Provisionally accepted for publication

REVIEW

Diagnosis of congenital airway abnormalities in children

Pierre **Goussard** PhD[1], Delano Rhode FC **Paed**[1], Andre **Gie** PhD[1], Jacques **Janson** PhD[2]

[1]Department of Paediatrics and Child Health, Faculty of Medicine and Health Sciences, Stellenbosch University and Tygerberg Hospital, Cape Town, South Africa

[2]Department of Surgical Sciences, Division of Cardiothoracic Surgery, Stellenbosch University and Tygerberg Hospital, Tygerberg, South Africa

Doi: 10.56164/PediatrRespirJ.2023.15

Address for correspondence:

Prof Pierre Goussard, Department of Paediatrics and Child Health, Faculty of Medicine and Health Sciences, Stellenbosch University, PO Box 241, Cape Town 8000, South Africa. Tel +27-21-938-9506; Email pgouss@sun.ac.za

ORCID identification: 0000-0003-1146-1307

Conflict of Interest

The authors have declared no conflict of interest.

Financial Support

There were no institutional or private funding for this article.

Authorship

Prof P Goussard, Dr D Rhode, Dr A Gie and Prof Jacques Janson are the only authors responsible for the conception and design of the work as well as the acquisition, analysis and interpretation of data presented.

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 present 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.

Keywords: 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 apnoea (11). Double aortic arch is the most common vascular abnormality, causing both tracheal and oesophageal 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 abnormalities; (5) vascular abnormalities; and (6) other lesions, including vertebral anomalies, gastrointestinal anomalies, and cardiac anomalies (12).

Chest X-ray: Looking at the airways (Figure 1 a)

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 1 b)

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.

Contrast bronchography (Figure 1 c)

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 (15).

Bronchoscopy

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.

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 (18):

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

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.

AIRWAY AGENESIS/STENOSIS (Table 1)

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 oesophagus 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 oesophagus (19).

Tracheobronchial or oesophageal 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 cardiomediastinal shift to that side with overinflation of the contralateral lung.

This shift produces distortion of cardiomediastinal 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, oesophageal atresia, tracheal stenosis, lung and vertebral anomalies (20,21).

Bronchial atresia (Figure 2)

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.

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 oesophagus.

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 a 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 oesophagus, 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).

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 oesophageal atresia and a fistulous connection between the trachea and oesophagus 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.

Oesophageal 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 oesophageal 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 oesophageal stenoses are associated with TOF (21).

Congenital broncho-oesophageal 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 oesophagus to a main or segmental bronchus.

Conventional barium oesophagography 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 oesophageal diverticulum (type I), an intralobar cyst (type III) or a pulmonary sequestration (type IV) (43).

EXTERNAL AIRWAY COMPRESSION

Bronchogenic Cyst (BC)

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

Oesophageal 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, oesophageal duplication cyst, or "bronchopulmonary foregut malformations" (BPFM) (56).

Duplication of the cervical portion of the embryonic foregut accounts for 23% of all oesophageal 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 oesophagus constitute 17% of oesophageal duplications and in the lower third in 60%. Oesophageal 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).

Oesophageal 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 oesophagus.

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 oesophagus.

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 (DAA) **(Figure 4)**

DAA 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 oesophagus. 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 nonvisualized 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 present 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 (LPA) **(Figure 5)**

In the PA sling anomaly, the 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. 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).

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 (LMB) vascular compression

The 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).

Innominate artery compression (IAC)

Anterior compression of the trachea by the brachiocephalic trunk (innominate artery) 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 oesophageal 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 (TBM)

Tracheomalacia (TM) is the most common congenital tracheal abnormality with a reported incidence of 1 in 2,100 children (82,83).

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.

REFERENCES

1. Thacker PG, Rao AG, Hill JG, Lee EY. Congenital lung anomalies in children and adults: current concepts and imaging findings. Radiol Clin North Am. 2014 Jan;52(1):155-81. doi: 10.1016/j.rcl.2013.09.001. Epub 2013 Oct 2. PMID: 24267716.
2. Lee EY, Boiselle PM, Cleveland RH. Multidetector CT evaluation of congenital lung anomalies. Radiology. 2008 Jun;247(3):632-48. doi: 10.1148/radiol.2473062124. PMID: 18487532.
3. Yikilmaz A, Lee EY. CT imaging of mass-like nonvascular pulmonary lesions in children. Pediatr Radiol. 2007 Dec;37(12):1253-63. doi: 10.1007/s00247-007-0637-4. Epub 2007 Oct 31. PMID: 17972072.
4. Goussard P, Schubert P, Janson J, Naidoo S, Rhode D, Parker N, Andronikou S. "To the Editor" recurrent wheezing caused by the combination of bronchogenic cyst and congenital lobar emphysema in a young infant. Pediatr Pulmonol. 2023 Jan;58(1):340-344. doi: 10.1002/ppul.26181. Epub 2022 Oct 11. PMID: 36193810.

