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Original Article

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# Primary anastomosis with or without T-tube enterostomy in jejuno-ileal atresia-a comparative study

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## ABSTRACT

**Aims:** Various surgical procedures are in practice for the management of jejuno-ileal atresia. Most used technique is resection of dilated segment of gut and primary anastomosis. But mortality and morbidity still remains high. Primary anastomosis with T- tube enterostomy is a recent concept for the treatment of jejuno-ileal atresia. It seems to offer important advantages over other surgical techniques. The aim of this study was to evaluate the effectiveness of T-tube enterostomy to reduce morbidity and mortality of neonate suffering from jejuno-ileal atresia.

**Methods:** It was a prospective comparative interventional study which was conducted in Bangladesh Shishu (Children) Hospital from March 2017 to September 2019. A total of 54 patients with uncomplicated jejuno-ileal atresia were selected for this study after fulfillment of inclusion and exclusion criteria. They were randomly assigned to primary anastomosis with T-tube enterostomy group (group A=26) and primary anastomosis without T-tube enterostomy group (group B=28). The comparative parameters between two groups were start of post-operative bowel movement, establishment of complete oral feeding, duration of hospital stay, anastomotic leakage and mortality. All patients were followed up for 6 months post operatively.

**Results:** The age range of the patients was 2 to 10 days where majority of the patients were male. Most of the patients were male in both groups. After operation, mean time to start bowel movement of group A 4.68±1.21 days and group B 5.29±1.82 days and there was no significant statistical difference. Time to establish complete oral feeding of group A (9.04±1.43 days) and group B (10.38±2.18 days) showed significant difference (p=xv 0.016). Rate of anastomotic leakage in group A was 7.7% and group B was 32.1% which was statistically significant (p=0.041). Rate of mortality was higher in group B (32.1%) compared to group A (7.7%) which was statistically significant (p=0.026). Time of post- operative hospital stay in group A (14.19±2.22 days) and group B (16.39±3.97 days) showed significant difference (p=0.015).

**Conclusion:** Primary anastomosis with T-tube enterostomy was found as an effective procedure to reduce morbidity and mortality of patients with jejuno-ileal atresia.

**Keywords**: T-tube enterostomy, jejuno-ileal atresia, intestinal obstruction

## INTRODUCTION

Management of jejuno-ileal atresia remains a major challenge for neonatal and paediatric surgeons worldwide, requiring rapid resuscitation and timely surgical intervention. Despite advances in neonatal care, morbidity and mortality rates remain high, particularly in developing countries.[1,2] Intestinal atresia refers to congenital complete obstruction of the intestinal lumen, most commonly affecting the duodenum, jejunum, or ileum, and rarely the colon.[3] Jejuno-ileal atresia is one of the leading causes of neonatal intestinal obstruction.[4] The marked discrepancy between the dilated proximal and narrow distal segments contributes to significant anastomotic complications,[5] making this condition a frequent indication for intestinal resection with associated high morbidity and mortality (**Figure 1**).[6] Over recent decades, improvements in neonatal intensive care, operative techniques, and the use of total parenteral nutrition (TPN) have reduced mortality in developed settings.[1,7] However, in resource-limited countries, outcomes remain poor.[8] Current treatment options include resection with primary anastomosis or exteriorization with delayed anastomosis.[9]

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The key principle is bowel preservation, management of the dilated proximal segment, and early initiation of oral feeding.[6] The most widely used approach—resection and end-to-back anastomosis—often requires removal of the dilated proximal segment, risking short bowel syndrome. Moreover, disparities in diameter and peristalsis between proximal and distal segments predispose to anastomotic leakage, sepsis, and bacterial overgrowth.[7,10] Short bowel syndrome remains a major postoperative concern.[11] To minimize bowel loss, techniques such as tapering enteroplasty, antimesenteric plication, and the Bianchi procedure have been described, though each carries risks of electrolyte imbalance and metabolic complications.[4,12] Alternatively, stoma-forming procedures such as the Mikulicz, Bishop-Koop, and Santuli methods allow decompression of the dilated bowel and reduce anastomotic leak rates.[11] Yet these techniques entail high-output stomas, fluid-electrolyte disturbances, malnutrition, and morbidity from subsequent stoma closure.[13,14] Use of transanastomotic tubes in duodenal atresia has shown favorable outcomes, facilitating early recovery.[15,16] The intraluminal T-tube, first used in hepatobiliary surgery by Hans Kehr, functions as a stent providing both internal and external drainage.[17,18] Its application in uncomplicated meconium ileus has proven safe and effective.[19,20] Chaet et al.[21] first reported T-tube use in jejuno-ileal atresia. Primary anastomosis with transanastomotic T-tube enterostomy offers dual benefits: internal drainage and decompression, functioning as a low-output stoma without the need for secondary surgery. The tube can be easily removed, and the site heals spontaneously.[22] Furthermore, preserving the dilated proximal segment prevents short bowel syndrome.[11] Previous studies demonstrated improved outcomes with this approach.[6,11] Given these promising findings and successful T-tube use in meconium ileus in Bangladesh, this study was undertaken to evaluate the role and effectiveness of primary anastomosis with T-tube enterostomy in the management of jejuno-ileal atresia.



**Figure 1.** Per operative picture of jejuno-ileal atresia (type-IIIB)

## METHODS

### Ethics

Ethical clearance was obtained from the Ethical Review Committee of Bangladesh Institute of Child Health & Bangladesh Shishu Hospital (Date: 21.05.2018, Decision No: BICH-ERC-12/01/2018). All procedures were carried out in accordance with the ethical rules and the principles of the Declaration of Helsinki. Written informed consent was obtained from guardians.

### Study Design

This was a prospective comparative interventional study.

### Study Place

This was a prospective comparative interventional study. The study was conducted in the Division of Paediatric Surgery, Bangladesh Institute of Child Health & Bangladesh Shishu (Children) Hospital, Dhaka.

### Study Duration

The study period was two years and six months, from 1st March 2017 to 30th September 2019.

### Study Population

Patients diagnosed with jejuno-ileal atresia at Bangladesh Shishu Hospital during the study period were included.

### Sampling Technique

Randomized sampling was applied. Patients preliminarily diagnosed with jejuno-ileal atresia were enrolled after obtaining informed consent from guardians, with final diagnosis confirmed during surgery.

### Selection Criteria

- **Inclusion:** All neonates with uncomplicated jejuno-ileal atresia confirmed during surgery.
- **Exclusion:** Major congenital anomalies affecting outcome and multiple atresia.

### Sample Size

Calculated using the formula:
$n=(P1(1-P1)+P2(1-P2))/((P1-P2)^2)\times(Z\alpha+Z\beta)^2$, with P1=0.44, P2=0.14, Zα=1.96, Zβ=0.85. The required sample was 32 per group, totaling 64 patients.

### Randomization

Sixty-four cards (32 per group) were used. Guardians picked cards to allocate patients to group-A (primary anastomosis with T-tube enterostomy) or group-B (primary anastomosis without T-tube enterostomy).

### Data Collection

Information was recorded using a pre-designed semi-structured questionnaire. Variables included time to bowel movement, time to complete oral feeding, postoperative morbidity and mortality, and duration of hospital stay.

### Preoperative Evaluation and Treatment

Patients were assessed for general condition, associated anomalies, and intestinal obstruction. Investigations included CBC, electrolytes, bilirubin, chest X-ray, and echocardiogram as indicated. Preoperative care included NPO, nasogastric decompression, intravenous fluids, antibiotics, and vitamin K.

## Operative Procedures

- **Group-A:** Minimal bowel resection, end-to-back anastomosis with 12 Fr T-tube for decompression and stenting (**Figure 2-4**).

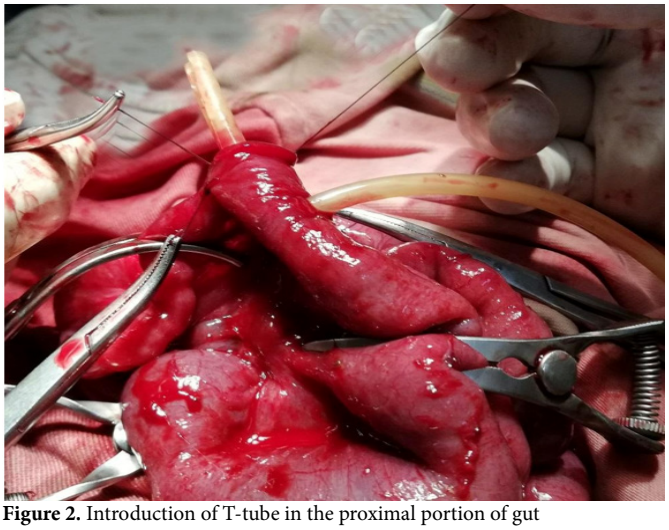


**Figure 2.** Introduction of T-tube in the proximal portion of gut

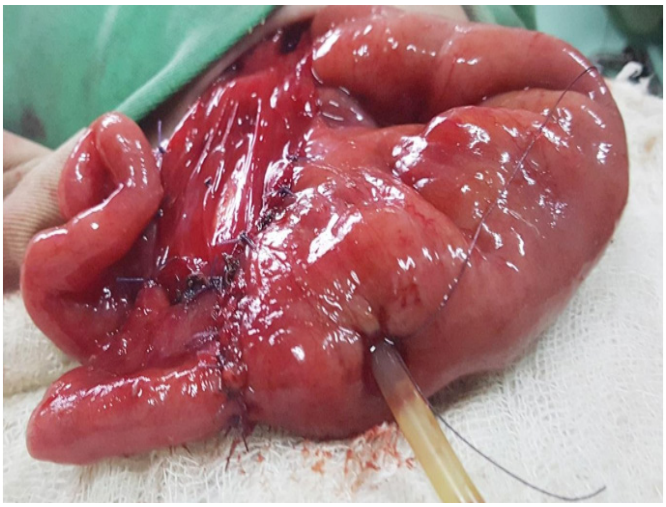


**Figure 3.** Primary anastomosis with placement T-tube proximal to anastomotic site



**Figure 4.** Primary anastomosis with T-tube enterostomy

- **Group-B:** Resection of dilated proximal bowel followed by end-to-back anastomosis (**Figure 5**).

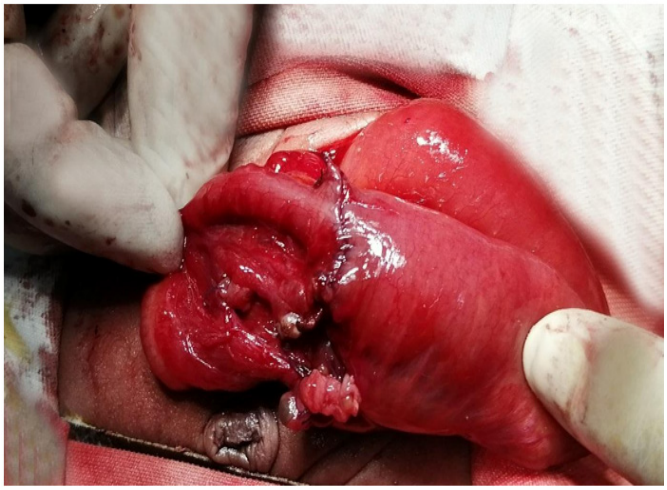


**Figure 5.** Primary anastomosis without T-tube enterostomy

## Postoperative Care and Follow-up

Patients received NPO, nasogastric suction, IV fluids, antibiotics, and analgesics. Oral feeding was started after bowel movement. Group-A T-tube output was monitored and removed after clinical readiness (**Figure 6**). Patients were followed for vital signs, abdominal condition, wound status, and complications.



**Figure 6.** Spontaneous closure of wound after removal of T-tube

## Diagnosis and Management of Complications

Anastomotic leakage, bowel obstruction, sepsis, wound infection, and wound dehiscence were identified based on clinical and radiological findings and managed appropriately.

## Discharge and Follow-up

Patients were discharged after tolerance of complete oral feeding and wound healing, with advice on signs of complications. Follow-up visits were scheduled at 2$^{nd}$, 4$^{th}$ week, 3$^{rd}$ and 6$^{th}$ month postoperatively.

## Statistical Analysis

Data were analyzed using SPSS v25. Continuous variables were assessed with Student's t-test, categorical variables with Chi-square or Fisher's exact test. $p<0.05$ was considered significant.

## RESULTS

A total of 64 patients meeting inclusion and exclusion criteria were enrolled and randomly allocated into two groups: group A (n=32) and group B (n=32). Twenty-six patients in group A and 28 patients in group B completed the six-month follow-up. The results of these 54 patients are presented below.

### Demographic and Baseline Characteristics

The mean age at presentation in group A was 4.04±1.75 days and in group B was 4.14±1.88 days, with no significant difference between groups (p=0.834, Student's t test, **Table 1**). Gender distribution was similar, with 15 (57.7%) males in group A and 17 (60.7%) males in group B (p=1.000, Chi-square test, **Table 2**). Type IIIA atresia was most common in both groups (42.3% in group A, 39.3% in group B), with no significant difference in type distribution (p=0.973, Fisher's exact test, **Table 3**).

**Table 1.** Age at operation

| Age (days) | Group A (n=26) | Group B (n=28) | p value |
|---|---|---|---|
| 2-3 | 12 (46.2%) | 11 (39.3%) | |
| 4-5 | 10 (38.5%) | 11 (39.3%) | |
| >6 | 4 (15.4%) | 6 (21.4%) | |
| Mean±SD | 4.04±1.75 | 4.14±1.88 | 0.834 |
| Range | 2-10 | 2-9 | |

SD: Standard deviation

**Table 2.** Gender distribution

| Gender | Group A no. (%) | Group B no. (%) | p value |
|---|---|---|---|
| Male | 15 (57.7%) | 17 (60.7%) | 1.000 |
| Female | 11 (42.3%) | 11 (39.3%) | |

**Table 3.** Type of atresia

| Type | Group A no. (%) | Group B no. (%) | p value |
|---|---|---|---|
| Type I | 4 (15.4%) | 4 (14.3%) | 0.973 |
| Type II | 9 (34.6%) | 11 (39.3%) | |
| Type IIIA | 11 (42.3%) | 11 (39.3%) | |
| Type IIIB | 2 (7.7%) | 2 (7.1%) | |

### Post-Operative Outcomes

The mean time for establishment of bowel movement postoperatively was 4.68±1.21 days in group A and 5.29±1.82 days in group B (p=0.166, Student's t test. Group A patients initiated oral feeding earlier than group B (6.96±1.39 vs. 8.29±2.18 days), with this difference being statistically significant (p=0.015. Similarly, the mean time to achieve complete oral feeding was shorter in group A (9.04±1.43 days) compared to group B (10.38±2.18 days), which was significant (p=0.016, **Table 4**). In group A, the mean time of T-tube removal was 10.67±1.52 days (range 8-13 days, **Table 4**).

### Complications

Anastomotic leakage occurred in 2 patients (7.7%) in group A and 9 patients (32.1%) in group B, showing a significant difference (p=0.041, Chi-square test, **Table 5**). Postoperative obstruction occurred in 2 patients (7.1%) in group B, while none occurred in group A (p=0.491. Wound infection was observed in 1 patient (3.8%) in group A and 3 patients (10.7%) in group B (p=0.612, **Table 6**). Wound dehiscence occurred in 1 patient in group A and 2 patients in group B (p=0.528, **Table 6**).

**Table 4.** Post-operative outcomes

| Parameter | Group A (n=26) | Group B (n=28) | p value |
|---|---|---|---|
| Time to bowel movement (days) | 4.68±1.21 | 5.29±1.82 | 0.166 |
| Time to start oral feeding (days) | 6.96±1.39 | 8.29±2.18 | 0.015 |
| Time to complete oral feeding (days) | 9.04±1.43 | 10.38±2.18 | 0.016 |
| T-tube removal (days) | 10.67±1.52 | - | - |
| Post-op hospital stay (days) | 14.19±2.22 | 16.39±3.97 | 0.015 |

**Table 5.** Anastomotic leakage

| Anastomotic leakage | Group A no. (%) | Group B no. (%) | p value |
|---|---|---|---|
| Absent | 24 (92.3%) | 19 (67.9%) | 0.041 |
| Present | 2 (7.7%) | 9 (32.1%) | |

**Table 6.** Other complications

| Complication | Group A no. (%) | Group B no. (%) | p value |
|---|---|---|---|
| Post-op obstruction | 0 (0%) | 2 (7.1%) | 0.491 |
| Wound infection | 1 (3.8%) | 3 (10.7%) | 0.612 |
| Wound dehiscence | 1 (3.8%) | 2 (7.1%) | 0.528 |
| Mortality | 2 (7.7%) | 9 (32.1%) | 0.026 |

### Mortality and Hospital Stay

Mortality was significantly lower in group A (2 patients, 7.7%) compared to group B (9 patients, 32.1%; p=0.026, Chi-square test. The mean postoperative hospital stay was also significantly shorter in group A (14.19±2.22 days) than in group B (16.39±3.97 days; p=0.015, (**Table 6**).

