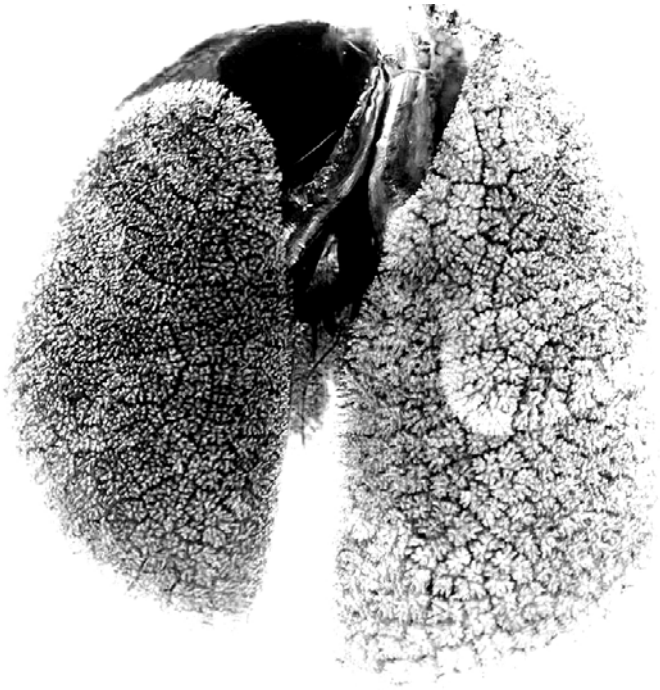
SWISS SOCIETY OF NEONATOLOGY

Hyperlucent lung in an extremely preterm girl

February 2017



Eberhard R, Arlettaz Mieth R, Clinic of Neonatology (ER, AMR), University Hospital Zurich, Switzerland

This female infant was born by Cesarean section at 24 2/7 weeks of gestation to a 41-year-old G2/P2 because of suspected chorioamnionitis (later confirmed by histology) following premature rupture of the membranes at 22 4/7 weeks of gestation. Two doses of antenatal corticosteroids had been given at 23 5/7 and 23 6/7 weeks of gestation.

After delivery, positive pressure ventilation was provided by mask with a T-piece resuscitator (Perivent®) with maximum settings of 30/5 cmH_2O. Once spontaneous breathing set in, adequate oxygenation was maintained on nCPAP with an FiO_2 of 0.25. After insertion of umbilical catheters, empiric antibiotic therapy was started. Arterial umbilical cord pH was 7.39 and Apgar scores were 3, 4, and 6 at 1, 5, and 10 minutes, respectively. Birth weight, length and head circumference were 700 g (P 50 – 75), 32 cm (P 50 – 75), and 22 cm (P 25 – 50), respectively.

The initial chest X-rays (Fig. 1A, B) showed well aerated lungs without obvious signs of surfactant deficiency. For the first week of life, the infant was supported on nCPAP with an FiO_2 of 0.3 – 0.4. When treatment of increasing episodes of apnea and bradycardia despite caffeine therapy failed to respond to non-invasive positive pressure ventilation (NIPPV), the patient was intubated and put on conventional mechanical ventilation on day of life (DOL) 11. At this point, a chest X-ray confirmed correct position of the endotracheal

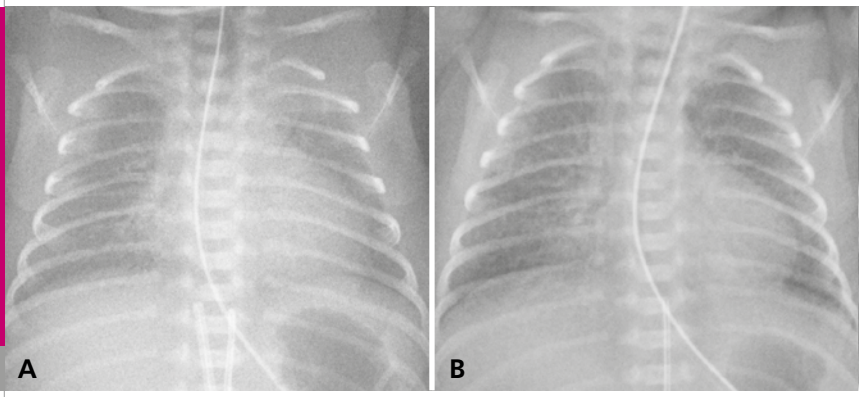


Fig. 1 A B

Chest X-ray on DOL 1 (A) and DOL 2 (B): no clear evidence of hyaline membrane disease while on nCPAP.

