Current Management of Infants and Children With Single Ventricle Anatomy

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Children with single ventricle anatomy are among the most complicated and challenging patients encountered in pediatric cardiology. Current management involves staged surgical procedures, beginning with neonatal palliation and followed by a bidirectional cavopulmonary anastomosis in infancy and culminating in the Fontan procedure. The Fontan procedure, despite separating the circulation, remains a palliative procedure with many long-term concerns. This report discusses the staged surgical management of patients with single ventricle anatomy and the nursing issues relevant to each stage.

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MANY CHILDREN WITH complex congenital heart disease have only one functional ventricle. Single ventricle anatomy (SVA) is the term used to describe a functional single ventricle regardless of anatomic subtype (Tweddell, Litwin, Thomas, & Mussatto, 1999) (Table 1). The functional ventricle provides a common mixing chamber and must pump both the pulmonary and systemic circulations; it is thus volume overloaded. The majority of defects are diagnosed within the first days or weeks of life because of cyanosis or congestive heart failure. An increasing number are diagnosed prenatally by fetal echocardiography. Without intervention, all patients with SVA will develop sequelae associated with chronic cyanosis (i.e., clubbing, polycythemia, increased risk of cerebral emboli, etc.) and eventually will develop decreased ventricular function. The goal of therapy for these children is to improve hemodynamics by surgically separating the systemic and pulmonary circulations and thereby relieving cyanosis and volume overload. This goal is ultimately accomplished with the Fontan procedure.

Children with SVA and their families are frequent visitors to the health care setting throughout their lives. Because of their unique cardiac anatomy, they challenge their caregivers to fully understand their hemodynamics and to plan every intervention with an eye toward the future. Because they must undergo multiple tests, catheterizations, and surgeries, they demand compassionate, individualized care. This report discusses the staged surgical management of patients with SVA and the nursing issues relevant to each stage.

There has been a shift in emphasis from selection for Fontan procedure to preparation for Fontan procedure (Jacobs, 2000). In the current era, patients with SVA are carefully managed from birth with a combination of staged surgical procedures to minimize future complications; these are outlined in Figure 1.

Initial neonatal palliation seeks to provide complete relief of systemic obstruction and manipulation of pulmonary blood flow to achieve a balanced circulation. An acceptable balance between the pulmonary and systemic output provides enough pulmonary blood flow for adequate oxygen delivery to prevent acidosis without an excessive volume load on the single ventricle. Some infants with SVA have a well-balanced circulation and need no neonatal palliation.

A second palliative procedure often consists of a cavopulmonary anastomosis in the form of a bidirectional Glenn shunt or Hemi Fontan procedure. This intervention provides passive pulmonary blood flow and decreases the volume load on the single ventricle.

The final stage is the Fontan procedure. Critical
to the success of the Fontan procedure is an unobstructed pulmonary circulation with low pulmonary artery pressures and pulmonary vascular resistance (PVR) and normal ventricular function. Risk factors for poor outcome after the Fontan procedure include ventricular hypertrophy and abnormal ventricular systolic or diastolic function, elevated right atrial pressure or PVR, pulmonary artery distortion, and atrioventricular valve regurgitation (Tweddell, 1999) (Wernovsky & Bove, 1998).

NEONATAL PALLIATION

SVA patients may be divided into three broad categories: (a) Those with unobstructed systemic flow and obstruction to pulmonary flow, (b) those without obstruction to either systemic or pulmonary blood flow, and (c) those with obstruction to systemic flow and unobstructed pulmonary flow. In most patients, there is obstruction either to the pulmonary or systemic outflow tracts. Having no obstruction or obstruction to both outflows is rare. All exhibit some degree of cyanosis because of mixing in the single ventricular chamber.

Obstruction To Pulmonary Blood Flow

Infants with unobstructed systemic flow and obstruction to pulmonary flow are cyanotic at birth. The degree of cyanosis is determined by the severity of pulmonary obstruction. Children with severe pulmonary stenosis or pulmonary atresia may be completely dependent on the patent ductus arteriosus (PDA) to provide pulmonary blood flow and may become critically ill when the ductus begins to close. Administration of prostaglandins keeps the ductus open temporarily. Children with moderate obstruction to pulmonary flow may do well without intervention for years. These children are vis-

Table 1. Types of Single Ventricle Anatomy

<table>
<thead>
<tr>
<th>Type of Single Ventricle Anatomy</th>
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<tbody>
<tr>
<td>Tricuspid atresia</td>
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<tr>
<td>Hypoplastic left heart syndrome</td>
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<tr>
<td>Heterotaxy with common AV valve</td>
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<tr>
<td>Unbalanced AV canal</td>
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<tr>
<td>Double outlet right ventricle</td>
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<td>Double inlet left ventricle</td>
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Abbreviations: AV, atrioventricular.
ibly cyanotic but are able to grow and thrive. For children with SVA, the ideal pulmonary blood flow is between one and two times the systemic flow and produces an arterial saturation that averages from 76% to 85% (Fyler, 1992).

