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REVIEW

Diagnosis of congenital airway abnormalities in children

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ABSTRACT

Congenital lung lesions are relatively rare but are an important consideration in any child with lung disease especially if the disease is recurrent or not resolving. Some of these lesions cause severe symptoms shortly after birth while others may not present with symptoms for years. Antenatal ultrasound has made it possible to diagnose some of these lung lesions early, which were not possible to diagnose in the past.

Most congenital airway lesions are not diagnosed antenatally especially if they are not associated with cardiac lesions. Not every mother has an antenatal ultrasound in the developing world, which leads to late diagnosis of many congenital lesions with significant consequences.

There is a large number of different types of congenital airway abnormalities, but can be divided in structural abnormal airway, external compression and airway fistula. Long segment congenital tracheal stenosis presents early especially if associated with left pulmonary sling.

It is important that airway lesions are diagnosed early to determine the correct diagnosis and management. Plain chest X-ray may be very indicative of airway pathology, and these should be evaluated to determine if the airways are visible, to determine narrowing, displacement or abnormal branching.

The diagnosis and management of children with airway pathology needs a team approach with skills needed in airway management, imaging, cardiology, bronchoscopy and airway surgery.

IMPACT STATEMENT: Congenital lesions are rare, but if undiagnosed, they may have significant consequences. These lesions are associated with abnormalities including congenital heart disease, which will complicate their diagnosis and management, particularly congenital airway malformations. Bronchoscopy plays an important role in diagnosis, intra-operative and post-operative management. Interventional bronchoscopy is useful in treating congenital airway abnormalities.

KEY WORDS
Congenital airway abnormalities; bronchoscopy; tracheal stenosis; chest CT-scan; contrast bronchography.

INTRODUCTION

Congenital lung lesions are relatively rare but are an important consideration in any child with lung disease especially if the disease is recurrent or not resolving. Some of these lesions cause severe symptoms shortly after birth while others may not present with symptoms for years. Antenatal ultrasound has made it possible to

diagnose some of these lung lesions early, which were not possible to diagnose in the past. However, many of these lesions will have disappeared at term.

Congenital lesions involve the airways, parenchyma, arterial tree, chest wall and the diaphragm. Sometimes these lesions are not diagnosed in childhood and present in adults as episodes of recurrent pneumonia or malignancies (1-3). In the developing world there is limited access to antenatal ultrasounds and some of these congenital lesions will only be diagnosed much later or once they have become complicated and infected (4).

WHEN TO CONSIDER A CONGENITAL LUNG LESION?

Neonatal period

A neonate with a significant lesion will present with respiratory distress. The lesions which present early are congenital cystic adenomatoid malformations (CCAM) and diaphragmatic hernias because they are space occupying (5). The rest of the ipsilateral lung may be hypoplastic.

It may be very difficult to distinguish these two lesions because both are cystic on the chest X-ray. Children with mayor airway pathology may present early. Factors that determine the time of presentation include severity of narrowing, combination of lesions and the presence of congenital heart disease. Difficult intubation and feeding issues may indicate airway pathology.

Infant and older child

The infant and child with a congenital lesion will present with recurrent pneumonia or non-resolving pneumonia that is localized to the same region of the lung (6).

During acute infection it will be difficult to identify the lesion radiographically. The lesions that present with recurrent infection are CCAM, bronchogenic cysts, duplication cysts and sequestrations.

WHY ARE CONGENITAL LUNG LESIONS IMPORTANT?

Some of these lesions have a very high mortality if not diagnosed early and treated correctly (diaphragmatic hernia, tracheal stenosis). These children are susceptible to recurrent infections that may lead to bronchiectasis. Some of these lesions are associated with other congenital abnormalities and congenital cardiac lesions.

Persistent/recurrent wheezing

Children who present with persistent wheeze, not responding to asthma treatment have a high incidence of congenital abnormalities. Congenital abnormalities have been reported in up to 45% of cases in bronchoscopy studies (7-10).

The airway narrowing is due to abnormalities of the wall of the airway or external compression of the airway. Vascular compression of the airways was observed in 13%-26% of children who underwent bronchoscopy for persistent wheezing, stridor, and apnea (11). Double aortic arch is the most common vascular abnormality, causing both tracheal and esophageal compression.

DIAGNOSIS OF AIRWAY PATHOLOGY

- Chest X-ray.
- Chest Computed Tomography (CT) scan.
- Bronchoscopy.
- Bronchograms.
- Echocardiography (ECHO).
- Contrast swallow study.
- Magnetic Resonance Images (MRI).

How does the chest X-ray help?

It is important to evaluate previous chest X-rays as that may indicate if the lesion is congenital or acquired. If the X-ray was never normal it makes the diagnosis of a congenital lesion more likely. CCAM may rarely present bilaterally, as most congenital lung lesions are unilateral. Combinations of congenital lesions do occur but are more commonly seen on the same unilateral side. It is important to determine if lesions are cystic or solid from the X-ray presentation. Solid lesions may cause more airway compression *versus* cystic lesions which can cause mediastinal shift.

Make sure that the trachea and bronchi are clearly visible on the X-ray especially in children who present with stridor and wheeze.

Some of these congenital lesions are recognizable on the chest X-ray as they are preferentially located in specific areas of the lung, *e.g*. congenital lobar emphysema (CLE) in the left upper lobe.

