

# SURGICAL STRATEGIES AND OPTIMAL MANAGEMENT OF ADULT HIRSCHSPRUNG’S DISEASE

Lory Hage<sup>1,2</sup>, Remy Amory<sup>1</sup>, Gianluca Pagano<sup>1</sup>, Rodrigue Chemaly<sup>3</sup>, Roy Ibrahim<sup>4</sup>, Daniel Mahfoud<sup>4</sup>, Gilles Manceau<sup>1</sup>, Sabine Irtan<sup>5</sup>, Georges Al Hajj<sup>3</sup>

(1) Department of Digestive Surgery, Georges Pompidou European Hospital, Assistance Publique-Hôpitaux de Paris, Paris, France; (2) Department of Paediatric Surgery, Assistance Publique-Hôpitaux de Paris, Université Paris Cité, Paris, France; (3) Department of General Surgery, Lebanese American University Medical Center-Rizk Hospital, Beirut Lebanon; (4) Department of Medical Imaging, Lebanese American University Medical Center-Rizk Hospital, Beirut Lebanon; (5) Department of Paediatric Surgery, Trousseau Hospital, Assistance Publique-Hôpitaux de Paris, Paris, France

## INTRODUCTION

- **Adult Hirschsprung's disease** (HD), defined as diagnosis **after the age of ten**, is a rare congenital disorder often misdiagnosed as refractory constipation.
- **Diagnostic delay** leads to emergency presentation and suboptimal management
- **Challenges** due to extensive colonic dilation, risk of anastomotic complications, and need for **personalized treatment**
- Paediatric **pull-through techniques** may be adaptable to adult anatomy
- Evidence guiding **optimal surgical strategy** in adults remain **limited**

The **aim of this study** is to evaluate surgical strategies and outcomes in adult HD through systematic review

## METHODS

### Systematic Review

- In accordance with PRISMA guidelines (Fig.2)
  - Pubmed/MEDLINE library
- Inclusion**
- Case reports or case series
  - > 10 years old
  - Histologically confirmed HD (Fig.1)

