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Great Ormond Street Hospital for Children



NHS Trust

ENT on ITU

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Associated clinical guidelines/protocols:

- Tracheostomy
- ENT guidelines – gosh intranet

Fundamental Knowledge:

List of topics relevant to PIC that will have been covered in membership examinations.

They will not be repeated here.

Anatomy:

- Upper airway and oropharynx
- Embryonic development of the upper airway.

Information for Year 1 ITU Training (basic):

Year 1 ITU curriculum

ITU management of the following Clinical Conditions:

- Tracheobronchomalacia
- Subglottic stenosis including laryngeal reconstruction/cricoid split
- Indications for MLB
- Laryngeal clefts

Investigations:

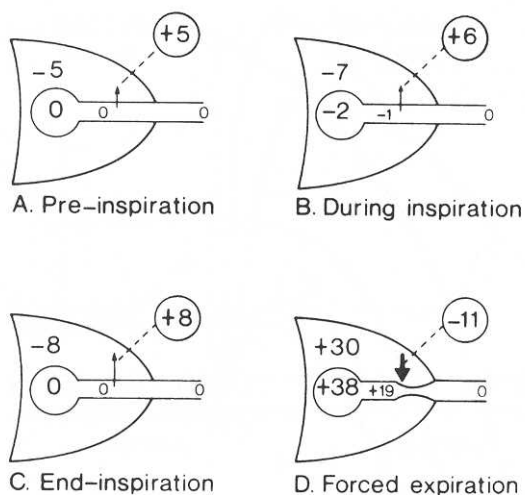
- Bronchoscopy
- Bronchogram

Curriculum Notes for Year 1:

Tracheobronchomalacia

Tracheobronchomalacia (TBM) is a condition of dynamic airway collapse during expiration. The diagram from West's textbook of respiratory physiology explains the airway dynamics during respiration and why collapse occurs in expiration.

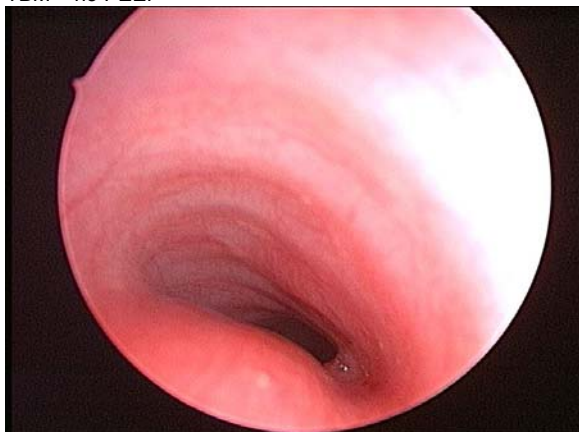
TBM can be congenital or acquired, an isolated lesion or associated with other abnormalities e.g. prematurity, gastroesophageal reflux, tracheoesophageal fistula, laryngotracheoesophageal clefts or tracheal stenosis. The exact pathophysiology is unclear. In preterm babies it is associated with ventilation at higher averaged mean airway pressures in the first week of life, longer duration of intubation and lower gestational age.



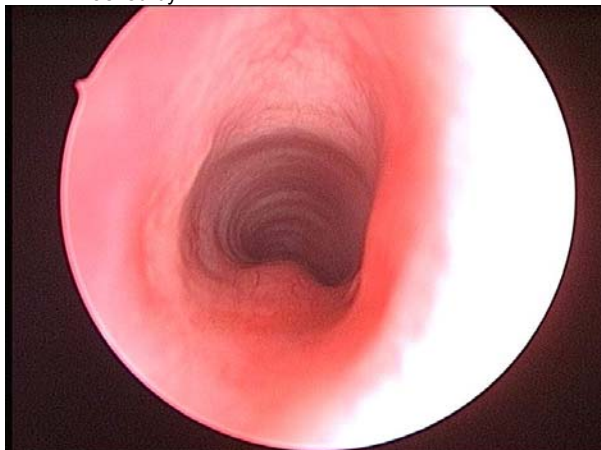
Dynamic compression of airways pg 109-113. In: Respiratory Physiology – the essentials. Ed John B West. Williams and Wilkins

Diagnosis requires dynamic assessment of the airway during various phases of spontaneous respiration and the demonstration of airway collapse during expiration. Various investigation modes are used. Bronchoscopy allows direct visualisation of the airway, but malacia may be masked by positive pressure, leading to a false negative result.

