Indian Journal of Thoracic and Cardiovascular Surgery (July–September 2018) 34(3):335–336
https://doi.org/10.1007/s12055-017-0642-7

INVITED EDITORIAL

Outcome of Norwood operation for hypoplastic left heart syndrome

Smruti Ranjan Mohanty[1]

Received: 9 December 2017 / Revised: 15 December 2017 / Accepted: 20 December 2017 / Published online: 15 January 2018

In 1938 with ligation of patent ductus by Dr. Robert Gross at Boston Children's Hospital, a new chapter was written in the history of cardiac surgery followed by correction or palliation of all variety of heart defects over the next five decades. The last surgical marvel in pediatric cardiac surgery was the Norwood operation in 1981.

When I asked Dr. Norwood what inspired him to design a surgical solution for hypoplastic left heart syndrome (HLHS), his reply disrobed the secret of this operation. In his early days at Boston Children's Hospital ICU, there was a corner where a few neonates with HLHS were kept. It was seldom visited by the staff and deliberately covered by dark shades. Dr. Norwood used to frequently go to that corner only to be dejected since little could be done for these unfortunate kids. Singling out these babies bothered him constantly. It was the perseverance of this young cardiac surgeon, Dr. William Norwood, who was so intrigued by the complexity of the cardiac entity that today we have an answer for correcting this wide spectrum of cardiac anatomical constellations, known as "Hypoplastic Left Heart Syndrome." It has been 37 years since the first successful Norwood operation was done, and the feasibility of the procedure is beyond doubt as thousands of kids with HLHS are surviving to adulthood in the western world leading a nearly normal life (though the word "normal" is a misnomer).

HLHS is defined as a congregation of lesions due to underdevelopment of the left side of the heart, i.e., the mitral valve, left ventricle, aortic valve, and aortic arch. HLHS occurs in 0.16–0.36 neonates per 1000 live births and comprises 1.4–3.8% of congenital heart disease. Despite the relatively low incidence, it is responsible for around 23% of all cardiac deaths occurring in the first week of life! [1, 2].

✉ Smruti Ranjan Mohanty
smrutiranjan.mohanty@relianceada.com;
smrutiranjanmohanty699@gmail.com

[1] Kokilaben Dhirubhai Ambani Hospital, Four Bunglows, Andheri (West), Mumbai 400053, India

The challenges of treating a baby with HLHS are multi factorial, an important one is the realization that it is not as uncommon as is thought to be. If the cardiac anomaly is diagnosed in utero, it is easy to counsel and prepare the family for a tertiary center delivery, with subsequent stabilization and surgery. If the diagnosis is suspected in a new born, a good echocardiography will clinch the diagnosis. The combination of the different lesions of this spectrum dictates the ultimate line of management. This is one condition where the pre operative stabilization is extremely important in terms of avoiding use of 100% oxygen, starting Prostaglandin E1, and ensuring the adequacy of atrial level communication. It is wiser to subject the baby to the stage I (Norwood) surgery after stabilizing the other organ functions which have a bearing on the ultimate outcome.

As far the surgical technique goes, it has been a constant modification of techniques since the 80s and the ultimate goals are threefold:

1. Ensuring an unobstructed systemic outflow with appropriate coronary supply and normal arch
2. Unrestricted inter atrial communication and
3. Maintenance of pulmonary blood flow with a Modified Blalock Taussig (BT) Shunt/Sano shunt/bilateral branch pulmonary artery (PA) banding.

The end result is to preserve the single right ventricle to maximize its efficiency and durability.

Surgical techniques for the arch reconstruction include excision of coarcted segment and all ductal tissue, a generous amalgamation of smallish ascending aorta and main PA to ensure unobstructed coronary flow, and appropriate augmentation of the arch of the neo aorta with a homograft patch of pulmonary homograft/bovine pericardium/treated autologous pericardium. At the end of the repair, the arch should look like "a normal arch with a gothic architecture". Fortunately, at the origin of the cranial vessels, the arch is usually generous and thus ensures normal flow through those arteries.

Atrial septectomy is a must to ensure unobstructed flow through the interatrial septum.

Controversy lies in the options of providing the pulmonary blood flow. Modified BT shunt is still practiced in high volume centers producing excellent outcomes. It has the advantage of not tampering with the systemic ventricle with a ventriculotomy which has an impact on the long-term ventricular function. The Sano shunt has the advantage of avoiding the coronary steal and a less stormy post operative course, however, at the expense of a ventriculotomy whose long-term consequence is not yet fully defined. Currently, more and more units are opting for it especially one with less volumes with the hope of less interstage mortality. [3–8].

