No gut syndrome in paediatric patients

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Introduction/Background

No gut syndrome or near total enterectomy (NTE) is defined as **the removal of the entire jejunum and ileum**, a rarely performed procedure, indicated in very specific situations.

This condition is associated with parenteral nutrition (PN) dependency, multiple complications and poor quality of life in general. To date, only few cases have been reported in literature, all in the adult setting.

Aim

The aim of this report is to raise awareness of this condition in paediatric patients and the difficulties encountered in their management.

Subjects and methods

Two male patients (2 and 5 year old) are presented. One secondary to an ischaemic event of unknown cause and the second post a failed small bowel transplant, both suffering complications specific to this condition.

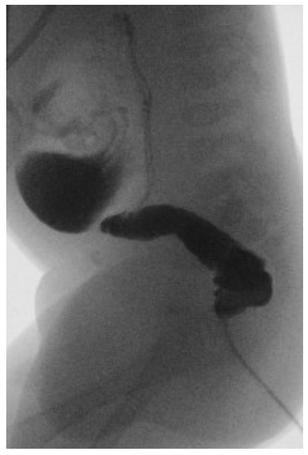
Summary and conclusion

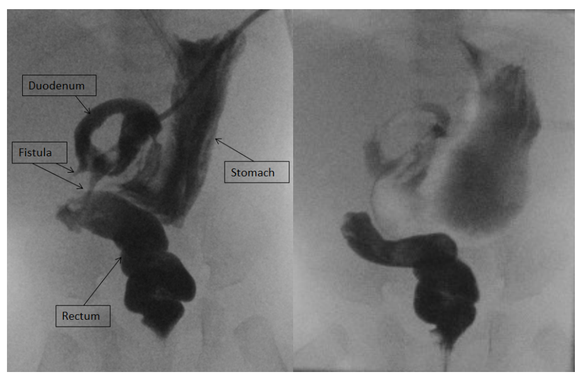
- **Advances in PN have made possible the long-term survival of patients with no gut syndrome in specialized centres.**
- **Complications, patient and family quality of life, use of health resources, medical and surgical challenges and overall outcomes are important aspects to be considered and more paediatric data is necessary.**

Results

Case 1: 2 years old boy

- First-born to non-consanguineous parents at 37 weeks of gestation.
- Other pathologies: 22q11 deletion and large VSD, frequent respiratory infections.
- 2.5 months of age: intestinal infarction from duodenum to sigmoid colon -> D1-D2 (D2 showed patches of ischemia). Duodenal tube in the duodenal stump and started on PN.





- 3.5 months of age: 2 fistulas between the duodenum and the rectum that resolved spontaneously.

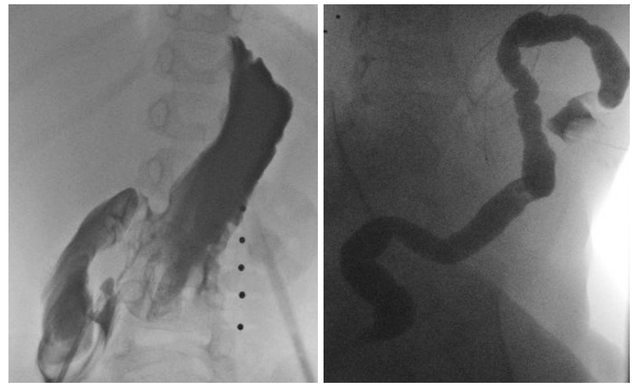
Currently:

- Total PN dependent, only some water orally (thicker consistency can block the tube).
- Balloon G/J tube pushed into the stomach due to a narrowed duodenal stump to try and minimise vomiting.
- Issues with leakage around the tube.
- Intermittently abnormal liver function test.
- Liver US: hepatomegaly and gallstones.

No conflict of interest to declare

Case 2: 5 years old boy

- Third child to non-consanguineous parents born at 35 weeks of gestation.
- Initial diagnosis: complex gastroschisis, jejunal atresia and hypoplastic colon -> remaining anatomy 15 cm from GJ junction.
- Initial treatment: placement/removal of a silo followed by bowel expansion, formation of a jejunostomy and a mucous fistula closed a year later when he had a jejunocolonic anastomosis -> unable to tolerate minimal enteral amounts with a high stoma output despite treatment with octreotide.
- 4 years of age: isolated bowel transplant followed by severe rejection not responsive to ATG/Campath and complicated with PTLD.
- 2 months later: graft enterectomy with remaining anatomy 5 cm of duodenum and leakage from the duodenal stump; large bowel length 35 cm, maximum diameter 1.5 cm.



Currently:

- Mucus fistula, abdominal drain and gastrostomy.
- Type 1 Chiari malformation incidentally found and recent onset seizures.
- Intestinal failure associated liver disease and renal stones.
- Frequent admissions due to his health needs and frequent complications.