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Welcome to Volume 9 Issue 1 of Journal of ENT Masterclass® 2016

This volume arrives in my second year as the Chairman of the Editorial Board of the Journal of ENT Masterclass®. Taking this role last year made me realise the magnitude of hard work that had been taken in the past years and I am very pleased that we have continued with the previous successes. The 8th Journal of ENT Masterclass® was very well received by the ENT fraternity in UK and the free subscription has been extended to all the members of ENT-UK following its official accreditation by our national body.

This year, we have expanded the Editorial Board with the addition of Professor Zhiqiang Gao and Professor Shusheng Gong from China. To keep with the growing body of scientific knowledge and improve on the editorial scrutiny, new co-editors for each of the four sections have joined the board. Andrea Thirwall and Hasnaa Ismail-Koch have taken the role of editing the Paediatric Section from Michael Kuo, to whom we are extremely grateful for producing outstanding articles for volume 8. Michael had to step down because of his extensive commitments. Ricard Simo will be joined by Ian Nixon as a Head & Neck Section co-editor, Charlie Huins joins Richard Irving in Otology and Alwyn D’Souza joins Shahzada Ahmed in Rhinology and Facial Plastics. The Rhinology Section has always encompassed Facial Plastics articles and we have incorporated this in the title.

The quality of articles remains of a very high standard and we hope to keep this resource free for all ENT surgeons in UK and abroad. To this end, the journal has also been distributed in symbolic numbers to some overseas countries like Germany, India, Saudi Arabia and China. On the other front, over the last 12 months the ENT Masterclass® platform has continued to expand its academic profile. Within UK, the National Audiology & Balance Masterclass was well received, as was the Masterclass for General Practitioners. Audiology & Balance Masterclass was well received, as was the Masterclass for General Practitioners.

The free ENT Masterclass educational resources have expanded beyond our expectation when it started as a revision course in Doncaster in 2005. It is all down to the generous and selfless contributions from UK and international faculty, and the continuing support and interest of the ENT community. Once again we hope that we continue to provide a free educational value that is accessible to all.

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December 2016.

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Intracranial complications of suppurative ear disease

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Abstract
Intracranial complications of suppurative ear disease are typically caused by spread of infection to surrounding areas. This can be direct, local or to distant areas via haematogenous spread. The pattern of disease has changed in developed countries in recent years with these now being more frequently seen as a complication of AOM in young children with only rare cases presenting in adults and complicating chronic ear disease. Before the antibiotic era, acute otitis media (AOM) commonly resulted in intracranial complications, with mortality of 75% or more being reported. In advanced health systems intracranial complications can be seen in approximately 3% with published mortality rates now approximately 5%. Although this represents a huge advance, it should be noted that this is still 1 patient in 20.

Intra cranial, suppurative ear disease, complications

Pathogenesis
Infection spreading beyond the mucopituitosum and bony walls of the middle ear and mastoid exits via a number of routes. These are vascular, through thrombophlebitis of small valveless venules, following bone erosion or via pre-existing pathways, such as the oval and round windows, IAM, endolymphatic duct and sac, dehiscent tegmen, around the jugular bulb or through fracture lines.

Diagnosis and investigation
In their early stages, intracranial complications can be surprisingly silent. Typical symptoms are fever & headache. Clinical mastoiditis is not a prerequisite for development of an intracranial complication; many patients have only clinical signs of AOM, which may include a discharging ear. Despite the irradiation of adjacent structures, dizziness and hearing loss are rare as presenting symptoms of intracranial complications. Imaging is recommended as a first-line diagnostic modality in all cases. CT should be contrast-enhanced. CT has the advantage over MR that images can be acquired quickly, raising the possibility of a feed-and-sleep scan in a young child, and light sedation in an older child. Contrast-enhanced MRI has a higher sensitivity for fluid collections, parenchymal oedema, and vascular complications. For presence and propagation of thrombi, MR is superior to CT, but MR venography is best.

Pus or tissue samples for microbiology are very important for empirical antibiotic therapy. If surgical intervention is planned, then pus can be retrieved from the middle ear via a myringotomy in order to identify the causative organism. Intracranial infection can often be diagnosed from culturing cerebrospinal fluid (CSF) obtained via lumbar puncture, but cross-sectional imaging is mandatory first, in order to examine for the possibility of a space-occupying lesion.

Meningitis
A degree of meningitis may co-exist with other complications of AOM however isolated meningitis is relatively uncommon. Congenital anomalies or acquired injuries of the temporal bone, which provide a more ready passage to the meninges, will increase the likelihood of meningitis as a complication of AOM. This includes dural exposure in previous mastoid surgery. Middle ear injury complicated by infection may lead to acute suppurative meningitis, which can then easily lead on to meningitis.

Meningitis as a complication of AOM can be a challenging diagnosis, as the symptoms of headache, vomiting and severe fever are non-specific. Signs such as photophobia and seizures are less common in older children and adults. Infants may present with convulsions. Nuchal rigidity and conscious level are not considered reliable signs early in the disease course. Kernig’s and Brudzinski’s signs can suggest meningeal irritation, but are not considered reliable in young children. Lumbar puncture (LP) is usually required to provide a definitive diagnosis via CSF chemistry and culture, but raised intracranial pressure (ICP) may make this procedure perilous, and imaging is usually considered mandatory before LP is undertaken. Analysis of obtained CSF will show cloudy fluid, >1000 cells per ml, elevated protein, reduced glucose, and organisms. Meningitis may provide an alternative and less hazardous route to identification of the causative organism. If the suppurating disease is acute and limited to the middle ear, the mainstay of treatment is intravenous antibiotic therapy and further surgery is unlikely to be necessary. If the suppurating disease extends to the mastoid or is chronic in nature, mastoid exploration may be required when the patient’s condition allows. If such intervention is contemplated, CT and MR are recommended in order to investigate for further intracranial complications. It has been reported that meningitis is more frequently lethal when secondary to chronic otitis media, as opposed to the acute form. Patients with meningitis usually require high-dependency care, a prolonged course of intravenous antibiotic therapy, and involvement of several clinical teams.

Extradural abscess
This is an abscess between the dura and the endosteum of the cranial vault more commonly located in the posterior rather than the middle cranial fossa. An extradural abscess closely associated with the sigmoid sinus is termed a peri-sinus abscess. Symptoms are a spiking fever and headache despite appropriate antibiotic therapy, deep pain in the mastoid region and otorrhoea. Extradural abscess is frequently associated with other complications of acute infection, such as a brain abscess or dural sinus thrombosis (Fig 1). Localised meningitis is expected, and the involved dura can thicken in response to the intense inflammatory stimulus (this is termed pachymeningitis). Treatment is surgical drainage via mastoidectomy combined with antimicrobial therapy. Wide mastoid exposure is recommended, maximising access before addressing the area adjacent to the abscess.

Empirical antimicrobial regimens are targeted against traditional AOM organisms. A typical regimen is: Metronidazole with Ceftriaxone or Cefotaxime. Ceftazidime may be substituted where there is a suspicion of involvement with pseudomonas (for example in the context of chronic ear disease). Vancomycin can be used for coverage of MRSA although achieves only low concentrations in the central nervous system, therefore Rifampicin may be added for its superior CNS penetration.

Antimicrobial therapy is usually continued for 6 weeks (8 if slow progress), with a repeat MRI at 4-6 weeks, or earlier if there is clinical deterioration.

Subdural empyema
This is a serious complication with infection between the dura and the arachnoid. The majority of such infections develop secondary to trauma or neurosurgery, but in the context of otitis, infection is typically transmitted by extension of disease through an emissary vein or other breach in the dura, and it may also develop secondary to meningitis and localised osteitis. Pus in the subdural space spreads rapidly resulting in progressive thrombophlebitis, encephalitis, raised ICP and cortical infarction causing focal neurology and seizures. The majority of cases occur in the second decade of life, and more commonly in males.

Headache is universal, and the typical patient will also be unwell, febrile and vomiting. Late signs include worsening fever, nuchal rigidity, focal neurology and fluctuating consciousness. Paralysis of contralateral conjugate gaze may result in deviation of the eyes towards the side of the lesion. The presence of focal neurology and declining consciousness in the context of otitis media demands decisive action. Empyema localised to the posterior fossa may show fewer localising signs, but will produce signs of meningeal irritation and raised ICP.

Figure 1: Contrast enhanced CT on soft tissue setting demonstrating an extradural abscess. Cerebellar abscess and surrounding cerebellar oedema.
Early in the disease course, radiological signs may be subtle, particularly with unenhanced CT and contrast-enhanced scans are recommended. MRI is more sensitive than CT but has less impact on surgical planning.

Lumbar puncture is contraindicated in the presence of clear subdural empyema owing to raised ICP. An apparently-reassuring scan early in the disease course may lead to consideration of LP as a diagnostic test, and in the case of bacterial meningitis without empyema this would represent correct management. Therefore, if LP is considered as a diagnostic modality, it should be performed as soon as possible after a scan demonstrates no evidence of raised ICP.

Treatment of subdural empyema is emergent neurosurgical drainage, washout and empirical antibiotic therapy (see “Extradural abscess” section for appropriate agents).

Percentage chance of survival correlates with conscious level at the time of surgical intervention. If acute mastoiditis is present, treatment is wide exploration of the mastoid with evacuation of infected material. Patients will often require high-dependency care and joint management with neurosurgery.

**Dural venous thromboses**

Sigmoid sinus thrombosis can arise via spread through the mastoid and clearance of infected material. Patients will occur without the presence of clinical mastoiditis2, possibly anaerobes5,11. In a review of paediatric cases Au et al. (2013) reported rates of partial or complete recanalisation in 25%34,35. Deteriorating mental state indicates worsening cerebral oedema and is a poor prognostic sign.

**Surgery**

A small minority of cases in the literature are treated without surgery although this is not standard management. Au et al. (2013) suggest that this be considered only for patients without intracranial signs and who respond rapidly to antibiotic therapy11.

**Brain abscess**

Intraparenchymal abscesses form a higher proportion of the abscesses found in developing countries, and carry a potentially high mortality, whilst in developed countries incidence is decreasing with time25,26, and mortality is low15. A report of 122 consecutive cases of brain abscesses in a Taiwanese hospital stated that “otitis” was the third-most common underlying cause of intraparenchymal brain abscess22. Various studies and reports note that such lesions are more common in males, in those of low socioeconomic status, and more common ipsilateral to the otic lesion20,21.

These lesions typically present with non-specific symptoms and can be surprisingly silent for a long time resulting in a delay in diagnosis21. The most common symptom is severe headache (insidious or sudden in onset), which is usually ipsilateral to the abscess. Neck stiffness occurs in only 15% of patients, and vomiting generally only as a result of raised ICP. Fever occurs in only 45 to 53% of cases and is therefore not a reliable sign. Focal neurology is documented in 50% of cases and is often delayed whilst seizures occur in 25%41. Deteriorating mental state indicates worsening cerebral oedema and is a poor prognostic sign. A brain abscess is a space-occupying lesion and in the context of focal headache, focal neurology (eg unilateral cranial nerve deficits, hemiparesis) or papilloedema, LP is contraindicated. Imaging is recommended and care must be taken in the interpretation of imaging. Complications with patients with an abscess will have a second intracranial complication. An abscess appears as an area of low density or low signal, surrounded by a ring of enhancing tissue, and is seen more clearly on MRI than CT, although CT may be easier and quicker to obtain.

**Otogenic abscesses**

are typically either cerebellar or temporal lobe. A cerebellar abscess may additionally present with horizontal nystagmus, tremor or ataxia, and a temporal lobe abscess with seizures, speech and visual disturbance. Symptoms and signs of cerebral abscess are typically more constant and definite in the case of a cerebellar abscess, owing to the restricted space in the posterior fossa and proximity to the brainstem. It can be the result of extension from posterior fossa dura, and in theory is then in continuity with an infected mastoid and middle ear cleft. It is possible to perform needle drainage of a cerebellar abscess through the posterior fossa dura via the opened mastoid cavity. A direct brain abscess may have arisen secondary to petrositis, but is more likely to be a result of retrograde thrombophlebitis. It is likely to develop over a longer timescale than the local posterior fossa abscess, and should be managed by a neurosurgeon. If indicated, mastoidectomy should be performed contemporaneously, through a separate surgical field. UK guidelines recommend that imaging be performed if an abscess has been drained or 6-8 if not. Again, empirical treatment with metronidazole with ceftriaxone and anhydrase inhibitors such as Acetazolamide can be used to reduce CSF pressure.

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Otitic hydrocephalus

This is a condition in which symptoms and signs of acute hydrocephalus followed some form of otitis. There is a spectrum of opinion on the nature of this entity; ranging from a distinct form of benign intracranial hypertension related to acute otitis media but unrelated to a space-occupying lesion, to sigmoid thrombophlebitis, to the increasingly accepted pathophysiological consequences of sigmoid sinus thrombosis related to any cause. 23 In this former definition, the cause of hydrocephalus is unknown, other than being related in some way to acute otitis media.

The predominant symptom is a diffuse headache, and imaging of cerebral ventricles will suggest the diagnosis. CSF collected at LP is normal. In the simplest disease course, collateral venous outflow opens over approximately 3 to 7 days, and the headache improves as ICP begins to fall. Signs of worsening disease include visual disturbance, reduction in mental ability and reduction in conscious level. Such developments constitute an emergency, and high doses of corticosteroids and diuretics should be administered and consideration for CSF diversion.

References

loss, tinnitus and vertigo as seen in Meniere’s disease and LVAS.1

Pathogenesis
In normal development, by the age of four years old, the growth of the posterior fossa pulls the endolymphatic duct from a short broad column into an elongated inverted ‘J’ shape.1 The outcome is a vestibular aqueduct diameter of 0.4 – 1.00 mm (mean of 0.62) in adults. In LVAS there is aberrant development in the seventh week of gestation which plays a crucial role in its function.13 The enlarged hypofunctioning sac leads to increased hydrostatic pressure which in turn leads to enlargement of the vestibular aqueduct. This is supported by histopathological findings of bony erosion leading to some authors referring to the syndrome as EVAS.2,11

Epidemiology
LVAS is the most common radiological and morphogenetic abnormality of the inner ear among children accounting for 13-15% of sensorineural hearing loss. Patients with unknown causes of sensorineural hearing loss, the prevalence of LVAS ranges between 0.64% to 13%.8 The malformation is overwhelmingly bilateral and has a female to male preponderance.7

Diagnosis
Computer-assisted tomography (CT) scanning of the temporal bones is the best tool to assess the bony VA (Figure 1). Magnetic resonance imaging (MRI) is most suited for visualising the endolymphatic duct and sac (Figure 2). The size criterion for diagnosis of LVAS, as suggested by Valvassori and Clemens13, is a VA or endolymphatic duct measuring >1.5mm at the midpoint between the common crus and the external aperture. Whilst this has been the standard, it is the subject of much debate: some have defined LVAS in cases where the midpoint is greater than 2mm,14 others when greater than 4mm.15 More recently the criterion has been revised down to 0.9 mm at the midpoint and 1.9 mm at the operculum. This has resulted in classifying previously unexplained sensorineural hearing loss as LVAS.16

Association with other syndromes
The SLC26A4 (PDS) gene encodes Pendrin, a protein involved in the transport of chloride, iodine, and bicarbonate anions across cell membrane. Mutations in the PDS gene are associated with both syndromic LVAS in Pendred’s syndrome,17 and nonsyndromic LVAS. Pendred syndrome has an autosomal recessive inheritance and is characterised by thyroid goitres and hearing loss. LVAS has also been associated with distal renal tubular acidosis, X-linked congenital mixed deafness, Waardenburg syndrome, otosaccro-ovalveal syndrome, branchio-oto-renal syndrome (BOR), and Noonan’s syndrome. Differences in hearing loss fluctuation and other clinical features have been reported across the different associated syndromes: patients with BOR and LVA are described as suffering from neither fluctuating hearing loss nor vertigo in contrast to those with Pendred syndrome and LVAS.11

Clinical Features
The symptoms are reported to manifest before the age of 10 years in 90% of patients. The most prevalent symptom at onset is hearing loss. Dizziness, vertigo and imbalance are only present in a minority of patients.11

Head trauma is most commonly reported as the precipitating event for hearing loss in up to 26% of patients.2,3,11,13 This is thought to be due to increased cerebrospinal fluid (CSF) pressure. This is corroborated by barotrauma and the Valvassori manoeuvre also precipitating hearing loss in up to 17% of patients.13 Other factors include upper respiratory tract infections, high fevers, acoustic trauma and physical exercise. Vestibular symptoms are usually disequilibrium or episodic attacks of vertigo. Younger children may present with poor coordination or an inability to walk.

Relation to hearing loss
Hearing loss severity has been found to have a linear correlation of 6 dB HL per unit of vestibular aqueduct diameter (95% CI 2-10; p = 0.003).1 All three types of hearing loss have been reported in LVAS (sensorineural, mixed and conductive). The majority are either mixed or sensorineural. There is often an air bone gap (ABG) in the lower frequencies (see Figure 3). The ABG at times has been misdiagnosed as osseous.14 Many theoretical models have been proposed to account for the ABG.2 Some studies have shown a gradual hearing loss of around 4 dB per year whilst others have reported the majority as stable. The hearing loss quite often fluctuates and is most often severe to profound with a recent large nationwide study finding a mean pure tone average (PTA) of 83.7 dB HL.12

Mechanisms of hearing loss
These have been widely theorised in the literature:1,4,8,10

1. The hyperosmolar reflux theory postulates that the large endolymphatic sac contains hyperosmolar fluid which refluxes through the LVA. It enters the inner ear thereby causing damage.

2. The electrolyte imbalance theory proposes that the endolymphatic sac is dysfunctional in its homeostatic role. This results in electrolyte derangement which damages the ion pump system of the stria vascularis.

3. The back pressure wave theory was initially suggested by Valvassori. It suggests that the conductive component is due to a back pressure of perilymphatic and endolymphatic fluid which results in decreased stapes mobility and an ABG particularly in the low frequencies (Figure 3). This is supported by the increased incidence of perilymphatic gushers. This is also said to explain head trauma and barotrauma as precipitating factors.

4. The third window lesion theory hypothesises that the change in the dimensions of the bony labyrinth affects impedance and compliance of the auditory system. This results in a third window loss of acoustic energy away from the cochlea through the VA - this leads to reduced air conduction thresholds. The enlarged VA is thought to furthermore lower impedance at the scala vestibuli. The resulting pressure difference between scala tympani and scala vestibuli leads to improved cochlear responses to bone conduction (Figure 3). This explains the supranormal bone conduction thresholds seen in some patients.

Management
Patients should be advised to minimise risk of head trauma by avoiding contact sports. Patients should also be advised not to scuba dive, to avoid sneezing with their nose pinched as well as weightlifting, and straining as these may lead to barotrauma due to increased CSF pressure.2,10

Hearing aids are usually beneficial in patients whose hearing loss is established. Cochlear implants are also commonly used in LVAS. Auditory skills of infants with LVAS have been demonstrated to develop rapidly after cochlear implantation similarly to those with normal inner ears.13

Figure 1: CT scan of the temporal bones depicting bilateral vestibular aqueducts (see red arrows) in a 43 years old with congenital deafness. The midpoint diameters are 3.4 on the right and 2.1 mm on the left

Figure 2: T2 weighted MRI scan of the internal acoustic meati of the same 43 year old with congenital deafness. Her large endolymphatic ducts and sac are depicted (see red arrows)

Figure 3: Audiogram of the same 43 year old patient with congenital hearing loss prior to cochlear implantation. Her BKB score was 3%. The low frequency ABG is well demonstrated
Corticosteroid therapy has been reported in small case series and case-control studies to produce hearing improvement in patients with LVAS. It is thought to be due to a reduction in pressure. Hyperbaric oxygen therapy has been administered in patients with LVAS and sudden hearing loss after head trauma. It is thought to lower CSF pressure and reflux of hyperosmolar endolymph into the cochlea, as well as increase oxygen tension in the inner ear fluid. It has been shown to improve hearing thresholds in small case series. However, the natural history may have been that of fluctuating hearing loss. Endolymphatic sac surgery has been attempted to improve or stabilise hearing in LVAS. This is controversial and most studies have found it to have a deleterious effect to hearing.

**Conclusions**

LVAS is the most common morphological cause of sensorineural hearing loss in childhood, usually presenting before the age of 10 years old. It is associated with many syndromes, in particular Pendred’s syndrome due to a mutation in the SLC26A4 gene. Hearing loss severity has a linear correlation with VA diameters, it may also be mixed with a large ABG. There are many valid theories that explain hearing loss in LVAS.

**References**


Management of sudden sensorineural hearing loss

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This paper was presented by the author at the British Society of Otology Annual Meeting, Bristol, February 2016

**Conflicts of Interest/ Financial associations**

None declared

**Introduction**

Sudden loss of inner ear hearing is a devastating occurrence and patients must therefore be managed promptly and thoroughly; most specialists consider sudden sensorineural hearing loss (SSNHL) to be an otological emergency. Careful investigation may identify an underlying cause, though most cases are idiopathic. Our understanding of SSNHL is poor and this is reflected in the literature where there is a paucity of high quality studies and conflicting opinion on the aetiology, definition, and optimal treatment of the condition. Clinicians must therefore have a good understanding of the best available evidence and offer patients treatment on an individualised basis.

