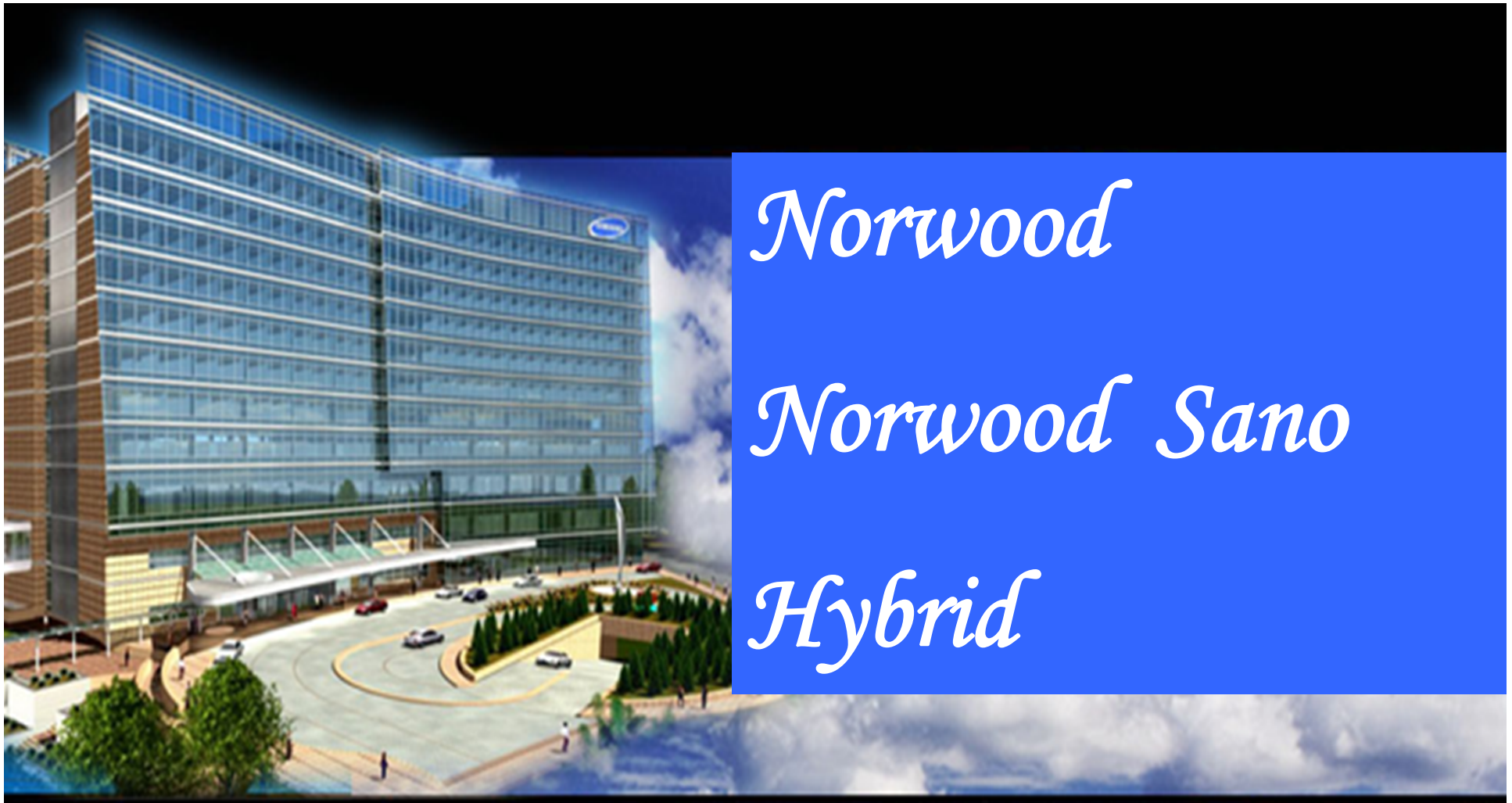


Norwood

Norwood Sano

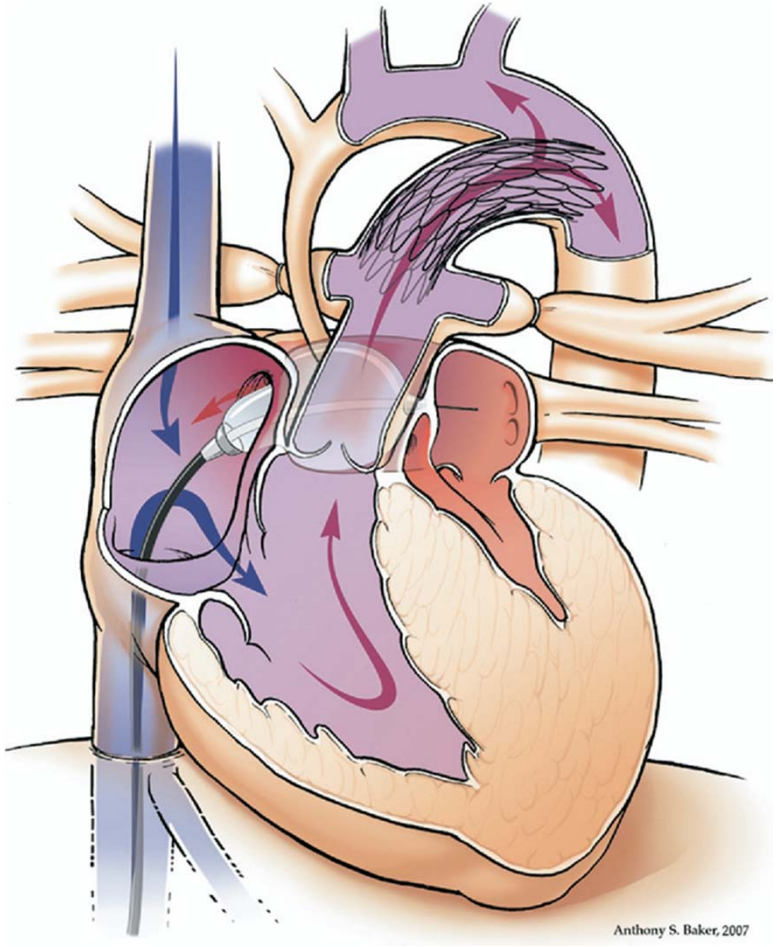
Hybrid

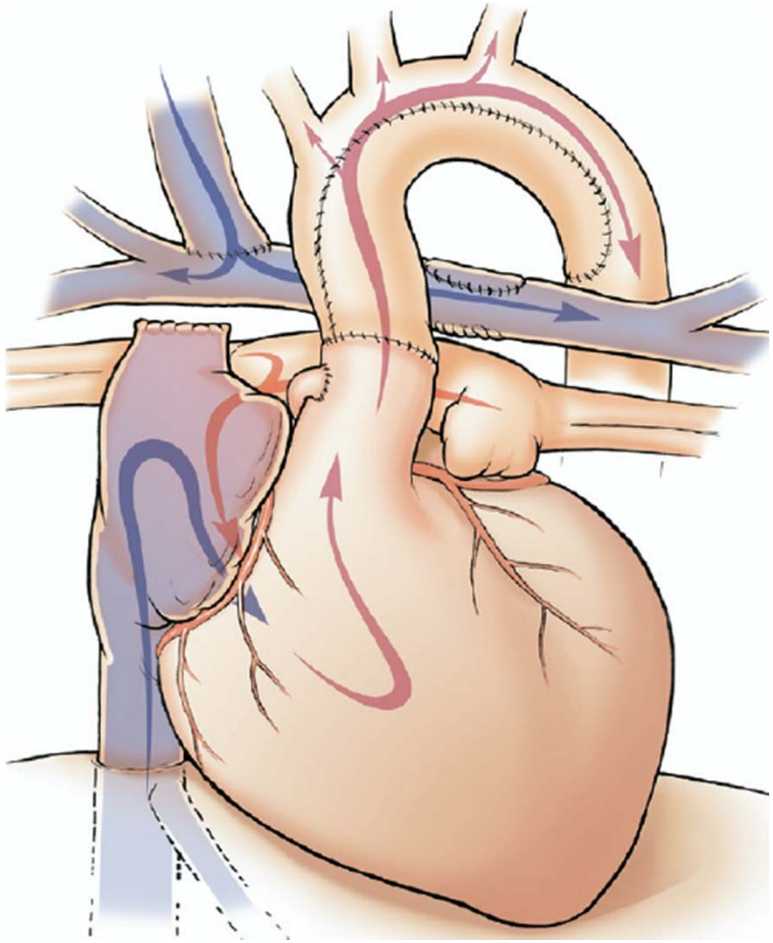
Tae-Gook Jun

Samsung Seoul Hospital

Sungkyunkwan University School of Medicine

Hybrid procedure





Hybrid procedure

Advantage

- Avoidance of neonatal CPB
- Avoid mutiple transfusion

Hybrid procedure

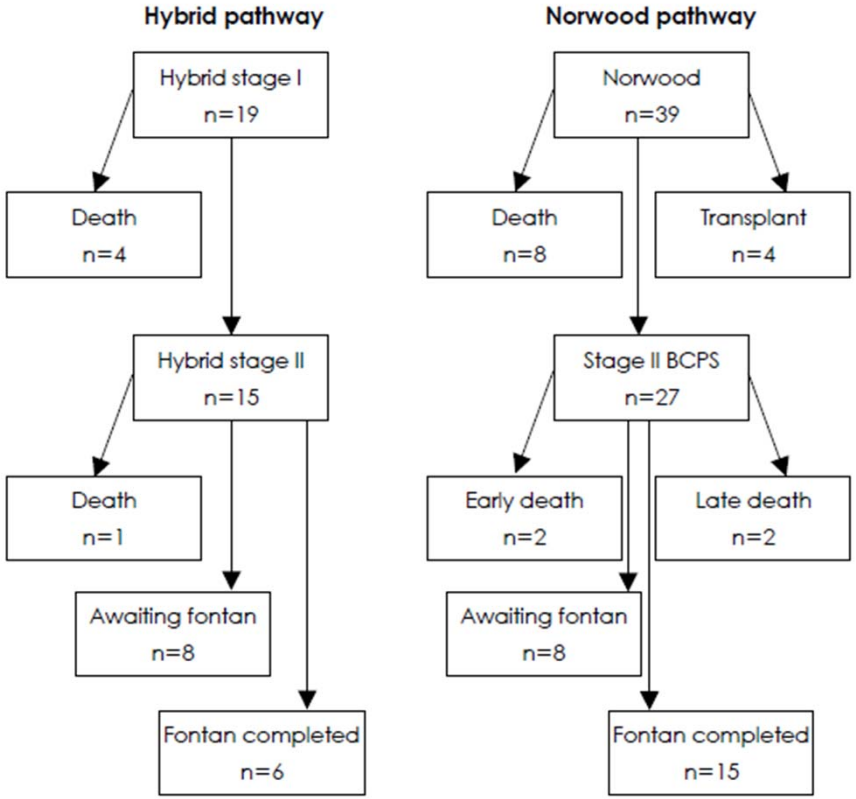
Disadvantage

- Development of retrograde coarctation from ductal tissue
  - Neurologic concern – decreased cerebral blood flow
  - Coronary ischemia
- Challenging second operation
  - Stent removal
  - Arch reconsturction
  - BCPS
- Complications related with pulmonary artery banding
  - Pulmonary artery distortion
  - Band migration

Hybrid Palliation for Neonates With Hypoplastic Left Heart Syndrome: Current Strategies and Outcomes

Osami Honjo, MD and Christopher A. Caldarone, MD

Division of Cardiovascular Surgery, Hospital for Sick Children, Toronto, Ontario, Canada



Korean Circ J 2010;40:103-11

Hybrid Palliation for Neonates With Hypoplastic Left Heart Syndrome: Current Strategies and Outcomes

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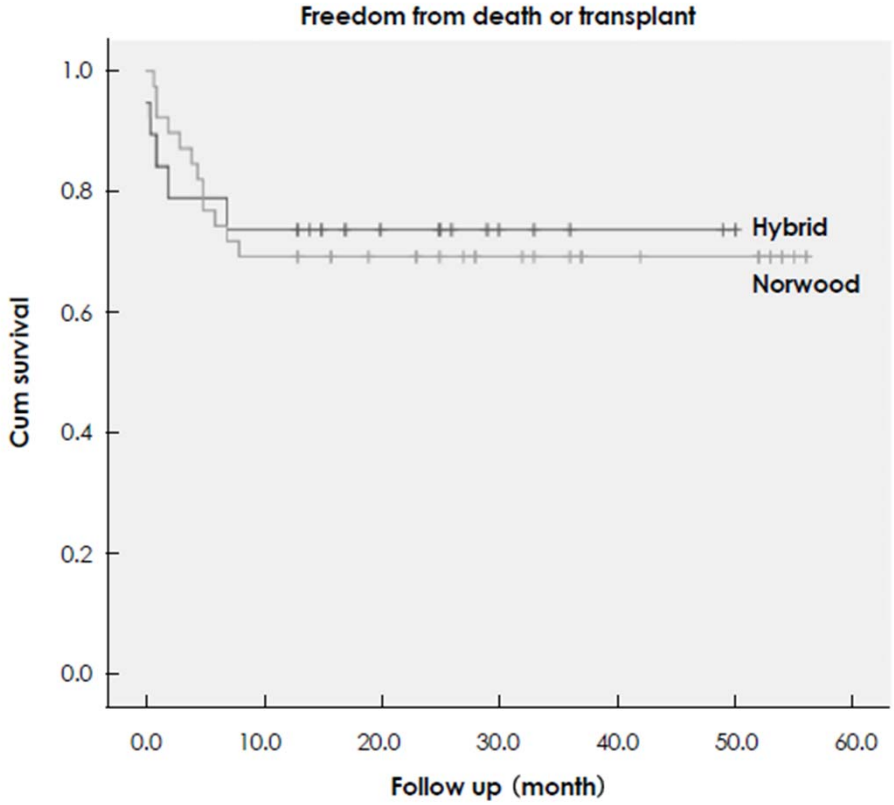

