Editorial

Definitive therapy for hypoplastic left heart syndrome — Indian scenario

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When most of the centers in India do not entertain the patients with hypoplastic left heart syndrome, the original authors of the article in the present issue should be congratulated for the attempt of treating this patient with hybrid procedure. It’s a spectacular beginning.

The development of staged reconstructive surgery for hypoplastic left heart syndrome (HLHS) by Norwood and colleagues in early 1980 has radically altered the prognosis of this most complex heart problem.¹ The important anatomical features of HLHS are severe aortic and mitral valve hypoplasia or atresia with underdevelopment of left ventricle and varying degrees of hypoplasia of aortic arch. Reconstructive surgery for this condition, which is essentially a long-term palliation, is traditionally performed in three stages. The first stage is performed within the first few days of life which consists of reconstruction of ascending hypoplastic aorta using proximal main pulmonary artery, removal of interatrial septum and insertion of systemic to pulmonary artery shunt or a right ventricle to pulmonary artery shunt (Sano shunt) to maintain pulmonary blood flow. The second stage is bidirectional Glenn shunt which is carried out between 4–12 months and the third stage is completion Fontan which is performed by 2–6 years of age.

Recently an innovative technique has been described in which stenting of ductus arteriosus is performed in the cardiac catheterization laboratory and bilateral pulmonary arterial banding is done by the surgeons.² This technique is labeled as “Hybrid Procedure”. In this issue of Indian Heart Journal Anuradha et al from Frontier Lifeline Hospital, Chennai have described a case in which they performed Hybrid Stage 1 palliation for hypoplastic left heart syndrome in a two month old male infant working in a conventional cathlab (2012;64:333–337). In addition to placement of a stent in the duct and pulmonary artery banding, the authors have placed a stent in the ascending aorta because of extensive arch hypoplasia. Potential advantage of the hybrid technique is avoidance of circulatory arrest in the early neonatal period and shifting the major surgical stage to later period until the brain is more developed. The primary goal of the procedure is to create a stable and balanced circulation without the use of open-heart surgery with its associated risk in a neonate. By shifting the age of bypass, there may be a dramatic impact to the patients neurologic development. Both these strategies have a common goal. They provide unobstructed systemic cardiac output, controlled source of pulmonary blood flow, reliable source of coronary blood flow and unobstructed egress of pulmonary venous drainage. The initial experience of hybrid technique is still developing and will have to demonstrate superior outcomes to the more traditional strategy. We have to study the hemodynamics of this child after 3 months as it is sometimes very difficult to control the pulmonary blood flow with this technique. In addition, extensive arch repair is a major advantage by surgery which determines the long-term outcomes of the Fontan procedure.

The procedure described in this issue was performed in the conventional cardiac catheterization laboratory. Ideally these procedures are performed in “Hybrid Suites”. These suites are designed around the cardiac surgeons and cardiac surgical patients offering advanced access and imaging in the operating room. There is no need to shift these critically ill patients from catheterization laboratory to surgical theaters. The suites accommodate collective catheterization and surgical team and facilitate rapid information sharing. These

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suites are expensive and we may have to justify their utility with the present numbers of patients who may need this facility.

Diagnosis of HLHS can be made easily in fetal life. It is a silent disease in utero and most of these fetuses complete the gestational term. Inherent tricuspid regurgitation may exist and might progress with gestation and is an important risk factor for poor post-surgical outcome. Size of the interatrial septal defect has a bearing in the development of the pulmonary vasculature and ultimate outcome. The ability to diagnose the lesion with certainty in fetal life and the extremely poor outcome in HLHS with intact or restrictive interatrial septum becomes a potential prenatal intervention for these babies in the form of balloon atrial septostomy. Babies born with HLHS need stabilization with PGE1 infusion immediately after birth to keep the ductal patency and maintain the perfusion of lower body and retrograde perfusion of cerebral and coronary circulation.

As corrective surgery for many biventricular heart lesions has become a real possibility, more and more neonates with biventricular hearts are undergoing the required procedure early in life with excellent long-term outcomes. There has been steady improvement in terms of miniature hardware, micro instrumentation, advanced miniature cardio pulmonary bypass circuits with oxygenators along with improvement in care of these newborns in the postoperative phase. Today there is a pattern of predictability even in preterm and low birth weight babies with congenital heart defects with good chances of survival after corrective surgery.

