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## Intestinal malrotation causing chylous ascites in an adolescent: a case report

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### ABSTRACT

**INTRODUCTION:** Intestinal malrotation is a rare etiology of chylous ascites in adolescents. Chylous ascites is caused by lymphatic system disarrangement, which can result in an anomalous build-up of a lymphatic fluid rich in lipid, namely chyle in the peritoneal cavity.

**PRESENTATION OF CASE:** We present a case of a 16-year-old Saudi Arabian male who came to the emergency department with right upper quadrant pain associated with difficulty in passing stool for one day and a history of congenital diaphragmatic hernia (CDH) repair at the age of 4 months. Free fluid in the abdomen was noticed in the bedside ultrasound. Abdominopelvic computed tomography revealed dilated small bowel loops and a whirl sign of the mesentery, which indicated intestinal torsion. The patient was treated using Ladd's procedure, and a large volume of chylous fluid was removed from the abdomen. Postoperatively, the abdominal drain revealed no chyle, and the patient was followed-up as an outpatient at which point, he reported no abdominal pain.

**DISCUSSION:** Intestinal malrotation is more commonly reported in children and associated with congenital chylous ascites. Chylous ascites by itself is a rare occurrence, and very few cases attribute it to intestinal malrotation in adults. Surgical management with Ladd's procedure is a well-documented surgery in pediatric patients, and yet it was successfully performed in our adolescent patient.

**CONCLUSION:** Surgical management of chylous ascites secondary to intestinal malrotation in an adolescent, which is considered novel in this age group, including peritoneal lavage of chyle, resulted in favorable outcomes.

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## 1. Introduction

Chylous ascites is a rare yet critical condition caused by lymphatic system disarrangement, which can result in an anomalous build-up of a lymphatic fluid rich in lipid, namely chyle in the peritoneal cavity. The etiology behind this disruption can be attributed to the rupture within the lymphatic system or an increase in the peritoneal lymphatic pressure as a result of intestinal obstruction. Consequently, chylous ascites etiology is determined on the basis of trauma, malignancy, infections, postoperative complication, or cirrhosis [1]. Two-thirds of chylous ascites cases in developed countries result from cirrhosis and malignant tumors; whereas, in developing countries, tuberculosis is responsible for a large number of cases [1,2]. Malignancy is the leading cause of chylous ascites in

adults, followed by postoperative complications. One of the rarest etiologies to cause accumulation of chyle in the peritoneum cavity is intestinal malrotation, which results in compression of the Ladd's band in the mesentery, leading to lymphatic obstruction. This condition is usually diagnosed and managed in the early years of the patient's life using Ladd's procedure, which is a surgery that involves intestinal derotation in a counterclockwise direction and separation of the bands that connect the colon to the duodenum [3].

Moreover, diagnosing chylous ascites depends on the obtainment of complete disease history and appropriate physical examinations. The patient typically presents with progressive painless distension of the abdomen for a few weeks or months. Other symptoms might manifest as weight gain and shortness of breath due to compression of the lungs caused by accumulating chyle in the abdominal cavity [1]. Chyle appears milky and contains triglyceride levels exceeding 200 mg/dL, which is considered significant for diagnosing chylous ascites [1]. The main goal in assessing and managing chylous ascites is to treat the underlying disease and to relieve symptoms. The management plan is dependent on the

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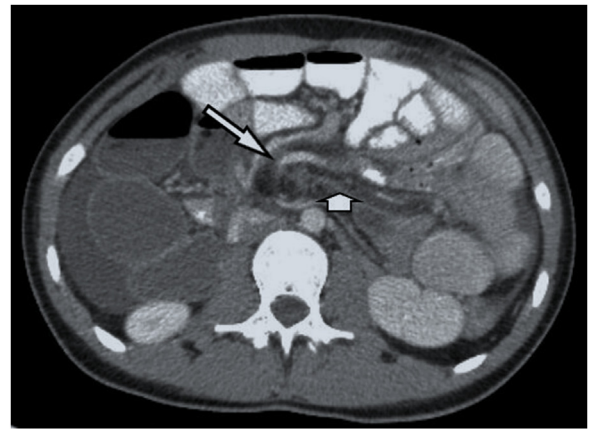
**Fig. 1.** Abdominal plain X-ray (erect anteroposterior view) showing dilated malrotated jejunal loops in RUQ with air-fluid level (arrow).

