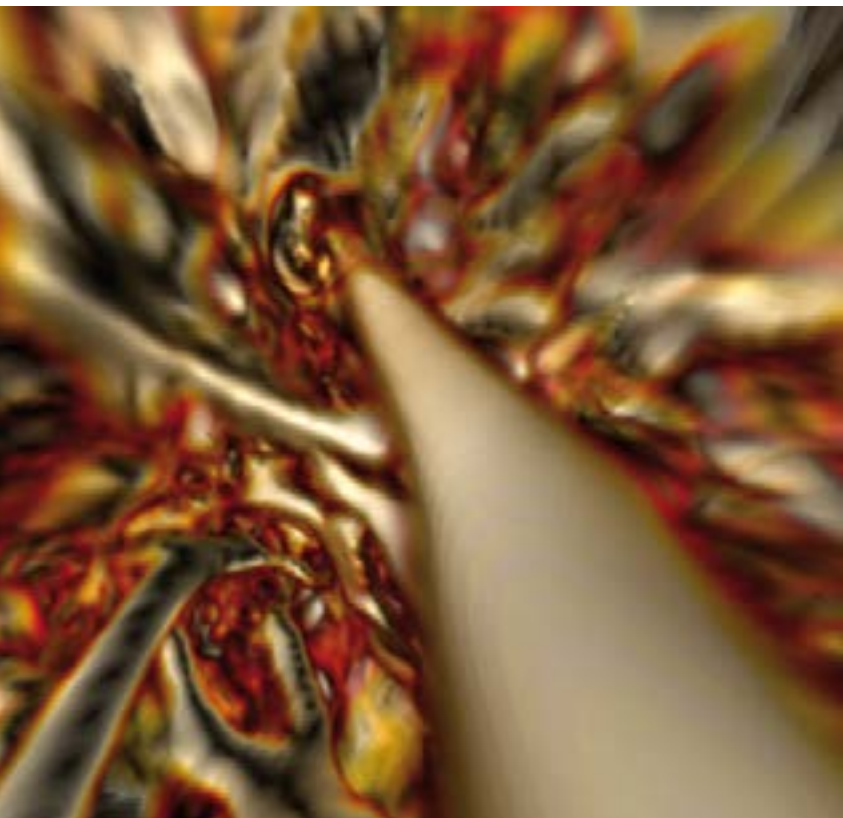


Persistent tachypnea
following respiratory distress
syndrome in a near-term
infant – The Macklin effect?

January 2009



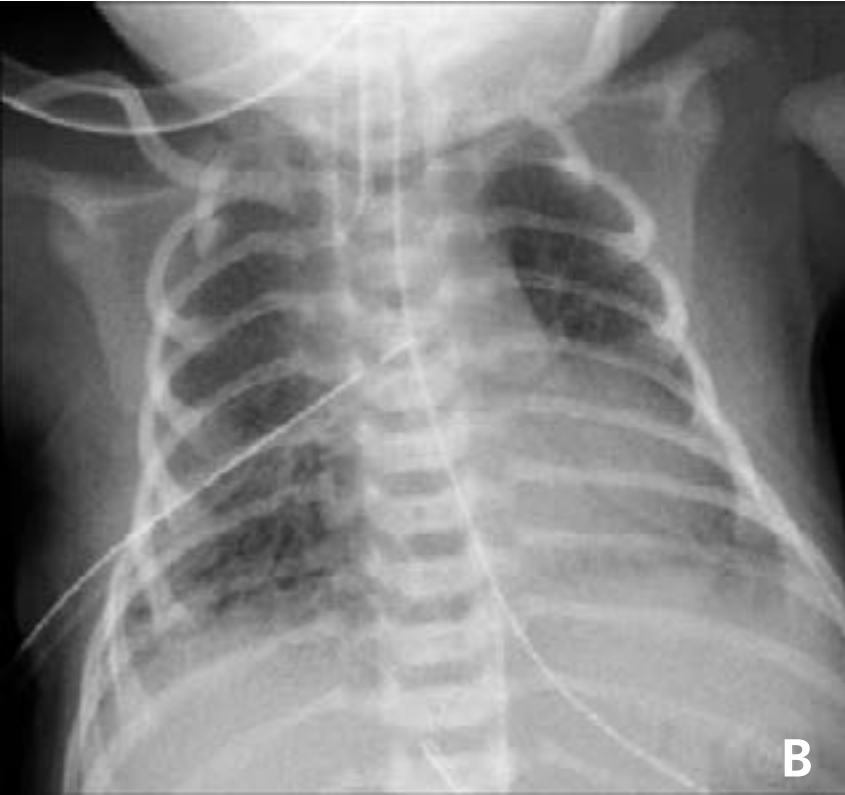
Steurer M, Berger TM, Caduff JH, Neonatal and
Pediatric Intensive Care Unit (SM, BTM), Children's
Hospital of Lucerne, Department of Radiology (CJH),
Cantonal Hospital of Lucerne, Switzerland

