Pediatr Surg Int (2008) 24:633–636
DOI 10.1007/s00383-008-2127-7

CASE REPORT

# A newborn with chylous ascites caused by intestinal malrotation associated with heterotaxia syndrome

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Accepted: 27 February 2008 / Published online: 11 March 2008


**Abstract** Chylous ascites is a rare entity in infants. An uncommon cause of chylous ascites is intestinal malrotation. We report a case of a neonate with heterotaxia-polysplenia syndrome and intestinal malrotation who presented with chylous ascites and subsequently developed duodenal obstruction from midgut volvulus. He was successfully treated with peritoneal drainage, surgical intervention, and nutritional support. A brief overview of the association of chylous ascites, intestinal rotational anomalies and heterotaxia syndromes is discussed.

**Keywords** Chylous ascites · Intestinal malrotation · Heterotaxia

## Introduction

Chylous ascites is a rare entity in infants that is most often due to congenital abnormalities of the intestinal lymphatic system. An uncommon cause is intestinal malrotation. We report an interesting case of chylous ascites in a neonate with intestinal malrotation who was also found to have heterotaxia-polysplenia syndrome. We discuss the association of situs anomalies with intestinal malrotation and provide a brief overview of the management of chylous ascites.

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## Case report

A 2 week old male infant presented with a 2 day history of abdominal distension. He was breast fed and tolerating his feeds well prior to his presentation. There was no history of fever. Urine output and stools were normal. On examination, he was in no apparent distress and his vital signs were normal. He was noted to have right sided heart sounds. His abdomen was tensely distended. Bilateral hydroceles were present. Neurological exam was unremarkable.

Abdominal ultrasound showed massive ascites. In addition, there was evidence of a pre-duodenal portal vein and polysplenia. Computed tomography of the abdomen showed ascites, right-sided stomach, left-sided liver, and multiple right sided spleens (Fig. 1). Dextrocardia with normal intra-cardiac anatomy was confirmed by echocardiogram. Our patient was felt to have heterotaxia syndrome, polysplenia variant.

On hospital day 2, a diagnostic paracentesis was performed and milky fluid was obtained. With laboratory confirmation of chylomicrons, the diagnosis of chylous ascites was made and the patient was started on a medium-chain triglyceride formula (Portagen). However, his abdominal distension worsened causing respiratory distress. A peritoneal drain was placed with subsequent removal of about 1,000 ml of chyle over a 48 h period (Figs. 2, 3). His respiratory status and abdominal examination markedly improved.

As chylous ascites may be caused by intestinal malrotation an upper gastrointestinal series was performed. Whereas normal rotation of the bowel would place the

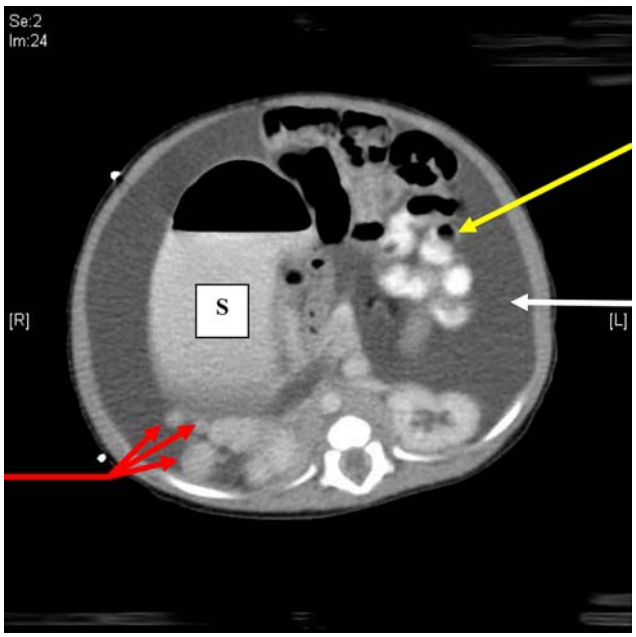


**Fig. 1** CT scan showing massive ascites (*white arrow*), right-sided polysplenia (*red arrows*), right-sided stomach (S), and left sided loops of bowel (*yellow arrow*)

duodenojejunal (D-J) flexure across the midline at the level of the pylorus, our patient's D-J flexure clearly did not cross the midline. A contrast enema was performed which showed the cecum positioned in the left lower quadrant suggesting a broad-based mesentery. Although this was felt to indicate a lower risk of midgut volvulus, on hospital day 15 our patient developed sudden onset of bilious emesis. An emergent repeat gastrointestinal series confirmed obstruction at the level of the third portion of the duodenum suggesting midgut volvulus (Fig. 4). A surgical consult was obtained.

