1-week old neonate with HLHS

GA 38 wk, Bwt 3.0 kg

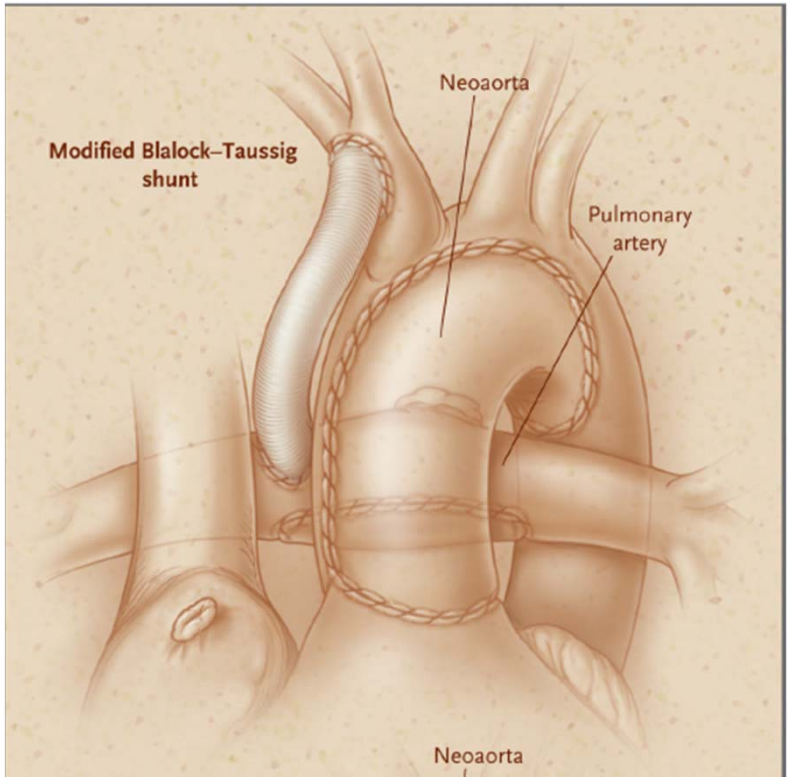
No other risk factors

Norwood operation with RV-PA shunt

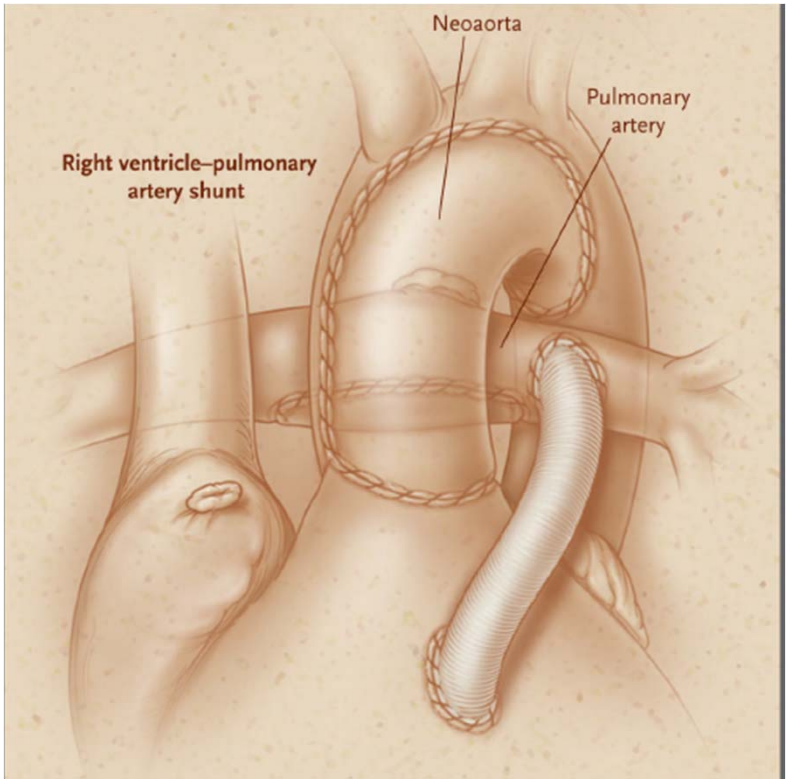
세종병원 흉부외과
이창하

Norwood operation

Classic - BT shunt



Modified - RV-PA shunt



William I. Norwood

- Hypoplastic left heart syndrome: Experience with palliative surgery (AJC, 1980)
- Experience with operations for hypoplastic left heart syndrome (JTCS, 1981)
- Physiologic repair of aortic atresia-hypoplastic left heart syndrome (NEJM, 1983)