Clues to the presence of congenital lung anomalies include: (1) thoracic asymmetry; (2) focal mass/consolidation; (3) focal hyperlucency; (4) airway abnor-

malities; (5) vascular abnormalities; and (6) other lesions, including vertebral anomalies, gastrointestinal anomalies, and cardiac anomalies (12).

Chest X-ray: looking at the airways (Figure 1A)

The following may be indicative of airway pathology (13):

- Lack of visibility.
- Narrowing.
- Abrupt discontinuity.
- Displacement of bowing.
- Abnormal branching.
- Focal or unilateral overinflation.

CT/MRI (Figure 1B)

Both CT and MRI are used for detailed visualization of airways as well as vessels when thin slices are acquired, and an intravenous contrast agent is used. Three-dimensional (3-D) and minimum-intensity projection reconstructions can be obtained with both modalities (14). Inspiration/expiration or dynamic cine can be used for airway evaluation and to determine if malacia is present. It is helpful to evaluate the airway without an endotracheal tube (ETT) in place as the ETT can obscure the airway lesions and prevent accurate assessment of airway narrowing (13). If an ETT is in place, it should be positioned as high as possible above any stenosis (**Figure 1B**).

Contrast bronchography (Figure 1C)

Bronchography enables the assessment of the trachea and the more distal bronchi. Unlike bronchoscopy, and since it is performed in real time with the infant spontaneously breathing, the airways are assessed throughout the respiratory cycle. It is simple to perform in infants who are already intubated and is also useful in diagnosing fixed airway stenosis. Contrast bronchography is used in children with prolonged airway compression who develops secondary malacia (14).

Bronchoscopy is still necessary to identify complete tracheal rings. Examination of the airway under direct vision by the surgeon at operation has been more useful than imaging in deciding which surgical approach is most suitable for several patients.

Bronchography can be performed by injecting isotonic non-ionic contrast down the working channel of a flexible bronchoscope (**Figure 1c**) (15).

Bronchoscopy (Table 1)

Bronchoscopy has a role in the diagnosis of airway pathology, the intraoperative management, postoperative management, and long-term follow-up. To evaluate both upper and lower airways, rigid and flexible bronchoscopy should be combined. The role of bronchoscopy in both anterior and posterior aortopexy have increased during the last number of years (16, 17). During these operations bronchoscopy is used to guide the improvement of airway size and stiches are inserted under bronchoscopy vision. Persistent wheezing, stridor and difficult breathing are indications to perform bronchoscopy and may be indicative of airway pathology.

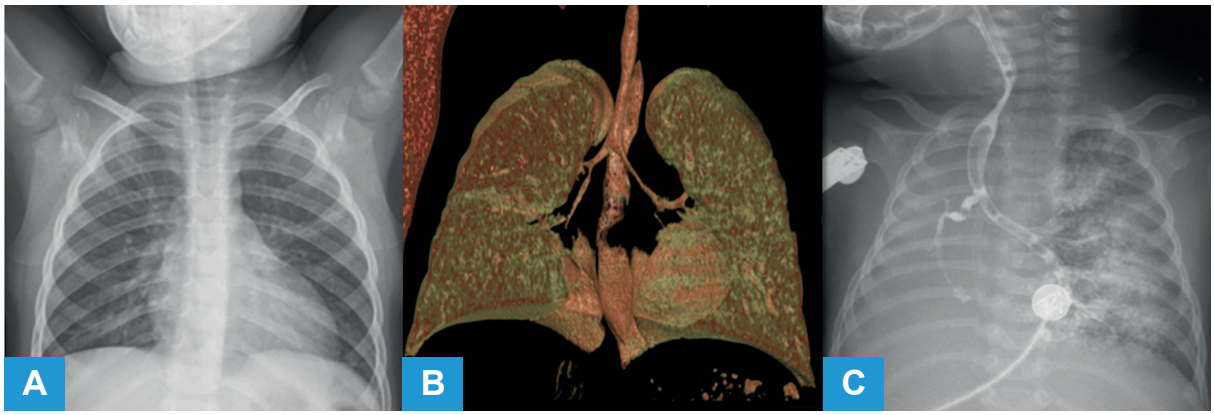

***Figure 1**. 3-year-old baby with noisy breathing: (**A**) chest X-ray demonstrates that the distal part of the trachea is not clearly visible; (**B**) CT scan reconstruction showing the tracheal stenosis but also narrow main bronchi with the right especially narrow; (**C**) tracheobronchogram done with water soluble contract medium demonstrates tracheal narrowing just below ETT. The right main bronchus is also narrow with absent RUL and RML bronchus.*

In cases of difficult intubation and ventilation, bronchoscopy is essential to exclude congenital airway pathology.

Echocardiography

Congenital airway lesions are associated with cardiac abnormalities (14). Echocardiography needs to be performed before airway surgery to plan the correct surgical procedures and cardiac lesions may have to be corrected at the same time. Long segment tracheal stenosis is associated with left pulmonary artery (LPA) sling.

Contrast study

Contrast swallow studies may be helpful in identifying different types of vascular compression (**Table 2**) (18).

Contrast swallow studies is also important in the diagnosis of laryngeal clefts, H-type tracheoesophageal fistula (TOF), swallowing incoordination and reflux. It is important to identify gastroesophageal reflux as the additional inflammation my worsen airway pathology and reduce the chance of successful surgical repair.