**Definition, Incidence and Presentation**

The most universally applied definition of SSNHL is a loss of at least 30dBHL in three contiguous frequencies on the audiogram, occurring within three days (72 hours). Hence this includes cases of sudden immediate hearing loss as well as those that progress rapidly. There is no consensus on how to define recovery from SSNHL. Previous papers have suggested systems for grading recovery. Unfortunately these are not well utilised in the literature, making meta-analysis of studies in this field impossible. Most cases of SSNHL are unilateral, the incidence of which is thought to be 5 to 20 per 100,000 per year. The true incidence may be higher, as mild cases or those that recover quickly are often unreported. The commonest age group affected is adults in their forties and fifties, with an equal gender distribution. Bilateral cases usually occur sequentially and account for less than 2% of all SSNHLs. In addition to hearing loss, one quarter to one half of patients will have vestibular symptoms. Tinnitus and fullness are also common. In non-idiopathic cases there may be other symptoms to suggest the underlying diagnosis. Cases of Wiklund's hearing loss (not discussed further here) may present with a rapid onset and must be ruled out by careful history taking and examination.

**Aetiology and Pathophysiology**

The pathophysiology of SSNHL is unknown. Various theories have been proposed, including viral, vascular, autoimmune and cellular response mechanisms. There may be multiple pathophysiological mechanisms that each culminate in the same end condition. Between 70 and 90% of cases of unilateral SSNHL are idiopathic. One review found the commonest aetiologies to be idiopathic (71%), infection (12.8%), otologic disease (4.7%), trauma (4.2%), vascular/haematological (2.8%), neoplastic (2.3%) and ‘other’ (2.2%)9. A more complete list of previously recognised aetiologies for SSNHL is shown in Table 1 (adapted from Chau et al). Bilateral SSNHL, though rare, is more commonly due to an underlying systemic pathology, many of which are associated with significant morbidity and mortality. Bilateral SSNHL tends to present with more severe hearing loss and has a worse prognosis.

**Investigation of SSNHL**

In the presence of an appropriate history, the diagnosis of SSNHL is confirmed with the pure tone audiogram. In the majority of cases there is no need to perform additional audiological or vestibular tests. Given the wide range of possible aetiologies for SSNHL (Table 1), it is impractical to perform additional investigations for each of them.
Hence the extent of investigation required will be dictated by the history and the availability of tests locally10,11. A magnetic resonance imaging (MRI) scan (or computerised tomography (CT) scan if MRI is contraindicated) is recommended in all cases, as 10 to 20% of vestibular schwannomas will experience SSNHL at some stage, often as the presenting feature12.

An MRI scan will also detect many cases in which there is a vascular compromise, neurological condition (e.g. multiple sclerosis, secondary hearing loss, bone or cartilage tumours), or inner ear abnormality. Although basic investigations (such as blood tests, blood count, erythrocyte sedimentation rate, urea and electrolytes) have a low yield, they are often recommended as they are readily available, inexpensive and may point towards the need for more detailed investigation. Raised cholesterol has been demonstrated in 35 to 40% of cases of SSNHL, and elevated serum glucose in 18 to 37% of cases; initiation of treatment may prevent future cardiovascular morbidity13.

Abnormal thyroid function (hyperthyroidism) may be detected in as many as 15% of cases of SSNHL and has also been suggested as a routine test7. There is little evidence to support a routine infectious or auto-immune screen, though many clinicians routinely perform a screen for syphilis as the possibility of treatable latent infection may not be evident from the standard history14.

Table 1. List of potential causes of SSNHL (adapted from Chau et al 2010).

<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic</td>
<td>Adenovirus, Epstein-Barr, hepatitis C, herpes simplex, measles, mumps, rubella, varicella, streptococcus, pertussis, meningitis, syphilis, toxoplasma, Lyme disease, Lassa fever, mycoplasma, HIV</td>
</tr>
<tr>
<td>Infectious</td>
<td>Merieux’s disease, secondary hydrophobia, otosclerosis, automicine inner ear disease (AIED), post-op (otologic surgery), ototoxicity, inner ear abnormality</td>
</tr>
<tr>
<td>Otolologic</td>
<td>Acoustic trauma, barotrauma, acoustic trauma, traumatic irritation</td>
</tr>
<tr>
<td>Traumatic</td>
<td>Head injury, perilymphatic fistula, baro-trauma, neglect of muscular trauma, traumatic injury</td>
</tr>
<tr>
<td>Autoimmune</td>
<td>AIED, Behcet’s, Cogan’s, systemic lupus, anti-phospholipid, temporal arteritis, granulomatosis/ vasculitis</td>
</tr>
<tr>
<td>Vascular</td>
<td>Cardiovascular event, subdural/ subarachnoid haemorrhage, transient ischaemic attack, saddle cell disease, haemostasisis, coagulopathy</td>
</tr>
<tr>
<td>Tumour</td>
<td>Acoustic neuroma, meningoma, cerebellar abnormality, multiple myxoma, metastasis</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Multiple sclerosis, migraine</td>
</tr>
<tr>
<td>Other</td>
<td>Post-op (non-otologic surgery), pregnancy, rabies vaccination, CO poisoning, functional/ maligning</td>
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Bilateral SSNHL is a special case, as the potential for an underlying life-threatening pathology warrants additional investigations including an urgent MRI scan, autoimmune screen and tests for infection (including HIV test)6. A full discussion of the management of recognised conditions causing SSNHL, as well as bilateral SSNHL, is outside the scope of this article and the sections that follow apply to the management of idiopathic cases.

### Natural History and Prognostic Factors

Before treatment can be discussed, the natural history of idiopathic SSNHL must be considered. It is recognised that many cases of SSNHL recover without treatment, with 32 to 65% (average of 46.7%) showing recovery within 2 weeks15. A meta-analysis of SSNHL studies showed a 14.3dB recovery on the pure tone audogram following treatment with placebo compared to 13.8dB after active medical therapy16. A less severe hearing loss at presentation and low frequency hearing loss are thought to be positive prognostic factors. Age over 60 or under 15 years, co-existing vertigo at presentation, and more severe hearing loss are negative prognostic factors. In addition, there is some evidence that early treatment may improve the prognosis, with spontaneous recovery rare after two months17,18. These factors must all be considered when determining how to treat an individual patient, as in some cases the most acceptable form of treatment may be supportive measures rather than medical therapy.

### Treatment of Idiopathic SSNHL

#### Supportive Measures

It is crucial to remember that SSNHL is a devastating experience for the patient, and a high level of support, counselling and reassurance from the treating clinician is required24. In the author’s experience, long-term support for patients with incomplete recovery is often lacking; adequate access to hearing rehabilitation including provision of hearing aids and treatment for single-sided deafness is important, and in selected cases tinnitus therapy, vestibular rehabilitation or formal psychological support may be required.

#### Medical Therapies

A variety of medical and alternative therapies have been suggested for SSNHL (Table 2). Steroids have been the mainstay of treatment for SSNHL for some years, and are recommended as first-line treatment by the majority of ENT consultants in the UK21,22. Despite this fact, there remains a paucity of evidence for their efficacy and there is no firm consensus on which is the best steroid agent nor on the optimal dose, timing (including the role of salvage therapy) or route of administration.

### Oral steroid therapy

Some of the earliest published trials examining SSNHL suggested that oral steroid therapy may have an advantage over placebo treatment23. More recently, other studies, systematic review, meta-analysis and a Cochrane review on this subject have shown contradictory evidence and an uncertain benefit from oral steroids25,26. Whilst there is significant variation, prednisolone seems to be the most widely used agent, commonly a short (e.g. 5 days) course at 1mg/Kg daily. Potential side effects must be considered and clearly caution is required in patients who are very elderly or who have relative contra-indications for steroid treatment. Although SSNHL is considered an emergency, there is some evidence that oral steroid treatment at 1 week may be as effective as that given within 24 hours27. However, there is additional evidence that there may be a worse outcome after 10 days, and little benefit from treatment after 4 to 6 weeks28,29. Common sense therefore dictates prompt treatment whenever possible.

### Intra-tympanic steroid therapy

Based on theoretical and animal studies that have shown an increased concentration of steroid in the perilymph of the inner ear when compared to oral or intravenous treatment, interest in the use of intra-tympanic (IT) steroids in SSNHL has grown in recent years. In the majority of studies dexamethasone or methylprednisolone is injected through the tympanic membrane into the middle ear cavity in the perilymphatic space.

**Figure 1. Technique for intra-tympanic steroid injection**

1. A bleb of EMLA cream is applied to the tympanic membrane (figure 1a). An inferior site is preferred for easy access for injection (and for perforation repair in case of perforation). Targeting of the round window niche is not required when filling the entire middle ear with fluid.
2. The cream is aspirated after 5 minutes using microsuction with fine tip.
3. A 27G spinal needle (bent so slightly angled) is used to puncture the eardrum (figure 1b).
4. Steroid solution (dexamethasone in highest available dose) at room temperature is infused slowly under direct vision. Normal volume instilled is approximately 0.5mls.
5. Patient is asked to remain with head turned to side for 30 minutes, and avoid talking/swallowing as much as possible.
middle ear space, though some advocate use of a round window wick or steroid drops instilled through a grommet. The technique favoured by the author is described in Figure 1. Randomised controlled trials and systematic review of the literature have shown that IT steroids are no more effective than high dose oral steroids when used as primary treatment \(^{12,13}\). There is limited evidence that combination therapy with oral and IT steroids together is more effective than oral treatment alone\(^{12,13}\). For this reason, most clinicians take a practical approach and use oral steroids alone as the preferred initial therapy unless contra-indicated, and this is reflected in published guidelines for SSNHL management\(^{19,34}\). When considering IT steroids as a salvage treatment following failed systemic steroid treatment, the balance of evidence from systematic reviews and meta-analysis seems to be towards showing some additional benefit\(^{12,13}\). The meta-analysis by Ng et al. pooled the results from 5 RCTs and showed an improvement in hearing outcomes in patients undergoing IT steroid injection (dexamethasone or methylprednisolone) following failed systemic steroid treatment (defined as persistent SNHL worse than 30dB or more than 10 to 20dB than the contra-lateral ear)\(^{36}\). In these studies, treatment consisted of repeated injections (e.g. 4 times over 15 days) commenced within 4 weeks of onset of the illness and it was felt to be justified to discuss the options before deciding on a treatment plan, which should include adequate counselling, support and rehabilitation.

### References


25. Narazaki K, Sat0 S, Nakasui K, et al. Anti-viral treatment, vasodilators/ vasoactive substances and hyperbaric oxygen have been suggested as treatments but Cochrane reviews of their efficacy have demonstrated little or no evidence to support their use\(^{11,37}\).

Future Therapies

Currently, there is much interest in the use of insulin-like growth factor in the inner ear and this may provide a promising treatment for SSNHL. It is clear that as our understanding of inner ear biology improves, new treatments (e.g. stem cell and gene therapies) will emerge to treat sensory hearing loss including SSNHL.

### Conclusion

Sudden sensorineural hearing loss is a rare but potentially devastating condition. Our understanding of the pathophysiology of SSNHL remains poor, with only a minority of cases reported to have an identifiable cause\(^{13}\). The evidence for medical treatment in SSNHL is weak. Nonetheless, there is some consensus that the potential benefit of oral steroids as primary treatment outweighs the risks in most cases. Intra-tympanic steroid therapy is emerging as a potential salvage treatment and as primary treatment when oral steroids are contra-indicated. Clinicians treating SSNHL must take an individualised approach to managing each patient and be prepared to discuss the treatment options before deciding on a treatment plan, which should include adequate counselling, support and rehabilitation.
Patulous eustachian tube

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Key words
Patulous Eustachian tube, augmentation, injection

Introduction
The Patulous Eustachian Tube (PET) can be defined as an abnormal patency of the Eustachian Tube (ET)¹. A normally functioning ET is closed in its resting state and only opens briefly during maneuvers such as swallowing and yawning. The ET opening is understood to be achieved by a coordinated contraction of the peritubal muscles; tensor veli palatini and levator veli palatini². In PET, the ET can remain patent for hours, allowing for a direct communication of sound and air pressure between the nasopharynx and middle ear.

Aetiology
The increased incidence of PET following weight loss (such as in malignancy) supports the theory that loss of volume in Ostman’s (peritubal) fat pad may be implicated in the aetiology of this condition. In addition, loss of volume to the mucosa or submucosa in this valve region has been suggested as the explanation for its occurrence following radiotherapy. Hormonally induced changes to nasal mucosa are also thought to explain the increased incidence of PET in pregnancy (which resolves after delivery), and the onset with hormonal treatment of prostate cancer and the OCP. However, paradoxically high levels of oestrogen are also believed to cause mucosal oedema as seen in rhinitis of pregnancy, in addition, topical oestrogen drops have been used as a treatment for PET. The mechanisms here remain to be clarified. Other risk factors identified include neuromuscular disorders and scarring from previous surgery such as adenoidectomy³.

Presentation
Typically patients present with autophony to voice and respiration (also termed breath synchronous tinnitus), due to reflux of sound via the column of air between the nasopharynx and middle ear. Patients can also complain of aural fullness, which may be misleading in suggesting a dilatory dysfunction of the ET. Although other otological symptoms have been reported, PET does not classically cause hearing loss or vertigo. The key alternative differential diagnosis to exclude is superior semicircular canal dehiscence (SSCD) that has some overlap in presentation. Table 1 shows the key presenting features of both for comparison with the most important differentiating features in red. Ultimately if there is any doubt in the diagnosis a CT temporal bones should be sought to assess for SSCD.

It is not uncommon for patients to present to more than one ENT doctor prior to receiving the correct diagnosis. In the senior authors series of patients treated for PET, all 12 had been seen by at least one ENT doctor without their PET being diagnosed and the mean duration of symptoms prior to treatment was 6.7 years. This delay may be due to a reduced awareness of PET and subtle clinical signs without abnormalities on routine otoscopy, pure tone audiometry and standard tympanometry.

Examination of the ear is best performed with magnification (microscope/endoscope) to detect breath synchronous medial and lateral excursions of the tympanic membrane (TM). These may be more obvious in an atrophic segment...
Sonotubometry can also detect a patent ET but is not recommended. Figure 1: shows a long time based tympanometry over a 20 second period with the typical ‘saw-tooth’ trace as seen in PET. Courtesy of Gemma Crunchell, Specialist Audiologist, Addenbrooke’s Hospital, Cambridge.

of the TM. Some authors have suggested occlusion of the contralateral nostril and forced breathing with the aim of exaggerating this sign. Nasendoscopy may show a scalloped concavity to the appearance of the antero-lateral wall of the ET orifice.

Frequently patients describe certain exacerbating and relieving factors. Common exacerbating factors are weight loss, dehydration, alcohol/caffeine intake, prolonged talking and exercise. Relieving factors include lying supine and nasal irrigation such as in the shower or when swimming. It is thought that the cessation of symptoms on lying supine is due to venous engorgement, induced temporary compression of the internal jugular vein can lead to prompt resolution of symptoms. This should be borne in mind when performing otomicroscopy, ensuring it is done in the upright position. Conversely, if the symptoms are not present in clinic, asking the patient to exercise may bring on symptoms, allowing confirmation on examination.

It is the senior authors opinion that one should be wary of offering surgical treatment for symptoms of PET in cases where respiratory TM excursions have not been seen.

Investigations Tuning fork tests and a pure tone audiometry are typically normal. The most useful objective test is continuous auditory synchronous compliance pattern which appears as a ‘saw-tooth’ type trace, reflecting changes in compliance in time with inspiration and expiration. See Figure 1 for example. Sonotubometry can also detect a patent ET but is not usually available in routine clinical practice. A CT scan may be indicated to exclude SSCD but also to identify a dehiscence of the carotid canal if ET augmentation surgery is planned.

Management The management of PET should be tailored to each individual patient depending on the severity of symptoms. We know that PET does not progress to cause hearing loss or any other otological complication and can therefore be safely left untreated. For many patients, an explanation and reassurance is sufficient and they can be discharged with their PET being merely an annoyance. However we know from more recent published studies using patient reported outcome measures (PROMs) that for some patients PET symptoms have a significant impact on their quality of life.

All patients may receive initial management with conservative measures aimed at addressing potential exacerbating factors including, encouraging hydration, reducing diuretics (caffeine and alcohol), stopping topical nasal steroids/decongestants and regaining lost weight. For those with persistent problematic symptoms, there are no RCTs demonstrating optimal management of PET, however, a wide range of treatments have been reported. These can broadly be divided into 4 different treatment strategies.

1 Topical nasal medications The simplest of these is instillation of saline into the nasopharynx presumably creating a meniscus of fluid over the ET orifice although this is clearly very temporary. Other agents aim to induce a chemical thinnings causing increased mucous production or mucosal swelling (such as potassium iodine drops with boric acid powder instillation). Some of these irritant solutions are available commercially as over the counter preparations. In addition to these, topical oestrogen drops (Premarin) have been used to induce mucosal hypertrophy.

Although worthwhile trialing as a non-invasive first line option, benefit is often limited and temporary meaning their use may be limited to patients with less severe PET.

2 Altering tympanic membrane mechanics Mass loading of the tympanic membrane with layers of paper or even Blue Tac has been shown to reduce symptoms of PET. This has led to surgical manipulation of the tympanic membrane to stiffen any potentially flaccid segments of the TM with modest benefit. The rationale for this is that in PET there is loss of the middle ear air cushion dampening effect with a resulting increased compliance of the TM to sound pressure waves transmitted via the ET. Whilst stiffening the TM may allow for some improvement in symptoms, it does not appear to address the underlying pathophysiology of the condition and has the potential risk to damage middle ear structures and hearing.

Interestingly 3 of the patients in our series had previously undergone grommet insertion as an attempted treatment with no improvement and in 2 the symptoms were worse.

3 Augmentation of the ET via the middle ear Given that patients with PET often present to otologists, it is not surprising the middle ear has been used as the route to augment the ET. This has been performed via myringotomy and tympanotomy to insert a shim/silicone plug with good results. Again there, is clearly a risk to middle ear structures and hearing. One of the patients in our series had previously undergone attempted augmentation of the ET via this route resulting in TM injury and a conductive hearing loss.

4 Augmentation of the ET via the nasopharynx This approach is probably the most established strategy and its use dates back to Zollner who injected paraffin into the nasopharynx presumably creating a meniscus of fluid over the PET. Whilst stiffening the TM may allow for some improvement, it is not surprising the TM is not the ideal route for this procedure.

Over the last 6 years the senior author has performed this procedure with the following technique using Vox® (a silicone elastomer implant material previously termed Bioplastique). Following topical nasal preparation with Moffett’s solution, the patient is placed supine with the mouth held open with a Boyle-Davis gag and a catheter passed through the nose and out through the mouth to retract the soft palate on the contralateral side to the ET being treated.

A 30 degree 4mm Hopkins rod is used through the ipsilateral nasal passage to view the Eustachian tube orifice as the angle instruments are introduced into the post nasal space via the mouth. Initially the eustachian tube orifice is opened using the Kujawski 800 angled instruments (Karl Storz). See Figure 2. This permits an improved view of the lateral wall of the Eustachian tube, facilitating injection into the more superolateral narrower segment of the eustachian tube rather than simply the eustachian tube cushion. Sufficient Vox® is injected until the typically concave lateral wall is filled thereby becoming convex and occluding the eustachian tube (usually 1-2ml per side).

All patients are followed up with PTA and tympanometry to assess for the development of glue ear and those with only temporary improvement are offered a repeat procedure.

Outcome measures It is well recognized that patients can be observed to have breath synchronous tympanic membrane excursions without symptoms of PET, analogous to the incidental finding of dehiscence of the superior semicircular canal without symptomatic correlation. With this in mind, objective outcome measures may be less useful, leaving us to rely more reliant on patient reported outcome measures (PROMs) to compare efficacy of different treatments. In recent years Poe has developed a PROM symptom scoring scale to determine benefit from treatment that has been adopted by other authors.

Treatment benefit PROM adapted from Poe

1 = Complete relief (no symptoms at all)
2 = Significant improvement, satisfied (no longer experience under normal circumstances, only on exercising/prolonged talking)
3 = significant improvement, dissatisfied (desired further medical/surgical treatment)
4 = unchanged
5 = worse
In addition, Rotenberg et al have devised a scoring system that measures impact on quality of life before and after treatment.1-5

1. Absence of autophony
2. Occasional autophony but not enough to affect activities of daily living
3. Consistent autophony throughout activities of daily living
4. Problematic autophony affecting ability to perform activities of daily living
5. Distracting symptoms leaving patient unable to cope on a daily basis

We used these measures to determine effectiveness of treatment in our series of patients. After a mean follow up of 6.3 months, 73% (8/11) were satisfied gaining either complete or significant improvement in symptoms. One patient had an improvement but was dissatisfied and two patients derived no persisting benefit from the procedure.

