

**RESEARCH ARTICLE**

**Acute life-threatening respiratory distress syndrome in an infant due to a bronchogenic cyst.**

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## **Abstract**

An 8-week-old infant was admitted to the hospital after an initially normal postpartum course with pronounced shortness of breath. Progressive hypoxia and a loss of consciousness occurred during the computer tomography examination, whereby the massively increased airway resistance hardly allowed ventilation. During an emergency thoracotomy, a bronchogenic cyst which had compressed the left main bronchus, was successfully extirpated.

Bronchogenic cysts in infants are relatively rare and they are often clinically silent, but with symptoms, like acute respiratory distress or hypoxia, the situation can be a big challenge for the surgeon and the anesthetist. Early diagnosis prevents life-threatening complications and early surgery is technically easier and reduces mortality and morbidity.

## **Introduction**

Bronchogenic cysts are rare malformations and arise from an abnormal “budding“ of the tracheobronchial tree during embryonic development from the so-called foregut in the 4th to 6th fetal week. The cysts are mostly located in the mediastinum and are usually not connected to the bronchial system [1-5]. However, they are lined with respiratory ciliated epithelium, whereby the cyst wall contains components of the airway [1-7]. Bronchogenic cysts account for only 5-10% of pediatric mediastinal masses and the incidence is equal between the sexes.

The symptoms are variable and age-dependent. Dyspnea is the main symptom in infancy, but occasionally there are “unclear“ infections in older children and adults. Mostly the cysts are asymptomatic and a co-incidence.

They are usually not recognizable in conventional x-rays and only become visible in a computed tomography. A bronchogenic cyst should be in the differential diagnosis of

pneumomediastinum or medial pneumothorax, even in premature infants who are on ventilators. Other differential diagnosis includes lung abscess, tuberculosis, vascular malformations, neoplasms and fungal disease as well as hydatidosis [7].

Due to possible complications (infection, bleeding, malignant degeneration), surgical removal is always indicated, even if the cysts are asymptomatic. Frequency of operative difficulty is higher in symptomatic cases than in asymptomatic cases. Early surgery is technically easier and helps to prevent complications that may occur later. Complete excision may prevent recurrence. Awareness may assist in the perioperative management of infants with bronchogenic cysts [5,7].

## **Case report**

In a female twin, prenatal sonographic diagnostics showed a mediastinal cyst, which was located below the aortic arch (Fig.1). However, the intrauterine lung development was normal and there were no signs of

compression of the intrathoracic neighbor structures. After birth, the clinical course was

normal as well as a postpartum chest x-ray (Fig.2).

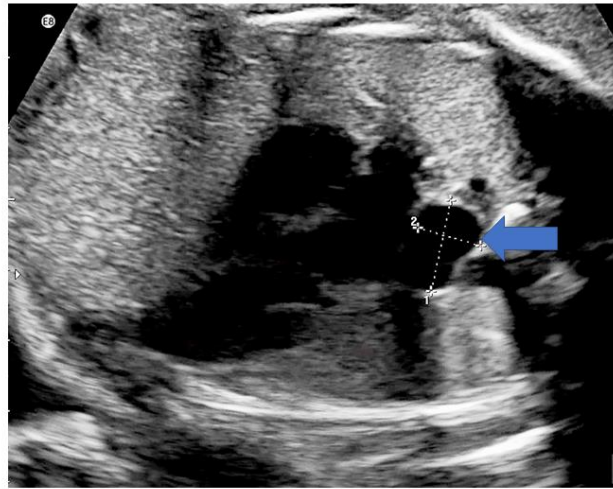


Fig.1

**Figure 1:** prenatal sonography

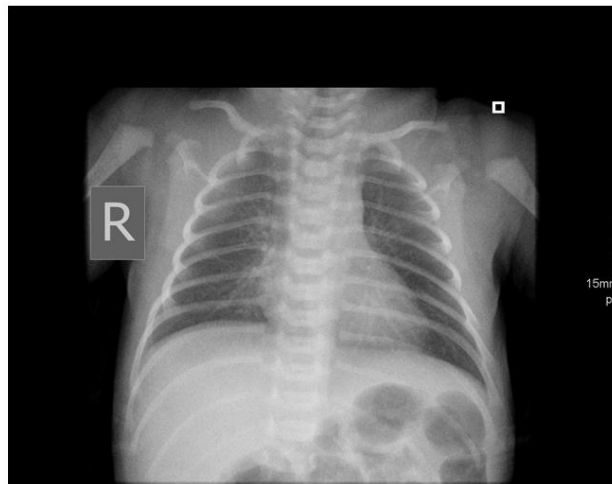


Fig.2

**Figure 2:** x-ray postpartum

After 8 weeks, the infant (body weight 3890 g) was admitted to the emergency room in a significantly reduced general condition with severe tachypnea and intercostal “retractions“. A refractory obstructive had lasted for about 4 weeks. The clinical examination showed a weakened breathing sound, an extended expirium on the left side and a “silent lung“ on the right. A CT-scan of the chest was performed immediately, based on the patient`s previous history and on the acute symptoms. During this period there was a dramatic deterioration with progressive hypoxia (cyanotic discoloration) and a loss of consciousness.

