Is the Extracardiac Conduit the Preferred Fontan Approach for Patients With Univentricular Hearts?

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Brian Kogon, MD

With improvements in technology, operative technique, and critical care medicine, more and more children with congenital heart disease are surviving into adulthood. As they age, these patients are providing physicians with new challenges, and we are realizing the long-term consequences of our previous interventions. This oftentimes has changed the mode of treatment for subsequent children.

Fontan palliation for single-ventricle patients was first described in 1971. The original approach was in the form of a classic atriopulmonary Fontan connection, bypassing the nonfunctional right ventricle. After recognizing the long-term complication of an atriopulmonary connection (right atrial dilation, arrhythmias, and thrombus formation), modifications have been made. The atriopulmonary connection has been abandoned in favor of modified Fontan anatomy and physiology.

The modified Fontan can be performed by the use of an extracardiac conduit approach or a lateral tunnel approach. The extracardiac Fontan uses an external conduit to anastomose the inferior vena cava into the pulmonary arteries, whereas a lateral tunnel Fontan uses a baffle within the right atrium (Figure 1). Because the systemic venous pathway is completely separated from the right atrium with the extracardiac Fontan, it is more challenging for the electrophysiologist to access the heart for ablation. My aim is to demonstrate why “the extracardiac conduit is the preferred Fontan approach for patients with univentricular hearts.”

The extracardiac Fontan has several advantages over the lateral tunnel Fontan.

**Advantage 1: Reduced Cardiopulmonary Bypass and Ischemic Times**

Minimizing cardiopulmonary bypass and ischemia may optimize early postoperative outcomes. In multiple studies, both prolonged cardioplegic arrest and extended cardiopulmonary bypass times have been associated with an increased risk of early postoperative death or Fontan failure.

The technique of Fontan construction often dictates whether cardiopulmonary bypass or a period of ischemic arrest are required. With the lateral tunnel Fontan, the right atrium is opened for creation of the inferior vena cava baffle, necessitating the use of cardiopulmonary bypass. In addition, a period of cardioplegic arrest is required to stop the heart to prevent air embolic events until the atrium is closed. With the extracardiac Fontan, the right atrium is not opened. Vascular control is achieved at the right atrial-inferior vena cava junction. The inferior vena cava is transected and sewn to the extracardiac conduit, and the cardiac opening is oversewn. Vascular control of the pulmonary arteries is then achieved, and the conduit is sewn to the right pulmonary artery. Last, side-biting clamps are placed on the right atrium and conduit to create a fenestration if desired. With the use of this...
technique, perfusion to the myocardium is maintained throughout the operation, and cardioplegic arrest is not required.

Interestingly, several recent studies did not show a significant difference in the total cardiopulmonary bypass times between the 2 techniques. Respective bypass times for the lateral tunnel and extracardiac Fontans were 124 versus 119 minutes, 107 versus 117 minutes, and 109 versus 102 minutes in several such studies. The time saved by eliminating an ischemic period and subsequent reperfusion is likely offset by the need to suture 2 anastomoses and a fenestration, as well. Nonetheless, the ischemic times can significantly reduced. One study shows average ischemic times of 4 minutes with the extracardiac Fontan and 48 minutes with the lateral tunnel.

There have also been isolated reports of extracardiac Fontan operations being performed without the use of cardiopulmonary bypass altogether. For the inferior vena cava, a passive bypass system between femoral vein (or low inferior vena cava) and the right atrium was used to maintain venous drainage of the lower body, while the inferior vena cava was controlled, transected, and anastomosed to the extracardiac conduit. In 1 case, the conduit was then anastomosed to the right pulmonary artery, while a left-sided superior cavopulmonary connection (of a bilateral) maintained pulmonary blood flow. In the other case, the conduit was then anastomosed to the central pulmonary artery, while the right-sided superior cavopulmonary connection maintained pulmonary blood flow.

**Advantage 2: Improved Hemodynamics**

Many studies have been performed to compare Fontan hemodynamics between various Fontan designs. Figure 2 shows the standard T-shaped Fontan connection (Figure 2, left), Lateral tunnel Fontan shows the Fontan pathway inside of the right atrium. Right, Extracardiac Fontan shows the conduit completely separated from the right atrium.
left) that is more characteristic of the lateral tunnel Fontan, and the offset connection (Figure 2, center) that is more characteristic of the extracardiac Fontan.

These studies used explanted animal Fontan models studied within in vitro flow loops, manufactured transparent stereolithographs reconstructed from patient MRIs, and models derived from MRI data together with computational fluid dynamics. With pulmonary resistances and cardiac outputs held constant, comparisons were made between different anatomic configurations. Studies typically examined right- and left-branch pulmonary artery flow distribution, superior and inferior vena cava pressures, and energy dissipation. The optimal Fontan configuration would show equal distribution of Fontan flow between the right- and left-branch pulmonary arteries, low vena caval pressures, and minimal energy loss. The extracardiac Fontan provided superior hemodynamics in comparison with the lateral tunnel Fontan. A further benefit was added with the conduit-superior vena cava offset in comparison with the T-type connection. These hemodynamic advantages were enhanced at high physiological flows that are representative of exercise.

More recent studies have evaluated a novel Y-shaped extracardiac Fontan, one in which the extracardiac conduit is constructed with 1 limb to the right pulmonary artery and 1 limb to the left pulmonary artery (Figure 2, right). In simulations, there was more high-velocity turbulent flow with the T-shaped connection than with the offset connection and the y connection. In addition, there were also more high-pressure zones with the T-shaped connection than with the offset connection and the y connection. The Table shows the relative rankings and superiority of the extracardiac Fontan designs, with respect to efficiency, pressure level, and flow distribution.

<table>
<thead>
<tr>
<th>Fontan Design</th>
<th>Efficiency</th>
<th>Lower Superior Vena Cava Pressure</th>
<th>Lower Inferior Vena Cava Pressure</th>
<th>Equal Branch Pulmonary Artery Flow Distribution</th>
</tr>
</thead>
<tbody>
<tr>
<td>T-shaped connection</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>++</td>
</tr>
<tr>
<td>Offset connection</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
<td>+</td>
</tr>
<tr>
<td>Y-shaped connection</td>
<td>++++</td>
<td>++++</td>
<td>++++</td>
<td>++++</td>
</tr>
</tbody>
</table>

The designs are ranked (+ +++ = best, + = worst) according to performance for efficiency, pressure level, and flow distribution.

Advantage 3: Reduced Arrhythmias

Arrhythmias are a persistent source of morbidity and mortality after the Fontan operation. Sinus node dysfunction and tachyarrhythmias are most common. The extracardiac Fontan may decrease the incidence of atrial arrhythmias by minimizing factors that predispose patients to arrhythmia generation. These factors include (1) exposure of the right atrium to elevated systemic pressures, (2) extensive atrial incisions and suture lines, (3) surgery in the vicinity of the sinus node, and (4) ventricular dysfunction resulting from ischemic arrest and long cardiopulmonary bypass times.

Figure 3 shows the lateral tunnel Fontan baffle being placed within the right atrium. It shows the portion of the right atrium within the Fontan pathway that remains exposed to elevated pressures, it shows the right atriotomy, and it shows the baffle suture lines. In particular, it shows the superior aspect of the baffle where these stitches are in close proximity to the sinus node. These aspects of the lateral tunnel Fontan construction are eliminated with the extracardiac Fontan design.

The incidence of atrial arrhythmias following Fontan-type surgery increases steadily with the postoperative interval, with at least 50% of patients experiencing problematic atrial tachycardia by 20 years of follow-up.

Figure 3. Lateral tunnel Fontan demonstrating the position of the intra-atrial baffle, the segment of right atrium exposed to elevated Fontan pressures, and the location of the suture lines near the atrial conduction system.
The arrhythmia profile is more favorable with the extracardiac Fontan in comparison with other Fontan designs. With nonextracardiac Fontans, the reported incidence of postoperative atrial arrhythmias (including supraventricular tachycardia and sinus node dysfunction) ranges between 14% and 32%. In a direct comparison, at 6-year follow-up, the freedom from arrhythmia was 83% for the lateral tunnel Fontan and 92% for the extracardiac Fontan ($P=0.022$). Amodeo et al also demonstrated a 5-year freedom from arrhythmia rate of 92% with the extracardiac Fontan. Another study looking specifically at the extracardiac Fontan reported new-onset transient supraventricular arrhythmias in 10% of patients in the early postoperative period. Most were short lived (<10 minutes), or they occurred when the patients were under the influence of endogenous (pain) and exogenous (inotropic agents) catecholamines. No patients were discharged on antiarrhythmic medications. The same study reported transient sinus node dysfunction in 8% of patients in the early postoperative period. Most of these resolved within 48 hours of surgery, and no patients required a permanent pacemaker at the time of discharge.

Despite these data, there is considerable variability in definition and criteria for arrhythmia reporting, making direct comparison between techniques and studies challenging. As such, not all studies show an improved arrhythmia profile with the extracardiac Fontan. Some studies show no statistical arrhythmia difference between lateral tunnel and extracardiac Fontan configurations. Some of the differences relate to arrhythmia classification: total arrhythmias, arrhythmias continued from the early postoperative period, or newly diagnosed arrhythmias at follow-up. Some of the differences relate to the variable thresholds for postoperative atrial pacing.

