Chapter 30
Single Ventricle: General Aspects

Eduardo M. da Cruz, Jonathan Kaufman, Brian Fonseca, Harma K. Turbendian, and James Jaggers

Abstract This chapter provides an overview of the very basic aspects of the diagnosis and management of patients with single ventricle anatomy or physiology. Specific chapters elsewhere in this book will further discuss the Fontan physiology and the hypoplastic left and right heart syndromes.

30.1 Introduction, Anatomy, and Physiology

Single ventricle anatomy characterizes a large spectrum of complex congenital cardiac defects, with an incidence of 5/100.000 alive newborns, and an equal gender distribution. *Single ventricle physiology* can also be found in some biventricular hearts affected by congenital malformations.

Single ventricles, of left, right, or undefined morphology, are often associated with cardiac malpositions and heterotaxia with levo- or dextro-isomerism. These congenital malformations, complex by definition, may have atrioventricular concordance or discordance, different types of atrioventricular connections, and concordant or discordant ventriculoarterial connections and may be associated with multiple other defects with or without aortic (less frequent) or pulmonary (more frequent) valvular or subvalvular obstruction or both (exceptional).

In the context of heterotaxia, anomalous systemic venous returns (i.e., interrupted inferior vena cava with azygos continuation) or pulmonary venous connections (partial or total anomalous pulmonary venous return with or without obstruction) may also be present and significantly complicate the surgical and medical management of these patients, as well as their outcome.

The atrioventricular connection may be a single inlet (i.e., mitral or tricuspid atresia), a double inlet (two functional atrioventricular valves), or a common inlet (similar to the complete atrioventricular septal defect). Atrioventricular valves may be overriding or straddling across large interventricular communications. Often, the normal and functional ventricle communicates with a rudimentary or undeveloped ventricle through the bulbo-ventricular foramen which plays an important role in the pathophysiology of the single ventricle, when restrictive. Such restriction may be the source of a sub-vascular aortic or pulmonary obstruction depending on the ventriculoarterial relationships. The same principle applies to anatomic forms where complex sub-vascular obstructions, often due to septal muscular conus or fibrous structures, are present.

Ventriculoarterial connections may be concordant or discordant and aortic or pulmonary stenosis or atresia may be found.

E. M. da Cruz (✉)
Associate Medical Director, CHCO Heart Institute,
Head, Pediatric Cardiac Critical Care Program & Inpatient Services,
Director, Cardiac Intensive Care Unit, Children's Hospital Colorado,
Tenured Professor of Pediatrics, Pediatric Cardiology and
Intensive Care, University of Colorado Denver, School of Medicine,
Aurora, CO, USA
e-mail: Eduardo.dacruz@childrenscolorado.org

J. Kaufman
Pediatric Cardiac Critical Care Program, Heart Institute,
Department of Pediatrics, Children's Hospital Colorado, University
of Colorado Denver School of Medecine, Aurora, CO, USA
e-mail: jonathan.kaufman@childrenscolorado.org

B. Fonseca
cMRI and Clinical Informatics, Heart Institute, Department of
Pediatrics, Children's Hospital Colorado, University of Colorado
Denver School of Medecine, Aurora, CO, USA
e-mail: Brian.Fonseca@childrenscolorado.org

H. K. Turbendian
Pediatric Cardiovascular Surgery, University of Pittsburgh Medical
Center, Wolfson Children's Hospital, Jacksonville, FL, USA

J. Jaggers
Pediatric Cardiothoracic Surgery, Heart Institute,
Children's Hospital Colorado, University of Colorado Denver
School of Medicine, Aurora, CO, USA
e-mail: james.jaggers@childrenscolorado.org

R. A. Munoz et al. (eds.), *Critical Care of Children with Heart Disease*, https://doi.org/10.1007/978-3-030-21870-6_30

From the functional standpoint, *a single ventricle physiology characterizes any cardiac defect that does not allow a biventricular repair,* including the following:

1. Complex congenital cardiac malformations with a single functional ventricle, with right, left, or undetermined morphology:
   (a) Hypoplastic left heart syndrome (HLHS)
   (b) Right-sided heart malformations:
      (i) Pulmonary atresia with intact interventricular septum and severe right ventricular hypoplasia (non-tripartite ventricle)
      (ii) Tricuspid atresia
      (iii) Severe forms of Ebstein malformation of the tricuspid valve
   (c) Heterotaxia syndromes with left or right isomerism (i.e., Ivemark syndrome)
   (d) "Criss-cross" type anomalies
2. Cardiac defects with significant ventricular unbalance and/or with straddling of the atrioventricular valve:
   (a) Atrioventricular septal defect
   (b) Double outlet right ventricle
3. Cardiac defects with multiple interventricular septal defects ("swiss-cheese" type ventricular septal defects)

Depending on the combination of anatomic features, multiple physiological scenarios may be found, alone of combined:

1. Decreased systemic flow in the presence of left subvalvular and/or valvular and/or vascular obstruction
2. Decreased pulmonary flow in the presence of right subvalvular and/or valvular and/or vascular obstruction
3. High pulmonary flow with pulmonary hypertension if there is no pulmonary protection; this may coexist with left-sided obstructions
4. Restrictive intracardiac mixing at the atrial and/or the ventricular level may be associated with any of the above

30.2 Diagnosis

30.2.1 Clinical

Clinical manifestations of single ventricle depend on the anatomic and physiological characteristics. Most patients are diagnosed in the neonatal period if not on fetal evaluation, particularly those who have a ductal-dependent circulation because of right- or left-sided heart obstruction. These patients present with profound cyanosis or progress toward cardiogenic shock, once the ductus arteriosus becomes restrictive or closes.

Depending on the associated malformations, patients may have a cardiac murmur, usually ejective in nature if there is a vascular obstruction, regurgitant in case of incompetent atrioventricular valves, or continuous if there is a largely patent ductus arteriosus or in the presence of collateral circulation. The first heart sound is usually normal but may be split in severe Ebstein malformation of the tricuspid valve, and the second sound is unique and loud whenever pulmonary hypertension is associated. When there is an obstructive systemic physiology, pulses will be weak, absent, or asymmetrical and the patient will display signs of low cardiac output or shock with progressive letargia, diaphoresis, difficulty to feed, tachypnea and tachycardia, and ultimately poor peripheral perfusion, vasoconstriction, or pallor and profound hypotension and lactic acidosis. When an obstructed total anomalous pulmonary venous return is concurrent, patients rapidly progress to cardiogenic shock in the context of a "white lung" syndrome. An immediate workup is vital to establish a differential diagnosis, namely, with severe pneumonia, streptococci B infection, lung lymphangiectasias, or persistent pulmonary hypertension of the newborn. As a matter of fact, many neonates are admitted with primary suspicion of noncardiac disease – frequently sepsis – and the diagnosis of cardiac disease is evoked in the setting of refractoriness to medical therapy. A similar pathophysiology may be found in patients with hypoplastic left heart syndrome in whom the atrial septal defect is restrictive or even absent, requiring an emergent intervention at birth to enlarge the communication.

30.2.2 Chest X-Ray

Radiological findings are very heterogeneous and depend of the anatomic characteristics and the pathophysiology. Heterotaxic or isomeric forms often show radiological features of situs inversus or ambiguous with left of right isomerism, levo- or dextrocardia. Forms with pulmonary obstruction will present with oligemic lungs. When the pulmonary bed is unprotected, the chest X-ray will display excessive blood flow with increased vascular markings and eventually lung edema as the pulmonary vascular resistances decrease over the first few days of life. The presence of diffuse interstitial infiltrates or a "white lung" aspect strongly suggests an obstructed pulmonary venous return. There may be various degrees of cardiomegaly.

30.2.3 Electrocardiogram

The ECG, although unspecific, may provide information regarding axis deviation and predominance that may be useful in steering the diagnosis. It is also useful to rule out any associated arrhythmias or conduction disorders and is particularly important in patients with heterotaxia or with Ebstein malformation of the tricuspid valve.

30.2.4 Echocardiography

Transthoracic echocardiography remains the cornerstone of diagnosis in single ventricle patients, allowing the fine identification of the anatomical and physiological characteristics of the disease [1]. Echocardiographic evaluation has to be exhaustive, thorough, and meticulous. Figures 30.1, 30.2, 30.3, 30.4, 30.5, 30.6, 30.7, and 30.8 demonstrate some

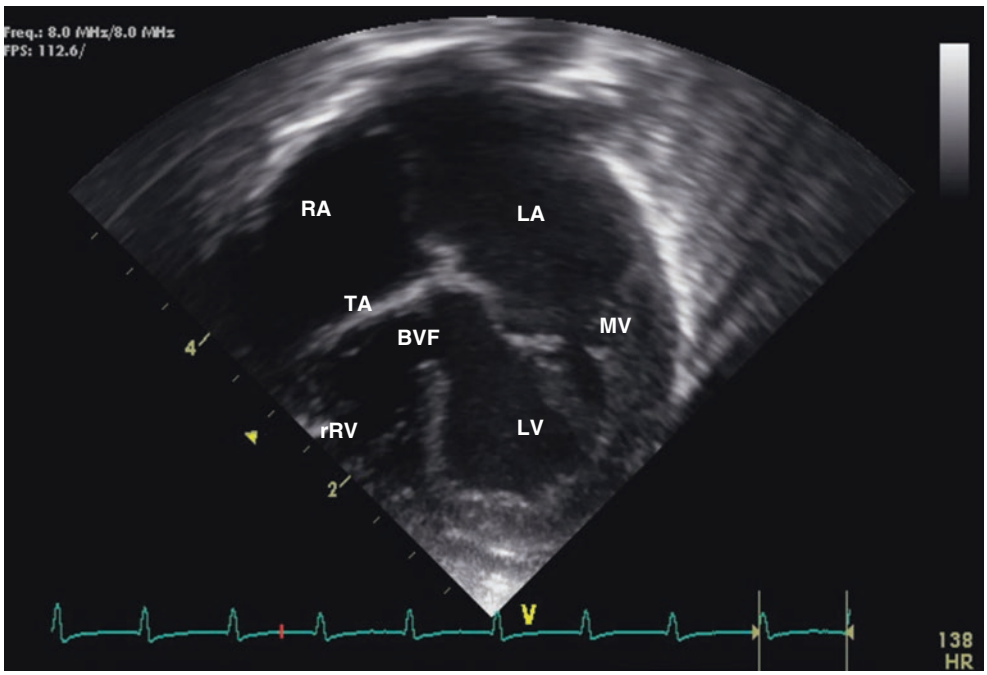

Fig. 30.1 Echocardiography documenting a tricuspid atresia; the ventriculoarterial concordance is not seen in this view. RA right atrium, LA left atrium, MV mitral valve, TA tricuspid atresia, LV left ventricle, rRV rudimentary right ventricle, BVF bulbo-ventricular foramen

