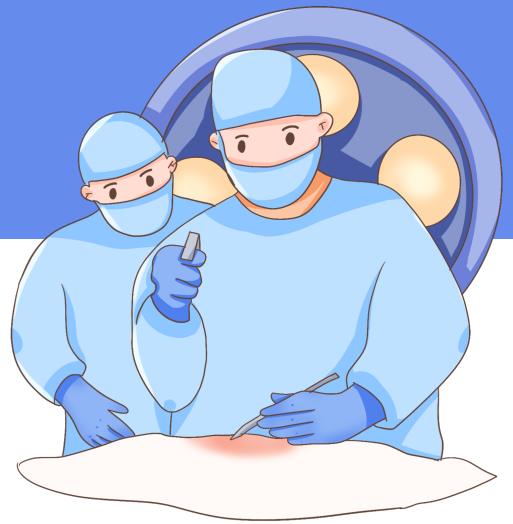
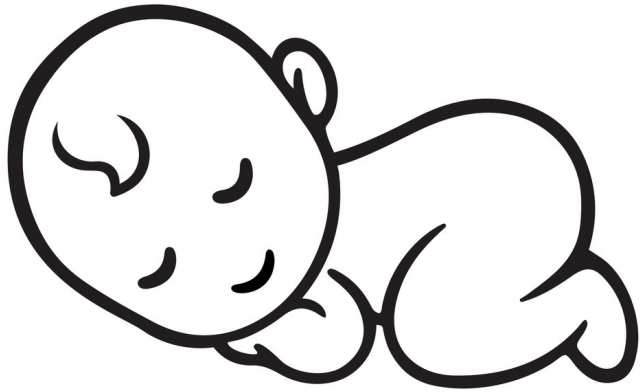
BELAPS session



A rare case of a newborn male presenting the association of Hirschprung's disease (HD) and Eosinophilic myenteric plexitis (PME)

