. Presently, management is directed toward a modified Fontan operation. When the patient is younger than 6 months, palliative procedures, including banding of the pulmonary artery or systemic to pulmonary artery shunts, are performed if necessary. After 4 to 6 months of age, a bidirectional cavopulmonary shunt could be considered to effectively provide sufficient pulmonary blood flow while reducing ventricular volume loading. This, in conjunction with additional pulmonary flow, either through a residual systemic to pulmonary artery shunt or through antegrade pulmonary flow, may provide adequate palliation for at least the next few years of life. A more final form of palliation is a Fontan-type operation in which the entire systemic venous return is directed to the pulmonary circulation, bypassing the heart. The 20-year survival of patients with a Fontan circulation is about 90% to 95% in the current era. The life expectancy of patients with this type of palliation is still uncertain.