

# Zespół Ivemarka

**Kod Orpha: 97548 Kod OMIM: 208530**

## 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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## Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.

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