Surgical management of the single ventricle

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Abstract

The vast majority of neonates with single ventricle physiology require some form of surgical intervention to realize long term survival. Surgical options are limited as septation of the single ventricle has been described but is an option available for only a small subset of patients with ideal anatomy. Orthotopic cardiac transplant is another approach which because of limited organ availability eliminates many potential candidates by attrition on the waiting list. The most common approach to palliation in the infant with single ventricle is to intervene surgically in a manner that ultimately culminates in an effective, successful Fontan. This typically requires a staged approach of successive operations which optimally preserve pulmonary vasculature and ventricular function while providing a milieu of adequate oxygenation to allow for normal growth and development of the infant.

Keywords: Single ventricle; Palliative surgery; Congenital heart defects; Hypoplastic left heart syndrome; Pediatric cardiac surgery

1. Introduction

There are a number of congenital heart defects in which there is only a single functional ventricle. This group includes tricuspid atresia, double-inlet ventricles and other forms of single left and single right ventricles. In the vast majority of patients with hypoplastic left heart syndrome, there is a single functional right ventricle. Some patients have two ventricles but possess anatomic features that may preclude a successful biventricular cardiac repair. An example of a ‘single ventricle equivalent’ is the patient with double-outlet right ventricle and a ventricular septal defect remote from either semilunar valve. Another example is the patient with double-outlet right ventricle and ‘criss-cross’ atrioventricular valves, preventing either ventriculoseptal defect (VSD) closure or intraventricular baffle creation. These patients, whether they physically or functionally possess a single ventricle, are triaged to a surgical treatment strategy that culminates in the Fontan circulation, wherein all systemic venous return flows directly into the pulmonary arterial circulation without the assistance of a ‘pulmonary’ ventricle and without mixing with pulmonary venous return. The Fontan circulation thereby allows these patients to have a normal systemic arterial saturation. Regardless of the precise anatomic diagnosis, therefore, the goals of the surgical treatment of all single ventricle patients are the same, that is, the successful achievement of the Fontan circulation with minimal morbidity, and optimal long-term cardiovascular performance.

2. Background

After several decades of experimental work into total right heart bypass by a number of investigators, Fontan reported his first three successful repairs of tricuspid atresia in 1971. These original ‘Fontan’ operations only conceptually resemble current modifications of this circulation that will be described below. Blood flow to the right pulmonary artery was provided by a classical Glenn shunt, and homograft valves were implanted at the entrance of the IVC into the right atrium and at the anastomosis of the right atrium to the pulmonary artery (Fig. 1).
The Fontan operation as originally described by F. Fontan for tricuspid atresia was comprised of five steps: (1) Glenn shunt to the right lung; (2) harvest of the main pulmonary artery from the right ventricular outflow tract; (3) anastomosis of main pulmonary artery to the right atrium with an interposed homograft valve; (4) placement of a homograft valve in the inferior vena cava-right atrium junction; and (5) closure of atrial septal defect. (Reprinted with permission from Fontan F, Baudet E, Thorax 1971;26:240.)

A few years later, Kreutzer modified this to a direct atriopulmonary anastomosis in consort with closure of the atrial septal defect [6]. Over the ensuing decade, atriopulmonary anastomoses were used to palliate tricuspid atresia and gradually became the approach of choice for most single ventricle variants.

Total cavopulmonary connection (TCPC) as an alternative to either the classic or modified atriopulmonary Fontan was popularized in the late 1980s with the growing recognition that right atrial contraction did not enhance pulmonary blood flow and that leaving the entire right atrium in the systemic venous circulation could contribute to some of the morbidity of the Fontan operation [7]. The long-term consequences of atriopulmonary connection had become evident in longer-term survivors, with right atrial dilation, atrial arrhythmias, atrial thromboembolism and compression of the pulmonary veins by the giant atrium. Thus there was further impetus to find alternative cavopulmonary connections. The lateral tunnel Fontan was first reported in clinical practice in the late 1980s (Fig. 2) [8,9].

Indeed, the success of the TCPC performed as a ‘lateral tunnel’ in the systemic venous atrium, demonstrated convincingly that the contribution of a pulsatile right atrium in the Fontan circuit was not necessary. Strongly suggestive evidence that this would be successful was provided by Kawashima who demonstrated that all systemic venous return (with the exception of hepatic venous blood) could be directed to the pulmonary arteries in patients with an interrupted IVC and azygous continuation without hemodynamic consequence [10]. In addition, this modification dramatically improved outcomes for single ventricle patients with stenosis or atresia of the left-sided atrioventricular valve. For these patients, the lateral tunnel allows for pulmonary venous blood to cross over freely to the right-sided atrioventricular valve, without interfering with the systemic venous pathway [11].

Two additional advancements in the surgical approach to the Fontan circulation occurred soon after the introduction of the lateral tunnel modification. First, the concept of adding a fenestration in the lateral tunnel was introduced [12]. The rationale for lateral tunnel fenestration was to provide a limited degree of right-to-left shunting after the Fontan. The fenestration was believed to allow for an increase in cardiac output at the expense of a tolerable degree of desaturation [13,14]. This fenestration has further improved early outcomes after the Fontan operation, a fact that has recently been demonstrated in a prospective randomized trial [15]. The fenestration has been seen to close spontaneously in some patients but it has been customary to perform a transcatheter device closure at some intermediate-term point in those patients with patent fenestrations. Delayed closure of the fenestration has been associated with acceptable hemodynamics and reliable improvement in the patients’ systemic saturation [16].

The second therapeutic advance introduced soon after the introduction of the lateral tunnel modification was the increasing use of the bidirectional cavopulmonary anastomosis (BDCPA) as an intermediate palliative step prior to the Fontan operation (Fig. 3) [17]. This procedure, also performed as a ‘hemi-Fontan’ operation in some centers, allows for direct flow of systemic venous blood from the superior vena cava to confluent pulmonary arteries (Fig. 4). It has been proposed that this intermediate arrangement allows for a more gradual transition toward the ventricular loading characteristics peculiar to the Fontan circulation, specifically, the necessity of a single ventricle to pump a cardiac output through the systemic and pulmonary resistance beds in series [18]. Meeting these demands acutely can be difficult for a number of reasons. In patients living with an aortopulmonary shunt, the ventricle is abnormally volume overloaded. The rapid removal of this volume load, with subsequent resistance loading, may result in dramatic changes in ventricular geometry (expressed as volume-to-mass ratio) [19]. In patients with a pulmonary band, or equivalent native pulmonary stenosis, the chronic imposition of an abnormally high afterload on the single ventricle can have deleterious effects on ventricular compliance. Ventricular compliance is an important determinant of pulmonary vascular resistance for these patients. Removal of the pulmonary artery

Fig. 1. The Fontan operation as originally described by F. Fontan for tricuspid atresia was comprised of five steps: (1) Glenn shunt to the right lung; (2) harvest of the main pulmonary artery from the right ventricular outflow tract; (3) anastomosis of main pulmonary artery to the right atrium with an interposed homograft valve; (4) placement of a homograft valve in the inferior vena cava-right atrium junction; and (5) closure of atrial septal defect. (Reprinted with permission from Fontan F, Baudet E, Thorax 1971;26:240.)
Fig. 2. The lateral tunnel total cavopulmonary connection is depicted after division of the main pulmonary artery (PA) and bidirectional cavopulmonary connection at a previous operation. (a) A lateral atriotomy is performed. The superior vena cava (SVC) stump is anastomosed to the inferior surface of the right pulmonary artery (RPA). (b) A conduit is fashioned from a synthetic tube graft and then secured around the inferior vena cava and superior vena cava orifices (IVC, SVC) skirting the widely opened atrial septal defect (ASD). Approximately 50% of the circumference of the lateral tunnel is the posterolateral wall of the right atrium. Ao, Aorta. (Reprinted with permission from Nichols DG, Cameron DE, Greeley WJ, et al. (eds.), Critical heart disease in infants and children, Mosby, St. Louis, MO, 1995;754.)

Regardless of the relative contributions of these theoretical considerations, the bidirectional cavopulmonary anastomosis is thought to have played a major role in improving the outcomes for historically high risk Fontan subgroups, such as those patients with hypoplastic left heart syndrome [20]. On a more practical note, the 3.5 mm modified Blalock–Taussig shunt has emerged in our experience as preferable to the 4mm diameter in neonates requiring an aortopulmonary shunt. This smaller shunt, however, usually means that a number of these patients become prohibitively cyanotic at an age (4–8 months) when most surgeons would be unenthusiastic about performing the Fontan operation, even in the era of fenestration.
Fig. 3. After Stage I palliation a bidirectional cavopulmonary connection is created. (a) The Blalock–Taussig shunt and azygous vein are ligated and divided after going on cardiopulmonary bypass. The superior vena cava is divided and the cardiac stump oversewn. A right pulmonary arteriotomy is performed to match the width of the superior vena cava. (b) The superior vena cava-to-right pulmonary artery (bidirectional cavopulmonary) anastomosis is performed with a running absorbable suture. (Reprinted with permission from Castaneda AR, Jonas RA, Mayer JE Jr, Hanley FL (eds.), Cardiac surgery of the neonate and infant, Philadelphia: W.B. Saunders, 1994;262.)