Hypoplastic Left Heart Syndrome: Experience With Palliative Surgery

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

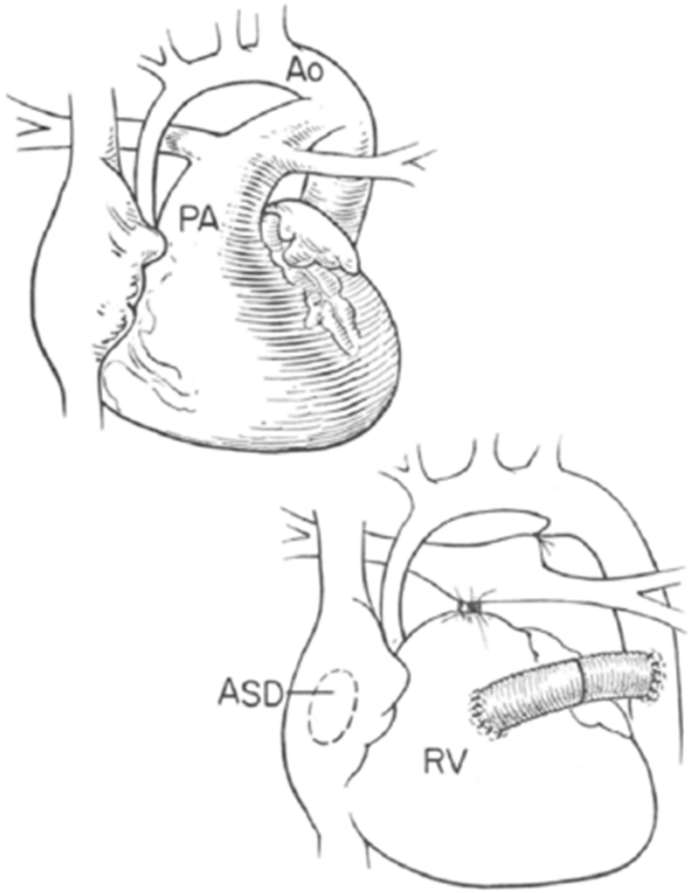
Boston, Massachusetts

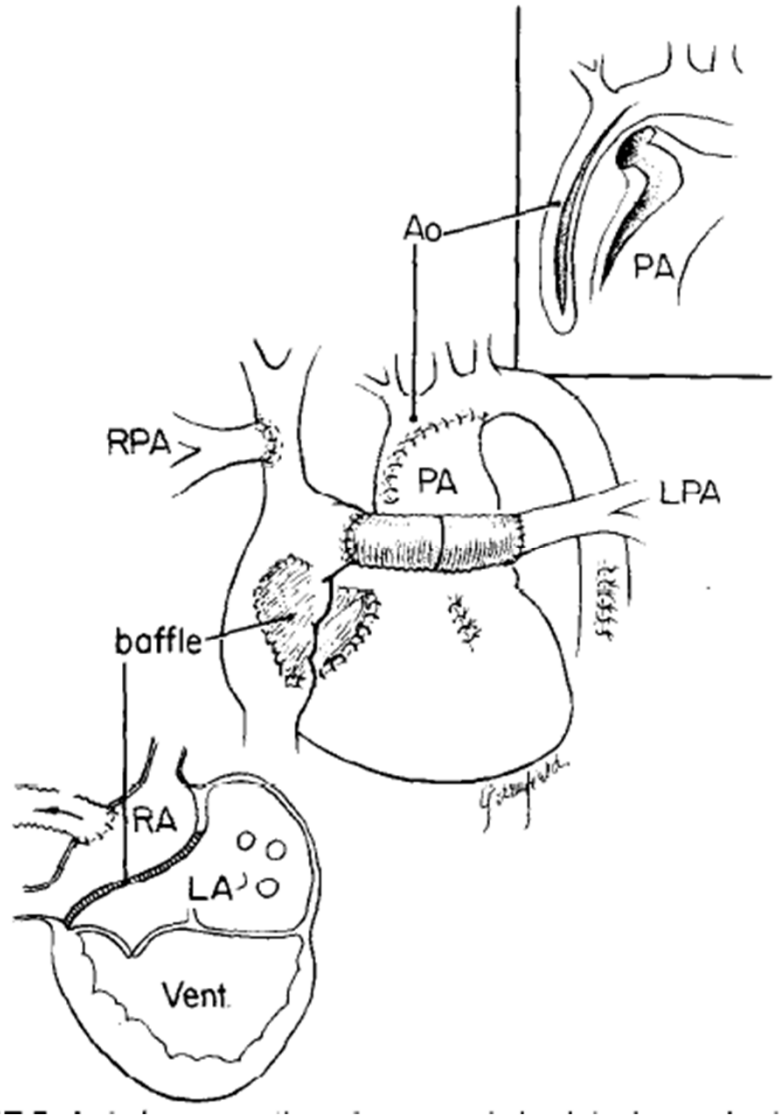
Aortic atresia is a form of hypoplastic left heart syndrome always complicated by associated severe hypoplasia of the ascending aorta and various degrees of mitral valve and left ventricular hypoplasia. At present it is a universally fatal lesion in early infancy. This is a report of a new palliative 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

Hypoplastic Left Heart Syndrome: Experience With Palliative Surgery

RV-Ao conduit





J THORAC CARDIOVASC SURG 82:511-519, 1981

Experience with operations for hypoplastic left heart syndrome

Aortic atresia is a form of congenital cardiac disease complicated by associated severe hypoplasia of the

