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Fontan's operation: evolution from a procedure to a process

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It was in 1971, Fontan and Baudet[1] published their experience using a surgical procedure to restore a physiologic circulation in three patients with tricuspid atresia. Of the three patients, two survived, thus ushering a new era in the treatment of children with complex forms of congenital cardiac disease. This operation itself, however, was the culmination of a series of experimental and clinical observations that underlined the clinical possibility totally to bypass the right side of the heart.

History

As long ago as 1943, Starr et al.,[2] in a series of experiments in dogs, were able to show that damage to the pumping capability of the right ventricle did not result in venous congestion and hypertension. Then, in 1949, Rodbard et al.[3] demonstrated, using acute and chronic experiments, that venous pressure was sufficient to provide flow to the lungs "without benefit of the pumping action of the right ventricle". Shortly thereafter, in 1950, Carlon et al.[4] published their experience with a canine preparation in which a direct end-to-end anastomosis was made between the divided azygous vein and the right pulmonary artery, the superior caval vein having been ligated as it entered the right atrium. They speculated briefly as to the possible utility of this operation in some of the cyanotic forms of congenital cardiac disease.

In 1958, Glenn[5] published his first clinical experience with the superior cavopulmonary anastomosis, which became known as the "Glenn shunt". He had performed an end-to-side anastomosis of the right pulmonary artery to the superior caval vein, with ligation of the proximal superior caval vein as it entered the right atrium. He speculated on the applicability of this intervention to "a number of congenital malformations of the heart, not previously amenable to adequate treatment". Furthermore, he correctly pointed out that "it is believed to be essential" that the vessels be of adequate size, and that the resistance to flow of the pulmonary circulation be normal or decreased. In the decade that followed, the "classic" Glenn shunt was widely applied, with good results, to a variety of patients having cyanotic forms of congenital cardiac disease, predominantly those with functionally single ventricles. The physiologic importance of this intervention was not lost on researchers and clinicians, who understood that the shunt increased the flow of blood to the lungs, and thereby systemic saturation, without imposing an increased volume load on the functionally single ventricle.[6] Over time, nonetheless, the limitations of this procedure became apparent and further modifications were explored. In 1961, Dogliotti et al.[7] published their report on 11 patients who underwent a cavopulmonary anastomosis for treatment of cyanotic cardiac disease, predominantly tetralogy of Fallot. Almost all of the patients had either end-to-end or end-to-side anastomoses between the superior caval vein and right pulmonary artery, but one patient had a " 'double' end-to-side anastomosis" of both sides of the right pulmonary artery, with ligation of the superior caval vein as it entered the right atrium. This allowed flow into both lungs from the superior caval vein. Dogliotti et al.[7] also made a passing reference to the experiments carried out in Russia, by Bakulev, in the attempts "completely to exclude the right heart" by connecting the superior caval vein to the right pulmonary artery and the inferior caval vein to the left pulmonary artery.

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As discussed above, it was in 1971, that Fontan and Baudet[1] first reported clinically successful "complete bypass of the right heart" in patients with tricuspid atresia. This was accomplished by the combination of a "classic cavopulmonary anastomosis" between the superior caval vein and right pulmonary artery, and channeling the inferior caval venous flow into the left pulmonary artery by using the proximal end of the right pulmonary artery and right atrium as a conduit, with interposition of an aortic valvar homograft. In light of the more current understanding of the operation, it is interesting to read their speculation as to why their intervention was successful. They suggested that the pump function of a "hypertrophied right atrium, as in tricuspid atresia", coupled with valvar homografts "inserted into the inferior vena cava", and the other at the "exit of the right atrium" to the left lung, provided the necessary "additional work represented by a pulmonary arterial pressure higher than the left atrial pressure".

The pace of growth and modifications to the original surgical procedure continued to increase, and subsequently other authors published the results of their modifications and experience of the application of Fontan's operation to patients with tricuspid atresia. In 1973, Kreutzer et al.[8] reported their modification using a direct anastomosis of the pulmonary valve to the right atrial appendage, without the concomitant use of a cavopulmonary anastomosis or a valve in the inferior caval vein. In 1979, Bjork et al.[9] published their experience creating a direct and valveless communication between the right atrium and the right ventricle.

As experience accumulated with the operation and its variants, it also was applied to other forms of congenital cardiac disease where the anatomy made a biventricular repair impossible. In 1976, Yacoub et al.[10] reported their experience in patients with "single ventricle and pulmonary stenosis". In the early 1980s, Norwood et al.[11] reported their experience in children with aortic and mitral atresia physiologically "corrected" using the Fontan procedure as part of their surgical management.

