

Opis choroby *

Definicja

Pulmonary atresia with ventricular septal defect (PA-VSD) is a rare cyanotic congenital heart malformation characterized by underdevelopment of the right ventricular outflow tract and atresia of the pulmonary valve, ventricular septal defect (VSD) and pulmonary collateral vessels. Clinical features depend on the anatomic variability of the lesion and patients may be minimally symptomatic, severely cyanotic or may develop congestive heart failure. PA-VSD may represent a severe form of Tetralogy of Fallot (see this term).

Dane

Klasyfikacja

Wada morfologiczna

Kod ORPHA

1207

Kod OMIM

178370

Kod ICD10

Q25.5

Kod ICD11

LA88.21

*Źródło

orphanet