

# Successful Treatment of Neonatal Idiopathic Chylous Ascites with Total Parenteral Nutrition and Somatostatin

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## Abstract

In this paper we report a case of neonatal chylous ascites the diagnosis of which was made by lymphoscintigraphy and exclusion of all other organic diseases. Initial treatment with skimmed milk feeding was not effective, hence repeated abdominal paracentesis was required to relieve the distension. Total parenteral nutrition combined with intravenous somatostatin eventually resulted in long-term remission.

**Key words** Chylous ascites; Somatostatin

## Introduction

Chylous ascites is not commonly seen in the neonatal period. The etiology of most cases of chylous ascites remains unknown. Congenital chylous ascites is primarily related to inadequate lymph drainage as a result of maldevelopment of the intra-abdominal lymphatic duct. Here we describe an infant with congenital chylous ascites who improved after total parenteral nutrition (TPN) and somatostatin administration.

## Case Report

A 50-day-old male baby was referred to our hospital because of abdominal distention. He was born full term by cesarean section and the immediate post-natal course was unremarkable. No evident dysmorphic features were found on physical examination. However, abdominal distention was noted on D10 and the abdominal girth increased

gradually. Ultrasonogram revealed massive ascites, the liver and spleen appeared normal, and no pleural effusion was found. Blood count showed: WBC  $4.9 \times 10^9/L$  with 57% lymphocytes, PLT  $368 \times 10^9/L$ , and Hb 111 g/L. Routine urinalysis was normal. Serum calcium was 2.62 mmol/L. There was no hypoalbuminaemia with a serum albumin content of 37.6 g/L. Triglycerides and cholesterol in the blood was 1.47 mmol/L and 3.09 mmol/L, respectively. Abdominal paracentesis yielded milky ascitic fluid with the following data: WBC 4000 cells/mm<sup>3</sup> with 20% neutrophils, 80% lymphocytes, protein 35 g/L, glucose 4.9 mmol/L. Chylous test (direct visualisation of fat globules under the microscope after extraction with ether) was positive. The diagnosis of chylous ascites was confirmed. Further tests were performed. There was no evidence of congenital TORCH infections in this infant. Serum electrolytes, liver and renal functions were normal. Abdominal CT scan revealed no abnormality.

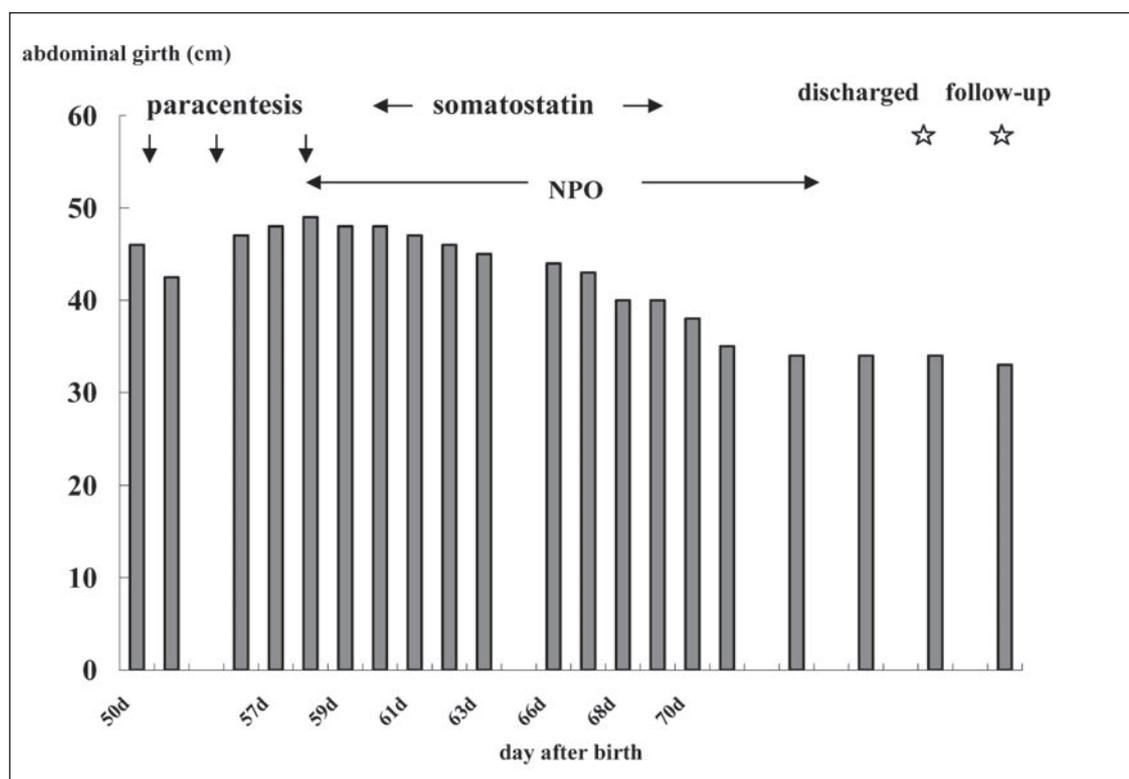
The baby was fed skimmed milk (Bright Brand skimmed milk 光明牌脱脂奶粉, Bright Dairy Products, Shanghai, China with fat content of less than 2 g/100 g) and partial parenteral nutrition was given at first. Abdominal paracentesis was done twice a week. However his abdominal girth continued to increase markedly (Figure 1). At 10 days after admission, the maximum girth reached 49 cm. Doppler color ultrasonogram showed absence of stenosis of the inferior vena cava and venous thrombosis. Lymphoscintigraphy with technetium (Tc) <sup>99m</sup>-dextran revealed an accumulation of radioactivity bilaterally in the