5. Thacker PG, Rao AG, Hill JG, Lee EY. Congenital lung anomalies in children and adults: current concepts and imaging findings. Radiol Clin North Am. 2014 Jan;52(1):155-81. doi: 10.1016/j.rcl.2013.09.001. Epub 2013 Oct 2. PMID: 24267716.
6. Montella S, Corcione A, Santamaria F. Recurrent Pneumonia in Children: A Reasoned Diagnostic Approach and a Single Centre Experience. Int J Mol Sci. 2017 Jan 29;18(2):296. doi: 10.3390/ijms18020296. PMID: 28146079; PMCID: PMC5343832.
7. Gu W, Jiang W, Zhang X, Chen Z, Yan Y, Huang L, Wang M, Shao X, Wang S, Ji W. Refractory wheezing in Chinese children under 3 years of age: bronchial inflammation and airway malformation. BMC Pediatr. 2016 Aug 27;16(1):145. doi: 10.1186/s12887-016-0680-0. PMID: 27568177; PMCID: PMC5002096.
8. Boesch RP, Baughn JM, Cofer SA, Balakrishnan K. Trans-nasal flexible bronchoscopy in wheezing children: Diagnostic yield, impact on therapy, and prevalence of laryngeal cleft. Pediatr Pulmonol. 2018 Mar;53(3):310-315. doi: 10.1002/ppul.23829. Epub 2017 Sep 14. PMID: 28910519.
9. Saglani S, Nicholson AG, Scallan M, Balfour-Lynn I, Rosenthal M, Payne DN, Bush A. Investigation of young children with severe recurrent wheeze: any clinical benefit? Eur Respir J. 2006 Jan;27(1):29-35. doi: 10.1183/09031936.06.00030605. PMID: 16387932.
10. Boyer D, Barsky E, Papantonakis CM, Pittman J, Ren CL, Esther CR Jr, Wilson KC, Thomson CC. Diagnostic Evaluation of Infants with Recurrent or Persistent Wheezing. Ann Am Thorac Soc. 2016 Nov;13(11):2057-2059. doi: 10.1513/AnnalsATS.201607-575CME. PMID: 27831796.
11. Goussard P, Pohunek P, Eber E, Midulla F, Di Mattia G, Merven M, Janson JT. Pediatric bronchoscopy: recent advances and clinical challenges. Expert Rev Respir Med. 2021 Apr;15(4):453-475. doi: 10.1080/17476348.2021.1882854. Epub 2021 Feb 10. PMID: 33512252.
12. Lee EY, Dorkin H, Vargas SO. Congenital pulmonary malformations in pediatric patients: review and update on etiology, classification, and imaging findings. Radiol Clin North Am. 2011 Sep;49(5):921-48. doi: 10.1016/j.rcl.2011.06.009. PMID: 21889015.

13. Newman B. Airway abnormalities associated with congenital heart disease. Pediatr Radiol. 2022 Sep;52(10):1849-1861. doi: 10.1007/s00247-022-05429-0. Epub 2022 Jul 2. PMID: 35778574.
14. McLaren CA, Elliott MJ, Roebuck DJ. Vascular compression of the airway in children. Paediatr Respir Rev. 2008 Jun;9(2):85-94. doi: 10.1016/j.prrv.2007.12.008. Epub 2008 May 13. PMID: 18513668.
15. McLaren CA, Elliott MJ, Roebuck DJ. Tracheobronchial intervention in children. Eur J Radiol. 2005 Jan;53(1):22-34. doi: 10.1016/j.ejrad.2004.07.022. PMID: 15607850.
16. Torre M, Carlucci M, Speggiorin S, Elliott MJ. Aortopexy for the treatment of tracheomalacia in children: review of the literature. Ital J Pediatr. 2012 Oct 30;38:62. doi: 10.1186/1824-7288-38-62. PMID: 23110796; PMCID: PMC3502176.
17. Arcieri L, Serio P, Nenna R, Di Maurizio M, Baggi R, Assanta N, Moschetti R, Noccioli B, Mirabile L, Murzi B. The role of posterior aortopexy in the treatment of left mainstem bronchus compression. Interact Cardiovasc Thorac Surg. 2016 Nov;23(5):699-704. doi: 10.1093/icvts/ivw209. Epub 2016 Jul 5. PMID: 27382044.
18. Burch M, Balaji S, Deanfield JE, Sullivan ID. Investigation of vascular compression of the trachea: the complementary roles of barium swallow and echocardiography. Arch Dis Child. 1993 Feb;68(2):171-6. doi: 10.1136/adc.68.2.171. PMID: 8481037; PMCID: PMC1029228.
19. Strouse PJ, Newman B, Hernandez RJ, Afshani E, Bommaraju M. CT of tracheal agenesis. Pediatr Radiol. 2006 Sep;36(9):920-6. doi: 10.1007/s00247-006-0231-1. Epub 2006 Jun 21. PMID: 16788813.
20. Newman B, Alkhori N. Congenital central pulmonary artery anomalies: Part 2. Pediatr Radiol. 2020 Jul;50(8):1030-1040. doi: 10.1007/s00247-020-04703-3. Epub 2020 Jun 4. PMID: 32500159.
21. Caffey's pediatric diagnostic imaging. 13th ed. Coley BD, editor. Philadelphia: Elsevier Saunders; 2019.
22. Riedlinger WF, Vargas SO, Jennings RW, Estroff JA, Barnewolt CE, Lillehei CW, Wilson JM, Colin AA, Reid LM, Kozakewich HP. Bronchial atresia is common to

extralobar sequestration, intralobar sequestration, congenital cystic adenomatoid malformation, and lobar emphysema. Pediatr Dev Pathol. 2006 Sep-Oct;9(5):361-73. doi: 10.2350/06-01-0023.1. PMID: 16953677.

