

Opis choroby *

Definicja

A rare, genetic, developmental defect during embryogenesis characterized by a partial mirror-image transposition of intra-thoracic and/or intra-abdominal organs across the left-right axis of the body. Intra-organ variations and other malformations, such as ciliary motricity anomalies (e.g. Kartagener syndrome), biliary atresia and cardiac defects, are frequently associated. Left (polysplenia syndrome) or right (asplenia syndrome) isomerism are usually observed.

Dane

Klasyfikacja

Wada morfologiczna

Synonimy

Incomplete situs inversus
Częściowe odwrócenie trzewi
Niecałkowite odwrócenie trzewi
Pośrodkowe położenie trzewi
Partial situs inversus
Situs ambiguous

Kod ORPHA

157769

Kod OMIM

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Kod ICD10

Q89.3

Kod ICD11

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*Źródło

orphanet