The Borderline 2 Ventricle Heart: When is a good Fontan better than a bad biventricular circulation?

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How to define “better”

Parents of newborns with congenital heart disease are appropriately focused on:
• long-term survival
• freedom from multiple operations
• quality of life

These are the quality indicators that should determine which surgical approach congenital teams take and not early mortality.
What is “quality of life”

• Exercise capacity/ VO$_2$ max
  – Systolic function
  – Diastolic function
  – Pulmonary and systemic vascular resistance

• Achieve optimal neurodevelopmental potential
• Early hospital procedural mortality is inherently misleading as a QUALITY INDICATOR of a congenital heart program and CANNOT be “risk-corrected”
Early hospital mortality as a Quality Measure is biased against 2 Ventricle repair

• Hospital A
  – not confident with complex 2 V repair
  – Usually chooses Fontan track over 2V
  – Example
    • 10 patients with DORV with remote VSD, no PS
    • 10 PA bands, 10 Bidirectional Glenns, 10 Fontans
    • 1 death, Hospital Mortality 3%

• Hospital B
  – Skilled at complex baffle repairs
  – Chooses 2V whenever possible
  – Example
    • 10 identical patients with DORV with remote VSD
    • 10 biventricular repairs
    • 1 death, Hospital Mortality 10%
Requirements of a Reliable Quality Improvement Tool for Congenital Heart Surgery

- Denominator must include all patients admitted to a program with structural heart disease
- End point must be at least one year survival
Our Mission

To build a comprehensive multidisciplinary institute focusing on surgical outcomes research, in order to analyze and improve outcomes of surgical therapies and their impact on patients, society (at local, national, and global levels), and the healthcare system.

Learn more about KIRSO
Advantages of World Society Database

• Designed as a Quality Improvement tool
• One year survival end point
• Free to members of World Society
• Compatible with Clinical Research tools like STS (no duplicate data entry)
• Global enrolment
  – Comparison with regional, national and global outcomes
  – Improved risk adjustment through size of database
Cumulative Surgeries and Hospital Enrollment

<table>
<thead>
<tr>
<th></th>
<th>Hospitals Enrolled</th>
<th>Surgeries Entered</th>
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<tr>
<td>Jan-17</td>
<td>7</td>
<td>29</td>
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<tr>
<td>Feb-17</td>
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<tr>
<td>Mar-17</td>
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<td>Apr-17</td>
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<tr>
<td>May-17</td>
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<tr>
<td>Jun-17</td>
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<td>Sep-17</td>
<td>26</td>
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We don’t know the long-term outcome for all *current* Fontan patients

- Reports from different centers contain highly variable numbers of high risk subgroups, e.g. HLHS, heterotaxy
Comparative freedom from failure (death, heart transplantation, reoperation on the Fontan circuit, poor functional status) for patients with and without hypoplastic left heart syndrome (HLHS; log-rank test P<0.001).

Log-rank test P<0.001
Why we don’t know the long-term outcome for all *current* Fontan patients

- Variable high risk subgroups, eg HLHS, heterotaxy
- Multiple technical modifications of the Fontan procedure since 1970
Kaplan–Meier Survival by Fontan type.

Log-rank test P<0.001
Fontan Generation I

Surgical repair of truncus arteriosus has been carried out in three patients. Two of these operations have been successful. A new surgical procedure has been used which transmits the whole venous blood to the lungs, while only oxygenated blood returns to the left heart. The right atrium is, in this way, “centralized,” to direct the inferior vena caval blood to the left lung. The pulmonary arteries receiving the superior vena caval blood through a cava-pulmonary anastomosis. This technique depends on the size of the pulmonary arteries which must be large enough to allow a cava-pulmonary anastomosis. The indications for this procedure apply only to children sufficiently well developed. Younger children or those whose pulmonary arteries are too small should be treated by palliative surgical procedures.

Only palliative operations (systemic veno to pulmonary arteries anastomosis, systemic arteries to pulmonary arteries anastomosis) have been performed in truncus arteriosus. Although these procedures are valuable, they result in only a partial clinical improvement, because they do not suppress the mixture of venous and oxygenated blood.

We have initiated a corrective procedure for truncus arteriosus, which completely suppresses blood mixing. The entire vena caval return goes arterialized in the lungs and only oxygenated blood comes back to the left heart. This procedure is not an anatomical correction, but would require the creation of a right atrium, but a procedure of physiological pulmonary artery restoration with suppression of mixing.

