

Single-ventricle (SV) heart defects are a unique and heterogeneous group of complex congenital heart defects characterized by stenosis and/or atresia of the semilunar and/or AV valves, hypoplasia of the ventricles, and/or a segmental cardiac arrangement not conducive to a biventricular repair. The result is a “functional” SV that provides parallel support to the pulmonary and systemic circulations. Examples, to name a few, include hypoplastic left heart syndrome (HLHS), tricuspid atresia, double inlet left ventricle (DILV), and unbalanced atrioventricular septal defects (AVSD). Although initial presentation and management varies, most will require staged palliations to allow free flow of blood from the ventricle out to the body, protect the lungs from high pressure and extra blood flow, and create separate pathways for blood flow to the lungs and to the body.

Anatomical Considerations

There are several possible combinations of SV defects and clear anatomical descriptions are used to guide management. Initial and ongoing considerations in the evaluation of the anatomy of a patient with functional SV include:

- Ventricular morphology and function
- AV connections and function
- Ventriculoarterial connections
- Source of systemic and pulmonary blood flow
- Source of systemic and pulmonary venous return
- Atrial communication

Anatomically, most defects can be classified as follows:

- **Single-inlet AV defects** are characterized by atresia of one AV valve resulting in a functional SV with a single AV valve. Specific examples include HLHS, double-outlet RV (DORV) with mitral atresia and tricuspid atresia (See Chapter 27 for HLHS and Chapter 20 for tricuspid atresia).
- **Common-inlet SV defects** are characterized by a common AV valve connected to the ventricles in an unbalanced manner. Although there are commonly 2 complete ventricles, distribution of the common AV valve may be such that it is not possible to achieve a biventricular repair. The appropriate SV pathway will be determined by associated defects and blood flow obstruction. These defects often have associated systemic and pulmonary venous return abnormalities. Those with obstructed anomalous pulmonary venous return and unbalanced AVSD are a high-risk group and many were previously considered inoperable.
- **Double-inlet defects** are characterized by a SV that receives both AV valves. The ventricle may be of left (common), right, or indeterminate morphology (rare). Typically, there is a rudimentary or incomplete ventricle that is connected to the dominant chamber by a VSD (or bulboventricular foramen). A common example is

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DILV. There are often associated lesions such as transposition of the great vessels, DORV, aortic coarctation, and varying degrees of subvalvar or valvar obstruction.

- **Other functional SV defects** may include situations in which a biventricular arrangement is not desirable such as patients with DORV and a remote VSD.

Pathophysiology and Clinical Presentation

The physiology associated with SV defects will depend on the relationship of pulmonary and systemic blood flow and the relative resistance of each pathway. Many infants with SV defects will have ductal-dependent systemic (SBF) or pulmonary blood flow (PBF) and require PGE infusion prior to initial palliation. They will often develop CHF symptoms secondary to excess PBF as the PVR falls. Infants with ductal-dependent SBF may also have signs of decreased systemic perfusion. Symptoms are worsened if there is a restrictive atrial-level shunt causing pulmonary venous congestion and limiting complete mixing of deoxygenated systemic and oxygenated pulmonary venous returns.

Infants with SV defects and ductal-dependent PBF are admitted to the NICU for preoperative evaluation and management while those with ductal-dependent SBF and/or heterotaxy with concern for pulmonary vein issues are admitted to the CICU. Preoperative feeding practices will vary based on type of SV defect and clinical status. However, most will remain NPO with TPN for nutrition. For those allowed to feed preoperatively, trophic PO feeds (20-40 mL/kg/day) with breast milk is preferred. The infant requires close monitoring of respiratory, GI, and circulatory status as the risk for necrotizing enterocolitis is increased in the setting of unbalanced PBF and SBF.

