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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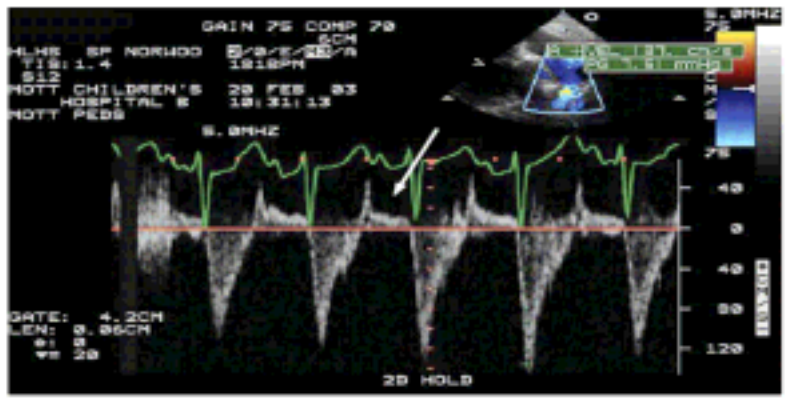
Prologue

A case report

- **Reported by dr. Ohye**
  Ann thorac surg 2004;78:1090–3
- **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
  - Po_2 ; 30 to 35 mm hg
  - Extubated on postoperative day 5
- **2 weeks after surgery**
  - Oxygen saturations ↓; 68% to 72% despite supplemental oxygen
- **Preoop. Findings** ; prior to 2nd OP.
  - Stenosis at the origin of the right carotid from the aorta
  - And a widely patent shunt



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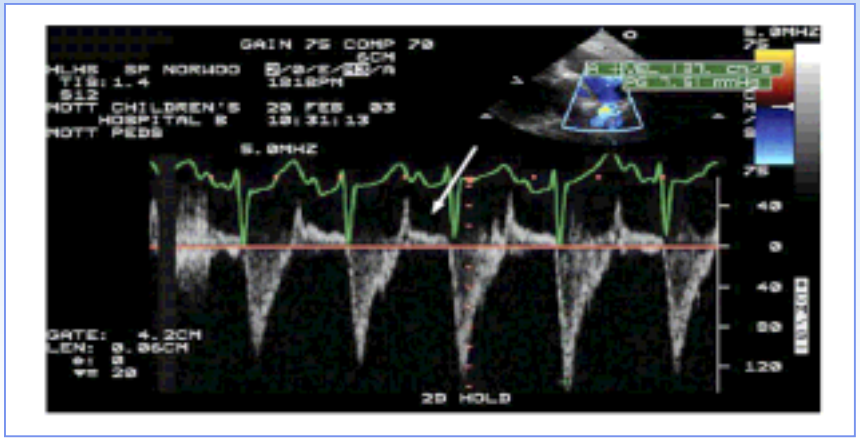
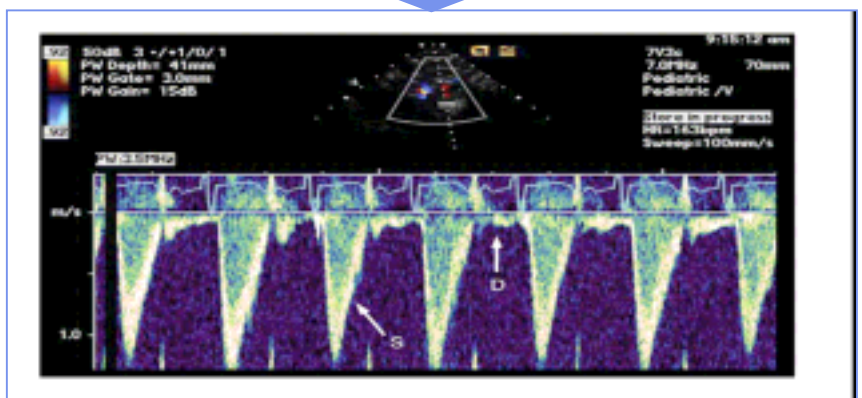
Prologue

A case report

- **Conversion of MBTS to a 4.0-mm RVPAC at postoperative day 14**
- **Operativefindings**
  - Stenosis at the origin of the rightcarotid artery, possibly related to a snare injury.
- **Postoperative course**
  - Weaned from bypass without difficulty
  - BP ; 75 to 80/40 to 45 mm hg
  - Po_2 ; 35 to 38 mm hg





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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

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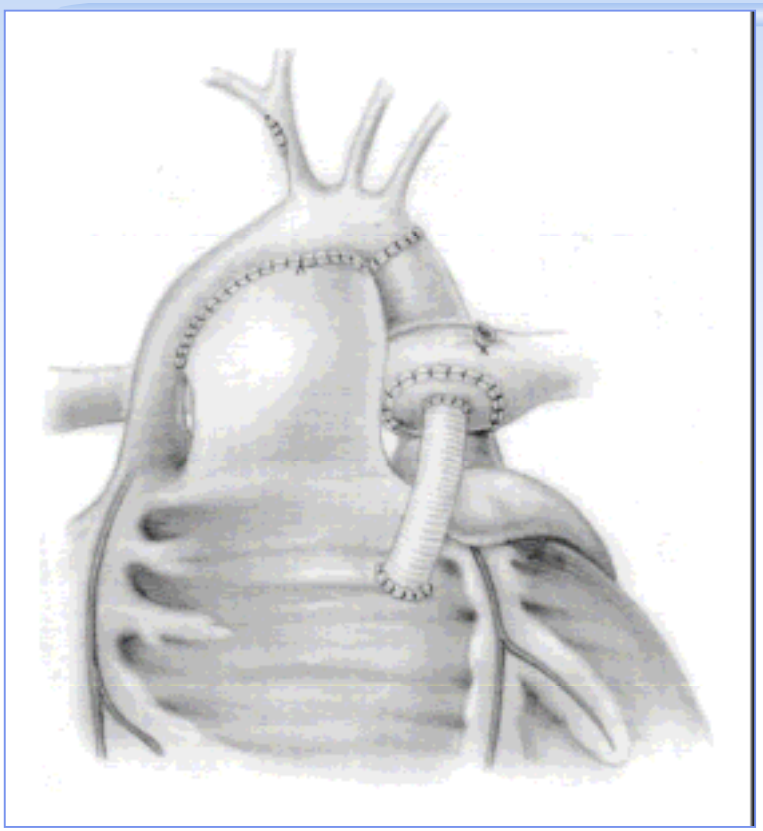
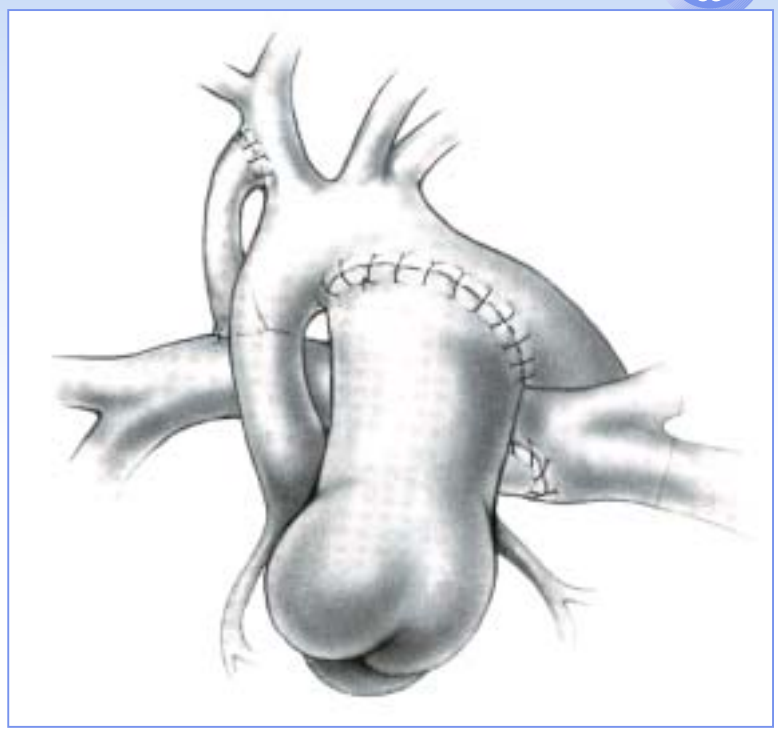
SPA shunt & RV/PA conduit

History





In 1981, the 1st successful surgical palliation by *Dr. Norwood*

In 1999, Revived by *Dr. Kishimoto*

Since 2001, Popularized by *Dr. Sano*

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SPA shunt & RV/PA conduit

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 ← intimal hyperplasia and increased oxygen demand
  - Volume overload ← reversal flow
  - Limited PA growth ← 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**

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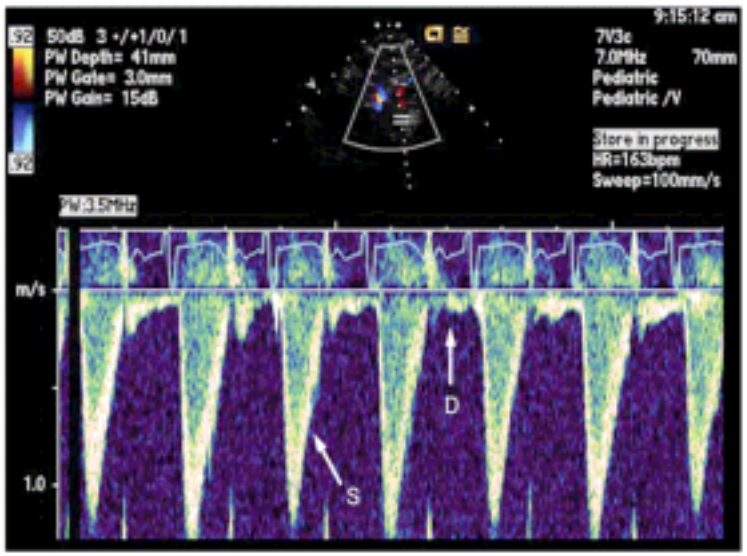
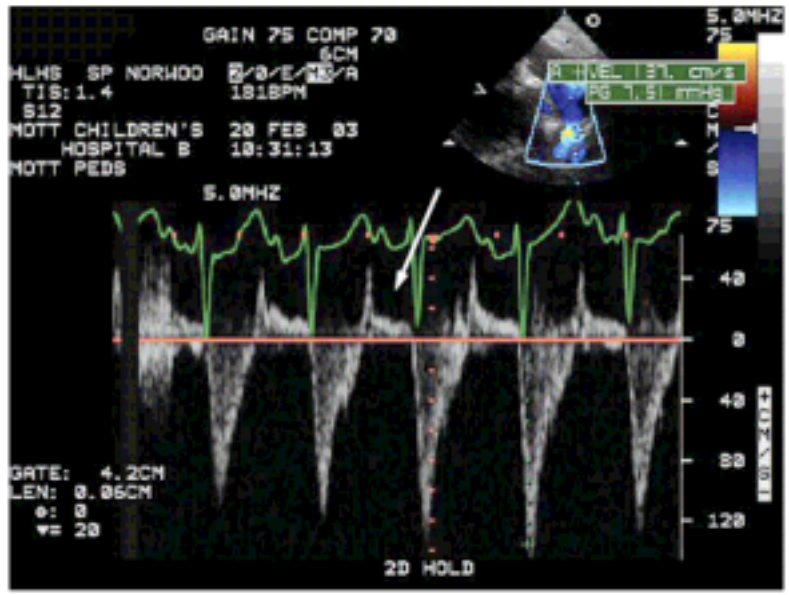
Echocardigraphic evaluation



RV/PA conduit



SPA shunt

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Echocardigraphic evaluation

Ventricular function

	Group 1 (BT shunt): Median [quartiles]	Group 2 (RV-PA conduit): Median [quartiles]	*P* value
Saturation SVC (%)	51 [43;53]	49 [43;53]	.682
Saturation aorta (%)	81 [78;86]	74 [72;79]	.036
PAP (mm Hg)	14 [14;17]	13 [12;17]	.415
Qp/Qs	1.6 [1.2;1.6]	0.9[0.8;1]	.005
EDP	12 [10;16]	11 [8;14]	.381
dp/dt (mm Hg/s^2)	776 [615;907]	955 [773;1110]	.018

BT, Blalock-Taussig; *dp,dt*, rate of pressure rise; *EDP*, end-diastolic pressure; *PA*, pulmonary artery; *PAP*, pulmonary artery pressure; *Qp/Qs*, pulmonary/systemic flow ratio; *RV*, right ventricular; *SVC*, superior vena cava.

J Thorac Cardiovasc Surg 2003;126:1378-84

- **Reduced workload of the right ventricle because of**
  - Less qp/qs
  - Improved coronary perfusion due to higher diastolic pressures.

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Echocardigraphic evaluation

Ventricular function

Table 2 Clinical and echocardiographic characteristics of each patient listed in chronological order of surgery

Patient	Echo post-op (days)	Heart rate (beats/min)	SaO_2 (%)	Inotrope (μg/kg/min)	TR	RV %A-Ch (%)	Peak systolic SR (1/s)	Peak systolic ε (%)	Outcome
S-PA shunt									
1	50	149	80	None	Mod	29	−1.11	−16.2	A
2	47	167	79	None	Mod	25	−0.69	−12.2	A
3	41	136	80	None	Mild	29	−0.77	−11.9	D (60 days)
4	28	177	82	Dobutamine 5	Mild	17	−1.08	−13.4	D (29 days)
Mean (SD)	41.5 (3.4)	157 (18)	80 (1)			25 (6)	−0.91 (0.21)	−13.4 (2.0)	
RV-PA conduit									
5	34	147	85	None	Mild	49	−1.54	−20.4	A
6	27	138	88	None	Mod	62	−1.06	−16.5	A
7	36	154	86	None	Mild	55	−1.18	−16.7	A
8	31	170	85	Dobutamine 2.5	Mild	53	−1.29	−16.5	A
9	32	133	82	None	Mild	62	−1.11	−19.0	A
Mean (SD)	32 (9.7)	148 (15)	85 (2)			56 (6)	−1.24 (0.19)	−17.8 (1.8)	
p Value	0.07	0.4	<0.01			<0.01	0.048	0.01	

ε, systolic strain; A, alive; D, died; mod, moderate; RV % A-Ch, right ventricle percentage area change; SaO_2, percutaneous oxygen saturation; SR, strain rate; TR, tricuspid valve regurgitation.

Heart 2004;90;191-194

- **First to compare two surgical strategies during the convalescent phase after S1P for HLHS.**
- **To quantify RV performance noninvasively with strain doppler echocardiography**

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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

- **Christian Pizarro, Circulation. 2003;108:ii-155.**
  - Recent results of two procedures (2000-2003).
  - Eccs 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, Jthorac 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**
  - **Ross ungerleider,** *pediatric cardiac surgery annual of the seminars in thoracic and cardiovascular surgery, vol 7, 2004: pp 16-21.*
  - 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**
  - **Glen S. Van arsdell,** *pediatric cardiac surgery annual of the seminars in thoracic and cardiovascular surgery, vol 7, 2004: pp 11-15.*
  - 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

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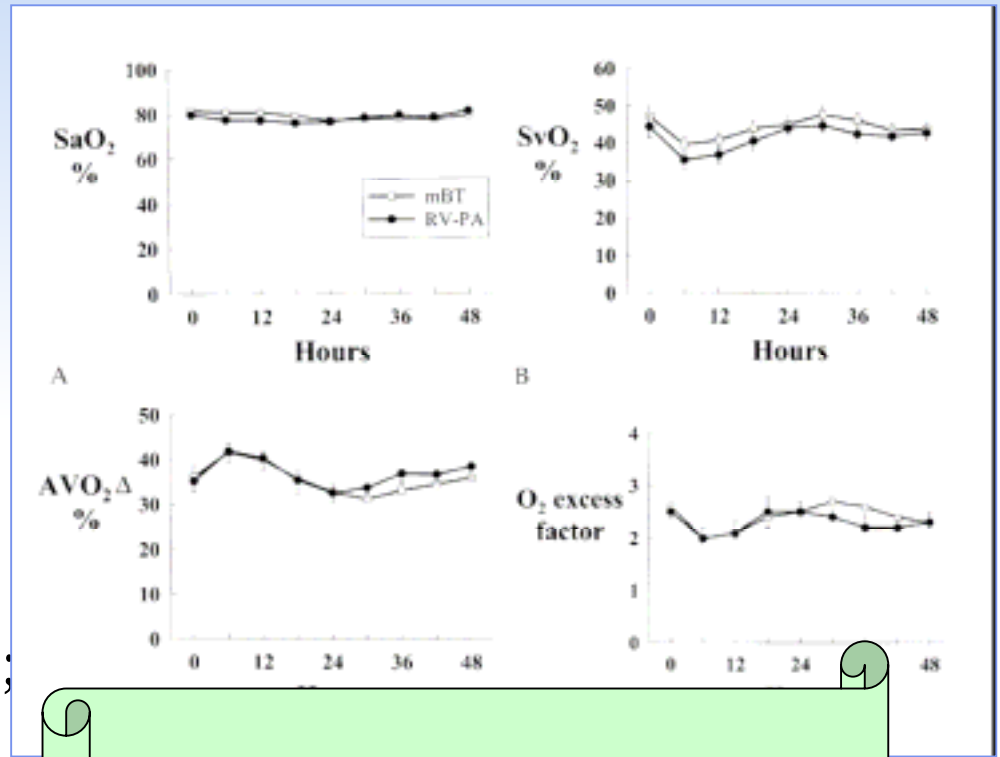
Postoperative hemodynamics & surgical outcomes

RV/PA conduit SPA shunt

- **Scott Bradley ,**

Ann thorac surg 2004;78:933- 41.

- 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

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Postoperative hemodynamics & surgical outcomes

Summary of other studies

Studies (SPA / RVPA)	Malec et al. (2003) (31/37)	Cua et al. (2005) –meta analysis (x 5) (72/84)	Azakie et al. (2004) (8/13)	Januszewska et al. (2005) (27/51)
Operation mortality (%)	35.5 .vs. 11 p =0.032	22.6 .vs. 9.7 p=0.04	12.5 .vs. 7.7 Not difference	11 .vs. 1.9 Not difference
Qp/Qs	1.41 .vs. 0.68 p=0.02	- - -	1.8.vs. 0.9 p=0.001	1.24 .vs. 0.80 p=0.011
Oxygen saturation (%)	75.1 .vs. 67.38 p =0.003	- - -	76 .vs. 77 Not difference	75.3 .vs. 67.4 p < 0.001
Mixed venous oxygen Saturation (%)	49.34 .vs. 47.2 Not difference	- - -	46 .vs. 47 Not difference	49.7 .vs. 43.5 p=0.024
Length of ICU stay Duration of ventilatory support	- - -	- - -	Not difference	Shorter in RV/PA conduit

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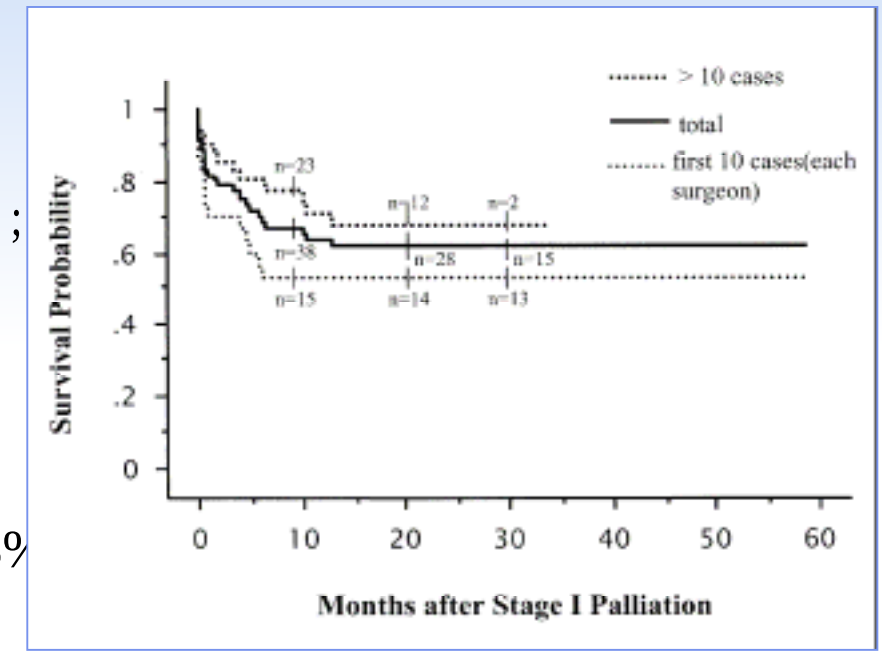
Mid-term outcomes on subsequent palliation

Overall survival on RV/PA conduit

- **Shunji Sano,** *Ann thorac surg 2004;78:1951- 8.*
  - 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



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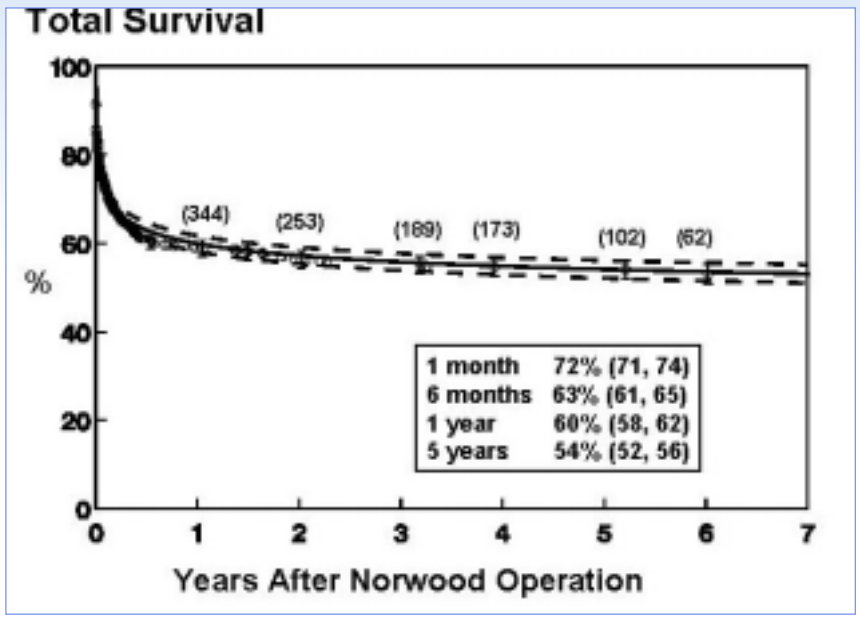
Mid-term outcomes on subsequent palliation

Overall survival on SPA shunt

- **David A. Ashburn, CHSS**

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

  - Between january 1994 and dece mber 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%, 6 0% , 54% at 1 month, 6 mont hs, 1 year, and 5 years.**



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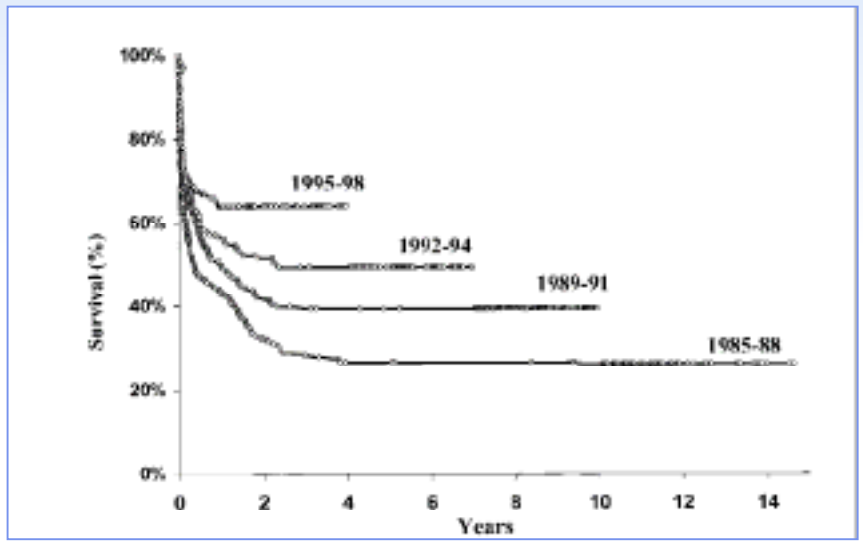
Mid-term outcomes on subsequent palliation

Overall survival on SPA shunt

- **William T. Mahle,**

  Circulation. 2000;102[suppl III]:III-136-III-141.

  - The largest cohort of chop ; 840 patients (1984 ~ 1999)
  - The overall hospital mortality for stage I surgery - 36.3%.
  - 3 year survival 66% (1995~1998) .vs. 28% (1984~1988)
  - Risk factor ;
    - Earlier era
    - Age > 14 days at stage I
    - Weight < 2.5



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Mid-term outcomes on subsequent palliation

Shunji Sano, *Pediatric Cardiac Surgery Annual of the Seminars in Thoracic and Cardiovascular Surgery, Vol 7, 2004: pp 22-31.*

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

Variable	Before Stage II: BT (N = 5) (mean value ± SD)	Before Stage II: RV-PA (N = 25*) (mean value ± SD)	Before Stage II: *P* value (Mann-Whitney's Test)	Before Stage III: BT (N = 5) (mean value ± SD)	Before Stage III: RV-PA (N = 9*) (mean value ± SD)	Before Stage III: *P* value (Mann-Whitney's Test)
RVEDP (mm Hg)	10 ± 5	6 ± 3	.12	7 ± 2	8 ± 3	.610
Ao-Sat (%)	77 ± 4	72 ± 5	.126	82 ± 5	82 ± 3	.806
Qp:Qs	0.88 ± 0.1	0.66 ± 0.2	.008*	0.56 ± 0.2	0.76 ± 0.2	.086
RPA/m²	27 ± 7	21 ± 4	.196	22 ± 1	18 ± 3	.011*
LPA/m²	21 ± 8	20 ± 6	.785	14 ± 3	15 ± 4	.954
Rt/Lt ratio	1.3 ± 0.2	1.1 ± 0.3	.083	1.7 ± 0.4	1.3 ± 0.4	.395
Nakata index	335 ± 200	201 ± 86	.103	232 ± 28	209 ± 44	.395
RVFS (%)	28 ± 2	35 ± 7	.018*	32.9 ± 2.7	32.5 ± 5.1	.806
RVEDd (m/m)	32 ± 5	26 ± 5	0.063	34 ± 7	34 ± 3.3	.713

Abbreviations: RVEDP, right ventricular end-diastolic pressure; Ao-Sat, aorta oxygen saturation; Qp:Qs, pulmonary-to-systemic blood flow ratio; Rt/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.

Dr. Sano ; 4th WCPCCS , Argentina

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

*In angiogram, depressed wall motion was Limited in the vicinity of ventriculotomy

Benefits for preparing the stage II

Variable	SPA shunt (n=27)	RV/PA conduit (n=51)	*P* value
LPAI (m^2/m^2)	49.63 ± 20.9	134.91 ± 48.1	0.006
RPAI (m^2/m^2)	61.24 ± 5.6	182.29 ± 73.3	0.001
VC-O_2 (%)	49.7 ± 7.3	43.5 ± 6.6	0.024
- O_2 (%)	75.3 ± 4.4	67.4 ± 6.6	<0.001
Qp:Qs	1.24 ± 0.43	0.80 ± 0.47	0.011
VH (n)	7/27	2/51	0.012
ICU stay (days)	15.0 ± 18.9	6.8 ± 10.0	0.004

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Mid-term outcomes on subsequent palliation

Inter-stage mortality

- **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

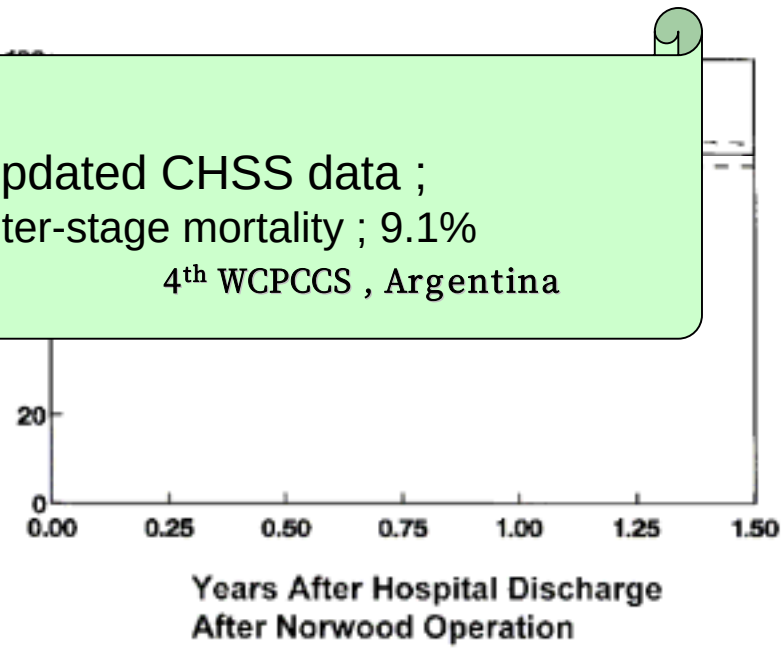
Inter-stage mortality

- **David A. Ashburn, CHSS**

  J Thorac Cardiovasc Surg 2003;125:1070-82.

  - 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



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Mid-term outcomes on subsequent palliation

Inter-stage mortality

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

	Azakie A et al [2]	Bradley SM et al [1]	Mair R et al [5]	Pizarro C et al [4]	Mahle WT et al [11]	Total
Discharged from hospital (n)						
NW-BT	7	20	13	14	18	72
NW-RVPA	12	17	13	33	9	84
Crossover (n)						
NW-BT		22[b]			20[b]	
NW-RVPA		15[b]			7[b]	
Mortality (n)						
NW-BT	1	4	3	2	4	14
NW-RVPA	1	0	0	1	0	2
Mortality (%)[a]						
NW-BT	14.3	20.0	23.1	14.3	22.2	19.4
NW-RVPA	8.3	0	0	3.0	0	2.4
Mortality with crossover (%)[a]						
NW-BT		18.2[b]			20.0[b]	
NW-RVPA		0			0	
Nonsurgical candidate/transplant						
NW-BT	1	0	0	0	0	1
NW-RVPA	0	0	0	0	0	0
Awaiting stage II (n)						
NW-BT	0	0	0	0	NR	
NW-RVPA	4[b]	3[b]	0	0	NR	
Time to stage II (months)						
NW-BT	Median	NR	NR	3.1	~6	6.3
NW-RVPA	Median	NR	NR	3.4	~6	6.3
Completed stage II (n)						
NW-BT		5	18	10	12	NR
NW-RVPA		7	12	13	32	NR

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Mid-term outcomes on subsequent palliation

Dr.Cua's meta-analysis

- **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.

→ **Ghanayem et al.'s report** *J Thorac Cardiovasc Surg 2003;126: 1367-77.*

Mid-term outcomes on subsequent palliation

Ghnayem's report s – Home surveillance

Home surveillance program prevents interstage mortality after the Norwood procedure

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N. A. Rudd, MSN[c]
M. M. Stoltzer, MSN[c]
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S. J. Frisbee, MS[f]
R. D. B. Jaquiss, MD[e]
S. B. Litwin, MD[e]
J. S. Tweddell, MD[e]

See related editorial on page 1257.

Objective: To determine whether early identification of physiologic variances associated with interstage death would reduce mortality, we developed a home surveillance program.

- **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.

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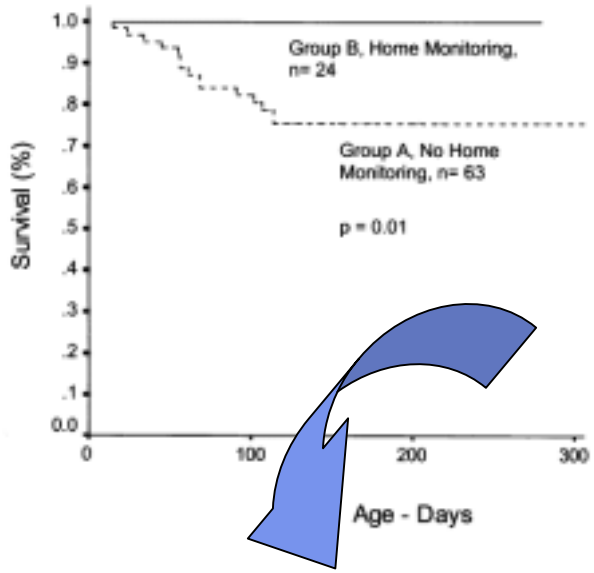
Mid-term outcom
palliation



"Hawthorne effect" ; described by Elton Mayo of the Harvard BusinessSchool ;

❖ 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.

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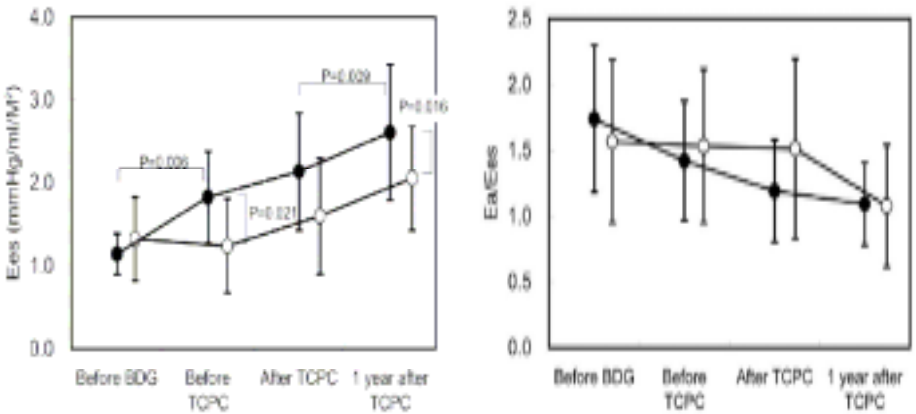
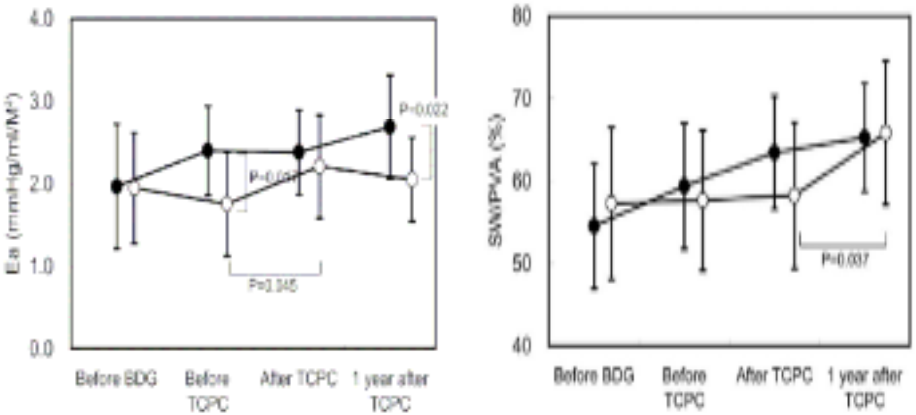
Mid-term outcomes on subsequent palliation

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**

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Mid-term outcomes on subsequent palliation

Summary of other studies

Studies (SPA/RVPA)	Pizarro et al. (2003) (20/36)	Bradley et al. (2004) (25/19)	Azakie et al. (2004) (8/13)	Mair et al. (2003) (18/14)	Rumball et al. (2005) (18/12)	Januszewska et al. (2005) (27/51)
Mortality before stage II operation	0.14 / 0.03 Not difference	18 / 0 p=0.1	0.25 / 0.15 Not difference	23 / 0 no comment	- - -	?? /1.9
		- - -	79% (1 year, total pts.) Not difference	- - -	- - -	- - -
PA index	- - -	- - -	- - -	- - -	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
Remarks	- - -	- - -	- - -	dp/dt is better in RV/PA conduit	No difference ; time interval to cath., Qp/Qs , Hb level	- - -

Dr. Pizarro ;
0.13(6/46) .vs. 0.02 (1/50) (p <0.01)
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Summary

- Theoretical advantages of new strategies may not produce true benefit. (Extracardiac Fontan, Continuous cerebral perfusion , Hybird 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 procedure.
- 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 =

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

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