This female infant was born at a gestational age of 35 3/7 weeks by a cesarean section for maternal indications at another hospital. Her birth weight was 2520 g (p 25-50). The Apgar scores were 8, 9, 9 at 1, 5 and 10 minutes, respectively. In the first hour of life, she developed signs of respiratory distress. At the age of 3 hours, she was transferred to our institution for the further management. Because her respiratory status worsened and there was radiographic evidence of surfactant deficiency, she was intubated and surfactant (Curosurf®) was administered. At the age of 20 hours she developed a right-sided pneumothorax and a chest drain was inserted (Fig. 1). The pleural drain was removed on the 3rd day of life and she was extubated two days later. She required no supplemental oxygen but remained tachypneic. On the 10th day of life, a chest x-ray was felt to be consistent with pulmonary interstitial emphysema (PIE) of the right lower lobe (Fig. 2). The diagnosis of persistent PIE was confirmed by a chest CT scan (Fig. 3-5). Despite strict right lateral decubitus positioning, the PIE enlarged leading to progressive mediastinal shift to the left (Fig. 6). A decision was made to attempt single lung ventilation following selective intubation of the left main stem bronchus (Fig. 7A). Within 24 hours, the infant became increasingly difficult to ventilate (severe respiratory acidosis with pCO₂ reaching 14.9 kPa) and was therefore switched to high frequency oscillatory ventilation (HFOV). This was only temporarily successful and after 30 hours of single lung venti-



Fig. 1

Chest X-rays at the age of 20 hours showing hyaline membrane disease and a right-sided pneumothorax (A) before and B) after the insertion of a chest tube).



