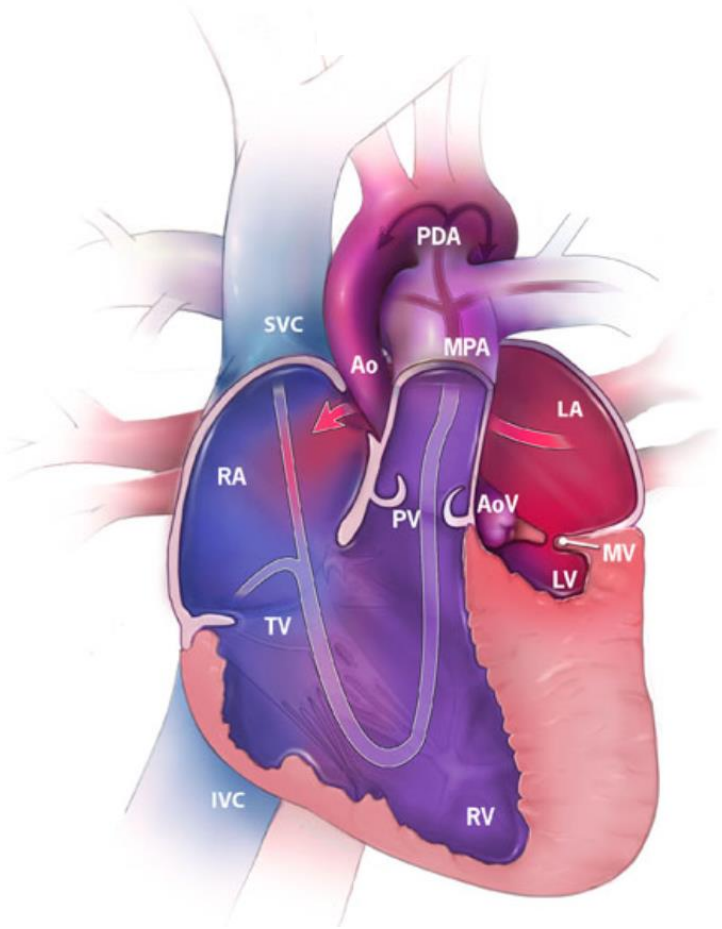


Norwood stage I

Bram van Wijk

Congenital cardio-thoracic surgeon

UMC Utrecht
Wilhelmina Kinderziekenhuis

Congenital cardiothoracic surgeons Utrecht

Prof Dr. Paul Schoof

Dr. Bram van Wijk

Dr. Hanna Talacua

Hypoplastic Left Heart Syndrome: Experience With Palliative Surgery

WILLIAM I. NORWOOD, MD, FACC*
JAMES K. KIRKLIN, MD*
STEPHEN P. SANDERS, MD†

Boston, Massachusetts

Aortic atresia is a form of hypoplastic left heart syndrome always complicated by associated severe hypoplasia of the ascending aorta and various degrees of mitral valve and left ventricular hypoplasia. At present it is a universally fatal lesion in early infancy. This is a report of a new palliative procedure for hypoplastic left heart syndrome that has resulted in early ongoing survival of two infants with aortic atresia. On the basis of experience with a third patient, an operation for future physiologic correction is proposed.

January 1980 The American Journal of CARDIOLOGY Volume 45 87

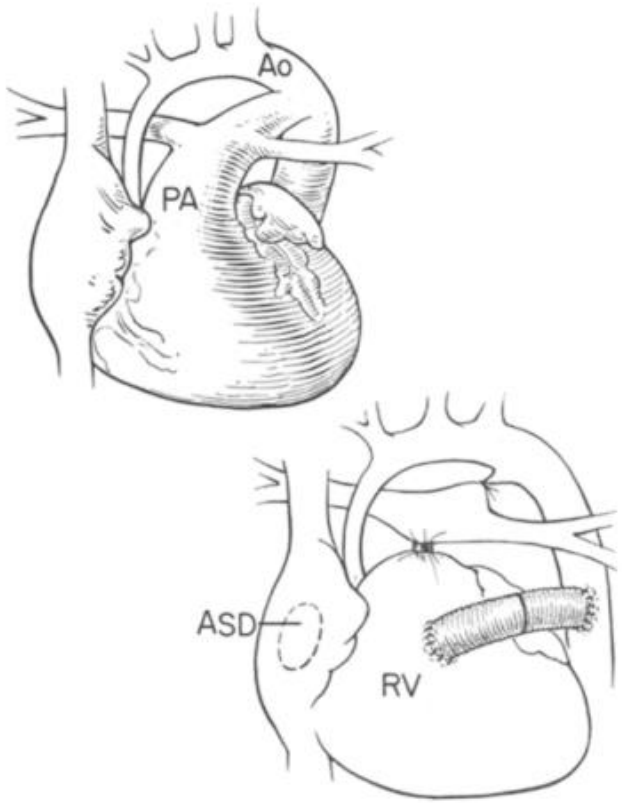


FIGURE 4. Case 3. Artist's conception of aortic atresia (**top**) and palliative procedure (**bottom**). Lower rendition illustrates ligation of patent ductus arteriosus, band on the main pulmonary artery (PA) and valved conduit between the right ventricular (RV) outflow tract and the descending aorta. Ao = aorta; ASD = atrial septal defect.

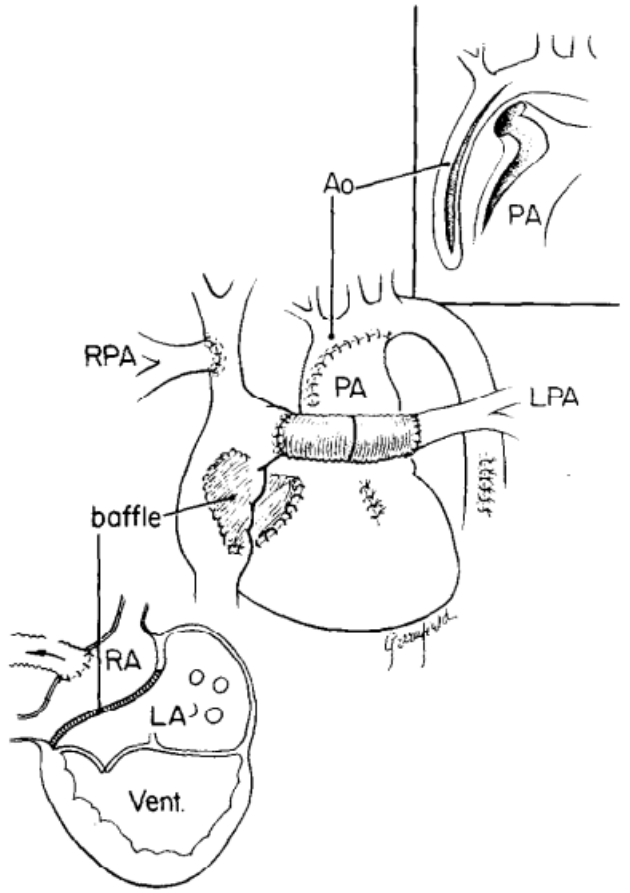


FIGURE 5. Artist's conception of proposed physiologic repair of aortic atresia (stage 2). The main pulmonary artery (PA) and the aorta (Ao) are anastomosed using the proximal stump of the right pulmonary artery (RPA) (**upper panel**). The right pulmonary artery is anastomosed end to side with the superior vena cava and a valved conduit placed between the right atrium (RA) and left pulmonary artery (LPA). An interatrial baffle ensures filling of the right ventricle (Vent.) with pulmonary venous blood from the left atrium (LA). A schematic drawing of the baffle and conduit is shown in the **lower panel.**

Norwood operation

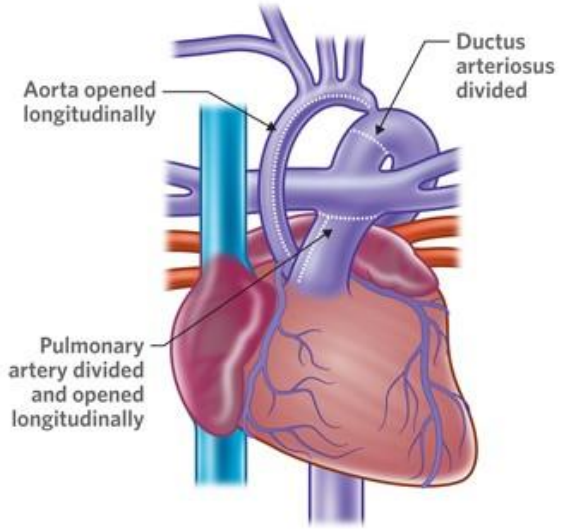
Basic principles:

Unobstructed systemic outflow of the right ventricle to a reconstructed aorta and coronary arteries

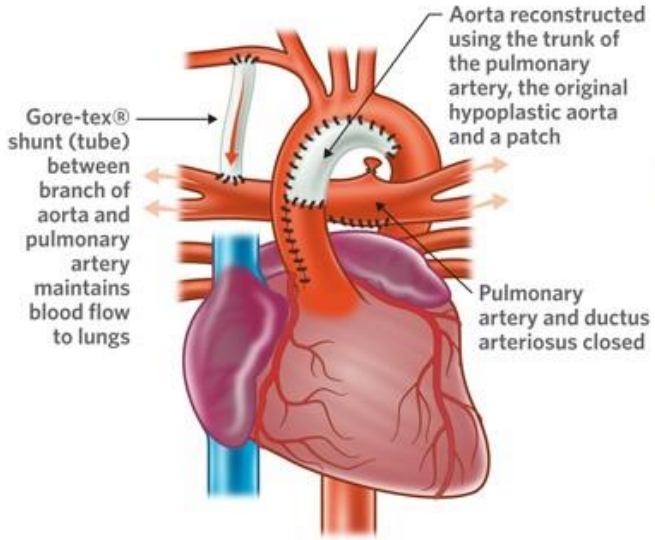
Unobstructed pulmonary venous return into the right atrium