## DISCUSSION

Management of jejuno-ileal atresia remains a surgical challenge. Various techniques such as resection with end-to-back anastomosis, tapering enteroplasty, Mikulicz, Bishop-Koop, Santulli, and T-tube enterostomy are employed. This study compared outcomes of primary anastomosis with or without T-tube enterostomy. About 38% of patients were operated within 4-5 days of age, while 18.5% presented after 6 days, often due to referrals from other hospitals. The mean age was approximately 4 days in both groups, consistent with Shahjahan et al.[23] and Rouzrokh et al.[11] Most neonates were male, similar to findings of Shakya et al[10] and Shahjahan et al,[23] although jejuno-ileal atresia is typically gender-neutral.[24] Type III A atresia was the most common, with no significant difference between groups. Postoperative bowel movement occurred earlier in the T-tube group (4.68±1.21 days) than without T-tube (5.29±1.82 days), though this difference was not statistically significant. Early bowel movement facilitated earlier initiation of oral feeding. Primary anastomosis with

T-tube enterostomy allowed proximal gut decompression and gradual flow to the distal bowel, stimulating peristalsis. Consequently, first and complete oral feeding started earlier in the T-tube group, consistent with Rouzrokh et al,[11] though absolute times were shorter in this study due to lack of TPN facilities. T-tube removal occurred on average at 10.67 days without complications, similar to Al-Zaiem et al.[25] Anastomotic leakage was significantly higher in the primary anastomosis without T-tube group, likely due to increased intraluminal pressure. Rates of postoperative obstruction were also higher without T-tube, though not statistically significant, consistent with Ali and Saleem[26] and Shakya et al.[10] Dilated proximal bowel and disturbed intestinal transit contribute to such complications.[27,28] Wound infection and dehiscence were lower with T-tube enterostomy (3.8% each) compared to without T-tube (10.7% and 7.1%), though not statistically significant. Secondary closure was required in cases of dehiscence. Stasis and bacterial overgrowth in the non-T-tube group likely contributed to higher wound complications. Mortality was 20.3% overall, with higher rates in the non-T-tube group, consistent with Rouzrokh et al.[11] Delayed presentation, systemic sepsis, metabolic disturbances, and absence of antenatal diagnosis contributed to mortality. Limited NICU availability and lack of TPN support at Dhaka Shishu Hospital may have influenced outcomes. Anastomotic leakage, atonic proximal bowel, and bacterial overgrowth are key factors in postoperative sepsis.[29] Postoperative hospital stay was significantly shorter in the T-tube group due to early feeding and fewer complications, aligning with Rouzrokh et al.[11] Overall, T-tube enterostomy was associated with reduced complications, earlier feeding, and shorter hospitalization compared to primary anastomosis alone.

### Recommendations

- Primary anastomosis with T-tube enterostomy should be the preferred surgical option in the management of jejuno-ileal atresia.
- Multi-center based, larger prospective studies are needed to validate our findings.

### Limitations

- Surgical procedure was performed by different surgeons which can affect the outcome.
- Because of lack of logistic support, financial constraints and lack of availability of coagulation profile, S. bilirubin, chest X-ray, Ultra sonogram of whole abdomen and Contrast X-ray could not be done in all patients. Result could have been influenced due to lack of these facilities.
- Unavailability of surgical ICU and TPN facility could also have influenced the outcome.

## CONCLUSION

This study concludes that, use of T-tube enterostomy proximal to primary anastomosis is helpful to the healing of the anastomotic site in jejuno-ileal atresia. This procedure can improve the outcome and overcome the difficulties regarding the management of jejuno- ileal atresia.

## ETHICAL DECLARATIONS

### Ethics Committee Approval

Ethical clearance was obtained from the Ethical Review Committee of Bangladesh Institute of Child Health & Bangladesh Shishu Hospital (Date: 21.05.2018, Decision No: BICH-ERC-12/01/2018).

### Informed Consent

Informed consent was obtained from a parent or legal guardian. Where appropriate, age-adjusted assent was also obtained from the child. The inclusion of vulnerable populations in this study adhered to national and international ethical guidelines. Extra care was taken to ensure voluntary participation, understanding, and protection of participant dignity and autonomy.

### Peer Review Process

This manuscript was subject to external peer review.

### Conflict of Interest

The authors declare no conflicts of interest related to this study.

### Financial Disclosure

The authors received no financial support for the conduct or publication of this research.

### Author Contributions

Concept: AAF; Design: AAF, TT; Control: AAF, AR; Resources: AAF, TT, AR; Materials: AAF, TT; Data Collection and/or Processing: AAF, TT; Analysis and/or Interpretation: AAF, TT; Literature Review: AAF, TT; Writing the Article: AAF, TT; Critical Review: AR.

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# Fecal secretory immunoglobulin A (sIgA) levels in Hirschsprung's disease and healthy controls: a preliminary study

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## ABSTRACT

**Aims:** Hirschsprung's disease (HD) is a congenital neurocristopathy characterized by distal colonic aganglionosis and is associated with an increased risk of enterocolitis and other postoperative complications. Impaired mucosal immunity, particularly alterations in secretory immunoglobulin A (sIgA), has been proposed as one of the contributing mechanisms. This study aimed to compare fecal sIgA levels between children with HD without enterocolitis and healthy controls.

**Methods:** This exploratory cross-sectional study enrolled pediatric patients aged 2 months to <18 years between September and December 2024 from two referral hospitals and one primary health center. A total of 32 subjects were included: 16 children with histopathologically confirmed HD without enterocolitis and 16 healthy controls. Control participants had no congenital gastrointestinal abnormalities or recent gastrointestinal symptoms and showed normal findings on physical examination, routine stool analysis, and fecal occult blood testing. Fecal samples were analyzed using enzyme-linked immunosorbent assay to quantify sIgA levels. Data normality was assessed using the Shapiro–Wilk test, and group comparisons were performed using the Mann–Whitney U test.

**Results:** The median fecal sIgA level was 1551.56 µg/ml (27.69-35988.75) in the HD group and 771.87 µg/ml (31.27-11250.52) in controls. Although the median value was numerically higher in the HD group, the difference was not statistically significant (p=0.72).

**Conclusion:** Children with HD without enterocolitis showed a numerically higher median fecal sIgA level than healthy controls, but no statistically significant difference was observed. These findings should be interpreted as exploratory baseline data rather than confirmatory evidence of clinical or mechanistic significance.

**Keywords**: Hirschsprung's disease, Hirschsprung-associated enterocolitis, secretory immunoglobulin A (sIgA)

## INTRODUCTION

Hirschsprung's disease (HD) is a congenital gastrointestinal disorder characterized by the absence of ganglion cells in the distal colon, resulting in functional intestinal obstruction. The estimated global incidence is approximately 1 in 5,000 live births, with a male-to-female ratio of 4:1. Although national incidence data in Indonesia are currently unavailable, extrapolation based on population and birth rate estimates suggests a considerable annual number of affected newborns.[1]

Since the first successful surgery by Swenson and Bill in 1946, surgical management of HD has evolved significantly. These include procedures such as Duhamel, Rehbein, Soave, and Boley techniques. Advances toward single-stage and minimally invasive procedures have reduced hospital stay and improved postoperative quality of life.[2-4] Nevertheless, postoperative complications remain a major concern. Hirschsprung-associated enterocolitis (HAEC) continues to be the most serious complication, with reported early prevalence of 28.73% and late prevalence of 21.37%. Soiling and incontinence are also reported in 20.99% of early and 19.66% of late postoperative cases.[2]

Recent evidence suggests that impaired mucosal immunity may contribute to the development of the complications. Huang et al.[2] demonstrated that low serum immunoglobulin A (IgA) levels were significantly associated with the occurrence of enterocolitis (OR=2.766) and soiling/

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incontinence (OR=3.885). Reduced serum IgA is believed to reflect decreased secretory immunoglobulin A (sIgA), which plays a crucial role in intestinal mucosal defense. Lower sIgA levels may weaken gastrointestinal immune protection, facilitating infection, altering the intestinal milieu, and increasing the risk of enterocolitis and incontinence.

Secretory IgA is produced by plasma cells along the intestinal mucosa and functions as a primary immunological barrier by coating microbiota and preventing pathogen adherence. It has been detected in fecal samples of patients with enteritis, where elevated levels are thought to represent a response to infection or inflammation.[5] However, current literature has not specifically examined fecal sIgA expression in patients with HD without enterocolitis. In HD, aganglionosis of the rectum and sigmoid colon that presented in approximately 80% of cases, causes functional obstruction which may promote bacterial translocation and influence sIgA production as part of an immune response.[6]

There is no study specifically evaluating fecal sIgA levels in children with HD without enterocolitis compared with healthy controls. This represents an important gap in understanding baseline mucosal immune status in HD prior to inflammatory complications. Therefore, this study aimed to determine the differences in fecal sIgA levels between children with HD without enterocolitis and healthy controls. The findings of this study are expected to serve as preliminary data for future investigations evaluating sIgA expression in HD.

## METHODS

### Ethics

Ethical approval was obtained from the Ethics Committee of Cipto Mangunkusumo National Hospital (Date: 11.10.2024, Decision No: KET-1437/UN2.F1/ETIK/PPM.00.02/2024). All procedures were carried out in accordance with the ethical rules and the principles of the Declaration of Helsinki. Written informed consent was obtained from parents or legal guardians of all participants prior to enrollment.

### Study Design

This study was a descriptive analytic cross-sectional study using primary data collection. The study was conducted between September and December 2024 at the Pediatric Surgery Clinic of Cipto Mangunkusumo National Hospital, Persahabatan General Hospital, and Senen District Primary Health Center, with laboratory analysis performed at the Integrated Laboratory of the Faculty of Medicine, Universitas Indonesia. Subjects were recruited prospectively using a consecutive sampling technique from eligible patients who met the predefined inclusion and exclusion criteria during the study period. Healthy controls were recruited during the same period from the participating outpatient/primary-care setting.

### Study Population and Sample Size

The target population consisted of pediatric patients aged 2 months to <18 years. The case group included children diagnosed with HD based on clinical evaluation and histopathological confirmation. The control group consisted of healthy children recruited during the same study period from the participating outpatient/primary-care setting and had no congenital gastrointestinal abnormalities or active gastrointestinal infection. Sample size calculation was performed using Sample Size Calculator version 2.0.4 for comparison of two independent numerical groups with 1:1 allocation, a two-sided alpha of 0.05, and 80% power. Because no prior fecal sIgA data in HD were available at protocol development, previously reported serum IgA data were used as surrogate parameters. The minimum required sample size was 16 subjects per group. Given the use of surrogate assumptions and the wide variability subsequently observed in fecal sIgA values, the present study should be interpreted as an exploratory pilot study.

### Inclusion and Exclusion Criteria

Children were included in the HD group if they were aged between 2 months and <18 years, had a confirmed diagnosis of HD based on histopathological findings, had no current or prior history of enterocolitis, and had not undergone definitive pull-through surgery. All patients in the HD group had previously undergone colostomy formation. Children were included in the control group if they were aged between 2 months and <18 years, had no congenital gastrointestinal abnormalities, had no symptoms of gastrointestinal infection within the preceding two weeks, and demonstrated normal findings on physical examination, routine stool analysis, and fecal occult blood testing.

Participants from both groups were excluded if they had known immunoglobulin A deficiency, signs of acute abdomen, severe intestinal obstruction, pneumoperitoneum, or confirmed HAEC. Additional exclusion criteria included long-term immunosuppressive therapy exceeding two months, gastrointestinal infection within two weeks prior to sample collection, other gastrointestinal disorders such as anorectal malformation, necrotizing enterocolitis, neuronal intestinal dysplasia, or acute gastroenteritis, and consumption of Lactobacillus supplements within the previous two weeks. Stool microbiology and viral screening were not performed. Therefore, asymptomatic infection could not be completely excluded.

### Data Collection and Variables

Demographic and clinical data were collected through medical records and structured interviews, including age, sex, histopathological confirmation of HD (for the case group), and body weight. Only variables that were systematically collected in both groups were included as formal baseline comparators. Feeding exposure was recorded descriptively, whereas breastfeeding duration, formula composition, nutritional/environmental variables, and colostomy duration were not systematically quantified for valid comparative analysis.

### Fecal Sample Collection and Analysis

Fresh fecal samples (approximately 15 grams) were collected without specific time restriction, ensuring that samples were not contaminated with urine. In patients with colostomy, samples were collected directly during stoma output. Samples were transported under appropriate temperature conditions and stored at –20°C until analysis. Fecal sIgA levels were

measured using enzyme-linked immunosorbent assay (ELISA) with the IDK® sIgA ELISA kit (Immundiagnostik AG, Germany). Samples were extracted and diluted to a final dilution of 1:75,000 according to the manufacturer's instructions. All measurements were performed in duplicate. Absorbance was read at 450 nm with reference at 620-690 nm, and concentrations were calculated using a standard calibration curve. Results were expressed in µg/ml.

### Statistical Analysis

Data distribution was assessed using the Shapiro–Wilk test. Continuous variables were presented as median (range) because fecal sIgA values were not normally distributed. Gender was compared between groups using Fisher's exact test. Age and other descriptive baseline variables were summarized in **Table 1**, while fecal sIgA levels between groups were compared using the Mann–Whitney U test. A p-value <0.05 was considered statistically significant.

**Table 1.** Baseline characteristic of the study subjects

| Variables | HD (n=16) | Healthy controls (n=16) |
|---|---|---|
| **Age (month)** | 24 (7-96) | 27 (2-72) |
| **0-12 months old** | 6 (37.5%) | 4 (25%) |
| **>12-24 months old** | 3 (18.8%) | 4 (25%) |
| **>24 months old** | 7 (43.8%) | 8 (50%) |
| **Gender** | | |
| Male (%) | 12 (75%) | 10 (62.5%) |
| Female (%) | 4 (25%) | 6 (37.5%) |
| **Body weight (kg)** | 20.01 (7.5-22.2) | 12.55 (4.8-35) |

Data are presented as median (min-max) for abnormally distributed numeric variable or n (%) for categoric variable. HD: Hirschsprung's disease, Min: Minimum, Max: Maximum

## RESULTS

A total of 32 subjects were included in this study, consisting of 16 children with HD and 16 healthy controls. The baseline characteristics of the study subjects are summarized in **Table 1**. The median age was 24 months (range 7-96) in the HD group and 27 months (range 2-72) in the healthy controls. In the HD group, 6 subjects (37.5%) were 0-12 months, 3 subjects (18.8%) were >12-24 months, and 7 subjects (43.8%) were >24 months. In the healthy controls, 4 subjects (25.0%) were 0-12 months, 4 subjects (25.0%) were >12-24 months, and 8 subjects (50.0%) were >24 months. Male predominance was observed in both groups, with 12 subjects (75.0%) in the HD group and 10 subjects (62.5%) in the healthy controls being male. Female subjects accounted for 4 (25.0%) and 6 (37.5%) in the HD and healthy controls, respectively. The median body weight was 20.01 kg (range 7.5-22.2) in the HD group and 12.55 kg (range 4.8-35) in the healthy controls.

The comparison of fecal sIgA levels between groups is presented in **Table 2**. Data normality testing using the Shapiro–Wilk test demonstrated that fecal sIgA levels were not normally distributed in both groups (p<0.05). Therefore, results were expressed as median (range), and comparisons between groups were performed using the Mann–Whitney U test. The median fecal sIgA level in the HD group was 1551.56 µg/ml (range 27.69-35988.75), whereas the median level in the healthy controls was 771.87 µg/ml (range 31.27-11250.52). Although the median fecal sIgA level was higher in the HD group, the difference between the two groups was not statistically significant based on the Mann–Whitney U test (p=0.72).

**Table 2.** Comparison of fecal sIgA levels between groups

| Variables | HD (n=16) | Healthy controls (n=16) | p value |
|---|---|---|---|
| Fecal sIgA (µg/ml), median (range) | 1551.56 (27.69-35988.75) | 771.87 (31.27-11250.52) | 0.72 |

Data are presented as mean±SD for normally distributed numeric variable and median (min-max). Statistical test: Mann-Whitney U test. sIgA: Secretory immunoglobulin A, HD: Hirschsprung's disease, SD: Standard deviation, Min: Minimum, Max: Maximum

**Table 3** presents the comparison of sIgA levels according to age group and sex between children with HD and healthy controls. In the 0-12 months old group, the median fecal sIgA level was 6271.5 µg/ml (range 95.93-35988.75) in the HD group and 9373.82 µg/ml (range 205.63-10817.06) in the healthy controls, with no statistically significant difference (p=1.00). In the >12-24 months old group, the median fecal sIgA level was 2877.73±3734.98 µg/ml in the HD group and 3888.72±5249 µg/ml in the healthy controls with normally distributed data, which was also not statistically significant (p=1.00). In the >24 months old group, the median fecal sIgA level was 1005.82 µg/ml (range 27.69-24086.25) in the HD group and 365.65 µg/ml (range 33.48-10817.06) in the healthy controls, with no statistically significant difference (p=0.77).

