Pulmonary artery sling: An overview.

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Abstract

Pulmonary artery sling is a rare childhood vascualr ring that is frequently associated with tracheal stensois. Consequently, neonates may present with a crictal airway obstruction if tehre is long segemt tarcheal stensosi and complete rings. Rapid diagnosis of this cardiac vascular malfomation and extent airway involvement is essentail as longterm outcoem can be excellent follwoing surgicla repair. In this review we focus on airway invetsigation and management for this challenging congential condition.

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Introduction:

Pulmonary artery sling (PAS) is a condition in which one of the branch pulmonary arteries originates from the contralateral sided pulmonary artery. Pulmonary artery sling was first described by Glaevecke and Doehle in 1897.[1] The disease spectrum ranges from asymptomatic state to severe life-threatening airway obstruction requiring high pressure mechanical ventilation, or in extreme cases, extracorporeal support. A "ring-sling" complex describes when there is a congenital anomaly of the major airways including complete tracheal rings with tracheal stenosis and an associated pulmonary artery sling. Morbidity and mortality are related to the extent of airway involvement including tracheobronchial stenosis, lung development, genetic, cardiac comorbidities and the need for intensive care unit admission preceding the surgery.[1,2] In more complex cases with long segment tracheal stenosis, diagnostic workup and airway surgery may require a highly specialised complex airway service and multidisciplinary team approach for optimal results.[3-7]

Anatomy:

Left pulmonary artery sling (LPAS) describes the aberrant path of the left pulmonary artery, instead of arising from the main pulmonary artery, it takes its origin from the postero-superior aspect of the right pulmonary artery. The aberrant left pulmonary artery then courses between the trachea and oesophagus (retrotracheal) to the left hilum, causing tracheobronchial compression at the level of the distal trachea and carina. The trachea is caught between the main pulmonary artery abutting anteriorly and the left pulmonary artery bordering the posterior aspect. Wells et al classified LPAS in two types based on tracheobronchial anatomy, Type 1 (normal bronchus) and Type 2 with a bridging bronchus (Table 1).[8,9] In type-1A LPAS, the carinal anatomy is normal and bifurcates into left and right mainstem bronchi, usually with mild airway compression (figure 1 b). In type-1B LPAS, the carinal position is normal (T4/5 level) however the right upper lobe bronchus is tracheal (arising from the trachea) above the carina and supplies either a single subsegmental apical right upper lobe bronchus (RUL) or rarely the entire RUL (figure 1c 1d) . The eparterial tracheal RUL bronchus may be malacic and can also have a blind ending as a diverticulum.[10] Type-II LPAS has an abnormal tracheobronchial tree without a well defined carina and is generally associated with tracheal stenosis distal to the tracheal RUL bronchus takeoff. This tracheal narrowing leads to a lower pseudo carina (T6) from which the left main bronchus originates (LMB), figure 2. Additionally the RML and RLL are supplied by a single bridging bronchus that crosses the midline from the left (Type-IIA). Type-II B is characterised by a complete absence of the right bronchial tree, with the right lung being supplied entirely by a bridging bronchus from the left mainstem bronchus and mostly, the right lung is hypoplastic.[11,12]In some case series type-II lesions predominate.[8] The narrowed tracheal section often contains complete tracheal rings (CTR) with an absent trachealis muscle.[13]

CTR have been reported in approximately two thirds of LPAS.[3] This combination is often described as 'sling-ring complex.[5,6] The number of complete rings may vary from one or two rings (focal stenosis at the LPA site) to long segment tracheal stenosis involving the proximal trachea as well. CTR may extend distally into the bridging bronchi, creating significant surgical challenges for repair. The trachea may also taper from proximal to distally in a funnel shape as the "rat's tail" trachea.[14] CTR are associated with genes encoding cartilage signalling pathways including Sonic Hedgehog and Wnt.[13] These genetic embryonal developmental pathways are well described in animal models but the exact association of complete tracheal rings and LPA sling remains unknown in humans.[9]

Embryologic Development:

The proximal portion of the pulmonary artery is derived from the ventral sixth aortic arch and the distal part is derived from the post branchial vessels, which develop from the capillary plexus surrounding each lung bud. Normally, the left post branchial vessels connect to the left sixth branchial arch to form the left pulmonary artery, and the right post branchial vessels take the vascular supply from the right sixth branchial arch to make a right pulmonary artery.[14] A left pulmonary artery sling develops when the left post branchial component of the pulmonary artery fails to connect to the left sixth aortic arch, instead connects to the neighbouring right post-branchial component of the pulmonary artery, which in turn connects to the right sixth aortic arch.[15] LPAS is much more common than right pulmonary artery sling. The likely explanation is the "space-available" theory in the development of embryonic foregut mesoderm. The embryonic structures including respiratory diverticulum, lung buds, bronchial buds, sixth branchial arch and left common cardinal vein, all compete for the same space. In the setting of LPA sling, during embryogenesis**,** there might be an early branching of the right upper bronchus, leaving a wider space around the lower developing primitive trachea causing left post bronchial pulmonary vessel to approach the right ventral sixth branchial arch caudally resulting in LPA sling.