Table 1. *Bronchoscopy findings in congenital airway abnormalities.*

CONGENITAL AIRWAY ABNORMALITIES		BRONCHOSCOPY FINDINGS
Airway agenesis/ stenosis	Tracheal agenesis	• Confirming esophageal intubation and TOF or BOF
	Bronchial agenesis	• Absence of bronchus or rudimentary bronchus • Deviation of trachea • No carina visible • Airway narrowing due to shift of mediastinum • Compression by surrounding vessels
	Bronchial atresia	• Blinding-ending bronchus or segmental bronchus
	Hypoplastic lung	• Abnormal bronchial configuration • Absent RUL and RML
	Tracheal stenosis	• Solid tracheal rings • LPA compression • Tracheal bronchus • Abnormal position of carina
Airway branching anomalies	Tracheal bronchus	• Displaced RUL bronchus, • supplying a normal RUL about 2cm above carina • Supernumerary bronchus, existing in addition to a normal RUL bronchus • Rudimentary blind ending RUL bronchus • LUL tracheal bronchus rare
	Bridging bronchus	• Pseudocarina at lower level T5-7 • T-shape pseudocarina due to angle of BB from LMB • Anormal bronchus, originating from the LMB crosses • Airway stenosis • LMB vascular compression
	Heterotaxy syndromes	
	o *Right isomerism*	• Bilateral right-side atria • The abnormal bronchial branching patterns includes bilateral right-side bronchial branching
	o *Left isomerism*	• Bilateral left-side bronchial branching
	Scimitar syndrome	• Absent RUL bronchus
CONGENITAL AIRWAY ABNORMALITIES		**BRONCHOSCOPY FINDINGS**
Airway fistula	Laryngotracheoesophageal cleft	• Redundant posterior mucosa herniating into the laryngeal lumen • Determine type according to the level and extend of connection
	Congenital trachea-oesophageal fistula (TOF)	• Position of fistula • Commonly close to carina • Tracheomalacia • Vascular abnormalities
	Congenital broncho-oesophageal fistula (BOF)	• Fistula is usually short and is running directly from the esophagus to a main or segmental bronchus

→

Airway function abnormality	Primary ciliary dyskinesia	
External airway compression	Bronchogenic Cyst (BC)	• Compression of trachea • Prominent carina • Compression of bronchi
	Foregut duplication cyst	• Compression of trachea • Prominent carina • Compression of bronchi • Shifting of mediastinum
	Vascular compression	
	o *Double aortic arch (DAA)*	• Compression and indentation of the right wall of the right wall of the distal trachea • RMB opening can be narrowed
	o *Left pulmonary artery (LPA) sling*	• Distal tracheal compression with significant pulsations on both anterior and posterior wall • Difficult to find carina • Tracheal stenosis may be present
	o *Left main bronchus (LMB) compression*	• Narrow LMB just below carina • Pulsation both from medial and lateral • Distal to narrowing LMB is patent
	o *Innominate artery compression*	• Anterior tracheal wall compression with pulsation • Tracheomalacia • Coming from left lower to the right upper portion of trachea
	o *Interrupted aortic arch*	• Compression of the LMB
	o *Right sided aortic arch with aberrant subclavian artery*	• Tracheal compression from the right side as well as posterior
	o *Absent pulmonary valve*	• Compression of the lower trachea, LMB and RMB or bronchus intermedius
Others	Horseshoe lung	• Abnormal right bronchial branching
	Mediastinal mass causing airway compression	• Anterior or posterior compression of trachea
	Tracheobronchomalacia (TBM)	
	o *Congenital (Primary)*	• Spontaneous respiration under general anaesthesia needed to detect more than 50% bulging of the Membranous s trachea during coughing or during expiratory phase of the respiratory cycle • Widening of the posterior membranous wall with a crescent shaped lumen of the trachea. Associated TOF
	o *Acquired (Secondary)*	
	Airway compression associated with congenital heart disease	• Compression of different parts of bronchial tree. LMB compression by enlarge left atrium • PA can compress both main bronchi

Table 2. *Types of vascular compression as was identified on contrast swallow.*

Anterior tracheal, posterior esophageal indentations	double aortic arch
Normal tracheal, posterior esophageal indentation	aberrant subclavian artery
Posterior tracheal, anterior esophageal indentation	pulmonary sling
Anterior tracheal indentation, normal esophageal	Innominate arterial compression

AIRWAY AGENESIS/STENOSIS

Tracheal agenesis

Tracheal agenesis is a rare and usually a fatal malformation. Tracheal agenesis can be divided into three types: type 1, the proximal trachea is absent, and a short distal trachea is present and connected to the esophagus via a TOF; type 2 (most common), most of the trachea is absent with a short carina dividing into

right and left bronchi and usually but not always TOF and type 3, the right and left bronchi arise separately from the esophagus (19). Tracheobronchial or esophageal stenosis might be present.

Bronchial agenesis

Bronchial agenesis more commonly affects the left rather than right side and is associated with absence of the ipsilateral lung and pulmonary artery (PA) resulting in cardio-mediastinal shift to that side with overinflation of the contralateral lung.

This shift produces distortion of cardio-mediastinal structures, which is more marked when the right lung is absent because of greater shift of the normally left-side heart, associated vessels, and airways. Most cases of unilateral pulmonary agenesis are associated with other anomalies, including congenital heart disease, other vascular abnormalities, pulmonary sling, other PA anomalies, anomalous origin of arch great vessels, esophageal atresia, tracheal stenosis, lung, and vertebral anomalies (20, 21).