Korean Circ J 2010;40:103-11

THE HYBRID STAGE 1 OPERATION IN HYPOPLASTIC LEFT HEART SYNDROME: A NEW ALTERNATIVE

*Emile Bacha, M.D.

Harvard Medical School and Children's Hospital, Boston, MA, USA

Table 1: Complications encountered with the hybrid stage I procedure (in parenthesis number of patients)

Intra-operative stage I	Post-operative	Interstage	Intra-operative Stage II	Follow-up
Cardiac arrest during wire passage into descending aorta (coronary clot) (1)	PA band revision (4) PA band migration (1)	Retrograde coarctation (2)	Difficult ductal stent removal (4) LPA stent (2)	LPA plasty (2)
Insufficient ductal coverage necessitating another stent (1)	Ductal stent distal migration (2)			
	Atrial stent migration (2)			

Heart view 2006;7:105-110

THE HYBRID STAGE 1 OPERATION IN HYPOPLASTIC LEFT HEART SYNDROME: A NEW ALTERNATIVE

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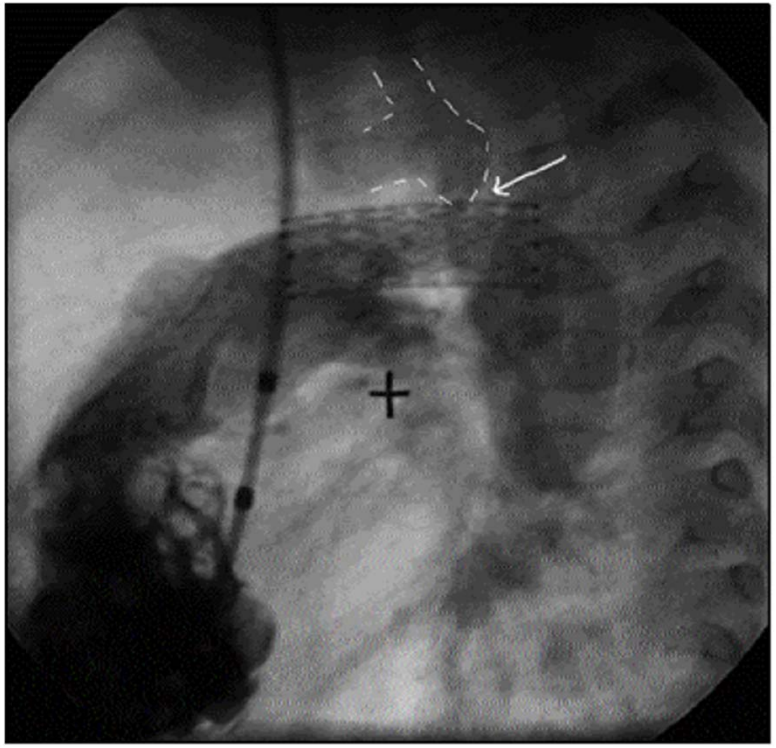

Fig.4: Pre-stage II angiogram at 3 months shows preductal coarctation. This patient was taken to the OR the next day.

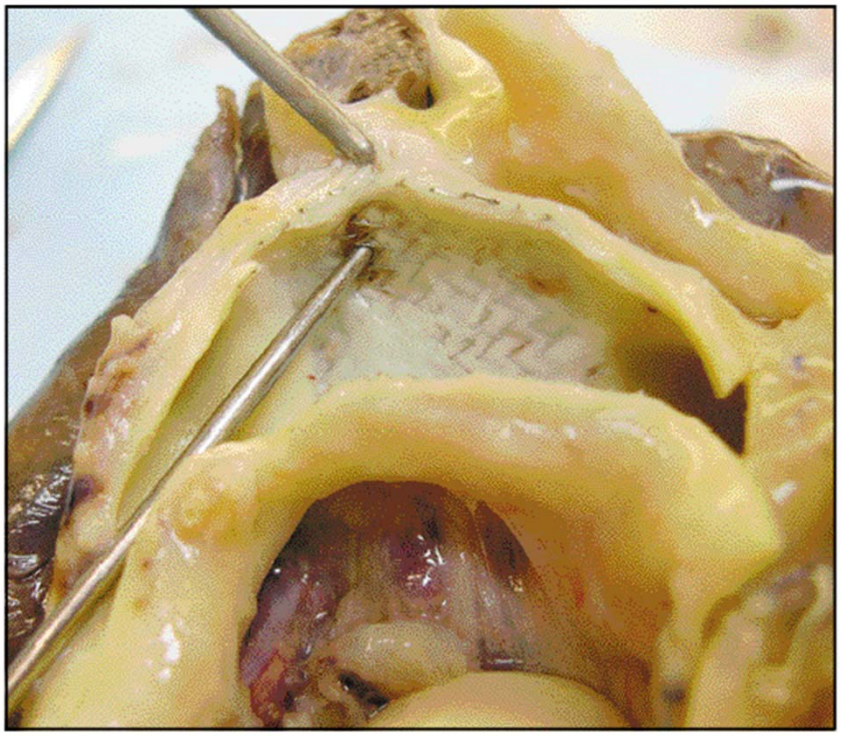

Fig.5: Ductal stent well embedded after 5 months. The opening into the transverse arch is shown with a probe.