Furthermore, during development, the left sixth branchial arch may get vascular supply from a nearby right bronchial artery. Compression of the primitive right lung structures and flow competition from the left post-branchial primitive pulmonary vessel (future LPA) which causes the sling can result in hypoplasia of the right lung and associated tracheal stenosis. The right pulmonary artery sling is possible only in an isomeric setting with an associated left lung hypoplasia.[16,17] Many associated congenital heart defects in LPAS can be explained on the basis of space availability theory. Since there will be an extra space available due to

abnormal origin of the LPA during the development, this would leave roomier left side in the splanchnic mesenchyme for the unrestricted growth of the primitive left sixth arch and the left common cardinal vein resulting in increased occurrence of patent ductus arteriosus and left superior vena cava respectively.[18]

Epidemiology:

LPA slings are quite rare, accounting for less than 5% of vascular rings. The prevalence of LPAS in a large screening study using echocardiography has found a prevalence of 59 per million school-aged children.[19] Cases are reported in patients with Down syndrome, associated oesophageal and lower gastrointestinal anomalies and VACTERL spectrum (vertebral anomalies, imperforate anus, cardiac anomalies, tracheoesophageal fistula, renal and limb anomalies.[20-22] There is evidence of genetic risk factors in LPA sling in identical twins and in patients with trisomy 18.[23,24] Other genetic syndromes associated with LPAS include Holt-Oram and Kartagener syndrome.[24]

Associated congenital heart disease:

Many congenital heart diseases have been reported in association with LPAS. Commonly observed defects are ventricular septal defects, atrial septal defects, coarctation of aorta, persistent arterial duct, persistent left superior vena cava, tetralogy of Fallot and total anomalous pulmonary venous connection.[25-27] Associated heart disease can aggravate the clinical course of the airway anomalies and may increase the complexity of the surgical repair which is commonly performed under cardiopulmonary bypass.[2]

Clinical Manifestations:

The spectrum of clinical manifestations of LPAS is very wide ranging from no symptoms to mild respiratory distress and noisy breathing on exercise, recurrent chest infections and swallowing problems.[28,29] At the other end of the spectrum neonates and infants may present with life-threatening disease due to critical airway narrowing. The diagnosis should be suspected if intubation results in an abnormally high endotracheal tube that cannot be pushed down due to undiagnosed tracheal stenosis (figure 3). Often hypercarbia and ventilation difficulties may be out of proportion to oxygenation issues.[28] Neonates may, however, be symptom-free even in the presence of severe tracheal narrowing due to lower respiratory flow rates which do not lead to turbulent flow.[30] Symptoms of upper airway obstruction and stridor often present after 6 months of age When the patient becomes more ambulant for example following an intercurrent chest infection where the diagnosis can be made as an incidental finding.Older patients may present with an asymptomatic murmur, chronic cough,dyspnea, wheeze or choking sensation.[28,29] The most important prognostic factor is the extent of trachea bronchial disease, especially a long segment of tracheobronchial stenosis, rather than the associated cardiac defects.[29]

Diagnosis:

There are two diagnostic goals in LPAS - firstly to identify the retrotracheal aberrant LPA arising from the RPA and secondly to establish the degree of airway compromise and stenosis and if they are complete tracheal rings. The former can be reliably ascertained with echocardiography showing the main pulmonary artery extending directly to the right pulmonary artery, and the left pulmonary artery arising from the right pulmonary artery (figure 4). Other intracardiac lesions may also be identified by echocardiography.

CT scan and MRI:

The classic tell-tale sign of the LPA wrapping around the back of the trachea is easily seen on contrasted CT (figure 5, 6) or MRI; however these image modalities play a more significant role in the assessment and extent of airway stenosis. MRI provides a good definition of vascular and intracardiac anatomy which takes much longer which is often an issue in an unstable patient. Multidetector CT is much quicker and demonstrates the position and extent of tracheobronchial compression, anatomy of the pulmonary arteries and the spatial relationships among the pulmonary arteries, trachea, bronchi and oesophagus. This is important for surgical planning. Inspiratory and expiratory phase CT scans are also useful for the detection of air trapping and fix tracheal stenosis with a diameter of the trachea remaining the same in both respiratory phases.[31]

Complete rings are not visible on CT scan; however there may be strong suspicion if the trachea is round and not D- shaped with no surrounding air around the endotracheal tube beyond the subglottis (Figue 7). Tracheobronchial airway abnormalities may be quite complex especially with Type II LPAS where there may be a bridging bronchus or blind ending diverticula with lung agenesis. There is a suggestion that CT findings may determine operability and that a conservative approach may be feasible if the trachea calibre is more than 40% compared to reference values.[32-34] CT may overestimate the degree of obstruction in the presence of mucus plugs or where diameters are only 1 to 2 mm.[35] Bronchoscopy and bronchograms may be very useful in this setting to clearly delineate the extent of the stenosis(figure 8).[7,35,36]

Bronchoscopy:

Bronchoscopy is an essential and useful tool to assess the airway and identify the site and number of complete rings. This however needs to be performed by a skilled operator as iatrogenic oedema of the tracheal mucosa may completely obstruct an already narrow airway. Rigid endoscopy can also be used, however views may be limited if there is significant tracheal stenosis. Assessment of the airway via laryngeal mask is ideal as it avoids instrumentation in the larynx.[8,37,38]Maintaining the airway and ensuring ventilation and oxygenation are paramount tasks for the anaesthetist especially in critical cases.[38] In neonates and infants, typically a 2.8mm flexible bronchoscope is used.[37] If the obstruction is severe, a 2.2 mm suction fiberoptic scope can pass through distal tracheal stenosis to visualise the carina. Topical vasoconstrictor epinephrine may reduce procedural risk.