Bronchial atresia (Figure 2A, B)

Bronchial atresia is increasingly being reported in conjunction with congenital pulmonary airway malformation and CLE as a diagnosis depicting an abrupt interruption of the airway with a distal mucus plug (22, 23). Atresia is an abrupt complete interruption of the bronchus which can be identified on CT-imaging as a mucus plug distal from the atresia and a local hypodense region distally (12). The mucus plug is formed due to accumulated mucus produced in the patent distal bronchus (mucocele).

The hypodense region is a result of hyperinflation of the excluded lung parenchyma by collateral ventilation through the pores of Kohn.

In younger children, atresia may be difficult to diagnose due to the small diameter of distally located airways in relation to the resolution of the CT-scanner.

Hypoplastic right lung

Hypoplastic lung occurs due to a decreased number of branching generations of the airways, with decreased number of acini and alveolar size (1). Right sided hypoplasia is much more common than left sided lesions.

Tracheal stenosis

Congenital tracheal stenosis (CTS) is a rare condition with an estimated incidence of 1 in 64,500 births (24). CTS is an embryological abnormality of the tracheal skeleton, with the presence of complete tracheal rings along the stenotic segment and creating a fixed narrow tracheal lumen. CTS could be focal or present over a long segment and is usually associated with absence of the posterior trachealis muscle with complete cartilaginous rings in the affected regions.

The symptoms start in a few days after birth and symptoms are associated with the severity of the stenosis more than stenotic length. The symptoms are variable and are directly related to the degree of narrowing of the lumen and stenotic length.

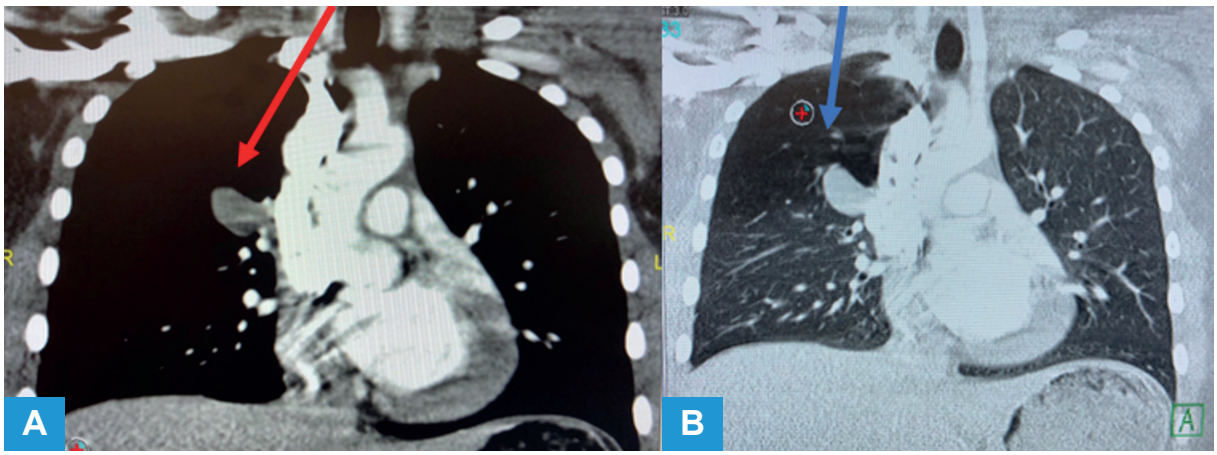

***Figure 2A, B.**. 9-year-old girl: chest CT scans demonstrate RUL bronchial atresia with mucocele and hypodense area visible in the RUL area. Lobectomy was done due to recurrent infections.*

CTS is associated with several other lesions including tracheal bronchus, other airway/lung anomalies as well as pulmonary sling, other structural cardiovascular disease, H-type TOF and Down syndrome.

Long-segment tracheal stenosis is strongly associated with type 2 pulmonary sling, occurring in 2 out of 3 cases (25). In the PA sling anomaly, the left PA arises from the right PA and courses to the left hilum between the trachea and esophagus.

There is a variety of airway abnormalities accompany the type 2 sling, including separate right upper lobe (RUL) bronchus or diverticulum at the normal carinal level resembling a tracheal bronchus, long-segment airway stenosis, low horizontal pseudocarina and bridging right bronchus arising from the left bronchus (20, 26).

In the type 1 pulmonary sling, the airway is usually not stenotic, but the right bronchus might be compressed by the sling or malacic, resulting in air trapping in the right lung. A tracheal bronchus might be present, and the carina is at the normal level (20, 27).

Congenital cardiac anomalies are often also associated with pulmonary sling (mostly type 2) including ventricular septal defect (VSD) or more complex heart disease. Lung abnormalities also occur, including hypoplastic or even absent lung (usually right), as well as bronchopulmonary malformations such as bronchogenic cyst, pulmonary sequestration, and scimitar syndrome; gastrointestinal anomalies might also be present (27).

The gold standard for the diagnosis of CTS is the rigid and flexible airway endoscopy. Sometimes the airway is so narrow that even an ultrathin flexible scope cannot be passed through the stenosis. CT or MRI are essential to study the associated vascular malformations.

The left PA is reimplanted onto the left side of the main PA. Sliding tracheoplasty is the technique of choice for long-segment tracheal stenosis, allowing for tracheal repair without the use of graft or prosthetic materials, promoting easier postoperative recovery, fewer infectious complications, and better long-term outcomes. In most of the cases, there is not a transition zone between the normal and complete rings. Only in few cases it is possible to identify a transition tracheal segment with rings from normal horseshoe shape to a complete ring (28-30).

