

[Open Peer Review on Qeios](#)

# Pulmonary atresia with ventricular septal defect

INSERM

## Source

INSERM. (1999). *Orphanet: an online rare disease and orphan drug data base. Pulmonary atresia with ventricular septal defect. ORPHA:1207*

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).