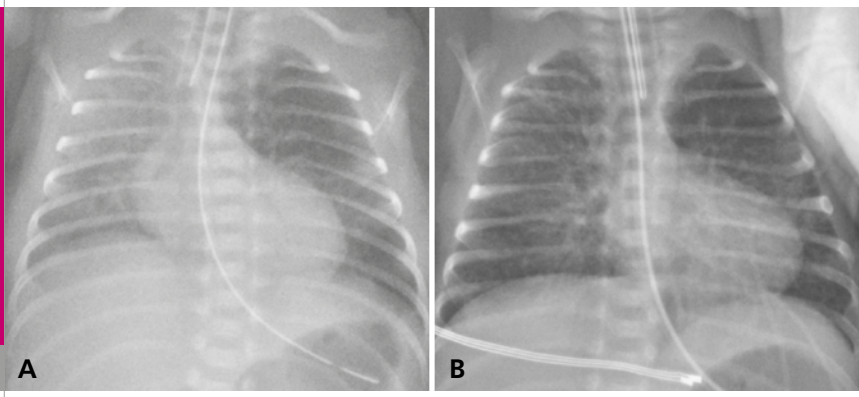


Fig. 2 A B

Chest X-rays following intubation: on DOL 11 (A), there is asymmetric lung aeration despite correct position of the endotracheal tube; and on DOL 15 (B) no obvious lobar overinflation can be recognized.

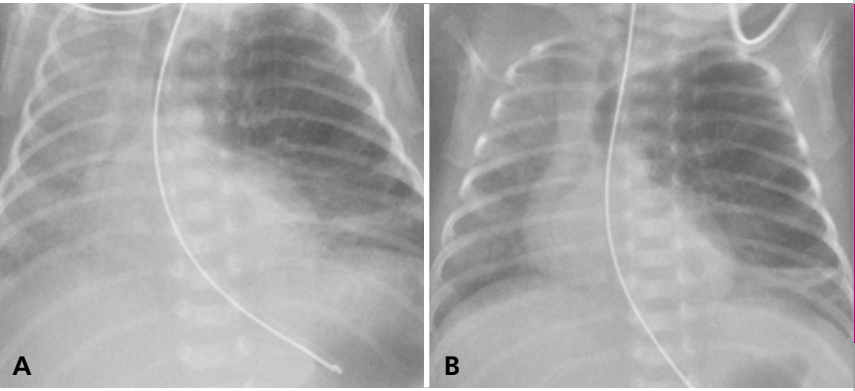


Fig. 3

Chest X-rays on DOL 30 (A) and 31 (B): persistence and even increase of a hyperlucent area (likely corresponding to the left upper lobe) with herniation of lung tissue to the contralateral side and opacification of the right lung.

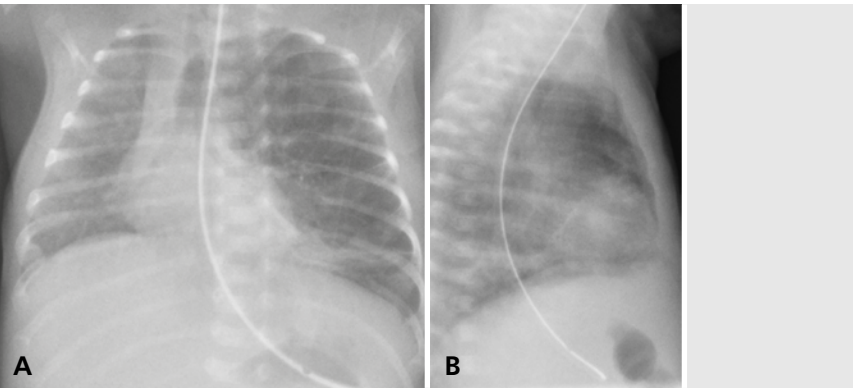


Fig. 4

The ap Chest X-ray on DOL 31 (A) was complemented by a lateral film (B): the findings were not consistent with a left-sided pneumothorax.

