Poster No.9

A CASE OF LEFT LUNG APLASIA WITH ESOPHAGEAL ATRESIA WITH DISTAL TRACHEOESOPHAGEAL FISTULA

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Introduction:

Pulmonary aplasia is a rare condition in which there is absence of the lung parenchyma and pulmonary vasculature. It has a dismal prognosis as it is commonly associated with other congenital anomalies.

Case Summary:

A baby boy born at term was noted to have facial asymmetry with left mandible hypoplasia and macrostomia. He had respiratory distress and copious oral secretions. Resistance was felt at 9cm from incisor during Ryle's tube insertion. Chest radiograph revealed coiling of Ryle's tube at T1 level with left lung collapse, homogenous opaque lung left hemithorax and well distributed bowel gas. Diagnosis of Gross Type C esophageal atresia was established.

On day 2 of life, he underwent rigid bronchoscopy, tracheo-oesophageal fistula division with repair of tracheal defect and Stamm gastrostomy. Rigid bronchoscopy showed tracheoesophageal fistula opening 2cm above carina. Left main bronchus opening was small but patent with copious mucus. Intra-operatively, the upper pouch of the oesophagus was not seen despite dissection over the thoracic inlet.

The baby then underwent a subsequent cervical exploration, proximal esophageal circular myotomy, right thoracotomy and delayed esophageal anastomosis at day 17 of life. Post operative, the child developed multiple pulmonary infections and required high ventilation settings, hence a contrasted computed tomography scan of the thorax performed and showed a left lung aplasia with rudimentary left main bronchus and absent left pulmonary vessels. An oral contrast study showed no anastomotic site stricture. He was treated with judicious ventilator care, antibiotics and gradually weaned to nasal prong.

At present, the baby is stable on oral and gastrostomy feeds, and gradually gaining weight.