Finally, the use of extracardiac conduits to carry the inferior vena caval blood to the pulmonary arteries has been introduced in the past decade [21]. The extracardiac Fontan modification has become increasingly popular as it is relatively easier to perform than the lateral tunnel operation (Fig. 5). Some surgeons have even performed extracardiac Fontans without the employment of cardiopulmonary bypass [22]. The longer-term benefits of this procedure when compared to the lateral tunnel modification are, however, yet to be defined [23–25].

3. Anatomy

There are many anatomic lesions that are ultimately similar in that they provide only one functional ventricle. These ‘single ventricle equivalents’ may in fact have two reasonably well developed ventricles but because of intracardiac pathology (such as VSD location, AV valve orientation and chordal attachments) a two-ventricle repair is precluded. They are therefore preferentially managed using a single ventricle surgical algorithm. Hereafter, all patients with either true anatomic single ventricles or functional single ventricles (two ventricles but because of the anatomy can only be offered single ventricle surgical palliation) will be referred to as ‘single ventricle’. Dr Weinberg’s chapter covers in detail the anatomic single ventricle variants.

4. Management

Management of single ventricle patients requires coordinated medical and surgical efforts throughout their course. In all but a few instances, these patients are managed in three stages. While the neonatal operations (Stage I) are variable depending on the nature of pulmonary and systemic blood flow, the second and third ‘Stage’ operations are more stereotyped. The following is a discussion of the three stages of management followed by some of the unique management concerns posed in these patients.

4.1. Stage I Management

The vast majority of single ventricle patients are diagnosed either in utero or in the neonatal period. Likewise, the majority of these patients will require surgical intervention early in life. Infants with a single ventricle will manifest one of four general physiologic patterns in the neonatal period depending on the underlying anatomy. Typically, infants will present with a combination of insufficient or excessive pulmonary blood flow with or without obstruction to systemic outflow. There are four major physiologic presentations of neonates and infants with single ventricles and each
Fig. 4. Hemi-Fontan technique: (a) The pulmonary arteries are opened widely to address any focal stenoses. The superior vena cava (SVC)-right atrial junction is opened longitudinally. (b) The back wall of the SVC-RA junction is approximated and the front wall augmented with a large patch of homograft or synthetic material. A dam is created between the SVC and right atrium with the baffle material to separate SVC and IVC venous blood. (Reprinted with permission from Karl TR, Semin Thorac Cardiovasc Surg 2001;64–65.

presentation is associated with a unique group of pathologies (Table 1) [26].

Whatever the physiologic presentation, the goals of the first palliative operation in a single ventricle neonate are to address the state of balance between pulmonary and systemic blood flow. Achieving this balance requires that the patient have a stable (non-ductal) source of pulmonary blood flow. It also requires that the pulmonary vascular bed be protected from high pressures and flows that could result in elevated pulmonary vascular resistance, ultimately precluding or reducing the likelihood of achieving a successful Fontan circulation. In addition to providing for stable and controlled pulmonary blood flow, any significant obstruction to systemic
outflow or pulmonary venous return must be addressed at the first palliative stage.

4.1.1. Inadequate pulmonary circulation (or ductal-dependent)

In the interval between birth and their first surgical intervention, patients who have been diagnosed antenatally or those presenting with cyanosis are stabilized with a prostaglandin E1 intravenous infusion to maintain ductal patency [27]. Ductal patency can be reestablished in the vast majority of patients under the age of 7 days within minutes to hours of initiating intravenous prostaglandin E1 therapy. In rare patients in whom it is impossible to reestablish ductal patency using a prostaglandin infusion, invasive transcatheter ductal dilation and stenting has been described, but these techniques are associated with variable success, significant risk of morbidity and mortality and are not techniques employed routinely in our institution [28]. Emergent shunting can be performed but if the patient progresses to extremis, extracorporeal membrane oxygenation (ECMO) should be considered.

Ductal patency can be maintained for prolonged periods with intravenous prostaglandin E1 allowing patients to be fully resuscitated and stabilized prior to operative intervention. It is not possible to precisely gauge the amount of pulmonary blood flow that will result when the duct is reopened. Balancing pulmonary and systemic blood flow ratios often requires manipulation of systemic and/or pulmonary vascular resistances in the intensive care unit [29].

Patients who present with either inadequate or ductal dependent pulmonary circulation (Table 1, No. 1) or with excessive pulmonary circulation with systemic outflow obstruction (Table 1, No. 3) will be managed as described above with a prostaglandin E1 infusion, metabolic and hemodynamic stabilization, followed by an appropriate operative intervention that will entail pulmonary artery banding or a pulmonary artery-to-aorta anastomosis (Damus–Kaye–Stansel, DKS) with an aortopulmonary shunt, respectively. In patients with intracardiac or extracardiac obstruction to pulmonary blood flow, ductal patency assures adequate pulmonary blood flow. In patients with systemic outflow obstruction, ductal

### Table 1
Clinical presentations of single ventricle patients

<table>
<thead>
<tr>
<th>Clinical presentation</th>
<th>Anatomic example</th>
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<tbody>
<tr>
<td>1. Inadequate pulmonary circulation (or ductal-dependent)</td>
<td>–[S,D,S] Tricuspid atresia with pulmonary stenosis (IB)</td>
</tr>
<tr>
<td>2. Excessive pulmonary circulation without systemic outflow obstruction</td>
<td>–Single right or left ventricle with pulmonary atresia</td>
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<tr>
<td></td>
<td>–[S,D,D] Tricuspid atresia, with transposition of the</td>
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<tr>
<td></td>
<td>great arteries and unrestricted pulmonary blood flow (IIC)</td>
</tr>
<tr>
<td>3. Excessive pulmonary circulation with systemic outflow obstruction</td>
<td>–Hypoplastic left heart syndrome (MS/AS; MA/AS; MS/AA; MA/AA)</td>
</tr>
<tr>
<td></td>
<td>–[S,D,D] tricuspid atresia with restrictive VSD</td>
</tr>
<tr>
<td></td>
<td>–[S,L,L] Double outlet left ventricle with restrictive VSD</td>
</tr>
<tr>
<td></td>
<td>–[S,D,S] Tricuspid atresia with pulmonary stenosis (IB)</td>
</tr>
<tr>
<td>4. Balanced pulmonary blood flow</td>
<td>–[S,D,D] DORV uncommitted VSD with pulmonary stenosis</td>
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AA, atrial atresia; AS, atrial stenosis; DORV, double outlet right ventricle; MA, mitral atresia; MS, mitral stenosis; S,D,S solitus/dextrorotated/dextroposed, expression of segmental anatomy whereby visero-atrial situs is solitus (usual); ventricular looping is D-loop (right-handed); ventriculo-arterial situs is dextroposed (aorta is anterior and rightward of the pulmonary artery); S,D,S solitus dextro solitus, expression of normal segmental anatomy with solitus visero-atrial situs, D-looped ventricles (right-handed, or usual), situs solitus ventriculo-arterial alignment; VSD, ventriculo-septal defect.
patency allows for adequate systemic blood flow prior to operative intervention.

For patients presenting in the neonatal period with inadequate or ductal dependent pulmonary blood flow, the palliative intervention of choice is the aortopulmonary shunt of which there are central and peripheral types.

4.1.1.1. Central shunts. Central shunts performed without cardiopulmonary bypass have been used in the past but have largely been abandoned in favor of peripheral shunts. Central shunts are described here for historic completeness only. The Waterston shunt is a direct anastomosis between the ascending aorta and right pulmonary artery, performed via a right thoracotomy [30]. The Potts shunt is a descending aorta to left pulmonary artery anastomosis performed via a left thoracotomy [31]. Because of difficulties in controlling the amount of pulmonary blood flow through these shunts, the frequent distortion of the central pulmonary arteries, and the difficulty encountered when taking down these shunts, central shunts have been abandoned in favor of the more reproducible results of the modified Blalock–Taussig shunt.

4.1.1.2. Peripheral shunts. The classic Blalock–Taussig (BT) shunt was first described in 1976 [32]. The classic BT shunt consisted of an end-to-side anastomosis of the subclavian artery to the pulmonary artery on the side opposite the aortic arch. The classic BT shunt usually grows proportional to the somatic growth of a child and, in an era when a shunt was the only palliation available to cyanotic patients, growth of the shunt was a desirable feature. On occasion, though, these classic BT shunts have become dilated and oversized, resulting in excessive pulmonary blood flow and pulmonary vascular obstructive disease. A major weakness of the classic BT shunt is the irreversible harvest of the subclavian artery. Subclavian artery harvest has been associated in some cases with compromised development of the ipsilateral upper extremity.

