

PROCTOCOLECTOMY WITH ILEOANAL ANASTOMOSIS IN THE TREATMENT OF EXTENDED AGANGLIONOSIS IN HIRSCHSPRUNG DISEASE

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Aim: We assessed complications and functional outcomes of restorative proctocolectomy with ileoanal anastomosis (IAA) performed on children with Hirschsprung disease (HD) in relation to patients with ulcerative colitis.

Methods: Medical records of all HD patients (n=16) who underwent proctocolectomy with IAA during 1997-2015 in a single center were retrospectively reviewed. Data on complications, stool frequency, day- and nighttime continence, enterocolitis/pouchitis and fecal calprotectin levels were collected and compared to the patients, who underwent IAA for pediatric-onset ulcerative colitis by the same surgeons.

Results: Median age of HD patients was 1.6 (IQR, 0.8-11.5) months at IAA and 5.6 (IQR 2.5-12) years at follow-up. Fourteen patients received J-pouch with median length of 4.0 (range 3.0-10) cm and two straight IAA. Diverting ileostomy was closed after median 11 (4.3-33) months. Three patients were re-operated due to complications: ileostoma prolapse, fascial rupture and adhesive obstruction, while no case of anastomotic leakage occurred.

As shown in table, daytime and nighttime stool frequency was 3.5 and 0, respectively. Two children below three years wore diapers. Of the rest, one patient aged 2.3 years used diapers during nights, while 11/14 (79%) were continent without fecal soiling or accidents. At least two treated (antibiotics and/or intersphincteric botulinum) enterocolitis/pouchitis episodes occurred in 13/16 (81%) HD patients, while histologically verified pouch inflammation was observed in only 4 (27%). Median fecal calprotectin was in normal range, while increased (>100 µg/g) value was observed in four.

Comparison of different parameters of stool frequency, fecal continence and enterocolitis/pouchitis between HD and UC patients is shown in table.

Conclusion: Proctocolectomy with IAA provides encouraging bowel functional outcomes in TCA. In relation to pediatric onset UC, stool frequency and fecal continence appeared better preserved. Although episodes of enterocolitis were common, frequency of histological pouch inflammation and fecal calprotectin levels were lower than in UC.

Variable	HD (n=16)	UC (n=32)
Follow-up age (y)	5.6 (2.5-12)	24.1
Stool frequency/day	3.5 (2.3-4.0)	5.8
Stool frequency/night	0 (0-1.75)	1.5
Fecal soiling (%)*	21%	22%
Totally continent (%)*	79%	44%
Enterocolitis/pouchitis (%)	81%	69%
>two episodes (%)	81%	22%
Histological pouch inflammation (%)	27% (n=15)	60%
Fecal calprotectin (µg/g)	68 (20-286) (n=10)	340

Table. Data are median/mean (IQR) or frequencies. *Patients weaned from daytime diapers included (n=14). HD=Hirschsprung disease, UC=ulcerative colitis.