Opis choroby *

Definicja

A rare heterotaxia characterized by complex congenital heart malformations and abnormal lateralization of other thoracic and abdominal organs due to embryonic disruption of the left-right axis development. Cardiac defects include dextrocardia or mesocardia, common atrioventricular valve associated with complete atrioventricular septal defect or common atrium, transposition or malposition of the great arteries, and total anomalous pulmonary venous drainage, among others. Cardiac arrhythmias are frequently observed. Typical abnormalities of other organs are bilateral trilobed lungs, midline liver, and asplenia. Patients present in the newborn period with severe cardiac failure and cyanosis. Prognosis is poor.

Dane

Klasyfikacja
Zespół wad wrodzonych

Synonimy
Isomerism of right atrial appendage
Ivemark syndrome
RAI
Isomerism of right atrial appendage
Ivemark syndrome
RAI

Kod ORPHA
97548

Kod OMIM
208530

Kod ICD10
Q20.6

Kod ICD11
LA8Y

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*Źródło

orphanet