The use of a synthetic conduit to create an aortopulmonary shunt was first described in 1976 [33]. This procedure, now known as a modified Blalock–Taussig shunt, is performed by interposing a tubular synthetic conduit between the innominate or subclavian artery and the pulmonary artery. One of the principal advantages of the modified BT shunt over the classic BT shunt is precise control of the diameter and length of the conduit. Diameter is the primary determinant of flow in a tube as described by Poiselle’s law which describes flow in a conduit as proportional to the fourth power of the radius, expressed by the formula $Q = \Delta P \pi r^4 / 8 \eta l$, where $Q$ is flow, $\Delta P$ is change in pressure between inflow and outflow of the tube, $\eta$ is viscosity of the fluid and $l$ is the length of the tube.

The ability to choose the length, diameter and proximal insertion site of the interposition conduit allows accurate regulation of the amount of flow through the shunt and, secondarily, the pressure in the pulmonary artery. Additional advantages of the modified BT shunt include the sparing of the subclavian artery, high early patency rates, and reduced risk of pulmonary artery distortion. The material commonly used to construct modified BT shunts is expanded polytetrafluoroethylene (ePTFE).

Assuming complete atrial mixing, an ideal early post-shunt saturation (80–85%) is followed by mild congestive heart failure (during the next several weeks as pulmonary vascular resistance falls) and then the gradual outgrowth of the shunt with diminishing saturations until the next stage of palliation. The shunt diameter chosen depends on: (1) the size of the patient; (2) the estimated or known pulmonary vascular resistance; (3) the anticipated time interval between the first and second operative stages (that is, how much patient somatic growth will occur). Currently we use 3.5 mm ePTFE grafts in neonates requiring aorto-pulmonary shunting (weight range 2.0–4.0 kg). Occasionally for a larger infant a 4.0 mm graft is used. The modified Blalock–Taussig shunt may be performed via thoractomy or sternotomy. We favor a median sternotomy approach. The advantages of a sternotomy approach to the BT shunt are:

1. Sternotomy allows for central placement of the BT shunt on the branch pulmonary artery. It is then easily approached at subsequent surgeries and is less likely to distort distal lobar branch pulmonary arteries.
2. Cardiopulmonary bypass can be instituted readily if necessary.
3. The concern about an initial sternotomy causing subsequent reoperative sternotomy with mediastinal adhesions has not been a source of increased morbidity.

4.1.1.3. Technique: modified Blalock–Taussig shunt. The aorta, brachiocephalic vessels and branch pulmonary artery are mobilized thoroughly on the side on which the shunt is to be constructed. For technical reasons, shunt construction is usually opposite the side of the aortic arch. If the patient’s oxygenation is tenuous or is hemodynamically unstable we place the patient on cardiopulmonary bypass. The vast majority of modified BT shunts performed at TCH (excluding the Stage I patients) are performed without cardiopulmonary bypass. A vascular clamp is applied to the distal innominate artery/proximal subclavian artery and an arteriotomy of equal diameter to the shunt is created. An end-to-side anastomosis is created between the graft and the systemic artery. An arteriotomy is created in the branch pulmonary artery and the distal anastomosis between graft and pulmonary artery is performed.
4.1.2. Excessive pulmonary circulation without systemic outflow obstruction

The goals of neonatal intervention in single ventricle patients with excessive pulmonary blood flow and no concomitant systemic outflow obstruction are two-fold. First, one seeks to reduce pulmonary artery pressure, which over time, has a deleterious effect on the developing pulmonary vasculature leading to pulmonary vascular obstructive disease. Second, one attempts to reduce the pulmonary shunt ratio ($Qp:Qs$). A large $Qp:Qs$ imparts a significant volume load on the single ventricle, which may have deleterious effects on ventricular performance and/or atrioventricular valve function [34]. Pulmonary artery banding is the technique traditionally used to address excessive pulmonary blood flow. To provide controlled pulmonary blood flow, though, a pulmonary-aortic anastomosis with a modified Blalock–Taussig shunt is increasingly used in this group of neonates.

Pulmonary artery banding, like all non-definitive palliations, has pros and cons. The benefits, as mentioned above, are the control of volume load to pulmonary circulation with a ‘lesser’ operation than, for instance, a DKS and a modified BT shunt (Fig. 6). The disadvantages of a pulmonary artery band are numerous. The band can be locally destructive to the pulmonary valve or, more importantly to the single ventricle patient, it may distort the branch pulmonary arteries. Pulmonary artery bands, while thought of as simple operations are actually technically difficult in that it is quite difficult to achieve a band diameter that ideally regulates pulmonary blood flow. Ventricular hypertrophy that results from pulmonary artery banding increases the risk of subsequent subaortic obstruction in a circulation pathway in which the aortic blood flow must pass from the systemic ventricle through a ventricular septal defect (also known as the bulboventricular foramen, BVF). Ventricular hypertrophy also contributes to an increased mass-to-volume ratio at the time of BDCPA and elevated end diastolic pressure at the time of TCPC, which may predispose to eventual Fontan failure.

Out current indications for pulmonary artery banding are limited. We would preferentially treat a single ventricle patient with a pulmonary artery band (rather than a Norwood-type pulmonary artery-to-aorta anastomosis) only if the following conditions exist:

1. Excessive pulmonary blood flow.
2. Normal aortic arch without evidence of coarctation or aortic arch hypoplasia.
3. A sufficiently large ventricular septal defect (BVF) that it is unlikely to result in subaortic obstruction (typically a BVF diameter indexed to body surface area $> 2.0 \text{cm}^2/\text{m}^2$) [35]. The patients with, or at risk, of developing subaortic stenosis will be discussed in more detail in the next section.

An alternative to pulmonary artery banding is pulmonary artery division and modified BT shunt. This operation has recently been validated in a prospective trial in which patients who were candidates for pulmonary artery banding underwent instead pulmonary artery division with a modified BT shunt. With a series of 19 patients, there were no operative deaths and 14 patients have gone on to uncomplicated BDCPA and five have had successful TCPC [36]. Pulmonary artery division with modified BT shunt can clearly be performed with low mortality and yields pulmonary physiology that is favorable for later BDCPA and TCPC.

4.1.2.1. Technique: pulmonary artery banding. Pulmonary artery banding can be performed via a thoracotomy or a median sternotomy. We prefer the median sternotomy approach for pulmonary artery banding for many of the same reasons as outlined for the modified BT shunt. Most importantly, the band can be carefully positioned on the main pulmonary artery so as not to impinge on the origin of either branch pulmonary artery. The determination of the proper circumference of the pulmonary artery band is a combination of calculated and clinically observed data. The initial band circumference is calculated based on retrospective observations of Trussler et al. who determined that the ideal band circumference was 20 mm + patient’s weight (kg) in mm (so in a 3 kg infant, the starting band diameter would be 20 mm + 3 mm = 23 mm) [37].

After conventional median sternotomy, a minimal upper pericardial window is opened. A limited segment of the main pulmonary artery is dissected free for encirclement by the band, to prevent proximal or distal migration of the band. A silastic band is cut to 3 mm encirclement by the band, to prevent proximal or distal impingement of the main pulmonary artery is dissected free for upper pericardial window is opened. A limited segment circumference was 20 mm of Trussler et al. who determined that the ideal band encirclement is calculated based on retrospective observations and clinically observed data. The initial band circumference was 20 mm of pulmonary artery banding can be performed via a thoracotomy. Pulmonary artery division and modified BT shunt can act as a nidus for thrombus formation. The defect in the pulmonary confluence is closed either primarily, or with a pericardial or homograft patch.

While some advocate pulmonary artery banding preferentially, the operation is not without significant risk. Current mortality rates for isolated pulmonary artery banding have remained persistently high, with a recent report indicating a 30-day mortality risk of 13.7% in patients palliated with a pulmonary artery band in the past decade [38]. It is evident then that pulmonary artery banding is not an insignificant procedure and carries with it a substantial mortality and morbidity risk in infants with a single ventricle.

4.1.3. Excessive pulmonary circulation with systemic outflow obstruction

Patients with unrestricted pulmonary blood flow frequently have concomitant systemic outflow obstruction and are treated with a different algorithm than those with unobstructed systemic outflow [26]. Some single ventricle patients present with severe systemic outflow obstruction at multiple levels. Patients with hypoplastic left heart syndrome (HLHS) predominate in this group. Clearly these patients could not be managed with a PA band alone, or even a PA band with aortic arch augmentation. The necessities presented by this group spurred Norwood to pioneer a first stage palliative procedure that appropriately bears his name [39]. The Norwood Stage I palliation addresses the systemic outflow tract obstruction problem with a pulmonary artery to aorta anastomosis, and an aortic arch augmentation. An atrial septectomy is routinely performed if pulmonary venous return must traverse anything less than a very large native ASD to reach the dominant atrioventricular valve. A controlled source of pulmonary blood flow is provided by a modified Blalock–Taussig shunt (Fig. 7).