Regarding the quality of life outcome scores (as devised by Rotenberg et al), when comparing pre-operative with post-operative, the 8 satisfied patients reported an improvement where the remaining 3 patients had no change to their score see Fig 3. This improvement was statistically significant when tested with the Wilcoxon Signed-Rank Test p<0.01 (W=4).

Conclusion
Patulous eustachian tube causes symptoms of autophony to voice and respiration, sometimes with aural fullness and is due to an abnormally patent eustachian tube. Diagnosis is often delayed and whilst the majority can be managed with conservative measures and reassurance, a minority have more severe symptoms which lead them to seek further treatment.

There is a lack of evidence to support any one treatment strategy over another however consensus in the use of outcome measures will allow for more direct comparison of techniques in the future.

Table 2 lists and compares some of the more recently published series of surgical treatment strategies for PET showing success rates and outcome measures used.

<table>
<thead>
<tr>
<th>Series author</th>
<th>Year journal</th>
<th>Technique</th>
<th>Activities of daily living</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boedts et al</td>
<td>2014 Laryngoscope</td>
<td>Transnasal shim into ET (wax filled catheter) secured with suture</td>
<td>100% complete resolution of symptoms (score 1/5)</td>
</tr>
<tr>
<td>Brace et al</td>
<td>2014 Laryngoscope</td>
<td>Tympanic membrane manipulation with laser myringoplasty (LM) or cartilage tympanoplasty (CT)</td>
<td>73% overall</td>
</tr>
<tr>
<td>Pence et al</td>
<td>2014 Otol Neuroutol</td>
<td>ET injection augmentation with Vax-Implant</td>
<td>11 ears in CT group improved but symptoms worse in 71% of LM group</td>
</tr>
<tr>
<td>Kong et al</td>
<td>2011 Am J Otolaryngology-Head and Neck Surgery</td>
<td>Auris cheloplasty injection to ET</td>
<td>Resolution of symptoms 100%</td>
</tr>
<tr>
<td>Poe et al</td>
<td>2007 Otol Neuroutol</td>
<td>Endoluminal cartilage shim ET reconstruction</td>
<td>Improved and satisfied (1 or 2)</td>
</tr>
<tr>
<td>Sato et al</td>
<td>2005 Acta oto-laryngologica</td>
<td>Transmypanic ET silicone plug</td>
<td>Relief from symptoms 71%</td>
</tr>
<tr>
<td>Ohashi et al</td>
<td>2003 Otolaryngol-HNS</td>
<td>Autologous fat injection to ET</td>
<td>Resolution of symptoms 100%</td>
</tr>
</tbody>
</table>

Figure 3: Pre and Post Operative Autophony QOL Impact Scores

References
Principles of facial reanimation

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Abstract:
Long-term facial nerve paralysis can be devastating for the patient. It not only has associated morbidity due to its cosmetic and functional consequence but there is also a significant psychosocial impact. Although many surgical options are available, the choice of treatment should take into consideration age and medical condition of the patient as well as both the duration and the type of paralysis.

In this paper, we discuss the general principles of facial nerve reanimation as well as static and dynamic procedures available to the patient.


Key words:
Facial reanimation, Reconstruction, facial paralysis.

Introduction:
A functioning facial nerve is not only important to providing conscious and subconscious control of the facial musculature but also provides ocular protection, nasal airflow, oral continence and clear articulation. Loss of facial function is not only devastating from a functional musculature but also provides ocular protection, nasal airflow, improve nasal airflow, prevent drooling and spontaneous facial movement. Choosing a dynamic technique (table 3) for facial reanimation depends on the duration of palsy and facial muscle condition.

Facial paralysis leads to weakness of the external nasal valve. Tarsorrhaphy (lateral or medial) narrows the palpebral fissure by approximating parts of the eyelids. It helps eyelid closure preventing dryness of the eye and exposure keratitis. It can be temporary or permanent.

2. Gold weights
Eye closure may be aided by insertion of a gold weight into the upper eyelid. Lid magnets, palpebral springs and platinum chains have also been described. These techniques are easy techniques for relief of exposure keratitis (Fig 1). The gold weight is inserted through an incision in the upper eyelid and sutured to the tarsal plate. Long-term extrusion can occur.

3. Brow lift
Paralysis of frontalis muscle causes ptotic/drop eye brows. Brow lift improves the aesthetic and function as it removes obstruction of upper visual field. Brow lift can be done endoscopic or open with minimal scars.

Static procedures:
Static procedures do not reproduce dynamic movement of the face however, they are used to provide corneal protection, improve nasal airflow, prevent drooling and provide facial symmetry

Dynamic procedures:
Dynamic procedures are utilized to improve facial symmetry and tone as well as providing learned and spontaneous movement. Choosing a dynamic technique (table 3) for facial reanimation depends on the duration of palsy and facial muscle condition.

Table 1: List of cause of facial palsy

<table>
<thead>
<tr>
<th>Causes</th>
<th>Example</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital</td>
<td>Mobius Syndrome, Goldenhar Syndrome, Facial nerve shwanoma</td>
</tr>
<tr>
<td>Inflammatory</td>
<td>Malignant otitis externa, Ramey Hunt syndrome, Acute and chronic otitis media, Deep neck space infection</td>
</tr>
<tr>
<td>Traumatic</td>
<td>Temporal bone fracture, Parotid injury</td>
</tr>
<tr>
<td>Irritative</td>
<td>Skull base surgery, Mastoectomy, and Parotidectomy</td>
</tr>
<tr>
<td>Metabolic</td>
<td>Diabetes Mellitus, Hyperlipidemia, Hypertension, Vitamin A Deficiency, Pregnancy</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Opular Syndrome, Millard-Gubler Syndrome, Guillain Barre</td>
</tr>
<tr>
<td>Hematologic</td>
<td>Leukemia</td>
</tr>
<tr>
<td>Toxic</td>
<td>Diphtheria, Tetanus, Arsenic Intoxication, Alcoholism</td>
</tr>
</tbody>
</table>

Table 2: Examples of static procedures

<table>
<thead>
<tr>
<th>Anatomical area</th>
<th>Procedure examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eye Brow</td>
<td>Brow Lifts</td>
</tr>
<tr>
<td>Eyelid</td>
<td>Tarsorrhaphy, Canthopexy, Gold weight</td>
</tr>
<tr>
<td>Nasal valves</td>
<td>Nasal valve surgery i.e. Alar button graft, suspension sutures</td>
</tr>
<tr>
<td>Oral commissure and lip</td>
<td>Static facial sling, Lip resection</td>
</tr>
</tbody>
</table>

Table 3: Dynamic facial reanimation procedures

<table>
<thead>
<tr>
<th>Procedures examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary nerve repair</td>
</tr>
<tr>
<td>Cable grafting</td>
</tr>
<tr>
<td>Cross-facial nerve grafting</td>
</tr>
<tr>
<td>Nerve transfers (jump grafts)</td>
</tr>
</tbody>
</table>

Static Procedures

1. Tarsorrhaphy
Tarsorrhaphy (lateral or medial) narrows the palpebral fissure by approximating parts of the eyelids. It helps eyelid closure preventing dryness of the eye and exposure keratitis. It can be temporary or permanent.

2. Gold weights
Eye closure may be aided by insertion of a gold weight into the upper eyelid. Lid magnets, palpebral springs and platinum chains have also been described. These procedures are easy techniques for relief of exposure keratitis (Fig 1). The gold weight is inserted through an incision in the upper eyelid and sutured to the tarsal plate. Long-term extrusion can occur.

3. Brow lift
Paralysis of frontalis muscle causes ptotic/drop eye brows. Brow lift improves the aesthetic and function as it removes obstruction of upper visual field. Brow lift can be done endoscopic or open with minimal scars.

4. Nasal valve Surgery
Facial paralysis leads to weakness of the external nasal valve. Multiple surgeries have been described, the most common are Batten grafts where a cartilage graft is inserted into a small pocket to support nasal cartilage. Suspension sutures are also used.
5. Static facial sling
Static sling is an excellent option in case of comorbidities that prevent active muscle transfer to maintain facial symmetry. Tensor fascia lata (TFL) is most commonly used with acceptable results. Synthetic materials as Gore-Tex/polytetrafluoroethylene and Alloderm have been described. The static slings are inserted between the corner of the mouth, ala of the nose or cheek and anchored onto the deep temporals fascia.

Dynamic Procedures
1. Facial nerve anastomosis and cable grafting
This is direct end-to-end repair of the facial nerve or the use of a cable graft to allow for a tension-free anastomosis. In a recent literature review, Barns et al. concluded that early repair before 2 months had the best results. However, grafting can occur anytime between 3 weeks up to a year after the injury. Therefore, there is no clear time frame or end plate regeneration and facial muscle fibrosis. After about a month, the proximal viable nerve starts to regenerate about 1 mm a day. Depending where the insult to the nerve occurred will determine the rate of recovery which can be up to 24 months.

The principles for repair include: finding the proximal and distal ends of the nerve under magnification, mobilizing them in order to facilitate a tension free repair (not > 2 mm risk of devascularization), freshening the edges, orientating the ends and using 8/0 suture material to suture the epineurium. In the case of intraoral injuries placing the two ends of the nerve together, surrounding it in fascia or without applying tissue glue will suffice. For all those defects that result in a significant distance between the proximal and distal nerve end then a cable graft using the great auricular nerve (GAN) or sural nerve (SN) can be used with minimal morbidity to the patient (GAN parasthesia angle mandible and lower 2/3 of ear; SN parasthesia outer aspect of foot). The most commonly used crossover technique is the hypoglossal-facial anastomosis due to proximity to the facial nerve, multiple nerve fibers, and accepted morbidity. The most commonly used crossover technique is the hypoglossal-facial anastomosis due to proximity to the facial nerve, multiple nerve fibers, and accepted morbidity. In the past, the hypoglossal-facial anastomosis was done by completely transecting the hypoglossal nerve and anastomosing it end to end with the distal facial nerve. However split hypoglossal transfer using 30% of the diameter of the hypoglossal nerve with an end to side anastomosis with the distal facial nerve can also be performed to prevent hemiglossal weakness. Similarly, a hypoglossal-jump graft can be performed (Figure 1). This is achieved by opening the hypoglossal nerve 30% and performing an end-to-side anastomosis with a mobilized distal facial nerve or a cable graft between the hypoglossal and distal facial nerve.

Recent the masseter branch from the trigeminal nerve has been used to anastomose the facial nerve with the buccal nerve on the functioning side. A tunnel is created between the face and the other end is anastomised onto the distal end of the facial nerve or a selected branch (usually buccal branch to allow for a spontaneous smile).

In a two-stage procedure, the distal end of the graft is placed in a pocket above the canine tooth. Nine to twelve months are allowed to pass until a positive Timel’s sign is elicited when tapping over the distal end of the graft indicating the presence of healthy neural fibers. The site is opened, the neuroma excised and an anastomosis is performed with the distal facial nerve trunk or a selected branch.

3. Nerve transfers
Nerve transfer is a reasonable choice in cases where there is irreversible proximal facial nerve injury with intact distal nerve and viable musculature detected on EMG. Ideally, the repair should be within 12 to 18 months to avoid muscle fiber atrophy. There are two situations where the transfer is indicated: First is in skull base surgery were the proximal stump is lost but the distal stump is intact and anastomosis or interposition is not achievable. The second is where the nerve is anatomically intact but with no evidence of function after 12 months. Advantages of this technique include a lower degree of technical difficulty, shorter time to movement (4 to 6 months), one suture line and reasonable motion with practice. However, this technique does not allow for spontaneous movement as with CFFNG.

4. Muscle transposition (dynamic slings) Regional muscle transfer has been described in the situation where the crossover techniques are not applicable such as congenital facial nerve absence as in Möbius Syndrome, cranial nerve sacrificed in skull base surgery or loss of motor end plates in long-standing facial palsy (> 2 years). For quick results or a low life expectancy the most reliable are the temporals muscle CT followed by the masseter muscle CT. By insertion of the muscle fibers onto the orbicularis oris, there is static support of the lower face and a trigeminal induced smile. This should be discussed with the patient as the muscle will need retraining, as it is associated with the clenching of teeth.

Also, it is important before planning surgery to be sure that the neurovascular supply to the muscle flap is intact as it may be affected by neurotological surgery of congenital disorders. Muscle transposition can also be used whilst waiting for natural facial nerve recovery following skull base surgery and cross facial nerve grafting.

5. Free muscle transfers
Free muscle transfer has been described as a reliable technique for active restoration of the facial symmetry and smile. According to the facial nerve status, it can be done in a second stage procedure once the axons have migrated across the graft. The latter will allow for dynamic facial motion. The use of the masseter and hypoglossal nerve will rely on retraining and eventual cerebral plasticity.

Figure 1: Hypoglossal-facial jump nerve anastomosis. This is the right side demonstrating a greater auricular nerve jump graft with an end-to-side anastomosis onto the hypoglossal nerve (partly obscured by the reattached digastric muscle) and an end-to-end anastomosis onto the facial nerve.

References

Both the cosmetic and functional loss following facial nerve weakness can have a significant effect on the quality of life of the patient. However through careful planning in a multidisciplinary setting, numerous surgical techniques can be explored in order to provide the patient with an achievable outcome.
Abstract

This article explores the concept of the multidisciplinary team (MDT) approach in the management of children with drooling. We discuss the management strategies such as behavioural, oral motor therapy, pharmacotherapy, Botulinum toxin, surgical treatment and the effective use of the MDT in this complex group of patients.

A multidisciplinary approach to the management of paediatric drooling

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Keywords

multidisciplinary team, paediatric drooling, pharmacotherapy, surgical management

Introduction

Drooling is generally defined as the involuntary loss of saliva from the mouth. As the daily production of saliva ranges between 1-1.5 L, drooling can lead to severe functional, social and clinical consequences for the children, their family and care providers. The physical and psychological complications of drooling include, but are not limited to skin maceration, aspiration and recurrent chest infections, hollistis, dehydration and social stigmatisaion. In the unstimulated state, it is estimated that approximately 70% of the total saliva produced is from the submandibular gland, with the parotid and sublingual glands producing 25% and 5% respectively. Conversely, in the stimulated state, saliva flow increases fivefold (7ml/min), with the parotid gland providing the majority of saliva. Drooling can be due to excessive production of saliva or poor control of saliva which may be due to poor head control, open mouth posture, disorganised tongue mobility, abnormal swallowing, macrognathia, dental malocclusion, nasal obstruction and decreased tactile sensation. In children with neurologic disorders, especially cerebral palsy, drooling appears to be mainly due to inefficient tongue and/or bulbar control, rather than increased saliva secretion. In children with normal neurology, drooling beyond four years of age is considered abnormal. In this group of children, it is worth considering nasopharyngeal or oropharyngeal obstruction and oromotor dyspraxia. Drooling in children is a complex problem and therefore requires a multidisciplinary team approach for effective management.

Multidisciplinary Team Approach

Paediatric drooling is a complex clinical problem which presents various challenges for both patients and healthcare providers alike and a multidisciplinary approach is often necessary. The team usually includes otoaryngologists, paediatricians, radiologists, speech and language and occupational therapists, paediatric dentists, nurses, teachers and social workers. Evaluation of these patients is undertaken in a multidisciplinary setting using various caregiver reported subjective qualitative scales such as the Teachers, Modified Teachers scoring and drooling severity and frequency scales. Currently, there is no specific scoring system for posterior drooling, however frequency of oropharyngeal suctioning and recurrent chest infections can be used as a tool for assessment. It is important to distinguish between anterior and posterior drooling as this impacts treatment options.

Management

As with many interventions in children, the evidence for non-medical treatment is scarce. Although there are no randomised controlled or controlled clinical trials to support the use of the various behavioural, physical and oral motor therapies for patients with drooling, there is increasing level 3 evidence to support the use of these interventions. Various programmes are currently being utilised in different centres and there is no particular evidence for or against any particular programme. Full assessment of the individual is often required to identify the needs of the child and the programme tailored to their specific needs. Good team work between occupational and
stimulation. These programmes can be laborious and include exercises to improve oral control and programme, to target the jaw, lips and tongue movements in improving symptoms in children12.

Prosthetics and Oral Care

A number of prostheses have been developed by dentists and speech and language therapists to help with jaw stability and lip seal. These devices include the chin cup developed by orthodontists to decrease jaw protrusion13. There are children who genuinely produce too much saliva and the causes of this may include habitual finger chewing and dental decay, hence appropriate oral care and dental evaluation may be necessary in some cases. Functional appliances using the principles of Castillo-Morales; consisting of an acrylic palatal plate with vestibular and lingual stimulators which induce sucking and subsequent tongue retraction, have been used successfully to manage drooling14,15.

Pharmacotherapy

The most frequently studied drugs in the treatment of drooling have been those that inhibit the secretion of saliva, typically anticholinergics. A systematic review described the benefits of anticholinergics in drooling16.

A review undertaken by the National Institute for Health and Care Excellence (NICE) in 2013, showed moderate evidence for oral glycopyronium bromide in reducing hypersalivation in children with neurological conditions when compared with placebo, however there is no evidence of its long term efficacy or safety in treating hypersalivation. Although oral glycopyronium bromide is unlicensed in the UK, NICE supports its use where there are good clinical indications for its use17. Studies have shown 70-90% response rates, but with a high rate of antimuscarinic side effects in approximately 30-35% of patients, hence their guardians may prefer to discontinue the medication due to dry mouth, excessive sweating, urinary retention, irritability and behavioural changes18.

Hycosine provides good results in the treatment of paediatric drooling, with the advantage of a single transdermal application and is believed to render adequate serum concentrations, lasting three days. This offers a good treatment option, especially in children with neurological impairment. Side effects include skin reactions, dry mouth, constipation, blurring of vision, behavioural abnormalities and mild sedation19,20.

Trihexyphenidyl has been used in the treatment of patients with cerebral palsy, as it offers the additional benefit of treating dystonia as well as drooling. Studies have shown significant improvement with some sources quoting improvements of up to 90% in patients with cerebral palsy. However acute/chronic colonic obstruction in patients using this therapy has been reported21.

Botulinum Toxin

A Cochrane review by Walsh et al (2012) found six randomised controlled trials using either Botulinum toxin (Btx-A) or oral medications to decrease drooling22. All studies demonstrated effectiveness up to one month’s duration for both treatments23. Another systematic review by Rodwell et al (2012) found sixteen articles on the use of Btx-A for paediatric drooling and also concluded that Btx-A was an effective but temporary therapy24. Of importance there have been mixed results, with some studies showing no improvement in over 30% of patients using Btx-A25.

The dose of Btx-A administered for injection varies widely, due to variations in practice at different centres. The recommended dose in children is 30-40units of Btx-A, which is injected into the parotid and submandibular glands, and is usually enough to control symptoms26. Although several studies have suggested that ultrasound guidance is not required, it is generally recommended that injections are given under ultrasound guidance to ensure injection into the salivary gland and avoid injury to nerves, muscles and vessels. The main complications of Btx-A treatment are dysphagia (due to diffusion into adjacent bulbar musculature), weak mastication, infection, facial nerve injury and dental caries27.

Surgical Management

Surgery is the treatment of choice in patients with severe drooling unresponsive to medical management. Surgery is generally deferred until 5-6 years of age. However it may be considered earlier in cases of severe posterior drooling with recurrent chest infections and intensive care unit admissions. There are several surgical procedures described in the treatment of drooling. These include, those aimed at reducing the amount of saliva (tympanic neurectomy, submandibular and parotid duct ligation, submandibular gland excision and sublingual gland excision) and those that redirect the flow of saliva (submandibular and parotid duct transposition), or a combination of these procedures. High overall subjective success rates for surgical options (81.6%) have been
In a child with no neurological impairment and no neuromuscular pathology and adenotonsillar hypertrophy, adenotonsillectomy may improve drooling.

In our experience bilateral submandibular gland excision and single parotid duct ligation is the preferred surgical treatment option, with optimal outcomes, in paediatric drooling unresponsive to a medical management in children with neurological and neuromuscular conditions.

Radiotheraphy

The use of radiotherapy as a treatment modality in drooling has only been described in adult patients with Amyotrophic Lateral Sclerosis and Parkinson’s disease. Oncologists are understandably cautious of administering radiotherapy for drooling in children due to the long-term risk of malignancy.4,14

Conclusion

A multidisciplinary multiprofessional team based approach together with guardian involvement is essential, in order to fully evaluate and institute the best management approach, with the best outcomes, in children with drooling. Various treatment strategies are described. In our experience when drooling is refractory to conservative measures and pharmacotherapy in children with neurological and neuromuscular conditions, bilateral submandibular gland excision and single parotid duct ligation is the preferred surgical option.

