

## CONGENITAL CHYLOUS ASCITES: REPORT OF FOUR CASES AND REVIEW OF THE LITERATURE

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This is a report of four neonates with chylous ascites, three of whom were recognized antenatally. The neonates were born at 34, 35 and 37 weeks' gestation, respectively. Their birth weights ranged from 1270 g to 3500 g, and all had abdominal distention at birth. Analyses of the ascites fluid revealed raised levels of triglycerides, cholesterol and protein. Three of the cases responded to low-fat diet and medium-chain triglycerides with multiple paracentesis drainage. In the fourth case, the chylous ascites was associated with a macrodigit involving the right index and medius. The condition persisted despite 10 weeks of parenteral alimentation, and only resolved after an exploratory laparotomy done at the age of two months.

### Case Reports

#### Case 1

The patient was a 1390 g male born after 34 weeks' gestation to a 25-year-old gravida 3 para 3. The ascites was detected by ultrasonography at 31 weeks of gestation (Figure 1). At birth, the abdomen was very distended. Paracentesis was performed and 200 mL of ascitic fluid was collected, showing a yellowish fluid rich in lymphocytes, with triglycerides of 1.4 mmol/L, cholesterol of 1.6 mmol/L, protein of 2.4 g/L and a specific gravity of 1.015. Because of the severe abdominal distention, a second paracentesis was done after three days, and enteral feeding was started at the age of six days by a formula rich in medium-chain triglycerides. The chylous ascites resolved completely within two weeks, and after 10 years of follow-up, no complications have been noticed.

#### Case 2

This patient was a 1270 g male born after 34 weeks' gestation to a 32-year-old unbooked gravida 7 para 7. All his siblings had died in the neonatal period in another hospital. At birth, the patient presented with hypotonia, bilateral cataract and facial dysmorphism. Cytogenetic study

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of peripheral blood lymphocytes cultured by G-binding technique showed a mosaic (47xy+ marker chromosomes at 80%, and 46xy at 20%) chromosomal constitution in the analyzed cells. The abdomen was distended, and paracentesis showed a 150 mL milky fluid rich in lymphocytes, with triglycerides of 3 mmol/L, cholesterol of 2.2 mmol/L, and protein of 3.6 g/L. Ultrasound and CT scan confirmed the presence of hamartoma of the liver (Figure 2). The chylous ascites resolved completely after 10 days of parenteral alimentation, but the patient remained ventilator-dependent and died of respiratory problems at four months of age.

#### Case 3

This patient was a 3500 g female born after 37 weeks' gestation to a 32-year-old diabetic gravida 6 para 5. The ascites was detected by ultrasound at 35 weeks, with polyhydramnios. At birth, the patient presented with a small interventricular septal defect and a huge abdominal distention and respiratory distress. An urgent intra-peritoneal drain was inserted and a yellowish fluid was extruded which was rich in lymphocytes, with triglycerides of 3.3 mmol/L, cholesterol of 2.3 mmol/L, protein of 2.7 g/L, and a specific gravity 1.061.

In spite of complete and strict parenteral alimentation, the peritoneal drainage produced more than one liter of fluid in the first two weeks. The drainage was removed at the end of the third week, and multiple needle aspirations were done until the age of two months, when an exploratory laparotomy was performed. No macroscopic anomalies were found, but the origin of the superior mesenteric vessels was explored as recommended in the literature. At the age of two months, a macrodigit was also evident involving the index and medius. The index was amputated at two years of age. Following surgery, the patient completely recovered and continued to grow and develop normally at five years' follow-up.

#### Case 4

This patient was a 2600 g male born after 35 weeks' gestation to a gravida 1 para 1. The ascites was detected at 32 weeks with polyhydramnios. At birth, the abdomen was

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very distended, and the patient also had mild respiratory distress. A peritoneal drain was inserted which collected about 700 mL of yellowish fluid rich in lymphocytes,

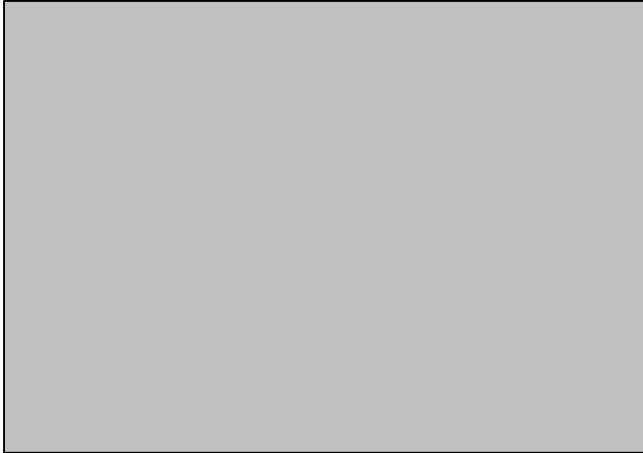


FIGURE 1. Prenatal ultrasound showing evidence of ascites.



FIGURE 2. CT scan showing hamartoma of the liver after complete resolving of the chyloous ascites.

cholesterol of 2 mmol/L, triglyceride of 2.1 mmol/L and protein of 3.2 g/L during the first week. The patient was put under complete parenteral alimentation for two weeks, and the chyloous ascites completely resolved during the third week, with no recurrence over a one-year period of follow-up.

Common features of all the cases of chyloous ascites mentioned above include the following:

- Uneventful pregnancy
- Spontaneous normal vaginal delivery
- Polyhydramnios found during delivery
- Ultrasound which revealed the presence of ascites before initial paracentesis, and was confirmed by laboratory results
- A yellowish color of ascitic fluid initially, becoming milky after paracentesis was done five to six days later, mainly when the patient had been fed orally.