“Atriopulmonary Fontan”
Generation III

Extracardiac Conduit Fontan Procedure

Atrial incision avoids
- Sinus node
- Crista terminalis
- Sinus node artery
The “Intra/Extracardiac Conduit” Fenestrated Fontan

Ring-supported Goretex conduit

Goretex suture tacks atrial wall to conduit

Fenestration in short intra-atrial segment
The “Intra/Extracardiac Conduit” Fenestrated Fontan
Intra/extracardiac fenestrated modification leads to lower incidence of arrhythmias after the Fontan operation.


N=134

**Conclusion:** Intra/extracardiac conduit with limited atriotomy Fontan modification associated with a significantly lower incidence of abnormal rhythm compared with lateral tunnel modification (odds ratio, 0.28; 95% confidence interval, 0.10-0.84; P = .015).
The Borderline 2 Ventricle Heart

- **Borderline RIGHT HEART**
  - Pulmonary atresia with intact ventricular septum
  - Tricuspid valve stenosis
  - Unbalanced AV canal

- **Borderline LEFT HEART**
  - Shone’s syndrome
  - Unbalanced AV canal
  - Corrected TGA with unprepared LV
Pulmonary Atresia with Intact Septum

- **Borderline RV/TV** can be managed with:
  - Single ventricle track
  - Biventricular repair OR intermediate forms of repair

*(Coronary anatomy (RV dependence) may mandate single ventricle track)*
Intermediate forms of “repair” for PA/IVS, no RVDCC

- 1.5V, no ASD  
  very small RV and TV
- 1.5V, fenestrated  
  smaller RV and TV
- 1.25V  
  smallest RV and TV but open RVOT
The One and Half Ventricle Repair

Bidirectional Glenn Shunt

ASD closure with or without fenestration
The One and a Quarter Ventricle Repair


Surgical alternatives to the Fontan procedure incorporating a hypoplastic right ventricle.

Gentles TL, Keane JF, Jonas RA, Marx GE, Mayer JE Jr

Additional cavopulmonary anastomosis
Long-term functional health status and exercise test variables for patients with pulmonary atresia with intact ventricular septum: a Congenital Heart Surgeons Society study


1987 and 1997
448 neonates with PAIVS
multi-institutional study

Late exercise capacity and health status were assessed following repair (mean 14 years). Relationships between health status, exercise capacity, morphology, and 3 end states (i.e., 2V, single V or 1.5-ventricle repair) were evaluated.
Long-term functional health status and exercise test variables for patients with pulmonary atresia with intact ventricular septum: a Congenital Heart Surgeons Society study


For those with smaller initial tricuspid valve z-score, achievement of survival with BV repair may be at a cost of late deficits in exercise capacity, emphasizing that better outcomes may be achieved for borderline patients with a 1.5V or Single Ventricle repair strategy.
Single V track may achieve better longterm functional outcomes for some patients with borderline LEFT HEART.
Postnatal *left ventricular diastolic function* after fetal aortic valvuloplasty


Echocardiographic evidence of LV diastolic dysfunction is common in patients with biventricular circulation after fetal aortic valvotomy and persists in short-term follow-up. **LV diastolic dysfunction** in this unique population may have important implications on long-term risk of left atrial and subsequent pulmonary hypertension.
Neonatal Assessment and Decision: Single V (Norwood) vs 2 Ventricle (Aortic Valvotomy) for Critical Neonatal Aortic Valve Stenosis

• Duct closed
  – LV is supporting circulation alone
  – Hemodynamics are meaningful
    ✓ Cardiac index
    ✓ MV gradient
    ✓ LVEDP
    ✓ LVOT gradient
**Neonatal Assessment and Decision:**
Single V (Norwood) vs 2 Ventricle (Aortic valvotomy) for Critical Neonatal Aortic Valve Stenosis

- **Duct dependent**
  - Hemodynamics meaningless
  - Use morphology and CHSS calculator

[www.chssdc.org](http://www.chssdc.org)
Critical Aortic Stenosis in the Neonate
Independent Factors Predictive of Survival Benefit

Independent Factors Associated with Survival Benefit [Norwood – Two Ventricle Repair] at 5 Years

