47 The Adult Fontan

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Coordinated management of patients suffering from single-ventricle anatomy and physiology has improved outcomes dramatically over the past several decades. As such, survival to adulthood following the final stage of palliation, the Fontan operation, is expected for the great majority of single-ventricle patients. The unusual nature of Fontan physiology as well as the changing modes of Fontan palliation present a multitude of challenges in caring for the adult Fontan patient. Most patients who have undergone Fontan palliation within the past 20 years have received a "modern-style" Fontan operation in the form of a total cavopulmonary connection (TCPC). This has been accomplished most commonly through the construction of a lateral atrial tunnel or more recently, through the use of extracardiac conduits. This is in distinct contrast to those Fontan connections constructed more than 25-30 years ago, which were predominantly atriopulmonary (A-P) Fontan connections. Given the long-term necessities of care and the frequent transitory nature of many patients, it is incumbent on the healthcare team to obtain surgeons' records of previous Fontan operations (or if unavailable through the use of advanced imaging) to determine the exact type of Fontan connection in a given patient. In general, patients with A-P Fontan connections tend to present with more frequent atrial arrhythmias, particularly reentrant tachycardias, than patients with TCPC Fontans, although this may be a feature of length of follow-up. Even in the absence of arrhythmias, most A-P Fontan patients should be considered for Fontan conversion surgery (see below).

Fontan Complications

Fontan palliation presents many challenges related to chronic cardiac and noncardiac complications. At approximately 20-25 years following Fontan palliation, many patients start to display some signs of these issues. As discussed in Chapter 39, close follow-up and longitudinal testing should be used to monitor Fontan patients. TCH has developed a long-term pathway care plan for these patients (Figure 39-4).

Cardiac Complications

Cardiac complications can be divided into several categories including structural, functional, and electrical. In the complex constellation of anatomic variants undergoing Fontan operations, there are many opportunities for residual or new structure abnormalities. These may include semilunar valve insufficiency, systemic outlet obstruction, pulmonary venous drainage obstruction, coronary sinus obstruction, AV valve regurgitation or stenosis, anastomotic strictures, or other pathway distortions. Any of these problems can result in compromised Fontan physiology and may be sufficient indication for a revision operation. AV valve regurgitation is an independent predictor of poor prognosis.

Fontan patients frequently develop diastolic dysfunction often leading to elevated end-diastolic pressures (EDP). This exacerbates a multitude of issues leading to elevated Fontan pressures. Systolic dysfunction tends to be less frequent, but if present, should

be treated with optimizing heart failure medications and considering mechanical/surgical support in extreme cases.

From an electrical perspective, bradycardia occurs eventually in many adult Fontan patients due to sinus node dysfunction. This can lead to a marked decline in functional status due to chronotropic incompetence and should be addressed, in many cases, with pacemaker implantation. Tachycardia is common, especially intra-atrial reentry tachycardia (IART). This is often difficult to diagnose as telemetry appears similar to a sinus tachycardia with a heart rate of 120-130s. However, anytime a Fontan patient is noted to have a relatively fixed heart rate above 110s, IART should be considered.

Noncardiac Complications

A multitude of noncardiac complications in adult Fontan patients can occur stemming from elevated systemic venous pressures. Any obstruction in the Fontan pathway or branch PAs should be aggressively managed. Congestive hepatopathy is the rule, often leading to signs of cirrhosis. Routine liver surveillance is required and a hepatology team, with particular understanding and experience of Fontan physiology, should probably follow most adult Fontan patients. While liver fibrosis is commonly seen, it is relatively rare to see any classic findings of portal hypertension, and hepatic synthetic dysfunction is not typically present, particularly in early stages.

Fontan patients are also at an elevated risk for thromboembolic events on both the systemic and pulmonary sides of the circulation. Most practitioners believe that Fontan patients should be treated, at the very least, with daily aspirin and many of the more complex patients may require full oral anticoagulation. Other complications that should be monitored, and treated if present, include protein-losing enteropathy (PLE), chronic kidney disease, pulmonary venovenous collaterals, and coagulopathy.