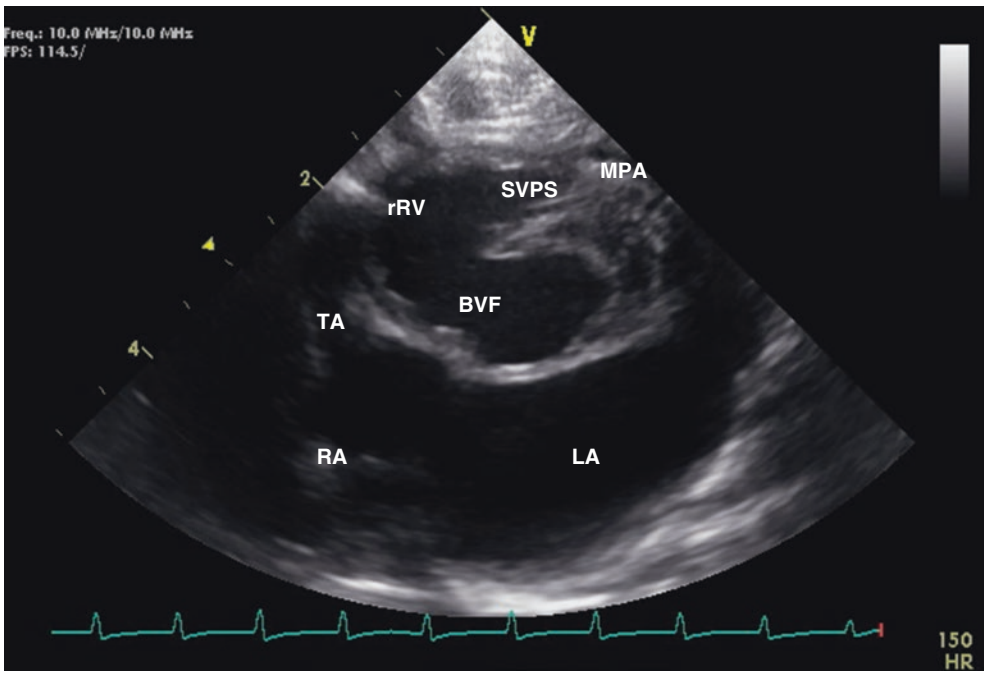

Fig. 30.2 Tricuspid atresia type 1b. LA left atrium, RA right atrium, TA tricuspid atresia, rRV rudimentary right ventricle, BVF bulbo-ventricular foramen, PMA main pulmonary artery, SVPS severe subvalvular pulmonary stenosis

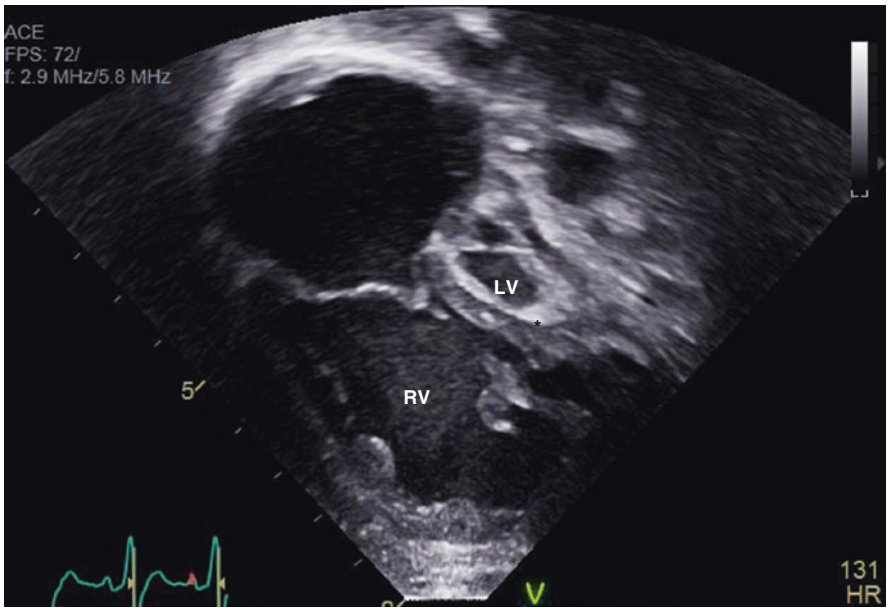

Fig. 30.3 Apical horizontal long-axis view in a patient with hypoplastic left heart syndrome. The systemic right ventricle (RV) can be seen to be appropriately dilated and hypertrophied. The left ventricle (LV) is hypoplastic with an echo-bright myocardium consistent with endomyocardial fibroelastosis

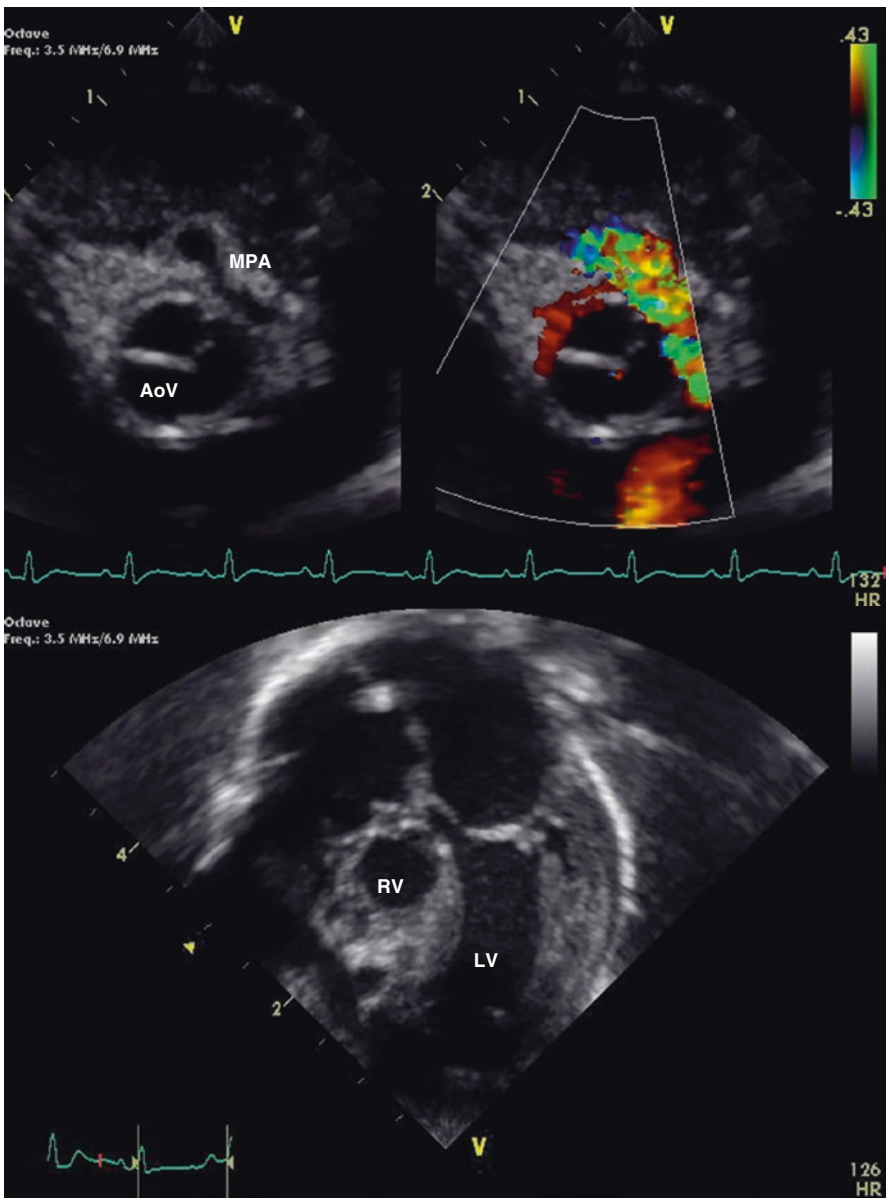

Fig. 30.4 (Panel **a**) Parasternal short-axis view with color Doppler comparison showing an atretic main pulmonary artery (MPA) anterior to the aortic valve (AoV) with retrograde filling from the patent ductus arteriosus (turbulent retrograde flow). (Panel **b)** Apical horizontal long-axis view of the same patient shows a diminutive right ventricle (RV) with no ventricular septal defect

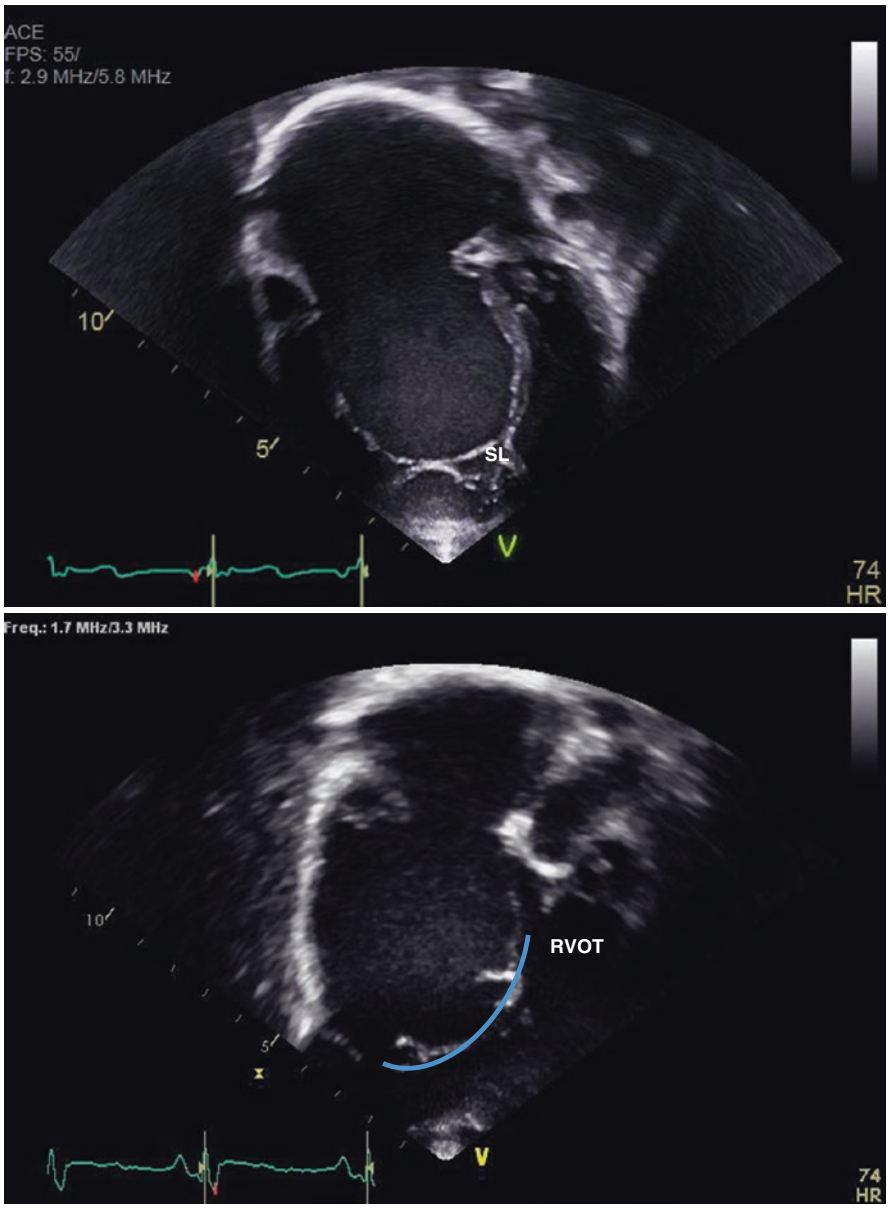

Fig. 30.5 (Panel **a**) Apical horizontal long-axis view showing severe apical displacement of the septal leaflet (SL) of the tricuspid valve. If the displacement is severe, the coaptation plane (line) will not be apparent in apical horizontal long-axis view and will only be apparent in long-axis views of the right ventricle in the right ventricular outflow tract (RVOT) (Panel **b**)

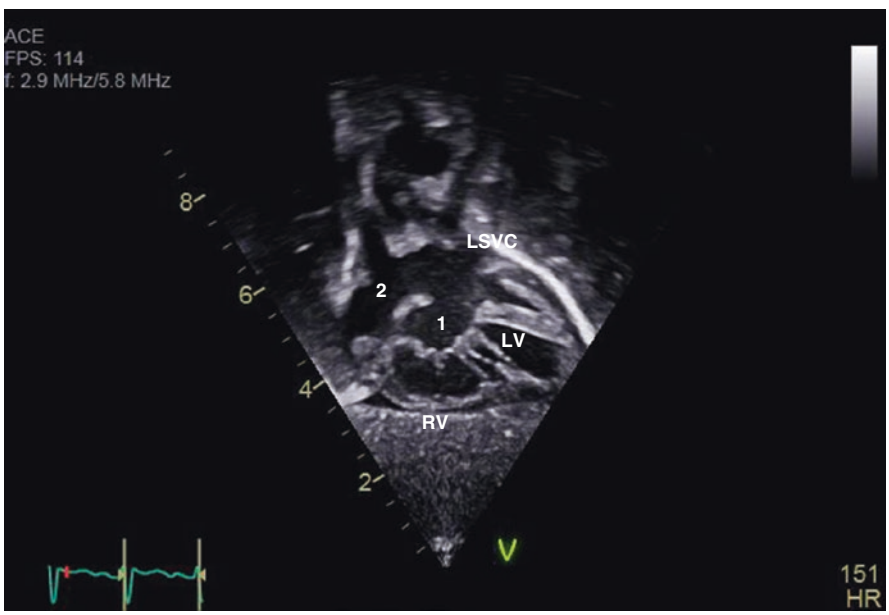

Fig. 30.6 Subcostal coronal view of a patient with right atrial isomerism. This view demonstrates a complete, unbalanced atrioventricular canal with right ventricular (RV) dominance. There is an ostium primum (1) and an ostium secundum (2) atrial septal defect. Also, typical of this lesion, there is a left superior vena cava (LSVC) which connects directly to the roof of the left atrium

