

Pediatric liver transplantation in Situs inversus and other vascular abnormalities

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No disclosures

Biliary atresia polysplenia syndrome

Some cases of biliary atresia can be subcategorized as syndromic which means that they coincide with malformations such as situs inversus, isolated dextrocardia and or vena cava aplasia with continuity of vena azygos or hemiazygos. This syndrome is also known as biliary atresia polysplenia syndrome.

We here report of three cases with biliary atresia and situs inversus for whom we performed liver transplantation. All of them had preduodenal portal vein and polysplenia, one had vena cava aplasia

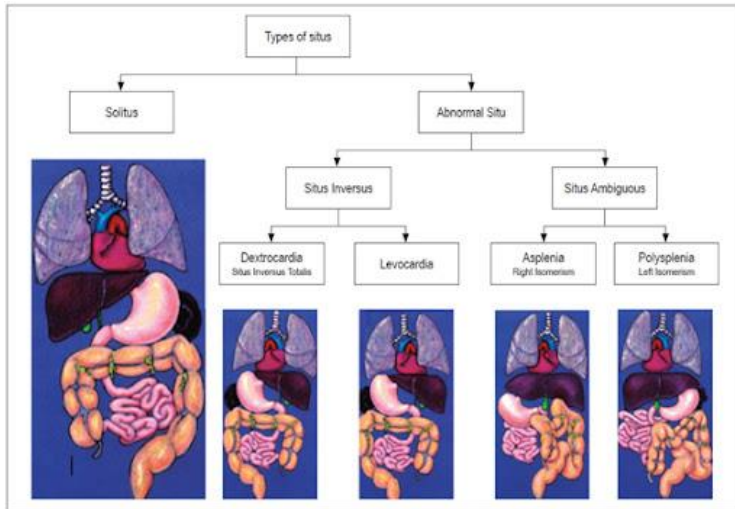


Figura 3. Types of situs. Source: Abdominal Manifestations of Situs Ambiguus abnormalities in adults based on four cases. Chilean Review of Radiology, 2013

Patient	Situs inversus	Polysplenia	Predudodenal portal vein	Cava aplasia
6mo m	✓	✓	✓	
8mo f	✓	✓	✓	
8y	✓	✓	✓	✓

Results

Patient 1:

