17

Univentrcular Heart & Fontan Circulation

Any congenital cardiac anomaly in which one ventricle is hypoplastic or absent is considered as a single ventricle or univentricular heart. Fontan operation remains a palliative procedure for single ventricle patients with a low mortality and morbidity. Outcomes are more favorable than for those patients who have had systemic to pulmonary artery shunts alone. Associated defects such as bulboventricular foramen obstruction and atrioventricular valve regurgitation should be treated concomitantly at the time of Fontan operation.

17.1 Morphology & Pathophysiology

Single ventricle correction is required for significant hypoplasia of either of the atrioventricular valves or hypoplasia of the apical portion of either ventricle.

Biventricular repair is feasible for hypoplasia of either of a semilunar valve or a ventricular outflow tract.

Initial operation for the single ventricular correction is focused towards:

1. Relief of systemic outflow tract obstruction to provide adequate perfusion of the body.
2. Provision of adequate pulmonary blood flow to provide adequate oxygenation, but the flow should be limited to prevent development of elevated pulmonary vascular resistance.

The final operative correction is creation of Fontan circulation in which systemic venous return is directed to the pulmonary arteries without a pumping chamber. This requires low pulmonary vascular resistance so that blood will flow to PA (pulmonary artery) passively, at an acceptable venous pressure.

17.2 Operative Procedures

Fontan circulation is usually accomplished in two stages.

17.2.1 Stage I

A cavopulmonary shunt (bidirectional cavopulmonary shunt or hemi-Fontan) is constructed, at 3 to 6 months of age. (see Figure 17.1). The shunt directs the superior vena caval blood flow to the confluent pulmonary arteries.

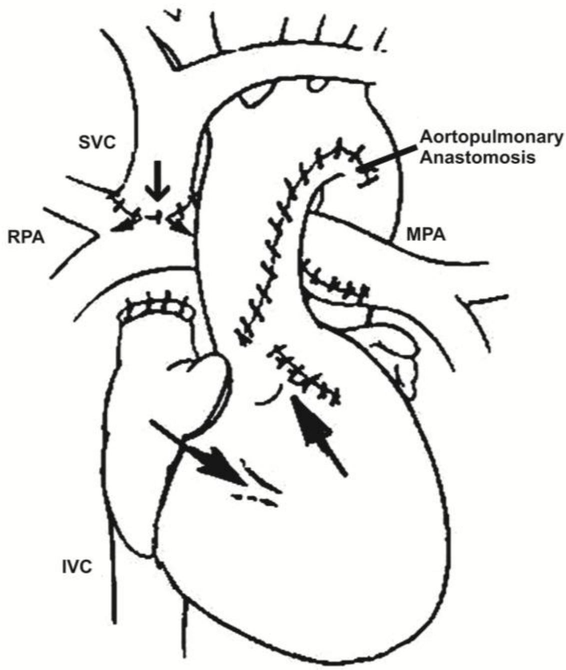

Figure 17.1 *Bidirectional superior cavopulmonary anastomosis (bidirectional Glenn). The superior vena cava is divided and the divided upper end is anastomosed to side of the right branch pulmonary artery. The divided lower end of SVC is closed. One third of systemic venous blood flows into pulmonary circulation. Two thirds of systemic venous blood from the inferior vena cava flows into systemic circulation through the aorta. The proximal end of the main pulmonary artery is closed. SVC=superior vena cava, RPA=right branch pulmonary artery, IVC=inferior vena cava, MPA=main pulmonary artery.*

Only a third of systemic venous return (through SVC) traverses through pulmonary vascular bed. The shunt, therefore, decreases the volume load of the ventricle, prevents the development of pulmonary hypertension, and elevated pulmonary vascular resistance.

17.2.2 Stage II

Fontan completion is undertaken at approximately at 2 years of age. It is done by redirecting the inferior vena caval blood flow to the PA with an intracardiac baffle (lateral tunnel) (see Figure 17.2) or by an extracardiac conduit.

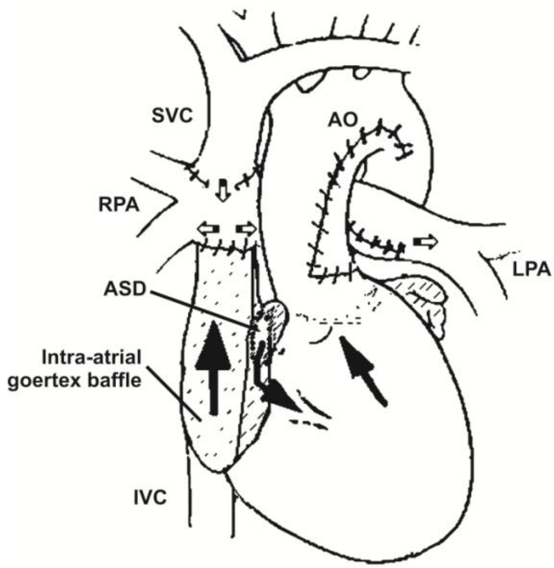

Figure 17.2 *Lateral tunnel Fontan. The goertex baffle is sutured in the posterior and lateral right atrium to tunnel the blood from the inferior vena cava to the right branch pulmonary artery. The Fontan circulation is thus completed on a previously performed bidirectional Glenn anastomosis. The blood from the left atrium flows freely through an enlarged atrial septal defect into the systemic ventricle and through aorta to systemic circulation. Arrows indicate the direction of blood flow from SVC and IVC to pulmonary circulation. AO=ascending aorta, ASD=atrial septal defect, IVC=inferior vena cava LPA=left branch pulmonary artery, RPA=right branch pulmonary artery, SVC=superior vena cava.*

17.3 Postoperative Management

The postoperative course following a bidirectional cavopulmonary shunt or hemi-Fontan is usually uncomplicated. The hospital stay averages five to seven days. The postoperative course following a completion Fontan can be more complex. The hospital stay averages ten days to three weeks.