Heart view 2006;7:105-110

THE HYBRID STAGE 1 OPERATION IN HYPOPLASTIC LEFT HEART SYNDROME: A NEW ALTERNATIVE

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Harvard Medical School and Children's Hospital, Boston, MA, USA

Table 2: Current indications for the Hybrid Stage I in hypoplastic left heart syndrome (HLHS)

Good indications: Neonate with HLHS and:	Possible indications: Neonate with HLHS and:	Questionable indication: Neonate with HLHS and:
1. Major non-cardiac defects	1. Lintact or restrictive atrial septum	1. Very diminutive ascending aorta and aortic arch
2. Cerebral hemorrhage	2. Poor ventricular function	
3. Late presentation	3. Severe tricuspid regurgitation	
4. End-organ damage	4. Additional cardiac defect	
5. Sepsis		

Heart view 2006;7:105-110

Hybrid Approach for Hypoplastic Left Heart Syndrome: Intermediate Results After the Learning Curve

Mark Galantowicz, MD, John P. Cheatham, MD, Alistair Phillips, MD, Clifford L. Cua, MD, Timothy M. Hoffman, MD, Sharon L. Hill, ACNP, and Roberta Rodeman, RN

- 2002 – 2007
- 40 patients
- Exclusion:
  - ✓ non-HLHS univentricular anatomy
  - ✓ those bridged to a two ventricle repair
  - ✓ Intact atrial septum
  - ✓ weight less than 1.5 kg
  - ✓ significant extracardiac malformations (CDH)
  - ✓ those previously reported as part of the learning curve.
- Overall survival 82.5% (33/40)

Ann Thorac Surg 2008;85:2063-71

The Retrograde Aortic Arch in the Hybrid Approach to Hypoplastic Left Heart Syndrome

Serban C. Stoica, MD, Alistair B. Philips, MD, Matthew Egan, MD, Roberta Rodeman, RN, Joanne Chisolm, RN, Sharon Hill, ACNP, John P. Cheatham, MD, and Mark E. Galantowicz, MD

Nationwide Children's Hospital, Columbus, Ohio

- 2002 – 2008
- 66 patients

- Group I :patients requiring RAAO intervention (n=16)
- Group II: no RAAO (n=50)

Ann Thorac Surg 2008;85:2063-71

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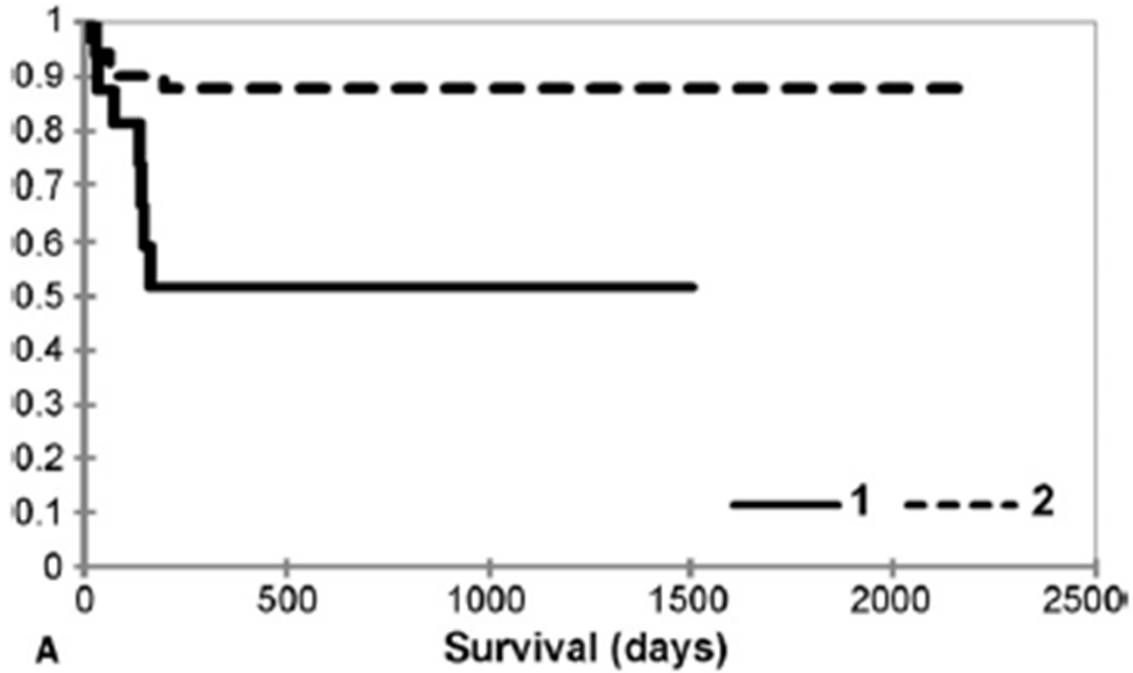

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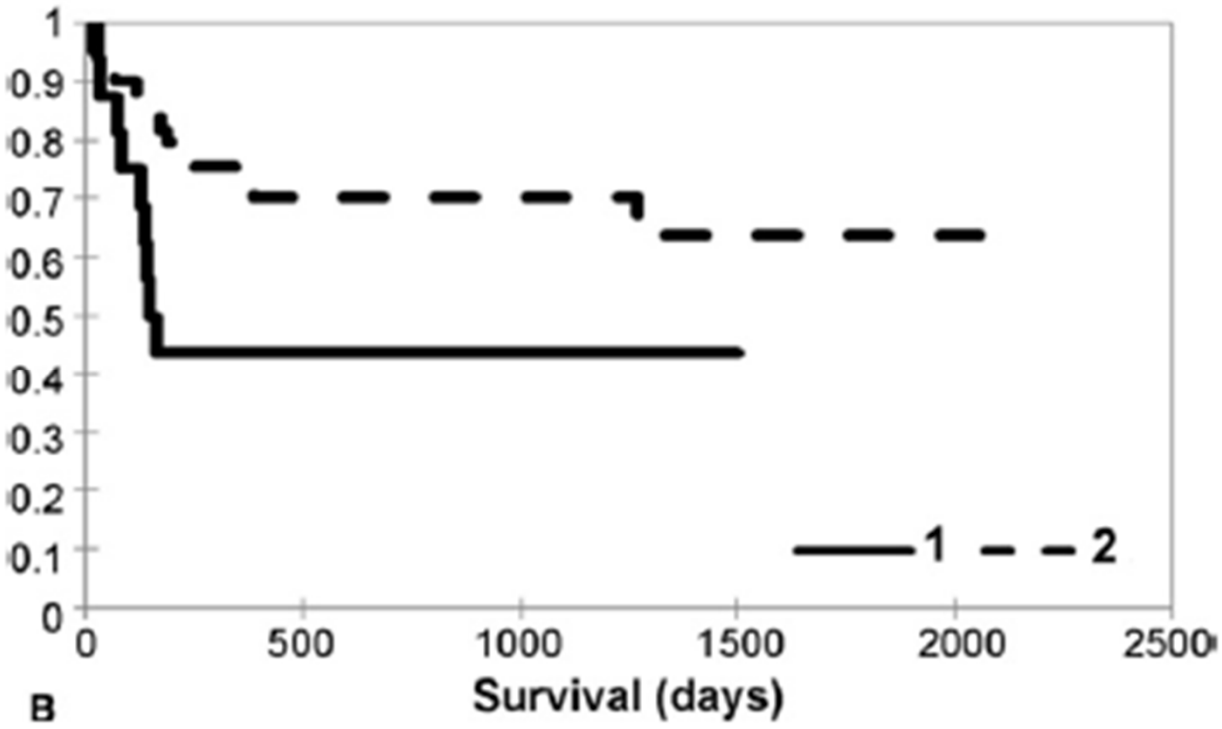

Ann Thorac Surg 2008;85:2063-71

Predictors of Retrograde Aortic Arch Obstruction After Hybrid Palliation of Hypoplastic Left Heart Syndrome

Matthew J. Egan · Sharon L. Hill · Bethany L. Boettner · Ralf J. Holzer · Alistair B. Phillips · Mark Galantowicz · John P. Cheatham · John P. Kovalchin

- 2002 – 2009
- 96 patients underwent hybrid stage I

- 68 patients (47 boys) included in study
- 20 patients had RAAO (29%)
- Risk factors for RAAO

Pediatr Cardiol 2011;32:67-75

Predictors of Retrograde Aortic Arch Obstruction After Hybrid Palliation of Hypoplastic Left Heart Syndrome

Matthew J. Egan · Sharon L. Hill · Bethany L. Boettner · Ralf J. Holzer · Alistair B. Phillips · Mark Galantowicz · John P. Cheatham · John P. Kovalchin