B

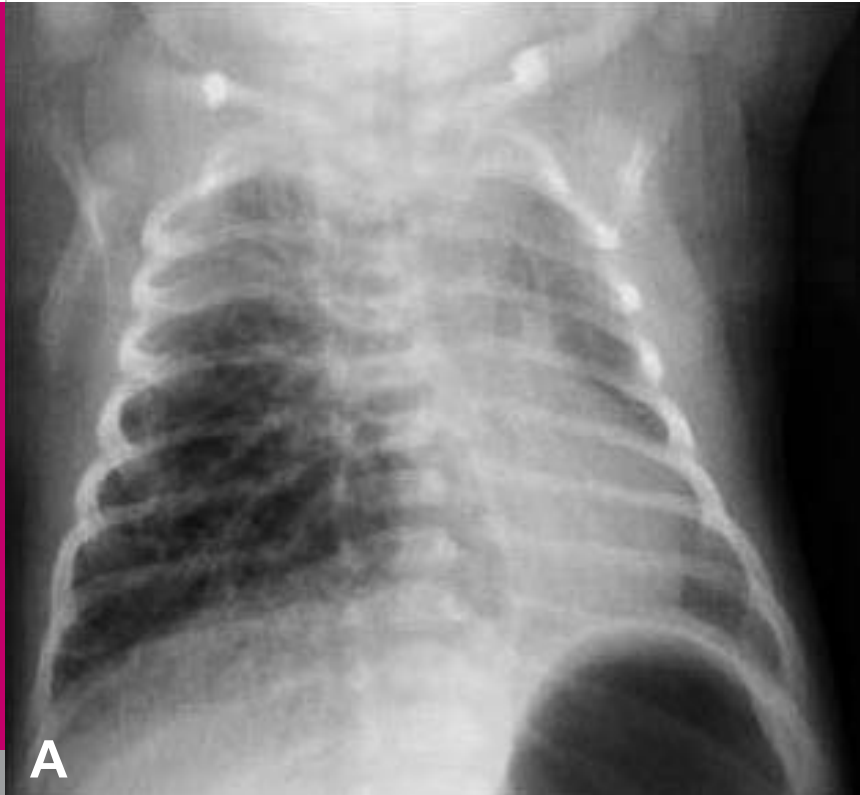
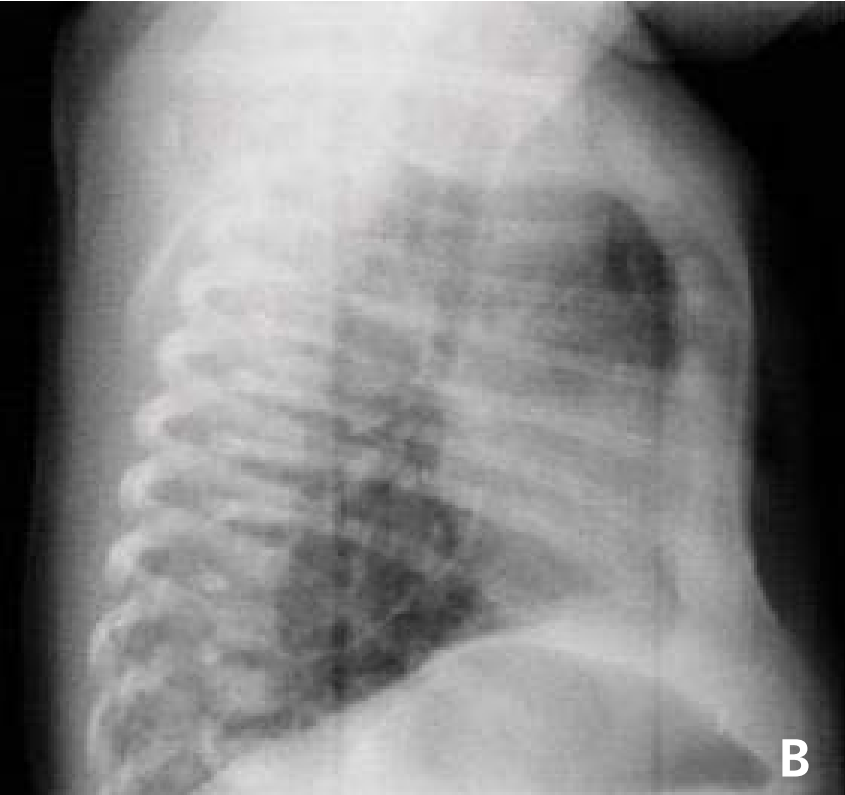


Fig. 2

A

Chest X-ray at the age of 10 days, demonstrating an expansive radiolucent lesion in the right lower lobe consistent with PIE (A) anterior and B) lateral view).



B

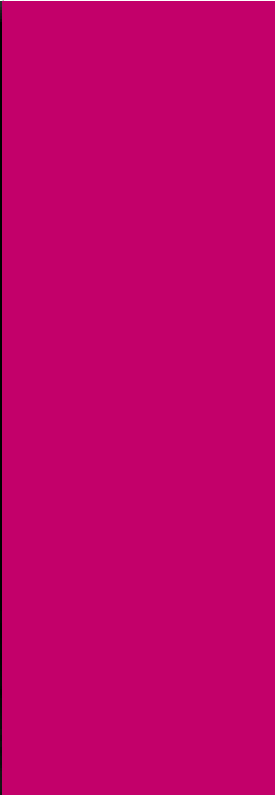
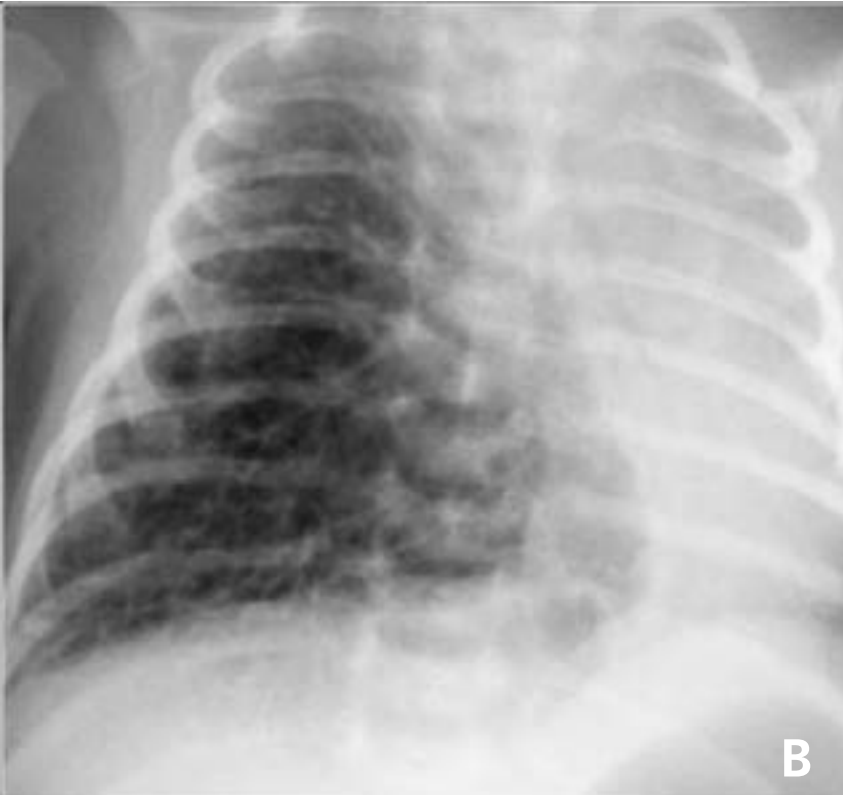