AIRWAY BRANCHING ANOMALIES

Tracheal bronchus

There is a 13-fold increase in the incidence of tracheal bronchus in children with congenital heart disease including complex cardiac anomalies (31). Especially Down syndrome is commonly associated with a tracheal bronchus. A tracheal bronchus is almost always on the right side but is occasionally on the left or even bilateral, especially in the context of right isomerism (32).

Tracheal bronchus can be completely asymptomatic or be associated with recurrent pneumonia, stridor, and respiratory distress (21). It is a displaced bronchus supplying the whole RUL rather than a supernumerary accessory bronchus. The origin of the bronchus can be stenotic, and occasionally the anomalous branch ends blindly as a tracheal diverticulum.

Bridging bronchus (BB)

The abnormality consists of an abnormal bronchus, originating from the left main bronchus (LMB), which crosses (bridges) the mediastinum and supplies the right lower lobe (RLL), and often the right middle lobe (RML) (33). It is associated with a LPA sling, where the LPA arises from the right PA and passes posteriorly, above the right main bronchus, between the trachea and esophagus, to the hilum of the left lung (34).

Heterotaxy syndromes

Heterotaxy syndromes are associated with abnormal abdominal situs, organ anomalies, aberrant airway, and pulmonary vascular branching as well as variable simple to complex underlying cardiovascular abnormalities (35, 36).

Right isomerism

Characterized by asplenia, horizontal liver, complex heart disease and bilateral right-side atria. The abnormal bronchial branching patterns includes bilateral right-side bronchial branching (eparterial, short mainstem bronchi) and bilateral trilobed (right) lungs (13).

Left isomerism

Characterized by poly-splenia, absent intrahepatic inferior vena cava (IVC) with azygous continuation. Simple to complex cardiac anomalies along with bilateral left-side atria can be present. The abnormal bronchial branching patterns includes bilateral left-side bronchial

branching (hyparterial, long mainstem bronchi) and bilateral bi-lobed (left) lungs (21).

Scimitar syndrome

Scimitar syndrome is characterized by hypoplastic lung (almost always on the right side), aberrant airway branching and hypoplastic right pulmonary artery (RPA). Ipsilateral anomalous pulmonary vein (vertical curved scimitar vein) usually draining to the IVC, and pulmonary sequestration or anomalous systemic vessel from the upper abdominal aorta to the RLL. Right diaphragmatic eventration or hernia may be present.

The airway branching anomalies include absent RUL bronchus with only two lobar bronchi, often accompanied by an aberrant RLL bronchial branch supplying a horseshoe segment of right lung extending behind the IVC across the midline to the left side, abutting or even fused with the left lung (37, 38).

AIRWAY FISTULA

Laryngotracheoesophageal clefts (Figure 3)

They are associated with the VACTERL (vertebral defects, anal atresia, tracheoesophageal fistula with esophageal atresia, cardiac anomalies, renal anomalies, and limb anomalies) spectrum as well as several syndromes including CHARGE (coloboma, heart malformation, choanal atresia, mental retardation, genitourinary and ear anomalies) and DiGeorge syndromes (39). Laryngotracheobronchial malacia frequently coexists with the clefts in these children. Laryngotracheoesophageal clefts are divided into four types depending on the length and location of the defect (40). A cleft should be considered in the cases of recurrent pneumonia or suspected aspiration. The aspiration into the airway through the cleft might be seen on an esophagram but can be mistaken for pharyngeal aspiration or a TOF. A larger cleft might be recognized on CT; however, the diagnosis and extent are defined by laryngoscopy.

The Benjamin-Inglis classification system published in 1989 describes four types of laryngeal clefts: type I involves an interarytenoid defect to the level of the true vocal folds; type II, partial extension through the posterior cricoid cartilage; type III is an extension completely through the posterior cricoid cartilage and possible extension into the cervical trachea and type IV involves extension into the intrathoracic trachea (40).

***Figure 3**. Bronchoscope image of a 3-week-old baby with recurrent aspiration: Connection is visible between the trachea and the esophagus creating a laryngotracheoesophageal cleft.*

The diagnosis can be missed with flexible bronchoscopy as it may be difficult to see the defect due to abundant tissue. Flexible and rigid bronchoscopy should be combined to exclude laryngotracheoesophageal clefts (41).

Congenital TOF

Most cases of congenital TOF are associated with esophageal atresia and a fistulous connection between the trachea and esophagus that can be proximal, distal (most common type, near the carina) or both (21, 42). A laryngeal cleft might also be present in these cases.

Esophageal atresia occurs more rarely without a fistula, and in this situation, there is absent gas in the gastrointestinal (GI) tract after birth. The diagnosis of an H-type fistula without esophageal atresia is often delayed and can present much later. This may be a difficult diagnosis as it can easily be missed on both contrast study and bronchoscopy.

Anomalies of the VACTERL spectrum as well as both tracheal and esophageal stenoses are associated with TOF (21).

Congenital broncho-esophageal fistula (BOF)

Congenital BOFs were first described by Negus in 1929 (43). In 1965, Braimbridge and Keith classified

it into four categories (44). This is a rare diagnosis and even more rare in young children with about 100 cases have been reported in the literature in mostly adult patients. The presentation may be delayed until childhood or adult life, with a median age of 33 years old, while the duration of symptoms can vary from 6 months to 50 years, with a mean of 17 years (45, 46). The majority (90%) of fistulas are type II according to the Braimbridge classification (47).