Table 4 Echocardiographic measurements of the aorta in all patients

		RAAO (mm)	Non-RAAO (mm)	*p* Value
Aortic annulus		2.3 ± 1	2.8 ± 0.8	0.066
	z-score	−3.9 ± 0.8	−3.5 ± 0.8	0.065
	Index BSA	5 ± 2.1	6.3 ± 1.9	0.041
Aortic root		3.6 ± 1.3	4.4 ± 1.4	0.036
	z-score	−3.1 ± 0.8	−2.5 ± 0.9	0.024
	Index BSA	7.7 ± 2.7	9.9 ± 3.3	0.018
Ascending aorta		2.2 ± 1.2	2.7 ± 1.4	0.170
	z-score	−3.7 ± 0.8	−3.3 ± 1.1	0.128
	Index BSA	4.8 ± 2.3	6.2 ± 3.3	0.105

Pediatr Cardiol 2011;32:67-75

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Table 4 Echocardiographic measurements of the aorta in all patients

		RAAO (mm)	Non-RAAO (mm)	*p* Value
Proximal transverse aortic arch		3.7 ± 0.6	3.9 ± 0.9	0.401
	Index BSA	8.1 ± 1.2	8.6 ± 1.8	0.260
Distal transverse aortic arch		4.2 ± 0.8	4.1 ± 0.9	0.699
	z-score	−2.2 ± 0.5	−2.3 ± 0.5	0.943
	Index BSA	9.1 ± 1.8	9.1 ± 1.8	0.957
Aortic isthmus		3.6 ± 0.6	4 ± 1	0.108
	z-score	−2.5 ± 0.4	−2.1 ± 0.8	0.059
	Index BSA	7.7 ± 1.1	8.9 ± 2.2	0.045
Descending aorta		6.3 ± 1.2	5.9 ± 0.7	0.127
	Index BSA	13.6 ± 2.1	13 ± 1.6	0.237

Pediatr Cardiol 2011;32:67-75

Predictors of Retrograde Aortic Arch Obstruction After Hybrid Palliation of Hypoplastic Left Heart Syndrome

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Table 6 Peak retrograde aortic arch velocities

Retrograde velocity	RAAO ($n = 20$) n (%)	Non-RAAO ($n = 47$) n (%)	p Value
≤3 m/s	11 (55)	29 (62)	0.036
3.1–3.5 m/s	5 (25)	17 (36)	
>3.5 m/s	4 (20)	1 (2)	

RAAO retrograde aortic arch obstruction

Pediatr Cardiol 2011;32:67-75

Predictors of Retrograde Aortic Arch Obstruction After Hybrid Palliation of Hypoplastic Left Heart Syndrome

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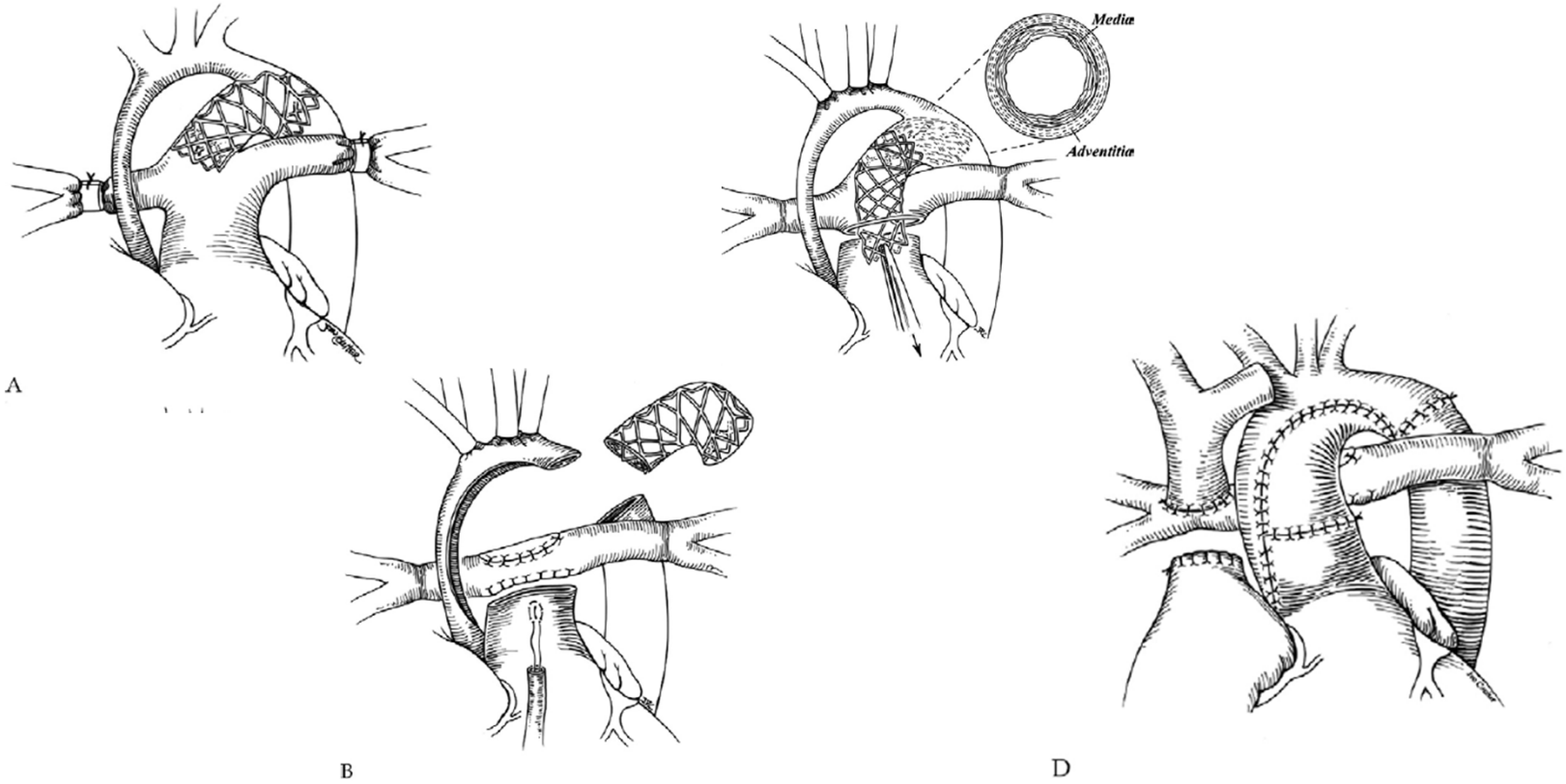
Additionally, studies to evaluate the histopathologic response of the surrounding tissue to the PDA stent are ongoing. There may be a subset of patients who have a heightened immunologic response to stent placement, which could lead to obstruction of flow in the retrograde aortic arch as well. Because the etiology of RAAO likely is multifactorial, it continues to require a high index of suspicion to prevent its possible sequelae.

Pediatr Cardiol 2011;32:67-75

Stage II Reconstruction After Hybrid Palliation for High-Risk Patients With a Single Ventricle

Christian Pizarro, MD, Kenneth A. Murdison, MD, Christopher D. Derby, MD, and Wolfgang Radtke, MD

The Nemours Cardiac Center; Alfred I. duPont Hospital for Children, Wilmington, Delaware



Ann Thorac Surg 2008;85:1382-8

Bilateral banding + PGE1 infusion

Advantage to ductal stenting

- Easier arch reconstruction at the stage 2
- Better retrograde arch flow for cerebral perfusion or coronary circulation
  - Prevention of isthmic stenosis

Bilateral banding + PGE1 infusion

Disadvantages

- Complication of PGE1
  - ✓ Vasodilation
  - ✓ Apnea
  - ✓ Cost
- Problem with a lonstanding catheter line
  - ✓ Sepsis
- Longer hospitalization

Early results of bilateral pulmonary artery banding for hypoplastic left heart syndrome☆

Takahisa Sakurai [a], Hideaki Kado [a,*], Toshihide Nakano [a], Kazuhiro Hinokiyama [a], Akira Shiose [a], Masaki Kajimoto [a], Kunihiko Joo [a], Yuichi Ueda [b]

[a] *Department of Cardiovascular Surgery, Fukuoka Children's Hospital, 2-5-1 Toujinmachi, Chuuo-ku, Fukuoka, 810-0063, Japan*
[b] *Department of Cardiothoracic Surgery, Nagoya University Graduate School of Medicine, 65 Tsurumai-cho, Nagoya 466-8550, Japan*

- 2004-2007
- PAB group (n=18), PA banding + PGE1 infusion
- Norwood group (n=25), (RV – PA conduit n=14)

Anatomical diagnosis.

