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Welcome to Volume 10 Issue 1 of Journal of ENT Masterclass® 2017

I very much welcome our readers to this edition of the Journal of ENT Masterclass®. We have two events to celebrate this year. It is the 10th birthday for the Journal which has continued to provide high quality up-to-date educational material free of charge. In this 10th edition, I am very happy to present you with articles that cover a breadth of subjects which we have not covered before. Latest developments such as auditory brainstem implants and other cutting edge topics have been covered. Detailed reviews on management of known conditions are also there to update our knowledge.

We very much appreciate the immense efforts of our editors and authors to present you with such comprehensive material and are certain it would be of great benefit.

We proudly celebrate our founding editor Shahed Quraishi's Royal appointment as an Officer of the Order of the British Empire (OBE) in recognition of his role in medical education and the NHS. In 2005, Mr Quraishi started with a local revision course that was held in Doncaster. The course was named “ENT Masterclass” and there was something special about it. It was the first ever full course to be provided to trainees free of charge.

Years have gone by and not even in our wildest dreams we would have expected it to be what it is today. Not only that the main course continued to date but other courses on the same theme were created. Now we run a Consultants’ Masterclass, GP Masterclass, Nurses Masterclass, Thyroid Masterclass…and more. More so, the course has been picked up internationally and we have run it in Sydney, Delhi, Mumbai, Jeddah, Cape Town, Hong Kong, Berlin and Beijing. Next editions are planned in Karachi, Lausanne, Paris…and more to come.

I would specially like to express my sincere gratitude to our editors and authors who selflessly continued to support the journal for all these years and got it to its 10th anniversary. The journal was officially accredited by ENTUK in 2016 and is now distributed free of charge to all members of ENUK. I am confident that the Masterclass platform is there to stay as an outstanding unique venture that will continue to support doctors in all corners of our planet.

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ENT aspects of cystic fibrosis management

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Abstract
Cystic Fibrosis occurs in approximately 1 in 2,500 births although the incidence varies between countries according to the frequency of healthy autosomal recessive carriers in the local population. Respiratory symptoms are the most important cause of morbidity and there is an important relationship between the occurrence of upper airway disease and lower respiratory infection. The presence of polyps should raise concerns about the possibility of cystic fibrosis and should prompt referral for appropriate diagnostic testing. Active management of sinus infection might significantly impact the occurrence of lower respiratory disease although the optimal approach to achieving this is not yet clearly defined. Functional endoscopic sinus surgery to clear reservoirs of infection from the upper airway and wide opening of the maxillary antra to ensure better ongoing drainage are appropriate measures to achieve improvements in symptoms amongst those with significant disease morbidity. New genotype specific precision medicines are likely to have a positive impact on disease progression including the occurrence of upper airway problems.

Key words
Cystic Fibrosis, Polyps, Sinusitis, Endoscopic Sinus surgery

Introduction
Whilst lung disease is the most important cause of morbidity and early mortality amongst individuals with cystic fibrosis (CF), this disease can also cause significant early mortality amongst children with cystic fibrosis (CF) although, as discussed below in the imaging section, one of the consequences of the persisting mucosal inflammation is the formation of polyps, although the extent to which this occurs is highly variable between patients. Whilst CF polyps are histologically similar to polyps in non-CF patients, there are several characteristic features which are more peculiar to CF. These include a thin basement membrane, less eosinophilic infiltration and a preponderance of more acid staining mucin.

There should be a high index of suspicion for CF in any young person presenting with nasal polyps as these are rare outside of the CF population in this age group. In older patients there is a wider range of underlying inflammatory conditions that can lead to polyps including allergic rhinitis and asthma, primary ciliary dyskinesia, Churg-Strauss syndrome and aspirin and alcohol intolerance. CF should always be considered if there are accompanying lower respiratory symptoms such as chronic wet sounding cough. Sinusitis alone is rarely a presentation of ongoing inflammation in the upper airway but does not provide the necessary bony detail for surgical planning and so its use is far more limited.

Pseudomonas aeruginosa, Haemophilus influenzae and Staphylococcus aureus are the three most commonly grown pathogens from sinuses. Genotyping studies of these bacteria indicate that these are often the same clonal species as those found in the lower airway. Fungi are also commonly cultured from the sinuses. The relevance of these organisms is less certain, but there is a move towards more aggressive treatment of fungi when isolated from sinus aspirates.

Diagnosis
The gold standard diagnostic test for CF is the sweat test and a sweat chloride of >60mmol/L is diagnostic with few reasons for a false positive result. Unfortunately some individuals and in particular older patients will have equivocal results between 30-60mmol/L that are best referred to CF services for clinical interpretation and further investigation to determine whether the patient has CF related pathology. Curiously, CF database analyses have shown that the presence of nasal polyps as reported in registry data is associated with better lung function although the reasons for this are unclear. Paradoxically it has also been reported that the presence of polyps is also associated with a higher frequency of infection with Pseudomonas aeruginosa.

Airway examination
It is quite common to see swollen turbinates on initial clinical examination. These are sometimes mistaken as polyps by the inexperienced physician. There might be nasal discharge and the more pathognomic pale glistening polyps which can occasionally prolapse out of the nasal cavity (Figure 1). More detailed endoscopy might reveal further polyoidal disease obstructing the nasal cavity and the sinuses. There might also be bulging of the ucinatic process causing further airways obstruction. The prevalence of polyps varies between reported studies. One prospective study in which all CF patients in a paediatric centre underwent detailed endoscopic examination found polyps in 19% of children aged up to 6 years rising to 45% among adolescents. Surgery was only deemed to be justified in less than half of these patients. In an adult CF centre in Ireland, polyps were only identified in 20% of patients.

It is always good practice to carry out a complete upper airway evaluation to look for adenotonsillar hypertrophy as an additional cause of airways obstruction although this is no more common amongst CF patients. Patients with Primary Ciliary Dyskinesia typically have chronic secretory otitis media with dilated blood vessels overlying their tympanic membranes but in general CF patients have little in the way of middle ear problems.

Imaging studies
Plain sinus X-Rays invariably show extensive opacification and are of little value in directing management. Although there are legitimate concerns about radiation exposure, a CT scan provides more useful detail about the extent and nature of the disease process and is the radiological investigation of choice (Figure 2). Whilst not recommended as a routine investigation for all patients, CTs are a necessary prerequisite to surgery in those individuals with significant symptoms despite medical management. Characteristic features less commonly seen in sinuses arising from other causes include agenesis of the frontal sinuses, hypoplasia of the maxillary and antral sinuses and medial bulging of the lateral nasal wall which can cause facial deformity. MRI imaging can be used to locate sites of ongoing inflammation in the upper airway but does not provide the necessary bony detail for surgical planning and so its use is far more limited.

Microbiology
Pseudomonas aeruginosa, Haemophilus influenzae and Staphylococcus aureus are the three most commonly grown pathogens from sinuses. Genotyping studies of these bacteria indicate that these are often the same clonal species as those found in the lower airway. Fungi are also commonly cultured from the sinuses. The relevance of these organisms is less certain, but there is a move towards more aggressive treatment of fungi when isolated from sinus aspirates.

Figure 1: Nasal endoscopy showing nasal polyps

Figure 2: CT scan showing extensive mucous filled ethmoid sinuses.
sputum samples which might be relevant to treating these organisms when found in the upper airway.

**Treatment**

Medical management includes the use of nasal douching, appropriate courses of oral antibiotics and empirical use of topical nasal corticosteroids. Concomitant seasonal allergic rhinitis is common amongst individuals with CF and should be enquired about and treated accordingly. Nasal irrigation with warmed saline solutions can be well tolerated even amongst young children and might be of benefit in early disease as well as part of post-operative management to optimise the long-term benefits of endoscopic surgery.

In recent years, there has been interest in the possibility that the upper airway and the sinuses in particular, are an important site for harbouring CF pathogens. There is accumulating evidence that bacteria including Pseudomonas aeruginosa are able to persist and change phenotypically to a biofilm mode of growth within the sinuses. This reservoir of organisms might then seed the lower airway with more difficulty to treat resulting in chronic lung infection and more rapidly progressive lung damage10. This has caused some CF centres to treat sinus infection more aggressively either on the basis of empirical antibiotic choices or the results of cultures from sinuses aspirates. Neubulisers have been adapted for nasal deposition of antibiotics although the extent to which these drugs are able to effectively penetrate the sinuses is uncertain11. The highly regarded Copenhagen CF centre in Denmark has led on a more aggressive approach using endoscopic sinus surgery in many of their patients. Their data suggests that this strategy resulted in a delayed onset of lower airways infection with gram-negative bacteria and impacted positively on lung function12. Whilst there is a growing rationale for these more invasive strategies, they have resulted in only limited success in some reported cases so far13.

Outcomes such as pain, discharge, fever or postnasal drip. This is thought to occur as a result of endonasal topical douching, creation of large drainage channels into the maxillary antra. Post surgical infection with tobramycin has been recommended by some centres to help reduce the microbial load and increase the time to recurrence of sinus problems.

Sinus surgery has been performed in those patients with severely impaired lung function as part of optimisation prior to heart lung transplantation to try and prevent recolonisation of lung grafts with bacteria such as Pseudomonas14. Unfortunately this approach does not appear to offer any advantages in terms of subsequent infection or survival benefit. More recently it has been suggested that sinus surgery performed post transplantation and accompanied by daily nasal douching can confer a survival advantage as a result of a reduction in post transplant pseudoamonia acquisition and an associated decrease in the occurrence of broncholiths obliterans syndrome15.

**Future Considerations**

There is currently great excitement within the CF community in response to clinical trials of new medications treating CF at a protein expression and function level. A surgical approach to managing patients with significant upper airway symptoms is widely accepted as good clinical practice. Systematic reviews of endoscopic surgery for sinus disease have identified good evidence for relief of upper airway symptoms although little evidence for impacts on lower respiratory status16. One important indication for operative intervention is when there is evidence of expansive polypos in the ethmoid sinuses resulting in widening of the nasal bridge and hypertelorism. This can occur with little else in the way of symptoms and should be treated by endoscopic sinus surgery.

Whenever surgery is contemplated careful consideration should be given to the risks of prolonged general anaesthesia on the lower airways. Pre-operative intravenous antibiotics might be necessary to optimise lung function and the input of a CF physiotherapist post-operatively is essential to minimise as far as possible any respiratory deterioration. A full blood count and clotting studies should be performed pre-operatively given the risks of CF related liver disease and/or hypersplenism in relation to haemostasis.

Simple polypectomy can provide short-term relief, but recurrence is common and more definitive interventions are likely to result in more long-term benefit. Surgery might also provide careful enucleation of the bony components within the ethmoid labyrinth and the creation of large drainage channels into the maxillary antra. Post surgery infection with tobramycin has been recommended by some centres to help reduce the microbial load and increase the time to recurrence of sinus problems.

The extent to which these treatments might correct the abnormalities within the upper airway is not yet understood, but one study in patients with the so-called G551D mutation treated with the corrector drug Ivacaftor had significant improvements in radiological evidence of sinus disease within 6 months of therapy17. Other drugs in the pipeline offer great potential for similar effects in patients with more common gene mutations18.

**Conclusion**

Extensive clinical experience has made it clear that ENT input is a valuable addition to the multi-professional team management of the complex multifaceted problems encountered in CF patients. Early medical treatment of microbial infection of the sinuses might usefully reduce the microbial load of the upper airway and its impact upon lower airways infection. Evidence would suggest that more definitive surgical treatment achieves significant benefit in those with more advanced symptomatic disease. Sinus disease tends to be recurrent, but it would appear that the time to recurrence depends upon which good clearance of pus and obstruction is achieved through surgical excision. Good multi-disciplinary care with support to patients performing daily nasal hygiene procedures such as saline douches would appear to be of additional benefit in achieving more prolonged symptomatic relief.

**Conflicts of interest**

None

**References**


Paediatric swallowing disorders

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Abstract
A normal swallow is a complex co-ordinated function vital to the development of a child. Infants and children with a disordered swallow may be referred to a Paediatric Otolaryngologist working within a Multidisciplinary Team for diagnosis and intervention. This article discusses the normal swallow, aetiology, common and important causes of swallowing disorders. These conditions affecting paediatric patients are discussed with their management.


Key words
Dysphagia, aspiration, laryngomalacia, adenotonsillar hypertrophy

Introduction
Paediatric Otolaryngologists are often asked to help assess and treat a child’s swallowing problems.

Swallowing is a highly complex biomechanical function dependent on structural integrity and mature developed neuronal reflex and control of the oral, pharyngeal and oesophageal muscles. Safe swallowing is also dependent on protective sensorimotor reflexes around a patent airway.

Swallowing can be disordered as a result of malformations, poor neuronal control, congenital or acquired infection, cardiovascular or respiratory compromise, prematurity, physiological or behavioural factors or a combination of these. In addition paediatric swallowing assessment and management requires consideration of other health issues, parent-child interaction and assessment of the child’s environment.

The multifactorial aetiology of swallowing disorders mandates an interdisciplinary team for diagnosis and management of these problems. Members of this team may include those listed in Table 1. Other health professionals that have a close link with the team include the gastroenterologist, psychologist, lactation consultant, child health nurse, general practitioner (GP), occupational therapist and immunologist.

Feeding disorder vs Swallowing disorder
It is important to differentiate between a feeding and a swallowing disorder. A feeding disorder is a problem associated with eating activities that may or may not be associated with a swallowing disorder. It may include a child who has difficulty picking up their food, a child with rigid food preferences or one that is disruptive during mealtimes. An oral aversion disorder is one that is disruptive during mealtimes. An oral aversion disorder is a problem associated with eating activities that may or may not be associated with a swallowing disorder. It may include a child who has difficulty picking up their food, a child with rigid food preferences or one that is disruptive during mealtimes. An oral aversion disorder is one that is disruptive during mealtimes.

Normal swallow
Swallowing can be broadly divided into four phases as shown in Table 2.

Feeding, screening and clinical examination of swallowing in children
A multidisciplinary team is necessary to investigate and treat patients with complex swallowing disorders. At our institution caregivers complete a Feeding and Mealtime questionnaire. The children’s weights are plotted on a growth chart with comparative percentiles. A detailed history of perinatal and early feeding history follows, supplemented by the completed clinical questionnaire. Assessment should determine the phase of swallow that is of greatest concern. This requires a systematic approach to each phase asking questions relevant to the phase. For example, for the “Oral Phase” – assessment of dentition, mouth closure, chewing habits and drooling is undertaken. Each phase of swallow should be explored for clues of possible dysfunction.

If feasible, a Fibreoptic Endoscopic Evaluation of Swallow (FEES) should be conducted. This involves the child being appropriately positioned and secured. Using a fiberoptic endoscope to visualise the supraglottic and glottic structures during a swallow, can provide a detailed view of the structural abnormalities (with greater detail than what would be seen on a Video Fluoroscopic Swallow Examination). In addition it can provide feedback for the utility of compensatory exercises introduced by the Speech Therapist.

Children may also undergo a video fluoroscopic swallow (VFSS) examination. This provides an overall view of the phases of swallowing. This is assessed and scored by the Speech pathologist and radiologist. This test can be useful to determine the phase of swallow that is most concerning. It can also indicate the extent and level of aspiration; with the additional benefit of allowing the speech pathologist to make recommendations about safe consistencies for oral intake. It is important to acknowledge that VFSS and its findings do not entirely rule out aspiration. This investigation in isolation cannot illustrate the impact of the swallowing problem on the child or predict the progression or timing of resolution.

From these investigations it is important to make an assessment of the child’s safety to swallow. If all textures are consistently aspirated the parents need to be counselled that the child is not safe to swallow and an alternative route (most commonly a nasogastric tube) needs to be introduced.

One of the most useful and universally used tools in grading a Video Fluoroscopic Swallow Examination is Rosenbek’s Penetration Aspiration Scales (Table 3) as they relate closely to the definitions presented below.

Penetration: When material enters the supraglottic larynx above the vocal folds. It is usually identified on Video Fluoroscopic swallow and is an indicator of high risk for aspiration.

Aspiration: The entry of any material below the level of the vocal folds into the trachea.

Silent Aspiration: The entry of material into the trachea without any effort to expel (cough or choke) or signs of problems. This is the most concerning as parents may not know that their child is at high risk of soiling the airway.

Origin of paediatric dysphagia
The multiple aetiologies of swallowing disorders are too broad to cover in this paper but a good tabular summary is presented in Table 4.

Table 1: The interdisciplinary team

<table>
<thead>
<tr>
<th>Role</th>
<th>Department</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speech language Therapist</td>
<td>Otolaryngologist</td>
</tr>
<tr>
<td>Neurologist</td>
<td>Otolaryngologist</td>
</tr>
<tr>
<td>Dental/Oromaxillofacial</td>
<td>Otolaryngologist</td>
</tr>
<tr>
<td>Paediatrician</td>
<td>Otolaryngologist</td>
</tr>
<tr>
<td>Teacher</td>
<td>Otolaryngologist</td>
</tr>
<tr>
<td>Dietician</td>
<td>Otolaryngologist</td>
</tr>
</tbody>
</table>

Table 2: The four phases of swallowing. Adapted from Schrotter-Morasch H & Graf S, 2014

<table>
<thead>
<tr>
<th>Phase</th>
<th>Function</th>
<th>Duration</th>
<th>Control</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral preparatory phase</td>
<td>Intake, mastication, saliva, bolus, closure of the mouth</td>
<td>Variable</td>
<td>Voluntary</td>
</tr>
<tr>
<td>Oral transportation phase</td>
<td>Transportation of the bolus to the posterior oropharynx</td>
<td>&lt;1s</td>
<td>Voluntary</td>
</tr>
<tr>
<td>Pharyngeal phase</td>
<td>Triggering of the swallowing reflex</td>
<td>&lt;1s</td>
<td>Reflex</td>
</tr>
<tr>
<td>Oesophageal phase</td>
<td>Transportation of the bolus through the oesophagus by peristaltic waves</td>
<td>4-40s</td>
<td>Reflex</td>
</tr>
</tbody>
</table>

Table 3: Penetration Aspiration Scales. Adapted from Rosenbek et al, 1996

<table>
<thead>
<tr>
<th>Phase</th>
<th>Function</th>
<th>Duration</th>
<th>Control</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Material does not enter the airway</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 Material enters the airway, remains above the vocal folds and is not ejected from the airway</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3 Material enters the airway, remains above the vocal folds and is not ejected from the airway</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4 Material enters the airway, contacts the vocal folds and is not ejected from the airway</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5 Material enters the airway, passes below the vocal folds and is not ejected from the airway</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6 Material enters the airway, passes below the vocal folds and is ejected into the larynx or out of the airway</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7 Material enters the airway, passes below the vocal folds and is not ejected from the trachea despite effort</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8 Material enters the airway, passes below the vocal folds and no effort is made to eject</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
This paper will focus on the common and important aetiologies affecting infants and children:

The common causes

Infant

During the first 12 months of life there are specific orofacial motor functions that develop. These milestones are required for a coordinated safe swallow. The most common reason for infant dysphagia is laryngomalacia.

Laryngomalacia is the most common congenital anomaly of the larynx. These patients typically present with inspiratory stridor, which starts in the first weeks of life and may progress for several months. Symptoms may be present up to 2 years of age.

Examination findings consistent with laryngomalacia include collapse of supraglottic tissues on inspiration and may progress for several months. Symptoms may be present up to 2 years of age.

One of the most common causes of dysphagia in a child encountered by an ear, nose and throat (ENT) surgeon is cleft palate.

Cleft palate

Commonly children with cleft palates will have difficulty maintaining sucking pressures during swallowing. Most strategies for cleft palate babies are used to overcome the negative pressures developed during swallowing. These include:

- Haberman nipple
- Squeezing a soft bottle to help with the flow of milk
- The use of specialist feeding teams monitoring and assisting with feeding

In patients with a cleft palate and associated anomalies or syndromes the feeding difficulties are often compounded with respiratory difficulties. These should be recognised and addressed at an early stage.

Clayburgh et al demonstrated in a study that swallowing quality of life scores improved significantly after tonsillectomy in a cohort of patients with dysphagia (SWAL-QOL mean 58.4 pre to 82.4 post). Within this group significantly more patients could tolerate a regular diet and they had a significantly increased weight percentile for age. Interestingly the control cohort – who had tonsillectomy for other indications – also had a significant improvement in swallowing quality of life scores too (SWAL-QOL mean 80.8 pre to 91.7 post). There was also a significant increase in the number of patients tolerating a regular diet and increased weight percentile for age. This indicates that dysphagia may be prevalent in patients with adenotonsillar hypertrophy – even when this is not the primary problem.

Adenotonsillectomy results in a significant improvement in dysphagia in a large number of patients with dysphagia secondary to adenotonsillar hypertrophy. However the complex nature of swallowing means that not all patients will experience a complete resolution of symptoms and in these cases further investigation is indicated.

The important causes

Syndromic and Congenital

Cleft palate

Commonly children with cleft palates will have difficulty maintaining sucking pressures during swallowing. However patients with isolated cleft palates rarely have problems with the upper airways and the remainder of their swallowing phases are usually normal. Therefore once milk enters the oropharynx the swallow is normally safe and successful.

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Treatment: Most strategies for cleft palate babies are used to overcome the negative pressures developed during sucking. These include:

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In patients with a cleft palate and associated anomalies or syndromes the feeding difficulties are often compounded with respiratory difficulties. These should be recognised and addressed at an early stage.

Macroglssia

Enlargement of the tongue can cause functional issues with swallowing. The oral phase of swallow is affected. Mastication, oral closure and movement of the food bolus may be affected leading to dis-coordinate transition of the food bolus into the oropharyngeal phase. The airway may also be compromised because of glossoptosis further compromising swallow. Causes include vascular malformations, Beckwith Wiedemann syndrome and Trisomy21 (relative macroglssia).
Sialorrhoea

A significant health issue in children with neurological impairment is excessive drooling. This can be divided into anterior and posterior drooling – that is the unintentional loss of saliva from the mouth due to malfunction of the oral phase or posterior drooling, which occurs when the saliva spills over the tongue into the faucial isthmus. In a neurologically impaired child chronic salivary aspiration is a major cause of morbidity as these patients develop frequent lower respiratory tract infections. These chronic infections can then lead to bronchiectasis and worsening pulmonary function. In addition, these infections result in prolonged admission to hospital and reduction in quality of life.

Treatment: Treatment can be divided into conservative, pharmacological and surgical. Conservative treatments include behavioural modifications and dental devices. Anticholinergics can be helpful for the reduction in salivary flow as they block the muscarinic receptors. Options include glycopyrrrolate bromide or hyoscine bromide patch. A Cochrane review supports the option of Botulinum toxin injection into the salivary glands. Rarely there can be deterioration in swallow after this procedure with one proposed mechanism for this being the extravasation of the toxin into the pharyngeal muscles or pharyngeal plexus.

For patients with refractory drooling surgical options will be discussed. Surgical management includes submandibular gland excision, submandibular duct ligation and parotid duct ligation. Other options include tracheostomy and laryngotracheal separation. A study of twelve patients over seven years showed that bilateral submandibular gland excision and bilateral parotid duct ligation was effective in control. The disadvantage is that it is likely to be temporary.

Surgical options include:
- injection in the inter-arytenoid defect with a filler (calcium hydroxyapatite, hyaluronic acid and gel foam have been used). The advantage of this is it is a quick simple procedure that will give information on symptom control. The disadvantage is that it is likely to be temporary.
- Endoscopic suture repair. This is a longer more complex procedure but it has better long-term success.

Tracheal and Oesophageal

Oesophageal and tracheal diseases are a factor in dysphagia for a reasonable number of patients. Although severe cases are primarily managed by gastroenterologists and paediatric surgeons, the diagnosis of reflux disease and eosinophilic oesophagitis is often made by paediatric otolaryngologists. Motility problems may also be diagnosed during the evaluation of swallow. Rare conditions such as H-Type tracheoesophageal fistulae can cause chronic aspiration symptoms and cough. Diagnosis can be suspected on contrast swallows but is confirmed on tracheoscopy and require surgical treatment.

Conclusion

The complexity of a paediatric swallow necessitates the assessment and treatment of swallowing disorders by a Multidisciplinary Team. Infants and children often have different conditions causing a swallowing problem and therefore management needs to be tailored to meet the needs of the individual child.

References

Paediatric tongue-tie

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Abstract
Tongue-tie, also termed ankyloglossia, can be anterior, posterior, or rarely superior in nature, resulting in breastfeeding difficulties, speech articulation errors or social problems. Assessment includes visualisation and palpation of the tongue and frenulum, observation of breastfeeding, and evaluation of growth and development. In older children, assessment of speech with a speech and language therapist may be required. Surgical options include frenotomy and frenuloplasty.

This article provides an overview of the current knowledge base regarding the management of tongue-tie in children.

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Key words
Ankyloglossia, tongue-tie, frenotomy, frenuloplasty, breastfeeding

Definitions and Anatomy
Ankyloglossia (ankylós: ‘crooked’ tongue), more commonly known as ‘tongue-tie’, is a congenital abnormality of an unusually short or thickened midline connection between genioglossus in the midline and the intrinsic musculature of the tongue. The exact embryological mechanism by which it occurs is unclear but it may result from failure of apoptosis or ‘over-fusion’ of the lingual prominences during tongue development.

The majority of ankyloglossia is anterior in nature and visible as a tight or thickened frenulum running from the floor of mouth to the anterior ventral surface of the tongue, inserting at or near the tip (Figure 1a). Less frequently, and probably under-diagnosed, is a posterior ankyloglossia (Figure 1b). This results from midline tethering from the floor of mouth inserting more posteriorly on the ventral surface of the tongue, and can be submucosal. Although these subdivisions are widely documented in recent literature, some individuals still challenge the concept of posterior ankyloglossia and feel that functional problems instead result from other issues.

Very rarely the tongue can be connected from its dorsal aspect to the hard palate (ankyloglossia superior).

Presentation
Ankyloglossia is a common anatomical abnormality with a documented frequency in the literature of 2-16% of neonates. The range of documented prevalence is accounted for partially by variation in detection rates and definitions for posterior ankyloglossia, but genetic factors may also account for geographic variation. There is evidence for a male preponderance with a relative frequency of 1.5-3:1. Over recent years there has been a marked increase in prevalence in several countries, with increases of over 800% in 15 years recorded in the USA, thought to be due to increased facilitation of breastfeeding and resultant awareness of ankyloglossia.

Although ankyloglossia is usually a sporadic finding, familial ankyloglossia with either X-linked or autosomal dominant inheritance has been postulated. It can also be associated with Optiz’s, Beckwith-Wiedemann, van der Woude, Simosa, X-linked clef palate, Orofaciodigital and other syndromes. There may also be an association with maternal cocaine misuse. Many cases of ankyloglossia are identified at newborn baby checks but only a proportion of these will develop functional disability either in neonatal life or later childhood.

Human milk is widely accepted to be the optimum form of infant nutrition and the World Health Organisation recommends exclusive breast feeding for the first six months of life. Breastfeeding also strengthens an emotional bond between mother and child. The normal process of breastfeeding requires the infant’s tongue to help pull the whole nipple and some of the breast into the mouth, to provide compression and suction to the nipple to encourage milk expression, and to control the milk bolus.

There is a reasonable body of evidence indicating that ankyloglossia can contribute to breastfeeding difficulties. A prospective study of neonates with ankyloglossia identified feeding difficulties in 44% of cases, and small case control studies have demonstrated a much greater incidence of both latching and feeding difficulties in the presence of ankyloglossia. Furthermore, the prevalence of ankyloglossia in infants with breastfeeding difficulties has been shown to be higher than the prevalence of ankyloglossia in the local population. The severity of the limitation of tongue movement secondary to ankyloglossia is felt to be proportional to the impact on breastfeeding. The infant with ankyloglossia may generate a shallower latch as a result of reduced tongue mobility, focussing pressure on the tip of the nipple and encouraging the infant to use their lips to clamp onto the nipple to generate extra force. The resulting pain for the mother, in addition to prolongation of infant feeding and failure to thrive due to reduced milk let down, can result in discontinuation of breastfeeding, and use of bottle or formula feeds. Evidence suggests that for every day of nipple pain in the first 3 weeks there is a 10-26% risk of cessation of breastfeeding.

Some children with ankyloglossia are breastfed without reported difficulty and present later during language acquisition with speech articulation concerns. The effect of ankyloglossia on speech is more controversial than that...
on breastfeeding, and can, at most, only account for articulation errors rather than global speech delay, or other abnormalities of language development26. Ankyloglossia is thought to affect the production of lingual sounds (such as "s" and "d") and sibilants (such as "z", "s", and "th")3. In one study Speech and Language Therapists identified roughly 70% of children over 1 year of age with ankyloglossia as having articulation errors ascribable to ankyloglossia27.

Older children with ankyloglossia may present with parental concerns regarding social development. Children may not be able to lick their lips or perform tasks requiring significant tongue protrusion such as licking an ice cream cone, and may present in later childhood or adulthood with an impaired ability to kiss1,3,11. Amongst other causes, ankyloglossia has been cited as contributing to abnormal dentition, deficiencies in oral hygiene, and difficulty playing wind instruments.

**Assessment**

The assessment of a child with suspected ankyloglossia requires an understanding of the age-appropriate history and performance of an attentive clinical examination. For neonates and infants, a detailed breastfeeding history attending particularly to the quality of the latch, presence of maternal discomfort and lactation, and the infant’s growth trajectory is imperative.

Intraoral examination includes inspection of the whole oral cavity including the integrity of the palate, as well as examination for an upper lip tie (tight maxillary labial frenum), which may potentially contribute to breastfeeding difficulties30. Examination of the tongue involves inspection of both dorsal and ventral aspects, opportunistic observation of tongue movements or tethers especially during crying, and palpation with a gloved finger along the deep ventral surface of the tongue from one side to the other (‘Murphy Manoeuvre’)2. Palpation of a midline restrictive band in the absence of a visible anterior frenulum indicates the presence of posterior ankyloglossia.

In neonates, an up-turned gloved finger inserted between the tongue and palate can facilitate examination of the tongue along its length against the palate28, and facilitates assessment of the quality of muscular opposition of the tongue along its length against the palate28. In many cases, an observed breastfeed attempt with a lactation specialist may demonstrate functional problems unresolved by breastfeeding issues29. Older children may follow commands to protrude or elevate their tongue which can aid in the assessment of tongue mobility. If referred with articulation concerns then formal assessment with a speech therapist is beneficial.

**Classification System & Severity Scores**

Various scoring systems have been developed to help classify and grade ankyloglossia but none have been shown to reliably predict the benefit of surgical intervention. The Corylles30 and Kotlow systems31 anatomically classify ankyloglossia according to the antero-posterior point of insertion of the lingual frenum into the ventral tongue, but the graded classification does not correlate with the severity of functional disability.

Other scoring systems have attempted to assess severity by grading the maximum degree of tongue elevation32,33 or protrusion34. Although they may have some merit in older children who can follow commands, these are practically very difficult to use in the neonatal setting. They also fail to suitably assess movement restriction of the posterior tongue resulting from posterior ankyloglossia, nor do they predict the outcomes of treatment.

The Hazeltaker Assessment Tool for Lingual Frenum Function (HATLFF) consists of both appearance and function criteria, with a diagnosis confirmed with appearance scores ≤ 8 or function scores ≤ 114. Although this system has been shown to have reasonable inter-rater reliability for many of its items34, it is time-consuming to perform and its usefulness has been questioned27,14. A more simple 4 criteria tool, the Bråtst Tongue Assessment tool (BTA), has also been developed and its validity confirmed against HATLFF44. Neither tool is currently used in routine clinical practice, nor has been shown to predict which patients require intervention.

Various other scores have been developed that identify and grade breastfeeding problems, although these tools are not specific for ankyloglossia. The LATCH score assesses breastfeeding efficiency and identifies high risk patients for breastfeeding discontinuation45,46. The short form McGill Pain Questionnaire (SF-MPQ) validated pain score has been used to grade nipple pain47, and the Infant Breastfeeding Assessment Tool (IBBAT) score assesses breastfeeding efficiency and satisfaction48.

**Types and Timing of Intervention(s)**

Asymptomatic ankyloglossia, identified during routine neonatal examination, but not impacting on breastfeeding, does not require automatic surgical intervention. This is supported by social VSFCE and numerous national organisations around the world14. Between 50% and 83% of neonates diagnosed with ankyloglossia are able to breastfeed without significant difficulties30,34. There is currently insufficient evidence that prophylactic treatment prevents significant numbers of tongue-tied infants developing speech articulation or mobility-related social issues in later life. Whether this is due to weakening of the retaining lingual frenum or the development of compensatory behaviour is unclear26,30.

Ankyloglossia-induced breastfeeding difficulties and maternal nipple pain are likely to present within the first few days of attempted breastfeeding. Initial management, particularly for new mothers, may include review by a trained breastfeeding specialist who may facilitate the establishment of a deeper latch, tongue stretching, or the use of nipple shields to normalise feeding1,23.

It can be problematic deciding when initial non-surgical interventions have been given sufficient opportunity and surgery should be considered. Delay in surgical intervention can result in emotional stress for the mother and an avoidance of breastfeeding45,46. While premature intervention may expose infants to unnecessary surgical risk32. Although there is little evidence for timing of surgical intervention many suggest action within the first few weeks of life is sensible, if conservative measures are failing.

Frenotomy, or division of the retaining lingual frenum, has historically been performed by appropriately trained staff from a range of clinical backgrounds and is the most commonly performed procedure in neonates. The infant is not fed for an hour prior to the procedure and is swaddled in a sheet or blanket. The tongue is elevated and protected, ideally with a grooved retractor (Figure 1a), prior compression of the frenum with a haemostat further maximising the area for division. Examination of the tongue involves inspection of both dorsal and ventral aspects, opportunistic observation of tongue movements or tethers especially during crying, and palpation with a gloved finger along the deep ventral surface of the tongue from one side to the other (‘Murphy Manoeuvre’)2. Palpation of a midline restrictive band in the absence of a visible anterior frenulum indicates the presence of posterior ankyloglossia.

Frenotomy was not found to stop any bleeding and an early breastfeed is encouraged. Postoperative stretching of the ventral surface of the tongue is sometimes recommended32 but there is little evidence for its efficacy in increasing mobility and preventing re-adhesion.

Frenectomy describes complete excision rather than division of the frenum. Frenuloplasty is performed under general anaesthesia in older children with thicker and more posterior ankyloglossia (Figure 2). It can also be used for revision cases following frenotomy. Following release of the frenum the diamond-shaped defect is closed vertically with absorbable sutures, maximising tongue mobility33. In addition, a z-plasty can be performed to provide additional release37.

**Efficacy**

A Cochrane review of frenotomy for ankyloglossia in newborn infants identified 5 RCTs that met inclusion criteria (n=302) but significant methodological concerns were noted. It concluded that while there was evidence of reduced maternal nipple pain in the short term following frenotomy, there was no ‘consistent positive effect’ on breastfeeding from these studies and that no long-term outcomes were available34. There is a marked placebo effect for frenotomy on maternal nipple pain and feeding scores, highlighting that caution should be demonstrated in reporting outcomes of uncontrolled and non-blinded studies of frenotomy35. This is especially true over the longer term as little is understood about the natural history of untreated ankyloglossia38. No robust studies, as yet, have proven that early frenotomy mitigates breastfeeding cessation.

Lower level evidence does at least support the role of frenotomy in breastfeeding. A study using sub-mental ultrasound documented improved sucking dynamics post frenotomy, along with increases in milk transfer, intake and subsequent maternal satisfaction demonstrated efficacy via reduced length of feeds, number of feeds per day, and increased time between feeds39,44 while infants can resultantly climb growth centiles post frenotomy40,41. Speech development is often normal in the presence of ankyloglossia39. There is only very limited evidence, using small sample sizes, for tongue tie division in infancy for ankyloglossia-related speech articulation difficulties42,43. Furthermore there is insufficient evidence to support the prophylactic use of frenotomy in neonates to prevent potential later articulation difficulties43,44.

**Complications**

Ankyloglossia release is a low risk procedure in trained hands. Bleeding is the most frequent complication, although with frenotomy this is normally minimal in volume45. In neonates it is not normally an issue with 8-18% of patients even sleeping through frenotomy16,45. Searing, resulting in recurrent ankyloglossia, occurs in 2.6-6.5% of cases37,39 and may be more common in cases of posterior ankyloglossia45. With appropriate anaesthetic care, injuries to the submandibular ducts, sublingual glands or intrinsic tongue muscles should be minimal40.
Service delivery
Ankyloglossia is managed across the world by teams composed of varied personnel. It is important for the multidisciplinary team to have timely access to experienced staff, trained in the assessment of breastfeeding issues, and equipped with the necessary non-surgical and surgical tools. When surgical intervention is deemed necessary it should be undertaken in a location where treatment can be escalated if required.