Controlled pulmonary blood flow

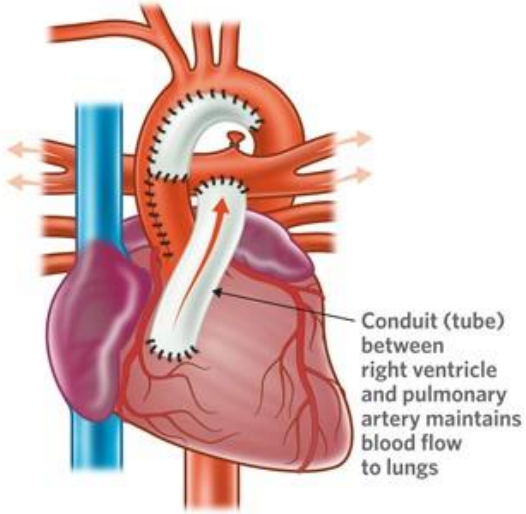
Norwood operation



Pre-operative anatomy



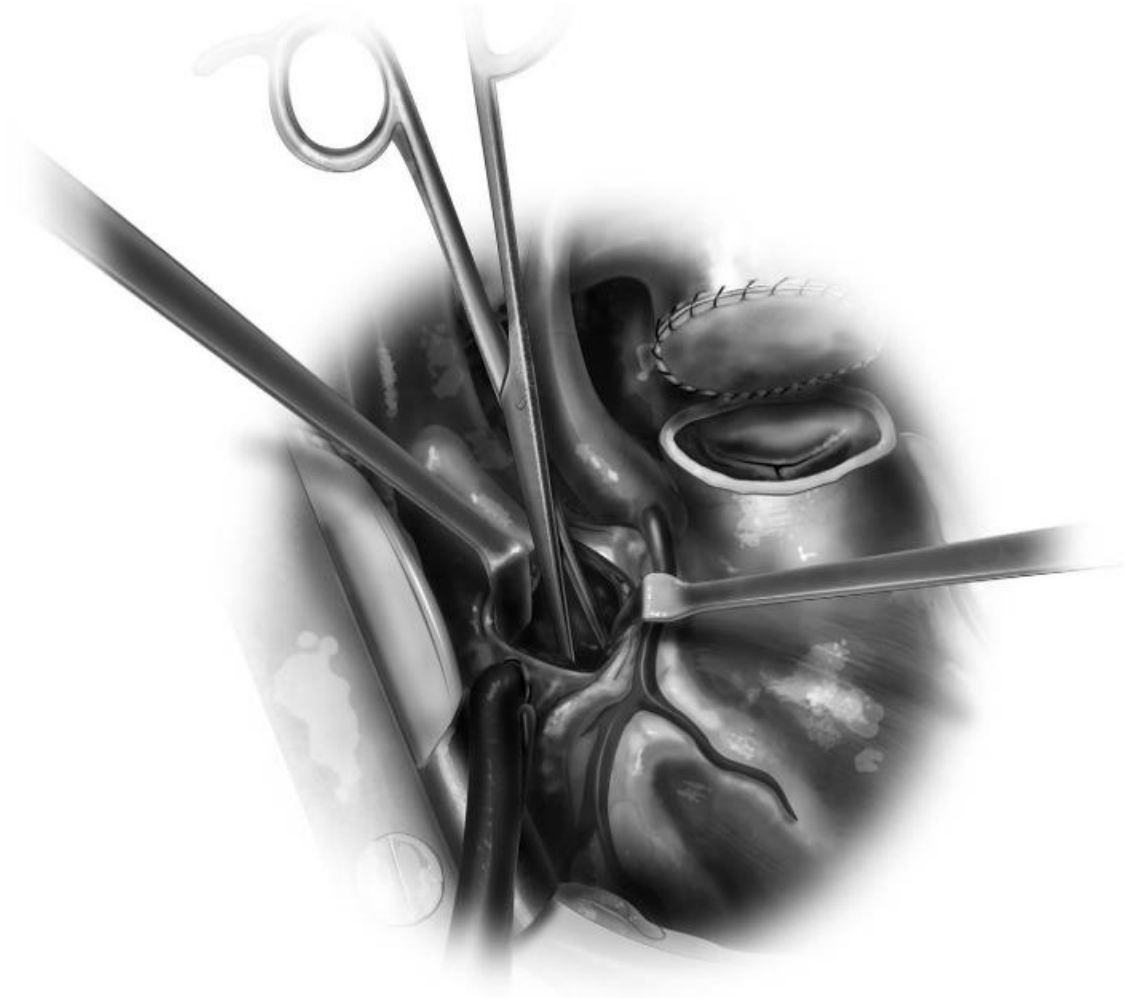
Norwood operation



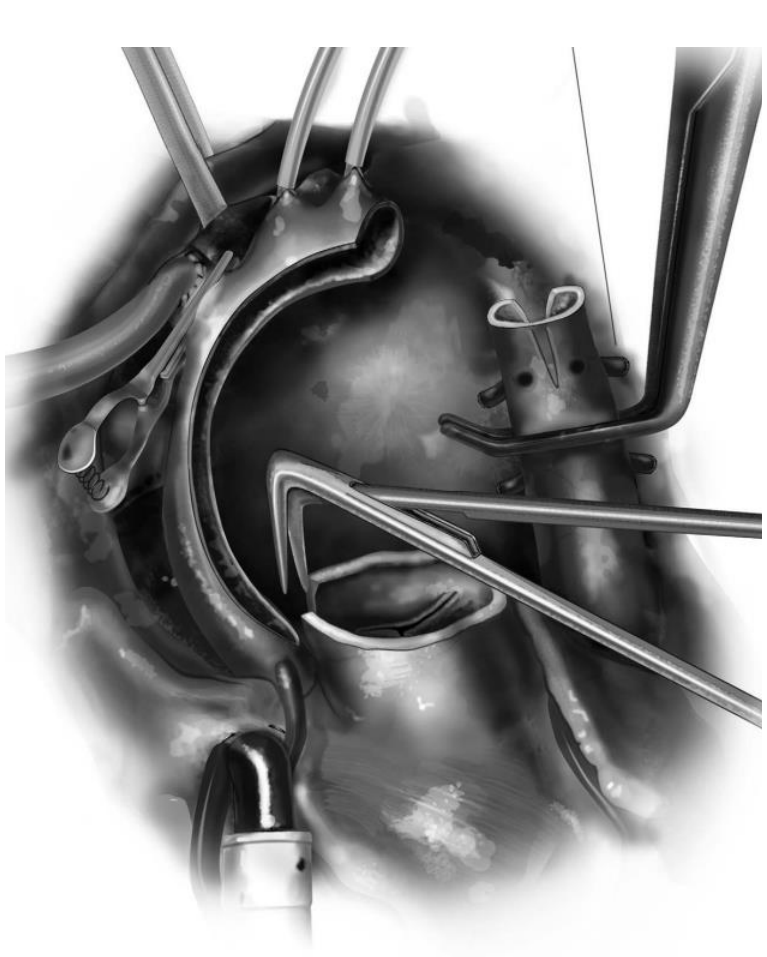
Norwood operation (Sano)

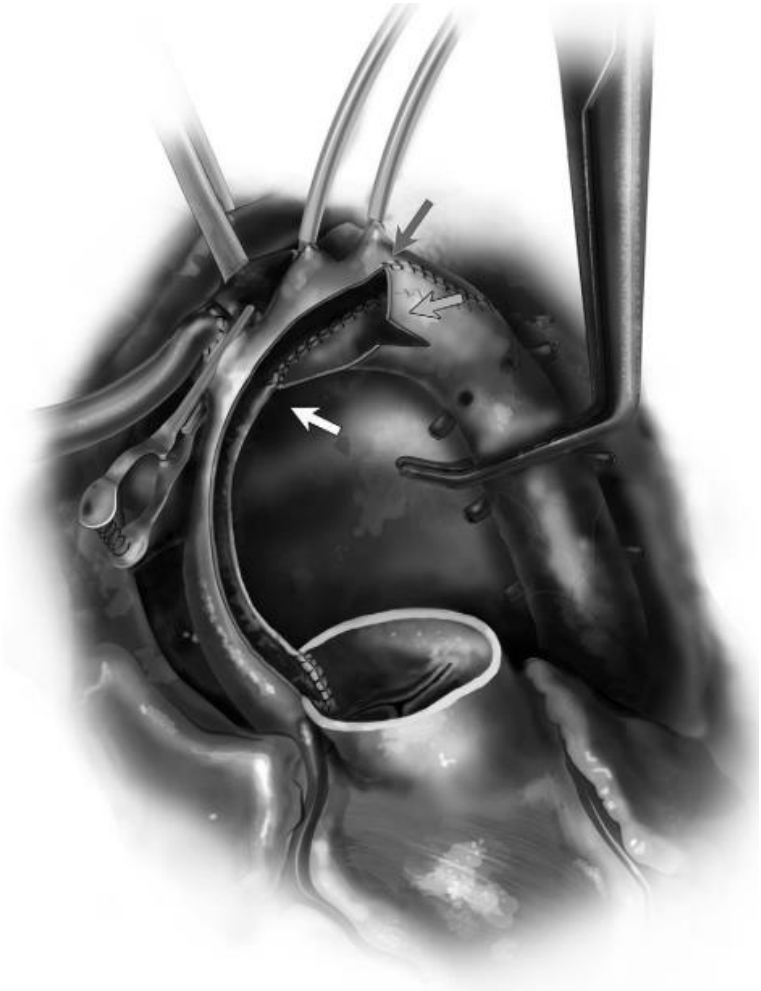
Unobstructed pulmonary venous return into the right atrium

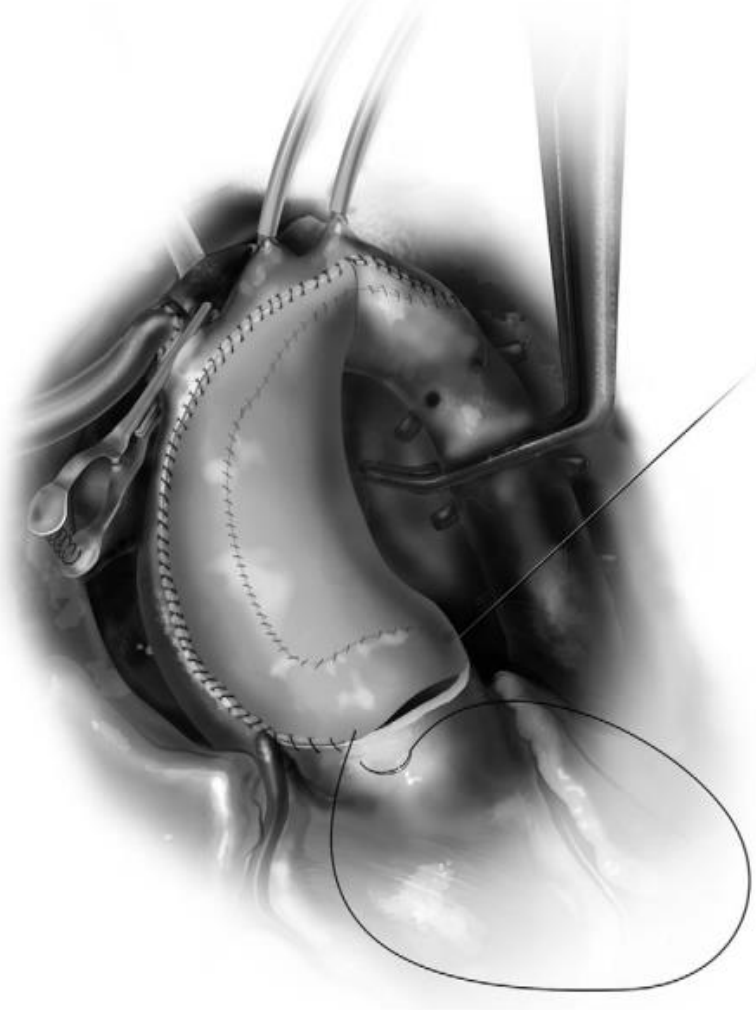
atrioseptectomy



Unobstructed systemic outflow of the right ventricle to a reconstructed aorta







Patch material

Pulmonary allograft (most used)

Cut out of a pulmonary bifurcation that shows a similar shape
Good sewing qualities
Expensive
Not always (accutely) available

Autoloog pericard (glutaraldehyde fixed)

Xeno pericard (Equine, porcine, bovine)

PTFE (Goretex)

Recoarctation After Norwood I Procedure for Hypoplastic Left Heart Syndrome: Impact of Patch Material

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Background. The development of recoarctation after the Norwood I procedure is a known complication in patients with hypoplastic left heart syndrome (HLHS).

Methods. We reviewed data on 194 consecutive patients with HLHS who underwent the Norwood I procedure between 2000 and 2015. All patients who survived until the second stage of the procedure were included. Recoarctation was defined by the need for intervention, either catheter based or surgical. The aim of our study was to determine if the patch material used for the initial arch reconstruction had an influence on the development of recoarctation.

Results. The study population consisted of 145 patients. The patch material used for aortic arch reconstruction was either a homograft (n = 87), autologous pericardium (n = 23), equine pericardium (n = 28), or other material (n = 7). Recoarctation was documented in 27 patients (18%) at a median time of 4.3 months (range, 1 to 28 months) after completion of Norwood I. Freedom from recoarctation was 85% ± 4%, 86% ± 7%, and 30% ± 22% at 2 years for homograft, autologous pericardium, and equine pericardium, respectively. Thirty-six percent of patients with equine pericardium exhibited recoarctation, significantly more than homograft patch ($p < 0.01$) or autologous pericardium ($p = 0.002$). Treatment for recoarctation was percutaneous balloon angioplasty in 19 patients, stent implantation in 3, and operative enlargement of the aortic arch in 5. On univariate analysis, arch reconstruction with equine pericardium was the only risk factor for recoarctation (hazard ratio, 6.0; 95% confidence interval, 2.6–14; $p < 0.01$).

Conclusions. The patch material used for reconstruction of the aortic arch in HLHS influences the rate of recoarctation following the Norwood I procedure. Equine pericardium cannot be recommended.

(Ann Thorac Surg 2017;103:617–21)

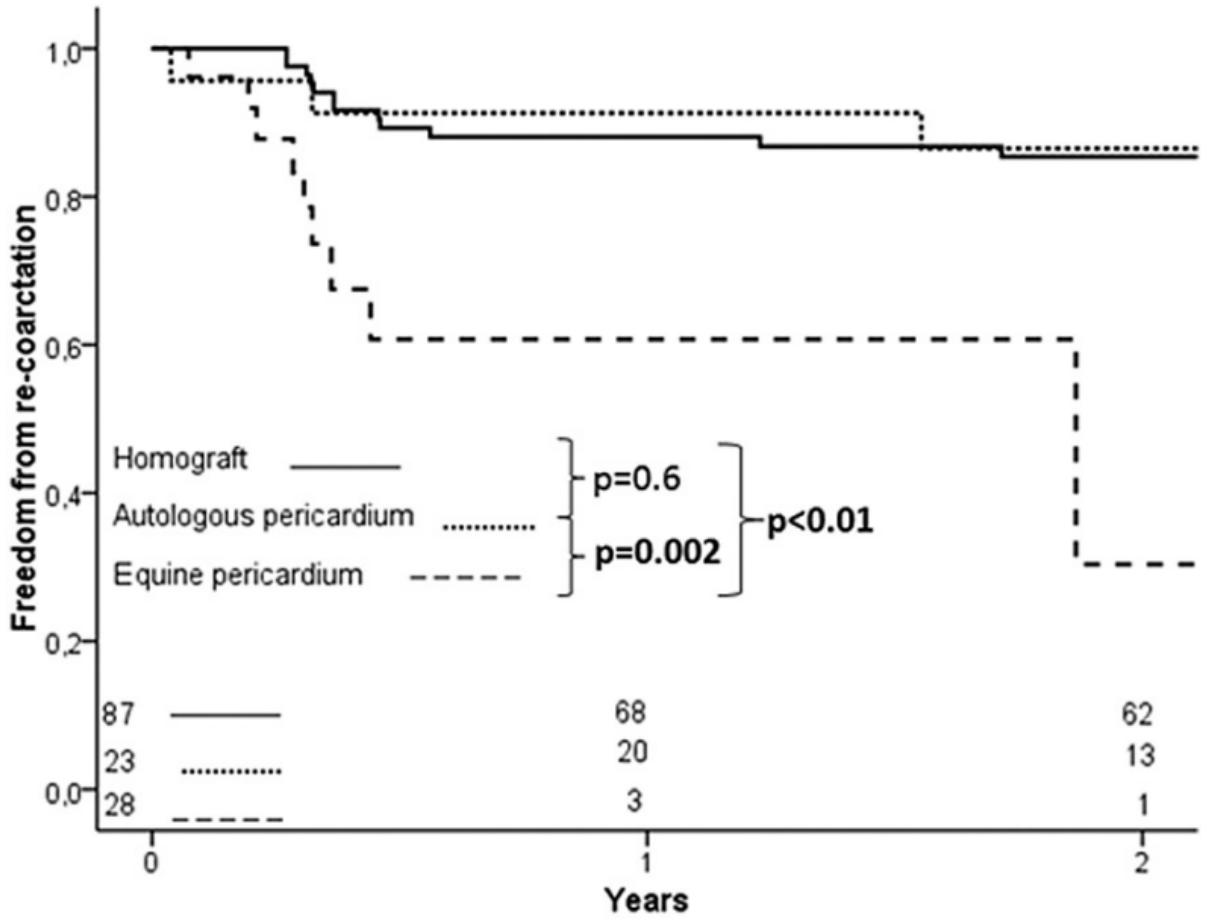

Fig 2. Freedom from recoarctation in 87 patients with homografts, 23 with autologous pericardium, and 28 with equine pericardium used for aortic arch reconstruction during the Norwood procedure.