Fig. 3

Chest X-rays at the age of 18 (A) and 20 (B) days demonstrating worsening PIE, leading to progressive mediastinal shift to the left.



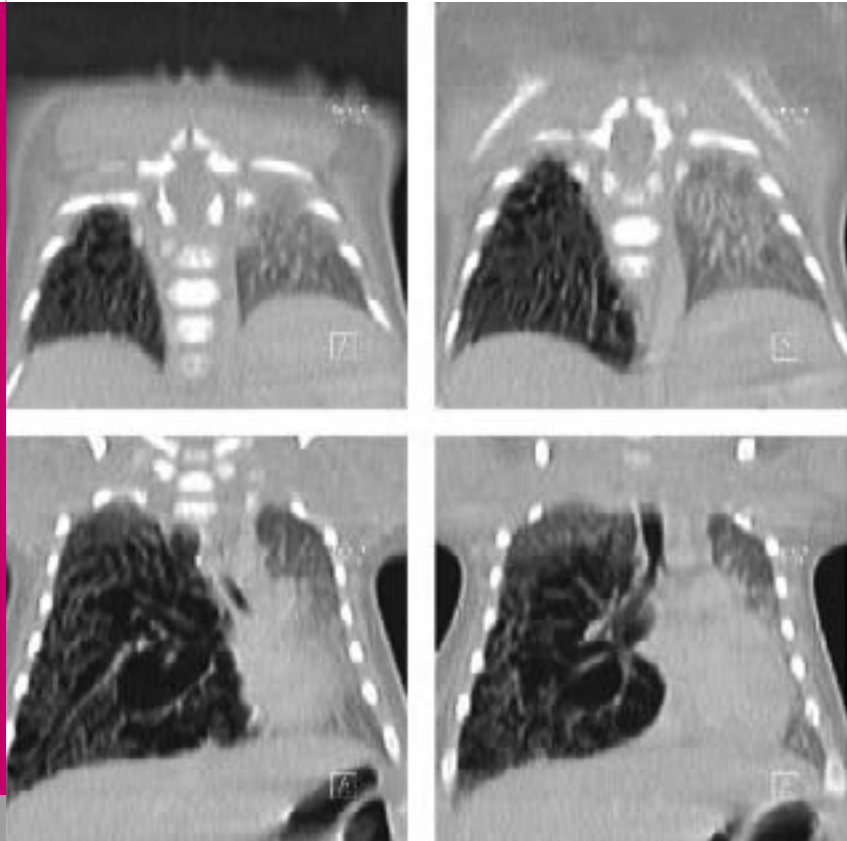
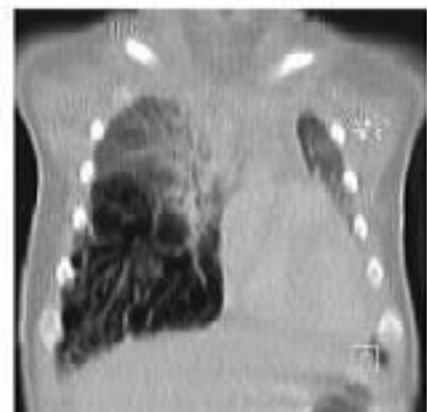
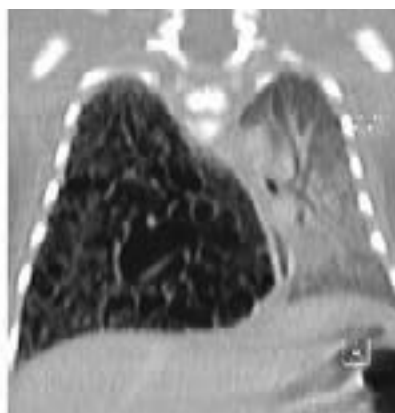


