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# Congenital bile acid synthesis defect type 2

INSERM

## Source

INSERM. (1999). *Orphanet: an online rare disease and orphan drug data base. Congenital bile acid synthesis defect type 2. ORPHA:79303*

Congenital bile acid synthesis defect type 2 (BAS defect type 2) is an anomaly of bile acid synthesis (see this term) characterized by severe and rapidly progressive cholestatic liver disease, and malabsorption of fat and fat-soluble vitamins.