The communication is usually short and is running directly from the esophagus to a main or segmental bronchus. Conventional barium esophagography is the most sensitive and most "rewarding" tool in the diagnosis of BOFs (45, 47). In other types, the fistula is communicating with a congenital esophageal diverticulum (type I), an intralobar cyst (type III) or a pulmonary sequestration (type IV) (43).

EXTERNAL AIRWAY COMPRESSION

Bronchogenic cyst

Bronchogenic cyst (BC) are mostly single, unilocular cysts which is filled with fluid or mucous. The BC arise from abnormal budding of the primitive trachea-bronchial tree during airway development, but do not branch further and end as a blind pouch (48).

BC most commonly occur in the mediastinum adjacent to one of the mainstem bronchi but can occur anywhere throughout the thoracic cavity including the retroperitoneum, neck, tongue, and subcutaneous tissue. About 65% to 90% of BC occur in the paratracheal, subcarinal, or hilar regions (48, 49). The most common location is subcarinal followed by the right paratracheal region (50). Intraparenchymal BC are found in approximately 12% of cases (51). They are lined by pseudostratified ciliated columnar respiratory epithelium and contain hyaline cartilage plates (48). Parenchymal BC have no communication between the cysts and the tracheobronchial tree. Communication of mediastinal BC with airways have rarely been reported (52). Expanding BC can cause central airway obstruction due to mass effect with or without distal lung hyperinflation. Due to mucus built-up, the cyst can enlarge and lead to infection. Infected BCs are more often found in older children and adults than in neonates or infants (12, 53). BC can be diagnosed on antenatal ultrasound and confirmed on CT scan after birth.

Foregut duplication cysts

Esophageal duplication cysts are rare congenital cystic masses which result from an error in foregut budding in the 4th to 6th week of embryonic development. The incidence remains unknown but the reported incidence from autopsies is 1 in 8200 autopsies (54).

William E Ladd used the term "duplications of the alimentary tract" in 1937 and he applied the term to congenital lesions having: (1) the presence of a well-developed coat of smooth muscle; (2) an epithelial lining representing some type of intestinal tract mucosa, and (3) intimate anatomic association with some portion of the gastrointestinal tract (55).

The foregut budding defects may lead to either a BC, esophageal duplication cyst, or "bronchopulmonary foregut malformations" (BPFM) (56).

Duplication of the cervical portion of the embryonic foregut accounts for 23% of all esophageal duplications and present as either an enlarging neck mass or with upper airway obstructive symptoms. They can also be asymptomatic. Duplication cysts of the mid esophagus constitute 17% of esophageal duplications and in the lower third in 60%. Esophageal duplication cysts can present with respiratory distress due to mass effects and compression on the large airways.

Airway compression is more often seen with BC compared to foregut duplication cysts due to their close location to the large airways (57).

Esophageal duplication cysts are difficult to diagnose on chest X-rays as the features are similar to that of the BCs but with the wall of the esophageal duplication cyst in more close contact with the esophagus.

Duplication cysts may be a rounded mass, with uniform density similar to that of the cardiac shadow and located close to the mediastinum. Air-fluid levels may be seen if the lesion communicates with the tracheobronchial tree (58).

A CT or MRI is needed for diagnosis and soft-tissue characterization. A CT scan can show a water attenuation structure which is located close to the esophagus. Early surgical resection is indicated to prevent recurrent or persistent pulmonary infection (59).

Vascular compression

The airways of young children are more pliable and smaller than those of adults. This makes them more likely to be symptomatic from extrinsic airway compression

(60). Vascular airway compression occurs with several entities associated with enlarged, malpositioned or encircling central vessels (so called rings and slings) (21, 61).

Double aortic arch

Double aortic arch (DAA) (**Figure 4A-F**) is the most common cause of vascular compression of the airway in children (62-64). DAA is determined by the presence of both left- and right-sided aortic arches, which together surround the trachea and esophagus. The right arch is usually the larger ('dominant') one and the left arch is usually small ('hypoplastic') or forms a fibrous cord ('atretic' segment) beyond the origin of the left common carotid or subclavian artery (65). The fibrous cord tethers the patent part of the left-sided arch to the descending aorta, completing the ring. A ductal ligament connects the distal left arch to the proximal LPA and form a ring. The fibrous cord is normally not seen on imaging and the diagnosis is made on the presence of an incomplete left arch (65). The presence of this diverticulum implies the presence of a non-visualized ductal ligament along with a vascular ring (66).

Depending on the type, the descending aorta may be left- or right-sided or may run in the midline anterior to the vertebral column. With the descending aorta midline, the structures of the mediastinum are 'stacked', resulting in compression between the spinal column and the sternum (67).

DAA presents in infancy, with symptoms including dysphagia, stridor, wheezing and respiratory distress (68). Transection of the non-dominant arch is required to relieve the airway compression (69). It is important to diagnose the arch anatomy before surgery because this determines the operative approach. Thirty percent of children will have residual symptoms despite surgical treatment. The residual symptoms may be due to persistent airway compression or severe malacia of the lower trachea (67).

Left pulmonary artery sling

In the PA sling anomaly, the Left pulmonary artery sling (LPA) arises from the RPA and courses to the left hilum between the trachea and esophagus.

There are 2 types of LPA: Type 1, position of the carina is usually normal, at the T4-T5 level and Type 2, low position of the carina, typically at the T6 level. In type 1 the airway is usually not stenotic, but the right bronchus might be compressed by the sling.