**Table 3.** Comparison of fecal sIgA levels between variables

| Variables | sIgA levels in HD (n=16) | sIgA levels in healthy controls (n=16) | p value |
|---|---|---|---|
| **Age** | | | |
| 0-12 months | 6271.5 (95.93-35988.75) | 9373.82 (205.63-10817.06) | 1 |
| >12 months-24 months | 2877.73±3734.98 | 3888.72±5249 | 1 |
| >24 months | 1005.82 (27.69-24086.25) | 365.65 (33.48-10817.06) | 0.77 |
| **Gender** | | | |
| Female | 648.50 (94.47-24086.25) | 9373.82 (88.61-10817.06) | 0.61 |
| Male | 3267.75 (27.69-35988.75) | 210.37 (31.27-11250.52) | 0.28 |

Data are presented as mean±SD for normally distributed numeric variable and median (min-max) for abnormally distributed numeric variable or n (%) for categoric variable. Statistical test: Mann-Whitney U test. sIgA: Secretory immunoglobulin A, HD: Hirschsprung's disease, SD: Standard deviation, Min: Minimum, Max: Maximum

Based on gender, the median fecal sIgA level in female subjects was 648.50 µg/ml (range 94.47-24086.25) in the HD group and 9373.82 µg/ml (range 88.61-10817.06) in the healthy controls, with no statistically significant difference (p=0.61). In male subjects, the median fecal sIgA level was 3267.75 µg/ml (range 27.69-35988.75) in the HD group and 210.37 µg/ml (range 31.27-11250.52) in the healthy controls, which was likewise not statistically significant (p=0.28).

## DISCUSSION

The main finding of this study was that children with HD without enterocolitis showed a numerically higher median sIgA level than healthy controls, although the difference was

not statistically significant. This finding should therefore be interpreted cautiously as preliminary baseline data rather than evidence of diagnostic utility or a mechanistic role of fecal sIgA in stable HD. Recent literature has increasingly recognized that mucosal immune dysregulation is only one component of the multifactorial pathogenesis of HAEC, together with intestinal dysmotility, microbial dysbiosis, and epithelial barrier dysfunction. Therefore, while the present result may suggest altered mucosal immune activity in HD without overt enterocolitis, it does not establish fecal sIgA as a clinically applicable biomarker and should instead be regarded as hypothesis-generating for future larger and better-controlled studies.[6-8]

The baseline characteristics in **Table 1** were interpreted primarily as descriptive context rather than as main explanatory variables for fecal sIgA differences. The median age was broadly similar between groups, and this age profile remains clinically plausible because, although HD is often recognized early in life, recent studies have shown that diagnosis may still occur beyond infancy with considerable variation in age at presentation.[9,10] Male predominance in the HD group was also consistent with recent reports showing that HD remains more frequent in boys than girls.[10] The higher median body weight observed in the HD group should be interpreted cautiously because body weight in children is strongly age-dependent, and nutritional status in HD may vary substantially across patients. A recent multicenter cross-sectional study further showed that undernutrition is not uncommon in children with HD and that weight-related anthropometric indices may worsen with more extensive aganglionosis.[11] Therefore, age, gender, and body weight in the present study should be regarded as background sample characteristics rather than primary determinants of fecal sIgA variation, although residual confounding related to age structure and nutritional status cannot be completely excluded.

Age- and gender-stratified comparisons are presented in **Table 3**. No statistically significant differences were observed between the HD and healthy control groups within any age category or gender stratum (all $p>0.05$). Because subgroup sizes were small and variability was wide, these findings should be interpreted as descriptive only. In a recent cohort study, fecal SIgA levels in infants differed according to birth mode and breastfeeding status at 3 and 12 months, supporting the concept that early-life SIgA maturation is strongly age- and exposure-dependent.[12] Similarly, Granger et al.[13] showed that stool IgA/sIgA in preterm infants followed a developmental trajectory, with earlier detection in breast milk–fed infants and delayed detection in formula-fed infants, further indicating that intestinal IgA expression varies across early postnatal stages. In addition, Li et al.[14] reported that fecal sIgA responses differed between younger (6-12 months) and older (13-24 months) children, suggesting that age may modify intestinal immune readouts even within relatively narrow pediatric age ranges.

In contrast, no clear sex-related pattern was observed in the present study. This is not entirely unexpected, because recent pediatric IgA-related studies have generally highlighted age, feeding, and perinatal exposure as more important determinants of mucosal IgA variation than gender.[13,14] Therefore, the subgroup analysis in **Table 3** should be viewed mainly as descriptive and exploratory, and the absence of significant differences across age and gender categories should not be interpreted as definitive evidence that these factors are unrelated to fecal sIgA in children with HD.

The absence of a statistically significant difference in fecal sIgA levels between groups should be interpreted with caution. First, although the minimum calculated sample size was achieved, the number of subjects remained small for a biomarker study with substantial biological variability, and the very wide range of fecal sIgA values observed in both groups likely reduced the effective statistical power to detect moderate between-group differences. Therefore, the negative result may reflect limited power and a possible type II error rather than a true absence of biological difference. Second, fecal sIgA is influenced by several major determinants of mucosal immunity that were not systematically controlled in the present study, including feeding-related exposures, microbiota-related factors, environmental conditions, asymptomatic infection, and nutritional status. Recent studies have shown that infant gut SIgA levels vary according to breastfeeding and perinatal exposures, while stool IgA expression also differs between maternal milk-fed and formula-fed infants. In parallel, recent HD/HAEC literature continues to support the role of dysbiosis and impaired mucosal defense as part of a multifactorial disease process rather than an isolated immune abnormality. Therefore, the observed numerical difference in fecal sIgA in the present study cannot be attributed solely to HD status and should instead be regarded as a preliminary signal that requires confirmation in larger and better-controlled studies.[8,12,13,15]

## Limitations

The present study does not establish direct clinical utility for sIgA in children with HD without enterocolitis. Because the observed difference was not statistically significant and the study was limited by small sample size, wide data variability, and uncontrolled confounding factors, fecal sIgA cannot currently be considered a clinically applicable biomarker in this setting. Another limitation is that stool microbiology and viral screening were not performed in the control group. Although control participants had normal physical examination, routine stool analysis, and fecal occult blood testing, asymptomatic infection could not be completely excluded and may have contributed to variability in fecal sIgA levels. In addition, baseline comparability between groups was not optimal. Differences in body weight and heterogeneity in age distribution may have resulted in residual confounding. Because of the limited sample size, formal adjustment for these variables was not considered feasible, and this should be taken into account when interpreting the findings. Nevertheless, the study provides preliminary baseline data on fecal sIgA expression in stable HD without enterocolitis, a population that remains insufficiently characterized. These findings may therefore be useful for generating hypotheses and informing future studies with larger sample sizes, age-stratified analysis, longitudinal follow-up, and more rigorous control of nutritional, microbiota-related, infectious, and environmental confounders.

## CONCLUSION

In this study, fecal sIgA levels were numerically higher in children with HD without enterocolitis than in healthy controls, but no statistically significant difference was observed. Accordingly, the present findings should be interpreted as exploratory baseline data rather than confirmatory evidence of clinical or mechanistic significance. Further studies with larger sample sizes, longitudinal design, age-stratified analysis, and more rigorous control of major confounding variables are needed to clarify the role of fecal sIgA in HD.

## ETHICAL DECLARATIONS

### Ethics Committee Approval

Ethical approval was obtained from the Ethics Committee of Cipto Mangunkusumo National Hospital (Date: 11.10.2024, Decision No: KET-1437/UN2.F1/ETIK/PPM.00.02/2024).

### Informed Consent

Informed consent was obtained from a parent or legal guardian. Where appropriate, age-adjusted assent was also obtained from the child. The inclusion of vulnerable populations in this study adhered to national and international ethical guidelines. Extra care was taken to ensure voluntary participation, understanding, and protection of participant dignity and autonomy.

### Peer Review Process

This manuscript was subject to external peer review.

### Conflict of Interest

The authors declare no conflicts of interest related to this study.

### Financial Disclosure

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### Author Contributions

Concept: JCR, RPT; Design: RPT; Control: RPT; Resources: RPT, YY; Materials: RPT, YY; Data Collection and/or Processing: JCR, VAM; Analysis and/or Interpretation: JCR, RPT, YY; Literature Review: VAM; Writing the Article: VAM; Critical Review: All Authors.

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MHA MEDIHEALTH ACADEMY

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Original Article

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# Diagnostic performance of the Alvarado score for identifying low-risk pediatric patients with suspected acute appendicitis

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## ABSTRACT

**Aims:** Acute appendicitis (AA) remains a major diagnostic challenge in children because of its variable clinical presentation and overlap with other causes of abdominal pain. This study aimed to evaluate the diagnostic performance of the Alvarado score (AS) in children with suspected AA.

**Methods:** This retrospective diagnostic accuracy study included 737 pediatric patients presenting with abdominal pain between January 2019 and December 2023. Patients were classified into the AA and nonspecific abdominal pain (NAP) groups. The diagnostic performance of the AS was assessed using receiver operating characteristic (ROC) analysis. Clinically relevant rule-out and rule-in thresholds based on previously published literature were evaluated.

**Results:** A total of 104 patients were diagnosed with AA and 633 with NAP. The median AS was significantly higher in the AA group than in the NAP group (8 [IQR 7-8] vs 3 [IQR 2-4]; p<0.001). ROC analysis demonstrated excellent diagnostic performance of the AS, with an area under the curve (AUC) of 0.979 (95% CI 0.961-0.996; p<0.001). An AS threshold of ≤4 demonstrated a sensitivity of 98.1% and a negative predictive value (NPV) of 99.6%, whereas a threshold of ≥6 demonstrated a specificity of 96.5% and a positive likelihood ratio of 27.2.

**Conclusion:** The AS demonstrated excellent diagnostic performance and may serve as a useful adjunctive tool for the initial risk stratification of children with suspected AA. In particular, an AS ≤4 was associated with a very low probability of AA and may help identify selected patients who could potentially avoid immediate imaging. Prospective multicenter studies are needed to validate these findings.

**Keywords**: Appendicitis, child, emergency service, diagnostic imaging, decision support techniques

## INTRODUCTION

Acute abdominal pain is one of the most common reasons for pediatric emergency department visits and may result from self-limiting conditions or from acute appendicitis (AA). The diagnosis of AA in children remains challenging because of overlapping clinical features and variable presentation. Inadequate evaluation may lead to negative appendectomy, whereas delayed diagnosis increases the risk of complications.[1,2]

Several clinical scoring systems have been developed to assist in identifying children at risk for AA. These tools are considered sufficiently sensitive to exclude AA in low-risk patients, thereby reducing unnecessary imaging and negative appendectomy rates.[3,4] However, the routine use of abdominal and pelvic computed tomography (CT) in children remains controversial because increased CT utilization has not consistently reduced negative appendectomy rates, while concerns regarding ionizing radiation exposure persist.[5] Ultrasonography (US) is generally recommended as the preferred first-line imaging modality in children because it avoids radiation exposure. Nevertheless, its diagnostic performance may vary depending on operator experience, patient characteristics, and appendix visualisation rates.[6,7]

The Alvarado score (AS) is a simple, widely used clinical tool that incorporates symptoms, physical examination findings, and laboratory parameters.[8,9] Despite its practicality, the diagnostic accuracy of the AS in pediatric populations remains variable across studies. A meta-analysis by Bai et al.[10] reported pooled sensitivity and specificity values of 76% and 71%, respectively, suggesting moderate overall diagnostic accuracy and supporting its role primarily as an adjunctive diagnostic tool.

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Therefore, this study aimed to evaluate the diagnostic performance of the AS in a pediatric population and to investigate clinically applicable cut-off values for risk stratification in children with suspected AA.

## METHODS

### Ethics

The study protocol was approved by the Ethics Committee for Non-interventional Clinical Researches at Çanakkale Onsekiz Mart University (Date: 10.12.2025, Decision No: 2025-382) and conducted in accordance with the Declaration of Helsinki.

### Study Design and Setting

This retrospective diagnostic accuracy study was conducted at the pediatric emergency and pediatric surgery department between January 2019 and December 2023.

### Patients

A total of 737 children were included in the final analysis (**Figure 1**). Children aged 0-17 years presenting with acute abdominal pain and evaluated for suspected AA were eligible. Patients with incomplete clinical or laboratory data, incomplete medical records, or an alternative acute abdominal diagnosis were excluded from the study. Because this was a retrospective diagnostic accuracy study, all consecutive eligible patients presenting during the predefined study period were included in the analysis. Therefore, no formal a priori sample size calculation was performed.

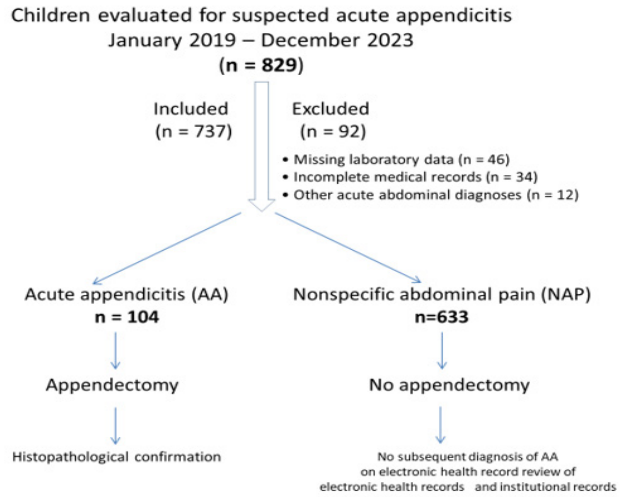



**Figure 1.** STARD flow diagram of patient selection

STARD flow diagram illustrating patient selection, reasons for exclusion, final study groups, and the reference standards used for diagnosis. AA: Acute appendicitis, NAP: Nonspecific abdominal pain

The patients were then divided into two groups: nonspecific abdominal pain (NAP) (n=633) and AA (n=104). AA was histopathologically confirmed in patients with appendectomy. Negative appendectomy was defined by postoperative histopathologic examination as the absence of inflammation in the appendix wall or polymorphonuclear leukocytes, including normal appendix and lymphoid hyperplasia. Non-operated patients were classified as NAP if they improved without surgery and had no subsequent diagnosis of AA based on review of the available electronic health records and institutional medical records.

The patient data analysed included age, gender, presence of migratory pain, anorexia, nausea/vomiting, right lower quadrant (RLQ) tenderness, and rebound pain. Body temperature, white blood cell count (WBC), neutrophil and lymphocyte percentages, neutrophil-to-lymphocyte ratio (NLR), C-reactive protein (CRP) levels, and US findings were also analysed. AS was calculated according to the Alvarado scoring system and is summarised in **Table 1**.

**Table 1.** The Alvarado scoring system (ASS)

| Component | | Score |
|---|---|---|
| Clinical symptoms | Migration of pain to the RLQ | 1 |
| | Anorexia | 1 |
| | Nausea/vomiting | 1 |
| Physical examination | RLQ tenderness | 2 |
| | Rebound tenderness | 1 |
| | Fever (>37.5 °C) | 1 |
| Laboratory findings | Leukocytosis (WBC >10×10$^9$/L) | 2 |
| | Elevated neutrophil count >75% | 1 |
| Total | | 10 |

RLQ: Right lower quadrant, WBC: White blood count

### US Evaluation

US reports were retrospectively reviewed and categorised as positive or negative based on the documented radiological findings. A normal appendix was defined as a compressible, blind-ended tubular structure with a maximal outer diameter of <6 mm, displaying preserved wall stratification and no evidence of hyperemia or surrounding inflammatory changes. An inflamed appendix was defined as a non-compressible tubular structure with a maximal outer diameter ≥6 mm, with or without the presence of an appendicolith. Secondary sonographic signs of appendicitis included increased echogenicity of the periappendiceal fat, localised free fluid, phlegmon, abscess formation, or an inflammatory mass.[11-13]

US was not performed routinely in all patients. The decision to perform US was made at the discretion of the treating pediatric surgeon or emergency physician according to the patient's clinical presentation and routine clinical practice. All US examinations were performed by board-certified general radiologists as part of routine clinical care.

A US was classified as negative when a normal appendix was clearly visualised without sonographic features of inflammation, or when the appendix was not visualised. No secondary signs suggestive of appendicitis were documented.[14-16] US was considered positive when either an inflamed appendix was directly visualised or when secondary inflammatory findings were present, regardless of direct visualisation of the appendix.