At the time of its introduction in the early 1980s the Norwood operation was a technical tour de force, with mortality rates of greater than 50%. Over the ensuing two decades, a number of centers have reported a reduction in operative mortality down to the 10–20% range [40]. As such, most centers consider this surgical strategy preferable to transplantation for these single ventricle patients [41].

While originally devised to treat patients with HLHS, the Norwood operation was rapidly adapted to other single ventricle defects with similar degrees of systemic outflow tract obstruction. Examples of this are (S,D,D)
DORV with mitral atresia, (S,D,D) tricuspid atresia and severe aortic stenosis, (S,L,L) double inlet left ventricle with severe aortic stenosis, and (S,D,S) unbalanced complete atrioventricular canal defects with severe left- or right-sided hypoplasia. The results in these patients compare favorably to patients with HLHS [42].

The Norwood operation is ideally performed in the first week or two of life. Often the HLHS diagnosis is made prenatally, or early in the postnatal period. Those not diagnosed prior to ductal closure typically present with varying degrees of cardiovascular collapse. All patients require a prostaglandin E1 infusion to maintain adequate systemic perfusion via the ductus arteriosus. These patients typically manifest some degree of pulmonary overcirculation. This can be deleterious to both the single ventricle and multiple other organ systems. The volume loading of the single ventricle can be detrimental to ventricular performance, which may increase the risk of poor ventricular function post-Norwood. Moreover, poor preoperative systemic perfusion can result in complications such as necrotizing enterocolitis, renal insufficiency and hepatic dysfunction [43]. Because of these considerations, it is our practice to move expeditiously to the operating room in the first days of life, once the patient’s anatomy has been defined echocardiographically, and they have been stabilized from any perinatal insults or injuries suffered subsequent to ductal closure.

An exception to the approach is utilized for patients who are significantly cyanotic preoperatively because of a highly restrictive atrial septal defect that prevents adequate flow from the pulmonary veins to the dominant atrioventricular valve. This serves to impair overall pulmonary blood flow, resulting in hypoxemia. These patients, in a manner similar to that seen in obstructed total anomalous pulmonary venous return, can possess a highly reactive pulmonary vascular resistance following cardiopulmonary bypass, contributing significantly to their post-Norwood morbidity [13]. We therefore favor preoperative interventional catheterization in these patients to address the restrictive atrial septum. This usually takes the form of a balloon atrial septostomy, but stenting of the atrial septal defect has been required in a subset of patients with heavily muscularized atrial septa. After this intervention, patients are allowed to stabilize for usually more than 48 hours, after which they are treated in a standard fashion [44].

4.1.3.1. Technique: Norwood operation/Stage I. The Norwood operation has been historically performed utilizing hypothermic circulatory arrest to perform the neoarch reconstruction and atrial septectomy. As the potentially deleterious effects of circulatory arrest have become increasingly recognized, and patients following the Norwood operation have been identified as being at risk for worse cognitive outcomes, efforts have been made to reduce, or eliminate the use of circulatory arrest in the Norwood operation. Circulatory arrest time can be reduced by placing the arterial cannula in the ductus arteriosus distal to the branch pulmonary arteries. This allows for the division of the main pulmonary artery, with closure of the distal central pulmonary artery.
while remaining on cardiopulmonary bypass. Additionally, the atrial septum can be rapidly excised working through the right atrial cannulation site after an interval of reperfusion following the arrest period during arch reconstruction. This saves the time required for creating, and subsequently closing, a separate atriotomy as well as providing for a period of perfusion between two episodes of arrest which has been associated with improved neurologic outcome in animal models [45,46].

Alternatively, strategies employing selective unilateral antegrade perfusion of the brain during the Norwood operation are being employed. In this approach, the arterial cannula is placed in the BT shunt graft after it is anastomosed to the innominate artery. With innominate artery occlusion, then, perfusion to the right common carotid and subclavian arteries can be maintained throughout the procedure (Fig. 8) [47,48]. While this technique is conceptually attractive, optimal selective perfusion conditions have not been defined, and the benefits of this technical modification of the Norwood operation have not been rigorously demonstrated [49].

Regardless of the perfusion strategy employed, we have found that the 3.5 mm modified BT shunt from the innominate or subclavian artery, as in non-Norwood neonates, appears to be the optimal size source of pulmonary blood flow. We have favored a homograft neoaortic arch augmentation, though some authors favor an entirely autologous repair in which the proximal descending aorta is anastomosed to the proximal transverse aortic arch, and the usually large main pulmonary artery is anastomosed to the underside of the aortic arch thus created [50,51]. While this approach has some attractive characteristics, it was trialed and abandoned at this institution in the 1980s. The particular weakness of the autologous neoaortic reconstruction is the fact that the left pulmonary artery and left mainstem bronchus, without an adequate arc to the aortic arch, are required to traverse potentially compressive region between the ascending aorta and descending aorta. We have seen significant stenoses of one or both of these structures, resulting in significant morbidity.

Stage I operative (30 day) mortality rates and the rate of attrition between Stage I and Stage II have declined significantly over the past two decades. Stage I operative mortality for hypoplastic left heart syndrome in series from the 1980s was reported between 37.5 and 58% [40,52–54]. At that time, operative mortality accounted for approximately 70% of all pre-Stage II mortality. Mortality for the Norwood operation in the current era should approach 10–20%. Achieving these results has been found to require significant input from all participants in a pediatric cardiovascular program. Results continue to improve incrementally at this and other centers. The Norwood 30-day mortality for all anatomic diagnoses at Children’s Hospital, Boston was 10.7% for the 3-year period from January 1999 to December 2002 (unpublished data). Risk factors for early mortality remain somewhat controversial, but there is agreement that earlier date of surgery is a contributor to early mortality risk [40,42,54]. A report from this institution found that HLHS patients were at greater risk for operative death compared with non-HLHS patients treated with the Norwood operation [42]. Other variables that have been identified by multivariable analysis as independent risk factors negatively affecting outcome.
include age at operation greater than 1 month, single right ventricle, synthetic tube graft used for aortic arch reconstruction, and HLHS subtypes with aortic atresia.

The interval between Stage I and Stage II has been identified as a period in which the infant remains at some considerable risk of mortality. The rate of pre-Stage II attrition in Stage I survivors is significantly lower in the current era (with most recent reports indicating a pre-Stage II attrition of 11–13%) compared with the previous decade (when reported pre-Stage II attrition rates were 24–28%) [42,52–54]. We speculate that earlier recognition and correction of reparable defects such as recurrent coarctation or AV valve regurgitation, as well as the move to earlier BDCPA in patients with significant ventricular dysfunction, may have contributed to this reduction in pre-Stage II attrition. Some of the reduction in the mortality for the pre-Stage II era is also almost certainly because of the heightened awareness of the fact that these infants are at increased risk. We communicate this fact clearly with the patient’s family and counsel them to have a low threshold to seek assessment by their cardiac care team. The cardiac care team then has a low threshold for admitting and investigating these ‘interstage’ infants for any recurrent or residual anatomic defects.

4.1.4. Excessive pulmonary blood flow and potential systemic outflow obstruction

In contrast to patients with dramatic systemic outflow obstruction, such as is seen in hypoplastic left heart syndrome, there are single ventricle patients with excessive pulmonary blood flow who demonstrate more subtle forms of systemic outflow obstruction, or who possess anatomic substrates now recognized as risk factors for future development of systemic outflow obstruction. The anatomic pattern in which outflow tract obstruction may be subtle, or merely anticipated, is an aorta arising from an underdeveloped infundibular chamber with a bulbocentric foramen (BVF) as the only source of systemic blood flow. Examples of this are (S,L,D) tricuspid atresia or (S,L,L) double-inlet left ventricle with transposition of the great arteries. Classically, these patients were palliated with pulmonary artery bands and relief of any significant aortic arch obstruction. More recently, however, these patients are increasingly triaged to a Norwood-type palliative strategy as the significant potential for later development of subaortic obstruction has been recognized after pulmonary artery banding in these patients [35,36].

Planning a first stage palliative procedure in these infants therefore requires a systematic, usually echocardiographic evaluation of the systemic outflow tract moving from the BVF to the proximal descending aorta. It is usually not possible to confirm any pressure gradient at the BVF in the neonate with an open ductus. Any resistance to flow out the aorta will merely divert more blood into the pulmonary artery. Morphologic hints that the subaortic area should be suspect are hypoplasia of the aortic annulus or aortic arch, or the presence of a coarctation. If any of these features are apparent in a neonate, we favor a Norwood operation, rather than pulmonary artery banding. The wisdom of this strategy was borne out in a retrospective analysis from Matitau et al. that examined the natural history of the BVF. Patients with aortic arch obstruction were much more likely to have a small BVF, and patients with a small BVF at presentation (< 2 cm²/m²) were more likely to develop significant obstruction at 2–5 years of follow-up [35].