<table>
<thead>
<tr>
<th>Factor</th>
<th>p</th>
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<tbody>
<tr>
<td>Higher grade of EFE</td>
<td>.0001</td>
</tr>
<tr>
<td>Lower z score AV at sinuses</td>
<td>.0001</td>
</tr>
<tr>
<td>Younger age at entry</td>
<td>.0001</td>
</tr>
<tr>
<td>Larger AscAo diameter</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Absence of mod/severe TR</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Lower z score of LV length</td>
<td>.002</td>
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*J Thorac Cardiovasc Surg 2001; 121:10-27*
Grade of Endocardial Fibroelastosis ("EFE") from Echo Review
N=320
1994-2000

0  none
1  involvement of papillary muscles only
2  papillary muscle with some endocardial surface involvement
3  extensive surface involvement
Primary *left ventricular rehabilitation* is effective in maintaining two-ventricle physiology in the borderline left heart


In patients with borderline left hearts, primary left ventricular rehabilitation with endocardial fibroelastosis resection and mitral and aortic valvuloplasty results in improved left ventricular systolic and diastolic performance and decreased right ventricular pressures. This approach may provide an alternative to single-ventricle management in this difficult patient group.
Single V track may be better for:

- Inadequate ventricle (e.g., small LV with aortic valve stenosis, Shone’s syndrome, small RV with PA/IVS)
- Congenitally corrected transposition
- Unbalanced AV canal
Late Survival after Traditional Surgery for Corrected TGA
N=123, 1963-1996

Fontan or septation
Double Switch

Senning Plus Arterial Switch
Senning Procedure
Arterial Switch Component of Double Switch
“Double Switch”
Mustard Plus Rastelli
Mustard Procedure
Mustard/Rastelli Procedure
Anatomic repair for congenitally corrected transposition of the great arteries: a single-institution 19-year experience


N=113 1991-2011

- Double-switch (DS) 68
- Rastelli–Senning (RS)-type 45
- Pulmonary artery banding for retraining 23
Need for Reintervention After Double Switch

- High incidence with no difference at 10 years between the two groups (DS, 50.3%; RS, 49.1%; p = 0.44).
- In the DS group, Lecompte maneuver was associated with late reinterventions on the pulmonary arteries.
- Surprisingly high incidence of aortic regurgitation among the true double-switch patients, including need for aortic valve replacement.
- High incidence of reoperation also for late baffle obstruction and pacemaker placement.

Murtaza et al 2011
Late LV Dysfunction after Double Switch

• High incidence not predicted by age at retraining.

• Quinn et al. (also Birmingham group) previously emphasized the important risk of late LV dysfunction after pulmonary artery banding in an earlier report in 2008.

• Similar concerns about late LV dysfunction expressed by Bautista-Hernandez et al. from Boston who found that pacemaker implantation rather than pulmonary artery banding was a risk factor for ventricular dysfunction after a double switch.
Double Switch Procedures for L-TGA

- Late results troubling in several reports
  - *Late LV dysfunction*
    - Murtaza et al 2011
    - Quinn et al 2008
    - Bautista-Hernandez 2006
  - *High rate of reoperation*
    - Murtaza et al 2011
  - *Aortic regurgitation*
    - Murtaza et al 2011
Anatomic Factors Complicating a Double Switch or Senning/Rastelli for L-TGA

- Dextrocardia, particularly for Senning/Rastelli (conduit compression, small atrium/difficult exposure)
- Anomalous coronary (conduit anastomosis)
- Predominant inlet rather than conoventricular VSD
Pulmonary Artery Band as Destination Therapy


Single V track may be better for:

- Inadequate ventricle (e.g., small LV with aortic valve stenosis, Shone’s syndrome, small RV with PA/IVS)
- Inadequate atrioventricular valve (e.g., tricuspid stenosis with PA/IVS)
- Congenitally corrected transposition
- **Unbalanced AV canal**
Unbalanced AV Canal

• In favor of 2V repair:
  – Down syndrome

• In favor of single V track:
  – <40% of common AV valve overlies LV
  – Straddling chords

Multi-institutional CHSS study in progress
Conclusions

• Many patients with complex congenital heart disease require a difficult decision between a biventricular versus a single ventricle approach.

• Early hospital survival is not proof that a biventricular approach was preferable.

• Late exercise testing (VO₂ max) will be the key to improving outcomes for marginal patients.