Conclusions
Ankyloglossia is a common condition, the natural history of which is poorly understood. A significant proportion of infants born with ankyloglossia never suffer a significant functional disability and so automatic surgery in the first weeks of life is not recommended. Ankyloglossia may however contribute to nipple pain and the discontinuance of breastfeeding, and a careful assessment of these issues may result in the decision to release the frenum. However, maintaining breastfeeding if non-surgical intervention is unsuccessful. Surgery is safe in trained hands, but further research is necessary, in the form of well-designed trials, to assess the optimum timing of surgery, and to identify which patient groups are most likely to benefit significantly.

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Paediatric oesophageal foreign bodies

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Abstract

There have been many reports of morbidity and mortality after foreign body ingestion in children. Paediatric oesophageal foreign body impactions may cause diagnostic uncertainty and there still remains some controversy over how best to manage these patients. It is important for the otolaryngologist to be aware of the issues and pitfalls involved in managing these children, although in tertiary centers paediatric surgeons, paediatric gastroenterologists and multidisciplinary tracheal teams may also be involved. This article discusses the presentation and investigation of ingested foreign bodies, details procedures in their removal and highlights the dangers of button battery ingestion. Current guidelines and management consensus are also discussed.

Key words

Foreign body, oesophagus, battery

Background

Foreign body ingestion by children is a common phenomenon with the potential for significant morbidity or even mortality. A large proportion of foreign bodies will transit through the gastrointestinal tract without consequence. Problems may arise if the object becomes impacted along its course, an event that is more likely to occur at particular sites of anatomical narrowing, notably the cricopharyngeus. Other oesophageal regions of narrowing are the level of the aortic arch, the left main bronchus and the gastro-oesophageal junction. Sites that lodge at the cricopharyngeus are likely to bring the child to the attention of both paediatric surgeons and otolaryngologists.

In 2014, NHS England issued a patient safety alert regarding the risks of delay in recognising button battery ingestion1. The complications of button battery ingestion regarding the risks of delay in recognising button battery ingestion is increasing and a current review of the management of paediatric oesophageal foreign bodies with a focus on button battery ingestion is warranted.

Presentation of Foreign Body Ingestion

Foreign body ingestion may not always be witnessed. If a caregiver has witnessed the ingestion of a foreign body or a child describes it, the exact nature of the object must be determined if possible. Coins remain the most common ingested foreign body in Western paediatric populations1,2, whereas fish bones are more common in Asian populations, reflecting the diet of that culture3. However, bones from fish or other meat are more likely to become lodged in the oropharynx4 or tonsill1 rather than the oesophagus. Food bolus obstruction occurs more frequently in children with previous pathology such as oesophageal stricture (e.g. after oesophageal atresia) or achalasia. Children are able to insert foreign bodies into the mouth themselves from the age of around 5 months. Any child presenting with a foreign body in a younger age group should raise the suspicion of non-accidental injury1. There have been reports of oesophageal foreign bodies in neonates as young as 3 weeks5.

Foreign bodies that become impacted in the oesophagus are likely to do so in the upper third6,7. Complications that may arise at this level include tracheo-oesophageal fistulae, vascular fistulae, abscess formation and mediastinitis. Distal sites of oesophageal foreign body impaction include at the level of the aortic arch, the left main bronchus and the gastro-oesophageal junction. It is rare for a foreign body to impact once it has passed the oesophago-gastric junction.

The acute presentation may be with choking, cough, wheeze, or cyanosis followed by dysphagia and increased work of breathing. A foreign body impacted in the oesophagus will then likely cause swallowing difficulties. Symptoms may have been attributed to a viral upper respiratory tract infection and consequently the foreign body may be missed on initial presentation8. Symptoms such as stridor, may be misinterpreted as coughing9. If an impacted oesophageal foreign body has not resulted in an acute presentation, it is likely that symptoms of dysphagia, odynophagia, chest pain, drooling, refusal to eat, and regurgitation will follow. There may be associated respiratory symptoms such as stridor, coughing or wheezing. Airway compromise may develop from an oesophageal foreign body if it causes external compression, either directly or from later abscess formation, or if a tracheo-oesophageal fistula develops. A proportion of children may be asymptomatic.

Late presentation increases the potential for morbidity from oesophageal impaction, in particular with button batteries due to the longer duration of contact time10. Even relatively inert objects, such as coins, may cause perforation of the oesophagus by pressure necrosis11. Chronically impacted foreign bodies can lead to oesophageal "pouch" formation12. There have been cases of inert foreign bodies migrating into the mediastinum requiring open thoracotomy removal13 and other cases of migration into the paraesophageal space14.

When to x-ray?

Any child with unexplained symptoms of airway obstruction, drooling, dysphagia, chest pain, persistent vomiting and coughing or choking when eating, could potentially have a foreign body, and a chest radiograph is indicated. Anterior-posterior and lateral views are required to identify the location (airway vs. oesophagus), morphotype and total number of foreign bodies15. If the object is radiolucent and the patient has ongoing symptoms with a normal x-ray, they should proceed to endoscopy. Contrast swallow is not recommended because of the risk of aspiration16.

If there is any concern that the child may have swallowed a button battery, even if asymptomatic, a chest radiograph should be performed, because of the potentially catastrophic risk of missing this diagnosis. A button battery will appear with a classic radiolucent ring ("halo" sign) on anterior-posterior chest x-ray (Fig 1) and a "step-off" sign on lateral x-ray. Button batteries may be mistaken for coins and if there is any diagnostic doubt, the child should proceed to endoscopy.

Management of Oesophageal Foreign Bodies

Once the diagnosis has been established from a definite history or chest radiograph, the next decision is whether the foreign body can be managed expectantly. If the child has presented with the foreign body in their upper oesophagus, it is less likely to pass spontaneously and endoscopic removal should be considered. Sharp objects and button batteries should always be removed urgently because of the risk of complications. Inert and smooth oesophageal foreign bodies are more likely to pass spontaneously17. Management of these patients should consist of observation and confirmation that the object has passed below the gastro-oesophageal junction. A randomised trial of asymptomatic patients with an ingested oesophageal coin found that 25% would pass spontaneously within 16 hours and this was more likely to occur the more distal the foreign body was18. Another cohort study showed 46% of foreign bodies were eliminated spontaneously. A smooth foreign body that has passed into the stomach is very likely to pass through without complication, and no further imaging is indicated unless significant gastrointestinal symptoms develop. Glucagon administration (as a smooth muscle relaxant) has been proposed, but this was not shown to be effective for oesophageal coin dislodgement in a small placebo-controlled trial19.

Oesophagoscopy (flexible or rigid) allows definitive identification of the foreign body and direct retrieval. Flexible nasoendoscopy prior to taking the child to theatre.
for a general anaesthetic is likely to only show pooling of saliva in the piriform fossa and is not helpful, except to exclude other causes for the symptoms. Magill forceps may be used to extricate foreign bodies if they are in the oropharynx and can be seen on direct laryngoscopy. A systematic review identified that the majority of foreign bodies can be removed endoscopically with no clear trend towards either flexible or rigid oesophagoscopy, however the benefit of flexible endoscopy with less risk of perforation was suggested.

Ideally, forceps should be used to grasp objects from the oesophagus. If direct retrieval cannot be achieved, an alternative is to push the foreign body into the stomach from where it is likely to pass spontaneously. Use of a balloon catheter, such as a Foley or angiography catheters to trawl the foreign body from the distal end has been described. Some metal objects can be removed with a magnetic probe. Blind removal of foreign bodies in this way does not allow visualisation of the oesophageal mucosa. The Foley catheter method, even under fluoroscopic guidance, has a risk of airway compromise, as it is difficult to control the passage of the foreign body out of the hypopharynx. Smooth muscle relaxation instigated by general anaesthesia occasionally may in itself be enough to allow the foreign body to drop into the stomach.

If no foreign body is found after examination of the trachea and oesophagus, it is pertinent to look in the nasopharynx, as a foreign body lodged here may also present similarly to other aerodigestive tract foreign bodies. Very rarely open surgery is required to remove foreign bodies if they have migrated beyond the field of the oesophagoscopy. This may include a foreign body within the wall of the oesophagus, or into the mediastinum. Several authors have developed management guidelines and protocols for various ingested foreign bodies, but the diversity of the paediatric population and ambiguity surrounding diagnosis means that it is difficult to apply these to all cases and clinical judgment should be utilised.

Button Battery Ingestion
Litzovitz and colleagues have analysed a large number of button battery ingestion incidents from a national database in the USA. Their initial findings reported in 1992, from 2382 cases, indicated a peak incidence in 1 to 3 year olds. Since then, a repeat analysis in 2010 of 8648 cases, identified 13 mortalities and 73 major adverse outcomes. They found that the incidence of button battery ingestion had not show any upwards or downwards trend over a 25 year period, but that the proportion of significant morbidity and death has significantly increased over that time.

Conversely, other studies have suggested that more children are currently presenting with button battery ingestion. Nevertheless, this increase in morbidity has been attributed to the use of a more hazardous 20mm lithium cell battery type, which is more likely to become lodged in a child’s oesophagus because of the larger diameter. Moreover, it has a higher voltage (3V) and therefore risk of caustic damage.

Button batteries lodged in the oesophagus should ideally be removed within 2 hours of ingestion, to prevent serious harm. This should occur regardless of the time from last oral intake. Battery batteries may cause oesophageal injury by three mechanisms - leak of corrosive alkaline substance (sodium or potassium hydroxide), direct pressure necrosis, and by inducing an electrical current that hydrolyses tissue fluid. The position that the battery lies in may influence this, as the negative pole generates the current and leads to hydrolysis production. As with other foreign bodies, batteries that have passed into the stomach usually pass through the remaining gastrointestinal system without complication. It is common practice to repeat an x-ray of the abdomen at 3 to 4 days after battery ingestion to ensure the battery has passed. Intervention is only indicated if significant gastro-intestinal symptoms occur. There is evidence to suggest that routine imaging may not be required in children over the age of 12 years who are asymptomatic after battery ingestion provided the battery is known to be <12mm diameter.

Features of button battery ingestion at initial oesophagoscopy include mucosal ulceration and necrosis, however it can be difficult to assess the depth of the injury. Therefore, note should be made of the position of the negative pole and relation to anatomical structures in order to help aid prognosis. Following removal of a button battery, there is ongoing risk that the caustic injury may extend. Tracheo-oesophageal and aorto-oesophageal fistulae have both been reported to present several days after battery ingestion. This might be due to the weakness of the scarred tissue or from ongoing presence of leaked alkali.

Reported complications of button battery ingestion include oesophageal stricture, perforation, tracheo-oesophageal fistula (Fig 2), aorto-oesophageal fistula, vocal cord palsy, pneumonia, mediastinitis, spondyloisclasis and death. In the reported cases of complication following button battery ingestion, the large majority (80%) of batteries were over 20mm in diameter. Previously there was concern that metals, particularly mercury, may be absorbed into the systemic circulation. Since mercury use in batteries has now stopped and other batteries do not tend to leak metals systemically, there is very little risk that metal poisoning will occur following battery ingestion. Tracheo-oesophageal fistula, if not apparent on initial endoscopic examination, is likely to present with aspiration pneumonia. This can be confirmed on repeat endoscopy.

Figure 2: Appearances on rigid bronchoscopy day 14 following oesophageal button battery removal on day 6, showing a tracheo-oesophageal fistula

Case 1: Delayed recovery of a button battery
A 20 month old presented with increased work of breathing, with a preceding 6 day history of temperatures and wheeze. Following a chest x-ray, an oesophageal lithium button battery was removed from 2 cm below the cricopharyngeus in the local hospital. The child developed a large aspiration pneumonia resulting in significant airway compromise, requiring ventilatory support and retrieval to the tertiary unit. The child suffered persistent aspirations and several failed attempts of extubation. Thus laryngotracheobronchoscopy and flexible bronchoscopy were undertaken eight days later and a tracheo-oesophageal fistula (TOF) in close proximity to the carina, despite normal laryngeal appearances (FIGURES 3A-C). The child was intubated with an endotracheal tube placed under direct vision beyond the fistula for safe transfer to the quaternary centre for repair of the TOF and underwent a complicated repair with multiple procedures. Initially a slide tracheoplasty with Gortex patch and primary oesophageal repair was undertaken. Post-operatively the child developed mediastinitis, a mediastino-cutaneous fistula and severe tracheomalacia. Subsequent tracheal and oesophageal dilatations were undertaken and biodegradable tracheal stents were inserted. The mediastinal wound was debrided with excision of the mediastino-cutaneous sinus tract and partial excision of the patch. The child underwent two subsequent balloon dilatations of the trachea. Five months following the first repair, the tracheo-oesophageal fistula recurred and was evident on tube oesophagogram. Therefore further repair was undertaken using an autologous pericardial patch tracheoplasty and primary oesophageal repair. Ten months after the second repair the child has undergone bronchoscopy, bronchogram and further airway dilatation and remains under follow up.
Conclusions

Ingested foreign bodies may present inconspicuously and the diagnosis should be considered in any child presenting with feeding or breathing difficulties. If a button battery is identified in the oesophagus, urgent endoscopic removal should be undertaken with surveillance for any complications. Due to the potential for significant consequences, all clinicians who manage the paediatric population should be cognisant of the morbidity of a swallowed foreign body, so as not to miss an oesophageal button battery.

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References


Biofilms in paediatric otolaryngology

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Abstract
Bacteria exist in planktonic (free-floating) and biofilm forms. Biofilms are communities of microorganisms that live in a protective glycocalyx matrix and share metabolites and genes allowing them to survive in many diverse environments. A significant number of children worldwide suffer from common ear, nose and throat conditions such as otitis media, chronic adenoiditis, recurrent tonsillitis, and rhinosinusitis. There is clear evidence that bacterial biofilms are implicated in these chronic conditions. Antimicrobial resistance and new appreciation of the biofilm lifecycle has lead to the development of new antimicrobial compounds including nitric oxide, silver nanoparticles and baculovirus therapy that compliment antibiotics and will augment our approach to biofilm treatment in the future.


Key words
Biofilm, otitis media, adenotonsillitis, rhinosinusitis

Introduction
Biofilms are collections of microorganisms (bacteria and fungi) with an extracellular matrix that live in colonies, sharing protective mechanisms, genetic material and metabolic products. They interface with a fluid or solid surface such as mucus or tissue, and behave distinctly from bacteria in the planktonic, free floating state such as mucus or tissue, and behave distinctly from bacteria in the planktonic, free floating state. Docking involves reversible contact between planktonic bacteria and the host surface. Locking anchors bacteria to the host surface and adjacent organisms using various ligands, pile, curly and fimbriae. Maturation involves the development of the extracellular matrix and development of complex intercellular signaling networks. Dispersal involves shedding of the biofilm to other sites.

This review aims to appraise the relevant literature regarding biofilms and their involvement in paediatric otolaryngological conditions, and present the current perspective on future research and treatments.

Biofilm Physiology
Current models of the biofilm lifecycle suggest stages that include surface conditioning, docking, locking, maturation and dispersal (Figure 1). Surface conditioning involves the binding of polysaccharides, salts, DNA, bile mucins and plasma proteins that change the physiochemical properties of the surface and allow planktonic bacteria to adhere to the surface. Docking involves reversible contact between planktonic bacteria to the surface dictated by Van der Waals forces, electrostatic and hydrophobic interactions, nutrients, steric hindrance and temperature overcoming the repulsive forces between the surfaces. Locking anchors planktonic bacteria to the surface and adjacent organisms by producing a complex of receptor specific ligands and pile, curl or fimbriae (projections involved in cell-cell / cell-substrate adherence and mobility which vary depending on the organism). As bacterial cells attach they produce various autoductors that spread signals regarding cell density and alter gene transcription. This induces production of extracellular matrix components and can change bacteria from the planktonic state to the sessile (biofilm) state. Maturation of biofilm involves replication of the organisms but also further development of the extracellular matrix including production of mature glycolalys, a complex of proteoglycans and glycoproteins that traps nutrients, metabolites and organisms. Mature biofilms can release organisms in a process known as dispersal. Signalling molecules such as nitric oxide have been identified which lead to transcriptional changes that resemble that of the planktonic state and mediate biofilm dispersal in S. epidermis, P. aeruginosa and E. coli.

Biofilms are advantageous for bacteria as they allow bacteria to tolerate harmful conditions, evade host defences and protect bacteria from antimicrobial compounds or toxins. The biofilm extracellular matrix reduces the diffusion rate of antibiotics by acting as a size exclusion barrier, binding or repelling charged antibiotic molecules thereby reducing the effective concentration of antibiotics in the deep layer. More superficial organisms may sense the attack signalling to deeper organisms in the biofilm to adapt and produce antibiotic resistance compounds such as...
as beta lactamase and catalase, or to induce a low metabolic state resistant to antibiotic effect\(^1\).

The host's immune system often mounts a futile and damaging response to biofilms. Given the diverse nature of the organisms present, biofilms can change their surface antigens regularly. The large mass of the extracellular matrix blocks macrophages ability to phagocytose foreign biofilms\(^2\). The polysaccharide coating hinders activation of complement and opsonisation\(^3\). Consequently, persistent activation of both the innate and adaptive immune system leads to collateral damage of the surrounding tissues by cytotoxic, proteolytic and proinflammatory responses\(^4\).

**Ear infections**

Otitis media with effusion is commonly thought of as a sterile inflammatory process as microbiological cultures of effusions are frequently negative. More recently, polymerase chain reaction (PCR), nucleotide amplification fluorescence in-situ hybridization (FISH) and confocal laser microscopy (CLSM) have demonstrated genomic material, mRNA, bacterial proteins and components of biofilm matrix in large proportions of chronic otitis media effusions (Figure 2)\(^5\)-\(^14\). This suggests both metabolically active bacteria and a possible role for biofilms in the disease process. Typical otopathogens include *H. influenzae*, *S. pneumoniae*, *M. catarrhalis*, *S. aureus* and *P. aeruginosa*. All have the capability of forming biofilms both individually and with other prokaryotes.

Hall-Stubley et al (2006) examined middle ear effusions within a paediatric population during grommet insertion for otitis media with effusion (OME) and recurrent acute otitis media (RAOM). Culture demonstrated growth in 22% of samples. PCR techniques examining DNA all tested positive for at least one ototopathogen. *H. influenzae* was positive in 70.8% of cases, with *S. pneumoniae* in 50% and *M. catarrhalis* in 25%. All three ototopathogens were identified within the same sample in 16% of middle ear effusions.

Middle ear biopsies from these patients and patients suffering from RAOM were analysed using CLSM and BacLight probes showing 92% of samples had biofilms. In total, 80% of middle ear mucosal biopsies contained *H. influenzae* using specific FISH probes, and 88% demonstrated biofilm features including presence of microcolonies, elaboration of extruded exopolysaccharide and tower formation. All samples were positive for *S. pneumoniae* and 71% demonstrated features of biofilm. These findings suggest an element of chronic infection with acute exacerbations.

The adenoid has often been attributed to recurrent infections of the middle ear and this is presumed to be due to mechanical obstruction. Van Hoecke et al (2016) investigated the adenoid as a reservoir for infection in children undergoing adenoidectomy and ventilation tube insertion for bacterial chronic otitis media with effusion. They established the same bacterial species colonised the adenoid and the middle ear effusion simultaneously in 84.6% of cases. In 81.2%, the same bacterial species genotype was identified using randomly amplified polymorphic DNA analysis\(^15\). Furthermore, Hoa et al (2009) identified 85% of the surface area of the adenoid was covered in biofilm in children with RAOM. Further FISH analysis showed multiple common otopathogens were involved\(^16\).

Comparison between adenoids of children with a history of chronic suppurrative otitis media and those removed for nasal obstruction demonstrated a significant difference in biofilm presence (100% vs. 65% respectively). The group with nasal obstruction showed a significantly greater adenoid to nasal airway ratio suggesting that mechanical obstruction may not be as important a factor as biofilm load on the adenoid surface of patients with OME/RAOM\(^17\). A similar study compared the surface area affected and location of biofilms on the adenoid in relation to OME and RAOM. Hoa et al (2010) identified 27.7% of the adenoid surface area was covered in biofilms in OME, whereas 97.6% was covered in RAOM\(^18\). Moreover, it has been shown 72.2% of patients with RAOM and/or OME have organisms capable of producing biofilms in vitro on the adenoid close to the eustachian tube orifice, whereas only 53.3% of organisms located on the nasopharyngeal dome were capable of forming biofilms\(^19\). This may suggest not only colonisation of the adenoid with biofilm-forming organisms, but also their proximity to the eustachian tube orifice is important factor causing chronic infection of the middle ear.

Biofilm components have also been identified in chronic ventilation tube otorrhoea. Idicula et al (2016) cultured tympanostomy tube otorrhoea finding 73.3% specimens were culture positive and 81.8% of those stained positive for biofilm components including DNABII and eDNA\(^20\). Indeed common otopathogens including *S. aureus*, *P. aeruginosa* and *H. influenzae* have been demonstrated in vitro to form dense biofilms on tympanostomy tube materials. Electron microscopy has further identified loci points for biofilm formations on various tympanostomy tubes that tend to occur around the perpendicular junctions of the structures\(^21\). Despite many modifications to tympanostomy tubes including various materials, changing the shape and coating with antibacterial/antibiotic agents no single modification has been shown to completely inhibit biofilm formation\(^22\).

**Nasal infections**

Recurrent upper respiratory tract infections affect around 18% of children between the ages of 1-4 years and can be associated with recurrent acute otitis media and recurrent acute sinusitis. There is limited data regarding the incidence of chronic rhinosinusitis (CRS) in the paediatric population. Furthermore, many children diagnosed with CRS in fact have chronic adenoids, which can mimic CRS in presentation\(^23\). Using scanning electron microscopy, Zuliani et al (2006) identified significant adenoidal biofilm in 100% of children with CRS symptoms compared to 0% in children with obstructive sleep apnoea (OSA). Surface area of the adenoids covered in biofilms were compared between children diagnosed with CRS and OSA who had an adenoidectomy, noting the CRS group had a mean adenoidal surface area coverage of 94.4% with biofilms compared with 1.9% in the OSA group (Fig 3)\(^24\). Vandenbengel et al (1997) prospectively studied symptom scores of patients treated with adenoidectomy for CRS symptoms noting a significant reduction in symptom score at 8 weeks and between 5-24 months after the procedure. Furthermore, 74% of patients using frequent antibiotic

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**Figure 2:** Example of confocal light microscopy with BacLight probe demonstrating surface-related bacterial biofilms on middle ear mucosa and control tissue. (A & B) Representative CLSM images of middle ear mucosa showing viable aggregates of bacteria (biofilms) (white arrows) attached to the underlying surface epithelium (red arrows). (C) Representative CLSM images of control middle ear mucosa showing epithelial cells (yellow arrows) with no evidence of any associated bacterial biofilms.
CRS is typically diagnosed in children between the ages of 4-7 years. CRS is believed to be a multifactorial condition involving osteomeatal abnormalities, hypersensitivity, mucociliary dysfunction and chronic infection. Studies investigating biofilms in CRS have been primarily adult-based as the condition is much more common in this age group. Furthermore, adults demonstrating biofilm features within the sinuses and lungs have been shown to have higher levels of biofilm forming pathogens compared to children. In the paediatric population, as immunopathological studies have drawn attention to the role of biofilms in the development of chronic rhinosinusitis in the paediatric population.

Cystic fibrosis (CF) is an autosomal recessive disorder resulting in a defect in the cystic fibrosis transmembrane regulator protein that leads to viscous mucus, which is difficult to clear from the lungs and sinuses. Bacteria are commonly identified as the cause of lung infections in cystic fibrosis patients, with P. aeruginosa and S. aureus being the most commonly identified species. Antimicrobial therapy for P. aeruginosa in cystic fibrosis includes long duration antibiotic therapies, surgical debridement and removal of prosthesis. Long duration antibiotic therapy is often less effective and can lead to increased resistance within the biofilm. Surgical debridement and prosthesis removal is not often feasible and can involve significant complications. Recent appreciation of the biofilm lifecycle has lead to many technologies under development to destroy biofilms and potentiate the effects of antibiotics.

Antimicrobial compounds such as silver have long been used in production of catheters, dressings, tracheostomy tubes and disinfectants. More recently studies have tested their efficacy against biofilms in both vitro and vivo models. Silver particles 40 nanometers in diameter were effective in killing S. aureus biofilms in vitro and a C. elegans in vivo model while causing minimal cytotoxicity in macrophage cell lines. Silver nanoparticles have been shown to disrupt antibiotic resistance through the production of silver nanoparticles.

Throat infections
Sore throat is a common presentation to general practitioners with an annual incidence of 100 per 1000 of the population. Recurrent tonsillitis often requires a number of courses of antibiotics and a significant number of these patients will require adenoidectomy and/or tonsillectomy. Despite penicillin, 20% of those treated for tonsillitis with beta-haemolytic streptococci remain carriers. This bacterial resistance may be a result of biofilms and invasion further investigation. Using CLSM and TEM 73-85% of children with recurrent tonsillitis have been shown to demonstrate biofilms within the tonsillar crypts surrounded by inflammatory cells. Kania et al (2007) used CLSM to assess for the biofilm component glyocalyx in tonsils removed for recurrent tonsillitis in adults noting 70.8% stained positively in crypts of 25 tonsils. It has been suggested biofilms may have a role in recurrent tonsillitis by intermittent dispersal of planktonic bacteria causing acute episodes of infection. Interestingly the same studies identified 41-75% of patients with obstructive sleep apnoea also have biofilms within the tonsillar crypts. Biofilms may also have a role in chronic tonsillar hypertrophy and immune surveillance. However, without information from age matched non-pathological controls it would be difficult to propose a causative association between chronic biofilm infection and recurrent tonsillitis.

Treatments
Current management of biofilms in routine practice includes long duration antibiotic therapies, surgical debridement and removal of prosthesis. Long duration antibiotic therapy is often less effective and can lead to increased resistance within the biofilm. Surgical debridement and prosthesis removal is not often feasible and can involve significant complications. Recent appreciation of the biofilm lifecycle has lead to many technologies under development to destroy biofilms and potentiate the effects of antibiotics.

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Quorum sensing inhibitors have great potential to disrupt intercellular signalling between bacteria that can instigate stress pathways and a variety of responses to increase virulence and protect biofilms from host defences. Sugars produced by seaweed can interfere with N-acetylhexosamine lactone and N-butaryl-L-homoserine signalling pathways involved in bacteria-host interaction and bacterial co-ordination to produce complex biofilm structures. It also promotes the production of virulence factors such as exoproteases, siderophores and exotoxins. A synthetically engineered furanone demonstrated downregulation of virulence associated genes in P. aeruginosa and was shown to reduce...
Nitric oxide has been shown to directly have bactericidal effects on in vitro S. pneumoniae biofilms in a dispersion independent manner. It also greatly potentiates the bactericidal effects of amoxicillin in vitro when treating S. pneumoniae biofilms.

Bacterial treatment utilises viruses that infect bacteria and eluding intercellular messengers and transcription messenger c-di-GMP and initiate dispersal and potentiate the bactericidal effects of tobramycin and H_{2}O_{2} in P. aeruginosa biofilms.

References:

Conclusions
Detecting organisms and biofils in widespread paediatric disease states such as OME, RAOM, adenotonsillitis and CF has renewed interest in a microbial cause for these pathologies. New technologies in detecting biofils and eluding immune system access are targeting transcription patterns are beginning to suggest biofils have a large role to play in paediatric otolaryngology. Biofils have evolved to a multi-component mechanism to protect themselves from insults including current antibiotics and the immune system. As a consequence they are becoming increasingly antibiotic resistant. Newer therapies show great promise in targeting intercellular messangers, removing enzyme cofactors and encouraging metabolism that induce stress responses and make biofils more susceptible to antibiotic effects. Future studies will likely elucidate a clear causative role for biofils in paediatric otolaryngology.
Intracranial complications of ear, nose and throat infections in childhood

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Abstract
Early detection and diagnosis of intracranial complications of ear, nose and throat infections in childhood may be difficult. There is no consensus regarding the specific management of paediatric intracranial infections. However, the literature suggests several risk factors, including immunosuppression, antimicrobial resistance and otolaryngology due to the potential for otogenic and sinoaerogenous causes of brain abscesses. Early multidisciplinary team discussion and input is recommended to manage cases on an individual basis and optimise clinical outcomes in children.


Key words
Intracranial, childhood, infection, thrombosis

Introduction
Children frequently develop infections of the ear, nose and throat (ENT). Most resolve either spontaneously or with antibiotics and limited surgical intervention. Intracranial complications are rare but can develop as a result of direct extension to adjacent brain or meninges, thrombophlebitis or haematogenous spread. Early recognition and management of intracranial complications is essential to reduce further morbidity and in some cases mortality1. This review focuses on recognising the signs of intracranial complications in children and how to manage them.

Presentation of Intracranial Infections
Intracranial complications are sometimes initially hard to detect in a distressed child with an upper respiratory tract infection, but it is important to consider them if there is evidence of any of the red flag signs (Table 1).

It is important to remember that symptoms of raised intracranial pressure (ICP) from intracranial pathology can be insidious in onset and can fluctuate. This is due to the early infection causing an impairment of flow and resorption of cerebrospinal fluid (CSF). Older children are able to report a headache, which will be worse on lying down or will wake them from their sleep. In younger children there may be a change in behaviour; with increased irritability even when apyrexial. They may be lethargic, nauseous or vomit persistently. Any additional symptoms will depend on the site and spread of infection2.

Types of Intracranial Infections
In a study by Kraus & Tovi (1992), which comprised 39 children (age range 1-15 years) with central nervous system (CNS) complications of otorhinological infections, 11 children (28%) were found to have more than one complication. Leptomenigitis was the most common paediatric intracranial complication (54%). Lateral sinus thrombosis occurred in 10 children (26%). Mortality in the paediatric group was 12.8%, with the highest mortality rate reported in those with a brain abscess1. This study demonstrates the high morbidity and mortality associated with intracranial infections secondary to ENT infections. It is therefore very important to be alert to the signs and symptoms of each individual complication.

Meningitis
As well as the initial symptoms of headache, vomiting and lethargy, examination reveals a sick and irritable child who dislikes being touched or moved. Younger children will demonstrate meningeal irritation by not wanting to move their head or neck, in addition to wanting to lie still in bed. Meningeal irritation causes photophobia, and the child may become more distressed in bright light. In older cooperative children it may be possible to elicit a positive Kernig and Brudzinski sign. Kernig’s sign is positive when the thigh is flexed at the hip and knee at a 90 degree angle, and subsequent extension in the knee is painful leading to resistance3. Brudzinski’s sign is positive when severe neck stiffness causes a patient’s hips and knees to flex when the neck is flexed4. A third of children with bacterial meningitis develop seizures5.

Lumbar puncture (LP) is performed if there are no contraindications to this test and there is confidence from neuroradiology studies that a space occupying lesion or infection has been excluded.

Cerebral venous sinus thrombosis
Venous sinus thrombosis often has a delayed diagnosis. This is because the initial ear or sinus infection and fever may have subsided. The thrombophlebitic veins are associated with an increased risk of venous thrombosis and frequently there are additional risk factors such as dehydration, further increasing prothrombotic risk.

Without early anticoagulation small clots may propagate and become more extensive. Again, symptoms of raised intracranial pressure predominate. If the early signs of headache, lethargy and vomiting are missed, or have a more insidious onset, papilloledema will develop. Raised intracranial pressure is transmitted to the optic nerve via the optic nerve sheath, leading to optic nerve disc swelling. Initially, symptoms of optic nerve involvement may be intermittent visual blurring. Left untreated, optic nerve dysfunction leads to a loss of colour vision, loss of visual fields and a deteriorating visual acuity. This may be irreversible and children with untreated raised intracranial pressure may be left blind6.

In a study by Maltik et al. (2009) 21 children aged between 1 month - 17 years were identified with cerebral venous sinus thrombosis. The presenting symptoms included headache (15/21), vomiting (14/21) and visual disturbance (8/21). Most had papilloedema. Importantly, the most common underlying condition was middle ear infection (13/21).

Gradenigo’s Syndrome
Gradenigo’s syndrome is a rare, but recognised, complication of acute otitis media (AOM). Petrous apicitis in the acute setting is thought to arise from abscess formation in a well pneumatised petrous apex. The child presents with a unilateral trigeminal and abducens nerve palsy. Pathophysiology of the abducens nerve palsy is due to contact of this nerve with the apex of the petrous part of the temporal bone as its runs beneath the petroclinoid ligament and through Dorelo’s canal. The trigeminal nerve and ganglion also lie against the petrous apex. Irritation of these nerves by inflammation of the underlying bone results in cranial neuropathies, which characterise Gradenigo’s syndrome.

Clinically children present with ipsilateral severe retro-orbital pain (due to involvement of trigeminal ganglion) and an abducens (VI) nerve palsy. The child will have developed a failure of lateral gaze to the affected side with diplopia (Figure 1). Younger children will not be able to communicate that they have diplopia. Instead, they may have a compensatory torticollis, cover or close one eye to minimise the double vision.

Cavernous sinus thrombosis
Cavernous sinus thrombosis results from an extension of infection and inflammation intracranially into the cavernous sinus7, due to anterograde spread of infection from the mouth, face, nose or sinuses.

The child is primarily unwell with a fever and usually has a frontal headache. Signs and symptoms may be unilateral or bilateral with an orbital cellulitis. Children may present with proptosis, visual loss of colour vision, loss of visual fields and a deteriorating visual acuity. This may be irreversible and children with untreated raised intracranial pressure may be left blind6.

Table 1: Red flags suggesting intracranial complications in children with recent or current ENT infections

<table>
<thead>
<tr>
<th>RED FLAGS</th>
<th>ADDITIONAL SYMPTOMS / SIGNS</th>
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<tbody>
<tr>
<td>Headache</td>
<td>Older children: Worse on lying down, Wakes them from sleep</td>
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<tr>
<td>Intensity</td>
<td>Inconsolable, even when with carer</td>
</tr>
<tr>
<td>Nausea and / or Vomiting</td>
<td>Persistent, Worsening, May be present only on waking</td>
</tr>
<tr>
<td>Lethargy / Level of consciousness</td>
<td>May be fluctuating or deteriorating</td>
</tr>
<tr>
<td>New squint / New diplopia</td>
<td>The child may cover or close one eye to prevent double vision, There may be new head tilt or torticolis</td>
</tr>
<tr>
<td>Deteriorating vision</td>
<td>An older child may complain of ‘blurring’, A younger child may stop focusing e.g. on faces, toys, television etc.</td>
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<tr>
<td>New limp</td>
<td>Weakness</td>
</tr>
<tr>
<td>New coordination / Imbalance difficulties</td>
<td>The child may not be able to sit or stand, Unsteady gait</td>
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References
ptosis, chemosis, sensory disturbance (due to cranial nerve V involvement) and ophthalmoplegia. Initial signs are those of a sixth nerve palsy, with redness in lateral gaze but evolve as the oculomotor and trochlear cranial nerves become involved, with limitation of eye movement and ptosis.

Unresponsive pupils and loss of vision may follow initial symptoms, caused by increased intracranial pressure and traction on the optic nerve and central retinal artery. Infection can spread from one cavernous sinus to the other and therefore focal cortical ischaemia. Children may present with headache, vomiting, seizures and fluctuating levels of consciousness. When the motor cortex is involved, the child may present with an acute stroke.

**Brain abscesses**

Children with brain abscesses usually present with signs of raised intracranial pressure, such as headache, lethargy and vomiting. There may be seizures or new focal neurological deficits, depending on the site of the lesion.

At our tertiary children’s hospital in the south of England, a retrospective study found that over a 33-month period 17 children were diagnosed with a brain abscess. All (17) the children presented with a headache. The proposed origin of the brain abscess based on imaging appearances, was thought to be sinogenic in six (6/17), otogenic in eight (8/17) and of unknown origin in three (3/17) of the cases (Figures 2-4). Ten microbiological culture specimens taken at the time of surgery grew streptococcal bacteria.8

**Extradural, epidural and subdural collections**

These extra cranial infections are characterised by signs of raised intracranial pressure. Vasculitis as a consequence of a subdural empyema may cause thrombosis of cortical veins and therefore focal cortical ischaemia. Children may become involved, with limitation of all eye movements and ptosis.

In a child presenting with signs and symptoms of acutely raised intracranial pressure, CT with CT venography should be performed urgently, to exclude cerebral abscess and dural venous sinus thrombosis [DVST].

**Overview of Investigation and Management**

If a child with an ENT infection develops new and evolving neurological signs, advice from a paediatric neurologist should be sought immediately. The management of intracranial complications requires a multidisciplinary approach involving ENT surgeons, paediatric neurosurgeons, paediatric neurologists and infectious disease paediatricians. Early discussions between the above teams will ensure that the correct investigations are arranged and management initiated (Cases 1 & 2; Figures 5-6).