This strategy requires that these patients be subjected to the risk of a Norwood operation, but they are then likely to be better prepared for subsequent palliation. It also important to note, that the early mortality from pulmonary artery banding in single ventricle patients, even in the current era, is not insignificant [38]. Patients who do not undergo a pulmonary artery-to-aortic anastomosis as a neonate, and subsequently develop subaortic obstruction have less than optimal surgical options. The performance of a pulmonary artery to aortic anastomosis after pulmonary artery banding is frequently impossible because of damage suffered by the pulmonary valve. Single ventricle to aortic conduits have been described, but clearly have limited long-term utility [55,56]. Surgical enlargement of the BVF can be performed, but this carries a significant risk of complete heart block, which is particularly deleterious for the hypertrophied single ventricle. Indeed, BVF enlargement has been found to be an ongoing independent risk factor for worse outcome following Fontan completion [13,57]. It is our strong opinion that these scenarios should be avoided, if at all possible. Daebritz and colleagues, based on retrospective data from our institution, concluded that the mortality risk of the Norwood operation was becoming incrementally lower during the 1990s [42]. These authors also determined that the Stage I mortality for non-HLHS patients was significantly lower during that time frame. It does appear, then, that for patients at risk of subaortic obstruction, a more liberal application of the Norwood operation is justified.

4.2. Stage II management: bidirectional cavopulmonary anastomosis (BDCPA)

The use of the Fontan operation as a primary or secondary procedure during the management of single ventricle was more common earlier in the surgical experience with this pathology. Increasing familiarity with the effects of long-term pulmonary artery banding or aortopulmonary shunting, and efforts to widen the scope of Fontan candidacy, led many centers to introduce an intermediate step in the partitioning of systemic venous return directly to the pulmonary arterial system.
The classic Glenn shunt, consisting of an SVC anastomosis to the divided right pulmonary artery, was a procedure that could be performed without cardiopulmonary bypass [58]. It was somewhat successfully employed as long-term palliation in some single ventricle patients beginning in the late 1950s. Problems with the classic Glenn included the creation of pulmonary artery discontinuity, and the development of arteriovenous malformations in the right lung. It was subsequently noted that a bidirectional cavopulmonary anastomosis, wherein the superior vena cava is anastomosed end-to-side to the pulmonary artery, could provide adequate pulmonary blood flow and, hence, adequate systemic arterial oxygenation in patients with relatively low pulmonary vascular resistance [59].

As part of a staged approach to single ventricle surgical management, the BDCPA was introduced initially as an interim palliation for patients who were high-risk candidates for the Fontan procedure. Patients judged to require second stage palliation with the BDCPA were those thought to be poor candidates for a Fontan procedure on the basis of systemic ventricular function, systemic atrioventricular valve function, pulmonary vascular resistance or pulmonary artery distortion. In some of these situations, by reducing the volume load to the systemic ventricle and/or by repairing the pulmonary arteries at the time of a BDCPA, there was improvement in the physiology such that a Fontan could be considered at a later date. An early review of this strategy from this institution confirmed the successful application of the BDCPA to high risk Fontan candidates [17].

Since the late 1980s, BDCPA has been increasingly employed as intermediate palliation prior to completion of a Fontan procedure. The BDCPA offers some distinct advantages to the single ventricle vis-à-vis aortopulmonary shunting or pulmonary artery banding. First, it reduces the abnormal volume loading of shunted single ventricles, or the abnormal resistance loading of banded single ventricles. Second, it eliminates any excessive pulmonary blood flow, as seen in over-shunted, or underbanded patients. This reduces the risk of developing chronically elevated pulmonary vascular resistance. Third, this intermediate operation allows the surgeon to address any pulmonary artery distortion that might exist after the patient’s initial palliative procedures. Atrioventricular valve regurgitation or aortic arch obstruction can also be addressed at the time of this second stage palliation. The BDCPA offers all of these benefits, while usually maintaining the patient’s arterial oxygen saturation in the 80–90% range.

The bidirectional cavopulmonary anastomosis is usually performed at approximately 6 months of age, although it can be performed at significantly earlier ages if there is catheterization data demonstrating undistorted pulmonary arteries and low pulmonary vascular resistance. This has been chosen as the optimal time for intervention based on the phenomenon of reduced pulmonary vascular resistance in non-neonates and the clinical experience of high mortality in neonates treated with a BDCPA [60–63]. Primary BDCPA’s have been attempted in the near neonatal period with less than optimal results [64,65]. We favor primary BDCPA only for those single ventricle patients with naturally limited pulmonary blood flow who are at least 8–10 weeks of age. The timing of the BDCPA may also be altered by a number of clinical factors, including the early post Stage I presentation of a single ventricle infant with ventricular dysfunction, atrioventricular valvar dysfunction, or significant desaturation.

Our current practice is to perform a cardiac catheterization prior to performing the BDCPA. It is important to define the pulmonary artery anatomy, and rule out any pulmonary venous obstruction. Pulmonary artery pressures should be measured, and pulmonary vascular resistance calculated. The presence of any obstruction to systemic outflow should be identified. Recurrent coarctations can be treated with balloon angioplasty at this time, or may be addressed at the time of BDCPA, according to institutional preference. Significant venovenous collaterals from the superior vena cava to inferior vena cava (other than the azygous vein, which is routinely ligated at the time of BDCPA) should be identified. These can be surgically addressed, or embolized at the time of catheterization. Any significant atrioventricular valve regurgitation is best detected on preoperative echocardiogram. It is important to note that, at this relatively young age of 4–6 months, very few patients are excluded from proceeding to BDCPA based on these catheterization data. A successful BDCPA can be expected in patients with PVR as high as 4 Wood units m², and mean pulmonary artery pressures in the 20 mmHg range. It is also not uncommon for shunted or banded patients to have ventricular end diastolic pressures of 10–15 mmHg prior to their successful BDCPA.

There are two general approaches taken to achieve a direct connection between the superior vena cava and the pulmonary arteries in continuity, the BDCPA and the hemi-Fontan operation. The purported advantages of the hemi-Fontan over the BDCPA are that the pulmonary arteries are routinely widely patched at the time of hemi-Fontan, and that the completion Fontan is technically easier as both cavopulmonary connections have been created at the time of the hemi-Fontan. The disadvantages of the hemi-Fontan are the fact that the hemi-Fontan requires a cardiotomy with or without a period of profound hypothermic circulatory arrest, whereas the BDCPA is entirely extracardiac and can be performed on the beating heart. Furthermore, in the hemi-Fontan the sinoatrial node is theoretically in greater jeopardy because of the more extensive dissection at the SVC-
RA junction. There has been, however, a prospective randomized trial looking at sinoatrial node function following these two operative techniques. In this study there was no difference in the incidence of sinoatrial node dysfunction between the BDCPA and hemi-Fontan groups [66].

4.2.1. Concomitant procedures

Occasionally at the time of BDCPA there is need for pulmonary arterioplasty, revision atrial septectomy, repair of residual or recurrent aortic coarctation, or atrioventricular valvuloplasty [67–69]. The performance of an additional procedure at the time of the BDCPA has not been found to increase morbidity or mortality in outcomes analyses [20,70].

4.2.2. Accessory pulmonary blood flow

Controversy exists as to the value of providing or allowing accessory blood flow to the pulmonary arteries at the time of the BDCPA or hemi-Fontan. This question is most commonly addressed in those patients with some degree of native pulmonary stenosis who are undergoing a BDCPA as their primary palliative procedure. Those in favor of leaving antegrade flow argue that the inclusion of some amount of blood containing hepatic venous effluent in the pulmonary arterial circulation will preclude the development of pulmonary arteriovenous malformations (AVMs). Pulsatile pulmonary blood flow is also theorized to improve pulmonary artery growth. It is then further argued that this will allow the patient to remain relatively better saturated, allowing the Fontan operation to be performed at a later age [71].

Although it is true that this maneuver reduces the likelihood of developing significant pulmonary arteriovenous malformations, it has not been our anecdotal experience that leaving antegrade pulmonary blood flow defers the Fontan operation significantly, if at all. With continued somatic growth, we have noted these patients to become prohibitively cyanotic, even without any evidence of either SVC to IVC veno-venous collaterals or pulmonary AVMs. Moreover, we, and others, feel strongly that the pulmonary valve and pulmonary artery stump must be appropriately oversewn in patients with a Fontan circulation. This blind-ended pulmonary artery valve and stump are thought to be a significant source of thromboemboli and have been blamed for a number of cerebrovascular accidents. As such, we feel it is easier to access this pulmonary valve at the time of a primary BDCPA, when the left main coronary artery course, posterior to the main pulmonary artery may be easily visualized. This visualization is not as easy at the time of the reoperation for the Fontan, and the left main coronary may be at some risk for injury while oversewing the pulmonary valve and pulmonary artery stump at the level of the pulmonary valve annulus. For these reasons, we do not favor maintaining accessory sources of pulmonary blood flow at the time of BDCPA or hemi-Fontan. In addition to our anecdotal institutional experience, Mainwaring et al. found worse actuarial survival in single ventricle patients who were left with accessory sources of pulmonary blood flow at the time of their BDCPA [72].