When the Fontan procedure was initially described, Fontan et al. established 10 criterions for selection of patients, which became known as the "Ten Commandments". With time, it became apparent that many of these "commandments" were not true requirements for a successful procedure. Although initially predicated for patients older than 4 years, we now know that it can be done successfully on patients aged 2 years, or even younger. Ideally, the patient can walk or at least crawl. While sinus rhythm is ideal, atrioventricular sequential pacing can be employed if sinus rhythm is not present. Normal caval venous drainage is not necessary, as many creative surgical approaches have now been developed to successfully utilize the Fontan process in patients with abnormalities of venous return. An enlarged right atrium can be managed with a reduction atrioplasty. Elevated pulmonary arterial pressure may be acceptable in states of high flow of blood to the lungs. The crucial issue is pulmonary vascular resistance. Impairing effects from previous shunts typically do not prohibit the Fontan procedure, and the pulmonary arteries can be rehabilitated either surgically or in the catheterization laboratory prior to the completion Fontan. Thus, in the final analysis, the three crucial issues that remain from these 10 commandments are low pulmonary vascular resistance, satisfactory systemic ventricular function, and competence of the atrioventricular valve(s).

It did not take long to appreciate that the physiologic and clinical impact of the Fontan procedure, as a single event, was important and associated with considerable morbidity and mortality. Mair et al.[12] published the early experience at the Mayo clinic for the years 1973–1979, and documented a significant early and late mortality. By this time the basic physiologic principles necessary for successful outcomes were becoming clearer, specifically acceptable function of the "systemic" ventricle and inlet valve(s) with avoidance of chronic volume overload, widely patent pathways and surgical anastomoses, and physiologic and anatomic preservation of the pulmonary vasculature.

Unfortunately, in a variety of patients, these basic requirements were not always "perfectly" available, and the risk of Fontan's operation remained high. This stimulated further refinements of the operative technique itself, such as the pioneering experimental work undertaken by de Leval et al.[13] on the hydrodynamics of flow that spawned the concept of the total cavopulmonary anastomosis. In addition, it also brought the increasing recognition that a staged or sequential approach would ultimately improve morbidity and mortality. DeLeon et al.[14] reported as early as 1983 that a previous Glenn shunt reduced the morbidity and mortality of the Fontan procedure. As experience accumulated, further reports confirmed the value of a staged approach to the treatment of the patients with functionally univentricular hearts. Whether the preferred approach is by the use of the bidirectional Glenn Anastomosis,[15] or the hemi-Fontan operation,[16] with subsequent construction of a fenestrated lateral tunnel,[17] or an extracardiac conduit with or without a fenestration,[18] the evolution of Fontan's operation from a single intervention into a staged, sequential process, has become widely accepted. Creation of the Fontan circulation, therefore, now represents the most common, if not universal, approach[19] to patients with functionally univentricular hearts, regardless of their anatomy.

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Current approach

The most important consideration in planning the clinical management of a patient unlikely to ever be a candidate for a biventricular repair is preservation of the architecture and function of the pulmonary vascular bed, with control of volume and pressure overload of the systemic ventricle.

At The Congenital Heart Institute of Florida, our preferred initial palliation, in those patients with decreased pulmonary blood flow in the neonatal period, is to construct a modified Blalock–Taussig shunt, reconstructing the confluence of the pulmonary arteries as needed. In those patients with increased pulmonary blood flow, and/or obstruction to systemic outlet, our preference is to create a Damus–Kaye–Stanzel anastomosis with a modified Blalock–Taussig shunt as the initial palliation. We rarely band the pulmonary trunk to prevent excessive flow to the lungs, although we may use this approach in patients with an unbalanced atrioventricular septal defect, where it is difficult to determine the long-term possibility of a biventricular repair. In recognition of the benefits of a staged approach, our next intervention is a bidirectional Glenn anastomosis, again reconstructing the pulmonary arteries if needed. This is accomplished at 4–6 months of age. We tend not to suppress additional sources of flow to the lungs unless they are judged to likely result in significant elevation of the superior caval venous pressure postoperatively, or if effusive complications cannot be controlled postoperatively with relatively simple measures. In cases of significant postoperative cyanosis, we tend to search aggressively for important venous decompressing collaterals, embolizing them as needed.