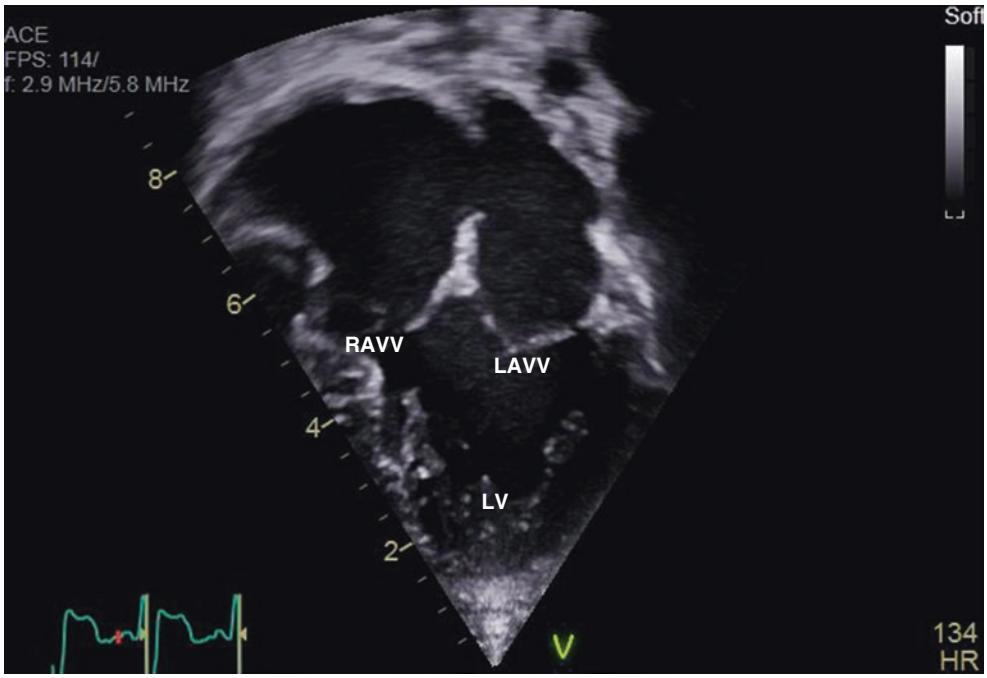

Fig. 30.7 Apical four-chamber view of a patient with double inlet left ventricle. Typical for this lesion, both the right (RAVV) and the left (LAVV) atrioventricular valves connect to a single left ventricle (LV)

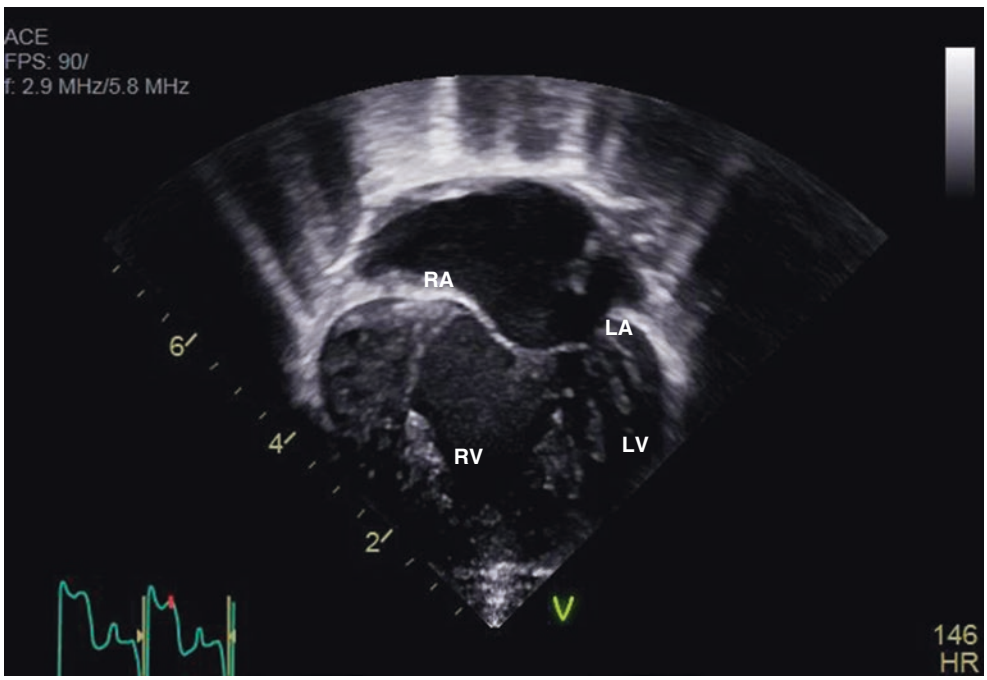

Fig. 30.8 Apical horizontal long-axis view showing a complete, unbalanced atrioventricular septal defect with right ventricular (RV) dominance. There is a common atrioventricular valve committed to the right ventricle and a large ostium primum defect. Note the atrial septum which is shifted to the left resulting in a hypoplastic left atrium (LA). The hypoplastic left ventricle (LV) is difficult to appreciate in this view due to the large inlet ventricular septal defect

examples of single ventricle anatomy. Transesophageal echocardiography is often used in the operating room to further elucidate intracardiac anatomic details and postoperative results.

30.2.5 Cardiac Catheterization

In the neonatal patient, diagnostic cardiac catheterization of patients with single ventricle is seldom indicated unless there is a complex venous return requiring further anatomic clarification or when there are doubts about the pulmonary vascular resistances. Many of these indications for cardiac catheterization are being progressively replaced by advanced noninvasive cardiac imaging like CT scan, CTA, or CMR. Nevertheless, cardiac catheterization remains an important diagnostic assessment of patients awaiting a partial or a total cavopulmonary connection.

Interventional procedures play a very important role in the collaborative management of these anomalies with the

surgical team, or else for electrophysiological studies or interventions. Not infrequently, patients with single ventricle benefit from embolization of collateral vessels and/or balloon dilatation and stent implantation particularly in the pulmonary arteries. Patients with pulmonary vein pathology may also need recurrent interventional procedures. Moreover, hybrid procedures may be indicated in some circumstances, particularly but not exclusively in patients with hypoplastic left heart syndrome.

30.2.6 Advanced Cardiac Imaging

Both cardiac MR (CMR) and cardiac CT have taken a more important role in noninvasive imaging of patients with single ventricle in recent years.

In general, cardiac MRI (Figs. 30.9 and 30.10) produces similar information to echocardiography and CT without the use of ionizing radiation. Using gadolinium contrast, an MR angiogram (MRA) produces a three-dimensional dataset with excellent visualization of both arterial and venous anatomy, without the limitations of acoustic windows. The three-dimensionality of the dataset also allows reformatting in multiple planes which is invaluable in understanding complex anatomy. Cardiac MRI can also produce cine images which are used to visualize cardiac anatomy, valve function, and ventricular function. Unlike volumetric measurements with echocardiography, MRI uses a highly reproducible contiguous slice measurement of ventricular volumes which is not dependent on geometric assumptions. CMR sequences can specifically image for intracardiac or vascular thrombus and myocardial infarction and quantify intracardiac shunts and aortopulmonary collateral flow and lymphatic abnormalities.

Although CMRs produce a wealth of data, they can be lengthy and require multiple breath holds. In patients under 8 years of age, CMR may require sedation and/or mechanical ventilation. It is important to note that a majority of adverse events at two high-volume congenital CMR centers involved single ventricle patients with either BT shunts or right ventricle to pulmonary artery conduits [2–4]. CMR in this patient population requires specialized expertise and should be done in centers with knowledgeable CMR practitioners and experienced anesthesia providers. Pacemakers are common in single ventricle patients and historically have been an absolute contraindication to CMR. Nonetheless, with modern pacemakers and leads, CMR is becoming more a routine in these patients [5].

Similar to a MRA, CTA (Figs. 30.11 and 30.12) produces a three-dimensional dataset which shows venous and arterial anatomy with excellent resolution (e.g., pulmonary arteries, BT shunt, cavopulmonary anastomoses, and coronary arteries). Unlike MRI, CT is generally performed with a single phase (arterial versus venous). Therefore, CT must be carefully planned to highlight the anatomy of interest. Multiple phases can be acquired but this increases the dose of ionizing radiation. With a gated technique, enough phases can be acquired to collect a single cardiac cycle that can be used to calculate ventricular volumes and systolic function. The temporal resolution of this technique is low, so it should be used with caution in smaller patients with higher heart rates. CTA is also particularly useful for visualizing small vessels such as coronary arteries in patients with pulmonary atresia with intact ventricular septum or with major aortopulmonary

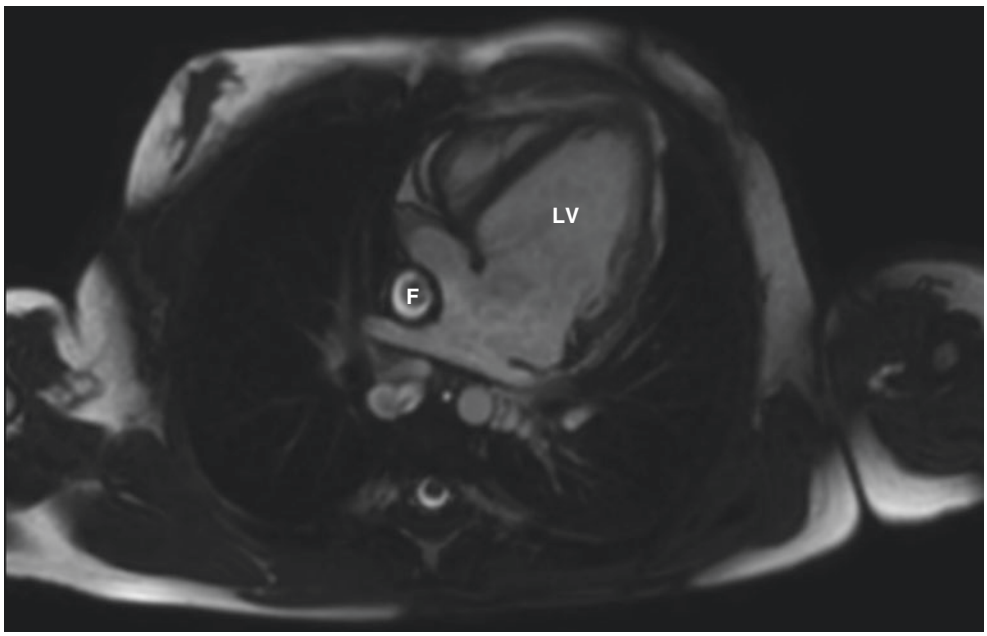

Fig. 30.9 Cardiac MRI showing the apical four-chamber view of a patient with tricuspid atresia. The extracardiac Fontan (F) can be seen in cross section. This kind of imaging can be used to quantify cardiac function and visualize anatomy, valve function, and vascular stenosis