Diagnosis

- **Fetal echocardiogram.** Prenatal diagnosis of SV defects is common during routine fetal anatomy scans typically done at 18-20 weeks. Most families receive extensive prenatal counseling from multiple pediatric specialties based on perceived postnatal needs. The cardiologist will also determine if the infant requires postnatal PGE infusion for ductal-dependent lesions or urgent postnatal intervention such as septostomy to relieve atrial-level restriction. In high-risk patients who may require immediate intervention, the cardiologist attends the delivery and coordinates transfer to the cardiac catheterization laboratory for atrial septostomy and stenting.
- **CXR.** Useful to show cardiac position, size, and pulmonary vasculature markings (overcirculated or underperfused).
- **ECG.** Useful to evaluate for rhythm and measure axis and intervals. Ventricular hypertrophy, sometimes with a strain pattern, is a common finding.
- **Postnatal TTE.** Used to confirm the anatomy in addition to obtaining information about ventricular and valve function, and ductal- and atrial-level shunts. In some cases, additional imaging such as CTA is used to confirm systemic and pulmonary venous return, arch anatomy, or coronary artery origins.
- **Cardiac catheterization.** Typically not necessary unless atrial septostomy is needed or there is concern for coronary artery abnormalities and/or fistulae.
- **Renal ultrasound.** Routinely obtained to assess for renal anomalies. Renal

consultation is obtained in newborns undergoing surgery with CPB in anticipation of the need for peritoneal dialysis.

- **Preoperative brain MRI.** Obtained to evaluate for ischemic lesions or brain matter abnormalities sometimes seen in infants with CHD.
- **Genetics testing.** Tailored to syndromic features and known associations. For example, HLHS has a known association with Turner syndrome, Jacobsen syndrome, and trisomies 13 and 18. However, many SV defects are not associated with an identified genetic abnormality or there is unknown significance. Chromosomal microarray (CMA) is common as it allows detection of a wide array of chromosome gains and losses. Whole-exome sequencing may be obtained if the CMA is negative and there is a high suspicion for a genetic syndrome. More information about genetics testing can be found in Chapter 50.

Indications / Timing for Intervention

Symptoms vary based on defects, but most require close monitoring for cyanosis, CHF, or cardiogenic shock. Symptoms may worsen as the neonate transitions from fetal circulation.

The current paradigm of SV palliation includes generally 3 stages: an initial palliation at the newborn period that varies depending on the degree of pulmonary and systemic blood flow, a second-stage palliation consisting of a superior cavopulmonary shunt (bidirectional Glenn) at 4-6 months of age, and a third-stage palliation with a completion total cavopulmonary anastomosis (Fontan) at 3-5 years of age.

First-Stage Palliation

Most SV defects will fall into 1 of 4 categories: increased PBF, decreased PBF, decreased SBF, and balanced circulation. Patients within the first 3 categories will typically require a first-stage procedure with the goal of providing adequate systemic and pulmonary blood flow, and at the same time protect/prepare the lungs for the second stage of palliation. Those with a balanced circulation may be observed over time and later undergo a bidirectional Glenn as their initial palliation.

Increased PBF

Patients with increased PBF may require pulmonary artery banding (PAB) to limit flow and pressure to the lungs. Medically managing a SV patient with significant overcirculation is not desirable as the excess flow to the lungs may alter pulmonary vascular reactivity and make the patient a less suitable candidate for the second stage of palliation.

The PAB is typically placed using a median sternotomy. The upper pericardium is opened and the main PA (MPA) encircled. A small catheter is inserted into the distal MPA to measure pressure and help with adjusting of the PAB. Classically, the opening PA pressure will be equal to systemic pressure. A piece of umbilical tape is trimmed and marked at a distance corresponding to Trussler's rule (20 mm plus the weight in kg of the patient). The PAB is placed around the MPA with care not to impinge on the right PA or the pulmonary valve, and secured with a fine horizontal-mattress suture passed through both sides of the PAB. The PAB is then adjusted by placing additional

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sutures or clips while monitoring PA pressure, oxygen saturation, and PaO_2 . The TEE is also used to monitor for PI, velocity across the PAB, and alterations in ventricular function. In general, the PAB is adjusted to achieve approximately 1/3 systemic PA pressure with oxygen saturation (SaO_2) levels in the 80s and PaO_2 in the 40s on an FiO_2 of 40–50%. When the PAB is applied to patients with a biventricular circulation (e.g., patients with multiple VSDs), the PAB is left slightly looser, with PA pressures approximately 35–50% systemic.

