

periventricular or basal ganglia changes. MRI of the cervical spine (**Fig. 1b**) (confirmed atlanto-axial dislocation (AAD) causing compressive myelopathy at C1 level, without any other spinal malformations. Neurosurgeon prescribed neck collar, and advised follow-up for cervical spine stabilization. In view of the truncal shortening and AAD, he was also advised evaluation for skeletal dysplasia, but the parents deferred it to a later date.

Conditions which mimic CP should be considered – when there is absence of definite preceding perinatal insult; there is family history of developmental delay and spasticity; there is developmental regression or onset of new clinical signs of upper motor involvement; and when there is associated significant ataxia, muscle atrophy, or sensory loss [2]. Although neuroimaging is not essential for making a diagnosis of CP, MRI brain is abnormal in more than 80% of children with CP [4]. Current Western guidelines recommend MRI in children suspected to have CP [1]. The imaging not only uncovers the pathogenic patterns responsible for the CP but can also detect structural malformations of the brain and neuro-metabolic problems which resemble CP [4].

Despite significant perinatal risk factors, the intermittent abnormal neck stiffness warranted meticulous examination and evaluation [3], which revealed AAD can be idiopathic or due to traumatic, inflammatory or genetic disorders like Down syndrome, achondroplasia, cleidocranial dysplasia and Morquio syndrome [4]. Neurological manifestations of congenital AAD in children result from progressive compression of the cervicomedullary junction and present as progressive quadriplegia. Patients with myelopathy may go undiagnosed for a long period because of very slow progression of the

disease process [3] and maybe mistakenly diagnosed as CP. Trauma or sudden movement can worsen symptoms in AAD. In the reported child, the increase in stiffness upon getting from sleep could possibly be due to the fact that while he was lying down, neck positioning could have caused an increase in stiffness. Poor cervical posture during sleep could cause increased biomechanical stresses on the structure of the cervical spine and could result in cervical pain and stiffness [5].

This case highlights compressive myelopathy as a differential for CP, and underscores the importance of a good history-taking in all patients, especially those labelled as cerebral palsy.

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Congenital Chylothorax with Lymphatic Malformation and Successful Antenatal and Postnatal Management

Neonatal chylothorax is an abnormal accumulation of lymphatic fluid in the pleural space which can be either congenital or acquired. Nearly 90% of all *in utero* pleural effusions are chylothorax [1]. The estimated incidence of congenital chylothorax is 4 per lakh [2], with mortality ranging from 30-50%. [3]. We herein report a late preterm

girl identified antenatally at 31 weeks of gestation with severe bilateral pleural effusion for which thoracoamniotic shunt was placed and subsequently diagnosed with congenital chylothorax after delivery.

A 37-year-old lady, G3P1A1L1 was admitted at 36^{5/7} weeks for delivery of hydropic fetus. Antenatal follow up had been uneventful till 31 weeks when ultrasonography showed hydropic changes in the fetus with bilateral pleural effusion and subcutaneous edema. A therapeutic fetal pleurocentesis was done with amniocentesis. Chromosomal analysis and microarray on amniotic fluid was negative. Mother had a negative indirect coomb's test, with serology negative for VDRL, and TORCH. Parvo

Virus PCR was negative, and she had a normal HbA1C and Hemoglobin electrophoresis. Pleural fluid examination showed 205 leucocytes per cu.mm, 80% lymphocytes, LDH 87 U/L and a protein of 1.8g/dL. Follow up scan at 33 weeks showed a return of significant pleural effusion. Rather than opting for preterm delivery, a right Rodeck thoracoamniotic shunt was placed. Subsequent USG showed resolution of effusion on the Right side with lung expansion and satisfactory interval growth with normal fetal Dopplers. The left pleural effusion was drained just prior to the delivery.

A female baby with birth weight of 3015 g was delivered at 36^{5/7} week gestation by elective LSCS. Baby had signs of labored breathing at birth, and was intubated and ventilated. Chest X-ray showed right side pneumothorax and left side pleural effusion for which bilateral intercostal tube drains (ICD) were inserted. Pleural fluid was clear exudate (Protein – 2.6 g/dL) with 3638 cells/mm³, predominantly lymphocytes and 1.9% neutrophils. Post ICD insertion baby improved and was extubated to high flow nasal cannula (HFNC). Echocardiography and ultrasound abdomen and cranium were normal. Once feeds were started pleural fluid became milky in nature. Pleural fluid sent for analysis showed rise in triglyceride level from baseline 35.8 mg/dL on Day 1 to 134 mg/dL on day 6 confirming the diagnosis of chylothorax.

