Communications

Hypoplastic Left Heart Syndrome

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Hypoplastic left heart syndrome is one of the common causes of mortality in infants with congenital heart disease, accounting for approximately 22 percent of infant deaths from congenital heart disease in the neonatal period.[1] The syndrome is probably more common than realized, since the majority of the infants with this syndrome die within the first week of life, sometimes without definitive diagnosis. This report describes a documented case in a newborn male infant, demonstrating how the diagnosis can be made and confirmed at the bedside using echocardiography and a flush aortogram.

Case Report

A 5-lb, 8-oz male term infant was born to a 21-year-old gravida 2, para 1 white woman under saddle block anesthesia. The mother's prenatal course and birth history were normal. The baby was delivered from a left occiput anterior position with an Apgar of 9 at one and five minutes. Bulb suction was used for resuscitation. On examination the baby was found to have a cleft of the soft palate. There were no other apparent abnormalities. The cardiovascular examination was normal. The following morning a grade 1 to 2 systolic murmur was heard over the left precordium. The baby was pink and comfortable in room air. The liver was 2 cm below the right costal margin. At approximately 38 hours of age the baby was reexamined and found to have a respiratory rate of 40/min and a heart rate of 130 beats/min. The liver was felt to be 3.5 cm below the right costal margin, and a chest film was obtained. As a result of the enlargement of the heart and the increase in pulmonary vascularity, the baby was transferred to Kosair-Children's Hospital Tertiary Care Nursery for evaluation and treatment. Upon arrival the baby was observed to be slightly tachypneic with a heart rate of 150 beats/min. The liver was noted to be 5 cm below the right costal margin. The patient was diagnosed as being in congestive heart failure, and he was digitalized using digoxin (Lanoxin) at 0.04 mg/kg as the total daily dose. He was also given a dose of furosemide (Lasix) at 1 mg/kg by intravenous route. At approximately 52 hours of age the patient was seen by the pediatric cardiologist. Upon examination the baby was observed to be in mild cardiorespiratory distress with no overt cyanosis. The first heart sound was normal. The second heart sound was loud and single at the upper left sternal border. The liver was 5 cm below the right costal margin. The peripheral pulses in the upper extremities were very feeble, and the pulses in the lower extremities were not palpated. The electrocardiogram revealed a heart rate of 150 beats/min and an abnormal frontal axis of +35°. The baby was clinically judged to have severe left ventricular outflow tract obstruction. An echocardiogram showed an absence of the mitral and aortic valves with severely hypoplastic ascending aorta (Figure 1) and left ventricular cavity. The echocardiographic diagnosis of hypoplastic left

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sions and conditions using two-dimensional echocardiography, most physicians continue to use single-film retrograde umbilical aortography to confirm the diagnosis.

A few centers are experimenting with palliative surgical procedures,[12] but most subscribe to the philosophy that the severe nature of the anatomic malformation in this cardiac lesion prohibits a reasonable survival, with or without surgery. In these centers only supportive therapy is recommended. Once the diagnosis has been made and confirmed, the parents are informed of the lethal nature of their infant's cardiac malformation with as much tenderness and caring as possible. Every effort is made to keep the baby comfortable while at the same time discontinuing life-support drugs such as digitalis and avoiding cardiopulmonary resuscitation. When possible, the babies are transported back to the referring hospital so that they can be with their mothers. In those rare babies that continue to do well after the mother is discharged, hospice is involved and the baby is permitted to die a natural death at home.

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