Dynamic bronchoscopy may also be useful to differentiate fixed stenosis from distal malacia , the latter occurring between 20 and 40% of cases. Tracheobronchomalacia contribute significantly to long-term morbidity and need for long-term ventilation, tracheostomy or highly specialised intervention such as tracheal and bronchial stents.[36,39,40] The typical bronchoscopic features of LPA sling are shown in figure 9 with mild cranial compression and left bronchus origin narrowing posteriorly by the retro-tracheal LPA (Type Ia) to significant carinal and right distal lateral tracheal wall compression obscuring the view of the carina into the right main bronchus. The severe end of the spectrum with multiple complete rings and an absent trachealis muscle (figure 9c). These are best seen with a well suctioned airway clear of secretions as these may obscure the presence of complete rings along the trachea floor. If tracheal stenosis is severe it may not even be possible to view the carina - in this instance bronchograms are very informative.[7]

Bronchograms:

Bronchograms should be performed under fluoroscopy preferentially with biplane imaging with AP and lateral views or with the recent 3D dynaCT reconstructions. Bronchogram can accurately determine the exact calibre of the smaller airways which may not be accessible by bronchoscopy. Figure 8 demonstrates the accuracy of this technique in comparison to a simultaneous CT that shows a left main bronchus artefact on CT only - this may be misleading especially for surgical planning. If contrast extends into the alveoli it may also give valuable information on lobar and sub segmental bronchial supply. Whilst only a small volume (typically 0.25 to 0.5ml/kg) of contrast agent are used (Omnipaque or Visipaque) it is rapidly resolved and does not usually cause bronchial irritation. Radiopaque per fluorocarbon is the ideal contrast agent as this is inert and rapidly evaporates following intracheal administration.[41] Gentle balloon dilation to access the tracheal calibre and if there is fixed stenosis can also be done with a low pressure balloon (figure 9) or to assess the airway for potential stenting. Bronchogram and balloon dilations form the cornerstone of post-operative care to treat recurrent stenosis at the operative site.

A single static bedside AP bronchogram in an intubated patient can be invaluable to rapidly diagnose long segment tracheal stenosis and the extent of tracheobronchial involvement (figure 11). Here, a standard chest X-ray is performed with 0.25 to 0.5ml/kg of contrast agent via a thin 6 Fr feeding tube positioned at the end of the endotracheal tube. The contrast is injected rapidly and an image is obtained 3 to 5 seconds after intratracheal injection under expiration. Dense hold-up of contrast indicates significant tracheal obstruction as normally the contrast agent dissipates rapidly into the bronchi creating the typical tram track appearance that lines the airway. This can be performed at the bedside in intensive care if the patient is too unstable

to move or the stenosis is too small for even a bronchoscopy to assess. Barium swallow is to delineate the oesophagus very limited diagnostic value.

Optical coherence tomography:

Optical coherence tomography (OCT) is a very informative imaging technique that uses infrared light to image tissue in a similar way to ultrasound. Due to the wavelength of infrared light, it is able to visualise mucosa and cartilage with extreme accuracy below 1 mm giving comparable views to tissue histology.[42,43] OCT requires a special 1 mm dragonfly catheter (St Jude Medical, Abbott U.S.) which is passed through an endotracheal tube on a monorail system under fluoroscopy - it can then image the exact calibre of the bronchi and trachea including the mucosa and cartilage, which appears as a black acoustic shadow. This technology can accurately identify if the cartilage rings are complete or almost complete (pseudo-complete ring), the latter being potentially balloon dilatable and subject to growth(figure 12). OCT is also able to identify scar tissue and may be able to assess the degree of healing around the tracheal suture lines.

The combination of echocardiography,bronchoscopy,bronchography and CT imaging provide excellent information to plan the surgical approach. It is also worth mentioning that antenatal foetal echocardiography can detect pulmonary artery sling.[44] There are increasing reports on the diagnostic value of antenatal MRI for detection of vascular and cardiac abnormalities in utero.[45]

Finally a genetic workup is also useful to detect major chromosomal abnormalities such as Down syndrome or VACTERL which may be associated with LPAS, especially if spinal and vertebral abnormalities or imperforate anus are present.

Management and outcome:

The management of LPA sling is beyond the scope of the review but is primarily to determine the exact relationship between the tracheobronchial abnormalities and the presenting symptoms and to delineate the extent of airway involvement and stenosis as this significantly influences the outcome. Assisted ventilation or cardiac arrest prior to surgery, associated genetic abnormalities and major cardiac defects are known risk factors which adversely affect the outcome. The most challenging cases which require assisted ventilation,the pressure control ventilation using inspiratory plateau pressure rather than peak pressure is essential as 30 to 50% of the airway pressure is lost and dissipated through the trachea obstruction. This is similar to the permissive hypercarbia strategy originally described by Darioli and Perret for acute severe asthma.[46] However; in contrast to asthma, long inspiratory times of 0.8 to 1 second allow better ventilation as the obstruction is during the inspiratory phase.[47]Usually, lower respiratory rates prevent air trapping and high frequency oscillation is of no benefit. In extreme cases, an extracorporeal membrane oxygenator (ECMO) is required.

Correction of tracheal stenosis is usually under cardiopulmonary bypass especially if there is long segment tracheal stenosis requiring tracheal surgery or tracheoplasty. The left pulmonary artery is re-implanted and any major congenital heart lesions are usually repaired at the same time.[27,28,48-51]

Over the years, there have been many different surgical procedures to treat tracheal stenosis including end-to-end anastomosis for short segment stenosis, patch tracheoplasty (with autologous pericardium, tracheal autograft, aortic homograft) and more recently the slide tracheoplasty. The slide tracheoplasty technique is performed by tracheal division at the midpoint of stenosis, longitudinal incisions on the opposite side of proximal and distal tracheal segments and sliding oblique anastomosis of the segment.[27,28,48,49] Success of this procedure depends on freeing and mobilising the trachea which will then be foreshortened with the slide but increase its diameter without the risk of ischaemia. This innovative technique has shown impressive results all round the world in specialised airway centres with over 70% survival.[29-32,51-59] Length of ICU stay is generally significantly shorter for slide tracheoplasty with less complications.[51,56-59] Average age of surgery is around six months of age which is similar to the time of presentation.