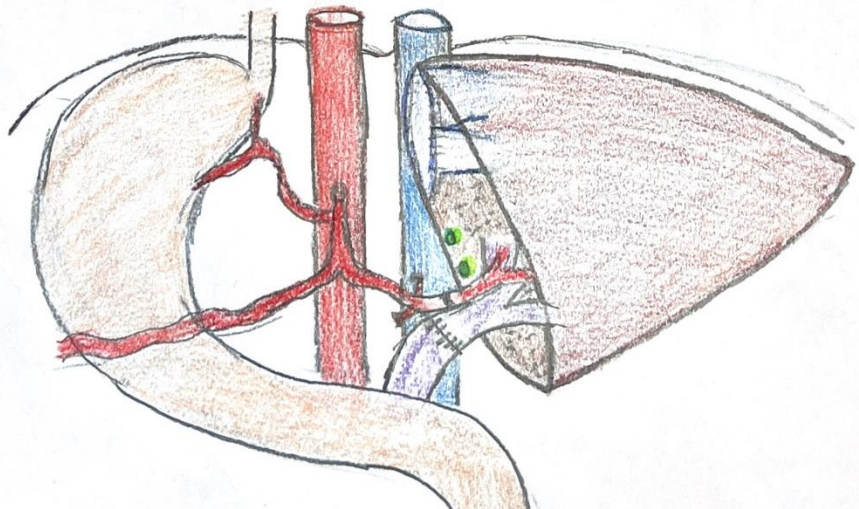
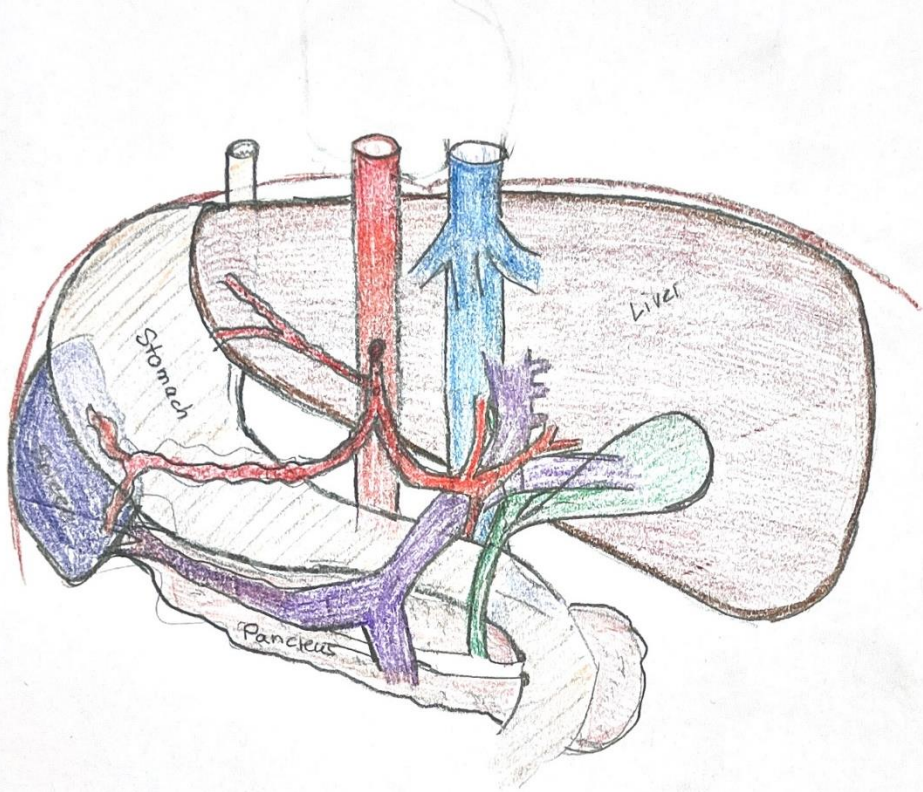
LLS from father. No complications. Discharged d21.

Patient 2:

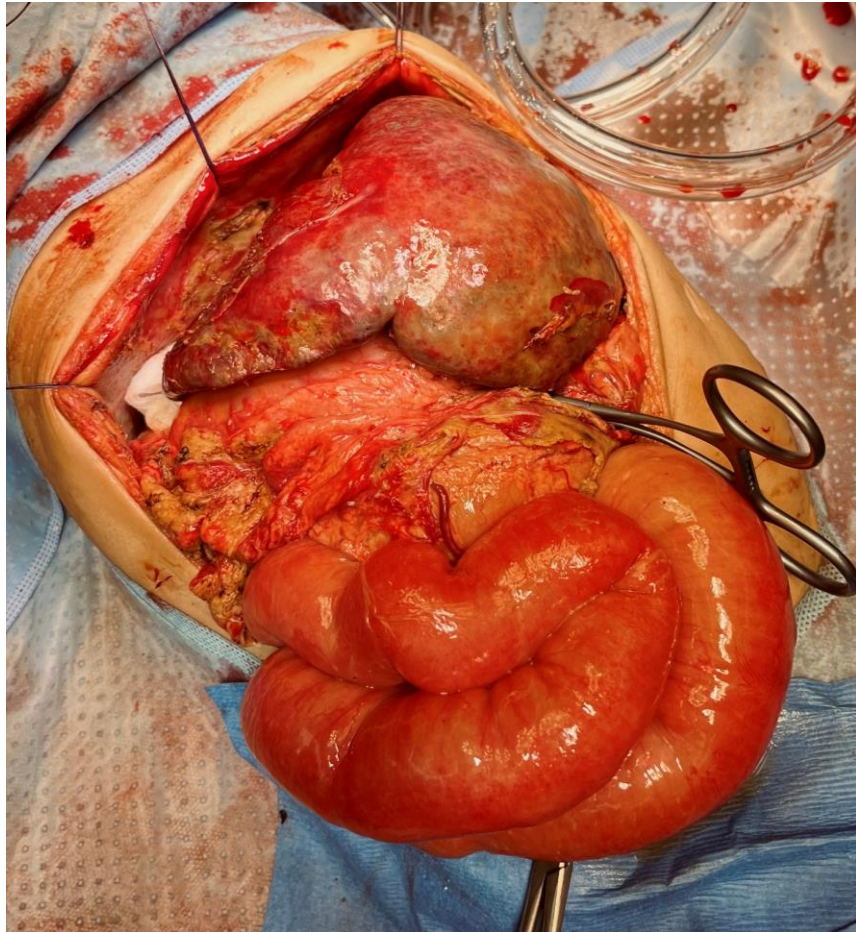
LLS from mother. D12 bile leak and consecutive arterial thrombosis. Re-transplant with jump graft to Aorta