However the story with HLHS is more complicated as there is a great variability in the anatomic substrate. The anatomy varies from severe hypoplasia of mitral, aortic valves, aortic atresia with hypoplastic aorta to reasonable sized left ventricle with severe left ventricular outflow obstruction where surgical options can be quite different. Mortality figures for stage I Norwood is center specific with mortality rates vary from 5–10% in experienced hands to in excess of 20% in developing countries. The improved survival after stage 1 Norwood has resulted in acknowledging the existence of interstage mortality upto 10%. We need to see whether the hybrid procedure addresses these issues on long-term follow-up. There has been a tilt toward early second stage in the form of a bidirectional Glenn or a hemifontan operation around 5–6 months of age. Morbidities such as exercise intolerance, arrhythmias, thromboembolic diseases, protein losing enteropathy and neurocognitive disorders have come to light in some of these children reaching teenage like other Fontan procedures. The eventual long-term survival after 3 stage reconstructive procedure for HLHS depends upon the right (neo-systemic) ventricular function and the degree of tricuspid regurgitation. Some of these survivors will need heart transplant owing to a failing Fontan in the years to come. In the long term there is uncertainty with the patients with HLHS with Fontan circulation as the right ventricle remains as systemic pump. Continuing improvements in surgical techniques may not only minimize peri and postoperative mortality but also reduce the risk of complications in the medium and long term. Otherwise heart transplantation remains the only option for these failing Fontan circulation.

Antenatal diagnosis has a major important role which allows the parents to consider treatment options and to arrange for a elective delivery in a specialized center. It can define the unfavorable intracardiac anatomy and presence of extracardiac defects which help parents to take decision. It also allows for a coordinated services to be provided between fetal cardiology, neonatology, pediatric cardiology and cardiac surgery. In order to counsel the parents antenatally, it is essential to understand the natural history of this condition.3,4 Confirmed or suspected genetic syndromes are frequent in patients with HLHS. Because early surgical palliation is necessary, identifying risk factors for worsened neurodevelopmental outcomes can be difficult. After the surgery, about 8–10% patients need ECMO support. There is a need to see long-term neurodevelopmental outcomes of these patients. Larger series have shown significant neurodevelopmental disabilities in upto 65% of patients.5 The hybrid procedures may prove superior to avoid these complications.

As far as the Indian scenario is concerned, with the incidence of HLHS about 0.1–0.25/1000 live births, we should expect about 2000 babies with HLHS born every year. In reality the total number of babies with HLHS seen in major centers in India will not be more than 100 children/year. Most of the fetuses with diagnosis of HLHS get terminated by parents options and counseling offered to them by perinatologists. The termination rates are quite high with additional extracardiac abnormalities with abnormal karyotyping. As these neonates are very sick after birth, unless a correct diagnosis is made and appropriate treatment is started for stabilization, there is a very high chance of early neonatal loss. As a result, very few neonates reach the tertiary care centers for evaluation and further management options. In the absence of prenatal diagnosis, most of these newborns present in a very sick and metabolically compromised state where the success rates of 1st stage Norwood repair are very low.

There are certain ethical, social and financial issues to be considered with us in India. Only a fraction of population has insurance cover. The young parents have to fund these operations from their meager savings and whether they will be able to fund all the three stages of operations is a big question mark. After counseling and knowing the need for multistaged repair, most of the parents opt for compassionate care and will not like do active management. They instead look forward to have a normal child in future. Very few parents want to go ahead with the complex management options in a precious child especially if the baby is born with assisted reproductive techniques. Unfortunately we have not been able to develop enough expertise to treat this condition as each center deals with a small number of babies. We need a team approach to take care of these complex anomalies which include expertise in pediatric cardiology, cardiac surgery, anesthesia, intensive care and perfusion. It’s an expensive affair.

Few centers in India have performed classical Norwood procedures. Our center has performed total six classical Norwood procedures till date. The surgeons performed extensive arch repair with Damus Kaye Stansel procedure and modified Blalock-Taussig shunt. Four patients had classical anatomy of HLHS i.e. mitral atresia with aortic atresia and hypoplastic aortic arch. Two had univentricular hearts with severe
subaortic stenosis. Out of six patients, three patients survived the first stage. Out of the three patients who did not survive the first stage, one baby was in severe low cardiac output with metabolic acidosis on presentation and the other two had severe tricuspid regurgitation. Retrospectively we thought that they were not the ideal candidates in the beginning of the program. Two patients underwent the second stage of Norwood repair i.e. bidirectional Glenn shunt and the third is awaiting.

The western world has already completed three decades of taking care of this complex defects and their earlier operated patients have already reached in twenties. They have overcome the initial hitches. What strategy should we follow? All the centers may not be able to offer the same survival statistics based on their own experiences. This raises the question of regionalization of care and referral to centers of excellence specifically identified as able to treat this anomaly. A regional referral center which has a track record of handling neonates successfully should gradually develop the needed expertise. Survival rates can improve when the procedure and postoperative care is taken care of by experienced hands.

REFERENCES