A massive increase in airway resistance prevented sufficient oxygenation. The CT-scan showed a cystic mass (Fig.3), which led to compression of the left main bronchus and by that to a massive overinflating of the left lung with additional mediastinal displacement (Fig.4). Due to the life-threatening situation, a left lateral thoracotomy was performed immediately. During the inspection, a cystic structure was found below the aortic arch (Fig.5), which “sat“ on the pulmonary artery and occluded the left main bronchus. The fluid-filled cyst could be completely extirpated after ligature of a small fistula to the esophagus (Fig.6).

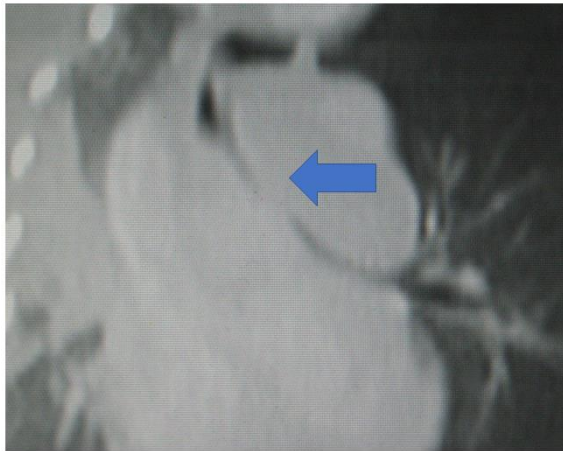


Fig.3

**Figure 3:** compression of the left main bronchus

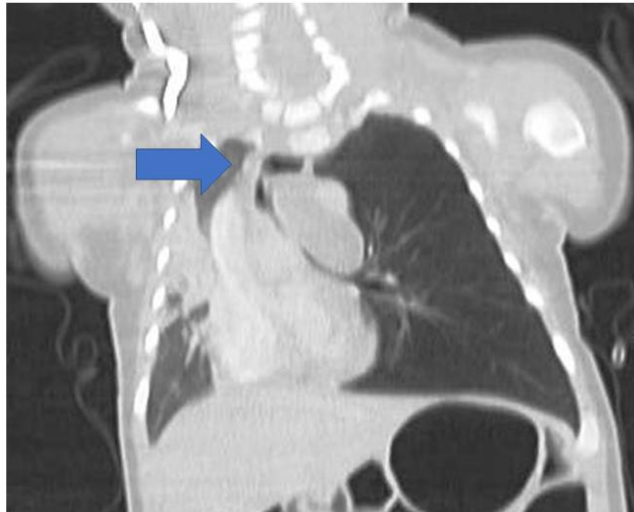


Fig.4

**Figure 4:** mediastinal displacement



Fig.5

**Figure 5:** cyst on the pulmonary artery

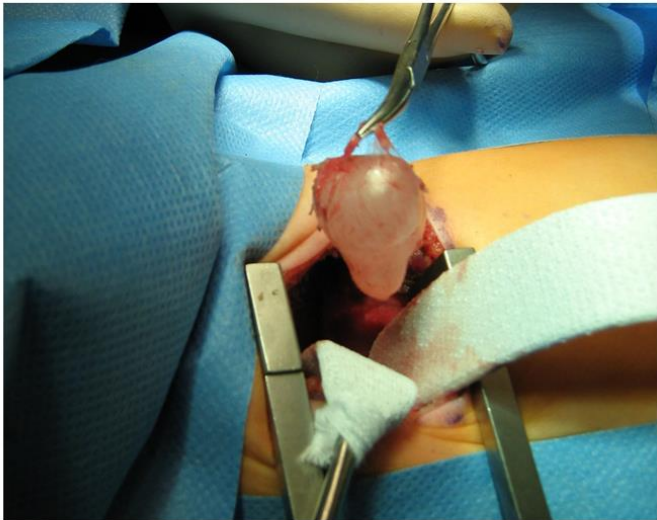


Fig.6

**Figure 6:** extirpated cyst

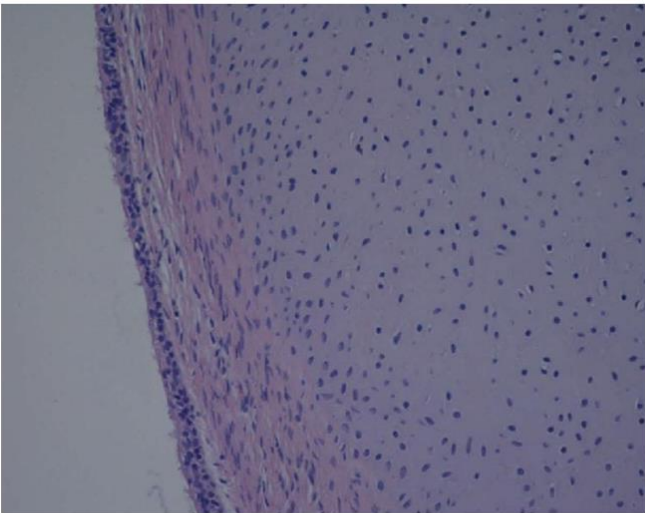


Fig.7

**Figure 7:** respiratory cylindrical epithelium

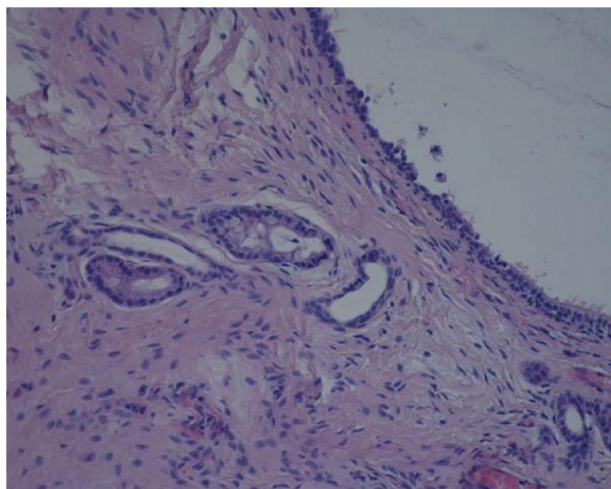


Fig.8

**Figure 8:** cyst wall, i.e. bronchial glands

Afterwards there an immediate improvement in ventilation with normal oxygen saturation could be reached.

The histological examination confirmed the suspicion of a benign bronchogenic cyst. On the second postoperative day, an extensive skin emphysema developed as well as a reduced breathing sound on the *right* side. The tension pneumothorax could be safely controlled by inserting a pleural drain (Charrière 12).

The further course of healing was uncomplicated. The child could be discharged home after 14 days. Several follow-up examinations showed a normal development without any consequential damage.

### **Discussion**

Bronchogenic cysts develop in addition to the lobar emphysema, the cystic adenomatoid

malformation or the intralobar sequester [2] through an abnormal “budding“ during the development of the tracheal tree from the “foregut“ in the 4th to 6th fetal week [1-5].

