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# Congenital bile acid synthesis defect type 1

INSERM

## Source

INSERM. (1999). *Orphanet: an online rare disease and orphan drug data base. Congenital bile acid synthesis defect type 1. ORPHA:79301*

Congenital bile acid synthesis defect type 1 (BAS defect type 1) is the most common anomaly of bile acid synthesis (see this term) characterized by variable manifestations of progressive cholestatic liver disease, and fat malabsorption.