Is it really better than **systemic-to-pulmonary shunt** for **right ventricular-to-pulmonary artery conduit** in stage I Norwood procedure of **HLHS**?

*Crossover with 4th WCPCCS in Buenos Aires*

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Dong-A University Hospital
Prologue

Reported by dr. Ohye
Ann thorac surg 2004;78:1090-3

Preop. Findings; prior to 2nd OP.

4-day-old, 2.6-kg, full-term, female neonate
- Shock to an outside emergency room
- HLHS with mitral & aortic atresia, an aberrant right subclavian artery, mildly depressed RV function, and TR(II~III)

Norwood procedure (with 3.5mm MBTS) on day of life 15

Uneventful recovery after surgery
- BP; 75 to 80/25 to 30 mm hg
- Po2; 30 to 35 mm hg
- Extubated on postoperative day 5

2 weeks after surgery
- Oxygen saturations ↓; 68% to 72% despite supplemental oxygen
Prologue

Conversion of MBTS to a 4.0-mm RVPAC at postoperative day 14

Operative findings
- Stenosis at the origin of the right carotid artery, possibly related to a snare injury.

Postoperative course
- Weaned from bypass without difficulty
- BP: 75 to 80/40 to 45 mm hg
- \( \text{Po}_2 \): 35 to 38 mm hg
Prologue

Limitation on this comparison

- Not enough data ⇐ relatively small number of patients
- No prospective randomized design clinical study
- Historical controls populations; The errors due to different surgical techniques according to the time of surgery in the patient and control study
Contents

- Introduction; SPA shunt & RV/PA conduit
- Comparison
  - Echocardiographic evaluation
  - Postoperative hemodynamics & surgical outcomes
  - Mid-term outcomes on subsequent palliation
In 1981, the 1st successful surgical palliation by Dr. Norwood
by 1999, Revived by Dr. Kishimoto
Since 2001, Popularized by Dr. Sano
SPA shunt & RV/PA conduit

Theoretical and observed advantages
- Higher postoperative diastolic blood pressure
- Improved coronary perfusion
- Balanced pulmonary and systemic circulations
- Avoidance of poor systemic perfusion?

Potential disadvantages
- Cardiac dysfunction due to ventriculotomy
- Ventricular arrhythmias
- Early or progressive hypoxemia $\leftarrow$ intimal hyperplasia and increased oxygen demand
- Volume overload $\leftarrow$ reversal flow
- Limited PA growth $\leftarrow$ low Qp/Qs, decreased in effective pulmonary blood flow

Most of the advantages or disadvantages are theoretical

No clear conclusion on the long and short-term clinical outcomes & inter-stage surgical results
Contents

- Introduction; SPA shunt & RV/PA conduit
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  - Mid-term outcomes on subsequent palliation
Echocardiographic evaluation

RV/PA conduit

SPA shunt
Echocardiographic evaluation

Ventricular function

- Reduced workload of the right ventricle because of
  - Less $q_p/q_s$
  - Improved coronary perfusion due to higher diastolic pressures.

J Thorac Cardiovasc Surg 2003;126:1378-84
Echocardiographic evaluation

Ventricular function

First to compare two surgical strategies during the convalescent phase after S1P for HLHS.

To quantify RV performance noninvasively with strain doppler echocardiography
Contents

- Introduction; SPA shunt & RV/PA conduit
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Postoperative hemodynamics & surgical outcomes

- Studies directly compared the two procedures in a patient group and control groups
- Common denominator among these studies
  - Higher mean diastolic pressure, narrower pulse pressure in RV/PA conduit group
  - Less mean pao2 (< 35 mmhg) and less sao2 (< 60-65%) in RV/PA conduit group → however, peripheral perfusion was good and metabolic requirements were satisfactory
Postoperative hemodynamics & surgical outcomes