23. Newman B. Congenital bronchopulmonary foregut malformations: concepts and controversies. Pediatr Radiol. 2006 Aug;36(8):773-91. doi: 10.1007/s00247-006-0115-4. Epub 2006 Mar 22. PMID: 16552585.
24. Hewitt RJ, Butler CR, Maughan EF, Elliott MJ. Congenital tracheobronchial stenosis. Semin Pediatr Surg. 2016 Jun;25(3):144-9. doi: 10.1053/j.sempedsurg.2016.02.007. Epub 2016 Feb 19. PMID: 27301600.
25. Harada A, Shimojima N, Shimotakahara A, Azuma S, Ishizuka Y, Tomita H, Hirobe S. Surgical indication for congenital tracheal stenosis complicated by pulmonary artery sling. J Thorac Dis. 2019 Dec;11(12):5474-5479. doi: 10.21037/jtd.2019.11.31. PMID: 32030266; PMCID: PMC6987985.
26. Wells TR, Gwinn JL, Landing BH, Stanley P. Reconsideration of the anatomy of sling left pulmonary artery: the association of one form with bridging bronchus and imperforate anus. Anatomic and diagnostic aspects. J Pediatr Surg. 1988 Oct;23(10):892-8. doi: 10.1016/s0022-3468(88)80379-8. PMID: 3069994.
27. Newman B, Cho Ya. Left pulmonary artery sling--anatomy and imaging. Semin Ultrasound CT MR. 2010 Apr;31(2):158-70. doi: 10.1053/j.sult.2010.01.004. PMID: 20304323.
28. Maeda K. Pediatric airway surgery. Pediatr Surg Int. 2017 Apr;33(4):435-443. doi: 10.1007/s00383-016-4050-7. Epub 2017 Jan 28. PMID: 28132084.
29. Varela P, Torre M, Schweiger C, Nakamura H. Congenital tracheal malformations. Pediatr Surg Int. 2018 Jul;34(7):701-713. doi: 10.1007/s00383-018-4291-8. Epub 2018 May 30. PMID: 29846792.
30. Rutter MJ, Cotton RT, Azizkhan RG, Manning PB. Slide tracheoplasty for the management of complete tracheal rings. J Pediatr Surg. 2003 Jun;38(6):928-34. doi: 10.1016/s0022-3468(03)00126-x. PMID: 12778396.
31. Chassagnon G, Morel B, Carpentier E, Ducou Le Pointe H, Sirinelli D. Tracheobronchial Branching Abnormalities: Lobe-based Classification Scheme. Radiographics. 2016 Mar-Apr;36(2):358-73. doi: 10.1148/rg.2016150115. Epub

2016 Feb 1. Erratum in: Radiographics. 2016 Jul-Aug;36(4):1258. PMID: 26824513.

32. Lee EY, Restrepo R, Dillman JR, Ridge CA, Hammer MR, Boiselle PM. Imaging evaluation of pediatric trachea and bronchi: systematic review and updates. Semin Roentgenol. 2012 Apr;47(2):182-96. doi: 10.1053/j.ro.2011.12.002. PMID: 22370196.

33. Henry BM, Cheruiyot I, Wong LM, Keet K, Mutua V, Chhapola V, Tubbs RS. The bridging bronchus: A comprehensive review of a rare, potentially life-threatening congenital airway anomaly associated with cardiovascular defects. Pediatr Pulmonol. 2019 Dec;54(12):1895-1904. doi: 10.1002/ppul.24488. Epub 2019 Aug 29. PMID: 31468716.

34. du Plessis AM, Andronikou S, Goussaard P. Bridging bronchus and sling left pulmonary artery: a rare entity demonstrated by coronal CT with 3-D rendering display and minimal-intensity projections. Pediatr Radiol. 2008 Sep;38(9):1024-6. doi: 10.1007/s00247-008-0913-y. Epub 2008 Jun 6. Erratum in: Pediatr Radiol. 2008 Aug;38(8):919. Goussaard, Pierre [corrected to Goussard, Pierre]. PMID: 18535823.

35. Yim D, Nagata H, Lam CZ, Grosse-Wortmann L, Seed M, Jaeggi E, Yoo SJ. Disharmonious Patterns of Heterotaxy and Isomerism: How Often Are the Classic Patterns Breached? Circ Cardiovasc Imaging. 2018 Feb;11(2):e006917. doi: 10.1161/CIRCIMAGING.117.006917. PMID: 29444810.