**Transfer and Neuroprotection**

Children with diagnosed or suspected intracranial pathology should be nursed in an appropriate specialist paediatric setting. This should be at a centre with paediatric neurosurgery and paediatric neurology services. Urgent transfer (within 4 hours of diagnosis) to the specialist unit should be arranged, liaising with the regional Paediatric Intensive Care transport service. If a child has raised intracranial pressure, advice can be given regarding neurological protection including patient positioning (child nursed in a head up position at a 30 degree angle) and specific drug and fluid treatment, such as hypertonic saline.9,10

In their 2017 study, Lampariello and co-workers reviewed the stabilisation of critically ill children at district general hospitals prior to transfer to intensive care. Neurology related interventions were further analysed by the group. They showed that the majority of computerised tomography (CT) scans were performed by district general hospital staff, prior to the arrival of the tertiary hospital retrieval team.

However, administration of osmolar therapy (hypertonic saline) for raised ICP was deferred until the retrieval team’s arrival in nearly half of the patients in whom it was indicated.10 The findings of this study highlight the importance of early liaison with paediatric intensive care unit specialists thereby enabling the safe transfer of children with diagnosed raised ICP.

**Neuroimaging**

Following neurological or neurosurgical review, appropriate further imaging can be discussed with radiology colleagues and is guided by the child’s neurological presentation. Consideration regarding the need for general anaesthesia in order to obtain imaging should be discussed with the multidisciplinary team, so that potential interventional treatments can be considered in advance and ideally planned under the same anaesthetic.

In a child presenting with signs and symptoms of acutely raised intracranial pressure, CT with CT venography should be performed urgently, to exclude cerebral abscess and dural venous sinus thrombosis [DVST]. Brain magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) is preferable in a child who is clinically stable.

**Surgical management**

Intracranial collections usually require neurosurgical drainage in addition to intravenous antibiotics. Otorhinolaryngological infections may also require drainage, but frequently the primary infection may have resolved prior to presentation with an intracranial infection. Close liaison between neurosurgical and ENT colleagues is recommended to consider joint procedures where possible; reducing unnecessary general anaesthetics.

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Figure 1: Identifying a sixth cranial nerve (CN VI) abducens nerve palsy, a left sided abducens nerve palsy is illustrated

Figure 2: CT scan with contrast showing an otogenic brain abscess

Figure 3: A) Axial CT scan showing an opacified frontal sinus. In this case dehiscence of the posterior table of the frontal sinus was also noted, with direct spread of the infection. B) Axial CT scan with contrast showing a frontal lobe abscess of sinogenic origin, as a result of the frontal sinus infection

Figure 4: Brain abscess of unknown origin.
Case 1: Sphenoid sinusitis with cavernous sinus thrombosis

A 15 year old male was admitted urgently to our tertiary children’s hospital with a sudden right proptosis following a 5 day history of upper respiratory tract infection, frontal headaches, photophobia, dippiness, intermittent vomiting and fever. A CT scan at his local unit showed a right cavernous sinus thrombosis with sphenoid sinusitis (Figure A). On examination, he was found to have a right sided 3rd and 6th nerve palsy, with mild restriction in all extremes of gaze. Antibiotic treatment was commenced with intravenous Ceftriaxone and Metronidazole. Operative intervention was undertaken urgently on the day of admission and pus taken grew staphylococcus aureus. Bilateral wide endoscopic sphenoidotomies with washout, retrograde ethmoidectomies posterior to anterior with endoscopic septoplasty for access achieved good clearance of the skull base. Further treatment was guided by monitoring symptoms and bloods (CRP and WBC). A repeat CT scan was undertaken due to ongoing headaches and this showed improvement (Figure B). Two weeks of intravenous Ceftriaxone were continued followed by 2 weeks of high dose oral Augmentin. Following surgery, anticoagulation was commenced, low molecular weight heparin. Clirexane, was administered via subcutaneous injection for 3 months duration. Heparin Anti-Xa levels were used to confirm levels were within the therapeutic range. Advice was given to avoid contact sports for 6 months, strenuous exercise for 1 month and to drink at least 2 litres of fluid per day. A CT venogram arranged 3 months following admission (as an alternative to MRV in order to provide more comparative images to Figure 6A and B). Anticoagulation was commenced with intravenous Ceftriaxone and Metronidazole. This treatment regimen is supported by the literature5. Adequate monitoring of a child’s neurological status and CSF pressure is reduced to preserve visual function.

Figure 5 Case 1: (A) Coronal CT scan showing opacified sphenoids with right cavernous sinus enlargement, relatively low peripheral enhancement outlines a large low density filling defect consistent with thrombus. The right superior ophthalmic vein was also noted to be dilated with no thrombus in this series. (B) Axial CT scan day 5 following surgical drainage with reduction of fluid in the sphenoid sinuses.

Paediatric Infectious Diseases and Clinical microbiology

All children with complex ENT infections should have input from a paediatric infectious diseases and microbiology specialist. The paediatric infectious diseases and clinical microbiology team will advise on the correct empirical antibiotic therapy, followed by rationalising treatment when microbiology results are available. They will also advise on the optimal timing of an intravenous to oral antibiotic switch and the total duration of treatment. When the course of intravenous treatment is predicted to be longer than 7 days, a peripherally inserted central catheter (PICC line) should be considered.

Early insertion of a PICC line minimises the distress to the child caused by multiple cannulations and also ensures antibiotic therapy is given punctually with no delays between doses. At our tertiary children’s referral centre we prescribe ceftriaxone (80 mg/kg once daily) and metronidazole. This treatment regimen is supported by available evidence in the literature10.

Monitoring

Accurate monitoring of a child’s neurological status and assessment of evolving neurology is mandatory regardless of whether a surgical, medical or combined approach is undertaken. Blood tests, in particular observing the trends in the C-reactive protein (CRP) and full blood count (FBC); specifically the white blood cell count and neutrophil count can help monitor response to treatment and guide ongoing management.

In a child with a DVST, with symptoms of raised ICP, it is vital to monitor visual function and to liaise closely with ophthalmology colleagues. If there are any concerns regarding visual function, lumbar puncture and pressure monitoring is performed by the paediatric neurology team and CSF pressure is reduced to preserve visual function. Repeat lumbar puncture may be required in addition to medical management including acetazolamide therapy10.

Acute seizures associated with intracranial infection require management by the paediatric neurology team, and introduction of short-term anticonvulsant therapy over a six-week course if seizures then subside.

Children presenting with acute stroke require ongoing neurological rehabilitation within a neurosciences centre.

Anticoagulation and management of stroke (or risk of stroke)

As previously discussed, cerebral venous sinuses thrombosis is an associated complication of ENT infections that has an associated mortality. When indicated prompt surgical drainage of the primary infection and collection, relief of mass effect and providing a specimen for microbiology should be performed prior to anticoagulation. It is also important to correct other risk factors in addition to infection such as dehydration and hypovolemia.

Treatment regimens vary between centres and should be discussed with the regional paediatric neurology team. At our institution, children receive anticoagulation with subcutaneous low molecular weight heparin (LMWH), with monitoring of anti-Xa levels for a three-month period. Interval MRI and MRV is performed following this course to determine if the venous sinus has recanalised.

Conclusion

We present the management of intracranial complications based on our unit’s experience and review of the current

Case 2: Frontal sinusitis with Potts Puffy Tumour and frontal extradural empyema

A 13 year old male presented with a two week history of headache, forehead swelling (10 cm above left eyebrow), bilateral peri-orbital oedema and left cervical lymphadenopathy. A CT head scan with contrast showed pan-sinusitis and a large extradural frontal empyema (Figures 6A and B). He underwent combined neurosurgical and ENT surgery. Bilateral frontal minitrephines, uncinectomy, middle meatal antrostomy and image guided burr hole drainage of frontal extradural collection was performed. Pus taken cultured heavy growth of streptococcus intermedius. He was monitored post op with neuro-observations and bloods (WBC and CRP). At day 6, a post-operative CT scan showed an improved picture (Figure 6C). He continued on antibiotics (IV ceftriaxone and oral metronidazole) for 6 weeks delivered via a PICC line and then oral augmentin for a further month. At 2 months, an MRI showed a small residual intracranial collection that did not require any further intervention as he was asymptomatic. He has been advised against contact sports.

Figure 6 Case 2: (A) Axial CT scan with contrast showing a 4x2x7cm extradural collection and Pott’s Puffy tumour. (B) Sagittal view showing frontal sinus opacification and the extradural collection. (C) Day 6 post surgery improved appearances of the frontal sinuses.

A 15 year old male was admitted urgently to our tertiary children’s hospital with a sudden right proptosis following a 5 day history of upper respiratory tract infection, frontal headaches, photophobia, dippiness, intermittent vomiting and fever. A CT scan at his local unit showed a right cavernous sinus thrombosis with sphenoid sinusitis (Figure A). On examination, he was found to have a right sided 3rd and 6th nerve palsy, with mild restriction in all extremes of gaze. Antibiotic treatment was commenced with intravenous Ceftriaxone and Metronidazole. Operative intervention was undertaken urgently on the day of admission and pus taken grew staphylococcus aureus. Bilateral wide endoscopic sphenoidotomies with washout, retrograde ethmoidectomies posterior to anterior with endoscopic septoplasty for access achieved good clearance of the skull base. Further treatment was guided by monitoring symptoms and bloods (CRP and WBC). A repeat CT scan was undertaken due to ongoing headaches and this showed improvement (Figure B). Two weeks of intravenous Ceftriaxone were continued followed by 2 weeks of high dose oral Augmentin. Following surgery, anticoagulation was commenced, low molecular weight heparin. Clirexane, was administered via subcutaneous injection for 3 months duration. Heparin Anti-Xa levels were used to confirm levels were within the therapeutic range. Advice was given to avoid contact sports for 6 months, strenuous exercise for 1 month and to drink at least 2 litres of fluid per day. A CT venogram arranged 3 months following admission (as an alternative to MRV in order to provide more comparative images to Figure 6A and B). Anticoagulation was commenced with intravenous Ceftriaxone and Metronidazole. This treatment regimen is supported by the literature5. Adequate monitoring of a child’s neurological status and CSF pressure is reduced to preserve visual function.

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Monitoring

Accurate monitoring of a child’s neurological status and assessment of evolving neurology is mandatory regardless of whether a surgical, medical or combined approach is undertaken. Blood tests, in particular observing the trends in the C-reactive protein (CRP) and full blood count (FBC); specifically the white blood cell count and neutrophil count can help monitor response to treatment and guide ongoing management.

In a child with a DVST, with symptoms of raised ICP, it is vital to monitor visual function and to liaise closely with ophthalmology colleagues. If there are any concerns regarding visual function, lumbar puncture and pressure monitoring is performed by the paediatric neurology team and CSF pressure is reduced to preserve visual function. Repeat lumbar puncture may be required in addition to medical management including acetazolamide therapy10.

Acute seizures associated with intracranial infection require management by the paediatric neurology team, and introduction of short-term anticonvulsant therapy over a six-week course if seizures then subside.

Children presenting with acute stroke require ongoing neurological rehabilitation within a neurosciences centre.

Anticoagulation and management of stroke (or risk of stroke)

As previously discussed, cerebral venous sinuses thrombosis is an associated complication of ENT infections that has an associated mortality. When indicated prompt surgical drainage of the primary infection and collection, relief of mass effect and providing a specimen for microbiology should be performed prior to anticoagulation. It is also important to correct other risk factors in addition to infection such as dehydration and hypovolemia.

Treatment regimens vary between centres and should be discussed with the regional paediatric neurology team. At our institution, children receive anticoagulation with subcutaneous low molecular weight heparin (LMWH), with monitoring of anti-Xa levels for a three-month period. Interval MRI and MRV is performed following this course to determine if the venous sinus has recanalised.

Conclusion

We present the management of intracranial complications based on our unit’s experience and review of the current
The superior canal dehiscence syndrome

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Abstract
A dehiscence in the roof of the superior semicircular canal is the commonest cause of a third window effect within the otic capsule and may give rise to the superior canal dehiscence syndrome (SCDS). The SCDS can be defined as a combination of a radiological dehiscence in the roof of the superior semicircular canal dehiscence syndrome (SCDS). The SCDS can be defined as a combination of a radiological dehiscence and has an approximately equal gender distribution. Supportive evidence for this comes from the high rate of bilaterality (25%), the high association with other temporal bone abnormalities such as tegmen dehiscence and the lack of association of abnormal temporal bone remodelling in histological studies. No doubt a second hit during life is required which is why presentation occurs within middle age rather than childhood. Approximately a quarter of patients give a history of minor trauma or a Valsalva manoeuvre as the precipitating incident. For others, it may simply be gradual erosion secondary to the natural pulsations of the cerebrospinal fluid (CSF); gradual thinning of the temporal bones has been shown to be associated with advancing age. Much more rarely a specific cause such as a meningioma may be encountered.

Pathogenesis
An SCD is thought to cause a third window effect altering the natural biomechanics of perilymph flow within the inner ear. Under normal circumstances sound enters the inner ear via movement of the stapes at the oval window, causing a pressure wave within the vestibule. The mobility of the round window, when compared to the rigidity of the bony otic capsule, creates a pressure gradient with reduced impedance towards the scala vestibuli of the cochlea, creating a pressure wave within the vestibule. The mobility of the round window, when compared to the rigidity of the bony otic capsule, creates a pressure wave within the cochlea, leading to hyperacusis to these sounds and contributing to the audiometric air-bone gap.

Key words:
Review; surgery; vestibular diseases; superior semicircular canal dehiscence syndrome.

Introduction
The superior canal dehiscence syndrome (SCDS) was first defined by Minor et al in 1998 as a dehiscence in the roof of the superior semicircular canal (SSC) associated with specific clinical symptoms and audi vestibular investigations. SCDS usually presents during middle age and has an approximately equal gender distribution. Although the precise aetiology is unknown, it is thought to be predominantly congenital. Supportive evidence for this literature. Although rare, intracranial infections can complicate ENT infections. It is important to recognise the signs of intracranial infections that may manifest differently in children compared to adults. Early involvement of the paediatric neurology team can help interpret an evolving neurological picture and minimise the associated morbidity and mortality.

Acknowledgements
With thanks to the families of the patients for their consent in using their images to illustrate this article. With thanks to our multidisciplinary colleagues at Southampton Children’s Hospital for their specialist care.

References
Clinical presentation
SCDS tends to present with highly specific clinical features that can be directly related to the pathophysiology described above. Vertigo can arise secondary to loud noise (such as a Valsalva manoeuvre or digital pressure at the tragus) or raised intracranial pressure (such as coughing, sneezing or a Valsalva manoeuvre against a closed glottis). Non-specific vestibular symptoms more commonly associated with vestibular hypofunction, such as dysequilibrium, lethargy or “brain fog”, are also described. Patients typically suffer with bone conduction hyperacusis secondary to hearing their internal body sounds such as their voice (autophony), heartbeat (pulsatile timpanus), eyes or neck movements or footfalls when walking. They may have hearing loss and distortion of sound perception. Noise induced head movement or the feeling of neck strain have been reported.

Examination may show a positive Hennébert’s sign or Valsalva induced nystagmus. Tuning fork tests should be appropriate to the type of hearing loss, whether it be conductive, sensorineural or mixed. The Weber test may be heard in the affected ear even when air-conduction thresholds are symmetrical and normal. A tuning fork placed on a distal bony prominence such as the malleolus may be heard in the affected ear.

Audiology
Pure tone audiometry is likely to show a CDHL most prominent in the lower frequencies. There may be bone conduction with negative decibel (dB) thresholds. It is therefore important to ensure that bone conduction testing is completed even if air conduction thresholds appear symmetrical and normal. Sensorineural hearing loss (SNHL) may also be seen. Both tympanometry and stapedial reflexes should be normal which may help to differentiate SCDS from other pathologies.

Radiology
Computed tomography (CT) scans are the principle diagnostic modality. A high resolution scan with submillimetric collimation is essential to adequately diagnose the dehiscence. Reformatted views should include coronal, sagittal and the plain of the SSC (Figure 1) and Steners (orthogonal to the SSC) [1]. Even with high resolution imaging, overdagnosis is likely because of limitations surrounding the bony resolution due to volume averaging and other artefacts. Temporal bone studies have suggested the prevalence of SCDS is approximately 0.5% whereas the prevalence in CT studies is closer to 10% [2,3].

When evaluating the CT a number of points should be taken into consideration [1]. It has been suggested that a frank dehiscence should only be diagnosed if it can be viewed on 2 consecutive images. Others have found that dehiscences that do not meet this criteria (termed a near dehiscence) might still cause symptoms and require surgical management [4]. Other inner ear dehiscences should also be excluded. The site and size of the dehiscence should be noted as this may have a bearing on surgical access and approach. It is important to note the position of the superior petrosal sinus and its relation to the SSC, cases have been reported where the SCD is directly related to this sinus [5]. There are also a number of well-known temporal bone abnormalities that are associated with SCDS that may impact upon surgical management. The prevalence of bilateral SCDS is approximately 25%. Dehiscence of the tegmen mastoideum occur in approximately 60% and of the tegmen tympani in 15%. Although unusual, there may be associated encephalocoele formation or ossicular erosion. The geniculate ganglion may be dehiscent in the middle fossa floor in up to 50% of cases which will expose the facial nerve to injury when elevating the dura. A low lying middle fossa dura relative to the SSC occurs in approximately 40% of cases and may complicate surgical access.

Magnetic resonance imaging (MRI) is not usually required preoperatively but may be useful in cases where there is asymmetric sensorineural hearing loss or a suggestion of related pathology such as encephalocoele or meningioma. Postoperatively an MRI may help to determine adequacy of SSC obliteration. Heavily weighted high resolution T2 imaging sequences are useful to determine the position and patency of the SSC. Diagnosis of tumours will usually require T1 post contrast images with fat suppression.

Vestibular function tests
A number of vestibular investigations can be used to help confirm the diagnosis. Vestibular evoked myogenic potentials (VEMP) are widely considered the most sensitive and specific test for diagnosing SCDS [2]. They rely on acoustic stimulation (either a click or low frequency tone burst) of the staccato or utricle at a lower than normal thresholds because of the third window effect described above. C-VEMPs make use of the sacculo-colic reflex, whereby stimulation of the saccule causes contraction of the ipsilateral sternocleidomastoid muscle. The normal threshold should occur at approximately 95dB but in SCDS the amplitude increases and the threshold drops, usually less than to 75dB. O-VEMPs make use of the utriculco-ocular reflex whereby utricular stimulation causes contraction of the contralateral inferior oblique muscle. This test also principally uses an auditory stimulus and raised amplitudes are found in SCDS.

Electrocochleography (ECoG) can be used to help diagnose SCDS [6]. This test demonstrates an elevated summating potential when compared to the action potential in the same way as it does with Meniere’s disease but SCDS seems to have a greater sensitivity, particularly when associated with normal hearing thresholds. It has also been demonstrated to normalise following surgical repair.

Other vestibular function tests such as caloric or video head impulse testing are usually normal but may be useful in providing a baseline should concern arise postoperatively that the vestibular apparatus has been damaged.

Diagnostic confirmation
The diagnosis of SCDS should only be made when considering radiological, clinical and audiological investigations results together. Ward et al have suggested that the diagnosis should only be made according to the criteria described in Table 1. Most patients will fit these criteria but uncertainty may arise when symptoms are non-specific, such as chronic dysequilibrium, and vestibular investigations are not clear cut. Patients who are highly symptomatic may warrant exploration even with near dehiscences on CT scan as such patients have been described as improving following surgical treatment. Conversely, patients may have clear dehiscences on CT scan and yet remain asymptomatic and in these situations no further investigations or surgical treatment is warranted.

Management
When considering SCDS management the patient must be counselled carefully as there are significant risks to surgery. Patients with limited symptomatology and little impact on their quality of life may wish to pursue a conservative management course. Unfortunately there is no medical option.

When considering surgery, there are a number of options available both in terms of approach to the SSC and the method of repair used [7]. In essence the SSC can be accessed via either the middle fossa or transmastoid approach. The repair can be carried out via resurfacing of

TABLE 1: Proposed diagnostic criteria for SCDS [2].

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Examples</th>
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<tbody>
<tr>
<td>1. High-resolution computed tomography images (≤0.625-mm slice thickness) reformed in the plane of the superior semicircular canal</td>
<td>Frank dehiscence</td>
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<tr>
<td>2. At least one of the following symptoms consistent with SCDS</td>
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<tr>
<td>A. Bone conduction hyperacusis (in the form of autophony, audible eye movements, audible footsteps, etc.)</td>
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<tr>
<td>B. Sound-induced vertigo</td>
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<tr>
<td>C. Pressure-induced vertigo</td>
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<tr>
<td>D. Pulsatile tinnitus</td>
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<tr>
<td>3. All of the following diagnostic tests indicating a third mobile window</td>
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<tr>
<td>A. Negative bone conduction thresholds on pure tone audiometry</td>
<td></td>
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<tr>
<td>B. Enhanced VEMP responses (low cervical VEMP thresholds or high ocular VEMP amplitudes)</td>
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<tr>
<td>C. Elevated summating potential to action potential ratio on electrocochleography in the absence of a sensorineural hearing loss</td>
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</table>

Figure 1: CT Scan showing superior canal dehiscence.

Figure 2A: Operative photo showing dural elevation over the superior semicircular canal via a transmastoid approach.
obliteration can be carried out via either approach and may involve the use of either cartilage or bone dust with or without fibrin glue. Equally either approach and may involve the use of either cartilage hydroxyapatite cement and thus is only used via the middle fossa craniotomy is fixed in place using the canal, obliteration using a plugging technique, a tegmental defect with bone paté via a transmastoid approach. Figure 2C:

The middle fossa approach entails a suprameatal craniotomy and elevation of the dura. This usually permits direct visualisation of the SD, facilitating repair, at the expense of a small increased risk of intracranial complications such as stroke or seizure secondary to elevation of the temporal lobe, epidural haematoma and a slightly increased risk of facial palsy if the geniculate ganglion is dehiscent. The intracranial risks necessitate an extended hospital stay with an overnight stay in critical care. A transmastoid approach entails a cortical mastoectomy and exterritorization of the air cells lateral to the SSC. The dura can be still elevated off the SSC if required for the resurfacing method, often making use of the natural dehiscence of the tegmen mastoideum (Figures 2A-C). Exposure of the dura may need to be widened and even so access is still limited and visualisation can only be carried out indirectly using a mirror or an angled endoscope. As temporal lobe retraction is negligible with this method it can be performed as a day case procedure. The dura is at risk of tearing with both approaches if it is elevated so there is a small risk of CSF leak and subsequent meningitis. These risks are negated for the transmastoid obliteration which avoids dural retraction altogether. Finally there are a small number of studies evaluating round window reinforcement as a treatment for SCDS on the basis of reducing the overall compliance of the inner ear. This can produce long-lasting reduction of symptoms but has also been reported to provide only temporary relief or worsening of symptoms. Conclusions

The overall success rate of SCDS surgical repair is high; a systematic review carried out in January 2015 found greater than 90% achieved total or partial resolution of the symptoms with no significant difference between approach or repair method. However numbers were small with 90 patients in the obliteration group but only 16 in the resurfacing, 19 in the combined obliteration and resurfacing and 25 in the capping groups.

The decision as to which approach and repair method to use will often rely on the expertise of the surgical team. The site and size of the SCD, whether or not the dura is low-lying and the position of the superior petrosal sinus may influence this decision.

For example a low-lying dura combined with a medially placed dehiscence may make access considerably more difficult and a dehiscence associated with the superior petrosal sinus may influence the surgeon to avoid dural retraction altogether. Finally there are a small number of studies evaluating round window reinforcement for superior semicircular canal dehiscence syndrome. Otol Neurotol (2013) 34:121–6. 3. Park HI, Lee SY, Song J-H, Choi BY, Koo J-W. Electrophysiologic findings in superior canal dehiscence syndrome. Hear Res (2015) 323:61–7.


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Tymanosclerosis

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Abstract
Tymanosclerosis is a chronic irreversible condition of scarring with calcific plaques in the tympanum (middle ear). The extent of scarring may be limited to the eardrum alone (myringosclerosis) or involve the middle ear including ossicles and ossicular joints. Depending on the extent of involvement, the clinical features could vary. In this article, we will discuss an overview of tymanosclerosis and up-to-date evidence based management options available in dealing with this condition.


Key words
Tymanosclerosis, Myringosclerosis, hearing loss

Introduction
Cassohns1 described the condition of deposition of calcareous plaques in the eardrum initially in 1734. Later on in 1873, Von Trotzsch2 described the changes in the middle ear mucosa and tympanic membrane due to tymanosclerosis (TS) due to an inflammatory process within the middle ear, which could be secondary to infection, autosensitisation or traumatic3. The commonest cause of trauma is iatrogenic – post graft insertion. Hyaline degeneration of the fibrous layer of the tympanic membrane and lamina propria of the middle ear mucosa occurs, with subsequent calcific deposits forming plaques within the mucosa and/or tympanic membrane. Depending on site and extent of involvement, there have been various classifications; the commonly used classification is that of Wielinga and Kerr4 (Table 1). Patients can present with conductive hearing loss dependent on the degree of ossicular chain fixation caused by the TS. Type I tymanosclerosis may often be asymptomatic with normal hearing levels, whereas type IV TS can present with maximal conductive hearing loss.

Shelly and House reported that TS might be found in a third of patients with chronic ear disease5. The rate of TS in chronic otitis media is reported to vary from 9 to 38% and can be bilateral in 50% of these cases6. Treatment options depend on patient’s functional requirements. These include a conservative approach with no treatment or auditory rehabilitation with hearing aids or surgical restoration of hearing. Various surgical treatment options have been described depending on the site of involvement, including removal of TS plaques, mobilisation of the ossicular chain, tympanoplasty, ossicular reconstruction and stapedectomy. However, surgery for TS may not be as successful as for simple otitis media without TS, as surgery in itself can reinitiate the process of further TS.

Surgical procedures described in literature
Myringotomy is the most commonest surgery performed is myringoplasty and excision of plaque from within the tympanic membrane. The success rate of myringoplasty in patients with TS is postulated to be similar to cases without TS. Bayazid7 reported graft uptake of 95.2% in their group of patients with tymanosclerosis, who were operated with single stage underlay tympanoplasty using formaldehyde fixed temporalis fascia. However only 33% of their patients had air-bone gap closure within 20db.

Patients with type II TS involving the ossicular chain in the attic can present with conductive hearing loss depending on the degree of ossicular fixation. There are various surgical techniques reported in the literature, including simple excision of tymanosclerotic plaques and mobilisation of the ossicles to ossiculoplasty and ossicular reconstruction with ossicular replacement prostheses. Atticotomy (a combined approach) is required to access tymanosclerotic plaques in the attic; however, proficient endoscopic technique can obviate the need for mastoidectomy or wide atticotomy. In favourable cases, plaques can be gently teased off the ossicles, without excessive manipulation of the ossicular chain. Potassium titanyl phosphate (KTP) laser can also be used to clear tymanosclerotic plaques with minimal ossicular rocking8. If TS extends medial to the ossicles in the attic, the incus may need to be removed and occasionally the malleus stump or stapes head is removed. However, recently in the UK, the use of glass ionomer products (Seren-Cem granules) has been withdrawn due to risk of bone reabsorption. The choices for interposition with an intact stapes superstructure are multiple but the senior author prefers reconstruction using Kutz clip partial prosthesis and believes this provides the most consistent hearing results in his hands. Recurrent sclerosis can however still occur and could be significantly bothersome.

The experience of the senior author and that of his mentors is that the density, consistency and maneuverability of tympanosclerotic plaques is very variable. Risk of ossicular damage increases when attempting to remove more solid plaques with the inherent risk of worsening hearing. The risk of recurrent TS is ever present and candid informed consent is therefore a crucial part of the management of this challenging pathology. Exploratory tympanotomy +/- treatment is a reasonable procedure to undertake having a low risk of recurrence without the situation is unfavourable to close the ear and cause no harm. Auditory rehabilitation with a conventional hearing aid or referral for consideration of an implantable device are the available options if ossiculoplasty has been attempted or is unlikely to be possible.

In patients with type III TS and stapedial footplate sclerosis, the treatment options include simple mobilisation of the stapes or stapedotomy. The hearing gain with footplate mobilisation is considered to be temporary due to probable further scarring/sclerosis. Possible complications from removal of plaques from the oval window include floating footplate, penilymph fistula, sensorineural hearing loss and risk to the facial nerve. Studies reporting good success rates with stapes mobilisation are all in the short-term period; there are no long-term studies looking into refixation probabilities. Success rates with stapedotomy, stapedectomy and replacement with stapes prostheses are claimed to be good9,10. There have been concerns over possible higher rates of post-operative sensorineural hearing loss in TS patients, however these were more so when patients routinely underwent large fenestration stapedotomy11.

Type IV TS patients have significant sclerosis causing fixation of the entire ossicular chain. In such patients, some surgeons have offered malleostapedotomy, which was described by Fisch et al. for revision stapes surgery in Otosclerosis12. Elevation of tympanomeatal flap towards the anterior spine is performed via an endaural approach. A superior canalplasty is performed in order to have a wider exposure to the ossicular chain in the attic. After confirming ankylosis of incus-malleus joint and stapedial footplate, the incus and malleus head is removed. The anterior tympanomalleus ligament, which is often calcified and involved with tymanosclerosis, is cut to enable mobilisation of the manubrium. The tensor tympani tendon is preserved in order to provide stability to the reconstruction. The stapes is then perforated with a SMART piston (nitinol) is used from the manubrium through the fenestrum in the stapes footplate. Magliulo et al. described good hearing results (closure of air-bone gap within 20db) with this technique in 8 of their 10 TS cases. One of their patients had a subsequent sensorineural hearing loss of 10db and there was no recorded incidence of profound hearing loss in their series.

Recent Advances
With technological advances in middle ear implantation, the applications for these devices are being extended. There are reports of the use of Vibrant Soundbridge in patients with otosclerosis and tymanosclerosis with good functional gain13,14. Stapes coupler, round-window coupler or short/long process incus coupler could be used depending on the ossicular chain status and effect of TS. The indications for these active middle ear implants are however only for patients who have stable hearing levels and have failed a trial of hearing aid due to anatomical constraints or chronic external ear infections precluding use of conventional hearing aid.

Conclusion
Tymanosclerosis is a common condition involving the mesotympanum secondary to inflammation or trauma. Presentation primarily can vary from being asymptomatic and an incidental finding on otoscopy to maximal conductive hearing loss. Treatment options have evolved over the last few decades as described in this article. There is no curative treatment for this condition and management mainly revolves around patient’s functional hearing status and requirements apart from extent of middle ear involvement.

Table 1: Wielinga and Kerr classification

<table>
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<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>I</td>
<td>Ossiculoplasty using conventional hearing aid</td>
</tr>
<tr>
<td>II</td>
<td>Ossiculoplasty using conventional hearing aid</td>
</tr>
<tr>
<td>III</td>
<td>Stapes reconstruction with mobile malleus-incus complex</td>
</tr>
<tr>
<td>IV</td>
<td>Stapes reconstruction with mobile malleus-incus complex</td>
</tr>
</tbody>
</table>

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7. SMART piston (nitinol), 2013.
15. FRCS (ORL-HNS), 2017.
17. FRCS (ORL-HNS), 2017.
Endoscopic ear surgery

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Abstract

The endoscope allows the observer to place their field vision within the middle ear space, providing a contextual and relational view of the anatomy. This method of observing the disease/anatomy relationship often allows the surgeon a superior view and understanding of the pathology compared to the traditional microscopic view. At present, the strength of Endoscopic ear surgery (EES) lies in the exploration of the middle ear cavity, as a single method or as an aid to microscopic surgery, most commonly in cholesteatoma clearance and tympanic reconstruction. However, EES promises to surpass the limits of the middle ear in the future.

A brief overview of the history of EES, endoscopic anatomy of the middle ear and the approach to pathology through the use of the endoscope is provided. A summary of current advances in endoscopic ear surgery technology is included, as it goes hand in hand with the development and growth of this field.

Conclusions: The advantages brought by EES over the past decade to the field of otology are comparable to those displayed by minimally invasive surgery in other areas. A more detailed relational observation of the anatomy, the ability to reach anatomically challenging places, a new and enhanced teaching opportunity, faster post-operative recovery and less pain are just a few of the perceived benefits. Once the approach reaches a higher level of maturity, which will include the publication of long-term outcomes, the method will likely have a significant role alongside other techniques in the field of otology.

Key words
Endoscopic ear surgery, tympanoplasty, cholesteatoma, middle ear, translabyrinthine.


Introduction

Endoscopic exploration of the middle ear was first described in 1967 and endoscopic ear surgery (EES) in its current form has been performed since the early 1980s. However, the evolution of EES has significantly expanded over the last decade, in parallel with improvements in camera, scope and lighting technology and the development of specially designed surgical instruments. This has translated into a significant increase in publications and teaching of the method.

At present, EES is not to be seen as a replacement to traditional microscopic surgery, but rather as a complementary tool. The middle ear is likely to become the sole domain of the endoscope and microscope will still be required for extended middle ear, mastoid and lateral skull base surgery.

The evolution of EES has been facilitated significantly by parallel developments in technology with continual collaboration between otologists and industry. As a result, enhancement of operative field illumination, video fidelity and resolution, as well as specially designed surgical instruments have enabled applications for the transcanal approach not previously seen.

For the surgeon, the endoscope allows a somewhat unparalleled view of the ear’s anatomy allowing to “look around” anatomical hurdles. The positioning of the visual target closer to the objective lens and the source of illumination enhances visual quality. Even though evidence regarding outcomes is being gathered, less invasive procedures often translate into faster recovery times, better cosmesis and decreased levels of discomfort in patients. For trainees and supervisors, the ability to watch the operation in real time provides a valuable teaching opportunity, with trainees showing a preference for this technique and evidence suggesting that the learning curve is shortened.
The lack of incisions for surgical access and egress reduces the operative time. This along with the reduction in burr use often translates into a cost reduction for the surgeon. The technique has limitations, such as loss of depth perception, as well as the need for a secondary procedure for bone grafting.

Endoscopic Anatomy of the Middle Ear

The study of anatomy was the initial motivation when Pontarelli et al introduced a flexible endoscope into the middle ear (ME) in the late 1980’s. Until recently, anatomy and pathology viewing of the ME may well be the most popular use of the endoscope in otology. The lack of incisions for surgical access and egress reduces the operative time. This along with the reduction in burr use often translates into a cost reduction for the surgeon. The technique has limitations, such as loss of depth perception, as well as the need for a secondary procedure for bone grafting.

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traditionally obscure areas, often compromised by cholesteatoma, may have an important impact in the rate of recurrence.11

Understanding the pathological and surgical effects on ventilation routes of the ME in hopes of improving outcomes by reducing recurrence is of importance when performing EES. The endoscope provides a clear view of the epitympanic diaphragm and the possibility of restoring the patency of the isthmus. The isthmus is the primary ventilation route between the superior and inferior compartments of the ME. Figure 5 illustrates the main ventilation routes10,12,13. One of the main goals of endoscopic surgical treatment of middle ear pathology involves the unification of the upper and lower ventilation units of the Epitympanum through the enlargement of the tympanic isthmus and the creation of an accessory route through the unification of the upper and lower ventilation units of the ME in hopes of improving outcomes by reducing recurrence is of importance when performing EES. The endoscope provides a clear view of the epitympanic diaphragm and the possibility of restoring the patency of the isthmus. The isthmus is the primary ventilation route between the superior and inferior compartments of the ME. Figure 5 illustrates the main ventilation routes10,12,13. One of the main goals of endoscopic surgical treatment of middle ear pathology involves the unification of the upper and lower ventilation units of the Epitympanum through the enlargement of the tympanic isthmus and the creation of an accessory route through the tensor fold.12

Indications and Applications

The most appropriate surgical technique (i.e. endoscopic vs. microscopic) lies within a spectrum according to the anatomy and pathology encountered. It ranges between completely endoscopic to completely microscopic, with a combination of both in between (Figure 6).14

The scope of applications in EES ranges from simple in-office examination all the way to the complex exploration of the petrous apex and cerebellopontine angle. In some of these procedures the endoscope has yet to prove significant advantage over the microscope, being used mostly as an adjunct and in some cases only for inspection. Additionally, long-term outcomes are not available for most applications of the technique. Table 4 lists the procedures in which EES has been used and elaborates on the findings of currently available literature.