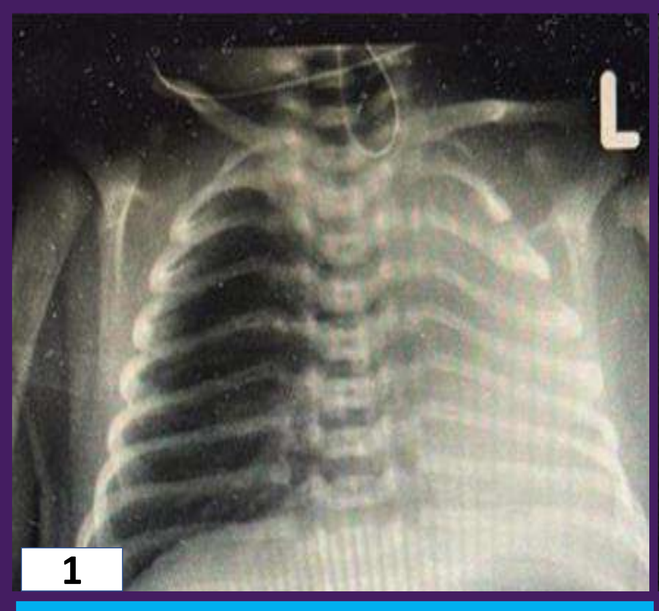


Figure 1 : Chest and Abdominal Radiography After Ryle's Tube Insertion

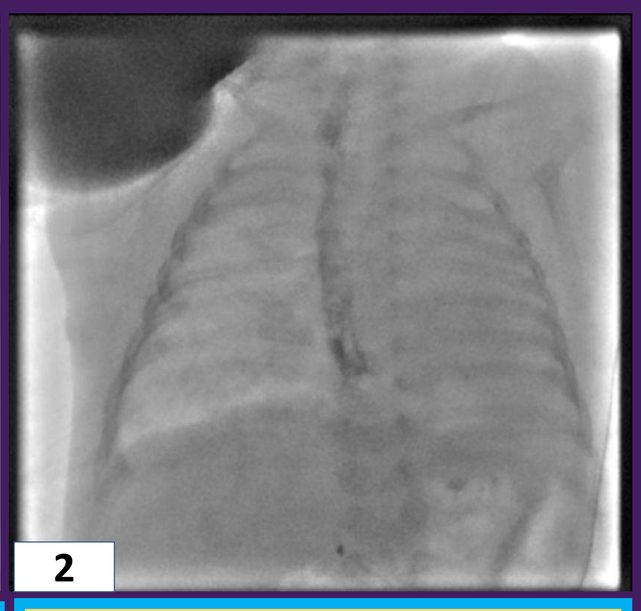
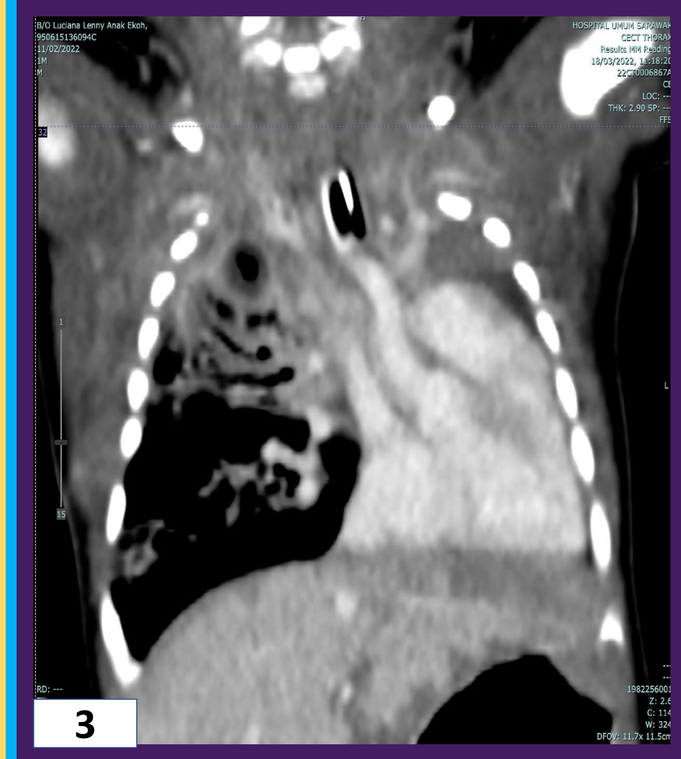
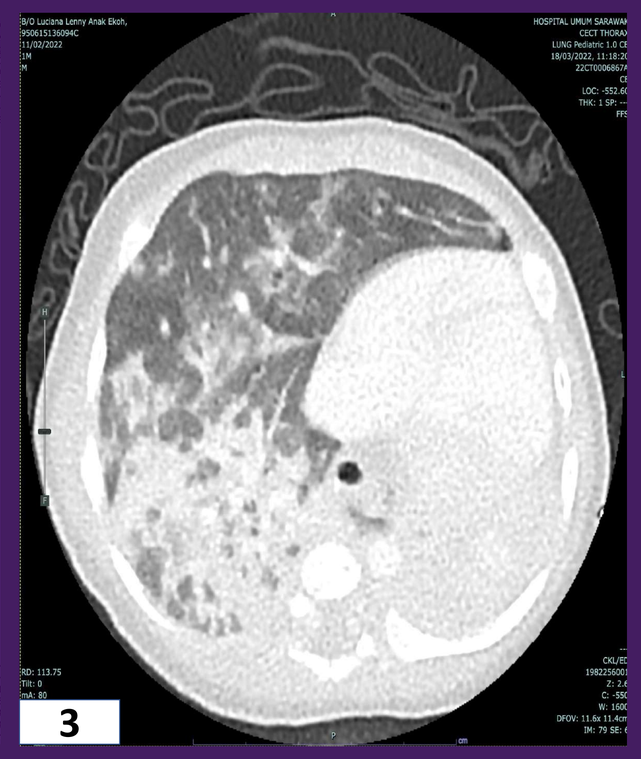
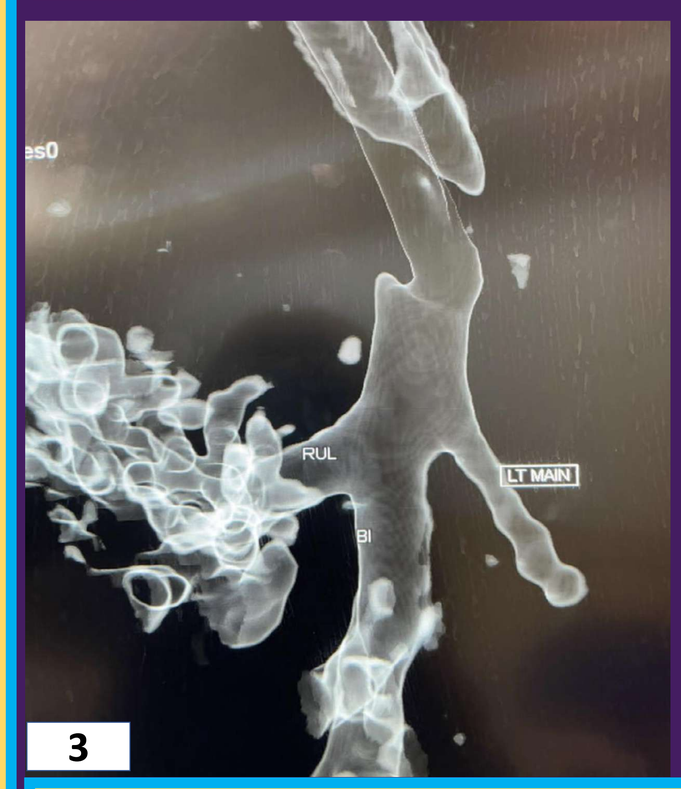


