

Case Report

Parenteral nutrition combined with rice soup can be a safe and effective intervention for congenital chylous ascites

Yi Cao MD^{1,2,3}, Weihui Yan MD, PhD^{1,2,3}, Lina Lu MD^{1,2,3}, Yijing Tao MD^{1,2,3},
Wei Lu MD^{1,2,3}, Yingwei Chen MD, PhD^{1,2,3}, Qingya Tang MD^{1,2,3}, Wei Cai MD, PhD^{1,2,3}

¹Department of Pediatric Surgery, Xin Hua Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai, China

²Shanghai Institute of Pediatric Research, Shanghai, China

³Shanghai Key Laboratory of Pediatric Gastroenterology and Nutrition, Shanghai, China

Congenital chylous ascites in the neonatal period is a rare entity. Total parenteral nutrition (TPN), medium chain triglyceride (MCT)-based diet, octreotide and repeated paracentesis are regarded as appropriate medical treatment for congenital chylous ascites, and surgery is recommended when conservative therapy has failed. We present two cases in which ascites were confirmed via an abdominal sonogram and diagnostic paracentesis. In our clinical experience, rice soup combined with PN can be a safe and effective intervention.

Key Words: congenital chylous ascites, neonate, optimal treatment, rice soup, parenteral nutrition

INTRODUCTION

Congenital chylous ascites are rare clinical entities that arise due to numerous factors and with changing dynamics of the disease. The treatment is multidirectional with several therapeutic steps, starting with the least invasive to surgical treatment, usually as a last resort. Because of the lack of therapeutic standards, this disease continues to be a medical challenge, and the therapeutic approach extremely individual. Here we present two infants with congenital chylous ascites. Experience with them suggests that rice soup combined with parenteral nutrition (PN) can be an efficacious and safe approach to management.

CASE REPORTS

Case 1

A four-month-old male infant was transferred to the Department of Pediatric Gastroenterology and Nutrition of our hospital due to abdominal distension. He was born full term by Caesarean section with a birth weight of 3,500 g. Fetal ascites was found by antenatal sonography performed at 34 weeks of gestation. The mother was a 29-year-old primigravida. The parents were not consanguineous, and family history was negative for lymphatic disorders.

On admission, physical examination revealed massive abdominal distension and bilateral hydroceles. The abdominal girth was 55 cm. Abdominal computerized tomography (CT) indicated massive ascites and bilateral hydroceles. Ultrasonogram showed massive ascites without pleural effusion or other abdominal anomalies. Both echocardiography and electronic gastroscopy revealed no abnormalities. Laboratory examination included: liver

function count as alanine aminotransferase (ALT): 18 U/L, aspartate aminotransferase (AST): 56 U/L, total protein: 55.9 g/L and albumin: 31.3 g/L; and normal blood count as white blood cells (WBC): 5,130/mm³, platelets (PLT): 684,000/mm³ and hemoglobin (Hb): 103 g/L. Renal and coagulation functions were normal. Abdominal paracentesis yielded the milky ascitic fluid with a raised WBC of 351×10⁶/L, predominantly lymphocytes (52%), and the Rivalta test was strongly positive. Combined with the positive result of the ascites chyle test, the chylous nature of the ascites was confirmed. The results of both the blood microfilaria and feces parasite eggs tests were negative.

The infant was initially fasted and was supported by total parenteral nutrition (TPN) [20% medium-chain triglycerides (MCT) / long-chain triglycerides (LCT) lipid: 2 g/kg/d, calorie: 72 kcal/kg/d]. Additionally, octreotide (a somatostatin analogue), which was to be continued until discharge, was started at a dose of 0.8 µg/kg/h. On days 1, 5 and 9 after admission, abdominal paracentesis was performed three times in total, and the volumes were 105 mL, 95 mL and 25 mL, respectively, accompanied by regular measurement of abdominal girth and review of

Corresponding Author: Prof Wei Cai, Department of Pediatric Surgery, Xin Hua Hospital, Shanghai Jiao Tong University School of Medicine, No. 1665, Kong Jiang Road, Yangpu, Shanghai, China.