The most common indications for EES are cholesteatoma clearance and tympanic and ossicular reconstruction. The strengths of EES are particularly evident in cases of congenital cholesteatoma, or in acquired cholesteatoma that occupies the mesotympanum primarily and extends adjacent with minimal mastoid extension and where the tegmen is low.11 In such instances the possibility of a mastoid-sparing procedure despite the presence of a large cholesteatoma is attainable which may be of great importance, as some authors believe that mastoid preservation maintains the physiology of the ear by upholding gas-exchange and ventilation between middle air-cells and the middle ear, thus improving the chance of a successful outcome and potentially lowering recurrence rates.10,13,14

Importance of Endoscopic Technology in safe surgery

Adequate visualization is paramount to the safe application of EES. Logically, the better image quality being delivered to the surgeon, the less likely complications will occur. An appropriate image depends on the integrity and quality of the components of the “optical chain”. The chain begins on the target tissue (ear) and ends on the operator’s eye (surgeon). It is composed by all the elements that transfer light from one end to the other, such as the endoscope, the camera, the light cord, the monitor and finally the surgeons’ eyes. The optical chain is only as good as its weakest link, so it is advisable to review each component before and during surgery and only proceed to operate in ideal conditions; updating the video system’s technology whenever possible is also worthwhile.

The medical technology industry is making great strides in the development of elements that provide unprecedented image quality. These include high-definition and ultra-high definition monitors, specialized video processing sensors in the camera units, faster interface between processors and monitors, processing of image that

Table 2: Description and classification of the Epitympanum10,12

<table>
<thead>
<tr>
<th>Epitympanum: Its divided into two compartments</th>
<th>The Cog</th>
<th>Posterior Epitympanum</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boundaries:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Anterior: Root of the Zygomatic arch</td>
<td>Divides the anterior and posterior compartments.</td>
<td>Bony septum arising from the Tegmen tympani in front of the malleus head in direction of the Cochleaiform process.</td>
</tr>
<tr>
<td>• Superior: Tegmen Tympani</td>
<td></td>
<td>Mostly occupied by the head of the malleus, body and short process of the incus.</td>
</tr>
<tr>
<td>• Lateral: Tympanic bone and Chorda Tympani</td>
<td></td>
<td>Its lateral portion is further subdivided by the Incudomalleolar fold into superior and inferior lateral attic spaces.</td>
</tr>
<tr>
<td>• Medial: Genuculate ganglion, separated from it by bony wall</td>
<td></td>
<td>• Superior lateral attic: Extends superiorly from the Tegmen tympani and inferiorty to the VII nerve (2nd portion). Ventilates f and communicates to the Mesotympanum through the isthmus anteriorly and the Aditus ad Antrum posteriorly.</td>
</tr>
<tr>
<td>• Inferior: Tensor fold (Incomplete in ~25% of cases) (12)</td>
<td></td>
<td>• Inferior lateral attic unit: Inferior to the epitympanic diaphragm, and communicates with the Mesotympanum directly.</td>
</tr>
<tr>
<td>• A complete and horizontal tensor fold has been found to be associated to attic cholesteatoma.12</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ventilation units (Or routes for the ventilation trajectories):</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Superior unit (or superior attic): formed by The lower lateral attic and the medial attic.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Inferior unit: Prussak’s space</td>
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Figure 4: Anatomy of the Protympanum


Figure 5: Independent ventilation routes in the middle ear

A–B: ventilation route to the epitympanic compartments through the isthmus (blue arrow) in close relation to the tensor tendon and the stapes; C: ventilation route to the Prussak’s space through the posterior pouch (orange arrow). D: Complete tensor fold (present in ~75% of the population). In this cases the isthmus provides the only ventilation pathway. E: Incomplete tensor fold, Allows ventilation from the Supratubal recess to the attic. F: (Osseous removed) Blue line: Main ventilation pathway. Green: Accessory ventilation pathway, when the tensor fold is incomplete.

M: Malleus, spm: short process of the Malleus, sc: stapes, ec: eustachian form process

Figure 6: Classification of endoscopic ear surgery and approximate proportion (indicative) of cases performed under that class.

Class 0: 100% Microscopic, Class 1: Endoscopy only for inspection, Class 2a: Endoscopy used in less than 50% of surgery, 2b: Endoscopy used in 50% or more of the surgery, Class 3: 100% Endoscopich

Table 4: Applications of Endoscopic Ear Surgery
data

In-office examination, debridement and wax removal:
- Positive response form patients when shown ear examination on the screen.
- Beneficial for training to view examination/procedures in real time.
- A study showed that endoscopic wax removal was quicker and better tolerated than microscopic removal.

Myringotomy and ventilation tube (grommet) placement:
- Useful in curved and stenotic ear canals.
- May require a longer procedure time when compared to the microscope.
- Danger of the surgeon did influence EAC trauma outcome (The contralateral EAC to the surgeon’s dominant hand suffered more trauma).

Expiratory tympanotomy and Ossicular Chain Reconstruction:
- Common indications include conductive hearing loss and secondlook procedures, but apply to OCM, ME, tympanostomy tube.
- A controlled tympanomastoid flap and its precise reposition help a quick and effective recovery.
- We will require cannula and removal of sodium less often when compared to the microscope.
- The use of laser and microtools is helpful in the management of fixations and adhesions.
- The placement of autologous tissues, bone cement and prostheses is quite achievable through the one-handed technique, but the need for two-handed placements is required on occasion. Reusing of the tool to the microscope should always be kept in mind.

Cholesteatoma:
- EES has shown to fulfill the goals of cholesteatoma surgery: complete removal of disease, assurance of long-term prevention of recurrence, and preservation of or improvement in hearing.
- Allows avoidance of mastoid clef for the sole purpose of approach.
- Usually achievable completely endoscopically when confined to the middle ear spaces, not going beyond the lateral semicircular canal, a ST type C or the deep into the mastoid.
- Requires excellent visualization and space optimization, complete degloving of the TM, if the ME is significantly occupied.

Tympanoplasty:
- Evidence suggests that EES can shorten the operative time without compromising the hearing outcome and graft uptake.
- The use of this technique can be helpful in these cases, especially in complex cases requiring complete ossiculoplasty.
- The narrowest segment of the ET is closest to the protympanum and not reachable transnasally.

Stapes surgery:
- Controversy regarding true role of EES.
- Opponents argue that as a one-handed technique may present more limitations than advantages.
- The main advantage of the cholesteatoma tympanotomy is to avoid another incision with reduced need for medial wall cutting, overcoming the limitation of the microscopic linear vision which improves to the anterior crus and less need for bone removal and manipulation which decreases damage..

Exustacan tube surgery:
- The use of this technique is still lacking and may be negligible in the hands of an experienced surgeon.

Cochlear implant:
- Applies to paragangliomas, cribriform tumours and benign ossous masses.
- Requires excellent visualization and space optimization, complete degloving of the TM, if the ME is significantly occupied.
- The use of microtools and bipolar cautery to achieve mass debulking and hemostasis are recommended.
- The use of bipolar helps hemostasis but thermal damage to V nerve must be actively avoided.

Benign tumors of the middle and inferior ear:
- Reported applications of EES include:
  - Suprapetrotympanar Approach: Allows access to the geniculate ganglion and periganglion area.
  - Transcanal Transpetrotympanar Approach: Access to the cocholeasigmoid region and is AC is granted without manipulation of the meninges.
  - Endoscope-assisted Vestibular Schwannoma Surgery
  - Infrachannel Approach: Provides access to the petrous apex.
  - Endoscope-assisted Superior Canal Dehiscence Management.

Endoscopic Ear Surgery:
- Optimalizes color, brightness and overall image fidelity and more efficient and light sources. Further progress with 3D imaging, infrared, and spectral imaging color enhancement cameras, will likely become routine surgical aids in the near future.

Complications and Pitfalls in EES:
- EES has several potential problems, and attention to possible risks and disadvantages to the technique is essential for safe practice. The microscope offers the possibility of a two-handed technique and binocular vision with depth of perception over the endoscope. In procedures these when elements are indispensable, it is advisable to opt against EES, or use it only in conjunction. However, particularly in the case of depth perception, these perceived downsides may become less important as the learning curve grows.

Potential risks can arise in EES secondary to poor image. These include thermal injury from heat originating from the endoscope’s tip or inadvertent vascular injury or disruption. The first can be avoided by frequently wiping and removing the endoscope shaft from the ME, utilizing suction often and maintaining a low intensity setting on the light source (<50%). The second complication can be avoided by maintaining screen visualization whilst the scope is in the middle ear, being particularly careful with angled endoscopes and introducing instruments ahead of the endoscope to keep them in the visual frame.

Other risks are not directly linked to poor image quality. These include thermal injury from heat originating from the endoscope’s tip or inadvertent vascular injury or disruption. The first can be avoided by frequently wiping and removing the endoscope shaft from the ME, utilizing suction often and maintaining a low intensity setting on the light source (<50%). The second complication can be avoided by maintaining screen visualization whilst the scope is in the middle ear, being particularly careful with angled endoscopes and introducing instruments ahead of the endoscope to keep them in the visual frame.

Conclusions:
- Endoscopic ear surgery is a promising approach for procedures that lend themselves for the minimally invasive technique. The method provides superior visualization to the microscope in several “hidden areas” of the middle ear. The authors’ observations of operating on middle ear pathologies with EES and observing trainees learning through the aid of the endoscope suggest that it provides global benefits. A methodical, stepwise, and careful approach to the technique is advised for those who are getting started, but results seem favorable once the learning curve has been achieved.

Disclosures/Conflicts of Interest
- No actual or potential conflicts of interest are involved in this paper.

References:
The purpose of vestibular function tests (VFTs) is to determine whether the peripheral vestibular system and associated central pathways are functioning normally. Tests may determine loss of unilateral or bilateral function and some aspects of testing can reveal central causes of balance symptoms.

VFT's examine the integrity of the vestibulo-ocular reflex (VOR) which is driven by information from the visual, vestibular and spinal afferents in the brain stem. The VOR is responsible for maintaining visual focus when the head is moving. The presence of spontaneous and gaze nystagmus, the accuracy of the pursuit and saccadic tracking systems which are controlled by central pathways, in addition to changes in the horizontal and vertical planes for each patient prior to testing. Recordings are taken with the patients eyes open in the dark. ENG utilises electrodes placed around the eyes to indirectly record the electrical potential difference between the front and back of the eye during eye movement. ENG still has a place in preference to VNG in certain situations in which the patient is claustrophobic or anxious and cannot tolerate the video goggles.

**Key words**

Vestibular function tests, Vestibular assessment, Balance, Dizziness.

**Introduction**

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**Assessment protocol**

The process always begins with a detailed patient history and examination. This will allow the clinician to determine the patient's presenting complaint and potential underlying pathology. The focus of the assessment will depend on the patient's presentation and any other relevant clinical findings. The assessment will be structured around a series of questions and tests designed to identify any underlying pathology and to determine the appropriate management plan.

**Pre-assessment questionnaires**

Self-reporting helps assess how the dizziness or unsteadiness impacts on the patients' functional, physical and emotional quality of life. One such questionnaire that is commonly used is the Dizziness handicap inventory (DHQ). The questionnaire is scored out of 100, the higher the score the more severe the handicap. Such questionnaires can be helpful since objective test results do not always correlate well with the patient's subjective experience. They are also helpful in measuring change after an intervention, such as balance rehabilitation.

**Vestibular function testing**

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**Abstract**

Balance disorders can be extremely debilitating and accurate diagnosis can prove challenging. Vestibular function tests are an often mysterious array of tests requested by the clinician to investigate patients in whom the initial cause of their disequilibrium is not clear.

In this paper, we outline the tests that are available, how they are performed and the information that they will provide to aid correct diagnosis and treatment.

**J ENT Masterclass 2017; 10 (1): 57 - 62.**
**Direct observation of eye movements**

This is essential prior to testing to ensure visual acuity, conjugate eye movement, a good range of eye movement in the horizontal, vertical and oblique planes and to check for congenital nystagmus, squints and spontaneous nystagmus. Disconjugate eye movements may result with certain central lesions. Presence and direction of nystagmus, as well as response to visual fixation, may provide information differentiating peripheral and central causes of vertigo.

**A basic screen of the VOR can be carried out at this time and includes:**

1. Dynamic Visual Acuity Test (DVA) assesses impairments in the patients’ ability to perceive objects accurately whilst actively moving the head in the horizontal plane. When the VOR is impaired, visual acuity degrades during head movement, resulting in retinal slip, producing a blurred image. Sitting two meters from a visual acuity chart the patient reads the lowest line where all the letters can be correctly identified. The head is dipped 30 degrees forward to bring the lateral canals into the plane for testing before moving side to side at a frequency of 2Hz (two side-to-side movements every second) and less than 20 degrees rotation. With the head in motion the patient is asked to maintain visual fixation. The patient visually fixates on a target for 15 seconds, with mental alerting to inhibit nystagmus suppression; visual fixation is then removed for a further 15 seconds. This is repeated, gazing 30 degrees to the right and to the left. Gaze nystagmus due to a peripheral cause is horizontal, beating in the same direction irrespective of gaze direction, and can be suppressed on fixation, following Alexander’s Law. Gaze nystagmus of central causes may be horizontal, torsional or pure vertical, can change direction in one gaze position, or is enhanced with visual fixation. Other types of eye movements maybe seen; for example, large amplitude square waves with fixation are always a central sign, whereas those of smaller amplitude with fixation are normal.

2. Head Thrust Test, first described in 1988 by Halmagyi and Curthoys, detects severe unilateral loss of semicircular canal function, where the canal paresis is greater than or equal to 63%. The test involves observing whether or not the patient can maintain visual fixation on a target after brief and rapid head thrusts in the planes of the semicircular canals, or whether corrective eye saccades are required to regain fixation on the target. A positive result is recorded if the patient needs to make a corrective saccade and is positive to the side that causes the corrective saccade to be made. The sensitivity of the test is 34% with 100% specificity.

**Modified clinical test of sensory interaction on balance (mCTSIB)**

The mCTSIB is an assessment of how well a patient integrates visual, proprioceptive and vestibular inputs. Patients are required to undertake 30 sec trials, with each trial involving compromising one of the inputs. Patients with uncompensated unilateral or bilateral peripheral vestibular deficits have been shown to have difficulty when visual and proprioceptive inputs are disrupted. The video head impulse test (vHIT) is an objective version of the Head Thrust Test. Using infrared goggles to measure the gain of the VOR, this observes any corrective saccades in response to short, brisk head impulses in the plane of the semicircular canal (SCC) whilst the patient attempts to maintain visual fixation on a small fixed target. The advantage over the subjective Head Thrust test is that objective data is recorded for analysis, with both overt and covert saccades being detected; only overt saccades can be seen with the naked eye in the Head Thrust test. A reduced VOR gain with the presence of overt and/or covert saccades suggests a weakness of SCC function in the plane of testing, on the same side to which the head was impulsed prior to the corrective response (Figure 1).

**POSITIONING MANOEUVRES**

The Dix-Hallpike Manoeuvre is a routinely used positioning manoeuvre performed to observe nystagmus associated with both peripheral and central positioning vertigo - both posterior and anterior canal Benign Paroxysmal Positional Vertigo (BPPV) are detected. The Roll Test is usually required to identify lateral canal BPPV, however occasionally the Dix-Hallpike will also reveal it. The Side Lying Test can also be used when the Dix-Hallpike manoeuvre may not be possible due to physical limitations. BPPV becomes more common with increasing age, with studies revealing that approximately 40% of patients over 65 years of age referred to a neuro-otology clinic for dizziness or imbalance were suffering from BPPV. If untreated in the elderly, BPPV is associated with increased risk of falls.

To perform the test, the patient’s head is turned 45 degrees to the test side and the patient lain from sitting to supine, with both eyes closed. The patient’s head is turned 45 degrees to the test side and the patient lain from sitting to supine, with both eyes closed. The test if repeated for both sides (Image 1 & 2). If torsional nystagmus is present, it’s latency of onset, direction and duration are noted. With repeated manoeuvres nystagmus is fatiguable - it’s intensity should reduce or be abolished. Abnormal results may indicate the need for canalith repositioning procedures, such as the Epley Manoeuvre.

**OCULOMOTOR ASSESSMENTS**

**Gaze Testing**

The purpose of the gaze test is to identify the presence of random or spontaneous eye movements, primarily nystagmus, which interferes with the eyes’ ability to maintain visual fixation. The patient visually fixates on a central target for 15 seconds, with mental alerting to inhibit nystagmus suppression; visual fixation is then removed for a further 15 seconds. This is repeated, gazing 30 degrees to the right and to the left. Gaze nystagmus due to a peripheral cause is horizontal, beating in the same direction irrespective of gaze direction, and can be suppressed on fixation, following Alexander’s Law. Gaze nystagmus of central causes may be horizontal, torsional or pure vertical, can change direction in one gaze position, or is enhanced with visual fixation. Other types of eye movements maybe seen; for example, large amplitude square waves with fixation are always a central sign, whereas those of smaller amplitude with fixation are normal.
Smooth pursuit
This is a tracking test that measures the ability of the patient to maintain fixation on a sinusoidal moving target over a range of frequencies, typically 0.2 - 0.7 Hz. It is the most sensitive test of central causes of disequilibrium. Smooth sinusoidal eye movements that correspond to the target motion are classified as normal; defective tracking suggests a central lesion. If symmetrical, this suggests that the cause is basal ganglia or cerebellar in nature; if asymmetric, a focal lesion involving brainstem or parieto-occipital region is suggested.

Saccades
Disorders of the central nervous system can sometimes be revealed by saccadic testing. Computer software allows the analysis of saccadic latency, velocity and accuracy of the eyes’ ability to track random movements of a target. Hypometric and hypermetric saccades imply a cerebellar lesion. Hypermetric saccades, when present more than 50% of the test time and greater than 2 degrees per second, are abnormal. Saccadic slowing can result from a variety of central lesions or, most commonly, from the effects of medication or tiredness.

Static Position tests
Static position tests are used to investigate the vestibular system’s response to a change in head or body position as compared with a neutral head or body position. Recordings are made with and without fixation; the presence or absence of nystagmus and/or changes in nystagmus, as well as the patient’s subjective report of symptom changes, are noted.

Head Shake Test
The head is shaken in the horizontal plane at a frequency of 2Hz to an angle of 30 degrees, after a baseline recording for 10 secs has been made. The test is considered positive if at least five nystagmus beats were seen. Any spontaneous nystagmus noted in the baseline recording is subtracted from the induced head shaking nystagmus. If its value is at least 3 deg per sec it is defined as significant. The sensitivity and specificity of testing for unilateral weakness of greater than or equal to 25% is 27% and 88% respectively. If the nystagmus changes direction in a single position it is a central indicator as is a loss of fixation suppression.

Caloric Tests
Following the work of Fitzgerald and Hallpike (1942) who described the technique, caloric tests were introduced into clinical practice in the 1940’s. The lateral semicircular canal in the test ear is stimulated by creating a temperature difference, using warm and cool air or water, seven degrees above and below body temperature respectively. The presence, direction and maximum slow phase velocity of any nystagmus that exists during and following irrigation of the right and left ears is compared (Figure 2). Responses to warm and cool stimulus differ: warm produces an excitatory response, cool an inhibitory response. Significant differences suggest a weakness of the canal having the lowest response. Although caloric tests are a test of peripheral function, failure of the patient to be able to adequately suppress the caloric response when fixating on a target may suggest a central lesion.

Vestibular Evoked Myogenic Potentials (VEMPs)
Vestibular evoked myogenic potentials (VEMPs) are an electrophysiological test of saccular, utricular and inferior and superior vestibular nerve function. A potential is generated in response to air or bone conducted tone bursts, typically a 500Hz stimulation. There are two VEMP tests, the cervical VEMP (cVEMP) that measures a relaxation response in the sternocleidomastoid muscle (SCM) which occurs in response to sound. The SCM must be contracted; this can be achieved typically by the seated patient turning the head or, from a lying position, lifting and turning the head (Images 3 & 4). The P1/N1 waveform complex is
recorded ipsilaterally to the stimulus side via one electrode on each SCM and one in the supra-sternal notch.

The ocular VEMP (oVEMP), N1/P1 waveform complex is recorded contra-laterally to the stimulus side. Two electrodes are placed under each eye and one on the forehead and an activation response from the ocular muscles is measured. The ocular muscles are activated by keeping the head in its natural resting position whilst the patient is seated and allowing the eyes to adopt an upward gaze. The waveforms are analysed for significant right versus left asymmetry.

Results of VEMP testing can provide information about pathology that affects balance and/or cause dizziness, such as superior semi-circular canal dehiscence and vestibular neuronitis (Figure 3).

References
considerations in NF2 management and decision-making. Gold standard care is delivered by multidisciplinary teams.

Over the last 15 years the indications for ABI have expanded. Colletti’s group in Verona led the way initially implanting non-tumour adults and subsequently children and infants that were not candidates for cochlear implantation (CI). This has led to a steady increase in ABI for patients with congenital aplasia of the cochlea nerve or cochlea, trauma cases including avulsion of the cochlea nerve(s) and situations where the cochlea is not implantable due to otosclerosis or ossification from infection.

Contraindications to ABI include severe developmental delay, a central cause of deafness, brainstem deformity and significant medical co-morbidities (Table 1).

The Multidisciplinary team (MDT) for ABI should consist of an otologist or neuro-otologist, paediatric neurosurgeon, implantation-experienced audiologist, electrophysiologist, speech and language habilitation/rehabilitation specialist, experienced neuroradiologist, and paediatric neuro-an aesthetist with a paediatric intensive care unit.

Outcomes

Outcomes of ABI are far more variable than outcomes of cochlear implantation (CI). Cochlear implant placement in the scala tympani naturally wraps the array around the modiolus close to the spiral ganglion neurons with automatic tonotopic organisation. In ABI there is comparatively uncertain positioning in the lateral recess of the fourth ventricle adjacent to the cochlear nucleus. There is then reliance on patient perception and feedback of auditory tones with pitch ranking for subsequent programming of the auditory brainstem implant. Patient factors, either NF2/tumour related or from concurrent medical problems or neurodevelopmental delay may cause disabilities that interfere with programming and auditory habilitation.

In tumour cases intra-operative factors either from mechanical or ischaemic damage to the cochlear nucleus or brainstem are also likely to influence outcomes. Furthermore, habilitation and performance take longer with ABI and may not plateau for several years compared to months for most cochlear implant users.

Initial habilitation benefits with ABI ranged from sound awareness, to identification of some environmental sounds, to face-to-face communication with test results of 43 paediatric (non-tumour) patients implanted with an ABI. 47.9% of ABI users achieved a degree of speech perception with Categories of Auditory Performance (CAP) scores >4 (5= Understanding common phrases without lip reading), however, patients with other disabilities did not benefit as much.

Assessment

The English consensus protocol for auditory rehabilitation in NF2 recommends annual audiological assessment as part of the regular review process for NF2. This involves pure tone audiometry (PTA) and speech discrimination testing with the Arthur Boothroyd (AB) word test as well as hearing aid review to maintain optimum performance. If speech discrimination scores (SDS) fall below 50%, Bamford-Kowal-Bench (BKB) sentence testing is recommended. This should trigger assessment for auditory implantation if scores are below 50% in best-aided conditions at the optimum sound intensity (identified by the SDS), in accordance with the National Institute for Health and Care Excellence (NICE) guidelines.

Auditory implant assessment additionally involves free field speech discrimination testing with and without lip-reading with hearing aids and potentially environmental noise discrimination tests as well. Sudden deteriorations or noticeable progressive hearing loss in NF2 require interim review and re-assessment.

For paediatric or non-tumour patients referral to an auditory implant centre is the first step (Figure 2). Infants with congenital deafness are assessed in the same manner as for CI with auditory brainstem response (ABR) testing, age appropriate behavioural testing and MRI. They have a developmental assessment to look for communicative intent and check whether they are meeting appropriate milestones. A psychologist, teacher of the deaf and speech and language therapist may carry out a combined baby assessment. A clinical geneticist arranges genetic testing in parallel. When there is abnormal anatomy with likely aplasia of the cochlear nerves or cochlea, computerised tomography (CT) is also carried out. General anaesthetic (GA) may be required for cross sectional imaging and/or ABR to obtain optimum results and confirm diagnosis. Uncertainty regarding suitability for cochlear implantation in the presence of abnormal anatomy or trauma, with a suspicion that ABI may be necessary, warrants referral to an ABI Centre depending on the skill set and expertise of the hearing implant centre.

Subsequent expert review of imaging and MDT discussion occurs in order to formalise an appropriate management strategy. The options are then discussed with the family. In depth and careful counselling is paramount.

Table 1: Indications, contraindications and surgical risk of ABI

<table>
<thead>
<tr>
<th>Indications</th>
<th>Contraindications</th>
<th>Surgical risks</th>
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<tbody>
<tr>
<td>Pre-lingual:</td>
<td>Central cause of deafness</td>
<td>Bleeding</td>
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<tr>
<td></td>
<td>Significant brainstem deformity</td>
<td>Haematomas</td>
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<tr>
<td></td>
<td>Severe developmental delay</td>
<td>Intracranial haemorrhage</td>
</tr>
<tr>
<td></td>
<td>Medical co-morbidities precluding neurosurgery</td>
<td>Infection</td>
</tr>
<tr>
<td>Post-lingual:</td>
<td>Low life expectancy</td>
<td>Meningitis</td>
</tr>
<tr>
<td>Pre-lingual:</td>
<td>Facial palsy</td>
<td>Cerebellopontine fluid leak</td>
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<tr>
<td></td>
<td>Lower cranial nerve injury</td>
<td></td>
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<tr>
<td></td>
<td>Cerebellar oedema</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hydrocephalus</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Cochlea aplasia / malformation</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Cochlea nerve aplasia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Trauma / cochlea nerve disruption</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Ossification/oblation due to meningo, supportive labyrinthis or other vascular inflammatory causes</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Tinnitus</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Imbalance</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Post-craniotomy headaches</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Epilepsy</td>
<td></td>
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<tr>
<td></td>
<td>Device failure</td>
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</tbody>
</table>

Figure 2: London ABI service pathway overview.
For patients with limited auditory response and either hypoplastic or an abnormal number or configuration of nerves within the internal auditory canal, transtympanic electrical auditory brainstem response (EABR) testing is useful in assessing the integrity of the cochlear nerve by measuring its ability to transmit an electrical stimulus. GA is required for this test which has been shown to correlate well with CI evoked EABR, and aids decision making in patients with CN aplasia/hypoplasia.15,16 It may also aid decision making pre-operatively in NF2 patients. A positive response is a good predictor of potential benefit from a cochlear implant (provided the implant can be inserted). If there is no response it does not necessarily mean there are no cochlear nerve fibres; no response could occur with poor electrode positioning or anatomical variation. Transtympanic EABR helps for pre-operative counselling of patients and families with abnormal anatomy. If there is a positive response it is appropriate to offer CI and the patient is likely to benefit. At the time of CI surgery, a CI evoked ABR can be carried out to confirm whether an ABR is present. It’s absence may not mean the patient will not benefit from the CI and a period of time to see if a patient develops some auditory stimulation may be necessary. More recently, a custom-made electrode array can be used to test for an ABR in a similar manner as CI evoked ABR but without the cost of opening a cochlear implant. Appropriate counselling and informed consent would have already been obtained for CI surgery and discussion regarding potential future ABI depending on results. Same surgery subsequent cochlear implant insertion occurs if the response is favourable. With ambiguous anatomy and a negative response the team may decide that ABI is the only option, which would then be arranged in due course.

For NF2 patients the decision to operate/offer an auditory implant is complex and is embolded with MDT decisions about patient tumour management.17,18 It involves consideration of patient factors, tumour factors - including size and growth rate, the contralateral ear/tumour and facial nerve function. CI may be possible and assessment of auditory nerve function is therefore important to try and predict whether a functional auditory pathway still exists from the cochlea. Promontory electrical stimulation in an apparent dead ear is crucial.19 A trans tympanic needle placed on the promontory to attempt to elicit an auditory percept in a conscious patient. Auditory perception suggests a functioning cochlear nerve (CN) but does not predict outcome with a cochlear implant.19

The presence of post-tumour resection ABR, cochlear nerve action potential (CNAP), EABR, or CI evoked EABR provides some prognostic information and is an indicator of residual auditory nerve transmission of electrical stimulation. In this situation CI may be worthwhile, in contrast if these are negative and/or the auditory nerve electrically definitely has been sacrificed then ABI is the auditory rehabilitation choice. Counseling of patients requires comprehensive explanation and may take multiple sessions. As well as an introduction to the implants and an explanation of the risks of surgery and alternatives as part of formal consent, there are a number of key areas that must be emphasised: pictorial explanation of the site/positioning of the implants is advised. In NF2 the possibility of the implant being a ‘sleepier’ device while serviceable hearing is present in the contralateral ear is important to address in specific cases: the range of possible outcomes and the difficulties with predicting prognosis/benefit; managing patient expectations is crucial; an explanation of the immense workload and commitment required by the patient and team for rehabilitation.15 Meningitis risk should be explained as part of the consent and pre-operative vaccinations two or more weeks before surgery are advised as for CI. Introducing the patient/family to one who has previously been through surgery and the process of auditory habilitation with an ABI is beneficial.

Surgical considerations

The aim of surgery in NF2 is total tumour removal with preservation of the facial and cochlear nerves. Surgical approach should be chosen based on experience and selecting the one most likely to enable optimal access to achieve the above. For unknown reasons VS in NF2 are ‘more aggressive’ than sporadic VS, which makes preservation of the cochlear and facial nerves more surgically challenging.15 In depth discussion of the surgical options in NF2 is beyond the remit of this article. Auditory implantation usually takes place after tumour resection in the same operation. If a retrosigmoid approach has been used then a cortical mastoidectomy and posterior tympanotomy to enable EABR testing and potential subsequent CI are necessary. Meticulous surgical technique is essential to minimise brainstem trauma from direct physical damage or collateral effects of electrocautery. A secondary effect of electrocautery is neural damage from activated stimulator and electrocautery. There is therefore an argument for early division of the vestibulocochlear nerve at the brainstem. This decision may be made early with a large tumour or cases where it is deemed unlikely to be able to preserve the cochlea nerve. ABI would then be performed. In paediatric ABI, low blood and cerebrospinal fluid volumes below the age of 1 and the inherent risks of the surgery have led to a consensus on the optimum age for surgery between 18 and 24 months.15 The rationale for surgery ‘as young as safely possible’ follows the same logic as for early CI in order to harness neural plasticity. Retrosigmoid is the preferred approach. To prevent migration of the receiver-stimulator, a well or anterior lip is usually drilled on the skull to recess the ABI, which is placed in a tight peristomal pocket. A channel for the array can also be drilled from the well to the edge of the cranioectomy.

Particular challenges for ABI in NF2 include distortion of the brainstem anatomy making placement more difficult than in non-tumour cases. Subsequent remodeling of the brainstem to fill the tumour void may lead to movement or displacement of the array. In paediatric ABI the brainstem may be congenitally abnormal. When the vestibulocochlear nerves are absent and/or the facial nerve has an abnormal course to the brainstem then identification of the foramen of Luschka is more challenging (Figure 3).

Intraoperative implant evoked ABR testing is a critical step in ABI surgery to optimise paddle position. Usually the four corners of the array are activated in succession and based on presence or absence of ABR, movement of the paddle is discussed with the surgeons. Subsequent re-positioning with further testing occurs until placement is optimal. Anaesthetic vigilance and close attention to the electrocardiogram is warranted in case of non-auditory stimulation. The silicone mesh on the ABI paddle can be trimmed prior to placement. The advantage of the mesh is that it aids adherence to the brainstem, conversely this is a disadvantage when re-positioning. Maintenance of the optimum position is crucial but can be especially challenging after tumour removal in NF2 cases. If the implant is placed deep in the lateral recess the brainstem itself may hug the implant. Fibrin sealant, Tissel (Baxter Biosciences, Vienna, Austria) and dural sealant DuraSeal (Covidien, Mansfield, MA, USA) can be used to help prevent displacement as retraction is relaxed and closure takes place. Anchoring the array in the cavity or at the cranioectomy edge should also be considered in tumour cases.

Device activation and rehabilitation

Adult ABI activation is done with the patient nil by mouth in either an anaesthetic room or recovery where there is cardiac monitoring and an anaesthetist present. This is due to the possibility of non-auditory stimulation and in particular the risk of vagal stimulation causing bradycardias, tachy arrythmias or the need for a secure airway. For paediatric patients in our centre, once the child has fully recovered from surgery (usually around 4 weeks post-operatively) they are admitted for further implant evoked ABR under GA. This is to establish thresholds and stimulation parameters, as feedback from the patient will be limited at switch on and impedances may have changed since surgery. The electrodes with the clearest responses

Figure 3: Choroid plexus being elevated to reveal the foramen of Luschka just superior to the root of cranial nerve IX in a left ABI case.

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can subsequently be targeted at switch-on, which is done a couple of weeks later in the same setting as for an adult.

Programming of the ABI involves stimulating each electrode in turn and then in groups across the array. Threshold and comfort levels are varied. For adults, auditory perception with feedback on loudness and pitch enable programming with the aim of access to a good range of frequencies whilst avoiding non-auditory stimulation. Speech and language therapists and habilitation specialists play a crucial role helping the patient to learn to recognise sounds. Motivation, commitment and perseverance are important for the patients to be able to maximise their potential benefit from their implant. Assistive listening devices may also help to optimise performance in day-to-day situations.

Conclusions

Auditory brainstem implantation has been shown to be safe and is now becoming more common for Non-NF2 patients. Specialist MDT assessment and counselling are crucial to inform decision-making and manage expectations. There are encouraging results reported particularly for non-NF2 patients and those without other medical problems or developmental delay, with almost 50% achieving some degree of speech understanding. There is a lack of long-term prospective outcome data but with experience and uniform reporting this will steadily increase. Reference to specialist centres is essential if management is uncertain in the auditory implant MDT. If there is a possibility that a cochlear implant may work then a trial of CI is a reasonable initial step.

Disclosures

No disclosures to be made.

Conflicts of interest

No conflicts of interest.

References:

Surgical management of temporal bone meningoencephalocele and CSF leaks

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Abstract

Cerebrospinal fluid leaks and brain herniation into the temporal bone are rare entities that can occur due to a spectrum of pathologies. The end result is a breach in the dural barrier, which exposes the confines of the cranial cavity to infection, leading onto serious complications including meningitis, intracranial sepsis and brain abscess. Sealing the CSF leak requires a surgical approach, the nature of which is dependent upon the location and size of the defect and any co-existent pathology. The morbidity of the approach and patient factors also need to be taken into consideration. Traditionally Otologists have approached these defects via a trans-mastoid route for small CSF leaks and dural defects, but this has not been universally successful in larger or multiple defects with sizable brain or meningeal herniation. The Middelfossa approach has been established as an alternative and reliable solutions for these complex defects.


Key words

Cerebrospinal fluid (CSF), CSF Otorhinorrhoea, Meningo-encephalocele, Tegmen, Dural defect, Intracranial hypertension, Skull-base, Trans-mastoid approach, Middle cranial fossa approach, Combined Trans-mastoid Middelfossa Approach.

Introduction

Temporal bone meningo-encephalocele is the herniation of membranous and bone through the cranial base defects, including the empty spaces within the tympanic or mastoid cavity. CSF fistulas are created between the subarachnoid spaces and the tympano-mastoid cavity.1 It is crucial to understand their intricate anatomy in order to plan the appropriate approach, anticipate the obstacles and successfully deal with them, while preserving all the surrounding vital structures. This review presents the nuances of dealing with these pathologies, highlighting the salient points which need to be kept in mind while planning surgical intervention.

Meningo-encephalocele of the temporal bone most commonly results from an idiopathic defect in the middle fossa floor but can also occur following traumatic or iatrogenic injury to mastoid tegmen or posterior fossa dura. They are less commonly associated with petrous pyramid disease. A congenital temporal bone encephalocele is very rare and the incidence is estimated at 1:10000 live births worldwide.2

Patients with meningo-encephalocele may remain asymptomatic or develop mild symptoms such as aural fullness, hypoacusis, pulsatile tinnitus, vertigo and rarely cranial neuropathy. In cases of chronic elevation of the intracranial pressure, morning headaches, sudden hearing loss, imbalance and blurred vision may manifest. The brain tissue over time usually becomes necrotic after herniation and this may cause a breach in the barrier leading onto CSF fistulas. Zaneti et al.3 have described the most common sites of meningoencephalocele and CSF leaks in the temporal bone being the tegmen tympani and tegmen antri. Posterior fossa defects are comparatively rare.