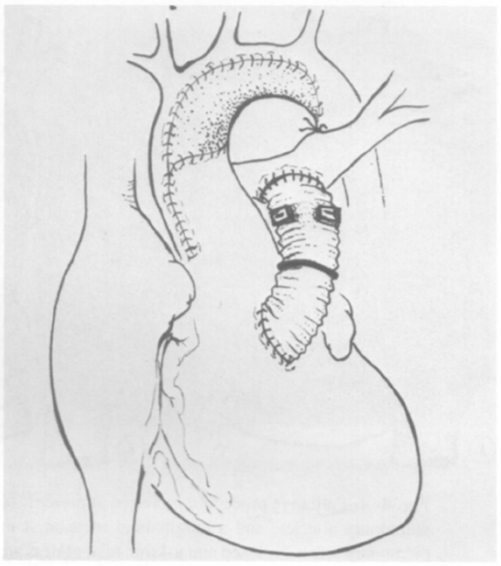
1979 – 1981

16 patients with HLHS

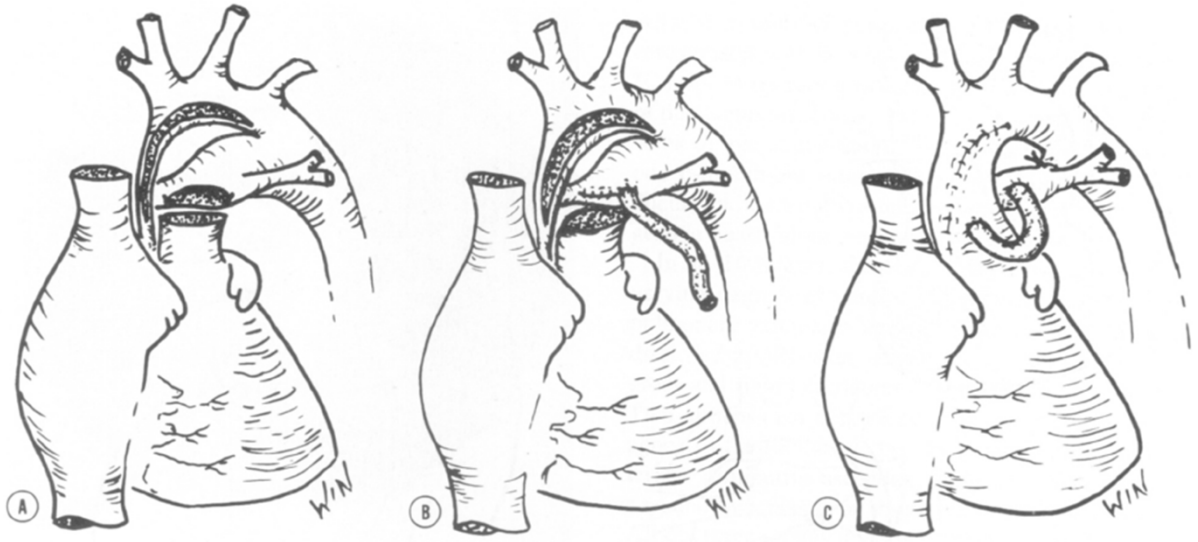
- ◆RV-Ao conduit (n = 3)
- ◆RV-PA conduit (n = 3)
- ◆Central shunt (n = 9)
- ◆LV-Ao conduit (n = 1)

J THORAC CARDIOVASC SURG 82:511-519, 1981

Experience with operations for hypoplastic left heart syndrome



RV-PA conduit



Central shunt

J THORAC CARDIOVASC SURG 82:511-519, 1981

Experience with operations for hypoplastic left heart syndrome

Table II. *Results*

Procedure	*Hospital deaths*	*Late deaths*
First stage		
RV-Ao conduit (a)	0/3	2/3
MPA-Ao, RV-PA conduit (b)	3/3	—
MPA-Ao, central shunt (c)	5/9	1/4
LV-Ao conduit, PAB	0/1	0/1
Totals	8/16	3/8
Physiological repair		
Modified Fontan	1/1	—
LV-Ao conduit, VSD closure	0/1	1/1
Totals	1/2	1/1

Legend: RV, Right ventricle. Ao, Aorta. MPA, Main pulmonary artery. PA, Distal pulmonary artery. PAB, Pulmonary artery band. (a), Fig. 2. (b), Fig. 3. (c), Fig. 4.

PHYSIOLOGIC REPAIR OF AORTIC ATRESIA–HYPOPLASTIC LEFT HEART SYNDROME

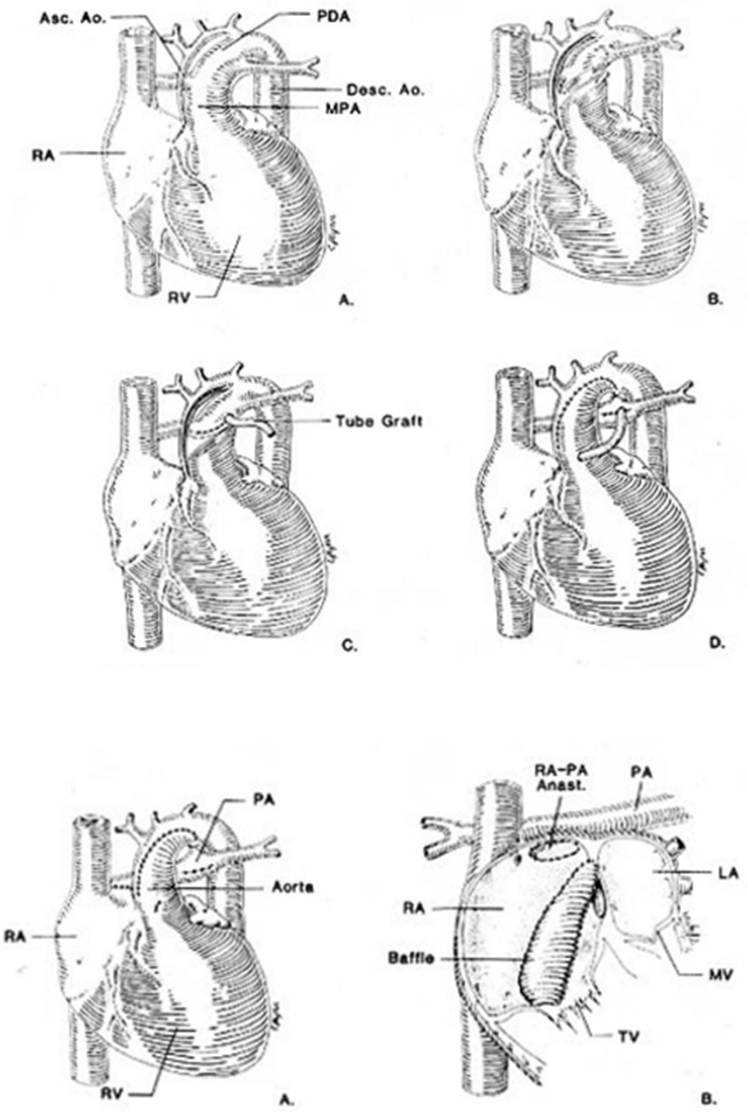
WILLIAM I. NORWOOD, M.D., PETER LANG, M.D., AND DOLLY D. HANSEN, M.D.