	PAB (*n* = 18)	NWD (*n* = 25)
HLHS	14	19
Shone syndrome	0	4
Unbalanced AVSD	0	1
DORV, hypoLV	1	1
TA, TGACoA	2	0
Aortic atresia, VSD	1	0

Eur J Cardio-thorac Surg 2009;36:973-9

Early results of bilateral pulmonary artery banding for hypoplastic left heart syndrome☆

Takahisa Sakurai [a], Hideaki Kado [a,*], Toshihide Nakano [a], Kazuhiro Hinokiyama [a], Akira Shiose [a], Masaki Kajimoto [a], Kunihiko Joo [a], Yuichi Ueda [b]

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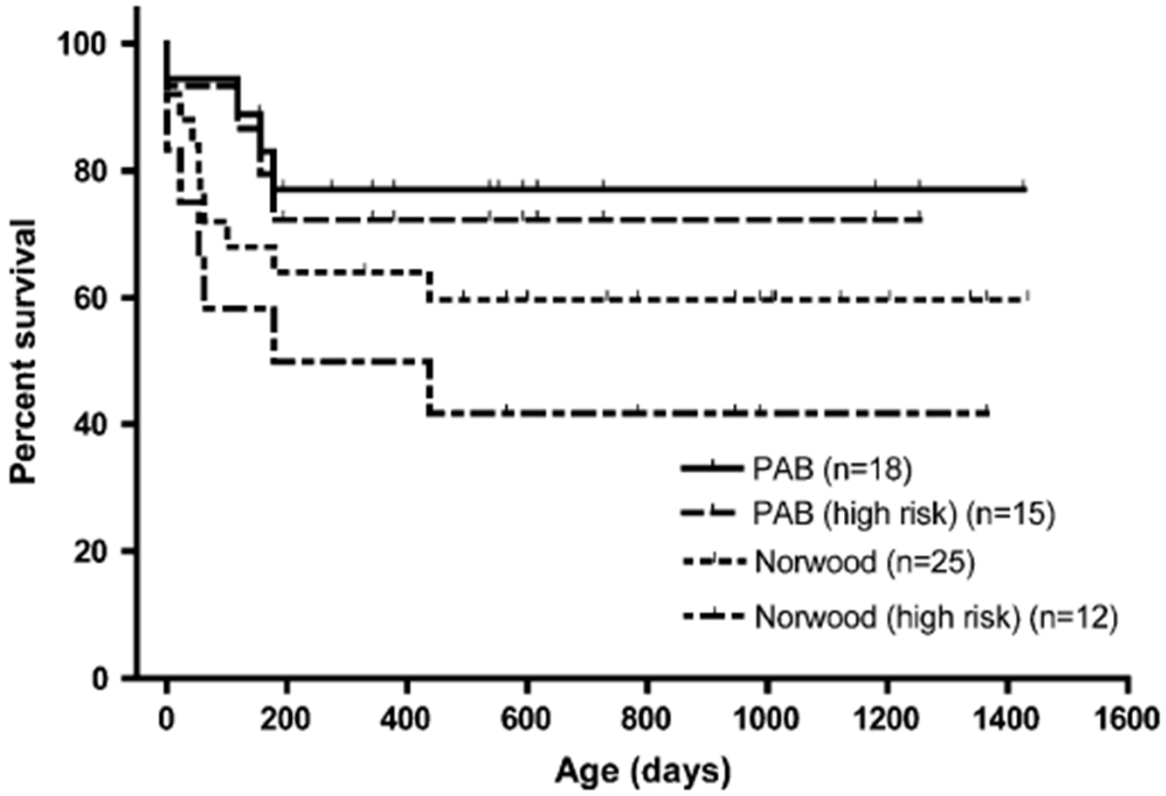


Eur J Cardio-thorac Surg 2009;36:973-9

Norwood procedure

RV – PA shunt vs. modified BT shunt

Sano modification (RV – PA shunt)

History

- Norwood
- Imato
- Sano

Sano modification (RV – PA shunt)

Advantage

- Higher diastolic pressure
- Higher coronary perfusion pressure
- More balanced pulmonary to systemic flow ratio
- Decreased ventricular volume
- Pulsatile pulmonary blood flow
- Improved end-organ perfusion

Sano modification (RV – PA shunt)

Disadvantages

- Consequence of ventriculotomy
  - Arrhythmias
  - RV dysfuntion
  - RV aneurysm
  - Tricuspid valve regurgitation
- Diastolic flow regurgitation – RV volume load
- Early cyanosis : lower Qp/Qs
- Earlier second stage procedure
- Increased number of intervention – inadequate PA growth
- Shunt stenosis/ thrombosis

Complications after Sano modifications

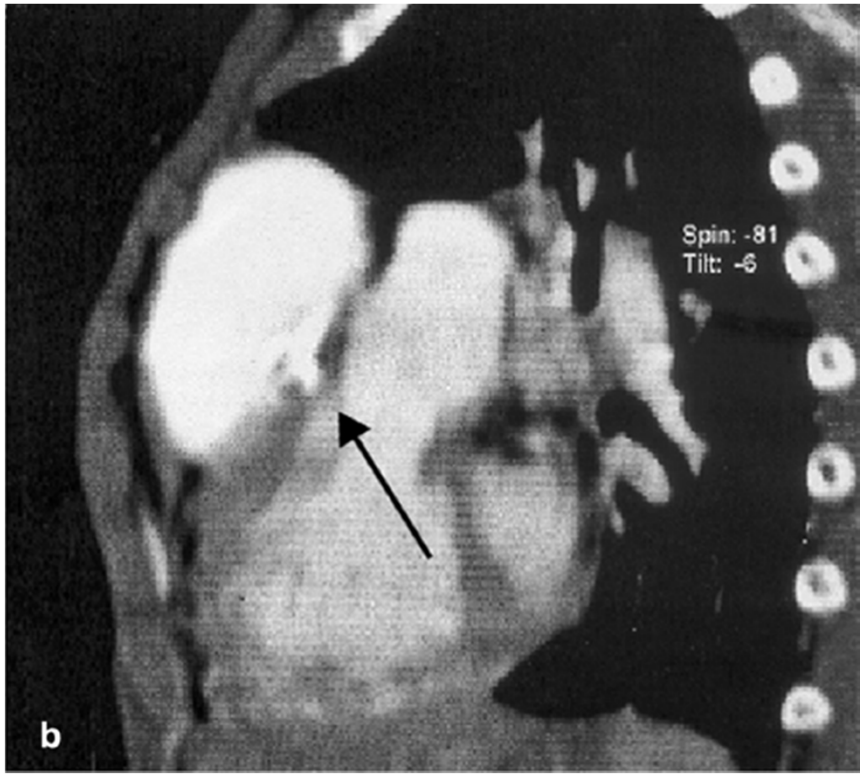
- Ventricular aneurysm
  - Cause: RV tomy

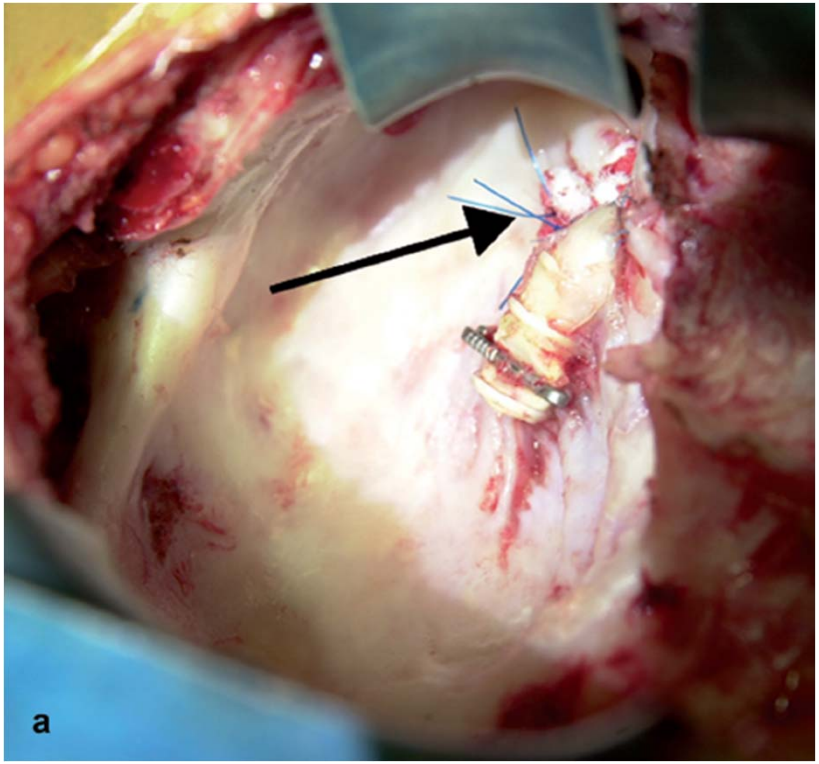
Crit Care Nurse 2010;30:54-9

False aneurysm origination from the proximal anastomosis of a right ventricular to pulmonary artery shunt following staged repair of hypoplastic left heart syndrome

Jürgen Hörer[a,*], Ivan Malcic[b], Christian Schreiber[a], Rüdiger Lange[a]

[a]Department of Cardiovascular Surgery, Deutsches Herzzentrum München an der Technischen Universität München, Lazarettstrasse 36, D-80636 Munich, Germany



