Opis choroby *

Definicja

A rare congenital heart malformation characterized by absence of the tricuspid valuvar annulus (absent right atrioventricular connection/junction) or an imperforate tricuspid valve leading to severe hypoplasia of right ventricle (functionally univentricular heart). The malformation is associated with normally related great arteries (70 to 80% of cases) or transposed great vessels, an obligatory interatrial connection that is crucial for survival (patent oval foramen or atrial septal defect ostium secundum type), ventricular septal defect (VSD), pulmonary outflow obstruction (pulmonary atresia, stenosis or hypoplasia), aortic coarctation and/or aortic arch interruption.

Dane

Klasyfikacja
Wada morfologiczna

Kod ORPHA
1209

Kod OMIM
605067

Kod ICD10
Q22.4

Kod ICD11
LA89.1

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*Źródło

orphanet