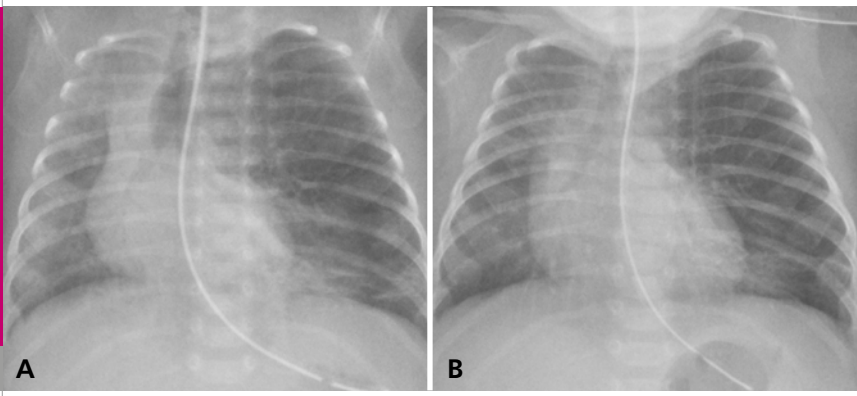


Fig. 5

Chest X-ray on DOL 41 (A) documents progression of hyperinflation, whereas a follow-up chest X-ray on DOL 68 (B) showed some improvement.

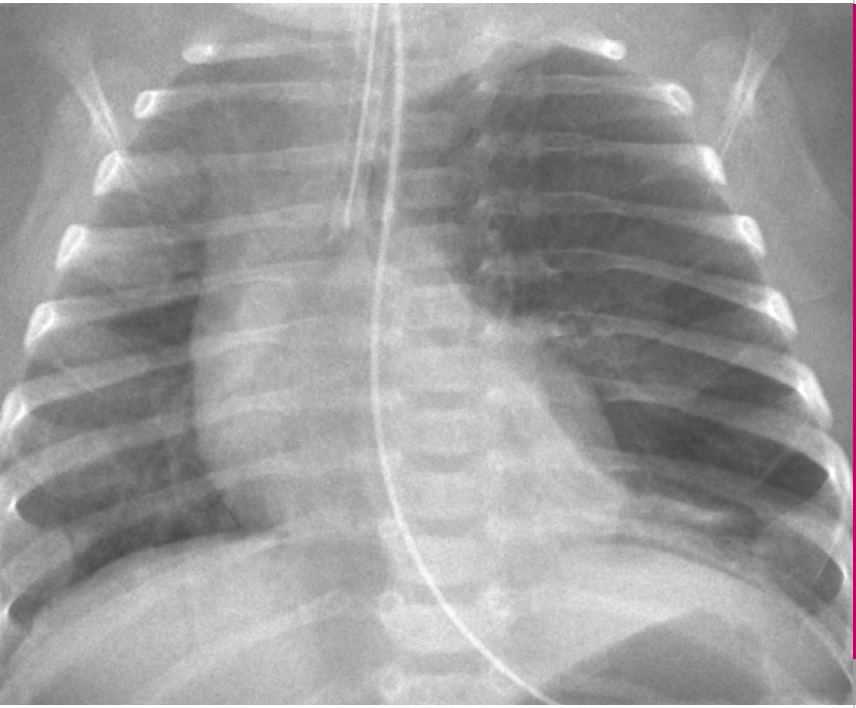


Fig. 6

Chest X-ray on DOL 95 following intubation for ROP laser therapy: persistence of lobar hyperinflation (also note low position of the endotracheal tube).

tube, but asymmetric aeration of the lungs was noted, which was interpreted as an atelectatic or infiltrated right upper lobe (Fig. 2A); this finding, however, appeared to be transient and was no longer apparent on DOL 15 (Fig. 2B).

The infant was extubated to nCPAP with a PEEP of 6 cmH_2O on DOL 17. Additional chest X-rays were obtained at 1 month of life because of an increasing oxygen requirement; they showed persistence of a hyperlucent area likely corresponding to the left upper lobe, herniation of lung tissue to the contralateral side and opacification of the adjacent lung (Fig. 3A, B). Because vascular markings were recognized in the hyperlucent area and a later chest X-ray was unremarkable (Fig. 4A, B), pneumothorax or emphysematous bullae were thought to be unlikely; therefore, overinflation of the left upper lobe with consecutive compression of the right lung was thought to be responsible for the imaging findings and a preliminary diagnosis of congenital lobar overinflation (CLO) or emphysema (CLE) was made. After consultation with members of the thoracic surgical team, a conservative approach was taken and the infant was preferentially positioned with the left side dependent. PEEP was adjusted to prevent further atelectasis without promoting additional overinflation. After additional progression, documented on a chest X-ray on DOL 41 (Fig. 5A), the situation finally appeared to stabilize with less overin-

flation and mediastinal shift shown on a chest X-ray on DOL 68 (Fig. 5B).