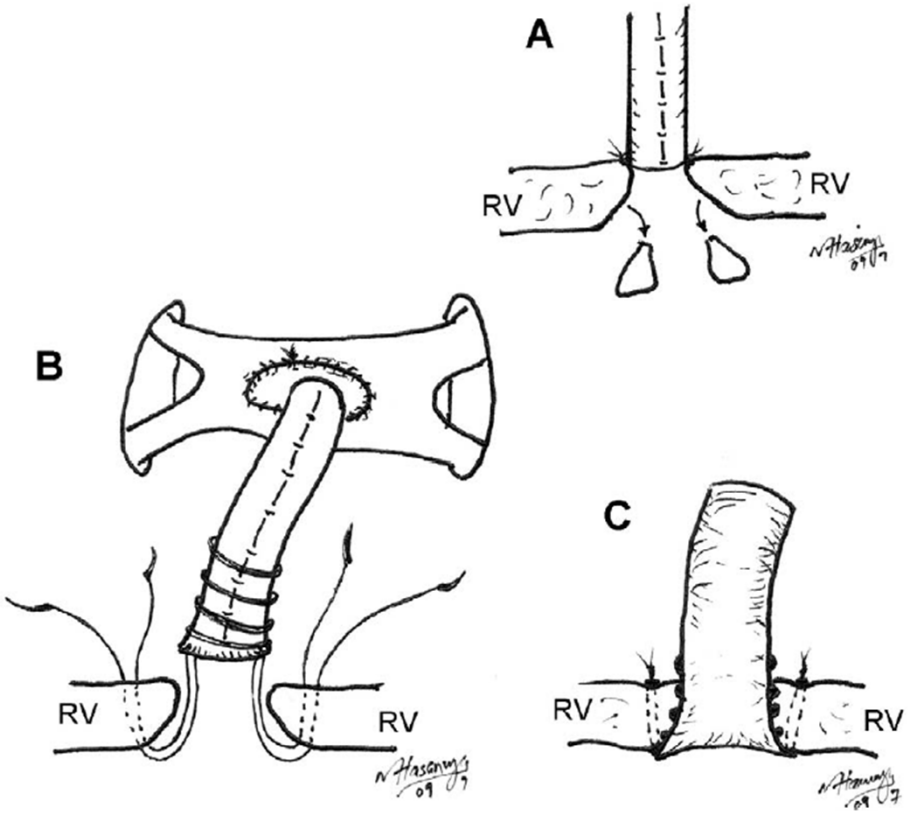


Int Cardiovas Thorac Surg 2011, in press

Modification of Ventricular-to-Pulmonary Shunt to Minimize Proximal Conduit Obstruction After Stage I Norwood Reconstruction

Nahidh W. Hasaniya, MD, PhD, Howard Shattuck, PA, Anees Razzouk, MD, and Leonard Bailey, MD

Department of Cardiothoracic Surgery, Loma Linda University Children's Hospital, Loma Linda, California



Ann Thorac Surg 2010;89:e4-6

Comparison of Norwood Shunt Types: Do the Outcomes Differ 6 Years Later?

Eric M. Graham, MD, Sinai C. Zyblewski, MD, Jacob W. Phillips, MD, Girish S. Shirali, MBBS, Scott M. Bradley, MD, Geoffery A. Forbus, MD, Varsha M. Bandisode, MD, and Andrew M. Atz, MD

Divisions of Pediatric Cardiology and Cardiothoracic Surgery, Medical University of South Carolina, Charleston, South Carolina

- Jan 2000 – April 2007
- 76 patients underwent Norwood procedure
- 35 with mBTS
- 41 with RV-PA conduit
- Pre-Fontan assessment
  - ✓ RV systolic function 1 – 4
  - ✓ TR 1 – 4
  - ✓ QRS duration

Ann Thorac Surg 2010;90:31-5

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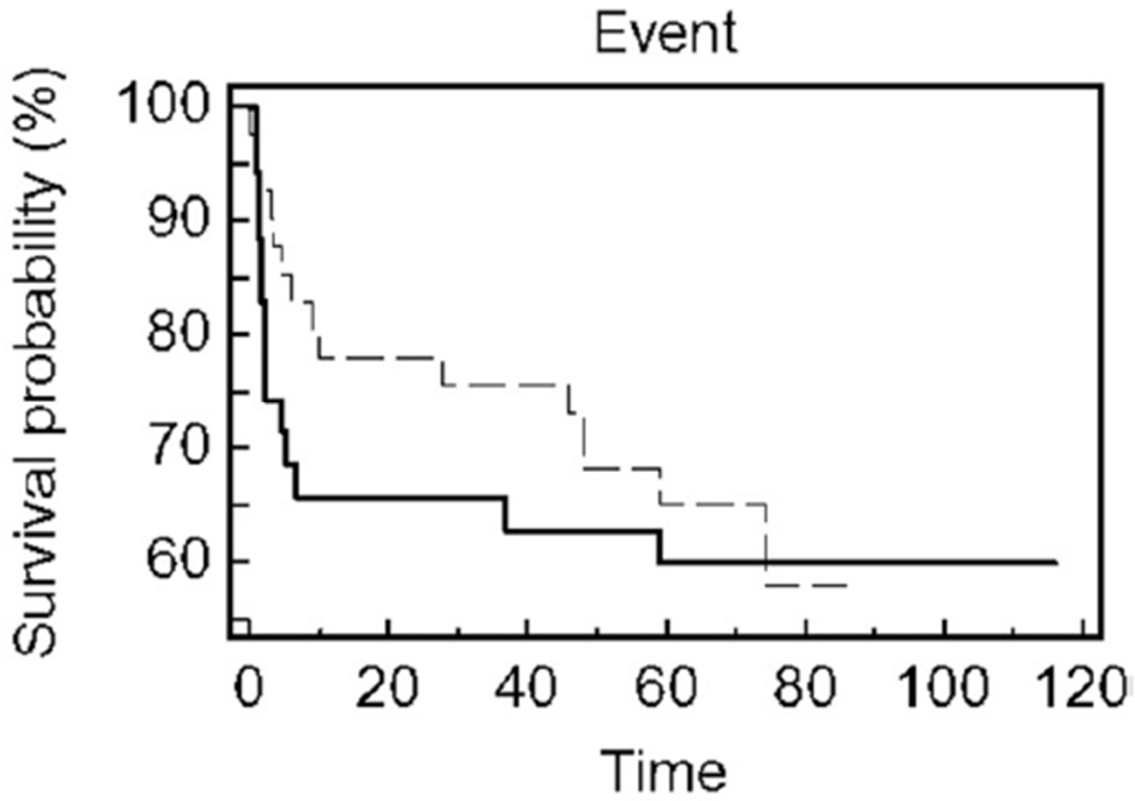

Ann Thorac Surg 2010;90:31-5

Differential branch pulmonary artery growth after the Norwood procedure with right ventricle–pulmonary artery conduit versus modified Blalock–Taussig shunt in hypoplastic left heart syndrome

Jay D. Pruetz, MD,[a] Sarah Badran, MD,[a] Fred Dorey, PhD,[b] Vaughn A. Starnes, MD,[c] and Alan B. Lewis, MD[a]

- 2000 – 2005
- 159 patients
- Group A : Norwood with MBT (n=103)
- Group B : Norwood with RV-PA shunt (n=56)

J Thorac Cardiovasc Surg 2009;137:1342-8

Differential branch pulmonary artery growth after the Norwood procedure with right ventricle–pulmonary artery conduit versus modified Blalock–Taussig shunt in hypoplastic left heart syndrome

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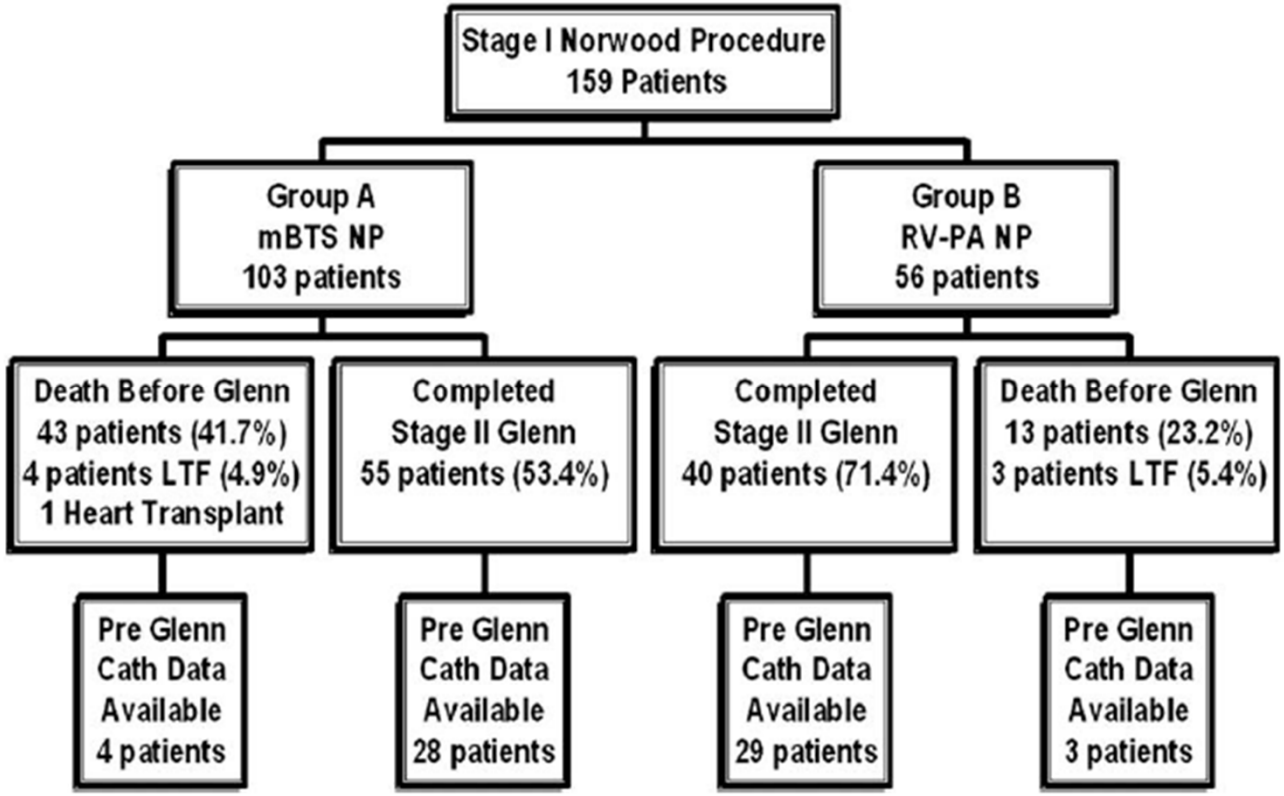

