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LIVER TRASPLANT IN A PATIENT WITH SYNDROMATIC BYLE ATRESIA WTHOUT POLYSPLENIA CASE REPORT

Dr. Ishtar Cabrera Lozano//Dr. Nidia De Monserrat Arreola Gutiérrez// Dr. Antonio Murillo//Dr. José Manuel Zertuche//Dr. Verónica Paredes Escalera

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Introduction

- Biliary atresia (BA) is obliterative cholangiopathy
- Syndromic BA is associated with other congenital anomalies.
- This occurs in 10% of patients
- it´s believed that there is a relationship between BA and maternal diabetes.

Interruption of the inferior cava vein	Preduodenal portal vein	Abnormal intestinal rotation
Complete visceral transposition	Cardiac anomalies	Polysplenia

Ruíz N, Patricia, Aguirre R, Karen, Mesa M, Catalina, & Lara L, Lilian. Atresia biliar sindrómica en un paciente pediátrico: Reporte de caso. *Revista colombiana de Gastroenterología*, 2015; *30* (4), 447-455.

Schwarz KB, Haber BH, Rosenthal P, et al. Extrahepatic anomalies in infants with biliary atresia: results of a large prospective North American multicenter study. Hepatology 2013; 58:1724

Objetive

- Experience of a patient with syndromatic bile duct atresia and a related living donor liver transplant (LDLT).

Clinical report

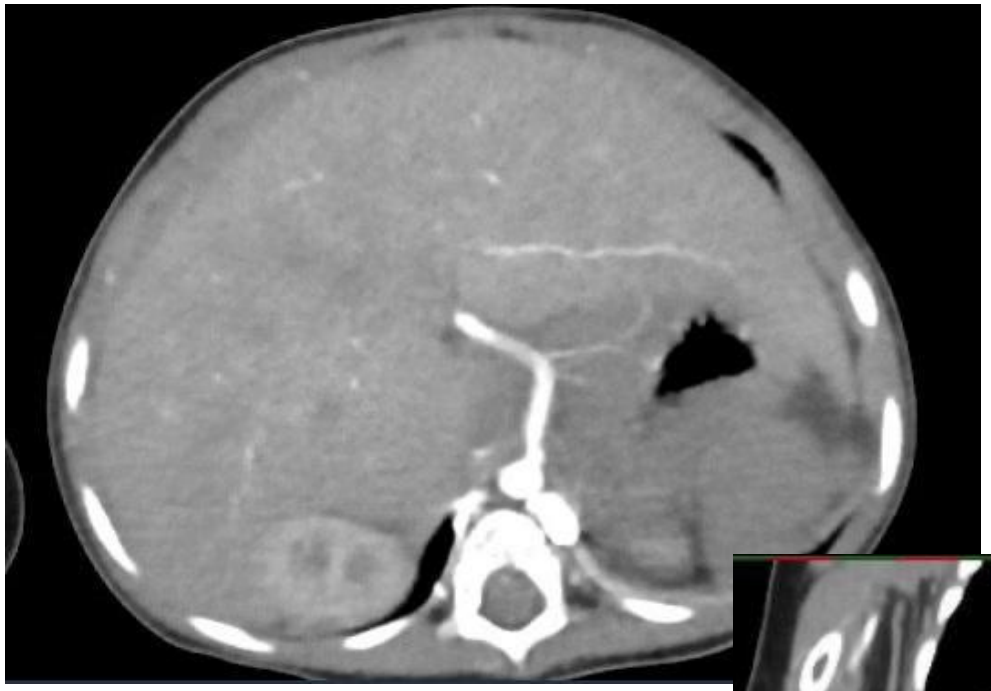
- Female 1 year 6 months old
- Maternal history of type II diabetes
- Started suffering from Cholestatic jaudince at 15 days of life
- Diagnosis of type III BA
- Plus another clinical features:

