ISSN: 2977-6139

Case Report

Open Access Journal of Pediatrics Research

Diagnostic and Therapeutic Approach to Refractory Constipation in a Pediatric Patient with Vater Association: A Case Report

Candela Lucía Poblete

Pediatric Clinic Resident, SAMIC Pediatric Hospital, Argentina

***Corresponding author**
Candela Lucía Poblete, Pediatric Clinic Resident, SAMIC Pediatric Hospital, Argentina

Received: December 15, 2025; **Accepted:** December 23, 2025; **Published:** December 28, 2025

Introduction

Refractory constipation is defined as constipation that persists despite conventional treatment at adequate doses, including dietary and behavioral modifications and the use of laxatives for at least three months[1]. It represents a complex clinical problem that can significantly impact the patient's quality of life and pose a diagnostic and therapeutic challenge for the medical team.

In patients with congenital malformations, such as those diagnosed with VATER association, refractory constipation acquires greater clinical relevance. VATER association is a polymalformative congenital disorder that includes vertebral, anorectal, cardiac, tracheoesophageal, and renal anomalies, which may coexist in different combinations, with at least three required for diagnosis[2]. The anorectal and gastrointestinal anatomical abnormalities inherent to this syndrome, in adittion to the frequent surgical history, contribute to the development of intestinal functional disorders that are difficult to manage.

The management of refractory constipation in patients with VATER association is particularly challenging due to the anatomical and functional complexity that characterizes this group. Anorectal malformations and frequent prior surgeries lead to alterations in intestinal motility, rectal sensitivity, and defecation, limiting the response to conventional therapies[3] and requiring a comprehensive evaluation by a multidisciplinary team.

This report presents the clinical case of a pediatric patient with VATER association and refractory constipation, with the aim of describing the diagnostic process, the therapeutic interventions implemented, and the outcomes obtained, as well as discussing the most appropriate management strategies within the context of the complexity of these patients.

Objectives

General Objective

To describe the diagnostic process and therapeutic management of refractory constipation in a patient with VATER association.

Specific Objectives

- To analyze the anatomical and functional factors involved in the patient's refractory constipation.
- To detail the therapeutic strategies implemented and their clinical effectiveness.
- To evaluate the clinical course and response to treatment during follow-up.
- To discuss the importance of a multidisciplinary approach in the management of complex cases of refractory constipation associated with congenital malformations.

Clinical Case

We report the case of a nine-year-old female patient who began follow-up for severe chronic constipation refractory to treatment. Her underlying condition was VATER association, including a diagnosis of type III esophageal atresia, imperforate anus with vestibular fistula, and sacral agenesis.

Surgical and Gastrointestinal History

The patient was born preterm at 26 weeks of gestation and was diagnosed with long-gap type III esophageal atresia, requiring

Citation: Candela Lucía Poblete. Diagnostic and Therapeutic Approach to Refractory Constipation in a Pediatric Patient with Vater Association: A Case Report. Open Access J Ped Res. 2025. 2(4): 1-3. DOI: doi.org/10.61440/OAJPR.2025.v2.34

gastrostomy placement and esophageal elongation using the Kimura technique, with delayed end-to-end anastomosis performed at four months of age. She required multiple esophageal dilations due to anastomotic stricture, the most recent one performed two years prior to the current evaluation.

As part of her esophageal atresia follow-up, upper videoendoscopy was performed and was normal, with no histological abnormalities. A 24-hour pH-impedance study demonstrated non-acid reflux with impaired clearance, for which she remained on omeprazole 1 mg/kg/day and domperidone 0.2 mg/kg/dose every 8 hours, with favorable clinical evolution.

Anorectal History and Evolution of Constipation

Regarding her anorectal malformation, which included imperforate anus and vestibular fistula, she initially required a sigmoid colostomy at five months of age, followed by fistula closure, rectal pull-through, and restoration of intestinal continuity at one year of age. Since then, the patient has experienced constipation that has been difficult to manage, requiring high doses of osmotic and stimulant laxatives, in addition to soap suds and phosphate enemas, with no response to oil-retention enemas.

The management of constipation in this patient was particularly challenging due to refractoriness to first-line treatments, requiring multiple hospitalizations for fecal impaction, which worsened at eight years of age. At that time, a contrast enema study showed dilation of the rectum and sigmoid colon with radiological signs of dolichosigmoid, without caliber changes and with adequate passage of contrast to the transverse colon (Figure 1).

