Hybrid Alternatives To Norwood Stage-1 May Not Be A Lower Risk Alternative

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American Heart Association
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Congenital Heart Surgeons’ Society: Critical Left Ventricular Outflow Tract Obstruction

Critical LVOTO
N = 692

Admission to CHSS institution
2005 – 2013
21 institutions
≤ 30 days
Ductal dependent circulation
Congenital Heart Surgeons’ Society: Critical Left Ventricular Outflow Tract Obstruction

Critical LVOTO
N = 692

- Ross-Konno n=3
- Yasui n=5
- Aortic Valvotomy n=107
- HTX n=5
- Single Ventricle Repair n=564
## Single Ventricle Repair

\( n = 564 \)

<table>
<thead>
<tr>
<th>Method</th>
<th>Percentage</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>BT</td>
<td>41%</td>
<td>232</td>
</tr>
<tr>
<td>SANO</td>
<td>39%</td>
<td>222</td>
</tr>
<tr>
<td>HYBRID</td>
<td>20%</td>
<td>110</td>
</tr>
</tbody>
</table>
Single Ventricle Repair
n = 564

BT (41%)  
n = 232

SANO (39%)  
n = 222

HYBRID (20%)  
n = 110

Norwood Operation:
Modified BT Shunt
Single Ventricle Repair
n = 564

- BT (41%)  
  n = 232

- SANO (39%)  
  n = 222

- HYBRID (20%)  
  n = 110

Norwood Operation:
SANO Shunt
## Single Ventricle Repair

### HYBRID

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Number (Percentage)</th>
</tr>
</thead>
<tbody>
<tr>
<td>BT (41%)</td>
<td>232 n = 232</td>
</tr>
<tr>
<td>SANO (39%)</td>
<td>222 n = 222</td>
</tr>
<tr>
<td>HYBRID (20%)</td>
<td>110 n = 110</td>
</tr>
</tbody>
</table>

- **Bilateral PA Band***: 110 (100%)
- **Ductal stenting**: 76 (68%)
- **PDA patency w/ PgE**: 35 (32%)
- **Atrial septostomy/stent**: 11 (10%)
- **Reverse BT shunt**: 18 (16%)

*4 Children with PA banding in 2 stages*
Is the HYBRID strategy a low risk alternative to Norwood Stage-1?
1) *Risk-adjusted* survival analysis:
   All 564 Children
1) *Risk-adjusted* survival analysis:  
All 564 Children

2) *Propensity matched* survival analysis:  
Subset of “comparable” children
1) Risk-adjusted survival: Competing Events
Competing Events:

**Alive without definitive operation**

% Proportion in each end-state

Years after procedure
Competing Events:
1V to 2V Conversation

Alive without definitive palliation

% Proportion in each end-state

Years after procedure
Competing Events:

**Transplant**

- Alive without definitive palliation

% Proportion in each end-state

Years after procedure

1V to 2V

8% 8% 8%

1%
Competing Events:

**Fontan**

- Alive without definitive palliation

Graph showing percent proportion in each end-state over years after procedure:
- Fontan
- Transplant
- 1V to 2V
- 8% Transplant, 8% Transplant
- 49% Fontan
Competing Events:

- Died without definitive palliation
- Alive without definitive palliation
- Died without definitive palliation
- 1V to 2V
- Fontan
- Transplant

% Proportion in each end-state

Years after procedure
Competing Events:

- Died without definitive palliation
- Alive without definitive palliation
- Died without definitive palliation
- 1V to 2V
- Fontan
- Transplant

% Proportion in each end-state

Years after procedure

- 49%
- 34%
- 8%
- 1%
Unadjusted Survival:
All 564 Children

<table>
<thead>
<tr>
<th>Years after procedure</th>
<th>% Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>100%</td>
</tr>
<tr>
<td>1</td>
<td>88%</td>
</tr>
<tr>
<td>2</td>
<td>80%</td>
</tr>
<tr>
<td>3</td>
<td>68%</td>
</tr>
<tr>
<td>4</td>
<td>66%</td>
</tr>
</tbody>
</table>

(323) (270) (179) (102)
## Risk factors for death: Multivariable

<table>
<thead>
<tr>
<th>Risk factor</th>
<th>P</th>
<th>Reliability</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low birth weight</td>
<td>&lt;.0001</td>
<td>64%</td>
</tr>
<tr>
<td>Small LVOT z-score</td>
<td>&lt;.0001</td>
<td>88%</td>
</tr>
<tr>
<td>Small MV annulus z-score</td>
<td>.065</td>
<td>41%</td>
</tr>
<tr>
<td>BT Shunt</td>
<td>&lt;.0001</td>
<td>72%</td>
</tr>
<tr>
<td>HYBRID</td>
<td>.003</td>
<td>72%</td>
</tr>
</tbody>
</table>
Risk-Adjusted Survival: By Initial Strategy (n = 564)

- **SANO**: 77%
- **HYBRID**: 62%
- **BT**: 60%

**Factors**:
- Low birth weight
- Small LVOT
- Small mitral valve annulus

Years after procedure
Risk-Adjusted Survival: Vs. Birth Weight (n = 564)
Risk-Adjusted Survival: 
Vs. *Birth Weight*  (n = 564)
Survival versus Birth Weight:
By Initial Strategy (n = 564)

Birth weight (kg)
Median (range):
SANO = 3.1 (1.8 - 4.4)
BT = 3.2 (1.6 - 4.5)
HYBRID = 3.0 (1.0 - 4.4)
Risk-adjusted survival