There was a prolonged nCPAP requirement (up to DOL 74, i.e., a corrected gestational age of 35 0/7 weeks); supplemental oxygen could be discontinued on DOL 99 at a corrected gestational age of 38 4/7 weeks. In addition, the baby's hospital course was complicated by a PDA, staphylococcus epidermidis septicemia, a submandibular abscess, and retinopathy of prematurity (ROP) stage III (right eye) and stage II (left eye) with plus disease, treated by laser coagulation at 38 weeks' postmenstrual age. Fortunately, all cerebral ultrasound examinations were normal.

A chest X-ray obtained after intubation for laser treatment of ROP documented persistence of the abnormal aeration pattern (Fig. 6); however, at that time, the baby was asymptomatic. Two weeks later, the infant was discharged home with a body weight of 3450 g (P 50). Follow-up with pediatric pulmonology has been arranged.

DISCUSSION

Congenital Lobar Overinflation (CLO) is a developmental anomaly of the lower respiratory tract resulting in hyperexpansion of one or more of the pulmonary lobes. It is a rare condition with a reported incidence of less than 1 in 20'000 infants, with an unexplained male to female ratio of approximately 3:1, and accounts for 5 – 25% of resected pulmonary malformations (1 – 4). As in the presented patient, it is typically diagnosed when characteristic hyperlucency on chest X-ray is noted. CLO can present in young infants either with rapidly progressive respiratory symptoms or as an incidental finding.

The pulmonary lobes are variably affected: 40 – 50% occur in the left upper lobe; approximately 30% in the right middle lobe, 20% in the right upper lobe and less than 10% in the lower lobes. CLO affecting multiple lobes is rare.

In the literature, CLO is more commonly called Congenital Lobar Emphysema (CLE), a historical designation probably owing to the surplus in pulmonary air; however, this term may not correctly reflect the histopathological findings. These range from uniformly enlarged distal airways and airspaces to a polyalveolar form with increased numbers of alveoli within the gas exchange units (5). In contrast, emphysematous changes like destructive rarefication of alveolar walls, inflammatory cells or fibrosis are lacking. Presumably,

these tissue changes result from various disturbances early in bronchopulmonary development, with postnatal overinflation as a final common pathway.

A common explanation for progressive lobar expansion is air trapping by a ball-valve mechanism caused by obstruction of the airways (6). Possible factors include deficiency of bronchial cartilage (most frequent), with consecutive airway collapse during expiration, or mucosal folds that can cause partial intraluminal obstruction. Extrinsic compression may be caused by vascular anomalies like pulmonary artery sling or anomalous pulmonary venous return, or intrathoracic masses, such as a foregut cyst or a teratoma. Often though, the definitive causative mechanism cannot be identified, neither by imaging nor by histopathology. Therefore, other unrecognized mechanisms promoting selective expansion of a predisposed lobe (e.g., elevated compliance due to developmental tissue alterations) may be responsible in some cases.

A possible inheritable component for the development of CLO has been suggested because of its occurrence in monochorionic twins, in a father and his son, and a mother and her daughter (7 – 9). The malformation is not described in association with syndromic diseases. However, an association with cardiovascular anomalies has been reported in about 15 percent of cases, the most common being ventricular septal defect, patent ductus arteriosus, and tetralogy of Fallot (10).

Similar to other pulmonary malformations, CLO is increasingly detected on prenatal ultrasound examinations. CLO lesions may involute during pregnancy, as do other pulmonary malformations (6). Postnatally, hyperinflation of the affected lobe leads to expansion and possibly herniation, mediastinal shift, as well as compression of the remaining lung tissue with resultant atelectasis. Some infants rapidly develop progressive respiratory distress, while others have a more gradual onset or even remain asymptomatic. The former typically have tachypnea and increased work of breathing, and often cyanosis (11). Recurrent pneumonia or poor feeding with failure to thrive are less frequent presentations that may occur in milder forms.

