

# AB153. SOH21AS115. Double check—a technical note on the management of a case of adult diagnosis Hirschsprung disease

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**Background:** Hirschsprung disease (HSCR) is a motility disorder of the gut usually diagnosed in infancy, caused by failed migration of neural crest cells during fetal development. It is characterised by a non-functional, aganglionic segment of colon which most commonly presents as a large bowel obstruction, but can have variable features which must be considered with adult presentations of chronic constipation.

**Methods:** We present the case of a 49-year-old male with large bowel obstruction, on a background of a left hemicolectomy in childhood for the same presentation. There was no preceding clinical or histological diagnosis of HSCR. On this occasion in adulthood, his whole colon was atonic from chronic constipation, with muscular hypertrophy and fibrosis. A full thickness rectal biopsy finally confirmed HSCR. His subsequent clinical course included formation of a defunctioning left sided colostomy to relieve his bowel obstruction. This ensured histological confirmation of ganglia in his left colon, and adequate colonic function via the colostomy, as an assessment of the proximal conduit for his planned delayed anastomosis. After preoperative counselling, he then underwent elective ultralow anterior resection, coloanal anastomosis and loop ileostomy with subsequent reversal.

**Results:** The patient made a full recovery and no longer experiences abdominal pain. His bowel movements

completely normalized.

**Conclusions:** This highlights an unusual approach to managing HSCR presenting in adulthood, taking into account the variabilities in aganglionosis which can occur, and the chronic effects of subacute obstruction on bowel function. These must be considered for any surgical approach, and our staged approach ultimately ensured an optimal functional outcome for the patient.

**Keywords:** Aganglionosis; constipation; Hirschsprung disease (HSCR); motility disorder; obstruction

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## Footnote

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

*Ethical Statement:* The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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