

Agnezja zastawki płucnej

Kod Orpha: 982 Kod OMIM:

Opis choroby *

Definicja

Pulmonary valve agenesis is a rare congenital heart malformation characterized by a total or partial absence of the pulmonary valve leaflets associated with stenosis of the pulmonary artery orifice and aneurysmal dilatation of the pulmonary arteries. It usually occurs in association with additional cardiovascular malformations such as tetralogy of fallot or ventricular septal defect, or can occur as part of a syndrome (e.g. 22q11.2 deletion syndrome). Clinical features depend on the presence of associated cardiac malformations and include pulmonary insufficiency, bronchial obstruction (secondary to compression by aneurysmally dilated pulmonary arteries), pulmonary stenosis, cyanosis, and cardiac failure.

Dane

Klasyfikacja

Grupa fenomenów

Synonimy

Absent pulmonary valve syndrome

PVA

Wrodzony brak zastawki płucnej

Congenital absence of the pulmonary valve

PVA

Kod ORPHA

982

Kod OMIM

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Kod ICD10

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Kod ICD11

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[*Źródło](#)

orphanet

Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.

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