

# Successful Treatment of Congenital Chylous Ascites with Total Parenteral Nutrition

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## Abstract

Congenital chylous ascites is a rare condition. It is predominantly idiopathic in etiology and presents a therapeutic challenge. A case of idiopathic congenital chylous ascites successfully treated with six weeks of total parenteral nutrition is described.

## CASE REPORT

A girl was born at 33-1/7-weeks to a 22-year-old Gravida 2 Para 2, mother. The mother's blood type was B Rh positive. Rest of her serology was unremarkable. Pregnancy was complicated by preterm labor, polyhydramnios, fetal ascites and maternal bipolar disease. An antenatal workup for viral, cardiovascular and congenital etiologies for the ascites was negative. The infant was delivered by C-Section 6 hours after rupture of membranes. Apgar score was 1, 6 and 9 at 1, 5 and 10 minutes respectively. She was intubated, given surfactant and transported to the NICU. Her birth weight was 3570 grams, OFC was 33.5 cm and length was 48 cms. Abdomen was distended and signs of free peritoneal fluid were elicited including a fluid wave. She was hypotonic with decreased muscle strength and tone but no focal neurological findings.

Rest of the physical examination was normal. She was mechanically ventilated for 6 days mainly due to decrease in the lung volumes due to the ascites. Hypotonia led to upper airway obstruction manifesting as episodes of bradycardia and desaturation, after her extubation which responded to placing of shoulder rolls and extension of her neck.

Initial evaluation of the ascitic fluid before enteral feeds were commenced was suggestive of a transudate with specific gravity of 1.019, albumin 1.9 (serum 2.5), amylase 5, total cell count 46,800, nucleated cell count 2800, 96% lymphocytes, LDH 190, and triglycerides 12, creatinine <1.0 and protein >0.8. In view of this the following work up was done: abdominal sonogram which revealed massive ascites with normal liver, kidney and urinary bladder and normal Doppler flow in the aorta and the inferior vena cava, normal

echocardiogram, no evidence of obstruction or malrotation on an upper GI study, negative work up for congenital infections (urine shell vial culture for CMV, polymerase chain reaction for parvovirus B19 and negative titer for toxoplasma), negative work up for lysosomal storage disorders (normal skeletal survey without evidence for dysostosis multiplex, normal ophthalmological exam and negative urine screen for mucopolysaccharide) and negative work up for congenital disorders of glycosylation (normal carbohydrate deficient transferrin levels). She was fed breast milk and her ascites and respiratory distress worsened. A therapeutic abdominal paracentesis to relieve respiratory symptoms was performed and the repeat ascitic fluid examination revealed a triglyceride of 436 confirming the diagnosis of chylous ascites. Further work up for chylous ascites included: normal karyotype and chromosomal microarray analysis and normal abdominal CT without evidence of intraabdominal masses or lymphatic duct abnormalities. Ascitic fluid had no evidence of malignancy or acid fast bacillus.

Enteral feeds were continued, now switched to Portagen with further increase in abdominal girth and respiratory distress requiring therapeutic abdominal paracentesis.

This was deemed as a failure of Portagen feeds and a decision was made to stop enteral feeds and total parenteral nutrition was commenced and continued for 6 weeks. The ascites gradually resolved clinically and radiologically by 6 weeks. Portagen feeds were recommenced without clinical or radiological reaccumulation of ascites and she was discharged. Her discharge weight was 5.455 kg, length 59 cm and OFC 42 cm. Her hypotonia had improved and a

magnetic resonance imaging of her brain was normal.

### DISCUSSION

Congenital chylous ascites is a rare condition. In one study, only 4% of neonatal ascites was attributed to CCA. Most cases are idiopathic. Modalities of treatment include medium chain triglycerides feeds (MCT) <sup>1,2</sup>, total parenteral nutrition <sup>3</sup>, somatostatin therapy <sup>4</sup>, thoracic duct ligation <sup>5</sup>, application of fibrin glue <sup>6</sup>, peritoneo venous shunts <sup>7</sup>, and definitive therapy of underlying cause, if any.

For severe cases, long-term total parenteral nutrition (TPN) has been a successful option. This method, which can be administered at home, provides proper nutrition, reduces the rate of chyle flow, and is superior to MCT. However, its major drawbacks are the risks associated with the use of central venous lines and the risk of disuse atrophy of the gut.

In the reported case, the diagnosis of chylous ascites was elusive till feeds were commenced. No etiology for the ascites was found despite exhaustive investigations. A leak in the lymphatic system was inevitable due to the nature of the fluid and hence lymphoscintigraphy was considered unnecessary. A decision to stop enteral feeds and treat with parenteral nutrition was made with the backup plan to use somatostatin in case the therapy failed. Surgical intervention was the last therapeutic option in case every other intervention failed.

At present, there are only a few reported pediatric cases of chylous ascites in which TPN was the primary therapeutic regimen. A TPN regimen of 4 – 10 weeks has been suggested <sup>3,8</sup>. Unresponsiveness of chylous ascites after a 4-week course of TPN has been suggested to be an indication for exploratory laparotomy <sup>8</sup>. However a 10 week course of TPN has been demonstrated to be effective in resolving the ascites <sup>3</sup>. The reported case may make it possible for care

givers to consider a 6 week course of TPN for successful amelioration of congenital chylous ascites. Mechanism of resolution following TPN is not known but may involve a maturation of the lymphatic system over the time course of the TPN. It is also possible that a leak in the lymphatic system would be repaired over the period of the treatment.

Since the majority of cases of CCA resolve spontaneously, surgical exploration is recommended for those unresponsive to conservative treatment or with correctable intra-abdominal pathologies. Surgery aimed at repair of lymphatic leaks, and relief of lymphatic obstruction is definitive but palliation with peritoneo-venous shunts has been attempted in resistant cases. Shunts are at risk of blockage due to the high viscosity of chylous fluids.

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