etiology and the severity of the ascites; therefore, the plan varies according to dietary needs, medical therapies, and surgical interventions. In cases of failure to respond to medical therapy, surgical intervention is advised [1]. In this report, we introduce a distinctive case of chylous ascites in an adolescent patient presenting with abdominal pain due to intestinal malrotation. This manuscript has been written in compliance with the SCARE 2018 criteria [4].

## 2. Presentation of Case

A 16-year-old Saudi Arabian male came to the emergency department complaining of pain in the right upper quadrant (RUQ) associated with difficulty in passing stool for one day. The patient was conscious, alert, and vitally stable. He had no significant medical or family history and was not on any medications. He had a history of congenital diaphragmatic hernia (CDH) repair in infancy but was otherwise medically sound (Table 1). Physical examination revealed a soft abdomen with no guarding or rigidity; however, tenderness was noted at the RUQ of his abdomen. During his stay in the emergency room, the patient was administered sodium chloride (0.9%; 1000 mL IV bolus), esomeprazole injection (40 mg IV over 30 min), acetaminophen injection (1000 mL IV over 15 min), and a fleet enema (133 mL rectal).

Over the following hour, the patient's symptoms mildly subsided, and laboratory investigations showed unremarkable results. However, free fluid in the abdomen was discovered using bedside ultrasound (Table 1). Urgent abdominal X-ray showed dilated malrotated jejunal loops in RUQ with air-fluid level (Fig. 1), and abdominopelvic computed tomography (CT) revealed loops in the small intestines and whirl sign of the mesentery (Fig. 2) [5]. Furthermore, the duodenojejunal junction was not visualized in its normal position; instead, it was located in the midline. The transitional zone was noted in the distal small bowel near the whirl sign with abdominal free fluid (chyle) (Fig. 3), and moderate chylous fluid was also seen in the pelvic area, as noted in Fig. 4. The cecum



**Fig. 2.** Abdominal CT (axial view) showing a whirl sign (arrow) and collapsed bowel (arrowhead).



**Fig. 3.** Abdominal CT (axial view) showing the transitional zone (arrow) with chylous ascites (arrowhead).

was seen in the RUQ, and collapsed ileal loops were occupying the left side of the abdomen.

The following morning, an urgent exploratory laparotomy was carried out by an associate consultant with over 20 years of experience in general surgery and yielded a considerable quantity of chylous fluid that was sent for triglyceride level evaluation after suctioning. Assessment of the drained fluid revealed a triglyceride level of 208 mg/dL (Table 1). Ladd's procedure was commenced by inspecting the bowel from the duodenojejunal junction until the rectum, with no identified perforation; however, multiple adhesions were discovered and released simultaneously. The remaining bowel was healthy, with no abnormalities other than visualization of the right colon and cecum in RUQ. Abdominal exploration was carefully performed in order to identify the transitional zone given that we found a collapsed bowel loop. The small intestines were thoroughly examined from the ligamentum teres to the ileocecal valve, which was in the RUQ with a band over it. Bowel dilation was noted, with multiple areas of dilatation and collapse at the transitional zone with band and adhesion over it. Sharp division of all adhesion was performed, and the entire small intestines were cautiously separated while hemostasis was secured, with no signs of active bleeding.