36. Harrison MJ, Shapiro AJ, Kennedy MP. Congenital Heart Disease and Primary Ciliary Dyskinesia. Paediatr Respir Rev. 2016 Mar;18:25-32. doi: 10.1016/j.prrv.2015.09.003. Epub 2015 Sep 26. PMID: 26545972.

37. Midyat L, Demir E, Aşkin M, Gülen F, Ulger Z, Tanaç R, Bayraktaroğlu S. Eponym. Scimitar syndrome. Eur J Pediatr. 2010 Oct;169(10):1171-7. doi: 10.1007/s00431-010-1152-4. Epub 2010 Mar 12. PMID: 20225123.

38. Korkmaz AA, Yildiz CE, Onan B, Guden M, Cetin G, Babaoglu K. Scimitar syndrome: a complex form of anomalous pulmonary venous return. J Card Surg. 2011 Sep;26(5):529-34. doi: 10.1111/j.1540-8191.2011.01309.x. Epub 2011 Aug 31. PMID: 21883462.

39. Leboulanger N, Garabédian EN. Laryngo-tracheo-oesophageal clefts. Orphanet J Rare Dis. 2011 Dec 7;6:81. doi: 10.1186/1750-1172-6-81. PMID: 22151899; PMCID: PMC3261097.
40. Benjamin B, Inglis A. Minor congenital laryngeal clefts: diagnosis and classification. Ann Otol Rhinol Laryngol. 1989 Jun;98(6):417-20. doi: 10.1177/000348948909800603. PMID: 2729823.
41. Boesch RP, Baughn JM, Cofer SA, Balakrishnan K. Trans-nasal flexible bronchoscopy in wheezing children: Diagnostic yield, impact on therapy, and prevalence of laryngeal cleft. Pediatr Pulmonol. 2018 Mar;53(3):310-315. doi: 10.1002/ppul.23829. Epub 2017 Sep 14. PMID: 28910519.
42. Hseu A, Recko T, Jennings R, Nuss R. Upper Airway Anomalies in Congenital Tracheoesophageal Fistula and Esophageal Atresia Patients. Ann Otol Rhinol Laryngol. 2015 Oct;124(10):808-13. doi: 10.1177/0003489415586844. Epub 2015 May 12. PMID: 25969571.
43. Savvoula S, Vasiliki P, Kyparisia K, Christina A, Ioannis M, Athanasios K. Congenital broncho-oesophageal fistula: An unusual cause of persistent pneumonia in a young adult. Respir Med Case Rep. 2013 Jan 12;8:21-4. doi: 10.1016/j.rmcr.2012.12.003. PMID: 26029609; PMCID: PMC3920443.
44. Zhang BS, Zhou NK, Yu CH. Congenital bronchoesophageal fistula in adults. World J Gastroenterol. 2011 Mar 14;17(10):1358-61. doi: 10.3748/wjg.v17.i10.1358. PMID: 21455337; PMCID: PMC3068273.
45. Dogan R, Farsak B, Yilmaz M, Tok M, Güngen Y. Congenital broncho-oesophageal fistula associated with bronchiectasis in adults. Report of two cases and review of the literature. Respiration. 1999;66(4):361-5. doi: 10.1159/000029390. PMID: 10461087.
46. Bekoe S, Magovern GJ, Liebler GA, Park SB, Cushing WJ. Congenital bronchoesophageal fistula in the adult. Chest. 1974 Aug;66(2):201-3. doi: 10.1378/chest.66.2.201. PMID: 4851152.
47. Su L, Wei XQ, Zhi XY, Xu QS, Ma T. Congenital bronchoesophageal fistula in an adult: a case report. World J Gastroenterol. 2007 Jul 21;13(27):3776-7. doi: 10.3748/wjg.v13.i27.3776. PMID: 17659748; PMCID: PMC4250660.

48. Aktoğu S, Yuncu G, Halilçolar H, Ermete S, Buduneli T. Bronchogenic cysts: clinicopathological presentation and treatment. Eur Respir J. 1996 Oct;9(10):2017-21. doi: 10.1183/09031936.96.09102017. PMID: 8902460.

49. McAdams HP, Kirejczyk WM, Rosado-de-Christenson ML, Matsumoto S. Bronchogenic cyst: imaging features with clinical and histopathologic correlation. Radiology. 2000 Nov;217(2):441-6. doi: 10.1148/radiology.217.2.r00nv19441. PMID: 11058643.

50. Suen HC, Mathisen DJ, Grillo HC, LeBlanc J, McLoud TC, Moncure AC, Hilgenberg AD. Surgical management and radiological characteristics of bronchogenic cysts. Ann Thorac Surg. 1993 Feb;55(2):476-81. doi: 10.1016/0003-4975(93)91022-f. PMID: 8431062.

51. Lee EY, Boiselle PM, Cleveland RH. Multidetector CT evaluation of congenital lung anomalies. Radiology. 2008 Jun;247(3):632-48. doi: 10.1148/radiol.2473062124. PMID: 18487532.