Infants who require early intervention because of inadequate pulmonary blood flow will receive some type of aorta-to-pulmonary shunt, usually a modified Blalock-Taussig shunt (MBTS) (Figure 2). An MBTS allows blood to flow from the aorta to the lungs through the placement of a tube graft (usually 3.5 mm in neonates) between the subclavian artery and the pulmonary artery. The size of the shunt limits pulmonary blood flow and the shunt is purposely kept small to prevent an increase in PVR. A shunt is a time-limited intervention, as it has no growth potential. As the infant grows, the shunt will become restrictive, and further palliation will be needed.

No Pulmonary or Systemic Obstruction

With no obstruction to pulmonary or systemic flow, infants with SVA may develop congestive heart failure (CHF) because of excessive pulmonary blood flow. Over the first few weeks of life, PVR falls with a subsequent increase in pulmonary blood flow. With no obstruction to pulmonary or systemic blood flow, blood will follow the path of least resistance and preferentially flow to the lower resistance pulmonary circulation. With the large pulmonary flow, cyanosis may be slight, and signs of CHF including tachypnea, tachycardia, diaphoresis, poor weight gain, and respiratory infections are prominent. If medical management of CHF is unsuccessful, pulmonary blood flow must be limited to control CHF and prevent further increases in PVR. A pulmonary artery band (PAB) may be placed around the main pulmonary artery and tightened to reduce pulmonary artery pressure to one-third to one-half the systemic pressure with a minimal reduction of systemic arterial saturation to between 75% and 90%. Potential disadvantages of banding include distortion of the main pulmonary artery, development of subaortic stenosis, and damage to the pulmonary valve (Wernovsky & Bove, 1998).

Obstruction to Systemic Flow

Hypoplastic left heart syndrome (HLHS) is a prime example of the third category of neonates with SVA: those infants with obstruction to systemic flow and unobstructed pulmonary flow. This syndrome comprises underdeveloped or atretic mitral and aortic valves, a very small left ventricle that is unable to sustain systemic circulation, and a hypoplastic ascending aorta. In these infants, systemic circulation is supplied entirely through the PDA. A left to right shunt at the level of the atrium, most often a patent foramen ovale, is necessary to allow oxygenated blood from the left atrium to enter the systemic circulation via the PDA.

The immediate treatment is administration of prostaglandins to maintain ductal patency and correct metabolic acidosis. Intubation, mechanical ventilation, and inotropic support are often needed. If pulmonary overcirculation is severe, ventilation with a hypoxic gas mixture (17% to 20% oxygen), which is achieved by adding nitrogen to the gas mixture, may be used to increase PVR and decrease pulmonary blood flow. Surgical intervention

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**Figure 2.** Right modified Blalock-Taussig shunt. Gore-tex tube graft (W.L. Gore & Associates, Flagstaff, AZ) from the subclavian artery to the right pulmonary artery to provide pulmonary blood flow.
is aimed at establishing systemic circulation and controlling pulmonary blood flow. Heart transplantation is another treatment option.

The stage 1 palliation developed by Norwood, Lang, and Hansen (1983) provides systemic circulation by transecting the main pulmonary artery and attaching it to the hypoplastic ascending aorta. This allows the right ventricle to pump blood to the systemic circulation. As the main pulmonary artery is now used in the systemic circulation, pulmonary circulation is established by placing a modified Blalock Taussig shunt. The existing PDA is ligated and the atrial septum is opened to allow flow of oxygenated blood from the left atrium to reach the right atrium and single right ventricle and then to the systemic circulation (Fig 3). This surgical technique is also performed in infants with other forms of SVA that include severe aortic obstruction or aortic arch hypoplasia.

Post-operative care. Postoperatively, hemodynamic stability is dependent on maintaining a balance between pulmonary and systemic flow. Complications present as symptoms of low cardiac output, CHF, and excessive cyanosis (Table 2). Providing adequate systemic blood flow may require inotropic support in the early postoperative period. As PVR continues to fall, there may be excessive flow through the shunt, resulting in CHF. Aggressive treatment with diuretics and afterload reduction may be required to manage CHF. Excessive cyanosis may result from structural limits to pulmonary blood flow or pulmonary causes of poor oxygenation such as edema or effusions.

Oxygen therapy for decreased oxygen saturations must be used with extreme caution in infants with HLHS both before and after repair. Oxygen is a potent pulmonary vasodilator, and blood will preferentially flow to the lungs at the expense of the systemic circulation. Effects of oxygen therapy must be carefully evaluated and, even during resuscitation attempts, 100% oxygen is rarely used in this population. Oxygen saturations of 75% to 85% are ideal and imply a good balance between systemic and pulmonary blood flow. Saturations above 90% indicate excessive pulmonary blood flow, and symptoms of CHF may be evident.

