Case Report

Surgical management of neglected anorectal malformation in an adult

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Abstract

Background: Neglected anorectal malformations in adults, often resulting from failed neonatal repairs and long-term loss of follow-up, present significant surgical challenges, including chronic constipation, megarectum, and fecal incontinence. Management of these rare cases requires a staged surgical approach, utilizing meticulous dissection to navigate hostile tissue and a tailored bowel management program to improve functional outcomes. Detailed radiological findings, such as MRI and contrast studies, are essential for mapping complex anatomy, identifying the sphincter complex, and planning necessary redo surgeries.

Key words: Anorectal Malformations (ARM); Tapering Rectoplasty

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1. Introduction

Anorectal Malformations (ARM) are congenital anomalies typically corrected in infancy but neglected or poorly followed cases in adulthood often lead to severe secondary complications, most notably acquired megacolon and megarectum. This condition is characterized by a massive, aperistaltic reservoir that causes chronic constipation and, eventually, overflow incontinence (encopresis) as liquid stool bypasses an inspissated fecaloma. This case presentation follows a 21-year-old female with a childhood history of ARM who presented with a 20-cm wide mega-sigmoid and extensive fecal impaction. The clinical challenge lies in the transition from pediatric to adult care, requiring a complex two-stage surgical approach—sigmoidectomy with tapering rectoplasty followed by anoplasty and colostomy reversal—to restore anatomical volume and functional bowel continuity.

2. Case Presentation

Initial Presentation: Patient: 21-year-old female with lifelong constipation.

History: S/P neonatal repair (? PSARP) at 3 months; lost to medical follow-up for 20 years.

Symptoms: RLQ pain (3 days), soiling/overflow incontinence (4 months).

3. Clinical Findings

- Abdomen: Massive, tender fecaloma; sigmoid extending to right subcostal area.
- P/R: Poor anal tone; hard, inspissated fecal impaction.
- Imaging: CT showed subacute large bowel obstruction; Sigmoid/Rectum dilated to 15–20cm; Right hydronephrosis.

3.1. Stage 1 surgery: 28/03/2025

Procedure: Sigmoidectomy with Tapering Rectoplasty and End Sigmoid Colostomy.

Operative Findings

- Sigmoid (20cm width) reaching the liver.
- Rectum (15cm width) displacing the uterus and bladder.
- Distal ileum adherent to the sigmoid.
- Surgical Technique: Resected redundant sigmoid utilized 100mm/60mm staplers for Rectoplasty to reduce rectal reservoir; created a left-sided stoma.

Outcome: 1-unit PRBC transfused for anemia. Patient discharged stable on POD-5.

3.2. Stage 2 surgery: 06/02/2026

Interval Assessment: Anal manometry (Normal); Sigmoidoscopy (Mild diversion proctitis); CECT showed rectum reduced to 4.3cm.

Procedure: Anoplasty, Anal Dilatation, and Colostomy Reversal.

4. Key Findings

- Severe anal stenosis (1cm fibrotic ring); dilated to Hegar size 20.
- Extensive adhesions (Uterus adherent to presacral fascia/rectum).
- Post-op Course: Managed postoperative ileus/vomiting (POD 5).

5. Discussion

- Pathophysiology: Chronic fecal retention leads to permanent dilation (Mega-rectum), rendering the bowel aperistaltic.
- Surgical Logic: Manual disimpaction is insufficient for a 20cm wide sigmoid. Resection and tapering (Rectoplasty) are mandatory to restore a functional diameter.
- The Transition Gap: This case highlights the danger of "losing" pediatric patients to follow up, resulting in complex adult presentations that require pediatric-adult surgical collaboration.

- Prognosis: Normal manometry suggests potential for continence, but physical stenosis (Anoplasty) was a critical barrier to overcome.

6. Take home message

- Lifelong Follow-up: ARM patients require transition-to-adult care programs to prevent "megabowel" complications.
- Surgical Volume Reduction: In extreme cases, simply diverting the bowel is not enough; the ectatic segments must be reduced (Rectoplasty) to facilitate future emptying.
- Bowel Management: Post-reversal success is not guaranteed by surgery alone; the patient requires a lifelong Bowel Management Regimen (laxatives and timed stools).