CHAPTER 28

Echocardiographic Assessment of Functionally Single Ventricles after the Fontan Operation

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Introduction

For the majority of patients with different types of functionally univentricular hearts, the treatment goal is to achieve palliation through the creation of a total cavopulmonary anastomotic connection known as the Fontan operation [1]. This operation essentially bypasses the right heart by directing the systemic venous return into the pulmonary circulation without passing through a ventricular chamber. The Fontan operation causes a unique type of circulation, which requires a specific echocardiographic approach. In the current chapter we will first review the specific physiologic characteristics of a Fontan circulation followed by a discussion of the echocardiographic evaluation.

The Fontan circulation

A variety of cardiac malformations are characterized by the presence of a hypoplastic ventricle that is too small to sustain one of the circulations resulting in a functionally single ventricular (FSV) heart that has to support both circulations. During fetal life and immediately after birth the systemic and the pulmonary circulations in a FSV are connected in parallel with one ventricular chamber pumping to both circulations usually by way of the patent ductus arteriosus (Figures 28.1a,b). In the pre-Fontan era, these patients were palliated by an aortopulmonary artery shunt (e.g., Blalock–Taussig shunt) or pulmonary artery banding (both outflows unobstructed) which resulted in persistence of the parallel circuit. This palliation caused persistent arterial desaturation and chronic volume loading of the FSV. These procedures contributed to progressive ventricular dysfunction, leading to FSV failure, congestive heart failure and death with few survivors beyond the fourth decade. In 1971, Francis Fontan reported on a new surgical approach [1], separating the systemic and pulmonary circulations and redirecting the systemic venous blood directly to the pulmonary circulation. In the Fontan circulation, mixing of the circulations is avoided and the post-capillary energy and systemic venous pressure are used as the driving forces to push the blood through the pulmonary circulation (Figure 28.1c) [2]. Advantages of a Fontan circuit include separation of both circulations with normalization of the arterial saturation, and abolishment of chronic ventricular volume loading. The primary disadvantage is the resultant elevated systemic venous pressure and passive flow through the pulmonary vascular bed which causes chronic systemic venous hypertension, reduced cardiac output and a limited reserve during exercise (Figure 28.2).

By creating a cavopulmonary connection, a new portal system is created. In a portal system a capillary bed pools blood into another capillary bed through veins without passing through the heart. This is most typically seen in the normal gastrointestinal system where the superior mesenteric vein and splenic vein are connected via the portal vein to the hepatic veins. The Fontan portal system pools the systemic venous blood in the pulmonary circulation and limits the pulmonary venous return to the heart due to different static resistances and dynamic impedances in the circuit. The optimal Fontan circuit requires unobstructed Fontan connections, good-sized branch pulmonary arteries without stenosis, low pulmonary vascular resistance, and unobstructed pulmonary venous connection to the atrium and low atrial pressures. Atrial pressures are influenced by atrioventricular valve function and by FSV diastolic function. Good systolic FSV function is required to generate sufficient systemic output; chronic volume load (residual shunts, atrioventricular valve regurgitation, aortic regurgitation) and/or pressure load (outflow tract obstruction, residual coarctation of the aorta), even if mild can be very detrimental in patients with this physiology.
Figure 28.1 Schematic representation of (a) normal cardiovascular circulation, (b) shunted palliation, and (c) Fontan circulation. (a) The normal circulation: the pulmonary circulation (P) is connected in series with the systemic circulation (S). The right ventricle (RV) is more compliant than the left ventricle keeping the right atrial (RA) pressure lower than the left atrial (LA) pressure. The RV functions at lower pressure than the LV which is related to lower pressure and resistance in the pulmonary artery (PA). (b) Parallel circulation in a univentricular heart. The systemic (S) and pulmonary (P) circuits are connected in parallel with one dominant single ventricle (V) pumping blood into both circulations. There is complete admixture of systemic and pulmonary venous blood, causing arterial desaturation. (c) Fontan circuit. The systemic veins (CV) are connected to the pulmonary artery (PA), without a subpulmonary ventricle or systemic atrium: the lungs are hereby converted into a portal system which limits flow to the ventricle. In the absence of a fenestration, there is no admixture of systemic and pulmonary venous blood, but the systemic venous pressures are generally elevated. Ao, aorta; CV, caval veins; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; V, single ventricle. Line thickness reflects output, color reflects oxygen saturation.

The functional single ventricle in a Fontan circulation

Ventricular dimensions and wall thickness

Figure 28.3 demonstrates the evolution of the FSV volume loading from fetal life until after completion of the Fontan circuit. During fetal life, the dominant ventricle is responsible for the combined output of the systemic and pulmonary circulations. This causes prenatal eccentric remodeling resulting in enlarged chamber dimensions in the single ventricle at birth [3]. After the initial neonatal palliation, (generally an aortopulmonary artery shunt or a pulmonary artery band), the parallel circulations cause a persistent, chronic volume load. The bidirectional Glenn shunt (a superior cavopulmonary anastomosis where only the
Cardiac output during exercise output: normal versus Fontan

Effect of different pre-Fontan palliative strategies on volume

Figure 28.2 Cardiac output during exercise output: normal versus Fontan circulation. A normal subject with a biventricular circuit can increase output five times compared to baseline (black line). In Fontan patients output this is significantly impaired both at rest and during exercise. In optimal Fontan patients (green line) the output is mildly decreased at rest, with a moderately reduced capacity to increase cardiac output during exercise. This limits exercise capacity in almost all Fontan patients. In failing Fontan patients (red line), the output is significantly reduced at rest with very limited increase during exercise.

