



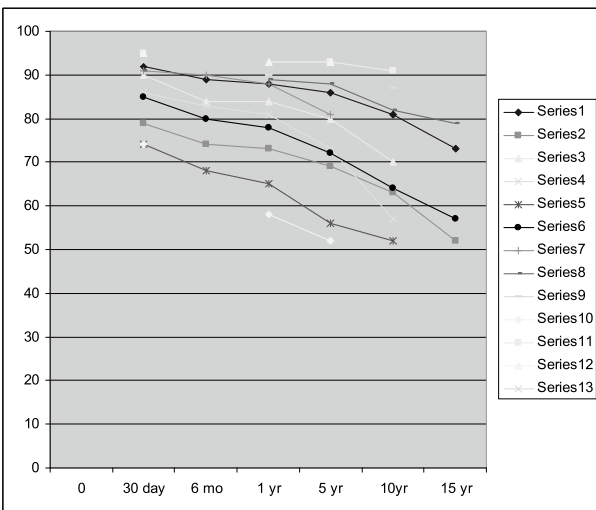
基調講演

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In 1971, Fontan and Baudet described a technique for definitive palliation of patients with tricuspid atresia. Since the original operation, the technique has evolved and changed. Changes have included elimination of implanted valves, staging of the operation with a bidirectional superior vena caval anastomosis, use of a fenestration, the, and the intraatrial and the extracardiac tunnel modifications

Survival

Initial operative survival (< 30 days) was approximately 80-85% in early reported series but has increased to as high as 95 % in more recent series. The Kaplan-Meier survival curves demonstrate difference in survival reported by a number of investigators. Survival at 10 years ranges from 50-90% and at 15 years from 50-80% .



Choussat and Fontan recommended 10 criteria for a successful low risk operation.

Table 1: Criteria of Choussat and Fontan

1. Age at operation 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 \leq 15 mm Hg
6. Pulmonary arteriolar resistance $<$ 4 u . m2
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 operation

These are relative criteria and it is clear that some are more important than others. Patients lacking one or more of these criteria can survive operation. However, the length of survival and quality of life, indeed, are related to many of these criteria. . Also, other investigators have identified additional important relative risk factors.

Table 2: Additional risk factors

Anomalous pulmonary venous connection	Left AV valve atresia	Non tricuspid atresia forms of single ventricle
Common AV valve	Ventricular hypertrophy	Early calendar year of op
Lack of fenestration	Non use of cardioplegia	Heterotaxia syndromes
None use of ultrafiltration	Longer ischemic time	Worse NYHA class
Preoperative diuretics	Valved RA-PA connection	Age > 18 years
Post operative renal failure	RA-PA connection with linear RA incision	
	Hypoplastic left ventricle	

Fenestration

In 1990, Bridges, described the concept and use of fenestration as part of the Fontan operation. In high-risk patients, the use of a fenestration improves survival. A fenestration reduces the duration of hospital stay, the duration and the volume of chest tube.

Arrhythmias

Gelatt reported incidences of atrial arrhythmia 5 years after a Fontan operation for patients who had an atrio-pulmonary connection, a total caval pulmonary connection, or right atrial to right ventricle connection as 33 % , 18 % , and 11 % respectively. Durongpisitkul reported incidences of atrial arrhythmias of 6 % , 12% , 17% , and 30% at 1, 3, 5 and 10 years after operation. The use of an extracardiac connection may be associated with a lower incidence of arrhythmias.

Protein Losing Enteropathy

Protein losing enteropathy has been reported to occur in 3-10% of patients after a non-fenestrated Fontan operation. In one study, the 5 year survival was 50 % after development of PLE. The cause of PLE in this setting is multifactorial. In 2 studies, PLE after the Fontan operation has been associated with longer cardiopulmonary bypass time, right ventricular forms of single ventricle, longer postoperative hospital stay and postoperative renal failure. The following treatment strategies have been employed to manage PLE: albumin infusion, heparin, steroids, somatostatin, and fenestration, cardiac pacing, Sildenafil, revision of the Fontan pathways, and cardiac transplantation. Each of these strategies may work in some but not in all patients. Infrequently, do they result in permanent resolution of the PLE. The use of sildenafil and oral budesonide has been effective in reducing protein lose in some patients with PLE. Thacker, et al (Ann Thorac Surg, 89:837, 2010) treated 9 patients with oral budesonide. Mean serum albumin levels increased from 1.9 g/dL to 2.9 g/dL. We have treated 7 patients with oral budesonide. Mean serum albumin levels increased from 2.7 g/dL to 3.3 g/dL. However, 5 patients had exacerbations of PLE while being treated with budesonide. In both studies, complications of Cushingoid features and infection were observed.

Thromboembolism

Occurrence of thrombosis after Fontan operation ranged from 2.2-19% depending upon the design of the study from which the data was abstracted.

Exercise Performance

Patients with functional single ventricle (prior to Fontan operation) have reduced aerobic capacity. In two different studies VO₂max was 20.5 and 23.1 ml/kg/min (Table 3). The major contributing factor to reduced aerobic capacity in these patients is the presence of a right to left shunt. There have been numerous studies of aerobic capacity after the Fontan operation. The values for VO₂max range from 15.9 to 28.5 ml/kg/min. Studies in which postoperative values were compared to preoperative values, have shown that there is a significant increase in postoperative VO₂ max

Quality of life

Malhle studied the outcome of 115 patients who had a Fontan operation for hypoplastic left heart syndrome. The parental assessment of the child's health and functional status was Excellent: 45%; Good: 35%; Fair: 12%; and Poor: 9%. The parental assessment of the child's school performance was: Above average: 42%; Average: 42%; and Below average: 17%. The parents' assessment did correlate with objective measures of cognitive function.

Pregnancy

We are aware of 71 pregnancies among 46 women. Maternal complications included atrial flutter/fibrillation in 7 patients, supraventricular tachycardia in 3 patients, and worsening NYHA classification in 6 and increased congestive heart failure in 2 patients. There were 39 live births, 8 therapeutic abortions, and 24 spontaneous miscarriages. One of the 39 live born children had an atrial septal defect. Although these results may seem remarkably good one must be suspicious that the women who became pregnant may have been patients who were doing very well. It probably is a mistake to extrapolate these results to all women who have had a Fontan operation.

Summary

The Fontan operation provides excellent long-term palliation for patients with a variety of functional single ventricle. Appropriate patient selection and selection of the appropriated surgical modification are crucial for insuring the best long-term outcome. Despite proper selection criteria protein losing enteropathy, arrhythmias, and ventricular failure remain therapeutic and management challenges.