4.2.2.1. Technique: BDCPA. It is our routine to perform the BDCPA on cardiopulmonary bypass with the heart beating. The modified BT shunt, SVC, right atrial appendage, and right pulmonary artery are dissected prior to instituting CPB. The azygous vein is ligated and divided. The ascending aorta, innominate vein, and atrial appendage are cannulated. With the initiation of CPB, the BT shunt and other sources of pulmonary blood flow are occluded. The superior vena cava is divided above the expected position of the sinoatrial node. The right atrial stump is oversewn above a clamp, so that the heart has not been entered. The distal BT shunt is then removed from the pulmonary artery and the arteriotomy is enlarged. The SVC is then anastomosed to the right pulmonary artery. If there is any significant pulmonary artery stenosis this is addressed with a patch angioplasty, usually utilizing pulmonary homograft. Concomitant intracardiac procedures such as atroioventricular valve repair or atrial septectomy can be performed during a period of cardioplegic arrest or induced ventricular fibrillation prior to performing the BDCPA. The patient is then weaned from CPB while measuring SVC pressure and common atrial pressure, to assess the transpulmonary gradient. Ideally, the transpulmonary gradient should be 10 mmHg or less, and the SVC pressure should be less than 20 mmHg. Arterial saturations may vary widely early on, but should be greater than 75% if the cavopulmonary pressures are in the acceptable range.

Patients with bilateral SVCs of nearly equal diameter and no innominate vein should undergo bilateral BDCPAs. Because these vessels are, in general, more diminutive than a single cava, it is our practice to perform these bilateral BDCPAs in patients at a somewhat later age if at all possible. In addition to allowing for more certainty about the quality of the vascular anastomoses, later operation also facilitates placing venous return cannulas in both SVCs. Patients with bilateral SVCs and an adequate innominate vein should undergo ligation of the non-dominant SVC (usually contralateral to the IVC) at the time of any first stage palliation.

A number of centers have reported the use of SVC-to-right atrial shunts for the performance of the BDCPA without CPB [73]. It is our opinion that this technique should only be employed in the setting of accurate SVC or jugular venous pressure monitoring. Extreme elevations in cerebral venous pressure while the patient is normotensive and normothermic may mean that the
cerebral perfusion pressure is dangerously low. We feel that the relative safety of performing the BDCPA without the use of CPB has not been adequately demonstrated with rigorous neurologic or cognitive outcome measures.

4.2.2.2. Technique: hemi-Fontan. A technical modification of the BDCPA has become known as the hemi-Fontan. This procedure is schematically a BDCPA, wherein all of the SVC flow is into the pulmonary arteries in continuity. In the hemi-Fontan, however, SVC to right atrial continuity is maintained. The cavoatrial orifice is occluded with a dam of prosthetic material or homograft. The most widely employed variant of the hemi-Fontan, as championed by Norwood, includes a pulmonary arterioplasty in all cases [18].

There are a number of purported advantages of the hemi-Fontan vs. the BDCPA. First, the completion lateral tunnel Fontan is significantly simplified, as it becomes a completely intra-atrial procedure wherein the dam is excised and the IVC orifice is baffled to the SVC orifice. Second, the requisite pulmonary arterioplasty provides a standardized approach to pulmonary artery stenosis after first stage palliations.

We routinely perform the BDCPA at our institution. Hemi-Fontans are occasionally performed if other intracardiac interventions, such as a septectomy, or atrioventricular valve repair are planned. We have also performed the hemi-Fontan in the setting of severe pulmonary artery stenosis at the BT shunt insertion site. In that case, the stenotic segment of pulmonary artery is resected. The remaining pulmonary arteries are anastomosed end-to-side to the SVC. We, like others, have had concerns that the hemi-Fontan incision in the area of the SVC-right atrial junction might harm sinoatrial node function. This, however, was not seen in a recent retrospective review comparing sinoatrial node function in the hemi-Fontan vs. the BDCPA [74]. In addition, despite the more extensive nature of the hemi-Fontan operation, comparable mortality rates of 1–3% are reported for the BDCPA and hemi-Fontan [75].

4.3. Stage III management: Fontan operation

The current weight of opinion favors the Fontan operation as definitive palliation for single ventricle patients. The above strategies for first and second stage palliative procedures are, therefore, designed to optimize the single ventricle patient’s candidacy for achieving a successful Fontan circulation. The original descriptors of this candidacy were elaborated by Fontan and colleagues as the ‘Ten Commandments’ [76]. With the evolution of a fairly systematic approach to single ventricle palliation over the past two decades, however, the Fontan circulation has been successfully achieved in a widening population of single ventricle patients [77]. Despite this more inclusive application of the Fontan operation, we do feel that several basic goals, thematically derived from the ‘Ten Commandments’ of Choussat and Fontan should be sought in every potential Fontan operation candidate:

1. undistorted pulmonary artery anatomy;
2. low pulmonary vascular resistance;
3. low ventricular end diastolic pressure;
4. absence of obstruction to systemic outflow; and
5. preservation of systemic atrioventricular valve function [34].

It is rare that a patient will not be a candidate for Fontan after the preceding algorithms for first and second stage surgical interventions have been followed, particularly in the era of Fontan fenestration. We do not feel that there are currently any solitary absolute contraindications to the performance of the Fontan. Each physiologic or anatomic variable in the preoperative assessment should be considered independently and examined for its accuracy. Relative contraindications include single lung physiology, very diminutive or diffusely stenotic pulmonary arteries, and diffusely stenotic pulmonary veins. Again, however, it is extremely uncommon for one of our staged patients to have a sufficiently elevated pulmonary vascular resistance (e.g. >3 indexed Wood units), irreducible atrioventricular valve dysfunction, or sufficiently impaired ventricular function (e.g. EDP ≥ 15 mmHg) to exclude the patient from consideration for the Fontan operation.

It is our practice to consider patients for the Fontan operation at 18–24 months of age. As alluded to earlier, this is a fairly arbitrary time-frame established over the years at this and other institutions. The optimal timing for converting a BDCPA patient to the Fontan circulation has not been rigorously determined. Each patient is evaluated with a preoperative transthoracic echocardiogram and cardiac catheterization. The echocardiogram provides information on atrioventricular valve function and a subjective assessment of ventricular function. The catheterization is used to visualize better the BDCPA and pulmonary artery system. Pulmonary vascular resistance is calculated. The ventricular end diastolic pressure is measured and the systemic outflow tract is evaluated appropriately to rule out obstruction. The pulmonary veins are assessed for any stenoses.

In addition to diagnostic information, occlusive interventions can be performed on any significant aortopulmonary collaterals, or accessible significant collaterals from the systemic venous or pulmonary artery systems to the pulmonary veins. Theoretically, significant aortopulmonary collaterals might impart an excessive volume load on the single ventricle. In practice, however, it has been our anecdotal surgical experience that, even after a comprehensive attempt to embolize these collaterals, there is still significant return from the pulmonary veins during cardiopulmonary bypass, which is indicative of
significant aortopulmonary ‘run-off’, presumably from residual aortopulmonary collaterals. The clinical utility of these pre-Fontan interventions on angiographically identified aortopulmonary collaterals remains unproven. Large collaterals from the systemic venous system to pulmonary venous system, on the other hand, may result in significant postoperative desaturation in Fontan patients. If identified preoperatively, they should be addressed in the catheterization laboratory if possible. Collaterals from the SVC to the IVC, which might result in significant desaturation in the BDCPA patient, will be rendered irrelevant at the time of Fontan. These should not be addressed if the patient is not profoundly cyanotic and will be proceeding to a Fontan in short order.

4.3.1. Fenestrated total cavopulmonary connection Fontan

It is currently our institutional practice to perform the Fontan as an intra-atrial lateral tunnel with a 4 mm fenestration in the prosthetic Fontan baffle. This procedure became established at our institution in the late 1980s [78]. There are numerous advantages of this TCPC technique compared with the Kreutzer-type Fontan. The TCPC offers laminar flow from the IVC into the pulmonary artery, avoiding the low velocity turbulence that may result with inclusion of the entire right atrium in the Fontan circuit. The resultant giant atrium in those patients has been seen to produce thrombi, atrial arrhythmias, and pulmonary venous obstruction [7]. In addition to reducing these complications, the lateral tunnel TCPC allows for the implementation of a standard surgical technique for the vast majority of single ventricle patients. Notably, patients with left sided atrioventricular valve atresia can be treated just as those patients with right-sided atrioventricular valve atresia [8]. Indeed, it was soon discovered that the lateral tunnel TCPC was a considerable technical advance for patients in whom the pulmonary venous return is required to traverse an atrial septal defect in order to reach the atrioventricular valve. Furthermore, the lateral tunnel allows the coronary sinus to be excluded to the low-pressure pulmonary venous pathway. Chronic coronary sinus hypertension has been experimentally related to reduced coronary blood flow which may have long-term deleterious effects on single ventricle function [79]. Finally, we also find that the lateral tunnel is preferable to the extracardiac variation [80]. The lateral tunnel has demonstrated that it can grow, and the late complications from baffle thrombosis are lower than we would expect from a circumferential prosthetic graft placed in the systemic venous system.