### Statistical Analysis

The data were analysed using the Statistical Package for the Social Sciences (SPSS) 18.0 software (SPSS Inc., Chicago, IL). The normality of continuous variables was assessed using the Shapiro–Wilk test. As the variables were not normally distributed ($p<0.05$), continuous variables were expressed as median and interquartile range (IQR), along with minimum

and maximum values.The Mann–Whitney U-test was used for non-normally distributed variables. Categorical variables were analysed using the chi-square test. A two-tailed p-value of <0.05 was considered statistically significant.

The diagnostic performance of the AS was evaluated using cutoff values of ≤4 and ≥6, selected based on previously published literature evaluating rule-out and rule-in thresholds in pediatric appendicitis. Sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV), overall accuracy, and likelihood ratios (LR+ and LR–) were calculated using cross-tabulation analysis. Receiver operating characteristic (ROC) curve analysis was conducted to assess the discriminatory performance of the AS, and the area under the curve (AUC) with 95% confidence intervals (CI) was reported.

## RESULTS

A total of 737 patients were included, comprising 633 with NAP and 104 with AA. As shown in **Table 2**, patients with AA were significantly older (median 12 (IQR 9-14.75) vs 10 (IQR 7-13) years; p=0.001), and male gender was more common in this group (63.5% vs 49.1%; p=0.007).

**Table 2.** Demographic characteristics of the study population

| Variable | NAP (n=633) | AA (n=104) | p value |
|---|---|---|---|
| Age, years (median, IQR) | 10 (7-13) | 12 (9-14.75) | 0.001† |
| Male gender, n (%) | 311 (49.1%) | 66 (63.5%) | 0.007‡ |

NAP: Nonspecific abdominal pain, AA: Acute appendicitis, IQR: Interquartile range, † Mann–Whitney U test, ‡ Pearson Chi-square test

Laboratory parameters and clinical scores demonstrated marked intergroup differences (**Table 3**). The median AS was significantly higher in the AA group (8 (IQR 7-8)) than in the NAP group (3 (IQR 2-4); p<0.001). Leukocyte count, neutrophil percentage, NLR, and CRP levels were significantly elevated in AA (all p<0.001), whereas lymphocyte percentage was significantly lower (p<0.001).

**Table 3.** Comparison of laboratory parameters and Alvarado scores between the NAP and AA groups

| Variable | NAP (n=633) median (IQR) | AA (n=104) median (IQR) | p value |
|---|---|---|---|
| Alvarado score (AS) | 3 (2-4) | 8 (7-8) | <0.001 |
| WBC (×10$^9$/L) | 10.03 (7.8-12.5) | 14.59 (12.1-17.4) | <0.001 |
| Neutrophil (%) | 68.5 (55.2-80.1) | 81.3 (70.2-87.4) | <0.001 |
| Lymphocyte (%) | 21.8 (12.4-33.7) | 11.0 (6.9-18.6) | <0.001 |
| NLR | 3.13 (1.64-6.57) | 7.16 (4.08-12.49) | <0.001 |
| CRP (mg/dl) | 0.36 (0.10-1.50) | 3.97 (1.31-10.14) | <0.001 |

NAP: Nonspecific abdominal pain, AA: Acute appendicitis, IQR: Interquartile range, WBC: White blood cell count, NLR: Neutrophil-to-lymphocyte ratio, CRP: C-reactive protein. Values are presented as median (25$^{th}$–75$^{th}$ percentile [IQR]).

ROC curve analysis demonstrated high diagnostic performance of the AS, with an AUC of 0.979 (95% CI 0.961-0.996; p<0.001) (**Figure 2**).

Using previously reported clinically relevant thresholds, an AS value of ≤4 demonstrated a sensitivity of 98.1% and NPV of 99.6%. Given the relatively low prevalence of AA in the study cohort, the observed NPV should be interpreted in the context of disease prevalence. In contrast, a threshold of ≥6 demonstrated a specificity of 96.5% and a positive likelihood ratio (LR+) of 27.2. Detailed diagnostic performance metrics, including sensitivity, specificity, positive and NPVs, likelihood ratios (LR+ and LR–), and overall accuracy, are presented in **Supplementary Table S1**.

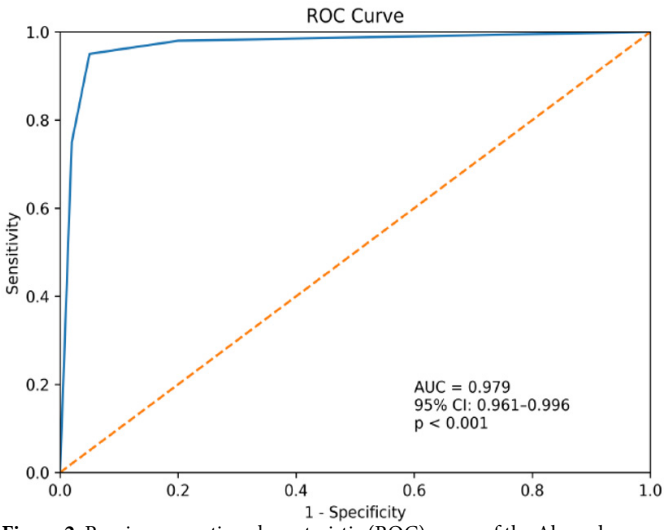



**Figure 2.** Receiver operating characteristic (ROC) curve of the Alvarado score (AS)

The ROC analysis demonstrated high diagnostic performance of the AS, with an area under the curve (AUC) of 0.979 (95% CI 0.961–0.996; p<0.001). The dashed diagonal line represents the line of no discrimination.

Among patients who underwent US (n=240), imaging demonstrated a sensitivity of 91.1% (95% CI 83.2-96.1) and a specificity of 38.5% (95% CI 13.9-68.4).

## DISCUSSION

In the present study, the AS demonstrated high diagnostic performance for identifying AA in children, with an AUC of 0.979. An AS threshold of ≤4 was associated with high sensitivity and a very high NPV, whereas a threshold of ≥6 demonstrated high specificity and a strong positive likelihood ratio. These findings support the potential utility of the AS as a risk stratification tool in the initial evaluation of children with suspected AA. The excellent diagnostic performance observed in our study was higher than that reported in most previous pediatric validation studies. This discrepancy may be explained by differences in patient selection, disease spectrum, and study design. In addition, the retrospective nature of the study, selective patient inclusion, and the use of different reference standards for operated and non-operated patients may have introduced selection bias, spectrum bias, and partial verification bias, potentially contributing to the observed diagnostic performance. Therefore, the reported diagnostic accuracy should be interpreted with appropriate caution until validated in prospective multicenter cohorts.

Previous studies have reported variable diagnostic performance of the AS in children. Reported sensitivities of approximately 72% and specificities of 79% suggest that an AS ≥6 alone may be insufficient for definitive diagnosis of AA, whereas lower scores may help identify patients at lower

**Supplementary Table S1.** Diagnostic performance of the Alvarado score at clinically relevant rule-in and rule-out thresholds

| Test | Cut-off/ definition | Sensitivity % (95% CI) | Specificity % (95% CI) | LR+ | LR– | PPV % (95% CI) | NPV % (95% CI) | Accuracy % (95% CI) | AUC (95% CI) |
|---|---|---|---|---|---|---|---|---|---|
| AS | ≥6 (rule-in) | 95.2 (89.1–98.4) | 96.5 (94.8–97.8) | 27.2 | 0.05 | 81.8 (73.8–88.2) | 99.2 (98.1–99.7) | 96.3 (94.7–97.6) | 0.979 (0.967–0.991) |
| AS | ≤4 (rule-out) | 98.1 (93.2–99.8) | 78.7 (75.3–81.8) | 4.61 | 0.02 | 43.0 (36.6–49.6) | 99.6 (98.6–100.0) | 81.4 (78.4–84.2) | 0.979 (0.967–0.991) |

AS: Alvarado score, LR+: Positive likelihood ratio, LR–: Negative likelihood ratio, PPV: Positive predictive value, NPV: Negative predictive value, CI: Confidence interval, AUC: Area under the curve. The 95% confidence intervals were calculated using the exact (Clopper–Pearson) binomial method.

risk.[4,17] In addition, a comprehensive meta-analysis evaluating the diagnostic value of the AS in children concluded that the score demonstrates moderate overall accuracy and should primarily be considered an adjunctive diagnostic tool rather than a standalone diagnostic method.[10]

Although other clinical scoring systems, such as the Pediatric Appendicitis Score (PAS) and the Appendicitis Inflammatory Response (AIR) score, have also demonstrated good diagnostic performance in children, we focused on the AS because it remains one of the most widely used and externally validated clinical prediction tools in both pediatric and adult populations. The PAS was specifically developed for pediatric patients, whereas the AIR score incorporates inflammatory biomarkers and may provide improved specificity in selected populations.[17,18,26] However, comparative studies have generally shown similar overall diagnostic performance among these scoring systems, and no single score has consistently demonstrated clear superiority across different clinical settings. Therefore, the AS remains a practical and clinically applicable tool for the initial evaluation of children with suspected AA.

Compared with these reports, the diagnostic performance observed in the present study was notably higher. An AS threshold of ≤4 was associated with high sensitivity (98.1%) and a high NPV (99.6%), suggesting that low AS values may help identify children with a low probability of AA. However, the high NPV should be interpreted in the context of the relatively low prevalence of AA in the study population, since predictive values are prevalence-dependent. Therefore, the diagnostic performance observed in this study may not be directly generalizable to populations with different disease prevalence. Similarly, a threshold of ≥6 demonstrated high specificity and a strong positive likelihood ratio, supporting its potential utility in identifying higher-risk patients.[18]

Careful physical examination remains a cornerstone of the evaluation of children with suspected AA. Previous studies have emphasized that inadequate physical examination may contribute to unnecessary imaging and radiation exposure.[19] The high diagnostic performance of the AS observed in the present study likely reflects the integration of key clinical findings, physical examination, and laboratory parameters into a structured risk stratification tool. Taken together, these findings suggest that structured clinical assessment remains an important component of pediatric appendicitis evaluation alongside laboratory and imaging findings.

Laboratory parameters also served as important adjunctive findings in our cohort. Leukocytosis, neutrophil predominance, and elevated NLR were significantly associated with AA. Previous studies have similarly reported moderate-to-high diagnostic performance for NLR in pediatric appendicitis.[20]

US remains the preferred first-line imaging modality in children with suspected AA because it avoids ionizing radiation exposure. However, its diagnostic performance may vary depending on operator experience, patient characteristics, and the rate of appendix visualisation.[16,21] In our study, only a subset of patients underwent US evaluation, most likely reflecting clinician-directed selection of patients with higher clinical suspicion. Consequently, the observed diagnostic performance of US should be interpreted cautiously because substantial selection and work-up bias may have been present. Furthermore, the relatively low specificity observed in our cohort may have been influenced by our retrospective classification strategy, whereby examinations with non-visualization of the appendix and no secondary inflammatory findings were categorized as negative. Because non-visualization of the appendix is generally considered a non-diagnostic rather than a negative US finding in pediatric practice, this approach may have underestimated the specificity of US. Together with the operator-dependent nature of US, these methodological considerations likely contributed to the observed diagnostic performance and should be taken into account when comparing our findings with previous pediatric studies.[17,22] Therefore, the diagnostic performance of US reported in this study should not be interpreted as the true diagnostic accuracy of US in children with suspected AA. Rather, it reflects the performance of US within the context of our retrospective study design and selective imaging strategy.

Previous studies have reported that CT does not improve diagnostic accuracy beyond that achieved through history, physical examination, and laboratory findings alone. They emphasised that a negative CT scan does not reliably exclude AA when clinical suspicion is strong.[23,24] At the same time, increasing CT utilization in pediatric populations has not consistently reduced negative appendectomy rates.[5,25] Given the potential long-term risks associated with ionizing radiation exposure in children, selective and clinically guided imaging strategies remain important.

Overall, our findings support the use of the AS as an adjunctive tool for the initial risk stratification of children with suspected AA. However, given the retrospective design and the potential for partial verification bias, these findings should be interpreted with caution.

## Limitations

This study has several strengths. First, it included a relatively large pediatric cohort evaluated using routinely collected clinical and laboratory parameters. Second, the diagnostic

performance of the AS was comprehensively assessed using ROC analysis and clinically relevant rule-out and rule-in thresholds. Third, the study evaluated practical cut-off values that may assist in risk stratification during the initial assessment of children with suspected appendicitis.

However, several limitations should be acknowledged. First, the retrospective single-center design may have introduced selection bias and limits the generalizability of the findings. Second, the use of different reference standards for operated and non-operated patients may have introduced partial verification (work-up) bias. Whereas AA was confirmed histopathologically in patients undergoing appendectomy, non-operated patients were classified based on clinical follow-up and review of medical records. Consequently, the true diagnostic accuracy of the AS cannot be determined with complete certainty, and the reported diagnostic performance should be interpreted in light of this limitation. Third, US was performed selectively rather than systematically in all patients, introducing potential work-up and selection bias in the imaging subgroup analysis. Fourth, the duration of abdominal pain before presentation was not consistently documented because of the retrospective study design and therefore could not be evaluated. Fifth, the relatively low prevalence of AA in the study population may have inflated the NPV, limiting the generalizability of this finding to populations with different disease prevalence.

Finally, the findings have not been externally validated. Prospective multicenter studies with external validation are warranted to confirm the generalizability and clinical applicability of these results.

## CONCLUSION

In this retrospective diagnostic accuracy study, the AS demonstrated high diagnostic performance and may serve as a useful adjunctive tool for the initial risk stratification of children with suspected AA. In particular, an AS of ≤4 was associated with a very low probability of AA and may help identify selected low-risk patients in whom immediate imaging could potentially be deferred. However, given the retrospective design and the use of different reference standards for operated and non-operated patients, these findings should be interpreted with caution. Imaging decisions should remain individualized and integrated with clinical assessment, laboratory findings, and physician judgment. Further prospective multicenter studies are required to confirm these findings before widespread clinical implementation.

## ETHICAL DECLARATIONS

### Ethics Committee Approval

The study protocol was approved by the Ethics Committee for Non-interventional Clinical Researches at Çanakkale Onsekiz Mart University (Date: 10.12.2025, Decision No: 2025-382).

### Informed Consent

This retrospective study used pre-existing anonymized patient data. No additional intervention was performed, and there was no direct patient contact. The study was approved by the Ethics Committee, and the requirement for written informed consent was waived by the ethics committee.

### Peer Review Process

This manuscript was subject to external peer review.

### Conflict of Interest

The author declare no conflicts of interest related to this study.

### Financial Disclosure

The author received no financial support for the conduct or publication of this research.

### Author Contributions

The author is solely responsible for the entirety of conception, execution, analysis, and writing of the manuscript.

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# Imperforate anus with rectopenile fistula: surgical experience with anterior sagittal approach and structured literature review

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## ABSTRACT

Imperforate anus with rectopenile fistula is a rare type of anorectal malformation (ARM) that is associated with diagnostic and surgical complexities due to its infrequent occurrence. We herein revisit this rare anomaly highlighting the advantages with anterior sagittal approach along with systemic review of literature. Two cases of imperforate anus with recto penile fistula underwent staged repair through anterior sagittal approach. In the Structured review using PRISMA's search and reporting principles, search for cases of ARM with recto penile fistula, seven articles describing nine patients with ARM and fistula to the penis. The fistula tract appeared extending anterior in perineum till ventral aspect of penis. All patients had distal rectal pouch below ischial spine thus classifying this anomaly into low variety on invertogram. Three patterns of fistulae to the penis were recognized and most patients underwent staged repair with colostomy. Thus, present report is unique where anterior sagittal approach appeared successful with its advantages for successful repair of this rare anomaly. The occurrence of a distal rectopenile fistula, although anatomically defined as a low ARM, is associated with considerable technical complexities. The anterior sagittal approach appears superior in dissection of the rectourethral plate, especially in cases of a distal penile fistula. The use of cystoscopic assistance in the lithotomy position is useful in obtaining clear anatomical definitions and ensures proper closure of the fistula without compromising urethral integrity.

**Keywords**: Imperforate anus, rectopenile fistula, anterior sagittal approach

## INTRODUCTION

Anorectal malformations (ARMs) encompass a broad group of congenital anomalies due to the abnormal development of the distal hindgut and cloaca.[1-4] Most ARMs are classified according to the location of the rectal pouch in relation to the sphincter complex. However, there are rare ARMs that may cause diagnostic and surgical challenges.[5] Rectopenile fistula is a rare form of ARM that involves the abnormal connection between the rectum and the ventral aspect of the penis, which may be associated with the communication with urethra.[1-8]

A systematic review done by Yang et al.[1] emphasized the anatomical anomalies associated with rectopenile fistula and the necessity of radiologic studies in defining the tract and its relationship to the urethra. Although this form of ARM is classified as a low-type ARMs, the anterior extension through the corpus spongiosum and its proximity to the distal urethra may pose a surgical challenge.[1,4,6]

Although the standard approach in the repair of most ARMs involves the posterior sagittal anorectoplasty, the anterior sagittal approach may be advantageous in certain situations where the distal penile fistula is involved. This approach provides a direct view of the rectourethral plane and aids in the precise dissection and ligation of the fistula.

