Surgery in the failing Fontan

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Fontan Types

Lateral Tunnel

Extracardiac
Normal vs. Fontan Circulation

A. Normal

LV

Lungs

RV

Body

P_{\text{Atrium}} = 5

P_{\text{PA}} = 15

P_{\text{IVC/SVC}} = 5

B. Fontan

SV

Lungs

Body

P_{\text{Atrium}} = 2

P_{\text{PA}} = 12

P_{\text{IVC/SVC}} = 15

Staged Fontan palliation
Fontan Failure

- Systolic vs diastolic failure
- Fontan = Severe Chronic Right Heart Failure
Major Long-Term Morbidity After the Fontan Operation

- Arrhythmia
- Thromboembolism
- Protein Losing Enteropathy
- Exercise Limitations
- Abnormal Health-Related Quality of Life
Fontan = Severe Chronic Right Heart Failure

- Venous hypertension
- Hepatic congestion $\rightarrow$ cirrhosis, acites
- Lymphatic congestion $\rightarrow$ PLE, PB
- Restrictive lung disease
- Altered pulmonary vasculature
- Single ventricle dysfunction
Post Fontan surgical options

- Repair of residual lesions
- Fontan Takedown
- Fontan Conversion
  - BiV Conversion
- Lymphatic Decompression
- Heart Transplant
  - Destination VAD
Residual lesions

• Unique anatomic and physiological impairments lead to myocardial dysfunction

  • Valves
  • Shunts
  • Flow obstruction
  • Electrical
Fontan Takedown to previous stage

- For early Fontan failure
- Low cardiac output/elevated CVP
  - Early post op
    - Early revision – if anatomic problem clearly established
    - ECMO – (primary arrhythmias, temporary ventricular dysfunction)
  - Immediate takedown to previous stage
Fontan Takedown to previous stage - outcome

Trezzi M et al, Bambino Gesù Hospital
Rome
Results:
n = 18, 1990 - 2015
  2 – immediate post op
  16 - within 2 months of Fontan
17 early survivors
3 underwent subsequent Fontan
4 underwent transplant
10 remained as BDG

Median O₂ saturation = 83% (76-89%)
Median follow-up = 7.0 years (0.9 – 26.5 years)

Long-Term Outcomes After Extra-Cardiac Fontan Takedown to an Intermediate Stage. STS 2017
Fontan Takedown to previous stage

- For late Fontan failure
- Hepatic congestion, protein losing enteropathy, variceal bleeding
- Poor outcome
Fontan Takedown to previous stage - outcome
Multi-institutional study

- n = 38, 1971 to 2012
- Fontan to takedown (0.6±1.9 years)
- Follow up (FU) of 6.7±6.7 years (0–23.7 years)
- Early mortality = 26.3% (10/15 patients)
- 5 Late deaths
- 4 patients (10.5%) underwent HTX after Fontan takedown After a mean period of 12.9±21.1 months, with two patients alive at the end of follow-up.
- 2 takedown patients (5.3%) underwent subsequent TCPC LT operation after a mean period of 34.1±14.1 months.

van Melle JP et al Heart 2016
• 17 patients (44.7%) had reached the primary end point mortality/HTX at the end of follow-up
Fontan conversion

• Who is a candidate for conversion?
  • Primarily an arrhythmia procedure
  • Preserved ventricular function
  • Preserved end-organ function

• Risk factors*
  • Protein losing enteropathy
  • Older age
  • Ascites
  • Right or indeterminate ventricular morphology
  • Bi-atrial arrhythmia operation

Fontan conversion

Backer CL et al Cardiol Young 2006
Fontan conversion

Backer CL et al Cardiol Young 2006
Fontan conversion

Backer CL et al Cardiol Young 2006
Fontan conversion
Fontan conversion – freedom from arrhythmias

The more complete the arrhythmia procedure the lower risk of recurrence

Fontan conversion - survival

Multi-center studies show early mortality of ~10%\(^2,3\)

3. van Melle JP et al Heart 2016
Fontan conversion

only extracardiac Fontan in Australia and New Zealand since 2007

d’Udekem Y et al Circ 2014
Biventricular conversion
Rationale: Why push the limits for 2V repair?