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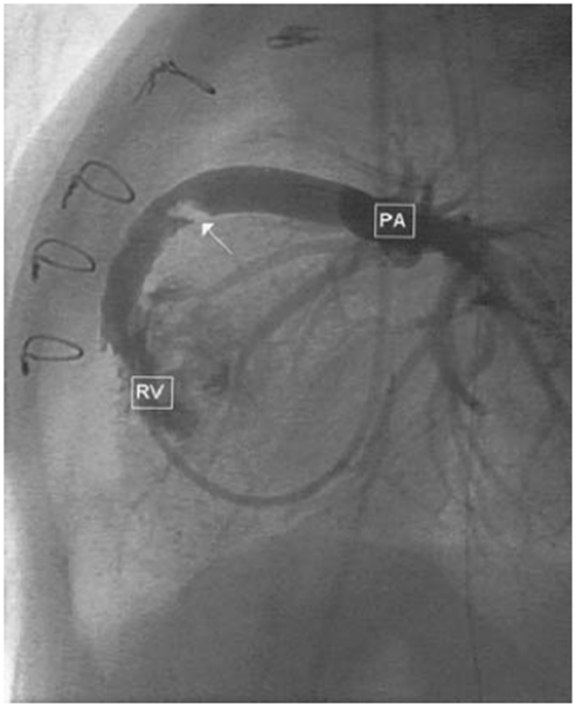
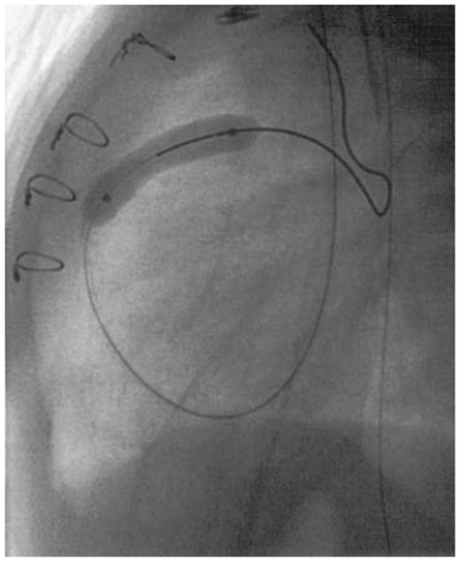
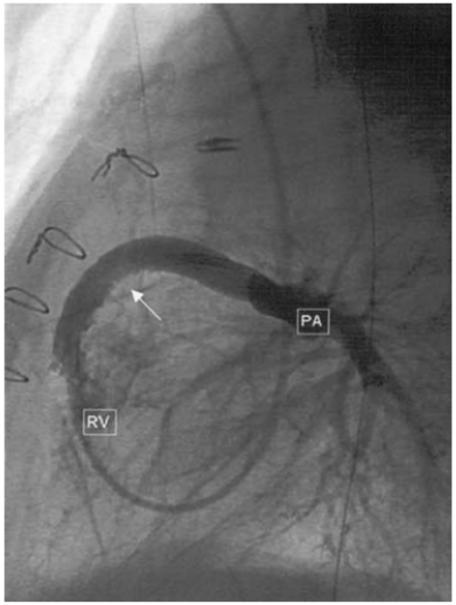
J Thorac Cardiovasc Surg 2009;137:1342-8

Thrombus Formation within the Right Ventricle-to-Pulmonary Artery Conduit (Sano Shunt) as a Cause of Acute Cyanosis in a Patient with Hypoplastic Left Heart Syndrome

M. R. Recto · W. L. Sobczyk · E. H. Austin III







Pediatr Cardiol 2008;29:656-8

Norwood procedure with BT shunt

Advantage

- No ventriculotomy
- Limits right ventricle overload
- Good pulmonary growth

Norwood procedure with BT shunt

Disadvantage

- Reduction in diastolic pressure due to a run off of systemic blood flow into the lungs
  - Decreased coronary perfusion
  - Decreased end organ perfusion
- Hemodynamic instability
- Shunt thrombosis

Outcome of the Norwood operation in patients with hypoplastic left heart syndrome: A 12-year single-center survey

Anke Katharina Furck, MD,[a] Anselm Uebing, MD,[a] Jan Hinnerk Hansen,[a] Jens Scheewe, MD,[b] Olaf Jung, MD,[a] Gunther Fischer, MD,[a] Carsten Rickers, MD,[a] Tim Holland-Letz, MSc,[c] and Hans-Heiner Kramer, MD[a]

- 1996 – 2007
- 157 patients underwent Norwood

General information	
Female	56 (35.7)
Male	101 (64.3)
Premature infants	10 (6.4)
≤2.5 kg	19 (12)
Prenatal diagnosis, yes	72 (45.9)
Anatomic subgroups	
MA/AA	70 (44.6)
MS/AS	42 (26.7)
MS/AA	34 (21.7)
MA/AS	11 (7)

J Thorac Cardiovasc Surg 2010;139:359-65

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- Actuarial survival 73.5% at 1 year, 68.4% at 5 year

- Risk factor for early death
  - ✓ Anatomic subtype (MS/AA)
  - ✓ Aorta less than 2 mm
  - ✓ Aberrant right subclavian artery
  - ✓ Female gender
  - ✓ Absence of ASCP

J Thorac Cardiovasc Surg 2010;139:359-65

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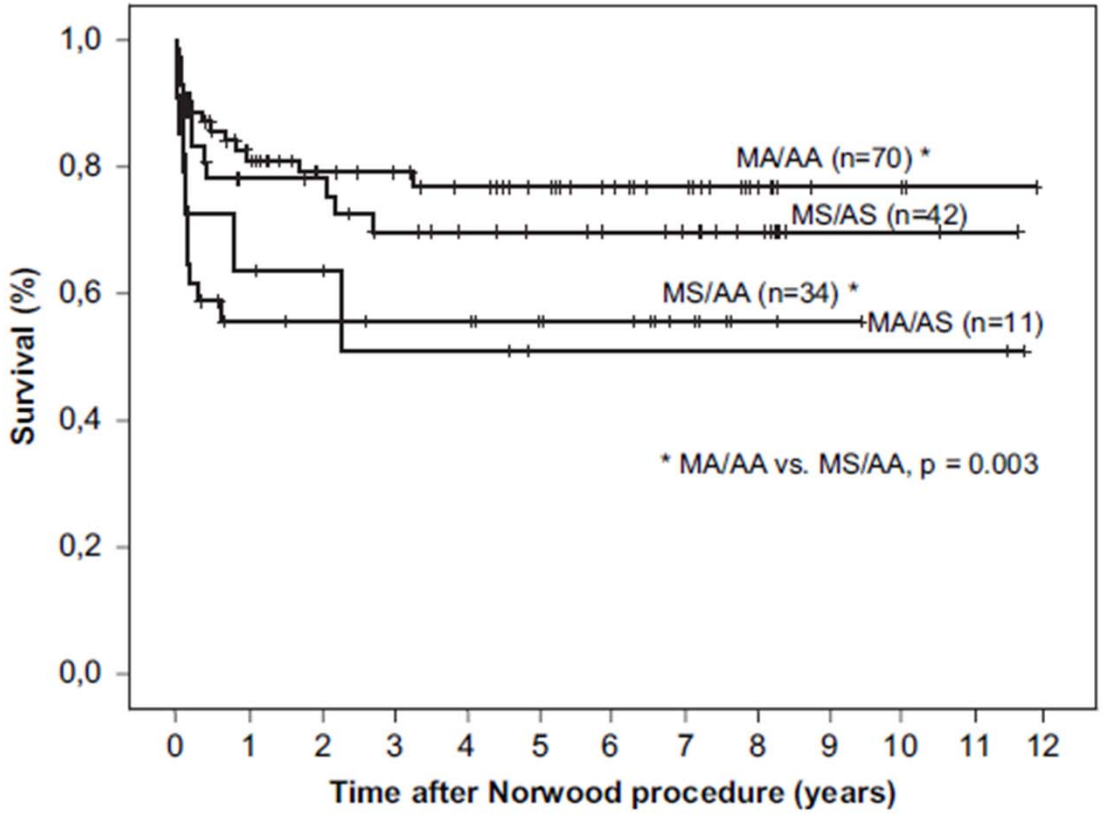

J Thorac Cardiovasc Surg 2010;139:359-65

Comparison of Shunt Types in the Norwood Procedure for Single-Ventricle Lesions

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- Pediatric Heart Network (15 North American Center)
- Randomized trial
- 921 patients May 2005 – July 2008
- 555(84%) enrolled in the study

