Pulmonary arterial hypertension associated with congenital heart disease

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Pulmonary arterial hypertension associated with congenital heart disease. ORPHA:275803

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.