Prenatal and postnatal counselling in hypoplastic left heart syndrome

Babey A. Zainab, MBBS, Arab Board, Giuffre R. Michael, FRCPC, FACC, Hasan U. Shabih, MD, Ian R. Lange, MD, FRCSC.

ABSTRACT

Hypoplastic left heart syndrome is an uncommon cardiac malformation, that if untreated, usually results in death within the first month of life. Counselling of parents choosing palliation may be complicated by prolonged survival without ongoing medical intervention. We present 2 neonates with hypoplastic left heart syndrome undergoing palliative care who survived for longer than 6 months.

Keywords: Hypoplastic left heart, counselling, survival, palliative care.

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Hypoplastic left heart syndrome (HLHS) is currently the most common cardiac malformation that results in neonatal death. Without treatment, 95% of these infants die in the first month of life, and almost none survive beyond 4 months. This information is commonly used when counselling parents and health care professionals on prenatal management and expectation for outcome. We report two cases of HLHS with prolonged neonatal survival with minimal intervention.

Case Reports.

Patient 1. A 28 year old gravida 3 para 1 patient was diagnosed at 32 weeks gestation to have a fetus with a hypoplastic left ventricle. The couple were informed that the anomaly was lethal without surgical intervention. They were offered a variety of management options for their baby. At their request, labor was induced at 34 weeks gestation. The infant weighed 1522 gm and was admitted to the NICU with mild respiratory distress. An echocardiogram showed a severely hypoplastic left ventricle, atretic mitral and aortic valves and a small aortic arch. At the parents request there was minimal intervention which included assisted ventilation and Prostaglandin E2. At 33 days of age with an uncomplicated course the baby was bottle feeding adequately and was discharged home. Prolonged survival occurred and treatment options were again discussed with the parents who later consented to the Norwood procedure. This was performed at 6 months of age. Unfortunately the baby died within the immediate post operative period in hospital.

Patient 2. After a normal pregnancy, a 35 year old primigravida gave birth at 39 weeks gestation to an infant weighing 4090 gm. The baby was cyanosed at birth and was admitted to the NICU for assessment. The echocardiogram demonstrated a moderately hypoplastic left ventricle, small atrial valve and left outflow tract, and a patent ductus arteriosus.

After appropriate counselling the parents chose palliative care. The prostaglandin infusion used to maintain ductal patency was discontinued. The baby was transferred to the special care nursery where he remained stable and was discharged home on day 9 of life. At the age of 6 weeks, the baby deteriorated and parents then requested intervention. The baby...
had a cardiac catheterization and aortic valvoplasty. The valvoplasty was subsequently repeated with good clinical response and the baby continued to do well until 6 months of age. The baby then presented with severe viral pneumonitis and died soon after hospital admission.

**Discussion.** HLHS is a congenital heart defect that ranges in severity from critical aortic stenosis with nearly normal left ventricular and mitral dimensions, to the most severe form where both the aortic and mitral valves are atretic and the left ventricle is absent or extremely underdeveloped.7 Our first case represents a more severe form of the condition while the second case was of a moderate severity.

The neonate with HLHS is dependent on persistent ductal patency to provide systemic blood flow. Interruption of ductal flow usually results in neonatal death within the first 3 days of life.3,4 Overall, the prognosis of the majority of pregnancies with in utero diagnosis of HLHS is poor and is negatively influenced by the presence of cardiac failure, aneuploidy or extra cardiac malformation or a combination of these.1,3,5,6

The antenatal diagnosis of HLHS necessitates counselling of the parents about the diagnosis and management options. These include termination of the pregnancy when the gestation is < 24 weeks, continuation of the pregnancy and neonatal surgical intervention with either neonatal cardiac transplantation or Norwood procedure, or no intervention with compassionate care of the baby after birth.1 As postnatal ductal patency can be achieved by Prostaglandin E2, its use may contribute to cases of prolonged survival.

The Norwood procedure necessitates multiple surgical interventions that result in a Fontan like repair with the right ventricle functioning as the systemic ventricle.7 Early neonatal transplant of the heart seems promising as there may be less organ rejection due to an immature immune response, however the potential for developing accelerated coronary arteriosclerosis in the allograft is disconcerting.3 Important ethical issues arise when choosing a Norwood procedure or to wait for heart transplantation, when to place a fetus on the waiting list for transplant, setting priorities between infants and fetal candidates for a donor organ, and when and where to deliver.

Many parents may consider compassionate care for their neonate in view of the high mortality and morbidity, including neurodevelopmental problems with survival and the final cost associated with surgical intervention.9,11 Rogers et al12 reported 64% of infants with HLHS who survived staged surgical repair had major developmental disabilities and that the quality of life outcomes must be considered when management options are evaluated.

In cases of prolonged survival, the options are still either palliative care or surgery. The ethical implication of either treatment or the refusal of treatment may evoke different and strong opinions among the family as well as health care professionals. The health care team is charged with the responsibility of maintaining an unbiased and non-judgmental approach to determine which treatment option is preferable. If surgical intervention is chosen, the parents must consider the overall family unit, their emotions and financial situation.

The counselling of parents and family pre and postnatally for HLHS is important and must include the possibility of prolonged survival with minimal intervention. This discussion may help prepare parents emotionally and socially for such an event and may avoid medical legal issues.

**References**