Aortic arch reconstruction in the Norwood procedure using a curved polytetrafluorethylene patch

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Key question

Is a curved PTFE patch suitable for aortic arch enlargement in the Norwood procedure?

Key finding(s)

A curved polytetrafluorethylene patch has excellent perioperative results and a low restenosis rate.

Take-home message

The PTFE patch can be used as an alternative to a pulmonary homograft in patients with an ascending aorta of at least 3 mm.

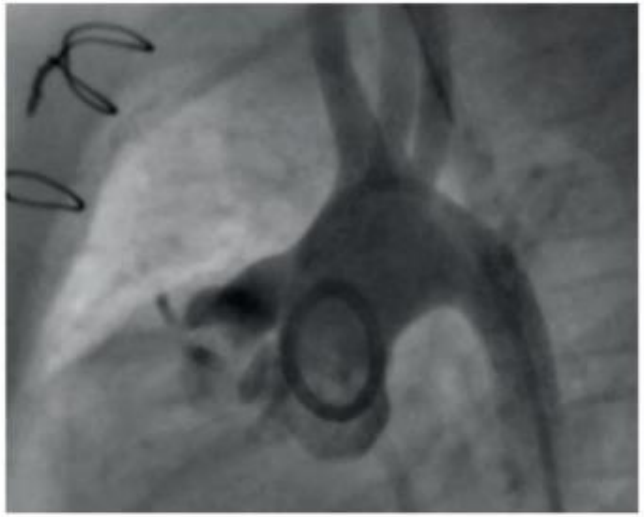


Table 3: Follow-up data

	PTFE group	Homograft group
N (%)	104 (100%)	120 (100%)
Median follow-up in years (range)	3.8 (0–11.4)	4.5 (0–11.3)
Late mortality, *n* (%)	6 (6%)	11 (9%)
10 years survival expectancy (%)	80,8%	78,1%
10 years survival: 95% confidence interval (%)	73,4–89,0%	70,8–86,2%
Median interval from Norwood until exitus in years (range)	0.2 (0,003–2.3)	0.2 (0-4.5)
HTX	0	2
Aortic arch restenosis	1	4
Aortic arch intervention only	1	1
Aortic arch reoperation	0	3
Median interval from Norwood until aortic arch restenosis treatment in years	1	0,3
Endocarditis	0	0
Aortic arch aneurysm	0	0

PTFE: polytetrafluorethylene.

Shape of the patch

E. Sames-Dolzer *et al.* / European Journal of Cardio-Thoracic Surgery

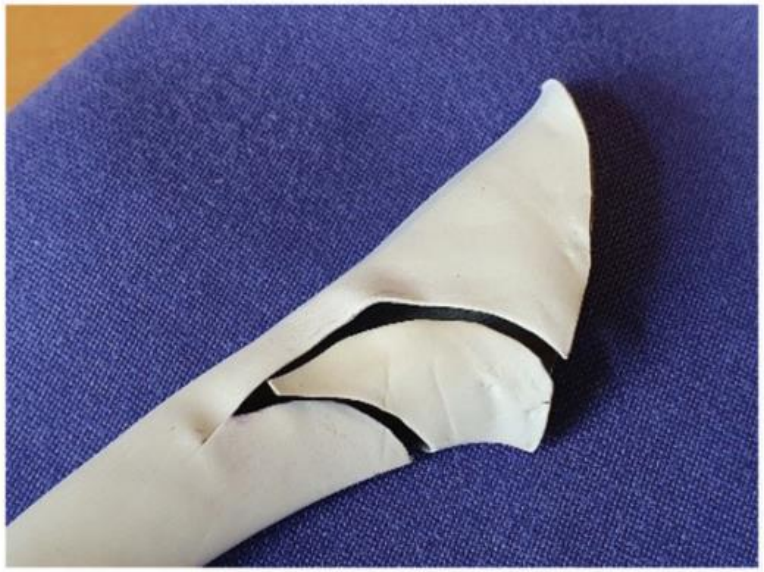


Figure 1: Polytetrafluorethylene prosthesis.

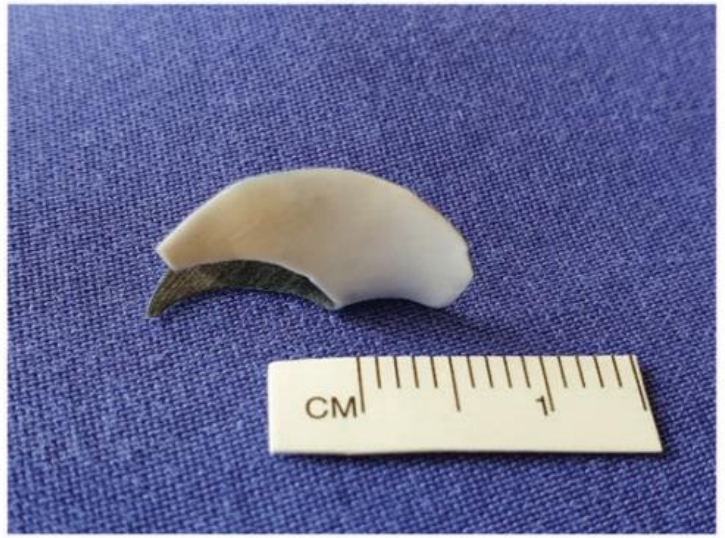
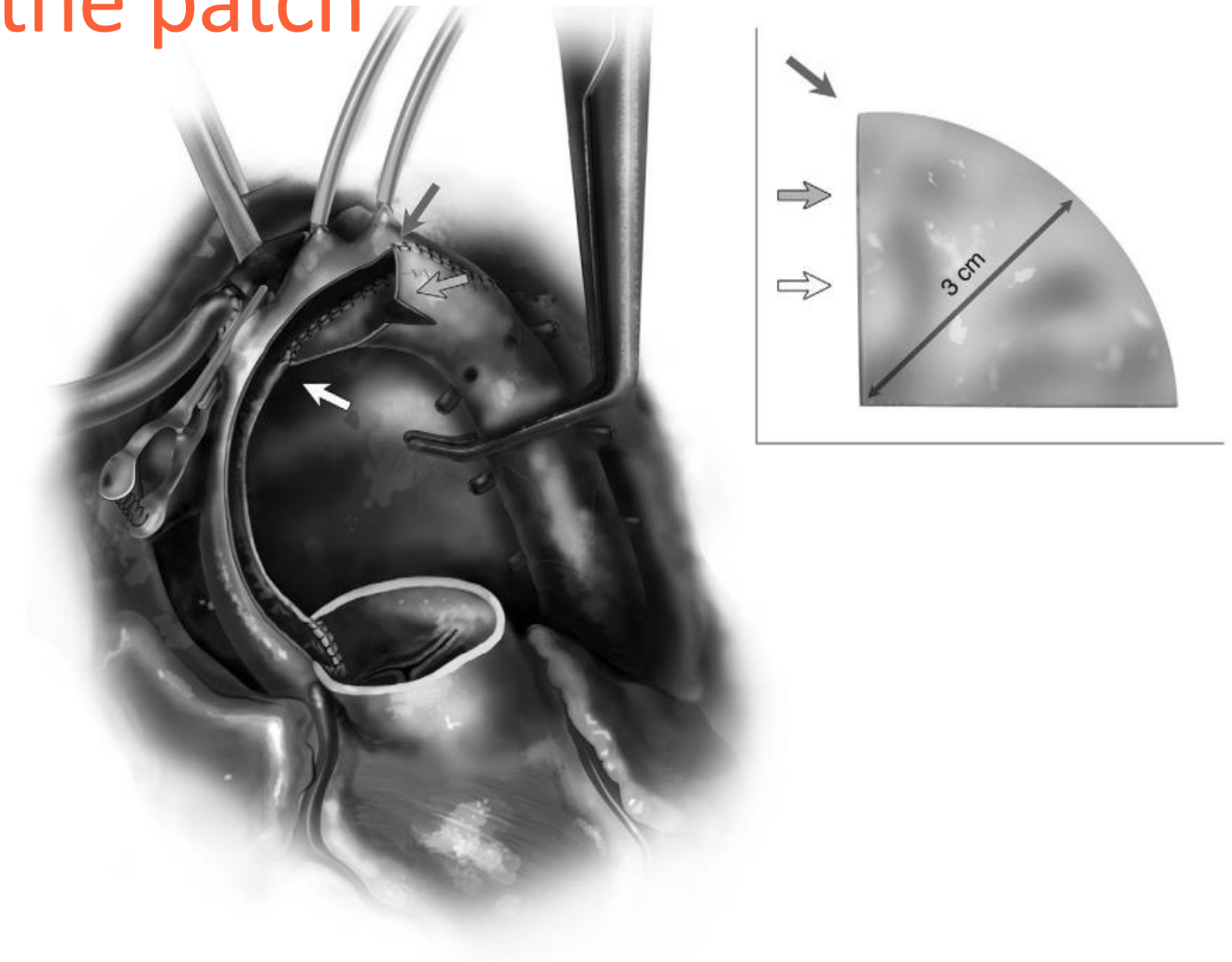


Figure 2: Polytetrafluorethylene patch.

Dynaflo® bypass graft; funnel shaped end

Shape of the patch



Operative Techniques in Thoracic and Cardiovascular Surgery

J.S. Tweddell et al.

Figure 10 Arch reconstruction, continued. A patch is used for completion of the arch reconstruction and creation of the neoascending aorta. This patch is a quarter circle with a radius of 3 cm and can be pulmonary homograft, pericardium, or porcine small intestinal submucosa (CorMatrix, Alpharetta, GA). The straight edge will be sewn down the left edge of the ascending aorta; the arrows in the figure indicate the alignment.

Size of the patch

Too small:
AortoPulmonary window narrowing
Residual ductal tissue; Recoartation rate 10-36% after Norwood I

Too large:
Kinking neck vessels
Folds in the patch which cause obstruction of the aortic arch
LPA/RPA compression

Technique of the arch reconstruction

Recoarctation After the Norwood I Procedure for Hypoplastic Left Heart Syndrome: Incidence, Risk Factors, and Treatment Options

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Departments of Cardiovascular Surgery and Pediatric Cardiology and Congenital Heart Disease, German Heart Center Munich, Technische Universität München, Munich, Germany

Background. Early survival after the Norwood I procedure has improved over the years, but subsequent morbidity is not yet well assessed. The aim of this study was to review the incidence of recoarctation, evaluate risk factors, and analyze treatment options.

Methods. We reviewed the medical records of 124 consecutive patients with hypoplastic left heart syndrome (HLHS) who underwent the Norwood I procedure. Reconstruction of the aortic arch was performed in a standardized manner, removing all visible ductal tissue and enlarging the distal anastomosis with a Y incision into the descending aorta. Angiographic assessment with measurement of the peak gradient across the aortic arch was performed before the second stage was performed.

Results. Recoarctation of the aorta was documented in 13 patients (13.4%) at a mean time of 6.4 ± 5 months after the Norwood procedure. One patient died before the recoarctation could be treated. Right ventricular function was good in all except 1 patient at the time of diagnosis. Ten patients underwent 16 percutaneous balloon angioplasties, and 2 patients underwent operative enlargement of the neoaorta. The pretreatment peak gradient of 24.1 ± 16 mm Hg (10–64 mm Hg) across the aortic arch was significantly reduced to 6.3 ± 4 mm Hg (0–14 mm Hg) after angioplasty or operation ($p = 0.003$). There were no procedure-related deaths. No risk factor for recoarctation could be identified.

Conclusions. A standardized surgical technique for reconstruction of the aorta leads to a low recoarctation rate. Balloon angioplasty can be performed in the majority of patients before the second-stage procedure.

(Ann Thorac Surg 2013;95:935–40)

Several options for reconstructing the aortic arch, in terms of patch material used and anastomosing techniques, have been described [9, 10]. Searching for a mechanism that may be responsible for the development of recoarctation, different possibilities have been discussed. Machii and associates [11] suggested that recoarctation is caused by ductal tissue remaining in the aorta. This is in accordance with other authors who recommend complete elimination of all ductal tissue during the Norwood I procedure [4, 5]. However as Machii and colleagues demonstrated, ductal tissue might expand out onto the inner wall of the aorta and may be detectable only by microscopy. Therefore in a recent postmortem study, which revealed that 33% of patients show inadequate resection of the ductal tissue located distal to the patch material after the Norwood I procedure, the authors recommended extending the patch further into the descending aorta [12]. It is also our aim to remove all visible ductal tissue during the Norwood I procedure, but we might miss some of it, as was the case in 1 patient who underwent a second operation. Additionally, we widen our anastomosis through a Y incision. We think that this magnification leads to a lower rate of recoarctation. Another suggestion for improving the surgical results is the "interdigitating technique," in which an incision is made in the anterior and posterior wall of

Intervention for arch obstruction after the Norwood procedure: Prevalence, associated factors, and practice variability

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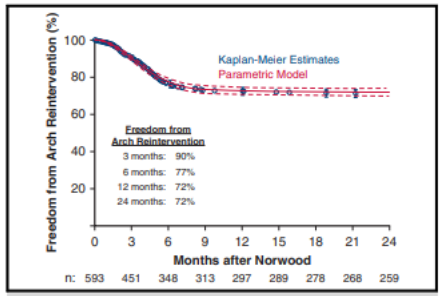
ABSTRACT

Objective: Arch obstruction after the Norwood procedure is common and contributes to mortality. We determined the prevalence, associated factors, and practice variability of arch reintervention and assessed whether arch reintervention is associated with mortality.

