Opis choroby *

Definicja

A rare, genetic, developmental defect during embryogenesis characterized by total mirror-image transposition of both thoracic and abdominal viscera across the left-right axis of the body. Congenital abnormalities, such as primary ciliary dyskinesia, Kartagener type, polysplenia syndrome, biliary atresia, congenital heart disease, and midgut malrotation, as well as vascular anomalies (e.g. absence of retrohepatic inferior vena cava, preduodenal portal vein, aberrant hepatic arterial anatomy) and malignancy, are frequently associated.

Dane

Klasyfikacja
Wada morfologiczna

Synonimy
Complete situs inversus
Całkowite odwrócenie trzewi
Odwrócenie trzewi
Complete situs inversus viscerum
Situs inversus

Kod ORPHA
101063

Kod OMIM
-

Kod ICD10
Q89.3

Kod ICD11
LA82

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*Źródło

orphanet