Two neonates with imperforate anus and distal recto penile fistula were successfully treated using the anterior sagittal approach, emphasizing the significance of surgical planning and the use of intraoperative cystoscopy to assess the integrity of the urethra.

## CASE REPORT

A full-term male newborn weighing 3,300 g was brought to our hospital shortly after birth with the absence of an anal opening. Physical examination showed absent anal opening with well-developed natal cleft and presence of opening on ventral aspect of distal penis with normal external urinary meatus and intact foreskin (**Figure 1**). There was no meconium seen from this opening initially but appeared on

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subsequent examination after 18 h of birth thus suggesting presence of fistulous connection between distal atretic rectum and penile raphe. There was no other congenital abnormality noted from the physical examination and further radiological survey. An invertogram done afterwards revealed that the distal rectal pouch was situated below the ischial spine, which is consistent with a low type of ARM. However, the abnormal location of the fistula on the penile shaft prompted us to consider the presence of rectopenile fistula. Due to the abnormal location of this fistula and considering the risk of contamination or injury to adjacent structures, a diverting divided sigmoid loop colostomy was performed in the neonatal period.

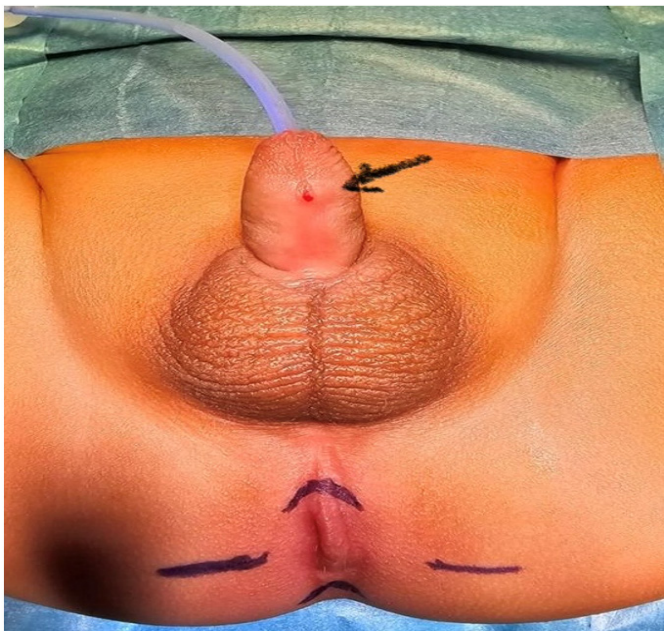


**Figure 1.** Distal recto penile fistula with imperforate anus

A distal loopogram at 4 weeks of age confirmed the presence of a rectopenile fistula that extends towards the ventral penile shaft without any communication with urethra (**Figure 2**).

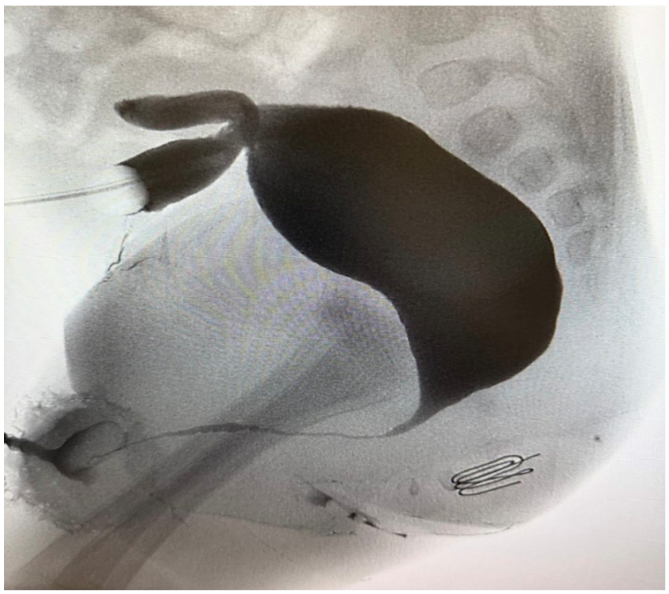


**Figure 2.** Distal Loopogram showing distal recto penile fistula with no urethral communication

Child underwent definite repair in form of anterior sagittal anorectoplasty (ASARP) at age of 8 weeks. The procedure was performed in lithotomy position and cystourethroscopy confirmed absence of any urethral communication. A catheter was introduced into the patient's urethra, and the fistula tract was catheterized with 3f catheter to aid in identification during dissection. An anterior sagittal incision in the perineum, through the midline, and towards the penile raphe was made. Skin flaps were raised to expose the patient's fistula tract. The patient's fistula was then carefully dissected from the surrounding tissues and separated from the corpus spongiosum. During procedure transillumination and direct visualization of penile urethra by intra operative cystoscopy facilitated in dissection where fistula trat appeared near spongiosum. After dissecting the patient's fistula, the procedure continued posteriorly. Muscle complex and site of neoanus were mapped by muscle stimulator followed by division of the perineal body in the midline. The distal rectal pouch was identified, mobilized and anorectoplasty was performed after flush ligation of fistula with rectum along with reconstruction of perineal body (**Figure 3**).



**Figure 3.** Anterior saggital anorectoplasty

The recovery was uneventful. The procedure for anal dilatation was started as per the standard protocol for postoperative anal dilatation. The colostomy was closed eight weeks later after ascertaining satisfactory healing and caliber of the neoanus. The patient has been followed for a period of one year. The child has a satisfactory neoanus, and the child is passing one to two bowel movements daily. There is no evidence of fecal soiling. The result has been satisfactory for the age group.

## CASE 2

A full-term male newborn weighing 2800 g presented with absence of an anal opening. On physical examination, there was a well-developed natal cleft with no visible anal orifice. An abnormal external opening was noted at the penoscrotal junction with presence of meconium at tip. External urethral meatus appeared normal in position with passage

of clear urine in good stream and normal phallus. No other congenital anomalies were identified on systemic radiological and cardiac examination. An invertogram performed subsequently demonstrated that the distal rectal pouch was located below the level of the ischial spine, consistent with a low type of ARM. However, the presence of meconium at the penile tip suggested a rectourethral fistula, most likely communicating with the distal urethra and hence a diverting sigmoid loop colostomy was performed in the neonatal period.

At 4 weeks of age, a distal loopogram confirmed the presence of fistula between distal rectum extending all the way anteriorly over ventral aspect of penis suggestive of rectopenile fistula without communication with urinary tract.

Definitive repair was planned at 8 weeks of age. The patient underwent ASARP in lithotomy position. Intra operative cystourethroscopy was performed, which confirmed absence of any communication with penile urethra. A catheter was placed in the urethra for identification, and the fistula tract was cannulated with a fine catheter to aid intraoperative dissection.

An anterior sagittal incision was made in the perineum extending toward the penoscrotal junction. Careful dissection was carried out to identify and isolate the fistula tract. The tract was meticulously separated from the surrounding tissues, including the corpus spongiosum and urethra, with the aid of transillumination and intraoperative cystoscopic guidance to prevent urethral injury. Following complete mobilization, the fistula was ligated and divided close to the rectal pouch. The muscle complex was identified using a muscle stimulator, and the perineal body was divided in the midline. The distal rectal pouch was mobilized adequately and brought down through the muscle complex to create a neoanus. Reconstruction of the perineal body was performed. The postoperative period was uneventful. Anal dilatation was initiated as per standard protocol. The colostomy was closed after 8 weeks, following confirmation of good healing and adequate caliber of the neoanus.

The child has been followed for two years postoperatively. At follow-up, the patient has a well-formed neoanus with satisfactory bowel function, passing one to two stools per day. There is no evidence of fecal soiling or urinary complications. Ultrasound scan showed normal kidneys and urinary bladder with no residual urine after voiding. The functional outcome has been appropriate for the age.

## METHODS

A structured literature search guided by PRISMA search and reporting principles of ARMs with associated rectopenile fistula was carried out by doing an extensive literature search both in Scopus and PubMed databases to identify relevant studies published in English from January 1992 to November 2025. The following keywords were used for the search: “anorectal malformation” OR “imperforate anus” and “penile fistula” or “rectopenile fistula.”

The titles and abstracts of all identified studies were reviewed to assess eligibility. If the relevance of a study could not be clearly determined from the abstract, the full text of the study was reviewed. The reference lists of the selected studies were manually searched to identify relevant studies.

Studies that had reported the presence of patients with ARM and the extension of the fistulous tract from the rectal pouch to the penis, especially involving the corpus spongiosum, with or without the involvement of the urethra, were included for the study. Articles that had focused on the clinical presentation, radiologic evaluation, and surgical management of the condition were also included. On the other hand, review articles, editorials, and studies that did not provide enough clinical and radiologic detail were excluded from the study.

Five authors independently reviewed the selected studies and extracted the relevant information using a standardized data collection form. The extracted variables included the clinical presentation, initial diagnosis, radiologic evaluation, surgical management, and the outcomes.

## RESULTS

Out of the 58 potentially relevant articles identified from the initial database search, four studies met the predefined inclusion criteria after the titles and abstracts had been scrutinized. Another two relevant reports were found from the manual review of the reference lists of the studies that had already been scrutinized. This brought the total number of studies included in the final analysis to seven with 9 patients diagnosed with recto penile fistula. In all the patients, the rectal pouch was situated below the ischial line, a feature typical of low-type ARM. However, the anatomical arrangement of the fistulous tract was different for different patients.

In three patients, the fistulous tract was from the rectum to the anterior urethra without any external skin opening. In one patient, the fistulous tract ran along the corpus spongiosum to the ventral surface of the penis without any urethral communication. In the remaining five patients, the fistulous tracts were to the ventral penile surface and had direct communication with skin.

Most of the patients underwent staged surgical management except two cases where cut back was performed in one case and other patient underwent limited posterior sagittal anorectoplasty. Despite the low anatomical level of the rectal pouch, the initial procedure in seven of the nine patients in literature consisted of the performance of a diverting colostomy because of the uncertainty of the complex course of the fistula tract. Definitive repair of the fistula involved the use of perineal techniques in six patients, the anterior sagittal technique in one patient, and the posterior sagittal technique in another two cases.

In two patients, the distal tract of the fistula was left in situ because of the absence of any urethral connection. Five patients presented with associated congenital anomalies, which included congenital heart disease, bifid scrotum,

solitary kidney, hypospadias, undescended testis, and hydronephrosis.

## DISCUSSION

Distal rectopenile fistula represents one of the rarest of the already heterogeneous group of ARMs and is considered as separate entity from rectoperineal fistula owing to its more anterior extent in close proximity to corpus spongiosum and penile urethra.[1-4,6] Although the incidence of ARMs is approximately 1 in 4,000 to 5,000 live births, fistulous extension to the corpus spongiosum of the penis is extremely rare, with handful of cases reported in English literature till date (**Table**).[1] The rarity of this condition, combined with its unusual anatomical course, often leads to diagnostic uncertainty, delay in recognition, and difficulty in surgical management. The current report adds two new cases successfully treated using the anterior sagittal approach for anorectoplasty (ASARP) with the aim to make physicians aware about this uncommon anomaly along with need for thorough examination for its diagnosis and advantages of anterior sagittal approach.

The hallmark of rectopenile fistula is the anterior extension of the rectal pouch into the corpus spongiosum, with or without fistula with urethra. Yang et al.[1] classified this anomaly with type 1 characterized by fistula between the rectum and the anterior urethra without any external cutaneous opening. In type 2 the fistulous tract passes through the corpus spongiosum to open on the ventral penile skin without any connection to the urethra.[1] In type 3, the tract communicates with both the urethra and the ventral penile skin via a short common channel.[1] In present and reported cases although fistula follows variable course, the rectal pouch is anatomically always situated below the ischial line and hence this rare anomaly is classified as a low-type malformation.

The embryological basis for the formation of recto penile fistula is not well understood. Normally, the cloaca is divided in the embryonic period by the formation of the descending urorectal septum into the anterior urogenital sinus and the posterior anorectal canal. Abnormal formation or improper alignment of the cloacal septum during embryonic development may result in the persistence of an accessory tract that runs anteriorly to the developing urethral plate.[1,8] Another theory for the formation of recto penile fistula is that the deep recto cutaneous tract is incorporated partially into the spongy urethra during the development of the penis. The fact that the tract is always located in the corpus spongiosum points to the developmental association with the formation of the urethra.[1] Whether or not this is a variant of the rectourethral fistula or an independent recto cutaneous tract with secondary involvement of the urethra is not well understood. However, the fact that the same three anatomical types are reproduced in the different cases points to the possibility that the same developmental process is involved.

One of the main difficulties in the management of distal recto penile fistula is preoperative diagnosis. Invertogram usually shows presence of low anomaly and hence it is important to carefully examine the perineal area and penis. As experienced in present cases presence of small opening on the ventral aspect of the penile shaft or meconium staining on the median raphe as experienced in present cases may

**Table.** Summary of reported cases

| Authors | Year | Age at diagnosis | Type of fistula | Level of distal rectum from skin | Surgical intervention | Management of fistula |
|---|---|---|---|---|---|---|
| Yang G[1] | 2016 | 4 hours | Parallel to the urethra from the rectal pouch and ventral penile skin | Below the ischium | One stage PSARP | Ligated flush with rectum |
| Ohno et al.[2] | 2008 | 6 months | Parallel to the urethra from the rectal pouch to the spongy urethra | Below the ischium (1 cm) | Transverse colostomy, ASARP, colostomy clousure | Severed from the rectum and ligated |
| Kumar et al.[3] | 2005 | 18 months | Between the anal canal and the skin in the peno-scrotal junction, with a small portion of common channel in the penile urethra | | Anoplasty, colostomy + fistula excision | Removed |
| Shah et al.[4] | 2003 | 9 months | From the rectum to the ventral aspect of the penis, no communication with urethra | Low | Transverse colostomy, PSARP, colostomy closure | Ligated, distal part was kept undisturbed |
| Currarino et al.[7] | 1999 | 9 months<br>2 days | Extending from rectum to cutaneous orifice near the penoscrotal junction, with communication with the bulbar urethra<br>A long rectocutaneous fistula open on the undersurface of the penis, communicating with the bulbar urethra | Below the ischial line<br>Below the ischial line | Perineal anoplasty, descending colostomy<br>Colostomy, sacroperineal rectal pull-through with ligation of rectal fistula, colostomy closure, | Fistula excision<br>Excision of urethrocutaneous fistula |
| Takamatsu et al.[6] | 1993 | 11 months<br>Unknown | Fistula between the anorectum and anterior urethra<br>Fistula between the anorectum and anterior urethra | Below the ischial line<br>Below the ischial line | Sigmoid colostomy, revision of fistula and perineal anoplasty<br>Sigmoid colostomy, revision of fistula and perineal anoplasty | |
| Asano et al.[5] | 1983 | 3 months | Fistula from rectum and open in the ventral surface of the penis, communication with urethra | Under the skin | Cutback procedure, excision of the fistula | |

be suggestive. A review of literature suggests that urinary symptoms in form of urine discharge through the fistula or presence of contrast through fistula tract on retrograde urethrogram or distal loop gram provides clues to the diagnosis.[1-4,8,9] It has been noted that the fistulous opening may be mistaken for a median raphe cyst, a small defect in the epithelium, or an inflammatory process However, absence of this symptom does not rule out a small connection and hence careful insight into this rare entity is important for pre-operative diagnosis.

As experienced in present and reviewed studies distal colostograms help to locate the course of the fistula tract and the type of fistula.[1,4,7,9] Voiding cystourethrograms has been considered in reported cases and has been found to be of special value in identifying the connection to the urethra and pinpointing the site of connection.[1-4] In the current cases, detailed contrast studies have enabled us to map the fistulous tract in the corpus spongiosum before surgery and intra operative cystoscopy confirmed absence of urethral fistula. Associated anomalies in rectopenile fistula patients are similar to those in other low-type ARMs. Renal anomalies, congenital heart disease, bifid scrotum, and undescended testes are the associated anomalies reported in the patients with rectopenile fistula. These findings correlate with the overall spectrum of associated anomalies in patients with ARM.