CSF leakage into the temporal bone most commonly occurs as a result of trauma and iatrogenic injury, although infectious and neoplastic aetiologies are also causes.4 It may arise spontaneously, in the absence of any precipitating factors. Spontaneous CSF leaks typically occur in two distinct populations: young children and middle-aged adults.5 Young children usually present with an episode of meningitis and this may be attributable to a specific congenital defect or secondary to enlargement of pre-existing bony pathways such as leaks through abnormal labyrinthine windows or a widened fallopian canal.6

The pathophysiology of adult-onset spontaneous CSF leak is less certain. Post-mortem temporal bone
studies have shown approximately 20% incidence of bony tegmen defects in the adult population. Ommaya suggested that small bony defects of the middle fossa tegmen resulting from abnormal embryologic development can enlarge over time with constant CSF pressure. It is thought that years of constant, pulsating intracranial pressure eventually causes dural thinning to the point of rupture through these areas of bony dehiscence. A second theory proposed by Gacek, implicates aberrant arachnoid granulations in the middle and posterior fossa dura. Persistent pressure at these arachnoid granulations results in thinning and subsequent erosion of the underlying bone leading onto Meningo-encephalocele associated with CSF leakage. Table-1 highlights the spectrum of causes which can lead to meningo-encephalocele and CSF leaks.

<table>
<thead>
<tr>
<th>Table 1: Aetiology of Lateral Skull base Defects &amp; CSF Leaks</th>
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<tbody>
<tr>
<td>Latrogenic:</td>
</tr>
<tr>
<td>- Accidental breach of tegmen during mastoid surgery</td>
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<tr>
<td>- Planned partial removal of middle fossa dural plate in</td>
</tr>
<tr>
<td>trans-petrous approach and sub-temporal + trans-mastoid</td>
</tr>
<tr>
<td>approach</td>
</tr>
<tr>
<td>Neoplastic:</td>
</tr>
<tr>
<td>- Temporal bone cancers</td>
</tr>
<tr>
<td>- Glomus tumours</td>
</tr>
<tr>
<td>- Meningioma</td>
</tr>
<tr>
<td>- Metastasis</td>
</tr>
<tr>
<td>Infective:</td>
</tr>
<tr>
<td>- Cholesteatoma / Chronic suppurative ear disease</td>
</tr>
<tr>
<td>- Otogenic Intracranial sepsis</td>
</tr>
<tr>
<td>- Skull base Osteomyelitis</td>
</tr>
<tr>
<td>Idiopathic:</td>
</tr>
<tr>
<td>- Spontaneous childhood or adult onset</td>
</tr>
<tr>
<td>- Congential dehiscence of skull-base suture plates</td>
</tr>
<tr>
<td>- Benign Intracranial Hypertension</td>
</tr>
<tr>
<td>- Mucoid Obdity</td>
</tr>
<tr>
<td>Trauma:</td>
</tr>
<tr>
<td>- Skull base fractures (acute or delayed onset)</td>
</tr>
<tr>
<td>- Whiplash injury with meningeal tear</td>
</tr>
<tr>
<td>Miscellaneous:</td>
</tr>
<tr>
<td>- Craniofacial syndromes</td>
</tr>
<tr>
<td>- Hydrocephalus</td>
</tr>
<tr>
<td>- Langerhans cell Histiocytosis</td>
</tr>
<tr>
<td>- Paget’s Disease</td>
</tr>
<tr>
<td>- Skull base Fibrous Dysplasia</td>
</tr>
<tr>
<td>- Degenerative metabolic disorders</td>
</tr>
<tr>
<td>- Post-radiation sequelae / Osteoradionecrosis</td>
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</table>

Spontaneous leaks have the highest rate of recurrence. Obesity with radiological evidence of empty sella in patients with spontaneous CSF otorhinorhrea requires exclusion of benign intracranial hypertension as a cause. Diagnostic lumbar puncture and neuro-ophthalmic examination of the fundi should be considered to rule out this diagnosis prior to surgery, although the diagnostic features may not become apparent until the leak is sealed. CSF leaks are often challenging to diagnose and manage. Patients typically present with complaints of aural fullness, hearing loss, pulsatile tinnitus, with a unilateral persistent or recurrent middle ear effusion along with post-nasal drip or unilateral watery rhinorhoea. Clinical presentations may be subtle and may be present for years. The diagnosis ultimately depends on a high degree of clinical suspicion and is sometimes made after myringotomy or tympanostomy tube placement results in persistent clear otorrhea. Investigations available to facilitate the diagnosis include ear fluid biochemistry, beta 2 transferrin assay / Tau-protein analysis and imaging with high-resolution computed tomography (HRCT) and magnetic resonance imaging (MRI).

Imaging is critical to diagnosis and the majority can be confidently identified by careful analysis of thin slice direct coronal T2 weighted MR images along with CT scanning. Imaging aids the differentiation between granulation tissue, cholesteatoma or brain hernia and aid in tracking the site of the CSF fistula precisely. In complex cases a CT cisternography with intrathecal contrast administration, digital subtraction myelography, MR cisternography or radionuclide scans are more specific tools to locate the defect, but these are rarely indicated.

Fig-1 & 2 represent comparative pre- and post- operative images of a patient who underwent successful repair of tegmen tympani defect for spontaneous CSF otorrhea. The most significant complication of CSF leakage is meningitis. Currently, there is no clear evidence to support prophylactic antibiotic use in the presence of a CSF leak due to skullbase fracture and for meningo-encephaloceles with or without CSF leaks. Definitive and expedited surgical repair is therefore critical to prevent this potentially devastating complication.

**Principles of Surgery**

Debate exists about the need for surgery in asymptomatic patients with a meningo-encephalocele but no CSF leak. If symptomatic, the herniated tissue can be either resected or retracted into the intracranial compartment. Herniated tissue is typically non-functional and hence resection is preferred to reduce the risk of intracranial contamination. In a trans-mastoid approach it is necessary to resect the redundant herniation in order to gain access to the skullbase defect for reconstruction. If a middle cranial fossa approach is selected, the herniated tissue can either be sectioned from above & left in middle ear or elevated from within the temporal bone. If left the isolated tissue progressively shrinks, becoming incorporated into scar tissue.

Defects should be repaired in a multi-layered fashion and better results and lower rates of recurrence are described when such a multi-layered approach is used. A thorough repair of both dural and bony defects is needed. Without a secure bony reconstruction, dural herniation and CSF leakage can recur. Various materials and techniques have been described to reconstruct the bony skullbase most using autologous calvarial bone or cartilage. The dural defect can be repaired using autologous pericranium or fascia as well as allogenic materials (Biodesign, Duragen). Tissue sealants and glues are useful in achieving a watertight closure. The use of tissue sealant does not appear to negatively affect the
ossicular chain and hearing outcome. 17 Alloplastic synthetic biomaterials including bone cements, bioglass ceramics, acrylic polymers, fluoroplastic, composite hydrogel with methyl methacrylate have been mainly reported as adjuncts with bone grafting for larger defects resulting from trauma, surgery, neoplasia and for recurrent defects.18,19

Per-operative CSF drainage with a lumbar drain is not routine but may be considered in larger defects of the skull base, in patients with raised intracranial pressure, BMI > 30 kg/m², high volume leaks and those with significant medical co-morbidities and anaesthetic risks.16,18

Post-operative care with neuro-ICU monitoring is recommended. Ample bed rest with head elevation and secure mastoid bandage, antibiotic cover, thromboembolic prophylaxis, avoidance of straining and sneezing, facial softeners and cough sedation are all help to prevent displacement of the graft.

Selection of Surgical Approach
Options for surgical repair include transmastoid, middle fossa craniotomy and combined approaches.14 Selection of approach is based on the location and size of defect, the ex-perience and preference of the surgeon.

The middle cranial fossa approach offers a direct route and improved exposure of the entire tegmen and petrous apex “from above,” it is however not appropriate for middle fossa defects. This approach may result in better hearing outcomes since the ossicular chain does not need to be manipulated to improve exposure.15 However, the middle fossa craniotomy approach is associated with greater morbidity and the risk of neurological complications.

The transmastoid approach avoids the risks associated with temporal lobe retraction and provides access to both middle and posterior fossa defects but potential disadvantages here include decreased exposure of the defect, lower initial success rate, and increased risk of hearing loss.

Many neuro-otologists recommend a combined transmastoid-middle fossa approach, especially in cases of large, multiple, or recurrent defects, or revision surgery for recurrent CSF leaks.20,22,23

Transmastoid Approach
This is a direct approach to the lateral cranial base and can address defects in both the middle and posterior fossae with minimal morbidity.16 Usually access to anterior middle fossa floor is limited by the ossicular chain in the attic manipulation of which often leads to a mild to moderate conductive deafness (Table-2).

Table 2: Trans-mastoid Approach to Tegmen Defects

<table>
<thead>
<tr>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
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<tbody>
<tr>
<td><strong>Technically easy to perform endoscopic procedure</strong></td>
<td>Higher incidence of graft failure and recurrence in high pressure leaks</td>
</tr>
<tr>
<td><strong>Safe procedure with fewer risks and complications</strong></td>
<td>Recurrence may need cavity obliteration with blind sac closure</td>
</tr>
<tr>
<td><strong>Ideal approach for posterior fossa defects</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Can simultaneously deal with chronic ear disease</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Not ideal for large meningo-encephaloceles or multiple CSF leaks</strong></td>
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</tbody>
</table>

A mastoidectomy with preservation of the outer ear canal wall is the preferred procedure. In case of chronic otitis media, eradication of the disease is carried out with appropriate reconstruction or blind sac closure.13

In rare instances, suturing the dura in an extradural fashion and cutting the redundant meningocele is required. The bone revision needed to be sealed by placement of a graft which can be done by an inline, overlay or sandwich technique and reinforced with tissue glue.20,23 Semaan et al.18, presented success with transmastoid extra-dural-intracranial approach for repair of transtemporal meningo-encephalocele in a review of 31 consecutive cases in which there was no significant hearing deterioration. Sanna et al.20 reported a much larger series on the treatment of meningo-encephalocele hernias with the transmastoid approach alone in 93/122 patients, including 55 with total obliteration of the middle ear cavities.

Middle Fossa Approach
The vast majority of spontaneous CSF leaks take place through the tegmen plate, rather than through a posterior fossa defect. Traumatic leaks are more unpredictable and many have both posterior fossa and middle fossa components. The major advantage of the middle fossa approach over the transmastoid is that the repair can be carried out without disturbing the middle ear ossicles (Table-3).15

The procedure is carried out through a ‘mini’ temporal craniotomy with a window of less than 4 cm².24 While drilling, bone dust is collected using a bone-paste collector that is essentially a filter-trap attached to the suction apparatus. After elevation of the dura, all the defects in the tegmen are exposed and sealed with an oblong slab of bone pate mixed with fibrin glue. In larger defects over 1 cm a solid bone chip is used in addition to bone pate. This pliable graft material is insinuated into every defect in the tegmen. A large free graft of temporalis fascia placed between the dura and the bone-pate slab provides additional reinforcement.

Combined Trans-Mastoid Middle Fossa Approach
The combined approach offers access to the entire middle and posterior fossa defect.20,23 O’Connell et al.,25 described a trans-mastoid-middle cranial fossa approach for repair of lateral skull base CSF fistula and encephalocele using a suture “pull-through” technique. This method facilitates reliable placement of a composite graft in the centre of lateral skull base defects through a small craniotomy that minimizes temporal lobe retraction.

Table 3: Middle Fossa Approach to Tegmen Defects

<table>
<thead>
<tr>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>Exposure and access to entire tegmen plate including anterior tegmen tympani</td>
<td>Higher incidence of graft failure and recurrence in high pressure leaks</td>
</tr>
<tr>
<td>Multiple defects / leaks can be sealed with larger graft securely to minimize chance of recurrence</td>
<td>Recurrence may need cavity obliteration with blind sac closure</td>
</tr>
<tr>
<td>Hearing preservation technique</td>
<td></td>
</tr>
</tbody>
</table>

Recurrence may need cavity obliteration with blind sac closure.

Conclusion
The successful management of meningo-encephalocele and CSF leaks requires skilled diagnostics with the majority being definitively diagnosed by high resolution conventional CT and MRI. The value of direct fine cut coronal T2-weighted views cannot be over-emphasised. Safe and reliable surgical outcomes can be achieved by using a multi-layered closure technique and recognition of associated pathology such as benign intracranial hypertension. This type of surgery needs to be performed in high volume tertiary referral centres with skull-base units with all the expertise and infrastructure is available.

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Ethical considerations
None

Conflict of interest
None

References
A review of middle vault reconstruction techniques

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Abstract
Management of the middle vault is an important aspect in rhinoplasty, as pathology in this area can lead to nasal valve obstruction and functional problems. This paper concentrates on techniques for management of the internal nasal valve, and aims to summarise the choices of treatments available.


Key words
Middle vault reconstruction, nasal valve collapse

Anatomy
The middle vault of the nose consists of the paired upper lateral cartilages (ULC) with its attached mucosa. The ULC are inserted under the caudal ends of the nasal bones, fusing with the bones. Medially they continued into the cartilaginous septum, their caudal end separating from the septum. The caudal end of ULC have attachments to the lower lateral cartilages (LLC), in a recircature or scroll fashion. Mucosa is tightly attached to the internal surface of the ULC and continuous with the lining of the septum and lateral nasal wall.

The term ‘nasal valve’ was first coined by Mink in 1903 to describe the slit-like opening of the junction between the upper and lower lateral cartilages or external valve. It is the most flexible and narrowest portion of the nasal airway. In a review of 500 patients, Elwany and Thabet found that nasal valve dysfunction accounts for 13% of patients complaining of nasal obstruction.

According to Poiseuille’s law, airflow is proportional to the radius of the nasal passages, raised to the fourth power. Thus a small change in the angle of the valve will have a large effect on airflow and resistance of the nasal cavity. When air flows through a narrow space, it speeds up and creates an inward pressure, causing in-drawing of any weakened area and nasal valve collapse.

Aetiology of nasal valve dysfunction
Causes of narrowing and collapsing of the nasal valve include congenital flaccidity, trauma, previous surgery and ageing. In primary rhinoplasty, there are certain characteristics to indicate that middle vault reconstruction is essential. This includes a narrow middle vault, dorsal hump reduction planned, middle vault deviation and asymmetry.

Constriction of the internal valve is a common reason for nasal obstruction in patients who have undergone rhinoplasty, with the inverted-V deformity a classic sign of middle vault pathology. Predisposing factors include full thickness mucocartilaginous transection of each ULC from the septum and over-rotation of the dorsal
septum or ULC. One of the most common manoeuvres in rhinoplasty is hump reduction. In traditional hump reduction surgery, the cartilaginous hump is removed en-bloc with the underlying mucosa. This separates the ULC from its natural attachment of the septum, interrupting the continuity of the mucoperichondrium, leading to posterior displacement of ULC. The effect of this may not be apparent immediately, but over the years could gradually present as inverted V deformity. Chronic nasal obstruction due to valve narrowing may also develop with time, where subsequent mucosal scarring with synechia in the valve region intrinsically decreases the nasal airway area. Grymer reported that there is a decrease of the internal valve cross-sectional area by 25% and the piriform region intrinsically decreases the nasal airway area.

Ageing also brings internal valve collapse as there is weakening of the muscular and fibrous support. Patients with internal valve collapse typically have a “pinching” appearance or medial collapse of the supra alar region.

Techniques of middle vault reconstruction
Reconstruction of the middle vault can be largely divided into 2 main groups: those where cartilage grafts were used, or where sutures were applied. Each group consists of different techniques described by various authors.

Methods using Cartilage Graft

1. Spreader Graft

Spreader grafts are probably the most common corrective technique used in middle vault rehabilitation. Popularised by Sheen, grafts of 3-4cm length and 3-4mm width are placed between the nasal septum and the ULC in order to lateralisate the ULC and widen the nasal valve. Most studies used an external rhinoplasty approach, as it allows direct visualisation of the middle third of the nose and easier to stabilise with direct suturing. The grafts can be harvested from the posterior septum or concha area. The ULC is separated from the septum in order for graft placement (see Fig 1) - resulting in a 3-layer construct held together by mattress suture (Fig 2).

André and Islam both described a variation using an endonasal approach and without detachment of ULC. André placed the graft as high as possible in a subperichondrial plane. Islam cut out a window on the dorsal septum so as to house the spreader.

The positive effect of spreader graft on nasal patency is well documented, with improvement rates from 81% to 100% being reported. Varadarajan and Cheang were able to demonstrate in their pilot study that using endonasal spreader grafts in patients with internal valve narrowing have their NOSE score improved from an average of 14.5 preoperatively to 5.2 postoperatively (unpublished data).

One drawback of spreader grafts is the effect on the external appearance of the middle vault. Using Adobe Photoshop pre- and post-operatively, Ingels showed that spreader grafts can widen the middle vault by 6%. Reassuringly in their study of 15 patients, none of them noticed this widening. Table 1 shows the summary of studies using spreader grafts, with their outcomes.

A modification of spreader grafts are spreader flaps - first described by Oneal and Herkowitz. This involves detaching the ULC from the septum and preserving them in some fashion. Following dorsal hump reduction, the redundant ULC are scored and folded medially alongside the nasal dorsum. This method mitigates the need for extra cartilage graft.

2. Butterfly graft

The butterfly graft over the nasal dorsum has been used in both primary and secondary rhinoplasty for internal and external valve collapse. It is designed to restore the physiological recoil of the nasal valve. Cartilage of approximately 1x2cm in size is usually harvested from the auricular concha. The graft is inserted into a sub-SMAS plane either via endonasal or open approach. The caudal end of the graft is sutured to the caudal end of ULC.

This method has shown to improve breathing in 80% to 100% of patients. In addition Ackam et al showed 65% patients had improved or stopped snoring. However 12-19% were dissatisfied with the cosmetic outcome of their nose, where there is added volume over the anterior septal angle. There is also a risk of graft fracture. It is important to provide adequate preoperative explanation and manage patients expectation when using this graft, as maintaining an aesthetic dorsum with a butterfly graft does possess a technical learning curve. Friedman advised to bevel the edges of the graft, and to place crushed cartilage over the graft and dorsum to maintain its smoothness. Eight to nineteen percent of patients are also not happy about the outcome of the ear.

Table 1. Summary of studies using spreader grafts. Grouped as techniques used.

<table>
<thead>
<tr>
<th>Study</th>
<th>Technique used</th>
<th>Approach</th>
<th>Outcome measures</th>
<th>Success rate</th>
<th>Complications rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Zijlstra1994</td>
<td>Spreader graft</td>
<td>Open</td>
<td>Nasal patency: Subjective assessed 10 point score</td>
<td>Nasal patency: 81% improved</td>
<td>Mean improvement 4.1 points</td>
</tr>
<tr>
<td>Stait2000</td>
<td>Absorbable spreader (Lactosorb)</td>
<td>Not specify</td>
<td>Nasal patency &amp; aesthetic: subjective report, not specified</td>
<td>100% improved. No recurrence even though graft absorbed.</td>
<td>None</td>
</tr>
<tr>
<td>André2004</td>
<td>Spreader graft (tucked in subperichondrial plane)</td>
<td>Endonasal</td>
<td>Nasal patency: Subjective assessed 4 pt scale</td>
<td>44% optimal, 44% improved</td>
<td>5.6% infection</td>
</tr>
<tr>
<td>Ingels2008</td>
<td>Spreader</td>
<td>Open</td>
<td>Nasal patency &amp; Aesthetic: Subjective assessed VAS, AR</td>
<td>Nasal patency: mean VAS increased by 3.4 points. Mean dorsal width increased by 6%. Not noticed by patients subjectively</td>
<td>None</td>
</tr>
<tr>
<td>Varadhajan1997</td>
<td>Spreader graft</td>
<td>Endonasal</td>
<td>NOSE score</td>
<td>NOSE pre op: 14.5</td>
<td>Not documented</td>
</tr>
<tr>
<td>Scutic1999</td>
<td>Spreader graft and ULC suspension suture</td>
<td>Open</td>
<td>Nasal patency: Subjective assessed 10 point score</td>
<td>Nasal patency: 100% improved</td>
<td>Mean improvement 4.4 points</td>
</tr>
<tr>
<td>Hoosh2004</td>
<td>Spreader, Batten or both</td>
<td>Either</td>
<td>Nasal patency &amp; aesthetic: subjective report, not specified</td>
<td>Nasal patency: spreader 88% improved. Batten 100%. Both 93%</td>
<td>Not documented</td>
</tr>
<tr>
<td>Fair2005</td>
<td>Combined Spreader and Batten</td>
<td>Open</td>
<td>Nasal patency: Subjective assessed VAS, OQL: Subjective assessed VAS, Aesthetic: Subjective assessed 3 point scale</td>
<td>Patency: mean improvement 55mm. OQL: mean improvement 49mm.</td>
<td>8.7% - 1 graft reabsorption 1 graft migration</td>
</tr>
</tbody>
</table>

VAS = visual analogue scale. AR = acoustic rhinomanometry. NOSE= Nasal obstructive symptoms evaluation scale. MCA = minimum cross-sectional area.

Figure 1: ULC detached with insertion of left spreader graft shown.

Figure 2: Bilateral spreader graft inserted fixed with suture.
3. Modified onlay spreader grafts
A variation of the Skoog method, Gassner et al. described a way of reconstructing the middle vault which simultaneously restores the nasal valve angle and the structural resilience of the nasal valve. After division of the ULC from the septum and reduction of septal dorsum, a cartilage graft of 8-9mm wide and 1.5cm long is fix to the midline, with the ULC sutured to the underside of the onlay graft. This allows lateral rotation of the entire ULC, before looping back lateral to the ULC and emerging endonasally to help deliver the caudal end of ULC.

Methods using Suturing
Most suturing methods aim to enlarge the narrowed valve area.

1. Nasal valve suspension sutures
Panisseau pioneered the nasal valve suspension suture in 1996, whereby the ULCs were suspended by placement of a permanent suture at the site of collapse and retracted laterally anchoring to the maxillary peristomeum. This technique serves to lateralise the alar side wall, expanding the nasal valve. There have been modifications of fixation methods to the infraorbital rim, either using a Mitek self-retaining screw or a permanent stay suture infraorbitaly as an anchor. André did not use a stay suture, anchoring the suspension suture directly onto peristomeum of inferior orbital rim.

The study of Wolford using nasal valve suspension sutures is shown in Table 2. Subjective symptoms were improved in 71% to 100% of patients. However long term data by Nusrat et al. reveals that the efficacy of this technique decreased over time, with satisfaction reduced to less than half of the patients followed up for more than a year. André similarly reported modest improvement that lasted for a short period of time, and a relatively high complication rate. Twenty-five percent of their patients experienced inflammation, swelling or pain under the eye, leading them to conclude that the nasal valve suspension suture is not recommended as a first line treatment for valve insufficiency. Roese et al. suggested that for the technique to be effective, the nasal and midfacial soft tissue need to be extensively undermined, mobilised and fixated in the new lateral position without tension.

2. Flaring suture
The flaring suture is placed through the caudal aspect of the ULC in a vertical fashion, and then passed on the contralateral side in a similar fashion. The caudal end of the ULC is typically hidden under the scroll, and the lateral crura will have to be retracted inferiorly for sufficient exposure or by the placement of a cotton bud endonasally to help deliver the caudal end of ULC. Tightening of the mattress suture across the dorsum will result in the ULC “flaring” laterally, increasing the angle of the internal nasal valve. All patients in Park’s study reported breathing improvement.

As noted above, relying on tension of sutures to permanently displace tissue may be ineffective over time. In this aspect, flaring suture may be more useful after the ULC has been detached and mobilised from the septum. Schlosser and Park used spreader grafts, alar batten grafts and flaring sutures in different combinations and did not present their result separately. However their cadaveric study in the same paper demonstrated that the combination of spreader grafts and flaring suture has the greatest impact on cadaveric nasal airway when measured with acoustic rhinomanometry.

3. Lateral Mattress Bending suture
Using 5-0 polypropylene, Ozurkan devised a horizontal mattress suture on each ULC without entering the mucoperichondrium. The tightened mattress suture causes the ULC to convert into a convex shape, thus increasing the internal valve angle. Using pre and post op endoscopic photos to measure the internal valve angle, they showed there was statistically significant increase from 9.1°± 4.2° to 25.3° ± 3.8°.

4. Concealed Lateralisering Suture Technique
A variation of the nasal valve suspension suture, Bizzi and Gauthier improved this technique to make a stab incision over the nasal bone laterally, anterior and inferior to the medial canthus. An endonasal intercartilaginous incision is also made. A 3/0 prolene suture is passed from the external incision under the SMAS layer, piercing the ULC, before looping back lateral to the ULC and emerging externally again inferior to the medial canthus.

They reported 10 year results in their later paper, 89.9% reported complete satisfaction with the improvement in their nasal airway. 10.1% failure rate as the suture cuts through the ULC, unraveled, or infection was encountered.

Methods using Implants
Turegan et al. fashioned a thin sheet of porous polyethylene into a saddle shape akin a conchal butterfly graft to correct dorsal deformity and internal valve collapse. Both groups demonstrated complete improvement in nasal patency as reported by patients, however 21% experienced extrusion and 17% infection in the former study. Wengen et al. fashioned this with a titanium graft, sutured fixated on top of the ULC. Preliminary data suggest that this implant may be tolerated.

Discussion
The main aims of surgery to the middle vault and nasal valves are threefold:

1) Increasing the internal valve angle if it was diminished; 
2) Stabilizing the free edge of the ULC to avoid its collapse; 
3) Strengthening the ULC and alar cartilages and increase resistance to negative pressure during inhalation.

It is important to decide whether the valvular insufficiency arose from a static or dynamic cause. This will serve to decide whether a widening or strengthening solution is needed. After saying that, many techniques overlap in form and function, and it is not possible to analyse a single technique in isolation.

The variety of techniques mentioned here serve to improve functional and aesthetic results. The aim of these techniques was to broaden the internal valve and re-establish the stiffness of the lateral nasal wall, but no technique is perfect, and each have their own drawbacks. Spreader grafts widen the middle nasal vault and aid in repositioning the ULC’s, but fail to support the lateral nasal walls. Butterfly grafts frequently require an open approach and...
adds bulk over the anterior septal angle. It also needs an extra procedure in the form of conchal cartilage harvesting. Upper lateral suspension procedure is technically demanding and requires specific equipment and infra orbital incision. Some suture techniques (flare suture, lateral expansion suture) claimed to be able to both widen and support the nasal valve. However as Gassner pointed out, tissues can only be repositioned properly if they have been adequately mobilized and fixated with the least amount of tension possible. Otherwise the repositioned pull would ultimately lead to tissue migration and cheese wire effect of the sutures10 – an important aspect of suture based techniques. It is also important to remember that techniques in thinplasty is only as good as its executioner.

A longstanding trend of the middle vault reconstruction has been the use of spreader grafts, but the functional value is increasingly being challenged38. Where there is also collapse of the middle vault, alternative or additional manoeuvres are recommended. The spreader flaps and onlay spreader graft are emerging alternatives.

Preoperative analysis and planning is important to prevent middle vault problems. Paying attention to details such as preservation of the middle vault mucosa, conservative osteotomies and maintaining soft tissue attachments of the nasal bones and ULC will minimize postoperative alterations, preventing the arising of future problems.

The last consideration is the approach used, and the preoperative counselling of patients. Many techniques had favoured an open approach, as it provides good access to the nasal valve38. Some of these overlap in form and function, but they all share the common goal of maintaining nasal aesthetics as well as preventing nasal obstruction. A combination of techniques may be necessary in some cases, and a knowledge of the techniques discussed in this review will hopefully provide the facial plastic surgeon with a variation of options in different circumstances.

References
The evolution of osteotomy in rhinoplasty

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Abstract
Osteotomies in rhinoplasty can be tailored to meet the aesthetic and functional needs of individual patients. We seek to review the evolution of osteotomies from the start to current practice in order to highlight the roles of various techniques and how they may be best utilised to meet patients’ requirements.

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Key words
Rhinoplasty; Septorhinoplasty; Nasal; Osteotomy

Background
In order to understand current osteotomy techniques, it is first important to review the historical development of rhinoplasty osteotomies. Throughout history, the aim has been to achieve desired aesthetic results whilst preserving nasal function.

The main indications to undertake nasal osteotomies during rhinoplasty include: reducing an open roof⁴, correcting deviated and asymmetric nasal bones⁵ and narrowing a bony base or dorsum⁶ including correction of bone convexity.

The earliest rhinoplasty osteotomy publications include Robert Weir’s rhinoplasty cases in 1892. Weir described the earliest rhinoplasty osteotomy publications include Robert Weir’s rhinoplasty cases in 1892. Weir described correcting deviated and asymmetric nasal bones and narrowing a bony base or dorsum including correction of bone convexity.

The dorsal nasal hump is frequently encountered during rhinoplasty surgery. It is composed of bony and cartilaginous components forming the bulk of the hump in majority of cases. Once adequately exposed it may be addressed using an osteotome.

One such modification was Maurice Cottle’s unique push down operation to preserve the nasal airway⁹. This involved lowering the nasal hump with lateral osteotomies, reducing the nasal septum and repositioning of the nasal dorsum. For large hump reductions, Cottle used bilateral intermediate osteotomies to remove the lateral nasal wall.

Over time the configuration of lateral osteotomies was modified. The starting point was approximately at the level of the inferior turbinate and was extended to a higher position on the nasal sidewall relative to the face of the maxilla creating a high-low lateral osteotomy in order to preserve Webster’s triangle and the nasal airway⁹. This was modified further by Farrar who recommended a high-low-high curved lateral osteotomy in order to preserve the lateral suspensory ligament at the level of the piriform aperture, therefore preventing iatrogenic nasal obstruction. The application of osteotomy techniques evolved further as surgeons tailored techniques according to patients’ anatomy whilst minimising complications.

Moving from the 1970s to 1990s saw a development of osteotomies tailored to patients’ anatomy rather than based on experience and training⁹. The methods were designed to reduce oedema, bleeding and nasal wall instability. More recent developments in the last 20 years include the movement away from the traditional en bloc hump reduction to component hump reduction with upper lateral cartilage preservation. Most recently the introduction of powered instruments and the variation in technique to accommodate their use has been popularised.

Osteotomy Techniques
The osteotomy forms part of the rhinoplasty surgeon’s armamentarium when addressing both functional problems and aesthetic nasal deformities. Judicious use of osteotomies should be exercised, as cases with minimal correction may not require unnecessary destabilisation of the nasal bones with excessive osteotomies and mobilisation.

Sources of discrepancy in practice include: 1) the type of osteotomies, 2) the size of osteotomies, and 3) the approach for each osteotomy.

Type of osteotomies:
Broadly speaking, the osteotomies can be divided into “non-powered osteotomies” vs “powered osteotomies”. Non-powered osteotomies are still the main method of performing osteotomy amongst most surgeons however there has been a move towards use of powered instrument for osteotomy in recent years. Electric and ultrasonic equipment have given the surgeons much better control in performing osteotomy and some of the risks associated with the non-powered osteotomies (such as unpredictable fracture lines) have been greatly reduced. Piezoelectric surgery has also allowed the bones to be remodelled after they have been mobilized. Piezorsurgery was introduced to rhinoplasty by Robiony et al following prior use in maxillofacial surgery¹⁰,¹¹.

The non-powered osteotomies are broadly divided into “guarded” vs “unguarded”, and straight vs curved osteotomies. The guarded osteotomy can be easily palpated through the skin. The curved osteotome allows a controlled curved path through the bone which is ideal in high-low-high lateral osteotomy and divergent medial osteotomy.

Approach to osteotomy:
Further variations exist with approaches to osteotomy: endonasal versus external (percutaneous).

The traditional teaching for non-powered osteotomies insists on preservation of mucosa and skin soft tissue attachment to the nasal bones to avoid collapsing of the nasal bones into the nasal cavity. This is often achieved by making subperiosteal tunnels and limiting the extent of soft tissue skin elevation. In powered osteotomies, the tendency is to have much wider skin elevation for ease of access. Comparison of traditional osteotomies with the piezoelectric surgery demonstrated a reduced incidence of postoperative pain, oedema and ecchymosis as the adjacent nasal mucosa was preserved more effectively with this technique¹²,¹³.

Osteotomies on the bony pyramid can be achieved by different types of osteotomy: 1) medial, 2) lateral, 3) transverse, and 4) intermediate (Fig 1). Generally the osteotomies are made from medial to lateral (medial first, then intermediate and then lateral osteotomy if all required).

![Figure 1: Types of Nasal Osteotomies](image)

Size of osteotomies:
Surgeons favouring small osteotomies would do so due to the reduced rate of oedema and ecchymosis, as well as avoiding disrupting the surrounding perioseum and mucosa. However, this method carries a greater risk of breaching the cribiform plate especially when undertaking medial osteotomies. This is due to the pressure being exerted over a smaller cross sectional area. For this reason, some surgeons prefer larger osteotomies to reduce such risks. The larger osteotomies are not the desired tool by some as there is more chance of tear of the underlying nasal mucosa producing further trauma and instability.

Some surgeons believe that smaller (ie narrower) osteotomies will be able to deliver more concentrated pressure to the bone resulting in a neater cut to the bone and reducing the chance of bone shattering and undesirable result or irregular contours. This is true when perecutaneous lateral osteotomy is employed. It is important to have a sharp osteotome to minimize the risk of a comminuted fracture.

The size of the osteotomies remains a topic of controversy and of surgeon’s choice as each school of thought claims their method of choice to be the safer and the superior one. There is no doubt that the delivery of the technique (whether with a small or large osteotome) remains the main determinant. Surgeon’s skills and control in placing and guiding the osteotome and the amount of pressure exerted by the hammer is crucial to achieve the desired result and reduce the risk of complications.

Dorsal Hump Reduction
The dorsal nasal hump is frequently encountered during rhinoplasty surgery. It is composed of bony and cartilaginous components; the cartilaginous component forming the bulk of the hump in majority of cases. Once adequately exposed it may be addressed using an osteotome.
Hump reduction was initially performed en bloc as described by Joseph. In this method, the bony cartilaginous hump was removed in one piece. En bloc hump reduction was later replaced by several modifications leading to component reduction where cartilaginous and bony parts are addressed separately. This technique was recommended by Roborich et al in order to protect dorsal aesthetic lines, preserve the upper lateral cartilages and internal valves and prevent inverted-V deformities. The use of spreader grafts in conjunction with this technique and subsequently spreader flaps have been recommended to handle functional and aesthetic problems involving the mid third of the nasal dorsum.

The nasal dorsal hump is best to be envisaged as a cartilaginous hump with a small bony cap. The method of hump reduction has evolved further to ovoid the open roof by preserving the underlapping upper lateral cartilage. This is undertaken by lowering the septal height and removing the bony dorsal cap without excision of the upper lateral cartilages. This technique facilitates the creation of upper lateral spreader flaps along their entire length, which will be used to reconstruct the dorsum and maintain the internal valve open.

Medial Osteotomy
Medial osteotomies indicate indications include nasal sidewall mobilisation, prevention of irregular back-fracture following lateral osteotomy and to widen a narrow upper third. They are often employed to correct the deviated nose or narrow the bony nose without a hump. They are carried out in an angulated fashion between the septum and nasal bones. Truly medial osteotomies will require a transverse osteotomy, especially when combined with a low-lower lateral osteotomy. Based on its angle, the medial osteotomy has been characterised as medial, paramedial and medial oblique and lateralised medial oblique (Fig 2). The vertical medial osteotomy can be used in situations that the surgeon wishes to widen the narrow bony vault. The medial oblique osteotomy will mobilise the sidewall and prevent an irregular back-fracture from the lateral osteotomy. The medial oblique was reported by Gruber et al to be effective when reducing the nasal dorsal width especially when placed laterally with an open roof. This eludes to the role of a lateralised medial oblique, which is approaching the indication for a truly intermediate osteotomy.

Transverse Osteotomy
This is usually indicated to effectively connect a medial or paramedial osteotomies with a low lateral osteotomy

The widely accepted path of the continuous osteotomy follows a high-low-high path. The path starts above the level of the attachment of the inferior turbinate taking care to preserve the lateral suprastructure ligament, which will help preserve the nasal airway. The osteotomy is continued along the nasal floor curve and anteriorly superior to the level of the mental canthus. Extending beyond the point of nasal bone divergence could create protrusion at the superior fracture site upon medialisation of the caudal part otherwise known as a rocker deformity. The percutaneous approach utilises a narrow osteotomy (eg 2 or 3 mm osteotome) to create a postage stamp perforations on the nasal bony side wall. This can facilitate more precise placement even in less experienced hands with minimal evidence of external scars. It also has been shown to provide greater preservation of periosteal support. An alternative approach is to undertake this intermediate osteotomy, which may be used to push out medially displaced nasal bones. The perforating osteotomy can be effectively used in revision cases or following trauma where maintaining structural support is vital.

Special consideration should be given to the extremely deviated nose, as the osteotomy starts with mobilising the bony side wall opposite the deviation followed by medial osteotomy and finally lateral osteotomy on the deviated side. This sequential series of osteotomies is analogous to turning the pages of a book with the nasal sidewalls and septum simulating the pages, which creates space for deviation realignment.