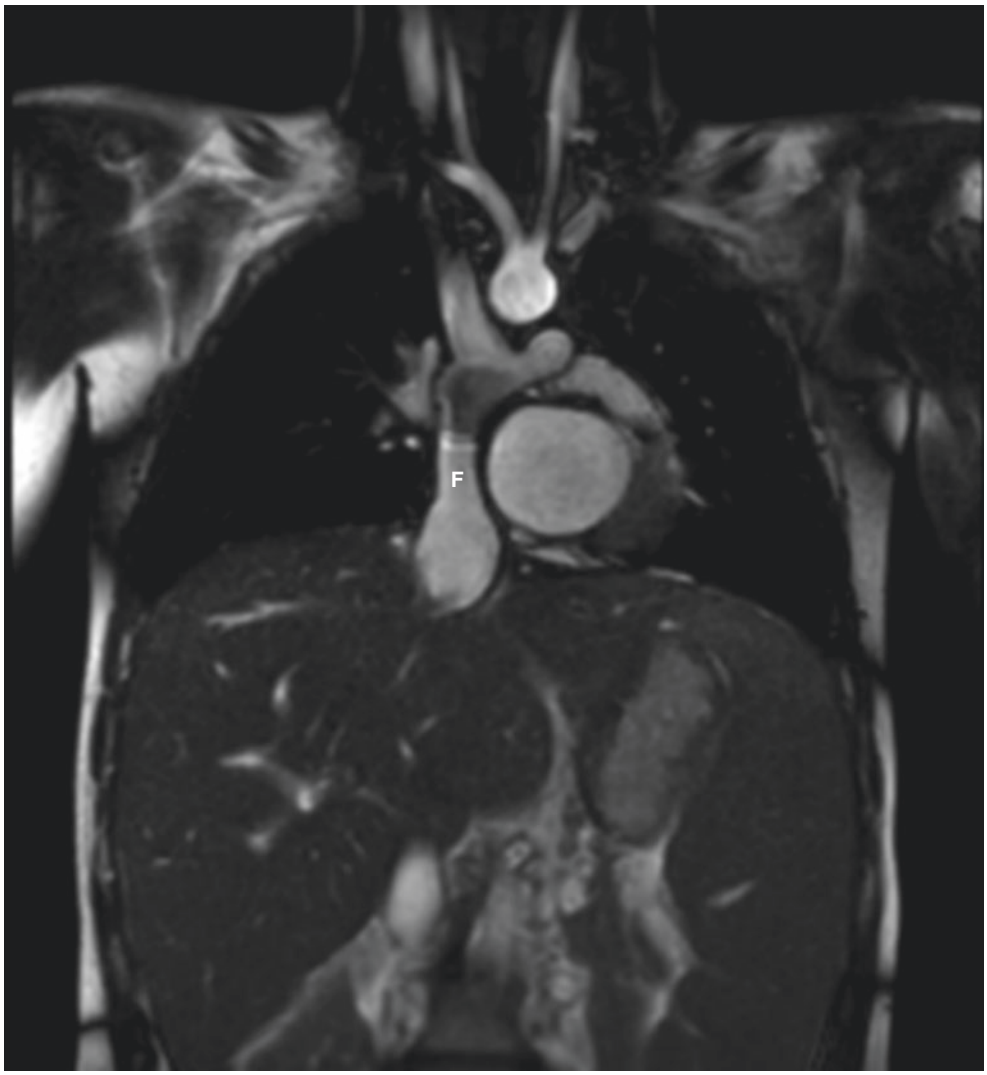

Fig. 30.10 Coronal MRI image showing the entire Fontan pathway. This is an excellent modality for screening for stenosis and thrombus

collaterals in the setting of pulmonary atresia with ventricular septal defect. A major benefit of CT angiography in this patient population who can be clinically unstable is that the scan can be performed very rapidly in a single breath hold. Smaller patients may need to have sedation and/or an artificial airway depending on institutional practices. CTA also produces excellent images of the airway which can be important in this population with a high incidence of airway anomalies. Additionally, CTs can be reformatted to show the three-dimensional relationship between the airway and the vasculature which is important for surgical or catheterization planning.

A drawback to CTA is the use of ionizing radiation in a patient population that will be exposed to numerous radiologic studies (e.g., cardiac catheterization and chest films), so it should be used judiciously.

30.3 Basic Principles of Single Ventricle Management

The concept of univentricular type "repair" implies that patients will need a sequence of palliative interventions steering the therapy toward a common pathway with the ultimate goal of a total cavopulmonary connection (Fontan-Kreutzer procedure) that separates circulations [6–9].

There are essentially three palliative phases or stages, but more interventions are often required through the interphases:

1. Neonatal palliation
2. Partial cavopulmonary connection: modified Glenn connection, bidirectional Glenn connection, hemi-Fontan, or

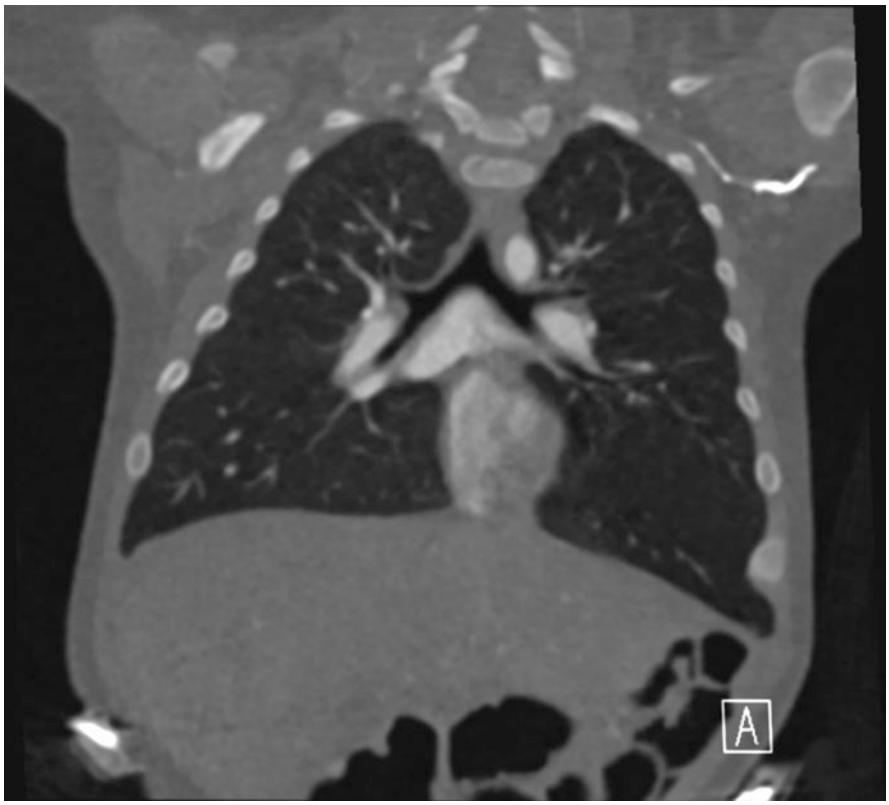

Fig. 30.11 Coronal reformat of a CT scan in a patient with right atrial isomerism which shows bilateral morphologic right airway anatomy with a steep angulation of the mainstem bronchus and an early takeoff of the upper lobe bronchus

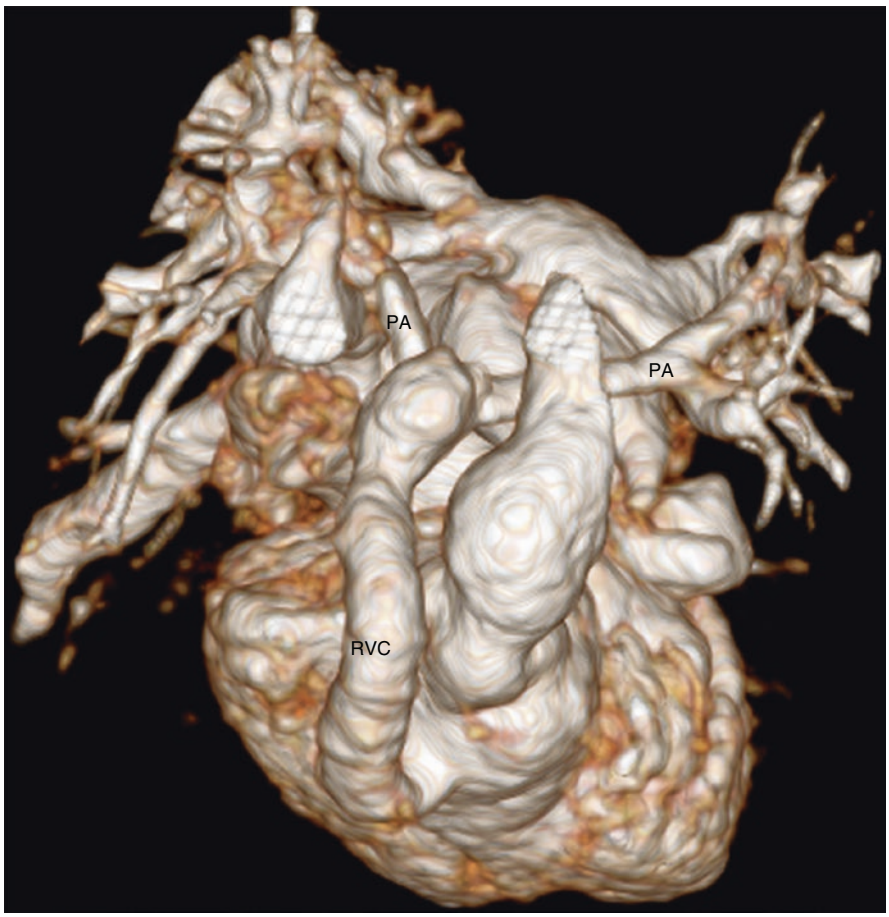

Fig. 30.12 Three-dimensional volume rendering of a CT scan of a patient with hypoplastic left heart syndrome after stage 1 palliation with a Norwood and right ventricle to pulmonary artery conduit (RVC). The right ventricle to pulmonary artery conduit can be seen originating from the anterior right ventricle and inserting into the pulmonary arteries (PA)

partial cavo-bi-pulmonary connection (or bi-cavo-bi-pulmonary, in case of right and left superior vena cava)
3. Modified intra- or extracardiac Fontan connection or total cavopulmonary connection

The most commonly followed therapeutic algorithm that varies with the specific diagnosis is as follows:

- *Stage 1: Neonatal palliation*
  - Modified Blalock-Taussig or a central shunt in case of a right obstruction. Alternatively, some patients may be candidates for stenting of the ductus arteriosus or the sub-pulmonary outflow tract.
  - Surgical repair of any left obstruction, i.e., aortic coarctectomy
  - Norwood, Sano, or Damus-Kaye-Stansel operations, hybrid approach, or orthotopic heart transplant
  - Pulmonary artery banding in case of high pulmonary flow with pulmonary arterial hypertension (unprotected pulmonary flow)
- *Stage 2: 3–6 months of age*
  - Partial cavopulmonary connection (modified Glenn procedure or hemi-Fontan procedure)
- *Stage 3: 2–6 years of age*
  - Total cavopulmonary connection (modified Fontan procedure; intra- or extracardiac; with or without fenestration)

30.4 Preoperative Management of the Neonatal Patient

Preoperative stability is key for good postoperative outcomes, notably in the neonatal patient. The main objective prior to intervention is to maintain the patient in a balanced physiological and homeostatic state with an efficient Qp/Qs ratio, allowing for adequate tissue perfusion. Very seldom do these patients need mechanical ventilation, except when diagnosed late and in cardiogenic shock. Maintaining the patients on spontaneous ventilation allows a more physiological stability, ad libitum feeding, and an opportunity to invest on mother-infant interactions. Safe venous lines are mandatory for the administration of drugs (i.e., PGE_1), blood sampling (to be minimized), and reliable access in case of decompensation. Close monitoring and follow-up of tissue perfusion markers is essential to evaluate trends anticipating a breach of the Qp/Qs balance with compromise of the systemic perfusion. Patients with physiology allowing pulmonary overcirculation to the detriment of the systemic perfusion may need systemic vasodilators and eventually the use of subatmospheric gas if refractory to medical therapy. In patients with such trend, the indication for intervention would be the only way to effectively address this physiology without exposing the patients to avoidable side effects and the risk of shock.