Postoperative management after PAB placement follows similar principles than postoperative Norwood palliation care (see Chapter 27) due to the need for precise partitioning of cardiac output but without the insult of CPB or, in some circumstances, reliance on an inferior systemic RV. Standardized monitoring for assessment of circulatory well-being, vasoactive support and ventilator management are similar to that used for Norwood palliation, though physiologic targets may vary depending on the underlying defect. For example, after Norwood palliation, the expectation is that SaO_2 that exceeds 90% places the patient at high risk for systemic hypoperfusion and persistent respiratory insufficiency. Whereas, a PAB placed to limit PBF in common-inlet SV defects might have persistent SaO_2 in the 90s with preserved lung function and good systemic output. Thus, therapeutic targets should include resolution of symptoms associated with CHF and pulmonary edema rather than targeting a specific SaO_2 . Though intraoperative PAB adjustment targets the aforementioned SaO_2 and PaO_2 , oxygenation often improves during the postoperative period due to resolution of the preoperative pulmonary edema-induced pulmonary venous desaturation. Oximetry targets ideally include narrowing of SaO_2 – cerebral rSO₂ (by NIRS) difference to <40% and SaO_2 – somatic rSO₂ difference to <20% (see Chapter 69). Widening of these oximetric gradients beyond the early postoperative period is an indication for systemic afterload reduction to optimize systemic oxygen delivery, and to further protect the lungs from excessive PBF and the subsequent development of irreversible pulmonary vascular disease.

Decreased PBF

Patients with decreased PBF will require an additional source of PBF in the form of a modified Blalock-Taussig-Thomas shunt (mBTTS) or a PDA stent. For details of these procedures and postoperative management, see Chapter 38.

Decreased SBF

Patients with impaired systemic outflow will usually require a Norwood-type operation that includes:

- **Creation of an unimpeded systemic outflow.** This usually entails creation of a Damus-Kaye-Stansel (DKS) anastomosis between the aortic and pulmonary trunks and an aortic arch reconstruction. For patients with a very small ascending aorta (such as those with HLHS and aortic atresia), the ascending aorta and aortic arch are opened longitudinally, an anastomosis is created between the transected PA and the side of the ascending aorta, and the entire ascending aorta/aortic arch is reconstructed with a homograft patch after removal of all ductal tissue (see Chapter 27). Patients with a sizable ascending aorta may undergo a modified Norwood

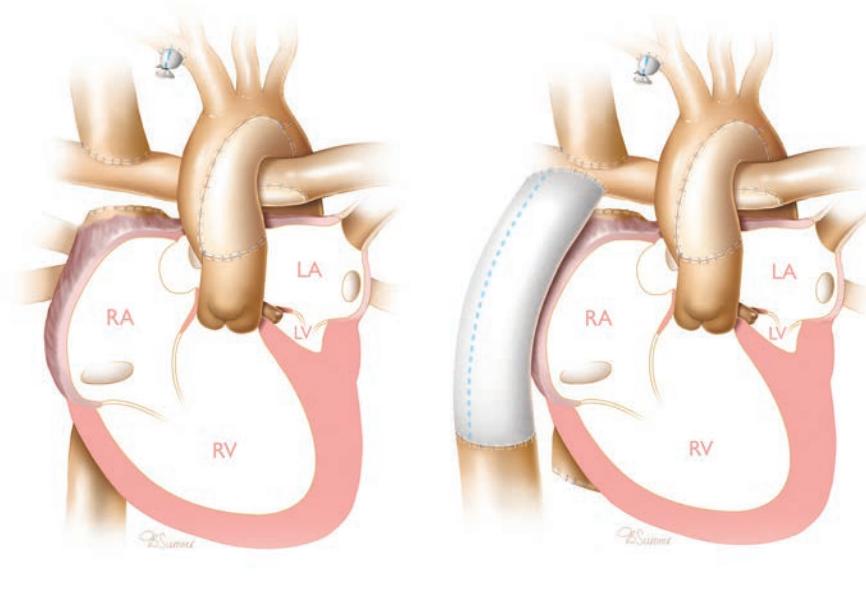


Figure 39-1. Bidirectional Glenn anastomosis in a patient after a Norwood procedure.

Figure 39-2. Extracardiac non-fenestrated Fontan procedure on a patient with a previous Norwood procedure and bidirectional Glenn anastomosis.

procedure (Lamberti modification). For these patients, both the ascending aorta and the PA are transected and anastomosed in a side-to-side fashion, therefore creating a double barrel. The aortic arch and ascending aorta are reconstructed using an aortic arch advancement technique (see Chapter 25) and a small patch. An end-to-end anastomosis is then created between the reconstructed ascending aorta and the double barrel. The advantage of this approach is that it decreases the time of antegrade cerebral perfusion since the DKS anastomosis and sometimes the ascending-aorta to double-barrel anastomosis is made under full-flow CPB.