The surgical approach to distal rectopenile fistula should be tailored to the individual case based on the pouch level and anatomy of the tract. Most of reported cases have undergone definitive repair by the posterior sagittal anorectoplasty (PSARP) procedure which remains gold standard for most ARMs because of its excellent visualization of the rectal pouch and pelvic musculature.[1-6,8,10] The surgical experience in present cases suggest that prone position during PSARP often makes dissection of fistula tract visually challenging owing to its anterior extension towards penile urethra. Hence the present two cases underwent definitive repair by anterior sagittal approach. In present cases it was experienced that with patient in lithotomy position a midline incision made anteriorly provides direct access to the ventral structures, including the corpus spongiosum and fistulous tract enabling precise dissection of fistula tract. Intra operative cystoscopy which was feasible only because patient is in lithotomy position and it was observed that cystoscopy remains beneficial in confirming urethral integrity and can aid in dissection of fistula in case it is close proximity to urethra. Additionally, this approach preserves the posterior sphincter complex, which would theoretically minimize damage to muscle fibers critical for urinary control. In our series, ASARP offered excellent visualization of the tract in the corpus spongiosum, allowing safe separation from the urethra. The straight-line approach to the lesion minimized tissue manipulation, making the procedure easier to control.

As experienced in present cases several technical considerations like careful dissection of fistula off urethra or corpus spongiosum with flush ligation with distal rectal pouch along with identification and reconstruction of neo anus in muscle complex results in satisfactory recovery. Although the anomaly is characterized as low variety based on radiological classification but owing to the course the fistula and technical challenges related to surgical repair it should be considered as intermediate anorectal anomaly and hence as reported in literature and experienced in present series staged repair with diverting colostomy remains surgical approach of choice.

Functional results in rectopenile fistula seem similar to those in other low-type ARMs. The reported patients show satisfactory bowel control in accordance with the low type of the lesions. The preservation of the posterior sphincter complex during the anterior sagittal approach might play a positive role in the continence mechanism. Urinary function is also an important aspect in rectopenile fistula repair, as the dissection is close to the urethra. In the reported cases, the urinary stream was normal without any signs of urethral stricture or leakage.

Rectopenile fistula should be differentiated from other low variants of ARM, such as perineal fistula, rectourethral fistula and H-type fistula.[1,9] The key differentiating point is the ventral position of the fistula on the penis. A misdiagnosis of simple perineal fistula may result in inadequate surgical exposure and incomplete excision of the fistula tract. The literature is comprised of individual case reports and small series. The lack of standardization in reporting, surgical methods, and follow-up duration makes generalization difficult. There is a paucity of information regarding long-term continence and urinary function into adolescence. Perhaps multicenter registry data will be required to provide a meaningful analysis of outcomes due to the rarity of this condition. With the available evidence and our experience, a practical approach is to carefully clinically evaluate all patients with imperforate anus, suspect those with a ventral penile discharge of meconium, and use imaging techniques to carefully delineate the anatomy of the tract.[1-6,9] Finally, a single versus staged anterior sagittal approach should be considered in those with low lesions and well-delineated anatomy. Staged repairs are best considered in those with undetermined anatomy or complex urethral involvement.

## CONCLUSION

In summary, distal rectopenile fistula is an extremely rare anomaly of ARM that is surgically correctable. It is essential that its anatomical characteristics are well appreciated and that radiologic delineation of this anomaly is carefully performed. The anterior sagittal approach can be considered as alternative approach in selected patients with low-type recto-penile fistulas without urethral communication.

## ETHICAL DECLARATIONS

### Peer Review Process

This review was externally peer-reviewed.

### Conflict of Interest

The authors declare no conflicts of interest.

### Financial Disclosure

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## Author Contributions

Concept: DM; Data Collection and/or Processing: GJ, VG; Analysis and/or Interpretation: DM, GJ, VG; Literature Review: DA, MR; Writing the Article: VG; Critical Review: DM, DA, GJ, MR, VG.

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MHA MEDIHEALTH ACADEMY

Surgery on Children

Case Report

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# The importance of perineal examination in adolescent girls presenting with abdominal pain: a case report[a]

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## ABSTRACT

All patients presenting with abdominal pain should be evaluated using a comprehensive approach, including perineal examination in adolescent girls. We report the diagnostic and treatment course of a 17-year-old girl with recurrent abdominal pain who had previously been hospitalized twice with a preliminary diagnosis of acute appendicitis but was not operated on due to family refusal. On admission to our clinic, laboratory findings (WBC, CRP, procalcitonin) were normal, and abdominal ultrasonography showed no appendicitis, although a 2 cm pelvic free fluid collection raised suspicion of pelvic inflammatory disease. Perineal examination revealed poor hygiene and foul-smelling, white curd-like vaginal discharge. Vaginal culture identified *Zygosaccharomyces* species, and symptoms resolved after itraconazole therapy. Uroflowmetry showed reduced flow rate and increased bladder capacity, suggesting bladder–sphincter dyssynergia. This case highlights the importance of perineal examination and careful evaluation of urinary symptoms to ensure timely diagnosis and appropriate management.

**Keywords**: Abdominal pain, perineal examination, vulvovaginitis

[a]This study was presented as an oral presentation at the 41st National Pediatric Surgery Congress and the 27th National Pediatric Surgery Nursing Congress, held on 7-10 November 2024 in Nevşehir, Turkiye

## INTRODUCTION

Abdominal pain is a common complaint in the pediatric and adolescent population and requires careful evaluation of its localization, associated findings, and underlying causes. All patients presenting with abdominal pain should be approached in a holistic manner. Therefore, appropriate and integrated interpretation of clinical and laboratory findings is essential to establish an accurate diagnosis, guide medical or surgical management, and prevent unnecessary investigations.[1,2] In addition to the assessment and follow-up of patients with abdominal pain, nurses and physicians play a crucial role in the selection, implementation, and optimization of both pharmacological and non-pharmacological pain management interventions.[2]

In most cases, the cause of abdominal pain is functional gastrointestinal disorders; however, conditions such as urogenital system anomalies and lower respiratory tract infections are also among the significant etiologies of abdominal pain.[1] At this point, it should be emphasized that perineal examination should not be neglected in adolescent girls. In this case report, we aimed to evaluate the diagnostic, follow-up, and treatment process of a 17-year-old girl who presented to our clinic with abdominal pain, in order to raise awareness and contribute to appropriate and timely interventions in clinical practice.

## CASE

A 17-year-old girl with recurrent hospital admissions for abdominal pain, persisting for approximately six months, had been hospitalized approximately one month earlier in two different centers with a preliminary diagnosis of acute appendicitis; however, she was not operated on during either admission due to lack of parental consent. As her complaints persisted, she was admitted to our clinic for further evaluation and treatment.

Physical examination revealed no findings suggestive of an acute surgical abdomen. Laboratory tests showed WBC: 6.37 mc/L, CRP: <0.60 mg/L, and procalcitonin: 0.039 ng/ml, all within normal limits. Urinalysis revealed increased bacteria (14) and amorphous material (98), but no erythrocytes, leukocytes, or epithelial cells. Abdominal ultrasonography demonstrated no signs of appendicitis, but approximately 2 cm of free fluid was noted in the pelvic cavity, suggesting pelvic inflammatory disease in correlation with the clinical findings.

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Review of her previous records revealed that no perineal examination had been performed during prior evaluations. Perineal examination at our clinic showed poor hygiene and abundant foul-smelling, white, curd-like vaginal discharge. The patient also reported intermittent urination. Vaginal culture yielded *Zygosaccharomyces* species. Following itraconazole treatment, the discharge resolved and hygiene improved. Itraconazole (Itraspor®; Nobel İlaç Sanayii ve Ticaret A.Ş., İstanbul, Turkiye) was administered at a dose of 200 mg once daily for three days, in accordance with the dosing regimen commonly used for vulvovaginal infections.

Uroflowmetry performed on the first day of hospitalization showed decreased urine flow rate and an increased bladder capacity compared with age norms (**Figure 1**). These findings indicated impaired voiding with low maximum and average flow rates, suggesting a functional voiding disturbance. A repeat test on the third day demonstrated normalization of bladder capacity, though urine flow rate and pattern showed minimal improvement (**Figure 2**). Despite improvement in bladder capacity, the persistence of low flow rates and an irregular flow pattern supported the suspicion of bladder–sphincter dyssynergia. Voiding cystourethrography was planned to evaluate bladder function and possible vesicoureteral reflux; however, the procedure could not be performed due to urine leakage around the catheter after insertion. Similarly, urodynamic testing was not completed because of poor patient cooperation and urine leakage around the urodynamic catheter. Considering the uroflowmetry findings together with the inability to perform urodynamic evaluation, bladder dysfunction with bladder–sphincter dyssynergia was suspected. Accordingly, the patient was instructed on double voiding and toilet training. Follow-up examinations revealed normal perineal findings and complete regression of pelvic fluid on ultrasonography. As uroflowmetry showed no significant improvement, propiverine hydrochloride 5 mg (Mictonorm®; Apogepha Arzneimittel GmbH, Dresden, Germany) daily was initiated. At the one-month follow-up, the patient reported reduced symptoms; bladder capacity was partially normalized, although urine flow rate remained suboptimal. Cystoscopy was recommended, but the patient declined further invasive evaluation.

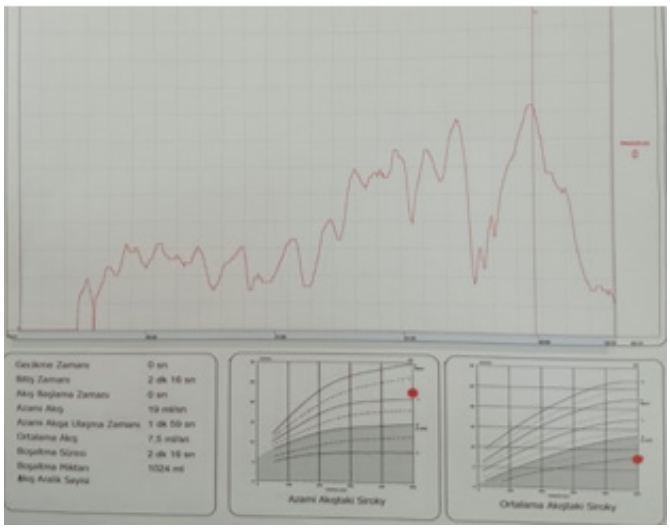


**Figure 1.** Initial uroflowmetry result

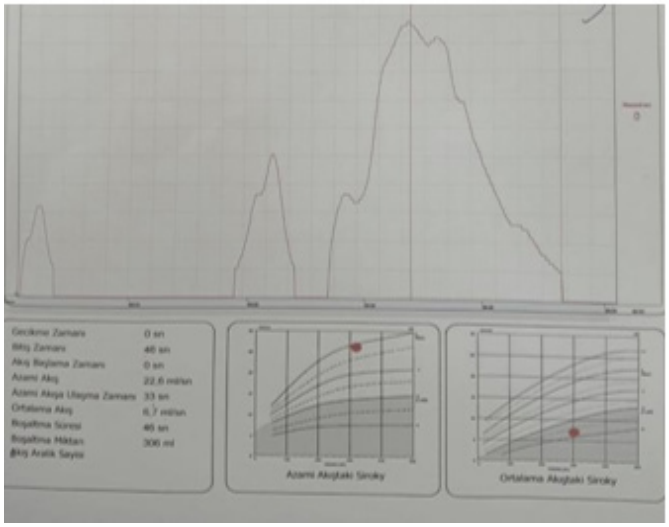


**Figure 2.** Follow-up uroflowmetry result

Written informed consent was obtained from the patient and her legal guardian for the publication of this case report.

## DISCUSSION

Abdominal pain is a common complaint in pediatric medicine and pediatric surgery clinics, and early, accurate diagnosis enables timely treatment and improved prognosis with reduced morbidity.[2] In pediatric patients, symptoms are often non-specific, and a complete physical examination may be challenging. Therefore, evaluation should include a detailed history and thorough physical examination, supported by laboratory tests and imaging when necessary. In adolescent girls, particular attention should be given to menstrual history, sexual activity, vaginal discharge, irregular spotting, and abnormal or painful menstrual bleeding.[3,4]

Inflammation of the vulvar and vaginal tissues and vaginal discharge represent common gynecological problems in childhood. Vulvovaginal inflammation and discharge are common in childhood, with infection risk influenced by age-related mucosal characteristics as well as hygiene and environmental factors.[3,5] During adolescence, rising estrogen levels promote epithelial maturation and increased resistance to infection.[6,7]

Vulvovaginitis may result from sexually transmitted or non–sexually transmitted etiologies, and identification of the causative organism is essential for accurate diagnosis. Normal vaginal flora typically includes *Staphylococcus epidermidis, diphtheroids, lactobacilli*, and *anaerobes*, which generally do not require treatment.[8] Commonly isolated pathogens in vulvovaginitis include *Streptococcus pyogenes*, *Staphylococcus aureus*, *Escherichia coli*, *Proteus mirabilis*, and *Gardnerella vaginalis*.[8]

In our case, detailed history-taking and physical examination of the adolescent girl presenting with abdominal pain revealed a foul-smelling, white, curd-like vaginal discharge. This characteristic discharge was considered an important clinical finding to be taken into account in the differential diagnosis of vulvovaginal infections. Vaginal culture yielded *Zygosaccharomyces* species, a member of the

*Saccharomycetaceae* family, known primarily as a spoilage yeast with notable environmental resistance. Although not classically considered pathogenic, its tolerance to extreme environmental conditions suggests that it may have potential clinical relevance as an opportunistic or colonizing organism.[9] Although no vulvovaginitis cases directly linked to *Zygosaccharomyces* species have been reported, infections caused by other members of the *Saccharomycetaceae* family have been described, indicating that pathogenic involvement cannot be completely excluded despite its rarity.[8,10,11] In our case, detailed history revealed that the patient lived in a rural area and that her family was engaged in agriculture and livestock farming. During physical examination, inadequate adherence to genital hygiene practices was noted. These environmental and hygienic factors may have facilitated colonization or opportunistic overgrowth by the identified yeast species. To date, no infections directly attributed to *Zygosaccharomyces* species have been reported in the literature. However, considering that this organism belongs to the *Saccharomycetaceae* family, previously reported cases have most commonly been treated with antifungal agents such as amphotericin B and fluconazole.[12] In the present case, antifungal therapy with itraconazole was selected although formal antifungal susceptibility testing was not performed, based on susceptibility patterns reported in the literature for *Saccharomycetaceae* species and the antifungal susceptibility profile provided in the culture report, and the observed clinical improvement following treatment suggests—but does not confirm—a possible pathogenic role.

## CONCLUSION

As a result, perineal examination with careful assessment of urinary function and menstrual history is crucial in children presenting with abdominal pain. This case underscores the importance of considering gynecological causes in the differential diagnosis within a multidisciplinary approach. Further well-designed studies are needed to better define vaginal microflora and optimize treatment strategies in symptomatic patients.

## ETHICAL DECLARATIONS

### Informed Consent

Informed consent was obtained from the legal guardians of the pediatric patient described in this report. Where developmentally appropriate, assent was also sought from the child.

### Peer Review Process

This report underwent external peer review.

### Conflict of Interest

The authors declare no conflicts of interest.

### Financial Disclosure

This case report did not receive any financial support.

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Concept: RS, SUT, AŞ; Design: RS, SUT; Control: AŞ; Resources: RS, SUT; Materials: RS, SUT, AŞ; Data Collection and/or Processing: RS, AŞ, SUT; Analysis and/or Interpretation: RS, SUT, AŞ; Literature Review: RS, SUT, AŞ; Writing the Article: RS, SUT, AŞ; Critical Review: AŞ.

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MHA MEDIHEALTH ACADEMY

Surgery on Children

Case Report

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# Chronic ventriculoperitoneal shunt-related costal destruction: a rare case report

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## ABSTRACT

Although ventriculoperitoneal (VP) shunts are widely used in the treatment of pediatric hydrocephalus, they are associated with various infectious and mechanical complications. The most frequently reported problems are shunt infections and catheter obstructions; rare complications, however, can pose diagnostic difficulties. To the best of our knowledge, this study presents one of the rare examples in the literature of costal destruction and osteomyelitis resulting from chronic mechanical contact with a VP shunt catheter. A five-year-old female patient, who had a VP shunt placed on the second postnatal day due to meningomyelocele and hydrocephalus, presented with an abscess in the right flank region and subsequent transparent discharge. Ultrasonography revealed infectious changes in the subcutaneous adipose tissue, while computed tomography showed dilation and lytic destruction in the distal segment of the right 10th rib, along with the shunt catheter located within the rib. The patient underwent surgical exploration, the infected costal segment was excised, and VP shunt revision was performed simultaneously. The patient received appropriate antibiotic treatment for osteomyelitis in the postoperative period and achieved clinical improvement. This case demonstrates that chronic mechanical contact along the course of VP shunts can, albeit rarely, lead to bone destruction and osteomyelitis; it emphasizes the need to consider bone involvement in cases of unexplained local infection findings along the shunt line.

