Congenital Idiopathic Bilateral Chylothorax in a Preterm Neonate

Zareen Fasih¹, Mahmoud Elsayed Attia Shamseldeen², Ignatius Edwin DSouza³ Anum Fasih³ and Ajit Gadekar⁴

¹Consultant Neonatologist, Thumbay Hospital, Ajman, UAE
²Paediatric Consultant, Thumbay Hospital, Ajman, UAE
³Paediatric Specialist, Thumbay Hospital, Ajman, UAE
⁴Specialist Neonatologist, Thumbay Hospital, Ajman, UAE

Abstract
Congenital chylothorax is the most common cause of pleural effusion in neonates. It is defined as abnormal accumulation of chyle or lymphatic fluid in the pleural cavity. Exact incidence of chylothorax in neonates is unknown. Among the several causes of chylothorax idiopathic cause has the highest incidence.

Chylothorax carries a high morbidity in terms of pulmonary complications, nutritional deficiencies, and immunodeficiency and also has a significant mortality rate.

Here we report a 33 weeks preterm neonate with bilateral chylothorax which was treated with bilateral thoracentesis and octreotide therapy.

Keywords: Chylothorax, Idiopathic, Premature, Neonate.

Introduction
Chyle in the pleural space was first described by Bartoletin 1633 [1]. 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 isolation [1]. The incidence of congenital chylothorax is reported to be 1 in 10,000 births [2]. In many cases no clear etiology is found and is considered as idiopathic congenital chylothorax. The diagnosis of chylothorax is considered when pleural fluid assay has a Triglyceride level >1.1mmol/L, the ratio of the pleural fluid to serum cholesterol is <1.0, and a total cell count of >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 complications [1-5]. Mortality rate is high with a case fatality rate of 15-57%.

Case Presentation
This neonate was delivered by spontaneous vaginal delivery at 32 weeks gestation due to premature onset of labour. Birth weight was 1.97 kg. Baby cried at birth with Apgar scores of 5/1,6/5 and 7/10 minutes, but soon developed severe respiratory distress and was incubated at 15 minutes of age and shifted to NICU and connected to the ventilator on IPPV mode. X-ray chest showed grade 3 HMD. Baby received 3 doses of surfactant in the first 24 hours. Baby had asymptomatic hypoglycemia and hypo calcinemia which were corrected. She was started on first line antibiotics and caffeine citrate. Echo done on day 3 showed small ASD and PDA. Baby was gradually weaned off ventilation and put on CPAP on day 3 of life. On day 5 baby again developed severe respiratory distress. Blood gages showed respiratory acidosis, so was again connected to ventilator on minimal settings. Due to rising CRP baby was shifted to 2 line Antibiotics (Meropenum and Amikacin) and was given a short course of Dexamethasone. Cranial u/s showed grade 3 IVH. Baby had high BP 106/79 a day 7 of life (>95 centile) with budging anterior fontanelle non pitting oedema on both legs with hyponatremia (Serum Na – 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 done and serum sodium returned to normal on day 10 of life.

On day 10 baby was extubated and kept on high flow O2 of 2L/ minutes with nasal prongs. She was started with Expressed breast milk with gavage feeding from 2 day of life which was gradually increased and the Fio₂ was gradually decreased to maintain Sio₂ above 95%. On day 16 day of life she suddenly desaturated and developed severe respiratory distress with, hypotonia and hyporeflexia and sluggish pupillary light reflex. X-ray chest showed moderate pleural effusion on left side with underlying collapse of lung. She was reconnected to ventilator and left thoracentesis was done by pediatric surgeon. 160 ml of turbid yellow white fluid was
drained. Biochemistry and microscopic exam revealed triglyceride level of 20.8 mg/dl, Cholesterol of 24.8 mg/dl with 980 cells/cm which were mostly lymphocytes. Gram staining and culture was negative.

The daily drain output was from 190-50 ml/24 hours of chylous fluid. Repeat echo showed moderate sized PDA with Pulmonary regurgitation and pulmonary hypertension. Repeat cranial U/S showed progressive dilatation of lateral ventricles and leukomalacia.

Baby was started on Fresh frozen plasma 10 ml/kg 12 hourly, 20% Albumin solution and feeding with milk with MCT oil. X-ray chest done on 29 day of life showed effusion on right side also, so chest drain was inserted on right. Side also. As both drains drained 30-210 ml of chylous fluid daily Octreotide was started on day 30 of life. She showed response to the therapy with decrease in the amount of fluid to 00-45 ml/day. All blood cultures were showing no growth, but lab. Reports showed severe hypo albuminemia, leukopenia and thrombocytopenia despite daily infusion of 20% albumin and FFP infusion. Doppler Ultra sound of chest and abdomen were normal except for bilateral pleural effusion. Plan for MRI chest and Lymphangiography was made for which baby needed to be shifted to another facility. On day 46 of life she suddenly developed abdominal distension, drop in saturations and BP. Plan for diagnostic abdominal tap was made but she had a cardio pulmonary arrest. Received 4 rounds of CPR but could not be revived.
Discussion
Chylothorax in neonates is most commonly associated with hydrops fetalis, chromosomal syndromes eg: turners, downs or congenital abnormalities of the lymphatics. Though it has been described in association with various causes such as post cardiac surgery injury to the thoracic duct, thrombosis, and trauma or iatrogenic with long line placements.

In our patient none of the above causes were present and may have been due to some underlying congenital malformation of underlying lymphatic channels or thoracic duct. As chromosomal analysis of our patient was normal and the baby expired before radiological investigations like MRI or lymphangiography could be carried out an underlying lymphangiomatous malformation could not be ruled out.

The baby showed a good to octreotide, a synthetic somatostatin analogue. Though its mechanism of action is not fully known, it is known to reduce splanchnic blood flow, decrease the production of intestinal lymph and decrease thoracic duct flow.

References