RV/PA conduit > SPA shunt

- Ecco usage, delayed sternum closure, and postoperative ventilatory manipulation are seldom in RV/PA conduit group (p<0.01)
- Postoperative mortality: 9.3% in RV/PA conduit vs. 30% in SPA shunt (p=0.03)

Rudolf Mair, J thorac cardiovasc surg 2003;126:1378-84
- More excellent postoperative outcomes in RV/PA conduit (n=14) than SPA shunt (n=18)
- Postoperative mortality: 7% in RV/PA conduit vs. 38% in SPA shunt (p<0.05)
- 3.5 mm shunt in all SPA shunt pts (< 3.0 kg, n=7/18 pts)
Postoperative hemodynamics & surgical outcomes

Improvement of postop care

- **Routine use of mechanical ventricular assist**
  - Since January of 2001, 23 patients have been managed using this strategy
  - Increasing cardiac output during assist → increase cerebral oxygen delivery and metabolism → translate into improved neurologic outcome
  - Overall hospital survival - 87%

- **Practical use of alpha blockade strategy**
  - Irreversible α-1 and α-2 adrenergic receptor blocker
  - Optimization of systemic cardiac output by maximal dilation of systemic circulation
  - Diminish myocardial oxygen consumption and manipulate the Qs side of the Qp:Qs balance
Scott Bradley,

- Relatively large study population (19 vs. 25) with homogeneity
- Postoperative hemodynamic and pathophysiological conditions were not better in RV/PA conduit group
- The postoperative mortality; 11% (2/19) in the RV/PA conduit group 20% (5/20) in the SPA shunt group

Updated CHSS data;
Postoperative mortality; 6.1%
with phenoxybenzamin and home monitoring program
4th WCPCCS, Argentina
# Postoperative hemodynamics & surgical outcomes

## Summary of other studies

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<tbody>
<tr>
<td>Operation mortality (%)</td>
<td>35.5 vs. 11, p = 0.032</td>
<td>22.6 vs. 9.7, p = 0.04</td>
<td>12.5 vs. 7.7, Not difference</td>
<td>11 vs. 1.9, Not difference</td>
</tr>
<tr>
<td>Qp/Qs</td>
<td>1.41 vs. 0.68, p = 0.02</td>
<td>- -</td>
<td>1.8 vs. 0.9, p = 0.001</td>
<td>1.24 vs. 0.80, p = 0.011</td>
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<tr>
<td>Oxygen saturation (%)</td>
<td>75.1 vs. 67.38, p = 0.003</td>
<td>- -</td>
<td>76 vs. 77, Not difference</td>
<td>75.3 vs. 67.4, p &lt; 0.001</td>
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<tr>
<td>Mixed venous oxygen Saturation (%)</td>
<td>49.34 vs. 47.2</td>
<td>- -</td>
<td>46 vs. 47, Not difference</td>
<td>49.7 vs. 43.5, p = 0.024</td>
</tr>
<tr>
<td>Length of ICU stay</td>
<td>- -</td>
<td>- -</td>
<td>- -</td>
<td>Shorter in RV/PA conduit</td>
</tr>
</tbody>
</table>
Contents

- Introduction; SPA shunt & RV/PA conduit
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  - Mid-term outcomes on subsequent palliation
Mid-term outcomes on subsequent palliation

Overall survival on RV/PA conduit


- Multi-institutional study (n=73, RV/PA conduit only)
- Actuarial survival of the total patients: 65%, 63% at 1 & 2 year
- Each surgeon’s first 10 cases; 53% at 1 & 2 year
- Each surgeon’s recent cases; 71%, 68% at 1 & 2 year
Mid-term outcomes on subsequent palliation

David A. Ashburn, CHSS

*J thorac cardiovasc surg* 2003;125:1070-82.

- Between January 1994 and December 2000
- 710 neonates (total 985 enrolled pts.) Underwent the SPA shunt
- Multi-institutional (29), longitudinal study
- Statistically “boot strap bagging technique”
- Overall survival: 72%, 63%, 60%, 54% at 1 month, 6 months, 1 year, and 5 years.

