Transplantation for Failed Fontan

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Last Major Surgical Operation before Heart Transplantation JAm Coll Cardiol 2009; 54:160-5

• 488 patients transplanted for CHD (CTRD & PHTS)



Fontan Failure

Bi-modal

- Early

Unsuspected risk factors

Intraoperative myocardial injury

- Late

Systemic-Ventricular Dysfunction Elevated PVR

- Lack of **pulsatility**
 - Pulmonary recurrent microemboli
 - Elevated pulmonary lymphatic pressure
 - Disordered NO synthesis/endothelial dysfunction
- Valve Dysfunction and Subvalvar Stenosis
- Lymphatic Derangement: PLE (3-15% incidence)

Plastic Bronchitis



Pulmonary AV Malformation

Thrombotic Circuit Occlusion (20% incidence, 25% mortality)

Intractable Arrhythmias

Outcome following "perfect" Fontan Circulation 1990; 81:1520-36



Indications for Heart Transplantation

Failing Fontan Physiology

- No obvious remediable lesions
- Poor ventricular Function

Timing

- Early
- Difficult due to disparate modes of failure
- Evidence of systemic mal-perfusion
- Lack of reliable mechanical assist methods

Transplant Evaluation

Analysis of anatomy

- Cardiac Situs
- Great vessels
- Location of pulmonary and systemic venous returns
- Fontan type
- Aortopulmonary collateral vessels
- **Patent vessels** for both hemodynamic monitoring and peripheral cannulation site.

Pulmonary Vascular Resistance in Failed Fontan

: Difficult to assess and often "unreliable"

- Systemic to pulmonary collateral arteries
- Low cardiac output
- Non pulsatile flow
- Insidious Pulmonary Emboli
- (Presence of pulmonary AV malformation)

Donor Procedure

- Knowledge of recipient anatomy is critical
- Vast majority of recipients require vascular reconstruction (PA, systemic or pulmonary veins)
- Retrieval of:
 - Aortic arch and head vessels
 - Branch pulmonary arteries to the hilum
 - SVC & innominate vein (for bilateral SVC)
 - IVC

Recipient Procedures

Safe re-sternotomy

- Multiple reoperation
- Anterior aorta
- Peripheral CPB may be necessary (> 40% of patients)
- Bi-caval venous anastomosis (less TR)

Bilateral SVC





Beiras-Fernandez et al. J heart Lung Transplant 2007; 26 : 290-2

Post - Transplant Survival



JAm Coll Cardiol 2009; 54:160-5 (data from CTRD & PHTS)



Circulation 2006; 114:273-280 (data from PHTS)



JAm Coll Cardiol 2004; 44:2065-72 (data from Philadelphia)

Causes of death after Transplantation

	Fontan Patients (n=23)	CHD (n= 47)	Non - CHD (n=85)
*Infection	7 (30)	10 (21)	11 (13)
*Early graft failure	4 (17)	5 (11)	7 (8)
Rejection	3 (13)	6 (13)	15 (18)
Sudden death	3 (13)	10 (21)	13 (15)
Graft CAD	2 (9)	0	10 (12)
*Hemorrhage/tech. /Op.	2 (9)	1 (2)	1 (1)

Circulation 2006; 114:273-280 (data from PHTS)

Risk factors of Death



Ann Thorac Surg 2009; 88:558-64 (data from Boston)



Circulation 2006; 114:273-280 (data from PHTS)

Summary

- Outcomes are acceptable.
- Transplantation for "failed Fontan" is technically challenging.
- Knowledge of recipient's cardiac morphology is very important.
- Measurement of pulmonary vascular resistance is unreliable.
- Clear understanding of its risks and pitfalls is necessary to maximize survival and quality of life among Fontan patients.