TBM – no PEEP



TBM – masked by PEEP



GOSH Patient. Reproduced with permission.

Tracheobronchography (TBG) is therefore the preferred investigation in GOSH especially in neonates with borderline respiratory function, as this can be performed without the need for respiratory support. Any pressure support during the investigation may mask malacia and we showed that the diagnosis can be missed with bronchoscopy alone. TBG also defines distal malacia better than CT and allows determination of opening pressures, which aids in the management decision. We also showed in a study that TBG was far superior compared to CT as it was associated with a much lower radiation dose.



PEEP 0



PEEP 10 cmH₂O

GOSH Patient. Reproduced with permission.

In isolated tracheomalacia e.g. in tracheoesophageal fistula aortopexy is successful in preventing life threatening obstructive episodes. However management is more difficult if the malacia is extensive or associated with other abnormalities. Stenting with CPAP or an internal tracheal stent improves dynamic compliance and reduces airway resistance. The outcome is good especially in preterm babies, where malacia resolves with growth and time. However this can take months, depending on the severity of malacia. The group in Australia suggested that patients who were ventilated for more than 3 weeks or who had any degree of bronchomalacia had a 100% mortality. However review of our patients showed that the prognosis is not associated with the duration of ventilation but with the underlying pathology. We showed that it was worse in the patients with associated complex cardiac lesions and syndromes (see below).

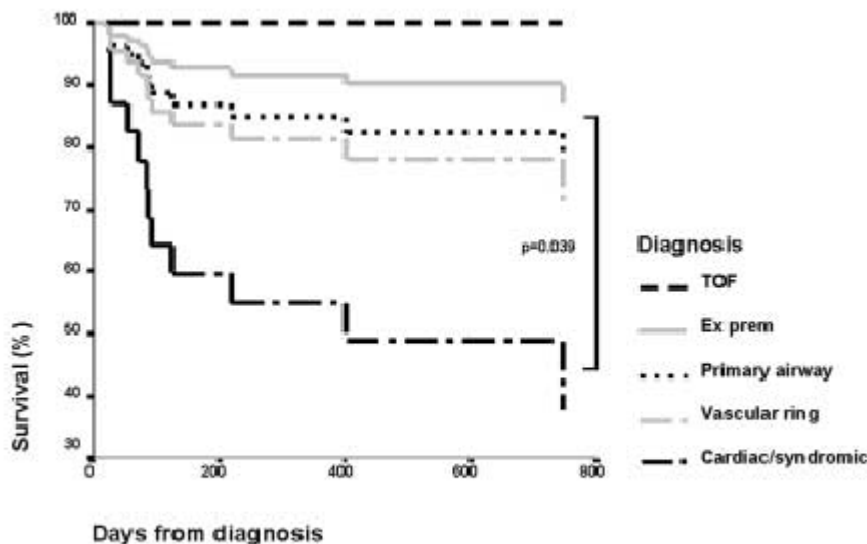
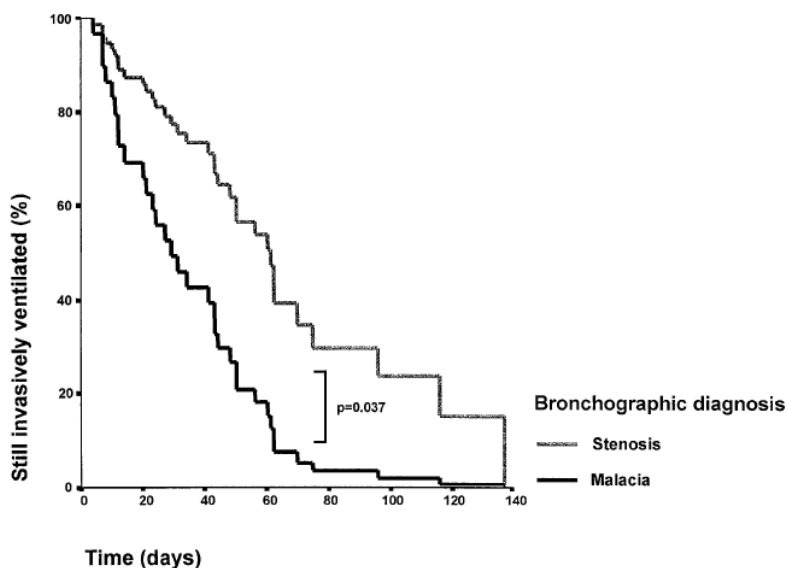