Fig. 4

Selected coronal views of a chest CT at the age of 16 days showing the expansive cystic lesion in the right lower lobe and mediastinal shift to the left.



lation, the ETT was pulled back above the carina. This led to slow improvement in ventilation but also to a recurrence of the PIE (Fig. 7B). Again, the baby was put in a right lateral decubitus position. Fortunately, near total atelectasis of the right lung was achieved (Fig. 7C) and normal lung inflation occurred when positional therapy was discontinued (Fig. 7D). The girl was extubated after 7 days of mechanical ventilation and made an uneventful recovery. She was discharged home at a corrected gestational age of 40 6/7 weeks.

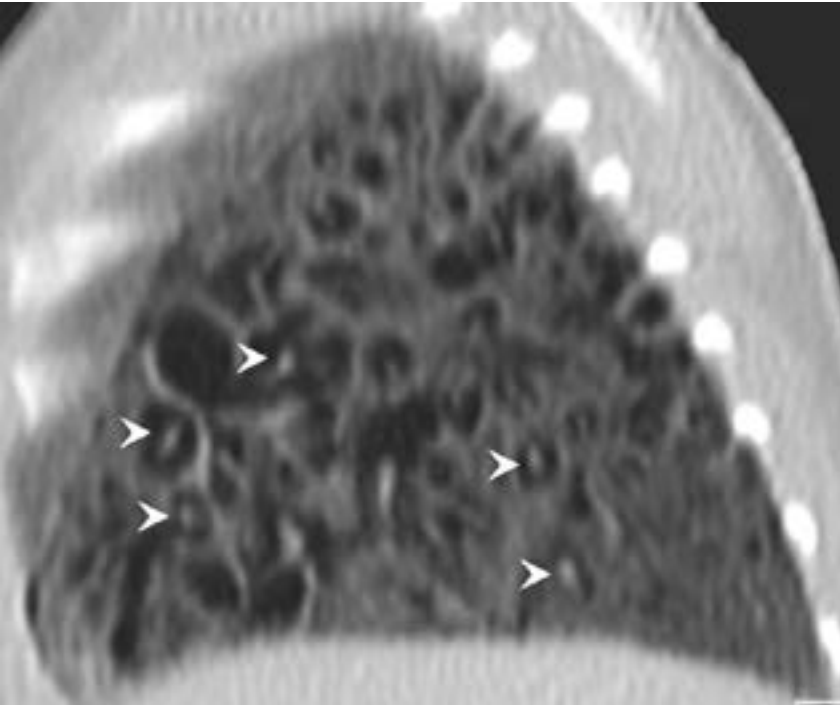


Fig. 5

Chest CT with bronchovascular bundles (arrow heads) appearing as white dots in the center of the air-filled cysts („hanging dots“ sign).