Although mortality for patients with HLHS who undergo stage 1 palliation is relatively high compared with other neonatal repairs, it has decreased significantly in the last decade. Hospital survival after repair at a large volume center was recently reported at 76% (Bove & Lloyd, 1996), and survival to second stage repair was 77% at another center (Tweddell, Hoffman, & Fedderly, 2000). Patients with cardiac defects other than HLHS who undergo Stage 1 repair have better outcomes (Daebrtit, et al., 2000).

Nursing Care

Infants with SVA are fragile and require knowledgeable nursing care to attain the best outcomes. Conservation of energy, maintenance of normothermia, and aggressive treatment of fevers are important. Increased body temperature increases oxygen consumption, which causes decreased arterial saturations. The tachycardia associated with fever places added stress on an already overworked
ventricle. Dehydration, which can increase the risk of thrombosis, should be avoided.

Infants with SVA are notoriously poor feeders. They tire easily, may have poor or inconsistent sucks, and may be unable to meet their caloric needs orally. Nurses can encourage smaller, more frequent feedings and help parents to position infants in a more upright position to increase the ease of feeding. Healthy infants require about 110 kcals/kg per day for normal weight gain. Infants with SVA have higher caloric needs—at least 130 kcals/kg per day. Breast milk, which contains essential immunologic properties, balanced nutrients, and digestive substances, is the recommended infant food and should be used whenever possible. Although some infants with SVA can nurse easily, those who cannot nurse can benefit from receiving breast milk by bottle or gavage feedings. High calorie formulas can meet increased caloric needs with less fluid volume. Calories can be added to pumped breast milk when needed. Nutritionists can help parents with recipes for formulas and nutritional assessments of their infants. Many infants will require assistance with nasogastric or nasojejunal tube feedings until adequate oral feeding can be established. Some infants will require placement of gastrostomy tubes for longer-term nutritional support.

**Parent Support and Education**

Parents are understandably stressed and overwhelmed when their newborn is diagnosed with severe congenital heart disease. Nurses can make a significant impact on how effectively parents cope with the stresses brought on by the diagnosis and hospitalization of the infant. Transition to home is also a stressful time as families become independent in their child’s care. Nurses are in a unique position to provide parental education about the care of these fragile infants and role-model interactions with these infants that will promote parent-infant bonding. Family education includes well baby care and nutritional concerns, as well as medication administration and assessment of signs and symptoms of cardiac problems and respiratory distress. Maintaining open communication with the health care team is important. Referrals to community agencies and early intervention programs (EIP) can help ease the transition from hospital to home and promote the infant’s growth and development. EIP can be especially helpful in promoting gross motor skills, which tend to be delayed in this population.

**CAVOPULMONARY ANASTOMOSIS**

The second palliative procedure for SVA usually occurs between 4 and 9 months of age. Preoperatively, a cardiac catheterization is done to assess pulmonary pressures, PVR, and ventricular function, and interventional procedures may be performed. The operative procedure is a bidirectional cavopulmonary anastomosis (BCPA), either bidirectional Glenn shunt or hemi-Fontan. The bidirectional Glenn shunt (BDG) involves a direct end-to-side anastomosis of the superior vena cava (SVC) to the right pulmonary artery (Fig 4). With the hemi-Fontan, the lower SVC is opened vertically to the right atrial appendage and anastomosed to the right pulmonary artery. A patch is placed in the superior vena cava-right atrial junction to block blood flow to the atrium and divert blood into the pulmonary arteries (Fig 5). In cases in which pulmonary artery stenosis is present, pulmonary artery dilatations and stenting may be done preoperatively in the cardiac catheterization laboratory, or pulmonary artery plasty may be done at the time of the bidirectional cavopulmonary anastomosis. Any
pre-existing aorto-pulmonary shunts or pulmonary artery bands are taken down at this time. The main pulmonary artery may be ligated and oversewn or left as an additional source of pulmonary blood flow, in some situations. Both procedures relieve the volume load on the single ventricle, which helps to prevent the development of atrioventricular valve regurgitation and preserve ventricular function. Effective pulmonary blood flow is increased. Numerous studies have shown that outcomes after the Fontan procedure are improved when the patient had a prior cavopulmonary anastomosis (Forbes, et al., 1997; Reddy, McElhinney, Moore, Haas, & Hanley, 1997; Van Arsdell, et al., 2000). An improvement in growth and development is usually noted in children after Stage 2 palliation.

The postoperative course following BCPA is usually uncomplicated, and the mortality rate is very low. Immediate postoperative fluid retention may result in edema and decreased oxygen saturations. Diuretic therapy usually resolves the edema over the first 48 hours or so, and oxygen saturations rise to between 75% and 85%. Increased SVC pressure may occur postoperatively either transiently secondary to volume overload or because of obstruction of the anastomosis or distal pulmonary artery distortion. The increased pressure can result in facial and upper extremity edema. Hypertension, headaches, and irritability are common. Diuretics and elevation of the head may help relieve increased pressures. Continuous analgesia may be needed for headache. Pleural effusions secondary to an increase in central venous pressure occur in some patients.

