JCRMHS

Case Report

Journal of Clinical Case Reports, Medical Images and Health Sciences

Volume 7 Issue 4, 2024

Article Information

**Received date:** 26/07/2024
**Published date:** 19/08/2024

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*Key Words:

Chyloperitoneum, Esophageal atresia, Infant.

*List of Abbreviation

Chylous ascites; CA
Esophageal atresia; EA
Middle chain triglyceride; MCT
Pediatric intensive care unit; PICU
Tracheoesophageal fistula; TEF
Total parenteral nutrition; TPN

# Unusual Cause of Abdominal Swelling in an Infant with Esophageal Atresia-Tracheoesophageal Fistula: Case Report

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## Abstract

**Background**: Chyloperitoneum is rare in children, the most common cause being primary lymphatic anomalies. However, it may also be an acquired disease, most commonly due to trauma and surgery. This is the first report of an infant who had chyloperitoneum because of thoracic duct compression by the dilated esophageal segment.

**Case**: A two-month 15-day-old boy was admitted to our pediatric intensive care unit for postoperative monitoring after endoscopic dilation of esophageal stricture. He was operated on the first day after birth due to the diagnosis of esophageal atresia. Circulatory insufficiency developed on the fifth day of hospitalization during postoperative follow-up. The patient required vasopressor and invasive respiratory support. Due to abdominal distension, total parenteral nutrition (TPN) was initiated, and under TPN support, abdominal distension regressed. Extubation was performed on the 14th day of hospitalization. Following the initiation of enteral nutrition, abdominal distension recurred. Chylous ascites was detected, and the etiology remained unclear. The patient, whose chylous ascites partially regressed with medical treatments and respiratory support was discontinued, was discharged under close monitoring. A week later, he presented with feeding difficulties. Chylous ascites was observed on bedside ultrasound. Dilation was performed, and on the same day, it was found on ultrasound that the chylous ascites had completely regressed.

## Background

Chyloperitoneum is rare in children, most common cause being primary lymphatic anomalies, followed by various uncommon causes. However, it may also be an acquired disease, most commonly due to trauma and surgery (1). Postoperative chylous ascites (CA) has been reported after surgery for esophageal atresia (EA) and tracheoesophageal fistula (TEF), cardiothoracic surgery, and liver transplantation. Cardiac disease, including heart failure, restrictive cardiomyopathy, and constrictive pericarditis, may also cause CA, both in adults and children. Extensive central venous thrombosis may impair lymphatic absorption and cause CA. Mortality may be as high as 40 - 70% (2).

However, chyloperitoneum occurred via a previously undescribed mechanism in our patient. We hereby present a male infant who had chyloperitoneum because of thoracic duct compression by the dilated esophageal segment after surgery for esophageal atresia.



DOI: 10.55920/JCRMHS.2024.07.001308

## Case Presentation

A two month 15 days 15-day-old boy was admitted to our pediatric intensive care unit (PICU) for postoperative monitoring after endoscopic dilation of esophageal stricture. He was born at week 36 of gestation with a birth weight of 2280 grams. He was diagnosed antenatally with EA, TEF, polyhydramnios, and malalignment-type ventricular septal defect. He had a left undescended testis and retrognathia on physical examination. His initial postnatal echocardiogram revealed a broad subaortic ventricular septal defect and dextroposition of the aorta. He underwent transpleural primary anastomosis of the proximal and distal esophageal stumps on his first postnatal day. During this operation, the azygos vein was ligated for better visualization of the type C TEF. Feeds were initiated on postoperative day 2 without problems. He was discharged uneventfully on postnatal day 17. However, he was admitted one week later due to aspiration pneumonia. A stricture at the anastomosis site was noted and dilated with no 18 Hegar dilator on day 5 of admission and was able to feed freely orally with breastmilk. He was discharged uneventfully and was evaluated regularly for recurrence of stricture, for which he had to undergo another endoscopic dilation procedure with no 15 Hegar dilator on postnatal day 77, after which he was admitted to the PICU for postoperative monitoring.

He was extubated and enteral tube feeds were commenced. Unfortunately, he became septic and needed invasive mechanical ventilation, vasoactive infusions (milrinone, noradrenaline), and hydrocortisone replacement due to hyponatremia, hyperkalemia, hypoglycemia, and persistent shock despite appropriate vasoactive support on day 6. He recovered and was extubated on day 14 of admission, whereafter persistent need for non-invasive ventilatory support and progressive abdominal distension. Point of care ultrasound revealed particulate fluid accumulation in the abdomen and no pleural effusion.

Diagnosis of CA depends on sampling of the peritoneal fluid and demonstrating increased triglycerides, low cholesterol, increased protein, and lymphocytic pleocytosis (3-9). Paracentesis revealed fluid with The triglyceride level was 238 mg/dL and a lymphocytic predominance was noted on microscopic examination and diagnosed CA.

An echocardiogram was repeated to rule out right heart failure as the cause of CA, and revealed normal systolic functions and ejection fraction, minimal pericardial effusion, and no mitral regurgitation. An abdominal X-ray with contrast was obtained to rule out leakage into the peritoneum (Figure 1). There was a dilated pouch proximal to the stenotic side, but no leak was seen. Lymphoscintigraphy was not possible due to the small size of our patient. Doppler ultrasound of the central venous system revealed no thromboses.

Chyloperitoneum may heal spontaneously with conservative management. Therefore, a conservative management is warranted. Conservative treatment modalities include a middle chain triglyceride (MCT) - based high protein diet, withholding enteral feeds, and providing TPN and octreotide, a long-acting somatostatin analog, which may be given as an infusion or subcutaneous injections. If chyloperitoneum persists for 1 - 2 months on conservative management, surgical options are considered (4). While it is possible to wait for healing only on an MCT and protein-rich diet, the literature suggests that improvement with octreotide may be faster (4-11).