52. Goussard P, Green LL, Janson JT, Schubert P. Subcarinal bronchogenic cyst communicating with tracheal bronchial tree, misdiagnosed as Pulmonary Tuberculosis. Pediatr Pulmonol. 2019 Mar;54(3):228-229. doi: 10.1002/ppul.24240. Epub 2019 Jan 10. PMID: 30632297.

53. Paterson A. Imaging evaluation of congenital lung abnormalities in infants and children. Radiol Clin North Am. 2005 Mar;43(2):303-23. doi: 10.1016/j.rcl.2004.12.004. PMID: 15737371.

54. Arbona JL, Fazzi JG, Mayoral J. Congenital esophageal cysts: case report and review of literature. Am J Gastroenterol. 1984 Mar;79(3):177-182. PMID: 6702802.

55. Lund DP. Alimentary tract duplications. In: Coran AG, Adzick NS, Krummel TM, Laberge JM, Shamberger RC, Caldamone AA, editors. Pediatric surgery. 7th ed. Philadelphia, PA: Elsevier Mosby, 2012. p 1155-1163.

56. Gerle RD, Jaretzki A 3rd, Ashley CA, Berne AS. Congenital bronchopulmonary-foregut malformation. Pulmonary sequestration communicating with the gastrointestinal tract. N Engl J Med. 1968 Jun 27;278(26):1413-1419. doi: 10.1056/NEJM196806272782602. PMID: 5652625.

57. Clements BS. Congenital malformations of the lungs and airways. In: Taussig LM, Landau LI, editors. Pediatric respiratory medicine. St. Louis, USA: Mosby; 1999: p 1106–1136.
58. Eber E. Antenatal diagnosis of congenital thoracic malformations: early surgery, late surgery, or no surgery? Semin Respir Crit Care Med. 2007 Jun;28(3):355-366. doi: 10.1055/s-2007-981656. PMID: 17562505
59. Tiwari S, Kothari P, Gupta A, Jayaswal S, Dikshit V, Kekre G. Thoracoscopic resection of foregut duplication cyst in a neonate. J Minim Access Surg. 2021 Jan-Mar;17(1):88-90. doi: 10.4103/jmas.JMAS_58_20. PMID: 32964888; PMCID: PMC7945632.
60. Yubbu P, Devaraj NK, Sahadan DZ, Latiff HA. Vascular compression of the airways: issues on management in children with congenital heart disease. Prog Pediatr Cardiol 2020;59:doi.org/10.1016/j.ppedcard.2020.101207.
61. Berdon WE. Rings, slings, and other things: vascular compression of the infant trachea updated from the midcentury to the millennium--the legacy of Robert E. Gross, MD, and Edward B. D. Neuhauser, MD. Radiology. 2000 Sep;216(3):624-32. doi: 10.1148/radiology.216.3.r00se40624. PMID: 10966687.
62. McLaughlin RB Jr, Wetmore RF, Tavill MA, Gaynor JW, Spray TL. Vascular anomalies causing symptomatic tracheobronchial compression. Laryngoscope. 1999 Feb;109(2 Pt 1):312-9. doi: 10.1097/00005537-199902000-00025. PMID: 10890785.
63. Woods RK, Sharp RJ, Holcomb GW 3rd, Snyder CL, Lofland GK, Ashcraft KW, Holder TM. Vascular anomalies and tracheoesophageal compression: a single institution's 25-year experience. Ann Thorac Surg. 2001 Aug;72(2):434-8; discussion 438-9. doi: 10.1016/s0003-4975(01)02806-5. PMID: 11515879.
64. Bakker DA, Berger RM, Witsenburg M, Bogers AJ. Vascular rings: a rare cause of common respiratory symptoms. Acta Paediatr. 1999 Sep;88(9):947-52. doi: 10.1080/08035259950168423. PMID: 10519334.
65. Schlesinger AE, Krishnamurthy R, Sena LM, Guillerman RP, Chung T, DiBardino DJ, Fraser CD Jr. Incomplete double aortic arch with atresia of the distal left arch: distinctive imaging appearance. AJR Am J Roentgenol. 2005 May;184(5):1634-9. doi: 10.2214/ajr.184.5.01841634. PMID: 15855130.

66. Gould SW, Rigsby CK, Donnelly LF, McCulloch M, Pizarro C, Epelman M. Useful signs for the assessment of vascular rings on cross-sectional imaging. Pediatr Radiol. 2015 Dec;45(13):2004-16; quiz 2002-3. doi: 10.1007/s00247-015-3424-7. Epub 2015 Aug 11. PMID: 26260202.

67. Fleck RJ, Pacharn P, Fricke BL, Ziegler MA, Cotton RT, Donnelly LF. Imaging findings in pediatric patients with persistent airway symptoms after surgery for double aortic arch. AJR Am J Roentgenol. 2002 May;178(5):1275-9. doi: 10.2214/ajr.178.5.1781275. PMID: 11959745.

