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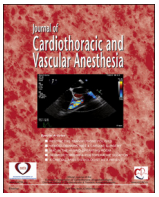

Expert Review

Fontan Palliation for Single-Ventricle Physiology: Perioperative Management for Noncardiac Surgery and Analysis of Outcomes

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Fontan Surgery: Historic Evolution and Indications

IN 1971, FRANCIS FONTAN and Eugene Baudet introduced a palliative surgical procedure for tricuspid atresia, describing the surgery as "a procedure of physiological pulmonary blood flow restoration, with suppression of right and left blood mixing."[1] This was accomplished by combining Glenn's procedure (1958) of anastomosing the superior vena cava (SVC) to the distal end of the right pulmonary artery (in order to perfuse the right lung) with an atriopulmonary connection between the proximal end of the divided right pulmonary artery and the anatomic right atrium. After closure of the atrial septal defect, inferior vena cava (IVC) blood flow would be guided through the right atrium and into the left pulmonary artery to perfuse the left lung. Ligation of the main pulmonary artery ensured complete bypass of the nonfunctional right ventricle. A similar procedure for the treatment of tricuspid atresia was introduced separately by Guillermo Kreutzer and his colleagues at the Children's Hospital in Buenos Aires during the same period.[2] The term "Fontan/Kreutzer procedure" is sometimes used to acknowledge the simultaneous, independent contributions of both groups.[3] The original procedures designed by both groups assumed that a pumping atrium was necessary to serve as a power source. Theoretically, atrial contraction could provide the pumping mechanism to propel blood flow into the pulmonary artery and ensure adequate perfusion of the pulmonary circulation.[3,4] Instead, this technique caused the atrium to dilate, resulting in blood stagnation and energy dissipation, leading to diminished flow.

Over the past 5 decades, a number of significant modifications to the original Fontan surgery have occurred. Most notably, the atriopulmonary connection has been replaced by a total cavopulmonary connection. Surgeons recognized that a pumping mechanism was not necessary to provide adequate blood flow and that, in fact, improved flow and hemodynamics were possible via this alternative arrangement.[4,5] Two primary forms of cavopulmonary connection have been developed. The first was the lateral tunnel technique using a Gore-Tex patch and a portion of the atrial wall to construct an intra-atrial baffle from the IVC to the right pulmonary artery. A second, more recent modification, uses a large Gore-Tex tubular extracardiac conduit to connect the IVC to the right pulmonary artery.[4] Both techniques are used currently in modern practice. A further modification fenestrates the Fontan circuit (direct connection between the Fontan circuit and the atrium) to allow for a decompressing, controlled right-to-left shunt. In the setting of increased pulmonary vascular resistance (PVR), flow across this fenestration permits filling of the systemic ventricle to aid in maintaining cardiac output while limiting elevation of the Fontan circuit pressure.[4–7]

The role of this surgery has been expanded to allow palliation of a broader spectrum of complex congenital cardiac diseases involving a single functional ventricle. In particular, Norwood's recognition that the right ventricle could be used to

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supply the systemic circulation opened the door for patients with hypoplastic left-heart syndrome to become candidates for the Fontan procedure, expanding its indication to patients with nonfunctioning left and right ventricles.[4] The surgery may be considered for a number of congenital cardiac defects that are not amenable to a biventricular repair including, but not limited to, the following: tricuspid atresia, mitral atresia, double-inlet left ventricle, hypoplastic left-heart syndrome, hypoplastic right ventricle, heterotaxia syndromes, unbalanced complete atrioventricular septal defects, pulmonary atresia with intact ventricular septum (with marked hypoplasia of the right ventricle), and double-outlet right ventricle with aortic atresia (or with left ventricle remote from aortic valve).[8,9] The far-reaching effects of the Fontan surgery allowed for the palliation of previously terminal conditions and laid the framework for the changing epidemiology of congenital heart disease, which now includes more adults than children and a significant proportion of complex pathology.[10]