![Total Survival Graph](image-url)
Mid-term outcomes on subsequent palliation

- William T. Mahle,
  

- The largest cohort of chop; 840 patients (1984~1999)

- The overall hospital mortality for stage I surgery - 36.3%


- Risk factor;
  - Earlier era
  - Age > 14 days at stage I
  - Weight < 2.5
**Mid-term outcomes on subsequent palliation**


**Table 1. Comparison of Hemodynamics Before Stage II and Stage III**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Before Stage II</th>
<th>Before Stage III</th>
<th>$P$ value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>BT (N = 5)</td>
<td>RV-PA (N = 25)</td>
<td></td>
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<tr>
<td></td>
<td>(mean value ± SD)</td>
<td>(mean value (Mann-Whitney’s Test)</td>
<td></td>
</tr>
<tr>
<td>RVEDP (mm Hg)</td>
<td>10 ± 5</td>
<td>6 ± 3</td>
<td>.12</td>
</tr>
<tr>
<td>Ao-Sat (%)</td>
<td>77 ± 4</td>
<td>72 ± 5</td>
<td>.126</td>
</tr>
<tr>
<td>Qp/Qs</td>
<td>0.88 ± 0.1</td>
<td>0.66 ± 0.2</td>
<td>.008*</td>
</tr>
<tr>
<td>RPA/m²</td>
<td>27 ± 7</td>
<td>21 ± 4</td>
<td>.196</td>
</tr>
<tr>
<td>LPA/m²</td>
<td>21 ± 8</td>
<td>20 ± 6</td>
<td>.785</td>
</tr>
<tr>
<td>RV/LT ratio</td>
<td>1.3 ± 0.2</td>
<td>1.1 ± 0.3</td>
<td>.083</td>
</tr>
<tr>
<td>Nakata index</td>
<td>335 ± 200</td>
<td>201 ± 86</td>
<td>.103</td>
</tr>
<tr>
<td>RVFS (%)</td>
<td>28 ± 2</td>
<td>35 ± 7</td>
<td>.018*</td>
</tr>
<tr>
<td>RVEDd (mm)</td>
<td>32 ± 5</td>
<td>26 ± 5</td>
<td>.0063</td>
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</table>

<table>
<thead>
<tr>
<th>Variable</th>
<th>SPA shunt (n=27)</th>
<th>RV/PA conduit (n=51)</th>
<th>$P$ value</th>
</tr>
</thead>
<tbody>
<tr>
<td>LPAI (mm²/m²)</td>
<td>49.63 ± 20.9</td>
<td>134.91 ± 48.1</td>
<td>0.006</td>
</tr>
<tr>
<td>RPAI (mm²/m²)</td>
<td>61.24 ± 5.6</td>
<td>182.29 ± 73.3</td>
<td>0.001</td>
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<tr>
<td>SVC-O₂ (%)</td>
<td>49.7 ± 7.3</td>
<td>43.5 ± 6.6</td>
<td>0.024</td>
</tr>
<tr>
<td>RV-O₂ (%)</td>
<td>75.3 ± 44</td>
<td>67.4 ± 66</td>
<td>&lt;0.001</td>
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<tr>
<td>Qp/Qs</td>
<td>124 ± 043</td>
<td>0.80 ± 0.47</td>
<td>0.011</td>
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<tr>
<td>RVH (n)</td>
<td>7/27</td>
<td>2/51</td>
<td>0.012</td>
</tr>
<tr>
<td>ICU stay (days)</td>
<td>15.0 ± 13.9</td>
<td>6.8 ± 11.0</td>
<td>0.004</td>
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</tbody>
</table>

**Abbreviations:** RVEDP, right ventricular end-diastolic pressure; Ao-Sat, aorta oxygen saturation; Qp/Qs, pulmonary-to-systemic blood flow ratio; RV/LT ratio, indexed right-to-left pulmonary artery diameter ratio; RPA/m², indexed right pulmonary artery diameter; LPA/m², indexed left pulmonary artery diameter; RVFS, right ventricular fractional shortening; RVEDd, right ventricular end-diastolic diameter.