Long-term issues after BCPA procedures include the development of venovenous collaterals and pulmonary arteriovenous malformations, which cause increasing cyanosis. Venovenous collaterals carrying blood from the higher pressure SVC to the lower pressure inferior vena cava (IVC) lead to increased cyanosis over time. The absence of hepatic venous return to the pulmonary circulation after BCPA results in the development of pulmonary arteriovenous malformations (AVMs). (Srivastava, et al., 1995).

Prior to the Fontan procedure (stage 3), patients have a cardiac catheterization to measure pulmonary artery pressure, PVR, end diastolic pressures and to assess atrioventricular valve insufficiency. Interventional procedures, such as balloon dilation of pulmonary artery stenosis or coiling of collateral vessels, may be done in preparation for the Fontan procedure.

**FONTAN PROCEDURE**

The underlying principle of the Fontan circulation is that the pulmonary circulation can be perfused without a ventricular pump. The Fontan procedure separates the systemic and pulmonary circulations to achieve normal (or near normal) oxygen saturations and decrease the volume load on the single ventricle. The single ventricle becomes the systemic pump, pumping blood through the aorta to the body. The pulmonary circulation receives passive nonpulsatile blood flow directly from the vena cava (SVC and IVC), without a ventricular pump. If pulmonary artery pressure is normal, the systemic venous pressure in the vena cava is able to exceed pulmonary artery resistance to allow forward flow into the pulmonary arteries.

First described by Fontan and Baudet in 1971 for repair of tricuspid atresia, the original operative technique has had many modifications and has been applied to all types of single ventricle anatomy. Currently, the Fontan procedure is often performed at 18 months to 4 years of age. There are several operative techniques used to create the Fontan circulation, which are outlined in Table 3.

A significant modification of the Fontan procedure was the addition of a fenestration into the atrial baffle (Bridges, Lock, & Castaneda, 1990). A similar idea was an adjustable intra-atrial communication with surgical ligatures (Laks, et al., 1991). A small hole is placed in the baffle to act as a “pop-off” to decrease high pressure in the lateral
tunnel. It also increases atrial blood flow, which provides adequate ventricular preload. A fenestration can also be placed in an extracardiac conduit. Oxygen saturations are decreased between 80% and 90% in the early postoperative period because deoxygenated blood flows right to left through the fenestration into the atrium. Placement of a fenestration has decreased mortality after the Fontan (Gentiles, et al., 1997) and has significantly decreased the incidence and severity of postoperative pleural effusions (Bridges, et al., 1992). The fenestration can be closed later by a device in the catheterization laboratory, although some will close spontaneously.

Modified ultrafiltration is another technical improvement that is widely applied for Fontan procedures. After cardiopulmonary bypass, modified ultrafiltration through the bypass circuit removes excess tissue water. This decreases myocardial edema, improves diastolic complianc and has been shown to decrease mortality after the Fontan procedure (Elliot, 1993, Koutlas, et al. 1997).

**Postoperative Management**

The general goal of treatment in the early postoperative period is to maintain cardiac output at the lowest central venous pressure possible (Wernovsky & Bove, 1998). The central venous pressure (CVP) is equal to the pressure inside the Fontan pathway. This pressure must be slightly greater than the pulmonary artery pressure for blood to flow forward into the lungs. Maintaining low pulmonary artery pressures becomes critical to maintaining a low CVP. Cardiac output is dependent on pulmonary venous return. If increases in pulmonary artery pressure and pulmonary resistance reduce blood flow into the pulmonary circulation, there will be less blood returning to the heart, causing a fall in cardiac output. A few millimeters of pressure difference in the Fontan path-

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**Table 3. Modifications of the Fontan Procedure**

<table>
<thead>
<tr>
<th>Operative Techniques</th>
<th>Comments</th>
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<tr>
<td><strong>Atriopulmonary Anastomosis</strong> (RA-PA anastomosis) (see Figure 6)</td>
<td>Entire RA subjected to increased pressure causing atrial distension</td>
</tr>
<tr>
<td>Direct anastomosis between RA and PA</td>
<td>Atrial distension and multiple suture lines contributed to atrial arrhythmias</td>
</tr>
<tr>
<td>Systemic venous return enters RA and exits through new anastomosis directly to the PAs</td>
<td>Systemic venous congestion and pleural/pericardial effusions common</td>
</tr>
<tr>
<td>ASD closed or LA blood flow baffled to the right AV valve (as in HLHS)</td>
<td>Swirling sluggish flow patterns impeded flow to PA increases risk of thromboembolism</td>
</tr>
<tr>
<td>Commonly performed in 1980s, rarely done now</td>
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**Total Cavopulmonary Connection** (see Figure 7)

An intra-atrial baffle creating a lateral tunnel within the RA is combined with a BCPA (may have been done at a previous operation)

