

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

[*Źródło](#)

orphanet