During the procedure, we identified and released a band in the cut-off of the superior mesenteric artery that was causing an obstruction. We then successfully released the bands connecting the ileocecal valve and the cecum. After adhesions were released, the small intestines were inspected for viability, and it was run

**Table 1**  
Timeline representation of the case report.

Date	Summaries from initial and follow-up visits	Diagnostic testing	Interventions
Relevant past medical history and interventions	Previous medical history revealed that the patient had a repair surgery of his diaphragmatic hernia at the age of 4 months. Occasional self-treatment with paracetamol for his intermittent abdominal pain.		
07 September 2019	Presented to the ER with one day history of RUQ pain associated with difficulty in passing stool	Vitally stable, Laboratory findings: unremarkable Bedside ultrasound: free fluid in the abdomen Abdomen X-ray: dilated segments of small intestines with large air-fluid levels Pelviabdominal CT: small intestines loops and whirl sign of the mesentery, cecum was seen in the RUQ	Treated with IV bolus of fluids, esomeprazole, acetaminophen, and fleet enema General surgeon consulted: urgent exploratory laparotomy was performed
08 September 2019	Transferred to the OR and put under GA	Chyle fluid suctioned from abdomen with TG levels of 208 mg/dL	Treated with Ladd's procedure, separation of bands, adhesiolysis, and detorsion counterclockwise of the bowels
09 September 2019	Admitted to the general surgery ward for monitoring	Patient was clinically stable on NPO and NGT with no active issues. SBP (mmHg) 114 DBP (mmHg) 63 HR (Freq./min) 100 RR (Freq./min) 18 T (°C) 37.0 SpO2 (%) 98	Treated with cefuroxime, morphine sulfate, and enoxaparin concurrently for 3 days
10 September 2019	Continued monitoring of the patient in the general surgery ward	Patient was clinically stable on soft diet SBP (mmHg) 111 DBP (mmHg) 56 HR (Freq./min) 82 RR (Freq./min) 20 T (°C) 36.9 SpO2 (%) 98	
11 September 2019	The patient was mobilizing and eating well, he was discharged	Patient was vitally and clinically stable SBP (mmHg) 126 DBP (mmHg) 77 HR (Freq./min) 87 RR (Freq./min) 18 T (°C) 36.9 SpO2 (%) 96	Given tramadol for 5 days, acetaminophen for 14 days, and cefuroxime for 7 days
16 September 2019	Followed-up as an outpatient with a history of repetitive vomiting for 4 days	Examination of wound revealed a clean, noninfected scar, abdomen was soft with no abnormalities	To be seen in the dressing clinic 3 days later, prescribed antiemetic medication

ER, emergency room; RUQ, right upper quadrant; CT, computed tomography; OR, operation room; GA, general anesthesia; TG, triglyceride; NPO, nothing by mouth; NGT, nasogastric tube; SBP, systolic blood pressure; DBP, diastolic blood pressure; HR, heart rate; RR, respiratory rate; T, temperature; SpO2, peripheral capillary oxygen saturation.

from the ligamentum teres to the ileocecal valve to inspect for further abnormalities. Finally, we checked the colon throughout the sigmoid and did not identify areas of obstruction or bands. The operation was well-tolerated, and the patient was in stable condition upon transfer to the post-anesthesia care unit. Postoperatively, the patient was started on a nothing by mouth (NPO) routine and was commenced on nasogastric tube (NGT) feeding, with gradual toleration of oral feeding. He was given cefuroxime injections (750 mg IV mixed with 5% dextrose in 50 mL water), morphine sulfate injections (10 mg/mL, 1 mg IV), and enoxaparin injections (40 mg subcutaneous) concurrently for three days (Table 1).

On postoperative day 4, the abdominal drain revealed no chyle, and the patient was discharged on tramadol capsules (50 mg orally for 5 days), acetaminophen tablets (1000 mg orally for 14 days), and cefuroxime tablets (500 mg orally for 7 days). The patient followed-up as an outpatient one-week post-discharge, at which point he reported neither abdominal pain nor any problems with continuing a normal diet; however, episodes of repetitive vomiting were reported. The patient was prescribed antiemetic medication, and no further imaging was performed (Table 1). No additional follow-up appointments were given, and the patient was instructed to



Fig. 4. Abdominopelvic CT (coronal view) showing chyle free fluid (arrow).

present to the emergency department upon experiencing further symptoms.