- Baffles the IVC blood up through the baffle to the SVC, then to the PAs
- Intra-atrial baffle frequently fenestrated

- Improved flow patterns within the baffle
- Increases velocity of PA flow
- Decreases thrombus risk
- Fewer atrial suture lines and less atrial distension may reduce incidence of atrial arrhythmias

**Extracardiac Conduit**

(see Figure 8)

- Prosthetic or homograft tube graft to carry IVC flow behind the heart to the PA
- Combined with a BCPA (may have been done at a previous operation)

- Short or no cardiopulmonary bypass
- Avoids atrial suture lines and RA distension, may decrease incidence of arrhythmias
- Preferred with complex pulmonary venous return (pulmonary veins to RA would be obstructed with lateral tunnel)
- Preferred with complex systemic venous connections (IVC not in alignment with SVC)
- Tube graft has no growth potential (procedure performed at 3 yrs/15 kg) so adult-sized conduit can be used
- Potential for conduit obstruction
- Potential for thromboembolism

Abbreviations: RA, right atrium; PA, pulmonary artery; SVC, superior vena cava; IVC, inferior vena cava; ASD, atrial septal defect; BCPA, bi-directional cavopulmonary anastomosis.

*Petrossian et al., (1999).*
way or pulmonary circulation can be the difference between a good outcome and a failing Fontan.

Comprehensive assessment and management by an experienced multidisciplinary team is critical in the early postoperative period. All caregivers need to understand the unique physiology of the post-Fontan procedure patient. There are many excellent reviews of the general principles of pediatric postoperative cardiac care in the literature. The following discussion focuses on issues unique to this patient population.

**Cardiovascular issues.** Patients are often unstable in the first 24 to 48 hours after the Fontan procedure because of the deleterious effects of cardiopulmonary bypass (especially increases in PVR) and fluid shifts. Pre-existing ventricular dysfunction or atrioventricular valve regurgitation place patients at greater risk for postoperative problems. Continuous monitoring of all hemodynamic parameters is essential in the early postoperative period. Intracardiac lines placed in the right atrium and the left atrium are very helpful after the Fontan procedure. The right atrial line assesses systemic venous pressure and pressure in the Fontan pathway. It is also very helpful in fluid management. The right atrial pressure should be 12 to 15 mmHg after the Fontan procedure. The left

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**Figure 6.** Atriopulmonary anastomosis (right atrium to pulmonary artery anastomosis). Direct anastomosis between the right atrium and pulmonary artery. All venous return enters the right atrium and exits through the new anastomosis to the pulmonary artery. Main pulmonary artery oversewn. Atrial septal defect closed.

**Figure 7.** Total cavopulmonary connection. Intra-atrial lateral tunnel baffle directs inferior vena cava flow to the superior vena cava which is anastomosed to the right pulmonary artery. Fenestration is placed in baffle to relieve elevated pressure in lateral tunnel in early postoperative period. Combined with bidirectional cavopulmonary anastomosis.

**Figure 8.** Extracardiac conduit. Conduit placed outside of right atrium, carrying inferior vena cava flow to the pulmonary artery. Combined with bidirectional cavopulmonary anastomosis.
atrial line assesses pulmonary venous return, ventricular preload, and ventricular function.

Low cardiac output is a common problem. Volume replacement to maintain adequate blood volume is initial management, along with inotropic support. Dopamine and dobutamine are the most common inotropes used in children. The addition of a phosphodiesterase inhibitor, milrinone, which offers both inotropic support and afterload reduction is becoming more common. Nitroprusside or other vasodilators may also be needed. Echocardiography should be done to rule out anatomic obstruction, effusion, or poor ventricular function in patients who do not respond adequately to therapy.

Arrhythmias result from pre-existing sinus node dysfunction, mechanical injury, injury to the conduction pathway, hypoxemia, or electrolyte imbalances. Atrial and junctional rhythms are most common. Junctional ectopic tachycardia and supraventricular tachycardia are managed with anti-arrhythmic medications, overdrive pacing, or other treatments. Slower arrhythmias are often managed with temporary pacing. Fontan patients rely on an adequate heart rate to maintain cardiac output. Persistent effusions and signs of low output are common with slow heart rates. Permanent pacing may be necessary to alleviate bradycardia and improve hemodynamics.

**Systemic venous hypertension and congestion.**
Acute systemic venous hypertension is a distinctive problem after the Fontan procedure. Increased venous pressure is the result of the sharp rise in right atrial pressure from a mean of approximately 5 mmHg preoperative to a mean of 12 to 15 mmHg postoperative to propel blood into the pulmonary circulation. Elevated right atrial, SVC, and IVC pressures result in increased pressure in the capillary bed, which causes accumulation of extracellular fluid. Common signs of systemic venous hypertension are an increased right atrial pressure, distended neck veins, and decreasing O₂ saturations. Systemic venous congestion results in pleural and pericardial effusions, hepatomegaly, ascites, and peripheral edema.

