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CASE REPORT

Congenital bronchial stenosis: a case report of an unusual diagnosis and its management

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ABSTRACT

Congenital bronchial stenosis is a rare bronchopulmonary anomaly characterized by significantly narrowing or blindly ending of a segmental or lobar bronchus, typically limited to a few rings, caused by developmental anomalies in the cartilaginous exoskeleton.

We describe a case of a six-month-old male infant with intermittent wheezing present since birth, with no signs of respiratory distress, which seemed to not respond to bronchodilators and to appear more evident with crying and during common colds.

He didn't present respiratory distress, but an expiratory wheezing was heard. A tidal flow-volume loop was recorded and appeared slightly deflected during the expiratory phase, with a normal tidal volume. A bronchoscopy detected a stenosis of the left main bronchus, confirmed by the chest CT. The patient underwent surgical treatment, consisting of resection of the stenotic tract and tracheobronchial anastomosis with a good result.

Congenital bronchial stenosis should be considered in newborns and infants with persistent wheezing that doesn't respond to bronchodilators; a tidal flow-volume loop could suggest the diagnosis, which should be confirmed with chest CT and bronchoscopy.

IMPACT STATEMENT

A case report of congenital bronchial stenosis, with the aim to review its diagnosis and management.

KEYWORDS

Congenital bronchial stenosis; Tidal breathing flow volume loop; Bronchoplasty; Infant; Case report

INTRODUCTION

The occurrence of wheezing in infants can be referred to a broad list of differential diagnoses, that include airways or vascular anomalies, cardiac, metabolic, and infectious diseases. A rare cause of persistent wheezing in infants could be congenital bronchial stenosis (CBS). CBS is a bronchial anomaly characterized by a significant narrowing of a segmental or lobar bronchus, usually limited to a few cartilaginous rings, caused by developmental anomalies in the cartilaginous exoskeleton (1) or by compressive vascular anomaly, cardiac anomaly, or congenital pulmonary cyst (2).

This report describes the case of a six-month-old boy who presented intermittent wheezing, and who was found to have congenital bronchial stenosis which was surgically corrected.

CASE REPORT

A six-month-old infant was referred to the pediatric respiratory outpatient clinic with a history of expiratory wheezing since birth, with no signs of respiratory distress, which seemed to not respond to bronchodilators and to appear more evident with crying and during common colds.

He was delivered at term; he had clinodactyly of the second finger of the left hand, waiting for surgical correction. His developmental history and his growth were on average; he was bottle-fed with no signs suggestive of aspiration. His family history was unremarkable.

At the examination, he was afebrile, his height was 65 cm (-1 SD), and his weight was 7.4 kg (-1 SD).

Oxygen saturation was 97% in room air. No signs of respiratory distress were present. At chest auscultation, bilateral wheezing was heard. The remaining physical examination was unremarkable, apart from mild pectus excavatum. A tidal flow-volume loop while asleep was recorded (**Figure 1A**), which appeared slightly deflected during the expiratory phase with a normal tidal volume (8 mL/kg).

The patient was electively admitted to the ward. A chest x-ray resulted normal; flexible bronchoscopy demonstrated severe stenosis of the left main bronchus, which was later confirmed by CT scan, showing a stenosis of 1.4 mm for a length of 2.2 mm with initial hyperinflation of the left lung. To avoid associated malformations, we performed the echocardiography and the ultrasound of the abdomen, which were normal. At 8 months of age, the patient was admitted to another tertiary hospital. At admission, sleep studies were performed showing no desaturations or apnea. After a multidisciplinary discussion, a CT scan with contrast medium under general anesthesia was repeated allowing to exclude any vascular malformation that could have determined the stenosis; the 3D volume rendering reconstruction and the virtual bronchoscopy reconstruction (**Figure 2**) showed an expiratory collapse of the bronchial lumen, suggestive of associated bronchomalacia. Rigid bronchoscopy was repeated (**Figure 3A**) and a 2.8 mm flexible endoscope was able to overcome the stenosis and explore the bronchial system beyond the stenosis, which appeared normal.