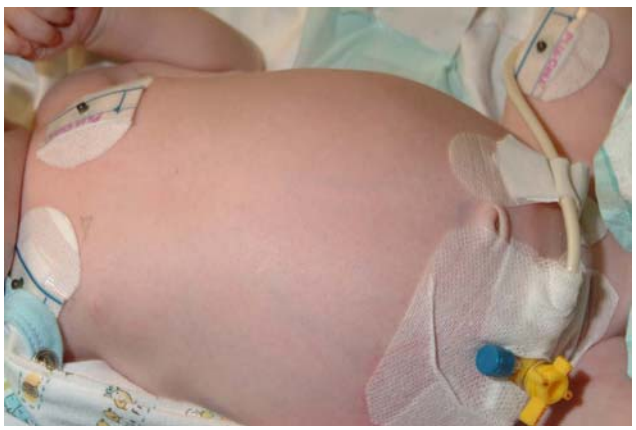

**Fig. 2** Marked abdominal distension due to chylous ascites in a newborn. Peritoneal drainage catheter showing milky fluid draining from the abdomen


**Fig. 3** Peritoneal drainage bag showing milky fluid due to chylous ascites coming from the abdomen

The patient was taken for an emergency laparotomy. A 270° midgut volvulus was found and the bowel was untwisted. The entire bowel was viable. A Ladd's procedure was performed and the peritoneal drain was removed. Post-operatively, the patient was started on a cow's milk based formula once bowel function resumed. No re-accumulation of ascites was seen and he was discharged home on full enteral feeds on post-operative day 7.

One week post-discharge (i.e. 2 weeks post-operation), the patient returned to the hospital with massive abdominal distension. A peritoneal drain was placed and confirmed the re-accumulation of chylous ascites. He was placed on bowel rest and total parenteral nutrition was instituted. One week later, Portagen feeds were started; the drain was subsequently removed and he continued on Portagen for 6 weeks before resuming a cow's milk based formula. He was seen at 6 and 9 months of age and remains both clinically and radiologically free of ascites.

## Discussion

Isolated accumulation of peritoneal fluid in the neonate is usually a result of urinary, chylous, biliary, or pancreatic ascites. In one study urinary ascites was the most

