

Opis choroby *

Definicja

A rare, cyanotic congenital heart malformation 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. 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.

Dane

Klasyfikacja

Grupa fenomenów

Kod ORPHA

98723

Kod OMIM

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Kod ICD10

Q22.6

Kod ICD11

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*Źródło

orphanet