Treating hypoplastic left heart syndrome in emerging economies: Heading the wrong way?

Krishna S Iyer
Department of Pediatric and Congenital Heart Surgery, Fortis Escorts Heart Institute, New Delhi, India

This issue of the Annals of Pediatric Cardiology contains an article entitled-''Norwood Procedure In An Emerging Economy-Initial Experience In A Single Centre.''[1] The authors have retrospectively analyzed and presented their experience with performing the Norwood procedure in seven patients with hypoplastic left heart syndrome (HLHS) (three patients) and other single ventricle situations with uncorrectable left ventricular outflow obstruction (four patients), with five survivors. This is probably the first report of its kind from the Indian subcontinent. The authors deserve credit for their results given the fact that the Norwood procedure has a steep learning curve and this was an initial experience. The title however highlights the setting of an ''emerging economy,'' which compels a critical discussion on whether or not the Norwood procedure should be offered as treatment for HLHS in countries like India which are at the cross-roads of economic development and are thus dubiously labeled as ''emerging economies.''

The Norwood procedure is now increasingly promoted as the standard of care for the initial palliation of children born with HLHS in the developed world (read North America, Europe, Japan, and Australia). However, it was not so long ago that non-intervention or comfort care was an option strongly put across to parents of children born with this fatal condition–despite the fact that the Norwood procedure had been in existence since 1981. The reasons for this were not difficult to understand. The procedure was essentially palliative in nature, operative mortality was high, resource consumption enormous, and survivors did have a significant risk of neuro-morbidity.[2] Neonatal heart transplantation was introduced as an alternative treatment strategy in 1985 by Leonard Bailey, however problems of donor availability and need for lifelong immuno-suppresion have restricted its application. In the past decade operative mortality has been brought down substantially in many of the high volume centers largely due to high rates of ante-natal diagnosis, better understanding of the post-operative management, and an improved understanding of pre-operative variables that predispose to a poor outcome. Given an early survival of upto 90% for standard risk HLHS in experienced institutions, western parents now find it increasingly difficult to choose comfort care as an option. Religious beliefs, ethical and medico-legal issues, and pressure from ''right to life'' organizations also play a role in parents opting for a surgical option even when they are not entirely convinced about its overall benefit. Issues relating to parental decision making nonetheless remain a subject of many a research study or internet blog.[3,4] When antenatal diagnosis has been made a sizeable proportion of parents still opt for termination of pregnancy. Operative mortality for second stage bidirectional Glenn and third stage Fontan conversion is currently extremely low, however interstage mortality still remains significant. All in all, currently, 70% of babies born with HLHS in the United States are expected to reach adulthood.

Improved survival has however come at a cost. HLHS was almost the last frontier for congenital heart surgeons in the developed world and having established uniform standards of care for all the other forms of CHD which was available to every child born with CHD, they were able to concentrate all their energies and resources into developing strategies for its management. In that sense the investment of resources into the management of HLHS did not come at the expense of other children requiring treatment for CHD. This is important to bear in mind considering the fact that HLHS is one of the most expensive CHD's to treat today. A recent analysis of costs for HLHS[5] showed that among 1941 neonates, stage 1 palliation (Norwood or Sano procedure) had a median length of stay (LOS) of 25 days and charges of $214,680. Stage 2 and stage 3 palliation (Glenn and Fontan procedures, respectively) had median LOS (length of stay) and charges of 8 days and $82,174 and 11 days and $79,549, respectively. Primary neonatal transplantation had an LOS of 87 days and charges of $582,920, and rescue transplantation required 36 days and $411,121. The median inpatient wait time for primary and rescue transplants was 42 and 6 days, respectively. Between 1998 and 2007, the LOS for stage 1 palliation increased

Address for correspondence: Dr. Krishna S Iyer, Department of Pediatric and Congenital Heart Surgery, Fortis Escorts Heart Institute, Okhla Road, New Delhi, India. E-mail: iyerks@hotmail.com

from 16 to 28 days and inflation-adjusted charges increased from $122,309 to $280,909, largely because of increasing survival rates (57% in 1998) and 83% in 2007. What is important to note is that not only are the absolute costs of treatment high, irrespective of the type, improved survival has been associated with longer lengths of hospital stays and substantially higher inflation adjusted costs. These costs are clearly unaffordable by individuals and these treatment options are feasible in these countries only because they are funded either by the state or by insurance. In the end however, we are still left with a cohort of palliated patients surviving on a Fontan circulation, which we are well aware is associated with its own limitations. As stated earlier there is also a substantial neuro-morbidity amongst the long term survivors, more so amongst those surviving stormy post-operative courses or requiring cardio-pulmonary resuscitation. The psycho-social impact of HLHS on the parents and on the survivors has been the subject of many studies reflecting the fact that the lesion apart from being a surgical challenge remains a constant source of emotional stress for the affected families.