Tel: +86 21 65791316; Fax: +86 21 65791316

Email: caiw1978@163.com

Manuscript received 05 May 2015. Initial review completed and accepted 26 May 2015.

doi: 10.6133/apjcn.092015.10

ultrasonogram. On the 11th day, the baby was given 50% MCT-based formula (Peptijunior®; Nutricia advanced medical nutrition, Cuijk, Holland), and the nutrient solution was decreased correspondingly due to evidence of remarkable reduction of ascites on an abdominal ultrasonogram and based on physical examination. On the 15th day, the formula increased while PN was stopped. Repeated ultrasonogram showed that ascites did not increase after four days of full enteral feeding. Then, the patient was discharged.

Two weeks after discharge, at the age of five months old, the baby was rehospitalized with recurrent ascites. Vital function was stable; however, the abdomen was again massively distended. TPN, octreotide and abrosia were restarted, and abdominal paracentesis was given twice during this period of hospitalization. On the 15th day, unfortunately, the liver function of the baby was impaired as indicated by elevated ALT (199 U/L) and AST (283 U/L). Considering that the impairment may be associated with TPN, the baby was given rice soup, and the nutrient solution was reduced by nearly one-half (20% MCT/LCT lipid: 0.8 g/kg/d, calorie: 25 kcal/kg/d). Meanwhile, liver-protection drugs, such as polyene phosphatidylcholine and compound glycyrrhizin, were injected. On the 25th day, the liver function returned to normal after ten days of the liver-protection drugs. On the 35th day, an abdominal ultrasonogram indicated that the number ascites decreased. Hence, the patient was discharged and then fed with 50% MCT-based formula and rice soup. At the age of eight months old, the patient was weighed 7.5 kg (<P₁₅) and had a height of 71 cm (P₅₀-P₈₅). Repeated abdominal ultrasonogram showed almost no ascites in the abdominal cavity.

Case 2

A five-month-old male infant was transferred to the Department of Pediatric Surgery because of abdominal distension. He was born full term with a birth weight of 3,100 g by eutocia. He had no history of injury or abdominal surgery. As his mother lived in a remote village, she did not bring him in for antenatal care regularly, and it could not be ascertained if there were any fetal ascites. The mother was a 23-year-old primigravida. The parents were not consanguineous, and the family history was negative for lymphatic disorders.

Before admission, the baby received diagnostic abdominal paracentesis at the local hospital. The paracentesis yielded 50 mL of milky ascitic fluid, and the chyle test was positive. On admission, physical examination revealed abdominal distension and shifting dullness existed. Abdominal CT and ultrasonogram showed moderate ascites without other abdominal anomalies. Echocardiography revealed no abnormalities. Laboratory examination included: liver function count as ALT: 102 U/L, AST: 172 U/L, total protein: 27.4 g/L and albumin: 18.2 g/L; normal blood count as WBC: 8,290/mm³, PLT: 688,000/mm³; and Hb: 141 g/L. Renal and coagulation functions were normal. The results of both blood microfilaria and feces parasite eggs test were negative.

The infant was initially fed rice soup and PN (20% MCT/LCT lipid: 1 g/kg/d, calorie: 31 kcal/kg/d) at the same time. During hospitalization, human albumin at a

dose of 0.8 g/kg/d was given four times in total, and each time, furosemide was injected thereafter. On the 18th day, ultrasonogram revealed no ascites in the abdomen. Liver function count was: ALT: 23 U/L, AST: 34 U/L, total protein: 35.6 g/L and albumin: 26.6 g/L; normal blood count was indicated by WBC: 3,390/mm³, PLT: 106,000/mm³ and Hb: 106 g/L. After 20 days of hospitalization, the patient was discharged at nearly six months of age. The baby was fed rice soup and 50% MCT-based formula (Peptijunior®; Nutricia advanced medical nutrition, Cuijk, Holland) after discharge. At the age of eight months, complementary feeding was added to the diet. The patient was fed a normal diet at 10 months of age. A 2.5-year follow-up study has revealed no recurrence of the ascites with a weight of 14 kg (P₅₀) and a height of 101 cm (>P₈₅). Nonspecific performance, including abdominal distension, edema and hypoalbuminemia, has not appeared after discharge.