Methods: From 2005 to 2017, 593 neonates in the Congenital Heart Surgeons' Society Critical Left Heart Obstruction cohort underwent a Norwood procedure. Median follow-up was 3.7 years. Multivariable parametric models, including a modulated renewal analysis, were performed.

Results: Of the 593 neonates, 146 (25%) underwent 218 reinterventions for arch



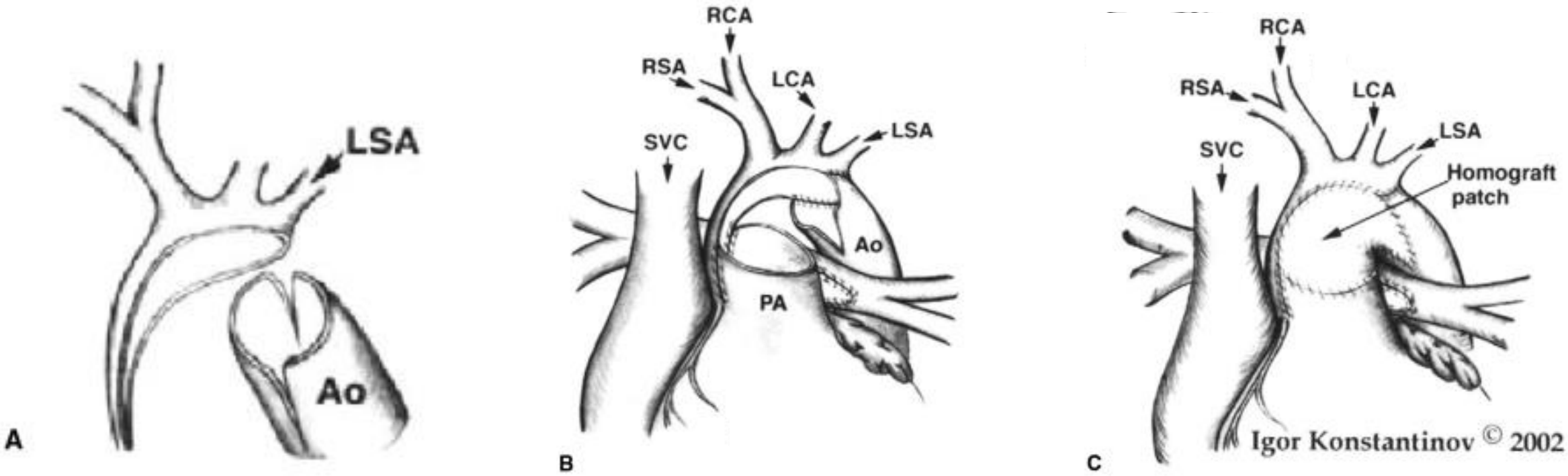
Freedom from arch reintervention after the Norwood procedure.

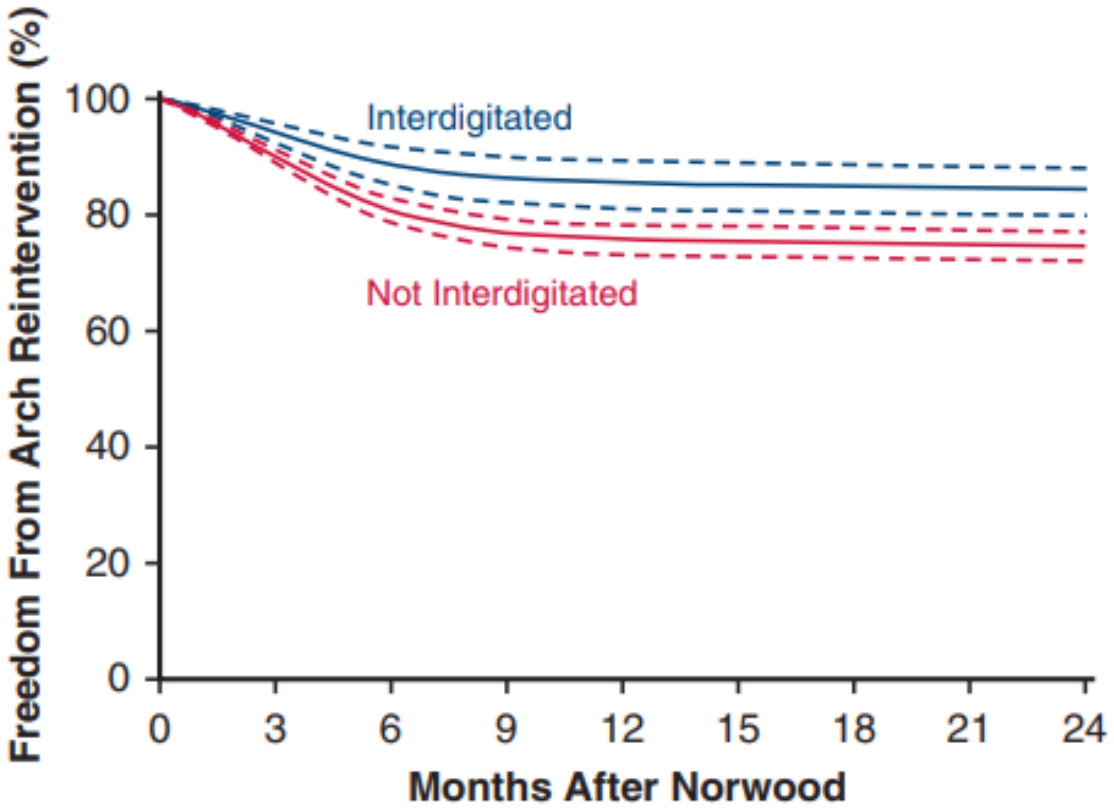
The Journal of Thoracic and Cardiovascular Surgery · February 2019

Interdigitation of the aortic arch anastomosis

Arch reintervention occurred in 25% the cohort. Interdigitation of the distal aortic anastomosis reduced the rate of arch reintervention.

Interdigitation of the aortic arch anastomosis



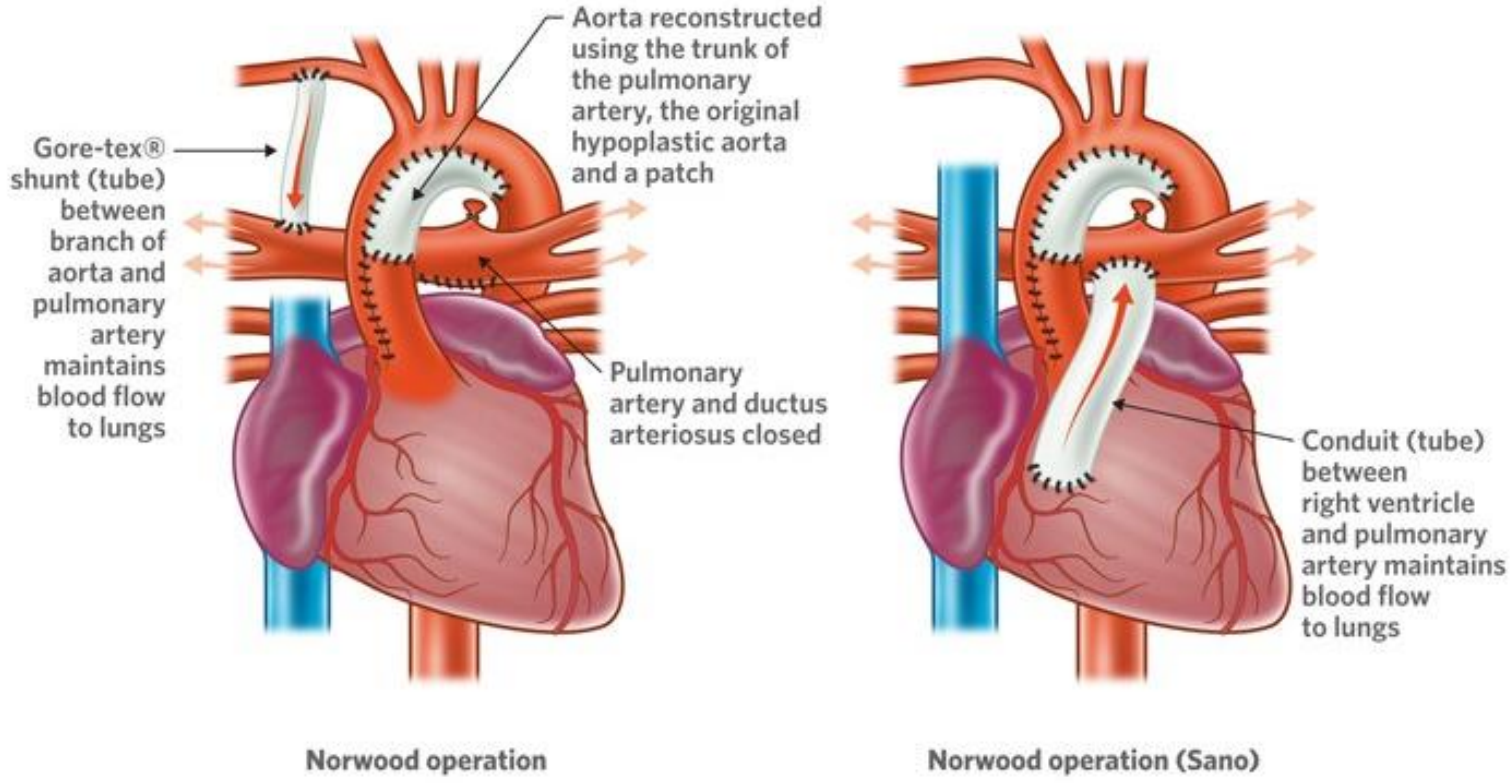


An interdigitating arch anastomosis protected against subsequent reintervention; 11% vs 25% with and without interdigitation.

Unobstructed systemic outflow

- Material of the patch: homograft, PTFE (Goretex)
- Size and shape of the patch
- Complete ductal removal
- Y incision or Interdigitation of the aortic arch anastomoses

Pulmonary blood flow; mBT versus RV-PA Shunt

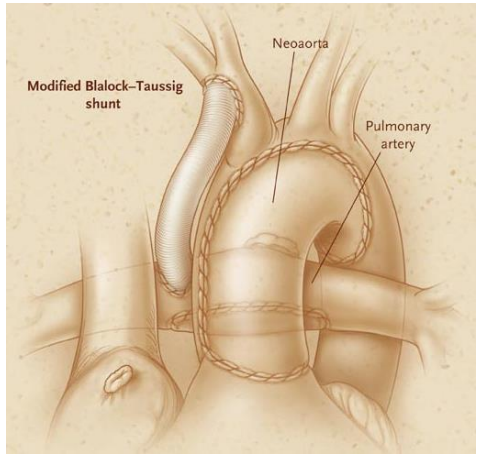


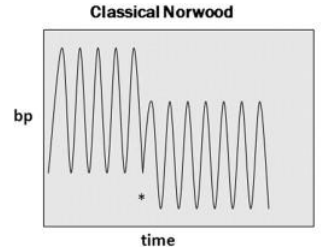
Pulmonary blood flow; mBT versus RV-PA Shunt

Modified BT-Shunt:

- Shunt from systemic to PA with continuous blood flow (syst/diast)

- Shunt size highly important
- Small size (3-3,5 mm) limit pulmonary bloodflow and diastolic hypotension



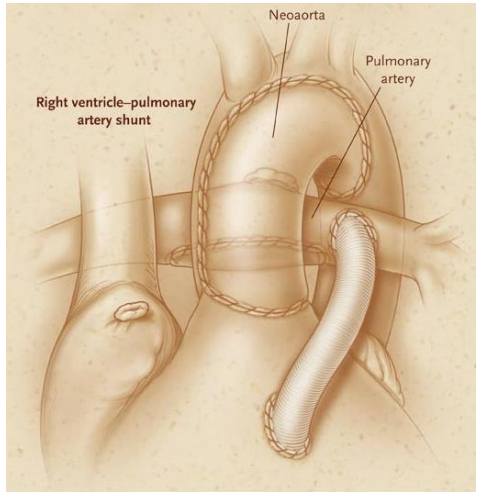


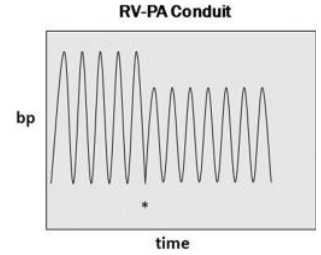
- Diastolic run-off, low diastolic BP and decreased CPP.
- RV-overload ↑, wall stress ↑ → RV dysfunction with low CO

RV-PAS (Sano):

- Systolic blood flow to the PA
- At the cost of a venticulotomy in the systemic ventricle
- Concern about short and longterm consequences.

- Greater need for cardiac reintervention (shunt stenosis)
- Poorer growth of branch pulmonary arteries





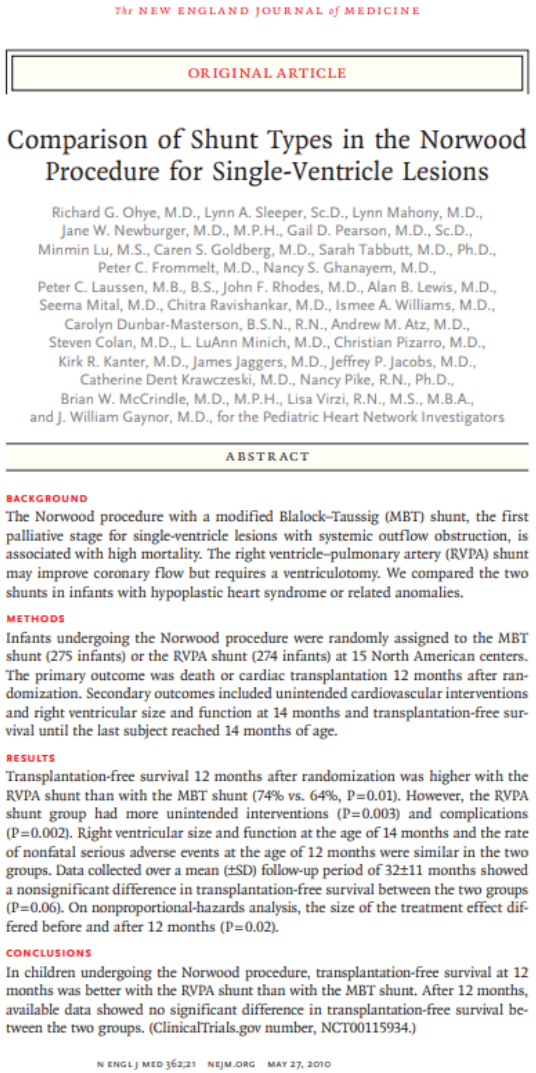
- Eliminates diastolic run-off.
- Maintenance of diastolic pressure with higher CPP.

