CHAPTER 43
Tracheomalacia

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Introduction

The normal trachea is supported by up to 20 horseshoe-shaped cartilage rings completed by a posterior membranous wall. In tracheomalacia, these cartilages may be abnormally shaped, small, or even absent, with a detrimental effect on the support of the trachea. The anteroposterior (AP) diameter of the tracheal lumen becomes reduced, especially during periods of increased airflow. The dynamic movement of the malacic segment becomes most pronounced during the exertion of feeding, crying, or coughing. Symptoms can range from mild to severe, the latter culminating in complete airway obstruction. Mild cases can be managed conservatively with the expectation of spontaneous recovery, usually within the first two years of life.

Associated medical problems including gastro-oesophageal reflux and pneumonia, require aggressive treatment. More severe cases of tracheomalacia require supportive therapy, diagnostic imaging and endoscopic evaluation, and a few may require early surgical intervention to prevent acute life-threatening airway collapse.

Aetiology

Tracheomalacia may be primary (congenital absence or deformity of tracheal rings; the cause is often unknown) or secondary (in conjunction with another pathology) (see Table 43.1). The latter group includes oesophageal atresia (OA), with or without tracheo-oesophageal fistula (TOF); a vascular ring (e.g., double aortic arch); vascular compression (aberrant innominate artery or pulmonary artery sling); or extrinsic compression from another source (e.g., a mediastinal mass). It may also occur in association with prolonged positive pressure ventilation or following a tracheostomy. It is rarely seen in association with connective tissue disorders (e.g., Larsen's syndrome). Tracheomalacia commonly affects the distal third of the trachea, but can rarely extend into the bronchi. When associated with TOF in infants with OA, the malacic segment is located in the middle third of the trachea. Isolated bronchomalacia is usually associated with major cardiac pathologies and is frequently fatal.

The incidence of tracheomalacia is unknown, but it is the most common cause of expiratory stridor in infants and children. It is most often identified secondary to OA/TOF.[1, 2, 3] In affected infants, the tracheal cartilage rings fail to develop normally, especially at the site of the previously ligated fistula.[4] This has long been thought to occur as a result of extrinsic pressure of the adjacent dilated upper oesophageal pouch, although more recent evidence suggests an early embryological disturbance of tracheal development.[5]

Localised tracheomalacia secondary to extrinsic compression, from either a vascular ring (double aortic arch) or an aberrant aortic arch or pulmonary artery (PA) vessel, form a small but important group of affected infants. A double aortic arch results from persistent left and right dorsal aortic segments, compared to the normal aortic arch in which there is regression of the right dorsal aorta by week 8 postconception. The extent and location of tracheal compression is variable; therefore, so is the degree of malacia. In addition, compression of the oesophagus may present with dysphagia. A double aortic arch will compress both the trachea and oesophagus.

Table 43.1: Causes of tracheomalacia.

Primary	Cause usually unknown. Absent or deformed tracheal cartilage rings.
Secondary	Oesophageal atresia with or without tracheo-oesophageal fistula. Extrinsic vascular compression: vascular ring (e.g., double aortic arch), aberrant vessel (e.g., anomalous innominate artery or pulmonary artery sling), or (mediastinal mass). Prolonged tracheal intubation and ventilation (especially cuffed tubes). Tracheostomy. Connective tissue disorder (e.g., Larsen's syndrome).

Chronic inflammation of the tracheal cartilages occurs with prolonged intubation or following a tracheostomy. This deleterious effect of mucosal ischaemia caused by the localised pressure of the intratracheal tubing, especially with cuffed tubes, will compound airway compromise and can delay successful decannulation.