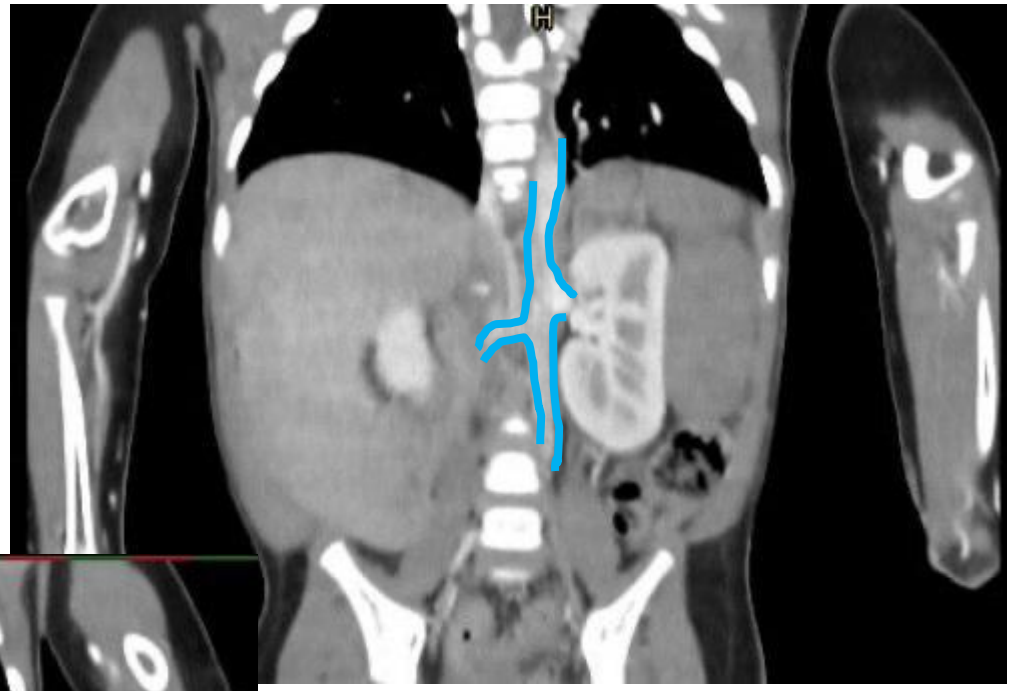
- ❑ Malrotation
- ❑ Pre-duodenal portal vein
- ❑Cardiac anomalie Intraventricular septal defect
- ❑shunt from left to right with a gradient of 60 mm without hemodynamic repercussion
- ❑Foramen ovale of 2 mm
- ❑Dyslipidemia

