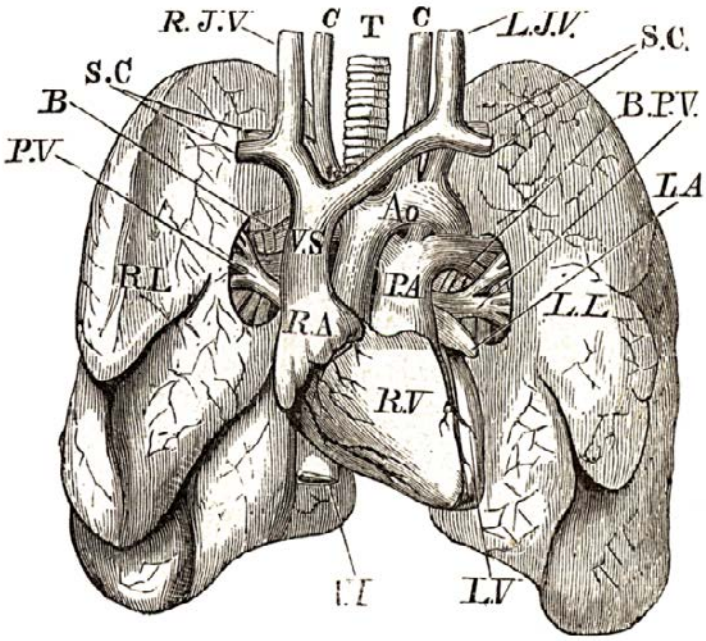
SWISS SOCIETY OF NEONATOLOGY

Challenges in postnatal management of congenital lung malformations

January 2019



Heyne-Pietschmann M, Stocker M, Regamey N, Hodel M, Szavay PO, Pediatric Surgery (HPM, SPO), Neonatal and Pediatric Intensive Care Unit (SM), Pediatric Respiratory Medicine (RN), Children's Hospital of Lucerne, Switzerland, Department of Obstetrics and Gynecology (HM), Women's Hospital of Lucerne, Switzerland

Title figure:
Anatomical drawing (source: www.clipartxtras.com)

INTRODUCTION

Congenital lung malformations (CLM) represent a heterogeneous group of rare developmental disorders including congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (BPS), congenital lobar emphysema (CLE), as well as hybrid lesions.

Nowadays, most of these lesions are detected by routine prenatal ultrasound; affected patients may either present with severe respiratory distress in the newborn period or remain asymptomatic until later in life.

Pre-, peri- and postnatal management of these patients remains challenging and requires interdisciplinary teamwork for decision making on the best individual treatment strategy. Minimally invasive surgical techniques for CLM have been developed over the last decades and are increasingly replacing open surgery.

We present two cases of congenital lung malformations, which illustrate the wide spectrum of clinical presentations of affected newborns and the ensuing differences in postnatal management including minimally invasive surgical techniques.

CASE REPORT 1

In this male fetus, prenatal ultrasound examination revealed a large, left-sided cystic lung lesion accompanied by severe mediastinal shift and dextroposition of the heart (Fig. 1, 2). A CPAM Volume Ratio (CVR) of 3:4 was calculated.

Due to massive expansion of the lesion, intrauterine drainage procedures, such as cyst aspiration or thoraco-amniotic shunting were discussed – but, as no signs of hydrops occurred, fetal intervention was not deemed necessary.

Elective Cesarean section was performed at 37 2/7 weeks of gestation. Apgar scores were 7, 8 and 8 at 1, 5 and 10 minutes, respectively. The male neonate showed signs of respiratory distress and was intubated shortly after birth because of a persistently high oxygen requirement. The patient was transferred to the NICU for further diagnostic evaluation and treatment.

On the first day of life, the infant remained stable on mechanical ventilation with a low oxygen requirement. A babygram showed a large cystic lesion in the left hemithorax (Fig. 3). On echocardiography, there was good biventricular function with mild persistent pulmonary hypertension and a small persistent ductus arteriosus. Delayed pulmonary left upper lobe resection was planned to prevent possible deterioration due to pulmonary hypertension.

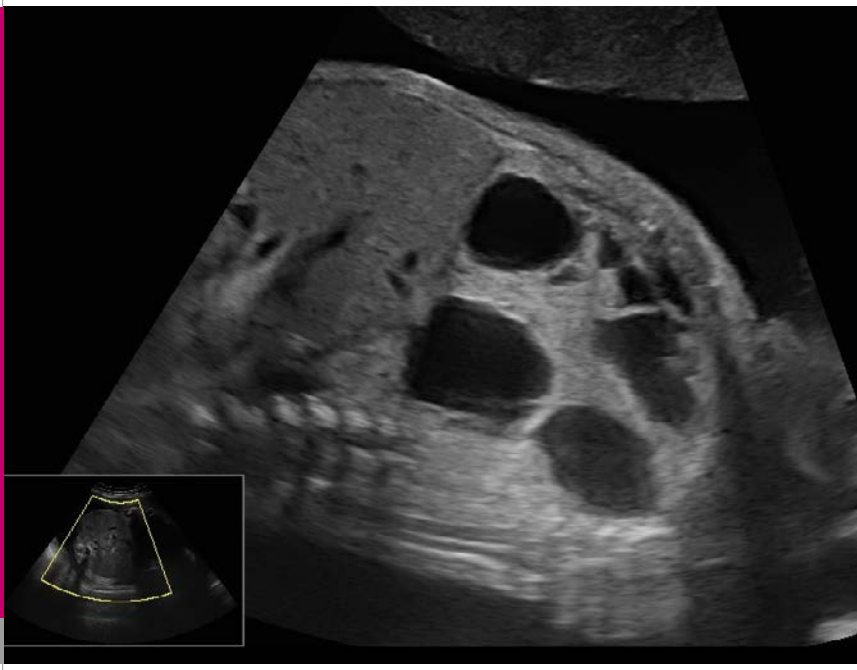


Fig. 1

Prenatal ultrasound (sagittal view) revealing a large, left-sided cystic lung lesion.