AORTIC-VALVE atresia associated with mitral atresia or stenosis, a diminutive or absent left ventricle, and severe hypoplasia of the ascending aorta and aortic arch make up a constellation of defects that was initially termed "hypoplasia of the aortic tract complexes" by Lev[1] and later called "hypoplastic left heart syndrome" by Noonan and Nadas.[2] The anatomic complexity of this cardiac malformation has been considered prohibitive of long survival, and generally, only supportive therapy has been recommended. Indeed, aortic atresia–hypoplastic left heart syndrome is uniformly fatal without surgical intervention, and prolonged survival after reparative surgery has not been reported. It is, however, an anomaly not easily ignored. Hypoplastic left heart syndrome is the fourth most common cardiac defect (7.5 per cent) listed by the New England Regional Infant Cardiac Program.[3] According to data collected between 1969 and 1979, 223 infants were identified as having aortic atresia in New England alone. None survived for a year; their deaths accounted for nearly 25 per cent of cardiac deaths during the first week of life, as recorded by the Program. The death of such a neonate is particularly poignant since most are well-developed babies with normal birth weights, and unlike patients with other forms of congenital heart disease, they have a very low incidence of associated extracardiac anomalies.[3]

These compelling facts and recent advances in cardiac surgery of the infant led us, in 1979, to initiate a program of staged surgical management of neonatal hypoplastic left heart syndrome. Our preliminary results with the palliative stage have been reported.[4,5] This paper relates our experience with the first child in this program who has been clinically well during six months of follow-up after physiologically corrective surgery for the syndrome.

CASE REPORT

The patient weighed 3.7 kg when he was born, after an uncomplicated full-term pregnancy. At 24 hours of age, a heart murmur and tachypnea were noted. No left ventricle was visualized on an echocardiogram. An infusion of prostaglandin E_1 (0.1 μg per kilogram of body weight per minute) was begun to maintain patency of the



N Engl J Med 1983;308:23-26

Norwood operation

Modified BT shunt

Balancing the Circulation: Theoretic Optimization of Pulmonary/Systemic Flow Ratio in Hypoplastic Left Heart Syndrome

OFER BARNEA, PHD, ERLE H. AUSTIN, MD, FACC,* BARBARA RICHMAN, MD,†
WILLIAM P. SANTAMORE, PHD*

Tel Aviv, Israel and Louisville, Kentucky

Objectives. **This study examined the effects of the pulmonary (Q_P)/systemic (Q_S) blood flow ratio (Q_P/Q_S) on systemic oxygen availability in neonates with hypoplastic left heart syndrome.**

Background. **The management of neonates with hypoplastic left heart syndrome is complex and controversial. Both before and after surgical palliation and before heart transplantation, a univentricle with parallel pulmonary and systemic circulations exists. It is generally assumed that balancing pulmonary and systemic blood flow is best to stabilize the circulation.**

Methods. **We developed a mathematical model that was based on the simple flow of oxygen uptake in the lungs and whole-body oxygen consumption to study the effect of varying the Q_P/Q_S ratio. An equation was derived that related the key variables of cardiac output, pulmonary venous oxygen saturation and the Q_P/Q_S ratio to systemic oxygen availability.**

Results. **The key findings are 1) as the Q_P/Q_S ratio increases, systemic oxygen availability increases initially, reaches a maximum and then decreases; 2) for maximal systemic oxygen availability, the optimal Q_P/Q_S ratio is ≤1; 3) the optimal Q_P/Q_S ratio decreases as cardiac output or percent pulmonary venous oxygen saturation, or both, increase; 4) the critical range of Q_P/Q_S, where oxygen supply exceeds basal oxygen consumption, decreases as cardiac output and percent pulmonary venous oxygen saturation decrease; 5) the relation between oxygen availability and Q_P/Q_S is very steep when Q_P/Q_S approaches this critical value; and 6) the percent oxygen saturation of systemic venous blood is very low outside the critical range of Q_P/Q_S and high within the critical range.**

Conclusions. **This analysis provides a theoretic basis for balancing both the pulmonary and systemic circulation and suggests that evaluating both systemic arterial and venous oxygen saturation may be a useful way to determine the relative pulmonary and systemic flows. When high systemic arterial and low systemic venous oxygen saturation are present, pulmonary blood flow should be decreased; conversely, when both low systemic arterial and venous oxygen saturation are present, more flow should be directed to the pulmonary circulation.**

(J Am Coll Cardiol 1994;24:1376–81)

RESTING CORONARY FLOW AND CORONARY FLOW RESERVE IN HUMAN INFANTS AFTER REPAIR OR PALLIATION OF CONGENITAL HEART DEFECTS AS MEASURED BY POSITRON EMISSION TOMOGRAPHY

Jon P. Donnelly, MD
David M. Raffel, PhD
Barry L. Shulkin, MD
James R. Corbett, MD
Edward L. Bove, MD
Ralph S. Mosca, MD
Thomas J. Kulik, MD

Objective: Coronary physiology in infants with congenital heart disease remains unclear. Our objective was to better understand coronary physiology in infants with congenital heart disease. *Methods:* We used positron emission tomography with nitrogen 13–labeled ammonia to measure myocardial perfusion at rest and with adenosine (142 μg/kg/min × 6 minutes) in five infants after anatomic repair of a congenital heart lesion (group I), and in five infants after Norwood palliation for hypoplastic left heart syndrome (group II). The groups were matched for age, weight, and time from the operation. *Results:* Resting coronary flow in the left ventricle in group I was 1.8 ± 0.2 ml/min/gm; resting flow in the right ventricle in group II was 1.0 ± 0.3 ml/min/gm ($p = 0.003$). Coronary flow with adenosine was 2.6 ± 0.5 ml/min/gm in group I and 1.5 ± 0.7 ml/min/gm in group II ($p = 0.02$). Absolute coronary flow reserve was the same in both groups (1.5 ± 0.2 in group I vs 1.6 ± 0.3 in group II, $p = 0.45$). Oxygen delivery was reduced in group II compared with group I at rest (16.1 ± 4.2 ml/min/100 gm vs 28.9 ± 4.42 ml/min/100 gm, $p = 0.02$) and with adenosine (25.5 ± 8.1 ml/min/100 gm vs 42.3 ± 5.8 ml/min/100 gm, $p = 0.02$). *Conclusions:* Infants with repaired heart disease have higher resting flow and less coronary flow reserve than previously reported for adults. After Norwood palliation, infants have less perfusion and oxygen delivery to the systemic ventricle than do infants with a repaired lesion. This may in part explain why the outcome for patients with Norwood palliation is less favorable than for others. (J Thorac Cardiovasc Surg 1998;115:103-10)

Modifications of Norwood op

The Journal of Thoracic and
Cardiovascular Surgery
December 1999

THE MODIFIED NORWOOD PALLIATION ON A BEATING HEART

Hidefumi Kishimoto, MD, Youichi Kawahira, MD, Hiroaki Kawata, MD, Takuya Miura, MD, Shigemitu Iwai, MD, and Toru Mori, MD, *Osaka, Japan*

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7 patients with HLHS (2.4 to 3.3 kg (2.8 kg))

3 deaths

◆Diastolic pressure of the systemic circulation after the Norwood palliation with an extracardiac conduit markedly increased compared with use of a Blalock-Taussig shunt.

