Opis choroby *

Definicja

A rare, life-threatening, congenital, non-syndromic, conotruncal heart malformation disease characterized by absent or severely undeveloped pulmonary valve leaflets (with a restrictive ring of thickened tissue at the place of the pulmonary valve annulus), associated with an intact ventricular septum and a patent ductus arteriosus, manifesting with marked respiratory insufficiency. Additional features include dilated main pulmonary artery (with or without dilatation of pulmonary artery branches), to-and-fro flow at site of the dysplastic pulmonary valve, and systolic pressure gradient across narrowed pulmonary valve. Tricuspid atresia and variable extra-cardiac anomalies (e.g. diaphragmatic hernia or cleft lip/palate), may be present.

Dane

Klasyfikacja
Zespół wad wrodzonych

Synonimy
APV/PDA, non-Fallot type
Brak zastawki płucnej - ubytek przegrody międzykomorowej - przetrwały przewód tętniczy
APV/PDA, non-Fallot type
PVA/PDA, non-Fallot type

Kod ORPHA
99048

Kod OMIM
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Kod ICD10
Q22.2

Kod ICD11
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*Źródło

orphanet