68. Brockes C, Vogt PR, Rothe TB, Arbenz U, Turina J. Doppelter Aortenbogen. Klinik, Diagnose und Therapie bei Kindern und Erwachsenen [Double aortic arch: clinical aspects, diagnosis and therapy in children and adults]. Z Kardiol. 2001 Feb;90(2):127-32. German. doi: 10.1007/s003920170199. PMID: 11263002.

69. ten Berge M, van der Laag J, van der Ent CK, Beek FJ. Clinical, radiological and functional follow-up after surgical decompression of double aortic arch. Pediatr Radiol. 2002 Aug;32(8):561-6. doi: 10.1007/s00247-002-0730-7. Epub 2002 May 25. PMID: 12136346.

70. Wells TR, Gwinn JL, Landing BH, Stanley P. Reconsideration of the anatomy of sling left pulmonary artery: the association of one form with bridging bronchus and imperforate anus. Anatomic and diagnostic aspects. J Pediatr Surg. 1988 Oct;23(10):892-8. doi: 10.1016/s0022-3468(88)80379-8. PMID: 3069994.

71. Last RJ. Anatomy, Regional and Applied. London: Churchill Livingstone, 1984, 226–7.

72. Abdel-Rahman U, Ahrens P, Fieguth HG, Kitz R, Heller K, Moritz A. Surgical treatment of tracheomalacia by bronchoscopic monitored aortopexy in infants and children. Ann Thorac Surg. 2002 Aug;74(2):315-9. doi: 10.1016/s0003-4975(02)03642-1. PMID: 12173806.

73. Jang WS, Kim WH, Choi K, Nam J, Kim JT, Lee JR, Kim YJ, Kim GB. Aortopexy with preoperative computed tomography and intraoperative bronchoscopy for patients with central airway obstruction after surgery for congenital heart disease: postoperative computed tomography results and clinical outcomes. Pediatr Cardiol. 2014 Aug;35(6):914-21. doi: 10.1007/s00246-014-0875-9. Epub 2014 Feb 8. PMID: 24509564.

74. Kosloske AM. Left mainstem bronchopexy for severe bronchomalacia. J Pediatr Surg. 1991 Mar;26(3):260-2. doi: 10.1016/0022-3468(91)90499-j. PMID: 2030470.

75. Wine TM, Colman KL, Mehta DK, Maguire RC, Morell VO, Simons JP. Aortopexy for innominate artery tracheal compression in children. Otolaryngol Head Neck Surg. 2013 Jul;149(1):151-5. doi: 10.1177/0194599813483449. Epub 2013 Mar 25. PMID: 23528271.

76. Mahboubi S, Gheyi V. MR imaging of airway obstruction in infants and children. Int J Pediatr Otorhinolaryngol. 2001 Mar;57(3):219-27. doi: 10.1016/s0165-5876(00)00448-1. PMID: 11223454.

77. Worhunsky DJ, Levy BE, Stephens EH, Backer CL. Vascular rings. Semin Pediatr Surg. 2021 Dec;30(6):151128. doi: 10.1016/j.sempedsurg.2021.151128. Epub 2021 Oct 23. PMID: 34930596.

78. Donnelly LF, Fleck RJ, Pacharn P, Ziegler MA, Fricke BL, Cotton RT. Aberrant subclavian arteries: cross-sectional imaging findings in infants and children referred for evaluation of extrinsic airway compression. AJR Am J Roentgenol. 2002 May;178(5):1269-74. doi: 10.2214/ajr.178.5.1781269. PMID: 11959744.

79. Kussman BD, Geva T, McGowan FX. Cardiovascular causes of airway compression. Paediatr Anaesth. 2004 Jan;14(1):60-74. doi: 10.1046/j.1460-9592.2003.01192.x. PMID: 14717876.

80. Nölke L, Azakie A, Anagnostopoulos PV, Alphonso N, Karl TR. The Lecompte maneuver for relief of airway compression in absent pulmonary valve syndrome. Ann Thorac Surg. 2006 May;81(5):1802-7. doi: 10.1016/j.athoracsur.2005.12.001. PMID: 16631676.

81. Mfingwana L, Goussard P, Andronikou S, Morrison J. Horseshoe lung in a young child at Tygerberg Hospital, South Africa. Afr J Thorac Crit Care Med. 2021 Dec 31;27(4):10.7196/AJTCCM.2021.v27i4.138. doi: 10.7196/AJTCCM.2021.v27i4.138. PMID: 35359695; PMCID: PMC8948481.

82. Boogaard R, Huijsmans SH, Pijnenburg MW, Tiddens HA, de Jongste JC, Merkus PJ. Tracheomalacia and bronchomalacia in children: incidence and patient characteristics. Chest. 2005 Nov;128(5):3391-7. doi: 10.1378/chest.128.5.3391. PMID: 16304290.

83. Fischer AJ, Singh SB, Adam RJ, Stoltz DA, Baranano CF, Kao S, Weinberger MM, McCray PB Jr, Starner TD. Tracheomalacia is associated with lower FEV1 and Pseudomonas acquisition in children with CF. Pediatr Pulmonol. 2014 Oct;49(10):960-70. doi: 10.1002/ppul.22922. Epub 2013 Oct 25. PMID: 24166775; PMCID: PMC4711356.

