Hypoplastic right heart syndrome

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Hypoplastic right heart syndrome. ORPHA:98723

Hypoplastic right-heart syndrome (HRHS) is a rare, cyanotic congenital heart malformation (see this term) caused by underdevelopment of the right-sided heart structures (tricuspid valve, RV, pulmonary valve, and pulmonary artery) commonly associated with an atrial septal defect, ostium secundum type (see this term). Pulmonary blood flow is diminished and right-to-left shunting occurs at the atrial level, leading to dyspnea, fatigue, atrial arrhythmias, right-sided heart failure, hypoxemia, repeated miscarriages that were mostly due to hypoxemia and cyanosis. Two subtypes of HRHS have been characterized: pulmonary atresia-intact ventricular septum and right ventricular hypoplasia (see these terms).