Patient 3

LLS from in situ split with 2 veins. Segment III vein had outflow problems. Re-Transplant with venous outflow modification.

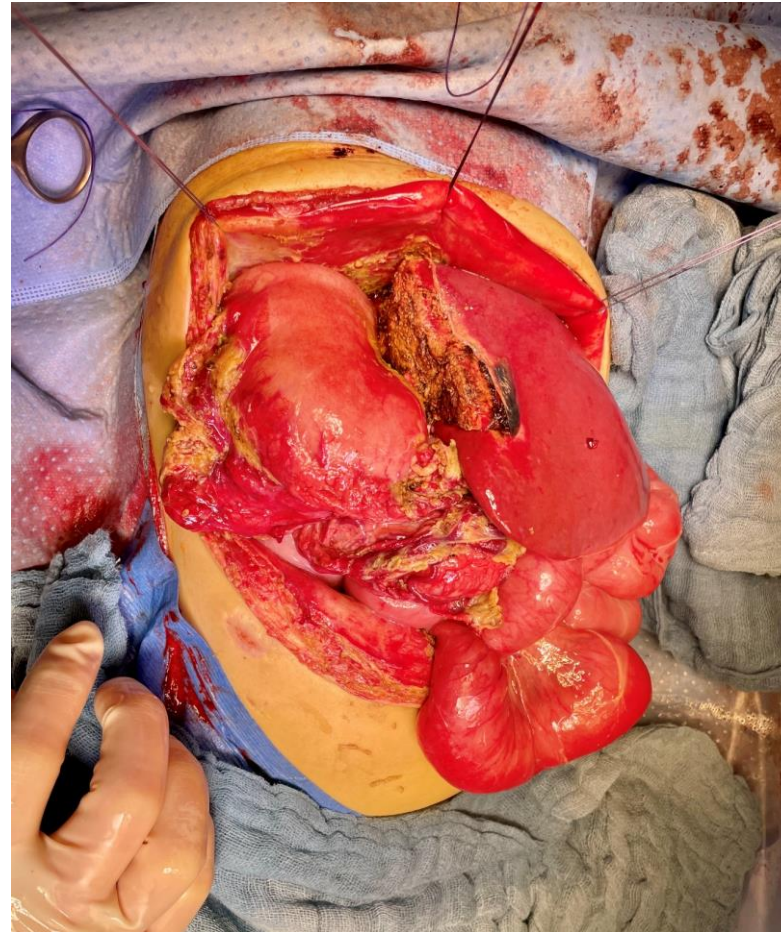


Patient No 2.

