Situs inversus totalis

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Situs inversus totalis. ORPHA:101063

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.