



IMAGE OF THE MONTH

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FROM YOUNG ESPGHAN

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Morbus Hirschsprung

A four-year-old boy presented with chronic constipation since birth suffering from recurring paradoxical diarrhea over the last year having 3-15 creamy, non-bloody stools a day. A colonoscopy three months prior was normal and ruled out IBD. Laboratory studies on admission were unremarkable. Abdominal ultrasound revealed dilatation of intestinal loops up to ~5cm with wall thickening up to ~6mm. The Colon appeared unsuspecting. MRI-Sellink imaging was performed demonstrating a substantially distended ileum from the ileocecal valve upwards, whereas the colon appeared small (see Figure 1). Since the mother reported that Hirschsprung's disease had been ruled out at the age of 8 months, a diagnostic laparoscopy was performed, which demonstrated an erythematous terminal ileum. 3cm of ileum were resected and a double-barrel ileostomy was created. Enzyme and immune histochemical assays revealed a complete aganglionic colon leading to the diagnosis of Hirschsprung's disease. Retrospectively, the mother revealed that the exclusion of Hirschsprung's disease at the age of 8 months was based on the absence of a dilated colon alone.

Hirschsprung's disease is an important differential diagnosis to chronic constipation, even in toddlers and pre-school children. Diagnosis can only be made by immune histochemical staining of rectum/colon biopsies.

Learn more about the medical management of chronic constipation in the ESPGHAN/NASPGHAN guidelines: www.naspgan.org/files/documents/pdfs/cme/jpgn/Evaluation_and_Treatment_of_Functional.24.pdf

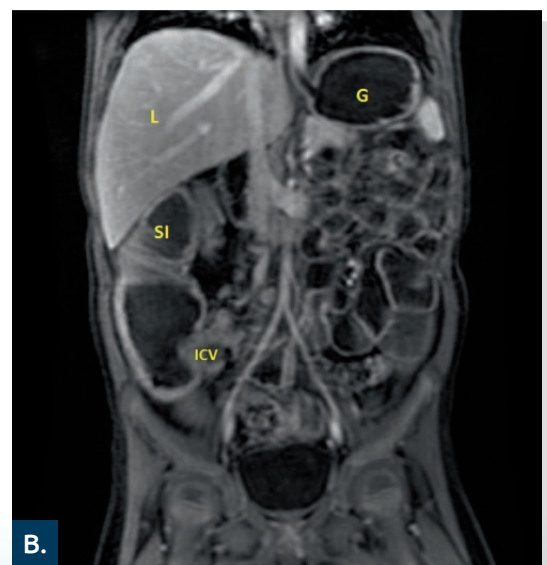


Figure 1 MRI Sellink

A. Dilated intestinal loops (L) liver, (G) gaster, (C) colon, (SI) small intestine

B. Slim colon before and dilated ileum after the ileocecal valve (L) liver, (G) gaster, (SI) small intestine, (ICV) ileocecal valve