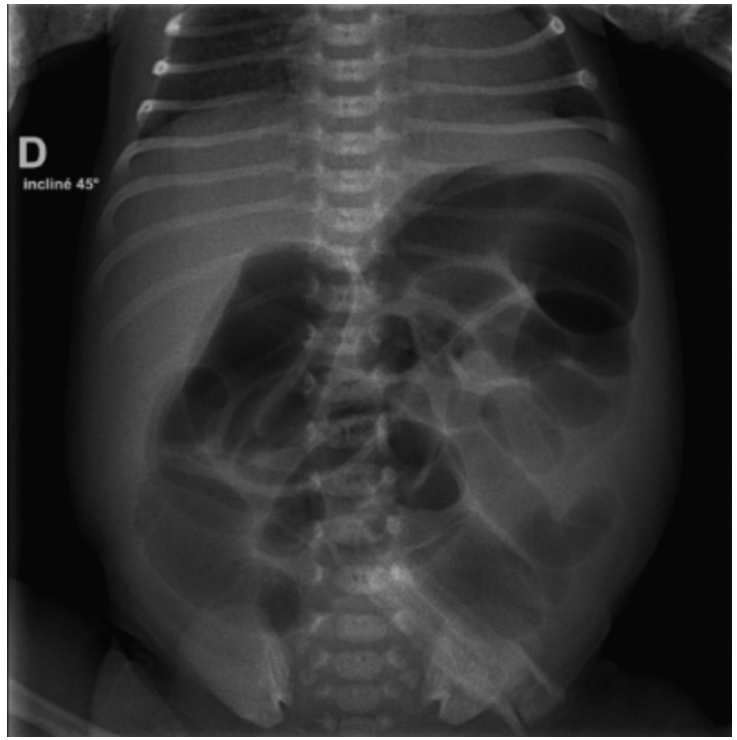


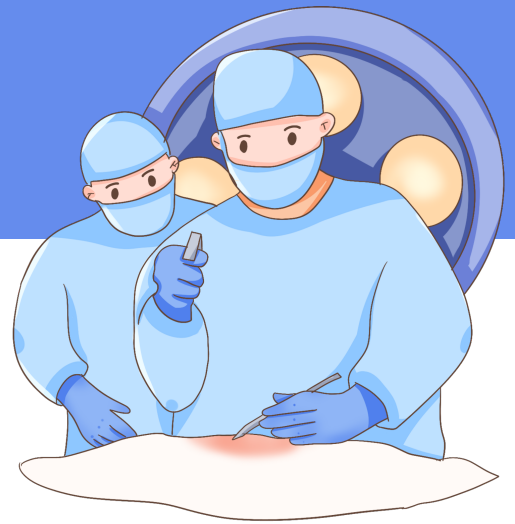
Belgian Surgical Week 2022 - Oostende

Shayan Madani , Emir Teftedarija, Emeline Bequet, Katty Delbecque, Martine Demarche

Case report

- Male newborn vaginal delivery 37 weeks 4 days amenorrhea
- Delayed meconium emission for 3 days
- Hydroaeric levels abdominal radiography
- Transfered in our institution for suspicion of **Hirschprung's disease (HD)**