- **Placement of a shunt or conduit to provide PBF.** This is usually achieved with either a mBTTS or an RV-PA (Sano) conduit. For patients with HLHS, an RV-PA conduit is favored unless there are prominent coronary arteries on the free wall of the RV. The conduit may be brought towards the left (traditional Sano) or towards the right (Brawn modification) of the aorta. Patients with other SV anatomy, such as those with DILV, usually undergo placement of a mBTTS.
- **Atrial septectomy** to provide unrestricted atrial-level shunting.

For details on the postoperative management of patients after a Norwood procedure, see Chapter 27.

Patients with DILV or tricuspid atresia with transposition of the great arteries may have a restrictive VSD leading to impaired SBF and aortic arch hypoplasia. Even though

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most centers will use a Norwood-type strategy to treat these patients, the anteroposterior arrangement of the vessels make a DKS anastomosis suboptimal and can lead to significant compression of the LPA by the dilated DKS with time. For these patients, if the coronary anatomy is conducive, a palliative arterial switch operation may be performed, usually with aortic arch reconstruction and sometimes requiring placement of a PAB on the neopulmonary trunk if the restriction at the VSD is not severe.

Interstage Period

The interstage is the period of time between the first and second palliation, which is associated with significant morbidity and mortality. Common issues include feeding difficulties, growth failure, residual defects, and difficulty achieving balanced circulation. Postoperative management will vary based on the specific SV defect, but generally the following clinical milestones should be achieved prior to transfer out of the CICU:

- Off continuous IV infusions
- All pacing wires and tubes removed (except PICC or peripheral IV access)
- Stable saturations on room air or minimal nasal cannula support
- Well-controlled arrhythmias with clear documentation of management plans
- Well-controlled BP with clear documentation of goals and medication plans
- All PO medications except IV antibiotic therapy as needed
- Tolerating narcotic weaning plan
- Tolerating at least 100 mL/kg/day of enteral feeds with no signs of feeding intolerance (emesis, bloody stools, etc.) and positive weight gain
- Recent postoperative echocardiogram with stable findings and no major residual lesions

Infants who have undergone a Norwood-type operation or have otherwise been identified for enrollment in SV home monitoring should be admitted to the Cardiology NP team. Generally, evening and weekend transfers should be avoided and face-to-face hand-off with members of the surgical, CICU, and cardiology teams should occur. A low threshold should be maintained for readmission to the CICU for red flag events, which include:

- Desaturation and/or cyanosis
- Tachypnea and increased work of breathing
- Feeding intolerance (emesis, hematochezia, loose stools)
- Increased irritability, fussiness, or fatigue
- Fever or other signs of illness

After discharge, surveillance should include biweekly echocardiograms to evaluate ventricular function, AV valve stenosis/regurgitation, shunt/conduit flow, atrial-level shunt, and systemic outflow tract/aortic arch. In addition, monthly ECGs are obtained and CXR imaging is performed as needed.

Discharge planning should start upon transfer out of the CICU with implementation of the standardized discharge checklist. Multidisciplinary collaboration is needed to achieve a successful discharge:

- Cardiology, CHS, and SV teams for treatment and surgical plan
- Dietician for growth goals and formula-mixing instruction

- Care management for home equipment (pulse oximetry, feeding supplies, Lovenox®, etc.)
- Social work for family support and resources
- Developmental pediatrics for neurodevelopmental evaluation and follow-up

Resources for difficult-to-obtain medications and formulas should be identified prior to discharge. Immunizations should be administered prior to discharge if >6 weeks postoperatively. Pavlizumab (Synagis®) should also be given during RSV season. Complete follow-up of genetic studies and newborn screens is needed for any abnormalities, and a plan should be documented.

The SV team and home monitoring program help families care for their infants at home with the support, resources, and clinical expertise needed to transition to stage 2 successfully. Before discharge, parents learn to measure daily weights, saturations, and heart rate. They enter information in a logbook along with feeding information. Once familiar with the red-flag action plan and comfortable with daily care, discharge occurs. Plans for pre-Glenn study (cardiac catheterization vs. CTA) and timing of stage-2 palliation should be tentatively decided/scheduled. Follow-up visits with Cardiology/SV team, pediatrician, and consulting services should be scheduled as well. Readmission during the interstage period is not uncommon.