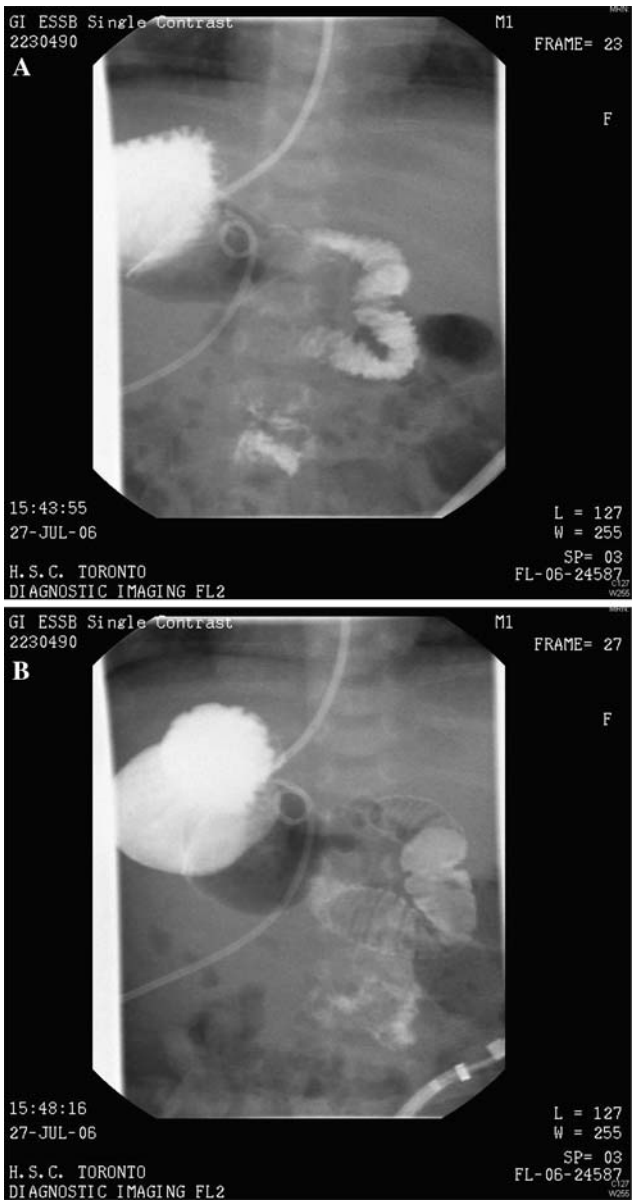


**Fig. 4** Images from the upper GI series taken of this patient in supine positioning. Given the patient's heterotaxy syndrome, his stomach is on his right side (i.e. opposite to the normal which is the left side). **a** This image showed that the patient's duodenojejunal flexure did not cross the midline and did not reach the level of the pylorus. Together, these two findings suggest the presence of a rotational anomaly. **b** The associated findings of a dilated second portion of duodenum and a corkscrew pattern distal to the duodenojejunal flexure suggested malrotation with midgut volvulus resulting in duodenal obstruction. This finding correlated clinically with the patient's symptoms of bilious vomiting

commonly reported etiology and only 4% of cases were attributed to chylous ascites [1]. Our patient had chylous ascites resulting from the leakage of lymphatic fluid into the peritoneal space. Causes of chylous ascites in children have been divided into three main etiologies: trauma, obstruction, and lymphatic abnormalities [2]. Although most cases of chylous ascites are due to lymphatic malformations, the cause in our patient may be secondary to mesenteric lymphatic obstruction associated with intestinal malrotation. Few case reports exist in the literature discussing the association of chylous ascites and malrotation [3]. In malrotation, there is incomplete retroperitoneal fixation of the mesentery of the bowel. Lymphatic obstruction from volvulus or potentially from the weight of the bowel loops may result in increased lymphatic pressure and contribute to the leakage of chyle into the peritoneum [3].

Treatment of chylous ascites is aimed at reducing lymphatic flow allowing the leak to close spontaneously. Infant formulas containing a higher proportion of medium chain triglycerides (which are directly absorbed into the portal system bypassing the lymphatics) or enteric rest with total parenteral nutrition support are the initial measures utilized. One to 2 months of conservative therapy has been suggested [4]. The response to conservative therapy has been variable [2] and controversy exists regarding the duration of this treatment modality before considering surgical intervention. In neonatal chylous ascites data on treatment is scant. Surgery is recommended for those unresponsive to medical management or with correctable intra-abdominal pathologies [5]. Our patient was successfully treated with a combination of peritoneal drainage, surgical correction of the malrotation, and use of a formula containing medium-chain triglycerides.

In addition to congenital chylous ascites and malrotation, our patient has heterotaxia syndrome, polysplenia variant. Three types of situs have been described depending on the arrangement of internal organs (lungs, liver, spleen, and atrium). In situs solitus, the viscera are in their normal positions. In situs inversus, this arrangement is reversed. Heterotaxia describes an arrangement of organs that cannot be readily determined and has two main variants, polysplenia and asplenia. Patients with heterotaxia syndromes have an increased risk of intestinal rotational abnormalities. In a recent review 3 of 45 patients with situs inversus, 1 of 16 patients with asplenia and 2 of 6 patients with polysplenia had malrotation [6]. The risk of volvulus in this setting, however, is not well characterized. It has been suggested that in cases of heterotaxia with non-rotation (cecum is in the left lower quadrant and the base of the mesentery is broad), asymptomatic patients may be followed clinically as the risk of volvulus is low [7]. However, the decision to initially manage non-operatively is highly controversial, and many pediatric surgeons would recommend surgical intervention at diagnosis to avoid the potential devastating risk of midgut volvulus. Although our patient had a contrast enema with findings consistent with a broad-based mesentery, volvulus occurred confirming that imaging studies are not definitive proof of adequacy of

bowel fixation. Therefore, cautious interpretation of radiology images should be taken when intestinal rotational abnormalities are identified in patients with situs anomalies. In addition, our patient was not asymptomatic as chylous ascites can be a manifestation of volvulus. Although our patient had an acceptable outcome, this case illustrates the hazards of delayed surgical intervention. In hindsight, urgent surgical consultation should have occurred once an intestinal rotational abnormality was identified. Surgical intervention could then have prevented the subsequent development of duodenal obstruction from midgut volvulus.

To our knowledge, this is the first reported case of a neonate with heterotaxia syndrome and intestinal malrotation presenting with chylous ascites. It also appears to be the first report in the literature of successful treatment of neonatal chylous ascites due to malrotation with the combination of peritoneal drainage, surgical correction of the malrotation, and appropriate nutritional support.

**Acknowledgments** We would like to acknowledge Dr. Kamaldine Oudjhane, Hospital for Sick Children for his assistance with interpreting the CT scan.

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