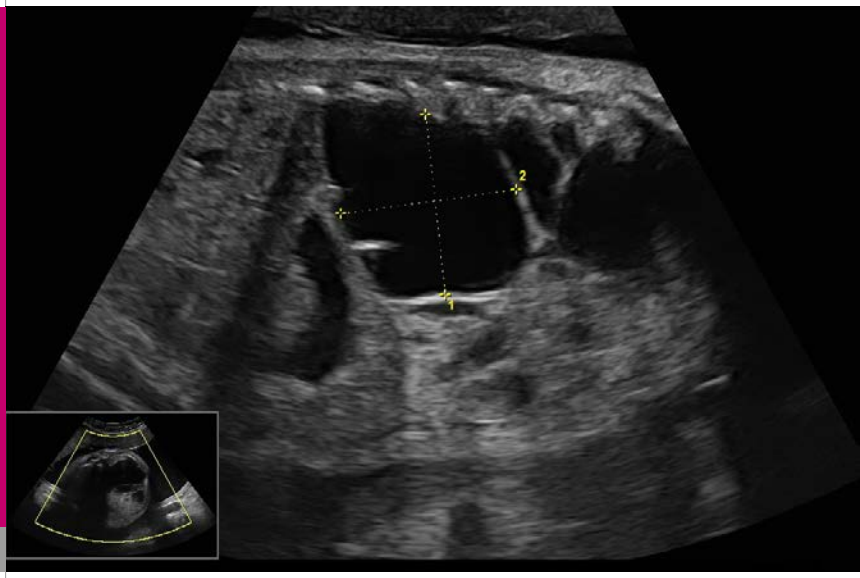


Fig. 2

Prenatal ultrasound (sagittal view) revealing a large, left-sided cystic lung lesion.

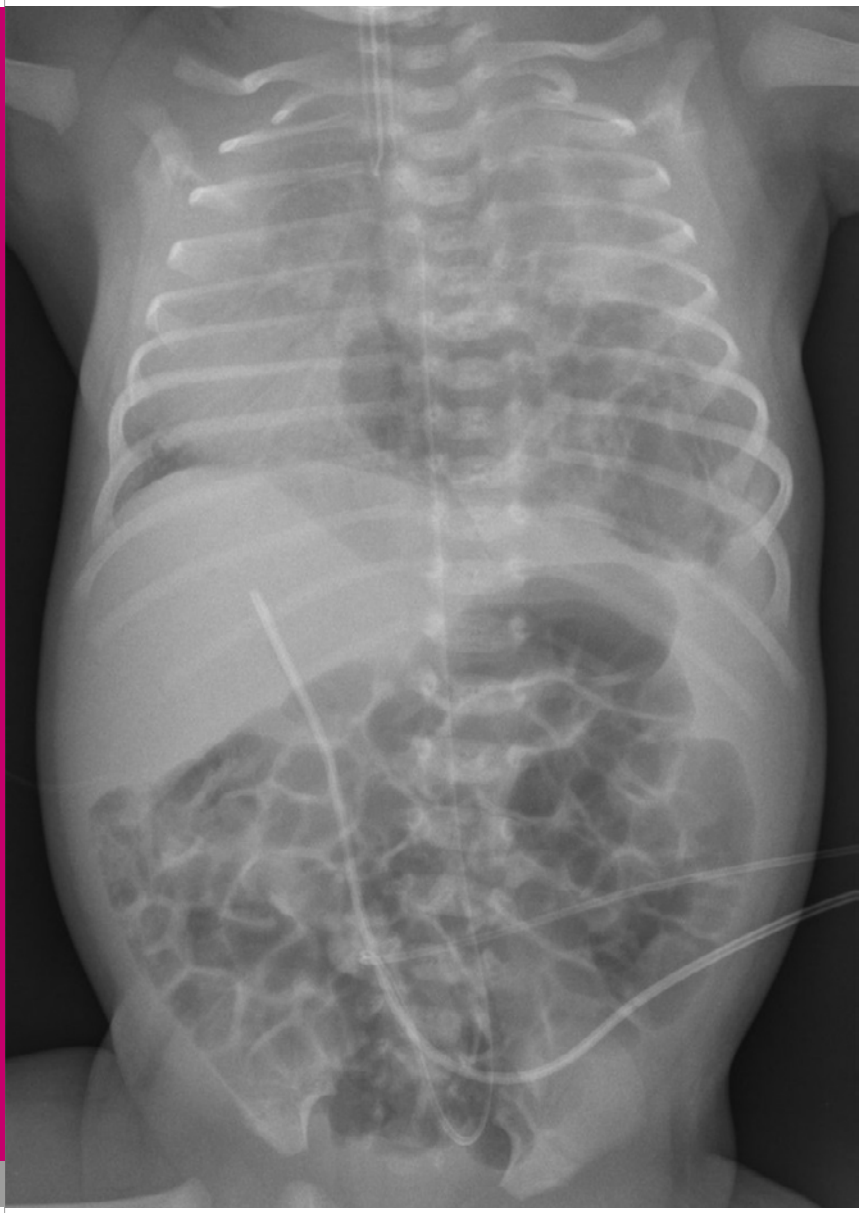


Fig. 3

Babygram on first day of life showing large cysts with mediastinal shift and compression of the contralateral lung.

On the second day of life, respiratory distress worsened due to increasing mediastinal shift and compression of the contralateral lung. Chest X-ray showed an increasing expansion of the cysts. A thoracic drain was inserted to drain the largest cyst, which lead to transient improvement of gas exchange. An attempt to extubate the patient on the sixth day of life failed and re-intubation was required shortly thereafter.

On the 7th day of life, a CT scan of the chest was obtained. It showed a progressively expanding CPAM with consecutive atelectasis of the left lower lobe and the right lung with severe mediastinal shift to the right and a small pneumothorax on the left (Fig. 4 – 6). Echocardiography revealed increasing pulmonary hypertension.

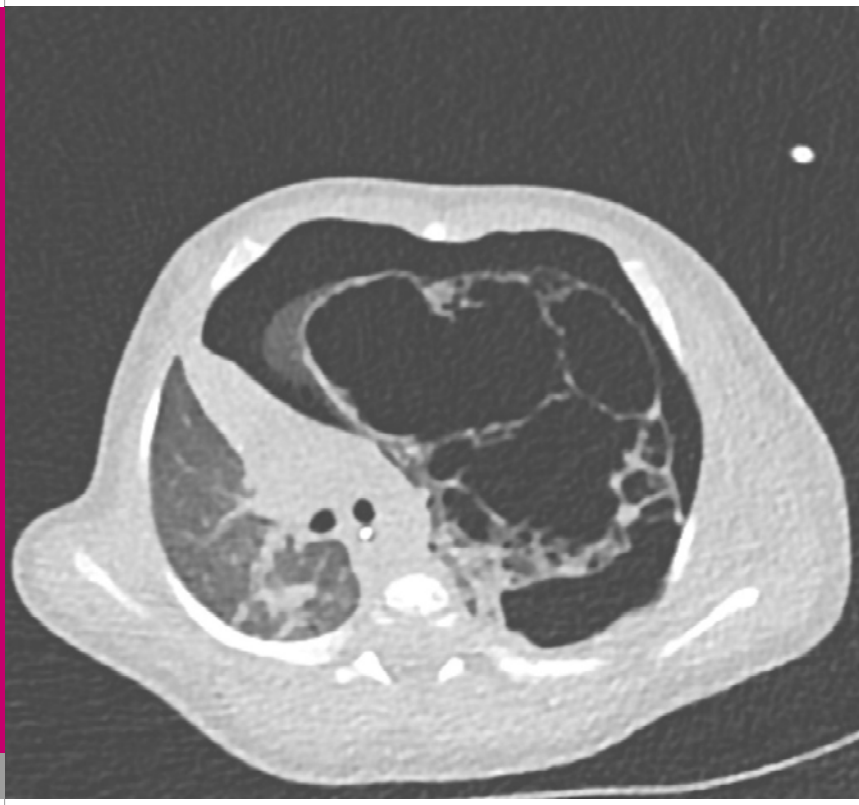


Fig. 4

CT-scan (axial view) on the seventh day of life demonstrating massive expansion of the CPAM.

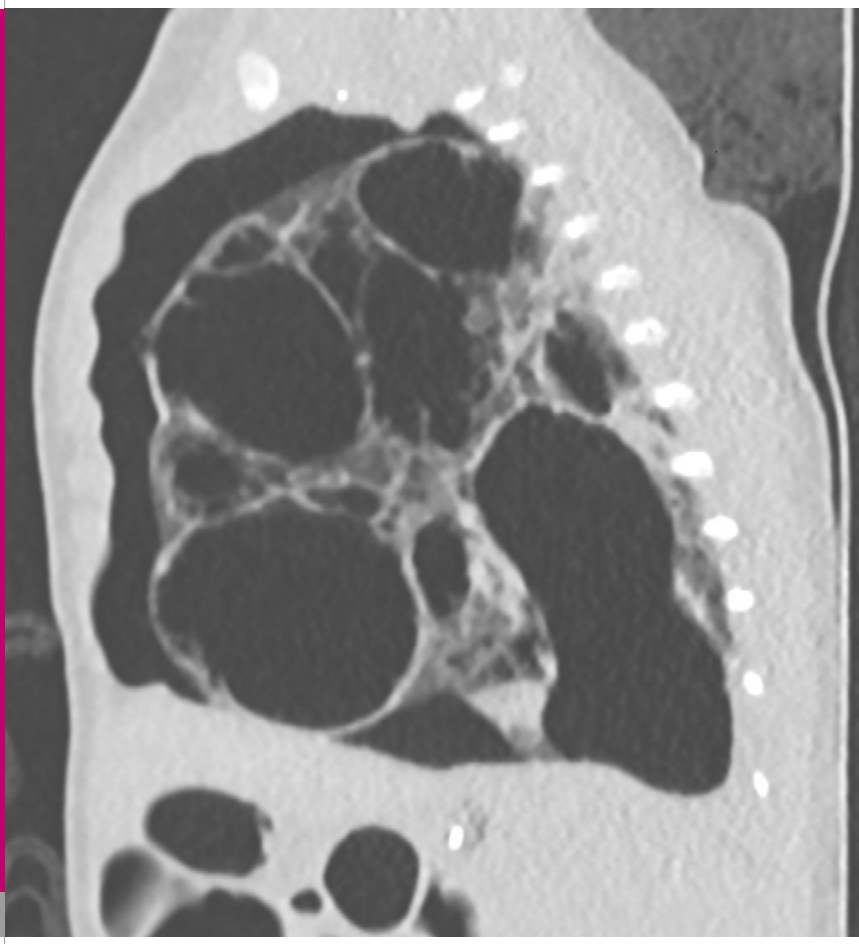


Fig. 5

CT-scan (sagittal view) on the seventh day of life demonstrating massive expansion of the CPAM.

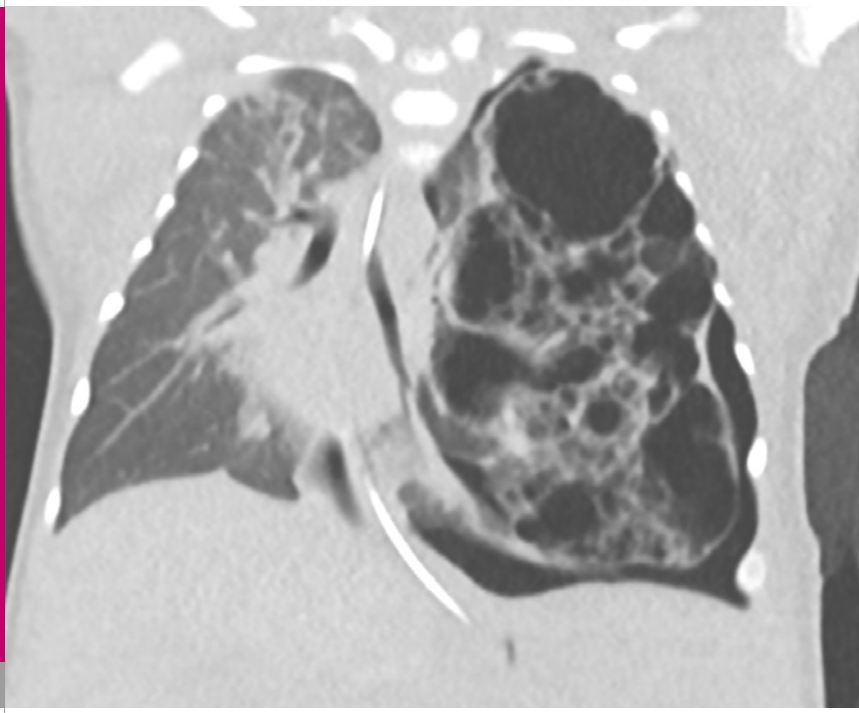


Fig. 6

CT-scan (coronal view) on the seventh day of life demonstrating massive expansion of the CPAM.

In the second week of life, further expansion and multiple spontaneous left-sided pneumothoraces occurred, further compromising the respiratory situation (Fig. 7, 8). Cyst ruptures as well as a broncho-pulmonary fistula were suspected, and multiple chest drains were inserted.

When further respiratory deterioration occurred, thoracoscopy was performed on day of life 11. A large cyst could be electrocoagulated, leading to intraoperative expansion and ventilation of the left lower lobe (Fig 9, 10). After surgery, regression of the cyst and the mediastinal shift could be observed on repeated chest X-rays.

Unfortunately, later on, respiratory distress worsened again with increasing oxygen and ventilation pressure requirements. Imaging revealed massive progression of cyst size and compression of the contralateral lung (Fig. 11), and, in the 4th week of life, left upper lobectomy was performed through a postero-lateral thoracotomy. The left lower lobe could be preserved. Histopathology of the resected lobe confirmed diagnosis of congenital pulmonary airway malformation type 1.

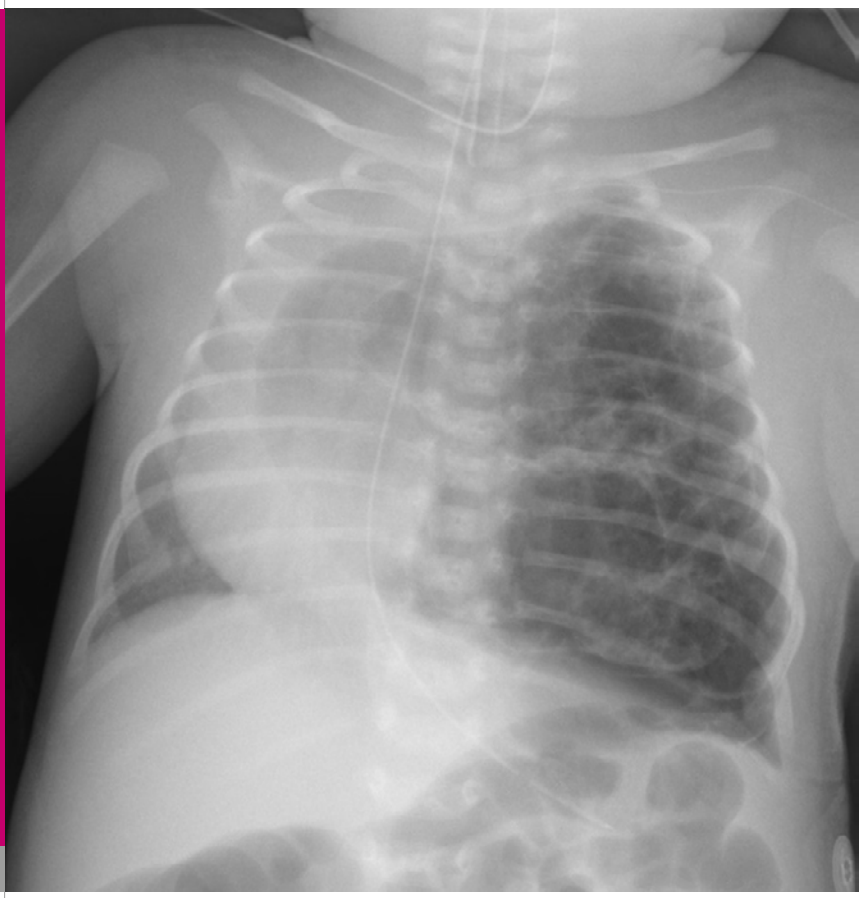


Fig. 7

Chest x-ray (2nd week of life) demonstrating further expansion of the left-sided lung malformation.

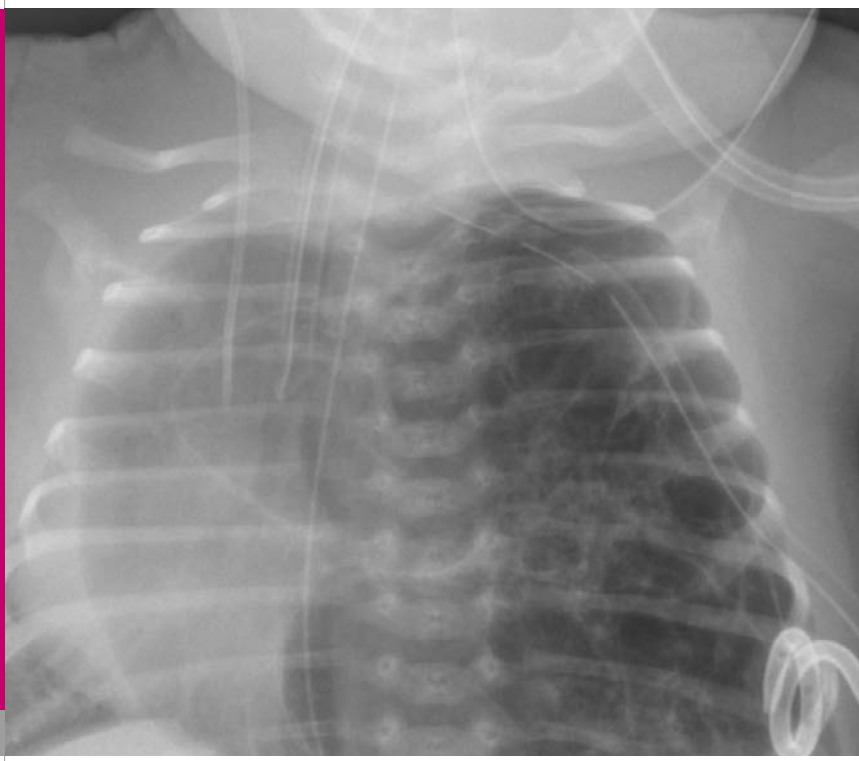


Fig. 8

Chest x-ray (2nd week of life) after drainage of spontaneously occurring pneumothoraces.

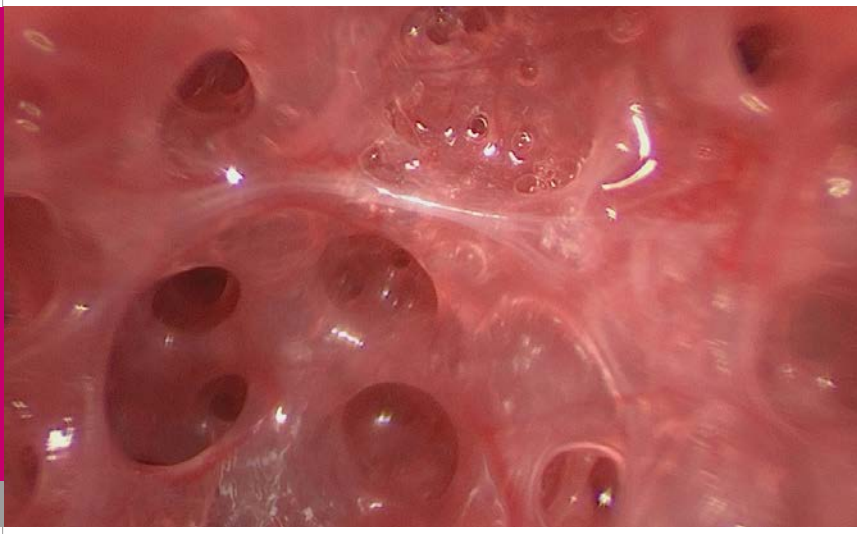


Fig. 9

Intraoperative view of the big cysts.

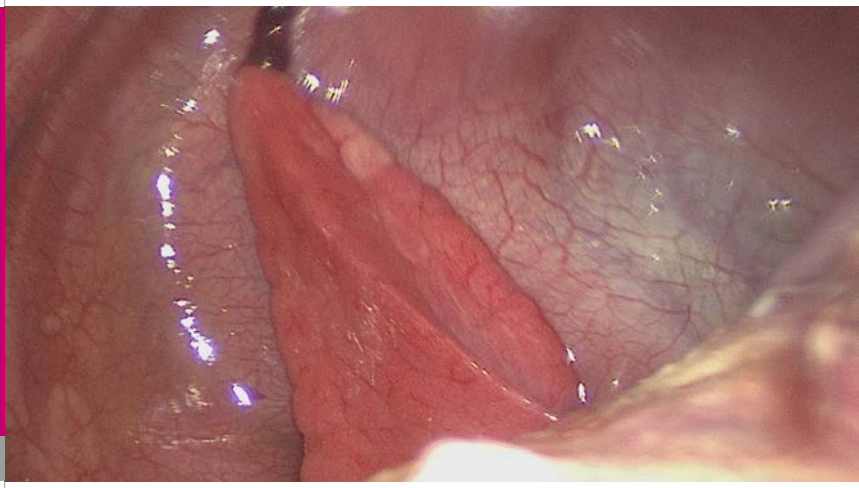


Fig. 10

Intraoperative view of the left lower lobe after cyst coagulation.

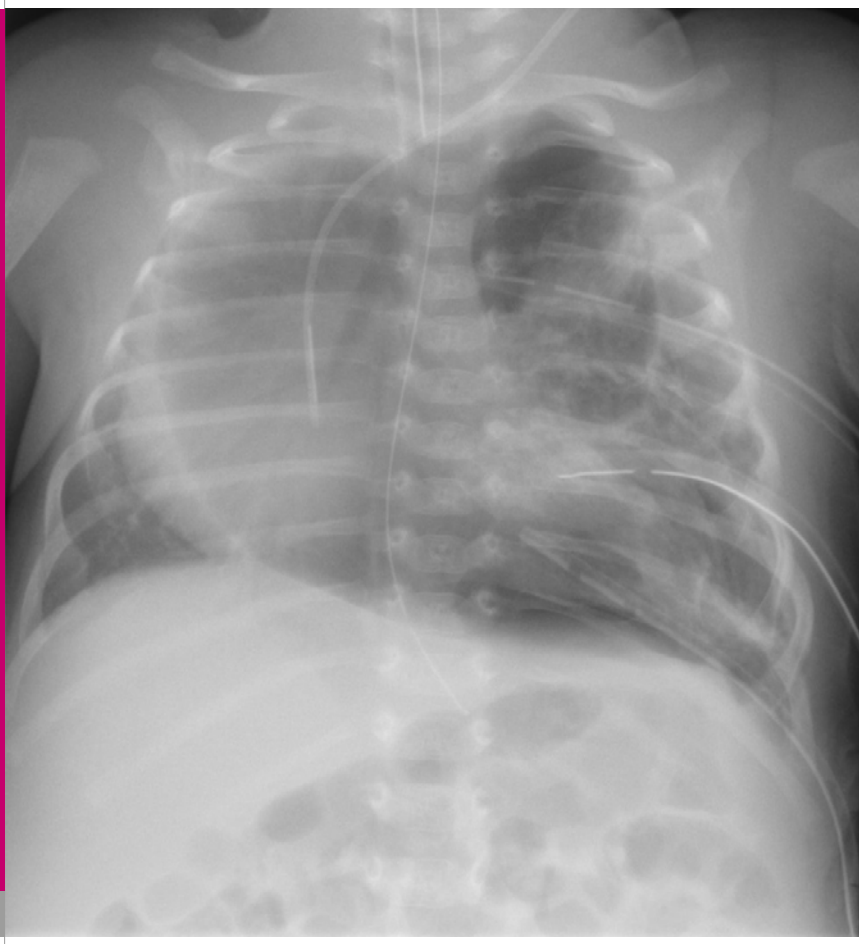


Fig. 11

In the third week of life, imaging revealed once again massive progression of cyst size with mediastinal shift and compression of the left lung.

The postoperative course was uneventful. A postoperative chest X-ray revealed good expansion of the contralateral lung with disappearance of the mediastinal shift (Fig. 12). The chest drain could be removed on the second postoperative day and the patient was extubated two days later.

The patient could be discharged home at six weeks of life. During long-term follow-up no respiratory tract infections occurred, and the boy remained asymptomatic.

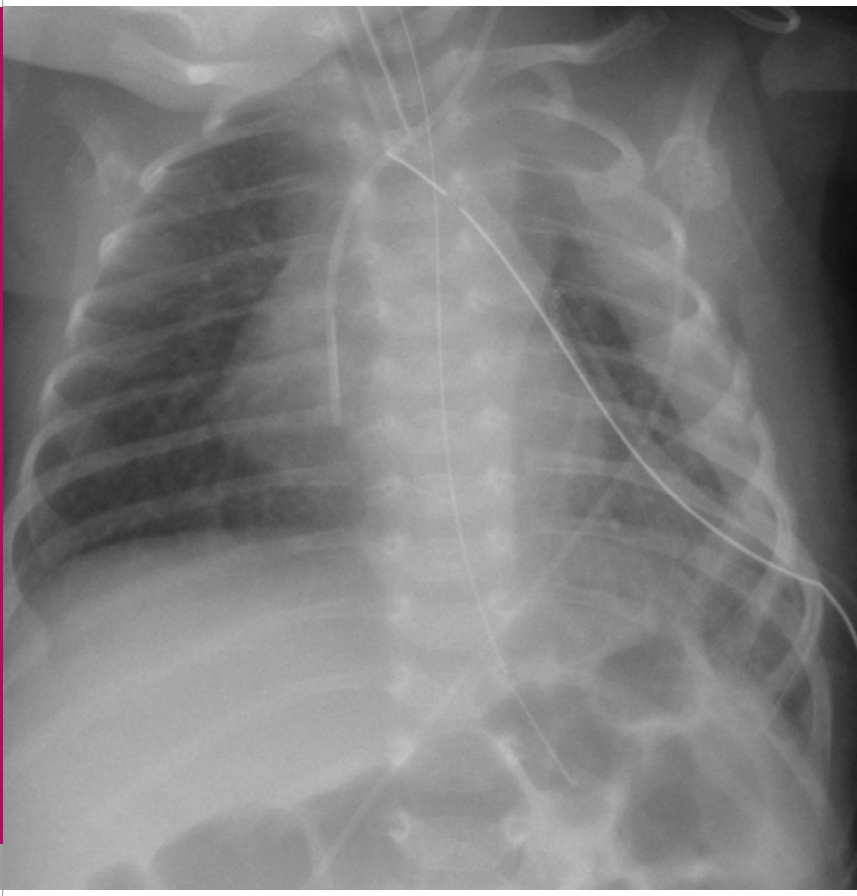


Fig. 12

Postoperative chest x-ray showing resolution of the mediastinal shift and good expansion of the right lung and.

CASE REPORT 2

In this female patient, pulmonary sequestration was suspected during routine prenatal ultrasound examination at 21 weeks of gestation. The left-sided lung lesion measured 22 mm × 21 mm × 16 mm and had an arterial supply deriving from the thoracic aorta (Fig. 13, 14). Over time, the lesion remained stable in size and configuration. The findings were felt to be consistent with intralobar sequestration.

The girl was born through vaginal delivery at 39 3/7 weeks of gestation and had a birth weight of 3100 g. She adapted well and showed no signs of respiratory distress.

On day 3 of life, there was no evidence of pulmonary sequestration or any other lung malformation on an ultrasound examination, and the girl was discharged home.

At 6 months of age, a CT scan of the thorax demonstrated an intralobar sequestration (size 30 mm × 25 mm × 10 mm) with an arterial supply from the thoracic aorta and venous drainage into lower pulmonary veins (Fig. 15).

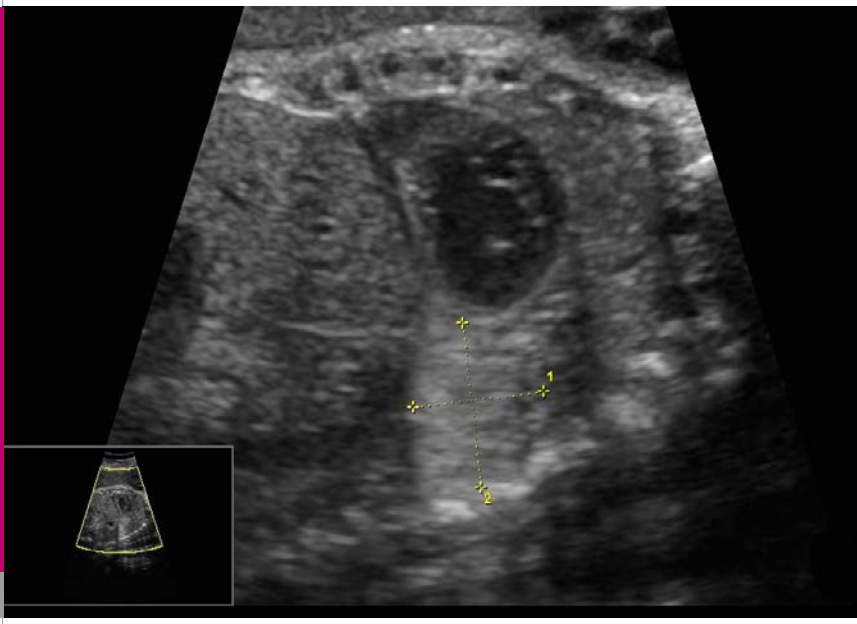


Fig. 13

Prenatal ultrasound (sagittal view) revealing left-sided pulmonary sequestration.