Second-Stage Palliation

Preoperative Evaluation

Stage-2 palliation typically occurs at 4-6 months. The SV team is responsible for obtaining and reviewing pre-Glenn imaging and collaborating with the surgical team on the type of imaging and timing of stage-2 palliation. Imaging targets that should be assessed include the aortic arch (in particular if a Norwood-type procedure was performed), the morphology and dimensions of the branch PAs, and the anatomy of the systemic veins, in particular the SVC.

CTA is usually adequate and it can usually be completed on an outpatient basis without anesthesia. Cardiac catheterization is reserved for patients who have residual defects that may require intervention such as recurrent coarctation, shunt/conduit stenosis, pulmonary vein stenosis, or restrictive ASD, or those with concerns regarding PVR (e.g., patients that had repair of anomalous pulmonary veins or prematurity with chronic lung disease). MRI is an option as well, but requires anesthesia. A preoperative Holter should be done to evaluate for ectopy and arrhythmias. Studies should be reviewed prior to surgical consultation.

Surgical Approach

Stage-2 palliation usually involves the creation of a cavopulmonary connection (SVC to PA) with takedown of previous shunts or conduits (Figure 39-1). If a patient has bilateral SVCs, bilateral bidirectional Glenn anastomoses are created unless the patient has a bridging innominate vein that is at least the size of the contralateral SVC. The azygous and hemiazygous veins are routinely ligated and divided. Any branch PA stenosis or hypoplasia is liberally addressed with patching.

Sometimes, an additional source of PBF is left for patients with heterotaxy, systemic

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POD	CV Meds	Diuretics	Fluid	Nutrition	Respiratory	Chest Tubes	Mobilization	Pain Meds	Disposition
0	Inotropes	None	Crystalloid / blood product replacement	NPO then clears if stable	OR extubation vs <6 hrs in ICU		Head of bed 30 degrees	PCA IV acetaminophen	CICU
1	Wean inotropes	IV furosemide (1 mg/kg q12, max 20 mg/dose)	50% maintenance	Start enteral intake	Min: 0.5 L/min	Mediastinal out	Encourage highest degree of mobilization	PCA IV acetaminophen Consider ketorolac	CICU
2	Initiate ASA Consider resuming home meds	IV furosemide (1 mg/kg q8, max 20 mg/dose) PO chlorothiazide	75-80% maintenance	PO encouraged	Min: 0.5 L/min	Mediastinal out Pleural to bulb	Walk in ICU at least once per shift	PO opioid + acetaminophen Consider ketorolac	Transfer to acute care floor
3	ASA Consider resuming home meds	PO furosemide (1.5 mg/kg q8, max 20 mg/dose) PO chlorothiazide	75-80% maintenance	Full PO	Min: 0.5 L/min	Pleural to bulb	Walk x3	PO opioid + acetaminophen PO NSAID	Transfer to acute care floor
4	ASA	Optimize PO diuretics	75-80% maintenance	Full PO	Stop O ₂ if sats >94%	Remove if <2 mL/kg/d each	Full mobilization as at home (min 3x)	PO NSAID PRN PO opioid	
5	ASA	Decrease or maintain	75-80% maintenance	Full PO	Stop O ₂ if sats >94%	Remove if <2 mL/kg/d each	Full mobilization as at home (min 3x)	PO NSAID PRN PO opioid	D/c criteria met?
6	ASA	Decrease or maintain	75-80% maintenance	Full PO	Stop O ₂ if sats >94%	Remove if <2 mL/kg/d each	Full mobilization as at home (min 3x)	PO NSAID PRN PO opioid	D/c criteria met?

Figure 39-3. Postoperative Fontan management protocol. Developed by the TCH Heart Center and the TCH Evidence-Based Outcomes Center. ASA: aspirin, CT: chest tube, NSAID: non-steroidal anti-inflammatory drug, PCA: patient-controlled analgesia, POD: postoperative day.