## Discussion

Morton first described chyloous ascites in an infant in 1691 after paracentesis on an 18-month-old male.<sup>1-2</sup> The incidence of the condition has always been underestimated, and is thought to be between 1 in 50,000 and 1 in 100,000 hospital admissions.<sup>3</sup> Congenital chyloous ascites is even rarer, but is the most frequent cause of nonimmunologic peritoneal effusion. More than 10 studies have been reported in the last few years.

Congenital chyloous ascites can be identified by antenatal ultrasonography (Figure 2), thereby eliminating an eventual traumatic result.<sup>4</sup> The presence of hydramnios is the major associated anomaly.<sup>5-6</sup> Other associations include chylothorax,<sup>7-9</sup> lymphedema,<sup>8,9</sup> pulmonary lymphangiectasis,<sup>10,11</sup> intestinal lymphangiectasis,<sup>12-14</sup> generalized lymphatic dysplasia,<sup>14</sup> omental cyst,<sup>15</sup> malrotation,<sup>5,16</sup> meconium peritonitis,<sup>18</sup> Down's syndrome, and other anomalies of genetic origin.<sup>16-20</sup>

Congenital chyloous ascites is due to an inadequate lymphatic drainage from the intestine, resulting from maldevelopment of the intra-abdominal lymphatic system.<sup>3,12,21</sup> The pathogenesis is poorly understood and genetic factors are discriminated. Consanguinity is a common feature,<sup>22,23</sup> and the male is more liable to the condition.<sup>24</sup> Cases of chyloous ascites involving twins have also been reported.<sup>24,25</sup> Fetal tachycardia accompanying a huge ascites has been reported,<sup>26</sup> and cesarean may be prompted by fetal distress during labor.<sup>27</sup> Prematurity is frequent,<sup>5-8</sup> and neonatal respiratory distress and abdominal distention are also common, with ultrasound confirming the presence of fluid and abdominal x-ray showing a coloparietal undermining.

Paracentesis is the most useful diagnostic step. The chyle is usually color free, however, its appearance and composition are not constant, and depend on multiple factors such as the size of fat particles, cellular content and diet. Microscopic examination of the fluid demonstrates fat globules, and Wright's stain shows a predominance of lymphocytes and concentration of triglycerides ranged from 1.4 to 3.8 mmol/L, cholesterol from 1.6 to 2.8 mmol/L, and protein level from 2.5 to 4.1 g/L. Chylomicron can be found at a high concentration if the patient has been fed normally, and the specific gravity usually ranges from 1.010 to 1.021.<sup>5,6,18,28</sup> After confirmation of the diagnosis, patients should be treated conservatively by dietary correction of any fluid, electrolytes and vitamin D deficiencies, and provided with a fat-free diet, with the fat being replaced by medium-chain triglycerides.<sup>5,28-31</sup>

In severe or complicated chyloascites or chyloascites that persists after a maximum of 10 weeks of diet, a maximum of 6-10 weeks' of complete bowel rest and total parenteral nutrition should be initiated. Parenteral nutritional support appears to decrease the hazard of

medical therapy by maintaining an adequate nutritional intake while eliminating obligate losses consequent to repeated paracentesis.<sup>31-33</sup>

Caty et al. have reported a successful treatment of congenital chyloascites by a somatostatin analog.<sup>34</sup> This first line of therapy, in conjunction with multiple needle aspiration or drainage, depending on fetal tolerance, has been successful in more than 65%-70%.<sup>21,35,36</sup> Prolonged use of low-fat infant formula has been associated with poor neurologic development, possibly from fatty acid deficiency, therefore, it should be limited to three to four months.<sup>5,20,30</sup>

In utero, evacuation of peritoneal fluid can be indicated in fetal distress.<sup>3,5</sup> After a 10-week trial of bowel rest, most authors agree on abdominal exploration.<sup>37</sup> Further imaging studies should also be considered—Tc 99m dextran lymphoscintigraphy,<sup>38,39</sup> lymphangiogram or preoperative ingestion of the lipophylic dye<sup>40</sup> can direct the surgical procedure.

Successful surgical treatment of congenital chylous ascites can be achieved in 80% of patients<sup>35</sup> by resection of the macroscopic localized anomaly, or by ligation of an identifiable lymphatic leak.<sup>41-43</sup>

The most prominent location of the lymphatic leak is at the base of the superior mesenteric vessel.<sup>15,35,36,42</sup> If no leak can be identified, the area around the root of the mesenteric vessels should be closely inspected after mobilization of the colon, duodenum and the head of the pancreas,<sup>35,36,44,45</sup> and hemostasis should be performed by multiple ligation.

A peritoneovenous shunt, either the Leveen or Denver type, has been reported to be successful at least temporarily, in children in whom repeated attempts of medical or surgical approach have failed.<sup>44,46</sup> Rector and Whittlesey reported a complete control of chylous ascites by creating a circuit of extracorporeal recirculation.<sup>47</sup>

In a large collecting series, the recurrence of chylous ascites is not unusual and can occur even after three years. The death rate from chylous ascites is estimated to be between 24% and 30%,<sup>42,45</sup> however, no recent report exists.

Congenital chylous ascites is an unusual and etiologically poorly understood entity. Prenatal paracentesis should be reserved for an unstable fetus. The surgical approach is indicated after failure of adequate medical treatment.

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