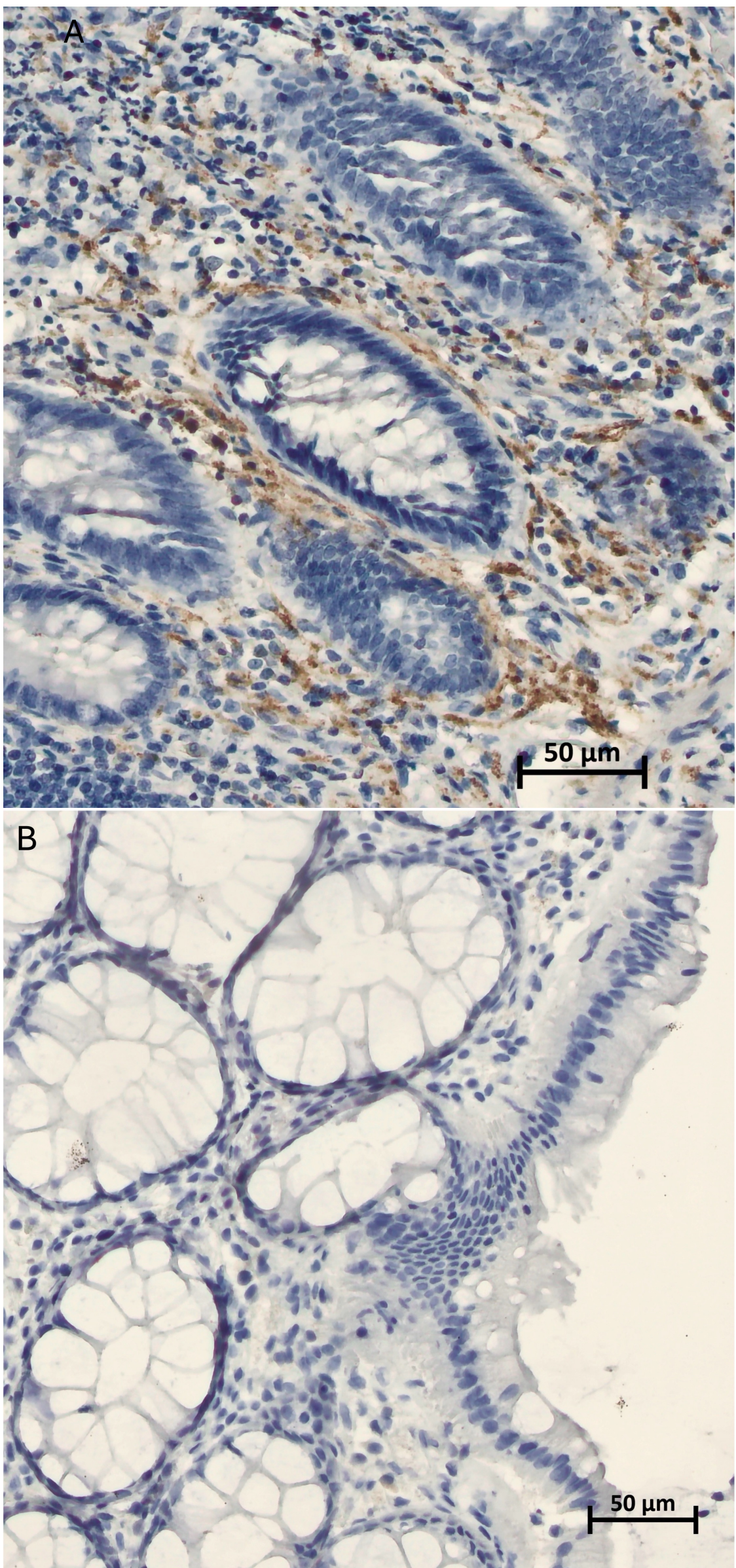
### Data collection

- Demographics, medical history
- Clinical presentation, initial diagnostic workup
- Management of the disease and follow-up