17.3.1 Hemodynamic Management

Monitor hemodynamic changes very carefully, by noting the principles of management outlined below:

Patients with univentricular physiology do not have as much hemodynamic reserve as patients with biventricular physiology, especially, during the first few days following surgery.

If there is restriction of free flow of blood from systemic venous return through pulmonary vascular capillary bed into the left atrium:

i) It results in cyanosis following a cavopulmonary shunt.

ii) It results in reduction of cardiac output after the Fontan, and any small increases in pulmonary vascular resistance can result in decreased cardiac output.

17.3.2 Transpulmonary Gradient

It is very important to monitor transpulmonary gradient during the postoperative period. The transpulmonary gradient determines the prograde blood flow across the pulmonary capillary bed, and is estimated by subtracting the left atrial pressure from central venous pressure (i.e., CVP-LAP).

Central venous pressure above fifteen to eighteen mm of Hg or a transpulmonary gradient above ten mm of Hg indicates difficulty with passive flow of blood across the pulmonary capillary bed.

Use the following formulae:

Transpulmonary gradient or Delta (d) / Flow = R (resistance), higher gradients (d) are required to maintain pulmonary flow in the presence of an elevated PVR.

(PBF= d/R, If R is elevated, d should be increased to maintain flow).

17.3.3 Restricted Transpulmonary Blood Flow in the Postoperative Period

The following are the other causes of restricted transpulmonary blood flow that may occur during the postoperative period:

1. High peak airway pressures.
2. Increased I: E ratio.
3. Excessive PEEP in a patient on a ventilator.
4. Rises in intrathoracic pressure caused by events such as in hemothorax or pleural effusion.
5. Poor diastolic compliance of the systemic ventricle. Probable agents that are useful to improve diastolic compliance are:

Nitric oxide, beta blockers, and Ace-inhibitors.

(Note: Endogenous NO (nitric oxide) released from the coronary endothelium may hasten LV (left ventricular) relaxation and may increase LV distensibility. Variations in the release of NO in accordance with prevailing cardiac workload, signaled via preload, coronary flow, mechanical forces, and heart rate may provide an acute auto-hyphon regulatory feedback that optimizes diastolic LV performance. Deficient production of NO in conditions such as pressure-overloaded heart (increased LV hypertrophy) and heart failure contributes to diastolic dysfunction. NO may be beneficial in these conditions, especially, for patients with heart failure and reduced inotropic reserve, who are dependent on the LV Frank - Starling response to maintain cardiac output).

All the five above, interfere with transpulmonary blood flow from systemic veins to the left atrium, leading to high transpulmonary gradients and should be corrected.

It is very important to monitor carefully, CVP and look for any physical evidence of systemic venous congestion.

If CVP is elevated with restricted flow through the pulmonary vascular capillary bed, the following physical findings will be apparent: Hepatomegaly, ascites or anasarca, or edema of the head and neck (in a cavopulmonary shunt).

17.3.4 Arterial Oxygen Saturation

Systemic arterial blood oxygen saturation (SaO_2) after a bi-directional cavopulmonary shunt should be 75% to 80 %.

Systemic arterial blood oxygen saturation (SaO_2) following a Fontan completion may be lower than normal (88% to 95%) since the coronary sinus is not included in the Fontan circuit.This coronary sinus blood return is allowed to mix with pulmonary venous return.

17.3.5 Invasive Monitors

Arterial, central venous, and LA (left atrial) catheters.

17.3.6 Vasoactive Drug Infusions

Dopamine or dobutamine, epinephrine, milrinone, and nitroprusside (see Section I Chapters 4 & 16).

17.3.7 Cardiac Rhythm Abnormalities

Sinus node dysfunction:

Extensive atrial suture lines and alteration of normal intra-atrial pressures can lead to a loss of sinus rhythm and cause sinus node dysfunction. It is common following a Fontan completion.

Sinus node dysfunction commonly presents as sinus arrhythmia, severe tachycardia, and loss of normal atrioventricular synchrony.

Temporary overdrive pacing:

It is needed as hemodynamic compromise is common in the Fontan patient subsequent to a loss of normal atrioventricular synchrony or in severe tachycardia.

Permanent pacemaker: It is occasionally required.

17.3.8 Postoperative Beeding

Excessive bleeding is very rare.

17.3.9 Anticoagulation

Slow venous blood flow through the long segments of prosthetic material make Fontana patients susceptible to thrombus formation in the early postoperative period.

Coumadin is started in Fontan patients once intracardiac lines have been removed.

Following the cavopulmonary shunt, anticoagulation is less rigorous and might include only aspirin or dipyridamole.

17.3.10 Pleural Effusion

Persistent pleural drainage following the bidirectional cavopulmonary shunt is rare. Persistent pleural effusion is common following the Fontan completion, so bilateral chest tubes may be routinely placed in both pleural spaces during Fontan completion.

Chemical Pleurodesis:

If pleural drainage continues more than 10 days or even a week, the chemical pleurodesis may be required to control protracted pleural drainage (see Section I, Chapter 12).

17.3.11 Discharge Medications

In uncomplicated Fontan patient or a patient with bidirectional cavopulmonary shunt, the discharge medications should include coumadin or antiplatelet agent, digoxin, lasix, and captopril.