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**Figure 1** Changes of abdominal girth during treatment. Initial poor response to paracentesis and skimmed milk is shown, followed by a good response to somatostatin and total parenteral nutrition. There was no recurrence of ascites after 4 months of follow-up.

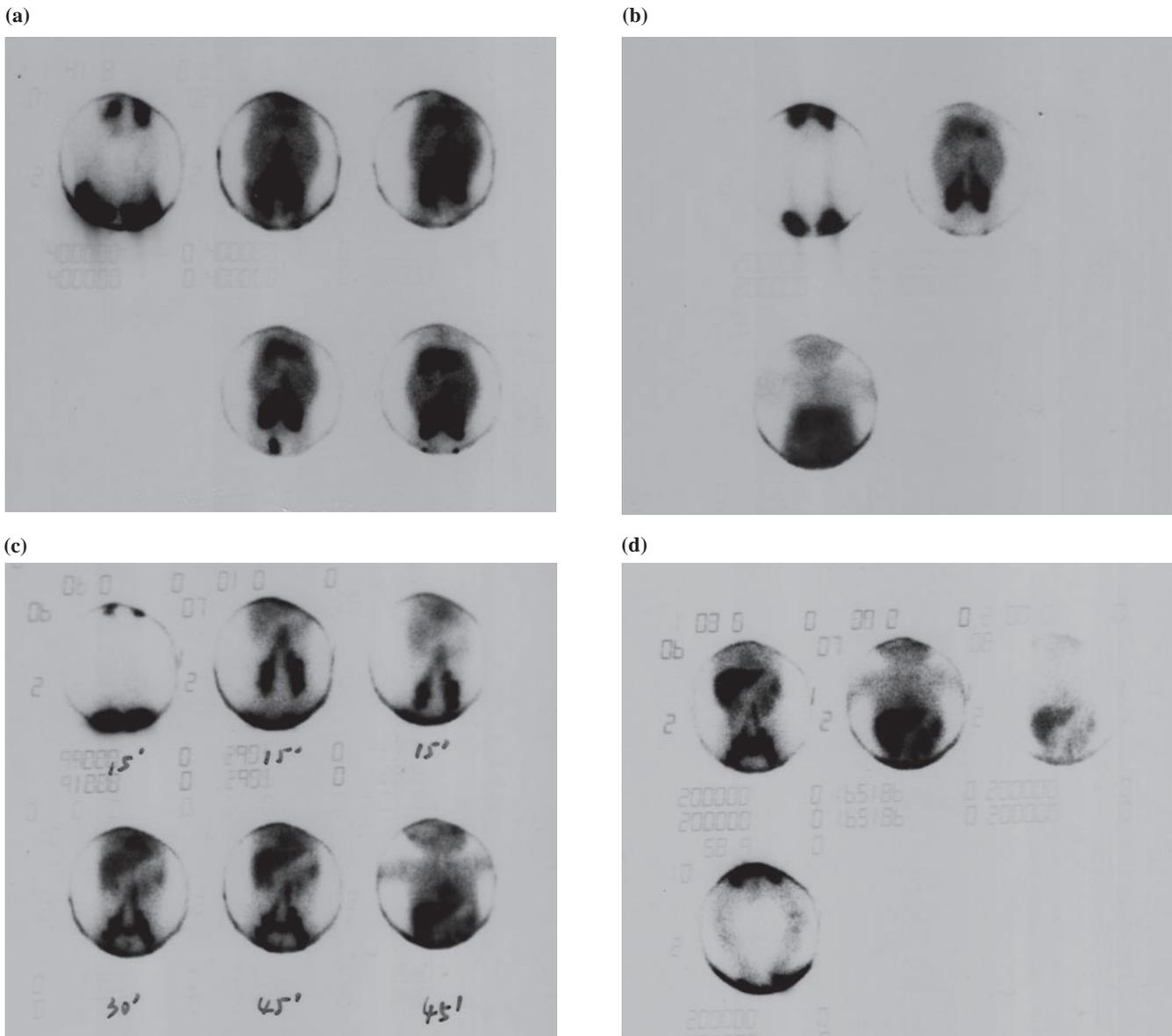
renal areas which eventually spread to the peritoneal cavity. The liver was not clearly visualised 6 hours after injection (Figures 2a and 2b). These results suggested bilateral obstruction of the lymphatic ducts and leakage into the abdomen.

One week after hospitalisation, enteral feeding was discontinued, and total parenteral nutrition was reinstated. After obtaining informed consent from the parents, the baby was fasted and intravenous infusion of somatostatin ( $1.5 \mu\text{g}/\text{kg}/\text{h}$ ) was initiated on the second day, to be continued over the next ten days. Abdominal girth and abdominal tension decreased gradually. Abdominal girth reduced markedly after seven days. From the 11th day post-infusion onwards, the girth became stable at 35 cm. Throughout the entire treatment course, no side effects of somatostatin, such as diarrhoea, hypoglycaemia or hypotension, were found. No ascites was seen upon abdominal ultrasonogram follow-up examination. Enteral feeding (skimmed milk) was re-introduced after fasting for 4 weeks, and full enteral feeding was established over the next two weeks. The patient was

discharged from our hospital shortly thereafter. Follow-up at 4 months showed no recurrence of chylous ascites on a normal diet.  $^{99\text{m}}\text{Tc}$ -dextran lymphoscintigraphy was repeated (Figures 2c and 2d). There was no accumulation of radioisotope in the body, and background radioactivity of the peritoneal cavity was very low. The liver was clearly visualised 6 hours after injection. These results indicated unobstructed lymph flow, without any lymphatic leakage.

## Discussion

Here we reported a case of congenital chylous ascites with suspected obstruction of the lymphatic duct. Affected neonates usually present with abdominal distention and respiratory distress.<sup>1</sup> The treatment and prognosis of the patients depend on the cause of chylous ascites. Chylous ascites may be the result of developmental defect of the lymphatic system, nonspecific bacterial, parasitic and tuberculous peritoneal infection, liver cirrhosis, malignant



**Figure 2** Lymphoscintigraphy before and after treatment. (a) (Before treatment)  $^{99m}\text{Tc}$ -Dextran was injected into the web between the first and second toe. Images of the lower limbs and whole abdomen were taken with a Parallel-hole Collimator 15, 30 and 45 minutes after the injection. At 15 minutes, uptake was seen at the bilateral groin and femoral lymph nodes which appeared to be enlarged. Bilateral lumbar trunks were faintly visualised. Diffusely distributed radiation was seen all over the abdomen. The liver was faintly discernible. (b) (Before treatment) Delayed image was taken 6 hours after injection of  $^{99m}\text{Tc}$ -Dextran. No significant changes visualised. Conclusion: Suggestive of bilateral lumbar trunk obstruction. (c) Scintigraphy (15, 30, 45 minutes after injection) taken 4 months later. In the 15-minute image, bilateral inguinal and femoral lymph nodes were clearly demonstrated. Bilateral lumbar trunks and cysterna chili were seen. Hepatic uptake was faintly visualised. In the 45-minute image, uptake at the inguinal and femoral lymph nodes has diminished while the hepatic uptake became more intense. (d) In this delayed (6-hour) image taken 4 months later, the liver was clearly visualised and no obvious changes were seen in the other organs. Conclusion: No significant abnormality seen in bilateral lymphatic imaging.

neoplasm, blunt abdominal trauma and surgical injury.<sup>2,3</sup> In adults the most common cause is abdominal malignancy, while in children, congenital lymphatic abnormalities are more common. Weber et al<sup>4</sup> reviewed 51 children with congenital chylous ascites from 1877 to 1972. The male to female ratio was 2:1. Seventy-seven percent had ascites at birth, the latest being diagnosed at 6 months. No specific cause of the chylous ascites was found in thirty of them. Seven underwent laparotomies with no abnormalities found. However, abnormalities were found in 21 patients, including: aplasia of the thoracic duct, occlusion of the thoracic duct, lymphangiectasia, band over the duct at the mesenteric root, tiny small vessels, large lymph nodes of the mesenteric root, etc. One child with lymphatic leak at the mesenteric root was operated on successfully at 2 weeks. Sixty-five percent survived from 3 months to 15 years. Twenty-seven percent died of infection, and this high mortality due to infection was not seen today because of advances in management.

Ultrasonogram of the abdomen confirms the presence of ascites. The most useful diagnostic method is paracentesis.<sup>5</sup> Although lymphangiography is the gold standard in defining the cause of the lymphatic obstruction, it is an invasive procedure. By injecting <sup>99m</sup>Tc-dextran into the interdigital web spaces, lymphoscintigraphy was able to evaluate the patency of the lymphatics. This more physiological technique is less invasive and should be used, especially when lymphangiography is contraindicated. More recently, there have been many reports on the successful identification of lymphatic leaks through lymphoscintigraphy. It is also more acceptable as a follow-up imaging procedure because it is safer and less invasive. In this case, we have used <sup>99m</sup>Tc-dextran as the contrast medium. <sup>99m</sup>Tc-labelled sulphur colloid or <sup>99m</sup>Tc-labelled albumin are also used in other countries. Compared with the colloidal preparations, <sup>99m</sup>Tc-dextran whole body uptake is faster and more intense while uptake at the lymph nodes may be less intense.

Previous investigators suggested that congenital chylous ascites should be treated conservatively at first.<sup>6</sup> In a series of 103 Japanese patients, 63.5% were cured by conservative treatment. The average treatment period was 63.5 days.<sup>7</sup> Most of the cases are functional defects of the lymphatic chain such as "leaky lymphatics", and structural abnormalities of the lymphatic system in infants are rare.<sup>1</sup> The goal of conservative treatment is to provide "gut rest" and decrease the intestinal secretions. Conservative management includes enteral feeding with a formula high in medium-chain triglycerides (MCT), low in long-chain

triglycerides (LCT) and enhanced in protein content; and repeated abdominal paracentesis. Although these methods are usually successful, enteral feedings, even just clear water, have been shown to increase thoracic duct lymph flow.<sup>8</sup> Liao et al<sup>9</sup> reported one case of neonatal chylous ascites that was put on a strict medium-chain triglycerides formula. The abdominal girth continued to increase, however, and warranted readmission at 8 months of age.

Theoretically TPN is superior to enteral feedings because the bowel is bypassed. Abdominal paracentesis can rapidly relieve respiratory insufficiency and abdominal discomfort. However, it may induce critical losses of fluid, lymphocytes, protein, coagulation factors and antibodies,<sup>10</sup> hence increases chances of infection, reduces immune function and may result in more ascites. It did happen in our patient. Some studies suggested prolonged nonoperative management, avoiding paracentesis if possible.<sup>11</sup> Surgical intervention was recommended if 1 to 2 months of conservative approach failed. The success of the operation depended on identifying the site of leakage of the lymphatic duct.<sup>5,7,12</sup>

In our patient, enteral skimmed milk feeding and repeated abdominal paracentesis did not have any significant effect. Total parenteral nutrition combined with intravenous infusion of somatostatin provided satisfactory relief. There have been reports of successful use of somatostatin in neonatal chylothorax,<sup>10,13-15</sup> chylous ascites after liver transplantation,<sup>3</sup> Kasai portoenterostomy,<sup>16</sup> or chylous ascites caused by various disorders.<sup>17</sup> It has been hypothesised that somatostatin arrests lymphatic flow through reduction in gastric, intestinal, and pancreatic secretions or by a decrease in hepatic venous pressure and splanchnic blood flow. Since the gastrointestinal secretory volume and enzymes are decreased, there is a resultant decrease in the volume and protein/triglyceride content of the fluid in the lymphatic duct.<sup>10,14</sup> Somatostatin has been used in children with pancreatitis, severe upper gastrointestinal bleeding, enteric fistula or neonatal hyperinsulinism. No serious side effect was found.

In this case, a marked reduction of chylous ascites was observed after somatostatin infusion and abdominal girth returned to normal within 11 days. The patient was in good general condition. Blood pressure remained stable during treatment with somatostatin. None of the reported side-effects were noted in our patient.<sup>18</sup> Total parenteral nutrition and administration of somatostatin shortened the time of hospitalisation and surgery was prevented. These results suggest that TPN along with somatostatin can stop lymphatic leakage, and clinical symptoms improve rapidly and

effectively. As the causes of chylous ascites are multifactorial, it is important to emphasize a patient-specific approach to appropriate treatment. Further investigation into the use of somatostatin in chylous ascites is needed.

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