The Failing Fontan

The term "failing Fontan" encompasses a broad spectrum of conditions from various hemodynamic and conduction abnormalities to extracardiac end-organ issues, including most frequently, the GI tract. A useful acronym – **FACET** – helps to address issues related to **F**unction, **A**rrhythmia, **C**yanosis, **E**nteropathy, and **T**hrombosis:

- **Function.** Systolic and diastolic dysfunction develop in many patients and initial anticongestive medical management is reasonable with limited data showing benefit.
- **Arrhythmia.** Brady- and tachyarrhythmias are common in adult Fontan patients. Antiarrhythmic medications can delay the need for ablation or Maze procedures. Most defibrillators are placed epicardially, but there may be a role for subcutaneous technology.
- **Cyanosis.** Cyanosis may develop from increasing Fontan pressures leading to systemic-to-pulmonary venous collaterals or due to prior surgical fenestration, both of which are amenable to catheter occlusion. However, cyanosis may recur with Fontan pressure elevation. Liver disease or misdirection of hepatic venous flow may contribute to the development of intrapulmonary arteriovenous malformations and intrapulmonary shunting, also leading to cyanosis. These may require redirection of the IVC baffle or liver transplantation.

- **Enteropathy.** Should PLE occur, reversible hemodynamic causes such as Fontan circuit obstruction or valvulopathy causing atrial hypertension should be addressed. However, this may be insufficient in advance stages. Multiple medical therapies and fenestration creation have been reported to be useful in some patients, but heart transplantation remains the mainstay in treatment of PLE.
- **Thrombosis.** As mentioned above, Fontan patients are at a higher risk for thromboembolic events. Antiplatelet therapy and anticoagulation are commonly used.

Certain patients with failing Fontan circulation may benefit from surgical interventions:

- The Fontan conversion (revision from older A-P to newer extracardiac or lateral tunnel versions combined with atrial Maze and epicardial pacemaker implantation) is an antiarrhythmic surgery for medication-refractory atrial arrhythmias best performed when ventricular function is preserved and in the absence of cyanosis.
- With newer reactive antitachycardia pacing devices, a dual-chamber epicardial pacemaker without Maze or Fontan conversion can benefit patients with chronotropic incompetence, poor hemodynamics from lack of AV synchrony, and/or medication-refractory atrial dysrhythmias.
- For medication-refractory systolic heart failure, a systemic ventricular assist device can be considered as a bridge to transplant or destination therapy after weighing the risks of thrombosis, anticoagulation, and infection.
- Heart transplantation is beneficial for pump failure, diastolic heart failure, PLE, or significant liver fibrosis prior to cirrhosis.

Anesthesia Considerations for the Fontan Patient

Anesthesia for the adult Fontan patient is aimed at assuring cardiopulmonary stability throughout the intraoperative period into the ICU transition. Because these patients have had multiple previous cardiac operations, they are at a much higher risk of bleeding during redo-sternotomy and from scar tissue during dissection. At least 1, preferably 2, large bore IVs (>18 gauge) are appropriate along with central venous access. Blood volume replacement should be geared to maintaining and/or restoring normal colloid osmotic pressure by the use of albumin 5% or FFP and PRBC as appropriate. Many of these patients have had chronic hepatic congestion and may have impaired liver synthetic function, thus impairing clotting factor production.

Some older Fontans may have had "classic" BT shunts with loss of the ipsilateral subclavian artery at some point in their care. It is necessary to know this ahead of time in order to guide noninvasive BP measurement and arterial-line location. Because of the high incidence of arrhythmias in these patients, it is appropriate to continue antiarrhythmic therapy through the day of surgery and to place external pacing/defibrillator pads on the patient once in the OR. Many of these patients also have pacemaker systems and it is strongly suggested that pacing devices be placed in a mode to avoid interference from the use of electrocautery. Ventilation should be adjusted to minimize intrathoracic pressure by utilizing as little PIP and PEEP as necessary to maintain alveolar patency and appropriate blood gases. If plastic bronchitis is present, the ETT should be irrigated and suctioned regularly to minimize airway obstruction.

Post-CPB care is directed towards normalizing intravascular status, assuring minimal

surgical bleeding by the use of blood products (including factor supplements and prothrombotic agents such as Kcentra® and activated Factor 7 as appropriate). Ideally, these patients should have a postoperative hemoglobin >13 g/dL in order to maximize oxygen carrying capacity. Many of these patients will benefit from the use of low-dose vasopressin infusions, especially if they have been on angiotensin-converting enzyme (ACE) inhibitors preoperatively. Epinephrine and milrinone should be titrated based upon cardiac function as determined by intraoperative TEE.