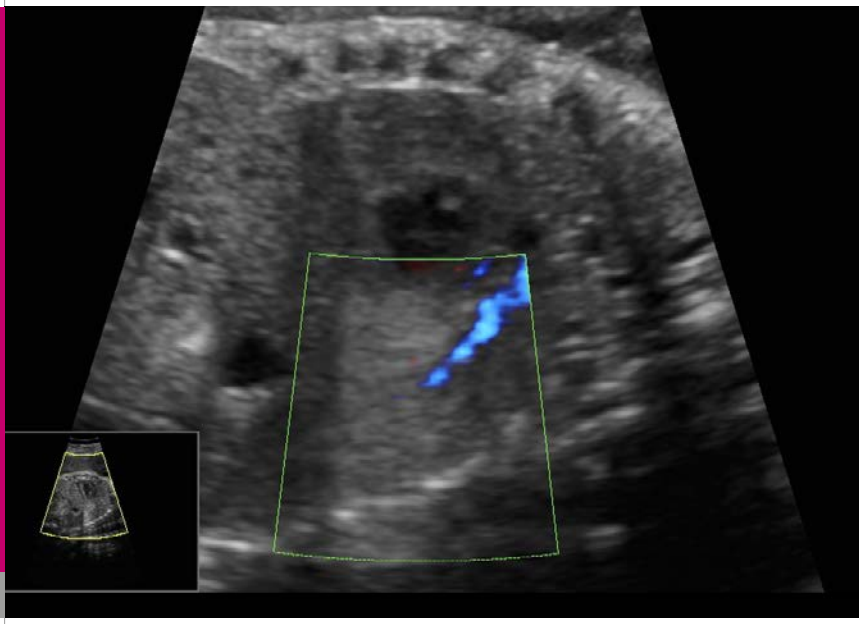


Fig. 14

Prenatal ultrasound (sagittal view with color Doppler) revealing a left-sided pulmonary sequestration.

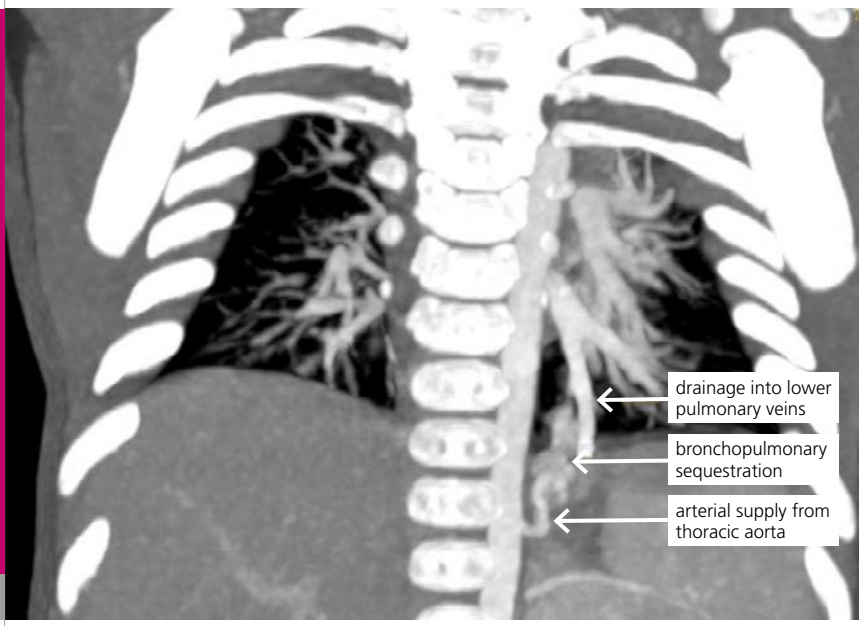


Fig. 15

CT scan (coronal view) at 6 months of age showing an intralobar sequestration with arterial supply from the thoracic aorta and venous drainage into lower pulmonary veins.

Thoracoscopic surgical resection of the sequestration was performed at 9 months of age. Intraoperatively, a feeding artery could be identified. In contrast to the findings of preoperative imaging studies, the majority of the sequestration was in an extralobar position. Uncomplicated and complete resection of the sequestration could be achieved (Fig. 16). The postoperative course was uneventful (Fig. 17), and the patient could be discharged home on the 4th postoperative day. During long-term follow-up, no infections occurred and the girl remained asymptomatic.

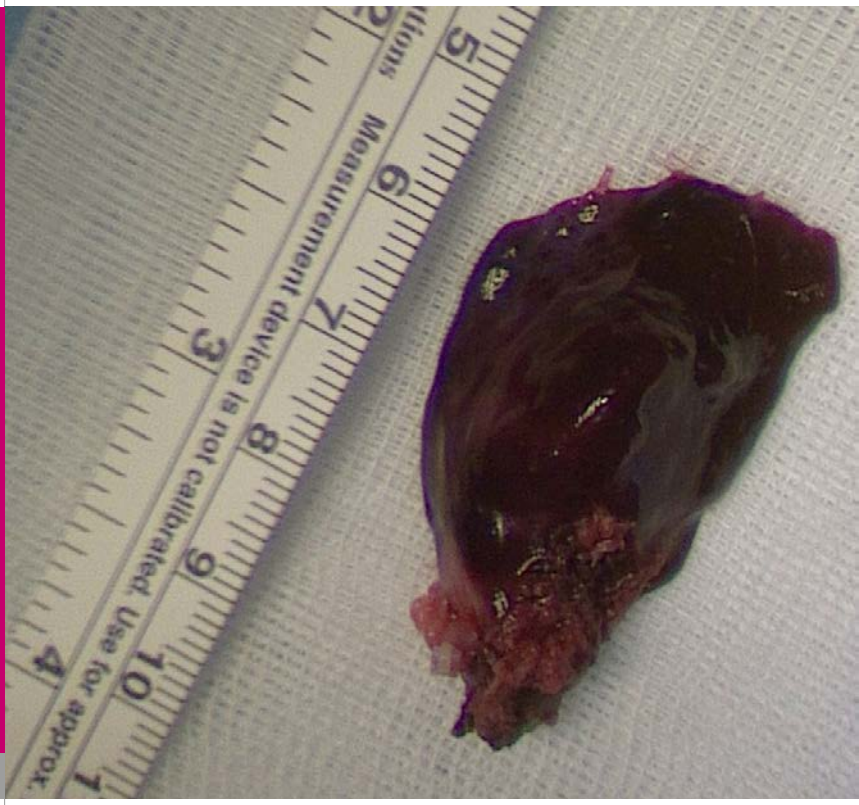


Fig. 16

Resected lung sequester.