30.5 Surgical Neonatal Palliations

30.5.1 Variants with Aortic or with Pulmonary Obstruction

30.5.1.1 Patients with Subvalvular or Valvular Pulmonary Obstruction

These patients have a significant ductal-dependent and therefore PGE_1-dependent physiology requiring a neonatal palliation. The classic intervention is a modified Blalock-Taussig or a central shunt. In some cases, the alternative of interventional percutaneous balloon dilatation of the sub-pulmonary outflow tract or ductal stenting may be considered.

30.5.1.2 Patients with Obstruction of the Aortic Arch

This anatomy is almost always associated with unrestricted pulmonary flow and pulmonary hypertension. Therefore, patients require a surgical palliation associating aortic arch repair (i.e., aortic coarctectomy) with a pulmonary artery banding. Surgical techniques are decided upon the surgeon's preference and the degree of aortic hypoplasia and may be performed through a thoracotomy or else through a sternotomy with cardiopulmonary bypass and hypothermic circulatory arrest or using specific cannulation techniques to ensure an adequate cerebral perfusion (selective brain perfusion).

30.5.1.3 Patients with Hypoplastic Left Heart Syndrome (HLHS) or with Complex Left Ventricular Outflow Tract Obstructions

Details concerning therapy of HLHS will be further discussed in a specific chapter in this book. HLHS is a complex disease for which different alternatives might be considered:

1. Therapeutic abstention: may be an option in certain environments or by family choice, or else in patients with unfavorable anatomy and reserved prognosis.
2. Stage 1 Norwood type intervention (with a modified Blalock-Taussig shunt) or Sano modification (with a systemic ventricle-to-pulmonary conduit).
3. The hybrid approach: following this approach, a stent is inserted in the ductus arteriosus to ensure its patency, and pulmonary circulation is protected by selective surgical pulmonary branch bandings. Patients may subsequently progress toward a combination of stage 1 and 2 procedures (associating the removal of the ductal stent, plasty of pulmonary arteries, the creation of a neo-aorta with repair of the isthmic aortic obstruction, and a partial cavo-pulmonary connection) or toward an orthotopic heart transplant, in which case the need for neonatal pulmonary protection is controversial. Indications for this approach vary depending on the institutions but may include team's preference, prematurity, low birth-weight, significant ventricular dysfunction, significant tricuspid regurgitation, lung disease or other associated malformations, contraindications to cardiopulmonary bypass, or religious principles (i.e., avoidance of the use of blood products).
4. Orthotopic heart transplant.
5. Other situations: in some anatomic forms like the type IIc tricuspid atresia with d-transposition of the great arteries, unprotected pulmonary flow, and partially restrictive bulbo-ventricular foramen at the origin of significant sub-aortic obstruction, the alternative of an arterial switch may be discussed, transforming the aortic obstruction into a protective sub-pulmonary obstruction.
6. Some borderline anatomic forms allow a biventricular repair often associated with endomyocardial fibroelastosis resection, mitral plasty, and resection of complex subaortic obstructions. Other patients with small left outflow tracts and ventricular septal defect may be candidates to the Yasui operation.

30.5.1.4 Patients with Hypoplastic Right Heart Syndrome

This group of cardiac malformations is initially approached as any severe right-sided obstruction, by creating a modified Blalock-Taussig or a central shunt associated with an atrioseptectomy. Depending on the degree of right ventricular hypoplasia, if the cavity is small but tripartite and the tricuspid valve considered viable, these interventions may be combined with the opening of the right ventricular outflow tract by surgery or by interventional catheterization in the hope that the patient becomes a candidate for a biventricular or a ventricle-and-a-half repair.

30.5.2 Variants Without Pulmonary Protection (May Be Associated with Left Obstructions)

Such patients need an intervention aiming to protect their pulmonary vascular bed from high flow and pulmonary hypertension. Hence, a pulmonary artery banding is the indication.

30.5.3 Complex Variants with Heterotaxia

In the context of heterotaxia, single ventricles are usually associated with cardiac malpositions and with anomalous systemic or pulmonary venous returns that require complex interventions at the time of the above-described palliations. Other associative defects have been described and are intimately linked to the type of isomerism, dextro or levo. These include anomalies of the atrioventricular concordance, the ventriculoarterial concordance, and often aortic or pulmonary subvalvular or valvular obstructions. Rhythm disturbances are not uncommon and have a significant impact on decision-making and on outcomes.

30.6 Postoperative Management of Neonatal Palliations

30.6.1 General Aspects

Management of HLHS, aortic coarctation, and anomalous pulmonary venous returns is discussed in specific chapters elsewhere.

Pulmonary artery banding and modified Blalock-Taussig or central shunts usually have an uneventful postoperative course, but in some patients, particularly in low weight or premature newborns, postoperative management may be fraught with hemodynamic challenges. Therefore, caregivers need to be meticulous and prudent. The Qp/Qs balance is often difficult to achieve and maintain. Most patients require some degree of inotropic or lusitropic support with low-dose epinephrine or dopamine combined with milrinone. The Qp/Qs balance is a complex equation that needs to be customized to patient's physiology. Interventions target the management of resistances in both the systemic and the pulmonary circulations with ventilatory maneuvers, control of pH, use of cardiovascular drugs, sedation, and core temperature control, to mention some. The ultimate objective is to maintain adequate systemic tissue perfusion while avoiding the

intracellular anaerobic threshold, achieve an oxygen saturation of around 75–80%, and protect the patient against multiorgan dysfunction.

Patients with *high pulmonary resistances* are characterized by pulmonary under-circulation (desaturation, oligemic lungs) and require specific management as described in the chapter dedicated to pulmonary hypertension, namely, with ventilation targeting a slightly alkalotic pH, and the use of inhaled pulmonary vasodilators. Other causes for under-circulation to be systematically ruled out and promptly addressed relate to significant systemic vasoplegia, to small pulmonary arteries, or to partial shunt or right ventricle-to-pulmonary artery conduit obstruction. Some patients may require cardiac catheterization to elucidate anatomic or physiological patterns behind the lack of Qp/Qs balance.

Patients with *high pulmonary flow* (overcirculation expressed by saturations above 80%, low diastolic pressures with a high pulse differential, radiological signs of pulmonary volume overload, progressive metabolic or lactic acidosis by lack of adequate tissue perfusion) may benefit from a "pharmacological banding" associating the maintenance of high blood viscosity (hematocrit above 40–45%), diuretics (usually loop diuretics eventually associated with hydrochlorothiazide or with spironolactone), and systemic IV vasodilators to optimize afterload reduction (milrinone, sodium nitroprusside, nitroglycerine, phentolamine, phenoxybenzamine). In patient refractory to medical therapy, a comprehensive anatomic assessment by echocardiography and eventually cardiac catheterization should be undertaken to assess function and the presence of residual lesions (i.e., residual aortic coarctation).

If the mixed venous and the pulmonary venous saturations are normal, a saturation of 80% reflects a Qp/Qs of approximately 1:1. Nevertheless, caution ought to be exerted in patients with desaturated pulmonary veins, in which case the same 80% of systemic saturation would reflect a much higher Qp/Qs predisposing the patient to a compromise of the systemic tissue perfusion.

The aim of this "pharmacological banding" is to decrease the Qp/Qs following the principles described in Poiseuille's law: flow is directly proportional to the ratio of resistances and the diameter of the shunt and inversely proportional to the length of the shunt and blood viscosity.

Pulmonary artery banding may sometimes be too loose or too tight at the origin of over or under-circulation, respectively. It may also migrate peripherally, producing distortion of the pulmonary arteries or even total occlusion. When the banding is too close to the pulmonary valve, it may also induce pulmonary regurgitation that must be addressed, particularly in those patients in whom the pulmonary valve will become a neo-aortic valve in the future.

Particular caution is necessary in patients with shunts, regarding the prevention of a hypercoagulability and risks of thrombosis. This includes avoiding dehydration or disproportionate negative hydric balance, using in the absence of bleeding, and introducing antiplatelet therapy once the patients resumes enteral feeding.

30.6.2 Monitoring

These patients require a peripheral arterial line ideally inserted in the right radial artery if an aortic coarctation was repaired and a central venous line, as indwelling catheters. Other parameters to be monitored are cardiac and respiratory rate, peripheral oxygen saturation, ECG, and cerebral and somatic near-infrared spectroscopy (NIRS).

30.6.3 Inotropic and Vasodilator Drugs

After a pulmonary artery banding or a shunt, patients may require inotropic support. *A cornerstone principle of management in these patients is to target the ultimate goal of circulation: tissue perfusion, rather blood pressures.*

Whenever necessary, an elective drug combination may include dopamine and milrinone. Nevertheless, it is essential to understand the patient's physiology to implement a goal-oriented therapy rather than universal "drug recipes". For that, a thorough physical examination, evaluation of invasive and noninvasive data, as well as biomarkers and indices of tissue perfusion usually provide the necessary information. The latter may need to be complemented by echocardiography and sometimes by cardiac catheterization in patients who do not respond as expected to medical therapy. Patients with overcirculation may require more selective systemic vasodilators such as sodium nitroprusside.

When an aortic coarctation is repaired, patients might have tachycardia and hypertension, needing therapy with systemic vasodilators and beta-blockers, usually sodium nitroprusside or IV nicardipine and esmolol (further details can be found on the chapter related to aortic coarctation).

Vasoplegia related to inflammation and/or side effects of some drugs, namely, milrinone, may need to be antagonized in some patients with vasopressors. Another indication for the use of these drugs may be the need to provide enough kidney or target-organ perfusion and a physiological trans-organ gradient, in patients with elevated central venous pressures. Vasopressin is gaining popularity in the world of cardiac intensive care for this purpose, all the more that it can be a selective vasodilator in some critical vascular beds, including the coronaries.

30.6.4 Respiratory Management

Neonatal patients after any surgery for single ventricle physiology need ventilation for at least the first postoperative hours while achieving an adequate hemodynamic balance. This is notably important in patients with low weight or prematurity, syndromic, or else with high pulmonary resistances or with overcirculation, who may require prolonged ventilation. Cardiopulmonary interactions are vital in these patients and will be discussed at length elsewhere in this book. The need for positive pressures does not necessarily justify mechanical ventilation and may be overcome with noninvasive positive pressure ventilation. In case of delayed chest closure, extubation can be accomplished within 12–24 hours after closure.

30.6.5 Sedation and Analgesia

Most patients achieve effective analgesic levels with nonopioid therapy associated with morphine or fentanyl and benzodiazepines (i.e., midazolam, clonazepam) at minimal efficient doses, for at least the first 48 hours, particularly in patients having required an intervention by thoracotomy. Dexmedetomidine is an attractive alternative in these patients. Children requiring prolonged use of opioids and benzodiazepines should be monitored for potential withdrawal syndrome. Also, caution is needed in patients requiring longer intensive care admissions with regard to delirium.

30.6.6 Anticoagulation

In case of systemic-pulmonary shunt, it is important to anticoagulate patients once postoperative bleeding is under control. Antiplatelet therapy (aspirin, dipyridamole, or eventually clopidogrel) ought to be prescribed as soon as patients resume enteral feeding, allowing to suspend the heparin.

30.6.7 Specific Problems

Specific problems are mostly those associated with the balance of the ratio between the systemic and the pulmonary resistances as previously discussed. Other occurrences and complications may be diaphragmatic palsy or paresis, chylothorax, or Horner's syndrome by phrenic or recurrent laryngeal nerve lesions.