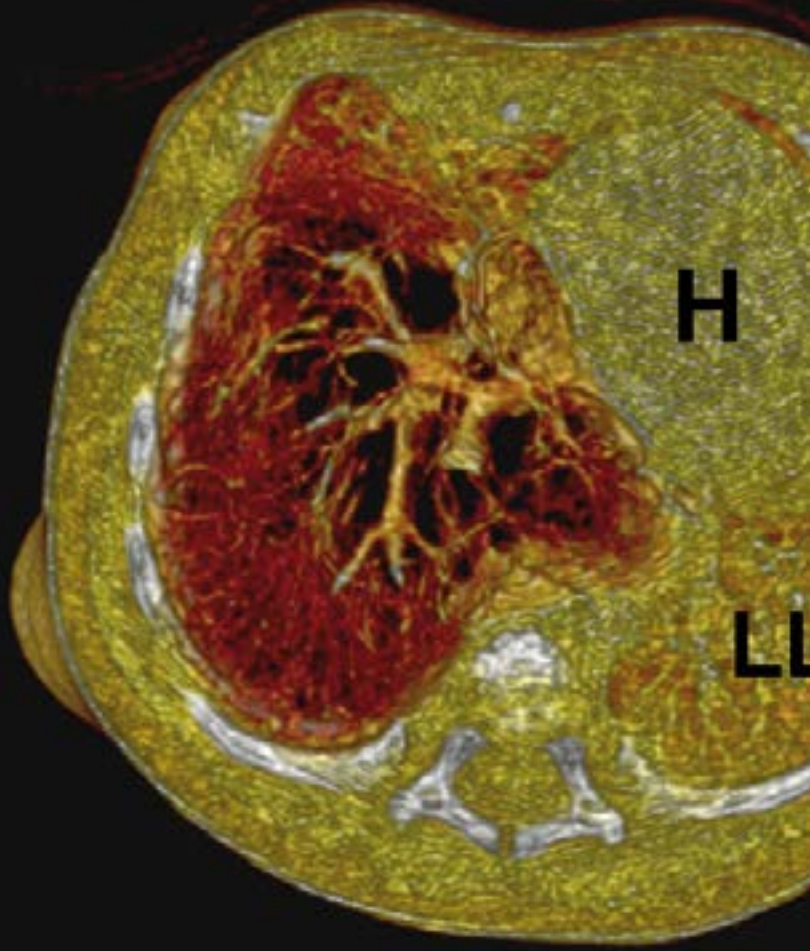
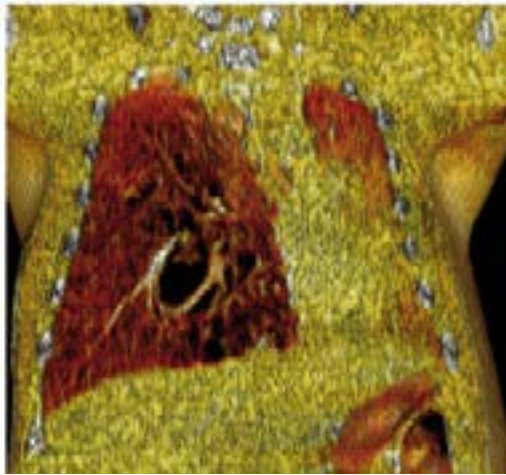
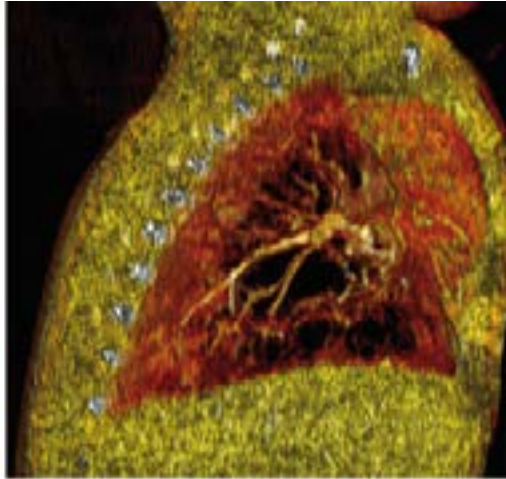
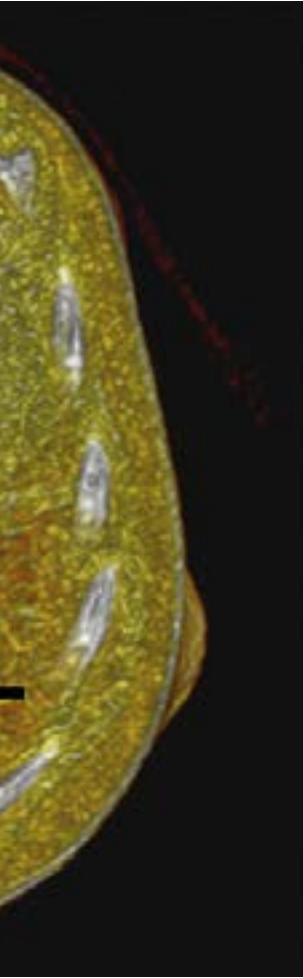
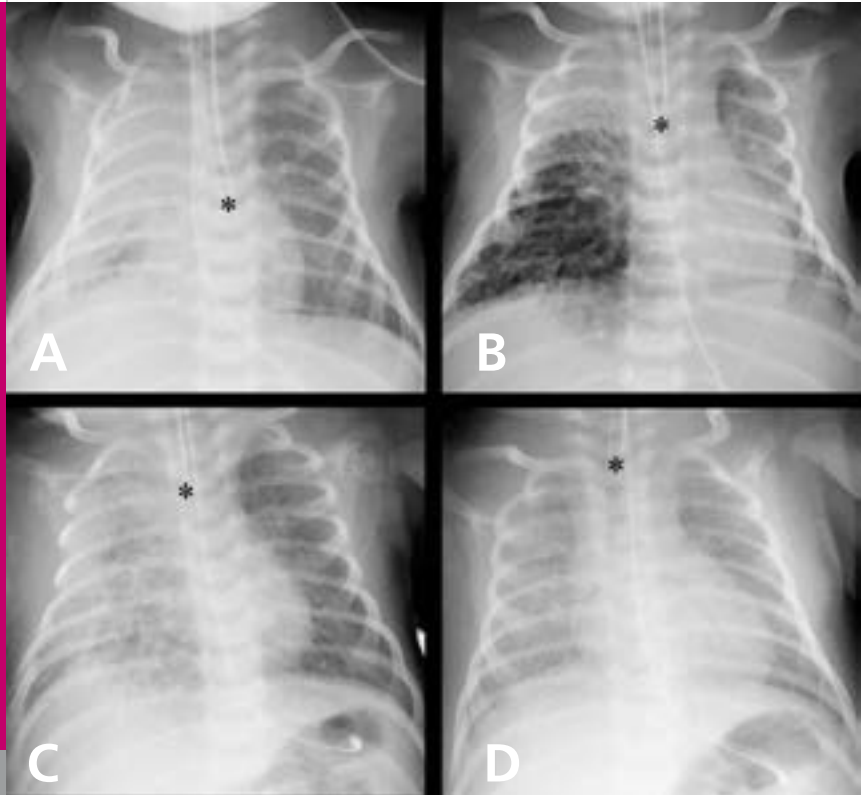