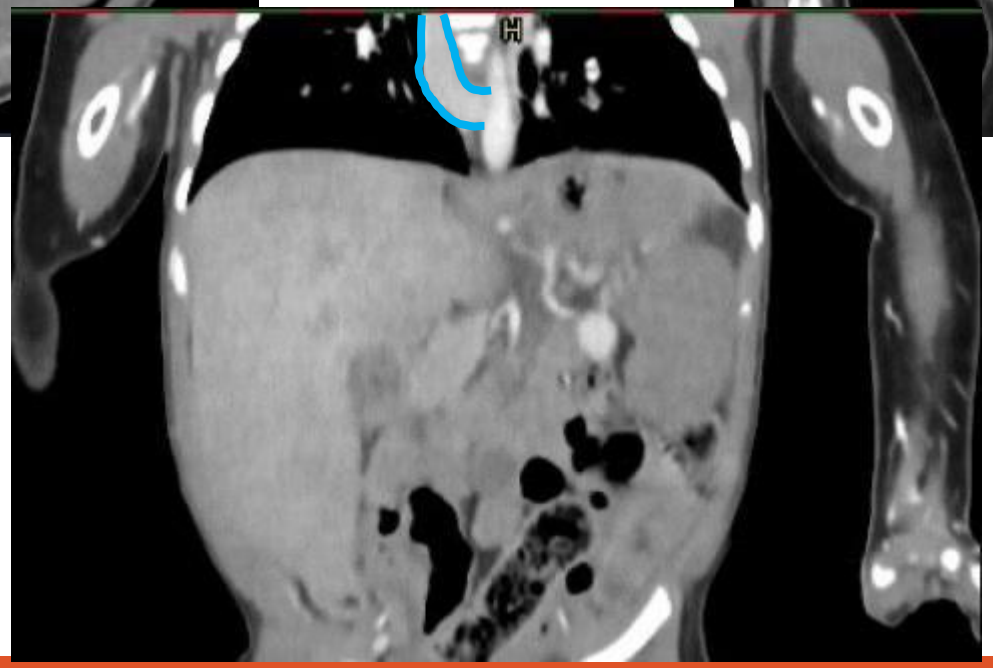
Evolution

- She did not have bile clearance
- With little growth with a weight of 7.5 kg
- Liver failure with a CHILD – PUGH B with 7 points and PELD 24 points
- It was decided to perform an LDLT.
- Appendectomy and placement of a biodegradable biliary stent in the roux.
- Presented partial thrombosis of the portal vein in hepatic segment II.

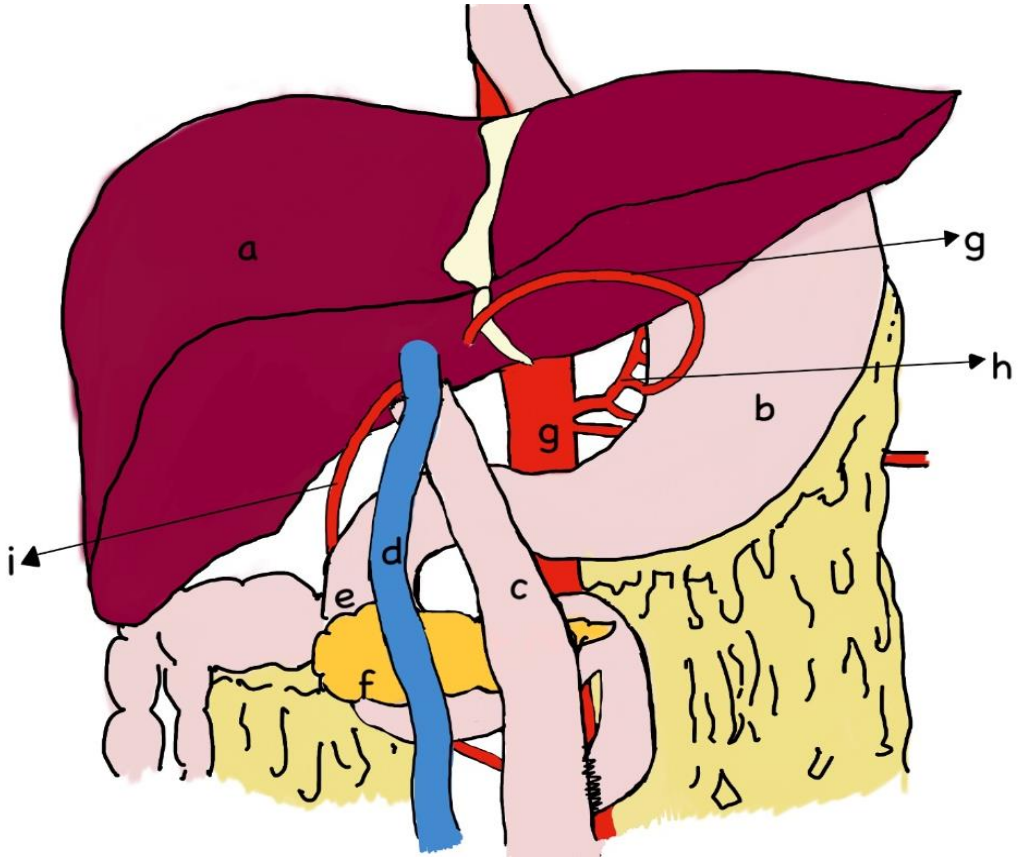
Angiotomography







Trasplant surgical findings



Liver anatomy of syndromatic biliary atresia without polysplenia.

a) Liver
b) stomach
c) jejunum
d) preduodenal portal vein
e) Duodenum
f) annular pancreas
g) left hepatic artery emerging from left gastric artery
h) left gastric artery
i) i) right hepatic artery emerging abdominal aorta.

Conclusion

The anatomical variants of a syndromatic atresia require great knowledge and vascular control so as not to lose the only possibility of arterial and portal anastomosis that the patient has, in this case the total release of the portal vein without injuring the pancreas or the duodenum represented a challenge.