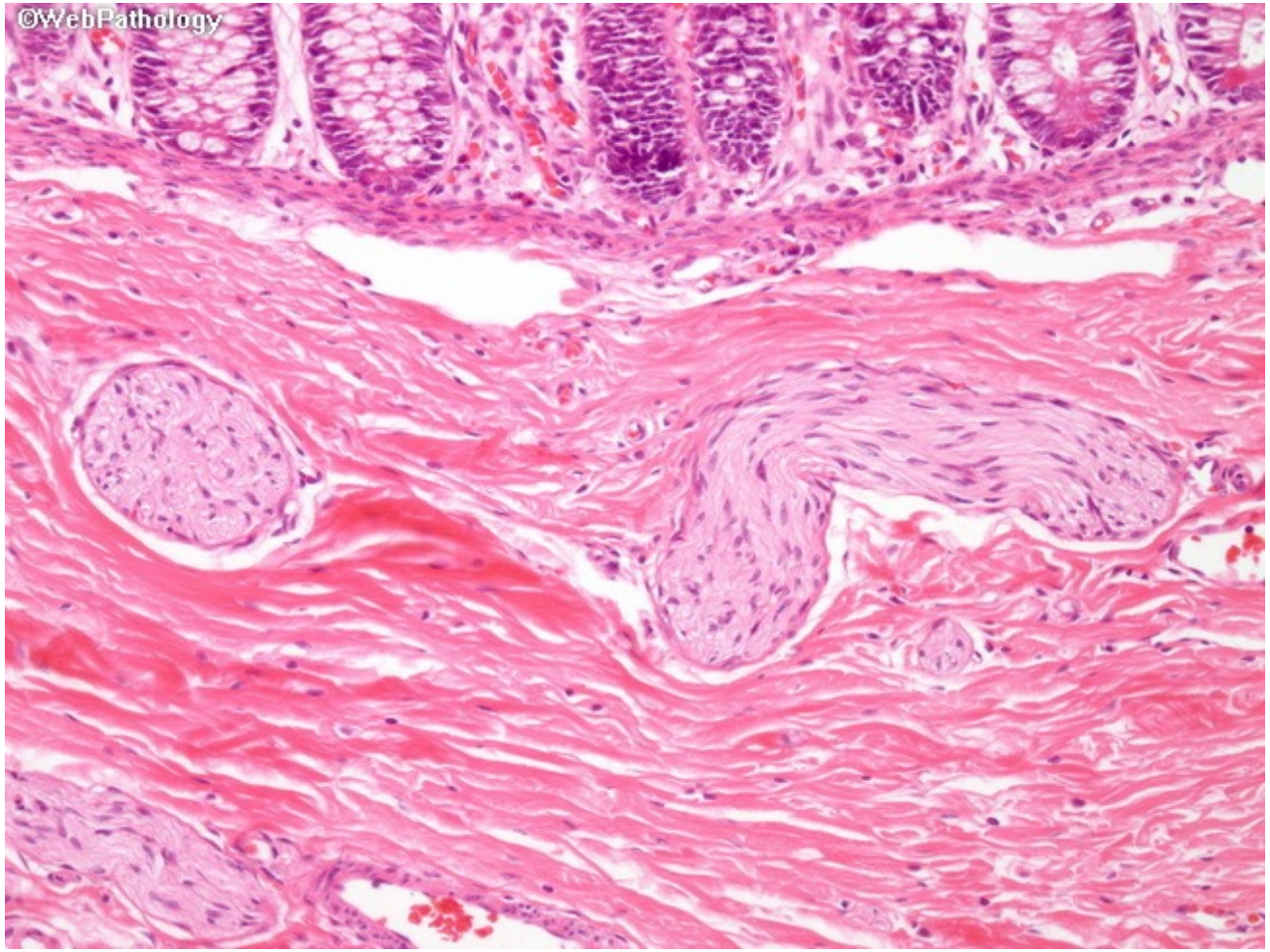


- Abdominal US : colonic dilatation > small bowel dilatation

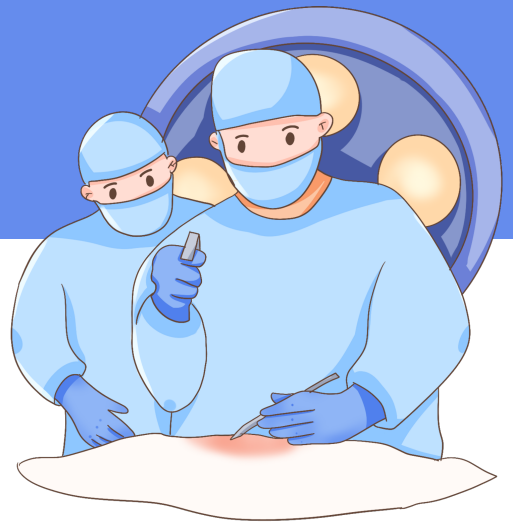
  No (in)direct signs of digestive ischemia

- Likely diagnosis : distal HD

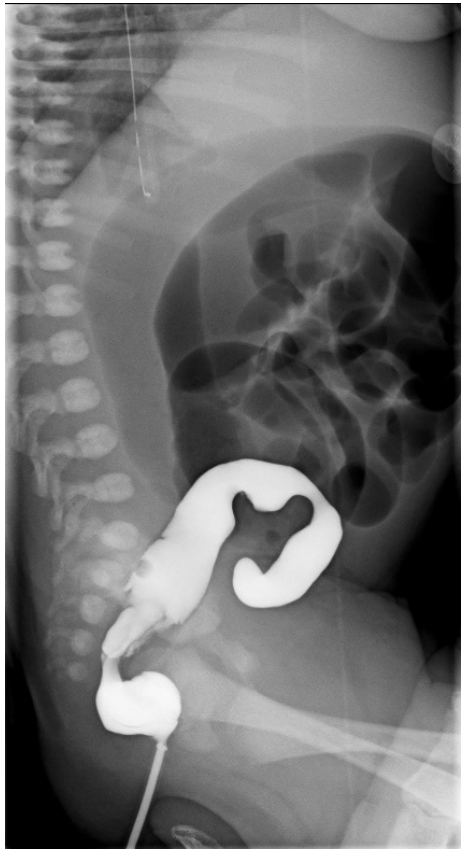
- **Rectal biopsy (next day)** : absence of ganglion cells in submucosal (Meissner's) and muscularis (Auberbach's) plexus + mild chronic colitis