Presentation

Signs and symptoms of tracheomalacia vary from mild to severe and life threatening (Table 43.2). Many infants exhibit a simple barking cough but otherwise are not troubled by their mild tracheomalacia. For those with OA/TOF, the term "TOF cough" is frequently used to describe the characteristic sound made. Expiratory stridor indicates increasing airway obstruction. Crying, agitation, and coughing make the degree of malacic collapse more pronounced. Signs of respiratory distress, including tachypnoea and intercostal recession, herald further airway compromise, and the stridor may become biphasic. Increasing severity with infections, including the respiratory syncytial virus (RSV) infection, is to be expected, and recurrent respiratory sepsis is common.

Feeding provides particular challenges, especially in an infant with tracheomalacia following surgery for OA. Distention of the proximal oesophagus, especially with a solid bolus, may cause compression of the posterior trachea and worsen the symptoms. This is further compounded by poor oesophageal motility, anastomotic strictures, and gastro-oesophageal reflux (GOR). Feeding difficulties may lead to poor weight gain.

The most severe tracheomalacia is complicated by hypoxia and cyanosis. With major airway collapse, following a period of significant respiratory distress, complete obstruction may supervene and the infant will lose consciousness. At this stage, the collapsed airway will relax and open again, but with no guarantee that normal ventilation will resume. These events are often referred to as "dying spells" or acute life-threatening events (ALTEs). The resulting hypoxia may be severe and prolonged, leading to bradycardia, cerebral anoxia, asystole, and even death. Immediate resuscitation, often by the parents, is vital and is an indication for prompt surgical referral.

Most infants will demonstrate a gradual improvement of symptoms over the first year or two of life as the tracheal cartilages become more

Table 43.2: Signs and symptoms of tracheomalacia.

Mild	Harsh, barking TOF cough
Exacerbating events	Crying Coughing Feeding (especially food bolus) Acute distress
Moderate	Expiratory stridor Wheeze Chronic cough Recurrent respiratory infections Feeding difficulties Failure to thrive Respiratory distress (tachypnoea, intercostal recession, hypoxia)
Severe	Severe hypoxia Biphasic stridor Cyanosis Reflex apnoea (vagal stimulation) "Dying spells" or acute life-threatening events, which may be fatal.

rigid and afford better support of the airway. However, it may take many years for a TOF cough to disappear, and for some this clinical sign will persist into adult life.

Assessment

The need for investigation should be guided by the severity of symptoms demonstrated by the child. For infants who have already had surgical correction for OA, a high index of suspicion should alert the clinician to signs of developing tracheomalacia. Close observation and timely investigation are recommended. For older children presenting with significant tracheomalacia, vascular or mediastinal compression should be considered. Other conditions that may cause diagnostic confusion, including a laryngeal cleft, laryngomalacia and H-type TOF (H-TOF), should be excluded or confirmed by laryngobronchoscopy.

A plain chest x-ray is of limited diagnostic value, although it may show a mediastinal mass. A lateral chest x-ray may demonstrate localised narrowing of the trachea. Flow volume loops are able to demonstrate major airway compromise, but the impracticalities of performing them in babies and infants limit their use except in specialist research facilities.

A bronchoscopy performed under general anaesthetic is the initial investigation of choice. This will both establish the diagnosis and assess the degree and location of any airway collapse. It is important to ensure that the child continues to breathe spontaneously and does not receive intravenous muscle relaxation. A rigid bronchoscopy will allow visualisation of the supraglottic, laryngeal, and tracheobronchial tree. Flexible bronchoscopy, ideally via a laryngeal mask, provides superior assessment of any airway collapse. The AP diameter of the airway reduces during expiration, and in severe cases, the anterior and posterior tracheal walls will touch and occlude the airway entirely. The site of collapse is confirmed by a typical "fish mouth" appearance (Figure 43.1).

An upper gastrointestinal (UGI) contrast study with both AP and lateral views of the entire oesophagus is recommended. This can clearly suggest a vascular ring and may demonstrate GOR. A double aortic arch is suggested by both a right and left lateral indentation of the oesophageal outline seen in the AP view and a posterior indentation on the lateral view. This differs from the normal left-sided indentation by the normal aortic arch.

Cross-sectional imaging of the chest with computed tomography (CT) or magnetic resonance imaging (MRI) will identify either abnormal vascular anatomy or a mediastinal mass. These methods are less helpful in identifying tracheomalacia, which is a dynamic process. Vascular anomalies may require further specialist investigations.

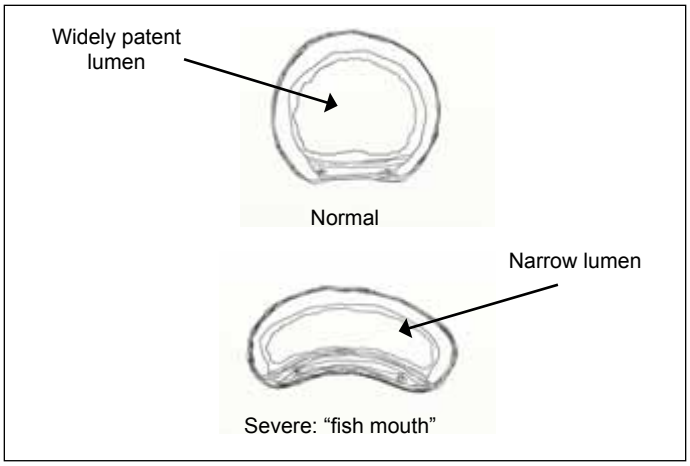

Figure 43.1: Airway in cross section showing varying degrees of tracheomalacia.

Table 43.3: Management of tracheomalacia.

Supportive	Frequent, small oral feeds NGT feeding (during times of acute respiratory infections) CPAP Intubation and positive pressure ventilation
Co-morbid pathology	Antibiotics for acute pneumonia Supplemental oxygen (pneumonia, RSV infections) Antireflux therapy for GOR
Surgical	Aortopexy Tracheostomy Correction of vascular rings or extrinsic compression (vascular, mediastinal mass) Endobronchial stenting Glossopexy Antireflux surgery (fundoplication)

Management

The management of tracheomalacia is summarised in Table 43.3. Treatment is initially focused on managing predisposing conditions. In cases of compression from a vascular ring or aberrant vessel, surgical correction may be required. This should be performed by a paediatric cardiothoracic surgeon and is tailored to the underlying vascular anomaly. Most commonly, surgical correction involves division of the smaller arch in cases of a double aortic arch, division of the ligamentum arteriosum when seen with other vascular rings, or reimplanting an aberrant vessel (typically the pulmonary artery in cases of a PA sling). However, tracheomalacia may persist or progress following correction of an underlying pathology, such as a OA/TOF.

Not all children will require intervention, especially when symptoms are mild. Appropriate medical treatment for GOR is started, and, when necessary, antireflux surgery may be undertaken. Respiratory infections require appropriate antibiotic therapy. RSV infections often require hospital admission and even respiratory support in the acute phase. Oral feeding may be problematic during this time, and nasogastric tube (NGT) supplementation may be required.

As the degree of tracheomalacia increases, conservative measures will not suffice. Supplemental oxygen may be required and should be available at home. The parents should receive resuscitation training. Adjustment of oral dietary regimens and periods of NGT feeding may be required. Support of the airway with continuous positive airway

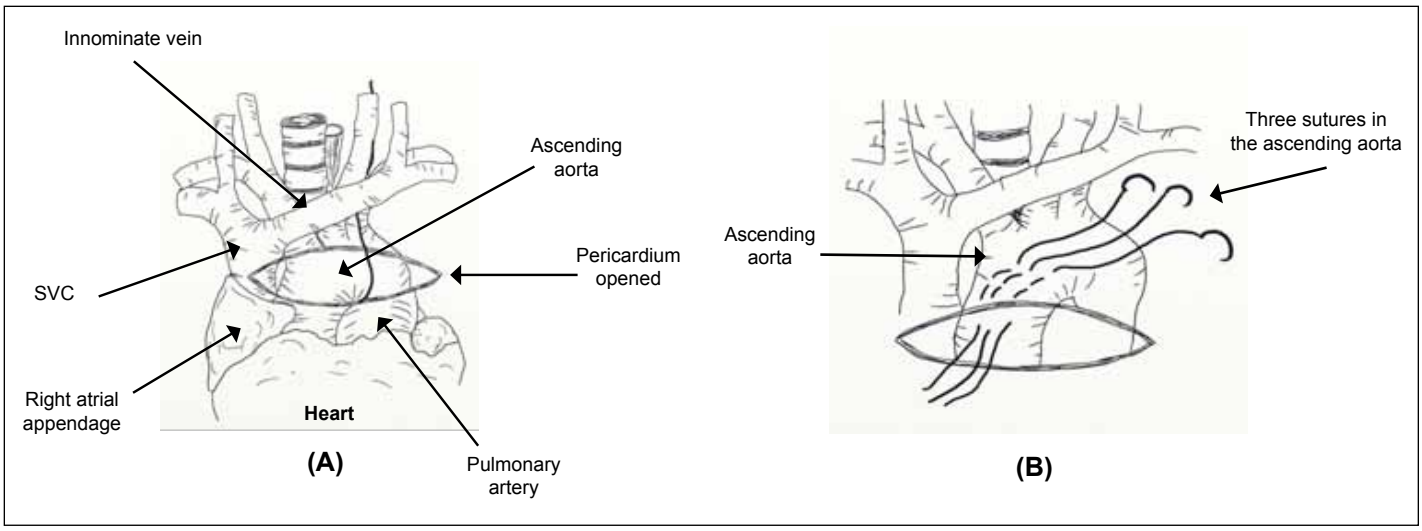

Figure 43.2: (A) Surgical approach to ascending aorta; (B) suture placement.

pressure (CPAP) may provide temporary assistance but is not suitable for long-term therapy.

Aortopexy

For severe tracheomalacia, especially for cases complicated by "dying spells" or ALTEs, and those infants who remain CPAP dependent, aortopexy offers an excellent surgical option.[6,7,8] The crucial step in an aortopexy is to ventrally suspend the ascending aorta, suturing it to the underside of the sternum, thereby creating space anterior to the trachea. Access to the aortic arch is achieved via either a median sternotomy or a left anterior thoracotomy (through the bed of the third rib), with resection of the thymus gland. Three nonabsorbable Prolene™ sutures are placed in the wall of the ascending aorta, each suture taking bites of the vessel from its intrapericardial segment to the innominate artery. These sutures can be passed through the infant sternum or sutured to its deep periosteum. The assistant depresses the sternum as the sutures are tied with minimal tension (Figure 43.2). Complications from surgery include bleeding from major vessel injury and phrenic nerve damage with subsequent ipsilateral diaphragm paralysis. Alternatively, a low cervical skin crease incision with a manubrial split affords excellent access for surgery under direct vision, with improved cosmesis.[9]

The surgical approach to aortopexy now includes thoracoscopy, with repair of the primary OA/TOF having already been undertaken endoscopically.[10] It has also been employed in aortopexy undertaken for vascular compression.[11]

In specialist cardiothoracic units, short segments of tracheomalacia may be resected and a primary anastomosis performed.

Glossopexy may offer an alternative surgical approach. This serves to anchor the tongue forward, although aortopexy may still be required.[13]

Endoluminal Stenting

Endoluminal stenting appears an attractive treatment modality, initially arising from a need to manage malignant airway compromise in the adult population. Technology used in endovascular stenting has further advanced the techniques. Balloon-expandable metallic or silicone-type stents placed at bronchoscopy are available in some specialist units. However, they carry potentially life-threatening complications of bleeding, granulation tissue formation, luminal obstruction, and erosion into adjacent blood vessels. Removal of these stents is also hazardous but they can offer an alternate mode of management in selected cases.[12]

Outcome

Long-term follow-up of children with significant tracheomalacia is mainly derived from studying infants previously treated with oesophageal atresia. Aortopexy leads to immediate relief of symptoms in the majority of infants.

Aortopexy may be required in up to 10% of infants following repair of OA/TOF, at a median age of 7 months. Ninety-five percent of these cases have resolution of their symptoms, although almost half require antireflux surgery (fundoplication) for severe reflux.[6] Overall, aortopexy affords good symptomatic improvement in such infants, with indications for surgery being "dying spells", inability to be extubated, expiratory stridor, and recurrent pneumonia.[14] When aortopexy fails, insertion of an airway stent or a tracheostomy may be required.

Evidence-Based Research

Tables 43.4 and 43.5 present case reviews involving management of tracheomalacia by aortoplexy.

Table 43.4: Evidence-based research.

Title	Management of tracheomalacia by aortopexy
Authors	E M Kiely, L Spitz, and R Brereton
Institution	The Hospital for Sick Children, Great Ormond Street, London, UK
Reference	Pediatr Surg Int 1987; 2:13–15
Problem	The problem is symptomatic tracheomalacia in infants with congenital tracheo-oesophageal anomalies. Indications for surgery included respiratory distress, recurrent apnoea, cyanosis or "dying spells", worsening stridor, or repeated hospital admissions for respiratory infections
Intervention	Aortopexy
Comparison/ control (quality of evidence)	Case review (level 4). A review of 210 infants with tracheo-oesophageal anomalies admitted over a six and a half year period. Twenty-five infants underwent an aortopexy, 22 having had repair of an oesophageal atresia and three who had primary tracheomalacia.
Outcome/ effect	Seventeen infants had immediate and dramatic relief of symptoms, and the other five were greatly improved. The operation failed in one patient.
Historical significance/ comments	Aortopexy had previously been described as a surgical option for the treatment of symptomatic vascular compression of the trachea. This was the first description of this surgical procedure for patients with congenital oesophageal anomalies. It demonstrated an excellent outcome from aortopexy for children with significant tracheomalacia, and recommended early surgery.

Table 43.5: Evidence-based research.

Title	Aortopexy for tracheomalacia in oesophageal anomalies
Authors	Corbally MT, Spitz L, Kiely E, Brereton RJ, Drake DP
Institution	The Hospital for Sick Children, Great Ormond Street, London, UK
Reference	Eur J Pediatr Surg 1993; 5:264–266
Problem	The problem is significant symptomatic tracheomalacia in association with repaired congenital oesophageal anomalies. Indications for surgery included recurrent apnoea/cyanosis (31), "near fatal episodes" (16), recurrent respiratory distress and infections (20), and worsening stridor (15).
Intervention	Aortopexy
Comparison/ control (quality of evidence)	Case review (level 4). A review of 48 patients over a ten-year period who underwent an aortopexy for tracheomalacia following repair of an oesophageal anomaly.
Outcome/ effect	Gastro-oesophageal reflux was also noted in 30 cases. Aortopexy cured near fatal episodes in all patients and resulted in improvement of airway obstruction in 95%. Failure in two patients was due to unrecognised bronchomalacia.
Historical significance/ comments	Aortopexy was recommended as the primary procedure of choice for significant tracheomalacia.

Key Summary Points

1. An association exists between oesophageal atresia and/or tracheo-oesophageal fistula (OA/TOF) and tracheomalacia.
2. Expiratory stridor should be investigated by bronchoscopy in a self-ventilating patient.
3. Anterior-posterior collapse of the tracheal lumen indicates severe tracheomalacia, and urgent intervention should be considered.
4. An aortopexy gives excellent results for localised tracheomalacia in association with OA and TOF.
5. Severe gastro-oesophageal reflux may be associated with tracheomalacia and may require a fundoplication.
6. Vascular anomalies associated with tracheomalacia require specialised investigations and management in a paediatric cardiothoracic unit.

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