**Keywords**: Ventriculoperitoneal shunt, shunt complications, costal destruction, osteomyelitis

## INTRODUCTION

Ventriculoperitoneal (VP) shunts are the most commonly preferred method in the surgical treatment of pediatric hydrocephalus. However, shunt systems are associated with high complication and revision rates. The literature reports that approximately one-third of shunts require revision within the first year, with the most common complications being infection, proximal or distal catheter obstruction, and mechanical failure.[1-3]

Although abdominal complications are less common, they can present with serious conditions such as peritoneal pseudocysts, bowel perforation, and intestinal obstruction.[4-6] Although cases of thoracic and abdominal migration have been reported, to the best of our knowledge, complications involving bone involvement, particularly those associated with costal osteomyelitis, represent a rarely reported entity in the literature.[7-9] This study presents a case of costal destruction and osteomyelitis resulting from chronic mechanical contact with a VP shunt catheter, discussing important diagnostic and therapeutic considerations.

## CASE

A five-year-old female patient underwent surgery on the second postnatal day for meningomyelocele and hydrocephalus, and a ventriculoperitoneal shunt was placed during the same session. The patient had no prior shunt revisions, and symptoms had been present for approximately two weeks before presentation. She was initially evaluated in the outpatient clinic for an abscess in the right flank region and was treated medically. During this period, the abscess drained spontaneously, and the patient presented again with clear discharge.

Laboratory tests revealed normal white blood cell count and acute phase reactants. Ultrasonographic evaluation showed infectious changes in the subcutaneous adipose tissue. Computed tomography (CT) imaging demonstrated cortical disruption and medullary involvement causing enlargement and lytic destruction in the distal segment of the right 10th rib. Furthermore, the scan revealed that the distal VP shunt catheter segment was closely abutting and partially extending into the affected bone structure (**Figure 1**). The patient underwent surgical exploration; intraoperatively,

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the purulent collection was drained, and the infected costal segment was excised (**Figure 2**). During the same session, the neurosurgery team performed a VP shunt revision. Oral feeding was initiated on the first postoperative day. On the second day, the patient was transferred to the pediatric infectious diseases department. Following the diagnosis of osteomyelitis and the detection of *Pseudomonas aeruginosa* growth in the culture, antibiotic treatment was continued accordingly. The patient showed clinical improvement and was discharged during the postoperative period.

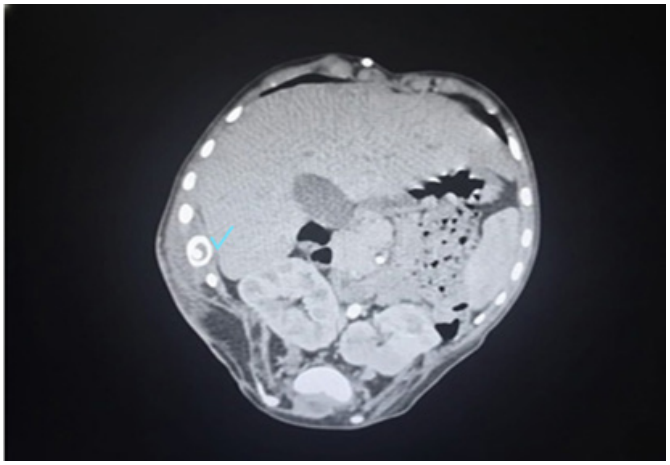


**Figure 1.** Computed tomography (CT) image demonstrating lytic destruction and enlargement of the distal segment of the right 10th rib. The blue arrow highlights the VP shunt catheter positioned within the costal bone.
VP: Ventriculoperitoneal



**Figure 2.** Macroscopic appearance of the excised infected costal segment. Specimen demonstrates the area of chronic mechanical contact where lytic destruction and osteomyelitis developed.

## DISCUSSION

The majority of complications associated with VP shunts manifest as infections and mechanical dysfunctions. Abdominal complications related to the distal catheter are rare but can lead to serious morbidity; pseudocysts, intestinal perforation, and obstruction have been reported as abdominal complications.[4,5] Although thoracic or abdominal organ involvement due to shunt migration has been reported in the literature, to the best of our knowledge, this case represents one of the rare examples of costal bone destruction and osteomyelitis associated with chronic mechanical contact.[7-9]

While instances of catheter migration and subsequent skeletal involvement have been documented as extremely rare events in the broader literature, they provide a crucial comparative reference for understanding such unique mechanical failures. Neto et al.[10] reported a case of a shunt catheter penetrating the sacral bone and extruding through the skin, demonstrating that mechanical contact can cause progressive damage to bone tissue. In our case, lytic destruction and osteomyelitis developing in the costal bone without extrusion represent a rare but possible spectrum of bone complications associated with VP shunts.

In the presented case, the underlying pathogenetic mechanism likely involves a cascade of mechanical and biological factors. Long-term continuous pressure from the shunt catheter against the costal margin may have impaired periosteal microcirculation, resulting in localized periosteal ischemia. Furthermore, repetitive microtraumas induced by respiratory excursions and physical activities could have weakened the structural integrity of the bone cortex over time. Concurrently, the catheter surface acts as a favorable substrate for potential biofilm formation, which can harbor pathogens and predispose the area to a low-grade infection. Based on this clinical picture, we hypothesize that the mechanical damage was the primary event; the progressive periosteal ischemia and cortical disruption severely compromised local tissue defenses, thereby paving the way for a secondary localized infection and subsequent osteomyelitis. As normal laboratory parameters in such low-grade, biofilm-associated infections may delay diagnosis, the use of advanced imaging techniques is crucial in the presence of unexplained local infection findings along the shunt line.[11,12]

Treatment involves the surgical removal of infected tissue, simultaneous shunt revision, and appropriate antibiotic therapy, forming the basis of a multidisciplinary approach.[13-15]

Written informed consent was obtained from the patient's legal guardians for the publication of this case report and any accompanying images.

## CONCLUSION

This case demonstrates that chronic mechanical contact during the course of VP shunts can, albeit rarely, lead to bone destruction and osteomyelitis. Importantly, it also highlights the need to consider underlying bone involvement in patients presenting with persistent or atypical infection along the shunt tract. Therefore, appropriate imaging and surgical intervention should be planned at an early stage.

## ETHICAL DECLARATIONS

### Informed Consent

Informed consent was obtained from the legal guardians of the pediatric patient described in this report. Where developmentally appropriate, assent was also sought from the child.

### Peer Review Process

This report underwent external peer review.

## Conflict of Interest

The authors declare no conflicts of interest.

## Financial Disclosure

This case report did not receive any financial support.

## Author Contributions

Concept: EBK, BT; Design: EBK, BT; Control: EBK, BT; Resources: EBK, BT; Materials: EBK, BT; Data Collection and/or Processing: EBK, BT; Analysis and/or Interpretation: EBK, BT; Literature Review: EBK, BT; Writing the Article: EBK, BT; Critical Review: EBK, BT.

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MHA MEDIHEALTH ACADEMY

Surgery on Children

Case Report

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# Acquired jejunal atresia in a neonate after repair of ileal atresia: a case report

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## ABSTRACT

Acquired small bowel atresia is unusual clinical entity and is rarely reported in the clinical literature. We report a case of acquired jejunal atresia in a neonate following surgical repair of ileal atresia. A premature 2-day-old neonate: not passing stool, abdominal distension, bilious nasogastric aspirates. Imaging revealed intestinal obstruction. Exploratory laparotomy identified type III ileal atresia with a mesenteric defect. An ileostomy was created due to marked luminal discrepancies. After ileostomy reversal, the infant developed bilious vomiting and abdominal distension. Imaging suggested intestinal obstruction. Re-exploration revealed a newly developed atretic segment in the jejunum comparable to type II jejunal atresia, while the previous anastomosis had remained intact. Resection of the atretic segment with end-to-end anastomosis was done, and the patient recovered well. Adhesive bands, postoperative vascular compromise, and previous surgery could have contributed to localized ischemia and eventual atresia.

**Keywords**: Atresia, acquired intestinal atresia, neonatal intestinal obstruction, stoma

## INTRODUCTION

Atresia is defined as complete congenital or acquired absence of continuity of the lumen of a hollow viscus or tubular structure. Intestinal atresia is most frequently congenital and is closely linked to embryologic developmental defects, usually due to intrauterine vascular occlusion resulting in ischemia, necrosis, and the resorption of segments of the developing intestine, with ultimate cause of bowel lumen discontinuity.[1,2] For the past decades there have been many investigations of pathogenesis and surgical treatment for congenital intestinal atresia. On the other hand, less attention has been given to acquired forms of postnatal atresia. Acquired intestinal atresia is a rare phenomenon, as the literature documents only limited reports of cases. Discovering possible contributing factors, such as possibly iatrogenic causes, might help minimize the incidence and enhance postoperative surveillance in neonates undergoing abdominal surgery. Here, we present a rare case of acquired jejunal atresia occurring after surgical repair of ileal atresia in a neonate, highlighting the diagnostic challenge and possible mechanisms responsible for this unusual presentation.

## CASE

A 2 day old infant in neonatal intensive care department admitted for the reason of prematurity was unable to pass stool. The abdomen was mildy full in upper aspect and patient had marked greenish aspirates on nasogastric decompression. The meconium was passed in first 24 hours after birth. Paediatric surgical consultation was sought on which after history and physical examination an X-ray abdomen was requested.

X ray of the abdomen revealed multiple air fluid levels and paucity of gas in rectum.

The diagnosis of small bowel atresia was made and patient was prepared for exploratory laparotomy. On surgical exploration the patient was found to have ileal atresia type III, where two blind ends were accompanied by a mesenteric defect. The small gut was followed all the way up to ligament of treitz and was found to be normal, similarly the distal gut was also examined and declared free of any abnormality and was found patent.

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The discrepancy between the two blind loops was remarkable and did not allow for primary anastomosis of two ends even if we opted for Benson and Nixon technique for that purpose. An ileostomy was thus formed with an idea of stool recycling to allow for distal lumen to increase in diameter with instillation of stool from proximal loop. After surgery the patient remained fine and was allowed orally on 4$^{th}$ post operative day when the diminished aspirates gave an evidence of normal bowel motility. Patient tolerated feed well and was gradually built up on feed till patient tolerated demand feed and was passing adequate stool. Patient was then discharged to home with detailed counselling of parents in regard of stool recycling of stoma. An elective stoma reversal date was scheduled in a month time. Patient revisited hospital in out patient department and had no complaints. After a month elapsed re admission was done in NICU for stoma reversal. Although the patient had crossed the neonatal period but owing to prematurity and low weight on centiles patient was entertained by NICU. Patient was taken for surgery where both the loops were mobilized, inspected thoroughly for patency and then meticulous single layer anastomosis was achieved (**Figure 1**).

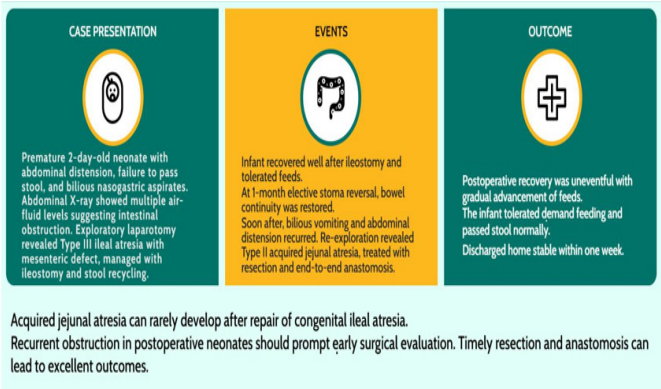



**Figure 1.** Clinical summary of the case of acquired jejunal atresia following repair of congenital ileal atresia

The illustration summarizes the initial presentation of a premature neonate with intestinal obstruction on day two of life due to type III ileal atresia with mesenteric defect, managed with ileostomy and stool recycling. After elective stoma reversal, the infant developed recurrent obstruction due to an acquired type II jejunal atresia. Surgical resection and end-to-end anastomosis resulted in successful recovery and discharge

Following stoma reversal, the patient developed recurrent bilious vomiting, abdominal distension, and failure to tolerate feeds. Abdominal radiography again showed multiple air-fluid levels suggestive of intestinal obstruction. Re-exploration was performed, which revealed a newly developed jejunal atretic segment proximal to the previous ileal anastomosis. The previously repaired ileal segment remained patent and intact. The jejunal lesion resembled type II jejunal atresia, with complete luminal obstruction and a fibrous connection between the blind ends. Segmental resection of the affected jejunal segment was performed followed by primary end-to-end jejunojejunal anastomosis. The postoperative recovery was uneventful, and the patient tolerated feeds well with successful discharge (**Figure 2**).

## DISCUSSION

Jejunoileal atresia is generally believed to be due to intrauterine vascular compromise of the developing midgut. Impaired blood supply results in ischemia and necrosis of the

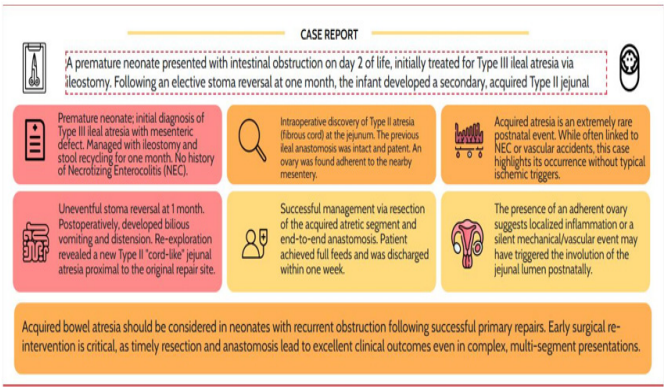



**Figure 2.** Timeline and key clinical events in acquired jejunal atresia after ileal atresia repair

This schematic provides a detailed schedule of events from neonatal presentation with type III ileal atresia, the management process through ileostomy and stool recycling, subsequent stoma reversal, and postoperative recurrence of obstruction. Later, re-exploration showed a new type II jejunal atresia proximal to the previous repair site, which was successfully treated with segmental resection and primary anastomosis, leading to full recovery.

intestinal segment, and reabsorption causing bowel lumen discontinuity. This mechanism is in contrast to duodenal atresia, which occurs when embryologic recanalization fails, not vascular disruption.[3] Jejunoileal atresia is further divided into four types according to anatomical attributes. Type I presents as an intraluminal membrane with preserved bowel continuity. Type II involves two blind ends joined by a fibrous cord that is without any mesenteric defect. Type III is defined by complete separation of bowel segments with a mesenteric defect, and type IIIb, the other form labelled as the "apple-peel" variant, has widespread mesenteric loss with distal bowel supplied by a single artery. Type IV comprises several atretic segments, characteristically appearing as a "string of sausages".[3] The usual surgical approach is resection of the dilated proximal portion, primary end-to-end anastomosis, and taper maneuvers to correct for luminal discrepancy. In cases of significant bowel diameter mismatch or questionable bowel viability, staged repair or temporary stoma formation may be necessary. However, advances in neonatal intensive care and surgical practices have led to drastically improved outcomes and in the majority of centers the survival rate is >90%.[3] Acquired intestinal atresia occurs in incredibly uncommon and isolated cases, in marked contrast to congenital forms. Han et al.[4] reported the case of acquired intestinal atresia in the wake of previous abdominal surgical procedures, indicating that subsequent interventions might have affected its pathogenesis. The mechanism proposed is that postoperative adhesions lead to localized vascular compromise with resultant ischemia, subsequent fibrosis, and ultimately destruction of the intestinal lumen.[4] Unlike congenital jejunal atresia, our patient developed a new proximal jejunal atresia after successful management of type III ileal atresia and subsequent stoma reversal. The intact distal anastomosis and previously normal proximal bowel strongly supported an acquired rather than congenital origin. Similar postoperative acquired intestinal atresia has been rarely described in literature, particularly following repeated abdominal interventions. Han et al.[4] reported acquired intestinal atresia secondary to postoperative adhesions causing vascular compromise and ischemic fibrosis. Our findings support a similar mechanism, where repeated laparotomy, mesenteric handling, and postoperative adhesions may have contributed to localized ischemia and subsequent atresia formation. Necrotizing enterocolitis

(NEC) is another well established etiologic mechanism of gastrointestinal injury in neonates, primarily in preterm infants. Intestinal wall erosion and its subsequent fibrosis and stricture formation occurs after severe and progressive period of healing during NEC, possibly presenting intestinal obstruction later in the healing process.[5] In our case, as our candidate postpartum infant with prematurity, NEC may provide an intellectual basis, but clinical or radiologic features were missing and intraoperative finding of discrete atretic segment, rather than stricture, limited the possibility for NEC as a theoretical explanation. Earlier abdominal surgery, including previous operative manipulation of the abdomen, is also a recognized risk factor for adhesive small bowel obstruction in children. Abdominal surgery is a further risk factor for childhood adhesion, leading to adhesive small bowel obstruction (ASBO). The peritoneal surgical interventions which occur can result in adhesion development and may also be a source of adhesion that has an effect both in distorting bowel loops, mobility and mesenteric blood supply. The possibility of adhesion generation and therefore, bowel obstruction is further facilitated by repeat abdominal surgery.[6] Here, the laparotomy path of previous ileostomy formation, followed by subsequent reversal of the stoma, may have formed adhesion-induced vascular compromise, therefore the new atretic segment was formed. One incidental intraoperative finding seen in our patient was adhesion of the left ovary to the mesentery around a fibrous band that joins together the segments of atresia. However, the ovary remained as normal as possible; the absence of signs of vascular compromise strongly suggests that it was not the main cause of the atresia. Likewise, though a family history of cystic fibrosis was mentioned, the lack of clinical features such as meconium ileus or inspissated intestinal contents only made this association less likely.