Figure 28.3 Effect of different pre-Fontan palliative strategies on volume loading. A: We assume a normal ventricle in a biventricular circulation functions on average at 100% of normal. Prior to birth the dominant single ventricle remodels and its size adjusts to the chronic volume load. The patient with a UVH generally is born with an appropriate ventricular size adjusted for chronic volume load. Prior to the 1990s (B, red line), the volume loading to the ventricle was augmented shortly after birth by a shunt procedure to ±150%. The patient slowly outgrows his shunt, and adapts his ventricle, thereby gradually reducing the volume overload to ±100% for its size. A second shunt was created, augmenting the volume overload again to 150%. As this patient again outgrows his shunt, a Fontan circuit is made, acutely reducing the volume load to around 25% of the preload it was exposed to prior to the procedure. This sudden preload reduction was often poorly tolerated hemodynamically. This resulted in changes in management strategy. After the 1990s (C, green line), a small neonatal shunt was created for a short time (generally 6 months); the patient slowly outgrows the shunt; the ventricle is progressively volume unloaded in two separate steps: first, at the time of the bidirectional Glenn shunt, and a second time, at the time of the Fontan operation. This stepwise approach is generally hemodynamically better tolerated.

superior vena cava is connected to the pulmonary artery and the inferior vena cava remains connected to the right atrium) is generally performed at 4 to 8 months of age to address this issue. After this procedure, the chronic volume load is reduced. A recent serial follow-up study using cardiac MRI in patients scanned before the bidirectional Glenn and again before the Fontan operation, demonstrated that the bidirectional Glenn shunt resulted in a reduction of indexed ventricular volume with an increase in ejection fraction (i.e., reverse remodeling) [4]. Quantitative measurements by cardiac MRI have also demonstrated that flow through aortopulmonary collaterals contributes significantly to pulmonary blood flow and cardiac output after the bidirectional Glenn shunt [5]; up to one third of the cardiac output and up to 40% of the pulmonary blood flow comes from collateral flow. A higher burden of collateral flow adversely influences clinical outcomes immediately after the Fontan operation [5,6]. Despite the presence of collaterals, the FSV is volume unloaded after the Fontan operation due to a reduction in pulmonary venous return (with a reduced preload reserve being one of the characteristic features of the Fontan circulation). After the Fontan operation, further reverse remodeling occurs but the FSV is characterized by a detrimental decrease in ventricular dilation with persistence of an abnormal wall thickness and incomplete resolution of hypertrophy (eccentric remodeling), often termed increased mass-to-volume ratio [7]. Concentric hypertrophy may also develop in the FSV in response to increased afterload, defined as ventricular wall stress. Studies have shown that increased afterload (i.e., outflow tract obstruction, residual coarctation of the aorta or systemic hypertension) may persist after the Fontan operation [8,9]. Residual ventricular hypertrophy may contribute to diastolic dysfunction and eventually to failure of the Fontan circulation.

Effect of loading changes on parameters of systolic function

The most commonly used parameter to assess global pump function is ejection fraction. This parameter is sensitive to changes in preload and afterload. Recent serial data obtained with either MRI or echocardiography has demonstrated a decrease in ventricular volumes and ejection fraction immediately after the bidirectional Glenn shunt [4,10]. During the remodeling process a compensatory decrease in end-systolic volumes may result in an increase in ejection fraction just prior to the Fontan procedure [4]. Despite these findings, ejection fraction is generally preserved in the majority of patients after the Fontan operation suggesting that adaptation or further reverse remodeling occurs in the post-Fontan period [7]. It also indicates that systolic dysfunction is uncommon in pediatric Fontan survivors. In adult patients, ventricular dysfunction seems to be more common and is an important cause of failure of the Fontan circuit [11]. Systolic dysfunction may be intrinsic to the FSV or may be related to other comorbidities in adult life.
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Diastolic pressure–volume relationship during different stages of pediatric Fontan patients have abnormal diastolic function. Recent cohorts suggest that the majority of these patients have abnormal diastolic function due to chronic volume unloading and eventually declining cardiac output. The Fontan syndrome (FSV) may enter a vicious cycle whereby the volume unloading results in adverse reverse remodeling, reduced compliance, and eventually declining cardiac output. In patients after Fontan procedures, the chronically volume-loaded ventricles were found to have abnormal diastolic and systolic abnormalities, especially affecting the isovolumic relaxation period and early filling. These diastolic changes have typically been observed in FSV patients who underwent a Fontan operation without a previous bidirectional Glenn shunt. In this era, the chronically volume-loaded ventricles were acutely unloaded and the observed diastolic and systolic abnormalities could be related to the ventricle being “too large” for the volume immediately after the procedure. In patients after the Fontan procedure, chronic volume reduction is thought to cause progressive stiffening of the ventricle with decreased compliance.

