Evolution of the Fontan Procedure: Early and Late Results

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This paper reviews the evolution of surgical technique that has occurred with the Fontan procedure since it was first introduced more than 25 years ago. Although there has been recent enthusiasm at some centers for a return to Fontan's original concept of use of a conduit to achieve the Fontan pathway, we continue to believe that the lateral tunnel with double cavopulmonary anastomosis is the preferred approach. The late incidence of arrhythmias with the lateral tunnel at 10 years follow-up is remarkably low. On the other hand conduits present a risk of outgrowth and pseudointima accumulation. Even small gradients, e.g. less than 4 mm, will be poorly tolerated over the longer term and may result in an increased incidence of cirrhosis and protein losing enteropathy.

Overall there has been a remarkable improvement in the early and late results of the Fontan procedure over the last decade. The role of the bi-directional Glenn shunt as either a staging procedure or definitive palliation when performed in conjunction with supplementary pulmonary blood flow needs to be defined by a prospective randomized study. Likewise the role of the fenestration also needs to be defined by a prospective randomized study including careful studies of late exercise capacity and maximal oxygen consumption. Another issue that needs to be defined by careful prospective randomized study is the importance of anti-coagulation with coumadin versus aspirin in reducing the incidence of thromboembolism.

Introduction

Since its introduction more than 25 years ago, a number of important modifications have been made to the Fontan procedure. These modifications can be thought of as being variations in the technique of anatomical connection of the systemic venous circulation to the pulmonary arteries and variations in the staging of the Fontan procedure.

Technical Modifications

Anastomotic Technique

Fontan's original procedure consisted of a classic Glenn shunt to the right lung and placement of a valved homograft conduit from the right atrium to the left pulmonary artery. Problems were soon noted with anterior compression of the homograft conduit as well as calcification and obstruction of the conduit. Kreutzer's modification of the posterior atriopulmonary anastomosis suffered from the disadvantage of potential damage to the sinus node artery. These problems led us and others¹,² to the concept of a double cavopulmonary anastomosis which remains our preferred anastomotic technique of the Fontan procedure.

Intra-Atrial Baffle Technique

In patients with left AV valve atresia or stenosis, most commonly those with hypoplastic left heart syndrome following a stage-1 Norwood procedure, pulmonary venous blood must be directed to the tricuspid valve. Early attempts at placing an oblique baffle from the ASD to tricuspid valve

frequently resulted in the development of pulmonary venous obstruction. This led us and others to the concept of a lateral cavopulmonary tunnel which we have applied since 1986. With this design it is unlikely that pulmonary venous obstruction will develop. de Leval et al. have also demonstrated that this design results in less turbulence and therefore less energy loss.

**Extracardiac Fontan**

An important late complication of the Fontan procedure has been atrial flutter. Although early indications suggest that the lateral tunnel will reduce the incidence of atrial flutter relative to previous techniques, some groups have taken an aggressive position in the hope of reducing this important late complication. Giannico et al. have recommended the placement of an extracardiac conduit from the divided IVC to the pulmonary artery. Thus, no atrial tissue is exposed to the high systemic venous pressure and the suture load within the atria is minimized. An important disadvantage of this approach is the absence of growth of the conduit. Furthermore, kinking and accumulation of pseudointima may result in gradients which are poorly tolerated on the venous side of the circulation. To address the issue of growth potential, Gundry et al. have suggested the use of an extracardiac pathway using the patient’s pericardium in situ. Although this technique reduces the intra-atrial suture load, nevertheless there is considerable suturing in the region of the sinus node and the pathway is quite irregular. Also there is a risk of compression of the right pulmonary veins. Careful follow-up will be necessary of both these alternative procedures before abandoning the more traditional intra-atrial baffle.

**Staging of the Fontan Procedure**

Fontan and Choussat in their original article recommended that the Fontan procedure not be performed until the child was at least four years of age. However this exposed the single ventricle to volume loading for at least four years because of the inherent inefficiency of the circulation when the pulmonary and systemic vascular beds are in parallel, as is the case in any child with a banded or shunted single ventricle or even with a balanced degree of pulmonary stenosis. However, attempts to perform the Fontan procedure early in infancy were accompanied by a higher risk of mortality and complications such as pleural effusions. Introduction of the bi-directional Glenn shunt as a staging procedure resulted in reduced mortality and morbidity for the second stage of palliation, particularly in patients with complex forms of single ventricle as hypoplastic left heart syndrome.

The fenestrated Fontan procedure was introduced in 1989 by Bridges et al. as a further stage towards the completed Fontan procedure (Fig. 1). Creation of a fenestration in the intra-atrial baffle allows right to left decompression and prevents excessive abdominal venous hypertension. It also allows cardiac output to increase at times when pulmonary vascular resistance is increased. The degree of cyanosis which results from this right to left shunt is generally well tolerated by these children who have been chronically cyanosed. Introduction of the fenestrated Fontan reduced the incidence of prolonged pleural effusions at Children’s Hospital in Boston from 40 to 10% (defined as need for chest tubes for greater than 1 week postoperatively).

**Early and Late Results**

A retrospective review of the first 500 patients who underwent the Fontan procedure at Boston Children’s Hospital between 1973 and 1991 by Gentles et al. revealed that the incidence of early failure had decreased from 27% in the first quartile to 7% in the last quartile. The probability of early failure was increased by a mean pulmonary artery pressure of 19 mm or greater, younger age at operation, heterotaxy syndrome, a right sided tricuspid valve as the only systemic AV valve, pulmonary artery distortion, an atrio pulmonary rather than cavopulmonary connection, absence of a baffle fenestration and longer bypass time. An increased probability of late failure was associated
with the presence of a pacemaker before the Fontan procedure. A morphologically left ventricle with normally related greater arteries or a single right ventricle (excluding heterotaxy and hypoplastic left heart syndrome) were associated with a decreased probability of late failure.

**Need for Prospective Randomized Studies**

It remains unclear whether all children should undergo an intermediate bi-directional Glenn shunt procedure prior to their Fontan. It is the current practice at Children’s Hospital in Boston to perform the bi-directional Glenn shunt only in situations where there is particularly complex anatomy, for example, status post stage-1 Norwood procedure or where a child is becoming excessively cyanosed by 4 to 6 months of age. If a child is relatively well balanced and can achieve 9 to 10 months of age, then a bi-directional Glenn shunt is avoided and a 1-stage Fontan procedure is performed. It is our current belief that all children should have a fenestration placed in order to reduce the risk of prolonged hospitalization secondary to pleural effusions. Only through randomized trials will it be possible to define the need for and optimal timing of a bi-directional Glenn shunt as well as the need for and timing of closure of a fenestration for patients undergoing the Fontan procedure.

**Conclusion**

There have been remarkable improvements in the outlook for the child with a single ventricle
over the last 25 years. Further improvements are likely to result with carefully designed large prospective clinical trials.

References