Typically, patients are ventilated for a few days to allow the tracheal sutures to heal and then the patient may require multiple balloon dilations in the ensuing 12 to 24 months at increasingly less frequent intervals.

Following slide tracheoplasty, the trachea usually grows with age with no limitations on exercise later in childhood or adulthood.[60] At the more complex end of the spectrum there may be associated tracheobronchial malacia which may be approached by a number of ways including long-term ventilation or airway stenting, more recently with biodegradable stents.[7,40,61]Untreated tracheal stenosis especially with complete rings may progress and require intervention, although mild cases may be managed without tracheal surgery. However this concept remains as a controversial subject in many centres.[34,62-65]

Reimplanted left pulmonary artery may get stenosis in the future. Ventilation-perfusion scan or cardiac MRI will help to quantify the differential flow to each lung when there is a stenosis of the re-anastomosed pulmonary artery.. Catheter based interventions in the form of ballooning or stenting of the LPA might be required in cases of significant LPA stenosis.[66]

Survival following isolated left pulmonary artery repair (type 1) is usually excellent, approximately 100%.[67,68] In most large case series there has been significant improvement over the last three decades with improving surgical techniques. Even at the severe end of the spectrum, LPA sling occurs in association with long segment tracheal stenosis; survival figures vary from 50 to above 90%.[34,69] There are even reports of reasonable survival for tracheal stenosis following slide tracheoplasty with a single lung.[61,70] Improved survival over the last decade also been due to the wider adoption of complex airway services and multidisciplinary teams which evaluate not only the heart and the airway but focus on other issues such as feeding, swallowing and neurodevelopment.[71] Management of this rare tracheobronchial vascular abnormality is better achieved in large specialised centres with highly skilled staff and availability of advanced diagnostic equipment.[71] Being a rare tracheobronchial vascular abnormality, its management experience with a low volume centre is expected to be less,hence it is advisable to perform the management in large specialised centres where highly skilled staff and advanced diagnostic equipments are available.

Table 1 Classification of LPA sling according to Wells.

Aberrant left pulmonary artery sling with the following features:

Feature	**Type 1 A**	**Type 1 B**	**Type**
Normal carina and tracheobronchial tree	Yes (T4/5)	Yes (T4/5)	No (p
Tracheal Right upper lobe bronchus	No	Yes - usually partial RUL supply (eg apical lobe of RUL)	Yes -u
Bridging bronchus	No	No	Yes -
Tracheal stenosis (complete rings)	Rarely	Rarely or short segment	Comm
Imperforate anus	Rare	Rare	Not u

RUL=right upper lobe T= Thoracic vertebra

Figure 1 Normal anatomy of trachea (a), tracheal stenosis with normal branching of major bronchi (b), tracheal stenosis with an accessory tracheal right upper lobe branch (c) and tracheal stenosis with partial supply of right upper lobe (apical branch) by tracheal bronchus (d). RUL = right upper lobe. LMB= left main bronchus. RMB = right main bronchus.

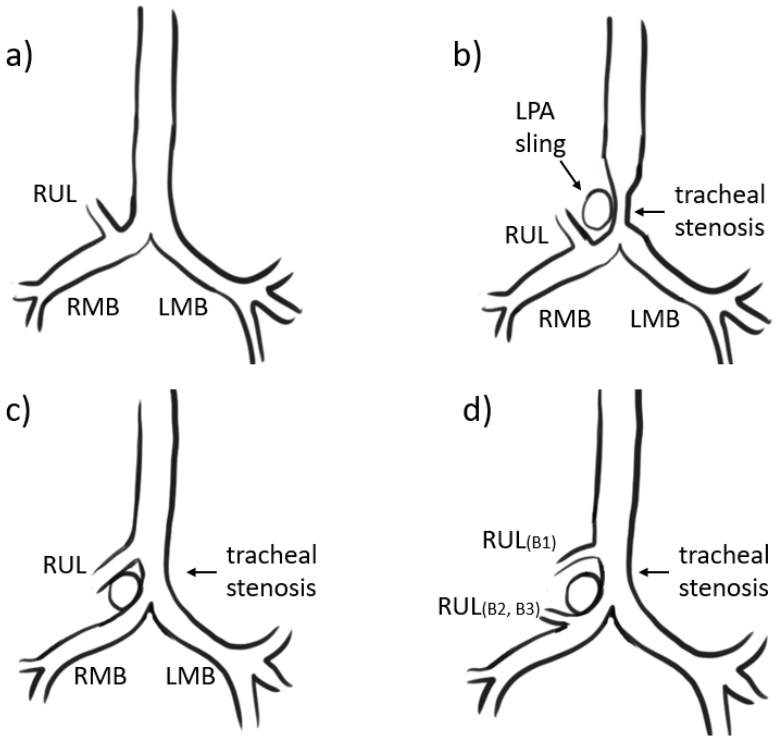


Figure 2 Type 2a LPAS with a pseudo-carina with the bridging bronchus originating from the left main bronchus origin. The right upper lobe is supplied by a tracheal bronchus.RUL = right upper lobe. LMB= left main bronchus. RMB = right main bronchus (bridging bronchus), LPA = left pulmonary artery.