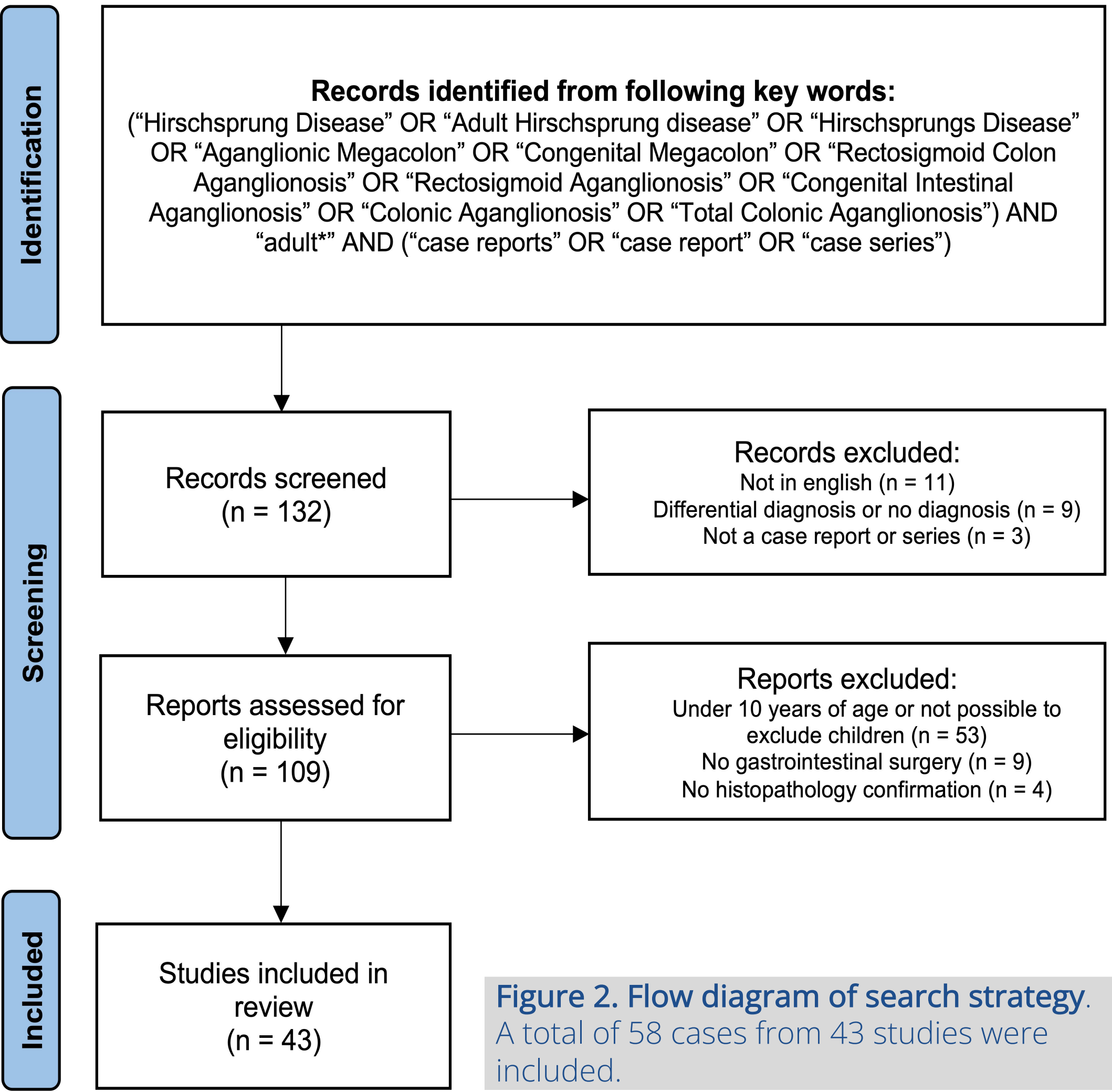
### Statistical Analysis

- Performed on Python version 3.10.16
- T-test, ANOVA, and Mann-Whitney T tests were used for continuous data
- Chi-squared or Fisher's tests were used for discrete data

**Figure 1.** Immunohistochemistry staining of Calretinin performed on a control case (A) and the rectal biopsies of the patient (B). Absence of Calretinin expression in the rectal mucosa confirmed aganglionosis



## RESULTS



### Clinical Presentation

- Mean age of 28.7±16.2 years; Male predominance of 72.7%
- Chronic constipation since childhood: 62.1%

### Disease Distribution

- Terminal rectum (type 2, 27.6%) and rectosigmoid (type 1, 27.6%) most frequent sites

### Diagnostic Work-up

- Imaging used in 87.9%; mostly CT-scan (44.8%)
- Preoperative rectal biopsy rare: ≤ 5%

### Surgical Management (Table 1)

- Commonly open approach (84.5%)
- Emergency surgery (69.0%); 67.2% of diverting stoma

→ Pull-through procedures were more frequent in younger patients (p = 0.001)