4.3.2. Extracardiac Fontan

While some centers routinely perform extracardiac TCPCs, we have favored extracardiac Fontans for a minority single ventricle patients [22]. Certain patients with heterotaxy syndrome, wherein the hepatic veins enter the atrium remote from the IVC, are better treated with an extracardiac conduit sewn to a large ‘button’ of atrium containing the hepatic veins and IVC. This avoids the difficult contours that an intra-atrial baffle requires in that anatomic scenario [81]. Another option is an intracardiac tube graft conduit carrying both the hepatic vein and IVC flow to the pulmonary artery [82]. This also minimizes the risk of the pathway ‘buckling’ because of its circuitous nature. The downside to this intracardiac tube technique is the fact that the outer coating of these vascular grafts is not designed to contact the bloodstream. It is potentially much more thrombogenic than the inner surface, increasing the possibility of developing thrombi in the systemic atrium.

Patients with significant anomalous pulmonary venous drainage to the SVC-RA junction are also better suited for the extracardiac Fontan operation. Patients with pulmonary atresia, intact ventricular septum, and right ventricular dependent coronary circulation are sometimes treated with an extracardiac conduit as this eliminates the need for delivery of antegrade cardioplegia, a technique that relies on continuity between the coronary arteries and the aorta. In the setting of right ventricle dependent coronary circulation, an extracardiac conduit also allows for the performance of the operation without ever decompressing the right ventricle, thus allowing for the maintenance of coronary perfusion from the right ventricular chamber. We typically perform extracardiac Fontans in patients at a somewhat older age, usually 3–5 years, as this allows one to implant a conduit of at least 20 mm in diameter.

We routinely place a 4 mm fenestration in the Fontan pathway, whether it is a lateral tunnel or extracardiac conduit. Some describe multiple fenestrations or an adjustable atrial septal defect, but in our experience, regardless of the size of the infant, a 4 mm fenestration is an adequate, and yet not too large, decompressing portal [83]. Fenestration of the lateral tunnel was introduced at this institution in the late 1980s and it appeared that this technique increased the success rate of the TCPC [84]. The benefits of fenestration in our practice appeared so profound that this rapidly became routine and was never prospectively studied. Recently, however, Lemler and colleagues reported the results of their trial evaluating the efficacy of Fontan fenestration [15]. They found that baffle fenestration reduced postoperative pleural drainage, hospital length of stay, and the need for early postoperative reinterventions.

4.3.2.1. Technique: lateral tunnel fenestrated Fontan operation. We perform the Fontan operation on cardio-pulmonary bypass at moderate hypothermia (24–28 °C). The innominate vein and IVC are cannulated for venous return. Ventricular fibrillation, if it does not occur
spontaneously, is induced mechanically after several minutes of cooling. A right atriotomy is performed parallel to the atrioventricular groove. The relationship of the ASD to the SVC stump is examined. A cardiotomy vent is placed across the atrioventricular valve, to prevent ventricular distension. The SVC stump is then reopened and anastomosed to the underside of the right pulmonary artery. After the cavoatrial anastomosis is completed the baffle from the IVC orifice to the SVC orifice is sewn in place. This can be done during a period of cardioplegic arrest or during continued ventricular fibrillation. We have tended to use fibrillation more frequently in patients who have undergone a Norwood-style neoaortic reconstruction. This eliminates the need to circumferentially dissect this neoaorta in preparation for application of the cross clamp. The baffle is fashioned from a 0.6 mm thickness ePTFE flat vascular patch. Cylindrical tube grafts of ePTFE sectioned longitudinally have been abandoned due to the potentially thrombotic outer coating on these conduits. Prior to completing the baffle suture line, a 4 mm punch is used to create the fenestration in the baffle. A monitoring catheter is placed in the pulmonary venous atrium. After emerging from cardiopulmonary bypass, pressures are measured in the SVC and IVC to confirm equivalent pressures in the TCPC pathway. A central venous monitoring catheter is usually left in the IVC after removal of that venous cannula. This is not necessary if the patient has an internal jugular catheter, but they are not routinely used in our practice. Acceptable early hemodynamics are a CVP less than 20 mmHg and a transpulmonary gradient that should not be much more than 10 mmHg. We typically aim to extubate Fontan patients within 24 hours of the operation. Patients are given aspirin long term to minimize the risk of thrombotic complications.

A recent report documented the late outcome of the first 220 lateral tunnel Fontans performed at this institution [80]. These operations were performed between 1987 and 1991 utilizing the treatment strategies delineated in this review. Only 63% of this group were fenestrated as this technique was introduced at this center in 1989. Operative mortality was 5%, and the 10-year actuarial survival was 91%. Freedom from Fontan failure at 10 years was 87%. The incidence of new supraventricular tachyarrhythmias was 91% at 10 years. The incidence of new bradyarrhythmias, indicative of sinus node dysfunction, was 79% at 10 years. Of note, only three patients were identified with protein losing enteropathy. The authors concluded that these results were superior to the more classical atrio pulmonary Fontan, and that they should provide a standard by which additional Fontan modifications (e.g. extracardiac Fontan) should be assessed. Interestingly, baffle fenestration at the time of Fontan was not associated with improvement of any of the late outcome measures examined in this study.

4.3.2.2. Technique: extracardiac Fontan operation. As mentioned above, we selectively perform extracardiac Fontan operations in particular anatomic situations. We perform this operation on cardiopulmonary bypass. Cardioplegic arrest is not necessary. Ventricular fibrillation is only necessary if the right atrium must be opened to prepare the IVC cuff, but this is not usually needed. The IVC is usually divided between clamps at the IVC right atrial junction. The right atrium is oversewn. An ePTFE tube graft of at least 20–22 mm in diameter is then anastomosed to the IVC orifice above the IVC venous cannula. We have favored the use of external ring-supported conduits for this technique. The graft is then cut to the appropriate length and anastomosed to the pulmonary artery. The creation of a baffle fenestration is accomplished by placing a 4 mm punch hole in the tube graft. A circular right atriotomy is then created that is significantly larger than 4 mm in diameter. This is then anastomosed to the conduit some distance from the edges of the fenestration in the conduit. We have found that this technique is superior to the simple suture of the atriotomy to the edges of the atriotomy as this often ‘crowds’ the fenestration with atrial tissue at the suture line. Others have described the interposition of small diameter vascular grafts between the conduit and the atrium to address this problem [85].

The short-term results of the extracardiac Fontan operation, as reported in several recent series, closely resemble those of the lateral tunnel Fontan [22–25,81]. We await longer term results to compare this modification with lateral tunnel TCPC. Outcome measures of interest will be conduit patency, and the incidence of thrombotic complications referable to the conduit. It will also be important to note whether or not the extracardiac operation reduces the long-term incidence of sinus node dysfunction or supraventricular tachyarrhythmias [23,25].

4.3.2.3. Management of the Fontan fenestration. We feel that the early postoperative benefits of a fenestration between the Fontan pathway and common atrium have been well demonstrated [13,86]. The optimal management of this fenestration in the longer term, however, is less clear. It has been our anecdotal impression that approximately 25% of these fenestrations spontaneously close at some point during the first several postoperative years. It has been our tendency to perform a catheterization on our Fontan patients 2–3 years postoperatively. The fenestration can be closed with the transcatheter delivery of a device at that time. The details of this procedure are described in detail in the review of Dr Marshall and colleagues in this volume. It is important to note, however, that firm data supporting this policy
of fairly routine device closure of Fontan fenestrations is scarce. A retrospective analysis of 181 patients who underwent fenestration closure at this institution has been recently published [16]. There were suggestions from those data that fenestration closure might improve somatic growth and reduce the need for anticongestive medications. We still do not know, though, if the potential benefits of the increase in arterial hemoglobin oxygen saturation outweigh the potential beneficial contribution right-to-left shunting may make to cardiac output with exercise, for example. The effects of an incremental increase in central venous pressure with device closure are also unknown.

4.4. Special situations in the surgical treatment of single ventricle

4.4.1. Mechanical circulatory support for palliated single ventricle patients

The application of mechanical support, usually in the form of venoarterial extracorporeal membrane oxygenation (VA-ECMO), has increasingly influenced the management of the entire range of patients with critical congenital heart disease at our institution. The introduction of rapid deployment ECMO has allowed us to include this in our resuscitation protocols for all of our patients, with rare exceptions. Palliated single ventricle patients can be successfully treated with this modality, with ‘success’ defined as a successful bridge to either recovery or cardiac transplantation. These single ventricle patients, however, require special technical considerations with regard to cannulation for mechanical support and perfusion strategies while on ECMO, depending on the particular nature of their palliation.