Powered instrument osteotomy
The powered instruments have made their way into many surgical specialties including rhinoplasty surgery. Further developments include the introduction of the piezoelectric surgical devices which use piezoelectric ultrasonic vibrations to undertake osteotomies. It has been shown to significantly lower pain, oedema and ecchymosis hence the increasing popularity amongst certain groups of surgeons.

The benefits of powered instruments are that they reduce the magnitude of postoperative complications in rhinoplasty and therefore improve recovery time. The cost of using such instruments should be considered, as these techniques increase operative time, so may be more difficult to justify unless significant benefit can be demonstrated with patient reported outcome measures in future.

Conclusion
The various osteotomies described here are utilised by different surgeons in different ways based on different patients and the variety of clinical situations in which they may present.

The variety of techniques and philosophies illustrate that there can be several different ways to achieve the desired functional and aesthetic goals. However all actions in rhinoplasty have another potentially undesirable reaction, which should be considered when selecting the most appropriate osteotomy. The underlying anatomy and the preoperative goals of surgery consulted with the patient regarding functional and aesthetic concerns should be considered prior to the final decision. The advent of powered-instruments has opened up new ways to perform osteotomy. The main determinant to achieve the desirable results in osteotomy rests with proper planning, understanding the anatomy and appropriate execution of the technique when performing osteotomy in rhinoplasty.

References
Navigation systems in endonasal endoscopic surgery – A review

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Abstract
The use of navigation systems in endonasal endoscopic surgery is relatively new, with the aim of improving operative outcomes. Image-guided navigation has been used increasingly as an adjunct during endoscopic endonasal surgery, with various navigation systems introduced worldwide. Therefore, this is an opportune moment to appraise this technology. This review summarizes the historical advances, key points and pitfalls of navigation systems, as well as providing potential future avenues, based on current literature and review of our own practice.


Key words
Navigation, Endoscopic Endonasal, Electromagnetic, Optical

Introduction
Over the past two decades, there has been a significant transformation in surgical management of sinus related disease and anterior skull base surgery.1-4 Having been initially introduced as a minimally invasive procedure to treat chronic rhinosinusitis, in patients whom medical management failed, endoscopic sinus surgery techniques have now expanded to treat orbital and skull base lesions, as well as both benign and complex malignant conditions.5,6,7,8 These disadvantages can be somewhat overcome through professional development of anatomical knowledge and cadaveric training, but not in entirety. As a result, intra-operative navigation systems have been developed to increase safety and efficacy of these procedures.9,10

Computer-assisted technology has allowed real-time anatomical navigation through the combination of tri-planar radiological images and intraoperative endoscopic views. Over the last 20 years, image-guided navigation has been used increasingly as an adjunct during endoscopic endonasal surgery. Various navigation systems have been introduced worldwide. Therefore, this is an opportune moment to appraise the existing navigation systems and offer an insight into the potential for future innovation.11-20

Historical Advances
Endoscopic endonasal procedures were first described in the late 19th century, but it was the improvement of optics, during the 1940s and 1950s, by Karl Storz and Harold Hopkins which ushered the new age of the Otorhinolaryngologist.21,22,23 Subsequent advances related to Messerklinger’s work in 1967, regarding sinus function

References
The use of a navigation system or technique was first introduced into the field of Otolaryngology in the late 1980s and early 1990s. Although fluoroscopy was formerly used as the primary image-guidance technique, nowadays computed tomography (CT) or magnetic resonance imaging (MRI) are used because of higher anatomical resolution. These allow isotropic, multiplanar, high-resolution, thin-section images of the head, with optical enhancement. Although MRI provides improved soft-tissue resolution, it has less information about bony structures, which are vital landmarks during endoscopic surgery. As a result, CT is the preferred current radiological imaging technique used amongst ENT surgeons, however neurosurgeons still continue to use MRI.29

Both navigation systems use a navigation screen, navigation module and tracking pointer.8,24 The navigation screen, as shown in Figure 1, helps in the registration, invasively (bone-anchored screws) or a combination of the two methods (hybrid). Photographic registration may also be used, which is potentially more accurate, as it can obtain more registration points (Fiagon Surgical Navigation (Fiagon, Berlin, Germany)).24 The non-invasive LED-mask-registration (Stryker® Instruments, Kalamazoo, Michigan, USA) is useful for routine endoscopic procedures, and has the advantage of automatic-surface registration. As the mask is applied to the midface during surgery, it typically cannot be used in open surgery of the midface or frontal region. This may be circumvented by transferring the mask registration to an additional patient tracker attached to the Mayfield frame (Stryker® Navigation System II-Cart, Stryker® Instruments, Kalamazoo, Michigan, USA). The hybrid registration technique can improve navigation accuracy significantly whilst invasive registration is reserved for selected skull base procedures which warrant a high level of precision.33

In the latter, bone-anchored screws are attached to the patient’s skull, which is an ideal substrate for registration, compared to the deformable skin or soft tissue.

Step 2 – Verification of accuracy. This is conducted using known landmarks, both on the patient’s face and intranasally. These landmarks are confirmed with pre-operative imaging.

Positives of Navigation System

Use of image-guidance systems have improved the scope of safe endoscopy. The endonasal surgery, with improvements in complication rates, especially major intracranial or intraorbital complications, as well as length of hospital stay.29,30 These seem to be related to improved identification of anatomical landmarks, especially in difficult, diseased or surgically revised anatomy.20,31

One of the critical issues is the navigation accuracy [more formally known as target registration error (TRE)]. For navigation to be useful, the TRE should be 2mm or better. The studies we reviewed showed navigation-guided surgery resulted in less invasive surgery, as well as subjective improvements in ‘comfort of surgeon’ and reduction of stress intra-operatively.3,14,17,29,38,39,40,41,42 In addition to this, there has been evidence of improved surgical performance and reduction in operative time, due to improved three-dimensional orientation.13,14

With respect to training, navigation systems improve the learning curve of juniors both intra-operatively and on transferring to stand-alone practice, and this is generally major difference in performance between different brands of navigation system, with all showing high system precision.31

Negatives of Navigation Systems

Although there are significant advantages with the advent of navigation systems, there are still some drawbacks. The learning curve of juniors both intra-operatively and on transferring to stand-alone practice. These shortcomings could further evolve into.

The first is augmented reality (AR) based, for example Scopis hybrid navigation (Scopis GmbH, Berlin, Germany).25 AR navigation system combines generated images of preoperative imaging data with real-time views of the endoscopic surgical field and has been particularly useful during frontal recess surgery. AR can also highlight structures to be avoided, such as the optic nerve and internal carotid artery, thus minimizing surgical morbidity. Although AR-based systems have their merits, inattentional blindness can be an issue. In a cadaveric study comparing AR augmented endoscopy and conventional endoscopy, users of AR failed to recognize a foreign body and skull base violation, which were clearly within view.28

Future navigation systems could also incorporate intraoperative updates, using portable CT scanners within the operative room. There can be misconceptions on the completeness of surgery as current navigation systems are based on pre-operative imaging and does not update with the extent of surgery. The need for an intra-operative update is all the more pressing in endoscopic skull base surgery.24

A combination of these factors necessitates training and experience prior to use, both by the surgeon and other operating staff.15 Electromagnetic navigation systems must be avoided in patients with electronic devices in the brain or nervous system.26 Previously, this was also true in patients with cardiac pacemakers, but recent advances have removed this as a contraindication. This cohort of patients, can still utilise optical tracking systems. Although optical systems are often described as more reliable than EM systems, the main setback involves requiring a direct line of sight between the tracking pointer and the tracker. If this line of sight is disturbed at any point intra-operatively the navigation screen is disturbed.19

Any potential crash to the computer system or the device can cause failure of the navigation system, as well as intermittently causing spurious results of location, almost at random. As a result, no technology should replace surgical vigilance and judgement.28 If in locations are not recognized by the surgeon, the results could be catastrophic. It must be acknowledged that navigation systems are an asset to surgical skills, not a substitute. Finally, there must be careful consideration of the initial and costs of purchasing equipment, as well as personnel costs.

Figure 1: Example of navigation technology – Stealth Station S7. Reproduced with permission of Medtronic, Inc.
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Scar revision: An overview and update

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Abstract
Scars can cause great distress to the patient and can have a significant psychological and social impact. Ideally, the need for scar revision should be avoided in the first place but may be indicated for cosmetic and/or functional reasons. Despite significant progress in the management of scars over recent years, scar revision remains challenging. There is a wide range of surgical and non-surgical therapies for scar revision but no universal consensus on best practice. The aim of this article is to provide an overview and an update on treatment options for scar revision.


Introduction
Scar formation is inevitable when the skin is breached beyond the deep reticular dermis. Skin appendages essential to wound healing (hair follicles and epithelial stem cells, sebaceous units and sweat glands) are disrupted and the resulting scar may not only affect cosmesis but also function. The ideal scar should be flat, narrow, should not compromise function and have a significant psychological and social impact.

Scars are not only caused by surgery but may also be the result of trauma, burns, acne or inflammatory processes. They can cause great distress to the patient with considerable psychological and social impact and reduced quality of life. Ideally, the need for scar revision should be avoided in the first place and the surgeon must take into consideration various factors to reduce the risk of an unfavourable scar (Table 1). The surgeon has most control over the intraoperative considerations which includes careful positioning of the incision, meticulous soft tissue handling, haemostasis, appropriate choice of suture materials, closure of any dead space, eversion of wound edges and avoiding tension on closure of the wound. The positioning of the incision is crucial and hiding incisions where possible is a very useful technique, such as hiding incisions within an orifice (e.g. transconjunctival, subhimal, intranasal), in hair, behind an anatomic prominence (e.g. postauricular, submental), at the junction of facial subunits, in a skin crease or in relaxed skin tension lines (RSTLs). When hiding a scar in hair, the edges of the incision should be bevelled so as not to jeopardize the hair follicles and increase the possibility of future balding. In planning the incision, it is also important to take into consideration the relationship of nearby structures in order to avoid distortion of these and subsequent morbidity, such as ectropion. Although many factors are also not directly within the surgeon’s control, it may be possible to influence some of the preoperative and postoperative factors through patient education.

Complications such as haematoma, infection, necrosis and dehiscence may interrelate with each other and the risks of these must be minimised to avoid increased scarring. Wound care is an essential aspect in reducing these complications. The use of Steristrips adds strength to the wound and reduces tension, whilst collagen is being deposited in the first few days of wound healing. Occlusive dressings promote re-epithelialization and better healing, although may be difficult to apply in the head and neck region. An alternative to occlusive dressings is the “open” technique which involves covering the wound with a generous amount of antibiotic ointment, thereby providing a moist environment which also acts as a physical barrier to dirt and reduces the risk of stitch abscesses. Appropriate wound cleansing to prevent the formation of crusts, which can predispose to infection, is also beneficial.

There is a wide spectrum of scar deformities which include the following subtypes:
- Elevated scar
- Atrophic scar with thinning of dermis and epidermis
- Rolling scar
- Boxcar scar
- Contracted webbed scar (thickened scar tissue replaces the epidermis)

Scars may also be described in further detail based on the following characteristics:
- Orientation
- Colour
  - Red-violaceous
  - White
  - Brown
- Contour
  - Elevated
  - Hypertrophic
  - Keloid
  - Depressed
- Atrophic or spread
- Shape
  - Linear or curved
  - Trap-door semicircular
  - Web or broken line
  - Stellate or broken line
  - Ice picked or pitted
  - Overhanging or avulsion

A variety of scar scales have also been described such as the Vancouver Scar Scale, the Patient and Observer Scar Assessment Scale, the Manchester Scar Scale and the Stony Brook Scar Evaluation Scale.

Having a clear history of the events leading to scar formation, the type of scar and any previous scar revision treatments are helpful in planning further management. The indication for scar revision may be cosmetic and/or functional. Prior to undertaking any form of scar revision, it is important to understand the patients’ goals and to manage their expectations. The objectives of scar revision should be discussed in detail with the patient and may include hiding the scar, improving the direction, redirecting tension, levelling contours, narrowing scar width, shortening linear components and/or camouflaging the scar. It is not uncommon for patients to have misconceptions about scar revision and they should understand the degree of improvement that may realistically be achieved and that it is not possible to make the scar completely disappear. Patient counselling and detailed informed consent is critical.

Good candidates for scar revision include those whose scars have the following characteristics:
- Longer than 20 mm
- Wider than 1-2 mm
- Scars disturbing function
- Fitzpatrick skin type I or II
- Scars over immobile areas

In contrast, poor candidates for scar revision, include those with the following characteristics:
- Fitzpatrick skin types IV to VI
- Scars against RSTLs
- Scars in mobile areas
- Scars over bony prominences

Traditional teaching on the timing of scar revision has been to wait at least 6 months to 1 year as remodelling continues and scars, in general, improve with time. In some cases, however, where it is clear that the scar will still be problematic even after significant maturation, it may be appropriate to undertake earlier scar revision. For example, a scar which lies perpendicular to RSTLs is unlikely to improve significantly with maturation alone and earlier scar revision may be appropriate.

Table 1: Factors to consider to reduce the risk of an unfavourable scar

<table>
<thead>
<tr>
<th>Preoperative Considerations</th>
<th>Intraoperative Considerations</th>
<th>Postoperative Considerations</th>
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<tbody>
<tr>
<td>Nutrition</td>
<td>Incision planning</td>
<td>Prevent infection</td>
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<td>Medication</td>
<td>Mitotixuous soft tissue handling</td>
<td>Sun avoidance</td>
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<td>Co-morbidity</td>
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<td>Smoking</td>
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Scar revision therapies have evolved in recent years and may be divided into 3 categories:

1. Invasive surgical techniques
2. Minimally invasive techniques
3. Non-surgical therapies

Individual techniques may be used alone or in combination. The choice of treatment is determined following careful analysis of the scar which includes its position, location and age of the scar, as well as taking into account the skin type and patient factors. The aim of this article is to provide an overview and an update on treatment options for scar revision.

**Invasive surgical techniques**

There are a range of surgical techniques based on scar excision and/or scar irregularization methods. Other options include regional flaps and free grafts. For depressed scars, soft tissue augmentation may also be required and this may be performed surgically or with injectable filler materials.

Adequate anesthesia and meticulous surgical technique are essential. The single most important step in wound closure is the precise re-approximation of the dermal layer. Other important requirements include proper instrumentation, appropriate suture materials, closure of any dead space and careful handling of tissue.

The method of scar excision can also affect the erosion of skin edges, for example bevelling the scalped away from the closure line helps erosion on wound closure. An M-plasty may be used to shorten a line of incision and spare tissue if necessary. Undermining can help reduce wound tension but extensive undermining is often counterproductive, as tension is not reduced any further beyond 6cm and the vascularity of the flap may be compromised.

**Scar excision techniques**

Simple excision

Simple full-thickness excision with primary closure in a RSTL or at the junction of facial subunits may be used to improve malaligned or malpositioned scars. To avoid standing cone deformities, the apical edge of the ellipse ideally should not be more than 30 degrees.

**Scar repositioning**

It may be possible to reposition a scar to a more hidden site such as within RSTLs, at the border of facial subunits or within hair-bearing skin. This technique can be used for scars lying close to these sites such as from rhinoplasty or parotidectomy.

**Serial excision**

This technique may be used to revise a scar which is too large to close in a single definitive procedure. Serial partial excisions of the scar are performed at intervals of 2 to 3 months and rely on the skin's ability to stretch over this time.

When revising large scars, other options that may be considered include skin expansion, skin grafts and flaps.

**Scar irregularization techniques**

Scar irregularization techniques include Z-plasty, W-plasty and geometric broken line closure. They can make a scar less conspicuous by forming an irregular pattern that is less noticeable to the eye. These techniques are often followed up with local dermabrasion 6 to 8 weeks afterwards to refine the final result.

**Z-plasty**

This is a double transposition flap (of two triangular flaps) which can be used to lengthen and realign a scar. The increase in length of the scar varies depending on the apical angle of the triangular flaps (Table 2).

The Z-plasty technique may be used for contracted scars, for scars that distort adjacent structures such as the eyelid, nostril or lips and to realign scars that are more than 30 degrees off the RSTLs. Multiple Z-plasties may be used for larger scars with the same benefits.

**W-plasty**

W-plasty involves excising the scar and adjacent tissue using multiple connected small triangles to form a regularly irregular pattern which is less noticeable (Fig. 1). The limbs of each triangle should be between 5mm to 7mm (longer limbs are more conspicuous) and ideally one of the two limbs should be parallel to the RSTL. The scar is therefore broken up into smaller parts with much of it parallel to RSTLs and this technique may be used to realign scars. To avoid a standing cone deformity, the angle at each end of the W-plasty should not exceed 30 degrees.

In contrast to the Z-plasty, the W-plasty involves excision of adjacent tissue but does not change the length of the scar. A further difference is that whereas the Z-plasty involves transposition flaps, the W-plasty involves advancement of the wound edges in an interdigitating pattern. A limitation of the W-plasty therefore is that it should only be performed in areas where there is sufficient laxity to allow excision and bilateral advancement. A further limitation is that as there is a regular repetitive element to the irregular pattern, it may become more noticeable with larger scars (greater than 4cm) and in these cases, the geometric broken line closure technique may be more appropriate.

**Geometric broken line closure**

Geometric broken line closure is a technique that evolved from the W-plasty and involves excising the scar and adjacent tissue using a series of alternating random shapes such as triangles, squares, rectangles, trapezoids and semicircles. The mirror image is created on the other side of the wound forming an irregularly irregular pattern which is less noticeable and can be used to realign and camouflage larger scars that may cross facial subunits.

**Minimally invasive techniques**

**Subcision**

Subcision is a technique which may be used for depressed scars or wrinkles. It involves puncturing the skin with a tri-bevelled hypodermic needle and manoeuvring its sharp edges under the defect to make subcuticular cuts, thereby releasing the tethered bound-down site with subsequent formation of new connective tissue. A history of keloids is a contraindication to the use of this technique.

**Recollagenation**

Recollagenation is the application of a stimulator material or collagen production which may be used effectively for small depressed post-traumatic scars or acne. Various bio-materials have been used although autologous fascia is optimal e.g. fascia lata.

**Dermal grafting**

Dermal grafting may be used for atrophic scars. It involves performing an initial subcision 2 weeks before grafting, following which a dermal graft is obtained from post-auricular skin (after excising the epidermis) and placed in a pocket or tunnel under the scar, improving the depression of the scar.

**Skin resurfacing techniques**

Resurfacing techniques remove the superficial skin (epidermis and part of the papillary dermis) to smooth the scar and allow it to blend with the surrounding tissue. After removal of the superficial skin, the area is allowed to re-epithelialize from the surrounding epithelium and deeper dermal appendages. The dermal appendages lie within the reticular dermis so great care must be taken not to penetrate this, which otherwise could lead to further scarring. The best time to perform resurfacing is thought to be at 6 to 8 weeks after surgery during the remodelling phase when there is still fibroblastic activity.

The three main resurfacing techniques are dermabrasion, laser and chemical peels. Dermabrasion, one of the oldest techniques, involves mechanical removal of the superficial skin, most commonly with a diamond fraise. Carbon dioxide and erbium lasers are the commonest lasers used in skin resurfacing. Chemical peels include glycolic acid, trichloroacetic acid, Jessner solution, salicylic acid and tretinoin. Antiviral prophylaxis against herpes is recommended by some to all patients, irrespective of whether there has been a previous history and should be started 0 to 2 days pre-operatively. Post inflammatory hyperpigmentation is more common in those with a higher
Corticosteroids
Intralesional corticosteroids are commonly used as first and second line treatments for keloid and hypertrophic scars respectively \(^{19,20}\). They are considered relatively safe and effective although some scars do not achieve a good response \(^{21,22}\). Injection of corticosteroid is into the dermal portion of the scar and it often has to be repeated, typically every 2 to 4 weeks depending on response. \(^{23,24}\) Triamcinolone acetonide (Kenalog; Bristol-Myers Squibb Company) is usually used at a concentration of 10mg/mL to 20mg/mL but may be used at a higher strength of 40mg/mL for more resistant lesions. \(^{25}\) There is a risk of irreversible epidermal atrophy if corticosteroid is injected too superficially. Other risks of intralesional corticosteroids include telangiectasia and hypopigmentation, which may be reduced by using a lower dose. Low recurrence rates of keloid and hypertrophic scars have also been reported when corticosteroids are combined with corticosteroid ointment after surgical excision \(^{26}\).

5-Fluorouracil
Studies have shown intralesional 5-Fluorouracil (5-FU) to be an effective and safe treatment of keloid and hypertrophic scars. 5-FU, a pyrimidine analogue and antimetabolite, acts as a 5% topical preparation for the treatment of keloid scars. \(^{27,28}\) Injection of the corticosteroid is into the dermis and it often has to be repeated, whilst others have not. \(^{29}\) The results on keloid and hypertrophic scars have also been mixed \(^{30,31}\). Onion extract may however improve the effectiveness of intralesional steroids when used in combination for hypertrophic and keloid scars. \(^{32}\)

Onion extract
Onion extract, also known as Allium cepa, has anti-inflammatory, antibiotic and fibroinhibitory properties \(^{33}\) and may be used as a topical gel on scars. There have been mixed results in studies with some reporting an improvement in scar symptoms and/or appearance \(^{34-36}\) whilst others have not. \(^{37}\) The results on keloid and hypertrophic scars have also been mixed \(^{38,39}\). Onion extract may improve the effectiveness of intralesional 5-FU in scars. \(^{40}\)

Bleomycin
Bleomycin, a cytotoxic antibiotic, has been reported to be effective and safe in the treatment of hypertrophic and keloid scars \(^{41,42}\). The exact mechanism of bleomycin in these scars remains unclear although the adverse reactions of intralesional bleomycin are much less extensive than systemic bleomycin. \(^{43}\) Larger studies are required to further assess the use of bleomycin in scars. \(^{44}\)

Mitomycin C
Mitomycin C, a chemotherapeutic agent that inhibits DNA synthesis and fibroblast proliferation, has had mixed results in keloid recurrence when applied as a topical agent after excision, from mainly small studies \(^{45,46}\). Laser therapy
Laser technology has greatly advanced in recent years and may be an effective treatment option for a variety of scars including hypertrophic scars, keloid scars, atrophic traumatic scars and acne scars. \(^{47}\) The principles of treatment are based on selective photothermolysis \(^{48}\) and requires proper scar categorization so that an appropriate laser wavelength and treatment parameters are chosen. Skin type classification is also important as the risks of adverse effects with laser therapy, particularly ablative lasers, are greater with a higher Fitzpatrick skin type. \(^{49,50}\)

Imiquimod
Imiquimod, an immune response modifier, has been used as a 5% topical preparation for the treatment of keloid and hypertrophic scars. Studies have reported lower recurrence rates with topical imiquimod after excision of keloids. \(^{51}\) Other studies, however, have not found topical imiquimod to be effective at reducing long term recurrence of keloid scars, particularly on the trunk. \(^{52}\) Another study showed that topical imiquimod was most effective at reducing recurrence of excised keloids at the pinna (where there is less tension) compared to the chest wall and neck. \(^{53}\) There is a need for larger high quality studies on the use of topical imiquimod in scars. \(^{54}\)

Pressure therapy
Pressure therapy, one of the simplest measures, has been used consistently in the management of scars, particularly hypertrophic scars from burns. \(^{55}\) It is thought that by immobilising the local muscles, this leads to reduced skin tension and therefore reduced microtrauma and subsequent inflammation. \(^{56}\) Other studies have also shown improved post-surgical scars with Botulinum toxin \(^{57}\). Botulinum toxin A has also been used in keloids although it has had mixed results \(^{58,59}\) and requires further investigation.

Cosmetics
Camouflage make-up can be very useful. It is effective and may be used instead of or in addition to surgery. It has the advantages of being very safe and allowing sooner return to function. \(^{60}\)

Emerging therapies
There are numerous emerging therapies for the prevention and treatment of scars which continue to be under investigation, such as Botulinum toxin A, Angiotensin-converting enzyme (ACE) inhibitors and Transforming growth factor α. \(^{61}\)

Botulinum toxin A has been reported to enhance healing and improve the appearance of scars from facial lacerations when injected into the musculature adjacent to the wound within 24 hours after wound closure. \(^{62}\) It is thought that by immobilising the local muscles, this leads to reduced skin tension and therefore reduced microtrauma and subsequent inflammation. \(^{63}\) Other studies have also shown improved post-surgical scars with Botulinum toxin. \(^{64}\) Botulinum toxin A has also been used in keloids although has had mixed results \(^{65,66}\) and requires further investigation.

Angiotensin II has been shown to induce fibroblast and keratinocyte migration and induce type I collagen gene expression in human dermal fibroblasts, playing an important role in wound healing. \(^{67,68}\) Angiotensin-converting enzyme (ACE) inhibitors reduce the formation of angiotensin II from angiotensin I and have been shown to bias. \(^{69}\)

Pressure therapy
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The transforming growth factor β (TGF-β) superfamily plays a key role in wound healing. Avotermin (recombinant human transforming growth factor-β3) has shown to significantly improve scar appearance following scar revision surgery compared with placebo in a randomized, double-blind, within-patient, placebo-controlled, phase II clinical trial. \(^{70}\)
Facial scars can cause great distress to patients and over recent years, scar revision remains challenging. There are a wide range of therapies also showing promise. Treatment options are undertaking any scar revision. There are a wide range of managing their expectations is important prior to best practice. Some treatments also appear to be more effective when used in combination and there is a need for more large scale high quality studies to further define best practice.

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References


Endoscopic transorbital neuroendoscopic surgery (TONES)

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Abstract

Transorbital neuroendoscopic surgery (TONES) is an evolving and exciting subspecialty of the endoscopic skull base surgeon. It uses the orbit as a surgical pathway to reach previously inaccessible areas allowing multiple trajectories of approach, improving visualisation and lesion manipulation. This new approach has resulted in the discovery of new endoscopic anatomy of the orbit, not described in the ENT literature.

Key words
Transorbital; Transconjunctival; CSF leak; Endoscope; Neuroendoscopy; Neurosurgery; Skull base; Anterior cranial fossa.

Introduction

Using the orbit as a surgical pathway to the lateral third of the skull base allows access to previously inaccessible areas that are difficult-to-access areas. These portals also provide excellent access to certain areas within the frontal and sphenoid sinuses that are difficult to reach with traditional endoscopic transnasal surgery.

History and rationale behind TONES

Kris Moe and his team from the University of Washington were one of the first to successfully use transorbital portals to manage lesions involving the orbit, the anterior skull base and middle cranial fossa.

Each bony orbit provides four portals, and with the nose providing two portals for manipulation of an endoscope and surgical instruments, ten portals are available to be used in various combinations (Figure 1). This allows for the target area to be approached from different trajectories, improving visualization and lesion manipulation.

Figure 1: The ten portals demonstrated (4 transorbital through each orbit and 2 transnasal).

Figure 2: The ten portals demonstrated (4 transorbital through each orbit and 2 transnasal).

neurosurgeons because of fear and uncertainty; uncertainty about the anatomy and fear of damaging neurovascular structures or muscles that would lead to visual impairment in a patient who often has little ocular symptoms.

There is no doubt that superior visualization is obtained using an endoscope within the orbital cavity. A similar evolutionary progression is emerging as was seen with the advent of endoscopic sinus surgery. The endoscopic anatomy of the eye is being described in new textbooks with structures being rediscovered by surgeons who previously accessed areas without appreciating structures not previously described in the ENT literature. A good example of this is Horner’s muscle – a little muscle in the medial aspect of the orbit that directs one to the anterior ethmoidal artery. The anatomy and function of this muscle was not appreciated by otolaryngologists when doing a Lynch-Howarth approach to the medial orbit.

Four transorbital pathways allow otolaryngologists, ophthalmologists and neurosurgeons to access the orbit for intracranial and extracranial lesions, the parasial sinuses and intracranial tumours.
The medial (precaruncular) portal provides easy, quick and safe access to the anterior and posterior ethmoidal arteries. For intractable epistaxis secondary to nasoethmoidal fractures, this approach avoids an external incision and allows for quick recovery without the need for suturing the wound (Figure 2). A medial 180 degree optic nerve decompression can be performed by either an ophthalmologist trained in endoscopic techniques or by an endoscopic sinus surgeon. This approach can also be used for medical decompression in thyroid eye disease. The advantage of the precaruncular approach is the potential to preserve normal paranasal sinuses and provide a slightly different trajectory to that obtained by the transnasal route. A contralateral precaruncular approach gives excellent access for repair of a cerebrospinal fluid leak secondary to a Sternberg canal defect in the lateral wall of a well-pneumatized sphenoid sinus. Even with the traditional transpterygoid approach, visualization and manipulation of instruments in a very lateral defect can be difficult and palatal numbness is often a consequence of the surgery due to dissection through the pterygopalatine fossa. The contralateral precaruncular approach allows for direct visualization of the defect using a zero degree 4mm endoscope. Conventional straight instruments can reach the defect (in our first case, was found to be 83mm from the medial canthus of the eye.)

The lateral portal can address pathology of the lateral orbit and will most likely supersedes a lateral bony orbitotomy for procedures that require removal of the lateral orbital wall. The advantage of this approach is that the orbital rim is left intact, obviating the need for reconstruction of the rim using plates. All three components of sphenoid wing meningiomas can be managed using a single lateral endoscopic approach (Figure 3)

- The bony hyperostotic lateral orbital wall can be drilled away to expose the temporalis muscle laterally, the dura superiorly and posteriorly. The dissection can continue to the superior and inferior orbital fissures. (Figures 4 & 5)
- The more hyperostotic bone present, the wider the surgical portal to access the intracranial tumour. Using aribbon retractor to medialize the orbit widens the surgical pathway. It is imperative that the pupil size and shape is monitored every few minutes during retraction of the orbital contents. A dilated or oval pupil indicates raised intraocular pressure or excessive traction on important neurovascular structures. All instruments should be removed from the surgical field to give the eye time to recover before continuing with surgery. It is advisable to have an ophthalmologist assist to help monitor the eye.

The superior eyelid crease approach provides good access to the superior orbit to remove orbital lesions, the frontal sinuses and the anterior cranial fossa. Repair of the orbital roof is mostly not indicated and the pulsation of dura on the orbital contents settles within 2 weeks. It is often impossible to access the lateral aspect of a well-pneumatized frontal sinus through the nasal cavity, even with an endoscopic Lothrop operation. The superior portal can therefore be used in conjunction with a transnasal approach for lesions / defects involving the lateral frontal sinus. This approach has been used to remove plexiform neurofibromas of the eye, hemangiomas, cystic lesions such as hydatid cysts and encephalocoeles and to repair CSF-leaks arising from the frontal sinus or anterior cranial fossa. Neurovascular structures such as the supratrochlear nerve and recurrent branch of the middle meningeal artery can be identified with clarity (Figure 6).

The inferior approach is useful for blow-out fractures of the orbit, especially in cases where the extraocular muscles are entrapped. The transconjunctival incision is made onto the rim of the inferior orbit, a minimum of 2mm inferior to the tarsus. With the orbit deflected superiorly, the endoscope has a wide surgical pathway with structures such as the infraorbital canal being clearly visualized. Endoscopic visualization allows for precise placements of grafts or plates to reconstruct the orbital floor.

The value of the 10 surgical portals is the fact that they can be used in many combinations to address specific areas that have been previously difficult to access. Examples illustrating the combinations that can be utilized are the following:
- Medial and lateral portal for a balanced two-wall decompression in thyroid orbitopathy.
- Medial portal + one or both nostrils (3 portals) for optic nerve decompressions, lesions involving the sphenoid sinus and encasing the optic nerve.

The most difficult area to dissect is the orbital component of sphenoid wing meningiomas. Due to invasion of the periorbita, especially at the level of the superior orbital fissure, differentiation between extraocular muscles, fat and tumour can be extremely challenging.

Three surgical incisions are available to the surgeon contemplating a lateral approach. The choice of incision depends on the indication for surgery:
- The extended superior eyelid crease approach. This approach is the workhorse approach for most lesions affecting the superior and lateral orbital walls, like sphenoid wing meningiomas and for accessing the anterior and middle cranial fossa
- Retrocanthal approach. This approach is useful if only a biopsy is required.
- The lateral canthotomy approach is used less commonly since the recovery time is longer than the other two approaches and the incision more bothersome than a larger extended superior eyelid crease approach.

The lateral approach (right eye) demonstrating the temporalis muscle laterally, the bony tunnel with dura superiorly and posteriorly and the periorbita medial to the retractors.

**Figure 2:** Precaruncular approach requiring no suturing - for access to the ethmoidal arteries, optic nerve and sphenoid sinus.

**Figure 3:** The lateral portal to address pathology of the lateral orbit.

**Figure 4:** The lateral approach demonstrates the temporalis muscle laterally, the bony tunnel with dura superiorly and posteriorly and the periorbita medial to the retractors.

- The most difficult area to dissect is the orbital component of sphenoid wing meningiomas. Due to invasion of the periorbita, especially at the level of the superior orbital fissure, differentiation between extraocular muscles, fat and tumour can be extremely challenging.

Three surgical incisions are available to the surgeon contemplating a lateral approach. The choice of incision depends on the indication for surgery:

- The extended superior eyelid crease approach. This approach is the workhorse approach for most lesions affecting the superior and lateral orbital walls, like sphenoid wing meningiomas and for accessing the anterior and middle cranial fossa
- Retrocanthal approach. This approach is useful if only a biopsy is required.
- The lateral canthotomy approach is used less commonly since the recovery time is longer than the other two approaches and the incision more bothersome than a larger extended superior eyelid crease approach.

The superior eyelid crease approach provides good access to the superior orbit to remove orbital lesions, the frontal sinuses and the anterior cranial fossa. Repair of the orbital roof is mostly not indicated and the pulsation of dura on the orbital contents settles within 2 weeks. It is often impossible to access the lateral aspect of a well-pneumatized frontal sinus through the nasal cavity, even with an endoscopic Lothrop operation. The superior portal can therefore be used in conjunction with a transnasal approach for lesions / defects involving the lateral frontal sinus. This approach has been used to remove plexiform neurofibromas of the eye, hemangiomas, cystic lesions such as hydatid cysts and encephalocoeles and to repair CSF-leaks arising from the frontal sinus or anterior cranial fossa. Neurovascular structures such as the supratrochlear nerve and recurrent branch of the middle meningeal artery can be identified with clarity (Figure 6).

The inferior approach is useful for blow-out fractures of the orbit, especially in cases where the extraocular muscles are entrapped. The transconjunctival incision is made onto the rim of the inferior orbit, a minimum of 2mm inferior to the tarsus. With the orbit deflected superiorly, the endoscope has a wide surgical pathway with structures such as the infraorbital canal being clearly visualized. Endoscopic visualization allows for precise placements of grafts or plates to reconstruct the orbital floor.

The value of the 10 surgical portals is the fact that they can be used in many combinations to address specific areas that have been previously difficult to access. Examples illustrating the combinations that can be utilized are the following:
- Medial and lateral portal for a balanced two-wall decompression in thyroid orbitopathy.
- Medial portal + one or both nostrils (3 portals) for optic nerve decompressions, lesions involving the sphenoid sinus and encasing the optic nerve.
Management of primary tracheal tumours

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Abstract
Primary tracheal tumours are rare accounting for <0.1% of tumours. Tumours are often diagnosed at an advanced stage requiring radical surgery and adjuvant therapy for disease control. Other management options include tracheal stenting or tumour debidement for symptomatic control. The aim of this article is to provide an updated overview of diagnosis and management of primary tracheal tumours.

Key words:
Primary Tracheal Tumour, Management

Introduction
Primary tracheal tumours (PTT) are extremely rare accounting for 0.1 – 0.4% of tumours1,2. Tumours are commonly benign in children and malignant in adults3. Diagnosis is often delayed, as the main presenting symptom is shortness of breath. This is commonly misdiagnosed as asthma or chronic obstructive pulmonary disorder and patients only undergo further investigations once the tumour has advanced in size to cause airway obstruction or hemoptysis3. Histological subtype and disease staging guide the management of tracheal tumours. Options include surgical resection, often with adjuvant treatment4. Radiotherapy may have a palliative role along with endotracheal stenting or tumour debidement for symptomatic relief in inoperable cases5. Stenting and debulking can also be used as a temporary measure prior to definitive treatment.

Pathology
Both benign and malignant PTT originate either from ciliated epithelial cells, minor salivary glands or mesenchymal components of the tracheal. In adults, 90% of PTT are malignant and the reverse is observed in the paediatric population, with the majority of PTT (80 – 90%) being benign. The predominant histological subtypes of PTT in adults are squamous cell carcinoma (SCC) and adenoid cystic carcinoma (AdCC), seen in 75% malignant cases. Squamous cell carcinoma of the trachea has a male predominance and is associated with tobacco smoking. Regional metastasis to the mediastinum and lungs can occur in up to 30% of cases. Adenoid cystic carcinoma has an equal sex distribution with no correlation to smoking. These tumours are associated with slow growth, perineural invasion and delayed presentation of distant metastasis independent of loco-regional disease control. Majority of tracheal tumours within the peadiatric population are benign in nature. Infantile hemangiomia, granular cell tumours, myofibroblastic tumour and juvenile xanthogranuloma, although all rare, make up the common subtype of benign tracheal tumours seen in children.