The location of the cysts depends on the period in which they occur. The mediastinal located cysts are the most common cysts (85% of the cases) and develop earlier during gestation [6,7]. They rarely communicate with the tracheobronchial system [7,8] and are lined with respiratory cylindrical epithelium (Fig.7). In the cystic wall (Fig.8) are parts of the airway, for example bronchial glands, hyaline cartilage plates and smooth muscle [1-5,7,9].

In our case, there was a fistula to the esophagus. This could be an explanation for the “reappearance“ of the cyst after disappearing postpartum without any clinical complaints. The cyst has probably filled up with fluid and air due to the fistula. In very

rare cases, the bronchial cysts can occur *ectopically* in the esophageal wall, in the pericardium or on the diaphragm [1,10,11]. Second most common the cysts can be found in the lung parenchyma (differential diagnosis: lung sequester) [9].

In infancy the cysts can cause a life-threatening respiratory distress syndrome due to compression of the “soft cartilage” [7,12,13,14]. Beyond this period, they usually cause no symptoms.

They are often radiological incidental findings or part of a complication assessment, for example in the case of sepsis due to an infected cyst or a bleeding due to a vascular erosion [5,8,9,12]. Although routine chest X-ray may be sufficient in some cases, it is diagnosed most often by sonography and computed tomography. In our case the mediastinal cyst already presented in the prenatal sonographic diagnostics. That is very rare and only possible with high experience in sonographic diagnostics. Occasionally, dysphagia can be a symptom due to the compression of the esophagus caused by the enlarging cyst [8]. Because of the possible complications [3,5,7,15] and also because of the malignant degeneracy risk (5,8,9), the surgical excision is indicated in any case [3,5,7-9,12] despite the absence of complaints. Morbidity and mortality increase significantly [5] with existing complications and if the excision is incomplete recurrence may occur. Early surgery is technically easier and helps to prevent complications that may occur later.

Transtracheal and percutaneous cyst aspirations have been proposed as alternatives to surgery, but are not widely accepted due to cyst recurrence [7].