Fig.1 Survival curves of different diagnostic groups

Inwald DP et al. Intensive Care Med 2001; 27(4): 722-729.

Patients who had tracheomalacia associated with tracheal stenosis also required longer duration of invasive ventilation than those with isolated malacia (see below). However various innovations in management have improved the outcome of the patients with tracheal stenosis (see later) and fewer are needing tracheostomies or long term ventilation.

Fig.2 Length of invasive ventilation by bronchographic diagnosis (survivors only)



Inwald DP et al. Intensive Care Med 2001; 27(4): 722-729.

Subglottic stenosis

This can be congenital or acquired. Subglottic stenosis is a known complication of prolonged intubation because of the anatomy of the paediatric airway, with the narrowest part being the cricoid ring. This is especially likely with shouldered ETTs (e.g. Coles tubes), if the ETTs are snug fitting or if there are repeated re-intubation events. It can also occur because of infection and inflammation of the airway, as seen with recurrent reflux. A study in the late 1980s reported that two thirds of preterm babies with chronic lung disease have an additional airway problem. The current estimate of the incidence of subglottic stenosis is less because of awareness of the problem and the increased use of nasal CPAP.

The Myer – Cotton grading is widely used to classify the stenosis, depending on the severity of the obstruction.

1. Grade I 0 – 50 % obstruction
2. Grade II 51 – 70 % obstruction
3. Grade III 71 – 99 % obstruction
4. Grade IV 100% obstruction

Cricoid split is the preferred surgical option in grade I stenosis. However, for the procedure to be successful, it is important that the larynx and trachea are otherwise normal and pulmonary function is adequate. Laryngotracheal reconstruction (LTR) using a cartilage rib graft to expand the airway lumen is the surgical option in moderate (grade II to III) stenosis. In severe stenosis both anterior and posterior grafts may be required. The airway is subsequently stented open for a period of 5-7 days with endotracheal intubation until healing occurs. It is essential that the ETT is fixed securely to avoid accidental extubation and to minimise tube movement. Developmentally appropriate children can be managed without physical or pharmacological restraints with fewer adverse events, and a shorter length of PICU and hospital stay. Antibiotics are commenced intra-operatively and continued for 7 days postoperatively. Anti-reflux therapy is routinely prescribed for 6 weeks. Steroids are started to cover extubation to reduce the need for reintubation which may cause displacement of the graft. A tracheostomy may be required while awaiting definitive surgical repair or when initial repair has failed.

Indications for MLB

This is requested via the ENT team when there are concerns about the upper airway. These are performed in theatre by the ENT surgeons using a rigid Storz bronchoscope. The Storz scope tends to have a larger internal diameter than the flexible scopes, and the patient can be ventilated through the instrument if necessary.

The indications include:

- Recurrent stridor
- Foreign body aspiration
- Failure to extubate
- Suspected vocal cord palsy
- Suspected compression of the airway
- Suspected upper airway abnormalities
- Evaluation of the airway in patients with tracheostomies

The rigid scope is used for assessment of the upper airway while the flexible bronchoscope is needed for assessment of the lower airways.

