Clinics in Surgery

Clinical Image
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# Congenital Diaphragmatic Eventration Complicated by Massive Peri-Operative Tension Pneumothorax

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## Clinical Image

A term male infant was born at 2800 g and established on CPAP for respiratory distress. Chest radiograph demonstrated a severe right sided eventration of the diaphragm (Figure 1). Ultrasound of the diaphragm was in keeping with the diagnosis. Both the child's mother and maternal grandmother had also been diagnosed with the same condition. Echocardiogram revealed a small central ASD/PFO but no pulmonary hypertension. Genetics workup is ongoing. As the child was unable to be weaned off CPAP, thoracoscopic plication of the diaphragm was planned for day eight of life. On the morning of the planned fixation, the baby was electively intubated in NICU with a 3.0 uncuffed ETT following difficulty passing a 3.5 uncuffed ETT. On arrival in theatres shortly after there was a poor $ETCO_2$ trace and increasing $FiO_2$ requirement. The child was reintubated using a video laryngoscope and a bougie, with difficulty passing the tube beyond the cords. There was ongoing difficulty ventilating the baby however cardiac output was maintained. Severe abdominal distension suspicious for a perforated viscous was also noted. Working diagnoses at this stage included severe subglottic stenosis, pneumothorax, pneumoperitoneum, severe bronchospasm or tracheal trauma. Bedside ultrasound of the chest was suspicious for pneumothorax but given the uncertainty an urgent plain X-ray of the baby was ordered before proceeding to needle decompression of either cavity. A massive tension pneumothorax with eversion of the diaphragm into the abdominal cavity was noted (Figure 2). Needle thoracocentesis was immediately attended followed by insertion of 10 Fr chest drain. The baby quickly stabilized and the procedure abandoned. Two days later the child had a persistent large air leak from the chest drain. Right sided muscle sparing thoracotomy through the sixth intercostal space was performed. A bronchopleural fistula at the anterior aspect of the oblique fissure was found and repaired with 5/0 and 6/0 PDS® sutures (Johnson & Johnson). Plication of the diaphragm was then performed with rows of pleated pledgeted 2/0 Ti-Cron® (Medtronic). Post operative CXR is shown in Figure 3. The child was weaned off CPAP but required nasal prong oxygen for a few weeks before being discharged home requiring supplemental nasogastric feeds. The exact cause of the pneumothorax remains unknown and the case presented a diagnostic dilemma in an unstable child. Congenital eventration results in replacement of the normal

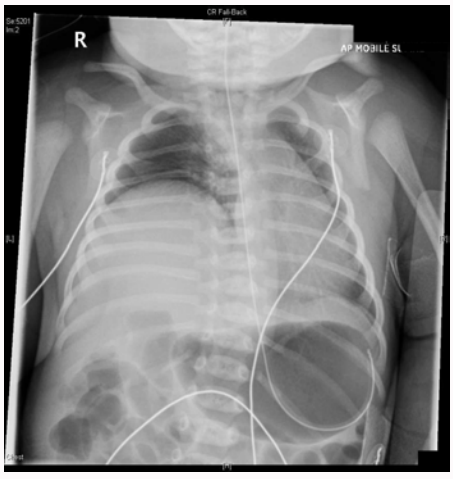


**Figure 1:** Pre-operative CXR. Severe congenital right diaphragmatic eventration.



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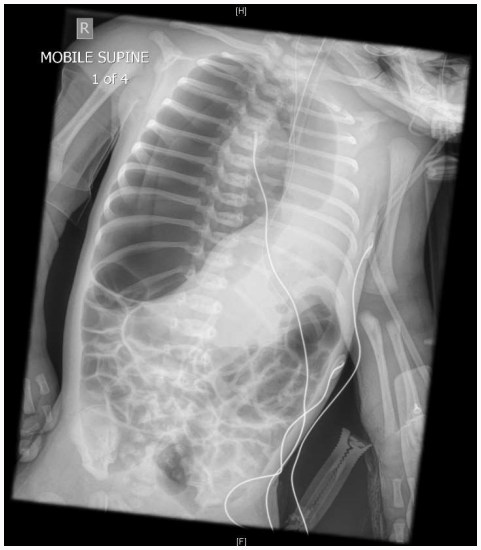


**Figure 2:** Perioperative CXR demonstrating tension pneumothorax.

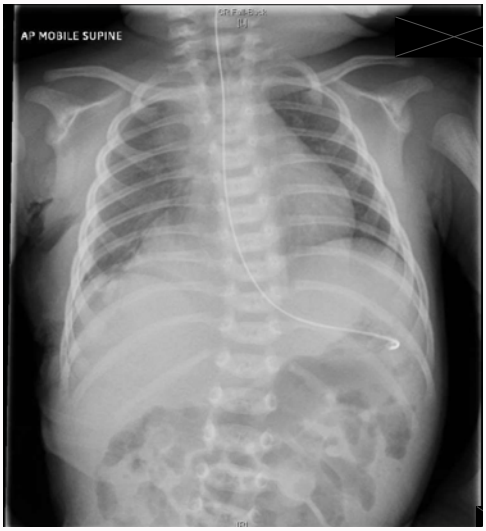


**Figure 3:** Post-plication CXR.

diaphragm muscle with fibroelastic tissue whilst retaining continuity and attachment at the costal margin [1,2]. This often leads to both respiratory (tachypnoea, distress, cyanosis) and gastrointestinal (vomiting, failure to thrive) symptoms [1,3]. The main differential diagnosis is congenital diaphragmatic hernia, with chest X-ray and ultrasound usually sufficient for diagnosis. In addition to supportive care, those infants with eventration requiring moderate to intensive respiratory support, unable to tolerate enteral nutrition or those with gastric volvulus, require surgical intervention [1,4,5]. Plication is usually done within the first weeks of life with good short- and long-term outcomes [3,4,6].

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