References


To date there is no specific procedure that is regarded as the gold standard for drooling. Compounding this issue is the difficulty in evaluating the surgical efficacy of these procedures and demonstrating their success. Our specialised tertiary multidisciplinary drooling clinic centre has noted recurrence of symptoms and ranula formation in children with salivary duct ligation; therefore this is not the preferred surgical modality for drooling in our centre.
Extra-uterine intrapartum treatment (EXIT) and upper airway obstruction of the newborn

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Abstract
Improvements in the antenatal diagnosis of congenital malformations have led to increased detection of foetal airway obstructing lesions, and paediatric ear, nose and throat (ENT) surgeons are increasingly involved in these cases. Traditionally, difficulty in obtaining a patent airway at delivery was a major factor in the dismal prognosis of these pregnancies. The EXIT procedure, which involves controlling partial delivery of the foetus whilst maintaining placental circulation, allows various airway manoeuvres to be performed to secure the airway in a controlled fashion. This article outlines the typical range of pathology seen, the logistics in providing support for anticipated deliveries and the multidisciplinary management of complex airway cases.


Key words
paediatric airway; EXIT; foetal; CHAOS

Introduction
The ex utero intrapartum treatment (EXIT) procedure is a multidisciplinary management plan that can be deployed following the prenatal diagnosis of foetal, and impending neonatal, airway obstruction. It allows for time at delivery for airway securement and other resuscitative measures whilst remaining on placental oxygen support, thereby converting an emergency neonatal airway situation into an elective controlled one. A successful EXIT outcome is heavily dependent on prenatal investigation techniques and a sufficiently high index of suspicion to detect potentially compromised infants. In addition to a surgical team with fetal medicine, ENT and anaesthetic neonatal airway expertise, highly specialist obstetric anaesthetic and surgical skill sets are required to ensure maternal safety and placental sufficiency throughout the procedure.

Indications for EXIT
Foetuses with airway obstruction may have one of a number of underlying defects affecting the head and neck. Obstructive lesions can be intrinsic, i.e. the so-called CHAOS infant (Congenital High Upper Airway Obstruction Syndrome), but are more commonly caused by extrinsic compression by pharyngeal, cervical or thoracic mass lesions. In principle and in practice, the EXIT procedure can be considered in any prenatally diagnosed situation where difficulty or compromise in neonatal cardiopulmonary resuscitation is anticipated, such as reversal of tracheal occlusion in congenital diaphragmatic hernia, pulmonary pathologies such as congenital cystic adenomatoid malformations or unilateral pulmonary agenesis, pre-separation of conjoined twins and as a bridge to extracorporeal membrane oxygenation (ECMO). Extrinsic lesions

Extrinsic lesions
Patient series at large tertiary treatment centres have demonstrated that the location of an extrinsic lesion is a more relevant factor in the development of airway distortion and compromise more than the absolute size of the mass. Lesions are likely to be isolated anomalies (Table 1), so securement of the airway at birth is key in allowing postnatal surgical treatment and good long-term outcomes. Benign lymphatic cysts commonly affect the neck (as cystic hygromas), likely following failure of the jugular lymph sacs to connect with lymph channels. Their overall UK incidence may be as high as 1 in 1000 pregnancies, making them the second most common type of neonatal soft tissue lesion after haemangiomas. Given their highly variable morphology, there is a wide spectrum of clinical severity. Posterior triangle lymphangiomas that are diagnosed early in the first or early-second trimester are more likely to be associated with underlying chromosomal defects and a poor pregnancy outcome. Anterior lesions, especially those diagnosed later in gestation, are not usually associated with other abnormalities and may even regress in utero. If the foetus is of a normal karyotype, without hydrops, and no sepsis are visible within the mass, then a good prognosis is expected and the main risk is that of perinatal airway compromise.

Head and neck teratoma
Teratomas are ectopic tissue neoplasms, far removed from their normal anatomical site, which contain elements from all three germ layers (the most common types are neural, cartilage and thyroid tissue). The head and neck is the fourth most common site for congenital teratoma. Tumours may be extensive, sometimes spanning from oral floor and mandible into the anterior mediastinum inferiorly or trapezius posteriorly, and can present with polyhydramnios from impaired foetal swallowing and oesophageal compression (Figure 1). Even in the presence of lymph node metastases, total tumour removal is achieved in the great majority of cases with excellent long-term follow-up results.

Intrinsic lesions
Most cases of CHAOS are due to laryngeal atresia, but can also be caused by obstructing laryngeal webs or cysts, subglottic stenosis or tracheal stenosis (Figure 1). Embryologically, the larynx and upper airway becomes occluded at around 10 weeks gestation with an epithelial membrane, which prevents the escape of accumulating lung fluid from the tracheobronchial tree. The use of the EXIT procedure to deliver these children is usually required to secure a tracheostomy at birth.

The sequence by which CHAOS worsens prognosis is by backpressure of accumulating lung fluid. This congestion of the foetal lungs causes compression of the heart within the mediastinum, impairing cardiac filling and leading to secondary heart failure and ascites. If the obstruction to lung fluid drainage is left for the duration of gestation, the lungs remain poorly developed and inefficient in gas exchange at birth. In the case of tracheal atresia, cardiopulmonary sequelae may spontaneously resolve over the course of the pregnancy if coexistent tracheoesophageal fistulae are present, which allow for lung

Table 1: Extrinsic Causes of Foetal Airway Obstruction. Adapted from Liechty & Crombleholme, 1999

<table>
<thead>
<tr>
<th>Cause</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cervical lymphangioma (lymph)</td>
<td>Lipoma</td>
</tr>
<tr>
<td>Cervical teratoma</td>
<td>Haemangiomata</td>
</tr>
<tr>
<td>Congenital goitre</td>
<td>Neuroblastoma</td>
</tr>
<tr>
<td>Haemangiomata</td>
<td>Parotid masses</td>
</tr>
<tr>
<td>Branchial cleft cyst</td>
<td>Nuchal oedema</td>
</tr>
<tr>
<td>Thyroglossal duct cyst</td>
<td>Chromitoma</td>
</tr>
<tr>
<td>Congenital thyroid tumour</td>
<td>Neural tube defects (e.g. cervical lymphangioma)</td>
</tr>
<tr>
<td>Laryngocele</td>
<td>Twin sac of blighted ovum</td>
</tr>
</tbody>
</table>

Figure 1: (A) Microlaryngoscopy demonstrating a laryngeal web with the endotracheal tube (ETT) intubated through a tracheoesophageal fistula. (B) Typical appearance of a neonate with a large cervical teratoma.
reported in several CHAOS cases following EXIT.30
manifestations, excellent long-term survival has been
in cases of isolated anomalies or otherwise mild syndromic
as Fraser Syndrome24,28 or Fragile X Syndrome29. This is a
sporadic isolated lesions, but may be associated with a
Most intrinsic airway anomalies leading to CHAOS are
atresia without CHAOS27.
the developing trachea and pharynx, potentially explaining
A suspected diagnosis of upper airway obstruction is
was born with laryngeal atresia without CHAOS30.
In cases of isolated anomalies or otherwise mild syndromic
manifestations, excellent long-term survival has been
reported in several CHAOS cases following EXIT.30
Prenatal assessment and diagnosis
A suspected diagnosis of upper-airway obstruction is typically made during standard prenatal ultrasonography
Screening can be visualized as early as 14 weeks gestation and is suggestive of a continuous aero-
digestive tract. As amniotic fluid is normally reabsorbed
via the gastrointestinal tract, any lesion that compresses the
oropharyngeal tract can lead to polyhydramnios due to
a partial or complete failure of foetal swallowing. In some
case series, 20% of pregnancies with polyhydramnios
were found to have an underlying head and neck abnormality41.
The foetal head may be held in an opisthotonic position,
even if the lesion is quite extensive. The classical
CHAOS US features are of large hypoechoic lungs,
distention of the trachea and bronchi, flattening or eversion
of the diaphragm, restriction to cardiac filling,
placentomegaly and non-immune hydrops fetalis30. In
severe cases, mothers can mirror this heart failure41.
Improved US resolution has meant these lesions can be
diagnosed by single modality imaging alone, and three-
dimensional reconstructions can provide more precise
anatomical information and extent of disease. Doppler is a
useful adjunct to provide information on the vascularity of
the lesion and clues to the degree of foetal heart strain
from umbilical cord blood flow patterns.
Foetal magnetic resonance imaging (MRI) has an
increasing role in defining exact sites of anticipated
obstruction, for confirmation of US features and for
follow-up monitoring of foetal lung volume30. Foetal
MRI is also excellent for diagnostic confirmation in
equivocal cases, e.g. to differentiate between herniation of
posterior fossa contents and posterior cleft hygroma31,
and in identifying syndromic features, without resorting to
invasive foetoscopy (Figure 2). It is also highly useful for
parent counseling and aids in delivery planning32, and is
an extremely useful tool for surgical planning where foetal
surgery may be under consideration33. Other specialised
tests include amniocentesis for karyotyping, and
to directly visualise the airway.
Prenatal management
Prenatal management is often conservative by monitoring the
pregnancy, such that foetos can be delivered when fully
developed. EXIT procedures are typically considered at a
gestational age of around 38 weeks. If there is a degree of
polyhydramnios, amniotamges may be required at certain
stages throughout the pregnancy. Direct management of the
obstructing lesion in utero is very limited; however foetoscopic
or laser techniques to relieve obstructive pressures are
reported in the literature with increasing frequency41.
Successful decompression is seen as an immediate decrease
in tracheal diameter on ultrasound, followed by normalisation
of cardiopulmonary parameters over the next few hours30. These
features should be monitored throughout the remainder of
the pregnancy, however; as punctures can close over time
necessitating repeat foetoscopic treatment or early delivery34.
Foetoscopic is also useful for confirming the presence
or absence of phenotypic features of other syndromic features if
ultrasound findings are inconclusive (such as in cases of large
maternal baby hydramnios)31.
Management
Following initial diagnosis, onward referral is
recommended to a designated tertiary care centre where
a multidisciplinary team (MDT) encompassing foetal,
obstetric and ENT expertise can further assess
and own scrub team and a paediatric airway team
that includes an airway anaesthetist and assistants.
ENT surgeons and dedicated scrub staff. Foetal medicine
specialists monitor foetal physiology during the procedure
and neonatologists are present for management of the
child after delivery.
To EXIT or not to EXIT
The MDT is essential in this decision making process, as
the risk to mother and foetus must both be considered
along with the child’s likely long-term outcome. In our
unit, all cases have assessment by foetal US and MRI.
Genetic analysis is performed as required. The severity of
associated anomalies is carefully evaluated and, if very
severe, may lead to a decision towards terminating the
pregnancy. Conditions such as Fraser Syndrome require
sensitive and in-depth counseling of the parents, who may
decline to continue with the pregnancy given the wide
spectrum of clinical severity in affected Fraser infants45.
Maternal medical conditions (e.g. pre-eclampsia) may
necessitate emergent delivery of the infant. Our unit
currently does not advocate an emergency EXIT procedure
in these situations, as considerable personnel and expertise
are required to be available. In these situations, the baby is
delivered through an emergency caesarean section and
standard neonatal resuscitative measures are performed.
When considering an EXIT procedure, the team has to
predict the likelihood of securing airway access. Much
information can be garnered from imaging alone, such as
whether the lesion is seen infiltrating the tongue base or is
particularly vascular in nature. Other anticipated
considerations beyond the immediate delivery of the
infant may need addressing, such as immediate cardiac
anomalies and/or early gastrointestinal and genitourinary malformation management. Early surgical
intervention to remove the lesion post-delivery may be
necessary to ensure continued control of the airway,
however, cardiac issues may take equal priority and require
correction in a combined procedure. A further consideration
of persistent in utero airway obstruction is the possibility
of an undervenous respiratory tree. The neonate may
therefore need prolonged respiratory support following
delivery.

 Theatre Procedure
Theatre set-up requires coordinated action by multiple
personnel from various disciplines. As there are ‘two
patients’ to consider in this procedure, surgical teams are
divided into those principally addressing the mother and
those addressing the child. Similar to other units that
deliver EXIT procedures, our team consists of an obstetric
team with two surgeons with their dedicated anaesthetists
and own scrub team and a paediatric airway team that
includes an airway anaesthetist and assistants. ENT
surgeons and dedicated scrub staff. Foetal medicine
specialists monitor foetal physiology during the procedure
and neonatologists are present for management of the
child after delivery.

The surgical approach to EXIT is similar to a standard
caesarean delivery but differs in the anaesthesia, as
ureteric hypotonia is the priority to prevent placental
transfusion. There is a low threshold to terminate the
EXIT procedure if the mother is at risk from haemorrhage
the child is then delivered before securing the airway.
Other considerations include the ‘lie’ of the placenta
where an anterior position may limit the incision (and thus
the position for the delivery of the foetal head). On
delivery, the head is rotated in an occipito-
position (Figure 3). The arm is also delivered with preservation of as much amnion as possible to limit amniotic fluid loss. Foetal continual monitoring equipment is reattached, which includes an oxygen saturation monitor and a pH scalp probe. An echocardiogram (ECHO) machine is made available but is not routinely used unless the other sensors fail. Foetal anaesthetic medications are immediately delivered which include intramuscular injection of Vecuronium (0.1mg/kg) and Fentanyl (10mcg/kg) into the deltoid.

We use the following algorithm for securing the airway in our centre (Figure 4). Following delivery of the head, the child can be immediately evaluated by direct laryngoscopy with an anaesthetic intubating laryngoscope (Miller 0 or 00 blades). Reliable suction needs to be on standby to evacuate thick secretions. A Benjamin operating laryngoscope can be used next if the above fails, followed by rigid bronchoscopy. Large masses can be retracted away from the airway to aid intubation. Standard intubating adjuncts (stylettes and bougies) should be deployed to aid intubation. Rigid bronchoscopy is attempted prior to front of neck access. We have found a y-splitting lead is particularly helpful for widening the scopes. Ultimately, if all of the above attempts fail, a tracheostomy is performed and, if lesions are infiltrating the front of the neck, a major neck tray is required.

Potential Complications
Foetal complications may occur if placental gas exchange cannot be preserved. This can occur secondary to loss of uterine relaxation, placental abruption or cord compression. Umbilical cord vessels are also at risk of vasospasm, particularly if exposed to changes in temperature. Inhalational anaesthetic agents are essential for uterine relaxation, but may lead to uterine atony. This can cause severe bleeding and could theoretically require hysterectomy to control bleeding in severe cases, though none have been reported in the literature or in our experience. Close surgical-anaesthetic communication is required to allow restoration of uterine tone as soon as the umbilical cord has been clamped. Uterine rupture may occur in subsequent pregnancies if vaginal delivery is attempted in the future, so mothers should be counselled during the EXIT decision-making process about the high likelihood of caesarean sections being required for all subsequent pregnancies. Lower uterine incisions may lower this risk but the feasibility of this is dependent on placental position6.

The Role of Foetal Surgery
Open foetal surgical techniques are becoming increasingly reported in several US centres for the management of time-sensitive life-threatening thoracic foetal conditions. The maternal risks surrounding open foetal surgery must be weighed in ethical terms against the likelihood of obtaining a more favourable foetal outcome than those obtained by delivery via EXIT and subsequent postnatal surgery (with or without foetoscopic interventions). The severe sequelae of CHAOS that could be avoided with early intervention, such as cardiac failure and impaired gas exchange development, may render foetal surgery ethically favourable but must be balanced against increased maternal risks and the likely presence of other foetal abnormalities, many of which are difficult to diagnose prior to birth even with the deployment of foetal MRI and foetoscopy. Open surgery would be of particular use in long-segment stenoses which are not amenable to foetoscopic treatment.

Conclusion
Foetal upper airway obstruction has been transformed with the use of the EXIT procedure, as congenital malformations that were previously fatal to the foetus at delivery can now be managed with a semi-elective procedure that allows time for securing the airway. Essential to this is a multidisciplinary team that can provide expertise from diverse disciplines to deliver specialised and individualised care to both mother and child. Paediatric ENT surgeons should have an awareness of the pathologies and management strategies, even if not directly involved with the EXIT procedure itself, to aid the long-term local management of these children.
The management of paediatric facial nerve palsy

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Abstract

The facial nerve is a mixed nerve containing motor, sensory and autonomic fibres. Facial nerve palsies may be upper motor neurone (UMN) or lower motor neurone (LMN), unilateral or bilateral. Incidence rates vary between 6.1-25 children per 100,000 per year. There are numerous causes both congenital and acquired, which vary from idiopathic to serious underlying pathology such as tumours. In approximately 50% of cases, the cause of the facial nerve palsy remains unknown. It is essential that a thorough history and appropriate investigations are undertaken to identify potentially treatable causes and minimise morbidity. The management of facial nerve palsy in the paediatric age group may prove challenging for children, caregivers and clinicians. The prognosis of paediatric facial nerve palsy depends on the aetiology, severity, and time of presentation. In this article, we suggest a strategy for assessment, investigation and management of paediatric facial nerve palsy with a multidisciplinary team approach.


Key words

Facial palsy, paediatric, congenital, management

Acknowledgements

We would like to acknowledge Mrs Hasnna Ismail-Koch, who greatly contributed to the text, editing, clinical cases, images and development of the management approach described. We would also like to recognise the valued contributions of Mr Tim Mitchell and Mr Sebastian Thomas for their expert advice on the management of paediatric nerve palsy.

Introduction

The facial nerve is a mixed nerve containing motor, sensory and autonomic fibres. It is the nerve of the second branchial arch, which supplies structures derived from Reichezt’s cartilage (table 1) and is responsible for essential functions including hearing, chewing, facial expressions, and facial movements. Approximately 58% of the facial nerve fibres in humans are motor, 18% sensory, and 24% autonomic.

The facial nerve arises from the facioacoustic primordium, which appears in the third week of gestation. The facial and acoustic primordium separate from each other by the end of the sixth week. By week 16 all communications of the facial nerve are established and by week 26 the formation of the fallopian canal is complete. At this gestation approximately 25-55% of fallopian canals show dehiscence, the commonest location being around the oval window. Incidence rates vary from 6.1-25 children per 100,000 per year.

Table 1: Derivatives of the second pharyngeal arch

<table>
<thead>
<tr>
<th>Pharyngeal Arch</th>
<th>Artery</th>
<th>Artery and Artery</th>
<th>Muscle</th>
<th>Derivative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reichert’s cartilage/ second pharyngeal arch</td>
<td>Facial nerve</td>
<td>Facial expression muscles</td>
<td>Maleus (mandibulum)</td>
<td></td>
</tr>
<tr>
<td>Cartilage</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Buccinator</td>
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<td></td>
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<tr>
<td>Stapedial Artery</td>
<td>Stapedius muscle</td>
<td>Stapes (except for footplate)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Stapedius muscle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pharyngeal Artery</td>
<td>Stylopharyngeal muscle</td>
<td>Facial canal</td>
<td></td>
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<tr>
<td>Pharyngeal Artery</td>
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<tr>
<td>Palatogastric</td>
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<tr>
<td>Posterior belly of digastric</td>
<td>Stylohyoid muscle</td>
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<td></td>
<td></td>
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<td></td>
<td></td>
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<tr>
<td>Stylohyoid ligament</td>
<td>Hyroid bone (lesser cornu &amp; upper part of body)</td>
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</tr>
</tbody>
</table>
**Epidemiology and causes**

Paediatric facial nerve palsy (partial weakness) or paralysis (complete weakness) refers to weakness of the facial nerve either present from birth (congenital) or developing after birth (acquired). Reported incidence rates vary between 6.1-25 children per 100,000 per year. Facial nerve palsies may be classified as upper motor neuron (UMN) or lower motor neuron (LMN), unilateral or bilateral. Facial nerve palsies may be isolated or occur as part of a syndrome, in conjunction with other cranial nerve palsies or other abnormalities, suggesting a developmental aetiology. In cases of congenital facial nerve palsy associated with congenital malformations, understanding the abnormality of the malformed organ or structures based on embryological development can indicate the fetal stage at which the insult occurred and development was arrested.

There are numerous causes of paediatric facial nerve palsy both acquired and congenital. In approximately 50% of cases, the cause of the LMN facial nerve palsy remains unknown, and these patients are collectively grouped under the term ‘Bells palsy’ much like their adult counterparts. The commonest cause of congenital facial nerve palsy accounting for 75-80% of the cases is birth trauma. Factors predisposing to birth trauma induced facial nerve palsy include prematurity, birth weight >3500 grams, forceps delivery, caesarean delivery and first child. Full recovery of facial nerve function occurs in the vast majority of these patients within a few months. Other congenital causes include a variety of genetic syndromes and congenital malformations (table 2).

There are many acquired causes of facial palsy and presentation depends on the underlying aetiology (table 3). Acquired facial nerve palsies, in general, have a relatively good prognosis compared to syndromic causes.

**DIAGNOSIS**

**History**

A thorough history should include the onset and progression of the facial nerve palsy. Specific questioning regarding associated otorhinological symptoms such as hearing loss, otorrhea, facial numbness or hyperacusis should be undertaken. Additionally, history of recent infections, tick bites, trauma, neck lumps, salivary gland pathology, weight loss and systemic symptoms should be sought.

**Clinical Examination**

It is essential that clinical examination includes general systemic aspects, assessment for syndromic features, complete neurological examination and examination of the cranial nerves to ascertain whether the facial nerve palsy is upper or lower motor neuron (figure 1). The severity of the LMN facial nerve palsy should be assessed and recorded according to the House Brackmann grading system (table 4). Other commonly used systems are the Sunnybrook and Yanagihara scales.

Systemic examination, for signs of potential underlying disease, should include checking for weight loss, bruises and organomegaly (oncological causes), tick bites and rashes (Lyme disease), and blood pressure measurement (by hypertension) which has been reported to be associated with recurrent facial palsy.

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Table 3: Acquired causes of Paediatric facial palsy

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bells palsy</td>
<td>Lumbar deformities aplasia of brachial and thoracic muscles, unilateral or bilateral facial nerve palsies</td>
</tr>
<tr>
<td>Goldenhar syndrome</td>
<td>Unilateral facial asymmetry, maxillary hypoplasia, microtia, microtia, facial nerve palsy</td>
</tr>
<tr>
<td>Moebius syndrome</td>
<td>Leptomeningeal aplasia of the cranial nerves, facial nerve palsies</td>
</tr>
<tr>
<td>Mekerson-Rosenthal syndrome</td>
<td>Recurrent facial nerve palsy, fissured tongue, oedema off the eyes and chin</td>
</tr>
<tr>
<td>Neurofibromatosis type 1 Von Recklinghausen’s disease</td>
<td>Autosomal dominant disorder, multiple neurofibromas, cafe-au-lait spots, vestibular schwannomas, facial nerve schwannomas (leading to facial nerve palsy)</td>
</tr>
<tr>
<td>Neurofibromatosis type 2</td>
<td>Autosomal recessive disorder, tubular muscle involvement, unilateral or bilateral partial facial nerve palsy, progressive sensorimotor hearing loss</td>
</tr>
<tr>
<td>Osteopetrosis</td>
<td>Deafness, abnormalities of the ear, facial nerve palsies</td>
</tr>
<tr>
<td>Sclerostenosis</td>
<td>Autosomal recessive disorder, mandibular, calvarial, clavicular, and pelvic osteosclerosis and hyperostosis, bilateral hearing loss, unilateral and bilateral facial nerve palsies</td>
</tr>
<tr>
<td>Dominant craniofacial dysplasia</td>
<td>Metaphyseal widening of limbs, unilateral or bilateral facial nerve palsies, hearing loss, scrotic mastoid, facial and bony skull growth</td>
</tr>
<tr>
<td>Recessive craniofacial dysplasia</td>
<td>Ophthalmoplegia, craniosynostosis, craniofacial malformations, facial nerve palsies, progressive hearing loss, blindness</td>
</tr>
<tr>
<td>Genetic or Hereditary</td>
<td>Pterygium, microphthalmia, micrognathia, microtia, hypoesthesia, hypogonadism, anhidrosis, anhidrosis, preauricular pits</td>
</tr>
<tr>
<td>Hereditary myopathies</td>
<td>3q21-22 mutation</td>
</tr>
<tr>
<td>10q21.3-22.1 mutation</td>
<td>10q21.3-22.1 mutation</td>
</tr>
<tr>
<td>Non-Syndromic</td>
<td>Birth Trauma</td>
</tr>
<tr>
<td>Birth Trauma</td>
<td>Prematurity, birth weight &gt;3500 grams, forceps delivery, caesarean delivery, first child</td>
</tr>
<tr>
<td>Sickle cell anemia</td>
<td>Diphtheria, Acute Otitis Media (Pneumococcus, H. influenza, meningococcus)</td>
</tr>
<tr>
<td>Poliomyelitis</td>
<td>Parotid, Tonsillitis, Sarcoidosis, Nodular (Pneumococcus, H. influenza, meningococcus)</td>
</tr>
<tr>
<td>Viral</td>
<td>Paralytic Facial Palsy, Ramsay Hunt Syndrome, Chronic Zosteropathy, Poliomyelitis, Bell’s Palsy</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Multiple sclerosis, Myasthenia gravis, Cystic fibrosis, Diabetes Mellitus</td>
</tr>
<tr>
<td>Metabolic</td>
<td>Diabetes Mellitus, Hypothyroidism, Acute Porphyria, Wilson’s disease</td>
</tr>
<tr>
<td>Drug/Toxin</td>
<td>Tricyclic antidepressants, Anticoagulants, Antihypertensive, Antiepileptic</td>
</tr>
<tr>
<td>Idiopathic/ Bells palsy</td>
<td>Prematurity, birth weight &gt;3500 grams, forceps delivery, caesarean delivery, first child</td>
</tr>
</tbody>
</table>

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**Figure 1:** Child with right LMN facial palsy.

**Figure 2:** Right acute otitis media.
A detailed head and neck examination should be performed to exclude parotid tumours or lesions. Thorough examination of the ear should be performed looking for any discharge, vesicles and/or granulations of the ear canal and to rule out any middle ear pathology including acute otitis media (figure 2), cholesteatoma or glomus tumours. A triad of ipsilateral facial nerve paralysis, auricular pain, and vesicles in the external auditory canal and on the pinna is typical for Ramsay Hunt Syndrome Type II19,20. Full cranial nerve examination should be performed to identify a central cause of the facial nerve palsy. Further examination of the peripheral nervous system may also be required. Bilateral facial nerve palsy suggests neurological causes such as multiple sclerosis and Guillain-Barre syndrome or infective causes such as Lyme disease21.

**Investigations**

The choice of investigations depends on the potential cause, associated symptoms, and mode of presentation. A full blood count and film should be undertaken to rule out rare malignant causes such as leukaemia. Lyme disease serology testing for Borrelia Burgdorferi is of great importance especially in areas with a high prevalence22. In the presence of otological symptoms and/or signs, a pure tone audiogram and tympanogram are required to identify the nature and severity of any associated hearing loss13,14. Children with normal otological findings and without any hearing loss may not need further audiological investigation.

**Imaging**

Radiological investigation is indicated in all patients with upper motor neurone signs as part of the initial workup21. These children should be referred urgently to a paediatric neurologist for further investigation. In those with lower motor neurone lesions indications for imaging include; a progressive facial nerve palsy over one month, no signs of recovery after six weeks, recurrent or bilateral facial nerve palsies and involvement of other cranial nerves12,13,14,15.

If a child presents with an asymmetric hearing loss or positive ear signs, then imaging should be performed. Magnetic resonance imaging (MRI) (figure 3) is the investigation of choice to rule out any neurological cause or a vestibular schwannoma (acoustic neuroma). Computerised Tomography scanning (CT) (figures 4, 5 & 6) may be required when bony erosion or chronic supplicative otitis media is present.

**Electrophysiological Testing**

Electrophysiological testing may prove useful, but interpretation very much depends on the operator23,24. Electrophysiological tests employed are nerve excitability testing, maximal stimulation testing, electromyography and electroneurography. These tests are carried out at specialised centres and are useful in determining the prognosis of the facial nerve palsy.

If there is uncertainty in diagnosis urgent multidisciplinary input is suggested including otorhinolaryngology, paediatric (general, neurology, paediatric infectious diseases and immunology), ophthalmology and radiology.

Bell’s palsy is a diagnosis of exclusion, once all possible causes have been ruled out. The prevalence of Bell’s palsy in children varies between 9-50%25.

**Management**

The management of paediatric facial nerve palsy depends on the aetiology. A complete facial nerve palsy at presentation correlates with a poor outcome when compared to patients presenting with a partial palsy. Treatment is directed towards the management of identifiable causes.

In the paediatric age group, treatment strategies should employ a multidisciplinary team approach, including specialists from otorhinolaryngology, paediatrics, neurology, ophthalmology, radiology, oromaxillofacial surgery, facial plastic subspecialist surgery, speech and language therapy and physiotherapy. Management comprises general, medical, and surgical treatment.

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**Table 4: House-Brackmann facial nerve grading system**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Characteristics</th>
<th>Gross</th>
<th>Forehead</th>
<th>Eye</th>
<th>Mouth</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Normal function</td>
<td>Slight weakness</td>
<td>Moderate to good function</td>
<td>Complete closure without effort</td>
<td>Slight weakness on maximal effort</td>
</tr>
<tr>
<td>II</td>
<td>Obvious weakness but not disfiguring</td>
<td>Moderate to good function</td>
<td>Complete closure without effort</td>
<td>Slight weakness on maximal effort</td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>Obvious weakness</td>
<td>Slight to moderate function</td>
<td>Complete closure with effort</td>
<td>Asymmetry on maximal effort</td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>Obvious weakness</td>
<td>No function</td>
<td>Incomplete closure</td>
<td>Asymmetry on maximal effort</td>
<td></td>
</tr>
<tr>
<td>V</td>
<td>Barely perceptible motion</td>
<td>No function</td>
<td>Incomplete closure</td>
<td>Slight movement</td>
<td></td>
</tr>
<tr>
<td>VI</td>
<td>No function</td>
<td>Normal symmetry at rest</td>
<td>Complete closure</td>
<td>Normal symmetry at rest</td>
<td>Complete closure without effort</td>
</tr>
</tbody>
</table>

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**Figure 4:** CT of a patient who presented with a right LMN facial nerve palsy showing a cholesteatoma with a discoloured facial nerve bony canal (arrow).
LOWER MOTOR NEURON FACIAL NERVE PALSY

**Routine Care**
- History, Examination & BP
- FBC, film & Lyme Serology

**Eye care is the priority in all cases** — use Hypromellose/Lacrifilm + tape at night if incomplete eye closure.

**Antibiotics** — Oral Amoxicillin 15-20mg/kg (max 500mg) tds for 14 days.
- If penicillin allergic:
  - < 12 years — Azithromycin 10mg/kg (max 500mg) od 3 days/week for 2 weeks
  - > 12 years — 100mg bd for 2 weeks

**Stereoids** — If symptomatic for < 72 hours prescribe Prednisolone 1mg/kg (max 40mg) for 10 days.

- **Red Flags**
  - Forehead sparing and/or other abnormal neurological findings
  - Acute otitis media, effusion, hearing loss, vertigo, ear discharge, vesicles
  - Parotid mass
  - Bilateral palsy
  - Severe pain
  - Bruising or oedema
  - Hypertension

- **Consider**
  - Ophthalmology — essential if eye closure is impaired
  - ENT — Refer any child < 3 years with ear symptoms and if there are red flags in all age groups
  - Neurology — if focal or evolving neurological signs
  - Speech & Language Therapy — if concerns about communication or swallowing
  - Physiotherapy — if no recovery at 6 weeks. See guideline for details.

- **Follow-up**
  - Arrange review in 1 week. Stop antibiotics if Lyme serology negative.
  - If progression of symptoms consider underlying diagnosis and need for referral.
  - Arrange further review in 3-6 weeks. If persistent symptoms reconsider underlying diagnosis and need for referral.
  - Long term prognosis is good with 85% resolution within 3 weeks. Complete resolution should occur within 3-5 months.

- **Complex or atypical cases should be referred to:** The Wessex Facial Nerve Centre

**Medical management**

### Eye care

If there is incomplete eye closure patients should be referred to ophthalmology and given eye protection advice. This includes taping the eye at night (figure 8), eye patch and artificial tears or lacri-lube ointment at night to prevent corneal ulceration.

#### Eye care

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  - If there is incomplete eye closure patients should be referred to ophthalmology and given eye protection advice. This includes taping the eye at night (figure 8), eye patch and artificial tears or lacri-lube ointment at night to prevent corneal ulceration.

- **Medical management**
  - The majority of idiopathic cases of facial nerve palsy (Bell’s Palsy) recover spontaneously. There is a lack of high quality evidence for the management of facial nerve palsy in children, most being inferred from adult studies.
    - In certain regions of the UK, for example the New Forest, Lyme disease is particularly prevalent. This caused 50% of LMN paediatric facial nerve palsy cases in our local 5-year review. Our local policy guideline recommends antibiotic treatment for all children whilst awaiting Lyme serology. The choice and dosage of the antibiotic depends on age. Amoxicillin three times daily for 14 days (15-20mg/kg) is recommended for children under eight years of age. Doxycycline 1-2mg/kg twice daily for 14 days is recommended for children equal to or older than eight years of age.
    - Children with a definite diagnosis of Ramsay Hunt Syndrome should be treated with acyclovir (> two years old) or valacyclovir (> 12 years old) to improve the chance of spontaneous recovery.

- **Surgical management**
  - Surgical management of paediatric facial nerve palsy (LMN) depends the cause. In acute infective cases such as acute otitis media, myringotomy, +/- grommet insertion, +/- cortical mastoidectomy should be considered.
    - Chronic supplicative otitis media with active squamous disease (cholesteatoma) requires urgent mastoid exploration and facial nerve decompression by a senior otologist. Vestibular schwannomas (acoustic neuromas) or any cerebellopontine angle tumours should be referred to the skull base MDT for further management.
  - There are no explicit algorithms for the optimal surgical management of acute facial nerve palsy in the literature. Gantz et al (1999) recommended performing facial nerve decompression in patients with > 90% loss on electrical activity on electroneuronography (ENoG) testing.
  - In acute paediatric facial nerve palsy, almost 100% of children recover spontaneously within a year, therefore surgical decompression of the facial nerve is not generally recommended, although there are no controlled trials to validate this.
  - In traumatic cases however, primary repair of the nerve has resulted in the best outcomes both aesthetically as well as functionally. In the paediatric age group due to the developing neural systems, early surgical intervention leads to better regeneration when compared to adults. Performing direct end-to-end anastomosis in a tensionless repair or using a cable graft from the greater auricular nerve or sural nerve when tensionless repair is not achievable is advocated. Reanimation procedures are considered for chronic irreversible causes of facial nerve palsy. The choice of reanimation procedure depends on the degree of damage and period of weakness for chronic irreversible causes. These procedures may be dynamic (when facial nerve palsy duration < 18 months) or static (when facial nerve palsy duration > 18 months).

- **Surgical management**
  - Surgical management of pediatric facial nerve palsy (LMN) depends the cause. In acute infective cases such as acute otitis media, myringotomy, +/- grommet insertion, +/- cortical mastoidectomy should be considered.
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  - Free muscle flaps or local rotational flaps are used for dynamic reanimation procedures. Ipsilateral/contralateral facial nerve, masseteric branch of the trigeminal nerve or sural nerves are used for innervation of the free muscle flaps. Dynamic reanimation procedures are aimed mainly at regaining the tone of the ocular and oral muscles, which greatly impacts on the functional and cosmetic results in children.
  - Static procedures are used where dynamic procedures have failed or cannot be used. Latissimus dorsi flap and a gold/platinum weight for the upper eyelid and medial canthopexy for the lower eyelid are mainly used in...
have failed, as there is limited evidence of when to
muscles. Active rehabilitation may be required in
oromotor tone due to the involvement of the facial
Children with facial nerve palsy may also suffer nutritional
Rehabilitation

References
2176.
Paediatric sleep physiology and sleep disorders—breathing

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Abstract
Good quality sleep is crucial for physical and mental health in children. Sleep disordered breathing is increasingly being recognised as having detrimental effects on neurocognitive development, academic achievements, behaviour, cardiovascular and metabolic health. A good understanding of sleep physiology, sleep architecture including sleep stages, control mechanisms of sleep, and developmental aspects and influences on sleep enables the clinician to embark on a more focussed investigative process and arrive at a more timely diagnosis. Parasomnias usually occur in the first third of the night and can occur either isolated or in the context of a snoring child with obstructive sleep apnoeas due to overall poor quality sleep. Obstructive events predominantly occur during REM sleep (when skeletal atonia occurs) and therefore more during the second half of the night. This increases pharyngeal collapsibility and therefore the likelihood of obstructive events increases. The length of individual REM periods increases across the night in children over one and adults the majority of REM occurs in the first half of the night. The second in the evaluation and scoring of sleep disordered breathing (SDB) in children. While parents may be vigilant during the early part of their child’s sleep they are more likely to miss, and therefore underreport, more significant symptoms such as snoring or apnoeas in the latter part of the night. Secondly in the evaluation and scoring of sleep studies: children who only achieve a short night’s sleep will sacrifice relatively more REM sleep. As SDB is defined according to an apnoea/hypopnoea index (AHI) where the denominator is total hours of sleep this will affect the severity grading of scoring.

Stage III sleep, conversely, occupies most of the first half of night but tends to disappear from later cycles when Stage II sleep increases. NREM parasomnias during Stage III sleep (e.g., confusional arousals and night terrors) typically occur therefore in the first half of the night. The absolute sleep requirements (hours per day) and sleep architecture (duration and frequency of different sleep stages) change throughout life (Figure 2).

In the absence of EEG, Electroencephalography (EOG) and Electromyography (EMG) measures of sleep stage, other correlates are used as surrogate markers to determine NREM (quiet) and REM (active) sleep. These include increased variability in respiration and heart rate during REM sleep.

Control of sleep
Two principle interacting mechanisms control our need for sleep. First of all the so called homeostatic drive (process S), which is a time dependent, linear and cumulative process. It depends on prior sleep and wakefulness and reflects the urge for sleep; it rises during times of waking, decreases during sleep and builds up with sleep deprivation. Secondly, the intrinsic circadian drive (process C) which is controlled by the “biological clock” located in the suprachiasmatic nucleus of the hypothalamus. The intrinsic circadian drive is synchronised with environmental cues, such as seasonally changing light-dark cycles, environmental sounds and meal times. Light is the most significant of these. It synchronises the intrinsic circadian clock to the 24 hour day of the surrounding environment. Exposure to blue light and white light late in the day can suppress endogenous melatonin secretion and delay bedtimes. A large cross-sectional general community-based study confirmed the widespread daytime and bedtime use of electronic devices and the associated risks of short sleep duration, long sleep onset latency and overall sleep deprivation in adolescents. Interestingly, and

Sleep
Sleep is a reversible state of reduced awareness of, and selective responsiveness to the environment. Arousals from sleep are clearly important in situations of external danger, or, indeed, with intrinsic problems such as severe obstructive sleep apnoea (OSA) where arousals triggered by abnormal levels of oxygen or carbon dioxide are crucial to enable protective airway responses. Sleep is not one entity but rather composed of multiple brain states called sleep stages, defined by neurophysiological parameters;
at first look counterintuitively, the circadian drive for wakefulness is at its peak just before bedtime and is lowest in the early morning before waking. The rationale behind this lies in the interaction between processes C and S. As the homeostatic sleep pressure increases at the end of a day, the body requires a stronger intrinsic signal promoting wakefulness. On the other hand, as the homeostatic urge for sleep disappears gradually during night-time sleep, changes in the circadian stimulus balance out to maintain sleep.

Why (good) sleep matters
Sleep is a process during which the brain is active. It affects both mental and physical health. Good quality sleep is crucial for the normal functioning of the body and its systems down to the level of gene expression and DNA synthesis. It forms an essential part of maintaining homeostatic regulatory and repair mechanisms. Poor quality sleep, which may be due to inadequate time in bed or sleep disorders such as OSA have potentially wide-ranging negative effects on many aspects of physical and psychological well-being as well as on aspects of behaviour and academic performance.

Bryant et al. have reviewed studies looking at the reciprocal relationship between sleep, infections and immunity and highlighted that many components of the immune system show, like sleep, circadian rhythmicity. Sleep restriction of 4 hours per night prior to influenza vaccination was associated with a 70% reduction in antibody titres 10 days post vaccination when compared to usual sleep controls in an adult study. A variety of bacterial and viral infections are associated with an increase in sleep duration and infants in the recuperation phase from upper respiratory tract infections often show impaired arousal from sleep, which also underlines the important healing and anabolic functions of sleep.

Poor quality sleep is also associated with other conditions. For example there is a linear relationship between total sleep time and obesity in children aged < 10 years. Those sleeping for less than 10 hours have a 60% increased risk for being obese or overweight. Each additional hour of sleep decreased the risk by 9%. Cross-sectional studies demonstrate an increased risk of the metabolic syndrome in association with increasing severity of obstructive sleep apnoea syndrome (OSAS) in obese children. Furthermore, OSA in children is linked to cardiovascular complications such as hypertension and blunting of the nocturnal fall in blood pressure. Investigating children with hypertension who also have clinically benign sleep apneas demonstrated significant positive correlation between sleep duration and cognitive performance. Sleep deprivation was associated with cognitive deficits and more behavioural problems.

Sleep disorders in children
Broadly defined, a sleep disorder is a condition or process that interferes with a child's previously established sleep-wake cycle. The International Classification of Sleep Disorders (3) defines the following categories:

1. Insomnia: primary sleep disorders that cause either a difficulty getting off to sleep, difficulty maintaining sleep, or non-restorative sleep.
2. Hypersomnias (increased need/amount of sleep or inability to stay awake).
3. Parasomnias (e.g. night terrors, sleep walking).
4. Circadian sleep-wake rhythm disorders (disturbance of biological clock).
5. Sleep related movement disorders (e.g. restless leg syndrome and periodic limb movement disorder).
6. Sleep related breathing disorders.

In the following we will focus on the two categories of sleep disorders that are common in paediatric clinical practice and appear most relevant to an ear, nose and throat (ENT) focussed readership: Sleep disordered breathing and Parasomnias. These two categories can also occur in one child at the same time as OSA can trigger parasomnias through the poor quality sleep mechanism.

Insomnia
Insomnia is defined as a sleep complaint that affects sleep quality, quantity or both and is associated with daytime consequences (e.g. daytime sleepiness). The prevalence of insomnia in children varies from 5% to 25% and is highly dependent on the criteria used for insomnia diagnosis. A meta-analysis of 10 studies (almost 9,000 children) revealed that children with more severe OSA (AHI > 5/hr total sleep time) appear to experience considerable neurocognitive impairment. The postulated mechanisms include sleep disruption and intermittent hypoxaemia which influence restorative processes that usually occur in sleep. A large meta-analysis of 86 studies (almost 36,000 children aged 5-12) demonstrated significant positive correlation between sleep duration and cognitive performance. Sleep deprivation was associated with cognitive deficits and more behavioural problems.

Sleep disordered breathing
Increased upper airway resistance and pharyngeal collapsibility lead to snoring and increased respiratory effort, the hallmarks of obstructive sleep apnoea which has a prevalence of 1-4% in the paediatric population. Depending on the severity four clinical entities have been defined: Primary snoring, upper airway resistance syndrome, obstructive hypopnoea and obstructive sleep apnoea syndrome. It is important to note that obstructive events and oxygen saturations below 90% rarely occur in normal children without SDB. The Apnoea-Hypopnoea index (AHI) – the number of mixed/obstructive central apnoeas and hypopnoeas per hour of total sleep time – is the most widely used and reported parameter to define the severity of SDB and informs the selection of the appropriate therapeutic intervention. See summary of definitions and scoring criteria in table 2.

Unlike in adults the presentation of SDB in children is more varied, symptoms change with age, and individual symptoms by themselves are of limited value for a conclusive diagnosis. Using symptoms and physical examination alone for diagnosis can lead to over- or undertreatment in children. Parental reports of frequent loud snoring, mouth breathing (leading to ‘adenoid facies’), witnessed apnoeas, and restless sleep are associated with presence of obstructive SDB. Attention deficit/hyperactivity symptoms rather than excessive daytime sleepiness appear to be more common in children with SDB. Nocturnal enuresis is linked to OSAS and improves after treatment. Increased respiratory effort can lead to night sweating. Tonsilar size (Brodsky score) is weakly related to presence or severity of obstructive SDB; some studies however suggest no association. A background of prematurity and parental/sibling history of OSAS, adenosinomellectomy or adenotonsillar hypertrophy are important risk factors to be taken into account.

Some complex genetic medical conditions (e.g. Achondroplasia, Down syndrome, Prader-Willi syndrome, neuromuscular disorders (e.g. cerebral palsy, Duchenne muscular dystrophy), and syndromes with midface hypoplasia and/or mandibular hypoplasia (e.g. Apert syndrome, Pierre Robin sequence) are associated with an increased risk for obstructive SDB. Obesity is an important emerging independent risk factor for OSAS in children. Importantly, adenosinomellectomy for OSAS in (normal and overweight) children leads to clinically significant weight gain which can place them at further risk for OSAS and therefore weight reduction strategies need to be included in the long-term management of this patient group.

Risk factors for persistence of untreated OSAS are male sex, obesity, obstructive AHI > 5/hr, narrow maxilla (which can lead to crossebite and malocclusion), and persistent tonsillar hypertrophy.

Objective investigations include flexible nasopharyngoscopy and lateral neck radiography. Cephalometry, computed tomography (CT) or magnetic resonance imaging (MRI) of the upper airways may be useful in selected patients. In comparison with nasopharyngoscopy cephalometry appears to underestimate...
Night-to-night variability is low. A PSG study in typically developing children is sufficient as called central pattern generators (CPGs). Essentially, (ambulatory) polygraphy and nocturnal pulse oximetry if PSG or polygraphy are laid out in the recently updated lateral neck radiography potentially overdiagnoses OSAS.

Parasomnias

Parasomnias are “undesirable behavioural, physiological, or environmental events that accompany sleep”11. They can be divided into NREM and REM related parasomnias. REM parasomnias are rare in childhood and will not be discussed further here. In NREM parasomnias, behaviours are triggered by activation of functional groups of neurons called central pattern generators (CPGs)12, 13. Essentially, they represent a state dissociation within the brain between wake and NREM sleep. They are usually brief but can last up to 30-40 minutes. Diagnostic criteria include: (ICSD III)11.

Sleep terrors typically occur occasionally in 3% of children aged 3-10 years and are characterized by abrupt waking in association with intense fear, loud vocalisation, and autonomic system activation leading to agitation, flushed appearance and sweating. Children do not respond to calm efforts, they may become more agitated and disoriented when woken, and may even leave their bed and run out of the room. Children, unlike adults, with sleep terrors do not have a higher incidence of psychopathology. Factors such as sleep deprivation, fever, and certain medications can be trigger factors22. Sleep terrors can be dramatic and terrifying events in parents but are usually not recalled by children themselves.

Confusional arousals usually occur in toddlers, and become less common in older children. They are characterised by a child sitting up in bed, appearing distressed, crying and vocalising. Again attempts to comfort may cause agitation. Confusional arousals are not associated with higher autonomic behaviour. Activities and prognosis are usually benign.

During sleepwalking a series of complex motor behaviours are instigated that lead to walking while the child is asleep. Eyes are open, perceptual elements of the environment can be remembered, however usually amnesia exists. Sleepwalking most commonly occurs after four years and peaks around twelve years of age. A pre-disposing factor is sleep deprivation which may be environmental or behavioural but could be exacerbates by OSA.

Treatment approaches include ensuring the safety of the child and providing reassurance about the overall benign nature of NREM parasomnias. OSA as a factor that disrupts sleep should be excluded and sleep quality needs to be improved. If NREM parasomnias happen very regularly anticipatory wakening may be attempted.

Sleep

Sleep affects mental and physical health and poor quality sleep in deprivation have far reaching consequences for the affected child. Knowledge of the basics of sleep physiology and SDB in children, the important aspects and historical background, and the contribution of sleep studies in the diagnostic process is important to achieve the best possible outcome for the snoring child in the ENT clinic.

References

Abstract
Paediatric tracheostomy is a procedure that all registered otorhinolaryngologists should be able to perform at least in an emergency situation. This review article examines how this procedure has changed over the years. It considers the indications, preoperative surgical checks, counseling, tracheostomy tubes, surgical technique, complications and outlines health care needs at home for children with long-term tracheostomies.

Introduction
The first paediatric tracheostomy was undertaken in 1620. Most early tracheostomies were performed as emergencies for relief of upper airway obstruction due to infection. In 1833 Trouseau performed around 200 tracheostomies due to the diphtheria epidemic. The indications for the procedure expanded when Galloway undertook tracheostomy during the poliomyelitis epidemics of the early 1950s, promoting the use of tracheostomy for positive-pressure ventilation. This highlighted the potential use of tracheostomy in optimising ventilation in other situations such as post cardiac surgery, high-grade burns, and in the care of preterm infants.

Nowadays, the majority of tracheostomies in the paediatric age group are not performed in the emergency situation. They are inserted for chronic non-infective upper airway obstruction or long-term ventilation. Airway obstruction can be congenital or acquired (table 1). One study showed that the most common indications necessitating paediatric tracheostomy were: long-term ventilation (20%), craniofacial abnormality causing airway obstruction (18%) and subglottic stenosis (14%).

Table 1: Congenital and acquired causes of upper airway obstruction

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Acquired</th>
</tr>
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<tbody>
<tr>
<td>Nasal</td>
<td>Infarction</td>
</tr>
<tr>
<td>Oropharyngeal obstruction (e.g. Pierre Robin sequence and other craniofacial abnormalities)</td>
<td>Trauma (e.g. physical, intubation, burns, chemical, occurring at any level)</td>
</tr>
<tr>
<td>Supraglottic (e.g. laryngomalacia)</td>
<td>Subglottic (e.g. subglottic stenosis, subglottic haemangioma)</td>
</tr>
<tr>
<td>Glottic (e.g. congenital vocal fold paralysis, laryngeal web)</td>
<td>Tracheobronchomalacia</td>
</tr>
<tr>
<td>Tumours/ Malignancy</td>
<td>Neurological/ Neuromuscular</td>
</tr>
</tbody>
</table>

Paediatric tracheostomy
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obstructive conditions such as subglottic haemangiomas, subglottic stenosis and laryngeal clefts.

**Preoperative counselling**

Looking after a child with a tracheostomy is life changing and has a huge impact on the carers including practical issues of tube management, worries about tube safety and social stigma. Where the tracheostomy is planned, preoperative counselling is essential and includes:

1. Multidisciplinary meetings
2. Reassurance about voice issues, swallowing and feeding
3. Educational material, videos and meeting other parents of children with a tracheostomy

As with any surgical procedure, the surgeon must have a clear and honest discussion with the family concerning the benefits, risks, and alternatives to tracheostomy.

**Preoperative checks**

In cases of airway obstruction, a full dynamic and static evaluation of the airway including laryngotracheobronchoscopy is performed. This is to assess anatomy, function and to determine whether any endoscopic surgical intervention could avoid the need for tracheostomy.

Prior to tracheostomy routine blood tests and a coagulopathy screen should be undertaken. A chest x-ray is mandatory to evaluate the respiratory system and may also be helpful in establishing the position of the trachea.

There are various anatomical differences between adult and paediatric airways and these should be considered when undertaking a paediatric tracheostomy (table 2).

**Tracheostomy tubes**

The selection of an optimal tracheostomy tube depends on the clinical indication and the size of the airway. Both the diameter and length should be considered. The internal diameters of paediatric tracheostomy tubes range from 2.5-5.5 mm and the lengths range from 30-36 mm for neonates and 39-56 mm for paediatric patients. The diameter of the tracheostomy tube can be calculated on the basis of the size (the inner diameter) of the child’s endotracheal tube. Great Ormond Street Hospital has produced a chart to aid tube selection. Neonatal tubes are equal to pediatric tubes in their both inner and outer diameters. As a result of their small size, paediatric tracheostomy tubes are only single-lumen.

Regarding tracheostomy tube length, a child under 12 months of age requires a shorter ‘neo’ tube; whereas a child over 12 months of age requires a longer ‘paed’ tube. Paediatric tubes were initially cuffless but low-pressure cuffed models have become available over the last few years to aid ventilation. Using too large a tube diameter or overinflation of a cuff tube may result in tracheal injury due to vascular compromise resulting in pressure necrosis, ulceration fibrosis and eventually stenosis.

As children grow they generally require progressively larger tracheostomy tubes to avoid nocturnal desaturations whilst allowing for speech.

**Open surgical tracheostomy technique**

The procedure is ideally undertaken on an intubated child under general anesthesia.

1. **Set up equipment**
   
   Figure 1 shows the equipment set up for a paediatric tracheostomy.

2. **Check Tracheostomy Tube size**
   
   It is necessary to check the tracheostomy tube size and ensure one size smaller is available. If a cuffed tube is used the cuff should be checked prior to commencing the procedure. The anaesthetic connectors connecting the tracheostomy tube to the anaesthetic circuit should also be checked (figure 2). The tracheal suction length should be measured by placing an appropriately sized tracheal suction catheter into the tracheostomy tube with the suction catheter tip just visible beyond the tracheostomy tube.

3. **Position the child**
   
   The child is positioned with the head extended using a shoulder roll and head ring. Excessive hyperextension should be avoided in children as this may result in mediastinal structures presenting in the neck and therefore making them vulnerable to injury. The head is then stabilised in the midline position using surgical tape placed across the chin attached to the operating table (figure 3). In certain cases neck extension may be contraindicated due to the risk of atlantoaxial subluxation e.g. with Trisomy 21 (Down’s syndrome) or achondroplasia.

4. **Skin Incision**
   
   The skin incision should be marked. A vertical or horizontal incision is made in the midline midway between the expected position of the cricoid cartilage and suprasternal notch (figure 4). The lateral limits of a horizontal incision are the anterior borders of the sternoclavicular muscles. Local anesthetic 2% lignospan special (2%...
Lidocaine with 1:80,000 adrenaline is infiltrated into the area of the planned incision. Alternatively in small neonates, 1 ml of 1,000,000 adrenaline can be used. A vertical skin incision is the preferred technique for emergency tracheostomies when preoperative ventilation and intubation is not possible and a surgical airway is urgently required.

5. Removal of subcutaneous fat, dissection and identification of trachea
The skin incision is continued through into the subcutaneous fat. The fat is debulked around the incision site using bipolar diathermy (figure 5). The removal of subcutaneous fat aids the placement of maturation sutures. In an emergency tracheostomy, where timing is critical, this is not necessary. The midline cervical fascia between the strap muscles is identified and divided (figure 6). The strap muscles can then be divided in the midline using bipolar diathermy or blunt dissection, maintaining meticulous haemostasis throughout. The strap muscles are retracted laterally to expose the thyroid isthmus. The thyroid isthmus is divided using bipolar diathermy to identify the trachea. It is essential to stay in the midline at all times particularly in neonates where the apex of the lung dome extends from the thorax into the root of the neck. The cricoid cartilage and tracheal rings are identified. It is essential to avoid injury to the first tracheal ring as this may lead to subglottic stenosis. A saline soaked pledget may prove useful in gently clearing the anterior tracheal wall to improve exposure of the trachea. Caution should be exercised when dissecting the lower trachea as the innominate artery normally passes below the 5th tracheal ring.

6. Insertion of stay sutures
Non-absorbable 3/0 or 4/0 prolene stay sutures are placed through the tracheal rings 3-5 on either side of the planned midline tracheal incision (figure 7). Stay sutures are beneficial in applying upward and lateral pull on the trachea, bringing it to the surface, and can be lifesaving during accidental decannulation.

7. Tracheal incision and maturation sutures
A vertical tracheal incision is made through the tracheal rings between the stay sutures (usually between the 3rd and 5th). In a non-emergency situation, maturation sutures are recommended and are placed prior to tracheostomy tube insertion. These are 4/0 vicryl absorbable sutures between the trachea and skin connecting the trachea directly to the skin, creating a safer stoma. These are placed superiority and inferiory on either side of the skin (figure 8). The inferior maturation sutures are the most crucial. Whilst applying the maturation sutures the endotracheal tube remains in place (access can be improved by withdrawing the tube slightly and/or deflating the cuff if present). In order to prevent anaesthetic gases escaping and to support ventilation the tracheal incision can be intermittently occluded with a small pledget or by carefully crossing over the stay sutures.

8. Tracheostomy tube insertion
Once the maturation sutures have been applied the tracheostomy tube can be inserted under direct vision (figure 9). The position of the tracheostomy tube may be checked intraluminally using a flexible endoscope. The distal end of the tube should ideally be situated above the carina. The tracheostomy ties are then applied and knotted; the tube should be well secured with the head flexed to avoid accidental dislodgement, but not too tight so as to cause vascular compromise and skin breakdown. The tube should not be sutured to the skin, as paediatric skin is very elastic therefore, should the tube dislodge reinsertion of a sutured tube would be more difficult. The stay sutures are then taped to the chest and labelled "RIGHT", "LEFT", and ‘DO NOT REMOVE’ (figure 10). The purpose and use of the stay sutures should be explained to all involved in the care of the child prior to the first tracheostomy tube change.

Postoperative care
Chest X-ray is required to exclude a pneumothorax, surgical emphysema and reconfirm the position of the tracheostomy tube. Paediatric or neonatal intensive care unit stay may be required in some instances until the first tracheostomy tube change. The child may be sedated and paralysed for 48 hours if necessary. Antibiotic prophylaxis may be indicated postoperatively. A tracheostomy box should be set up at the patient’s bedside containing a same size and smaller tube, lubricant, saline, suction catheters, scissors, ties, dressings.

Careful suctioning and humidification is essential especially in the first 3 days. The tracheocutaneous tract is usually well established at 72 hours. Nursing staff should be informed regarding the correct suction length in order to avoid any distal tracheal injury and granulations. The skin of the neck and stoma should also be checked on a daily basis.

The first tracheostomy tube change should be considered at 6-7 days, but may be performed earlier. The surgeon determines the exact timing of the first tracheostomy tube change. This should be performed in a safe environment where emergency reintubation is possible. Following tube change and confirmation of correct positioning the tube is secured. Finally the stay sutures are removed.

Laryngotracheobronchoscopy with review of the tracheal stoma and the original indication for tracheostomy by the multidisciplinary team should be undertaken at regular intervals. This may range from six monthly to yearly, however an individual case based approach should be employed.
Complications

The risks of tracheostomy can be classified as early postoperative and long-term complications.

Early complications

Complications directly related to the procedure itself include: losing airway control, insufficient ventilation, bleeding, pneumothorax and oesophageal injury. Many paediatric patients requiring tracheostomy have complex medical co-morbidities, which also increases the risks of cardiac arrest and death. The incidence of tracheostomy tube occlusion has been reported to be as high as 72% in premature and newborn children decreasing to 14% in paediatric patients older than 12 months. The higher rate of blockage in the younger age group is attributed to the narrower diameter of tracheostomy tubes and bronchopulmonary dysplasia resulting in viscous bronchial secretions in premature infants.

Accidental decannulation is a serious complication in tracheomalacia and vocal cord movement. Decannulation generally recommended if the tracheostomy has been in situ for several weeks. Pre-decannulation laryngotracheobronchoscopy is essential. Failure of decannulation can be related to different factors. These may be removed either endoscopically using bronchoscopes with cold steel, KTP laser techniques or laser techniques or these may be removed either endoscopically using bronchoscopes with cold steel, KTP laser techniques or laser techniques. These may be removed either endoscopically using bronchoscopes with cold steel, KTP laser techniques or laser techniques. These may be removed either endoscopically using bronchoscopes with cold steel, KTP laser techniques or laser techniques.

Tracheostomy impacts speech and language development. Earlier decannulation results in improved speech and language development. A multidisciplinary approach to management optimising initial hospital care and promoting effective communication has been shown to improve outcomes and avoid complications. This includes weekly care conferences, consensus guidelines, educational initiatives, care plans, and regular follow up with endoscopic assessment.

Home care preparation

Pre-discharge family and home environment planning are essential. During the hospital stay the child’s family or guardians undertake a structured and detailed training program, requiring the achievement of competencies and providing the necessary skills to manage the long-term care at home. Although complications can occur, several studies report a low rate of documented accidents and high carer satisfaction rates when managing the children at home with appropriate support.

Conclusion

Paediatric tracheostomy continues to have a significant role in paediatric airway management. There have been many changes in indications, tubes and surgical modifications over the years. The main development has been the multidisciplinary approach to tracheostomy management and the increasing role of the parents in caring for their children at home.

References

Paediatric vocal cord paralysis

Virginia Fancelli1, Matthew Ward1, Hasnna Ismail-Koch1 and Kate Heathcote1

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Abstract

Vocal cord palsy in children may occur in isolation or in association with other pathologies. It may be unilateral or bilateral, temporary or permanent and may present in a number of ways. This includes ill-defined feeding difficulties, weak cry or in an emergency situation with stridor. The advent of flexible nasolaryngoscopes has greatly improved our ability to diagnose these conditions. In infants and toddlers the primary concerns are focused on the airway and aspiration risk. Whereas the full impact of the vocal disability only reveals itself as the child gets older, effecting social and educational development. The focus of this review is to offer a step-by-step approach to diagnosis and management of both unilateral and bilateral vocal cord palsy in children.


Key words

vocal cord palsy, laryngeal electromyography, laryngeal reinnervation

Acknowledgement

Andrea Burgess, Paediatric Consultant ENT Surgeon, University Hospital Southampton NHS Foundation Trust. Who contributed to the clinical cases and management of the approach described.

Introduction

The term “vocal cold paralysis” (VCP) refers to the absence of vocal cord movement, typically due to lower motor neuron dysfunction. It may be unilateral (UVCP) or bilateral (BVCP) and be due to a central lesion or, more commonly, as a result of vagal or a recurrent laryngeal nerve (RLN) impairment. In recent decades the prevalence of paediatric VCP seems to have increased. This is likely to be, at least in part, due to an increase in diagnosis as more units routinely use paediatric flexible nasolaryngoscopes (FLN). Another important factor is the improved survival rates among pre-term infants. However, the true incidence of VCP in children remains unknown.

Aetiology

In 2006 Smith proposed a classification system whereby the causes of paediatric VCP can be identified as congenital, hereditary or acquired. Their prevalence of the different aetiologies varies widely between studies and over the decades, showing interesting changes in recent years. (Tab. 1)

Abstract of Table 1:

<table>
<thead>
<tr>
<th>CONGENITAL</th>
<th>ACQUIRED</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebral agenesis</td>
<td>Neoplastic (brain, neck and mediastinal)</td>
</tr>
<tr>
<td>Corpus callosum agenesis</td>
<td>Congenital vascular accidents</td>
</tr>
<tr>
<td>Nucleus ambiguous dystrophy</td>
<td>Traumatic</td>
</tr>
<tr>
<td>Arnold-Chiari malformation</td>
<td>Birth injury</td>
</tr>
<tr>
<td>Hydrocephalus</td>
<td>Post-surgical correction of cardiovascular</td>
</tr>
<tr>
<td>Encephalopathy</td>
<td>or oesophageal abnormalities</td>
</tr>
<tr>
<td>Meningomyelocele</td>
<td>Neck surgery</td>
</tr>
<tr>
<td>Meningocele</td>
<td>Otmar’s syndrome</td>
</tr>
<tr>
<td>Congential myasthenia gravis</td>
<td>INFECTIVE</td>
</tr>
<tr>
<td>Cardiomegaly</td>
<td>Mediastinitis</td>
</tr>
<tr>
<td>Interventricual septal defect</td>
<td>Pertussis encephalitis</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Polyneuritis</td>
</tr>
<tr>
<td>Vascular ring</td>
<td>Poliomyelitis</td>
</tr>
<tr>
<td>Dilated aorta</td>
<td>Diphtheria</td>
</tr>
<tr>
<td>Double aortic arch</td>
<td>Rubies</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>Syphilis</td>
</tr>
<tr>
<td>Transposition of great vessels</td>
<td>Herpes encephalitis</td>
</tr>
<tr>
<td>Aberrant innominate artery</td>
<td>Taterus</td>
</tr>
<tr>
<td>Cystic Hygroma</td>
<td>Botulism</td>
</tr>
<tr>
<td>Acute feotal suffuring</td>
<td>Tuberculosis</td>
</tr>
<tr>
<td>Neonatal benign hypotonia</td>
<td>Congenital Varicella Zoster</td>
</tr>
<tr>
<td>ASSOCIATED WITH OTHER CONGENITAL ANOMALIES</td>
<td>Congenital Rubella</td>
</tr>
<tr>
<td>Laryngeal abnormalities</td>
<td>Epstein Barr Virus infection</td>
</tr>
<tr>
<td>(subglottic stenosis, laryngomalacia, laryngeal cleft, larynx haemangiomia)</td>
<td>Guillain-Barre Syndrome</td>
</tr>
<tr>
<td>Bronchogenic cyst</td>
<td>NEUROTOXIC</td>
</tr>
<tr>
<td>Oesophageal cyst</td>
<td>Vincristine/Vinblastine</td>
</tr>
<tr>
<td>duplication, atresia</td>
<td>Heavy metal poisoning</td>
</tr>
<tr>
<td>Cricopharyngeal stenosis</td>
<td></td>
</tr>
</tbody>
</table>

HEREDITARY

Autosomal dominant

Autosomal recessive

Hlx-linked

Isolated mutation

Associated with other hereditery syndromes (e.g. Pelizaeus-Merzbacher syndrome, Cri du chat disease, Charcot Marie Tooth disease, Neurofibromatosis)

Clinical features and management

VCP is the second commonest cause of stridor in neonates and infants after laryngomalacia and accounts for more than 10% of laryngeal anomalies in children. Children with concurrent cardiac or neurological pathologies are particularly vulnerable to cyanotic episodes, intercostal recession, and apnoea. Thus, tracheostomy is more likely required for the vagus of the uvula or RLN. This is less and less described, most likely due to the increase rate of caescan section.

Diagnosis

A medical history with a careful physical examination is crucial. This should focus on symptoms of VCP, for example, abnormalities in the voice or cry, feeding difficulties, and stridor or cyanotic episodes. It is also important to establish any risk factors as discussed above.

FLN is an essential tool for assessing the dynamic movement of the larynx. A 2.8 mm paediatric fibroptic scope is reasonably tolerated by children. In newborns swallowing can be a helpful technique to ensure that the child remains still. Similarly, children older than 3-4 years can usually tolerate FLN after careful explanation of the technique. Between the ages of 1-2 years, awake FLN can be very difficult and is approached on a case by case basis.

Microlaryngobronchoscopy under general anaesthesia plays a fundamental role in differentiating VCP from other laryngeal defects such as cricarytenoid joint fixation, interarytenoid scar or vocal fold fusion, and to exclude associated laryngeal abnormalities for example subglottic stenosis.

Magnetic Resonance Imaging, and in selective cases Computer Tomography, may be necessary to investigate the aetiology and associated pathologies.

The clinical manifestations must be carefully evaluated before any surgical procedure is considered in view of the potential long-term impact some may have on voice. Direct observation of breathing and oxygen saturation monitoring are essential to plan the management. When feeding difficulties are present, a videofluoroscopy may be used to investigate silent aspiration, particularly in cases with cardiorespiratory co-morbidities.

Laryngeal Electromyography (LEMG) introduced into clinical practice by Hirano and Ohara in 1969, is a useful examination in children with vocal cord immobility. It can help to plan treatment in cases where the differentiation between paralysis and fixation may be difficult to make. Moreover, in children with BVCP it can be valuable in predicting recovery.

Laryngeal ultrasound was proposed by Friedman in 1997 as a way to assess vocal cord mobility. This technique is non-invasive, painless and requires neither sedation nor general anaesthesia. It may be of use as a screening tool to exclude VCP rather than to investigate a suspected case.

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to be required as well as in those with obstructive sleep apnoea due to conditions such as microretrognathia.

UVC

In neonates UVCP may present as a weak cry or choking episodes. Intermittent stridor may also be a feature and has been reported to occur in up to 77% cases. If stridor is present it is usually mild and without symptoms. As a result, microsurgical laryngoscopy may not, at first glance, appear indicated. However, it is vital to distinguish UVCP from aspiration, which may be caused by intubation. The appearance of both can be very similar, especially as patients with UVCP often have marked arytenoid prolapse. If dislocation is present, the dislocated arytenoid needs to be relocated as soon as possible in an attempt to prevent permanent asynchrony. During microsurgical laryngoscopy the mobility of the criocarotidarytenoid joint should be noted. In palsy the joint is mobile whereas in dislocation the arytenoid is firmly in place and the joint must be forcibly reduced in an attempt to restore function and prevent arytenoid ankylosis with permanent fixation. Intra-operative Laryngeal Electromyography (LEMG) may be helpful in this differential. (Case 1)

An alternative treatment for UVCP is non-selective reinnervation. In this technique a branch of the ansa cervicalis is anastomosed with the recurrent laryngeal nerve. This reinnervates the muscles of the hemi-larynx and can restore muscular tone and improve arytenoid position. There is no restoration of movement coordinate with respiration and phonation, hence “non-selective”. One advantage of this technique is that the laryngeal framework is not disrupted in any way and so the larynx will continue to grow without interruption or the need for revision of the procedure. The reinnervation may not be effective for up to 6 months, while axonal ingrowth occurs, and so injection augmentation with a temporary substance should be performed simultaneously. (Case 2)

Speech therapy is of benefit in all age groups. Feeding techniques may avoid aspiration and swallowing. Vocal cord voice exercises can improve compensation from the contralateral vocal cord.  

Tracheostomy is frequently performed in children with BVCP, with reported rates of between 68% to up 80% of BVCP. Despite the difficulties in managing a tracheostomy, in our opinion this option is preferable to procedures that are destructive to the laryngeal structures such as cordotomy, arytenoidectomy and vocal cord lateralisation. These procedures result in permanent degradation of voice which may be avoided. Recovery occurs if Selective Laryngeal Reinnervation, as described by Professor Jean-Paul Marie, is successful. The aim of this technique is to restore physiological vocal cord movement that is coordinated with respiration and phonation. The upper root of the phrenic nerve on one side supplies the thyrohyoid muscle on the other side. This nerve is active in inspiratory efforts and so during the action of the PCAs, Professor Marie has performed this technique in children of all ages, with excellent success rates. We are currently introducing the technique in the UK. Conclusion

VCP has an associated mortality rate due to respiratory compromise and complications from aspiration. Treatment is imperative to prevent mortality and morbidity. Technological developments have improved the diagnostic rate and new techniques, such as reinnervation, have the potential to improve outcomes.

References

Transoral laser surgery for advanced laryngeal cancer

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Abstract
Surgical treatment of laryngeal cancer has evolved progressively towards transoral resections. In early tumours, local control with transoral laser microsurgery (TLM) has proved to be as good as in open surgery and fully comparable to that achieved under radiation protocols, but at a lower cost. Consequently, TLM is presently considered a first line treatment in early laryngeal cancer.

These good oncological and functional results have pushed TLM indications to intermediate and advanced carcinomas. In this article we review the outcomes of TLM in the treatment of locally advanced tumours of the larynx, putting special emphasis on difficulties, appropriate patient selection and different technical considerations.

The oncologic outcomes published in the literature suggest that TLM is comparable to other treatment alternatives in appropriately selected patients. From the functional point of view, TLM reduces patient morbidity, provides faster recovery and avoids tracheostomy in a high number of patients.


Key words
transoral laser microsurgery, advanced carcinoma, laryngeal cancer, CO2 laser

Conflicts of interest
The authors declare no conflicts of interest.

Acknowledgements
To Dr. José Luis Blanch and to all the members of the Hospital Clinic ENT Department.

Introduction
In the last decades, TLM has changed the concept of laryngeal cancer surgery. The possibility to remove the tumour transorally, in a concept of minimally invasive surgery, has reduced postoperative morbidity, with a more rapid recovery for patients and avoiding a tracheostomy in most of them. From the oncologic point of view, TLM has proved to be as good as open surgery and totally comparable to radiation protocols in early laryngeal cancer, but at a much lower cost1. Thus in many centres, where external approaches or radiation therapy were established as a first line treatment, the treatment protocols have been reconsidered.

Subsequently, such good results obtained in early tumours have pushed the indications of TLM to intermediate-advanced laryngeal cancers. In that scenario, TLM shares a leading role with chemoradiation strategies and open surgery. According to the literature, there is increasing evidence that in appropriate cases, comparable oncological results can be achieved with TLM also for advanced laryngeal cancers. In that scenario, TLM also for locally advanced tumours, with a substantial decrease in morbidity and a very good postoperative quality of life2-4. Unfortunately, no randomized studies have been conducted in this area, and the decision process is in part dependent to the learning curve and to the availability of treatment alternatives.

The goals of TLM include the oncologic cure, the organ preservation with an acceptable voice, a regular swallowing recovery, and avoiding a permanent tracheostomy or gastrostomy. There are currently no absolute recommendations regarding the indications or contraindications for TLM in advanced laryngeal tumours. An appropriate selection of ideal candidates needs to foresee that after the final tumour resection the patient has

References
11. Dedo DD. Pediatric vocal cord paralysis. Laryngoscope 1979; 89(9): 1378-1384
all odds of an aspiration-free deglutition or a good chance of swallowing rehabilitation.

Authors with experience in TLM essentially include T3 laryngeal tumours and, exceptionally, selected T4a cases (usually with limited involvement of the tongue base, growth towards the membranous pyriform sinus or minimal extralaryngeal extension). Bilateral involvement of the posterior commissure, cricoid cartilage infiltration, extensive subglottic involvement and marked extralaryngeal tumour extension are considered as contraindications5-8.

Controversies for TLM in locally advanced tumours

Exposure of the tumour

Adequate exposure of the tumour is always needed to achieve a complete endolaryngeal sound resection. A useful approach requires a suitable anatomy, an experienced surgeon, adequate instruments (different size and types of laryngoscopes, forceps, etc) and the knowledge of tricks about how to improve the tumour view.

Piazza et al. have recently published a predictive scoring system “the laryngoscore” which is based on eleven anatomic variables, and permits to anticipate the difficulties with the exposure of the larynx before a transoral approach9. The laryngoscore may help in preoperative planning and counselling. According to the authors, it also seems to be related to the risk of close or positive margins.

During surgery, initial debulking with the laser in scan mode may be necessary to progressively improve exposure in large tumours. External pressure is extremely helpful in the anterior commissure and the ventricle. The pressure has to be exerted on the cricoid cartilage, to verticalize the larynx and to facilitate the perpendicular cut with the laser. Under these conditions, and after repositioning the laryngoscope many times, most advanced tumours can be exposed than early ones, but overall, only in 23 patients out of 1,109 consecutive patients treated with TLM in our center the laser achieved a complete exposure. Even in the best scenario, there are many postoperative situations in which the margin may be considered "uncertain". These are given when the specimen sent for the pathology analysis presents a wide area of carboximation, when the laser reaches the cartilage and the certainty of infiltration remains unclear, or when the surgeon has enlarged the resection by means of additional vaporization. The attitude to be adopted in this clinical situation varies according to each author.


table 1: exposure according to tumour location and tumour size.

<table>
<thead>
<tr>
<th>All tumours (T1-T4a)</th>
<th>Good</th>
<th>Difficult</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supraglottic</td>
<td>273/311 (87.8%)</td>
<td>38/311 (11.2%)</td>
</tr>
<tr>
<td>Glottic</td>
<td>55/535 (87.6%)</td>
<td>7/535 (12.4%)</td>
</tr>
<tr>
<td>Anterior commissure (vertical plane)</td>
<td>123/163 (75.5%)</td>
<td>40/163 (25.4%)</td>
</tr>
<tr>
<td>T3-T4a</td>
<td>109/131 (83.2%)</td>
<td>22/131 (16.8%)</td>
</tr>
<tr>
<td>Supraglottic</td>
<td>47/55 (85.5%)</td>
<td>8/55 (14.5%)</td>
</tr>
<tr>
<td>Glottic</td>
<td>26/33 (77.8%)</td>
<td>7/33 (21.2%)</td>
</tr>
</tbody>
</table>

Within the marrow should not be underestimated. Thus, limited resection of the area of focal involvement may be insufficient and the possibility of an open partial approach should be considered.

Even in the best scenario, there are many postoperative situations in which the margin may be considered "uncertain". These are given when the specimen sent for the pathology analysis presents a wide area of carboximation, when the laser reaches the cartilage and the certainty of infiltration remains unclear, or when the surgeon has enlarged the resection by means of additional vaporization. The attitude to be adopted in this clinical situation varies according to each author.

Table 2: Oncologic results after TLM for advanced supraglottic tumours (T3-T4)

<table>
<thead>
<tr>
<th>n</th>
<th>TMM</th>
<th>Local control with TLM</th>
<th>Final local control</th>
<th>DSS / DFS</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>48</td>
<td>T3-T4</td>
<td>63.3%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>17</td>
<td>T3-T4</td>
<td>47%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>10</td>
<td>T3</td>
<td>91%</td>
<td>79%</td>
<td>86%</td>
<td>–</td>
</tr>
<tr>
<td>18</td>
<td>T3</td>
<td>81%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>46</td>
<td>28 T2</td>
<td>97%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>15</td>
<td>T3</td>
<td>80%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>96</td>
<td>T3</td>
<td>69.8%</td>
<td>91.7%</td>
<td>61.8%</td>
<td>45.8%</td>
</tr>
<tr>
<td>20</td>
<td>T3</td>
<td>83%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>104</td>
<td>T3</td>
<td>77.3%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>48</td>
<td>T4a</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>154</td>
<td>120 T3</td>
<td>73.8%</td>
<td>93.9%</td>
<td>67.6%</td>
<td>55.6%</td>
</tr>
<tr>
<td>24</td>
<td>T3</td>
<td>75.5%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>22</td>
<td>T3</td>
<td>95.4%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
</tbody>
</table>

Table 3: Oncologic results after TLM for advanced glottic tumours (T3-T4)

<table>
<thead>
<tr>
<th>n</th>
<th>TMM</th>
<th>Local control with TLM</th>
<th>Final local control</th>
<th>DSS / DFS</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>37</td>
<td>T3</td>
<td>67%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>70</td>
<td>T3</td>
<td>87%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>51</td>
<td>T3</td>
<td>65%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>41</td>
<td>27 T3</td>
<td>74%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>10</td>
<td>5 T3</td>
<td>45%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>11</td>
<td>T3</td>
<td>71.6%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>51</td>
<td>T3</td>
<td>47.1%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>31</td>
<td>T4a</td>
<td>74.6%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>122</td>
<td>T3</td>
<td>71.5%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>40</td>
<td>T3-T4</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>19</td>
<td>T3</td>
<td>52.7%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>34</td>
<td>T3</td>
<td>70%</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
</tbody>
</table>
Table 4: Functional results after TLM for advanced supraglottic tumours (T3-T4)

<table>
<thead>
<tr>
<th>n</th>
<th>TNM</th>
<th>Site</th>
<th>LPR</th>
<th>LFS</th>
<th>FPR</th>
<th>Gastrostomy (definitive)</th>
<th>Tracheotomy (definitive)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ambrosch et al., 2001**</td>
<td>50</td>
<td>T3</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
<td>0.02%</td>
</tr>
<tr>
<td>Motta et al., 2004**</td>
<td>18</td>
<td>T3</td>
<td>93.7%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>–</td>
</tr>
<tr>
<td>Caballeros et al., 2004*</td>
<td>15</td>
<td>T3</td>
<td>86%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>–</td>
</tr>
<tr>
<td>Vilaseca et al., 2016*</td>
<td>96</td>
<td>T3</td>
<td>76.6</td>
<td>74.5%</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Peretti et al., 2010*</td>
<td>20</td>
<td>T3</td>
<td>88.2%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Canis et al., 2013†</td>
<td>48</td>
<td>T4a</td>
<td>–</td>
<td>–</td>
<td>8%</td>
<td>8.3%</td>
<td>2%</td>
</tr>
<tr>
<td>Canis et al., 2014†</td>
<td>104</td>
<td>T4</td>
<td>2%</td>
<td>–</td>
<td>–</td>
<td>2%</td>
<td>–</td>
</tr>
<tr>
<td>Pantazis et al., 2015*</td>
<td>24</td>
<td>T3</td>
<td>91.7%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Peretti et al., 2016*</td>
<td>22</td>
<td>T3</td>
<td>95.4%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Vilaseca et al, 2016†</td>
<td>154</td>
<td>T3</td>
<td>75.2%</td>
<td>1.3%</td>
<td>2.6%</td>
<td>–</td>
<td>–</td>
</tr>
</tbody>
</table>

Table 5: Functional results after TLM for advanced glottic tumours (T3-T4)

<table>
<thead>
<tr>
<th>n</th>
<th>TNM</th>
<th>Site</th>
<th>LPR</th>
<th>LFS</th>
<th>FPR</th>
<th>Gastrostomy (definitive)</th>
<th>Tracheotomy (definitive)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ambrosch et al., 2001**</td>
<td>167</td>
<td>T2b-T3</td>
<td>G</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0.5%</td>
</tr>
<tr>
<td>Motta et al., 2005**</td>
<td>51</td>
<td>T3</td>
<td>80.5%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>–</td>
</tr>
<tr>
<td>Ninno et al., 2007**</td>
<td>117</td>
<td>T3-T4</td>
<td>41 G</td>
<td>65 S</td>
<td>86%</td>
<td>51%</td>
<td>7% survivors</td>
</tr>
<tr>
<td>Grant et al., 2007†</td>
<td>10</td>
<td>T3-T4</td>
<td>G</td>
<td>–</td>
<td>–</td>
<td>10%</td>
<td>10%</td>
</tr>
<tr>
<td>Olthof et al., 2009†</td>
<td>39</td>
<td>T3-T4</td>
<td>S+G</td>
<td>–</td>
<td>–</td>
<td>89.7%</td>
<td>30% survivors</td>
</tr>
<tr>
<td>Vilaseca et al., 2010†</td>
<td>51</td>
<td>T3</td>
<td>–</td>
<td>58.9</td>
<td>51%</td>
<td>–</td>
<td>10% all series</td>
</tr>
<tr>
<td>Peretti et al., 2010*</td>
<td>11</td>
<td>T3</td>
<td>72.7%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Blanch et al., 2011†</td>
<td>26</td>
<td>T3</td>
<td>LAC</td>
<td>–</td>
<td>65.5%</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Canis et al., 2013†</td>
<td>31</td>
<td>T4a</td>
<td>G</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Canis et al., 2014†</td>
<td>122</td>
<td>T3</td>
<td>83%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Breda et al., 2015†</td>
<td>40</td>
<td>T3-T4</td>
<td>G</td>
<td>69.2%</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Pantazis et al., 2015*</td>
<td>19</td>
<td>T3</td>
<td>73.7%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Peretti et al., 2016†</td>
<td>34</td>
<td>T3</td>
<td>85.3%</td>
<td>–</td>
<td>–</td>
<td>0%</td>
<td>0%</td>
</tr>
</tbody>
</table>

LPR: laryngeal preservation rate; LFS: laryngectomy-free survival; FPR: function preservation rate; LAC: laryngeal anterior commissure.
Open partial surgery for primary and recurrent laryngeal cancer

Giuseppe Sprano MD, Giuseppe Rizzotto MD, Andy Bertolin MD, Giuseppe Mercante MD, Antonio Schindler MD, Erika Crosetti MD, and Giovanni Succo MD

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Otolaryngology Dept – Vittorio Veneto Hospital, Treviso, Italy
Biomedical and Clinical Science Dept "L. Sacco" – University of Milan, Milan, Italy
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Abstract
The evolution of treatment for advanced laryngeal carcinoma is focusing on maintaining locoregional control while also maintaining a functional larynx. Open partial horizontal laryngectomies may be a viable option for intermediate and selected advanced laryngeal carcinoma while maintaining laryngeal function. Strict selection criteria, based on locoregional tumor extent as well as on patient’s general condition, allow excellent oncological outcomes. Whereas, albeit slightly worse, but similar, outcomes are also obtained in radio-recurrent patients, these procedures must be included as an option in selected patients with radio-recurrent laryngeal cancer.