The case was discussed by a multidisciplinary board with the decision to proceed surgically.

After midline sternotomy, the patient was placed on normothermic cardiopulmonary bypass with the beating heart by right atrial and ascending aortic cannulation. The carina and left mainstem bronchus were exposed by dissecting the transverse sinus of the pericardium and removing the subcarinal lymph nodes. Hence a left bronchial transection distal to the site of the stenosis and an oblique section at the origin of the left main bronchus extended to the carina and distal tracheal portion were made. Finally, a terminal-lateral tracheo-bronchial anastomosis was performed using a running resorbable 6-0 polydioxanone (PDS) suture. An intraoperative fiberoptic control confirmed a satisfactory anastomosis with a wide and patent bronchus (**Figure 3B**), and the cardiac procedure of disconnection from bypass was undertaken. Mediastinal and right pleural drainages were inserted. The patient was extubated after the procedure and remained stable in high-flow-nasal-cannula. On postoperative day 2, he was started on empirical antibiotic therapy with ceftazidime for a right lower lobe pneumonia with effusion. He was

discharged on postoperative day 8 with the resolution of the effusion. Audiological assessment and eye examination resulted normal.

Two months after surgery, he presented intermittent noisy breathing and inconstant inspiratory stridor, with no signs of respiratory distress or infections. A bronchoscopy under general anesthesia was then performed, revealing mild laryngomalacia and raising suspicion of partial left vocal cord paralysis.

Moreover, a tidal flow-volume-loop was recorded, showing no alterations with a normal tidal volume (**Figure 1B**).

The patient is undergoing respiratory follow-up with tidal flow-volume loops, and a chest CT will be repeated one year after surgery.

DISCUSSION

Bronchial stenosis could be acquired or congenital.

Acquired stenosis in children is usually caused by infection, traumatic granulomas due to chronic intubation, lymphnode enlargement, atelectasis (3), tracheobronchomalacia and excessive dynamic airway collapse (4). In particular, in a retrospective study (5) the most common causes of endobronchial obstructions detected in 256 paediatric patients were aspirated foreign bodies (35.9%), endobronchial tuberculosis (31.6%), mucous plugs (16.7%) and granulation scars (6%) and other rare pathologies found were hydatid cysts, hemangiomas, tumors, submucosal nodules, and polyps.

CBS is rare, it can be occasionally diagnosed prenatally (6), but usually it becomes more apparent in the neonatal period (7).

CBS usually involves the left main bronchus (2). Other structural anomalies may also be found including subglottic stenosis, tracheoesophageal fistula, and esophageal atresia (1).

Symptoms can vary, based on the location of the stenosis and the residual lumen (2). In fact, CBS determines a “ball-valve” mechanism with ipsilateral air trapping and lung distention (7) leading to diminished unilateral breath sounds at the physical examination; inspiratory and expiratory stridor can also be present, due to fixed airway obstruction (7) as observed in our patient, which not respond to beta2agonists (7). Other clinical signs include “barking” or brassy cough, “washing machine” airway sounds, cyanosis, and breathing spells (2). In some cases, especially when tracheal stenosis is associated,

CBS could present at birth with significant respiratory distress, needing non-invasive or invasive respiratory support (7). Sometimes, patients could present only subtle symptoms of airflow limitation as wheezing and increased work of breathing, especially near the end of the first year of life when physical activity increases, until late childhood or early adolescence, when they tend to develop exercise-associated respiratory difficulties and, consequently, a CBS could be found as an incidental finding during the diagnostic work-up (8).

There is no consensus on the diagnostic approach. Since the same symptoms could be a sign of congenital tracheal stenosis (CTS), it is more cautious to perform a rigid bronchoscopy under general anesthesia (8) with extreme care, since a mucosal lesion could precipitate edema, leading to a critical obstruction. Even though the stenosis could be detected by a CT scan, the diagnosis is made usually by bronchoscopy, because it provides dynamic information about malacia (1), along with the location, the extension, and the severity (2) of the stenosis.

Other approaches include the virtual endoscopy, which evaluates distal airway anatomy using high-resolution multirow detector computed tomography (CT) scanning to obtain high-resolution 3-dimensional endoluminal images to the level of the segmental bronchi (5) distal to the stenosis which are impassable for flexible bronchoscopy. Virtual endoscopy has the advantage that it can be performed without general anesthesia, and there is no direct invasion of the airways (9). Moreover, it could be used as a complementary technique for the planning of the flexible bronchoscopy, in patients who could not tolerate the bronchoscopy, and for the follow-up of the stenosis (10).

In a retrospective study (11) patients with CBS were found to have similar high comorbidity of cardiovascular anomalies as children with CTS (55.6%), such as pulmonary artery sling, ventricular septum defect, atrial septum defect, patent ductus arteriosus. In view of the high proportion of patients with other congenital vascular and cardiac anomalies, echocardiography should be performed to rule out any cardiac defect, along with a contrast CT scan with 3D reconstruction, that will better depict the anatomy of the airways, the parenchyma, and the vessels (2), allowing to characterize any vascular rings as in the case presented. It should be noted that CT frequently underestimates the degree and length of airway narrowing (8). Magnetic resonance imaging enables the assessment of the vascular structures

(2) and it has been found to be equally sensitive to CT to detect congenital cardiovascular malformations associated with airway pathologies without any radiation exposure (12).

Furthermore, 3-dimensional printing in case of complex airway anomalies could help in the planning of the surgical approach and educate care providers and family members (13).

Additionally, our patient had pectus excavatum. The association between chest deformities and bronchial stenosis is described in the literature (14–16). It has been suggested that the costosternal retraction can be the consequence of the increased intrathoracic pressure during expiration to overcome the airway obstruction due to the CBS (14) and usually the deeper impression corresponds to the side of the CBS. Accordingly to this hypothesis, the pectus excavatum of the patient nearly resolved at the 6-month after-surgery follow-up evaluation. However, it is questionable whether pectus excavatum is solely due to CBS. CBS, in fact, can be also caused by an abnormal thoracic configuration (17) due to complex mechanisms of vascular compression (15). Based on the clinical presentation, it could be useful to perform a full genotyping as suggested by some Authors (1), along with further diagnostic work-up, especially in the presence of features suggesting skeletal dysplasia (18).

CBS should be managed at major tertiary centres with experience in complex airway malformation, involving a multidisciplinary team including cardiothoracic surgeons, otolaryngologists, cardiologists, pulmonologists, and anesthesiologists (8).

The surgical approaches vary in the literature and are better characterized for CTS rather than CBS. However, they should be tailored to the patient (19) and the presence of other anatomical abnormalities. Surgery includes resection and reconstruction, and bronchoplastic techniques (20,21); nonetheless, both are complex, especially in infants because of the smaller size of the airways (2). The post-operative bronchoscopy could show granulation tissue, mild restenosis, or formal scarring that could be treated with balloon dilatation (1); in these cases, it would be desirable to avoid laser techniques that could induce thermal damage to the tissue with the consequent risk of restenosis (1). Another postoperative complication is bronchial malacia, which could be addressed with a stent placement (22).

Ultimately, given the lack of international consensus on CBS diagnosis and management, raising awareness of this condition is important to allow a prompt diagnosis.

KEY TAKEAWAYS:

- CBS is a rare malformation usually diagnosed during the neonatal period, but even in late childhood.
- Presentation varies from unexplained neonatal respiratory distress to a late presentation with inspiratory-expiratory stridor, wheezing, increased work of breathing, noisy breathing, cyanosis, breath-spells, and apnea.
- Usual findings at chest examination are diminished unilateral breath sounds, wheezing or stridor.
- Chest X-rays usually show ipsilateral lung hyperinflation or could result in normal.
- CBS is usually diagnosed with bronchoscopy, which should be performed carefully to avoid any tissue damage.
- Echocardiography and contrast CT are needed to evaluate the presence of congenital cardiovascular malformations such as vascular rings that are usually associated with CBS; 3D CT reconstruction and virtual bronchoscopy are helpful and complementary to bronchoscopy to assess distal airways and for surgical planning.
- Generally, CBS is managed surgically; the approach should be tailored to the patient and discussed within a multidisciplinary team.
- Close follow-up with monitoring for postoperative complications, and parental education, can impact the outcome of the patient.

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Compliance with Ethical Standards

Conflict of interests

The authors have no conflicts of interest to declare that are relevant to the content of this article.

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Author contributions

LV: writing - original draft; GF, LT, AC1, FP, VB, AC2: writing, review, and editing; AC2: supervision; RC, GP: conceptualization, and supervision.

Informed consent

The caregivers gave their informed consent to the submission of this manuscript.

ABBREVIATIONS:

CBS congenital bronchial stenosis

CTS congenital tracheal stenosis

CT computed tomography

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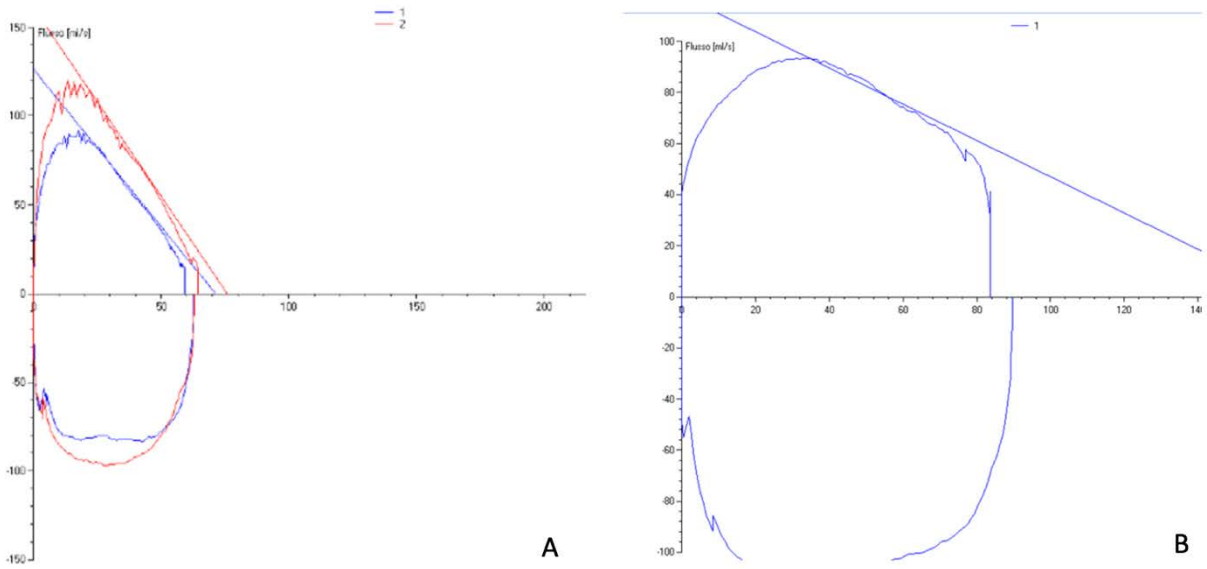

Figure 1. A. Tidal flow-volume loop recorded while asleep with slight deflection during the expiratory phase with a normal tidal volume (8 mL/kg); the loop recorded after the administration of albuterol didn't show any significant change. **Figure 1. B.** Tidal flow-volume loop recorded while asleep after surgery showed a normal morphology with a normal tidal volume (9.8 mL/kg).

