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授課主題

Hypoplastic Left Heart Syndrome: Case Discussion and Anesthesia

Relevance

摘要

Hypoplastic Left Heart Syndrome (HLHS) is one of the most severe forms of congenital heart disease (CHD). It is characterized by underdevelopment of the left-sided cardiac structures, resulting in an inability of the left ventricle to support systemic circulation. Without timely intervention, HLHS is associated with extremely high neonatal mortality. Advances in prenatal diagnosis and staged surgical palliation have significantly improved patient survival and long-term outcomes.

The management of HLHS relies on a series of staged reconstructive procedures rather than definitive anatomical correction. The first stage, the Norwood procedure, is typically performed during the neonatal period to establish adequate systemic circulation while maintaining pulmonary blood flow. This is followed by the bidirectional Glenn procedure at 4–6 months of age, which reduces ventricular volume overload by directing superior vena cava blood flow to the pulmonary arteries. Finally, the Fontan procedure, usually performed between 2 and 4 years of age, completes total cavopulmonary connection, allowing passive pulmonary circulation while the single ventricle supports systemic perfusion.

This presentation also discusses the perioperative anesthetic considerations in patients with single-ventricle physiology, including maintenance of balanced pulmonary and systemic blood flow, optimization of ventricular function, and prevention of postoperative complications. Modified ultrafiltration after cardiopulmonary bypass is highlighted as an effective strategy to reduce systemic inflammatory response and improve postoperative recovery. In addition, a



representative clinical case involving complex congenital heart disease with functional single-ventricle physiology is presented to illustrate individualized surgical planning and long-term management. Although HLHS remains an incurable condition, multidisciplinary care and staged palliation have markedly improved survival, functional outcomes, and quality of life for affected patients.