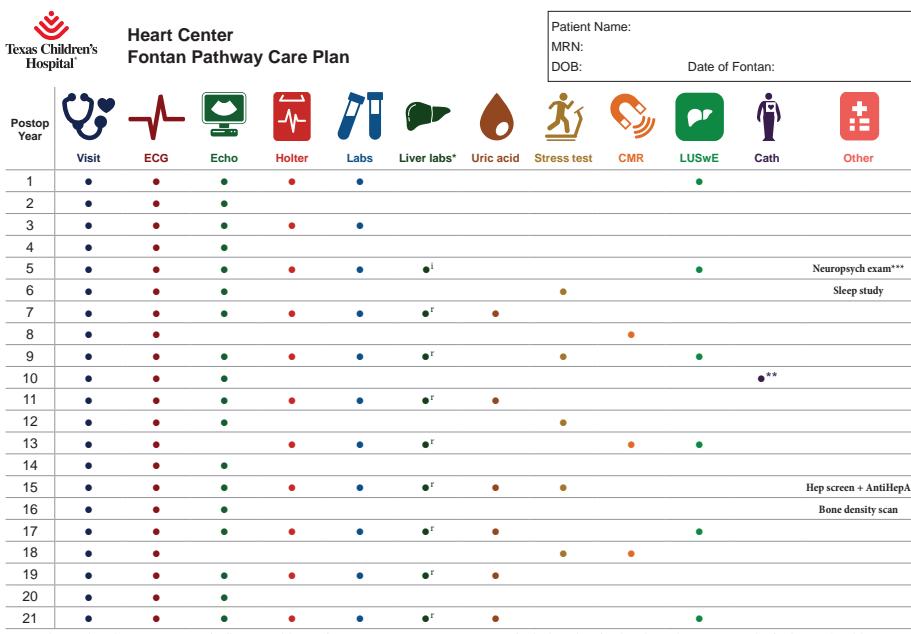


Figure 39-4. Heart Center Fontan Pathway Care Plan.

venous anomalies, genetic syndromes, or those in which it is unclear whether they will be adequate Fontan candidates in the future. This additional source of PBF is achieved by leaving some prograde flow across the RVOT; an existing PAB may be tightened or if there is not enough RVOT obstruction, a PAB may be placed. By providing additional PBF through the RVOT, these “pulsatile Glenns” may allow patients to be less desaturated as they grow, allow a delay in the next stage of palliation, and may decrease the creation of pulmonary arteriovenous malformations by allowing hepatic flow into the PAs.

The Glenn procedure is performed on CPB with the heart beating except in those patients that required intracardiac repairs, arch reconstruction, or pulmonary valve exclusion. If the RVOT is patent and a pulsatile Glenn is not planned, the heart is arrested, the main PA transected, the pulmonary valve oversewn, and the cardiac PA stump closed. Failure to oversew the pulmonary valve (or perform a valvectomy if the valve is very small) may lead to thrombus formation between the pulmonary valve and the MPA stump, which may then enter the ventricle and cause systemic embolization.

It is customary to place IJ and femoral venous catheters in patients undergoing Glenn procedures. This allows postoperative measurement of Glenn pressures and transpulmonary gradients. Adequate Glenn pressures at the end of the procedure are usually in the low-to-mid teens with a transpulmonary gradient between 3 and 8 mmHg. The IJ “Glenn” line is usually removed on the first postoperative day. Low-dose heparin may be started a few hours postoperatively and continued until the Glenn line is out if the SVC is small, especially in patients with bilateral SVCS.

Postoperative Management

Understanding cardiopulmonary interactions is implicit in postoperative management after the Glenn procedure. Because positive-pressure ventilation after cavopulmonary connection is associated with a decrease in PBF and thus cardiac output, early spontaneous breathing is desirable and extubation commonly occurs in the OR or shortly after arrival to the CICU. For those patients who are better served by positive-pressure ventilation in the early postoperative period (circumstances including ongoing bleeding, underlying airway disease, or depressed myocardial function), a ventilator strategy that allows for permissive hypercarbia at the lowest possible mean airway pressure (single digit) is optimal. Though acute hypocarbia and alkalosis have been shown to acutely reduce PVR, the benefit does not outweigh the negative impact of increased intrathoracic pressure on postoperative Glenn circulation. Alternatively, hypercarbia (pCO_2 45-55 mmHg) has been shown to increase PBF and oximetry measures likely through hypercarbia-induced increases in cerebral blood flow (cardio-pulmonary-cerebral interaction). In circumstances in which the transpulmonary gradient is wide and oxygenation concerning, iNO may be indicated.