Figure 2: Oral Contrast Study at Day 52 of life







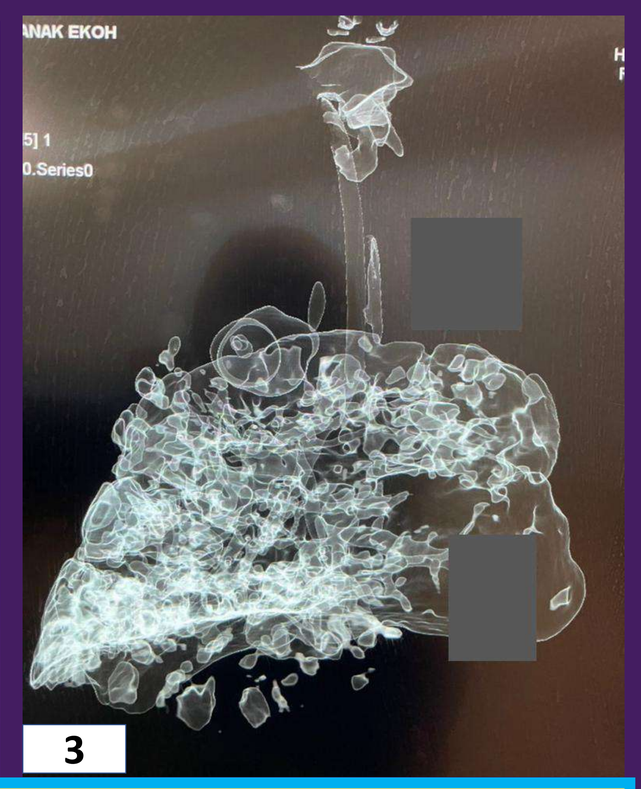


Figure 3: Contrasted computer tomography scan of the thorax at day 35 of life

Discussions:

1. The diagnosis of lung aplasia requires a high index of suspicion. Antenatal scans, serial chest radiographies, computer tomography and computer tomography angiography confirms the diagnosis.
2. Patients with pulmonary aplasia are associated with multiple systemic congenital abnormalities and often a challenge to treat in view of repeated pulmonary infections, post operative and long term care. However, multiple staged surgeries, aggressive intensive care unit care will aid in the survival of the patient.

Conclusion:

Despite a dismal prognosis, well coordinated and aggressive multidisciplinary management and parent awareness is essential in treating and providing better survival benefits for patients with multiple congenital abnormalities.

References:

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2. Venkatesh Harohalli Aswathanarayana Iyer, Siddu Charki, and Kumar Shanthigrama Ramachandra. Aplasia of the lung in a neonate—the myth is explored