Many of these patients will have been on aspirin therapy and will require platelet transfusion; transfusion goals may be directed by the use of advanced clotting studies such as ROTEM®. Attempts should be made to promote perioperative extubation as soon as possible, ideally in the OR, to take advantage of the supplementary effects of negative inspiration on pulmonary blood flow in the Fontan circuit. This is facilitated by the limited use of narcotics and benzodiazepines, and the use of dexmedetomidine and acetaminophen IV. Appropriate perioperative pain control with judicious use of a narcotics and nonnarcotic analgesia to minimize splinting and postoperative atelectasis is very important. As soon as the use of intraoperative electrocautery has been completed, the pacing system, if in place, should be adjusted to the most appropriate mode (typically DDD). Transfer of care to the CICU should include a complete handover including such factors as cardiac rhythm (and pacing mode), function postrepair, blood product usage, intraoperative pressures in the Fontan circuit, antibiotic and analgesia plans, and goals for airway management if not extubated.

Fontan Revision

In properly selected patients, a Fontan revision operation can confirm considerable benefit in allowing patients to have improved Fontan physiology, mitigation of secondary hemodynamic and symptomatic consequences of the failing Fontan, and the avoidance (or at least delay) of cardiac transplantation. As noted above, indications for a Fontan revision are centered on improving a correctable anatomic, functional, or electrical problem with the existing Fontan circuit. Historically, Fontan revision operations were predominantly utilized in patients with failing A-P Fontan connections. More recently, it has become clear that other forms of Fontan connection may also develop correctable problems that merit intervention.

In selecting patients for a Fontan revision, it is critical to conduct an exhaustive preoperative functional and physiologic assessment. As noted above, it is enormously useful to obtain and carefully study old operative records, including all previous palliations. Cardiac assessment should include detailed echocardiography (transthoracic or transesophageal), diagnostic (and where indicated interventional) cardiac catheterization, cardiac MRI and/or chest CTA, pulmonary function testing, and 24-hour Holter monitoring. Detailed blood work should include serum chemistries, complete blood count/indices, liver function testing, coagulation profile, and thyroid function testing (many patients are chronically treated with amiodarone and are at risk of hypothyroidism). In some patients, liver biopsy may be considered, particularly in the setting of borderline synthetic function, although interpretation of biopsy results in these patients may be very challenging. It is also critical to perform an extensive survey of vascular access

as all of these patients have undergone multiple previous interventions and there may be chronic occlusions of important access vessels including the femoral arteries and/or veins, issues that need to be understood for operative planning. In patients with any history of neurologic dysfunction, a detailed evaluation by a neurologist and in most cases, brain imaging, is mandatory prior to proceeding with a surgical procedure.

In counseling patients for the revision operation, the proposed operation is discussed usually during several lengthy preoperative sessions. Patients must be informed of the very complex nature of the operation. These are typically very long procedures; operations extending for as long as 10-12 hours are not infrequent. It is very important to inform patients that revision operations were formerly associated with a very high perioperative risk of morbidity and mortality. Fortunately, in the current era, in properly selected patients, several centers have reported outstanding outcomes (our overall perioperative survivorship for all Fontan revision operations from 1995-2018 was >99%). Nonetheless, patients should know that there are alternatives. The historical mainstay of the failed Fontan has been cardiac transplantation. While a complete review of this topic is beyond the scope of this chapter, cardiac transplantation in a Fontan patient can be a very challenging and risk-laden proposition with variable results being reported. Previous anatomic distortions, situs abnormalities, chronic multiorgan dysfunction (particularly hepatic insufficiency), patient sensitization with preformed circulating antibodies, and other challenges all combine to make transplantation in Fontan patients a potentially challenging intervention. Finally, bailout strategies are always discussed, including support with ECMO and temporary and durable VAD support.

The revision operations require detailed preparation by the anesthesia team as noted above. For the surgeon, carefully planned sternal reentry is paramount. Patients with failing A-P Fontan connections may have massively dilated systemic atria, often immediately underneath the sternum. In others, the aorta or PAs may be intimately associated with the sternum. Our approach has been to proceed with careful, deliberate sternal reentry without the routine use of preemptive femoral cannulation. In most cases, with meticulous direct vision dissection, reentry can be safely accomplished. The critical, immutable key features are meticulous dissection, under direction vision and without need of hurry. Breaching these principles can be catastrophic.