Diagnosis is usually based on chest X-ray findings. Computed tomography (CT), echocardiography, magnetic resonance imaging (MRI), bronchoscopy, esophagography and angiography have all been used to further delineate the disorder. Histopathology of resected lobes is helpful to confirm the diagnosis and to detect components of other pulmonary malformations that sometimes coexist in one lesion (11).

In the past, CLO has mostly been managed by pediatric surgeons. The majority of patients with CLO present with respiratory symptoms in the neonatal period or in infancy and require early resection of the affected lobe. Case series show good outcomes after lobectomy; in long-term follow-up, patients may have

generalized overinflation on chest radiographs and pulmonary function abnormalities, but most have few clinical symptoms (12).

Increasingly, CLO has also been diagnosed in asymptomatic or oligosymptomatic patients. Recently published case series have demonstrated clinical and sometimes radiological improvement in patients treated conservatively. Long-term outcomes in mildly symptomatic patients are also comparable between patients treated surgically or conservatively (12 – 14).

CONCLUSION

Congenital pulmonary malformations should be considered in the list of differential diagnoses for progressive respiratory distress in both term and preterm infants. We describe the case of an extremely preterm female infant with a gestational age of 24 2/7 weeks who presented with signs and symptoms of CLO in the first few weeks of life.

REFERENCES

1. Ramachandran P. Congenital malformations of the lung and mediastinum – A quarter century of experience from a single institution. J Pediatr Surg 1997;32:44 – 47 *(Abstract)*

2. Ng C, Stanwell J, Burge DM, Stanton MP. Conservative management of antenatally diagnosed cystic lung malformations. Arch Dis Child 2014;99:432 – 437 *(Abstract)*

3. Shanti CM, Klein MD. Cystic lung disease. Sem Pediatr Surg 2007;17:2 – 8 (no abstract available)

4. Laberge J-M, Puligandla P, Flageole H. Asymptomatic congenital lung malformations. Sem Pediatr Surg 2005;14:16 – 33 *(Abstract)*

5. Tander B, Yalçin M, Yilmaz B, Karadağ CA, Bulut M. Congenital lobar emphysema: a clinicopathologic evaluation of 14 cases. Eur J Pediatr Surg 2003;13:108 – 111 *(Abstract)*

6. Olutoye OO, Coleman BG, Hubbard AM, Adzick NS. Prenatal diagnosis and management of congenital lobar emphysema. J Pediatr Surg 2000;35:792 – 795 *(Abstract)*

7. Thompson AJ, Reid AJM, Reid M. Congenital lobar emphysema occurring in twins. J Perinat Med 2000;28:155 – 157 *(Abstract)*

8. Roberts PA, Holland AJA, Halliday RJ, Arbuckle SM, Cass DT. Congenital lobar emphysema: Like father, like son. J Pediatr Surg 2002;37:799 – 801 *(Abstract)*

9. Wall MA, Eisenberg JD, Campbell JR. Congenital lobar emphysema in a mother and daughter. Pediatrics 1982;70:131 – 133 (no abstract available)

10. Moideen I, Nair SG, Cherian A, Rao SG. Congenital lobar emphysema associated with congenital heart disease. J Cardiothorac Vasc Anesth 2006;20:239 – 241 (no abstract available)

11. Choudhury SR, Chadha R, Mishra A, Kumar V, Singh V, Dubey NK. Lung resections in children for congenital and acquired lesions. Pediatr Surg Int 2007;23(9):851 – 859 *(Abstract)*

12. Özçelik U, Göçmen A, Kiper N, Doğru D, Dilber E, Yalçın EG. Congenital lobar emphysema: Evaluation and long-term follow-up of thirty cases at a single center. Pediatr Pulmonol 2003;35:384 – 391 *(Abstract)*

13. Karnak I, Şenocak ME, Büyükpamukçu N. Congenital lobar emphysema: Diagnostic and therapeutic considerations. J Pediatr Surg 1999;34:1347 – 1351 *(Abstract)*

14. Mei-Zahav M, Konen O, Manson D, Langer JC. Is congenital lobar emphysema a surgical disease? J Pediatr Surg 2006;41:1058 – 1061 *(Abstract)*

SUPPORTED BY Vifor Pharma

CONTACT Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch