AB182. 141. Congenital lobar emphysema: an important diagnosis in the infant with respiratory distress

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Abstract: Congenital lobar emphysema (CLE) is a rare congenital malformation of the lung characterised by hyperinflation of one or more lobes which usually presents in the neonatal period or in early infancy. Males are three times more likely to be affected than females and the left upper lobe is the area most commonly involved. The diagnosis and management are often difficult and if not detected may be mismanaged leading to significant respiratory distress and haemodynamic instability. We present the case of a six-week old male who presented to the emergency department with respiratory distress and was admitted with a diagnosis of respiratory syncytial virus positive bronchopneumonia. However after an acute deterioration on the ward he required admission to the intensive care unit where he was ventilated for management of type two respiratory failure. Further deterioration while on convential ventilation lead to commencement of high frequency oscillatory ventilation and inotropic support for persistent respiratory acidosis and hemodynamic instability. A CT Thorax was later preformed showing mediastinal shift with hyperinflation of the right middle lobe confirming the diagnosis an underlying diagnosis of congenital lobar emphysema which was likely exacerbated by ventilation. An emergency thoracotomy and right middle lobectomy was preformed with extracorporeal life support on standby. The surgery was successful and the patient had an uneventful post operative recovery. Here we emphasise the importance of early consideration of underlying congenital lung disease in cases where the clinical picture does not correlate with the working diagnosis especially in the infant population. We also look at anaesthetic challenges associated with congenital lobar emphysema and how best they can be managed.

Keywords: Cardiothoracic surgery; congenital lobar emphysema; neonatal intensive care

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