References
The broad histological variant of primary tracheal tumours is summarized in Table 1.

**Table 1: Classification of primary tracheal tumours**

<table>
<thead>
<tr>
<th>Benign</th>
<th>Malignant</th>
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<tbody>
<tr>
<td>Papilloma</td>
<td>Squamous cell carcinoma</td>
</tr>
<tr>
<td>Papillomatosis</td>
<td>Adenoid cystic carcinoma</td>
</tr>
<tr>
<td>Salivary gland tumours</td>
<td>Mucoepidermoid carcinoma</td>
</tr>
<tr>
<td>Lymphoma</td>
<td>Carcinoma ex pleo</td>
</tr>
<tr>
<td>Mesenchymal tracheal tumours</td>
<td>Chondrosarcoma</td>
</tr>
<tr>
<td>Fibroma</td>
<td>Carcinoma</td>
</tr>
<tr>
<td>Hemangioma</td>
<td>Adenoid cystic carcinoma</td>
</tr>
<tr>
<td>Paraganglioma</td>
<td>Carcinoma</td>
</tr>
<tr>
<td>Glomus tumour</td>
<td>Adenoid cystic carcinoma</td>
</tr>
<tr>
<td>Condroma</td>
<td>Carcinoma</td>
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<tr>
<td>Leiomyoma</td>
<td>Carcinoma</td>
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</table>

**Surface epithelial tracheal tumours**

- Surface epithelial papillomatosis
- Surface epithelial papilloma

**Table 2: TNM classification for primary tracheal tumour**

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<thead>
<tr>
<th>T</th>
<th>N</th>
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<tr>
<td>T0</td>
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<tr>
<td>T1</td>
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<tr>
<td>T4</td>
<td>N0</td>
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**Management**

- Current practice in management of PTT involves surgical resection and adjuvant radiotherapy when indicated. There have been no randomized clinical trials to validate the best treatment option for the management of PTT due to the rarity of these cases and the limited number of patients available for study. Selecting the optimum treatment option should involve multidisciplinary team approach, with consideration of factors including patient fitness for surgery, tumour histology and disease staging.

- When patients present acutely with symptoms of airway obstruction, an endotracheal approach with tumour debulking or airway stenting may be necessary. Restoring patency and maintaining airway integrity are essential. The approach for malignant PTT may require an anastomosis and preserving meaningful respiratory function. The anaesthetic team should be informed prior to opening the trachea for inspection, which is often done distal to the tumour. Once distal margins are deemed adequate, the anastomosis is performed, and the proximal segment of trachea can be excised for complete resection.

**Diagnosis**

Due to the non specific nature of symptoms associated with tracheal tumours, a high index of suspicion by clinicians to investigate patients further is required, particularly in cases of non resolving symptoms with initial medical management or in patient with a high risk profile.

**Endotracheal debriement and stents**

Endotracheal tumour debriement is often performed to provide symptomatic relief of airway obstruction. This may be required for control of acute tracheal tumour associated haemorrhage. Tracheal lumen patency can be established with cold steel tumour debriement, electrocautery, coblation or laser debriement, prior more definitive treatment. In patients with advanced unresectable tumours, palliative symptomatic airway control can be achieved with endotracheal stenting. Tracheal stents can be placed under sedation with flexible bronchoscopy or rigid bronchoscopy under general anaesthesia. Radiotherapy Current evidence on adjuvant therapy is drawn from outcomes based on retrospective cohort reports. Current indications for radiotherapy post surgical resection of PTT are for cases with positive margins and intermediate or high grade malignant tumours. The observed outcomes date to are encouraging as both squamous cell carcinoma and adenoid cystic carcinoma are relatively radiosensitive tumours. There is a role for radiotherapy in the management of unresectable tracheal tumours or for symptomatic palliation. For microscopic retained disease, 60 Gy photon radiation is administered over a period of 6–8 weeks. The dose is increased to 60–70 Gy for macroscopic residual tumour control.

**Chemotherapy**

There are currently no recognized treatment regimes to support the use of concurrent chemotherapy in addition to surgery and radiotherapy for PTT. Combined chemoradation has been described in several case reports of inoperable PTT. A combination of cisplatin, 5-fluorouracil, etoposide was used with 60 Gy of radiation in 30 fractions for an inoperable tracheal SCC. The patient was reported to be free of disease 24 months post treatment. There is evidence to support role of palliative chemotherapy with radiotherapy to improve locoregional control of advanced tracheal adenoid cystic carcinoma, however with no improvement of overall survival.

**Conclusion**

Primary tracheal tumours are rare neoplastic lesions of the airway with a wide mix of both benign and malignant tumour histopathology. Most patients with PTT are...
diagnosed at an advanced stage due to the non specific nature of presenting symptoms. Primary surgical resection of disease with adjuvant radiotherapy in select cases prolongs improves survival. Radiotherapy can be used in managing inoperable cases and symptom palliation.

References

Abstraction
Poorly differentiated thyroid carcinoma (PTDC) represents a subgroup of aggressive thyroid cancers that occur in a histological mid-ground between differentiated thyroid carcinoma and anaplastic thyroid carcinoma. There is little evidence as to the optimal treatment strategy for patients with this diagnosis. This paper explores the current evidence and offers advice on how to maximize survival and function in these patients. PDTC is locally aggressive and often presents with distant disease. A thorough work up should include comprehensive imaging of the primary disease and the sites of likely metastasis. Vocal cord palsy is a prognostic indicator and function should be documented pre-operatively.

Cross sectional disease clearance is the priority of treatment. Radioactive iodine should be trialed despite loss of iodine avidity associated with the process of de-differentiation. Other adjuvant therapies can be explored on an individualized basis in the setting of a specialized multidisciplinary team. With microscopic clearance, individualized adjuvant therapy and close monitoring patients can expect an overall survival of around 70%.

Keywords
Thyroid carcinoma; poorly differentiated.

Introduction
Poorly differentiated thyroid carcinomas (PTDC) represent a distinct subgroup of aggressive thyroid cancers. Despite their aggressive behaviour they do not display the universally lethal outcomes apparent in anaplastic thyroid cancers. Their diagnosis and management present significant challenges. However, most patients should be given the opportunity to be treated as over 60% of them can be cured and henceforth live normal lives. PTDC demonstrate significantly heterogeneous pathological features ranging from well differentiated areas to elements of anaplastic change. Their incidence ranges from 4 to 7% of all thyroid cancers1. Detailed studies on PDTC are lacking in the published literature and controversy exists regarding pathological diagnosis and management. In terms of prognosis they represent a mid-ground between differentiated thyroid carcinomas (DTC), which have an excellent long-term prognosis2, and anaplastic thyroid carcinomas which are almost invariably fatal3. Despite their potentially aggressive behavior most patients with PDTC can be adequately treated with a combination of surgery and adjuvant therapies. It is thus important to understand the tumour biology, natural progression and prognosis of such cancers to determine the appropriate level of intervention required.

Differentiated thyroid carcinomas represent 1% of all malignancies and are usually curable with surgery and radioactive iodine1. PDTCs although representing a small proportion of thyroid carcinomas account for a significant proportion of the morbidity and mortality related to this disease spectrum4. It has been shown that in comparison with follicular thyroid carcinoma, the presence of 16% PDTC within a given tumour affects prognosis significantly5. However with appropriate management 5-year locoregional and distant control of 81 and 59% respectively can be achieved6. The only recognized prognostic indicators of worse disease specific survival are T4a or M1 disease with patients more frequently succumbing to the effects of distant disease in comparison to those with anaplastic thyroid carcinoma in whom mortality is due to more commonly secondary to upper airway obstruction7.

Tumour heterogeneity makes pre-treatment diagnosis of PDTC difficult, which is concerning in a condition associated with high rates of locally advanced disease and metastasis on presentation8. Those initially under-staged risk a delay in treatment in a condition known to be locally

Poorly differentiated thyroid carcinoma
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invasive and potentially aggressive. Those over-staged risk being categorized as inoperable and/or palliative due to comparison with the more aggressive and almost universally fatal anaplastic subtype. Poor outcome from PDTC is statistically more likely if the patient is aged over 45 years at presentation and/or the histopathology reveals the presence of necrosis and a mitotic index of ≥3 per 10 HPF.

The WHO Classification of Endocrine Tumours in 2004 defined PDTC as a thyroglobulin-producing non-follicular, non-papillary thyroid carcinoma having an intermediate behavior between well-differentiated and anaplastic carcinomas.

The purpose of this review is to highlight the clinical presentation, evaluation, management and outcomes of patients diagnosed with PDTC.

Clinical Presentation.
PDTC often presents at an advanced stage. Up to 80% of patients have nodal involvement and 30% gross extra-thyroidal extension (ETE) at the point of diagnosis. Metastasis (M1) at presentation is also far more common in PDTC with ETE than in those with purely DTC and ETE at 37% compared to 9%. High rates of M1 disease at presentation in addition to high rates of distant failure account for the fact that most patients who have a fatal outcome will die of distant disease.

Clinical Evaluation
Clinical evaluation should include a full head and neck examination including fibreoptic laryngoscopy. In a study of 365 consecutive patients undergoing thyroidectomy, 21 had invasive disease. Of those 70% had a recurrent laryngeal nerve palsy pre-operatively compared to only 0.5% with non-invasive disease. Given the propensity for vocal cord palsy at presentation in PDTC, evaluation and surgical planning should include documentation of vocal cord function at initial assessment. In addition to fibreoptic laryngoscopy, patients with suspected visceral involvement should undergo laryngo-tracheo-bronchoscopy and oesophagoscopy to accurately plan surgical management. Significant tracheal invasion would require tracheal resection with either direct closure or tracheostomy placement to secure the airway during recovery.

Biochemical evaluation
Biochemical evaluation should be as for differentiated thyroid carcinoma and should include thyroid function testing and a baseline of thyroid antibodies. Although once de-differentiated, the ability to concentrate iodine is markedly reduced; it has been shown that PDTC often has a differentiated component and may respond to radioactive iodine ablation. It has also been shown that those with an undetectable post-operative thyroid antibodies have a low rate of recurrence. As such, obtaining a baseline count of circulating thyroid antibodies can help monitor the effectiveness of treatment which is particularly useful in a disease characterized by a high rate of distant failure.

Radiological evaluation
Ultrasound. PDTC displays similar features on ultrasound to anaplastic thyroid cancer. These include heterogeneous echogenicity (93%), solitary nodules (80%), a circumscribed margin (63.3%) and oval-to-round shape (63.3%). However, in comparison to anaplastic thyroid cancer, the circumscribed margin and oval-to-round shape were significantly more frequent in PDTC in a retrospective study of 55 patients. 

Cross Sectional Imaging. Due to the locally invasive and aggressive behaviour demonstrated in PDTC, cross sectional multipleplanar views with contrast three-dimensional imaging of the neck and thorax is recommended in the diagnostic work-up of these patients. CT with contrast is recommended to demonstrate structural involvement including tracheal invasion, nodal involvement, particularly in the mediastinum and pulmonary metastasis (figure 1). As the majority of metastasis are to the lung or bone, CT imaging is ideally placed to diagnose these lesions. In tumours with extra-thyroidal extension MRI is of additional benefit when exploring invasion of musculature and vessels (figure 2).

Functional imaging may also be helpful in the initial evaluation of PDTC. Most studies have demonstrated that PDTC have low thyroglobulin production and are often avid on FDG-PET scanning reflecting their undifferentiated nature although it has been found that at least 25% retain the ability to concentrate radioactive iodine.

As such radioactive iodine uptake imaging may be useful for disease detection in PDTC particularly when metastatic disease has not been detected by FDG-PET. Whole body scanning as part of a full diagnostic work up is further encouraged by the fact that PDTC can have multiple bony metastases at presentation, with the femur and skull being common sites for the occult primary in PDTC. Distant metastasis is present in 36-85% of patients with PDTC and those with M1 disease are three times more likely to die as a result of their disease. The recognized flip-flop phenomenon noted in progression from differentiated thyroid carcinoma to PDTC explains that whilst FDG-PET activity increases with progression of disease, radioactive iodine uptake decreases. As such, the two modalities may provide complimentary information.

Cytology
International standardization of the cytological criteria for diagnosing PDTC followed an international consensus meeting in Turin, Italy in 2006.1: This consisted of (1) the presence of solid/trabecular/insular pattern of growth, (2) absence of conventional nuclear features of papillary carcinoma, and (3) presence of at least one of two outlined nuclei; mitotic activity of at least 3 per 10 HPF or; tumour necrosis (Figure 3). Due to significant overlapping features on cytology PDTC is most commonly confirmed on post-operative thyroidectomy specimens. Severe nuclear overlapping and crowding is often seen in PDTC and can mimic papillary thyroid carcinoma. A micro-follicular pattern on cytology with nuclear grooving and overlapping are generally labeled as follicular-variant papillary thyroid carcinoma whereas those without nuclear grooving may be described as a follicular neoplasm and be classified as Thy3f in cases where PDTC should make the list of differentials. Medul lary thyroid carcinoma has several overlapping features with PDTC; an insular pattern, plasmacytoid cells, bimuleate cells and abrupt nuclear enlargement however immunological staining for thyroglobulin will reveal the true nature of the specimen and avoid progression down a completely different management path.

Prior to publication of the Turin criteria from the Departments of Pathology at the University of Turin and the Mauriziano Hospital in Italy, PDTC was considered any thyroid carcinoma termed solid, trabecular, insular, poorly differentiated, intermediate type, primitordial cell, less well differentiated or follicular carcinoma with insular component and as such outcomes from this heterogeneous group were difficult to measure accurately. Decaussin et all found that insular pattern on histopathological assessment was an independent prognostic indicator of poor outcome. The World Health Organisation recognizes an oncocytic variant of PDTC initially not included in the Turin classification, those with mitochondrio rich cells, these tumours display poorer overall and tumour specific survival and as such these patients may not respond to radioactive iodine in the same way and may be a cohort in whom novel adjuvant treatments may be more effective.

Even this issue however is contentious, as other retrospective studies into oncocytic variants of PDTC found no difference in overall and tumour specific survival on univariate analysis.

Histopathology
With overlapping features on cytology resulting in a risk of misdiagnosing patients with PDTC it is important to...
maintain a high index of suspicion when interpreting histopathological findings. Upper aerodigestive tract invasion is more prevalent in PDTC than DTC. Therefore the presence of this should alert the clinician to the aggressive nature of the disease. PDTC is also considered an intermediate in de-differentiation and full pathological specimens may contain elements of DTC, PDTC and ATC. The British Thyroid Association classifies PDTC as those where more than 50% of the tumour is PDTC22. With such heterogeneity found on final histopathology it is challenging to determine the most effective management and as such these patients must be managed in a setting whereby surgeons, oncologists and nuclear medicine physicians can discuss the relative benefits of multi-modality therapy.

Management
Patients with PDTC should be treated in dedicated specialist centres with expertise in the management of complex thyroid cancers. If PDTC is suspected, patients should be referred urgently so an appropriate diagnosis and management strategy can be put into place23.

Surgery
Given the inherent lack of radiosensitivity in thyroid malignancies and a varying ability to concentrate iodine in PDTC, the goal of initial treatment is to achieve adequate macroscopic clearance of disease. Comprehensive workup must then prepare the surgeon for the need to perform surgery to the trachea, larynx, major vessels and musculoskeletal structures of the neck. Although there are no internationally agreed guidelines for the management of PDTC, the argument for treatment of medullary or anaplastic thyroid cancer can be extrapolated when planning surgery as it should be presumed that the tumour will not be able to concentrate iodine ahead of surgery. The American Thyroid Association (ATA) has published guidelines for the surgical management of anaplastic thyroid cancer22. Tumours are stratified by the presence or absence of distant disease and the resectability of loco-regional disease. The aim of surgery thus should not be only to debulk but ensure macroscopic clearance, ideally with preservation of vital functioning structures. Appropriate imaging of the primary disease and distant sites of metastasis helps determine resectability. In patients with a pre-existing vocal cord palsy, every effort should be made to preserve the integrity of the contralateral recurrent laryngeal nerve. In this setting, the use of intraoperative neuromonitoring should be encouraged. If there is concern that the contralateral nerve may need to be sacrificed, the patient may require a tracheostomy in anticipation of impending airway compromise (figure 4).

Figure 3: Twin classification of PDTC on cytology.

Figure 4: Clinical photograph of patient in Figure 1 demonstrating the tumour abutting to the skin and causing marked tracheal displacement.

The British Thyroid Association (BTA) guidelines are in agreement with this and also address the fact that patients should be assessed in light of their performance status as to whether they will be able to withstand en-block resection (figure 5) and adjuvant treatments21.

Regional disease. The aim of surgery thus should not be only to debulk but ensure macroscopic clearance, ideally with preservation of vital functioning structures. Appropriate imaging of the primary disease and distant sites of metastasis helps determine resectability. In patients with a pre-existing vocal cord palsy, every effort should be made to preserve the integrity of the contralateral recurrent laryngeal nerve. In this setting, the use of intraoperative neuromonitoring should be encouraged. If there is concern that the contralateral nerve may need to be sacrificed, the patient may require a tracheostomy in anticipation of impending airway compromise (figure 4). The British Thyroid Association (BTA) guidelines are in agreement with this and also address the fact that patients should be assessed in light of their performance status as to whether they will be able to withstand en-block resection (figure 5) and adjuvant treatments21.

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Adjuvant radioactive iodine ablation (RIA)
Thyroglobulin (Tg) is a specific product of thyroid follicular cells and has been used widely to predict residual thyroid tissue following surgical management of differentiated thyroid carcinomas (DTC) during their follow-up. It has also been used to predict the relative benefit of RIA. In those treated surgically for DTC with a low post-operative Tg there is a very low risk of recurrence. Undetectable Tg both post-operatively and post-RAI ablation is associated with increased recurrence-free survival. However this is not absolute, most likely due to varying proportions of de-differentiated components within tumours. There remains limited data available to determine which patients with PDTC will benefit from radioactive iodine ablation21. The American Thyroid Association guidelines do not recommend the use of radioiodine for ablation of remnant thyroid tissue in high-risk thyroid carcinomas, however there is strong evidence that the use of radioactive iodine as an adjuvant therapy, following macroscopic clearance, in high risk differentiated thyroid carcinoma improves overall and disease-specific mortality24. Our institutional approach is to treat all those with PDTC using a single treatment dose of radioactive iodine and following that to monitor serum Tg. Response to the treatment or a rising Tg would then prompt further ongoing doses in an attempt to control both loco-regional and distant disease. Volante et al support this approach in a report of their study of 183 patients with PDTC in which they found that those treated with radioactive iodine shortly after surgery had a greater uptake in comparison to those treated at the time of local recurrence or distant spread24. As up to 85% of patients with PDTC display radio-avidity, radioactive iodine should be attempted in all those medically suitable for the treatment21. A stepwise de-differentiation from well differentiated to undifferentiated disease in thyroid carcinomas has been proposed as evidence for the intermediate level of aggression demonstrated in PDTC24. This process is
associated with loss of thyroid specific functions such as TSH receptor signalling and iodine uptake and may render the tumour resistant to conventional adjuvant therapies.

External Beam Radiotherapy

The evidence for external beam radiotherapy is retrospective and contradictory2. EBRT as a single modality is unlikely to be of benefit. As an adjuvant tool however, it is often used to reduce the risk of local recurrence. Those with unresetable residual disease may benefit from treatment to disease control due to relative radio-resistance our practise is to reset gross, accessible recurrent disease in patients deemed medically suitable for a general anaesthesia. Therefore those in who further resection is not possible and who have failed to respond to radioactive iodine ablation, EBRT can be trialed on an individualized basis following discussion in a specialized MDT (figure 6).

Chemotherapy

Chemotherapeutic agents are yet to proven as effective agents in the control of advanced thyroid malignancies. Anthracyclines have been used for many years for radioiodine refractory thyroid carcinomas with unsatisfactory results23. New molecular pathways and targets are being investigated to improve the pathological understanding of treatment outcomes for those with in de-differentiated thyroid malignancies26. Epidermal growth factor receptor (EGFR) is a member of the ErbB receptor tyrosine kinase (TK) superfamily27. EGFR signaling may be inhibited by monoclonal antibodies or TK inhibitors. Both chemotherapeutic agents have proven useful for differentiate between PDTC and ATC. Clinical studies into this area have shown that TK inhibitors have been shown to have minimally toxic whilst maintaining progression free survival at 11 months following onset of treatment for a patient with macroscopic and metastatic disease. FDG-PET avidity, low Tg and negative radioactive iodine uptake, medically unsuitable for surgery28.

As such prior to commencement of chemotherapeutic agents mutational analysis may prove prognostically beneficial on diagnosis of PDTC. At the time of writing TK inhibitors are considered the first line treatment for radioiodine refractory thyroid tumours29. A systematic review of sorafenib use in 159 patients revealed a partial response in 21% of patients although no differentiation between DTC and ATC. All were treated with curative intent with a median follow-up at 59 months, was required a median sternotomy for access and 22 underwent radioactive iodine ablation. The overall survival rate was 69.7%.

Follow up

Due to the propensity of PDTC to recur and metastasize we recommend lifelong follow up in a specialized MDT setting. PDTC has been shown to be associated with loss of thyroid specific functions such as TSH receptor expression and iodine uptake, medically unsuitable for surgery. As such prior to commencement of chemotherapeutic agents mutational analysis may prove prognostically beneficial on diagnosis of PDTC. At the time of writing TK inhibitors are considered the first line treatment for radioiodine refractory thyroid tumours. A systematic review of sorafenib use in 159 patients revealed a partial response in 21% of patients although no differentiation between DTC and ATC. All were treated with curative intent with a median follow-up at 59 months, was required a median sternotomy for access and 22 underwent radioactive iodine ablation. The overall survival rate was 69.7%.

Conclusions

A thorough diagnostic work-up consists of cytopathological analysis by a specialist head and neck or endocrine pathologist, imaging including three-dimensional and functional, a high index of suspicion when faced with local invasion. Treatment should consist of aggressive attempts to gain loco-regional control, providing access to radioactive iodine and further therapy dependent on findings in the operating theatre, biochemical assessment, imaging, molecular profiles patient co-morbidities and response to initial treatment.

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Abstract

Minor salivary gland carcinomas of the head and neck are a group of rare tumours with a diverse pathology. They can affect a number of locations in the upper aerodigestive tract. The most common pathologies are Adenoid Cystic Carcinoma and Mucoepidermoid carcinoma. Adenoid Cystic Carcinoma is unique because of the propensity for peri-nervous spread, distant metastasis and frequent local recurrence. Treatment and outcome for patients with these tumours is variable and in general is affected by tumour site, histological type and grade. In this article, we review the published literature describing incidence, the main histological types, investigations, clinical outcomes and evidence base for treatment strategies.

Key words
Minor Salivary Cancer, Outcome, Molecular Oncology

Introduction

Minor salivary gland carcinomas of the head and neck are a group of rare tumours with a diverse pathology. There is significant heterogeneity in patients with these tumours with respect to histology and ultimate clinical behavior. Our understanding and treatment of these tumours has traditionally been guided by retrospective studies, as multicenter and randomized controlled trials are difficult to carry out due to this diversity. With an increased understanding of the molecular biology of salivary malignancy the diagnosis of these tumours is now more accurate and has led to the identification of oncogenic alterations which may be targeted with novel therapies. In this review, we present an overview of the current literature of minor salivary gland cancers.

Incidence

There are an estimated 450-1,000 minor salivary glands within the human body, in addition to the major salivary glands, which are the parotid, submandibular and sublingual glands. They are distributed widely in the head and neck and upper aerodigestive tract. The majority are located in the oral cavity and oropharynx accounting for 70-90%. Smaller numbers are found in the nasal cavity, middle ear, nasopharynx, laryngopharynx, hypopharynx and trachea. They contribute 8-10% of both stimulated and unstimulated saliva production. When compared to the salivary tissue of the major glands, the minor salivary glands have a reduced volume, an abbreviated ductal system and lack a consistent glandular capsule.

Management of the minor salivary gland tumours in the head and neck

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The most common histological types reported in retrospective studies varies depending on the population, geographic location and reporting center's expertise\(^2\). However, the two most common histological types are Adenoid Cystic Carcinoma and Mucoepidermoid carcinoma\(^1\). Adenoid cystic carcinoma account for 32–69% and Mucoepidermoid carcinoma accounts for 15–35% of tumours\(^10\). Adenocarcinoma not otherwise specified, Acinic cell carcinoma, myoepithelial carcinoma and carcinoma ex-pleomorphic adenoma are less common. However, all the histological tumour types have been reported to occur in the minor salivary glands\(^5\).

### Histopathology- features and behavior

**Adenoid cystic carcinoma (ACC)**

Adenoid cystic carcinoma develops from the progenitor cells located in the intercalated ducts which develop into epithelial duct and myoepithelial cells. It has variable morphologic appearances, including tubular, cribriform and solid patterns\(^6\). When occurring in minor salivary glands, ACC is most frequently seen in the palate and is the most common malignant tumour type at this location. The next most frequent locations are the tongue, buccal mucosa, lip and floor of mouth. ACC occurs at other sites such as the larynx\(^1\) and in the nasal cavity and sinuses. This tumour affects all ages but is more commonly seen in middle aged and older people. It appears to affect men and women equally\(^1\).

ACC has a number of unique features. It has a high propensity for peri-neural invasion, even in small and early-stage tumours. Tumour invades and can spread along large nerves and skin lesions are common. ACC tumours are most commonly graded according to Szanto et al\(^11\) as cribriform or tubular (grade I), less than 30% solid (grade II), or greater than 30% solid (grade III). ACC with a more solid appearance has a worse prognosis and increased risk for distant spread\(^11\). The solid appearance has been linked to a larger number of epithelial duct cells compared to the myoepithelial cells. Identifying epithelial-dominant and myoepithelial-dominant variations may allow for a functional classification and facilitate new therapeutic agents and prognostic prediction\(^12\). The molecular understanding of these tumours has recently been reported in a large study looking at whole exome sequencing of 60 ACC tumours\(^13\). This highlighted the importance of mutations in SMARCA2, CREBBP, and KDM6A suggesting aberrant epigenetic regulation in ACC oncogenesis. MYB-NFIB translocations and somatic mutations in MYB- associated genes were also seen and support previous reports showing the importance of MYB in ACC\(^14\).

The clinical behavior of these tumours is also unique. They have a low risk of metastases to neck lymph nodes, but pulmonary metastases are seen more commonly with up to 30% of patients developing distant recurrence\(^15\). Pulmonary metastases are typically multiple small volume nodules. Local recurrence is also common and ACC is typically chemotheraphy resistant in most cases.

Other methods, such as nomograms, have been described to help in understanding the prognosis of patients with these tumours\(^16\).

**Mucoepidermoid carcinoma**

Mucoepidermoid carcinoma is characterized by epidermoid (squamous like) cells, mucus producing cells and cells of intermediate type\(^17\). These tumours are often multi-lobulated with solid components. They are the most frequent malignant tumour arising from intraoral minor salivary glands accounting for 36–59% of all malignant intraoral salivary gland tumours\(^18\). Palatal tumours can invade up into the skull base. Lip lesions tend to spread into submental nodes initially and intraoral tumours metastasize to submandibular, post auricular and upper neck lymph nodes. Distant metastases do occur and involve lung, liver, bone, and brain. There are a number of systems for grading MEC tumours with one using a points system to classify tumours into low, intermediate and high grade\(^19\).

Mucoepidermoid carcinoma (MEC) has a propensity for peri-neural invasion, presence of necrosis, 4 or more mitoses and anaplasia are the histological features used to score these tumours. High grade MEC tumours in the minor glands and parotid glands have a poor prognosis due to distant metastases. Patients with MEC tumours in the submandibular gland tend to act more aggressively independently of grade\(^20\). MEC have been reported to possess a translocation between the long arm of chromosome 11 and the short arm of chromosome 19 (11p11.9;2q13), an abnormality that is also shared by acute leukemias\(^21\).

**Polymorphous Adenocarcinoma (PAC)**

This entity was previous called Polymorphous Low Grade Adenocarcinoma (PLGA) but has been renamed in the most recent addition of the WHO classification of Head and Neck Tumours\(^2\). They have an infiltrative pattern and may show tubular, fascicular, cribriform, papilliferous or solid architecture. They classically demonstrate neurotropism and often show a targetoid appearance\(^22\). The histological appearance can be similar to ACC and it is important to distinguish these from ACC because they have very different outcome and prognosis. PAC almost exclusively occur in the lower lip and intraoral sites with the palate being the most frequent. They predominate in the 5th decade and show a female predilection of approximately 2:1. PAC occur with local recurrences rates of 10–33% and regional metastases of 9–15%. Distant metastases and death from disease are exceptionally rare\(^23\).

**Carcinoma ex pleomorphic adenoma (Ca-ex-PA)**

Carcinoma ex pleomorphic adenoma (Ca-ex-PA) is defined as a pleomorphic adenoma from which an epithelial malignancy is derived\(^24\). Ca-ex-PA usually presents in the 6th or 7th decades, approximately one decade later than the appearance of the pleomorphic adenoma. Histologically, there is usually a benign and malignant component to these tumours. The malignant component is frequently a poorly differentiated adenocarcinoma or an undifferentiated carcinoma but any form of carcinoma may be found\(^25\). Ca-ex-PAs can be classified into noninvasive, minimally invasive (≤1.5 mm extension from capsule), and invasive (≥1.5 mm of invasion from the tumour capsule into adjacent tissues)\(^26\). Non-invasive and minimally invasive tumour tend to have a good prognosis when adequately treated but invasive tumours have worse outcomes\(^27\).

**Acinic cell carcinoma**

Acinic cell carcinoma demonstrate serous acinar cell differentiation and are characterized by cytoplasmic zymogen and secretory granules. Salivary acinar cells are also seen\(^1\). These tumours are thought to arise from neoplastic transformation of the intercalated duct cells with differentiation towards serous acinar cells\(^2\). Acinic cell carcinomas have more aggressive behavior when they affect the submandibular gland but in minor salivary glands their behavior is less aggressive\(^28\). Acinic cell carcinomas do have the potential to metastasize to cervical lymph nodes and subsequently to more distant sites, most commonly the lung. In the largest molecular analysis of acinic cell tumours 21 of the 25 (84%) showed loss of heterozygosity in at least one of the 20 bcr tested on chromosomes\(^29\) and 1723. A correlation was seen with the histological grade. However, this is yet to translate into a clinically useful tool. Histological grading is inconsistent and the clinical stage is a better predictor of outcome. Other prognostic factors have been investigated, including frequent mitoses, focal necrosis, perineural invasion, pleomorphism, infiltration, and stromal hyalinization. The cell proliferation marker Ki-67 has shown promise as a predictor; when the percentage of Ki-67 positivity is below 5% the rate of recurrence was zero but when the Ki-67 positivity is above 10% there was a higher recurrence rate\(^30\).

### Tumour locations and etiology

In a study using SEER data, including 5,334 patients, the most common subsite of minor salivary gland malignancy was the oral cavity (58.7%) followed by the pharynx (21.2%), nasal cavity/ paranasal sinuses (15.8%) and larynx (4.3%)\(^31\). Within the oral cavity, the most common site is the palate, with buccal mucosa, retromolar trigone and the upper lip the next most common subsites, accounting for more than 75% of tumours\(^3\). Outcomes have traditionally been thought to be worse for the ‘hidden sites’, such as the larynx, nasopharynx and nose\(^32\). This was also reported in the recent SEER analysis reporting 5-year cancer specific survival in the larynx (HR, 2.42; 95% CI, 1.67-3.50) and nasal cavity/ paranasal sinuses (HR, 1.73; 95% CI, 1.29-2.32) confirming the importance of the anatomical sites on prognosis.

Tumors of minor salivary gland origin are more common in women (1.2:1 to 1.9:1) and some series suggest that there is a higher incidence in blacks than whites and in the South African population\(^33\) and in Ugandan\(^34\). Previous irradiation, and certain occupations with exposure to rubber manufacturing have been implicated in the etiology of salivary gland cancers but there is no evidence addressing these points specifically in minor salivary gland malignancies\(^35\).
The American Joint Committee for Cancer (AJCC) and the Union for International Cancer Control (UICC) do not have a TNM staging system for minor salivary gland tumours. However, when they are diagnosed as malignant the TNM system for the tumour location is used as in squamous cell carcinoma of the head and neck. Spiro et al. successively operated the criteria used for non-surgical salivary gland carcinoma of the oral cavity, pharynx, larynx, and sinuses of MEC carcinoma.

Minor salivary gland tumours often have dual or biphasic histology, with more than one histological pattern within the same tumour. There is also controversy in grading of salivary gland malignancy, particularly with minor salivary glands. Further complexity is caused by the high inter-observer variability in reporting these rare tumours and the reclassification of a large number of the histological subtypes.

Prognostic factors

The clinical tumour stage of minor salivary gland carcinomas has been shown on multivariate analysis to be the most important prognostic factor for outcome and recurrence. Tumor histology and grade of malignancy are also important predictors for survival. These are all independent variables but clinical tumour stage seems to be more important than grading. In ACC, the additional independent prognostic factors associated with poor outcome are peri-neural invasion, positive margins and solid histological variant.

Surgery

Management of patients with minor salivary gland carcinoma requires the involvement of a multi-disciplinary team with accurate radiographic imaging and histological diagnosis. Surgical resection with adjuvant post-operative radiation therapy, depending on risk factors, is the main treatment strategy. The treatment and the surgical approach needs to be individualized to the patient encompassing the tumours size and extension, likelihood of any peri-neural spread and the anatomical site. A wide resection with clear margins is deemed the optimal surgical treatment.

Adjuvant treatment

Post-operative radiotherapy for minor salivary gland malignant disease is indicated for high grade tumours and tumours with advanced stage. Postoperative radiotherapy has been reported to be safe to be omitted if a good clear margin resection is performed in early-stage disease without adverse prognostic factors, such as lymphovascular or peri-neural invasion.

Indications for postoperative radiotherapy include positive or close surgical margins, a high-grade malignant tumour and the presence of lymphovascular invasion, Eustachian tube dysfunction or airway impingement.

Presentation

The presenting features of patients with minor salivary gland malignancies vary depending on the underlying primary site and the extent of the tumour. The majority of tumours are found in the oral cavity and oropharynx. These typically present as a painless lump with a mass in the submucosa. The mucosal layer is often adherent to the mass and a small ulcer may develop. The minor salivary glands have a limited capsule and this should be understood prior to resection. Tumours in the nasopharynx, nasal cavity and larynx may present as incidental findings on imaging or from mass effects leading to nasal obstruction, Eustachian tube dysfunction or airway impingement. There is a high incidence of peri-neural invasion in some minor salivary tumours and patients with paresthesia or pain require further investigation with MRI. Initial presentation with a neck mass can occur in up to 15% of patients.

Investigations

The most important aim in the investigation of patients with minor salivary gland neoplasms is to determine if the lesion is benign or malignant and what the likely histological subtype is. However, the final histological diagnosis may not be available until after the formal excision biopsy. A core needle biopsy should allow a preliminary diagnosis on which the extent of resection can be determined. An excisional biopsy makes it more important than grading. In ACC, the additional independent variables but clinical tumour stage seems to be more important than grading. In ACC, the clinical tumour stage of minor salivary gland carcinomas has been shown on multivariate analysis to be the most important prognostic factor for outcome and recurrence. Tumor histology and grade of malignancy are also important predictors for survival. These are all independent variables but clinical tumour stage seems to be more important than grading.

Targeted agents have been investigated in salivary malignancy but not applied to minor salivary glands.

Outcomes

Outcome of minor salivary gland malignancies has recently been reported in a retrospective population-based study using the National Cancer Institute's Surveillance, Epidemiology, and End Results (SEER) data. Patients with a primary tumour of the oral cavity had the highest 5- and 10-year unadjusted Cancer Specific Survival (CSS) with rates of 90.1% and 84.7%, respectively. A primary site in the larynx had the lowest 5- and 10-year unadjusted CSS rates of 52.6% and 45.3%, respectively. A summary of previously published disease specific survival (DSS) outcomes is shown in Table 3. At 5 years, the DSS figures are similar but by 10 years there are some differences and this is a reflection on the proportion of adenoid cystic carcinomas in the cohorts as they have late local and distant failures.