Even though the total cavopulmonary connection technically can be performed as a single procedure, a staged approach has become standard practice. This begins with the SVC-to-pulmonary artery anastomosis (Glenn procedure) at approximately 3-to-6 months of age followed by a separate, later surgery to connect the IVC to the pulmonary circulation when the patient attains approximately 15 kg. The need for this stepwise palliation primarily is due to the naturally elevated PVR present in the neonatal/early infancy period, which precludes Fontan completion at that time.[11] The Fontan surgery typically is not performed until after the patient is 18 months old; however, it is important to note that there is no consensus regarding the ideal timing of the procedure.[11,12] A controversy exists, with some advocating for early performance of the surgery (ie, 18 months to 2 years), whereas others suggest delaying the surgery to postpone the onset of the inevitable long-term complications of the Fontan circulation.[11,12] Perhaps more important than the timing of the surgery is appropriate patient selection. In particular, Pundi et al reported decreased survival in those with preoperative pulmonary hypertension (>17 mmHg) and absence of normal sinus rhythm.[13] Even though the staged approach to single-ventricle palliation currently is embraced, the details regarding patient selection and procedural timing remain uncertain. An Australian study estimated a doubling in the single-ventricle population over the next 30 years[14] and a British study suggested 60% growth in only 10 years.[15] It is clear that the total single-ventricle population will continue to grow rapidly.

Physiology

The physiology of the post-Fontan state has been adequately described as a "man-made form of chronic heart failure" and is unavoidably abnormal.[16] In particular, chronic central venous hypertension and diminished cardiac output ultimately may contribute to the host of debilitating and life-threatening multiorgan complications. Simply put, the lack of a reservoir and pumping chamber (the right or subpulmonary ventricle) on the venous return side of the systemic circulation establishes a passive rather than pulsatile pulmonary circulation.

Passive pulmonary circulation means that blood flow through the lungs is determined by the PVR, which is the intrinsic impedance to flow offered by the pulmonary vasculature, and the capacitance and preload on the systemic venous return side of the circulation. Other important parameters of single-ventricle physiology are the systolic function and diastolic compliance of the single ventricle and the proper function of the semilunar valve; the atrial-ventricular valve (s) also are paramount to adequate cardiac output. The ability to increase cardiac output is limited by the absence of a subpulmonary ventricle because of a lack of reserve to increase oxygen uptake and systematic ventricular preload. Exercise capacity can be impeded severely in many of these patients and worsens as the patient ages.[17] Pulmonary blood flow is optimal when PVR is as low as possible and systemic venous capacitance and preload are generous (central venous pressure high-normal at 10-15 mmHg). When the surgery was first proposed for patients with tricuspid valve atresia, patient selection criteria included the following[1,18]:

1. Age at surgery between 4 and 15 years
2. Presence of sinus rhythm
3. Normal systemic venous return
4. Normal right atrial volume
5. Mean pulmonary artery pressure ≤ 15 mmHg
6. Pulmonary arteriolar resistance < 4 U/m^2
7. Ratio of pulmonary artery diameter-to-aorta diameter > 0.75
8. Left ventricular ejection fraction $\geq 60\%$
9. Competent mitral valve
10. No adverse effect from prior pulmonary artery surgery

The key objective of these early criteria and subsequent modifications of the Fontan procedure was to maximize laminar pulmonary blood flow in the absence of the pulsatility provided by a subpulmonary ventricle. Many of these criteria still are used today, including a low PVR. The fourth factor became irrelevant when contemporary techniques were used to either bypass the right atrium altogether (the extracardiac conduit Fontan procedure [ECC]) or only used a portion of the atrial wall to fashion an intra-atrial conduit (lateral tunnel) to the pulmonary artery.

Smaller pulmonary artery size may not have as much impact as previously believed in the short term,[19] but the ideal pulmonary artery size remains elusive.[20] Pulmonary artery flow and growth also are influenced by pulmonary vessel distensibility. Better flow is promoted by compliant vessels. The overall lack of pulsatility diminishes the shear stresses on the vasculature, predisposing to progressive weakening of the involved vascular tree.[21]

The technical choice also might influence pulmonary blood flow. One computer modeling study by Bove et al