Opis choroby *

Definicja

Pulmonary arterial hypertension associated with congenital heart disease (PAH-CHD) is a form of pulmonary arterial hypertension (PAH, see this term), characterized by elevated pulmonary arterial resistance leading to right heart failure occurring as a common complication of congenital heart malformations (see this term) with left to right cardiac shunts. Eisenmenger syndrome (see this term) is the most advanced form of PAH-CHD and is defined as the complete or partial reversal of an initial left-to-right shunt to a right-to-left shunt, causing cyanosis and limited exercise capacity. PAH-CHD also includes mild to moderate systemic-to-pulmonary shunts with no cyanosis at rest, patients with small defects, and those with residual PAH following corrective cardiac surgery.

Dane

Klasyfikacja
Grupa fenomenów

Synonimy
PAH associated with congenital heart disease
PAH związane z wrodzoną chorobą serca

Kod ORPHA
275803

Kod OMIM
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Kod ICD10
I27.2

Kod ICD11
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*Źródło

orphanet