Pleuraleffusions are the most common manifestation of systemic venous congestion. Chest tube drainage may last several days to several weeks after the procedure; sometimes for longer periods. Symptoms of pleural effusions include tachypnea, decreased breath sounds, decreased O₂ saturations, grunting respirations, decreased exercise tolerance, irritability, anorexia, nausea, and vomiting. Chest radiograph confirms the clinical findings. Aggressive diuresis is often used, and mechanical drainage with a chest tube or pigtail catheter may be needed. The child’s fluid balance, heart rate, blood pressure, weight, electrolytes, and physical examination must be closely monitored. Preventing atelectasis and encouraging ambulation are important. Adequate pain management is especially important for children with chest tubes. With a focus on pain prevention, analgesics are given on a regular schedule during waking hours. Providing adequate nutrition is a challenge. Nurses, parents, and the dietitian can plan small, frequent high-calorie and high-protein meals and snacks. Children who are losing weight or are significantly protein depleted may need nasogastric supplemental feedings at night. If patients have high volumes of chest tube drainage with a falling serum albumin, fluid replacement with albumin may be needed to maintain intravascular volume. All patients with persistent effusions should have a catheterization to assess for Fontan pathway obstruction, patency of the fenestration, and the presence of collateral vessels. Interventional catheterization techniques may resolve these problems and improve hemodynamics.

Pericardial effusions are also seen after the Fontan procedure and can be dangerous because of potential compression of the heart and reduction in cardiac output. Pericardial effusions can have an insidious onset and occur days or weeks after surgery. Symptoms include tachycardia at rest, hypotension, diaphoresis, weak pulses and poor peripheral perfusion, nausea, vomiting, and irritability. The chest radiograph shows cardiomegaly and an enlarging cardiac silhouette. Echocardiogram is the definitive study to assess the size of the effusion and the extent of hemodynamic compromise. Patients with known effusions need to be carefully monitored for symptoms of tamponade. Smaller effusions may be managed with diuresis, afterload reduction, and anti-inflammatory agents such as ASA or Motrin. Significant effusions may need to be drained with a flexible pigtail catheter. Percardiocentesis can be done under echo guidance in an intensive care unit setting with adequate sedation and monitoring or with fluoroscopy in the cardiac catheterization laboratory. Severe recurrent pericardial effusions may require surgical creation of a pericardial window to drain the pericardial space into the mediastinum.

Hepatomegaly, ascites, and peripheral edema are further evidence of systemic venous congestion and are generally seen in patients with significant effusions, ventricular dysfunction, or atrioventricular (AV) valve regurgitation. Management in-
cludes improving cardiac output with diuresis and afterload reduction. Comfort measures, such as elevating the head of the bed and loose-fitting clothing, are helpful. These patients are at increased risk for skin breakdown. Careful assessment and preventive care, such as the use of pressure reducing mattresses, position changes, and judicious use of tape, are important.

Respiratory issues. Increases in pulmonary vascular resistance after surgery are particularly dangerous in patients following the Fontan procedure because they are dependent on passive, non-pulsatile blood flow into the pulmonary circulation. Elevated PVR increases systemic venous pressures and decreases cardiac output. In the postoperative period, positive and expiratory pressure is avoided, and patients are extubated within 12 hours if possible. Following extubation, careful assessment of respiratory status includes respiratory effort, breathe sounds, and \( \text{O}_2 \) saturations. Children with fenestrations are expected to have oxygen saturations between 80% and 85% in the early postoperative period. Patients without fenestrations should be fully saturated. Preventing atelectasis with deep breathing exercises, ambulation, frequent position changes, and chest physiotherapy is important. Pulmonary complications (such as pleural effusions, pneumothorax, pneumonia or lobar collapse) increase pulmonary vascular resistance and may impair cardiac function. Supplemental oxygen may be needed if there is pulmonary compromise.

Thromboembolic complications. Thromboembolic complications can occur both early and late after the Fontan procedure. Sluggish blood flow through the Fontan pathway and extensive atrial suture lines and foreign material may predispose these patients to thromboemboli. Significant morbidity and mortality can occur from occlusion of the Fontan pathway, pulmonary embolism, and stroke. The incidence of thromboembolic events varies in the literature, ranging from 1% to 19% (Monagle, et al., 1998). The management of significant thromboembolic events may include surgical embolectomy, thrombolytic therapy, or anticoagulation. The outcome of aggressive treatment is not clear. Monagle et al. (1998), in a review of the available research studies, identified complete resolution of thrombosis in only half the patients, and subsequent death in 25% of the patients. Because of the poor outcomes from thromboembolic events, most programs use a regimen of prophylactic anticoagulation with either Coumadin (DuPont Pharmaceuticals Company, Wilmington, DE), aspirin, or other antiplatelet agents. No prospective randomized trials have been conducted to determine the efficacy of prophylactic anticoagulation agents or antiplatelet agents in preventing thromboembolic events or necessary length of treatment.

