CONGENITAL IDIOPATHIC BILATERAL CHYLOTHORAX IN A PRETERM NEONATE

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ABSTRACT
Congenital chylothorax is the most common cause of pleural effusion in neonates. It is defined as the abnormal accumulation of chyle or lymphatic fluid in the pleural cavity. The exact incidence of chylothorax in neonates is unknown. Among the several causes of chylothorax, the idiopathic cause is the most common. High morbidity in terms of pulmonary complications, nutritional deficiencies, and immunodeficiencies as well as significant mortality rate is associated with chylothorax. Our patient was a 33-week preterm neonate with bilateral chylothorax, who was treated with bilateral thoracentesis and octreotide therapy.

Keywords: Chylothorax, congenital, neonate, preterm

INTRODUCTION
Chyle in the pleural space was first described by Bartoletin in 16331. It results from an anatomical disruption of the thoracic duct and/or a major lymphatic tributary. Chylothorax can be congenital or acquired. It may be associated with certain syndromes, lymphatic disorders and prematurity, or may occur in isolation1. The incidence of congenital chylothorax is reported to be 1 in 10,000 births2. In many cases, no clear etiology is found and these are considered as idiopathic congenital chylothorax3. Chylothorax is diagnosed when pleural fluid assay has a triglyceride level >1.1 mmol/L, the ratio of the pleural fluid to serum cholesterol is <1.0, and total cell count is >1000 cells/mL with >80% lymphocytes or chylomicrons. Chylothorax has a very high morbidity in terms of nosocomial infections secondary to immune deficiency, nutritional deficiencies, and pulmonary complications1–5. The mortality rate is high, with a case fatality rate of 15–57%.

CASE PRESENTATION
A neonate was delivered by spontaneous vaginal delivery at 32 weeks gestation due to premature onset of labor. The birth weight was 1.97 kg. The baby cried at birth with Apgar scores of 5 (1 minute), 6 (5 minutes), and 7 (10 minutes); she soon developed severe respiratory distress, and was intubated at 15 minutes of age, shifted to the neonatal intensive care unit, and connected to the ventilator on intermittent positive pressure ventilation mode. A chest X-ray showed grade 3 hyaline membrane disease. The baby received three doses of surfactant in the first 24 hours. She had asymptomatic hypoglycemia and hypocalcemia, which were corrected. She was started on first-line antibiotics and caffeine citrate. An echo done on day 3 showed a small ostium scandinum defect and patent ductus arteriosus of 2 mm. The baby was weaned off ventilation and put on continuous positive airway pressure on day 3 of life. On day 5, the baby again developed severe respiratory distress. Blood gases showed respiratory acidosis; hence, she was again connected to the ventilator on minimal settings.

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Owing to C-reactive protein rising, the baby was shifted to second-line antibiotics (meropenem and amikacin) and was administered a short course of dexamethasone. Cranial U/S showed grade 3 IVH. The baby had a high BP of 106/79 on day 7 of life (>95th centile) with budging anterior fontanel non-pitting edema on both legs with hyponatremia (serum sodium: 116 mEq/L; serum potassium: 5 mEq/L).

A diagnosis of SIADH was made as serum osmolality was low, while urinary osmolality was high. Fluid restriction was applied and serum sodium returned to normal on day 10 of life.

On day 10, the baby was extubated and kept on high-flow O2 of 2 L/minute with nasal prongs. Expressed breast milk through gavage feeding was started from day 2 of life, which was gradually increased and Fio2 was gradually decreased to maintain Sio2 above 95%. On day 16 of life, the baby suddenly desaturated and developed severe respiratory distress with hypertonia and hyporeflexia as well as sluggish pupillary light reflex. A chest X-ray showed moderate pleural effusion on the left side with an underlying collapse of the lung. She was reconnected to the ventilator, and a left thoracentesis was performed by a pediatric surgeon. A total of 160 mL of turbid yellow white fluid was drained. Biochemistry and a microscopic exam revealed a triglyceride level of 20.8 mg/dL and cholesterol of 24.8 mg/dL with 980 cells/cmm, which were mostly lymphocytes. Gram staining and culture were negative.
The daily drain output comprised 50–190 mL/24 hours of chylous fluid. A repeat echo showed a moderate-sized PDA with pulmonary regurgitation and pulmonary hypertension. The baby was administered ibuprofen syrup for three days. A repeat echo showed the PDA size had reduced to 2 mm. Cranial U/S was repeated; it showed progressive dilation of the lateral ventricles and leukomalacia.

**DISCUSSION**

Chylothorax in neonates is most commonly associated with hydrops fetalis, chromosomal syndromes (e.g., Turner, Down, Noonan), and congenital abnormalities of the lymphatics. Chylothorax has also been associated with causes such as injury to the thoracic duct after cardiac surgery, thrombosis, trauma malignancy, and iatrogenic causes with long-line placements. These causes were not present in our patient and the condition could have been due to an underlying congenital malformation of the underlying lymphatic channels or thoracic duct. As the chromosomal analysis of our patient was normal and she expired before radiological investigations such as CT/MRI or lymphangiography could be conducted, an underlying congenital lymphangiomatous malformation could not be ruled out.

The baby responded well to octreotide, a synthetic somatostatin analog. Although its mechanism of action is not fully known, octreotide is known to reduce splanchnic blood flow, decrease the production of intestinal lymph, and lessen thoracic duct flow. A Cochrane review by Das et al. studied the use of octreotide in chylothorax, but the dose, duration of therapy, and frequency of doses varied significantly in different studies, with no randomized controlled trials conducted on its use. Perhaps a prospective multicenter randomized controlled trial is needed to assess the efficacy, safety, and appropriate dose of octreotide in the treatment of chylothorax in neonates.

Our patient was on partial TPN with 12.5% D/W and proteins; rare cases have been
reported of a development of TPN-related chylothorax\textsuperscript{13,14}. Surgical intervention is considered if, after conservative management, the daily loss of chyle exceeds 100 mL/day or 50 mL/kg/24 hours for over five days, chyle production fails to diminish after 14 days, or nutritional complications are present\textsuperscript{15}. Surgical procedures for refractory chylothorax include chemical pleurodesis, pleural ablation, thoracic duct ligation, apical pleurotomy, or pleuropertitoneal shunts. Surgical intervention was not considered for our patient due to the availability of a pediatric thoracic surgeon.

Early surgical intervention in refractory cases may minimize morbidity related to immune deficiency, infections, and nutritional deficiencies, and shorten the hospital stay.

Le Nué \textit{et al.} propose early surgical management of babies with significant prematurity, low birth weight, and massive chylothorax with chyle output of more than 50 mL/kg/day\textsuperscript{16}. Al-Tawil \textit{et al.} studied the long-term outcome of congenital chylothorax at two large neonatal centers over 13 years, and reported spontaneous resolution in 62\%, conservative management requirement for 10\%, and surgical intervention requirement for 28\%\textsuperscript{11}. No patient in his study was treated with octreotide. The survival rate in his study was excellent, even in babies presenting with hydrops.

\textbf{Figure 4.} Chest X-ray with chest tube

Our patient responded well to octreotide therapy, with chyle output decreasing from 100–120 mL/day to 15–20 mL/day within a week. A CT/MRI of the chest and lymphangiography or diagnostic thoracic surgery was planned for the baby. Our patient expired, probably due to complication of the disease (e.g., bleeding due to vitamin K deficiency, loss of clotting factors in chyle, nutritional deficiencies, pulmonary hemorrhage).

\textbf{REFERENCES}

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