Flexible bronchoscopy

Bronchoscopy should only be performed by trained practitioners, as the fiberoptics can be easily damaged and they are very expensive to repair or replace. The bronchoscopes in GOS are therefore only available to consultant members who are registered as bronchoscopists, and who have acquired and maintained these skills. The scopes need to be booked out

through the staff in the angio suite. There is a strict protocol for transporting, cleaning and storing the bronchoscopes.

The 2.8, 3.6 and 4.9 scopes have a suction channel and can be used for bronchoalveolar lavage or for instilling fluids or oxygen. The 2.2 scope does not have a suction channel and can only be used for inspection of the airway. It is important that the user is aware of the external diameters of the different scopes available, as well as the internal diameter of the ET or tracheostomy tube, so that they are not damaged while being passed through an ETT.

Scopes available

2.2 scope	needs	ETT size \geq 3.5
2.8 scope		ETT size \geq 4.5
3.6 scope		ETT size \geq 5.5
4.9 scope		ETT size \geq 6.5

Bronchogram

This is arranged with the interventional radiology department and is done in the angio or fluoroscopy suite. It is important that the patient is not heavily sedated or muscle relaxed as a dynamic assessment of the airway is necessary to ensure that malacia is not masked by positive pressure ventilation (see bronchograms above).

Water-soluble non-ionic contrast is injected into the endotracheal tube via a 4-French nasogastric tube inserted through an airtight connector. Aliquots of approximately 0.2 mL are used, to a maximum of about 1.0 ml. The ETT is withdrawn into the upper trachea, using fluoroscopic guidance. The images are acquired in posteroanterior and lateral projections, to ensure that airway collapse is detected. Images are obtained with different levels of continuous positive airway pressure, with the patient breathing spontaneously above this level of CPAP throughout the study. The opening pressure is the level of CPAP above which the malacia is overcome.

Other sources of information:

Tracheostomy Clinical Nurse Specialists Jo Cooke and Alex Kennett ext 8257, bleep 0712

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Tracheobronchomalacia

Dynamic compression of airways pg 109-113. In: Respiratory Physiology – the essentials. Ed John B West. Williams and Wilkins

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CM Bailey. Current approaches to the investigation and surgery of laryngotracheal disorders in children. In: *Recent advances in Otorhinolaryngology* Ed David Moffat. Churchill Livingstone

Jacobs BR, Salman BA, Cotton RT, Lyons K, Brill R. Postoperative management of children after single-stage laryngotracheal reconstruction. *Crit Care Med* 2001; 29: 164-168

Information for Year 2 ITU Training (advanced):

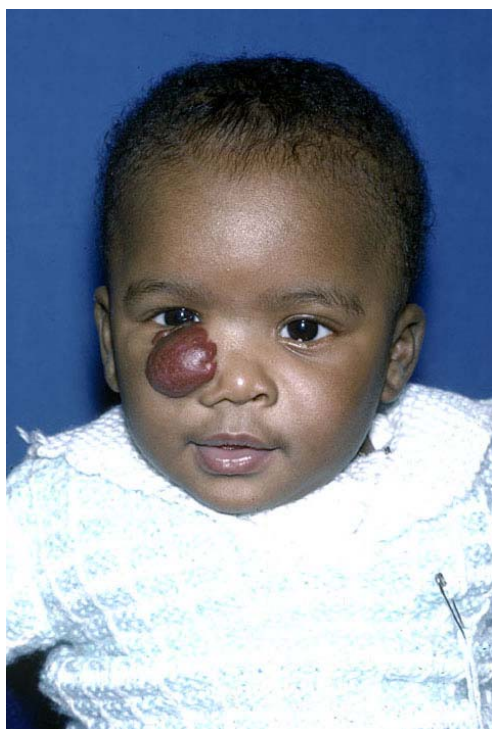
Year 2 ITU curriculum
ITU management of the following Clinical Conditions:

- Airway Haemangioma
- Tracheal stenosis
- Vocal cord paralysis.
- Retropharyngeal abscess

Curriculum Notes for Year 2:

Airway haemangioma

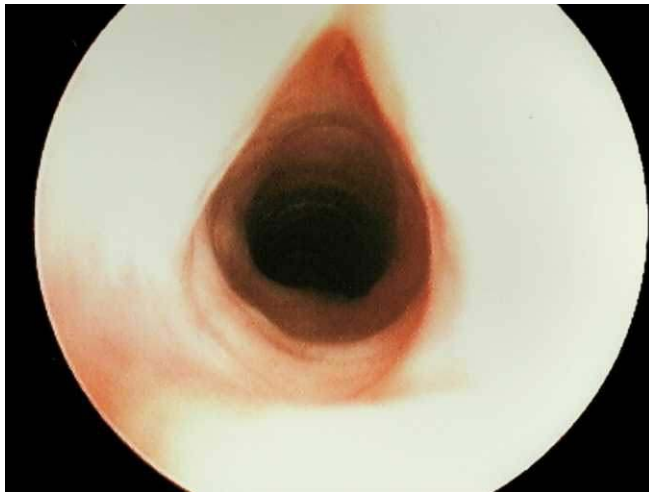
Consider this as a cause of stridor, especially when there are other haemangiomas elsewhere in the body. This can be treated successfully with intralesional steroid injection, and may require a period of endotracheal intubation to stent the airway. Systemic steroids can also lead to a reduction in the size of the haemangioma, and therefore the problem may be masked by the use of steroids for "croup". Smaller lesions can be treated with laser ablation while tracheostomy may be required for severe or extensive haemangioma. Multimodality treatment may be needed for recurrent or persistent lesions. Treatment with interferon-alpha as an anti-angiogenic agent is now not recommended especially in infants because of serious associated complications (neurotoxicity and spastic diplegia) occurring in 2.5 - 6% of cases treated. Extensive haemangiomas may require involvement of and management by a multidisciplinary team (dermatology, interventional radiology, surgery, ENT, oncology and ICU).



GOSH Patient. Reproduced with permission.

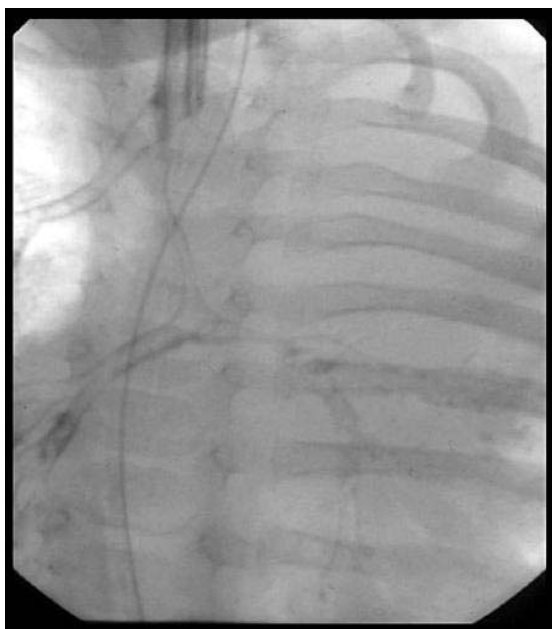
Tracheal stenosis

This is often associated with cardiac abnormalities especially a pulmonary artery sling, causing extrinsic airway compression and abnormal development of the airway. An echocardiogram is therefore important as part of the work-up. Complete tracheal rings are usually seen on bronchoscopy. Tracheobronchogram may show the typical T-shaped carina.



Complete tracheal rings on bronchoscopy

GOSH Patient. Reproduced with permission.



Tracheobronchogram showing typical T shaped carina

GOSH Patient. Reproduced with permission.

