



## Perinatal management of enlarged bronchogenic cyst causing hydrops fetalis

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### ABSTRACT

Bronchogenic cysts are rare congenital anomalies that they are usually diagnosed prenatally during the routine second trimester scan. We present such a rare case of bronchogenic cyst in a fetus. Our initial scan demonstrated a big cystic mass, which enlarged progressively causing shifting of the mediastinum and compression of the fetal heart. Consequent hydrops fetalis was treated with thoracoamniotic shunt and the pregnancy continued with no further complications. Postnatal period, surgery and long term follow-up was uneventful.

### 1. Introduction

Bronchogenic cysts are unusual developmental malformations caused by aberrant tracheo-bronchial tree budding or branching. They are classified as foregut malformations and similar to intestinal duplications because they are presumed to be produced from an abnormal division of the embryonic foregut [1]. They typically presents as unilocular mucus field lesions usually arising in the mediastinum or within the pulmonary parenchyma and, less frequently, below or within the diaphragm [2]. Their prevalence is 1 in 50,000 births, but in most cases antenatal diagnosis can be achieved by 2D ultrasound [3]. They found not to be associated with chromosomal abnormalities and genetic syndromes. Fetal MRI has been demonstrated to be beneficial in the evaluation of fetal chest masses as an addition to sonography [4]. Furthermore serial scans are advisable in order to monitor the evolution of the lesion. Increase of its size may progressively cause

mediastinal shift and compression of the fetal heart. Thus these lesions are associated with fetal hydrops and severe respiratory failure at birth. Moreover, if there is evidence of bronchial obstruction delivery should be achieved by cesarean section with Ex Utero Intrapartum Treatment (EXIT) Procedure [4]. Long term related complications such as infection, rupture, bleeding and compression are common. Risk of malignant degeneration also exists in these lesions. Postnatal, usually requires surgery in the form of cystectomy or lobectomy or thoracotomy and excision of lesions [5]. Postnatally, usually requires surgery in the form of cystectomy or lobectomy by thoracotomy or video assisted thoracic surgery (VATS).

### 2. Case report

A 37-year-old G3P2 Caucasian woman was referred for ultrasound examination at 20 weeks of gestation because of cystic mass at the right

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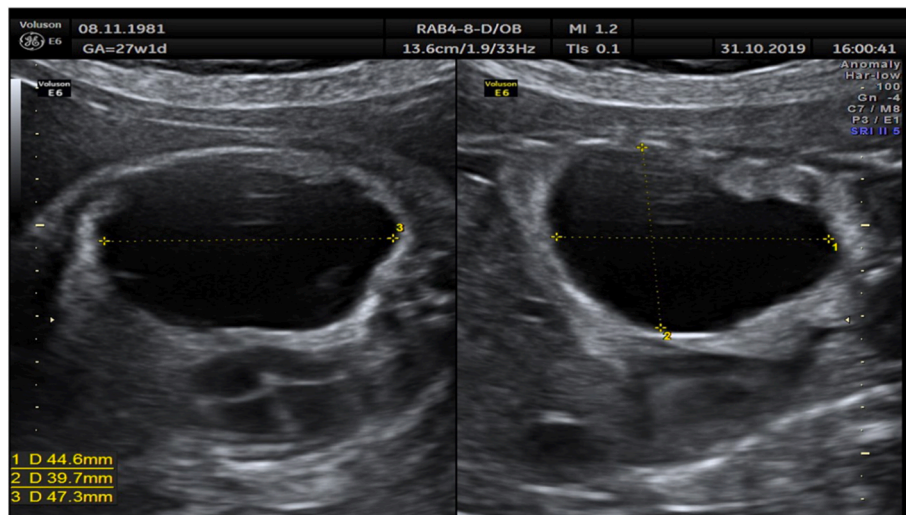
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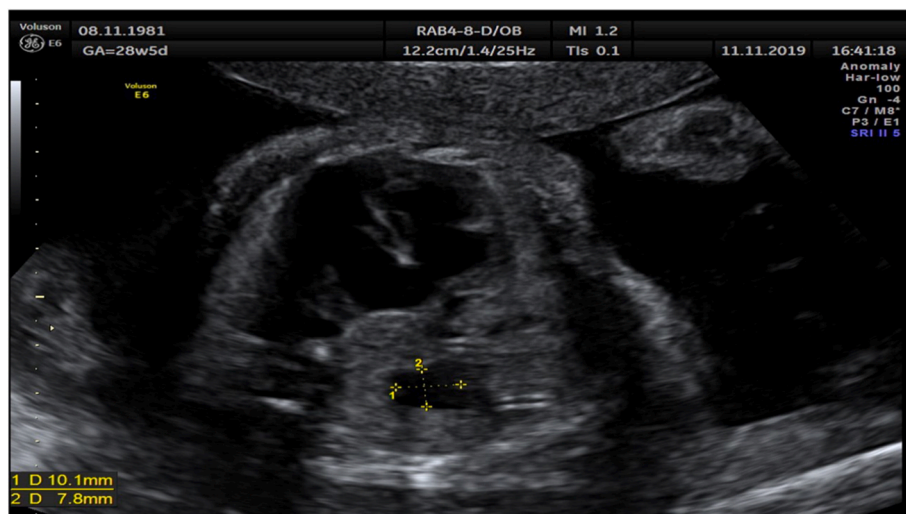
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**Fig. 1.** Ultrasound examination at 27w+1D, demonstrating enlargement of bronchogenic cyst (45 × 40 × 47mm), shift of the mediastinum and simultaneous compression of the heart.