Interestingly, although 1 additional article continues to show a higher incidence of arrhythmias in the lateral tunnel group, it suggests that the previous staging operation may be the most important factor. Arrhythmias were more common in the bidirectional Glenn/lateral tunnel approach, than either the hemi-Fontan/lateral tunnel or bidirectional Glenn/extracardiac approaches. The lateral tunnel approach following a bidirectional Glenn requires repeat dissection in the vicinity of the sinus node and artery, an area that is typically scarred and lacks the cavoatrial junction as a landmark, resulting in increased postoperative arrhythmias. The arrhythmia benefit of the extracardiac Fontan may be ameliorated by altering the pre-Fontan staging strategy when anticipating a subsequent lateral tunnel technique.

**Advantage 4: Atrial Access for Electrophysiology Study May Still Be Achieved**

Despite an extracardiac Fontan, the electrophysiologists will find a way to get into the heart. Khairy et al report “Transcatheter ablation via a sternotomy approach as a hybrid procedure in a univentricular heart.” In this case report, the authors describe a 6-year-old girl who developed unstable supraventricular tachycardia shortly after her Fontan operation. She was taken back to the operating room where her chest was reopened and access was obtained by direct atrial puncture. Obviously in this instance, this was only feasible because of a fresh sternotomy and dissection.

Brown et al report, “Transapical left ventricular access for difficult to reach interventional targets in the left heart.” In this report, one of the patients had an extracardiac Fontan and required ablation. Access to the heart was achieved through a per-ventricular approach. The authors concluded that “blind” percutaneous left ventricular puncture was effective and could be considered, but that puncture under direct visualization following minithoracotomy was preferable because of fewer complications.

More importantly, Pass et al report “A transfemoral approach to ablation in a child with an extracardiac Fontan.” Successful ablation was performed in an 11-year-old child with a double-inlet left ventricle, who underwent extracardiac Fontan at age 3. Dave et al also reported “Transconduit puncture for catheter ablation of atrial tachycardia in a patient with extracardiac Fontan palliation.” Successful ablation was performed in a 34-year-old patient with tricuspid atresia, who underwent Fontan conversion from an atriopulmonary Fontan to extracardiac Fontan at age 30. The authors concluded that transconduit puncture, although challenging, provides a minimally invasive option for definitive treatment of these highly symptomatic and often life-threatening arrhythmias. In the appropriate setting, this technique may prove to be a viable alternative to on- or off-pump arrhythmia surgery, hybrid atrial or ventricular puncture, or direct transthoracic puncture.

**Conclusions**

The advantages of an extracardiac Fontan outweigh those of an intracardiac Fontan even though it makes it more challenging for the electrophysiologist.

**Disclosures**

None.

**References**

Response to Kogon

Paul Khairy, MD, PhD; Nancy Poirier, MD

Dr Kogon is to be commended for his defense of the extracardiac conduit as the preferred Fontan approach. This perspective is, however, based on theoretical assertions unsubstantiated by clinical evidence. First, it is argued that the simpler surgery may improve early postoperative outcomes. Clinical evidence has refuted this hypothesis. As Dr Kogon admits, several recent studies have not shown significant differences in cardiopulmonary bypass times, and longer hospitalizations have been reported following extracardiac conduits. The second argument related to hemodynamics is likewise theoretical. To our knowledge, no study has convincingly demonstrated improved functional outcomes with extracardiac conduits in comparison with lateral tunnels. Hemodynamics are influenced to a far greater extent by a host of factors that extend beyond the type of cavo pulmonary connection. Finally, Dr Kogon views the 90% to 92% arrhythmia-free survival rate at 5 to 6 years as justification for the extracardiac conduit. Interestingly, a near-identical rate was reported with the atrio pulmonary Fontan, only to plummet below 50% with increased follow-up. Dr Kogon aptly concedes that a reduction in arrhythmias is far from established. Moreover, these arrhythmias are exceptionally problematic to manage and, as Dr Kogon acknowledges, may be “highly symptomatic” and “life-threatening.” To quote Henry Louis Mencken: “For every complex problem there is a solution that is simple, neat, and wrong.” Disadvantages of the extracardiac Fontan far outweigh any potential benefit. Relying on heroic high-risk bailout measures to treat common, predictable, and highly morbid complications is an unacceptable standard of care. The congenital community is challenged, once again, to display creativity and foresight in improving outcomes for future generations.
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