A case of congenital chylothorax

Introduction
Chylothorax is the commonest cause of pleural effusion encountered during the perinatal period. Congenital chylothorax (CC) is rare with a prevalence of 1:10,000. CC could be due to congenital lymphatic malformation and may be associated with syndromes. We present a neonate with isolated congenital chylothorax. To best of our knowledge there are no published reports of CC in Sri Lanka.

Case Report
The antenatal scan of a 25-year-old primigravida mother was detected to have a right side pleural effusion in the fetus. An extensive anomaly scan by a specialist in fetal medicine at 34 weeks of gestation confirmed the finding without any other associated congenital abnormalities. The baby boy was delivered vaginally at term with a birth weight of 3.1kg and good Apgar scores. His routine newborn examination was unremarkable except for reduced air entry in the right lower zone.

An ultrasound scan (USS) of the chest on day two of life demonstrated a right (R) sided pleural effusion. He was not in respiratory distress and the oxygen saturations were normal. His chest X-ray revealed (R) sided pleural effusion with collapse of the right lung (Figure 1) although there was clinically no respiratory compromise in the baby.

On the tenth day of life the baby developed respiratory distress and repeat USS of chest revealed a massive pleural effusion. An intercostal (IC) tube was inserted on the same day and the fluid was drained. The fluid was yellowish white in colour without blood staining which was in favour of chyle. (Figure 2)

The pleural fluid analysis confirmed chylothorax due to high triglyceride levels and low cholesterol levels. (Table 1) The intercostal tube was kept in situ for 7 days until cessation of the drain.
The baby was monitored for possible complications associated with chylothorax such as lymphopenia and hypoalbuminaemia. However, his lymphocyte count and serum albumin level remained within normal range. His basic biochemical investigations and inflammatory markers were normal. Serum cholesterol level was 18 mg/dl. His serum lactic dehydrogenase (LDH) was 625 U/L (0-248) and serum protein was 58 g/L (41-63). 2D-echocardiography revealed good biventricular functions and no pericardial fluid was noted.

The baby was kept nil oral and given intravenous (IV) fluid for 48 hours. He was commenced on medium chain fatty acid rich formula continued for 10 days. IV octreotide 1µg/kg/hr infusion was initiated and tailed off over a 9 days. The IC tube was kept in situ for 5 days and was removed 2 days later after drain became nil. The baby was commenced on breast feeding on day 21 and discharged after 2 weeks of hospital stay. He did not have any respiratory distress on discharge and repeat USS of the chest showed no effusion.

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### Discussion

The mortality rate of CC ranges from 20-60%\(^1\). Iatrogenic trauma, malignancies and some granulomatous infections cause secondary chylothorax in infants and children\(^1,2\). Antenatally detected CC can act as a space occupying lesion interrupting normal lung development\(^2\). Best method of diagnosing chylothorax is measurement of pleural fluid triglyceride and cholesterol levels\(^4\). If the triglyceride level is more than 110 mg/dl and the pleural fluid to serum cholesterol ratio is less than one, the diagnosis is established. Computerized tomography scan, magnetic resonance imaging, lymphangiography and lymphoscintigraphy will be helpful as imaging studies to visualize the lymphatic system to identify chyle leak and lymphatic system anatomy\(^5\).

The key aims in treatment of chylothorax include relief of respiratory symptoms by draining the pleural fluid, treating the underlying cause to prevent recurrences and management of associated complications like malnutrition and immunodeficiency\(^2\). Octreotide can be used to reduce the chyle production. It is thought to cause reduction in splanchnic circulation and thereby reduce chyle production\(^5,6\). Failed medical management of chylothorax warrants surgical intervention. Surgical options include thoracic duct ligation, pleurodesis and pleuro-peritoneal shunt placement\(^2\).

### References