Long-Term Results of the Fontan Operation

D. J. Driscoll

Evaluating Failing Fontans for Heart Transplantation: Predictors of Death

Eric R. Griffiths, MD, Aditya K. Kaza, MD, Moritz C. Wyler von Ballmoos, MD, Hugo Loyola, BS, Anne Marie Valente, MD, Elizabeth D. Blume, MD, and Pedro del Nido, MD

The Failing Fontan: Options for Surgical Therapy

C. B. Huddleston
Biventricular Conversion After Single-Ventricle Palliation in Unbalanced Atrioventricular Canal Defects

Meena Nathan, MD, Hua Liu, MS, Frank A. Pigula, MD, Francis Fynn-Thompson, MD, Sitaram Emami, MD, Christopher A. Baird, MD, Gerald Marx, MD, John E. Mayer, MD, and Pedro J. del Nido, MD

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- N=16pts.
  - 9/16 left dominant;
  - 7/16 right dominant;

- 31ml/m² (22-35)
- RVEDV of 42ml/m² (26-86)

- All had previous SV palliation
  - 2-Stage I; 12 – BDG; 2 - Fontan
Hemodynamic parameters predict adverse outcomes following biventricular conversion with single-ventricle palliation takedown

Melissa A. Herrin, BA, David Zurakowski, PhD, Christopher W. Baird, MD, Puja Banka, MD, Jesse J. Esch, MD, MSc, Pedro J. del Nido, MD, and Sitaram M. Emani, MD
Heart transplantation

Extracardiac Fontan

SVC

IVC

Aorta

RA

LA cuff

SVC

Aorta

LA cuff

IVC
Heart transplantation

Sughimoto K et al Ann Thorac Surg 2015
Heart transplantation

SVC

IVC

Aorta
Liver Abnormalities and Post-Transplant Survival in Fontan Patients

Sophie C. Hofferberth, MBBS\textsuperscript{1}, Tajinder P Singh, MD\textsuperscript{2}, Elizabeth D. Blume, MD\textsuperscript{2}, Francis Fynn-Thompson, MD\textsuperscript{1}

30 Fontan pts with pre-transplant hepatic ultrasound

- Group 1 - Normal: N = 8
- Group 2 - Mildly abnormal (Hepatomegaly or vascular congestion): N = 14
- Group 3 - Severely abnormal (Heterogeneous echogenicity or nodularity): N = 8

Log-rank test p = 0.114

Time since transplantation (years)
Fontan patient

Source of Fontan failure – ventricular vs other
Failing ventricle – systolic dysfunction vs elevated EDP
Cardiac cath
Options for systemic VAD +/- takedown to BDG +/- using oxygenator
Schematic of the patient's Fontan circulation and biventricular assist device cannulation
Right sided VAD support

Right-Sided Univentricular Cardiac Assistance in a Failing Fontan Circulation

René Prêtre, MD, Achim Häussler, MD, Dominique Betcx, MD, and Michele Genoni, MD

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Failed Fontan physiology
- ↑ CVP
- Liver congestion

- TAH
  - OR
  - Takedown to Glenn + Continuous Flow LVAD

Both
- ↑ CVP
- ↑ EDP
- Pulmonary congestion
- Poor C.O.

- TAH
- Continuous flow VAD

Systemic Ventricle Failure
- ↑ EDP
- Poor C.O.

- Continuous flow VAD
Extra-cardiac Conduit Fontan → TAH
Excision of Native Heart and Fontan Conduit

- SVC
- Reconstructed PA
- Remnant of RA
- IVC
- Aorta
- LA with cuff of ventricular muscle
Anastomosis of Atrial Connector to LA
Creation of Neo-Right Atrium  
Insertion of Artificial Left Ventricle
Thoracic duct decompression

- The lymphatic circulation drains into the venous system
- Venous hypertension will result in lymphatic hypertension
- Lymphatic hypertension
  - Protein losing enteropathy
  - Plastic bronchitis
Thoracic duct decompression

Lymph capillaries converge to become collecting vessels and end up as either Thoracic duct or right lymphatic duct
Thoracic duct decompression

Hraska V Ann Thorac Surg 2013
Summary and conclusions

- Fontan conversion is an option for individuals with an aortopulmonary Fontan, arrhythmias and preserved function
  - The future role of Fontan conversion is unclear as the number of individuals with an aortopulmonary Fontan decreases
- Lymphatic decompression may provide palliation for PLE and PB with preserved function
- Better mechanical support strategies
- Ultimately transplant remains the best chance for long term survival