Once sternal reentry has been achieved, the anatomy is carefully dissected, again with careful attention to hemostasis. CPB is then instituted with the use of separate, direct vena caval cannulation. Our routine has been to use mild hypothermia (nasopharyngeal temperature of 32 °C) for most cases. The operative plan should include complete takedown of the previous Fontan connection, often with considerable debulking of the massively dilated RA. Previous ASD patches are often calcified and should be removed, if possible. All structural deficiencies that are amenable to correction should then be addressed. These include AV valve repair or replacement, semilunar valve replacement, subaortic (systemic outlet) resection, pulmonary venous pathway obstruction relief, and repair of PA stenosis. A modified cryo-Maze procedure is performed to include isolation of potential reentrant pathways on both right and left atrial aspects. After reconstruction of the debulked common atrium, the new Fontan channel is constructed with an extracardiac tube graft (typically 24 mm Gore-Tex®) anastomosed between the

divided IVC and the branch PAs. If the patient did not have superior cavopulmonary connections previously, bidirectional (bilateral in appropriate situations) Glenn shunts are constructed. Finally, it is critically important to place a dual-chamber epicardial pacing system at the end of the operation. We have favored bilateral, epicardial steroid eluting "button" electrodes. Finding an acceptable lead site can be a challenging proposition in patients who have undergone multiple previous surgeries, but with diligence, the surgeon will be able to find appropriate locations. The pulse generator is selected in coordination with the collaborating cardiac electrophysiologist (a critical relationship in the management of these patients) and should be a sophisticated device with overdrive pacing capabilities. Despite the length of these operations, we favor early extubation (typically in the OR) and rapid progress through the CICU. The typical hospital length of stay is around one week. Despite the use of Maze operations and epicardial pacing, most patients are at risk of recurrent atrial reentrant tachycardias, particularly during the first 6 months after the revision operation. As such, many centers prefer to maintain patients on oral amiodarone therapy, but this remains a somewhat controversial subject.

Cardiac Replacement Therapy

In patients with refractory heart failure in the setting of systemic ventricular dysfunction, treatment with a VAD and/or cardiac transplantation may be the only options. While each subject could merit its own separate chapter, several points are worth mentioning. We first "discovered" the utility of placing continuous flow devices in patients with failing Fontans when we placed a HeartMate II™ continuous flow device in an adult Fontan patient with profound ventricular dysfunction. We were uncertain how the Fontan circulation would perform in this setting and were pleased to observe excellent hemodynamics with improvement in Fontan pressures. This reemphasizes the critical nature of ventricular dysfunction in overall Fontan circuit performance. The patient was quickly progressed through the hospital and this experience led us to become more liberal with the use of VADs as either a bridge to transplantation or chronic "destination" therapy in appropriate patients.

Cardiac transplantation in failing Fontan patients is variously reported by different centers as prohibitively risky or of acceptable risk profile. This fact, of course, has to do with patient selection and technique. By way of patient selection, it is important to emphasize the general rule that in a failing Fontan with preserved ventricular function, something else is wrong (distorted PAs, obstructed pulmonary veins, elevated PVR to name a few) and great caution should be exercised in proceeding with cardiac transplantation. The transplant operation itself can be very challenging. Patients with multiple reoperations have increased bleeding risk, exacerbated by hepatic dysfunction, arteriovenous and venovenous collateral burden, anatomic distortion, and other features. Patients with complex venous connections, distorted branch PAs, situs abnormalities (e.g., left-sided venae cavae) require creative technical solutions, but these patients are candidates for transplantation. We believe that an experienced congenital heart surgeon should be involved in complex transplantation operations

in patients with complex structural congenital heart anomalies, particularly in failing Fontan patients, to optimize outcomes.

Long-Term Follow-Up

Patients with a Fontan circulation require diligent, frequent, lifelong surveillance by an adult congenital cardiologist who works in close association with a congenital heart surgeon well versed in adult remedial operations. While the question of how long one can live with a Fontan circulation is, as of yet, unanswered, we have several patients who are now in their early 60s with very satisfactory cardiorespiratory physiology in the setting of a Fontan connection. It is incumbent on the medical community to properly shepherd these patients through the stages of life in an expectant, vigilant manner.