DISCUSSION

Chylous ascites may arise for a variety of reasons, which include developmental defects of the lymphatic system, nonspecific bacterial and parasitic infection, liver cirrhosis, malignant neoplasm, blunt abdominal trauma and surgical injury. Congenital chylous ascites is probably the result of maldevelopment and immaturity of lymphatic vessels, which are the most common cause in the neonatal period.¹ Dysplastic lymphatic vessels, both hypo- and hyperplastic, favor lymphatic leakage in the mechanism of increasing pressure in the lymphatic system and lymphatic congestion.² Delayed maturation of lymphatic vessels, which lead to abnormal structure of their walls, also lead to a condition called "leaky lymphatics", which results in lymph accumulation in the peritoneal cavity.³

The initial step to confirm the presence of ascites is ultrasonography of the abdomen. Due to its noninvasiveness, it can be repeated; thus, it is possible to evaluate the course of the pathological process. Analysis of the fluid obtained by abdominal paracentesis is the key stage in the diagnosis of this disease. The ascitic fluid, known as chyle, is usually color-free or milky white. The protein and triglyceride content is usually high with predominance of lymphocytes on differential count.⁴ The presence of chylomicrons is considered to be pathognomonic.² However, the next step to determine the underlying cause of the chylous ascites is the true challenge because it is technically difficult to carry out and a high risk exists in newborns and infants.⁵ The initial diagnostic investigation includes ultrasonography and CT or magnetic resonance imaging (MRI) of the abdomen to exclude conditions that necessitate immediate surgical intervention. Diagnosis of malformation of the lymphatics is suspected when all results are negative. Further diagnostic tests, such as lymphangiography and lymphoscintigraphy, are imperative if surgery is decided, with the purpose of identifying the site of the leakage of chyle preoperatively. However, it is usually difficult to clearly show leakage.

It has been reported that the primary therapeutic modality for congenital chylous ascites is abrosia and TPN.⁶⁻⁸ The mechanism of this resolution may involve maturation of the lymphatic system over the time course of the

TPN. It is also conducive to reduce lymph flow in the peritoneal cavity so that the leak in the lymphatic system may be repaired during the treatment period. According to Bhatia et al, a satisfactory result can be obtained after 3-4 weeks of treatment. The success rate is 60-100%.⁹ Although TPN is considered as the "first-line" intervention, there are obvious drawbacks, such as the risks of liver injury, the risks associated with the use of central venous lines and the risk of diffuse atrophy of the gut.⁵ TPN-associated liver injury was observed once in our case. Based on our experience with the second successful case, PN combined with rice soup, which is long-chain fats (LCT)-free, may be a better "first-line" intervention because it reduces the risk of long-term use of TPN, and it is also effective.

As chylous ascites has decreased or remained stable, the next stage is the withdrawal of PN, with introduction of enteral nutrition. Dietary management is an important treatment modality in chylous ascites. An MCT-based diet has even been accepted as the first measure for reducing chyle production.¹⁰ After it is absorbed by the intestinal mucosa, MCT bypasses the enteric lymphatics and directly enters the portal system. Not only can nutrients be provided by the MCT-based diet, but it also reduces lymphatic flow and pressure within the lymphatic system and decreases the amount of lymph leakage. However, it also has been found that long-term use of the MCT-based diet, which is necessary for the therapeutic effect, has an estimated success rate of approximately 30%.¹⁰ Liao et al reported one case of neonatal chylous ascites that was put on a strict MCT formula, but the abdominal girth continued to increase.¹¹ Consistent with these findings, our first case showed recurrent ascites after a period of feeding with 50% MCT-based formula. Feeding rice soup only or combined with an MCT diet seems to be more effective.

Paracentesis is not only diagnostic but is also a therapeutic method in the management of chylous ascites.² Despite several definite complications, repeated paracentesis is commonly included in conservative treatment regimens to relieve respiratory insufficiency and abdominal distention.¹² The operation should be aseptic to prevent bacterial peritonitis, and rapid or hard lower abdominal pressure should be avoided. Based on our experience, paracentesis is an important supplemental method when ascites are too difficult to manage; otherwise, it is not essential.