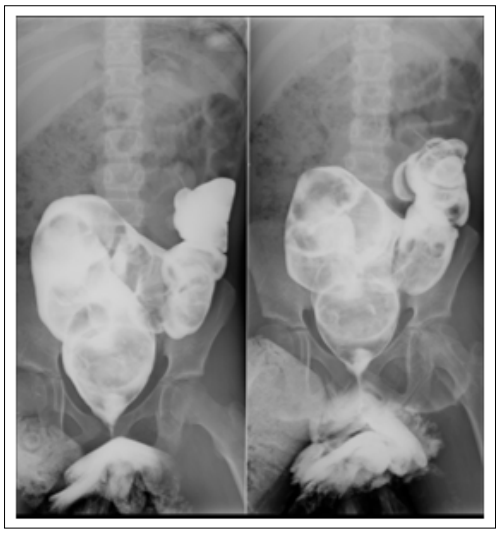

Figure 1: Contrast enema

Showed dilation of the rectum and sigmoid colon.

In this context, transanal colonic irrigations with water were indicated. Although an adequate response was achieved, the patient had poor tolerance to the procedure. A consultation with the general surgery team led to the decision to perform resection of the megarectosigmoid, considering that this dilated intestinal segment with loss of contractile capacity and sensitivity could promote chronic fecal accumulation and contribute to the unfavorable course. Histopathological examination of the resected specimen revealed the presence of ganglion cells, ruling out aganglionosis.

The postoperative course was uneventful; however, several months later, a follow-up contrast enema revealed recurrent rectal dilation, with no additional findings.

Functional Evaluation

A high-resolution anorectal manometry was performed, showing a resting pressure of 15.8 mmHg, voluntary contraction pressure of 42 mmHg, and absence of the rectoanal inhibitory reflex (RAIR) (Figure 2). These findings were consistent with a contractility disorder characterized by hypotonia of both sphincters, type I dyssynergia, and associated rectal hyposensitivity.

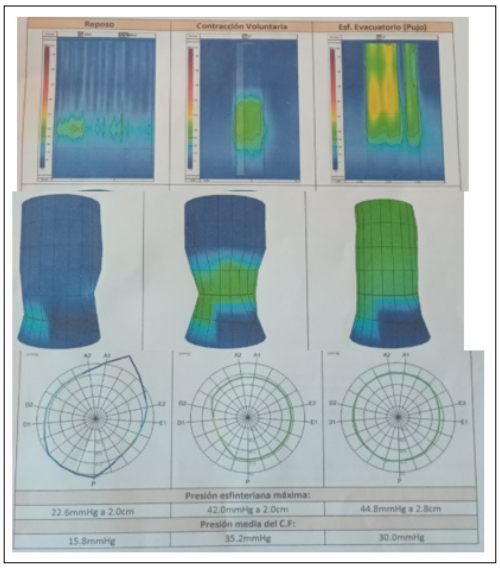

Figure 2: High-resolution anorectal manometry study

Demonstrating decreased resting pressures with moderately to severely hypotensive external anal sphincter.

Based on these results, it was concluded that, in addition to the anatomical component, the patient's constipation was associated with an anorectal motility disorder, explaining the poor response to conventional treatments and the recurrence of rectal dilation.

Management and Follow-up

Pelvic floor rehabilitation and anorectal biofeedback therapy were initiated, along with polyethylene glycol 50 g/day, bisacodyl 5 mg/day, and oil-retention plus phosphate enemas. Despite these interventions, the patient continued to experience severe constipation.

Given the persistence of symptoms despite comprehensive treatment aimed at improving intestinal motility and addressing all therapeutic aspects, a multidisciplinary decision was made to schedule the creation of a cecostomy with a CHAIT button for antegrade enemas.

The patient remains under follow-up by the treating team.

Conclusion

The present case illustrates the diagnostic and therapeutic complexity of refractory constipation in a patient with VATER association, in whom constipation is refractory to conventional measures due to the previously described clinical background.

This case highlights the need for a comprehensive and multidisciplinary approach, based on evidence and adapted to the anatomical and physiological particularities of each patient, demonstrating that treatment strategies must always be individualized.

Early recognition of refractoriness and consideration of advanced interventions are sometimes necessary to improve quality of life in this patient population. We consider the development of multidisciplinary teams essential, integrating the different specialists involved in the follow-up of patients with anorectal malformations, to enable joint evaluation, coordinated follow-up, and shared decision-making, thereby optimizing comprehensive care. Furthermore, such an approach would promote continuous professional development through the exchange of experiences and the creation of standardized protocols aimed at improving long-term outcomes [1-3].

References

1. Kilgore AL, Rogers Boruta MK, Ambartsumyan L, Gómez Suárez R, Patel D, et al. Evaluación y manejo del estreñimiento refractario pediátrico: recomendaciones del comité de neurogastroenterología y motilidad de NASPGHAN. Journal of Pediatric and Gastroenterology and Nutrition. 2025. 80: 353-373.
2. Tonni G, Koçak Ç, Grisolia G, Rizzo G, Araujo Júnior E, et al. Presentaciones clínicas y diagnóstico por imagen de la asociación VACTERL. Patología Fetal y Pediátrica. 2023. 42: 651–674.
3. Hakalmaz AE, Topuzlu Tekant G. Anorectal Malformations and Late-Term Problems. Turk Arch Pediatr. 2023. 58: 572-579.