30.7 Partial Cavopulmonary Connections

Single ventricle physiology is shared by various and heterogeneous entities. Their common characteristic is that there is a complete mixing of oxygenated and desaturated blood that will be distributed in both the systemic and the pulmonary circuits. The main objective of the univentricular type repair or palliation is to individualize the systemic and the pulmonary circulations to achieve almost normal systemic saturations with a more effective ventricular workload. The partial cavopulmonary connection performed with the modified Glenn intervention is the first step toward this goal. William E. Glenn firstly described his technique in 1958. The classic Glenn procedure consisted in completely dividing the left from the right pulmonary branches, connecting the latter to the right superior vena cava while preserving the anterograde flow from the heart toward the left branch. Some of these patients have survived to adulthood, but many have developed left pulmonary hypertension, having therefore become cyanosed, as an equivalent to the Eisenmenger's complex. In case of surgical intervention, these patients would require a double-lumen endotracheal tube allowing differential ventilation of the right and the left lung. The current modified Glenn connection (since the 1980s) allows passive flow from the superior vena cava to both pulmonary branches, and the continuity with the ventricular mass is ceased by sectioning the pulmonary trunk or restricted by further tightening the banding, in which case a degree of anterograde pulsatile flow will persist into the pulmonary arteries.

30.7.1 Preoperative Evaluation

Patients with single ventricle physiology will become candidates for partial cavopulmonary connections throughout the first months of life. To summarize, the required conditions to indicate a cavopulmonary connection are the confirmation of normal rhythm and conduction and the absence of any anatomical or functional obstructions throughout the future cavopulmonary circuit:

- Anatomic criteria
  - No significant stenosis or deformation of the pulmonary arteries
  - No pulmonary venous stenosis
  - Nonrestrictive communication between the atrial cavities (in case of stenosis or atresia of an atrioventricular valve)
  - No subvalvular or valvular aortic obstruction

  - No residual aortic coarctation
  - No thrombus within the vascular bed
- Functional criteria
  - Low pulmonary pressures and resistances
  - No significant atrioventricular valvular regurgitation
  - Normal ventricular function, both systolic and diastolic
  - Normal sinus rhythm and no conduction disorders

    The main general objectives of the cavopulmonary connection are:
    - To facilitate the diastolic unload of the systemic ventricle
    - To prevent deformation of the pulmonary arteries
    - To avoid the negative impact of the continuous diastolic "steal" on the myocyte which, associated with the diastolic volume overload, may induce irreversible myocardial changes

These objectives are accomplished by diverting venous blood from the superior vena cava toward the pulmonary artery with a modified Glenn connection, or else with a hemi-Fontan procedure. Usually, this intervention is proposed when the pulmonary arteries have an adequate diameter, and the pulmonary vascular resistances are low, around 4 months of age and ideally before 6 months of age.

30.7.2 Preoperative Cardiac Catheterization

Cardiac catheterization is a common procedure in preparation for the cavopulmonary connection [10–12]. The objectives of such procedure are as follows:

1. To measure saturations and pressures in the pulmonary branches and to estimate the Qp:Qs ratio and the pulmonary vascular resistances. In patients with borderline values, cardiac catheterization allows the performance of pharmacological tests aiming to reduce the pulmonary resistances.
2. To perform angiographies in the ventricular cavities, the innominate vein, the pulmonary arteries, and the aortic arch.
3. To perform interventional procedures like balloon dilatation and stent insertion in localized pulmonary stenosis or on residual obstructions of the distal suture of the neoaorta and also to dilate or stent any residual aortic coarctation. It may also be instrumental for the percutaneous embolization of veno-venous pulmonary collateral vessels, arteriovenous malformations, or aortopulmonary collateral vessels. Last, but not least, cardiac catheterization is useful to occlude fenestrations after completion of the total cavopulmonary connection.

30.7.3 Surgical Techniques

30.7.3.1 The Glenn Procedure

The modified Glenn anastomosis (Fig. 30.13) may be performed on cardiopulmonary bypass with beating heart or without cardiopulmonary bypass in patients with anterograde flow who do not require any intracardiac intervention. The superior vena cava is sectioned from the atrial mass after ligation of the azygos return, and its caudal portion is directly anastomosed onto the right pulmonary artery. Any previous aortopulmonary shunt is ligated and/or sectioned. When a previous pulmonary banding has been performed, some groups maintain a persistent anterograde flow from the ventricular mass, although tightening the banding, while others favor the section of the pulmonary trunk and even the resection of the pulmonary valve to remove any pouches that would become a potential source of thromboembolism. The theoretical advantage of the first approach is to provide the lungs with a hepatic angiogenesis inhibitor factor that would decrease the risk for pulmonary arteriovenous fistula formation, but there is no evidence confirming this theory. Potential disadvantages of preserving the pulmonary anterograde flow are pulmonary diastolic overload and potential for distortion of the pulmonary artery anatomy by the

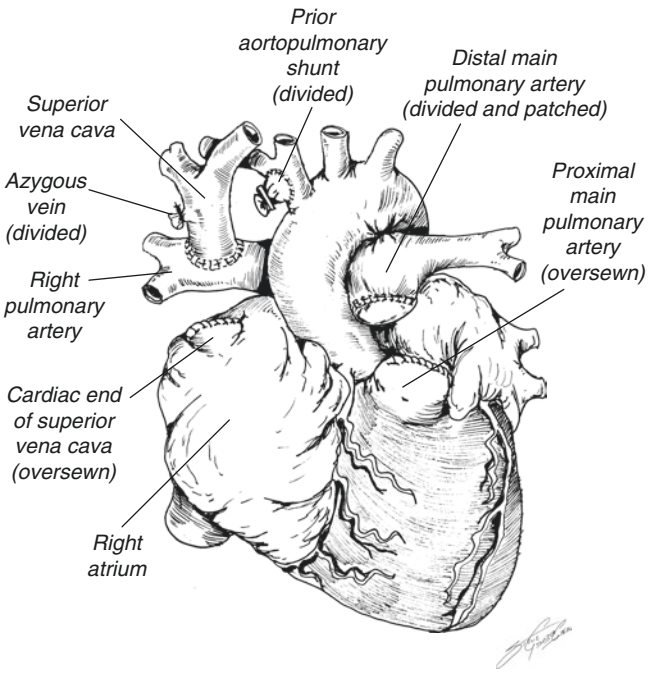

Fig. 30.13 The partial cavopulmonary connection (Glenn procedure)

banding, particularly if it migrates toward the pulmonary branches bifurcation. During the same operating time, other interventions might be indicated: atrioventricular valvular plasty, atrial septectomy, pulmonary artery plasty or patch enlargement or repair of any residual aortic arch obstruction. Arteriovenous fistulas have a significant hemodynamic impact on the Fontan circulation as a source of persistent hypoxemia and are more common in patients with heterotaxia and with interrupted inferior vena cava and azygos continuation in whom a Kawashima procedure is performed. If identified, such collaterals require ligation if not occluded in the catheterization laboratory.

The Glenn anastomosis is more laborious and may carry an increased morbidity when performed in patients with HLHS and previous hybrid approach, in whom it is required to remove the ductal stent and perform an atrial septectomy, a plasty of the pulmonary arteries, and an extensive reconstruction of the aortic arch.

30.7.3.2 The Hemi-Fontan Procedure

This is an alternative to the modified Glenn connection [13]. It might facilitate the second step toward the total cavopulmonary connection if the choice is an intracardiac tunnel. It consists in creating an anastomosis between the right atrium and the pulmonary artery by septalizing the atrial cavity with a Goretex® patch to divert the flow drained from the superior vena cava onto the pulmonary bed while ensuring the drainage of the flow arriving from the inferior vena cava toward the ventricular mass, across the atrioventricular valve.

30.7.4 Postoperative Management of the Partial Cavopulmonary Connection

30.7.4.1 Monitoring

These patients require the conventional postoperative monitoring (cardiac rate and ECG, respiratory rate, oxygen saturation, arterial pressure both invasive and noninvasive) eventually complemented by a transthoracic right atrial catheter to appraise filling pressures and an internal jugular indwelling catheter to assess the pulmonary artery pressure. The presence of these catheters allows the estimation of the transcapillary gradient that often proves useful in the postoperative management. Near-infrared spectroscopy (NIRS) is commonly utilized to assess trends on regional perfusion, and some centers also use technologies based on thermodilution and on pulse wave analysis.

30.7.4.2 General Measures

After a partial cavopulmonary connection, general measures to be undertaken do not change significantly from the measures adopted for any postoperative case, with a few important details to be highlighted:

(a) Patients should be positioned in a 45-degree semi-fowler decubitus, to promote passive venous drainage by gravity.
(b) Enteral feeding should be resumed as soon as possible.
(c) Early extubation and mobilization are essential.
(d) Any indwelling lines, particularly those in the superior venous segments, should be removed as early as possible.
(e) Caution should be taken with regard to the pleural drains; the trend is to be conservative and leave them in situ for at least 48 hours, considering the accrued risk for pleural effusion and chylothorax.

30.7.4.3 Inotropic and Vasodilator Therapy

Usually, these patients require low doses of inovasodilators (i.e., milrinone). Target saturations are between 75 and 80%, central venous pressures are around 5–8 mm Hg, and mean pulmonary pressures should be below 15 mm Hg with a low trans-pulmonary gradient. It is common to observe a transient systemic hypertension, quite likely of central origin.

30.7.4.4 Respiratory Management

Cardiopulmonary interactions are fundamental in the context of the cavopulmonary connections. Ventilatory parameters should be rigorously monitored to preserve both the pulmonary and the cerebral blood flow. Hyperventilation should be avoided, since although facilitating pulmonary flow (by raising pH), it may decrease cerebral flow (by decreasing CO_2) which provides the main preload of the cavopulmonary system. As a matter of fact, mild permissive hypercapnia should not preclude extubation as it increases the cerebral blood flow and therefore the Glenn flow. Also, positive intrathoracic pressures induce a reduction of both the pulmonary flow and the systemic ventricle preload with an increase of pulmonary vascular resistances. Therefore, most patients are extubated in the operating room or during the first 6 postoperative hours, once there is evidence of hemodynamic, neurologic, respiratory, and homeostatic stability and controlled bleeding. Ventilation is better tolerated after the partial connection rather than the total connection because in the first

case, flow from the inferior vena cava to the heart fills the systemic ventricle independently. Any respiratory complications like atelectasis, pneumothorax, or pleural effusions should be promptly rectified.

30.7.4.5 Sedation and Analgesia

Postoperative sedation and analgesia should target proper levels of comfort while ensuring spontaneous breathing autonomy allowing early extubation. A balance must be established to avoid pain, allow proper cough and airway protection (to reduce risks of atelectasis), and reduce the typical irritability that characterizes these patients, secondarily to transient cerebral venous congestion and changes in cerebral flow patterns.

30.7.4.6 Anticoagulation

Prophylactic anticoagulation with heparin should be started in the absence of bleeding. Once feeding is resumed, antiplatelet therapy with aspirin (3–5 mg/kg/day) may be started.

30.7.5 Postoperative Complications

The four main complications observed after the partial cavopulmonary connection are increased pressures in the cavopulmonary circuit, hypertension and bradycardia, low cardiac output syndrome (LCOS), and hypoxemia.

30.7.5.1 Increased Pressures in the Cavopulmonary Circuit

Immediately after surgery, there may be a transient increase of pressures in the cavopulmonary circuit, secondarily to the inflammatory changes induced by the cardiopulmonary bypass, volume overload, and the mechanical ventilation with positive pressure. The clinical expression of this complication is the development of a superior vena cava syndrome, associated with increased pulmonary pressures, progressive cyanosis, and decrease in the systemic stroke volume. It is therefore important to establish a spontaneous breathing pattern as early as possible, to aggressively manage any respiratory occurrence (atelectasis, pneumothorax, or pleural effusions), and to use pulmonary vasodilators as required, mostly nitric oxide and sildenafil [14, 15], and loop diuretics to induce diuresis and a negative fluid balance. When pressures remain high in spite of these measures, further investigations (echocardiography, cardiac catheterization) may be indicated to rule out stenosis at the anastomotic site or else, distally in the pulmonary arteries, thrombosis, or high pulmonary vascular resistances.