Somatostatin analogues, such as octreotide, have been demonstrated to be effective in reducing lymphorrhea. There have been reports of successful use of octreotide in neonatal chylothorax.¹³ The exact mechanisms by which somatostatin prevents lymphatic flow are not completely understood. It has been previously shown to decrease the intestinal absorption of fats, lower triglyceride concentration in the thoracic duct and attenuate lymph flow in the major lymphatic channels.¹⁴ Satisfactory results were achieved by the administration of the octreotide combined with TPN.¹⁴⁻¹⁵ Four case reports of patients with congenital chylous ascites described successful treatment with octreotide.¹⁶ The octreotide dose required for satisfactory outcome has been reported to be 1-2 µg/kg/h of infusion.^{1,16} However, the somatostatin dose in our first

case was below this minimum effective concentration, making it difficult to determine whether it is a helpful treatment. Furthermore, it was not used in our second case, who responded to the combination of rice soup and PN.

Surgery is advised in cases that are resistant to conservative treatment, which is usually undertaken for 4-8 weeks.¹⁷ The success of the operation depends on identifying the site of leakage of the lymphatic duct.¹⁸⁻¹⁹ A shunt joining the peritoneal cavity with the venous system is a last resort because of the possibility of severe complications and limited effectiveness.^{2,5} Due to invasiveness and difficulty, surgery is not the preferred treatment.

Rice soup is home-made for our patients. 50 g rice and 500 mL water are brought to a boil. Rice soup contains only carbohydrate with little fat, and one portion contains 169 kcal, while rice is 337 kcal/100 g. It can reduce lymphatic flow and pressure as a LCT-free formula, and it also decreases the risks associated with TPN, including liver injury and diffuse atrophy of the gut. Moreover, the combination of rice soup with an MCT diet seems to be more effective compared with the low success rate of an exclusive MCT-based diet.¹⁰⁻¹¹

In conclusion, the combination of PN and rice soup may be a better 'first-line' intervention because it decreases the risk of long-term TPN and is effective at the same time. After withdrawal of PN, feeding rice soup combined with an MCT diet, as a transition, is relatively nutritious and effective. When repeated, the ultrasonogram shows a gradual reduction in ascites or no recurrence, the proportion of MCT diet can gradually increase and complementary feeding can be added to the diet. A patient-specific approach is required for chylous ascites with its various causes should start as soon as possible. Further studies of more patients with chylous ascites are needed before a treatment algorithm can be in place with confidence.

AUTHOR DISCLOSURES

No competing interests are reported.

REFERENCES

1. Purkait R, Saha A, Tripathy I, Roy B. Congenital chylous ascites treated successfully with MCT-Based formula and octreotide. *J Indian Assoc Pediatr Surg*. 2014;19:175-7. doi: 10.4103/0971-9261.136480.
2. Campisi C, Bellini C, Eretta C, Zilli A, da Rin E, Davini D, Bonioli E, Boccardo F. Diagnosis and management of primary chylous ascites. *J Vasc Surg*. 2006;43:1244-8. doi: 10.1016/j.jvs.2005.11.064.
3. Beshay VE, Beshay JE, Rosenberg AJ. Chylous ascites: a case of child abuse and an overview of a rare condition. *J Pediatr Gastroenterol Nutr*. 2001;32:487-9. doi: 10.1097/00005176-200104000-00019.
4. Cardenas A, Chopra S. Chylous ascites. *Am J Gastroenterol*. 2002;97:1896-900. doi: 10.1111/j.1572-0241.2002.05911.x.
5. Karagol BS, Zenciroglu A, Gokce S, Kundak AA, Ipek MS. Therapeutic management of neonatal chylous ascites: report of a case and review of the literature. *Acta Paediatr*. 2010; 99:1307-10. doi: 10.1111/j.1651-2227.2010.01818.x.
6. Lee YY, Soong WJ, Lee YS, Hwang B. Total parenteral nutrition as a primary therapeutic modality for congenital chylous ascites: report of one case. *Acta Paediatr Taiwan*. 2002;43:214-6.