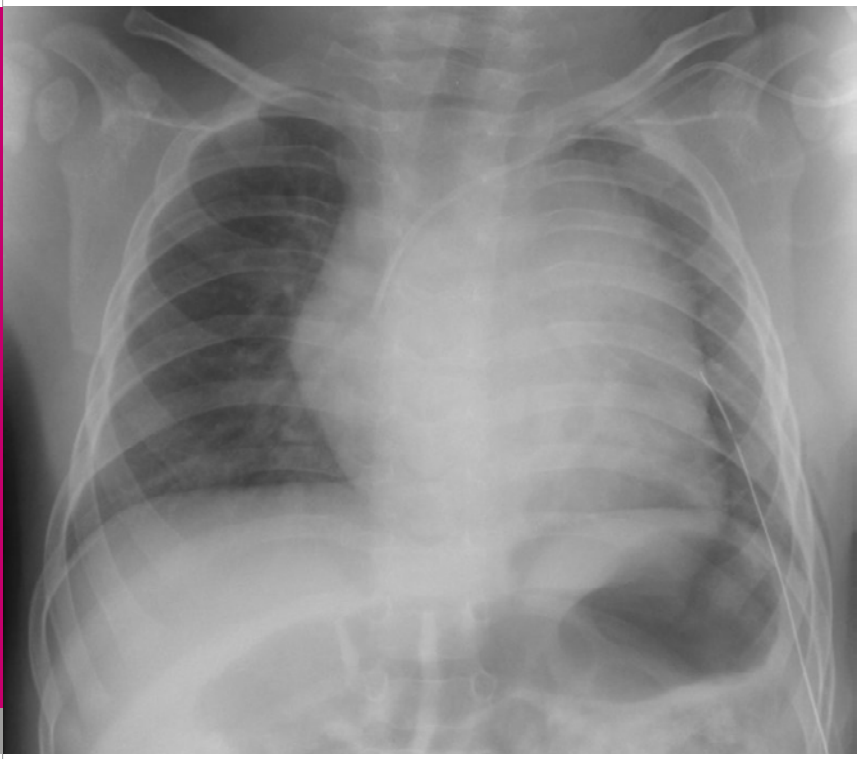


Fig. 17

Postoperative chest X-ray after resection of the left-sided lung sequester.

DISCUSSION

Pre- and postnatal management of antenatally detected CLM remains challenging and some aspects are still discussed controversially. In the neonate, CLM may present with a large spectrum of symptoms. Evaluation and management must be individualized to define the best treatment strategy for each patient.

It is widely accepted that symptomatic lung lesions should be resected (1, 2, 7). Timing of surgery depends on the infant's condition and the severity of respiratory distress. In patients with significant respiratory distress and large lesions, it may be necessary to perform surgery urgently. However, in general, surgery for CLM should be elective and performed later in the first year of life before patients become symptomatic (2, 5, 7, 8). Published literature supports surgical interventions in the first year of life; perioperative complications and technical difficulties during surgery occur rather more frequently after the onset of clinically apparent or subclinical infections (1, 5, 6, 9).

Thoracoscopic resection of CLM should be the standard approach as perioperative complications and outcomes are comparable to open surgery and the morbidities associated with thoracotomy can be avoided (3, 6, 9).

The management of asymptomatic lesions is still a subject of controversy. Some authors advocate an early resection strategy to avoid infections and

to facilitate potential compensatory lung growth. Furthermore, they argue that imaging may not provide a definite diagnosis and often cannot distinguish between benign lesions and lesions with the potential for malignant transformation. Early elective surgery also obviates long observation periods with repeated imaging studies with ionizing radiation (1, 2, 4, 7).

Other authors, however argue in favor of a conservative strategy with close observation of asymptomatic patients to avoid overtreatment and allow time for spontaneous resolution (7, 10).

With the presentation of the two cases, we would like to emphasize the importance of an individualized approach to patients with CLM. We recommend elective surgical resection also for asymptomatic patients as surgery has been shown to be safe and feasible even in infants. Thoracoscopic resection should be favored but might have its limitations in complicated cases or in larger lesions.

REFERENCES

1. Singh S, Davenport M. The argument for operative approach to asymptomatic lung lesions. Semin Pediatr Surg 2015;24: 187–195 *(Abstract)*

2. Stanton M, Njere I, Ade-Ajayi N, Patel S, Davenport M. Systematic review and meta-analysis of the postnatal management of congenital cystic lung lesions. J Pediatr Surg 2009;44:1027 *(Abstract)*

3. Adams S, Jobson M, Sangnawakij P, et al. Does thoracoscopy have advantages over open surgery for asymptomatic congenital lung malformations? An analysis of 1626 resections. J Pediatr Surg 2017;44:1027 *(Abstract)*

4. Morini F, Zani A, Conforti A, et al. Current management of congenital pulmonary airway malformations: A «European Pediatric Surgeons' Association» survey. Eur J Pediatr Surg 2018;28:1 – 5 *(Abstract)*

5. Jelin EB, O'Hare EM, Jancelewicz T, Nasr I, Boss E, Rhee DS. Optimal timing for elective resection of asymptomatic congenital pulmonary airway malformations. J Pediatr Surg 2018;53:1001 – 1005 *(Abstract)*

6. Kunisaki SM, POwelson IA, Haydar B, et al. Thoracoscopic vs open lobectomy in infants and young children with congenital lung malformations. J Am Coll Surg 2017;218:261 – 270 *(Abstract)*

7. David M, Limas-Pinheiro R, Henriques-Coelho T. Prenatal and postnatal management of congenital pulmonary airway malformation. Neonatology 2016;110:101 – 115 *(Abstract)*

8. Downard CD, Calkins CM, Williams RF, et al. Treatment of congenital pulmonary airway malformations: a systematic review from the APSA outcomes and evidence practice committee. Pediatr Surg Int 2017;33:939 – 953 *(Abstract)*

9. Rothenberg SS, Kuenzler KA, Middlesworth W, et al. Thoracoscopic lobectomy in infants less than 10 kg with prenatally diagnosed cystic lung disease. J Laparoendosc Adv Surg Tech A 2011;21:181 – 184 *(Abstract)*

10. Makhijani AV, Wong FY. Conservative postnatal management of antenatally diagnosed congenital pulmonary airway malformations. J Paediatr Child Health 2018;54:267 – 271 *(Abstract)*

concept & design by mesch.ch

SUPPORTED BY

VIFOR PHARMA

CONTACT

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