Surgery Gives Infants With Heart Defect Chance Of Survival

When prenatal diagnosis detects the severe heart defect hypoplastic left heart syndrome (HLHS) in a fetus, a comprehensive prenatal evaluation is important to provide parents an accurate prognosis. In HLHS, one of the heart's pumping chambers is severely underdeveloped. However, say researchers, in two-thirds of cases, reconstructive surgery affords the infant an excellent chance of early survival.

Researchers from The Children's Hospital of Philadelphia report on five years of experience at that hospital, in a review of 240 fetuses diagnosed with HLHS from 2004 to 2009. Children's Hospital has some of the world's longest and most extensive experience in performing staged surgical repair of HLHS.

"Because we have offered this type of reconstructive heart surgery for over 25 years, our goal was to establish a benchmark for perinatal and early surgical outcome in the present era," said Jack Rychik, M.D., medical director of the Fetal Heart Program at Children's Hospital.

Rychik was the primary investigator of the study, published in the October issue of the journal Ultrasound in Obstretrics and Gynecology.

In HLHS, one of the most common forms of congenital heart disease diagnosed before birth, the underdeveloped left ventricle is unable to properly circulate blood. Over the past 25 years, surgeons at Children's Hospital of Philadelphia and elsewhere have developed and refined reconstructive surgery—currently a series of three planned procedures, beginning in the newborn period and extending to 1 ½ to 4 years of age. Although outcomes vary broadly worldwide, overall survival rates for children with HLHS have steadily improved.

In the current study the researchers classified 162 (68 percent) of the 240 fetuses as standard-risk, and 78 of them (32 percent) as high-risk. In high-risk cases, in addition to the severely underdeveloped left ventricle, the fetus also had genetic and chromosomal defects, prematurity, or other heart abnormalities.

Of the 240 fetuses diagnosed with HLHS, 185 newborns underwent the first stage of surgery, called the Norwood procedure, resulting in 155 survivors and 30 deaths. Within those overall figures, 93 percent of standard-risk cases survived the first operation, compared to 57 percent of high-risk cases.

The Children's Hospital of Philadelphia typically employs a staged surgical approach for fetuses diagnosed with HLHS, says Rychik, but at that center and elsewhere, some parents choose to terminate a pregnancy or to decline medical intervention at

birth. The current study, he says, may provide clarity to families and caregivers in categorizing the degree of mortality risk from this condition.

"Surgical outcomes for HLHS are in-part related to patient volume, institutional experience, and the availability of dedicated resources," said Rychik. "However, we found a striking survival advantage for the standard-risk fetuses compared to the higher-risk cases. Two-thirds of fetuses with HLHS do not have a higher-risk form of the condition, and have a stronger chance of survival. After an initial prenatal diagnosis of HLHS, we strongly encourage families to receive a comprehensive evaluation including amniocentesis, so they may obtain a more accurate prognosis. In this way, families can have the best information during prenatal counseling by which to make their plans for the future of their fetus and newborn child."

"Our current research and clinical efforts are focused on improving the quality of life and long-term survival for this group of patients, including dedicated follow-up programs for the evaluation and treatment of cardiac and non-cardiac outcomes," said Gil Wernovsky, M.D., associate chief of cardiology and director of the NeuroCardiac Care Program at Children's Hospital. He added, "Many of these children will face life-long challenges as they mature, and we are committed to understanding and improving these on-going issues as our patients grow."

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