| Type of surgical intervention        | Duhamel (n = 14) | Soave (n = 2) | Swenson (n = 7) | Resection w/ anas. (n = 16) | Resection w/o anas. (n = 14) | p      | Open (n = 49) | Lap. (n = 8) | p    |
|--------------------------------------|------------------|---------------|-----------------|-----------------------------|------------------------------|--------|---------------|--------------|------|
| Intraoperative adverse events, n (%) | 2 (33.3%)        | 0 (0.0%)      | 0 (0.0%)        | 2 (33.3%)                   | 1 (16.7%)                    | 0.77   | 5 (83.3%)     | 1 (16.7%)    | 1    |
| Terminal Ostomy, n (%)               | 0 (0.0%)         | 0 (0.0%)      | 0 (0.0%)        | 0 (0.0%)                    | 8 (80.0%)                    | <0.01* | 8 (80.0%)     | 2 (20.0%)    | 1    |
| Early complications, n (%)           | 2 (20.0%)        | 1 (10.0%)     | 0 (0.0%)        | 2 (20.0%)                   | 4 (40.0%)                    | 0.60   | 8 (80.0%)     | 2 (20.0%)    |      |
| Clavien-dindo class, n (%)           |                  |               |                 |                             |                              | 0.04*  |               |              | 0.19 |
| II                                   | 2 (100.0%)       | 0 (0.0%)      | 0 (0.0%)        | 0 (0.0%)                    | 0 (0.0%)                     |        | 2 (100.0%)    | 0 (0.0%)     |      |
| IIIA                                 | 0 (0.0%)         | 1 (100.0%)    | 0 (0.0%)        | 0 (0.0%)                    | 0 (0.0%)                     |        | 0 (0.0%)      | 1 (100.0%)   |      |
| IIIB                                 | 2 (66.7%)        | 0 (0.0%)      | 0 (0.0%)        | 0 (0.0%)                    | 0 (0.0%)                     |        | 3 (100.0%)    | 0 (0.0%)     |      |
| IVA                                  | 0 (0.0%)         | 0 (0.0%)      | 0 (0.0%)        | 0 (0.0%)                    | 2 (100.0%)                   |        | 1 (50.0%)     | 1 (50.0%)    |      |
| IVB                                  | 0 (0.0%)         | 0 (0.0%)      | 0 (0.0%)        | 0 (0.0%)                    | 1 (100.0%)                   |        | 1 (100.0%)    | 0 (0.0%)     |      |
| V                                    | 0 (0.0%)         | 0 (0.0%)      | 0 (0.0%)        | 2 (66.7%)                   | 1 (33.3%)                    |        | 3 (100.0%)    | 0 (0.0%)     |      |
| Two-stage intervention, n (%)        | 1 (25.0%)        | 1 (25.0%)     | 0 (0.0%)        | 1 (25.0%)                   | 1 (25.0%)                    | 0.54   | 3 (75.0%)     | 1 (25.0%)    | 1    |
| Redo surgery, n (%)                  | 2 (40.0%)        | 0 (0.0%)      | 0 (0.0%)        | 0 (0.0%)                    | 2 (40.0%)                    | 0.42   | 4 (80.0%)     | 1 (20.0%)    | 1    |
| Functional outcome, n (%)            |                  |               |                 |                             |                              | 0.67   |               |              | 0.15 |
| Restored continence                  | 2 (22.2%)        | 1 (11.1%)     | 1 (11.1%)       | 4 (44.4%)                   | 1 (11.1%)                    |        | 6 (66.7%)     | 3 (33.3%)    |      |
| Improved continence                  | 2 (25.0%)        | 0 (0.0%)      | 0 (0.0%)        | 3 (37.5%)                   | 2 (25.0%)                    |        | 8 (100.0%)    | 0 (0.0%)     |      |
| Incontinence                         | 4 (80.0%)        | 0 (0.0%)      | 0 (0.0%)        | 1 (20.0%)                   | 0 (0.0%)                     |        | 5 (100.0%)    | 0 (0.0%)     |      |
| Worsened bowel function              | 0 (0.0%)         | 0 (0.0%)      | 0 (0.0%)        | 1 (100.0%)                  | 0 (0.0%)                     |        | 1 (100.0%)    | 0 (0.0%)     |      |
| Death, n (%)                         | 0 (0.0%)         | 0 (0.0%)      | 0 (0.0%)        | 2 (66.7%)                   | 1 (33.3%)                    | 0.64   | 3 (100.0%)    | 0 (0.0%)     | 1    |