Fig. 6

3-D reconstruction of chest CT images with large emphysematous interstitial spaces in the expanded right lower lobe, mediastinal shift (H: heart) and compressed left lung (LL).



**Fig. 7**

Series of chest X-rays (tip of ETT; asterisk): A) selectively intubated left main stem bronchus with atelectasis of the right lung; B) recurrence of PIE after withdrawal of the ETT; C) subtotal atelectasis of the right lung achieved by strict right lateral decubitus positioning; D) reventilated expanded right lung without evidence of PIE recurrence.

PIE was first described by Macklin in 1943 and its pathophysiology is still known as the „Macklin effect“ (1). This term relates to a three-step pathophysiologic process: 1) blunt traumatic alveolar rupture, 2) air dissection along bronchovascular sheaths, and 3) spreading of the pulmonary interstitial emphysema into the mediastinum (2, 3). The rupture of the alveoli is created by overdistension of the alveoli due to increased tidal volumes rather than high airway pressures (4). PIE is a well recognized complication of neonatal respiratory distress syndrome. It occurs predominantly in preterm neonates undergoing mechanical ventilation, but has also been described in both full-term and spontaneously breathing infants (5).

In some cases, PIE persists and is then referred to as persistent pulmonary interstitial emphysema (PPIE). Histologically, PPIE is characterized by giant cells felt to be a reaction to prolonged air trapping (5). Cyst size in localized PPIE can measure up to 3 cm while in diffuse PPIE the cysts are usually smaller (6).

The diagnosis of PIE may be suspected based on the clinical history as well as the occurrence of characteristic cystic changes and - in the case of localized PIE - mediastinal shift on the chest X-ray. Occasionally a CT scan may be required to confirm the diagnosis and rule out congenital malformations (see below). The bronchovascular bundles appear as a dot in the center of the air-filled cysts („hanging dot“ sign, Fig. 5). This

appearance is unique to PIE and distinguishes it from other radiolucent chest lesions in neonates (6, 7).