We applied different treatments sequentially in our patient based on response. A diet consisting of 50% MCT (Peptijunior; Nutricia advanced medical nutrition, Cuijk, Holland) was started, but there was no decrease in abdominal girth. The need for non-invasive respiratory support persisted, making abdominal volume a priority. Enteral feeds were withheld, and we started total parenteral nutrition and medical treatment, namely, octreotide infusion and propranolol (0.5 mg/kg/day initially, which was increased to 3 mg/kg/day). We also had to perform paracentesis on days 19, 21, and 27 to reduce abdominal volume. The progress of our patient on medical therapy for CA has been summarized in Figure 2. Although CA persisted, abdominal volume decreased sufficiently with medical treatment to allow weaning of the patient to room air. We considered the MCT-enriched diet to be ineffective and restarted enteral feeding with breastmilk. Our patient was stable on conservative management with subcutaneous

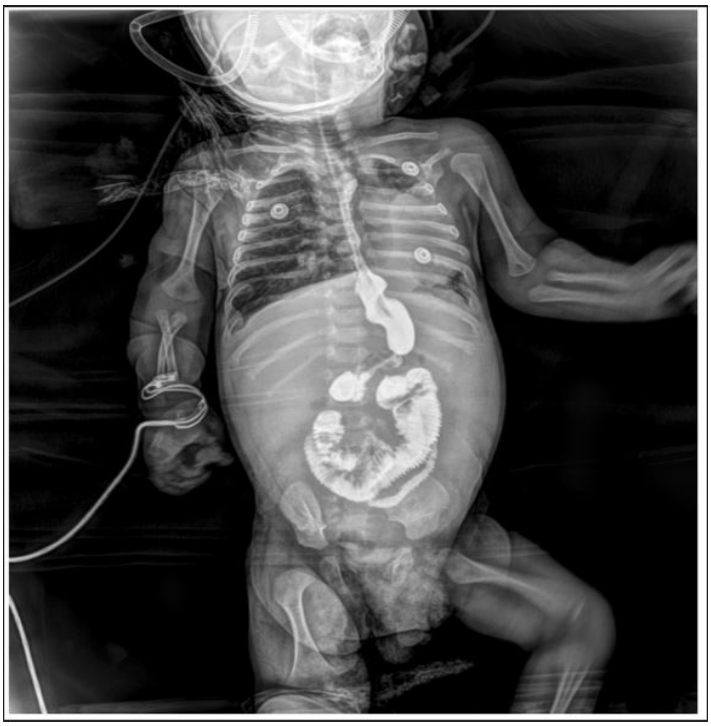

***Figure 1:*** *Upper gastrointestinal series showing the esophageal dilatation proximal to the anastomotic site and no leakage.*



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***Figure 2:*** *Timeline showing different procedures and medications. HFNC, high flow nasal cannula; IMV, intermittent mandatory ventilation; MCT, middle chain triglyceride; SubQ, subcutaneous.*

octreotide and breastmilk, pending healing of a presumed surgical thoracic duct damage, like the previous report by Sy et al (12). The patient was discharged with mild abdominal distension and persistent CA on propranolol and subcutaneous octreotide, with arrangements for close follow-up.

One week after discharge, he presented with difficulty feeding and was readmitted for respiratory distress. His chest X-ray revealed a massively dilated esophageal pouch and NG tube placement for enteral feeds proved impossible. He was started on intravenous fluids and dilation was performed. The day after dilation, the abdomen was found to be supple and completely free of fluid on ultrasound. The dilated pouch was no longer visible on X-ray. His beta-blocker dose was decreased to 1 mg/kg/day and octreotide treatment was tapered off over the ensuing days, after which CA did not recur one-and-a-half-year follow-up.

We hypothesized that CA was the result of compression of the thoracic duct by the dilated esophageal segment proximal to the stenotic area, rather than direct surgical damage to lymphatics. The patient is now enrolled in a program of frequent esophageal dilation and remains free of CA at the time of this report.

## Conclusion

The fact that CA disappeared after esophageal dilation suggests that the mechanism for CA in our patient was external compression of the thoracic duct by the proximal stenotic segment, rather than direct damage to the lymphatics during surgery. He underwent dilation during his second admission because he had difficulty with oral feeds, and we were unable to place an NG tube. The day after dilation, both the stenotic proximal segment and the chyloperitoneum disappeared, which strongly suggests that the accumulation of peritoneal fluid was the result of mechanical compression by said dilated segment. After the dilation procedure, we were able to quickly taper off the octreotide treatment and the patient remains free of chyloperitoneum on a frequent dilation program, which also supports the thoracic duct compression by dilated esophageal segment theory.

To our best knowledge, this is the first report of chyloperitoneum occurring via this mechanism. In dysfunction of the thoracic duct, compression by dilated viscera should also be considered, particularly in patients with a history of esophageal surgery and stenosis.

**Informed consent/ Patient consent**: Informed consent was obtained from the parents of the infant.

**Data availability statement**: Patient data can be shared upon request.

**Conflict of interest statement**: The authors state that they have no conflicts of interest relevant to this publication.

**Funding statement**: No financial support was received for this case report.

**Authors' Contribution**: Each author has contributed to the design and conduct of the work. The manuscript has been written, read, and approved by all authors.

**Acknowledgments**: The authors would like to acknowledge and thank Başak Erginel, MD and Erdem Özatman, MD for their contribution in surgical management of this patient.

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