European Respiratory Society
Annual Congress 2013

Abstract Number: 3264

Publication Number: P4348

Abstract Group: 8.1. Thoracic Surgery

Keyword 1: Children **Keyword 2:** Congenital lesion/malformation **Keyword 3:** Surgery

Title: Reliving Rudhe syndrome: The reversible shunt related lobar emphysema- Report of cases and systematic review of literature

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Body: Lobar emphysema (LE) is not quite uncommon in children with congenital heart diseases (CHD). In the year 2010, the name Rudhe syndrome was used to refer to reversible shunt related lobar emphysema, Rudhe proposed that the emphysema is secondary to the shunt thus surgical intervention be directed to the correction of the cardiac defect alone and not lobectomy. We reviewed 4 cases of patients with concomitant LE and CHD. The medical records were evaluated with reference to age, type of CHD, pulmonary function, radiographic findings, pulmonary artery pressure, clinical signs and symptoms, surgical management and outcome after surgery. We also reviewed 21 literatures on LE with CHD. Among the 4 cases we had, 3 were left to right shunts and 1 is a case of Tetralogy of Fallot (TOF) with an absent pulmonary valve. The 3 underwent correction of the cardiac lesion and repeat chest radiograph and CT scan showed almost complete resolution of the lobar emphysema with complete resolution of clinical symptoms 6months post cardiac surgery. The other one underwent TOF correction with lung tacking with radiographic resolution of the lobar emphysema however patient died of sepsis. The literature review consisted of 137 subjects.The three most common cardiac lesions associated with lobar emphysema are ventricular septal defect, patent ductus arterioles and TOF with absent pulmonary valve. One hundred out of the 137 underwent correction of the cardiac lesion without lobectomy. Resolution of airway obstruction as well as radiographic resolution of the emphysematous lung were noted as early as 3months to 1 year post cardiac surgery.