N Engl J Med 2010;362:1980-1992

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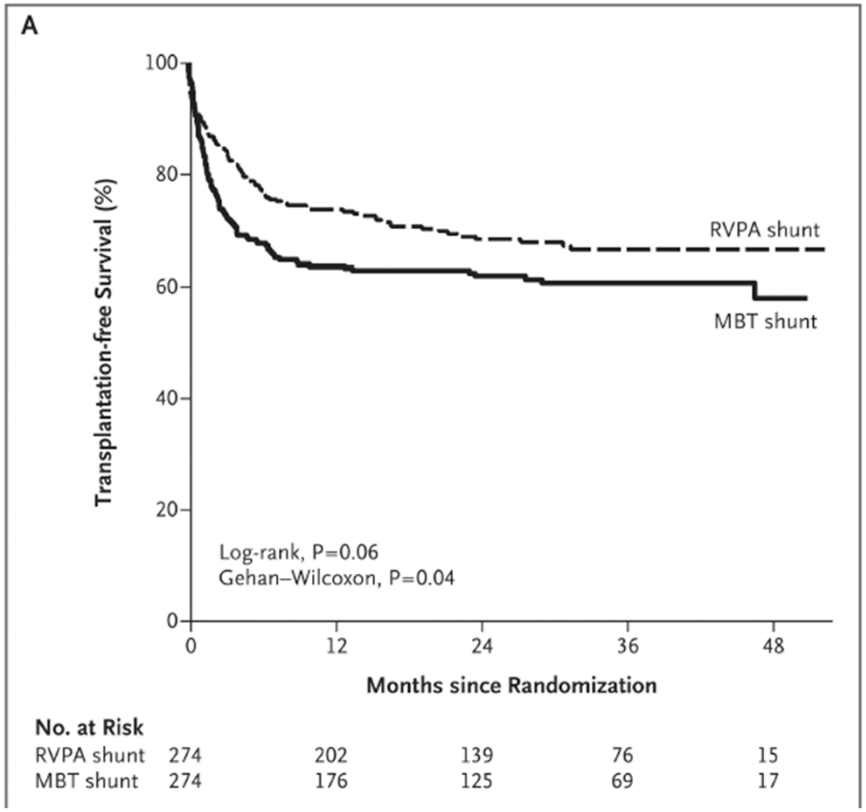

N Engl J Med 2010;362:1980-1992

Bilateral banding -> staged Norwood

Advantage

- Avoid neonatal CPB

Precise evaluation of bilateral pulmonary artery banding for initial palliation in high-risk hypoplastic left heart syndrome

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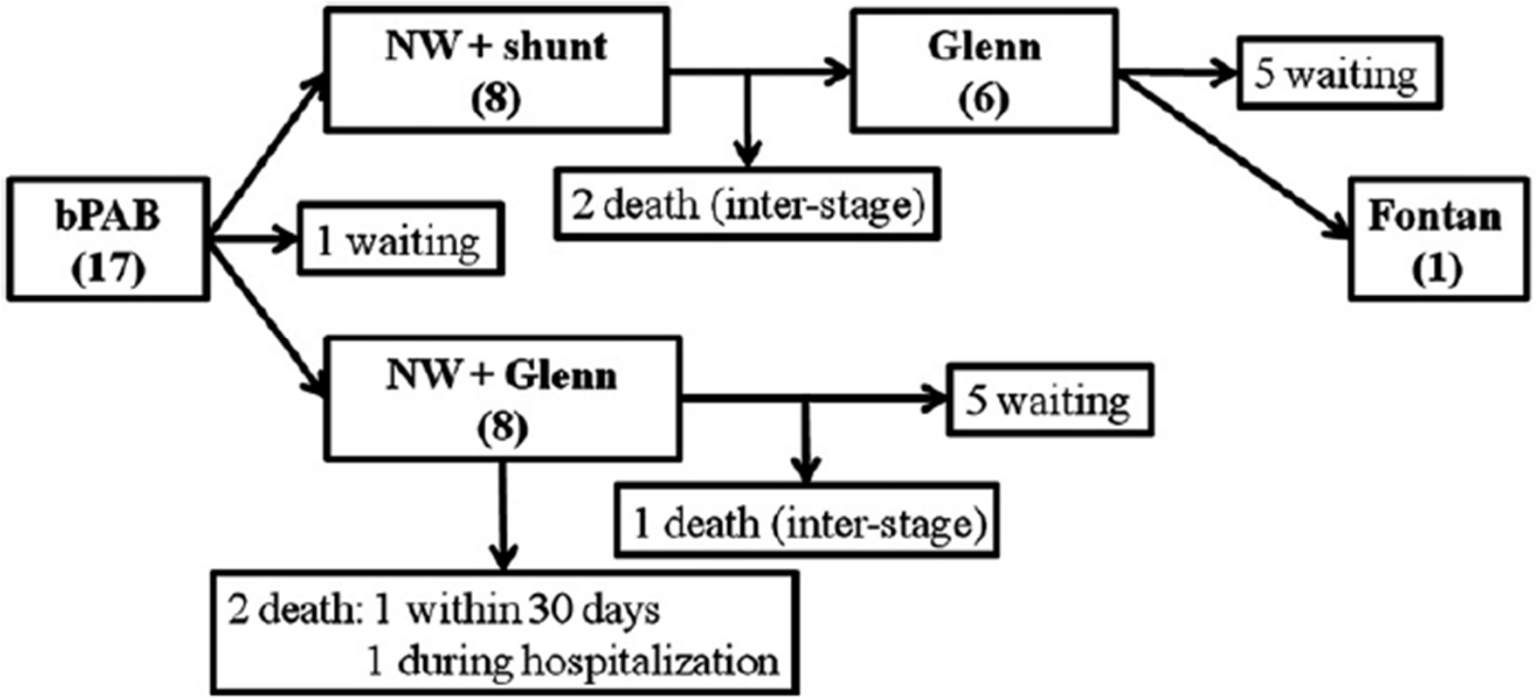

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Measurement of technical performance in surgery for congenital heart disease: The stage I Norwood procedure

Emile A. Bacha, MD,[a] Luis A. Larrazabal, MD,[a] Frank A. Pigula, MD,[a] Kimberlee Gauvreau, ScD,[b] Kathy J. Jenkins, MD,[b] Steve D. Colan, MD,[b] Francis Fynn-Thompson, MD,[a] John E. Mayer, Jr, MD,[a] and Pedro J. del Nido, MD[a]

Subprocedure	Optimal	Adequate	Inadequate
Proximal arch reconstruction	• No gradient • No evidence of coronary ischemia • Peak velocity $<$ 1.5 m/s by echo	• Mild gradient at proximal aorto-pulmonary anastomosis or proximal arch ($\Delta p \leq 20$ mm Hg by echo or by cath) • Peak velocity $\leq$ 2.5 m/s by echo • Successful intraop revision	• Need for reintervention during initial hospital stay • More than mild gradient ($\Delta p > 20$ mm Hg) • Peak velocity $>$ 2.5 m/s by echo • Clinical evidence (symptoms) of "neo-supra AS"
Distal arch reconstruction	• No narrowing or flow acceleration by echo • No BP gradient • Peak velocity $<$ 2 m/s by echo	• Peak velocity $\leq$ 3 m/s by echo • Mild gradient ($\Delta p \leq 20$ mm Hg by echo or by cath) • Successful intraop revision	• Need for reintervention during initial hospital stay • Peak velocity $>$ 3 m/s by echo • More than mild gradient ($\Delta p >$ 20 mm Hg) • Clinical evidence (symptoms) of coarctation
Coronary perfusion	• Unobstructed flow into proximal coronary arteries	• Unobstructed flow into proximal coronary arteries • Successful intraop revision	• Need for reintervention during initial hospital stay Evidence for obstructed coronary flow
Atrial septectomy	• No gradient • restrictive atrial septum left on purpose	• Mean gradient $\leq$ 4 mm Hg	• Need for reintervention during initial hospital stay • Mean gradient $>$ 4 mm Hg

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Subprocedure	Optimal	Adequate	Inadequate
Source of pulmonary blood flow			
a. Modified BT shunt	• Patent	• Patent • Downsizing of shunt because of pulmonary overcirculation	• Need for reintervention during initial hospital stay • Early symptomatic distortion of branch PAs • Symptomatic shunt narrowing (clot, suture line)
b. RV-PA conduit	• Patent	• Patent • Downsizing of shunt because of pulmonary overcirculation	• Need for reintervention during initial hospital stay • Early symptomatic distortion of branch PAs • Symptomatic conduit narrowing (clot, suture line)

BT, Blalock–Taussig; *RV-PA,* right ventricle–pulmonary artery. Reexploration for bleeding does not count as reintervention (ie, not coded as "inadequate").

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Stage I Norwood: Optimal technical performance improves outcomes irrespective of preoperative physiologic status or case complexity

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- 2004 – 2007
- 135 patients underwent stage I

TABLE 3. Technical performance scores and patient outcomes (n = 130)

	Overall	Optimal (n = 81)	Adequate (n = 26)	Inadequate (n = 23)
In-hospital mortality (no.)	19 (14.1%)	1 (1.2%)	5 (19.2%)	8 (34.8%)
Hospital stay (d, median and range)	18 (1–293)	15 (7–96)	20 (4–138)	46 (1–293)
Intensive care stay (d, median and range)	9 (1–293)	8 (3–60)	9 (3–58)	29 (1–293)
Ventilation time (d, median and range)	6 (1–293)	5 (1–47)	7 (2–58)	19 (1–293)
Major postoperative complications (no.)	34* (25.2%)	5 (6.2%)	9 (34.6%)	15 (65.2%)
Extracorporeal membrane oxygenation (no.)	25† (18.7%)	3 (3.7%)	5 (19.2%)	13 (56.5%)

All differences $P < .0001$. *Includes 5 patients without technical performance scores. †Includes 4 patients without technical performance scores.

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Summary

- HLHS
  38 weeks
  3.0 kg
  No risk factors such as
  - Intact or restrictive atrial septum
  - Additional cardiac anomalies
  - Non cardiac genetic malformation

- Norwood procedure with modified BT shunt