A recently published ''white paper'' on the current understanding of the management of HLHS elegantly summarizes the knowledge that has been gained over the last three decades.[6] While it generates a lot of optimism for infants born with HLHS today, the summary statement stresses continued caution, as much is yet to be learnt about this condition. To quote ''The dramatic improvements in the treatment and outcomes for HLHS over the past three decades have been accomplished through the efforts of many dedicated providers, families, and patients. Current successes and expectations that 70% of newborns born today with HLHS may reach adulthood are exciting, yet one must always remember that the current surgical strategies remain palliative. Caution must be exercised because a great deal remains to be understood as it relates to this group of patients, their QOL (quality of life), their long-term morbidities, and the sequelae of recent surgical modifications.''

In developing countries like India, CHD remains largely untreated. Available facilities and resources fall way short of the demand, as a result of which the majority of patients even with simple CHD perish without receiving surgery.[7] Most of the surgeries today are performed in the private sector where the costs have to be borne by the families of the affected infant. Although health insurance is gaining popularity in the country, it still remains largely unavailable for congenital heart disease. In public hospitals where free or subsidized treatment is available, the demand is so high that waiting lists are ever increasing and many children with easily correctable lesions miss the boat. In this milieu, there is need to allocate resources in a responsible way and thereby a need to take important decisions as to which conditions should be treated and which should probably be left alone at this stage of our development. The authors in this study have concluded that the cost to the patient has not been prohibitively high, however, they have taken into consideration only the material costs. What has not been computed is the costs of time, manpower, and other non-billed hospital resources that are consumed for each operation. Also costs for inter-stage management and subsequent surgeries have not been included and these can be substantial for the families in the Indian milieu. What is important is that unlike in the developed countries, resources consumed for the management of HLHS come at the expense of children with correctable lesions. So when we look at costs in the Indian milieu, we need to calculate how many corrective surgeries could have been performed with the same resources. Many would argue that every child deserves a chance to live and that by this logic we should not be performing open heart surgery at all in a land where hundreds of thousands of children die every year of diarrhea, malnutrition, and infectious disorders. While we have to accept that we live in a land of glaring anomalies and disparities where cardiac surgery itself may be viewed as a frivolous luxury, we do not have to add insult to injury by adding the Norwood operation to our surgical basket.

An interesting study in Norway[8] revealed that parents of babies born with HLHS who opted for comfort care over surgical treatment tended to be more educated and more likely to be associated with the healthcare profession. This would indicate that the more knowledgeable parents were about the long term outcomes of HLHS, the less likely they were to proceed with a surgical option. Indian families in general have limited awareness and generally tend to have blind faith in their physician's advice. Most tend to brush aside or gloss over long term concerns, believing that their child would by some miracle remain on the right side of the survival curve. A decision to proceed with surgery is often an emotional one, rather than one taken logically after weighing all the pros and cons. Often a surgical option is taken because it provides a short term solution ''one way or the other'' irrespective of the long term consequences. Comfort care is often not accepted because parents feel guilty about not doing something active to save their child's life and just watch them die helplessly. The responsibility therefore lies on the treating team as to whether to provide a surgical option or not. If a surgical option is offered many are likely to opt for it blindly, and bankrupt themselves in the process.

It is tempting for pediatric units to perform the Norwood operation in the belief that the challenges it poses helps sharpen the skills of the surgical, perfusion and intensive care teams, and outcomes for simpler operations would improve as a fallout. Professional growth for surgeons,

especially the younger ones, involves a steady increase in the level of complexity of the surgical procedures they perform. In the current era the Norwood procedure may well be considered to be one of the most challenging procedures in congenital heart surgery today and it is inevitable that for that reason alone more and more Norwood procedures will be performed in our country. However, what young surgeons need to understand is that there are many more corrective operations in congenital heart surgery which are equally challenging but much more rewarding e.g., an arterial switch with arch reconstruction for a Taussig-Bing heart with hypoplastic aortic arch or double-switch after left ventricular retraining for corrected transposition. Pediatric surgical units would be better rewarded by working towards improving their outcomes with complex but corrective procedures and using that as a marker of their proficiency rather than embark on doing an occasional Norwood procedure just to be able to say 'we can also do it'.

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