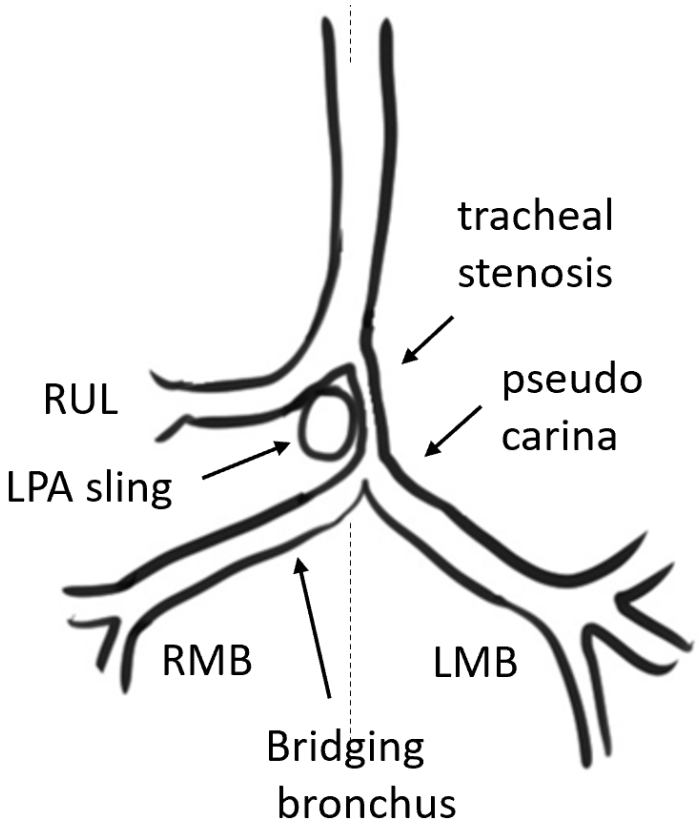


Figure 3 A chest X-ray showing a high tracheal tube at the level of the clavicle (arrow). There is some residual contrast following a bedside bronchogram showing the tracheal outline (tram tracking) with proximal tracheal stenosis and a normal distal trachea, carina and bronchi.

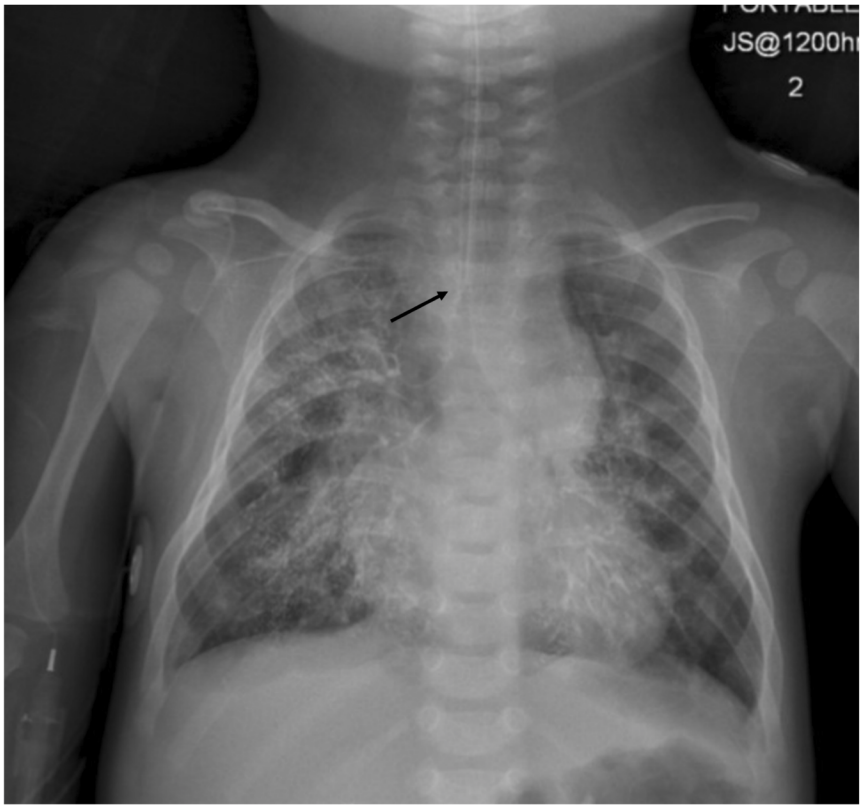


Figure 4 Parasternal short axis echo image showing main pulmonary artery continuing as right pulmonary artery (arrow) and left pulmonary artery (curved arrow) arising from right pulmonary artery.

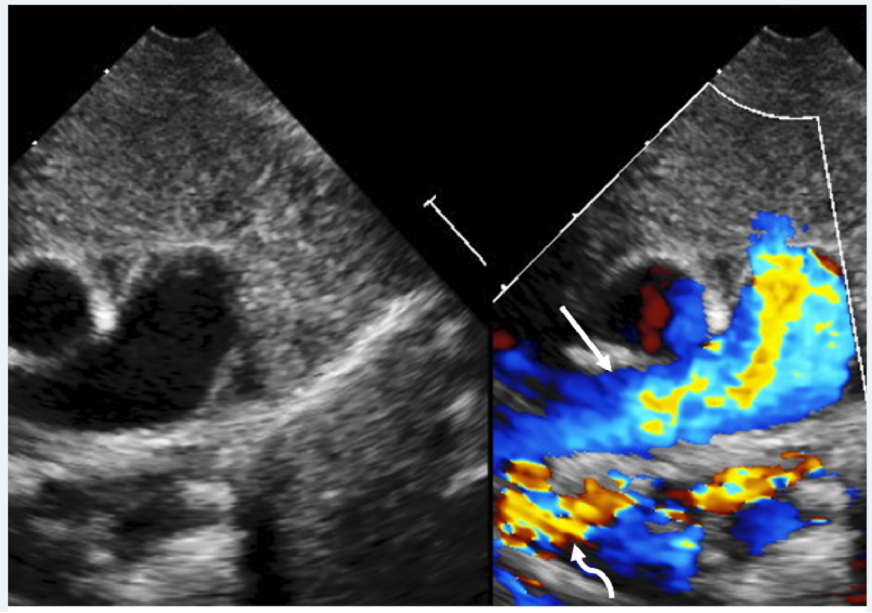


Figure 5 Axial contrast CT showing the left pulmonary artery (LPA, curved arrow) arising from the right

pulmonary artery (RPA, straight arrow) encircling the distal trachea which is very narrow (*). The LPA path is retro-tracheal between the trachea and oesophagus. RV= right ventricle, RPA = right pulmonary artery and LAP = left pulmonary artery