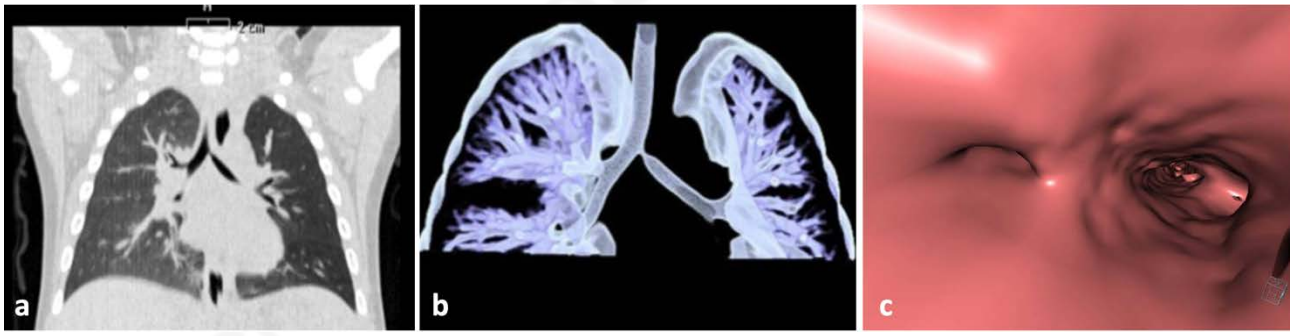

Figure 2. Coronal non-enhanced CT image with lung window (panel a) showing stenosis of the proximal left main bronchus and hyperinflation of the right lung; 3D volume rendering reconstruction of the tracheobronchial tree (panel b); virtual bronchoscopy reconstruction (panel c) demonstrating marked narrowing of the left main bronchus lumen.

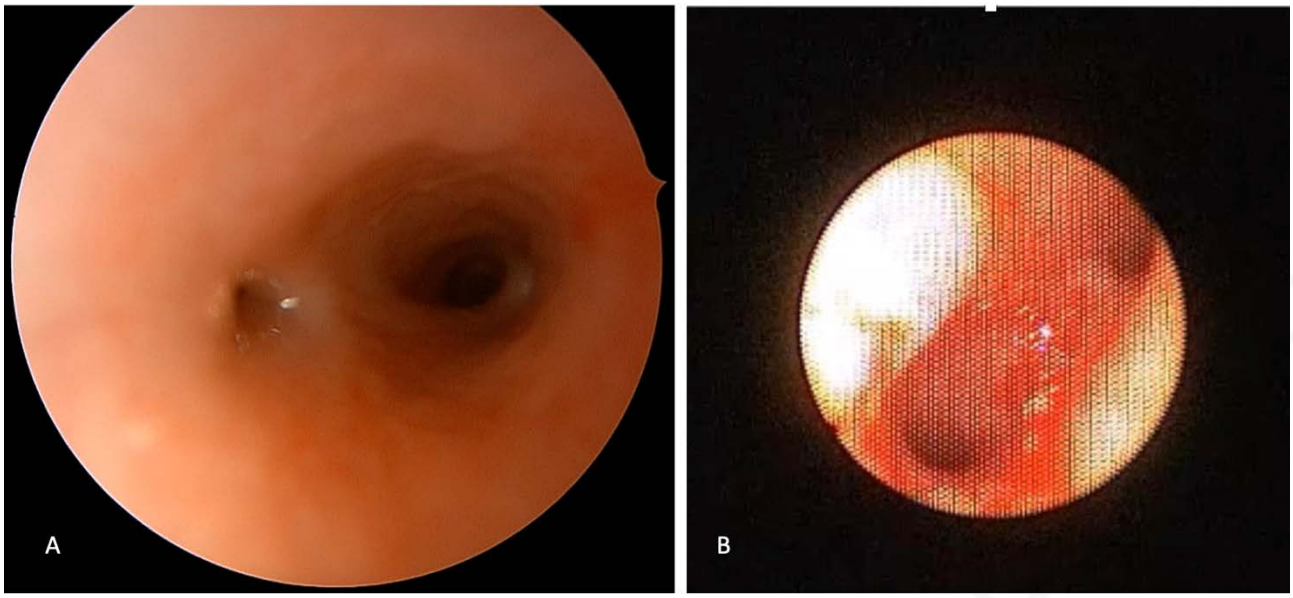

Figure 3. A. Rigid bronchoscopy showing a significant stenosis of the left main bronchus. **Figure 3. B.** Intraoperative fiberoptic control showing a wide and patent left main bronchus.