*One of 4th WCPCCS, Argentina

Dr. Sano; *mPAP (mmHg) 15.2 vs. 12.1 (p=0.008)

*In angiogram, depressed wall motion was limited in the vicinity of ventriculotomy.
Mid-term outcomes on subsequent palliation

Theological etiologies of inter-stage mortality

- Coronary perfusion
- Shunt obstruction
- Arrhythmias
- Right ventricular failure
- Residual arch lesions
- Acute inter-current illness
- Atrioventricular valve regurgitation.
Mid-term outcomes on subsequent palliation

- David A. Ashburn, CHSS


- Only SPA shunt patients
- Inter-stage mortality: 12% (63/512 discharged pts.)
- Risk factor:
  - Lower birth weight
  - Older age at operation
  - Total circulatory arrest ↑
  - Ascending aorta dimension ↓

Updated CHSS data; Inter-stage mortality: 9.1%

4th WCPCCS, Argentina
Mid-term outcomes on subsequent palliation

Inter-stage mortality

Clifford L. Cua, *Ann Thorac Surg* 2005;80:44-9; meta-analysis

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<td>NW-BT</td>
<td>7</td>
<td>20</td>
<td>13</td>
<td>14</td>
<td>18</td>
<td>72</td>
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<tr>
<td>NW-RVPA</td>
<td>12</td>
<td>17</td>
<td>13</td>
<td>33</td>
<td>9</td>
<td>84</td>
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<td>Crossover (n)</td>
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<td>NW-RVPA</td>
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<tr>
<td>Mortality (n)</td>
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<td>NW-BT</td>
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<td>14</td>
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<td>NW-RVPA</td>
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<td>1</td>
<td>0</td>
<td>2</td>
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<td>Mortality (%)</td>
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<tr>
<td>NW-BT</td>
<td>14.3</td>
<td>20.0</td>
<td>23.1</td>
<td>14.3</td>
<td>22.2</td>
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<td>NW-RVPA</td>
<td>8.3</td>
<td>0</td>
<td>0</td>
<td>3.0</td>
<td>0</td>
<td>2.4</td>
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<td>Mortality with crossover (%)</td>
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<tr>
<td>NW-RVPA</td>
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<td>Nonsurgical candidate/transplant</td>
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<td>Awaiting stage II (n)</td>
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<td>0</td>
<td>NR</td>
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<td>3^b</td>
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<td>0</td>
<td>NR</td>
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<td>NW-BT</td>
<td>Median</td>
<td>NR</td>
<td>NR</td>
<td>3.1</td>
<td>~6</td>
<td>6.3</td>
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<tr>
<td>NW-RVPA</td>
<td>Median</td>
<td>NR</td>
<td>NR</td>
<td>3.4</td>
<td>~6</td>
<td>6.3</td>
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<td>Completed stage II (n)</td>
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<td>18</td>
<td>10</td>
<td>12</td>
<td>NR</td>
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<td>NW-RVPA</td>
<td>7</td>
<td>12</td>
<td>13</td>
<td>32</td>
<td>NR</td>
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</table>
Mid-term outcomes on subsequent palliation

- **Cua et al. Suggested**
  - The larger conduits used for the NW-RVPA, less likely to occlude
  - RV/PA conduit procedure could be the higher diastolic blood pressure

- **Study's own limitation**
  - Overall improvement in quality of care for patients cannot be excluded
  - Two cohorts were from different time periods
  - Etiologies of inter-stage deaths were not documented → so they could only speculate on causes.

Mid-term outcomes on subsequent palliation

Ghnayem’s report s – Home surveillance

- All discharged patient with SPA shunt (not RV/PA conduit)
- Daily log of weight and arterial oxygen saturation at home
- Contact their physician
  - Arterial oxygen saturation < 70%
  - Acute weight loss > 30 g in 24 hours
  - Failure to gain at least 20 g during a 3-day period.