Barron, Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual, Volume 16, Issue 1, 2013, 52–58

Pulmonary blood flow; mBT versus RV-PA Shunt

Single Ventricle Reconstruction trial

- 2005-2008
- 15 North American centers
- 555 neonates with HLHS or FSRV lesions
- 275 MBTS vs 274 RV-PAS (randomized)
- Primary outcome: combined endpoint of death or HTX 12 months after randomization



The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

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

Richard G. Ohye, M.D., Lynn A. Sleeper, Sc.D., Lynn Mahony, M.D., Jane W. Newburger, M.D., M.P.H., Gail D. Pearson, M.D., Sc.D., Minmin Lu, M.S., Caren S. Goldberg, M.D., Sarah Tabbutt, M.D., Ph.D., Peter C. Frommelt, M.D., Nancy S. Ghanayem, M.D., Peter C. Laussen, M.B., B.S., John F. Rhodes, M.D., Alan B. Lewis, M.D., Seema Mital, M.D., Chitra Ravishankar, M.D., Ismee A. Williams, M.D., Carolyn Dunbar-Masterson, B.S.N., R.N., Andrew M. Atz, M.D., Steven Colan, M.D., L. LuAnn Minich, M.D., Christian Pizarro, M.D., Kirk R. Kanter, M.D., James Jaggers, M.D., Jeffrey P. Jacobs, M.D., Catherine Dent Krawczeski, M.D., Nancy Pike, R.N., Ph.D., Brian W. McCrindle, M.D., M.P.H., Lisa Virzi, R.N., M.S., M.B.A., and J. William Gaynor, M.D., for the Pediatric Heart Network Investigators

ABSTRACT

BACKGROUND

The Norwood procedure with a modified Blalock–Taussig (MBT) shunt, the first palliative stage for single-ventricle lesions with systemic outflow obstruction, is associated with high mortality. The right ventricle–pulmonary artery (RVPA) shunt may improve coronary flow but requires a ventriculotomy. We compared the two shunts in infants with hypoplastic heart syndrome or related anomalies.

METHODS

Infants undergoing the Norwood procedure were randomly assigned to the MBT shunt (275 infants) or the RVPA shunt (274 infants) at 15 North American centers. The primary outcome was death or cardiac transplantation 12 months after randomization. Secondary outcomes included unintended cardiovascular interventions and right ventricular size and function at 14 months and transplantation-free survival until the last subject reached 14 months of age.

RESULTS

Transplantation-free survival 12 months after randomization was higher with the RVPA shunt than with the MBT shunt (74% vs. 64%, P=0.01). However, the RVPA shunt group had more unintended interventions (P=0.003) and complications (P=0.002). Right ventricular size and function at the age of 14 months and the rate of nonfatal serious adverse events at the age of 12 months were similar in the two groups. Data collected over a mean (±SD) follow-up period of 32±11 months showed a nonsignificant difference in transplantation-free survival between the two groups (P=0.06). On nonproportional-hazards analysis, the size of the treatment effect differed before and after 12 months (P=0.02).

CONCLUSIONS

In children undergoing the Norwood procedure, transplantation-free survival at 12 months was better with the RVPA shunt than with the MBT shunt. After 12 months, available data showed no significant difference in transplantation-free survival between the two groups. (ClinicalTrials.gov number, NCT00115934.)

Pulmonary blood flow; mBT versus RV-PA Shunt

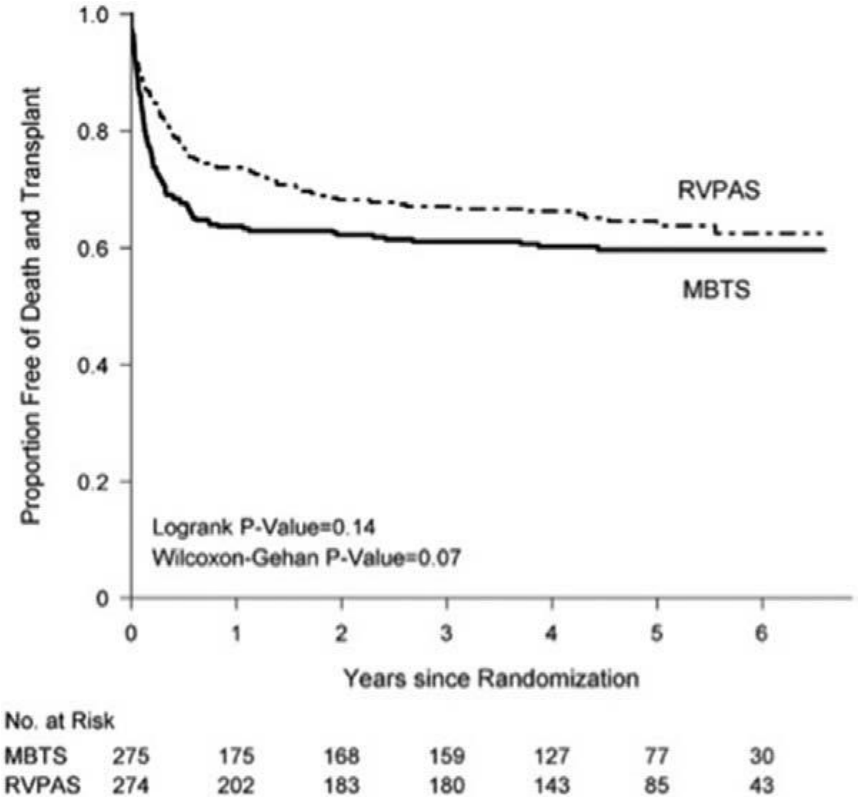
Results SVR trial

At 12 months:

- RV-PAS: 74% transplantation free survival
- MBTS: 64% transplantation free survival (p=0,01)

At 32 ± 11 months:

- No significant transplantation-free survival difference (p=0,06)
- Anatomic subtype (AA, AS, MA, MS) did not impact outcome
- RV-function at 14 months similar
- RV-PAS required more unplanned surgical or catheter interventions (p=0,003) (shunt stenosis, branch PA's, neoarta)



Circulation

ORIGINAL RESEARCH ARTICLE

Transplant-Free Survival and Interventions at 6 Years in the SVR Trial

Editorial, see p 2254

BACKGROUND: In the SVR trial (Single Ventricle Reconstruction), 1-year transplant-free survival was better for the Norwood procedure with right ventricle-to-pulmonary artery shunt (RVPAS) compared with a modified Blalock–Taussig shunt in patients with hypoplastic left heart and related syndromes. At 6 years, we compared transplant-free survival and other outcomes between the groups.

METHODS: Medical history was collected annually using medical record review, telephone interviews, and the death index. The cohort included 549 patients randomized and treated in the SVR trial.

RESULTS: Transplant-free survival for the RVPAS versus modified Blalock–Taussig shunt groups did not differ at 6 years (64% versus 59%, P=0.25) or with all available follow-up of 7.1±1.6 years (log-rank P=0.13). The RVPAS versus modified Blalock–Taussig shunt treatment effect had nonproportional hazards (P=0.009); the hazard ratio (HR) for death or transplant favored the RVPAS before stage II surgery (HR, 0.66; 95% confidence interval, 0.48–0.92). The effect of shunt type on death or transplant was not statistically significant between stage II to Fontan surgery (HR, 1.36; 95% confidence interval, 0.86–2.17; P=0.17) or after the Fontan procedure (HR, 0.76; 95% confidence interval, 0.33–1.74; P=0.52). By 6 years, patients with RVPAS had a higher incidence of catheter interventions (0.38 versus 0.23/patient-year, P<0.001), primarily because of more interventions between the stage II and Fontan procedures (HR, 1.72; 95% confidence interval, 1.00–3.03). Complications did not differ by shunt type; by 6 years, 1 in 5 patients had had a thrombotic event, and 1 in 6 had had seizures.

CONCLUSIONS: By 6 years, the hazards of death or transplant and catheter interventions were not different between the RVPAS versus modified Blalock–Taussig shunt groups. Children assigned to the RVPAS group had 5% higher transplant-free survival, but the difference did not reach statistical significance, and they required more catheter interventions. Both treatment groups have accrued important complications.

CLINICAL TRIAL REGISTRATION: URL: https://www.clinicaltrials.gov. Unique identifier: NCT00115934.

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*Investigators and centers in the SVR Trial (Single Ventricle Reconstruction) are listed in the Appendix in the online-only Data Supplement.

Key Words: cardiac surgery ■ congenital heart defect ■ congenital heart disease ■ Norwood procedure ■ single ventricle

Sources of Funding, see page 2252

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Circulation

ORIGINAL RESEARCH ARTICLE

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CLINICAL TRIAL REGISTRATION: URL: https://www.clinicaltrials.gov. Unique identifier: NCT00115934.

Clinical Perspective

What Is New?

- We compared transplant-free survival and other outcomes at 6 years after the Norwood procedure with right ventricle-to-pulmonary artery shunt compared with a modified Blalock–Taussig shunt in children enrolled in the SVR trial (Single Ventricle Reconstruction).
- The right ventricle-to-pulmonary artery shunt group had similar transplant-free survival at 6 years but required more catheter interventions before the Fontan procedure.
- Right ventricular ejection fraction, New York Heart Association class, and complications did not differ by shunt type.
- Cumulative incidence of morbidities by 6 years included 20% with a thrombotic event, 15% with a seizure, and 7.5% with a stroke.

What Are the Clinical Implications?

- The right ventricle-to-pulmonary artery shunt strategy carries a survival advantage before stage II surgery but a greater hazard of catheter interventions until the Fontan procedure is performed.
- After the Fontan procedure, there is no sustained advantage of the initial systemic to pulmonary artery shunt on transplant-free survival or catheter intervention.
- Morbidity begins early in life and steadily increases for children in both shunt groups.
- These data emphasize the importance of continued follow-up of this cohort and the need to find new strategies to improve the long-term outlook for those with single ventricle anomalies.

Circulation. 2018;137:2246–2253. DOI: 10.1161/CIRCULATIONAHA.117.029375

Influence of Shunt Type on Survival and Right Heart Function after the Norwood Procedure for Aortic Atresia

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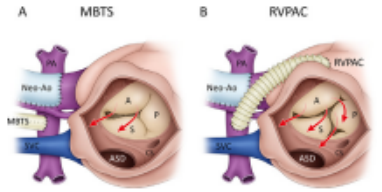
The study objective was to compare the results after Norwood procedure between modified Blalock-Taussig shunt (MBTS) and right ventricle-to-pulmonary artery conduit (RVPAC) according to Sano in patients with hypoplastic left heart syndrome (HLHS) and aortic atresia (AA). A total of 146 neonates with HLHS and AA who underwent the Norwood procedure at our institution between 2001 and 2020 were divided into 2 groups according to shunt type (MBTS or RVPAC). Survival after the Norwood procedure was compared between the groups. Longitudinal right ventricular and tricuspid valve function in each group were evaluated using cubic splines method. RVPAC was performed in 103 patients and MBTS in 43 according to surgeon preference. There were no differences in the 30-day mortality rates (16.5% vs 16.3%, $P = 0.973$). Survival at 0.5, 1 and 3 years was 79.6%, 74.6%, and 68.9% in RVPAC and 66.8%, 64.3%, and 58.5% in MBTS ($P = 0.293$). Among 23 patients undergoing tricuspid valve procedure, different mechanisms of tricuspid regurgitation were observed between the groups. Longitudinal analysis revealed greater prevalence of late right ventricular dysfunction in RVPAC patients. In 77 patients who completed Fontan procedure, the postoperative N-terminal pro B-type natriuretic peptide value was significantly higher in RVPAC vs MBTS (554 vs 276 ng/L, $P = 0.007$). No survival advantage of RVPAC over MBTS was observed in neonates with HLHS and AA undergoing the Norwood procedure. Longitudinal analysis demonstrated a greater prevalence of right ventricular dysfunction and higher N-terminal pro B-type natriuretic peptide values during late follow-up in patients with RVPAC.

Semin Thoracic Surg 34:1300–1310

Keywords: Hypoplastic left heart syndrome, Aortic atresia, Modified Blalock-Taussig shunt, Right ventricle to pulmonary artery conduit, Ventricular function, Tricuspid regurgitation



Different mechanisms of tricuspid valve regurgitation in MBTS and RVPAC.