The post operative management is based upon balancing the systemic vascular resistance and pulmonary vascular resistance and thus aiming a Qp: Qs close to 1. In today's practice, it is very much possible with the use of good inodilators, nitric oxide, and other selective pulmonary vasodilators. More and more kids are discharged with stable hemodynamic on full oral feeds to proceed for stage II around 6 months of age.

In the current publication (https://doi.org/10.1007/s12055-017-0603-1), the outcomes achieved by the Polish group are excellent. It shows that if pursued, the outcomes can be life changing in many kids with this extremely difficult lesion. Innovation has been the key to the success that has been achieved to date with HLHS. Importantly, the impetus to apply newer modifications with an aim to improve outcomes should be backed with careful analysis, teamwork, introspection, and reflection. Whether this type of innovative thinking can be maintained in today's global environment of increasing regulation and fear of reporting the "bad" outcome remains to be seen.

Currently, the situation in India is exactly as it was in Boston in 1981. If this anomaly is diagnosed in utero, it is terminated. If neonates are diagnosed with HLHS, they are invariably denied surgical treatment because the treating doctors, due to lack of knowledge regarding HLHS, play the role of soothsayers deciding things beyond their scope. [9–11]

In India, time has come for us to gear up to the challenge of repairing HLHS rather than brushing it below the carpet. It should be pursued holistically taking both the care givers and family into confidence and involving them in the process through all the stages. Nowhere in the field of congenital heart surgery, is a positive outlook and extremely close cooperation between cardiologists, cardiac surgeons, anesthesiologist, perfusionist, intensivists, nurses, and the family required, as it is for treating a child with HLHS. Personally I believe, if a child with HLHS survives till term; in most situations, the anatomy will be favorable for a stage I repair, and if done in the right way will have a good outcome.

In Dr. William Norwood's world, there is nothing called impossible rather it is "I'm possible" and I sincerely believe that with regards to HLHS.

References

1. No authors listed. Report of the New England Regional Infant Cardiac Program. Pediatrics. 1980;65:375–461.
2. Samánek M, Slavík Z, Zborilová B, Hrobonová V, Vorísková M, Skovránek J. Prevalence, treatment, and outcome of heart disease in live-born children: a prospective analysis of 91,823 live-born children. Pediatr Cardiol. 1989;10:205–11.
3. Photiadis J, Sinzobahamvya N, Haun C, et al. Does the shunt type determine mid-term outcome after Norwood operation? Eur J Cardiothorac Surg. 2012;42:209–16.
4. Ohye RG, Sleeper LA, Mahony L, et al. Comparison of shunt types in the Norwood procedure for single-ventricle lesions. N Engl J Med. 2010;362:1980–92.
5. Barron DJ, Brooks A, Stickley J, et al. The Norwood procedure using a right ventricle-pulmonary artery conduit: comparison of the right-sided versus left-sided conduit position. J Thorac Cardiovasc Surg. 2009;138:528–37.
6. Ballweg JA, Dominguez TE, Ravishankar C, et al. A contemporary comparison of the effect of shunt type in hypoplastic left heart syndrome on the hemodynamics and outcome at Fontan completion. J Thorac Cardiovasc Surg. 2009;138:528–37.
7. Sano S, Huang SC, Kasahara S, Yoshizumi K, Kotani Y, Ishino K. Risk factors for mortality after the Norwood procedure using right ventricle to pulmonary artery shunt. Ann Thorac Surg. 2009;87:178–85.
8. Sharma V, Deo SV, Huebner M, Dearani JA, Burkhart HM. In search of the ideal pulmonary blood source for the Norwood procedure: a meta-analysis and systematic review. Ann Thorac Surg. 2014;98:142–50.
9. Balachandran R, Nair SG, Gopalraj SS, Vaidyanathan B, Kottayil BP, Kumar RK. Stage one Norwood procedure in an emerging economy: Initial experience in a single center. Ann Pediatr Cardiol. 2013;6:6–11.
10. Iyer KS. Treating hypoplastic left heart syndrome in emerging economies: Heading the wrong way? Ann Pediatr Cardiol. 2013;6:12–4.
11. Peer SM, Sinha P. Re. Treating hypoplastic left heart syndrome in emerging economies: Heading the wrong way? Ann Pediatr Cardiol. 2013;6:206.