The major differential diagnoses of PIE are congenital cystic adenomatoid malformation (CCAM) and congenital lobar emphysema (CLE). CCAM is a developmental abnormality. Adenomatoid proliferation of bronchial structures leads to the formation of cysts at the expense of normal alveoli. On CT scan, cystic changes are associated with abnormal soft tissue. The treatment is surgical resection even in asymptomatic children in order to prevent infection and because rarely rhabdomyosarcoma can develop within the lesion (8). Congenital lobar emphysema (CLE) is a congenital over-expansion of a pulmonary lobe. The left upper lobe is most commonly affected. Immediately after birth, the chest x-ray is likely to show a radiopaque lobe because of retained fetal lung fluid. Following progressive clearing of the fluid the affected region appears hyperlucent. In contrast to PIE, where the air accumulates in the interstitial space, the alveoli themselves are distended in CLE. Therefore, in patients with CLE, the CT scan shows cysts with the vascular structures at the periphery rather than in the center (7).

Several reports have described the successful noninvasive management of unilateral PIE and even PPIE using strict lateral decubitus positioning with the emphysematous lung dependent. In ventilated infants, appropriate ventilator settings consisting of small tidal vo-

lumes, increased ventilator rate and short inspiratory times are crucial (9). In some cases high frequency oscillatory ventilation (HFOV) may be beneficial.

If these measures fail, more invasive procedures like single-lung ventilation are required. While right main stem bronchus intubation can easily be performed at the bedside for treatment of left-sided PIE, left main stem bronchus intubation for the treatment of right-sided PIE is technically more challenging because of the airway anatomy (10). The duration of single-lung ventilation required for resolution of PIE is unknown and reports in the literature describe durations between 1 to 10 days.

An alternative technique of single-lung ventilation is to use a Swan-Ganz catheter to block the main stem bronchus on the affected side (11). If this technique is used, it is important to deflate and reinflate the balloon every hour in order to prevent partial bronchial occlusion leading to a ball valve effect and worsening of air trapping.

Several case reports and a recent case series describe lung puncture with the creation of an artificial ipsilateral pneumothorax and subsequent drainage as a therapeutic option before proceeding to excisional surgery (12). However, because the experience with this approach is still limited, the safety of this procedure is difficult to assess.

See also **COTM 10/2000**: Selective bronchial intubation for unilateral PIE and **COTM 12/2002**: Selective bronchial occlusion in a preterm infant with unilateral pulmonary interstitial emphysema.

REFERENCES

1. Macklin MT, Macklin CC. Malignant interstitial emphysema of the lungs and mediastinum as an important occult complication in many respiratory diseases and other conditions: an interpretation of the clinical literature in the light of laboratory experiment. *Medicine* 1944;23:281–358
2. Jassal MS, Benson JE, Mogayzel PJ Jr. Spontaneous resolution of diffuse persistent pulmonary interstitial emphysema. *Pediatr Pulmonol* 2008;43:615-9
3. Dembinski J, Heep A, Kau N, Knöpfle G, Bartmann P. CT imaging of pulmonary lobar interstitial emphysema in a spontaneous breathing preterm infant. *Am J Perinatol* 2002;19:285-90
4. Caldwell EJ, Powell RD Jr, Mullooly JP. Interstitial emphysema: a study of physiologic factors involved in experimental induction of the lesion. *Am Rev Respir Dis* 1970;102:516-25
5. Freysdottir D, Olutoye O, Langston C, Fernandes CJ, Tatevian N. Spontaneous pulmonary interstitial emphysema in a term unventilated infant. *Pediatr Pulmonol* 2006;41:374-8
6. Jabra AA, Fishman EK, Shehata BM, Perlman EJ. Localized persistent pulmonary interstitial emphysema: CT findings with radiographic-pathologic correlation. *AJR Am J Roentgenol* 1997;169:1381-4

10. Chalak LF, Kaiser JR, Arrington RW. Resolution of pulmonary interstitial emphysema following selective left main stem intubation in a premature newborn: an old procedure revisited. *Paediatr Anaesth* 2007;17:183-6
11. Rastogi S, Gupta A, Wung JT, Berdon WE. Treatment of giant pulmonary interstitial emphysema by ipsilateral bronchial occlusion with a Swan-Ganz catheter. *Pediatr Radiol* 2007;37:1130-41
12. Dördelmann M, Schirg E, Poets CF, Ure B, Glüer S, Bohnhorst B. Therapeutic lung puncture for diffuse unilateral pulmonary interstitial emphysema in preterm infants. *Eur J Pediatr Surg* 2008;18:233-236

SUPPORTED BY



CONTACT

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