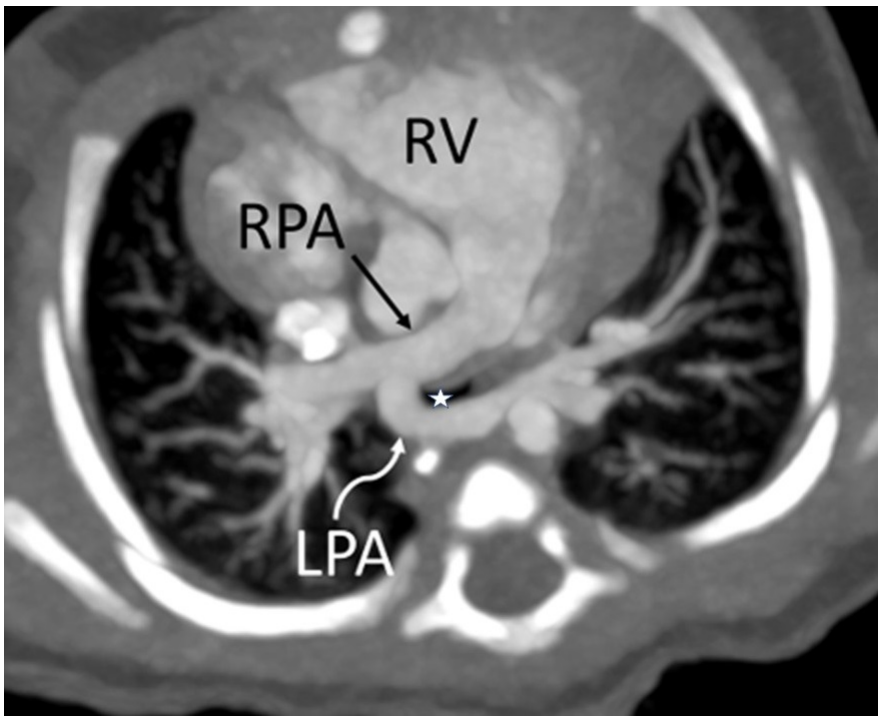


Figure 6 Posterior 3D reconstructed CT angiography image with airway rendered in blue, showing the LPA (curved arrow) encircling the posterior aspect of the right main bronchus coursing anterior to the nasogastric tube (NGT). There is a tracheal right upper lobe which ends abruptly. LPA = left pulmonary artery, RUL = right upper lobe.

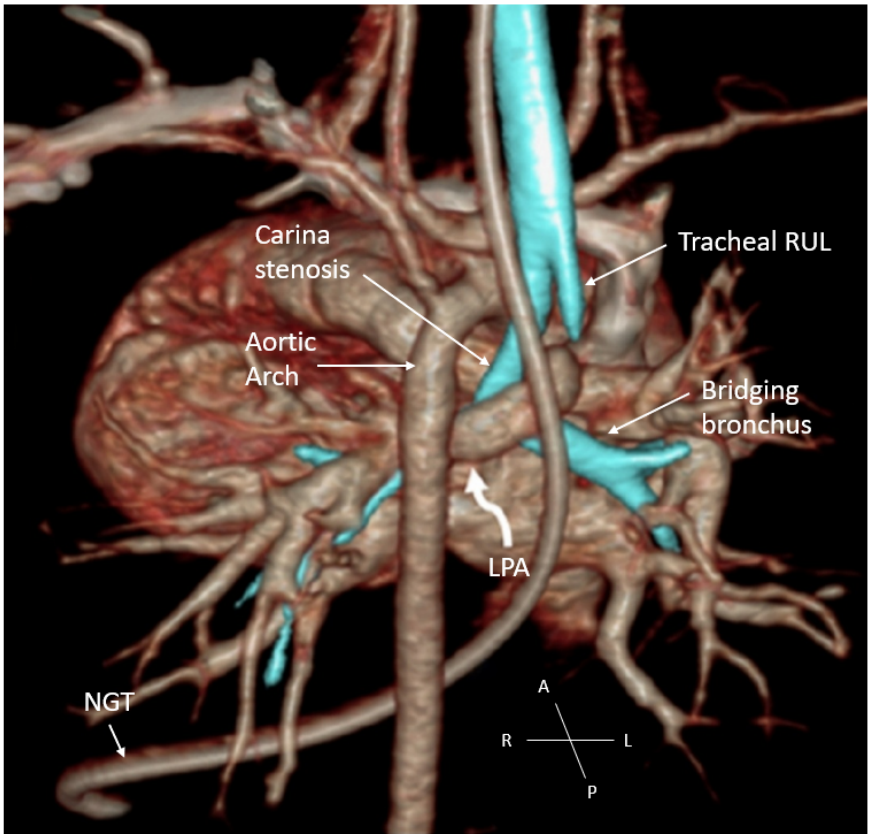


Figure 7 Axial contrast CT of the chest at mid tracheal level showing round endotracheal tube with no air in surrounding trachea indicating significant tracheal stenosis. Complete rings are not visible on CT.