Home surveillance program prevents interstage mortality after the Norwood procedure

N. S. Ghanayem, MD
G. M. Hoffman, MD
K. A. Mussatto, BSN
J. R. Cava, MD
P. C. Frommelt, MD
N. A. Rodi, MSHP
M. M. Stettiar, MSN
S. M. Benvando, BSN
S. J. Frisina, MS
R. D. B. Joquiss, MD
S. B. Linnis, MD
J. S. Tweedell, MD

Objective: To determine whether early identification of physiologic variables associated with intussucception would reduce mortality, we developed a home surveillance program.
Mid-term outcomes on subsequent palliation

“Hawthorne effect” ; described by Elton Mayo of the Harvard Business School ;

- The overt enlistment of the family in the pre-stage II home monitoring care of their child → increased the sophistication level of postoperative and predischarge teaching.
- Make the family feel more empowered to contribute positively to the care of their child.
- Minirevolution with a SPA procedure of Norwood op.

- Inter-stage mortality ; 15.8% (n = 9/57) without the home surveillance and 0% (n = 0/24) with home surveillance (P = .039)
  → This result emphasized the scrupulous follow-up of the discharged patients.

- HLHS who has undergone S1P appears to have limited growth potential with a plateau phase of weight gain after 150 days
  → This poor growth potential after 4 to 5 months calls into question the value of routinely delaying S2P beyond 5 months of age.
Mid-term outcomes on subsequent palliation

**Ventricular performance**

- Yoshihisa Tanoue,
  *Ann thorac surg 2004;78:1965-71.*

- Between April 1992 and August 2003
- SPA shunt group (n=11) & RV/PA conduit (n=10)
- Comparing the ventricular performance after BCPS & TCPC in HLHS patients
- Operative survival; SPA shunt - 78.3%, RV/PA conduit - 79.5%
- Inter-stage mortality; SPA shunt - 27.8%, RV/PA conduit - 19.4%
- Ees (contractility) & Ea; RV/PA conduit group
- No difference in ventricular efficiency (Ea/Ees), SW/PVA in both group
- Only SPA shunt has been performed until 1998 → historical control group
# Mid-term outcomes on subsequent palliation

## Summary of other studies

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<tbody>
<tr>
<td>Mortality before stage II operation</td>
<td>0.14 / 0.03</td>
<td>18 / 0</td>
<td>0.25 / 0.15</td>
<td>23 / 0</td>
<td>---</td>
<td>?? /1.9</td>
</tr>
<tr>
<td>Dr. Pizarro ; 0.13(6/46) .vs. 0.02 (1/50) (p &lt;0.01)</td>
<td>---</td>
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<td>79% (1 year, total pts.)</td>
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</tr>
<tr>
<td>PA index</td>
<td>---</td>
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<tr>
<td>Remarks</td>
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<td>---</td>
<td>---</td>
<td>dp/dt is better in RV/PA conduit</td>
</tr>
</tbody>
</table>

Combined PA diameter ; larger in RV/PA conduit $p=0.05$ 
LPA stenosis in SPA shunt 
Both PA ; larger in RV/PA conduit $P<0.05$
Summary

- Theoretical advantages of new strategies may not produce true benefit. (Extracardiac Fontan, Continuous cerebral perfusion, Hybrid stage I, RV/PA conduit)
- RV/PA conduit procedure shows stable hemodynamics in early postoperative period.
- No significant difference was found in mid-term survival between two procedures.
- Patient related risk factors would be more significant than shunt type in overall outcomes.
- Newer strategies must be evaluated over time by rigid analysis and data collection, and (ideally) contemporaneous control groups.
MBT Shunt is the Best Option for Norwood Stage I

Is the Best Option =

Is not the Best Option =
4th WCPCCS in Argentina

MBT shunt is the best option for Norwood stage 1 Palliation?