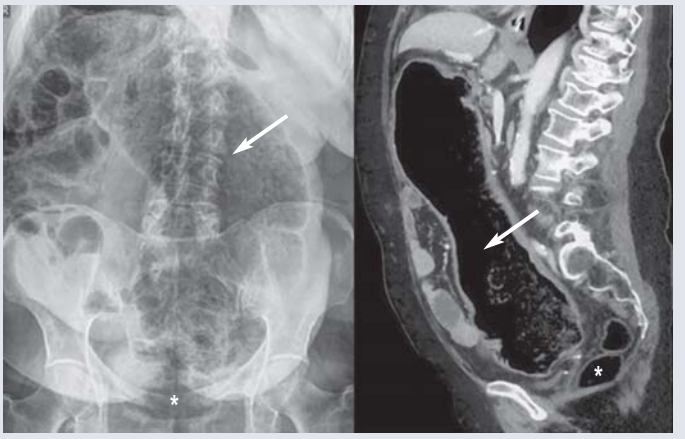
CLINICAL SNAPSHOT

Aganglionic Megacolon (Hirschsprung's Disease)—61 Years to Diagnosis



Abdominal radiography (left) and sagittal CT (right) (kindly provided by Prof. Dr. Hans-Jürgen Raatschen, Radiology Department, Helios Hospitals Schwerin) in a 62-year-old woman with aganglionic megacolon (Hirschsprung disease) and status post left hemicolectomy in infancy show enlargement of the residual colon to over 10 cm (arrow) and a typically narrow, deep rectum (asterisk).

A 62-year-old woman presented to the emergency department with painful stool retention. Because of chronic constipation since childhood, she regularly used macrogol and enemas to induce defecation. She had undergone hemicolectomy for "intestinal obstruction" in infancy and several laparotomies with adhesiolysis in adulthood. Radiological imaging showed a narrow rectum and a massively enlarged residual colon *(Figure)*. A full-thickness wall sample was obtained by endoscopy and aganglionosis was found (HE staining). Histological diagnosis was confirmed by a reference pathologist. Given the impairment of the patient's quality of life caused by the constipation, and with her agreement, we performed subtotal colectomy. Due to anal sphincter insufficiency, an ileostoma had to be created rather than the intended ileal pouch. Segmental aganglionosis (congenital Hirschsprung's disease: incidence 1:5000; men:women 4:1) was again diagnosed in the colorectal surgical specimen. The currently valid guidelines for pediatric Hirschsprung's disease recommend removal of the aganglionic rectum (with or without the sigmoid colon) followed by anastomosis of the colon to the anus (pull-through surgery).

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Conflict of interest statement: The authors declare that no conflict of interest exists.

Translated from the original German by David Roseveare.

Cite this as: Schmitz D, Ritz JP, Wöhlke M: Aganglionic megacolon (Hirschsprung disease)—61 years to diagnosis. Dtsch Arztebl Int 2024; 121: 344. DOI: 10.3238/arztebl.m2023.0103