SAVING DE'ANGELO: MONITORING LITTLE HEARTS AT HOME REPORT #2875

BACKGROUND: Hypoplastic Right Heart Syndrome, or HRHS, can be a range of right-sided congenital heart defects in which the right-sided structures are underdeveloped or not formed. These structures include the tricuspid valve, right ventricle and/or pulmonary valve. When these structures are too small or do not function properly, the right side of the heart cannot send enough blood to the lungs, which causes a baby with untreated HRHS to have abnormally low oxygen levels. The treatments for HRHS have developed considerably in the last few decades and are tailored to each baby's needs. HRHS is less common than hypoplastic left heart syndrome, or HLHS. Within the U.S., HRHS occurs in 1 in 60,000 births, whereas HLHS occurs in 1 in 4,300 births.
(Source: https://www.ssmhealth.com/cardinal-glennon/fetal-care-institute/heart/hypoplastic-right-heart-syndrome and https://en.wikipedia.org/wiki/Hypoplastic_right_heart_syndrome)

CURRENT TREATMENT FOR HRHS: Most babies with HRHS can be delivered at full-term. After delivery, the baby will need prostaglandin (PGE), an intravenous medication that keeps the patent ductus arteriosus (PDA) open. The PDA will allow blood to get to the lungs. A cardiac catheterization may be required to perform a balloon atrial septostomy in the first few days of life. This x-ray is a guided procedure done by placing a thin flexible tube, or catheter, through the umbilical vein and does not require an incision. A balloon atrial septostomy creates a larger hole between the top chambers of the heart. The interventions for HRHS vary depending on which structures are abnormal. In more severe cases, the right heart is too small to send enough blood to the lungs. Shortly after the baby's birth, surgeons perform the first of a series of surgeries done to increase blood flow to the lungs and bypass the poorly functioning right side of the heart. These surgeries are not able to cure HRHS but do provide for a functioning heart.
(Source: https://www.ssmhealth.com/cardinal-glennon/fetal-care-institute/heart/hypoplastic-right-heart-syndrome)

FURTHER RESEARCH: The Fontan procedure has allowed more people born with only one ventricle, the lower pumping chamber of the heart, to survive into adulthood. A new scientific statement published in the American Heart Association journal, *Circulation*, summarizes how best to care for these unique patients. People with Fontan circulation have chronically elevated pressure in their veins and less blood being pumped out of their heart, which can lead to circulatory failure because when the heart pumps less efficiently it is not able to provide enough oxygen to the cells in the body. "We need more research into the basic biology of single ventricle hearts and whether the damage to other organ systems, such as kidneys, liver and brain can be mitigated or reversed," said Jack Rychik, MD, Robert and Dolores Harrington endowed chair in cardiology and Professor of Pediatrics, Children's Hospital of Philadelphia. Although life expectancy for people born with one ventricle is lower than average, people with Fontan circulation can live a fulfilling life.
(Source: https://www.sciencedaily.com/releases/2019/07/190701144633.htm)

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