Congenital Chylothorax

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Congenital chylothorax is a rare cause of respiratory distress in the neonatal period and yet the most important cause of unilateral pleural effusion in the newborn (1). We present a case of chylothorax that was diagnosed antenatally and managed conservatively in the postnatal period.

Case Report

A booked 20-year-old primigravida mother, who had no antenatal risk factors, had a routine ultrasound scan of the abdomen at 34 weeks gestational age. The fetus was found to have an isolated right sided pleural effusion with no evidence of hydrops or other anomalies. She had an uneventful normal delivery elsewhere and the baby developed mild respiratory distress at 6 hours of age. The infant was breastfed immediately after birth and brought to our hospital at 16 hours of age.

On examination, the baby was found to have tachypnea with dullness to percussion over the right hemithorax. Chest roentgenogram confirmed the presence of fluid in the right pleural space. Aspiration of this fluid yielded 35 ml of xanthochromic fluid with disappearance of symptoms. Examination of the fluid revealed a lymphocyte predominance (96%) and elevated protein content (3 g/dl). Special stains to detect fat globules were negative. Hemoglobin level, total and differential WBC counts, total protein and albumin levels and echocardiography of the heart were normal. The cultures of blood and pleural fluid were sterile.

Within 48 hours of admission, the infant developed respiratory embarrassment with signs of reaccumulation of fluid in the right pleural space for which an intercostal drain was placed. The fluid aspirated this time was again tested for fat globules. Though the infant had been breastfed during this time, fat globules were not detected. However, the triglyceride level of the pleural fluid was markedly elevated (>100 mg/dl), strongly supporting the diagnosis of chylothorax.

The fluid drainage continued for a further 48 hours when breastfeeding was stopped and the infant was given a skimmed milk formula along with medium...
chain triglycerides to provide 150 kcal of energy and 4 g protein in 150 ml of fluid per kg body weight per day. With these measures, the lymph drainage progressively decreased and the chest tube was removed on day 7 with no subsequent reaccumulation of fluid.

**Discussion**

Congenital chylothorax is an important differential diagnosis for pleural effusion detected *in utero*(2). When the infant is unfed, the fluid is clear with elevated protein content and cell counts revealing lymphocyte predominance, which may be used as an important criterion for diagnosis of this condition(1). However, after commencing feeds, the fluid becomes milky and fat globules start appearing. In this instance, non appearance of fat globules was intriguing, though the triglyceride levels were high.

Since the first description of congenital chylothorax by Pisek in 1917, the treatment of this condition has improved. In most cases, the cause is unknown, but trauma to the thoracic duct during delivery (especially breech, face and brow presentations) or following thoracic surgery have been implicated(3,4). Management involves repeated thoracentesis or intercostal drainage and dietary manipulation. This can easily be done by substitution of milk fat with medium chain triglycerides, which are absorbed directly into the portal venous system thus bypassing the intestinal lymphatics, yet providing adequate calories. Adequate intake of protein and calories is mandatory as the infant tends to lose a large volume of protein and fat rich fluid. In most cases, the above management results in complete resolution of the effusion without subsequent recurrence. In refractory cases however, total parenteral nutrition and surgery may be required(4).

Prognosis seems excellent in cases where there are no associated hydrops or lymphangiectasia. Early detection of the effusion and prompt management results in better outcome. Congenital chylothoraces appearing at gestational age greater than 32 weeks at antenatal diagnosis, as in our case, also carry a better prognosis as compared to effusions developing earlier during intra-uterine life.

**REFERENCES**