◆The increase seemed to be advantageous in systemic perfusion, including the coronary circulation.

)

Experience with the Norwood procedure without circulatory arrest

Yutaka Imoto, MD[a]
Hideaki Kado, MD[a]
Yuichi Shiokawa, MD[a]
Kazu Minami, MD[a]
Hisataka Yasui, MD[b]

Objective: We evaluated a new cardiopulmonary bypass technique that allowed complete avoidance of circulatory arrest and deep hypothermia in the Norwood procedure for hypoplastic left heart syndrome.

Methods: A total of 10 patients were included in this study. The arterial line of the cardiopulmonary bypass circuit was divided in two in a Y shape; one branch was used for cerebral perfusion through the innominate artery and the other for lower body perfusion through the cannula inserted into the descending thoracic aorta. Moderate hypothermia (29°C-31°C rectal temperature) and high pump flow (150-180 mL · kg^{-1} · min^{-1}) were used. A valveless conduit between the right ventricle and the pulmonary artery was used in 6 patients as an alternative pulmonary blood source to a conventional Blalock-Taussig shunt (n = 4).

Results: Circulatory arrest was completely avoided throughout the operation in all cases, and no complications from the new cardiopulmonary bypass technique were seen. Early deaths occurred in 3 cases. Neurologic deficits were not seen among the survivors, and the postoperative course was stable and uneventful, including satisfactory renal function.

Conclusions: The Norwood procedure for hypoplastic left heart syndrome was successfully accomplished with complete avoidance of circulatory arrest by means of cerebral perfusion through the innominate artery combined with cannulation of the descending aorta. A conduit between the right ventricle and the pulmonary artery seems an excellent alternative pulmonary blood source, although right ventricular function needs to be carefully monitored.

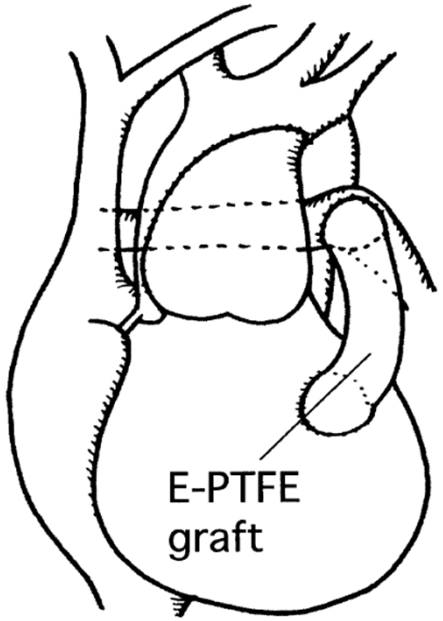
J Thorac Cardiovasc Surg 2001;122:879-82

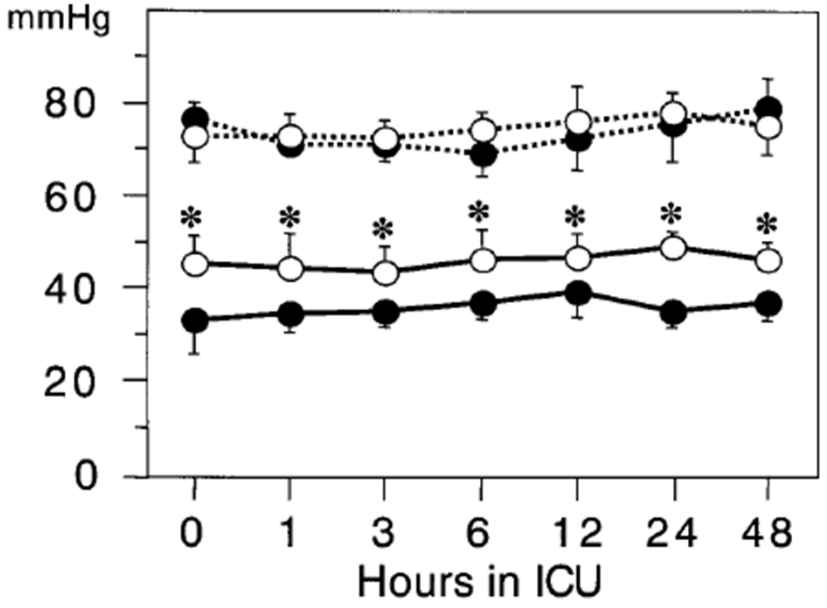
TABLE 1. Summary of the cases

Case	Age (d)	BW (kg)	AV/MV lesions	Diameter of AscAo (mm)	Pulmonary type	Blood source size (mm)	Result
1							
2							
3							
4							
5							
6							
7							
8							
9							
10							

AV, A… …duit.

◆A conduit between the right ventricle and the pulmonary artery seems an excellent alternative pulmonary blood source, although right ventricular function needs to be carefully monitored.





Right ventricle–pulmonary artery shunt in first-stage palliation of hypoplastic left heart syndrome

Shunji Sano, MD[a]

19 patients with HLHS

Modified Norwood operation with RV-PA shunt

Early mortality 2/19 (11%)

Survival rate 53% to 89% after modifications

◆Without delicate postoperative management to control pulmonary vascular resistance, the modified Norwood procedure using the right ventricle–pulmonary shunt provides a stable systemic circulation as well as adequate pulmonary blood flow.

◆This novel operation may be particularly beneficial to low-birth-weight infants with hypoplastic left heart syndrome.