**Fig. 2.** Ultrasound examination at 28w+5D, demonstrating shrink of the bronchogenic cyst (10 × 8 × 8mm) after shunt placement (yellow arrow). Cardiac function and position has been restored. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

lung. Previous obstetrical, medical and family histories were unremarkable. The first trimester scan (nuchal translucency assessment) was normal and gestational age was consistent with dates of last menstrual period. Examination was performed using a Voluson E6 scanner (GE Healthcare, Austria) and a 4–8 MHz abdominal convex probe. Our scan demonstrated a big cystic mass (13 × 15 × 12mm) and two smaller cysts locate peripherally, originating from the lower lobe of the right lung, without shifting of the mediastinum (Figs. 1 and 2). The initial polycystic appearance raised diagnostic dilemmas and the differential diagnosis included macrocystic type of congenital pulmonary airway malformation (CPAM). Further fetal anatomy appeared normal but the placenta was complete previa. The size of the mass and fetal wellbeing was monitored by serial ultrasound examinations. Later at 24 week the lesion had an appearance of a single cyst which was progressively enlarged (22x24 × 31mm) and the diagnosis of bronchogenic cyst was confirmed with fetal MRI. The mass gradually increased during the next couple of weeks, shifting the mediastinum and compressing the fetal heart. At 27 + 1 weeks (44 × 39 × 47mm), fetal hydrops was present, as manifested by ascites and the CVR of the entire mass was 1.55. An ultrasound-guided cyst-amniotic shunting operation was organized the

next day. A single shunt was placed on the larger cyst, successful drainage of cyst fluid was noted during the operation and the overall size of the lesion decreased markedly within 24hrs. Small sample of amniotic fluid was retrieved which revealed normal fetal karyotype. Serial ultrasound examinations were planned weekly, which evidenced the shunt tube to remain on site and cyst size to be stable. Furthermore, fetal growth was normal and the placenta appeared to be increta. At 36 + 1 weeks' gestation, an elective caesarean section was planned in order to facilitate neonatal resuscitation and to handle the previa increta placenta. A female neonate weighing 2410g with Apgar scores of 8 and 8 was delivered. An urgent hysterectomy took place because of post-partum hemorrhage. The neonate was transferred to the ICU and the shunt was removed. On the 2nd day of life, she developed respiratory distress and she was intubated. Chest x-rayed revealed right-side pneumothorax and a chest tube was placed. She was extubated on the third day of life but she required respiratory support by nCPAP with PEEP 3–4 mmHg. A CT scan was performed on the 7th day of life showed a 4.1 × 5.2 × 5.6cm cystic lesion and atelectasis of the upper and middle right lobes due to compression. The patient was submitted to right muscle sparing thoracotomy and the lesion was approached through the