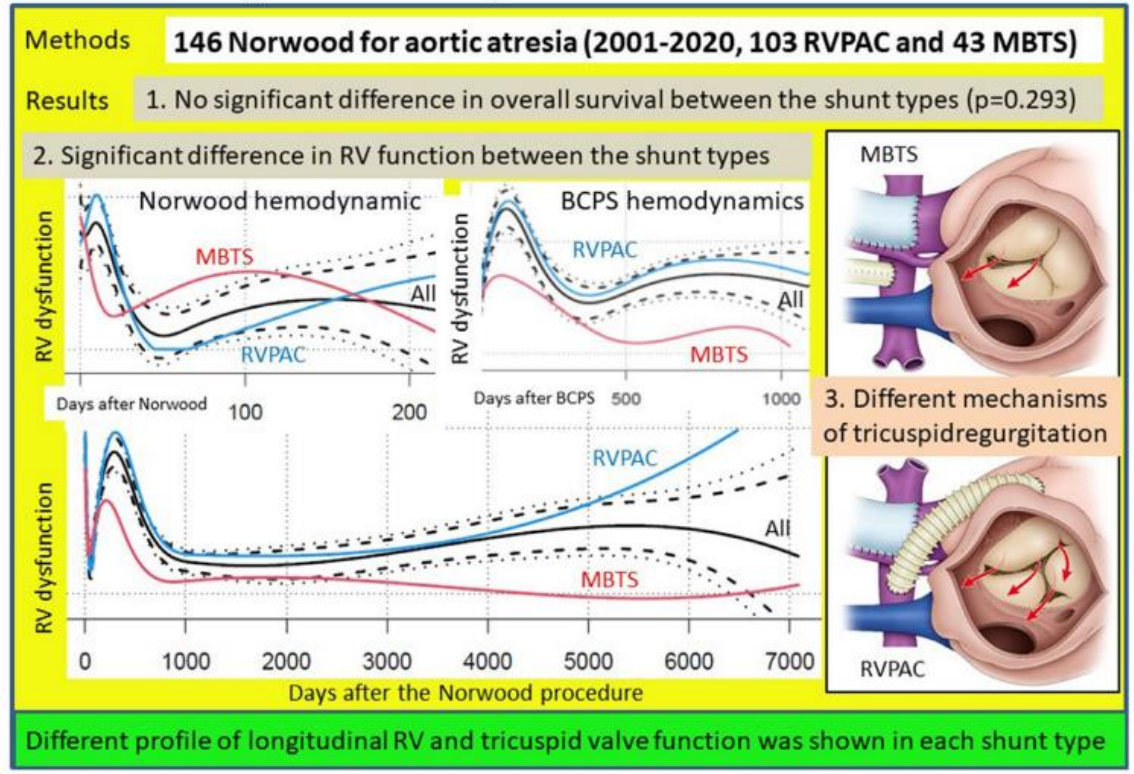
Central Message

Right ventricle to pulmonary artery conduit showed late RV dysfunction and tricuspid regurgitation more often than modified BT shunt after Norwood procedure in patients with HLHS and aortic atresia.

Perspective Statement

No survival advantage of RVPAC over MBTS was observed after the Norwood procedure in patients with HLHS and aortic atresia. Long-term results could be improved by (1) refining the surgical and medical management after the Norwood procedure with MBTS, and (2) introducing medical or novel surgical therapies aimed at improving right ventricular function after stage II palliation in patients with RVPAC.

Influence of shunt type on survival and right heart function after the Norwood for aortic atresia



RV(PAC): right ventricle (to pulmonary artery conduit), MBTS: modified Blalock-Taussig shunt, BCPS: bidirectional cavopulmonary shunt

Influence of Shunt Type on Survival and Right Heart Function after the Norwood Procedure for Aortic Atresia

Nicole Piber, MD,[*,#] *Masam*
Melchior Burri, MD,[†] *Christo*
Julia Lemmer, MD,[‡] *Daniel L*
Peter Ewert, MD, PhD,[‡] *Alfr*

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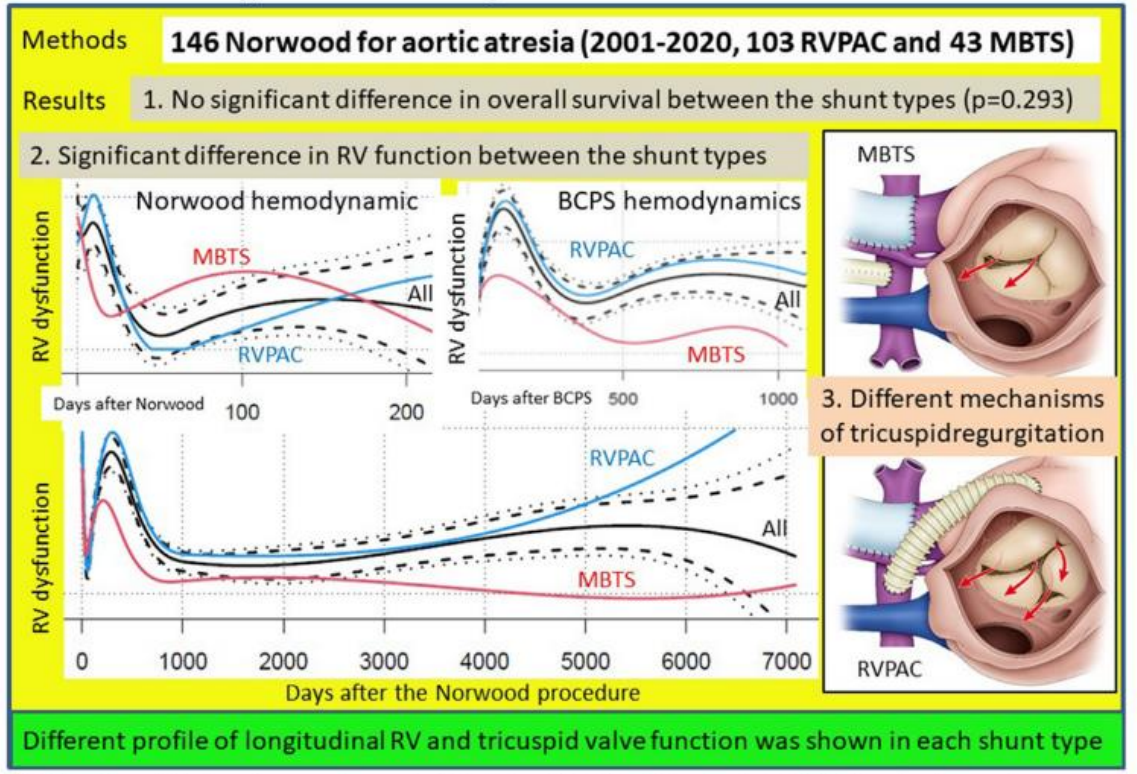
Influence of shunt type on survival and right heart function after the Norwood for aortic atresia

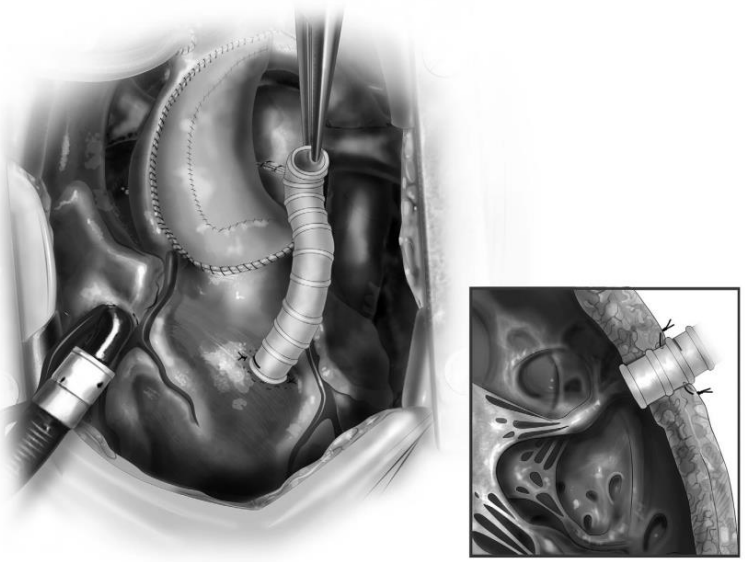


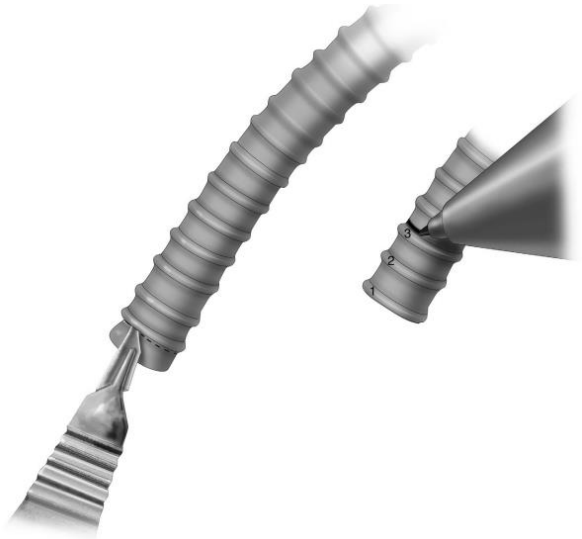
RV(PAC): right ventricle (to pulmonary artery conduit), MBTS: modified Blalock-Taussig shunt, BCPS: bidirectional cavopulmonary shunt

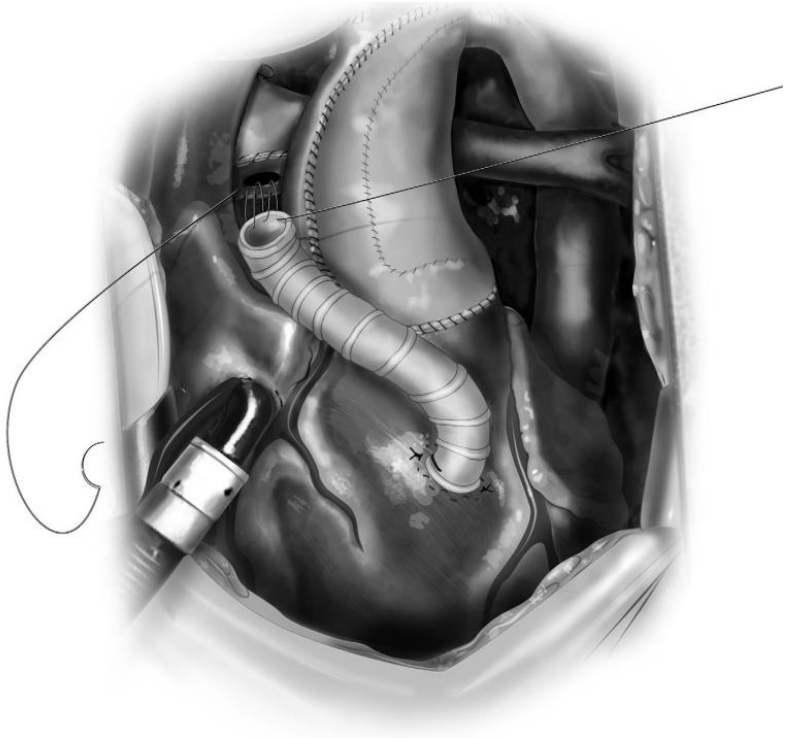
Operative Techniques in
Thoracic and
Cardiovascular Surgery

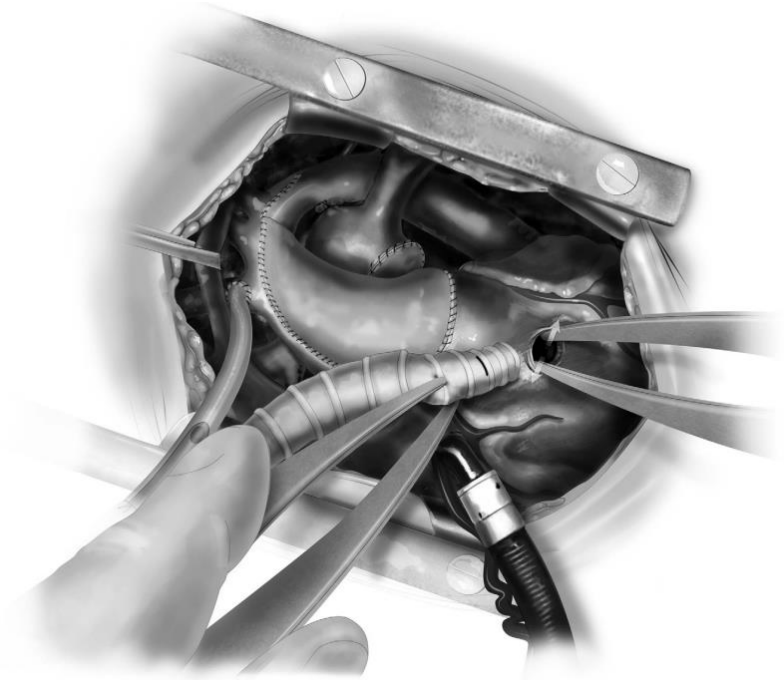
Construction of the Right Ventricle-to-Pulmonary Artery Conduit in the Norwood: The "Dunk" Technique

James S. Tweddell, MD,* Michael E. Mitchell, MD,* Ronald K. Woods, MD,*
Thomas L. Spray, MD,† and James A. Quintessenza, MD‡









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Periscopic technique in Norwood operation is associated with better preservation of early ventricular function

Puneet Bhatla, MD,[a,b] TK Susheel Kumar, MD,[c] Luv Makadia, MD,[d] Brandon Winston, MPH,[c] Catherine Bull, PNP-BC,[c] James C. Nielsen, MD,[a] David Williams, MD,[c] Sujata Chakravarti, MD,[a] Richard G. Ohye, MD,[c] and Ralph S. Mosca, MD[c]

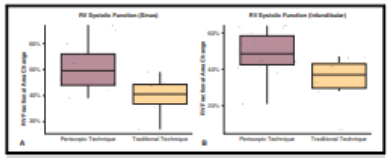
ABSTRACT

Objective: Although the right ventricle (RV) to pulmonary artery conduit in stage 1 Norwood operation results in improved interstage survival, the long-term effects of the ventriculotomy used in the traditional technique remain a concern. The periscopic technique (PT) of RV to pulmonary artery conduit placement has been described as an alternative technique to minimize RV injury. A retrospective study was performed to compare the effects of traditional technique and PT on ventricular function following Norwood operation.