SANO was associated with improved 4-year survival

Low birth weight was a strong predictor of mortality

HYBRID procedures mitigate some risk associated with low birth weight
2) Propensity Matched Survival: “Comparable” children
Propensity Matched Children:

HYBRID with BT

HYBRID=110
100%

BT=232
100%
Propensity Matched Children: HYBRID with BT
Propensity Matched Children:
HYBRID with BT

- HYBRID 25%
- BT 65%

Matched

- 82 (75%)
- 82 (35%)
Propensity Matched Children: HYBRID with BT

HYBRID: Unmatched (n=28)

Matched

C-statistic = .77

HYBRID
25%

BT: Unmatched (n=150)

BT
65%

82 (75%)

82 (35%)
<table>
<thead>
<tr>
<th>Variable</th>
<th>HYBRID</th>
<th>Value</th>
<th>BT</th>
<th>Value</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at operation (d)</td>
<td>8 ± 7</td>
<td>9 ± 7</td>
<td>.32</td>
<td></td>
<td></td>
</tr>
<tr>
<td>BSA at operation</td>
<td>.2 ± .02</td>
<td>.2 ± .02</td>
<td>.44</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Branch PA z-score</td>
<td>-.4 ± 1.7</td>
<td>-.8 ± 1.6</td>
<td>.24</td>
<td></td>
<td></td>
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<tr>
<td>PDA (cm)</td>
<td>0.67 ± 0.47</td>
<td>0.73 ± 0.83</td>
<td>.95</td>
<td></td>
<td></td>
</tr>
<tr>
<td>RV Index</td>
<td>6 ± 2</td>
<td>6 ± 2</td>
<td>.7</td>
<td></td>
<td></td>
</tr>
<tr>
<td>TV z-score</td>
<td>-1.1 ± 1.9</td>
<td>-1 ± 2.1</td>
<td>.74</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aortic valve sinus (cm)</td>
<td>.46 ± 0.2</td>
<td>.47 ± 0.2</td>
<td>.76</td>
<td></td>
<td></td>
</tr>
<tr>
<td>MV atresia</td>
<td>33%</td>
<td>28%</td>
<td>.51</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal MV morphology</td>
<td>20%</td>
<td>27%</td>
<td>.27</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Small/Restrictive ASD</td>
<td>51%</td>
<td>57%</td>
<td>.51</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aortic arch hypoplasia</td>
<td>94%</td>
<td>93%</td>
<td>.79</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal TV</td>
<td>90%</td>
<td>92%</td>
<td>.72</td>
<td></td>
<td></td>
</tr>
<tr>
<td>LV Dysfunction</td>
<td>56%</td>
<td>68%</td>
<td>.23</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aortic valve atresia</td>
<td>37%</td>
<td>45%</td>
<td>.42</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Survival in Propensity Matched Children:

HYBRID with BT (n = 164)

% Survival

Years after procedure

p = 0.13
Propensity Matched Children: HYBRID with SANO

Matched

C-statistic = .75

HYBRID: Unmatched (n=28)

SANO: Unmatched (n=140)

82 (37%)

82 (75%)

SANO 63%
<table>
<thead>
<tr>
<th>Variable</th>
<th>HYBRID</th>
<th>SANO</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at operation (d)</td>
<td>8 ± 6</td>
<td>7 ± 5</td>
<td>.56</td>
</tr>
<tr>
<td>BSA at operation</td>
<td>0.2 ± 0.02</td>
<td>0.2 ± 0.02</td>
<td>.75</td>
</tr>
<tr>
<td>MV z-score</td>
<td>-15 ± 8</td>
<td>-16 ± 8</td>
<td>.37</td>
</tr>
<tr>
<td>Normal MV morphology</td>
<td>24%</td>
<td>29%</td>
<td>.48</td>
</tr>
<tr>
<td>Aortic valve atresia</td>
<td>46%</td>
<td>44%</td>
<td>.75</td>
</tr>
<tr>
<td>Aortic arch hypoplasia</td>
<td>91%</td>
<td>91%</td>
<td>1</td>
</tr>
</tbody>
</table>
Survival in Propensity Matched Children:

**HYBRID** with **SANO** (n = 164)

<table>
<thead>
<tr>
<th>Years after procedure</th>
<th>HYBRID</th>
<th>SANO</th>
</tr>
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<tbody>
<tr>
<td>0</td>
<td>100%</td>
<td>100%</td>
</tr>
<tr>
<td>1</td>
<td>65%</td>
<td>77%</td>
</tr>
<tr>
<td>2</td>
<td>65%</td>
<td>77%</td>
</tr>
<tr>
<td>3</td>
<td>65%</td>
<td>77%</td>
</tr>
<tr>
<td>4</td>
<td>65%</td>
<td>77%</td>
</tr>
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</table>

p = 0.015
Summary

In neonates with critical LVOTO who underwent initial 1-V palliation:

Norwood operation with SANO is associated with improved survival prior to Fontan completion:

Risk-adjusted survival
Survival in propensity matched children
Summary

In neonates with critical LVOTO who underwent initial 1-V palliation:

Birth weight has a strong association with poor survival:

**HYBRID** strategies appear to mitigate risk associated with very low birth weight
Clinical Implication

In children with similar baseline characteristics, HYBRID strategies to Norwood Stage-1 may not be a low risk alternative:

However, they may currently provide an advantage in neonates with low birth weight.

The full extent of potential advantages associated with HYBRID strategies remains to be determined.