

TEACHING FILES (GRAND ROUNDS)

## TACHYPNEA IN A NEONATE AND DIAGNOSTIC CONUNDRUMS

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### KEYWORDS

Lung Hyperinflation, Tachypnea

### ARTICLE HISTORY

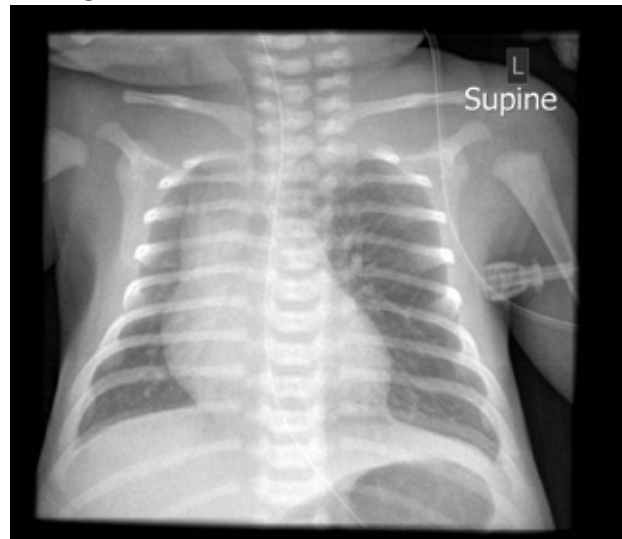
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### Clinical Problem

A term male infant was born by normal vaginal delivery following an uncomplicated pregnancy. He was admitted to the neonatal intensive care unit (NICU) at 33 hours of age with tachypnea [respiratory rate 105 breaths/min (bpm)]. Examination was otherwise normal including preductal oxygen saturation of 97% and post-ductal oxygen saturation of 96%. Full blood count showed hemoglobin 15.1 gm/dL, white cell count  $15.8 \times 10^9$  cells/cumm, platelets  $317 \times 10^9$  cells/cumm and C-reactive protein (CRP) was 40 mg/l. Chest x-ray was interpreted as a possible right upper lobe pneumonia in the presence of a degree of rotational artefact (Figure 1). Intravenous (IV) benzylpenicillin and gentamicin was commenced and given for 5 days. Cerebrospinal fluid (CSF) and blood cultures did not grow any organism. The presumptive diagnosis therefore was congenital pneumonia. He was commenced on 0.1L/min of low flow nasal cannula oxygen shortly after admission following a drop in his oxygen saturation to 93% and then escalated to high flow nasal cannula oxygen support (HFNC) within 3 hours due to worsening respiratory distress. (Figure 2).

**Figure 1.** Chest x-ray done shortly after admission on low flow oxygen. Image appears slightly rotated with possible right upper lobe collapse/consolidation and mild right sided mediastinal shift.



**Figure 2.** Run chart showing the respiratory rate pattern during admission.



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Capillary blood gas performed on 6L/min of high flow on 21% oxygen showed a pH 7.39, pCO<sub>2</sub> 5.6, bicarbonate (HCO<sub>3</sub>) 24.6. He remained on HFNC in air with stable blood gases with a significant drop in his