30.7.5.2 Hypertension and Bradycardia

The mechanism behind hypertension might be inadequate analgesia and sedation, stress response with release of endogenous catecholamines, or a down-regulator effect to maintain cerebral perfusion in the context of high venous pressure and congestion. This phenomenon is usually transient and well controlled with angiotensin inhibitors, but during the acute phase, these patients might need the use of sodium nitroprusside or intravenous calcium inhibitors.

Bradycardia is usually a reflex response to the sudden unload induced by the Glenn connection, although in a small percentage of cases, it may be due to a lesion of the sinus node, in which case it is unresponsive to drugs like atropine or isoproterenol.

30.7.5.3 Low Cardiac Output Syndrome (LCOS)

Significant low cardiac output syndrome is seldom observed after a partial cavopulmonary connection, except in patients with previous ventricular dysfunction or with atrioventricular valve regurgitation, in whom a sudden unload may have an impact in cardiac output.

30.7.5.4 Hypoxemia

Hypoxemia is the most common short- and long-term complication after a partial cavopulmonary connection. Initial saturations are expected to be between 75 and 85%. Persistent saturations below 70% justify further investigations. Etiology of persistent hypoxemia is variable and heterogeneous and may include decreased cerebral flow (hypocapnia, hypotension), ventilation/perfusion mismatch (pleural effusion, atelectasis, pneumothorax, pneumonia, arteriovenous malformations), increased oxygen consumption (sepsis, low cardiac output, ventricular dysfunction, anemia), or decreased pulmonary blood flow (increased pulmonary vascular resistances, stenosis of the cavopulmonary anastomosis, veno-venous collateral vessels, restrictive intra-atrial communication). Veno-venous collateral vessels between the superior and inferior venous territories induce a persistent desaturation by decreasing the effective pulmonary flow, and some may be occluded by percutaneous interventional catheterization. Arteriovenous malformations (AVM) are a common cause of late hypoxemia and are more frequently documented in heterotaxic syndromes with interrupted infe-

rior vena cava and azygos continuation. They are thought to develop because of the absence of a hepatic angiogenesis inhibitor factor. Diagnosis is suggested by echocardiography with contrast test and confirmed by angiography. AVMs tend to progress after the completion of the Fontan circuit. Patients are more prone to develop AVMs and chronic hypoxemia after a Kawashima intervention.

Currently, partial cavopulmonary connections carry a very low morbidity and mortality is close to 0%.

30.8 Total Cavopulmonary Connections

30.8.1 The Fontan-Kreutzer and Modified Fontan Procedures

Francis Fontan originally described a procedure [16–18] by which the right atrial cavity would be directly anastomosed onto the pulmonary artery. Guillermo Kreutzer described a similar technique contemporarily. This original Fontan-Kreutzer operation lately showed to have a number of disadvantages, due to persistent blood stasis in the right atrium. Concomitantly, it was recognized in the early 1980s that to proceed with a total cavopulmonary connection in one surgical step would have a significant mortality due to a significant acute remodeling of the systemic ventricle. Since inception of the original technique, three essential modifications were described and developed for patients who had already undergone the Glenn or the hemi-Fontan connection as a first step intervention [19, 20], in order to avoid or limit the remodeling phenomenon. Firstly, the anastomosis between the inferior vena cava, the hepatic veins, and the pulmonary artery was performed as an intracardiac shunt created with the wall of the right atrium and with Goretex® and diverting the blood draining from the inferior segment of the body toward the pulmonary artery [21]. Secondly, a more recent modification was proposed by diverting the blood with an extracardiac conduit [22–26] (Fig. 30.14), a less traumatic surgery and theoretically reducing the risks for arrhythmia, the latter not having been demonstrated so far. The third modification consists in the creation of a fenestration [27, 28] between the intra- or the extracardiac conduit and the atrial mass. This fenestration acts like a "pop-off" structure that is functionally useful in patients in whom the pulmonary pressures and resistances are above the desired levels. Although inducing cyanosis, this fenestration allows a more stable and adequate hemodynamic profile and decreases the risk of persistent "right failure" with superior vena cava syndrome, peripheral edema, pleural effusions, ascites, and protein-losing enteropathy. More recent modifications such as the one proposed by Viktor Hraska (diversion of the innominate vein to the atrial mass to reduce strain on the thoracic duct drainage) [29] have been proposed but have yet to show consistency or reproducibility.

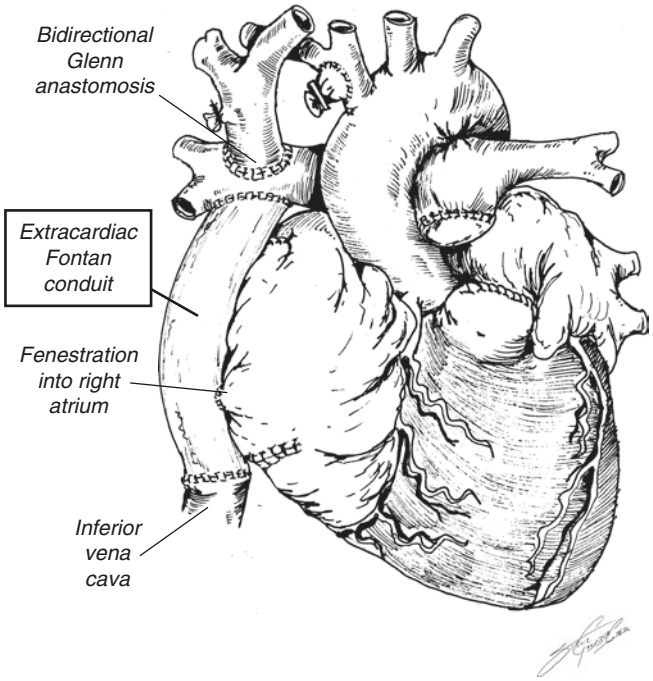

Fig. 30.14 Extracardiac fenestrated modified Fontan or total cavopulmonary connection

30.8.2 Postoperative Management

30.8.2.1 Monitoring

After a total cavopulmonary connection, patients are comprehensively monitored with heart rate and ECG, respiratory rate, oxygen saturation, and arterial pressure both invasive and noninvasive and also have a transthoracic atrial catheter and an internal jugular indwelling catheter to assess the pulmonary artery pressure. Indwelling catheters should be removed as soon as possible to minimize risks for thrombosis in the cavopulmonary circuit. Near-infrared spectroscopy (NIRS) and modern technology by thermodilution may also be instrumental in taking therapeutic decisions with these patients.

30.8.2.2 General Measures

As for the partial cavopulmonary connections, general measures are the same as for any postoperative case [30–32], but once again, there are a number of specific details to cautiously follow:

(a) Patients should be positioned in a 45-degree semi-fowler decubitus with partly folded legs, to promote passive venous drainage from both the superior and the inferior segments of the body.
(b) Enteral feeding should be resumed as soon as possible.
(c) Early mobilization is crucial.
(d) Any indwelling lines should be removed as early as possible.
(e) Caution should be taken with regard to the pleural and mediastinal drains since these patients are prone to develop pericardial and pleural effusions and chylothorax, mostly when the pulmonary pressures are high or in the upper normal range.

30.8.2.3 Inotropic and Vasodilator Therapy

After a total cavopulmonary connection, patients often need low to moderate doses of inovasodilators or lusitropic drugs (milrinone). Target saturations are expected to be above 90%, and central venous pressures that correspond to mean pulmonary pressures should be below 15 mm Hg with a low trans-pulmonary gradient. As for the Glenn procedure, it is common to observe a transient systemic hypertension or transient peripheral and central edema.

30.8.2.4 Respiratory Management

Cardiopulmonary interactions are fundamental after the total cavopulmonary connection. Ventilatory parameters should be rigorously monitored in order to preserve both the pulmonary and the cerebral blood flow. Hyperventilation should be avoided. Caregivers need to keep in mind that positive intrathoracic pressures induce a reduction of both the pulmonary flow and the systemic ventricle preload with an increase of pulmonary vascular resistances. Ventilation is better tolerated after the partial connection rather than the total connection because in the first case, flow from the inferior vena cava to the heart fills the systemic ventricle independently. Consequently, most Fontan patients are extubated in the early phase of their postoperative course, once there is evidence of hemodynamic, neurologic, respiratory, and homeostatic stability and controlled bleeding. Any respiratory complications like atelectasis, pneumothorax, or pleural effusions should be promptly rectified.

Patients with persistent low saturations ("right-to-left" shunt through the fenestration) and high pulmonary pressures may benefit from inhaled nitric oxide, although its administration has to be cautious when the function of the systemic ventricle is borderline.

30.8.2.5 Sedation and Analgesia

Postoperative sedation and analgesia should target proper levels of comfort while ensuring spontaneous breathing autonomy allowing early extubation. A balance must be established to avoid pain, allow proper cough and airway protection (to reduce risks of atelectasis), and reduce the typical irritability that characterizes these patients, secondarily to transient cerebral venous congestion and changes in cerebral flow patterns.

30.8.2.6 Anticoagulation

There is no current consensus regarding the potential benefit of long-term anticoagulation versus antiplatelet aggregation therapy after a total cavopulmonary connection. Nevertheless, acute prophylactic anticoagulation is universally ensured with heparin as previously described. Once feeding is resumed, heparin is replaced by antiplatelet therapy with aspirin (3-5 mg/kg/day), alternatively with dipyridamole or clopidogrel or by anticoagulation with antivitamin K agents as opposed to antiplatelet drugs.

Patients with dysfunctional Fontan physiology or with documented pro-coagulant status should be considered for active anticoagulation with oral antivitamin K agents and/or with subcutaneous heparin, taking into account their risk for thrombosis.

30.8.3 Complications

A*nticipated potential complications* after the total cavopulmonary connection are similar to the occurrences after the partial connection (increased pressures in the cavopulmonary circuit, low cardiac output syndrome, hypoxemia), but there is a higher prevalence and risk of pleural effusions, chylothorax, pericardial effusion, arrhythmias, and thromboembolic events [33, 34]. Other chronic complications are the development of venous collaterals, pulmonary arteriovenous malformations or systemic-pulmonary arterial collaterals, failure to thrive, protein-losing enteropathy (PLE) [35], low functional capacity, plastic bronchitis, and liver disease, cirrhosis, and neoplasia [36].

Management of acute complications follows the recommendations described herein in the section dedicated to the postoperative course of the partial cavopulmonary connection. There are a few peculiarities related to the Fontan operation though.

Significant low cardiac output syndrome may be observed after a total cavopulmonary connection, mostly in patients

with previous ventricular dysfunction, with severe atrioventricular valve regurgitation or with a tenuous hemodynamic stability. Treatment is based on the use of inotropic, vasodilator, or lusitropic drugs, induced hypothermia, diuretics, and eventually re-synchronization strategies. Patients with refractory LCOS may require mechanical assistance and may be considered for cardiac transplant.

Hypoxemia is rather common in patients with fenestrations and moderately or severely increased postoperative pulmonary resistances. Persistent saturations below 90% justify further investigations as described above for patients with increased pulmonary pressures, situation that usually coexists with the hypoxemia. A cardiac catheterization may also be necessary to rule out anatomic or functional obstructions, thrombosis, or high pulmonary vascular resistances. It may also identify veno-venous or arterial-venous fistula requiring embolization [37–39]. In some circumstances, it is useful to transiently occlude the fenestration and reassess hemodynamics. When the pulmonary pressures remain below 18 mm Hg, systemic saturations increase significantly, and there is no impact in the cardiac function, a definite occlusion of the fenestration with an intra-vascular device may be considered.

Acute and chronic effusions confined to the thorax (pleural or pericardial effusions, chylothorax) or extra-thoracic (ascites, peripheral edema) are common after total cavopulmonary connections [40]. Conceivably, the presence of a fenestration is a favorable preventive factor. These patients may require intra-thoracic drains for long periods of time. In case of persistent chylothorax, an adequate diet with middle-chain triglycerides and the use of parenteral feeding is indicated. In refractory cases, somatostatin or octreotide followed by pleurodesis may be considered. Selective thoracic duct embolization is a promising interventional technique that fosters encouraging prognosis in patients with refractory effusions, PLE, and plastic bronchitis [41, 42]. The partial hepatic vein exclusion reported by Yves Lecompte [43, 44] is not currently performed since most patients develop intrahepatic venous collaterals with significant right-to-left shunts. Nevertheless, recent case reports describe modifications of the technique with varying degrees of success, namely, in patients with associated PLE [45, 46].