Our current preference is to complete the Fontan circulation at about 2 years of age, utilizing an extracardiac conduit with a fenestration. This intervention is preceded by documentation of acceptable pulmonary vascular resistance and architecture of the pulmonary vascular bed, as well as normal or near normal ventricular and atrioventricular valvar function. If needed, significant aortopulmonary collaterals are embolized, and relief is provided for stenosis of either the pulmonary arteries or aortic arch by balloon dilation, particularly in the patients with a previous history of surgery to the aortic arch. If the fenestration does not spontaneously close after creation of the Fontan circulation, we often proceed to close it with a device about 1 year after the operation.

Outcomes

As experience over the short- and intermediate-terms continues to document improving outcomes,[20] the long-term follow-up of patients with the Fontan circulation continues to illustrate the physiologic impact of persistently elevated systemic venous pressures. Irrespective of whether the circulation has been created with a "classical Fontan" or the total cavopulmonary anastomosis, the long-term sequels increase over time. In a statistical study published by Fontan et al.[21] in 1990, the results after a "perfect" procedure predicted a survival of 73% 15 years after the surgery. Stamm et al.,[22] in 2001, reported their 10-year outcomes of 220 patients in whom they constructed a lateral tunnel with or without fenestration. There were 12 early and seven late deaths, four takedown operations, and four transplantations, with an estimated survival of 91% at 10 years, and freedom from failure of 87% at 10 years.

Most commonly, complications associated with late failure include atrial arrhythmia, thromboembolism, protein losing enteropathy, and late ventricular failure. Estimates of atrial arrhythmias vary from 10% to 40%,[19] most commonly manifesting as sinus nodal dysfunction. A significant population, which typically has undergone variants of the atriopulmonary connections, can suffer from disabling atrial re-entrant tachycardia. In those patients, conversion to a total cavopulmonary connection, coupled with surgical attack on the arrhythmic pathways, can result in significant improvement.[23]

Asymptomatic formation of thrombus in the Fontan circuit has been documented in as high as one-third of patients examined by transesophageal echocardiography.[24] Recognizing this phenomenon, most patients are routinely anticoagulated with aspirin. If the patient has been documented to have thrombus, or is at higher risk for thromboembolism because of arrhythmia, ventricular dysfunction, and borderline hemodynamics, or has suffered thromboembolic complications in the past, warfarin is then used. In addition, if possible, potential portals for right-to-left embolization are eliminated.

Significant protein-losing enteropathy and late ventricular failure remain as important problems, with generally poor outcomes. The physiology of protein-losing enteropathy is poorly understood, albeit that the incidence is likely to be low, in less than one-twentieth of patients, as suggested from an international multicentric study.[25] In those patients that are affected, it is associated with significant morbidity and mortality. Therapeutic interventions including pacing, glucocorticoid and heparin administration, and/or fenestration of the Fontan circuit have been effective in some. Mortality rates as high as 50–60%, nonetheless, have been recorded in the more seriously affected cases. Unfortunately, transplantation, as "rescue therapy" for the "failing Fontan" is associated with significant mortality,[26] leading some authors to recommend transplantation as an alternative only in patients deemed to be at very high risk.[27]

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At The Congenital Heart Institute of Florida, the largest programme for congenital cardiac surgery in Florida, we performed 237 Fontan procedures in the 15-year interval between October 1990 and September 2005, with an overall 30-day mortality of 5.1%. During the first 5 years of this time period, our 30-day mortality was higher, at 8.3%, but this 30-day mortality after the Fontan procedure has now decreased to less than 5% in the modern era. Our current mortality is consistent with national data. The congenital database of the Society of Thoracic Surgeons reports a mortality of 3.5% at discharge in 374 patients undergoing construction of a total cavopulmonary connection in 2002 and 2003.[28] We believe that our evolution in approach to the creation of the Fontan circulation from a single operation to an overall process is responsible for our decrease in mortality.

Summary

In patients with a functionally single ventricle, creation of the Fontan circulation remains the single best available option for physiologic palliation over the long-term. A staged approach, with strategies for preservation of normal or near normal pulmonary architecture, pulmonary vascular resistance, and avoidance of volume loading of the systemic ventricle, remains the most common and universal therapeutic option. Unfortunately, long-term problems are recognized, and are at times are difficult effectively to treat. Therapeutic strategies, nevertheless, continue to evolve and develop. Although it is sincerely hoped that in time creation of the Fontan circulation will be remembered as a footnote in the long history of medicine, for the moment, we must continue to understand and modify the chronic, deleterious effect of long-term elevation of systemic venous pressure.

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