Methods: A retrospective study of all patients who underwent Norwood operation from 2012 to 2019 was performed. Patients with baseline RV dysfunction and significant tricuspid valve regurgitation were excluded. Prestage 2 echocardiograms were reviewed by a blinded experienced imager for quantification of RV function (sinus and infundibular RV fractional area change) as well as for regional conduit site wall dysfunction (normal or abnormal, including hypokinesia, akinesia, or dyskinesia). Wilcoxon rank-sum tests were used to assess differences in RV infundibular and RV sinus ejection fraction and the Fisher exact test was used to assess differences in regional wall dysfunction.

Results: Twenty-two patients met inclusion criteria. Eight underwent traditional technique and 14 underwent PT. Median infundibular RV fractional area change was 49% and 37% ($P = .02$) and sinus RV fractional area change was 50% and 41% for PT and traditional technique ($P = .007$) respectively. Similarly qualitative regional RV wall function was better preserved in PT ($P = .002$).

Conclusions: The PT for RV to pulmonary artery conduit in Norwood operation results in better preservation of early RV global and regional systolic function. Whether or not this benefit translates to improved clinical outcome still needs to be studied. (JTCVS Techniques 2021;8:116-23)



Boxplot comparison of periscopic versus traditional technique for RV systolic function.

CENTRAL MESSAGE

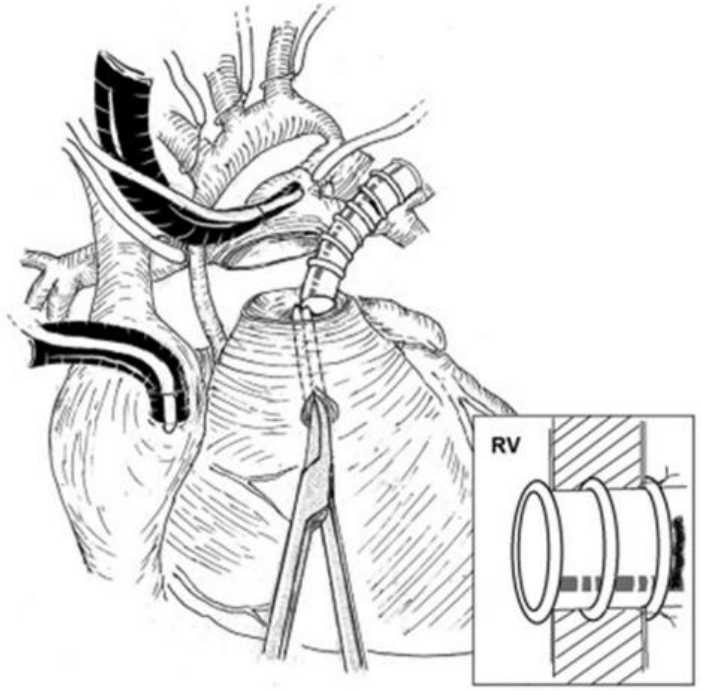
Use of periscopic technique for right ventricle to pulmonary conduit placement in stage 1 Norwood operation is associated with better preservation of right ventricular function.

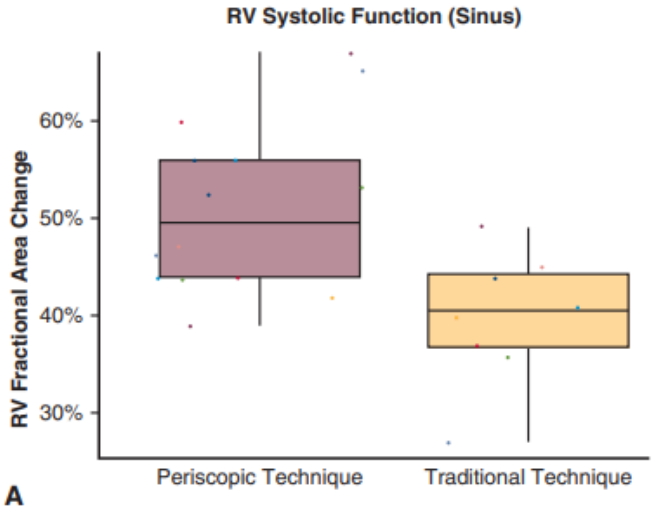
PERSPECTIVE

Although right ventricle to pulmonary artery conduit in Norwood operation results in improved interstage survival, the long-term adverse effects of ventriculotomy remain a concern. The periscopic technique in which a graft is placed inside from within the ventricle combines the advantages of right ventricle to pulmonary artery conduit along with preservation of ventricular function.

See Commentary on page 124.

From the [a]Division of Pediatric Cardiology and Departments of [b]Radiology, [c]Cardiac Surgery, and [d]Pediatrics, New York University Langone Medical Center, New York, NY; and [e]Department of Cardiac Surgery, Mott Children's Hospital, Ann Arbor, Mich.





Periscopic technique in Norwood operation is associated with better preservation of early ventricular function

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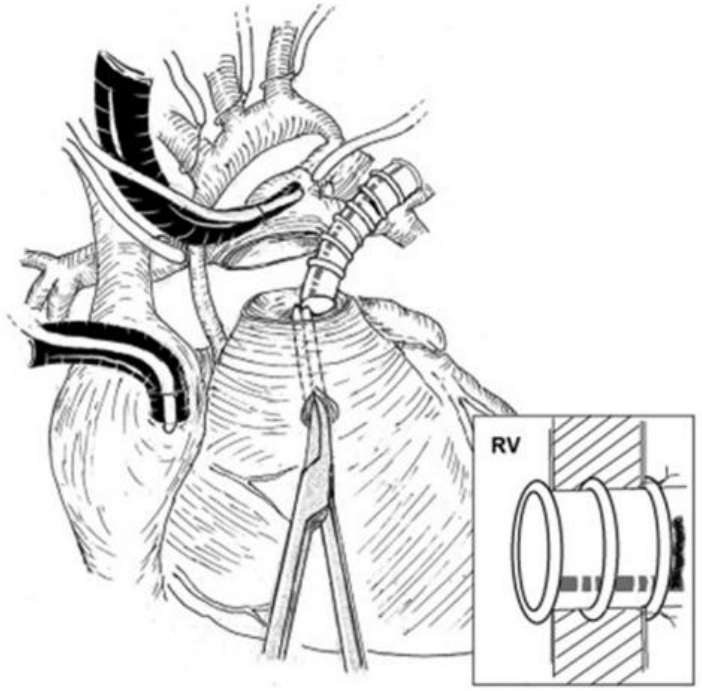

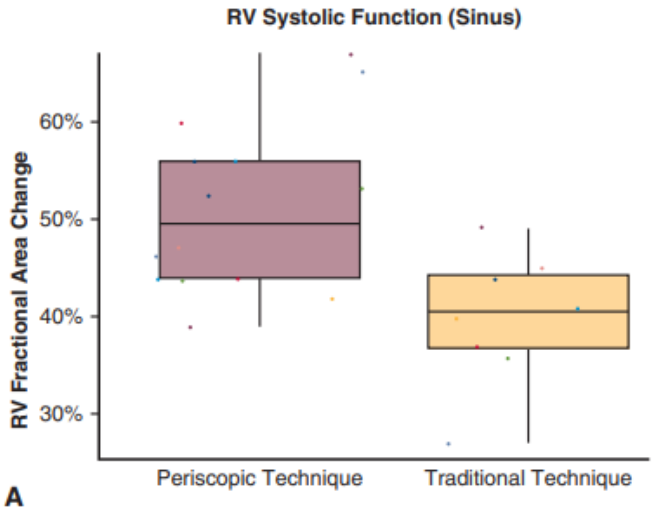

Pulmonary blood flow; mBT versus RV-PA Shunt

No consensus which shunt is the “Gold standard”

HLHS forms a complex and highly variable spectrum, which requires different surgical approaches according to the individual morphology and physiology of the patient.

Original Article

Selective Use of the Blalock-Taussig Shunt and Right Ventricle-to-Pulmonary Artery Conduit During the Norwood Procedure

World Journal for Pediatric and Congenital Heart Surgery
2016, Vol. 7(3) 329-333

DOI: 10.1177/2150135115625203
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Abstract

Background: The single ventricle reconstruction trial showed better one-year transplant-free survival for the right ventricle-to-pulmonary artery (RV-to-PA) conduit over the modified Blalock-Taussig shunt (mBTS) at Norwood operation. However, concerns remain about the long-term effects of a neonatal ventriculotomy. In our institution, we have used specific selection criteria for the use of mBTS in the Norwood operation. **Methods:** We reviewed 122 consecutive neonates undergoing the Norwood procedure from December 2006 to December 2013. We used the following criteria to select our source of pulmonary blood flow: (1) presence of a dominant morphologic left ventricle; (2) presence of antegrade blood in an ascending aorta that is greater than 3 mm; and (3) presence of significant large "crossing coronaries" on ventricle. All patients who met any of the above 3 criteria underwent an mBTS while the remaining patients underwent an RV-to-PA conduit. **Results:** Seventy-five (61.5%) patients had the RV-to-PA conduit and 47 (38.5%) patients had an mBTS. The overall surgical mortality was 9%. Mean follow-up interval was 23.5 months. Actuarial transplant-free survival was similar at 12, 24, 36, and 48 months in both the mBTS group and the RV-to-PA conduit group. In the RV-to-PA conduit group, actuarial transplant-free survival was 73% at 12 months, 71% at 24 months, 71% at 36 months, and 67% at 48 months, while in the mBTS group, actuarial transplant-free survival was 82% at 12 months, 75% at 24 months, 75% at 36 months, and 75% at 48 months. **Conclusion:** Our selection criteria for mBTS have allowed us to obtain equivalent transplant-free survival at 12, 24, 36, and 48 months when compared to the RV-to-PA conduit group.

CONG

Thirty years and 1663 consecutive Norwood procedures: Has survival plateaued?

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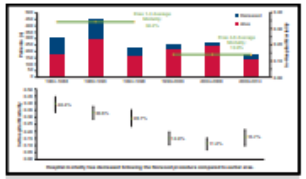
ABSTRACT

Objective: Hypoplastic left heart syndrome is one of the most common and challenging lesions requiring surgical intervention in the neonatal period. The Norwood procedure for hypoplastic left heart syndrome was first reported in 1983. The objective of this study was to describe early outcomes after the Norwood procedure at a single institution over 30 years.

Methods: This retrospective cohort study included all patients with hypoplastic left heart syndrome (and variants) who underwent the Norwood procedure between January 1984 and May 2014 at a single institution. The study period was divided into 6 eras: era 1, 1984 to 1988; era 2, 1989 to 1993; era 3, 1994 to 1998; era 4, 1999 to 2003; era 5, 2004 to 2008; and era 6, 2009 to 2014. The primary outcome was in-hospital mortality after the Norwood procedure. Binomial point estimates complete with 95% confidence intervals ($CL_{0.95}$) were computed for the entire cohort and by era.

Results: During the study period, 1663 infants underwent the Norwood procedure. Overall in-hospital mortality was 25.9% ($CL_{0.95}$, 23.8-28.0). Mortality by chronologic era was 40.4% ($CL_{0.95}$, 34.9-45.9), 33.6% ($CL_{0.95}$, 29.2-37.9), 28.7% ($CL_{0.95}$, 22.8-34.6), 14.9% ($CL_{0.95}$, 10.4-19.3), 11.2% ($CL_{0.95}$, 7.4-15.0), and 15.7% ($CL_{0.95}$, 10.3-21.1). Survival was improved in eras 4 to 6 compared with eras 1 to 3 (P all < .03). Anomalous pulmonary drainage, moderate to severe atrioventricular valve regurgitation, lower birth weight, earlier era, younger gestational age, genetic anomaly, preterm birth, race other than white or African-American, and lower weight at the Norwood procedure were associated with increased mortality. Mortality was greatest in patients with 3 or more risk factors. In the best-fitting multiple covariate model, anomalous pulmonary venous drainage, gestational age in weeks, genetic anomaly, and race other than white and African American were statistically significant contributors, after adjusting for era.

Conclusions: Survival after the Norwood procedure has plateaued despite improvements in diagnosis, perioperative care, and surgical techniques. Nonmodifiable patient characteristics are important determinants of the risk of mortality. (J Thorac Cardiovasc Surg 2019;158:220-9)



Hospital mortality has decreased after the Norwood procedure compared with earlier eras.

Central Message

Hospital mortality after the Norwood procedure has decreased in recent eras. We identified patient-specific factors that affect mortality. Patients with 3 or more risk factors have worse survival.

Perspective

The operative technique of the Norwood procedure today is nearly identical to the original description. Hospital mortality for patients undergoing this procedure has decreased in recent eras, but it remains unchanged for the last 15 years. Patient-specific risk factors continue to play a key role in survival. Lower-risk patients have lower mortality rates compared with their high-risk counterparts.

See Commentaries on pages 230 and 232.

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Thirty years and 1663 consecutive Norwood procedures: Has survival plateaued?

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ABSTRACT

Objective: Hypoplastic left heart syndrome is one of the most common and challenging lesions requiring surgical intervention in the neonatal period. The Norwood procedure for hypoplastic left heart syndrome was first reported in 1983. The objective of this study was to describe early outcomes after the Norwood procedure at a single institution over 30 years.

Methods: This retrospective cohort study included all patients with hypoplastic left heart syndrome (and variants) who underwent the Norwood procedure between January 1984 and May 2014 at a single institution. The study period was divided into 6 eras: era 1, 1984 to 1988; era 2, 1989 to 1993; era 3, 1994 to 1998; era 4, 1999 to 2003; era 5, 2004 to 2008; and era 6, 2009 to 2014. The primary outcome was in-hospital mortality after the Norwood procedure. Binomial point estimates complete with 95% confidence intervals ($CL_{0.95}$) were computed for the entire cohort and by era.