The shift from dual-distribution circulation to superior cavopulmonary connection commonly results in postoperative hypertension. BP control through vasoactive infusions (milrinone + nifedipine or nicardipine) in the early postoperative period will assist in augmenting cardiac output through afterload reduction as well as lowering the end-diastolic pressures, allowing for optimal transpulmonary circulation. All patients will likely begin transitioning to angiotensin-converting enzyme (ACE) inhibitors on the first postoperative day. Diuretics are needed to maintain low Glenn pressures

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and to reduce the risk of pleural effusions. Even though the “Glenn” IJ line is typically removed on postoperative day 1, thorough physical exams including attention to the anterior fontanelle and facial edema assist in titration of diuretics.

It is common for patients after a Glenn procedure to have some degree of transient upper body edema that may lead to headaches. Alternating acetaminophen and non steroidal anti-inflammatory is effective to manage pain.

Third-Stage Palliation

Preoperative Evaluation

The Fontan operation is the third palliation and is usually done at 3-5 years of age. Timing varies based on individual degree of cyanosis. Evaluation includes:

- **ECG and Holter.** Patients with a Fontan circulation benefit from adequate AV synchrony. It is thus important to rule out arrhythmias or AV block that may require treatment.
- **CXR.** Assess cardiomegaly and pulmonary abnormalities.
- **Echocardiogram.** A complete echocardiogram is essential to assess abnormalities that may lead to failure of a Fontan circulation. These include lesions such as AV valve regurgitation, ventricular function, aortic arch or ventricular outflow tract obstruction, and other residual lesions.
- **Cardiac catheterization.** A successful Fontan circulation is dependent on low PVR. For this reason, all patients undergo a pre-Fontan cardiac catheterization for hemodynamics and to address residual lesions. Important hemodynamic values include Glenn pressures, PVR, Qp:Qs, end-diastolic pressure, transpulmonary gradient, and other intracardiac or aortic gradients. In addition, it is important to assess the IVC morphology and orientation (in particular in patients with heterotaxy), branch PA anatomy, patency of the Glenn anastomosis, the presence of collaterals (venovenous and arteriovenous) and arteriovenous malformations. Significant collaterals may be occluded in the lab.

Surgical Approach

The Fontan completion involves the creation of an extracardiac conduit (usually 18- or 20-mm Gore-Tex® graft) that connects the IVC to the PA (Figure 39-2). The procedure is performed using CPB via bicaval cannulation with the heart beating, unless intracardiac repairs are necessary. Any stenosis or hypoplasia of the branch PAs is managed with a patch plasty. If the RVOT is patent (pulsatile Glenn), the pulmonary valve is oversewn and the MPA sutured closed using a brief episode of cardiac arrest.

A fenestration (a communication between the Fontan conduit and the atrium) is created selectively in high-risk patients with hypoplastic PAs or high PVR in order to serve as a pop-off for elevated Fontan pressures. Currently, more than 75% of patients at TCH undergo a nonfenestrated Fontan.

Due to an expected higher venous pressure as part of the Fontan circuit, patients may have significant chest tube output postoperatively. It is therefore routine to open both pleural spaces and place bilateral pleural chest tubes in addition to the mediastinal chest tube. The mediastinal chest tube will usually be removed on postoperative

day 2 while pleural chest tubes will remain in place until chest tube output decreases. Patients are routinely extubated in the OR since positive-pressure ventilation is unfavorable for the Fontan circuit.

Postoperative Management

Figure 39-3 depicts the TCH protocol for postoperative management of Fontan patients. The Fontan procedure “septates” the circulation in patients with SV physiology. PBF is a passive diastolic phenomenon (occurs during negative inspiration and is impeded with positive-pressure ventilation) and cardiac output is dependent on pulmonary venous return. This phenomenon dictates the postoperative management of patients with a Fontan circulation as pulmonary mechanics and volume status will determine cardiac output.

The transpulmonary gradient (CVP [which equals the PA pressure] minus LAP) is an important concept in Fontan management and must be low for good cardiac output. Strategies to decrease PVR will improve PBF and thus pulmonary venous return and cardiac output. Optimal Fontan physiology includes:

- CVP / PA pressure: 10-15 mmHg (most are >12 mmHg)
- LAP: 5-10 mmHg
- Transpulmonary gradient: 5-10 mmHg
- AV synchrony
- Lack of systemic hypertension

Some important tenants of postoperative Fontan management are:

- Negative-pressure ventilation improves hemodynamics; it is thus important to extubate early.
- Volume is needed in the early postoperative period (first 1-2 days) to maintain cardiac output. Avoid diuresis during this time. Low-dose vasopressin may help minimize excessive volume needs.
- Patients require eventual fluid restriction and diuresis to decrease chest-tube output toward discharge (days 2-3 until the time of discharge). Fluid restriction and diuresis is maintained at the time of discharge.
- AV synchrony is important to maintain cardiac output and avoid increases in atrial pressure that can impede pulmonary venous return.
- Avoid significant catecholamines as tachycardia decreases filling time and can lead to arrhythmias.
- A fenestration allows for maintenance of cardiac output in the face of less-than-optimal pulmonary hemodynamics. As such, it is used at TCH in high-risk patients with elevated PVR.
- Patients with SV physiology have high SVR and it is important to treat this to augment systemic perfusion.
- If there are persistent pleural effusions it is important to assure that they are not chylous (for management of chylothorax, see Chapter 77).

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- If a patient has unexpected cyanosis after a Fontan procedure, evaluation should include:
 - Ventricular function/AV valve regurgitation (decreased mixed venous oxygenation)
 - Shunting through a fenestration or baffle leak
 - Systemic venous collateralization
 - Intrapulmonary arteriovenous malformations
 - Pulmonary pathology/diaphragm paresis
 - Hepatic venous connection to the atrium (not incorporated into the Fontan circuit)
 - Coronary sinus flow

Long-Term Follow-up

The unique nature of the Fontan physiology presents patients with a multitude of long-term issues. The Heart Center Fontan Pathway Care Plan (Figure 39-4), was developed so that longitudinal, standardized testing can be used to monitor specific long-term complications commonly encountered in Fontan patients. These include:

- **Cardiac**

- *Structural* – There is universal diastolic dysfunction with any elevation in end-diastolic pressures leading to further elevation in Fontan pressures. Significant AV valve regurgitation independently predicts poor outcomes. Systolic dysfunction is less common, but still a noted concern.
 - *Electrical* – Bradycardia is common (mainly sinus node dysfunction). Tachycardia is common (atrial > ventricular), especially intra-atrial reentry tachycardia.

- **Liver.** Chronic congestive hepatopathy is the rule with most patients eventually developing some degree of cirrhosis. There are rare reports of hepatocellular carcinoma.

- **Fontan pathway obstruction.** Any obstruction in the Fontan pathway needs to be corrected since it will lead to elevation in Fontan pressures.

- **Pulmonary.** Any branch pulmonary stenosis needs to be corrected. Venovenous collateral vessels (from systemic to pulmonary veins) can lead to desaturations. Elevated PVR is rare but difficult to treat.

- **Vascular.** The diagnosis of protein-losing enteropathy (PLE) is made via clinical suspicion and can be confirmed with testing for fecal alpha-1-antitrypsin. Treatment is individualized (with medical treatment often unsuccessful) and ranges from medications (oral steroids, pulmonary vasodilators), to catheter interventions (fenestrating the Fontan circuit), to surgery (Fontan conversion, transplantation).

- **Hematologic.** Patients may present with coagulopathy. Thromboemboli are common with high risk of venous thromboemboli, pulmonary emboli, or stroke.

- **Renal.** Chronic kidney disease is likely underrecognized.

In a series of 610 patients undergoing Fontan procedures at TCH with a median follow-up of 7 years, freedom from any Fontan failure (defined as death, heart transplant, take-down or revision of the Fontan circuit, creation of fenestration, major reintervention, or development of plastic bronchitis or PLE), was 91% at 5 years and 89% at 10 years (Mery et al. 2019).

Suggested Readings

- Castellanos DA, Ocampo EC, Gooden A, et al. Outcomes associated with unplanned interstage cardiac interventions after Norwood palliation. *Ann Thorac Surg* 2019;108:1423-1429.
- Chacon-Portillo MA, Zea-Vera R, Zhu H, et al. Pulsatile Glenn as long-term palliation for single ventricle physiology patients. *Congenit Heart Dis* 2018;13:927-934.
- Heinle JS, Carberry KE, McKenzie ED, et al. Outcomes after the palliative arterial switch operation in neonates with single-ventricle anatomy. *Ann Thorac Surg* 2013;95:212-218.
- Mery CM, De León LE, Trujillo-Díaz D, et al. Contemporary outcomes of the Fontan operation: a large single-institution cohort. *Ann Thorac Surg* 2019;108:1439-1446.