**Figure 3.** Post intubation x-ray showing overinflated left hemithorax with significant right sided mediastinal shift.



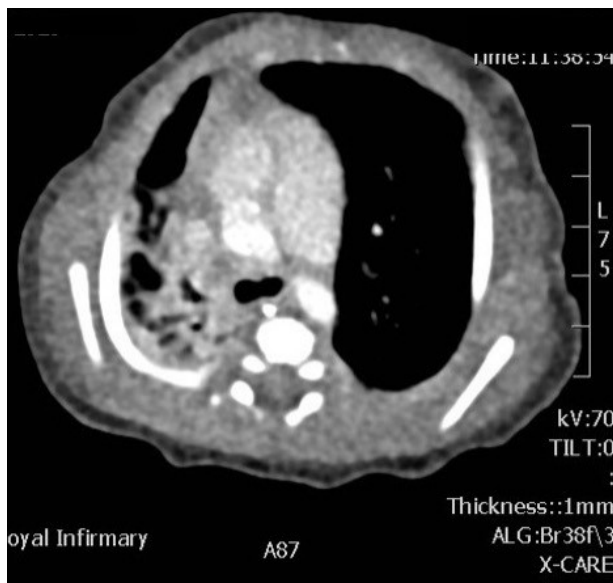
respiratory rate (figure 2) from 105 bpm to 65 bpm. HFNC was stopped between day 2 and 3 and the respiratory rate was seen to rebound to 85 bpm (figure 2). A grade 3/6 systolic heart murmur was heard around 80 hours of life and an echocardiography revealed a small outlet muscular ventricular septal defect (VSD) not thought to be a contributing factor to his tachypnea. The cardiovascular examination remained otherwise normal. Nasogastric tube feeding was commenced a few hours after admission but interrupted by vomiting spells, however this was subsequently well tolerated without incidence after a brief pause. At one week of age, the tachypnea persisted, and respiratory rate remained between 65 bpm and 80 bpm on HFNC oxygen with increased work of breathing. At this stage the oxygen requirement then increased by up to 50% with worsening respiratory acidosis on capillary blood gas (pH 7.24, pCO<sub>2</sub> 9.3, HCO<sub>3</sub> 24.5). He was intubated and ventilated and thereafter investigated and treated for sepsis a second time. The post intubation x-ray showed a right upper lobe collapse with an overinflated left lung and displacement of the left main bronchus (Figure 3). HRCT chest is depicted as Figure 4. A trial of dexamethasone 75 mcg/kg/dose was started on day 10 and subsequently increased to 200 mcg/kg/dose after 48 hours without improvement in oxygen requirement. In view of the failure to respond to medical treatment he was referred for surgery.

*What is the diagnosis?*

### Discussion

Investigations revealed a diagnosis of congenital lobar emphysema (CLE), and patient subsequently underwent successful surgery in the form of a left thoracotomy and left upper lobectomy on day 16 of life. Histology of the resected lobe confirmed the diagnosis. Our patient was extubated 4 days later to low flow

**Figure 4.** Lung CT showed partial or complete segmental collapse bilaterally with relative hyperinflation of the left upper lobe. The left main bronchus is partially narrowed as it passes between the aorta and heart.



oxygen and discharged well on day 24 of life.

CLE is a rare disease (1:20,000 to 1:30,000).<sup>1</sup> Diagnosis in the new-born period can be difficult and challenging. It should be suspected if a baby presents with sustained tachypnea and persistent increase in work of breathing, where alternative causes cannot be found. Babies with CLE may present with signs of severe respiratory distress although only 50% of these cases present with symptoms at birth. Only a third of cases are actually diagnosed at birth and males are three times more affected than females.<sup>1</sup> The left upper lobe is most commonly involved (43%) as in our patient and in 50% of cases, the cause is idiopathic in nature. Patients can be mistakenly diagnosed as pneumonia, pneumatocele, pulmonary hypoplasia or pneumothorax.<sup>1</sup> As in our patient, about 12-20% of cases are associated with congenital heart diseases with 50% of these being VSD.<sup>2</sup> Serial x-rays may show progressive hyperinflation of the lungs which occurs as a result of bronchial compression with air trapping on expiration.<sup>3</sup> Confusion may arise if the film is thought to be rotated or collapse/consolidation of one segmental lobe is thought to be the origin of the asymmetry. If this is mis-interpreted as a pneumothorax, inserting a chest drain can result in increased morbidity and mortality.<sup>4</sup> Chest CT as performed in our case is the gold standard for diagnosis and contrast enhanced CT provides information about vascular anatomy, which may be important for subsequent surgery and will highlight other mediastinal masses if present. CT findings include hyperinflation of the affected lobe with contralateral mediastinal shift, compressive atelectasis of adjacent lobes in addition to attenuation of vascular structures in the affected lobe.<sup>5</sup> Surgery in the form of lobectomy is the mainstay of treatment and is often reserved for severe cases or those with progressively

worsening disease. However, some patients with mild to moderate disease have been managed conservatively with regular follow up.<sup>6</sup> Post-operative pneumonia is reported as the most common complication leading to respiratory failure and need for prolonged mechanical ventilation. Overall mortality from surgery ranges between 0-21% with one case series reporting a mortality of 1.9% over a 15-year period.<sup>7</sup> Outcome for patients managed conservatively varies in several reports. A few will eventually require surgical intervention while others have shown gradual and complete resolution with no respiratory compromise and normal growth outcome at follow up.<sup>7,8</sup> Our baby showed an initial response to HFNC oxygen with a significant drop in his respiratory rate which appeared reassuring. However, the rebound once HFNC was stopped between day 2 to 3 was highly suggestive of on-going pathology. It is possible that we were dealing with a combined diagnosis of sepsis and CLE.

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#### **Compliance with ethical standards**

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Conflict of Interest: None

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