Results: During the study period, 1663 infants underwent the Norwood procedure. Overall in-hospital mortality was 25.9% ($CL_{0.95}$, 23.8-28.0). Mortality by chronologic era was 40.4% ($CL_{0.95}$, 34.9-45.9), 33.6% ($CL_{0.95}$, 29.2-37.9), 28.7% ($CL_{0.95}$, 22.8-34.6), 14.9% ($CL_{0.95}$, 10.4-19.3), 11.2% ($CL_{0.95}$, 7.4-15.0), and 15.7% ($CL_{0.95}$, 10.3-21.1). Survival was improved in eras 4 to 6 compared with eras 1 to 3 (P all < .03). Anomalous pulmonary drainage, moderate to severe atrioventricular valve regurgitation, lower birth weight, earlier era, younger gestational age, genetic anomaly, preterm birth, race other than white or African-American, and lower weight at the Norwood procedure were associated with increased mortality. Mortality was greatest in patients with 3 or more risk factors. In the best-fitting multiple covariate model, anomalous pulmonary venous drainage, gestational age in weeks, genetic anomaly, and race other than white and African American were statistically significant contributors, after adjusting for era.

Conclusions: Survival after the Norwood procedure has plateaued despite improvements in diagnosis, perioperative care, and surgical techniques. Nonmodifiable patient characteristics are important determinants of the risk of mortality. (J Thorac Cardiovasc Surg 2019;158:220-9)

Central Message

Hospital mortality after the Norwood procedure has decreased in recent eras. We identified patient-specific factors that affect mortality. Patients with 3 or more risk factors have worse survival.

Perspective

The operative technique of the Norwood procedure today is nearly identical to the original description. Hospital mortality for patients undergoing this procedure has decreased in recent eras, but it remains unchanged for the last 15 years. Patient-specific risk factors continue to play a key role in survival. Lower-risk patients have lower mortality rates compared with their high-risk counterparts.

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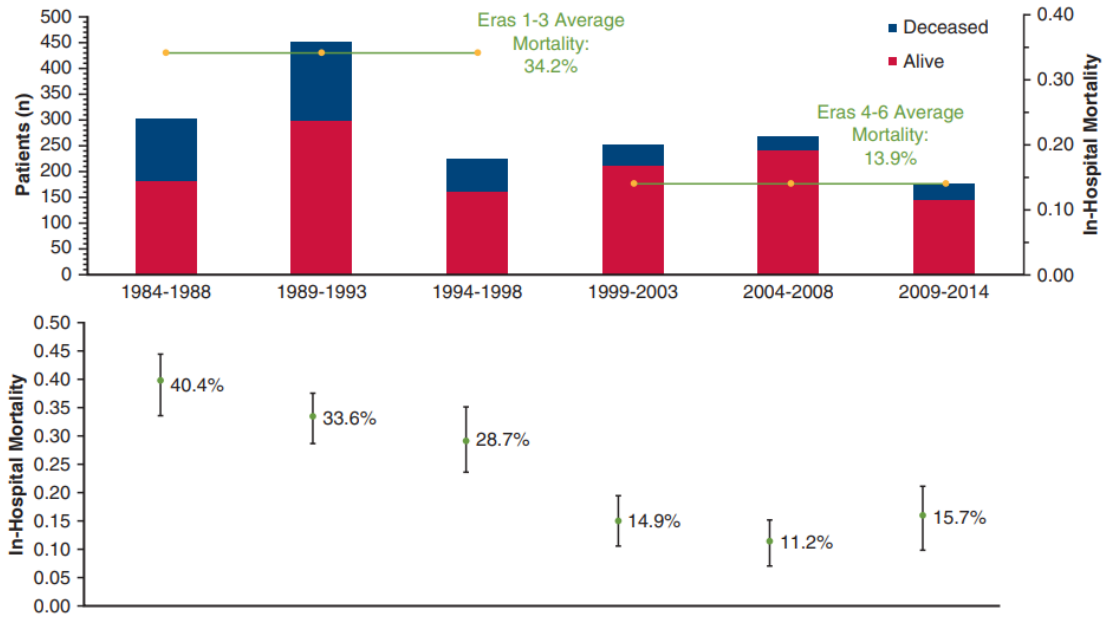

FIGURE 1. Hospital mortality has decreased after the Norwood procedure compared with earlier eras.

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UMC Utrecht Experience

"Patient tailored approach" for HLHS lesions

- Very limited use of bilateral pulmonary banding and Prostin;
  - patient weight <2,5 Kg,
  - hemodynamic instability in the first week
- Norwood with mBT shunt, Selection criteria for Sano:
  - Ascending aorta < 2mm,
  - Moderate/severe TVR,
  - Patient weight <2,5 Kg

Norwood operation in UMC Utrecht

- Since 2004-2022; 85 Norwood operations (2 bilateral pulmonary banding and prostin preceding the Norwood)
- 30 day mortality; 14%
- In hospital mortality 18%

Conclusions

Best approach for the Norwood operation

- Y- aortic (or interdigitating) anastomosis of the aortic arch
- Homograft or PTFE patch for the aortic arch reconstruction
- If RV-PA shunt; Dunck method

- “Patient tailored approach” may positively influence mortality in HLHS patients
- Nowadays patient specific factors mainly influence outcome of the Norwood operation in HLHS

Questions

Commentary: The Norwood operation: Can we modify the nonmodifiable?

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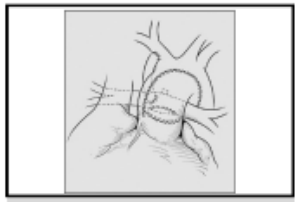
This is an important article reviewing 30 years of experience with the Norwood procedure at a center with largest experience reported to date.[1] The large number of patients operated at a single institution with (essentially) a uniform approach makes this a valuable contribution to our literature. The study's principle finding is that although hospital survival improved in their contemporary patient cohort it has essentially plateaued, a finding that the authors attribute to the high incidence of nonmodifiable patient-specific risk factors. Other interesting findings include no difference in mortality associated with ascending aortic size and HLHS anatomic subtype (mitral and/or aortic atresia vs hypoplasia). This reviewer, coming from a center with another large experience with this procedure (>1300 cases), found the results reported by the investigators very consistent with our experience.[2]

Despite the value of this report, the article lacks information that would have proved to be very useful to the reader. The authors imply throughout the report that the Norwood operation, as used in this review, is used for HLHS and yet they include what are referred to as "HLHS variants" such as double inlet left ventricle and tricuspid atresia with transposition of the great arteries. Although these conditions are often treated with a Norwood operation, they represent a different group of patients with a systemic left (vs right) ventricle. Despite this concern, no mention is made if the results for these patients differ from those with classic HLHS.[3] Additional valuable knowledge that might have been gained from this review include the effect of shunt type and size, interstage mortality, and individual surgeon outcomes.[4]

These comments should in no way detract from what is a valuable addition to the literature, one that suggests that further improvements in outcomes for the Norwood procedure have plateaued because of nonmodifiable conditions. Indeed, our own experience suggests that improved prenatal diagnosis and recommended delivery at a major cardiac center might have actually resulted in worse outcomes. This seemingly paradoxical finding is likely because patients who would not have survived hospital transfer or would not have been offered staged palliation because of associated risk factors such as intact atrial septum, poor right ventricular function and tricuspid regurgitation, significant prematurity and/or low birth weight, and major associated congenital anomalies, now undergo a Norwood operation. Additionally, newer treatment options such as in utero intervention, hybrid procedures, and shunt type have as of yet failed to convincingly show improved overall outcomes, and it remains unclear which patient subgroups might benefit the most from these therapeutic approaches.

Should we continue to offer patients with these associated nonmodifiable conditions a Norwood operation? This is a difficult question, certainly beyond the scope of this editorial, but one that we still struggle with when confronted with these newborns. Admittedly, other factors weigh on the decision to offer treatment, such as parental desires and individual physician preferences, but our own group has become more conservative in offering surgery when the outcomes are clearly poor. As the results for the Norwood procedure improve, the question still remains: can we modify the nonmodifiable?



The classic Norwood procedure shown with a modified Blalock-Taussig shunt.

Central Message

This report suggests that as outcomes for the Norwood operation improve they have plateaued because of patient-specific nonmodifiable conditions, raising the question: can we modify the nonmodifiable?

See Article page 220.

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Workflow Hypoplastic Left Heart WKZ

Zorgpad cardiochirurgische patiënt voor Norwood procedure

Prenataal	Geboorte	Preoperatief IC	Operatie	IC post-operatief	Leeuw
Echo verdenking cor vitium --> verwijzing WKZ HART-2 spreekuur Aanvullende diagnostiek en bespreken uitslagen CHD lifespan onderzoek **Aanspreekpunt: Gynecoloog / verloskundige**	Streef geboorte op a terme leeftijd op woensdag of donderdag Bij voorkeur in WKZ Indien buiten gebied WKZ--> streven naar inleiding **Aanspreekpunt: gyneacoloog / verloskundige**	Opvang op VK of OK door Neonatologie Start Prostin direct post partum via perifeer infuus Zo snel mogelijk opname op PICU **Aanspreekpunt na geboorte: kindercardioloog en kinder-intensivist**	Streef operatie bij voorkeur rond dag 5 (tot 7) **Aanspreekpunt: kinderthoraxchirurg**	Opvang patient volgens standard oparating procedure: Opvang cardio chirurgische patient op IC **Aanspreekpunt: kinderintensivist**	Laagdrempelig medebeoordeling IC bij achteruitgang Bespreek wanneer mogelijk retour UMCG indien patient daar vandaan komt Wekelijks e cho cor / lab / RR (4 extremiteiten) **Aanspreekpunt: kindercardioloog**

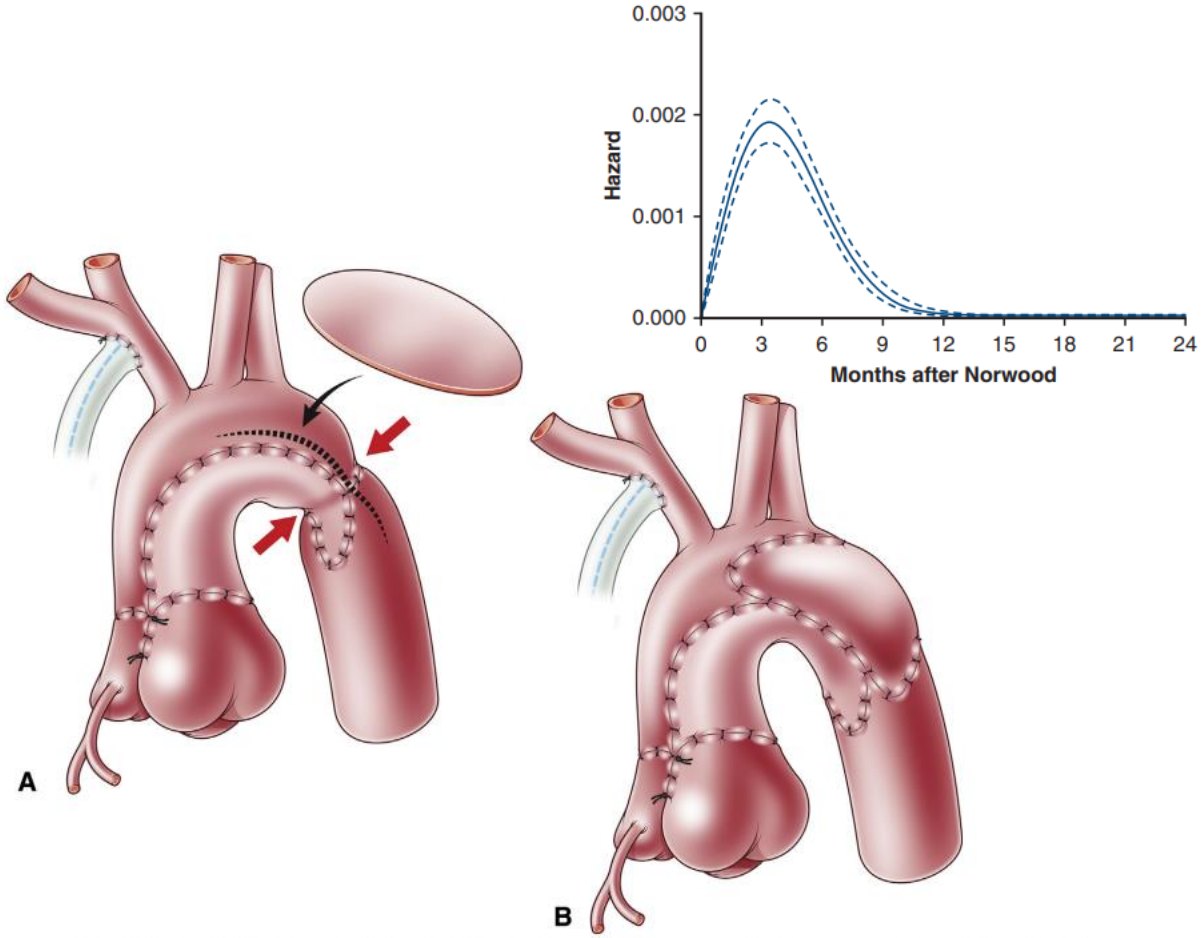

FIGURE 1. Effective surgical arch augmentation can be achieved simply by the addition of a patch. A, An incision is made across the obstruction in the axis of the aorta. B, After patch augmentation.

Devlin and colleagues1 clearly demonstrate the association between residual arch lesions and worse tricuspid valve and ventricular function, and this relationship is undoubtedly causal. The current threshold at which reintervention is warranted remains poorly defined, and it will remain poorly defined, because in a single-ventricular circulation, arch obstruction is likely to impair cardiac output before it generates a high gradient.

We should intervene on all arches that seem to have a suboptimal shape or size: it is time for cosmetic surgery on aortic arches after Norwood!