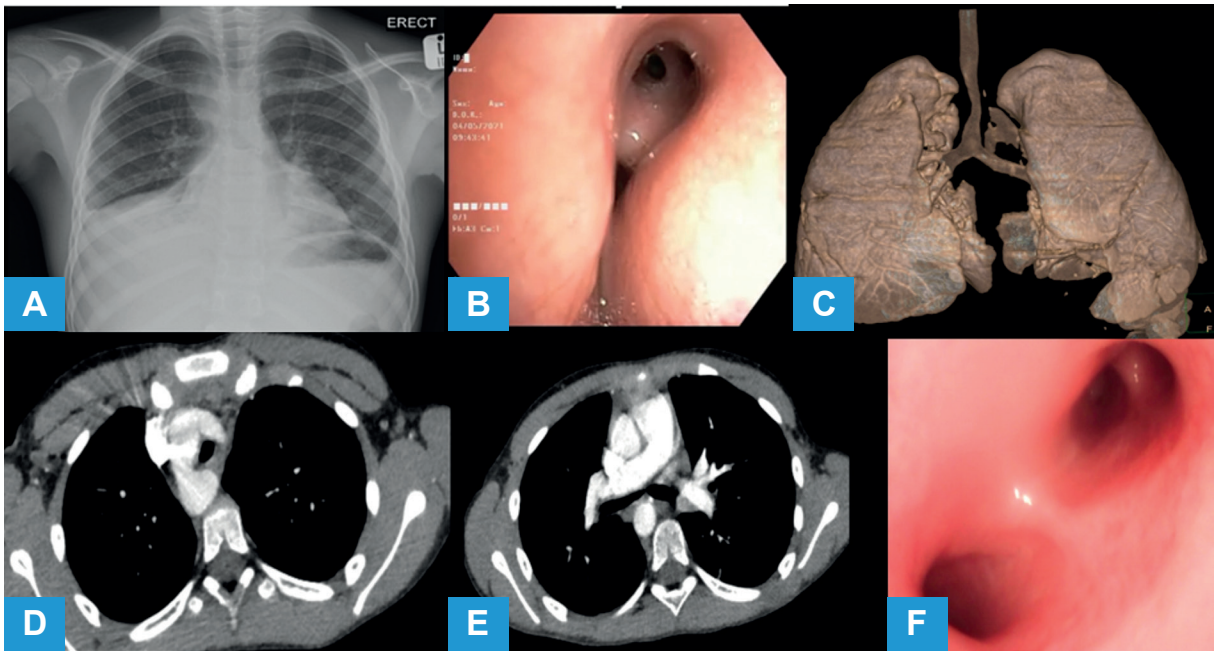

***Figure 4**. 8-year-old boy, who had previous surgery for double aortic arch presented with: (**A**) collapse of the RML and RLL on the chest X-ray; (**B**) bronchoscopy image demonstrates compression of the opening of the RMB; (**C, D, E**) chest CT scans confirms large right-side arch causing distal trachea and RMB compression. The descending aorta (**E**) running on the right side also causes compression of the right main bronchus; (**F**) bronchoscopy image during posterior aortopexy showing that the compression has significantly improved.*

Long-segment tracheal stenosis due to complete cartilaginous rings, T-shaped carina, and a bridging bronchus are often seen in patients with type 2 PA sling (20, 31, 70) (**Figure 5**). Type 2 is often associated with congenital cardiac anomalies.

LPA is suspected based on careful evaluation of plain radiographs and especially recognition of a narrowed or poorly visualized airway with a low horizontal bifurcation, right lung overinflation or hypoplasia. CT angiography is the most common study of choice to fully define the vascular and airway anomalies.

The LPA is reimplanted onto the left side of the main PA and sliding tracheoplasty is the technique of choice for long-segment tracheal stenosis.

Left main bronchus vascular compression

The left main bronchus (LMB) has a longer course and a smaller diameter than the right main bronchus and is wedged between the PA anteriorly and the esophagus, descending aorta (DA) and vertebral body posteriorly (71). Vascular compression of the LMB in the absence of cardiac disease is at least in part, related to an anteriorly positioned DA. The prespinal position of the DA has also been noted as a normal variant in asymptomatic children. Bronchoscopy demonstrate medial and lateral compression with pulsation of the LMB just after the off take of the LMB. The airway distal to the area of compression is normal (72, 73). At surgery a ligament or patent ductus arteriosus remnant can be identified pulling the PA and DA to each other and trapping the LMB (74).

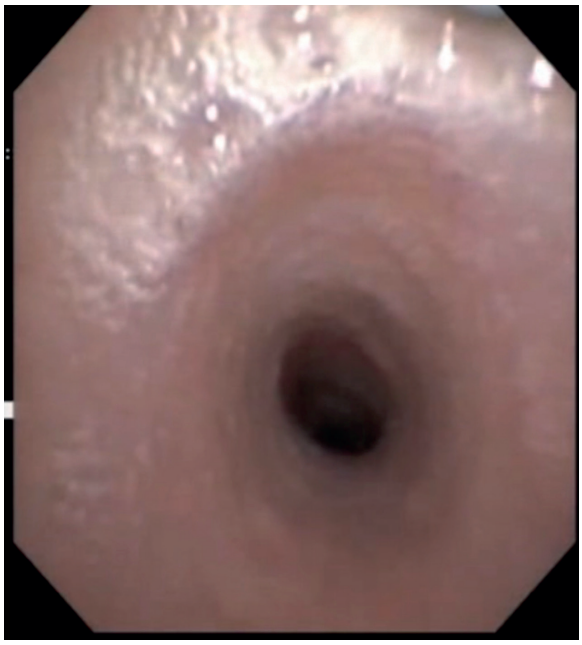

***Figure 5**. Bronchoscopy image demonstrate solid tracheal rings confirming congenital tracheal stenosis.*

Innominate artery compression

Anterior compression of the trachea by the brachiocephalic trunk (Innominate artery compression - IAC) is a commonly seen on bronchoscopy. This is due to the innominate artery as it crosses from left to right and symptoms similar to those who do have a vascular ring (75). This condition seems to have been over-diagnosed and possibly over-treated in the past. IAC needs to be differentiated from tracheomalacia as seen in esophageal atresia (76). Stacking may also play a role with a large thymus pushing the innominate artery onto the anterior part of the trachea.

