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# Tracheal bronchogenic cyst causing cyanosis in a neonate

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ABSTRACT

Bronchogenic cysts are congenital malformations of the tracheobronchial tree that are mainly located in the mediastinum or the pulmonary parenchyma. They often occur in early childhood, compressing the airway or the esophagus or by superinfection.

Bronchogenic cysts located in the tracheal wall are relatively rare. They present with recurrent episodes of cyanosis, stridor and progressive respiratory distress. The diagnosis is suspected by clinical observation and endoscopic and radiologic evaluation and confirmed by anatomopathological findings. The treatment of choice is the surgical resection of the lesion.

We here report the case of a newborn presenting with cyanosis and stridor due to an intramural tracheal bronchogenic cyst that was successfully treated by tracheal resection with end-to-end anastomosis. We highlight the importance of considering this rare entity in the differential diagnosis of cyanotic spells in this age group.



## 1. Introduction

The bronchogenic cyst (BC) is a remnant of the primitive foregut due to a malformation in the embryonic development of the tracheobronchial tree between the 26th and 40th days of gestation [1–3]. It is more common in male infants (2–4:1) and is often diagnosed in childhood [2,4]. Very few cases are documented in the neonatal period [5]. Most of them are located in the mediastinum (86%), mainly in the right side [1,2].

In relation to the trachea, a BC can be classified into paratracheal (the most common one), intraluminal or intramural [6–8]. The intramural BC is extremely rare [6–9]. It presents with biphasic stridor and recurrent cyanotic episodes and severe respiratory distress due to intermittent occlusion of the tracheal lumen caused by the intramural mass [8].

The presumptive diagnosis is made with a chest x-ray that may show an intrathoracic cystic image and with an esophagogram that allows to visualize the esophageal compressions. The computed tomography scan and magnetic resonance imaging help to better define the lesion location. The tracheobronchoscopy is highly effective in diagnosing an intramural cyst.

The treatment of choice for BC is the complete surgical excision of the lesion in order to prevent its recurrence. In the case of the intramural tracheal cyst, resection of the damaged tracheal wall with end-to-end anastomosis for the complete removal of the cyst is required [7].

## 2. Case report

We here describe the case of a female newborn that was referred to the Respiratory Endoscopy Service due to recurrent episodes of cyanosis and severe respiratory distress since she was 10 days old. Her symptoms worsened with feeding and crying.

The patient was born at full-term by normal delivery. No perinatal pathology history was reported.

The physical examination revealed biphasic stridor with expiratory phase predominance. The cardiologic and pulmonary evaluation was normal. The chest x-ray showed no alterations.

An esophagogram was requested, revealing extrinsic esophageal compression (Fig. 1A). Endoscopic evaluation of the airway was performed under general anesthesia, showing a non-pulsatile mass that was compressing the posterior membranous wall of the

*Abbreviations:* BC, bronchogenic cyst.

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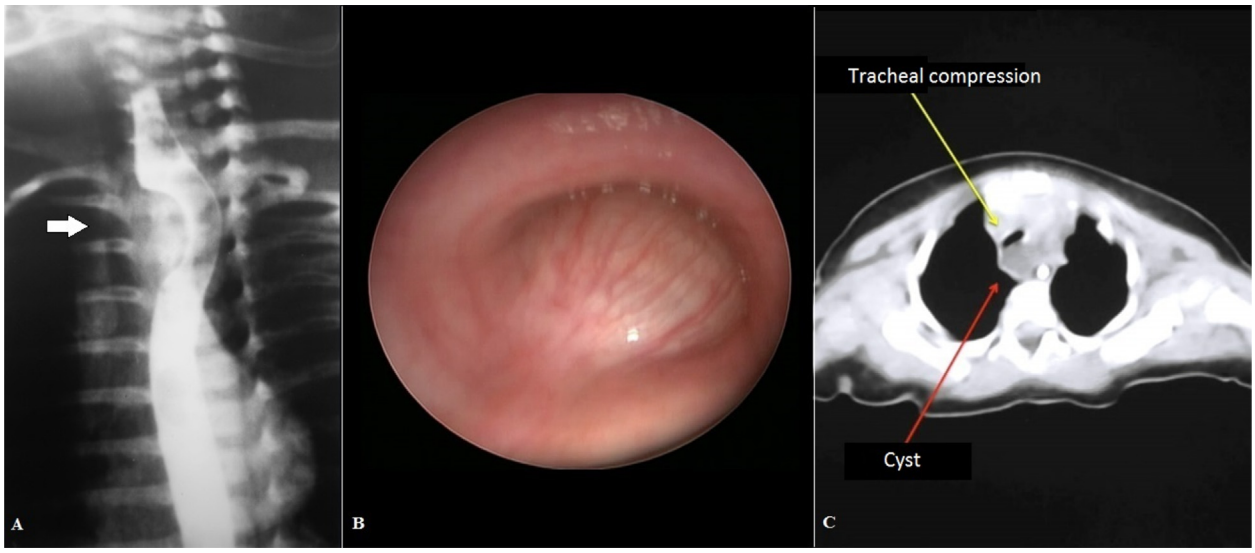