Outcome by histology

MEC had the highest unadjusted 5-year CSS (90.8%) compared with patients with ACC (79.1%), adenocarcinoma (80.8%), and other rare carcinomas (70.1%). Ten-year CSS patterns were similar to those of 5-year CSS for patients with ACC (68.8%), adenocarcinoma (75.5%), and other rare carcinomas (62.1%); however, the 10-year CSS for patients with ACC was much lower (62.4%).

Conclusion

Minor salivary gland cancers are rare and comprise a diverse group of cancers. Treatment needs to be tailored to the individual patient and is dependent of the tumour's site and extension, histology and grade. Post-operative radiotherapy is indicated in most cases apart from low clinical stage, low grade malignancies that have received surgery with clear margins. The clinical progression of these cancers is dependent on the histological type, with adenoid cystic carcinoma recurring locally, showing peri-neural spread and having late metastatic spread, usually to the lungs.

Due to the rarity and diverse nature of this disease the literature is based mainly on retrospective case series. With advancing knowledge of the molecular mechanisms underlying some of these cancers targeted therapy may be a useful option in the future.


Pharyngeal pouch and cricopharyngeal spasm: current indications and treatment modalities

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Abstract
Pharyngeal pouch refers to a false pulsion diverticulum of the hypopharynx. Cricopharyngeal spasm is associated with pharyngeal pouch formation, although can present independently. Cricopharyngeal spasm can be treated with dilatation, botulinum toxin injection or cricopharyngeal myotomy. Botulinum toxin injection for pharyngeal pouch is probably better reserved for patients with adverse surgical risk.

Pharyngeal pouches can be treated with transcervical cricopharyngeal myotomy. The transcervical approach, offers an established and dependable option in experienced hands. However, adverse surgical risks may mean endoscopic approaches, especially flexible endotherapy are more appropriate.

The recent general movement towards endoscopic approaches is based on surgical simplicity, seemingly equal efficacy and potentially lower morbidity. However, the predominantly retrospective evidence-base and total absence of suitably powered trials shows little difference between the most commonly employed techniques in clinical practice. Given this fact, the surgeon’s experience and patient’s opinion are the most important factors in interventional decision-making.


Key words
Zenker Diverticulum; Pharyngeal Muscles; Disease Management

Introduction
Pharyngeal pouch (also termed Zenker’s diverticulum) refers to a false pulsion diverticulum of the hypopharynx passing most commonly through Killian’s dehiscence.1 The UK incidence of pharyngeal pouch is two in 100,000 per annum, being most common in the eighth decade.1 The aetiology of pharyngeal pouch formation remains uncertain, although most theories centre on cricopharyngeal dysfunction, including spasm.2,3 Cricopharyngeal spasm (also termed cricopharyngeal achalasia) is associated with uncomplicated gobus, aphagia, aspiration and pharyngeal pouch formation.1 Some evidence links gastro-oesophageal reflux disease, cricopharyngeal spasm and pharyngeal pouch formation with cricopharyngeal inflammation and fibrosis.4 The classic presenting features of a pharyngeal pouch include oropharyngeal dysphagia, chronic cough, regurgitation and halitosis, complicated by aspiration.1 Cricopharyngeal pouch carcinoma is rare, but requires consideration if the pouch is not fully excised.5 Functional endoscopic evaluation of swallowing may suggest a pharyngeal pouch, although a videofluoroscopic swallow study is the diagnostic gold standard.6 A cricopharyngeal bar – a radiological sign characterised by posterior indentation at the pharyngoesophageal junction – is invariably found in pharyngeal pouch, but frequently found incidentally in asymptomatic individuals.7 In this review, we will outline the current indications and treatment modalities for cricopharyngeal spasm and pharyngeal pouch.

Cricopharyngeal spasm
Cricopharyngeal spasm can be treated with dilatation, botulinum toxin injection or cricopharyngeal myotomy.8 Indications for treatment are not well established. To confirm the diagnosis, most patients undergo a videofluoroscopic swallow study. The diagnostic use of manometry or manofluoroscopy is occasionally utilised despite controversy.9

Cricopharyngeal myotomy
A transcervical (open or external) cricopharyngeal myotomy was the traditional treatment for cricopharyngeal spasm. The use of endoscopic carbon dioxide laser was subsequently popularised to decrease morbidity.9 Classic transcervical cricopharyngeal myotomy involves intubation of the cervical oesophagus to provide internal distention. A left-sided skin incision is made, followed by dissection down to the cricopharyngeus. The incision is traditionally left sided as the recurrent laryngeal nerve is more medial on this side running truer to the tracheoesophageal groove, and hence less prone to damage. The omohyoid muscle and middle thyroid vein (if present) are divided, and then myotomy is performed. There are variable recommendations regarding the length of the myotomy and any additional elements of the procedure. In practice the myotomy is performed to include distal thyropharyngeus and proximal oesophageal musculature down to mcosa. This optimises the likelihood of a total myotomy as the boundaries of the cricopharyngeus are not always clear-cut. In many centres a drain is left in situ. Oral feeding may be commenced the same night following a period of observation to minimise the likelihood of mucosal perforation, which when small may not be immediately evident.

Botulinum toxin injections
Botulinum toxin injection can be used diagnostically to establish benefit from myotomy. Administration is technically challenging and repeat injection is required. Endoscopic injection under general anaesthesia is the standard approach. Percutaneous injection is possible under local anaesthesia with electromyographic guidance. Botulinum doses range from 10 to 100 units.10

Dilatation
Dilatation can take the form of blind or endoscopically assisted bougine utilising wire-guided polyvinyl, air-filled, pneumatic or water-filled balloon dilatation techniques.11

Indications, patient selection and comparative outcomes
Ashman et al.12 undertook a systematic review of the management of isolated cricopharyngeal dysfunction. They concluded from review of case series that botulinum toxin A injection and dilatation may be used first-line in elderly patients, and those with significant medical co-morbidities, for diagnostic purposes. Cricopharyngeal myotomy is more effective for refractory or recurrent disease. Endoscopic approaches to cricopharyngeal myotomy may offer lower morbidity, although they found a lack of evidence to favour a single technique.

A systematic review by Kockor et al.13 concluded that significantly higher success rates were seen with myotomy compared to botulinum toxin injection, but not dilatation. Complication rates were not significantly higher between all three interventions. Patient selection appeared to be the key. Higher success seemed to be related to factors including idiopathic or non-oncological iatrogenic aetioloogy; absence of the subatmospheric intrasphincteric pressure drop; favourable deglutition, phonation, tongue and laryngeal function; and the invasiveness of the procedure. However, studies included were retrospective or non-randomised and used small sample size and methodological issues.

A systematic review of the effectiveness of endoscopic cricopharyngeal myotomy in adults with neurological disease was unable to determine its effectiveness due to identifying only two studies which had significant methodological concerns.14 A single randomised prospective pilot study comparing balloon dilatation and laser myotomy exists. Azena et al.15 conducted this study on eight treatment-naive dysphagic patients with cricopharyngeal dysfunction without pharyngeal pouch. No difference was found in upper oesophageal sphincter sagittal diameter increment or six-month patient-reported outcome measure between treatment groups.

Pharyngeal pouch
The transcervical technique is the traditional method for pharyngeal pouch treatment. There has been movement towards endoscopic techniques based on the principle of seemingly equal efficacy and lower morbidity of endotherapy.16 Table 1 compares the outcomes from these two approaches using data from a recent systematic review.17 All patients undergoing surgery require a rigid or flexible upper oesophageal endoscopy and exclude carcinoma.18 In line with international guidelines, all geriatric surgical patients should have a comprehensive preoperative assessment including cognition, capacity and functional status.19 Co-morbidity should be evaluated and optimised and polypharmacy reviewed.20 Preoperative nutritional assessment and intervention is essential as many patients are malnourished.21 Patients should be given the best chance to be optimised preoperatively, as age alone should not be a contraindication to surgery. Many patients may benefit from preoperative (and postoperative) gastrostomy review, although this type of service is not yet widely widespread. Transcervical procedures involve longer periods of hospitalisation, but this is not the case when mucosal sparing techniques are employed.

Terminology
Diverticulotomy is excision of the pouch with oversewing. Diverticulotomy refers to division of the common septum between the pouch and the oesophagus. Diverticulopexy
Persistent symptoms, elevated complications and challenging than endoscopic procedures. Diverticulectomy required makes the transcervical approach more technically which can be stapled or hand-sewn. The neck dissection always

Table 1: Comparison of outcomes between endoscopic techniques and transcervical procedures for pharyngeal pouch surgery.

<table>
<thead>
<tr>
<th>Technique</th>
<th>Endoscopic procedures</th>
<th>Transcervical procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Failure rate</td>
<td>18.4%</td>
<td>4.2%</td>
</tr>
<tr>
<td>Short-term failure rate</td>
<td>14.5%</td>
<td>1.3%</td>
</tr>
<tr>
<td>Access difficulty</td>
<td>5.2%</td>
<td>0%</td>
</tr>
<tr>
<td>Complications</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type of complications</td>
<td>7%</td>
<td>Mediastinitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>11%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Fistula</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Recurrent laryngeal nerve</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Haematoma</td>
</tr>
<tr>
<td>Mean length of stay</td>
<td>8.9 days</td>
<td>8.4 days</td>
</tr>
<tr>
<td>Technique</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stapling</td>
<td>Pouch excision</td>
<td></td>
</tr>
<tr>
<td>Laser</td>
<td>Inversion</td>
<td></td>
</tr>
<tr>
<td>Coagulation</td>
<td>Suspension</td>
<td></td>
</tr>
<tr>
<td>Open neck dissection</td>
<td>Not required</td>
<td>Required</td>
</tr>
</tbody>
</table>

refers to inversion and suturing of the diverticulum in the retropharyngeal space to the anterior longitudinal ligament of the cervical spine. Diverticulum invagination refers to inversion into the oesophageal lumen with overswinging.14 Transcervical procedures

These can be performed under general anaesthetic (most commonly), local anaesthetic or C-5–C-7 selective spinal anaesthesia.15 Transcervical approaches nearly always include a cricopharyngeal myotomy (as described above) with either a diverticulectomy or a diverticulotomy, which can be stapled or hand-sewn. The neck dissection required makes the transcervical approach more technically challenging than endoscopic procedures. Diverticulectomy is most commonly performed. Cricopharyngeal myotomy or a diverticular procedure alone are associated with persistent symptoms, elevated complications and recurrence.14,17 The decision as to which additional procedures occur after cricopharyngeal myotomy is mainly based on the size of the pouch, determined from poor-quality retrospective studies: myotomy alone (4cm and symptomatic), myotomy with diverticulotomy (1-4cm) or myotomy with diverticulotomy (<4cm).16,18 Older patients with pouches ≤6cm may also be suited to myotomy with diverticulotomy to avoid division and suturing of the pharyngooesophageal structure.19

Following a cricopharyngeal myotomy with diverticulectomy, a nasogastric tube is usually placed (unless a linear cutting stapler is used) and kept in for 3-4 days.

Endoscopic techniques

Endoscopic approaches can be rigid or flexible; laser, stapled or coagulative. Trismus limiting instrumentation restricts all endoscopic approaches. Rigid endoscopic approaches are contraindicated in patients with reduced neck extension. The most common surgical technique for pharyngeal pouch performed in the UK is the rigid endoscopic staple-assisted diverticulotomy, which is often considered first-line (if feasible).16 In brief, a general anaesthesia is required and a diverticuloscope is positioned so that the anterior blade fits into the oesophagus, with the posterior blade intubating the diverticulum. An endoscopic linear cutting stapler is positioned to create a diverticulotomy between the posterior pouch and the anterior oesophagus. The creation of a common channel reduces the entrapment of food boluses.14 Carbon dioxide laser-based endoscopic diverticulotomy is an alternative to stapling. The procedure involves general anaesthesia and visualisation of the common septum using an operating microscope via a diverticuloscope. A CO2 laser, controlled by a micromanipulator, enables transmural mucosal incision and complete myotomy. This technique can be used to get around the issue of an incomplete cutting staple line and residual pouch. Pouches <3cm are generally suitable for treatment by endoscopic laser approach. Endoscopic diverticulotomy for pouches <2cm can also be performed using a harmonic scalpel.18

The National Institute of Health and Care Excellence has approved the use of flexible endoscopic treatment of pharyngeal pouches.15 This does not require neck hyperextension or general anaesthesia (it can be undertaken with sedation), making it useful for older, co-morbid patients or those with limited range of cervical motion. An endoscopic diverticulotomy and cricopharyngeal myotomy can be performed. Flexible endotherapy is generally undertaken by tertiary gastroenterology endoscopists. The failure rate is 29%.15

Butylinum toxin injections

As for cricopharyngeal spasm this may be used in symptomatic pharyngeal pouch as a sole treatment in patients with an adverse surgical risk profile.

Alternative feeding routes

In some cases of very frail elderly patients who are completely unsuitable for intervention, non-operative management is necessary and alternative feeding such as gastrostomy is required.

Indications, patient selection and comparative outcomes

Asymptomatic patients with an incidentally found pharyngeal pouch generally do not require surgery. Some argue towards intervention in younger patients who are medically suitable for surgery, as small asymptomatic pharyngeal pouches (e.g. <1.2cm) may enlarge over time. Others may elect to follow-up these patients over time. Evidence of aspiration is an absolute indication for surgery. As pharyngeal pouch tends to affect older patients, medical co-morbidity may influence the appropriateness or type of intervention.

Outcomes between transcervical techniques show diverticulotomy has a higher fistula rate and longer length of stay versus diverticuloplasty or diverticular invagination. Diverticuloplasty appears to offer the most favourable outcome profile with a mean rate of failure (2%), fistula (1.5%), recurrent laryngeal nerve (RLN) palsy (2.5%), haematoma (2.5%) and length of stay (5.5 days).11

Outcomes between endoscopic techniques show stapling to be superior, despite more issues with exposure (6.2%) and operative failure (19%) versus coagulation. This is mainly due to its favourable complication profile, including low mean incidence of mediastinitis (0.2%) and emphysema (0.8%), and mean length of stay of 3.2 days.11

The comparison of outcomes between staple-assisted endoscopic and transcervical diverticulotomy approaches is roughly equivocal. Verdonck et al concluded that transcervical approaches are preferred for surgically fit patients who desire a more definitive outcome. Open approaches may also be required where anatomical factors limit endotherapy: a transcervical diverticulotomy or diverticular invagination can be offered to anyone suitable for general anaesthetic. Patients with adverse general anaesthetic risk can be offered flexible endotherapy under sedation.

Our experience

The evidence-base is largely case-series derived and this fails to show demonstrable superiority of any technique. The surgeon’s experience and shared-decision making with the patient is therefore extremely important.

In the senior author’s experience, from a case series of 120 pharyngeal pouch procedures,22 cricopharyngeal myotomy is the single most important intervention wherever possible. A complete cricopharyngeal myotomy can only be reliably performed using a transcervical approach, which enables the surgeon to directly visualise the cricopharyngeus. In addition, an open approach enables direct visualisation of the pharyngeal and oesophageal mucosa. If mucosal integrity is maintained then fistula formation and mediastinitis cannot occur. The ability to accurately assess the maintenance of mucosal integrity with endoscopic techniques is not always possible, and as a result, rare cases of representation with undiagnosed mediastinitis do occur.23 Pouch size does not preclude this, nor is it an accurately measurable entity. In our experience, pouch excision is only necessary when the pouch is adherent in the mediastinum. A large pouch will still respond to a total cricopharyngeal myotomy without opening the mucosa. The key is to rescue the patient after the procedure, to ensure the myotomy is complete and the cricopharyngeal bar has been completely removed.

In our experience, endoscopic stapling outcomes are inferior to open approaches in the longer-term, demonstrated by both higher recurrence (26% vs 7.5%) and multiple procedure (32% vs 0%) rates.15

Conclusion

A variety of transcervical and endoscopic techniques have been described for the management of cricopharyngeal spasm and pharyngeal pouch, reflecting attempts over the years to both innovate and improve poor outcomes. For cricopharyngeal spasm intervention, the senior author’s experience favours cricopharyngeal myotomy over dilatation. Botulinum toxin injection for cricopharyngeal spasm is probably better reserved for patients with adverse surgical risk.

For pharyngeal pouch intervention, the open approach, especially mucosal-sparing techniques, offer an established and dependable option in experienced hands. However, the adverse surgical risk may mean endoscopic approaches, especially flexible endotherapy are more appropriate.

The predominantly retrospective evidence-base and total absence of suitably powered trials shows little difference between the most commonly employed techniques in clinical practice. Given this fact, the surgeon’s experience and patients’ opinion are the most important factors in interventional decision-making.

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None
References


Giant parathyroid adenomas in the neck: A minimally-invasive approach to 17 consecutive cases

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Abstract
Introduction: We present a case series of 17 giant adenomas and discuss the challenges of a minimally invasive surgical approach. The entity of “giant parathyroid adenomas” is defined as those with a pathological weight > 3.5 g. They are reported to pose a more challenging procedure.

Methods: A case note review of 17 giant adenomas operated on at a single institution by the senior author was performed (2006-2017). There were 8 males and 9 females, with a median age of 62.1 years. Data was collected on patient demographics, symptoms, biochemical, ultrasound and sestamibi results, operation outcomes, complications and histopathology.

Results: All giant adenomas were treated successfully with a single minimally invasive operation. The location of the glands in the pre-operative imaging was 100% concordant with the intra-operative location. The weight of the adenomas ranged from 3.5 g – 20 g with a mean of 6.36 g. Histology confirmed all the glands to be benign.

Discussion: Our case series of 17 demonstrates that adenomatous glands up to 20 g in weight and 60 mm in the largest dimension can be excised using a lateral skin incision via a minimal invasive open approach.

Key words
Giant Parathyroid adenoma, Minimally invasive surgery

Introduction
Sporadic primary hyperparathyroidism, characterised by hypercalcaemia and raised parathyroid hormone levels, is due to a solitary parathyroid adenoma in 80%-85% of cases. Patients typically present with symptoms including fatigue, exhaustion, weakness, polydipsia, polyuria, nocturia, joint pain, bone pain, constipation, depression, anorexia, nausea, heartburn, and associated conditions, including nephro lithiasis, and haematuria. Typically, normal parathyroid glands weigh 38 - 59 mg.

The American Association of Endocrine Surgeons parathyroidectomy guidelines are listed in table 1. There is a general agreement that this results in an improvement in bone density, reduction in fractures and the frequency of nephrolithiasis. Increasingly, the approach taken for single gland adenomas is that of a minimal invasive approach with pre-operative dual modality imaging to accurately localise the adenomatous gland.

The entity of “giant adenomas” is defined as those with a pathological weight greater than 3.5 g. They are thought to be a different genetic entity themselves and therefore behave differently to ‘normal’ sized adenomas.

Table 1: Indications for parathyroid surgery

<table>
<thead>
<tr>
<th>All patients with symptomatic pHPT</th>
<th>Serum calcium level is greater than 1 mg/dL above normal</th>
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<tbody>
<tr>
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<td>Evidence of renal involvement, including silent nephro lithiasis on renal imaging, nephrocalcinosis, hypercalcaemia</td>
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<td>Patients with pHPT and osteoporosis, fragility fractures, or evidence of vertebral compression fractures or spinal imaging</td>
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<tr>
<td>When pHPT is diagnosed at 50 years or younger regardless of whether objective or subjective features are present or absent</td>
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<td>Clinical or biochemical evidence is consistent with PCA</td>
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<td>Patients who are unable or unwilling to comply with observation protocols</td>
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<td>Patients with neurocognitive and/or neuropsychiatric symptoms that are attributable to pHPT</td>
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<tr>
<td>Patients with cardiovascular disease who might benefit from mitigation of potential cardiovascular sequelae other than hypertension</td>
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Spanheimer et al noted giant adenomas have a greater pre-operative calcium and PTH levels and hence have an increased risk of symptomatic postoperative hypercalcaemia. However in our series of 16 and individual case reports no significant differences in calcium and PTH levels were noted. The first documented parathyroid adenoma was in 1936 by Snell and this weighed 101 g. No Age Sex Duration from US to surgery (days) Durations of operation (mins) Adenoma size on USS (mm) Calcium Pre op PTH Pre op Calcium Post op PTH post op Adenoma Weight (g) Adenoma Size (mm)

| No | Age | Sex | Duration from US to surgery (days) | Durations of operation (mins) | Adenoma size on USS (mm) | Calcium Pre op | PTH Pre op | Calcium Post op | PTH post op | Adenoma Weight (g) | Adenoma Size (mm) |
|----|-----|-----|------------------------------------|-------------------------------|--------------------------|-----------------------|-------------|-------------|-----------------|-------------|-------------------|------------------|
| 1  | 60  | F   | 127                                | 43                            | 27                       | 3.1                    | 43.5        | 2.59         | 9.7             | 5.4         | 32                |                  |
| 2  | 57  | M   | 170                                | 47                            | 47                       | 2.96                   | 50.8        | 2.41         | 10.4            | 40          |                   |                  |
| 3  | 64  | F   | 84                                 | 79                            | 14                       | 3.2                    | 56.1        | 3.2          | 7.4             | 4          | 28                |                  |
| 4  | 63  | M   | 363                                | 42                            | 27                       | 3.56                   | 108.6       | 2.21         | 25.8            | 5.6         | 28                |                  |
| 5  | 65  | M   | 237                                | 81                            | 42                       | 2.91                   | 14.5        | 2.44         | 4.5             | 5.6         | 43                |                  |
| 6  | 44  | F   | 84                                 | 88                            | 27                       | 3                     | 13.3        | 2.4          | 7               | 4.2         | 25                |                  |
| 7  | 49  | M   | 200                                | 40                            | 20                       | 3.28                   | 45.9        | 2.28         | 6.6             | 4.8         | 30                |                  |
| 8  | 41  | M   | 150                                | 82                            | 60                       | 3.1                    | 50.9        | 2.4          | 4.5             | 20         | 60                |                  |
| 9  | 78  | M   | 89                                 | 59                            | 28                       | 3.6                    | 147         | 3.2          | NA              | 3.76        | 30                |                  |
| 10 | 56  | M   | 227                                | 47                            | 27                       | 3.2                    | 34.5        | 2.47         | 4.7             | 7          | 29                |                  |
| 11 | 63  | F   | 140                                | 77                            | 40                       | 3                     | 31          | 2.4          | 6.1             | 3.5         | 37                |                  |
| 12 | 68  | M   | 163                                | 110                           | 33                       | 3.77                   | 54.4        | 2.13         | 4.1             | 8.6         | 36                |                  |
| 13 | 74  | M   | 263                                | 48                            | 50                       | 3.29                   | 42.3        | 2.83         | 4.3             | NA          | 30                |                  |
| 14 | 77  | m   | 68                                 | 77                            | 12                       | 2.85                   | 11.6        | 2.48         | 9.6             | 14          |                   |                  |
| 15 | 19  | M   | 105                                | 90                            | 35                       | 2.96                   | 16.3        | 2.44         | NA              | 9          | 40                |                  |
| 16 | 80  | F   | 183                                | 80                            | 9                        | 3.1                    | 39.9        | 2.57         | 5.3             | 4.4         | 11                |                  |
| 17 | 56  | F   | 217                                | 100                           | 25                       | 2.74                   | 19.6        | 2.74         | 1.7             | 5          | 40                |                  |

Table 2: Patient results

adenomas both anatomically and functionally, respectively. They are then reviewed in the one-stop parathyroid ENT clinic, with their imaging results. The non-ectopic single gland parathyroid adenomas are booked for day-case minimally invasive parathyroidectomy. This approach is summarized in algorithm (Figure 3).

The surgical approach involves a 2-3cm laterally-placed horizontal skin incision located over the position of the gland, as determined pre-operatively with the dual modality imaging. Subplatysmal flaps are raised and using blunt dissection, the strap muscles are medialised and the sternocleidomastoid is lateralisated to expose the carotid bundle. The thyroid lobe is medially rotated and the carotid bundle is lateralised to reveal the adenomatous gland. Blunt dissection provides the advantage of a dry operating field and maintains the colour contrast between parathyroid and thyroid tissue. The parathyroid gland is then carefully dissected off the thyroid lobe and the vascular pedicle (Figure 4) is divided using bipolar diathermy. During the surgical approach, it is essential to divide the vascular pedicle (Figure 4) is divided using bipolar diathermy. The weight of the adenomas ranged from 3.5g – 20g with a mean of 6.36g. Histology confirmed all the glands to be benign. None of the patients were readmitted or had any complications such as bleeding, infection, haematoma, or recurrent laryngeal nerve injury. Follow up was at 6 weeks (range 13-75 days) and all were subsequently discharged after confirming biochemical cure.

These results are summarized in Figure 3.

Discussion

Giant parathyroid adenomas are classified as those weighing >3.5g14; these pose a challenge for surgical management via a traditional bilateral neck exploration let alone via a minimally invasive approach. Our series comprised of 17 patients with giant parathyroid adenomas out of a total 260 parathyroidectomies performed using a cervical neck incision. This proportion is consistent with other cohorts in the literature3. Spannheimer et al. found 15 giant adenomas in 300 consecutive parathyroidectomies4. Whereas Lalanne-Mustrith et al. reported on 26 giant adenomas in 225 cases5. Specific operative technique used to remove the giant adenomas in these published case series were not given.

Minimally invasive parathyroidectomy is mentioned within the American Association of Endocrine Surgeons Guidelines for management of primary hyperparathyroidism. It is also mentioned within the Consensus statement of the European Society of Endocrine Surgeons. A pan European survey suggested 59% of responders were using MIP6 but from the Scandinavian quality register only 33% were employing MIP7. But again it should be stressed these are not figures quoted for giant adenomas which are now widely accepted to represent a distinct clinical entity and as such are often managed differently8.

A literature search revealed we are the only giant adenoma series excised used a minimally invasive approach. Most reports of giant adenomas are single case reports where a bilateral neck exploration was performed. The rationale for MIP is that most patients (>85%) have a single parathyroid adenoma, which should be identifiable with the correct pre operative investigations and therefore excised with a selective cervical exploration9. We believe the key to success and our zero rate conversion to the traditional bilateral neck exploration is the pre-
operative work done by a dedicated multidisciplinary parathyroid team. This involves the pathway of diagnosis by the endocrinology team who refer appropriate patients to the parathyroid surgeon with dual modal imaging of the gland. The dedicated parathyroid radiologist who performs all ultrasounds with the patient supine and neck extended akin to the position during the operation to aide the surgeon’s approach. The same radiologist also performs and interprets all MIBI scans; the wealth of experience over the last 10 years allows a high accuracy of results.

Our pre operative radiological investigations are ultrasound and sestamibi scan. The positive predictive value of identifying an adenoma on ultrasound alone is 60-92% and sestamibi alone is 78-100% hence combining the two investigations gives a positive predictive rate which is over the last 10 years allows a high accuracy of results.

The preoperative imaging allows a focused approach through a small incision directed over the precise location for the gland and allowing minimal exploration within the neck. In addition, a dry operating field due to blunt dissection to locate the gland also aids a quicker recovery, with any drains used, removed within 24 hours. The key to the operative approach, having located the gland accurately with pre-operative dual modality imaging, is removal of the gland without rupture of it’s thin adherent capsule. Any rupture risks local seeding of parathyroid tissue into the neck and hence ongoing biochemical abnormalities in keeping with primary hyperparathyroidism despite the causative gland being removed. In our normal practice, a drain is not used for MIP and most patients have a day case procedure. In this series, the senior author felt that with the 3 largest adenomas (range 8.6 g – 20g) that a drain would be advisable due to extensive tissue dissection and these patients were kept overnight for observation and the drain removed the next morning.

A literature search of “giant” and “parathyroid” of the databases AMED,EHMA,HIMIC, MEDLINE, PsycINFO, BNI, CINAHL, Health business elite, revealed 111 results of which 48 were original articles. These were accepted. As well as producing a good cosmetic result, it also shortens the intra-operative time, length of post-operative stay and increases the likelihood of a day case procedure.

Conclusion
Our case series of 17 patients we have demonstrated that cervical adenomatous glands up to 20g in weight and 60mm in the largest dimension can be excised using a
Abstracts: ENT Masterclass Trainees’ Gold Medal 2017

The role of the novel cytokine IL-35 in the regulation of macrophage-mediated anti-tumour immunity against head and neck cancer.

Recipient of the ENT Masterclass Gold Medal 2017 for best paper
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Background
Cancer cells have the ability to induce immune tolerance, which aids in tumour survival and proliferation. Interleukin (IL)-35 is a novel immunosuppressive cytokine composed of two subunits: EBI3 and p35. Previously, we have demonstrated that head and neck squamous cell carcinoma (HNSCC) cell lines constitutively express IL-35 and this is up-regulated by the pro-inflammatory cytokines IFNγ and TNFα. The aim of this study was to investigate cytokine secretion in co-culture of macrophages/dendritic cells (DCs) with cancer cells.

Methods
Human THP-1 macrophages were co-cultured with human HNSCC cell lines (H137, FADU and C1) or their conditioned media with or without human dendritic KG-1 cells. THP-1 cells were analyzed for cytokine production by ELISA.

Results
Culture of THP-1 cells with increasing concentrations of HNSCC conditioned media increased TNFα production. Co-culture of THP-1 and KG-1 cells with HNSCC conditioned medium stimulated IFNγ production, indicating production of biologically active IL-18 in the co-culture system. THP-1 cell aggregation was observed after co-culture with HNSCC cell lines. Prolonged co-culture for 5 days resulted in a tolerogenic macrophage phenotype demonstrated by diminished TNFα production by THP-1 cells in response to Endotoxin stimulation.

Conclusions
Macrophages/DCs appear to be initially activated by head and neck cancer cells. However, head and neck cancer cells subsequently induce a tolerogenic phenotype in these cells, which may promote tumour survival and proliferation. Determination of the mechanisms by which this occurs may identify novel therapeutic targets.

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The use of a microfluidic device to investigate thyroid tissue response to treatment

Recipient of the ENT Masterclass Silver medal 2017
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Introduction
Incidence data shows increasing thyroid cancer presentation. Up to 15% of patients are refractory to radiodine ablation and for this group an alternative regimen is needed.

Microfluidic culture provides a biomimetic microenvironment enabling in vitro tissue studies under pseudo in vivo conditions. We sampled thyroid tumours in pseudo in vivo conditions. We sampled thyroid tumours in vivo. We then incubated these tumours in a microfluidic system for 9 days before processing for histology and immunohistochemistry.

Conclusion
The microfluidic system allows investigation of the effects of conditioners on tumour tissue histology.

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lateral skin crease incision via a minimal invasive approach. This is achieved by having a robust approach to pre-operative imaging to ensure optimal outcomes biochemically and in terms of patient satisfaction. Conflict of interest Authors declare that they have no conflict of interest and financial disclosure.

References
Methods: Following ethical approval and written, informed consent, thyroid tumour specimens were taken at time of surgery (n=9). These were placed into microfluidic devices within 1 h of resection. The specimens were perfused with complete Medium (2μl/min) with and without thyroid stimulating hormone. The tissue was maintained for up to 6-days with effluent being collected at 2 hourly intervals. Viability of tissue was measured both morphologically (with the architecture examined by a head and neck pathologist), and also via detection of lactate dehydrogenase within the effluent.

Results: Both follicular and papillary structure of papillary thyroid carcinoma were maintained following culture for up to 4 days. Initial LDH release was high following microfluidic set-up due to tissue manipulation but reduced to low levels for up to 4-days. The addition of lysis buffer induced sharp increases in LDH release, indicating tissue viability. No apparent differences were observed, in either morphology or LDH release, with and without TSH.

Conclusion: Using a microfluidic device it is possible to maintain viable thyroid tumour tissue for up to 4-days. This technology platform can be used for further studies investigating the effects of adjuvant treatment paving the way towards personalised treatment.

No Authors have any conflict of interest to declare and all authors are happy with the submission.

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Oral Rinse Testing for HPV in Oropharyngeal Squamous Cell Carcinoma: A Diagnostic Accuracy Study

Qureishi A (presenting author), Malik M, Mawby T, Fraser L, Moller H, Winter S
Work conducted in: Oxford University Hospitals NHS Foundation Trust.

Background
The number of cases of oropharyngeal cancer (OPSCC) is rising exponentially and is attributed to infection with human papilloma virus (HPV).

HPV screening is routine in new cases of OPSCC to help prognosticate and guide treatment. P16 immunohistochemistry (IHC) is commonly performed; this requires surgical biopsy. PCR has shown success in detecting HPV infection in the saliva of individuals. These tests have not been evaluated towards personalised treatment.

Aim
To determine the diagnostic accuracy of saliva testing against P16 IHC in patients with OPSCC.

Methods
A diagnostic accuracy study was designed using Quadas-2 and STARD criteria. Forty five consecutive patients with a new diagnosis of OPSCC had pre-treatment saliva collected and surgical biopsies performed. The index test of PCR on saliva was compared to the gold standard of P16 IHC on surgical biopsies. The tests were conducted independently and investigators blinded to the results to avoid bias.

Results
The mean age of participants was 59, 75% were male, 73% current or ex-smokers with tumours ranging in size from T1 to T4.

The sensitivity and specificity of oral rinse testing was 71% (52.6-85.8% 95%CI) and 83.3% (51.6-97.9% 95%CI) respectively. Whilst positive and negative predictive value were 91.7% (73.6-99.0% 95%CI) and 52.6% (28.9 – 75.6% 95%CI).

Conclusion
Oral rinse testing shows promise as an alternative to P16 IHC in patients with OPSCC. It may avoid the need for surgical biopsies in patients with a positive result and expedite both surgical and non-surgical treatment.

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Predictors of need for a thoracic surgical approach for excision of retrosternal goitre: A thoracic surgical perspective

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AUTHORS’ STATEMENT:
No conflicts of interest. We as authors have reviewed the abstract and have agreed its submission.

Background
There is no consensus regarding the best cross-sectional imaging predictors of need for a thoracic surgical approach in resection of Retrosternal Goitre (RG). We aim to clarify these logistically important predictors by examining our 10-year thoracic surgery series of RG-patients referred to the thoracic surgical service.

Methods
An anonymous data was prospectively collected for all our RG-thyroideotomy patients (March 2006-2016). Primary cervicotomy was always attempted first, prior to any subsequent decision for extra-cervical access. Data concerning patient demographics, post-medical history, pre-operative RG CT features, surgical approach, and tissue diagnosis. The Fisher-Exact Test was used to investigate for statistically significant predictors (significance level =0.05).

Results
Twenty-nine RG-thyroideotomy patients (mean age =67yrs; range =43-82yrs) were identified. Four had previous thyroid surgery. Three had malignant RGS. Fourteen were joint cases with ENT/General Surgery. Twelve required an extra-cervical approach (41%). All patients with either a rectangular RG (5pts; p=0.013), RG-thoracic-inlet transverse-diameter ratio of >1 (4pts; p=0.021), or ectopic RG nodules (4pts; p=0.021) required an extra-cervical approach. A RG-thoracic-inlet sagittal-diameter ratio of >1 (1pts; p=0.001) and infra-aortic arch extension (14pts; p=0.003) were also significant predictors. Infra-carinal extension (5pts; p=0.622) and posterior mediastinal extension (19pts; p=0.694) were insignificant.

Conclusion
The RG-thoracic-inlet dimensions ratio, ectopic nodules, RG-shape and infra-aortic extension are important predictors of an extra-cervical approach. A previous systematic review and other non-thoracic series describe infra-carinal and posterior mediastinal extension as significant predictors. This contradicts our experience.
MOE is a chronic infection of the external ear canal including the surrounding soft tissue and bone. It is typically caused by P. aeruginosa and tends to affect diabetic or immuno-compromised patients. It can compromise cranial nerves and has been associated with a high mortality rate. With the use of modern antibiotics this rate has decreased, however with the increasing prevalence of ciprofloxacin resistant pseudomonas, there is a requirement for alternative therapies. Hyperbaric oxygen therapy (HBOT) has been shown to be of benefit. The rational for this is that HBO is bacterio-static for pseudomonas, it stimulates leucocyte function and promotes soft tissue and bone healing.

Method
Retrospective review of the case notes of patients treated between 1995 – 2015 in a single hyperbaric centre. To ensure validity, where available, the hyperbaric centres’ notes were cross-referenced with hospital notes.

Results
There were 21 patients, all presented with otalgia and 43% had one or more cranial nerve palsy. 71% of patients had DM or were immuno-compromised and 93% of cases had confirmed P. aeruginosa. One patient had a complication of HBOT: Low grade unilateral tympanic membrane barotrauma. Following treatment 81% of patients had resolution of otalgia and 83% had resolution of cranial nerve dysfunction. The mortality rate was low with a 30-day mortality rate of zero and a 1-year mortality rate of less than 10%.

Conclusion
HBOT is a safe and useful adjunct for the treatment of MOE and should be considered early in the management of this disabling condition.