A delayed diagnosis can lead to the development of life-threatening complications, like infections, bleeding-related ruptures, compression of esophagus and large vessels (e.g. pulmonary artery) and furthermore a possible malignant degeneration [1,5,8,9,15].

In our case the knowledge of the existence of the bronchogenic cyst through the prenatal sonographic diagnostic made it easier to interpret the symptoms of the infant when it was admitted to the emergency room.

### **Conclusion**

Bronchogenic cysts are rare malformations (prevalence 0,04%-0,06% in the newborn period), which can develop into a life-threatening clinical pattern in infancy [2,14,16]. Because of the unspecific symptoms (dyspnea, dysphagia etc.) and the unreliability of conventional chest x-rays [6,9,14], computer tomography should be performed in children with “unclear shortness” of breath, especially in infants in the 1st year of life [3,8,16,17]. In older children and adults, these cysts are usually discovered accidentally and primarily show no symptoms [12,16,18]. Nevertheless, the detection of this pathological structure is *always* an indication for early surgery [5-9,12] due to possibly life-threatening complications [2-4,7,12,18].



## References

1. Gould SJ, Hasleton PS. Congenital abnormalities. In: Hasleton PS, Spencer's Pathology of the Lung. McGraw Hill 1996:69-71.
2. Haller JA, Golladay ES, Pickard LR, Tepas JJ III, Shorter NA, Shermeta DW. Surgical management of lung bud anomalies: Lobar emphysema, bronchogenic cyst, cystic adenomatoid malformation and intralobar pulmonary sequestration. *Ann Thorac Surg.*1979;28(1):33-43.
3. Rodgers BM, Harman PK, Johnson AM. Bronchopulmonary foregut malformations. *Ann Surg* 1986;203(5):514-524.
4. Stewart B, Cochran A, Iglesia K, Speights VO, Ruff T. Unusual case of stridor and wheeze in an infant: tracheal bronchogenic cyst. *Pediatr Pulmolol.* 2002;34(4):320-323.
5. Aktogu S, Yuncu G, Halilcolar H, Ermete S, Budunelli T. Bronchogenic cysts: clinicopathological presentation and treatment. *Eur Respir J.* 1996;9(10):2017-2021.
6. Heithoff KB, Sane SM, Williams HJ, Jarvis CJ, Carter J, Kane P, Brennom W. Bronchopulmonary foregut malformations. *Am J Roentgenol.* 1976;126(1):46-55.
7. Sarper A, Ayten A, Golbasi I, Demircan A, Isin E. Bronchogenic cyst. *Tex Heart Inst J* 2003;30:105-108.
8. Tiwari MK, Yadav R, Mathur RM, Shrivastava CP. Mediastinal bronchogenic cyst presenting with dysphagia and back pain. *Lung India.* 2010;27(2):86-88.
9. Tsunozuka Y, Shimizu Y, Tanaka N, Ishikawa N, Ohama K. Progressive intraparenchymal bronchogenic cyst in a neonate. *Ann Thorac Cardiovasc Surg.* 2007;14:32-34.
10. Wenig BL, Abramson AL. Tracheal bronchogenic cyst: a new clinical entity? *Ann Otol Rhinol Laryngol.* 1987;96:58-60.
11. Gomes MN, Hufnagel CA: Intrapericardial bronchogenic cysts. *Am J Cardiol.* 1975;36(6):817-822.
12. Johnston SRD, Allison DJ, Smith P, Ind PW. Recurrent respiratory obstruction from a mediastinal bronchogenic cyst. *Thorax.*1992;47:660-662.
13. Ramenofsky ML, Leape LL, McCauley RG. Bronchogenic cyst. *J Pediatr Surg.* 1979;14:219-224.
14. Lazar RH, Younis RT, Bassila MN. Bronchogenic cysts: a cause of stridor in the neonate. *Am J Otolaryngol.* 1991;12(2):117-121.
15. Miralles LF, Gonzales-Martinez B, Luna MS, Valencia RA. Carcinoma arising in a calcified bronchogenic cyst. *Respiration.* 1981;42:135-137.
16. Zedan M, Elgamal MA, Zalata K, Nasef N, Fouda A. Progressive stridor: could it be a congenital cystic lung disease? *Acta Paediatr.* 2009;98(9):1533-1536.
17. Busino RS, Quraishi HA, Cohen IT. Stridor secondary to a bronchogenic cyst in a neonate. *Ear Nose Throat J.* 2011;90(11):E8-E10.
18. St. Georges R, Deslauriers J, Duranceau A, Vaillancourt R, Deschamps C, Beauchamp G, Page A, Brisson J. Clinical spectrum of

bronchogenic cysts of the mediastinum and lung in the adult. *Ann Thorac Surg.* 1991;52(1):6-13.