ELSEVIER

European Journal of Cardio-thoracic Surgery 23 (2003) 991–995

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

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Right ventricle to pulmonary artery conduit has a favorable impact on postoperative physiology after Stage I Norwood: preliminary results☆

Christian Pizarro*, William I. Norwood

Nemours Cardiac Center, Alfred I,. duPont Hospital for Children, 1600 Rockland Road, Wilmington, DE 19899, USA

15 patients with HLHS

Stage 1 Norwood with RV-PA conduit

Early mortality 1/15 (6%)

◆RV to PA conduit eliminated diastolic runoff into the pulmonary vascular bed resulting in a higher diastolic blood pressure.

◆This physiology appears to be associated with a more stable postoperative course and improved hospital survival.

대흉외지 2010;43:364-374 □ 임상연구 □
DOI:10.5090/kjtcs.2010.43.4.364

변형된 노우드(Norwood) 수술의 결과: 단일병원의 지난 7년간의 경험

김형태* · 성시찬* · 김시호* · 배미주* · 이형두** · 박지애** · 장윤희***

Outcome of the Modified Norwood Procedure: 7 Years of Experience from a Single Institution

Hyungtae Kim, M.D.*, Si-Chan Sung, M.D.*, Si-Ho Kim, M.D.*, Mi-Ju Bae, M.D.*,
Hyoung Doo Lee, M.D.**, Ji Ae Park, M.D.**, Yun Hee Chang, M.D.***

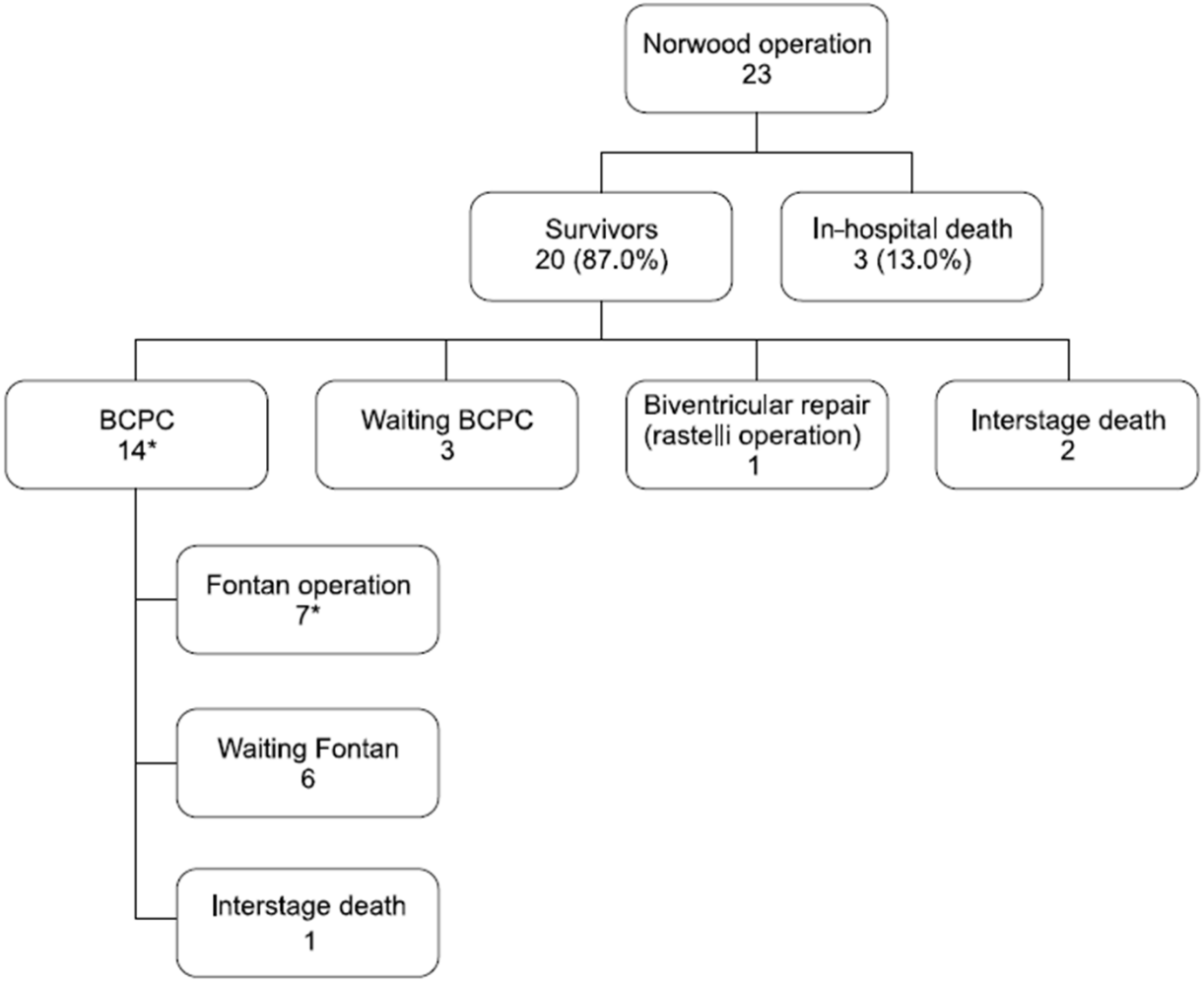
Background: We assessed the early and mid-term results of the modified Norwood procedure for first-stage palliation of hypoplastic left heart syndrome (HLHS) and its variants to identify the risk factors for hospital mortality.

2003 -2009
23 patients with HLHS & variants

tients (3/23, 13%), of whom 2 had TAPVC. Fourteen patients underwent subsequent bidirectional cavopulmonary connection (BCPC, stage 2) and seven underwent the Fontan operation (stage 3). Three patients died between stages, 2 before stage 2 and one before stage 3. The estimated 1-year and 5-year survival rates were 78% and 69%, respectively. On multivariate regression analysis, aberrant right subclavian artery (RSCA) and associated total anomalous pulmonary venous connection (TAPVC) were risk factors for hospital mortality after stage 1 Norwood procedure. ***Conclusion:*** HLHS and its variants can be palliated by the modified Norwood procedure with low operative mortality. Total anomalous pulmonary venous connection adversely affects the survival after a stage 1 Norwood procedure, and interstage mortality rates need to be improved.