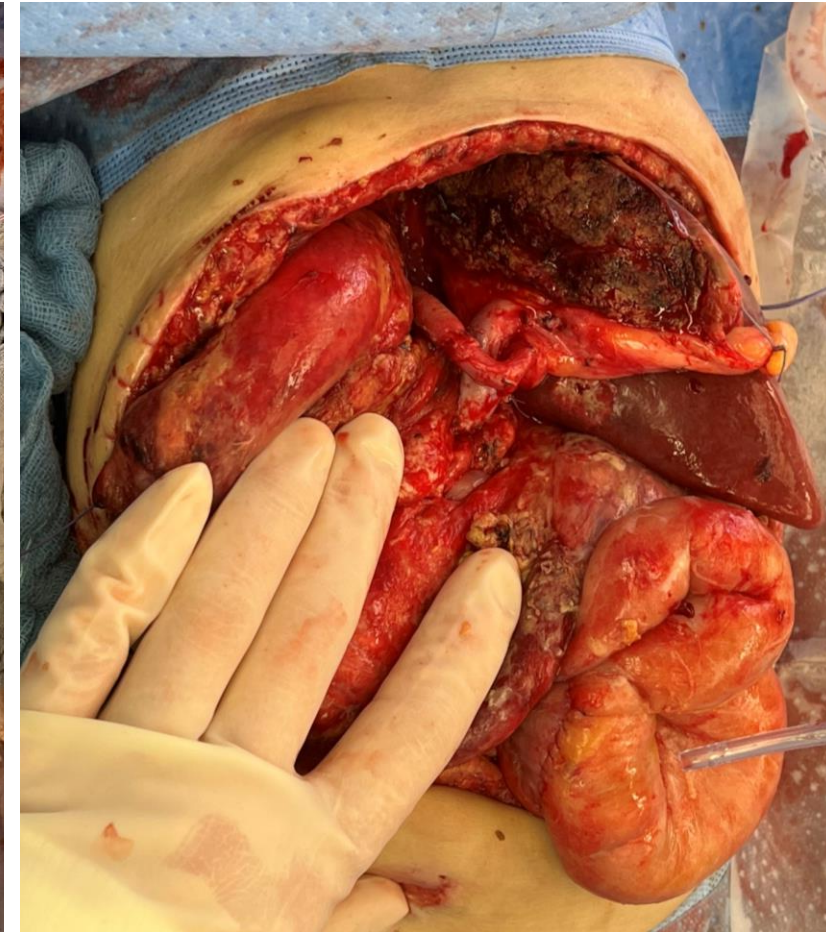


Operative situs

Bildnachweis: Istockphoto

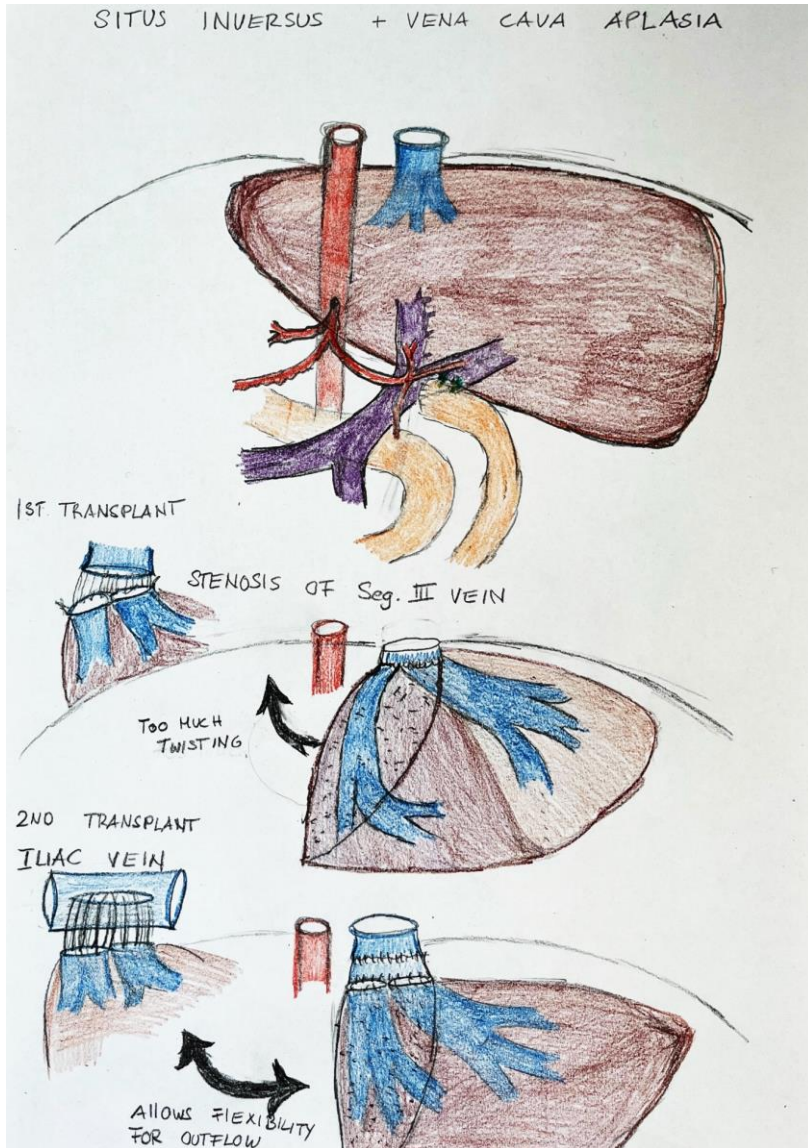


1st graft LLS from mother

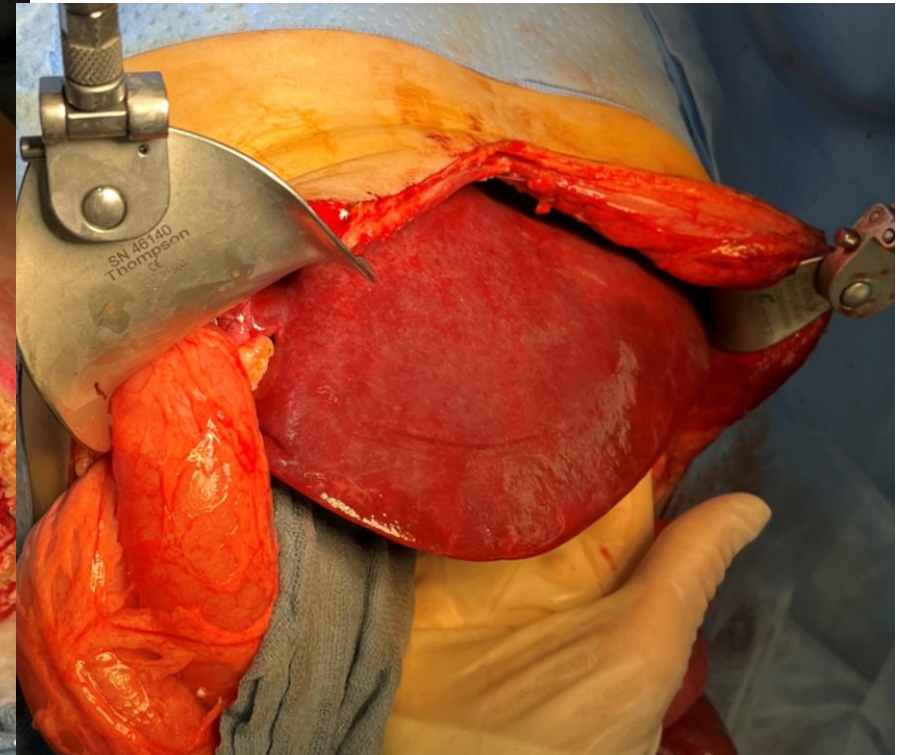


2nd graft with jump graft to aorta

Patient No 3.



LLS from in situ split



congestion of segment III

Conclusions

- Liver transplantation in children with syndromatic biliary atresia associated with Situs inversus and other vascular malformations can be technically challenging
- Preduodenal portal veins are often very thin
- Arterial reconstruction in situs inversus needs to cross the portal vein (anterior or dorsal)
- In vena cava aplasia venous reconstruction of a LLS vein should be considered
- Full grafts should be preferred for cava aplasia
- Vessels for reconstruction should be available