Diastolic function of the single ventricle

In the Fontan circuit, low atrial pressure is required for a low-pressure gradient across the pulmonary vascular bed (transpulmonary gradient). Atrioventricular valve regurgitation and single ventricular diastolic function are important determinants of atrial pressure. Diastolic function after the Fontan operation has been poorly studied due to the absence of good methods for assessment of ventricular filling in patients with this physiology. Experimental data have shown that acute volume unloading of the ventricle results in less recoil and decreased suction resulting in increased filling pressures (Figure 28.4) [12–15]. Echocardiographic studies in Fontan patients have suggested that the volume reduction associated with the Fontan operation results in early relaxation abnormalities and incoordinate wall motion abnormalities especially affecting the isovolumic relaxation period and early filling [13–15]. These diastolic changes have typically been observed in FSV patients who underwent a Fontan operation without a previous bidirectional Glenn shunt. In this era, the chronically volume-loaded ventricles were acutely unloaded and the observed diastolic and systolic abnormalities could be related to the ventricle being “too large” for the volume immediately after the procedure. In patients after the Fontan procedure, chronic volume reduction seems to cause a progressive decrease in ventricular compliance with a left upward shift in the end-diastolic pressure–volume relationship [16]. Moreover, the thicker myocardium may become stiffer with age. The FSV may enter a vicious cycle whereby the volume unloading results in adverse reverse remodeling, reduced compliance, reduced ventricular filling, and eventually declining cardiac output. A recent cohort study suggests that the majority of pediatric Fontan patients have abnormal diastolic function but the clinical and prognostic significance of these findings is currently unknown [7]. The primary issue regarding assessment of diastolic function is that the conventional echocardiographic parameters (atrioventricular valve inflow, pulmonary venous flow patterns, and tissue Doppler velocities) are difficult to interpret in Fontan patients. Progressive increase in ventricular stiffness and reduction in compliance may be one of the major mechanisms explaining Fontan failure with progressive age.

The pulmonary circulation limits cardiac output after the Fontan operation

A characteristic feature of the Fontan circuit is the lack of pulsatile pulmonary blood flow through the pulmonary vascular bed. In the normal biventricular circulation pulmonary blood flow is generally not a limiting factor for determining cardiac output. In ischemic heart disease and cardiomyopathy, decreased cardiac pump function limits cardiac output. After the Fontan procedure the pulmonary circulation limits the preload reserve to the FSV and determines the cardiac output response [8,17]. This is comparable to obstructed inflow after Mustard repair, primary pulmonary hypertension, constrictive pericarditis, and mitral stenosis.

Different factors influence the amount of pulmonary blood flow and cardiac output after the Fontan operation. The first limiting factor is the energy loss in the surgical connections. The presence of unobstructed surgical connections between the caval veins and the pulmonary arteries is a requirement for optimal Fontan function. Any degree of obstruction of the Fontan connections will reduce cardiac output. The pulmonary arteries need to be good-sized and unobstructed. Pulmonary vascular resistance (PVR) further determines pulmonary blood flow. Even a mild increase in PVR reduces pulmonary blood supply and cardiac output [18]. Recent data have shown that pulmonary vasodilators like sildenafil positively influence cardiac output and exercise capacity in Fontan patients [19,20]. Pulmonary venous obstruction is another important factor determining pulmonary blood flow. Some patients with FSV also have anomalous pulmonary venous connection that requires surgical intervention; they are at high risk for reobstruction. The extracardiac Fontan conduit may also compress the adjacent right pulmonary veins. Rarely, the left lower pulmonary vein may become compressed between the heart, the descending aorta and the spine.

In the Fontan circuit, pulmonary blood flow will increase with inspiration and decrease with expiration [21]. Studying respiratory variation in Fontan patients has provided interesting physiologic information. Increased intrathoracic pressure due to airway obstruction or positive pressure ventilation reduces pulmonary blood flow and is detrimental for Fontan physiology [22].
**Fontan modifications**

Since its original description, the Fontan circuit has undergone several surgical modifications (Figure 28.5) [23,24]. The original Fontan operation included the hypoplastic right ventricle in the circuit. This required a valved surgical conduit to connect the hypoplastic ventricle with the pulmonary artery. These conduits degenerated and conduit obstruction was a major problem resulting in a high reoperation rate. This initial approach from Francis Fontan was quickly abandoned and was replaced by the atriopulmonary connection. This variation became the most commonly used modification as it was thought that including the right atrium in the connection would be beneficial for pulmonary hemodynamics due to the presence of the atrial kick. However, elevated atrial pressure caused progressive right atrial dilation, reducing the energetic efficiency and becoming a substrate for atrial arrhythmia. Experimental and computational modeling was subsequently performed which showed that a total cavopulmonary connection (TCPC) would be hemodynamically more favorable and would result in less energy and power loss in the conduit when compared to atriopulmonary connection [25,26]. In this circuit, the vena cavae are connected directly to the right pulmonary artery. The superior vena cava is connected to the right pulmonary artery (bidirectional Glenn shunt) and the inferior vena cava is connected through an intraatrial (lateral tunnel) baffle or by an extracardiac conduit. The benefits of the lateral tunnel Fontan include use in small children and potential for growth. The possible drawback is that there is substrate for atrial arrhythmia as a result of the suture line in the atrium and the exposure of atrial tissue to high pressure. To avoid these problems, the extracardiac conduit was introduced. This procedure consists of a tube graft between the inferior vena cava and the pulmonary artery placed externally around the right atrium. This circuit leaves the entire atrium at low pressure, has minimal atrial suture lines, and can be performed without aortic cross-clamping. However, this tube graft has no growth potential and therefore is typically offered to larger patients (generally between 2–3 years of life). It remains to be determined whether the extracardiac conduit will ameliorate the arrhythmia risk. It is also more difficult to fenestrate (see later) this type of baffle.

In most centers the extracardiac conduit is currently the preferred technique though some are returning to the lateral tunnel method. When assessing a patient with a Fontan circulation, it is essential to know which type of connection has been made.

**Current strategy towards a Fontan circulation**

At birth, it is fatal to create a Fontan circulation because the pulmonary vascular resistance remains elevated for several weeks to months. Even when resistance falls, a staged approach to Fontan completion is preferred, with the superior and inferior caval veins incorporated into the systemic venous chamber in two separate stages. Thus, the body adapts progressively to the new hemodynamic condition and eccentric hypertrophy, reducing the overall operative morbidity and mortality. In the neonatal period, clinical management aims at achieving unrestricted flow to the aorta (if required: arch repair, Damus–Kaye–Stansel, Norwood procedure), appropriately limited flow to the lungs (if required: pulmonary artery band, aortopulmonary artery shunt, stenting of the arterial duct), and unrestricted return of blood to the ventricle (if required: balloon septostomy, pulmonary venous connection). Between 4 to 8 months of age, the
bidirectional Glenn shunt is performed. After the Glenn shunt, the patient remains mildly cyanotic because the desaturated blood from the inferior vena cava still connects to the systemic ventricle and the aorta. Between 1 to 5 years of age, the Fontan circuit is completed, incorporating the inferior vena cava into the baffle.

Frequently a small fenestration or hole is created between the systemic venous pathway and the pulmonary atrium, either routinely or only in “high-risk” patients. The fenestration allows a residual right-to-left shunt, thereby reducing systemic venous pressure and increasing preload of the systemic ventricle at the expense of cyanosis. A fenestration has been shown to reduce operative mortality and morbidity (e.g., pleural drainage). It can be closed by a percutaneous intervention weeks or months after adaptation of the body to the new hemodynamic condition, particularly if marked cyanosis occurs with exercise.

Complications after the Fontan operation are common and relate to the increased systemic venous pressure and chronic low cardiac output. There may be clinical important early and late mortality, mild to moderate exercise intolerance, ventricular dysfunction, rhythm and conduction abnormalities, hepatomegaly with secondary liver dysfunction and potential for development of liver fibrosis or cirrhosis, lymphatic dysfunction, protein-losing enteropathy, plastic bronchitis, thrombus formation, ascites, and peripheral edema [1,2].

**Echocardiographic assessment of the Fontan circulation**

Before scanning a patient who has undergone a Fontan operation, it is essential to first review the underlying cardiac anatomy and previous surgical history. The ventricular morphology (left, right, or indeterminate) needs to be determined as this influences measurements and interpretation of functional information. It is important to establish the type of Fontan connection that has been performed as well as any additional cardiac surgery before starting to scan. This includes information regarding arch reconstruction, the presence of a Damus–Kaye–Stansel connection (between the ascending aorta and the pulmonary trunk), previous valve surgery, type of previous shunt if performed (modified Blalock–Taussig shunt or other), interventions on the pulmonary arteries (patch, balloon dilatation, or stent implantation) and intervention on the pulmonary veins. Additionally, the echocardiographer should know the clinical indication for the scan. When interpreting the images it helps to know the patient’s clinical status and whether the study is being performed because of a concern or for routine follow-up. Some issues such as residual cyanosis, ascites, protein-losing enteropathy, or other signs of Fontan failure may require specific image acquisition. Access to previously performed imaging studies greatly facilitates interpretation of findings and detection of changes. Comparison between studies should be easy and should be routine in the review process. To facilitate this comparison, digital storage of clips and usage of standard scanning protocols is highly recommended. As these patients have frequently undergone multiple surgeries, acoustic windows may be challenging, especially with increasing age.

**Goals of imaging**

1. **Assessment of the Fontan pathway**
   - a. Assess for presence and size of fenestration and transfenestration gradient
   - b. Assess for presence and size of thrombus in Fontan pathway
   - c. Assess for obstruction of Fontan pathway to pulmonary artery.

2. **Assessment of pulmonary venous chamber**
   - a. Assess for adequate atrial communication
   - b. Assess for pulmonary venous obstruction.

3. **Assessment of atrioventricular valve function**
   - a. Severity of atrioventricular valve regurgitation, if present.

4. **Assessment of single ventricular function**
   - a. Systolic and diastolic performance
   - b. Hypertrophy and/or dilatation.

5. **Assessment of outflow tracts, semilunar valve(s), ascending aorta, and aortic arch**
   - a. Determine semilunar valve competency
   - b. Assess for residual or recurrent arch obstruction.

6. **Assessment of branch pulmonary arteries**
   - a. Determine narrowing, stenosis, competitive flow.

**Assessment of Fontan connections, pulmonary arteries, and pulmonary veins**

Typically it is recommended to initiate the scan with subxiphoid imaging. Coronal or subxiphoid sagittal views in particular allow for visualization of the inferior vena cava and its connection to the Fontan baffle. Color flow imaging and pulsed-wave (PW) Doppler tracings in this view are used to assess for obstruction (Figure 28.6; Videos 28.1 and 28.2). The inferior vena cava and hepatic veins are typically dilated and spontaneous contrast is often noted. Flow velocities should be low (generally <20–30 cm/s). For PW Doppler assessment, the sweep speed should be reduced in order to record flow throughout the respiratory cycle. Normally a low-velocity continuous systolic and diastolic flow pattern is detected (Figure 28.7). Usually, the flow increases with inspiration and decreases during expiration. The presence of respiratory variation suggests that intrathoracic pressure changes are well transmitted in the Fontan conduit, a sign that the Fontan connections are unobstructed. Absence of respiratory variation or the presence of reverse flow in the cardiac cycle is abnormal. Retrograde flow during systole may be seen in the setting of atrioventricular valve regurgitation or due to the presence of significant competitive collateral flow. Flow reversal during diastole may be present in the failing Fontan or in the presence of atrial arrhythmia (increased pressure in the Fontan circuit). Lack of respiratory variation should raise suspicion of conduit obstruction. In the intra-atrial tunnel or an extracardiac conduit, the connection between the
inferior vena cava may be visualized well using a subxiphoid long- and short-axis sweep. Subxiphoid, apical and parasternal views should be combined to assess the Fontan baffle. This assessment includes looking for tunnel dilation, the presence of clots and residual fenestration or baffle leaks (Video 28.3). If a fenestration is present, a PW Doppler tracing of the fenestration flow should be acquired (Figure 28.8). An estimate of the pressure gradient between the systemic and pulmonary venous chambers can be determined by obtaining the mean gradient across the fenestration (transpulmonary gradient). If the fenestration has been closed with a device, the position of the device should be evaluated as well as the presence of a residual shunt.

The superior connection between the tunnel/conduit and the pulmonary artery is often more difficult to image. The apical and parasternal long-axis views but particularly the suprasternal view are helpful in visualizing this region (Figure 28.9; Video 28.4). In the atroventricular Fontan, the connection is variable depending on the anastomosis. Typically, the atrioventricular connection results in progressive, marked atrial dilatation with low flow velocities, spontaneous contrast and potential for thrombus formation. Other types of connections involving valved conduits should be carefully inspected for valve dysfunction (especially stenosis) (Figure 28.10; Video 28.5) and thrombus. Color flow using low Nyquist settings should be used to visualize the surgical anastomosis sites of the Fontan connections. In the case of obstruction, the flow pattern in the baffle becomes continuous throughout the respiratory cycle as the gradient can persist during expiration. If TTE is insufficient, transesophageal echocardiography (TEE) may provide better imaging of the conduit, tunnel or atrium. Studies have demonstrated that thrombus that is present on TEE may not be visualized on TTE.

In patients with a lateral tunnel or extracardiac type Fontan connection, it is also important to evaluate the connection between the superior vena cava and the right pulmonary artery (Glenn anastomosis; Figure 28.11; Video 28.6). This can only be visualized from the suprasternal frontal view. In this view, the flow into the branch pulmonary arteries may be imaged although in older patients adequate images are challenging. The flow pattern in the superior vena cava resembles those in the inferior vena cava with increased flow with inspiration and decreased flow with expiration. If flow acceleration is noted at the anastomosis site, a PW Doppler tracing and mean gradient can be calculated. Thrombus may be present in the superior vena cava.
cava or the innominate vein, especially if a central line is present. In case of bilateral superior vena cavae, both anastomotic connections should be visualized. Generally, the left superior vena cava is smaller than the right, and obstruction of the left-sided anastomosis is more common. Also worth noting is that the pulmonary artery segment between both anastomoses sites may be small and become hypoplastic over time because it receives little flow.

The branch pulmonary arteries are also best visualized using suprasternal views (Figure 28.12; Videos 28.7 and 28.8). The proximal right pulmonary artery is visualized from the suprasternal frontal view. When moving the transducer into a sagittal (arch) view, the left pulmonary artery may be seen going towards the left hilum. Pulsed Doppler tracings should be obtained in both pulmonary arteries. If stenosis is present, a continuous flow pattern with an elevated mean gradient will be seen. If a pulmonary artery stent is present, the metal often causes shadowing and reverberations which can make it difficult to demonstrate flow. Color flow seen distal to the stent is reassuring. Flow reversal in the branch pulmonary arteries suggests increased pulmonary vascular resistance or the presence of significant aortopulmonary collaterals.

The pulmonary veins should also be interrogated after the Fontan procedure. An extracardiac conduit or significant atrial dilatation may cause compression of the pulmonary veins (Figure 28.13; Video 28.9). Pulmonary venous obstruction in a Fontan circuit can be difficult to detect by echocardiography and may require additional imaging by cardiac CT, magnetic resonance imaging, or cardiac catheterization.

**Assessment of atrioventricular valve function**

Careful evaluation of the atrioventricular valves should be performed in every Fontan patient. Significant atrioventricular valve regurgitation is associated with morbidity and mortality in this patient population.

Atrioventricular valve regurgitation is a common problem after the Fontan operation particularly in the presence of a common atrioventricular valve or a dominant tricuspid valve. Echocardiographic assessment should include evaluation of the
severity of atrioventricular valve regurgitation as well as identification of the mechanism (Figure 28.14; Videos 28.10 and 28.11). Evaluation of the severity of regurgitation is largely based on qualitative assessment using color flow. Different imaging planes should be used for assessing the size of the regurgitant jet in three dimensions. The jet width at the level of the leaflets generally is a good indicator of the severity of the regurgitation. Jet width assessment becomes more challenging when multiple jets are present. A combination of jet width, jet area and jet length into the atrium generally allows for subjective assessment as mild, moderate or severe. If more than mild, identifying the mechanism of regurgitation becomes important. A common mechanism for regurgitation is valve dysplasia with resultant valve leaflet irregularities at the zone of apposition and leaflet thickening [27]. Describing the mechanisms of regurgitation requires scanning the valve in multiple planes. Recent

Figure 28.10 Stenosis on a conduit between the right atrium and a hypoplastic RV in a patient with tricuspid atresia who underwent a valved conduit insertion between the right atrium (RA) and the hypoplastic right ventricle. This is a right parasternal view. The conduit is located just below the sternum and can be very difficult to image. The Doppler tracing allows to quantify the degree of stenosis (mean gradient 5–6 mmHg). Also notice that there is regurgitation during systole. Source: Courtesy of Dr. F. Meijboom.

Figure 28.11 Anastomosis between the superior vena cava (SVC) and the right pulmonary artery (RPA). Suprasternal notch view demonstrating the connection between the SVC and the RPA. The connection is widely patent with laminar continuous flow increasing with inspiration (Insp).
studies have suggested that 3D echocardiography may provide additional information on the atrioventricular valve in FSV [28]. Further improvements in spatial and temporal resolution of this technique will allow better defining of leaflet abnormalities and a more detailed description of the subvalvar apparatus.

After valve repair, careful follow-up of the surgical result is required. In case of prosthetic valve insertion, evaluation of prosthetic valve function is essential. Perivalvular leaks can be present and should be distinguished from regurgitation related to deficient prosthetic valve function. The low flow profile through the prosthetic valve makes it theoretically more prone to valve thrombosis. After valve repair, close monitoring of single ventricular function is required. The long-term outcome of patients undergoing atrioventricular valve repair in the context of single ventricle palliation is worse compared to case-matched patients not requiring valve surgery [29].

Atrioventricular valve stenosis is rare but can be present after atrioventricular valve repair or after prosthetic valve replacement. Inflow through the valve(s) should be assessed using color flow and PW Doppler from the apical views. If a gradient is present, CW Doppler should be obtained as aliasing may occur. The mean gradient should be calculated throughout the respiratory cycle. For prosthetic valves, a mean gradient is common and known for the specific valve types. If significant regurgitation is present, the mean inflow gradient may be overestimated due to increased flow.

Assessment of outflow tracts, ascending aorta, and aortic arch

In FSV, the outflow tract from the dominant ventricle connects the ventricle to the aorta. Outflow tract obstruction, if present, causes chronic pressure loading with secondary concentric hypertrophy, which is detrimental for the long-term preservation of ventricular function. It can further limit the output from the FSV. The mechanism causing outflow tract obstruction most commonly is subvalvar obstruction. The most typical example is double-inlet left ventricle with transposition of the great arteries where the VSD to the RV outlet chamber may become restrictive and thus, cause subaortic obstruction. In cases where a patient is at risk for this obstruction, a Damus–Kaye–Stansel (DKS) operation connecting both outflow tracts to the aorta is performed. The outflow tract needs to be imaged in all Fontan patients, especially in those who did not undergo a DKS procedure to assure unobstructed flow. Different acoustic windows (subxiphoid, apical views, and parasternal long-axis views) can be used to image the outflow tracts. Color Doppler, PW and CW Doppler traces should be obtained as well aligned with the outflow tract as possible. The peak and
mean gradients should be measured. If a gradient is detected, the mechanism causing the gradient should be defined. This can include subaortic obstruction due to a restrictive VSD, a membranous/fibrous ring, or valvar obstruction. In every patient the DKS connection between the ascending aorta and previous pulmonary trunk should be imaged. Obstruction at the connection between the aorta and the pulmonary can compromise coronary artery blood flow and result in ischemia and ventricular dysfunction. Semilunar valve function should also be evaluated. When the pulmonary valve is assigned to the aortic position (neo-aorta) regurgitation is prevalent. A DKS connection can distort one or both semilunar valves and cause regurgitation. Fortunately, important semilunar valve regurgitation is unusual in FSV. There have been reported cases of valve replacement.

In patients who underwent aortic arch reconstruction or coarctation surgery, assessment for residual arch obstruction is essential. Residual arch obstruction and coarctation of the aorta causes arterial hypertension and a significant increase in afterload to the FSV. Even a mild gradient can cause significant ventricular dysfunction. The suprasternal window is best to view the arch. Color Doppler and CW Doppler can be used to assess the peak gradient across the arch and the descending aorta. In a reconstructed arch, the proximal arch gradient should be incorporated into the Bernoulli equation to achieve the most accurate arch gradient. In adults the arch can be difficult to image. A good alternative for ruling out residual arch obstruction is obtaining a PW Doppler tracing of the abdominal aorta from the subxiphoid views. The absence of significant diastolic anterograde flow in the abdominal aorta rules out significant residual arch obstruction.

**Assessment of single ventricular function**

One of the most challenging parts of the echocardiogram in patients after the Fontan procedure is the assessment of ventricular function. Application of standard methods of assessment of the normal left ventricle to a FSV with variable ventricular morphology is a significant challenge. The FSV operates with reduced preload reserve and the pulmonary circulation is an important regulator of cardiac output. Assessment of ventricular function requires assessment of both systolic and diastolic function.

Systolic performance: The most widespread technique used to assess systolic ventricular function in patients after the Fontan operation, is *subjective assessment* or “eyeball technique.” Most
Part V Miscellaneous Cardiovascular Lesions

This index of combined systolic and diastolic time can be calculated. This preload-sensitive index is good amongst the core lab echocardiographers and ejection fraction [31]. The reproducibility of this method was found to be good amongst the core lab echocardiographers as well as the accuracy (agreement between echocardiographic assessment and cardiac MRI quantification) of the assessment. The reproducibility of qualitative assessment was found to be moderate for LV morphology but weak for RV morphology. Also the agreement between qualitative assessment by echocardiography and quantitative assessment by cardiac MRI was weak. Both studies found that image quality and reader’s experience influenced the results. In the mild to moderate range of dysfunction the disagreement is more pronounced. This is not surprising as most readers will be able to diagnose patients at the both extremes of the spectrum with either severely reduced function or a completely normal EF.

Quantitative approaches have been proposed and evaluated. A multicenter cohort study of the Fontan population used a modified biplane Simpson’s method to quantify FSV volumes and ejection fraction [31]. The reproducibility of this method was found to be good amongst the core lab echocardiographers evaluating the studies, with better agreement for the LV than the RV measurements. The agreement between echocardiographic and MRI quantitative assessments was weak, however, with systematic underestimation of FSV volumes by echocardiography. This is probably related to the different geometry of FSV when applying the Simpson’s formula. Given the relatively good reproducibility within the same laboratory, the biplane Simpson’s technique may provide a reasonably good method for quantitative serial follow-up in the same patient. As most EF calculations are based on the geometric assumption of an ellipsoid LV, there will be relative inaccuracy in cases of more complex geometry. A reasonable alternative is the use of fractional area change from an apical 4-chamber view as an easy method for quantifying function (Figure 28.15). Lately 3D echocardiography has been proposed as a potential emerging technique for evaluating FSV volumes and ejection fraction. The method of discs shows good reproducibility and accuracy [32]. However, it has recently been replaced by semi-automated (for the RV) or automated border detection and volumetric analysis programs that have been validated in biventricular hearts. These methods have yet to be validated for FSV [10]. A major issue with 3D echocardiography is that it can be very difficult to acquire a full volume of the entire FSV at reasonable frame rates; moreover, the anterior wall segments can be extremely difficult to visualize due to their anterior position in the chest just behind the sternum.

Since volumetric data are difficult to obtain in FSV, alternative nongeometric methods have been proposed. Using blood pool Doppler signals, \(dP/dt\) can be calculated. This preload-sensitive method is based on a valve regurgitation jet (Figure 28.16) or by using a modified method based on measurement of the isovolumic contraction time (IVCT) and aortic diastolic blood pressure and assuming FSV end-diastolic pressure (5 mmHg) [33]. The regurgitation method requires the presence of at least a mild to moderate amount of regurgitation in order to be able to acquire a reliable jet, which is not possible in every patient. The method based on IVCT requires measuring time intervals on the inflow tracing and the outflow tracing, which requires that there is no change in heart rate between the two traces. An additional problem with this method is that the estimation of end-diastolic pressure may be inaccurate. The reproducibility of \(dP/dt\) measurements is high but the value of \(dP/dt\) correlates only weakly with MRI-based measurement of EF [33]. An alternative nongeometric Doppler-based method is the myocardial performance index (MPI) or Tei-index. This index of combined systolic and diastolic ventricular performance is calculated as follows:

\[
\text{MPI} = \frac{\text{IVRT} + \text{IVCT}}{\text{ET}}
\]

where IVRT = isovolumic relaxation time and ET = ejection time. The index can be measured based on blood pool data (inflow and outflow) or tissue Doppler tracings. Tissue Doppler (Figure 28.17) has the advantage that the measurement can be performed on the same cardiac cycle [33,34]. The
Figure 28.15 Quantification of single ventricular function. From an apical 4-chamber view fractional area change (FAC) can be measured by tracing the end-diastolic and end-systolic area and calculating the FAC. In this patient FAC was 50%. Using the Simpson’s method volumes can be estimated and ejection fraction can be calculated. Ejection fraction in this patient was 72%.

Reproducibility of this method has been moderately good. In Fontan patients, MPI has been reported to be prolonged but the degree of prolongation does not correlate with EF as measured by MRI. MPI is preload and afterload sensitive and this may affect the measure [35]. A potentially more attractive concept is the ratio of systolic to diastolic duration [36]. This measure requires an atrioventricular valve regurgitation jet and measuring the duration of the regurgitation jet as the systolic duration and the inflow duration as the diastolic duration. When ventricular dysfunction develops, systolic duration prolongs, shortening diastolic inflow duration. In severe dysfunction the heart spends most of the time in systole with only a very short time in diastole thus limiting diastolic filling. In normal children the S/D ratio varies between 0.4 and 1.6 and is largely determined by heart rate [37]. In patients with hypoplastic left heart syndrome the S/D ratio has been shown to be elevated and higher in patients with RV dysfunction compared to patients with normal RV function [36]. When corrected for heart rate, however, the values were shown to be in the normal range [34]. All these recent data illustrate that the use of nongeometric Doppler-derived timing parameters requires further investigation before they can be recommended in routine clinical practice.

Alternatively tissue Doppler velocities can be used. Tissue Doppler traces are relatively easy to obtain and are generally

Figure 28.16 $\frac{dP}{dt}$ as calculated from the atrioventricular valve regurgitation. In this patient with single ventricular dysfunction $\frac{dP}{dt}$ was significantly reduced.
Figure 28.17 Measuring time intervals on tissue Doppler traces obtained in the AV-valve annulus. (i) represents the isovolumic contraction period with the IV A peak, (ii) is the ejection time, (iii) is the isovolumic relaxation time, and (iv) is the diastolic time or filling time. The myocardial performance index can be calculated by (ICT + IRT)/ET. (Bellsham-Revell EHJ-CVI 2012)

Diastolic performance

The development of diastolic dysfunction is probably one of the most important problems after Fontan palliation. Diastolic dysfunction has been shown to be highly prevalent with 72% of the patients having diastolic abnormalities in a large cohort study [7]. The study proposed a grading system for diastolic dysfunction with normal diastolic function defined as E/A between 1 and 2, DT ≥140 msec, and E/E′ ≤10. Impaired relaxation was defined on E/A ratio <1 only. Pseudonormalization was identified when E/A was between 1 and 2 but DT <140 msec or E/E′ >10 or (FP <55 cm/sec). Restrictive physiology was based on E/A >2. Of note, this grading system is extrapolated from adult data and has never been validated against invasive pressure data in Fontan patients. Moreover, there are limitations of these modalities related to nonsinus or paced rhythm, heart rate and atrioventricular valve function.

In the aging Fontan population, progressive decrease in ventricular compliance and increased end-diastolic pressure is often observed. This is difficult to detect using noninvasive methods. In the adult population with a biventricular circulation E/E′ has measurement of cardiac contractility. It is, however, highly heart rate dependent and difficult to measure.

Most recently, the introduction of speckle tracking technology has allowed the calculation of myocardial deformation or strain imaging in Fontan patients (Figure 28.18) [38]. This method was recently validated against myocardial strain measurements as obtained by MRI-based myocardial tagging [39]. Similar to other methods of function, strain measurements are dependent on loading conditions. Increased preload will increase myocardial deformation; increased afterload will result in decreased myocardial deformation. Presently, there is limited information on the clinical use of strain imaging in the Fontan population and how it can be used in clinical practice. The main application for this technology may be its use in detecting intraventricular dyssynchrony in patients who develop ventricular dysfunction in order to help to identify patients who might benefit from resynchronization therapy (Video 28.12) [40,41].

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been proposed as a useful parameter for identifying patients with increased filling pressures. In patients with FSV, the use of the E′/E ratio is problematic because E velocities are influenced by atrioventricular valve size and E′ velocities are often preserved. Though diastolic dysfunction is a primary cause of Fontan failure, thus far current echocardiography techniques are not capable of identifying these patients based on any criteria. Identification of FSV patients with reduced compliance is one of the remaining challenges in congenital cardiology. Hopefully methods will soon be developed which will provide information on tissue characteristics, such as MRI-based T1-mapping.

**Conclusion**

Echocardiographic imaging in patients after the Fontan operation remains a significant challenge. It requires a full understanding of the underlying complex morphology, the surgical techniques and the unique physiology of the cardiac circuit. While echocardiography is the standard routine follow-up technique, patients with Fontan failure or suspected complications will likely require additional imaging by MRI, CT, or angiography.

**Videos**

To access the videos for this chapter, please go to www.lai-echo .com.

**Video 28.1** The connection between the inferior vena cava (IVC) and the extracardiac conduit (ECC). The connection between the IVC and the conduit is nicely demonstrated from a subxiphoid view by two-dimensional imaging.

**Video 28.2** The connection between the inferior vena cava (IVC) and the extracardiac conduit (ECC). The color flow image shows increased flow in the conduit with inspiration related to negative intrathoracic pressures during inspiration with increased blood flow in the Fontan circuit.

**Video 28.3** Fenestrated extracardiac Fontan. In this subxiphoid view, color Doppler imaging demonstrates the presence of a continuous low-velocity jet through the fenestration.

**Video 28.4** Connection between the extracardiac Fontan and the pulmonary artery. In this suprasternal image slightly tilted to the right and anteriorly, the connection between the extracardiac conduit (ECC) and the right pulmonary artery are nicely demonstrated. The red flow indicates the flow in the direction of the pulmonary artery during inspiration.

**Video 28.5** Stenosis on a conduit between the right atrium and the hypoplastic RV. This patient with tricuspid atresia underwent a valved conduit insertion between the significantly dilated right atrium (RA) and the hypoplastic right ventricle which became significantly stenotic and regurgitant. Source: Courtesy of Dr. F. Meijboom.

**Video 28.6** Suprasternal notch view demonstrating the connection between the superior vena cava (SVC) and right pulmonary artery (RPA). The connection is widely patent with laminar continuous flow increasing with inspiration.

**Video 28.7** Imaging the left pulmonary artery (LPA). In a leftward tilted suprasternal view the LPA can be imaged below the aortic arch. The laminar continuous antegrade low velocity flow can be demonstrated in the proximal LPA.

**Video 28.8** Imaging the right pulmonary artery (RPA) from a rightward tilted suprasternal notch view. The continuous low-velocity flow in the proximal RPA can be seen.

**Video 28.9** Pulmonary venous obstruction caused by atrial dilatation. Atriopulmonary connection with important dilation of the right atrium (RA), causing compression on the right upper pulmonary vein with turbulent flow in the pulmonary vein and a mean gradient of 4–5 mmHg. Source: Courtesy of Dr. F. Meijboom.

**Video 28.10** Tricuspid regurgitation in a patient with hypoplastic left heart syndrome after the Fontan operation. This clip shows a color Doppler image obtained from the apical 4-chamber view. There is a wide jet of tricuspid regurgitation coming from the zone of apposition between the anterior and septal leaflets.

**Video 28.11** Tricuspid regurgitation in a patient with hypoplastic left heart syndrome after the Fontan operation. This clip shows an apical 2-chamber view of the RV with an additional jet coming from between the anterior and inferior leaflets of the tricuspid valve. Overall the regurgitation is severe in this patient with significant right atrial dilatation.

**Video 28.12** Dyssynchrony in a patient with a single ventricle. Speckle tracking was used to quantify longitudinal deformation. From the images and curves it can be observed that the left lateral wall is being stretched at the time the right lateral wall contracts.

**Video 28.13** Dyssynchrony in a patient with a single ventricle. This causes significant dysfunction of the single ventricle with the patient developing symptoms of heart failure. Resynchronization therapy resulted in significant improvement in global ventricular function and reverse remodeling.

**References**