84. Kamran A, Jennings RW. Tracheomalacia and Tracheobronchomalacia in Pediatrics: An Overview of Evaluation, Medical Management, and Surgical Treatment. Front Pediatr. 2019 Dec 12;7:512. doi: 10.3389/fped.2019.00512. PMID: 31921725; PMCID: PMC6922019.

85. Hysinger EB, Panitch HB. Paediatric Tracheomalacia. Paediatr Respir Rev. 2016 Jan;17:9-15. doi: 10.1016/j.prrv.2015.03.002. Epub 2015 Mar 17. PMID: 25962857.

LEGENDS

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.

Figure 2

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.

Figure 3

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

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 - 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.

Figure 5

Bronchoscopy images demonstrate solid tracheal rings confirming congenital tracheal stenosis.

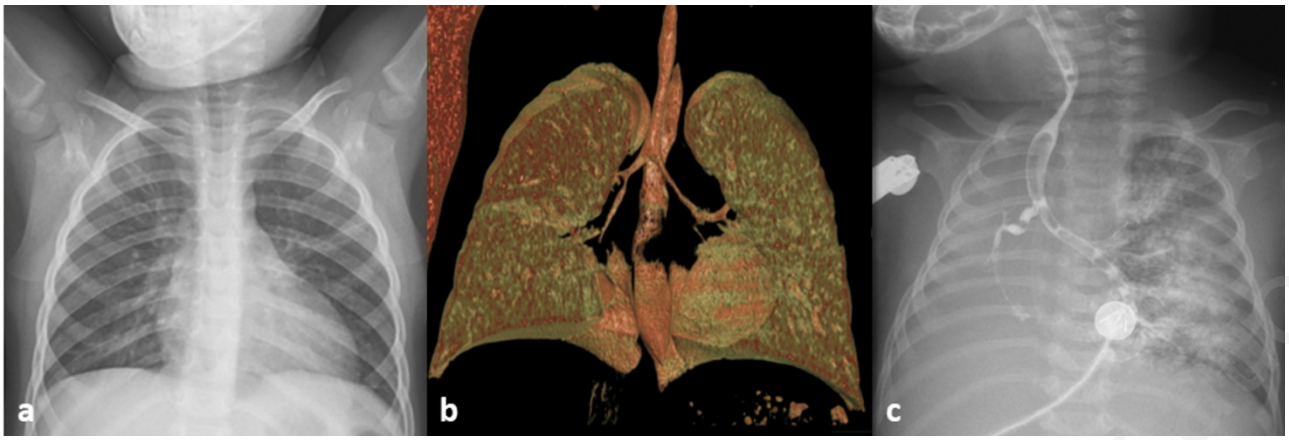

Figure 1.

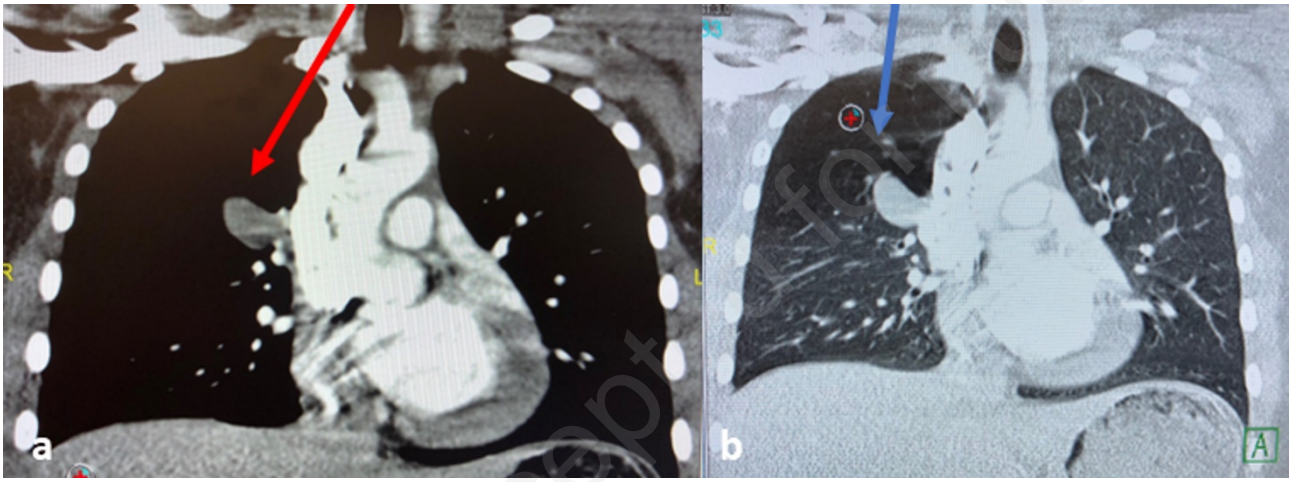

Figure 2.