Nursing care. Effective nursing care to provide psychosocial support to younger children and their parents following the Fontan procedure is vitally important to both a successful medical and emotional outcome. Care of the family as a unit must be the focus of nursing interventions during a sometimes prolonged hospital stay, as parents are the main source of emotional support for their children.

After surgery, parents are relieved but anxious and continue to fear death or a life-threatening event. They become tired, depressed, and frustrated as they attempt to balance outside family and work demands. Many express helplessness at seeing their child undergo multiple painful procedures. Frequent explanations and realistic reassurance about their child’s progress and eventual recovery are important to maintain hope and optimism. Parents need to regain their parental role by participating in activities of daily living with their child, having meaningful choices in care when possible and being a partner with the health care team in decision-making. Serious illness and prolonged hospitalization can exacerbate dysfunctional family dynamics. Early assessment and referrals to social work, psychiatry, or outside agencies should be facilitated.

Toddlers and preschoolers are emotionally vulnerable when hospitalized because of normal developmental concerns about separation from parents, mutilation, and punishment. They have many fears and fantasies and have limited verbal skills and intellectual ability to understand events. Following surgery, they are in an unfamiliar environment and are anxious, frightened and irritable. Pain, disruption in sleep cycles and normal rituals, withdrawal from anesthesia and narcotics, multiple monitoring lines, and periodic invasive procedures compound their discomfort. During a prolonged hospital stay, they show more signs of emotional upset such as panic, temper tantrums, depression, and withdrawal.

Consistent nursing care fosters a sense of familiarity, security, and trust in a strange environment. A calm, honest approach works well, and the child’s preferences should be incorporated in his plan of care when possible. Planning a daily schedule with the child and parent provides structure and control over daily events. Painful procedures
should be avoided in a child’s room, but rather performed in a treatment room. Adequate premedication for chest tube insertions and removals should be given. A sedative such as Versed (Roche Laboratories, Inc., Nutley, NJ) in combination with an analgesic works well. Whenever possible, children need days without tests or procedures; this can be accomplished by grouping necessary tests and treatments together. Children use play to cope with difficult situations. Child life specialists can provide frequent opportunities for hospital play, drawing, fantasy play, and other diversional activities to help children master new experiences. Enhancing a child’s ability to cope with the difficult experience of hospitalization and providing for his emotional well-being may be the most important nursing goal.

**Long-term Outcomes and Issues**

Results of the Fontan operation have steadily improved in the last three decades. Five hundred patients with SVA operated on at Boston Children’s Hospital from 1973 to 1991 showed a steady decline in Fontan failure during the time period, from 27% in the first quarter to 7% in the last quarter (Gentiles, et al., 1997). Functional outcomes were excellent, with 91% of patients free from cardiac symptoms or mildly symptomatic with exercise. Recent outcomes from 1990 to present for Fontan procedures using a total cavopulmonary connection have early mortality rates of less than 5% (VanArsdell, et al., 2000).

Even though there is excellent operative survival, the Fontan remains a palliative procedure for patients with SVA. Despite relief of cyanosis, reduction of the volume load on the ventricle, and division of the circulation, there remains only a single ventricular pumping chamber. A unique physiology is created with chronic systemic venous hypertension and chronic pulmonary artery hypotension (deLeval, 1998). The Fontan ventricle is chronically underloaded because of diminished pulmonary venous return, and this may contribute to diastolic dysfunction. Chronic systemic venous hypertension increases vascular resistance and, over time, is likely to contribute to ventricular dysfunction (Tweddell, et al., 1999). The ultimate fate of the single ventricle in the Fontan is unknown. Long-term problems are discussed in the following sections.

**Exercise.** Following the Fontan operation, patients have an abnormal cardiorespiratory response to exercise. They have a decreased aerobic exercise capacity that decreases further with age and a lower-than-normal anaerobic threshold. Multiple factors contribute to the exercise limits experienced by these patients. They have a blunted heart rate response, so their heart rate increases more slowly with exercise and they reach lower maximal heart rates. Their ability to increase stroke volume with exercise is limited and may be related to impaired ventricular function or residual Fontan obstruction. They also experience mild systemic desaturation with exercise, with O₂ saturations of approximately 90%. Those with an open fenestration have more pronounced desaturation with exercise. Children should be encouraged to be physically active and allowed to set their own exercise limits. Parents, teachers, and other adults should be aware of the child’s exercise limitations, especially with endurance sports, and provide alternate activities and adequate rest periods.

**Arrhythmias.** Atrial flutter is the most common arrhythmia seen post-Fontan, and the incidence increases with time after the Fontan procedure (Fishberger, et al., 1997). The multiple atrial suture lines near the sinus node and its blood supply, atrial enlargement, and elevated atrial pressures may contribute to the increased incidence of atrial arrhythmias. Atrial flutter is poorly tolerated in the Fontan circulation and can be fatal. Sinus node dysfunction is also common and is associated with a higher incidence of atrial flutter (Fishberger, et al., 1997). Multiple treatments, including antiarrhythmic medications, radio-frequency ablation, pacemaker placement, and surgical ablation, are used in these patients.