7. Alliet P, Young C, Lebenthal E. Chylous ascites: total parenteral nutrition as primary therapeutic modality. *Eur J Pediatr.* 1992;151:213-4. doi: 10.1007/BF01954387.
8. Smeltzer DM, Stickler GB, Fleming RE. Primary lymphatic dysplasia in children: chylothorax, chylous ascites, and generalized lymphatic dysplasia. *Eur J Pediatr.* 1986;145:286-92. doi: 10.1007/BF00439402.
9. Bhatia C, Pratap U, Slavik Z. Octreotide therapy: a new horizon in treatment of iatrogenic chyloperitoneum. *Arch Dis Child.* 2001;85:234-5. doi: 10.1136/adc.85.3.234.
10. Chye JK, Lim CT, Van der Heuvel M. Neonatal chylous ascites--report of three cases and review of the literature. *Pediatr Surg Int.* 1997;12:296-8. doi: 10.1007/BF01372154.
11. Liao HB, Hwang RC, Chu DM, Chu ML, Chu CC, Hwang EJ. Neonatal chylous ascites: report of two cases. *Zhonghua Min Guo Xiao Er Ke Yi Xue Hui Za Zhi.* 1990;31:47-52. (In Chinese)
12. Aalami OO, Allen DB, Organ CH, Jr. Chylous ascites: a collective review. *Surgery.* 2000;128:761-78. doi: 10.1067/msy.2000.109502.
13. Goto M, Kawamata K, Kitano M, Watanabe K, Chiba Y. Treatment of chylothorax in a premature infant using somatostatin. *J Perinatol.* 2003;23:563-4. doi: 10.1038/sj.jp.7210975.
14. Collard JM, Laterre PF, Boemer F, Reynaert M, Ponlot R. Conservative treatment of postsurgical lymphatic leaks with somatostatin-14. *Chest.* 2000;117:902-5. doi: 10.1378/chest.117.3.902.
15. Laterre PF, Dugernier T, Reynaert MS. Chylous ascites: diagnosis, causes and treatment. *Acta Gastroenterol Belg.* 2000;63:260-3.
16. Huang Y, Zhuang S, Li Y, Liu M, Chen H, Du M. Successful management of congenital chylous ascites in a premature infant using somatostatin analogue. *Indian J Pediatr.* 2011;78:345-7. doi: 10.1007/s12098-010-0256-1.
17. Romanska-Kita J, Borszewska-Kornacka MK, Dobrzanska A, Rudzinska I, Czech-Kowalska J, Wawrzoniak T. Congenital chylous ascites. *Pol J Radiol.* 2011;76:58-61.
18. Mitsunaga T, Yoshida H, Iwai J, Matsunaga T, Kouchi K, Ohtsuka Y, Okada T, Hishiki T, Ohnuma N. Successful surgical treatment of two cases of congenital chylous ascites. *J Pediatr Surg.* 2001;36:1717-9. doi: 10.1053/jpsu.2001.27973.
19. Antao B, Croaker D, Squire R. Successful management of congenital chyloperitoneum with fibrin glue. *J Pediatr Surg.* 2003;38:E7-8. doi: 10.1016/j.jpedsurg.2003.08.011.

Case Report

Parenteral nutrition combined with rice soup can be a safe and effective intervention for congenital chylous ascites

Yi Cao MD^{1,2,3}, Weihui Yan MD, PhD^{1,2,3}, Lina Lu MD^{1,2,3}, Yijing Tao MD^{1,2,3},
Wei Lu MD^{1,2,3}, Yingwei Chen MD, PhD^{1,2,3}, Qingya Tang MD^{1,2,3}, Wei Cai MD, PhD^{1,2,3}

¹Department of Pediatric Surgery, Xin Hua Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai, China

²Shanghai Institute of Pediatric Research, Shanghai, China

³Shanghai Key Laboratory of Pediatric Gastroenterology and Nutrition, Shanghai, China

静脉营养联合米汤优化治疗先天性乳糜腹

先天性乳糜腹在新生儿时期较为罕见，目前其主要治疗手段包括全肠外营养、中链脂肪酸为基础的饮食、奥曲肽以及反复腹腔穿刺引流术，当保守治疗失败时则建议手术。本文提供了两例通过腹部超声和诊断性腹腔穿刺确诊的先天性乳糜腹病例。根据我们的临床治疗经验，口服米汤结合肠外营养是先天性乳糜腹的一种安全有效的治疗方法。

关键词：先天性乳糜腹、新生儿、最佳治疗、米汤、肠外营养