MRI Chest was done on Day 8 of life for central lymphatic anatomy and intrathoracic mass lesions. It showed prominent tortuous lymphatic channels along with prominent azygous vein (**Fig. 1**). Baby was started on medium chain triglyceride formula on day 9 of life in view of chylothorax. Feeds had to be discontinued and parenteral nutrition restarted along with injection Octreotide on day 12 because of increasing chyle drainage. Following this, chyle formation reduced and the intercostal drains were removed on day 16. Post drain removal there was an increase in pleural effusion (right > left) which was organizing and non-tappable. Octreotide infusion was increased in view of persistent collection (at 10 mcg/kg/hr). Immunoglobulin levels were low for which single dose IVIG was given. On Day 25, lymphoscintigraphy was done to rule out lymphatic dysplasia, which was reported as normal. Feeds were restarted on day 28, after which there was worsening in respiratory distress but there was no increase in pleural effusion; as monitored by ultrasound. Keeping the possibility of leaky pulmonary lymphatics causing increase in pulmonary interstitial fluid, diuretics were added, to which baby responded well and feeds were gradually increased. Diuretics were continued till day 41 of life. Octreotide infusion was tapered gradually. Clinical

exome testing done which showed no pathogenic variants causative of the phenotype, but variants of uncertain significance were detected (lymphatic malformation-3, OMIM#613480). Baby was discharged on day 44 of life on MCT- based formula (Pregestimil).

Congenital chylothorax can be an isolated finding or may be associated with genetic conditions. Early antenatal detection and management by placement of fetal pleuroamniotic shunt improves perinatal outcome by avoiding complications due to pulmonary hypoplasia [4]. Irrespective of the cause, initial postnatal management consists of drainage of pleural fluid, appropriate ventilation, total parenteral nutrition and dietary modification (conservative approach). Medication and surgery may be required in refractory cases. Most chylothorax cases improve spontaneously because of the natural course of the disease. In newborns it is important to distinguish neonatal chylothorax from congenital lymphatic dysplasia, as the latter is difficult to treat and has poorer prognosis.

As increased chyle formation is associated with immunological and nutritional complications, we had started octreotide on day 12. On reviewing literature [5,6] we could not find any practice recommendations for use of octreotide. The lymphatic malformation-3 may explain the localized edema in neck, genitals and probably in lungs, which persisted at time of tapering of octreotide infusion.



Fig. 1 MRI image (T2 SPAIR, coronal diffusion weighted sequence) showing bilateral pleural effusion (R>L), prominent tortuous lymphatic channels in thoracic region extending into upper abdomen, with no evidence of any mediastinal mass.

To conclude, optimum management of such cases is still a matter of debate, but prenatal evaluation and management is associated with improved survival. Postnatally we should follow conservative approach for few weeks to give enough time for the lymphatics to heal and develop collaterals [6]. Refractory cases would require additional therapy.

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Stumped by Potassium: A Rare Case of Familial Pseudohyperkalemia

Hyperkalemia is a common electrolyte disturbance requiring emergent intervention to avoid potential fatal arrhythmias. Pseudohyperkalemia should be kept in mind in the absence of symptomatology and other associated laboratory abnormalities. We present a rare case of pseudohyperkalemia detected incidentally during evaluation of a child with acute respiratory infection. Familial pseudohyperkalemia is an asymptomatic condition that is detected incidentally during evaluation if serum is stored below room temperature prior to testing. It is characterized by spuriously high serum potassium levels due to cold-induced 'passive leak' of red blood cell (RBC) potassium ions into plasma [1,2]. *ABCB6* gene (2q36) has been identified as the causative gene of this rare condition [3].

A 2-month-old baby, first born of non-consanguineous parents, presented to a peripheral health care setup with history of cough and fever for 2 days with some lethargy and refusal to feed. On examination, the infant was found to have mild tachypnea and tachycardia and was admitted for monitoring and supportive therapy. Hematological and

biochemical parameters were sent to rule out sepsis and dyselectrolytemia. All parameters were within normal limits except for the elevated potassium levels of 6 mEq/L. In view of high potassium levels, repeat sample was sent, which had serum potassium values of 6.8 mEq/L. The ECG did not show signs of hyperkalemia. Echocardiography revealed a structurally normal heart with good biventricular function. However, chest X-ray showed diffuse non homogenous opacities suggestive of bronchiolitis and a large homogenous opacity silhouetting the left cardiac border with a linear translucency surrounding it. Tumor lysis syndrome was initially suspected to be the cause of hyperkalemia but computed tomography of chest revealed that this anterior mediastinal mass was an unusually large, hypertrophied thymus gland and not a malignant mass. During PICU stay, baby remained asymptomatic but continued to have hyperkalemia. There were no dysmorphic features or abnormal genitalia suggestive of any recognizable genetic syndrome. Baby was worked up further with plasma renin activity and aldosterone levels for possibility of pseudohypo-aldosteronism. However, all investigations were within normal limits. Common causes of pseudohyperkalemia (cell lysis, extreme leukocytosis or thrombocytopenia, or use of EDTA anticoagulant) were ruled out.

Clinical exome sequencing was done to rule out pseudohypoaldosteronism type II; as common causes had been excluded for the cause of hyperkalemia. It