**Fig. 1.** A. Esophagram. Esopahageal compression due to the cyst (arrow). B. Endoscopic image of the intramural tracheal cyst. C. Computed tomography, axial section. Tracheal compression due to the mediastinal cystic mass.

trachea and occluding more than 90% of the tracheal lumen (Fig. 1B). Computed tomography scan of the chest revealed a mediastinal cystic mass, to the right side of the midline, which compressed the trachea (Fig. 1C).

These findings suggested the probable diagnosis of a BC originating outside the tracheal wall and extrinsically compressing the tracheal lumen.

The cyst was excised by video-assisted thoracoscopy, being approached through the right side. When the cyst was dissected, a common wall between the cyst and the trachea was observed. Partial cyst excision was performed, leaving the mucous membrane shared with the posterior membranous wall of the trachea of approximately 1 × 1 cm.

The anatomopathologic analysis revealed a cystic lesion compatible with BC.

The patient remained asymptomatic for 30 days. One month after surgery the same symptoms reappeared. A laryngotracheoscopy was performed, confirming the lesion recurrence.

A new surgery was performed through a transverse cervical incision and manubriotomy. The affected tracheal segment was fully resected and end-to-end anastomosis was performed to restore the trachea (Fig. 2). Six tracheal rings were resected and anastomosis was performed at a 3-ring distance from the tracheal carina. The patient was extubated on postoperative day 3.

The histopathological analysis confirmed the diagnosis of intramural tracheal BC (Fig. 3).

The patient's recovery was uneventful. She was discharged 7 days after surgery. The endoscopic control performed 6 months after surgery revealed a normal tracheal caliber with no evidence of the lesion (Fig. 2D). At 2 years follow-up, the girl is asymptomatic and with no BC recurrence.

## 3. Discussion

The incidence of BC has been estimated to be 1 in every 42,000–68,000 newborns [3]. Most of these cysts are intrathoracic, located in the mediastinum or in the pulmonary parenchyma. BCs account for 10% of mediastinal masses in children [2]. They are often adjacent to the distal third of the trachea and are classified into paratracheal, carinal, hilar and paraesophageal cysts [2]. Rare locations for these types of cysts are the abdominal cavity, the neck, the subcutaneous tissue and the pericardium [3]. Cysts may be single, multiple or multiloculated and they often do not communicate with the airway [10].

Few cases of tracheal intraluminal or intramural BCs have been documented in children [2,6–9]. When the cyst is located between the mucosa and submucosa layers of the trachea, it is defined as intraluminal cyst, and when it is found between the cartilage and the adventitia it is an intramural cyst [7]. This distinction cannot be made before surgery.

The BC symptoms depend on their location and size and may be due to mass effect, compression of adjacent structures or a secondary infection [11]. Most cases occur in childhood, although the symptoms may develop at any age, being asymptomatic in up to 19% of cases. As the child grows and the airways stabilize, the BC becomes less compressive and is usually asymptomatic until it becomes infected [2].

Tracheal cysts cause airway obstruction symptoms; stridor, cough, dyspnea, substernal retractions and recurrent cyanotic crises being the most common ones [2,8]. Other symptoms may include thoracic pain, hemoptysis, recurrent infections and dysphagia (the latter due to esophageal compression).

Intramural cysts, when protruding intermittently into the tracheal lumen, cause recurrent events of severe respiratory obstruction and cyanosis [8]. Feeding and crying make symptoms worse.

The preoperative diagnosis of the tracheal BC is difficult [7]. This possibility should be considered when the cyst is in contact with the trachea. In documented cases, the diagnosis of an intramural lesion could not be made before surgery [7–9].

An x-ray of the chest is effective but not always can the cyst be visualized [8]. An esophagogram allows to observe the compressions of the esophagus, locating the cyst in the different sections of the mediastinum. The computed tomography scan and the magnetic resonance may show cysts that cannot be observed in a simple X-ray image, better delimit the lesions and show the compromise of adjacent structures.

Direct visualization using trachebronchoscopy is very effective for diagnosis, mainly for intramural and intraluminal cysts [2]. This technique helps differentiate other causes of endoluminal and