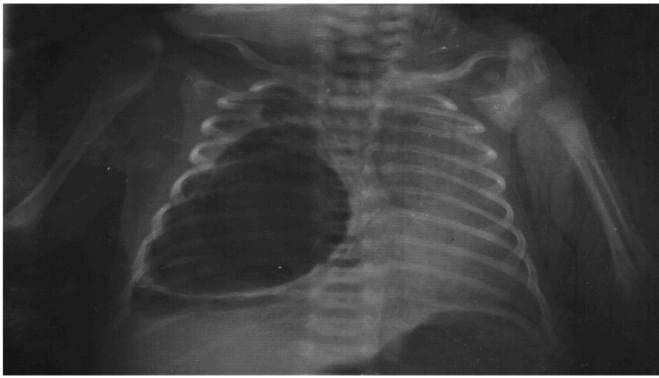


Fig. 3. Chest x-ray before surgery.

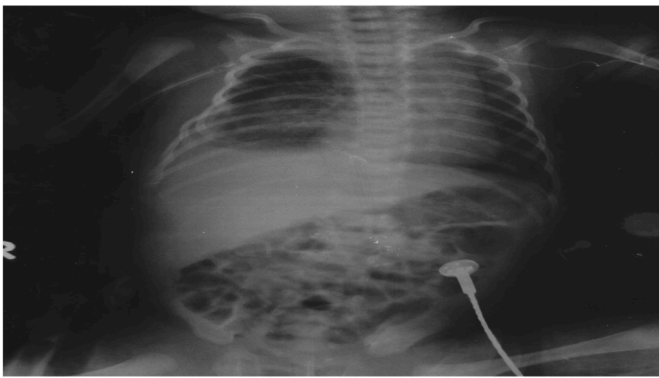


Fig. 4. Postoperative chest x-ray.

6th intercostal space. A multilocular cyst originating from the right lower lobe was bluntly dissected from the lung parenchyma. Hemostasis and lung sealing was achieved by bipolar coagulation. The cyst was removed in toto and the excision margins on the lung parenchyma were approximated by a continuous absorbable suture. Two chest tubes were placed in the thoracic cavity. The estimated blood loss was less than 10 cc. The patient was transferred to the NICU intubated. The postoperative course was uneventful. Histologic examination of the cyst showed a single multilocular cyst lined by respiratory epithelium and scattered gastric glands (Fig. 3). The patient was discharged on the 13th postoperative day (Figs. 4 and 5).

### 3. Discussion

The major factors affecting prognosis and survival are firstly the presence of hydrops, secondly the size of the lesion, thirdly timely diagnosis, and finally degree of hypoplasia of the remaining lung [6]. A paradox is observed for small solid pulmonary lesions where although small in size they tend to be associated with polyhydramnios, fetal anasarca, ascites and usually have a poor prognosis.

Ultrasound evaluation assists in the early prenatal diagnosis of these pulmonary malformations and allows timely intervention especially in the case of thoracic compression symptoms and hydrops [7]. An early minimal intervention for symptom relief is a series of needle aspirations or ultrasound-guided placement of thoracic-amniotic shunt. In the case where fetus thoracocentesis or shunt placement is not possible then fetus resection of the lesion is advocated. In a few cases of hydrops, open fetal surgery with excision of the lesion has been carried out with relatively good results. In the study by Peranteau WH et al. [9] improved survival benefit was observed with steroid administration to the mother if the fetus had hydrops and a CVR >1.6 [8].