A small group of patients with previous atrio-pulmonary anastomosis-type Fontan procedures develop worsening functional status after many years because of atrial arrhythmias refractory to therapy, obstruction of the Fontan pathway resulting in chronic effusions, and worsening ventricular function. Some of these patients have undergone conversion to a total cavopulmonary anastomosis Fontan (either lateral tunnel or extra cardiac conduit) with a low mortality and improvement in their clinical status (Kreutzer, et al., 1996; McElhinney, Reddy, Moore, & Hanley, 1996; Marcelletti, et al., 2000). Mavroudis, Backer, Deal, and Johnstrude (1998) have added additional arrhythmia circuit cryo-ablation and the placement of a prophylactic antitachycardia pacemaker to their Fontan conversion operations to achieve more effective arrhythmia control in patients with severe, refractory arrhythmias. Heart transplantation is another option for patients with a failing Fontan circulation.
Protein-losing enteropathy. Protein-losing enteropathy (PLE) is a poorly understood late complication of the Fontan procedure that entails loss of protein through the bowel wall as evidenced by elevated stool alpha1 antitrypsin levels, hypoalbuminemia, and hypoproteinemia. Edema, ascites, pleural effusion, and chronic diarrhea may also be seen. Chronic malnutrition may occur in severe cases. The pathophysiology of PLE is unclear; the common hypothesis is that elevated systemic venous pressure causes disturbed lymph production and drainage in the intestinal bed, leading to bowel wall edema and loss of protein and lymphocytes. The severity of symptoms varies widely and may be transient in some patients. The incidence has been reported as between 2.5% and 13% (Feldt, et al., 1996; Gentiles, et al., 1997; Mertens, Hagler, Saver, Somerville, & Gewillig, 1998) and increases with longer duration of survival. (Tweddell, et al., 1999). The prognosis is poor with about 50% of patients with identified PLE eventually dying. (Feldt, et al., 1996; Mertens, et al., 1998).

Multiple treatment approaches have been tried with variable success. Medical management usually includes diuretics, afterload reduction, and inotropic support in an attempt to optimize hemodynamics. Chronic albumin infusions and dietary adjustments (high protein, low fat) may be needed to combat chronic malnutrition. Sterioids (Rychick, Piccoli, & Barber, 1991; Mertens, et al., 1998) and heparin therapy (Donnelly, Rosenthal, Castle, & Holmes, 1997) have also been used with success in some patients and with no effect in others. Interventional catheterization techniques such as balloon dilation and stenting of pulmonary artery stenosis and, particularly, creating or enlarging the fenestration have been beneficial in some patients. Surgical approaches such as creating a fenestration, converting to a newer Fontan pathway, and transplantation have also been tried with some success.

Developmental outcomes. Several recent studies have evaluated neurodevelopmental outcomes following the Fontan procedure. Multiple factors place this patient group at risk for neurologic and developmental problems including chronic CHF and cyanosis, periods of acidosis or ischemia, co-existing neurologic deficits, failure to thrive, multiple surgical procedures with cardiopulmonary bypass and periods of deep hypothermic circulatory arrest, thromboembolic events, and other factors. Uzark and colleagues (1998) administered neurodevelopmental tests to 32 post-Fontan patients ages 26 months to 16 years (mean age 5.3 years). They found that intellectual development is essentially within the normal range, although visual motor deficits may be more prevalent. Wernovsky and colleagues (2000) studied 133 children with a median age of 11 years and median time since Fontan of 6 years. They reached a conclusion similar to Uzark’s that the majority of individual patients had cognitive outcomes and academic function within the normal range but the performance as a whole was lower than the general population.

Children with an initial diagnosis of HLHS appear to have more neurodevelopmental deficits than the Fontan group as a whole. Uzark, et al. (1998), Wernovsky, et al. (2000), and Goldberg, et al. (2000) identified those with HLHS who had deep hypothermic circulatory arrest with their initial surgical procedure as having worse outcomes. Mahle, et al. (2000) recently reviewed outcomes in the school-aged survivors of staged management for HLHS and found that the mean performance of the group was lower than the general population, although most individual patients were in the normal range. Mental retardation (IQ less than 70) was identified in 18%.

CONCLUSION

Children with single ventricle anatomy are among the most complicated and challenging patients encountered in pediatric cardiology. Current management involves staged surgical procedures in the early years, culminating in the Fontan procedure. The outlook for these children has greatly improved in the last 30 years since Fontan first described this surgical approach for single ventricle anatomy. The operative survival for all surgical repairs has improved, and the functional status of these children is very good, with most growing and developing normally. The Fontan procedure, despite separating the circulation, remains a palliative procedure with many long-term concerns. The ultimate fate of the single ventricle is unknown. Nurses have an important role in the care of these patients and their families throughout infancy and childhood and into adulthood.

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