The management depends on:

1. The narrowness of the trachea
2. The length of involvement of the trachea. The term long segment tracheal stenosis is used if there is involvement of more than two thirds of the trachea (LSTS)
3. Involvement of the bronchi
4. Presence or absence of complete tracheal rings

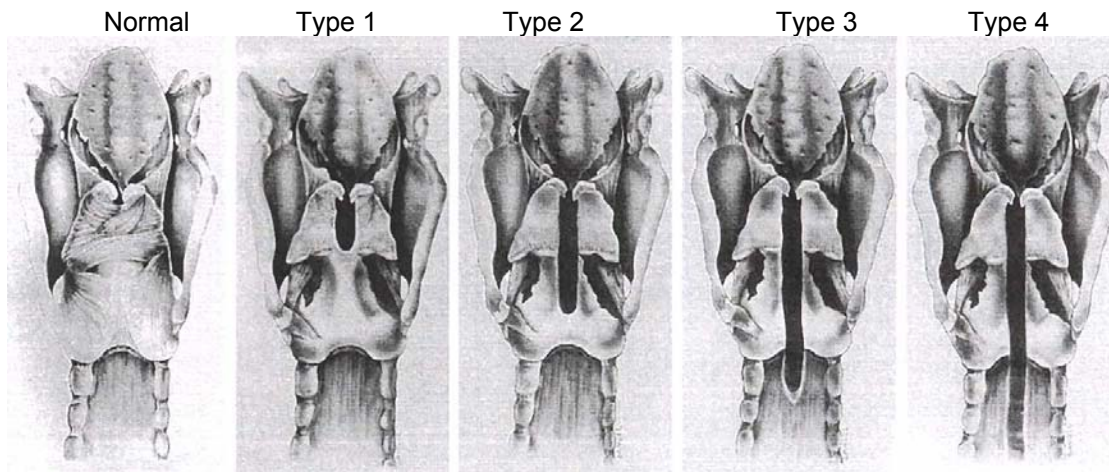
Short segments of tracheal stenosis may be resected or balloon dilated. Slide tracheoplasty is now the preferred surgical option for LSTS, as patch tracheoplasty using pericardium has been associated with more long term problems. There is often associated tracheobronchomalacia, which may be amenable to stenting. Difficult cases are discussed with the multi-disciplinary trachea team. The outcome is improving with multi-disciplinary team working, and the duration of intubation and length of hospital stay has reduced.

Laryngeal Clefts

In the embryo the separation of the respiratory system from the primitive foregut starts at about day 20 of gestation. Shortly after, the tracheoesophageal septum develops in a caudal to cranial direction and the cricoid cartilage fuses between 6 and 7 weeks in utero. Failure of the posterior cricoid lamina to fuse and incomplete development of the tracheoesophageal septum results in varying degrees of communication between the airway and oesophagus. This rare congenital abnormality is often associated with other congenital abnormalities, especially cardiac and gastrointestinal defects. Up to 50% of Opitz G syndrome children, where an autosomal dominant trait results in various midline defects, have a laryngeal cleft. The diagnosis requires a high index of suspicion and is made by microlaryngobronchoscopy.

Classification of Laryngotracheoesophageal clefts (Benjamin and Inglis)

1. Type 1 supraglottic interarytenoid cleft
2. Type 2 partial cricoid cleft extending below the level of the vocal cords
3. Type 3 total cricoid cleft extending into the cervical trachea
4. Type 4 cleft extending into the thoracic trachea



Benjamin B, Inglis A. *Ann Otol Rhinol Laryngol.* 1989;98:417-20

Successful treatment depends on early diagnosis, securing the airway, and protecting the tracheobronchial tree from aspiration. The management depends on the extent of cleft. The overall mortality is 14 – 46 %, although it is much higher in type 4 cleft (60 – 93 %). The morbidity and mortality is reduced by patients being managed in multidisciplinary teams. The incidence of revision surgery also increases with the severity of the cleft. Postoperative complications include reflux due to oesophageal dysmotility and microgastria, recurrent tracheoesophageal fistula at the suture line of the repair and severe tracheobronchomalacia.

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