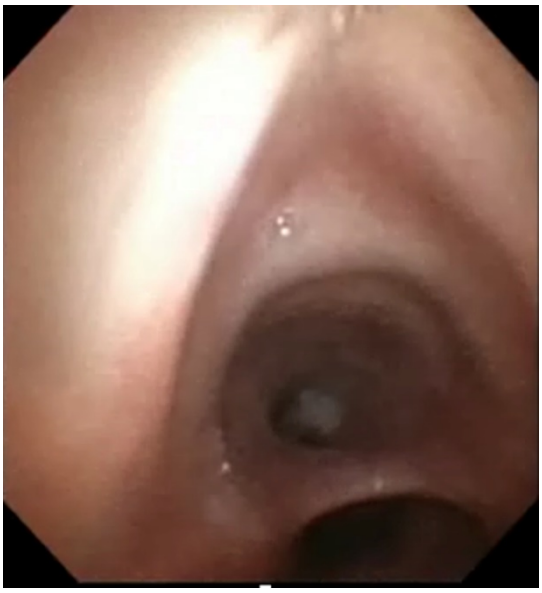

Figure 3.

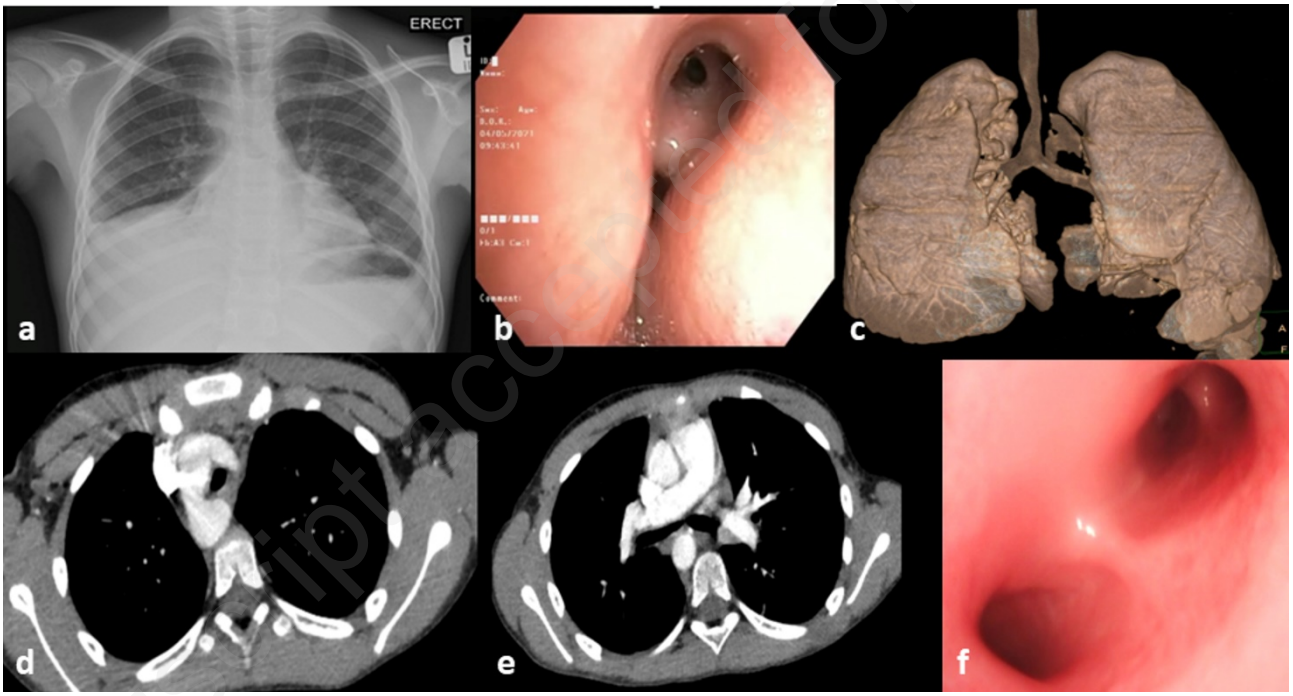

Figure 4.

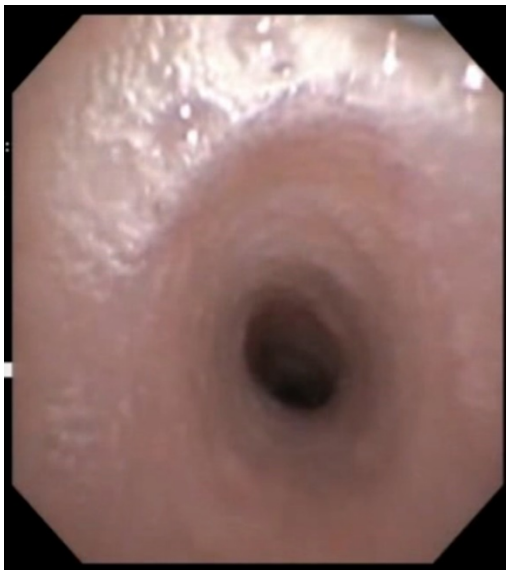

Figure 5.

TABLE 1

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
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)	• 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 *Congenital (Primary)*	
	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