Patients with total cavopulmonary connections tolerate very poorly any arrhythmia [47, 48] or conductive disorder and must be aggressively managed. Atrioventricular synchrony is vital in these patients. Nodal or junctional rhythm is a common immediate "benign" postoperative finding, usually well tolerated but sometimes requiring AAI pacing. Atrial flutter is the most common potentially life-threatening arrhythmia in this context and is often associated with sinus node dysfunction, which complicates its management. It often gives a sign of alert for serious hemodynamic complications. As previous mentioned, the use of extracardiac conduits has not reduced the incidence of arrhythmias or sinus node dysfunction. Atrial flutter may be preceded by sinus bradycardia in which case the use of a prophylactic epicardial pacemaker may be a benefit, although this has yet to be demonstrated. Technical complications for the use of mono- or dual-chamber pacemakers are related to the fact that these can only be epicardial, in patients who often have chronic adhesions secondary to multiple surgeries.

30.9 Conclusions

Single ventricle anatomy and physiology displays a very wide anatomic spectrum. Although medical and surgical management of these patients has been relatively standardized, there remain multiple outliers and challenges in the perioperative management of these complex patients. Constant innovation, data collection, patient- and family-centered interdisciplinary team collaboration focused on quality and safety, and multicentric research are key factors for further understanding and consistently and efficiently managing single ventricle patients.

References

1. Penny DJ, Redington AN. Doppler echocardiographic evaluation of pulmonary blood flow after the Fontan operation: the role of the lungs. *Br Heart J*. 1991;66:372–374.
2. Dorfman AL, Odegard KC, Powell AJ, Laussen PC, Geva T. Risk factors for adverse events during cardiovascular magnetic resonance in congenital heart disease. *J Cardiovasc Magn Reson*. 2007;9:793–798.
3. Fogel MA, Weinberg PM, Parave E, et al. Deep sedation for cardiac magnetic resonance imaging: a comparison with cardiac anesthesia. *J Pediatr*. 2008;152:534–539.
4. Fogel MA, Weinberg PM, Rychik J, et al. Caval contribution to flow in the branch pulmonary arteries of Fontan patients with a novel application of magnetic resonance presaturation pulse. *Circulation*. 1999;99:1215–1221.
5. Williamson BD, Gohn DC, Ramza BM et al. Real-World evaluation of magnetic resonance imaging in patients with a magnetic resonance imaging conditional pacemaker system results of 4-year prospective follow-up in 2,629 patients. *JACC Clin Electrophysiol*. 2017;3:1231–1239.
6. Marino BS, Spray TL. Separating the circulations: cavopulmonary connections (Bidirectional Glenn, Hemi-Fontan) and the modified Fontan operation. In: Nichols DG, Ungerleider RM, eds *Critical Heart Disease in Infants and Children*. Philadelphia: Mosby; 2006.
7. Rydberg A, Teien DE, Krus P. Computer simulation of circulation in patient with total cavo-pulmonary connection: inter-relationship of cardiac and vascular pressure, flow, resistance and capacitance. *Med Biol Eng Comput*. 1997;35:722–728.
8. Senzaki H, Masutani S, Kobayashi J, et al. Ventricular afterload and ventricular work in Fontan circulation: comparison with normal two-ventricle circulation and single-ventricle circulation with Blalock-Taussig shunts. *Circulation*. 2002;105:2885–2892.

9. Jonas R. *Single Ventricle. Comprehensive Surgical Management of Congenital Heart Disease*. 1st ed. Boca Raton: Hodder Arnold Publication. Hardcover; 2004. Cap 20 Pag 357–385.
10. Ro PS, Rome JJ, Cohen MS, et al. Diagnostic assessment before Fontan operation in patients with bidirectional cavopulmonary anastomosis: are noninvasive methods sufficient? *J Am Echocardiogr*. 2000;13:452.
11. Fogel M.A. Is routine cardiac catheterization necessary in the management of patients with single ventricles across staged Fontan reconstruction? No! *Pediatr Cardiol*. 2005;26:154–158.
12. Nakanishi T. Cardiac catheterization is necessary before Glenn and Fontan procedures in single ventricle physiology. *Pediatr Cardiol*. 2005;26:159–161.
13. Jacobs ML, Rychik J, Rome JJ, et al. Early reduction of the volume work of the single ventricle: the hemi-Fontan operation. *Ann Thorac Surg*. 1996;62:456–461.
14. Yoshimura N, Yamaguchi M, Oka S, Yoshida M, Murakami H, Kagawa T, Suzuki T. Inhaled nitric oxide therapy after Fontan type operations. *Surg Today*. 2005;35:31–35.
15. Agarwal HS, Churchwell KB. Inhaled nitric oxide use in bidirectional Glenn anastomosis for elevated Glenn pressures. *Ann Thorac Surg*. 2006;81:1429–1435.
16. Fontan F, Mouniccot F, Baudet E, Simonneau J, Gordo J, Gouffrant J. Correction de l'atrésie tricuspidienne: rapport de deux cas corrigés par l'utilization d'une technique chirurgicale nouvelle. *Ann Chir Thorac Cardiovasc*. 1971;10:39–47.
17. Kreutzer G, Galíndez E, Bono H, De Plama C, Laura JP. An operation for the correction of tricuspid atresia. *J Thorac Cardiovasc*. 1973;66:613–621.
18. Kreutzer GO, Schlichter AJ, Kreutzer C. The Fontan/Kreutzer procedure at 40: an operation for the correction of tricuspid atresia. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annua*. 2010;13:84–90.
19. Jacobs ML, Norwood WI Jr. Fontan operation: influence of modifications on morbidity and mortality. *Ann Thorac Surg*. 1994;58:945–951.
20. Reul GJ, Gregoric ID. Recent modifications of the Fontan procedure for complex congenital heart disease. *Tex Heart Inst J*. 1992;19:223–231.
21. De Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. Experimental studies and early clinical experience. *J Thorac Cardiovasc Surg*. 1988;96:682–695.
22. Ocello S, Salviato N, Marcelletti CF. Results of 100 consecutive extracardiac conduit Fontan operations. *Pediatr Cardiol*. 2007;28:433–437.
23. Marcelletti C, Corno A, Giannico S, Marino B. Inferior vena cava-pulmonary artery extracardiac conduit. A new form of right heart bypass. *J Thorac Cardiovasc Surg*. 1990;100:228–232.
24. Fiore AC, Turrentine M, Rodefeld M, Vijay P, Schawartz TL, Virgo KS, Fischer LK, Brown JW. Fontan operation: a comparison of lateral tunnel with extracardiac conduit. *Ann Thorac Surg*. 2007;83:622–630.
25. Kumar SP, Rubinstein CS, Simsic JM, Taylor AB, Saul JP, Bradley SM. Lateral tunnel versus extracardiac Fontan procedure: a concurrent comparison. *Ann Thorac Surg.* 2003;76:1389–1397.
26. Petrossian E, Reddy VM, McElhinney DB, et al. Early results of the extracardiac conduit Fontan operation. *J Thorac Cardiovasc Surg*. 1999;117:688–696.
27. Lemler MS, Sott WA, Leonard SR, Stromberg D, Ramiciotti C. Fenestration improves clinical outcome of the Fontan procedure. *Circulation*. 2002;105:207–212.
28. Kreutzer J, Lock JE, Jonas RA, Keane JF. Transcatheter fenestration dilatation and/or creation in postoperative Fontan patients. *Am J Cardiol*. 1997;79:228–832.
29. Hraska V. Decompression of thoracic duct: new approach for the treatment of failing Fontan. Ann Thorac Surg. 2013;96:709–711.
30. Gersony DR, Gersony WM. Management of the postoperative Fontan patient. *Prog Pediatr Cardiol*. 2003;17:73–79.
31. Bull K. The Fontan procedure: lessons from the past. *Heart*. 1998;79:213–4.
32. Meyer DB, Zamora G, Wernovsky G, Ittenbach RF, Gallagher PR, Tabbutt S, Gruber PJ, Nicolson SC, Gaynor W, Spray TL. Outcomes of the Fontan procedure using cardiopulmonary Bypass with aortic cross-clamping. *Ann Thorac Surg*. 2006;82:1611–1620.
33. Coon P, Rychik J, Novello R, Ro PS, Gaynor W, Spray TL. Thrombus Formation after the Fontan Operation. *Ann Thorac Surg*. 2001;71:1990–1994.
34. Monagle P. Thrombosis in children with BT shunts, Glenns and Fontans. *Prog Pediatr Cardiol*. 2005;21:17–21.
35. Cheung YF, Tsang HY, Kwok JS. Immunologic profile of patients with protein-losing enteropathy complicating congenital heart disease. *Pediatr Cardiol*. 2002;23:587–593.
36. Narkewicz MR, Sondheimer HM, Ziegler JW, et al. Hepatic dysfunction following the Fontan procedure. *J Pediatr Gastroenterol Nutr*. 2003;36:352–357.
37. Chang RK, Alejos JC, Atkinson D, et al. Bubble contrast echocardiography in detecting pulmonary arteriovenous shunting in children with univentricular heart after cavopulmonary anastomosis. *J Am Coll Cardiol*. 1999;33:2052–2058.
38. Sacuitto RJ, Ross Ascuitto NT. Systematic-to-pulmonary collaterals: a source of flow energy loss in Fontan physiology. *Pediatr Cardiol*. 2004;25:472–481.
39. McElhinney D, Reddy VM, Hanley FL, Moore P. Systemic venous collateral channels causing desaturation after bidirectional cavopulmonary anastomosis: evaluation and management. *J Am Coll Cardiol*. 1997;30:817–824
40. Cava JR, Bevandic SM, Stelzer MM, Tweddell JS. A medical strategy to reduce persistent chest tube drainage after the Fontan operation. *Am J Cardiol*. 2005;96:130–133.
41. Dori Y, Keller MS, Rome JJ, Gillespie MJ, Glatz AC, Dodds K, Goldberg DJ, Goldfarb S, Rychik J, Itkin M. Percutaneous lymphatic embolization of abnormal pulmonary lymphatic flow as treatment of plastic Bronchitis in patients with congenital heart disease. *Circulation*. 2016;133:1160–1170.
42. Itkin M, Rychik J, Piccoli DA. Reply: the need to define treatment goals for protein-losing enteropathy in Fontan Care and Research. *J Am Coll Cardiol*. 2017;70:2603.
43. Lecompte Y. Subtotal cavopulmonary connection. *J Thorac Cardiovasc Surg*. 1992;104:1500
44. McElhinney DB, Kreutzer J. Incorporation of the hepatic veins into the cavopulmonary circulation in patients with heterotaxy and pulmonary arteriovenous malformations after a Kawashima procedure. *Ann Thorac Surg*. 2005;80:1597–1603.
45. Brizard CP, Lane GK, Alex G, Cheung MM. Original surgical procedure for the treatment of protein-losing enteropathy in Fontan patients: report of two midterm successes. *Circulation*. 2016;134:625–627.
46. Antonio M, Gordo A, Pereira C, Pinto F, Fragata I, Fragata J. Thoracic duct decompression for protein-losing enteropathy in failing Fontan circulation. *Ann Thorac Surg*. 2016;101:2370–2373.
47. Durongpisitkul K, Porter CJ, Cetta F, et al. Predictors of early- and late-onset supraventricular tachyarrhythmias after Fontan operation. *Circulation*. 1998;98:1099–1107.
48. Hasselman T, Schneider D, Mdan N, Jacobs M. Reversal of fenestration flow during ventricular systole in Fontan patients in junctional or ventricular paced rhythm. *Pediatr Cardiol*. 2005;26:638–641.