Operative intervention is indicated for patients with more than 80% compression of the tracheal lumen, as measured on bronchoscopy (77). The innominate artery is suspended to the sternum, with multiple techniques. Anterior thoracotomy can be used with removal of part of the thymus.

Intraoperative bronchoscopy confirms successful pexy and opening of the compressed trachea.

Other causes of vascular compression

- **Interrupted aortic arch (IAA)**. Some part of the lumen of the aortic arch is discontinuous and is found in about 1% of all children with CHD. The airway compression seen in IAA is a consequence of surgical repair and mostly not related to the malformation (14).
- **Right-sided aortic arch (RAA)** with an aberrant left subclavian artery. RAA with an aberrant left subclavian artery and/or a left *ligamentum arteriosum* is reported in 12%-25% of children with vascular rings. These children are mostly asymptomatic. The left subclavian artery often originates from an outpouching of the descending aorta, called a Kommerell's diverticulum. If airway compression is present, it is due to airway compression in the RAA and is usually due to the enlargement of the Kommerell's diverticulum, a short *ligamentum arteriosum* or a midline descending aorta (78).
- **Absent pulmonary valve (APVS)**. It is characterized by the presence of enlarged PAs and hypoplastic pulmonary valve cusps. It is seen in association with ventricular septal defect and right ventricular outflow tract obstruction (14). Compression of the

lower trachea, LMB and right main bronchus or bronchus intermedius occurs due to enlargement of the PAs and left atrium (79, 80).

HORSESHOE LUNG

Horseshoe lung is a very rare congenital malformation in which the bases of the right and left lung are fused to each other by a narrow isthmus. Although rare, a hyperlucent area in the lower left lung, close to the vertebral column, may represent a horseshoe lung. Horseshoe lung is often associated with scimitar syndrome (81). There may be an aberrant RLL bronchial branch which supply the horseshoe segment of the right lung.

TRACHEOBRONCHOMALACIA

Tracheomalacia (TM) is the most common congenital tracheal abnormality with a reported incidence of 1 in 2,100 children (82, 83). Tracheobronchomalacia (TBM) has been often reported in infants and young children who underwent bronchoscopic evaluation for respiratory distress. TM is an abnormal softness of the tracheal wall due to structural anomalies of tracheal cartilaginous and/or posterior membrane. The ratio among cartilage and the floppy part of the normal trachea is around 4.5/1. The symptoms are thought to often be incorrectly attributed to asthma.

TM is difficult to diagnose due to dynamic airway narrowing. Diagnosis of TM is based on dynamic trachea-bronchoscopy with direct observation of the tracheal collapse during spontaneous breathing. The aim of bronchoscopy is to evaluate the percentage of reduction of airway lumen and the sites involved (upper, middle, distal trachea, proximal and peripheral bronchi). TM may be either primary (congenital) or secondary (acquired). Primary TBM is caused by impaired cartilage maturation or cartilage deficiency and is relatively uncommon. Primary or congenital TM/TBM can be found alone or in conjunction with other genetic and congenital disorders (84, 85). TM is associated with morphogenetic airway anomalies including tracheoesophageal fistula, esophageal atresia, mucopolysaccharidoses and polychondritis.

Bronchoscopy and inspiration/expiration CT imaging are used to evaluate airway collapse in expiration. Caliber change of >50% between inspiration and expiration is the criterion for diagnosis of TM (21).

AIRWAY ABNORMALITIES ASSOCIATED WITH CONGENITAL HEART DISEASE

Airway abnormalities are important but sometimes overlooked problems in children with congenital heart disease. It is often difficult to separate symptoms related to cardiac disease from those associated with airway or lung disease. Some of the lesions are incidental while others cause significant symptoms and are important in overall functional outcome. Congenital and acquired as well as intrinsic and extrinsic lesions occur and can overlap.

CONCLUSIONS

Infants with stridor, abnormal cry, feeding difficulties, and signs of airway obstruction should be evaluated with an awake flexible laryngoscopy and, if necessary, a direct laryngoscopy or bronchoscopy. The diagnosis of congenital airway anomalies requires a high degree of suspicion. Treatment often involves a multidisciplinary team approach given the rate of associated abnormalities and complexity of disease.

COMPLIANCE WITH ETHICAL STANDARDS

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Author contributions

Prof. PG, Dr. DR, Dr. AG and Prof. JJ are the only Authors responsible for the conception and design of the work as well as the acquisition, analysis and interpretation of data presented.

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Animal studies

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N/A.

Data sharing and data accessibility

The data underlying this article are available in the article.

Publication ethics

Plagiarism

All original studies are cited as appropriate.

Data falsification and fabrication

All the data correspond to the real.

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