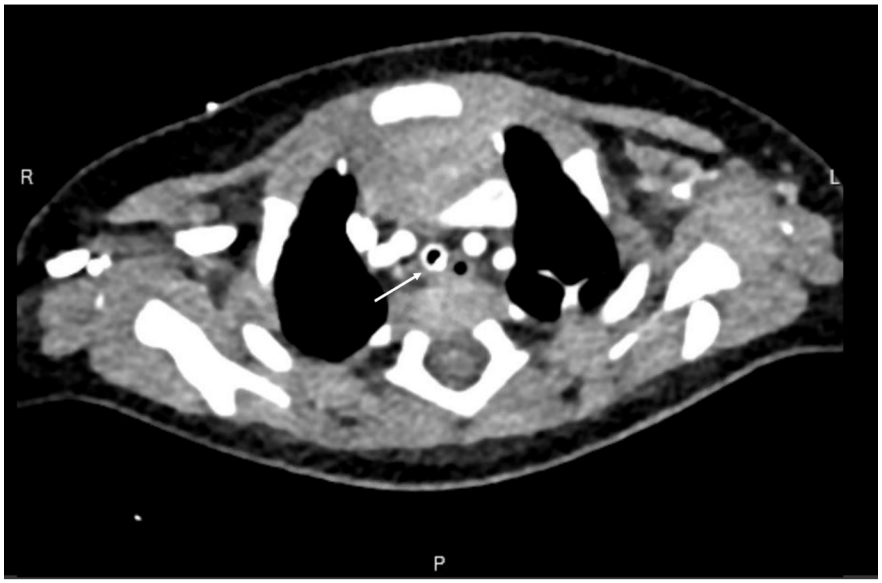


Figure 8 a) Dynamic bronchogram with digital subtraction clearly showing tracheal RUL bronchus, nar-

rowing to the carina and widely patent right and left bronchi. b) Simultaneous DynaCT of airway showing apparent narrowing of left main bronchus (artefact). Extra contrast was injected into the tracheal right upper lobe bronchus to create an alveolar gram indicating good right upper lobe supply. (the tracheal bronchus is not blind ending). LMB = left main bronchus. RUL = right upper lobe. RML = right middle lobe, RLL = right lower lobe.

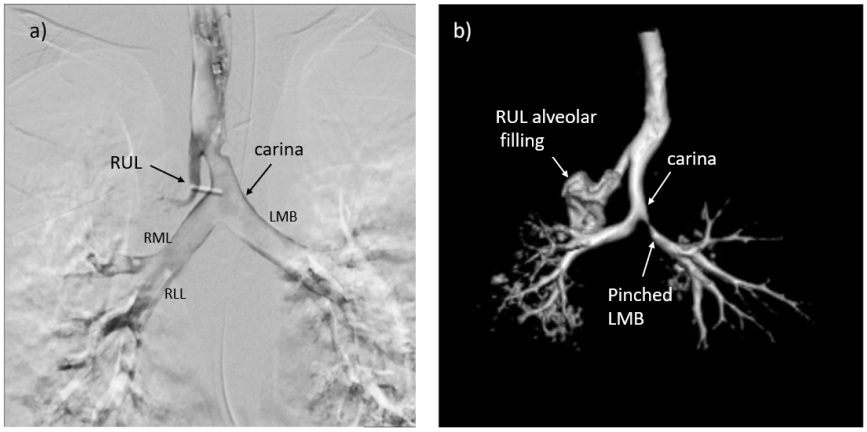


Figure 9 Typical bronchoscopic view of a) mild indentation of right lateral wall of trachea with posterior vascular compression of left main bronchus origin. b) Significant vascular compression of the right lateral tracheal wall by the right pulmonary artery as it wraps behind the trachea with cartilage distortion. Trachelais muscle is present. c) Complete rings with limited visibility of carinal bifurcation. The cartilage rings cross the midline on the floor of the trachea and there is no visible trachealis muscle.

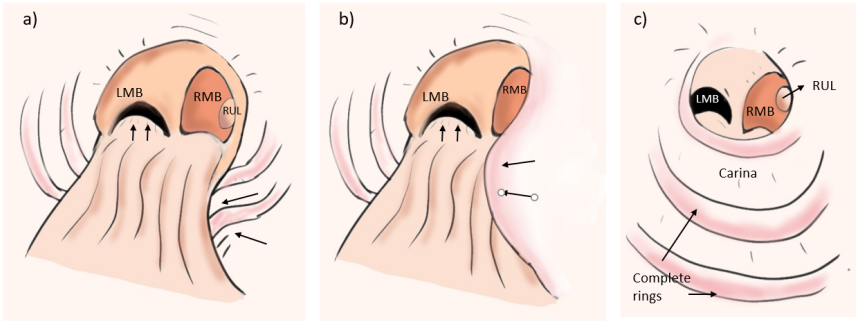


Figure 10 Gentle low pressure balloon dilation of the stenotic distal tracheal stenosis to determine if the obstruction is fixed (complete tracheal rings). Note the balloon fills easily without a waist. This allows for exact size and length of tracheal stenosis. The procedure is done via the endotracheal tube under fluoroscopy with the balloon placed over a guidewire

