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Positive outcome in a patient with hypoplastic left heart syndrome: Key factors for successful survival: A case report

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Background: Hypoplastic left heart syndrome (HLHS) is a rare, life-threatening congenital heart defect characterized by underdeveloped left-sided cardiac structures, severely impairing systemic circulation. Survival in the neonatal period depends on a patent ductus arteriosus (PDA), maintained pharmacologically using prostaglandin E1 (PGE1). Without timely intervention, HLHS is uniformly fatal and requires a staged surgical approach, typically beginning with the Norwood procedure.

Case Description: A full-term female neonate born at 39 weeks in Florida with a birth weight of 4.3 kg presented shortly after birth with cyanosis, tachypnea, low oxygen saturation, hypotension, and pallor. Prenatal echocardiography had revealed HLHS with mitral and aortic atresia, mild tricuspid regurgitation, large atrial septal defect, and severe hypoplasia of the ascending aorta. These findings were confirmed postnatally. Due to hemodynamic instability, an umbilical venous catheter was placed for PGE1 infusion to maintain ductal patency and support systemic perfusion. The patient underwent several interventions, including pulmonary artery (PA) banding, the Norwood procedure, bilateral Glenn PA plasty, and cardiac catheterization with PA stenting and collateral occlusion. Postoperative recovery was favorable. At discharge, she was prescribed aspirin, enalapril, famotidine, and furosemide. By 21 months, she demonstrated age-appropriate neurodevelopment without deficits, normal growth, and remained on aspirin with regular follow-up.

Discussion & Conclusion: This case highlights the importance of early prenatal diagnosis, prompt initiation of PGE1, and a coordinated surgical plan in achieving favorable outcomes in HLHS. While still high-risk, HLHS can be survivable with early intervention, meticulous perioperative care, and multidisciplinary collaboration.

Biography

Guiscenthia Rose Tharlene Auguste is a general physician who graduated in 2024 from Universidad Marista de Mérida, Yucatán, México. She is currently preparing for the USMLE Steps and aspires to pursue a residency in Pediatrics. Her main interests include pediatric cardiology, neonatal care, and medical research. She has collaborated on several research projects during her clinical rotations and academic training, resulting in published articles. In addition, she participated in a genetics symposium in 2019. She is passionate about advancing evidence-based pediatric care and is committed to continuous learning, academic growth, and contributing to interdisciplinary healthcare teams.

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