4.4.1.1. ECMO for the single ventricle infant with an aortopulmonary shunt. We have employed venoarterial ECMO in single ventricle neonates and infants with an aortopulmonary shunt for a number of indications [87]. The predominant subgroup of patients requiring mechanical support are those with cardiovascular collapse or evidence of shunt thrombosis following the Norwood operation. If the patient presents within several days of his operation, we favor open chest cannulation of the ascending aorta for arterial inflow and the right atrial appendage for venous return. If shunt thrombosis is suspected, as is the case if the episode is one of acute desaturation accompanied by undetectable levels of end-tidal carbon dioxide, the shunt can be explored after the institution of adequate circulatory support. Thrombus can be extracted directly from the shunt, and a small (3 Fr) Fogarty balloon thrombectomy catheter can be passed into the right and left pulmonary arteries, as well as the innominate artery, to extract additional thrombus. If there is no significant thrombus discovered, or there are concerns about the technical adequacy of the shunt, cardiac catheterization should be performed while the patient is still on ECMO for both diagnostic and possibly therapeutic intervention [88]. Once the patency of the shunt is established one must address the adequacy of systemic perfusion while on ECMO. It was historically thought that the shunt should be occluded once on ECMO, so that the lungs would not be ‘flooded’ by the arterial inflow, resulting in pulmonary edema and impaired pulmonary mechanics. It has been noted, however, that complete occlusion of the aortopulmonary shunt results in what is likely irreversible ischemic lung damage [89]. It appears that bronchial blood flow alone is insufficient to adequately nourish neonatal lungs. A more successful strategy has been to increase the pump flows to significantly higher than usual flow rates for infant ECMO. In a neonatal patient with a 3.5 mm modified BT shunt, we have found that flow rates of 200 ml/kg per min are usually required to maintain adequate systemic perfusion. Plasma lactates should be followed closely during the initial period of the ECMO course. If the lactate has not dropped steadily to less than 2–3 mmol/l, and the treatment team is uncomfortable with increasing ECMO flows further, the shunt can be narrowed slightly with some metal hemoclips placed along the length of the shunt to reduce the amount of pulmonary ‘run-off’ and improve systemic perfusion.

A total of 22 aortopulmonary shunt-dependent patients have been supported on ECMO for postoperative cardiovascular crises. Twelve patients were weaned from ECMO successfully. Nine patients survived to discharge, including one patient who was successfully bridged to transplant (unpublished data).

4.4.1.2. ECMO for the patient with a BDCPA or TCPC. In contrast to early postoperative use of ECMO in infants following first stage palliation, ECMO is usually implemented for BDCPA or TCPC patients as a late resuscitative measure in patients with a failing single ventricle. As the systemic venous system is divided in the BDCPA patient, we have found it important to obtain venous drainage from both the SVC and IVC systems in this setting. This usually requires both femoral and internal jugular venous cannula placement to provide adequate flow and venous decompression, even if the single ventricle is still ejecting. We have also found it necessary in TCPC patients to obtain venous drainage from above and below the Fontan pathway. TCPC patients, particularly those without a fenestration, must be closely observed for signs of common atrial hypertension while on VA-ECMO. This results from the inability of the weak single ventricle to empty fully against the afterload created by the now pump-driven arterial perfusion pressure. Atrial hypertension on ECMO can lead to further ventricular distension and myocardial injury, as well as pulmonary edema. These effects can preclude the patient from bridging to myo-
cardiac recovery or becoming a suitable heart transplant candidate. We have, therefore, aggressively decompressed the common atrium with either recreation of a baffle fenestration, or placement of an additional venous cannula across the baffle to actively empty the common atrium.

At our institution we have treated 18 patients with either a BDCPA or TCPC with ECMO. None of the four BDCPA patients survived. Five of the 14 TCPC patients were successfully weaned from ECMO, with four long-term survivors (unpublished data).

4.4.2. Surgical treatment of acute or chronic Fontan failure

4.4.2.1. Acute Fontan failure. Early failure of the Fontan circulation has become relatively rare since the advent of staged preparation was initiated in the neonatal period [90]. In addition, an earlier review from this institution found that the early failure rate fell significantly from 27.1% to 7.5% with the introduction of baffle fenestration [13]. Our recent experience is that early failure patients present pre-Fontan with borderline native anatomy and physiology (e.g. unilateral pulmonary vein atresia) or with the late results of improper palliation (e.g. severe pulmonary artery distortion from multiple aortopulmonary shunts, or ventricular non-compliance from a prolonged period with a PA band). Acute Fontan failure is manifest by prohibitively high central venous pressure and low cardiac output. Unfavorable hemodynamics should prompt an early investigation to rule out any residual anatomic problems. This investigation usually requires a diagnostic catheterization. If a specific cause is identified, the surgeon should address it in one of two ways, if the patient is in an extremely morbid state, the Fontan can be taken down to a BDCPA, and the Fontan deferred. Early takedown to an aortopulmonary shunt has been uniformly fatal in our historical institutional experience. If the problem is easily addressed, and the patient is relatively more stable, a revision of the Fontan can be performed acutely.

4.4.2.2. Chronic Fontan failure/Fontan conversion to TCPC with arrhythmia surgery. Despite improving techniques and early operative morbidity and mortality, a number of patients will experience debilitating symptoms referable to their Fontan circulation some years out from their operation [91]. These complications include low cardiac output states due to Fontan pathway obstruction or ventricular dysfunction, atrial arrhythmias, and protein-losing enteropathy [92]. Patients presenting with these scenarios are older patients who were treated some years ago with an atriopulmonary-style Fontan. It is the general consensus that these symptoms are more prevalent in this group because of the already discussed limitations of that technique, although this cannot be firmly asserted without longer-term follow-up in the TCPC group.

Because of the purported improved flow characteristics of the TCPC, and the success of maze-type procedures in addressing atrial tachyarrhythmias, this and other centers have begun to address the debilitating late symptoms of the atrioventricular Fontan by combining a conversion of the Fontan to a TCPC in combination with the maze procedure. In these conversion operations, the original Fontan is taken down, the atrial septum excised widely and a fenestrated lateral tunnel is created. As well, a series of atrial incisions and cryoablation lesions are used to eliminate reentrant circuits to prevent atrial tachyarrhythmias. An antitachycardia pacemaker is invariably implanted at the time of this procedure to address both the sinus node dysfunction in these patients and the possible failure of the arrhythmia surgery. The TCPC at the time of conversion is typically an extracardiac conduit, as this allows for significant right atrial reduction. A fenestration at the time of Fontan conversion can also be performed, but it is thought that these older patients tolerate the resulting cyanosis less well. Most surgeons have added a fenestration to the TCPC in only a minority of their Fontan conversions [93–99].

For patients with atrial re-entrant tachycardia, most surgeons favor a right-sided maze modification, in which a large portion of the right atrial body is resected and three general cryoablation lesions are placed. One line of ablation runs from the superior edge of the ASD to the base of the resected right atrial appendage, a second runs posteriorly from the ASD margin to the open free edge of the atrial wall, traversing the crista terminalis. The final line of cryoablation spans the isthmic area from the coronary sinus to the divided os of the inferior vena cava.

Patients with atrial fibrillation undergo the more extensive maze-Cox III procedure, which, in addition to the above right sided maneuvers, includes an incision that encircles the pulmonary veins. Cryoablation lesions are then carried from this incision up to the base of the resected left atrial appendage, and down to the posterior atrioventricular valve annulus [96].

A total of eight series have reported results of Fontan conversion to a TCPC. The total experience reported to date totals 116 patients [93–102]. The reported mortality includes nine patients (six early, three late). Three of the early mortalities occurred in patients with protein losing enteropathy. The early deaths were secondary to sepsis (n = 1), arrhythmia or low cardiac output (n = 1), respiratory failure (n = 1) and generalized hypoproteinemia with progressive ascites and peripheral edema (n = 3). Clinical features that improved in these series were the NYHA class, exercise capacity, and the occurrence of supraventricular tachyarrhythmias. Even with the inclusion of arrhythmia surgery, though, there is a recurrence of atrial arrhythmias in 10–15% patients.
Not all patients with late Fontan failure are offered conversion to TCPC. Patients without significant arrhythmias as part of their presentation, or those with primary ventricular dysfunction, should probably not be offered Fontan conversion. Likewise, deconditioned patients presenting with protein-losing enteropathy have had extremely poor outcomes. Note that, during the time that Mavroudis and colleagues converted 40 patients to the TCPC, five patients presented who were not converted, but referred for cardiac transplantation [96]. To date we have not established any absolute contraindications to conversion Fontan, although there is beginning to be a bias against older aged patients (>40 years of age) as these patients seem to have more profound ventricular dysfunction from longstanding single ventricle physiology with progressive hypertrophy and fibrosis.

5. Summary

The surgical treatment of single ventricle patients remains a formidable challenge for the congenital heart surgeon. The long-term outcomes for patients with the Fontan circulation also remain to be defined. The staged strategy described in this review, however, has resulted in vast improvement in the short- and intermediate-term outcomes. Only continued close follow-up will allow both surgeon and cardiologist to confidently determine if this strategy provides for optimal long-term cardiovascular performance.

References