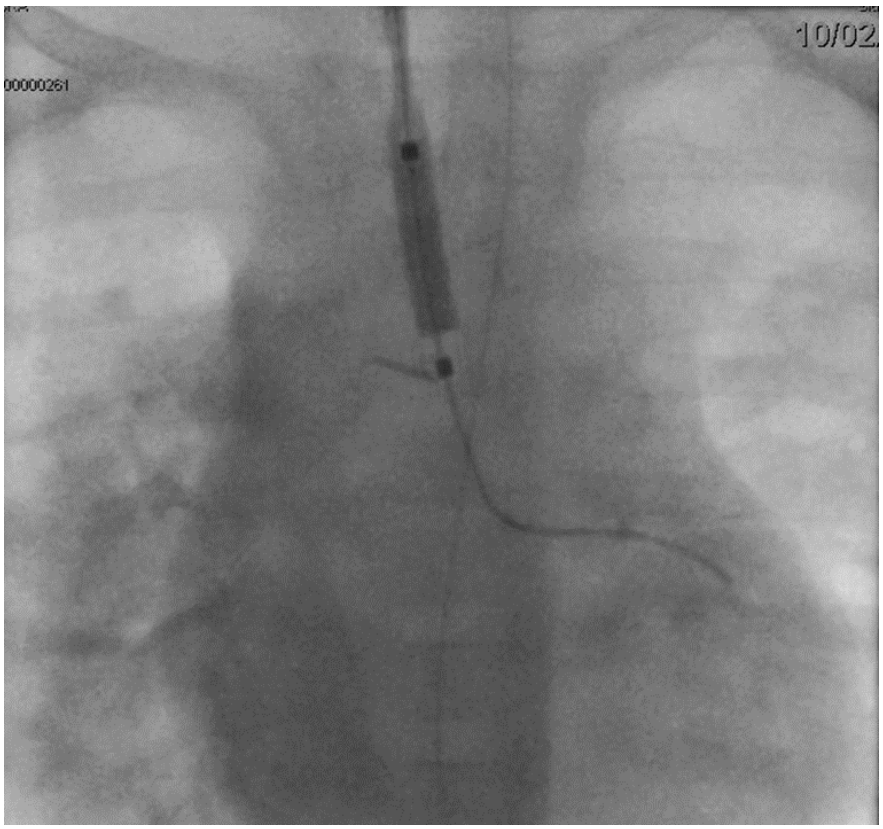


Figure 11 Rapid bedside bronchogram in a child with marked ventilation difficulties and high endotracheal tube (*) demonstrating extensive long segment tracheal narrowing (broken arrows). There is extreme narrowing at the carina with almost total loss of contrast at the carina. The contrast is dense, indicating severe tracheal obstruction with little contrast not passing through the carinal obstruction. The patient required ECMO to maintain gas exchange.

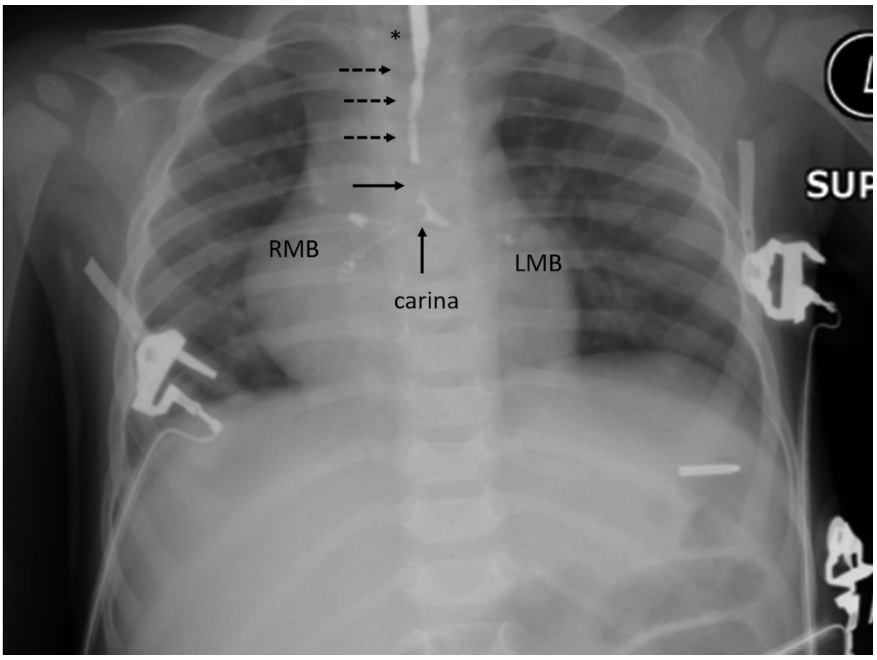
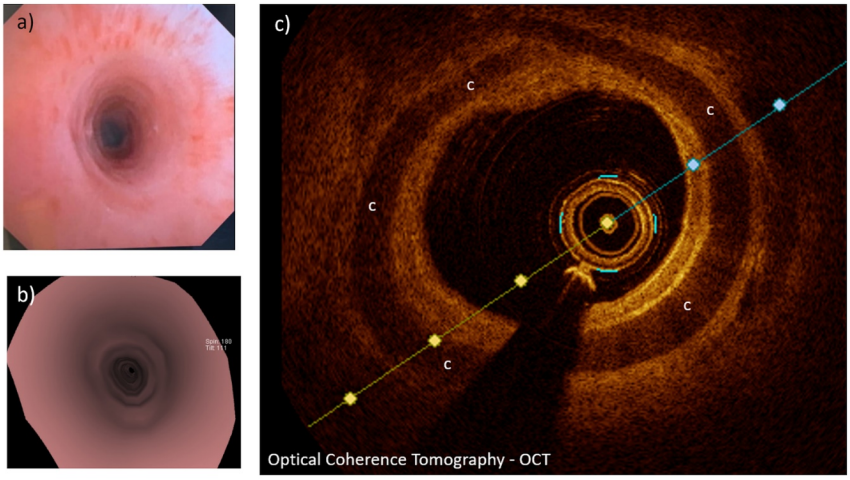


Figure 12 Bronchoscopic (a) and virtual 3D reconstructed CT images (b) of the trachea showing complete tracheal rings and long segment tracheal stenosis. Bronchoscopy is more accurate and can determine the exact number of complete rings. Image 12c shows optical coherence tomography (OCT) with a pseudo-complete ring that does not join in the midline. The cartilage is visible as a black acoustic shadow (C). Each grading dot on the OCT image is 1 mm.



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