### 3. Discussion

Chylous ascites is described as the leakage of lipid-rich lymphatic fluid into the peritoneal cavity as a result of traumatic or nontraumatic obstruction of the lymphatic system or one of its tributaries [6]. Numerous conditions have been reported to cause chylous ascites, which are divided into primary and secondary causes. Primary causes are often attributed to congenital lymphatic abnormalities. More common secondary causes usually consist of surgeries, traumas, or malignancies. Abdominal malignancies (e.g., lymphoma) and liver cirrhosis are accountable for most cases of chylous ascites in Western countries; whereas, infections such as filariasis are the main culprits in cases in Eastern countries [6]. In adults, the most frequent etiology of chylous ascites is malignancy (7%), while the leading etiology in pediatric patients is congenital lymphatic abnormalities (32%) [7,8].

Intestinal malrotation is uncommonly reported in adolescents and adults although it is a well-established disease in children and associated with congenital chylous ascites in infants [3]. Chylous ascites by itself is a rare occurrence, and very few cases attribute it to intestinal malrotation. In our case, the patient had a surgical history of CDH repair, with a possibility of oversight in his intestinal malrotation intraoperatively. Heiwegen et al. [9] retrospectively reported that CDH is commonly associated with intestinal malrotation found at the initial repair of CDH (39%) in infants. Moreover, the study reported that intestinal malrotation was missed at the time of CDH repair surgery in 6% of cases and was diagnosed years later when most of the patients presented with shortness of

breath; however, no patient was diagnosed with secondary chylous ascites. A case was reported of a 19-year-old Chinese male presenting with vomiting for one day associated with severe abdominal pain, in which, upon exploratory laparotomy, chylous fluid and intestinal malrotation were found. Unlike our case, the patient did not have a history of prior surgeries and had a sudden disruption of the superior mesenteric vein immediately distal to the splenoportal confluence [10]. Moreover, Ladd's procedure is a well-documented surgery that is usually reserved for pediatric patients, and that involves counterclockwise derotation of the intestines and separation of the coloduodenal bands, as seen in our case [11]. Nevertheless, the low incidence of intestinal malrotation and midgut volvulus in adolescents, especially with chylous ascites, raises the question of favorable outcomes of Ladd's procedure in this age group.

### 4. Conclusion

As illustrated in our case, the co-occurrence of intestinal malrotation and chylous ascites in an adolescent patient qualifies as nearly unprecedented. However, it is critically important to exclude life-threatening conditions in patients who present with a complaint of recurring abdominal pain. As demonstrated in this case, surgical management, which is considered novel in this age group, including peritoneal lavage of chyle in a patient who presented with chylous ascites secondary to intestinal malrotation, resulted in favorable outcomes. During follow-up, clinical judgment concluded full recovery from chylous ascites upon surgically correcting the intestinal malrotation.

### Declaration of Competing Interest

The authors report no declarations of interest.

### Sources of funding

None.

### Ethical Approval

This case report is approved by the Institutional Review Board of King Abdullah International Medical Research Center (SP19-547-J).

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Author contribution

Saad Mohammed Alshahrani - Writing of original draft and proofing.

Mughis Ahmed Saeed - Conceptualization and supervision.

Abdullah Saleh Alghamdi - Writing and proofing.

Mohammed Saad Alameri - Writing and proofing.

### Registration of research studies

Not applicable.

### Guarantor

Saad Mohammed Alshahrani.

## Patient's perspective

The surgeon explained the procedure well to me and my father, and we gave him consent. I was then taken to the operating room under general anesthesia. The surgery was successful, and there was no need to remove parts of my intestines. After the surgery, I started feeding from a tube and took antibiotics and painkillers with a gradual feeding from the mouth. I was discharged 4 days later and was feeling well.

## Provenance and peer review

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