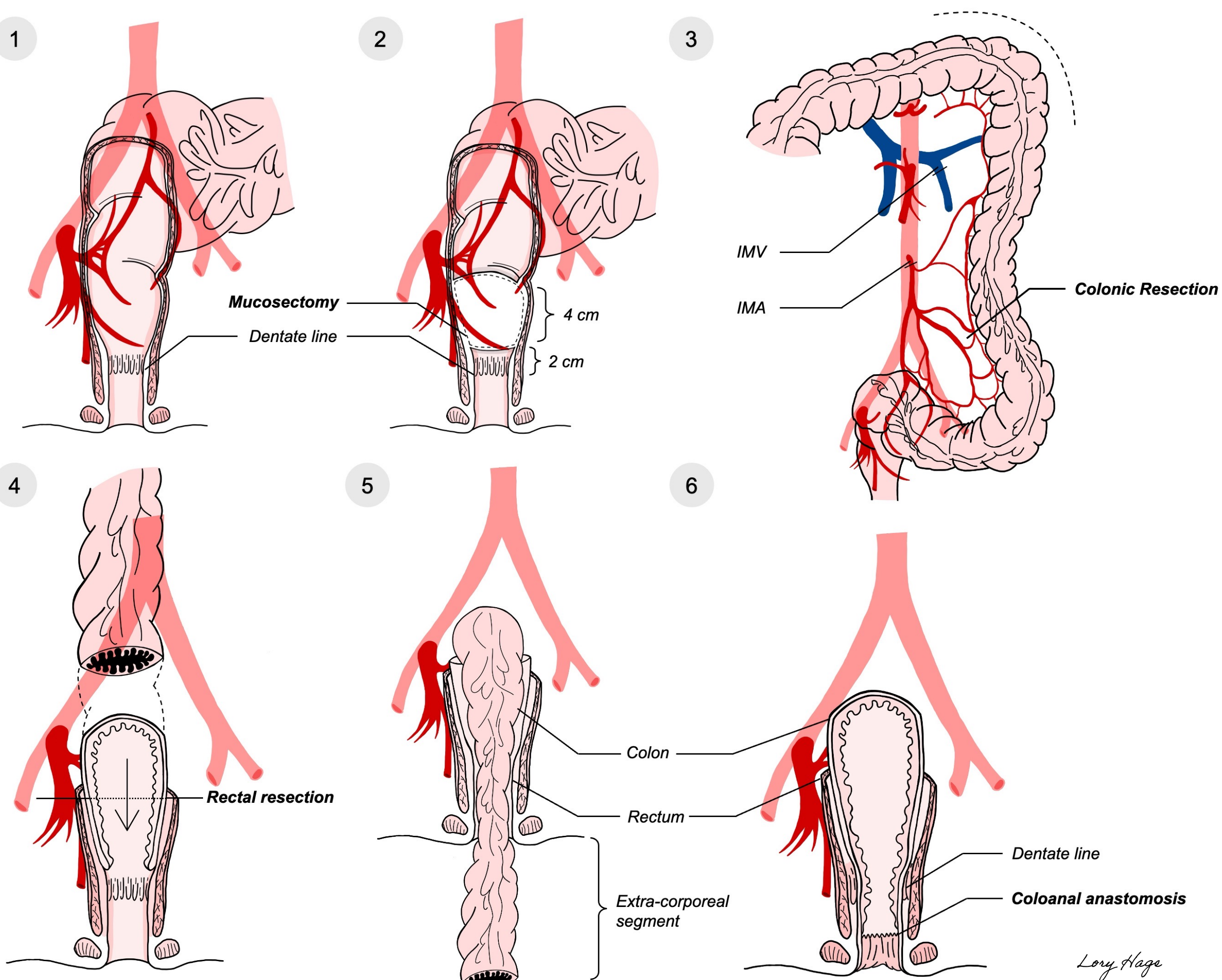
**Table 1. Differences in perioperative outcomes according to surgical intervention.** Resection w/ anas., bowel resection with primary anastomosis; Resection w/o anas., bowel resection without primary anastomosis; Lap., laparoscopic approach.

### Postoperative outcomes (Table 1)

- Early complications rate of 17.2%, with 90% being **severe** (Clavien-Dindo > 3A)
- Most common: faecal incontinence (9%) and subocclusion (5.3%), often requiring **redo surgery** (8.6%)

→ Pull-through techniques showed fewer severe complications than resection (p = 0.04)

## SURGICAL TECHNIQUE



**Figure 4. Surgical steps of the two-stage Soave laparoscopic endorectal pull-through (LERPT) procedure performed in a 38-year-old patient with delayed diagnosis of HD.**

- (1) Transanal mucosectomy
- (2) 2 cm above the dentate line for ± 4 cm cranially
- (3) Laparoscopic dissection of the mesentery up to the level of the mucosectomy, ligation of the inferior mesenteric vein (IMV) and inferior mesenteric artery (IMA) and resection of the proximal colon above the diseased segment
- (4) resection of the rectum at the level of the mucosectomy
- (5) 8 cm segment of healthy colon is pulled through transanally
- (6) 8 days postoperatively extracorporeal segment is resected and a coloanal anastomosis is performed.

## CONCLUSION

- **Early diagnosis** is critical to reduce morbidity
- **Tailored surgical intervention** significantly improve outcomes and quality of life in adult HD patients.
- The presented **two-stage LERPT** technique is feasible in adults and a promising alternative