Induction of labor aiming for vaginal delivery usually is planned at 38 weeks at a hospital with neonatal intensive care and pediatric surgery. However, if there is evidence of bronchial obstruction delivery should be achieved by cesarean section with Ex Utero Intrapartum Treatment (EXIT) Procedure [4].

Prognosis is good and the survival rate is >95% in cases with no hydrops [9]. Surgical treatment is controversial and it depends on the emergency, and development of mass effects, prenatal behavior, and postnatal presentation [10]. Improved postoperative surgical outcomes have been observed in children with small lesions undergoing elective surgery [11]. Clinical examination and careful follow up are essential. The usual clinical signs are: reduced breath sounds, tachypnoea, poor oxygen saturation, respiratory distress, chest in-drawing, and abnormal hemodynamic parameters [12,13]. In the case of unstable patients preoperative optimization and stabilization is necessary. The surgical management has to be delayed in the case of pneumonia or recurrent chest infection [12]. Hypotension with volatile agents is observed in patients with low cardiac reserve. In a meta-analysis of 9 studies pulmonary complications were revealed to be double the number of postoperative complications in symptomatic versus asymptomatic patients (risk ratio 2.8,  $P < 0.005$ ). Usually complications in emergency surgeries are higher (28%) than for elective surgeries in neonates and infants [14].

Most common early pulmonary complications are: desaturation atelectasis, air leak, hypoventilation, bleeding, pleural effusion, and infection. Successful management of these congenital pulmonary

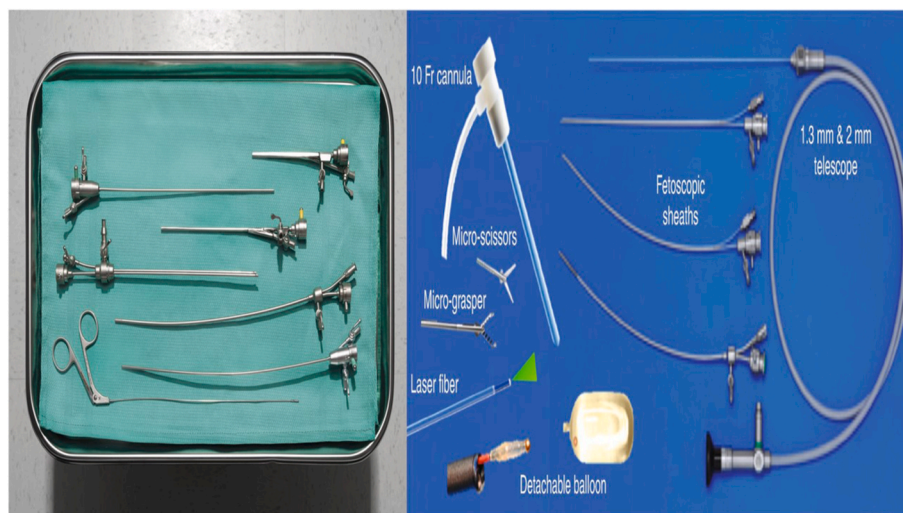


Fig. 5. Instruments during surgery.

malformations in babies needs multi-disciplinary care. Anesthetic management of such cases is challenging and lung protective ventilation strategies and thoracic epidural catheters are recommended. Early extubation is a strategy to avoid iatrogenic ventilator-induced bronchial stump dehiscence.

In conclusion, a follow-up by a multidisciplinary team with an ultrasound prenatal assessment every 2–3 weeks will be needed. If any sign of compression shows up, an intrauterine treatment should be discussed. The mode and timing of delivery depends on the size, location, and complications of the BCs. The patient should be transferred to a tertiary hospital with a neonatal surgical team to ensure maternal–neonatal safety. For the patient with asymptomatic BCs, surgical treatment may be performed at 3–18 months after birth.

#### Declaration of competing interest

All authors declare no conflict of interest.

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