## CONCLUSION

The existence of acquired intestinal atresia represents an exceptionally rare phenomenon; consequently, the evidence base is limited to sporadic accounts and definitive findings on its pathogenesis are difficult. This case presents a smaller study in the limited literature on postoperative acquired intestinal atresia and the potential role of it for recurrent intestinal obstruction in infants.

## ETHICAL DECLARATIONS

### Informed Consent

Informed consent was obtained from the legal guardians of the pediatric patient described in this report. Where developmentally appropriate, assent was also sought from the child.

### Peer Review Process

This report underwent external peer review.

### Conflict of Interest

The authors declare no conflicts of interest.

### Financial Disclosure

This case report did not receive any financial support.

### Author Contributions

Concept: FAZ: Design: MA, AI, AS; Control: AM; Resources: MA, AI, AS; Materials: MA, AI, AS; Data Collection and/or Processing: MA, AI, AS; Analysis and/or Interpretation: FAZ, MA, AI, AS; Literature Review: PM, SS; Writing the Article: FAZ, PM, SS; Critical Review: MM, AM; Supervision: AM

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Case Report

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# Progressively worsening headache due to a giant arachnoid cyst in a teenager: a case report

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## ABSTRACT

Headache is one of the most common neurological complaints in the pediatric population and encompasses a broad differential diagnosis. Although primary headache disorders constitute the majority of cases, secondary etiologies—including intracranial space-occupying lesions—must be considered, particularly in patients with persistent, progressive, or treatment-refractory symptoms. Arachnoid cysts are benign, cerebrospinal fluid (CSF)-filled, extra-axial lesions that are typically congenital in origin and arise from developmental abnormalities of the arachnoid membrane. While most arachnoid cysts are asymptomatic and incidentally detected, symptomatic cases may present with chronic headache due to mass effect or impaired CSF circulation. They account for less than 1% of intracranial space-occupying lesions. We report the case of a young female patient with long-standing, treatment-resistant headache without focal neurological deficits. Neuroimaging revealed an intracranial arachnoid cyst exerting mass effect consistent with increased intracranial pressure. Given the persistence of symptoms despite conservative management, surgical intervention was undertaken. The patient underwent cystoperitoneal shunt placement. Postoperative follow-up demonstrated marked clinical improvement, with near-complete resolution of headache severity and frequency. Radiological evaluation confirmed a significant decrease in cyst size and alleviation of mass effect. This case underscores the importance of comprehensive evaluation for secondary causes in pediatric patients presenting with refractory headache. Although arachnoid cysts are frequently incidental findings, symptomatic lesions may require surgical management. Cystoperitoneal shunting can provide favorable clinical and radiological outcomes in appropriately selected patients. Early recognition and timely intervention are essential to prevent prolonged morbidity.

**Keywords**: Arachnoid cyst, headache, shunt

## INTRODUCTION

Arachnoid cysts are benign, cerebrospinal fluid (CSF)-filled, extra-axial cystic lesions lined by flattened arachnoid cells and a collagenous membrane.[1] They are thought to arise from developmental splitting or duplication of the arachnoid membrane during embryogenesis. These lesions are typically located between the cortical surface and the skull base or within the arachnoid membrane itself, one of the three meningeal layers enveloping the central nervous system.[2]

The clinical presentation of arachnoid cysts is heterogeneous and largely dependent on cyst size, anatomical location, and their effect on adjacent neural structures and CSF circulation. Although the majority remain clinically silent and are incidentally identified on neuroimaging, symptomatic lesions may result from progressive cyst enlargement, mass effect, distortion of surrounding parenchyma, or obstruction of CSF pathways leading to elevated intracranial pressure.[3] Furthermore, secondary complications such as intracystic or subdural hemorrhage may precipitate acute neurological deterioration.[4]

Reported clinical manifestations include chronic headache, epileptic seizures, dizziness, gait ataxia, nausea and vomiting, tinnitus, diplopia, visual disturbances, and other signs consistent with increased intracranial pressure.[5]

In this report, we describe a young female patient presenting with long-standing, refractory headaches who was subsequently diagnosed with an intracranial arachnoid cyst. Given the persistence of symptoms and radiological evidence of mass effect, a cystoperitoneal shunt was placed to achieve cyst decompression and restoration of CSF dynamics. The patient demonstrated complete resolution of symptoms in the early postoperative period, accompanied by radiological

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evidence of cyst volume reduction and alleviation of mass effect.

## CASE

A 13-year-old female patient was referred to our neurosurgery clinic with complaints of persistent headache from the pediatric clinic. The patient had experienced recurrent pain attacks for approximately four years, which had progressively worsened over the preceding two months. The pain was predominantly right-sided, severe enough to awaken her from sleep, and was particularly pronounced during the morning and evening hours. The headaches occurred in recurrent attacks, each lasting at least 30 minutes. Over time, both the frequency and duration of the attacks increased. During headache attacks, pain intensity varied between 8 and 10 points on the Visual Analog Scale (VAS). According to the ICHD-3 criteria, the patient's symptoms were consistent with cluster headache. Initially, partial relief was achieved with 500 mg of paracetamol; however, as the symptoms progressed, the patient began taking an additional 100 mg of flurbiprofen. Despite this escalation in analgesic therapy, adequate pain control could not be achieved. Furthermore, the patient's academic performance gradually declined over the course of the illness. The patient did not receive any preventive or advanced medical therapy or other medical treatment at our clinic. Information regarding the medications used and their dosages was obtained from the patient's medical history and review of the medical records.

She denied associated symptoms such as nausea, vomiting, visual disturbances, or seizures. There was no history of head trauma, central nervous system infection, or previous neurosurgical intervention. Her personal and family medical histories were unremarkable, with no known chronic systemic diseases.

On admission, the patient was in good general condition and fully conscious, with a Glasgow Coma Scale (GCS) score of 15. Pupillary examination revealed bilaterally reactive pupils with intact direct and consensual light reflexes. Cranial nerve examination was normal. Motor and sensory examinations were within normal limits, and cerebellar function tests demonstrated no abnormalities. No focal neurological deficits were identified. Ophthalmological evaluation, including fundoscopy, revealed no abnormality despite radiological evidence of significant mass effect.

Non-contrast brain magnetic resonance imaging (MRI) demonstrated a large extra-axial cystic lesion exhibiting signal characteristics identical to CSF on all sequences. The lesion extended along the right fronto-parieto-temporal convexity and reached the Sylvian fissure. The maximal axial dimensions measured approximately 108×66 mm. Imaging findings were consistent with a giant arachnoid cyst. Significant mass effect was observed, including compression of the adjacent right fronto-parieto-temporal lobes and a 10-mm leftward midline shift. Additionally, a retrocerebellar cystic dilatation measuring approximately 26 mm was noted (**Figure 1**). The lesion was classified as Galassi type III.

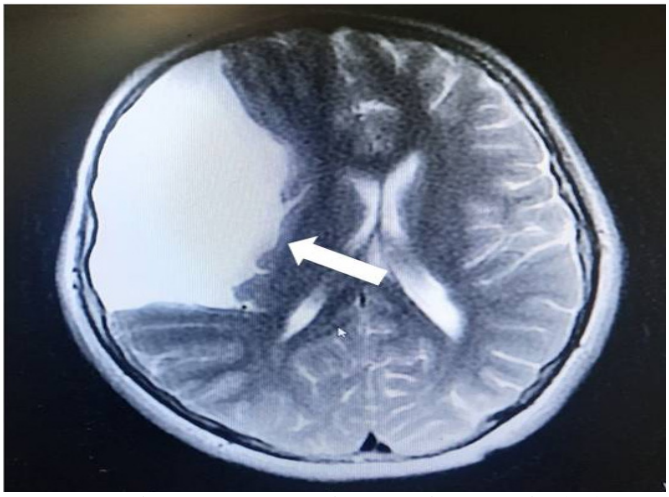


**Figure 1.** Axial non-contrast cranial computed tomography (CT) image obtained with a 2.5-mm slice thickness demonstrates a right-sided, well-defined extra-axial hypodense lesion with attenuation characteristics consistent with cerebrospinal fluid, compatible with an arachnoid cyst (white arrow). The lesion produces significant mass effect, resulting in a leftward midline shift and compression with effacement of the ipsilateral lateral ventricle.

Subsequently, a non-contrast cranial computed tomography (CT) scan was obtained, revealing a well-demarcated hypodense lesion with attenuation values consistent with CSF in the right frontal region, measuring up to 10 cm in maximal diameter (**Figure 2**). These findings further supported the diagnosis of an arachnoid cyst.



**Figure 2.** Axial T2-weighted magnetic resonance imaging (MRI) obtained with a 3-mm slice thickness demonstrates a right-sided, well-circumscribed extra-axial lesion exhibiting cerebrospinal fluid–like signal intensity, consistent with an arachnoid cyst (white arrow). The lesion exerts significant mass effect, resulting in a leftward midline shift and effacement of the ipsilateral lateral ventricle, with associated compression of the adjacent cortical and subcortical structures.

Given the progressive nature of the patient's symptoms (which were consistent with increased intracranial pressure), the substantial mass effect with midline shift, and the lack of satisfactory response to conservative treatment, surgical intervention was recommended. After evaluating available treatment options, cystoperitoneal shunting was selected because it was considered the most appropriate strategy to achieve continuous cyst decompression and rapid reduction of intracranial mass effect. The patient underwent cystoperitoneal shunt placement. A burr hole was created in the right frontoparietal region overlying the cyst. The catheter

was connected to a medium-pressure valve system equipped with an automatic antisiphon device to minimize the risk of overdrainage. No intraoperative complications occurred.

Postoperative neuroimaging demonstrated a marked reduction in cyst volume and significant improvement in the midline shift (**Figure 3, 4**). Compared with preoperative imaging, the midline deviation was substantially decreased. Clinically, the patient experienced significant relief of headache symptoms even in the early postoperative period. She remains under regular clinical and radiological follow-up for 3 years.



**Figure 3.** Postoperative axial computed tomography (CT) image obtained three days after surgery with a 2.5-mm slice thickness demonstrates partial reduction in the size of the lesion and improvement of the midline shift. The white arrow indicates the residual cyst, while the black arrow points to the shunt pump.

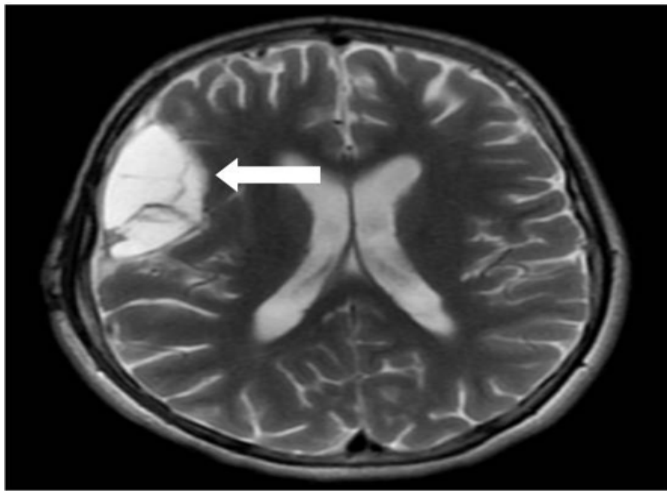


**Figure 4.** Axial T2-weighted magnetic resonance imaging (MRI) obtained three months postoperatively with a 3-mm slice thickness demonstrates marked reduction in the volume of the arachnoid cyst (white arrow). There is resolution of mass effect, with re-expansion of the previously compressed ipsilateral lateral ventricle and restoration of normal cortical and subcortical architecture.

## DISCUSSION

Although headache is a common complaint during childhood, it is frequently underestimated by caregivers; nevertheless, it may occasionally represent an underlying serious intracranial pathology.[6] Headaches occurring at an early age, those that awaken the child from sleep or are most pronounced in the early morning hours, the "worst headache ever experienced," or headaches accompanied by seizures, visual disturbances, focal motor deficits, or balance impairment should prompt careful evaluation and comprehensive neuroimaging.[7] In the present case, none of these red-flag features were observed. The neurological examination was entirely normal, and the patient's sole complaint was progressively worsening headache.

The etiology of pediatric headaches is broadly classified into primary and secondary categories.[8] Primary headache disorders include migraine, tension-type headache, and cluster headache syndromes.[8] In contrast, secondary headaches arise from structural, vascular, infectious, inflammatory, or neoplastic processes involving the central nervous system.[8] Among structural causes, arachnoid cysts represent a rare but clinically relevant entity.[9] Arachnoid cysts account for approximately 1% of all intracranial space-occupying lesions and are most commonly located in the middle cranial fossa, followed by the suprasellar and posterior fossa regions.[9] Although frequently asymptomatic and incidentally detected, these lesions may become symptomatic depending on their size, anatomical location, and associated mass effect or disturbance of CSF circulation.[9]

The optimal management of arachnoid cysts remains a matter of debate. Surgical intervention is generally reserved for patients with signs of elevated intracranial pressure, progressive cyst enlargement, or radiological evidence of significant compression of adjacent neural structures.[10] Microsurgical total excision or fenestration offers the theoretical advantage of definitive treatment without shunt dependency.[11] However, complete resection may not be feasible in all cases due to anatomical constraints, risk of recurrence, and the potential for injury to adjacent eloquent brain tissue.[11]

Minimally invasive approaches, including endoscopic ventriculocystostomy or cyst fenestration, have gained popularity due to reduced operative morbidity and shorter hospitalization periods.[12,13] Nevertheless, procedure-related complications—such as intraventricular or intracerebral hemorrhage—have been reported.[12] Endoscopic techniques are generally considered safe and effective particularly when cyst anatomy permits adequate fenestration and CSF diversion without the need for permanent shunting.[12] However, recurrence and the need for reoperation remain potential limitations with this procedure.[12] In addition, as is the case in our institution, this technique may not be available in all hospitals.

In the present case, cystoperitoneal shunt placement was preferred due to the large size of the cyst, the presence of a 10-mm midline shift, and the need for reliable long-term decompression. Considering these radiological findings, shunt placement was considered the most appropriate treatment strategy. The early postoperative course was uneventful, with marked radiological and clinical improvement. The use of a medium-pressure shunt system equipped with an automatic antisiphon device allowed controlled drainage, thereby minimizing the risk of overdrainage and subsequent subdural hematoma formation.

## CONCLUSION

Persistent or treatment-resistant headache in the pediatric population warrants thorough evaluation, even in the absence of focal neurological deficits. Significant intracranial mass lesions may be present despite a normal neurological examination. Although the optimal management of symptomatic arachnoid cysts remains controversial and should be individualized according to cyst characteristics and patient factors, cystoperitoneal shunting represents a technically straightforward, widely applicable, and effective surgical option. In appropriately selected patients, it can provide rapid symptom relief, significant reduction of cyst volume, and favorable radiological outcomes.

## ETHICAL DECLARATIONS

### Informed Consent

Informed consent was obtained from the legal guardians of the pediatric patient described in this report. Where developmentally appropriate, assent was also sought from the child. The inclusion of vulnerable populations in this study adhered to national and international ethical guidelines. Extra care was taken to ensure voluntary participation, understanding, and protection of participant dignity and autonomy.

### Peer Review Process

This report underwent external peer review.

### Conflict of Interest

The authors declare no conflicts of interest.

### Financial Disclosure

This case report did not receive any financial support.

### Author Contributions

Concept: HA, EC, SÖ; Design: HA, EC, SÖ; Control: HA; Resources: EC, SÖ, RNT; Materials: EC, SÖ, RNT; Data Collection and/or Processing: EC, SÖ, RNT; Analysis and/or Interpretation: EC, RNT, SÖ; Literature Review: HA, EC, RNT; Writing the Article: HA, EC, RNT; Critical Review: SÖ.

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