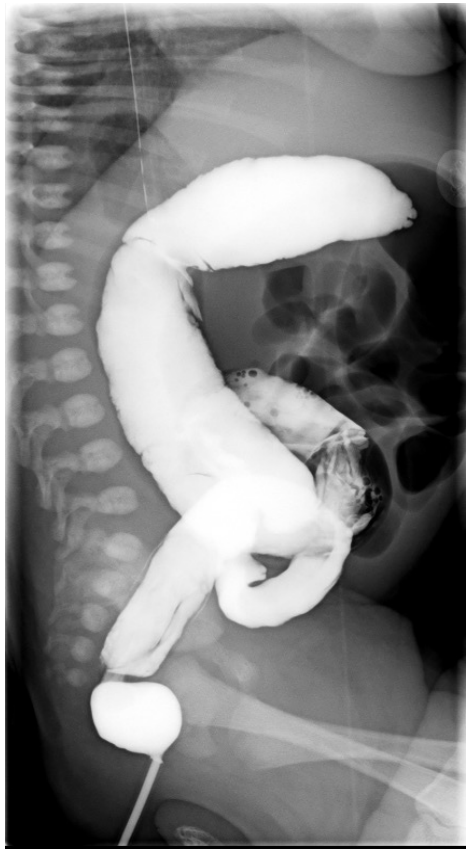


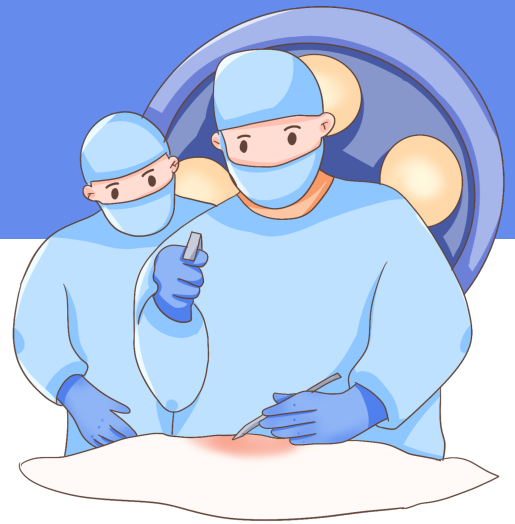
https://www.webpathology.com/image.asp?n=7&Case=224



- Lower GI tract radiography (barium enema)

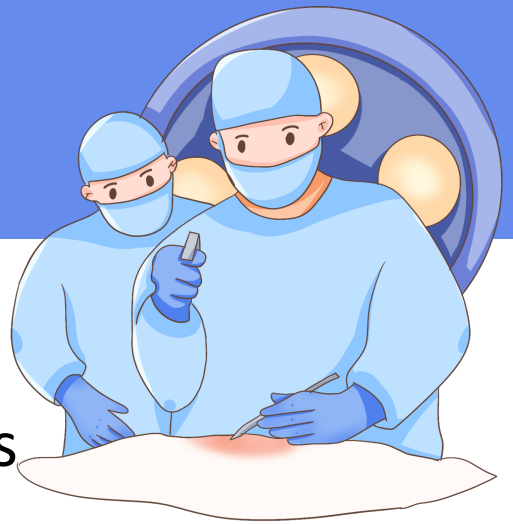






- Treatment
  - Day 11 : coloproctectomy with colo-anal anastomosis
    - Rectum + ¾ of the distal colon
    - Frozen sections : transition zone

Brief theoretical reminder



- HD = congenital disorder absence of ganglion cells in Meissner's (submucosal) and Auerbach's (muscularis) plexus of distal rectum → variable distance proximally
- 1 – 1,63/10.000 births → 4/1 male predominance
- Multigenic inheritance (weak penetrance)
- Diagnosis = combination of clinical, radiological and **histopathological** findings
- ***Management = surgical*** (Soave procedure)

Brief theoretical reminder

- Myenteric plexitis : abnormal inflammation of Auerbach's plexus (lymphocytes > eosinophils) → can be associated with :
  - Paraneoplasic syndromes
  - Infections
  - Allergies or auto-immune disease
  - Inflammatory bowel disease
  - ***Chronic intestinal pseudo-obstructions (CIPO)***
- ***Management = medical*** *(ant-inflammatory, immunosuppressors)*

CONCLUSIONS

- Differential diagnosis and treatment of HD and EMP is a challenge
- Clinical presentation can be similar when EMP is encountered within CIPO syndromes
- In the case of concomittant HD and EMP, post-operative signs of pseudo-obstruction can remain and if not recognized, could lead to diagnostic wandering
- Good anatomopathologist is the key in this case