(Korean J Thorac Cardiovasc Surg 2010;43:364-374)

Key words: 1. Congenital heart disease (CHD)
2. Hypoplastic left heart disease
3. Norwood procedure


Norwood operation
23
Survivors
20 (87.0%)
In-hospital death
3 (13.0%)
BCPC
14*
Waiting BCPC
3
Biventricular repair
(rastelli operation)
1
Interstage death
2
Fontan operation
7*
Waiting Fontan
6
Interstage death
1

RV-PA conduit (n = 15)
RMBT shunt (n = 8)

Concerns about impact of RV incision on later outcome

Best evidence topic - Congenital

In hypoplastic left heart patients is Sano shunt compared with modified Blalock–Taussig shunt associated with deleterious effects on ventricular performance?

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Received 29 October 2009; received in revised form 9 December 2009; accepted 11 December 2009

Summary

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◆Current available evidence, although weak, does not show any adverse effects of ventriculotomy on ventricular performance in patients with Sano shunt in the short- and medium-term.

◆However, all the existing studies are limited by small numbers, non-randomised design and retrospective nature with failure of correlation of echocardiographic indices to clinical outcomes.

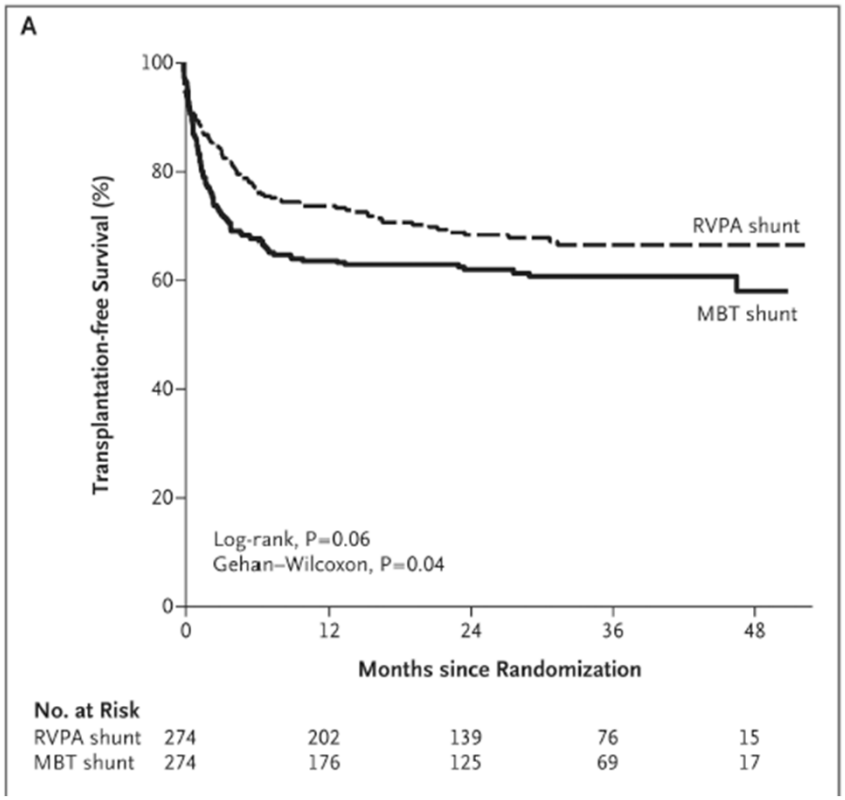
N Engl J Med. 2010 May 27; 362(21): 1980–1992. doi:10.1056/NEJMoa0912461.

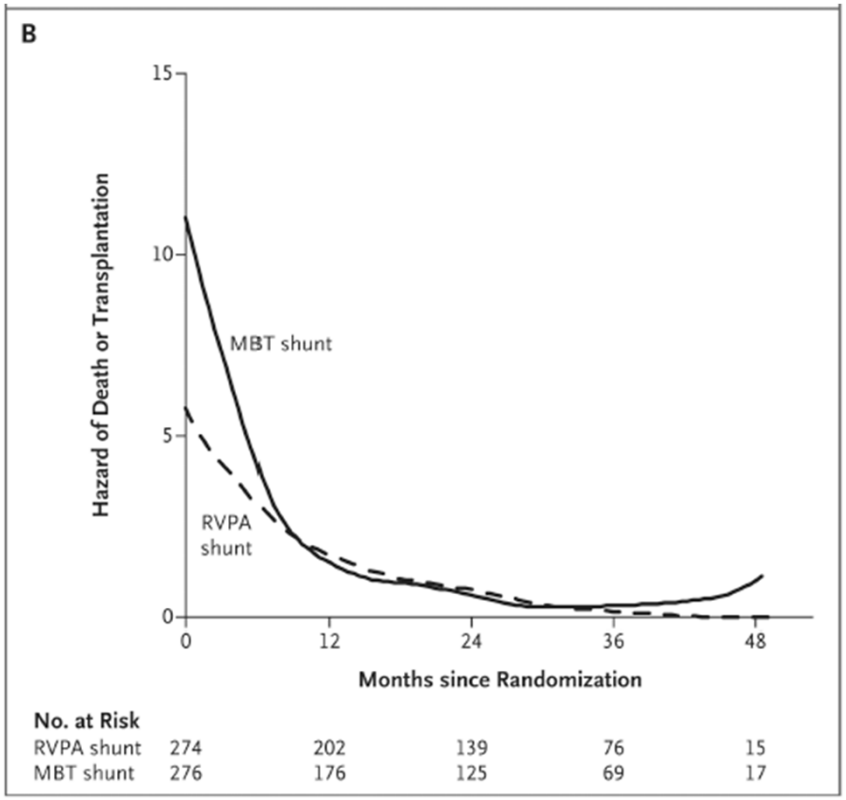
Comparison of Shunt Types in the Norwood Procedure for Single-Ventricle Lesions

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**
From the University of Michigan Medical School, Ann Arbor (R.G.O., C.S.G.); New England Research Institutes, Watertown, MA (L.A.S., M.L., L.V.); University of Texas Southwestern Medical Center, Dallas (L.M.); Children's Hospital Boston, Boston (J.W.N., P.C.L., C.D.-M., S.C.); National Heart, Lung, and Blood Institute, Bethesda, MD (G.D.P.); Children's Hospital of Philadelphia, Philadelphia (S.T., C.R., J.W.G.); Children's Hospital of Wisconsin and Medical College of Wisconsin, Milwaukee (P.C.F., N.S.G.); North Carolina Consortium: Duke University, Durham; East Carolina University, Greenville; Wake Forest University, Winston-Salem (J.F.R., J.J.); Children's Hospital Los Angeles, Los Angeles (A.B.L., N.P.); Hospital for Sick Children, Toronto (S.M., B.W.M.); Children's Hospital of New York, New York (I.A.W.); Medical University of South Carolina, Charleston (A.M.A.); Primary Children's Medical Center and the University of Utah, Salt Lake City (L.L.M.); Nemours Cardiac Center, Wilmington, DE (C.P.); Emory University, Atlanta (K.R.K.); the Congenital Heart Institute of Florida, St. Petersburg (J.P.J.); and Cincinnati Children's Medical Center, Cincinnati (C.D.K.)

N Engl J Med. 2010 May 27; 362(21): 1980–1992. doi:10.1056/NEJMoa0912461.

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





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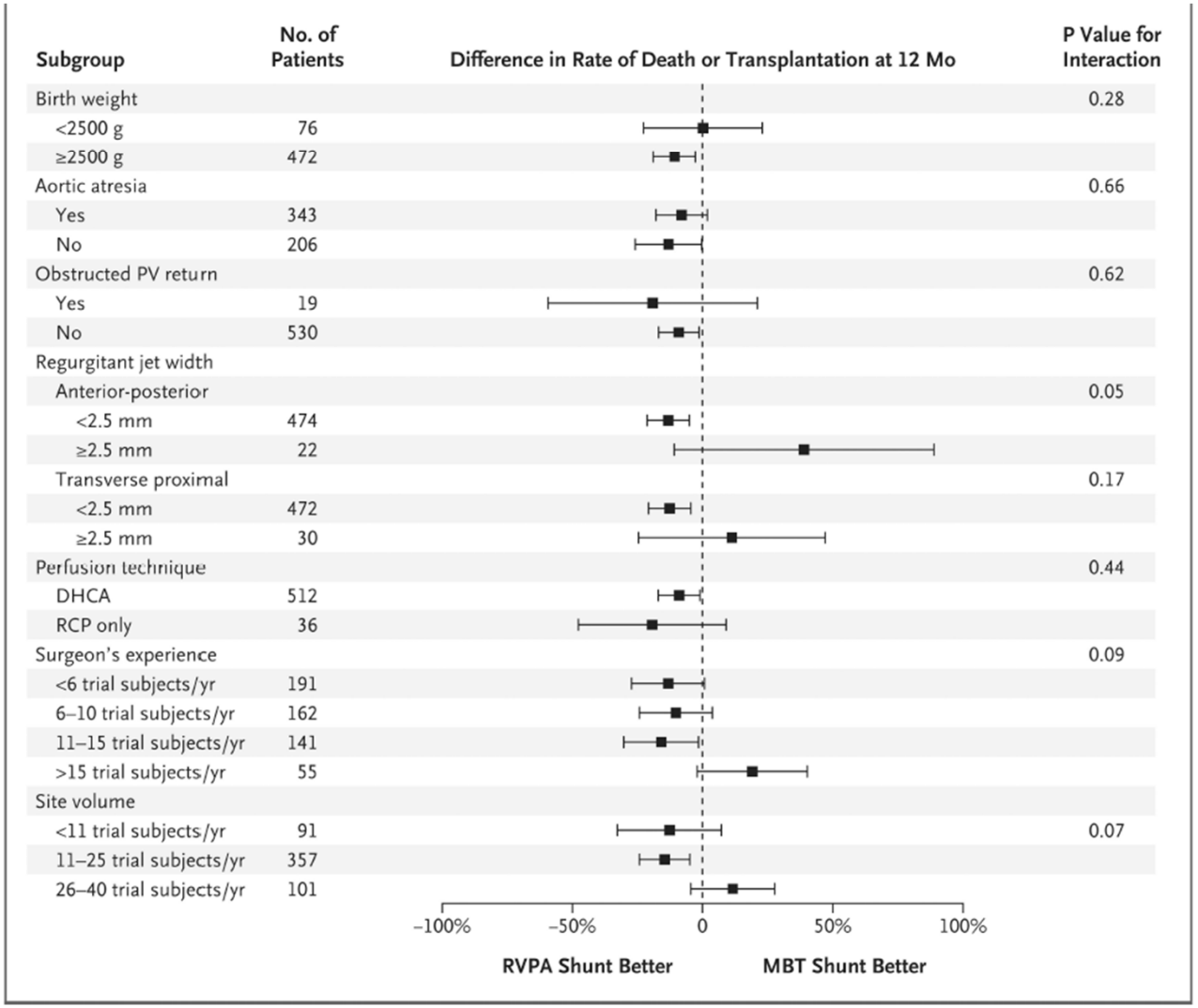
Comparison of Shunt Types in the Norwood Procedure for Single-Ventricle Lesions



RV-PA shunt

- Theoretic advantages
- Absence of diastolic blood flow (systemic to pulmonary)
  - A more stable postoperative course
  - Increased coronary arterial flow because of the lack of aortic diastolic runoff
  - Lower interstage mortality
  - Improved weight gain related to improved splanchnic perfusion

- Potential disadvantages
  - Ventriculotomy
    - Ventricular dysfunction
    - Arrhythmias
    - False aneurysms
  - Free PR - Ventricular dilation,
  - Decreased PA growth related to the lack of forward flow during diastole,
  - Need for earlier stage II procedure because of hypoxemia