Key words
Partial laryngectomy, laryngeal cancer, supracricoid laryngectomy, suprathyroid laryngectomy, salvage surgery

Introduction
The last two decades have gradually witnessed a paradigm shift in the treatment of laryngeal cancer with the primary focus now on organ and function preservation.2

Several approaches are available for the treatment of laryngeal cancer at the different primary tumour (T) and nodal (N) stages, with comparable rates of overall survival, locoregional control and laryngectomy-free survival.

The non-surgical approaches, primary radiotherapy (CRT) for early stages (I-II) and concurrent chemoradiotherapy (CT-RT) or induction chemotherapy followed by radiotherapy (IC-RT) for advanced stages (III-IV), have demonstrated that larynx preservation is feasible, even though the disappointing long-term results of CT-RT deserve further evaluation and investigation.

On the other hand, surgery can be offered to patients as a valuable method to preserve part of the larynx and its functions, avoiding the negative physical and psychological impact of a permanent tracheostomy. Open partial laryngectomies (OPHL) and transoral laser microsurgery (TLM) have been used extensively, with excellent results, as upfront treatment in early (I-II) as well as in more advanced stages (III). Even in the latter case, OPHLS have shown very good oncological outcomes, high laryngectomy-free survival, relatively low morbidity and mortality rates, and finally acceptable functional outcomes, as long as a careful selection of patients is carried out (good general and functional conditions, cN), patient compliance to an intensive rehabilitation protocol, different subcategories based on local T extent,3,4 After an appropriate selection, based essentially on pre-treatment and post-treatment tumour extent, OPHLS can also be used as salvage procedures in radio-recurrent and cancer, achieving comparable survival rates, an acceptable (although higher) morbidity, effective swallowing and a sufficiently intelligible voice.

Based on this scenario, a review of recent English language literature was carried out in an attempt to assess the changes which are occurring, based on indications, strategies and technique refinements. We focused only on OPHLS since vertical partial laryngectomies have now largely been replaced by TLM.
History and current classification
In the late nineteenth century, surgeons began to think about how to save part of the larynx and its functions, but was only in the second half of the last century that these interventions became established as an effective weapon in the treatment of laryngeal cancer, untreated or radioresistant in an intermediate stage and also in selected advanced stages.

Buck and Solis-Cohen in the USA, and Hautant and Leroux-Robert in France were the pioneers of vertical partial laryngectomies. Supraglottic laryngectomy, described by Alonso in 1947, opened the era of horizontal partial laryngectomies. Supraglottic laryngectomy, advanced stages.

It was only in the second half of the last century that these procedures and, in particular, OPHL type II.

In recent years, the treatment of laryngeal cancer has increasingly focused on tumour control as well as preservation of function and quality of life. Furthermore, diagnosis in such cases is more difficult because of the lack of exposure and in the case of bulky tumours. In patients affected by pT3a cancer, the 5-year locoregional control (LRC) drops to 64.8% and DFS to 52.7%. The functional results are comparable to those of the more studied type II OPHL.

Finally, a possible future role of type III OPHL could be that of a rescue intraoperative procedure if, during a type II OPHL, the resection margins are insufficient and thus allowing the surgeons to avoid shifting to an upfront total laryngectomy.

OPHL type II for recurrent laryngeal cancer after radiotherapy
In patients affected by pT7 cancer, the 5-year locoregional control (LRC) and DFS are 88.7% and 86.4%, respectively, while in patients with a pT4a, LRC drops to 64.8% and DFS to 52.7%. The functional results are comparable to those of the more studied type II OPHL.

In the 1990s, Laccourreye et al. reported a modification of conventional supracricoid partial laryngectomy (SCPL), removing the cricoid ring in the case of glottic tumours with poor control rates. Furthermore, diagnosis in such cases is more difficult because of the lack of exposure and in the case of bulky tumours. In patients affected by pT3 cancer, the 5-year locoregional control (LRC) drops to 64.8% and DFS to 52.7%. The functional results are comparable to those of the more studied type II OPHL.

Finally, a possible future role of type III OPHL could be that of a rescue intraoperative procedure if, during a type II OPHL, the resection margins are insufficient and thus allowing the surgeons to avoid shifting to an upfront total laryngectomy.

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Finally, a possible future role of type III OPHL could be that of a rescue intraoperative procedure if, during a type II OPHL, the resection margins are insufficient and thus allowing the surgeons to avoid shifting to an upfront total laryngectomy.
OPHL type II (including both supracricoid partial laryngectomy with cricohyoidoepiglottopexy, SCPL–CHEP or OPHL type Ia, and supracricoid partial laryngectomy with cricohyoidoepiglottopexy. SCPL–CIP or OPHL type Ib) can be adopted in many intermediate laryngeal cancers, strictly respecting tumour- and patient-related indications58-60.

Some reports have documented the feasibility of OPHL type II in terms of local tumour control and functional results after radiotherapy. However, there is a bias related to the small number of cases and the low number of centres performing this type of surgery. Paleri et al. published an interesting meta-analysis on radioimaging laryngeal squamous cell carcinoma in 560 patients treated with open conservative laryngectomy but not focused on OPHL51. In their reports, different partial laryngectomy techniques have been considered.

Since the first report of OPHL type II after radiation failure, many centres worldwide have adopted this procedure as salvage surgery. Different papers by authors from different countries have confirmed the feasibility of the technique51,61, and one was from Japan53.

A meta-analysis of those papers shows that the majority of cases occur in a low to intermediate stage (T1 27.5%, T2 42.2%, T3 21.3% and T4 7.2%). The surgical procedures were OPHL type Ia or type Ib.

Local control at 24 months may vary from 70% to 95%,5,3,4 and DFS at 36 months ranged between 70% and 90%5-9,23,69. As 5 years has been reported between 70% and 90%10,11,14,16,17,19.

Some cases require salvage or functional total laryngectomy because of re-recurrence or aspiration. The laryngeal preservation rate was 85.2%. The pooled mean decannulation rate was 91.2% based on data reported in the literature for over 200 patients. The incidence of laryngeal stenosis has to be considered to be a possible complication; this was described in 3.9% of cases and it may require further surgery to enhance airway patency and to facilitate tracheostomy decannulation. This event has been described in 6% of cases. An efficient swallowing was reported in over 90% of cases. The percutaneous endoscopic gastrostomy (PEG) dependence rate was 3.5%, while the aspiration pneumonia rate was 6.4% based on data reported for 221 patients.

Voice and speech outcomes are rarely reported in the literature, however, some data are available. In our earlier paper, we reported an ‘acceptable quality of voice for most patients’55. Voice quality was definitively hoarse in all patients and maximum phonation time (MPT) ranged from 3 to 18 s (mean, 8.3 s). Marchese-Ragona et al. and Leon et al. reported ‘satisfactory voice production in all cases’55. Similar results were also reported by Pellini et al. in a study in decannulated patients in which voice was evaluated as hoarse to varying degrees (19 patients grade 1, 49 grade 2, and 8 grade 3)55. MPT ranged from 2 to 18 s (mean, 7.9 s). Finally, Deganello et al. reported ‘satisfactory voice production that allowed normal social interactions’55.

Laryngeal recurrence represents the most salvageable area in head and neck with reported OS higher than 60% at 2 years54. Furthermore, it is more frequent in the primary site than in regional lymph nodes or distant sites55,56. Unfortunately, laryngeal salvage surgery corresponds to a decreased QoL and degradation of laryngeal function51.

Diagnosis of laryngeal recurrence after radiotherapy can be challenging because of chronic oedema and, occasionally, arytenoid fixation. Difficulties in obtaining a representative biopsy are common due to oedema, chronic inflammation and fibrosis with higher chances of false negative results55. Furthermore, imaging has low specificity after radiation therapy.

Recurrent and persistent laryngeal cancer after radiotherapy is associated with aggressive growth patterns, high extralaryngeal spread and subglottic involvement, and intravascular and perineural invasion51. As a result, total laryngectomy is the most performed salvage procedure in the case of recurrent/persistent LSCC after radiotherapy. However, salvage total laryngectomy is associated with an increased risk of wound and systemic complications. In the review of the literature performed by Goodwin51, major complications ranged from 5% to 48% while pharyngocutaneous fistula rates ranged from 30% to 80%51,52,56,64. In these scenarios, OPHLs have to be considered as a valuable alternative for selected recurrent laryngeal cancers.

Published papers showed a very good local control rate which was over 90% at 24 months. T1 stages in reports in the literature may range from 1 to T4 and the high rate of local control ensures that salvage OPHL type II can be a valuable treatment strategy in radioimaging tumours. The DFS rate at 36 months was 80%. This reflects the percentage of patients who are considered to be disease-free. Consequently, DFS evaluates not only local control but also regional control and distant metastases, and thus the functional consequence is a strained, deep and aesthetically altered voice, difficult to modulate and to raise; speech is composed of short sentences, because the patients are short of breath52. Maximum phonation time (MPT) implies adequacy of air support for speech and, in supracricoid laryngectomy and OPHL procedures, there is quite low probability of aspiration pneumonia due to a lower resistance of the neoglottis with consequent air loss during phonation51.

Thus, in order to compensate for the air wastage during phonation, the SCL patient needs to increase neoglottis resistance and subglottic pressure with consequent vocal fatigue because of the increased physiological effort required to phonate. Interestingly, inspiratory effort appears to be significantly affected by arytenoid removal, suggesting well-tolerated recovery of the glottal closure after removal of extralaryngeal arytenoid and reconstruction of the neoglottis47.

Only a few studies on functional outcome after OPHL included an assessment of QoL. Data are sometimes contradictory; it must be noted that QoL brings many factors into play, including the patient’s psychosocial traits, cultural and ethnic backgrounds. Therefore, it is not surprising that different authors report different QoL scores on a small number of subjects studied in different countries. When swallowing-related QoL was analysed, data suggest that swallowing difficulties have only limited impact on daily living activities. Moreover, it is to be noted that between 80% and 90% of patients undergoing OPHL achieve the ability to eat without restriction47.

Self-assessment data revealed a moderate impact on voice related QoL. (V-QoL) in terms of speech after OPHL type II and III, on the emotional, physical and functional levels of the voice handicap index (VHI), even if some authors reported high degrees of vocal handicap47. Since the voice is mainly used for everyday verbal communication, it is possible that VOCal is perceived by the patients as not being very compromised even if the voice per se is rather poor47.
Conclusion

Multiple single and multi-institutional series support the evidence that open partial laryngectomies represent a valid option for the treatment of laryngeal cancer in early, intermediate and also selected advanced stage, maintaining high OS, LRC and DFS as well as acceptable functional outcomes.

Whereas, albeit slightly worse, but similar, outcomes are also obtained in radiotherapy patients, these procedures must be included as an option in selected patients with radioresponsive laryngeal cancer.

References


Management of professional voice problems

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Abstract

Introduction: The term professional voice user encompasses a wide range of professionals for whom their occupation is dependant on having a functioning voice and is not limited to singers and actors.

Assessment: An adequate assessment of such patients with dysphonia requires a multi-disciplinary approach in a specialised voice clinic. A thorough history, clinical examination, perceptual evaluation of the voice is required to make an accurate and complete diagnosis as well as guiding treatment strategies.

Pathology: Causes of dysphonia can be broadly categorised into functional or organic/structural; however in clinical practice patients often have multiple aetiologies contributing to their voice disorder. Data on 1393 new patients seen in our voice clinic demonstrated that functional voice disorders, cysts and inflammatory laryngeal disorders were significantly higher amongst a subset of 255 singers.

Management: Multi-disciplinary management is essential to successfully treat dysphonia in professional voice users. Surgery can be helpful for pathology confined to the epithelium or superficial lamina propria but should not be considered a treatment in isolation. It is essential to elicit contributory psychological elements, as well as the impact the voice disorder is having on the patients professional demands.

Conclusion: Professional voice users with dysphonia require multi-disciplinary assessment and management. Their dysphonia is often multi-aetiological and all contributory factors must be addressed by the voice team in order to expedite successful, individualised treatment and manage patient expectations.
Assessment Of The Professional Voice User

Ideally all professional voice users should be seen in a dedicated voice clinic, of which there are now around 100 around the UK. As a minimum the patient will initially be assessed by an ENT surgeon with a special interest in voice along with a specialist speech and language therapist. The clinic should also be equipped with a range of laryngeal imaging equipment, including stroboscopy. The team can also include a singing teacher, psychotherapist or osteopath, voice scientist, psychiatrist and social worker. There should also be established referral pathways to relevant medical specialties such as neurology and gastroenterology.

The comprehensive assessment required of the patient with a voice disorder is detailed in Table 2. One of the most important features for the otolaryngologist is comprehensive history taking which can lead to a likely principal diagnosis prior to clinical examination. Key features of the assessment for the otolaryngologist are discussed below:

Voice History

It is important to determine the onset, duration and severity of voice symptoms. Enquiring about vocal load, and for singers in particular the frequency and duration of singing in their current schedule can provide important information, as well as the type of singer they are. Any upcoming major performances or specific career goals can dictate the urgency and intensity of therapy that may be required. Singers will often be more aware of fine changes in their voice production so enquiring as to whether there is a particular vocal range where their voice is maximally affected can help both the speech therapist and vocal coach focus their therapy appropriately.

Relevant Medical History

It is important to elicit any factors from the medical history that may be exacerbating the voice problem. These include:

- Preceding upper respiratory tract infection that may have triggered dysphonia.
- History of rhinitis, allergies, or chronic rhinosinusitis.
- History of gastro-oesophageal reflux disease or symptoms of laryngopharyngeal reflux.
- Smoking and alcohol history.
- Caffeine intake and hydration status.
- Stress, anxiety and psychiatric disorders.
- Musculoskeletal disease or trauma to the head and neck that may be affecting posture and muscle tension.

Examination

A general ENT examination should be performed to exclude contributory factors to the voice disorder such as tonsillar hypertrophy or nasal polyposis. Specifically, examination of the neck should include palpation of the extrinsic laryngeal musculature to assess for muscle tension and tenderness. The supra and infra hyoid muscles should be palpated. It can also be useful to palpate and displace the cartilage framework of the larynx to assess for tension and tethering. There is marked variability within published literature between these techniques with few validated objective tools to quantify degree of diagnostic accuracy of palpation but despite this palpation can provide valuable information contributing towards the diagnosis and treatment strategies.

The voice itself needs to be perceptually analysed. There are patient-centred or clinician-centred scoring tools. There are many patient-centred questionnaires with the most common being the VHI-10. The most commonly used clinician-centred analysis of voice is the GRBAS scale (Grade, Roughness, Breathiness, Asthenia, Strain). Visual examination of the larynx is imperative to assess for structural pathology and the dynamic function of the larynx. In addition vibratory patterns of the vocal cords can be assessed during phonation when voice-synchronised stroboscopy is adopted. Despite some modern advances, video-strobe-laryngoscopy is considered the gold standard in laryng imaging and can be performed using a 70° or 90° rigid stroboscope or a newer flexible chip-and-tip naso-laryngoscope, these have been shown to be nearly as good as the conventional rigid strob-laryngoscopy.

Pathology

Just as is the case with the general population, professional voice users can present with a wide variety of underlying pathologies and diagnoses for their dysphonia. In the broadest sense these can be categorised as functional or organic. It is important to also recognise that in many cases there may be more than one aetiological factor contributing to the voice problem.

The most common functional disorder that is seen is muscle tension dysphonia. This is a form of voice use / abuse / misuse that is characterised by excessive muscular effort and pressed phonation. This can also be referred to as muscle tension imbalance or laryngeal hyperfunction. Other causes of functional dysphonia can include conversion and psychogenic dysphonia. Due to the lack of uniformed terminology it is difficult to ascertain accurate estimates of the prevalence of such conditions.

The term organic dysphonia refers to either structural, neurological, endocrinological or laryngeal diseases. Structural causes can include vocal cord polyps, cysts, nodules, granulomas and many more. In clinical practice however it is clearly recognised that patients often have more than one pathology contributing to their voice problem and their dysphonia is a result of multiple aetiologies. According to work performed by Koufman and popularised by McGlashan these can be broken down into inflammation, neoplastic / structural, neuromuscular, and muscle tension imbalance. There is very often complex interplay between aetiologies; for example the patient with a vocal fold poly may then develop muscle tension imbalance as a compensatory mechanism. This categorisation and the interplay between pathologies are displayed in Table 3.

Table 2: Initial Assessment of Patient with a voice disorder

<table>
<thead>
<tr>
<th>Assessment</th>
<th>Approach</th>
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<tbody>
<tr>
<td>Joint Assessment</td>
<td>Visual examination</td>
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<td>Otolaryngologist &amp; SLT</td>
<td>Laryngolarygraphy synchronised stroboscopy</td>
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<td>Rigid endoscopy</td>
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<td>Flexible thermo-digital nasendoscopy</td>
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<td>Shimmer and jitter measurements</td>
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<tr>
<td>Perceptual Assessment</td>
<td>Study of the interrelation between speech subsystems</td>
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<td></td>
<td>Identification of parameters of voice contributing to the dysphonia</td>
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<td>Evaluation of each speech subsystem and potential for change</td>
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<td>Establishment of a baseline and a measure of overall severity</td>
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<td>Instrumental Assessment</td>
<td>Aerodynamics</td>
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<td>Pitch</td>
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<td>Intensity</td>
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<td>Resonance</td>
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<td>Vibratory Cycle</td>
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<td>Vocal Quality</td>
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<tr>
<td>Palpation of the Larynx: Laryngeal Muscle Assessment</td>
<td>Status of the extrinsic laryngeal musculature</td>
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<td></td>
<td>Position of the laryngeal cartilages at rest</td>
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<td></td>
<td>Position of the laryngeal cartilages during phonation</td>
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<td></td>
<td>Degree of muscle tension</td>
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<tr>
<td>Client Self-assessment</td>
<td>Self-perception of voice</td>
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<td>Impact of the voice disorder and symptoms on their life</td>
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<td></td>
<td>Profile of voice use Questionnaire</td>
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</table>

Table 3: Aetologies causing voice disorders and multifactorial causes – original figure – adapted from references detailed in separate table.

There is a paucity of evidence in the literature detailing the prevalence of structural laryngeal disorders amongst the general population, let alone amongst professional voice users. Our voice disorders unit has kept a database of all patients seen in our voice clinic since 2010, and whilst this data is as yet unpublished, we have analysed the data of 1393 new patients seen between 2010-2015. Of these patients 449 (32.3%) were classified as professional voice users and 255 (18.3%) were singers. Our data demonstrated around half of all patients were diagnosed with functional disorders, and the other half with structural pathology. This rate was not significantly different among the subgroup of professional voice users and was (p=0.0002*) and could be attributable to the differing expectations, training, and vocal ability in this group.

The most common pathology demonstrated in the whole cohort (19.1%) and amongst the subgroup of singers (33.3%) was vocal cord cysts (see Figure 1) and this was significantly higher among singers (p=0.003*). Inflammatory conditions including laryngitis, oedema and polyposid degeneration of the cords represented 31.0% of patients in the singers group and was their 2nd most
popular pathology, whereas it only represented 11.5% amongst the whole cohort. In our series singers were significantly more likely (p=0.001*) to have inflammatory pathology. There was no difference in the prevalence of vocal cord polyps (See Figure 2) between the two groups (p=0.4977*). Vocal cord nodules (see Figure 3) represented only 4.08% of structural diagnoses in our series, with this rate being 9.2% amongst singers, and this difference was not quite considered significant (p=0.052*).

The relevant data is detailed in Table 4. Overall vocal cord nodules were the 8th most common diagnosis in our series, whereas it only represented 11.5% amongst the professional voice users compared to 79 (11.5%) amongst the case series of voice users. A statistically significant difference was found (p=0.0001*) to the professional voice users with either a proton pump inhibitor14 or liquid alginate15. However, significantly more likely (p=0.0001*) to have inflammatory pathology. There was no difference in the prevalence of vocal cord polyps (See Figure 2) between the two groups (p=0.052*). Vocal cord nodules (see Figure 3) represented only 4.08% of structural diagnoses in our series, with this rate being 9.2% amongst singers, and this difference was not quite considered significant (p=0.052*).

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or on the free border of the vocal fold as this can cause tethering of the epithelium also affecting mucosal wave generation. Microsurgical and laryngeal surgery should never be seen in isolation as a definitive treatment and must always be followed (and in some cases preceded) with speech and language therapy. The saying “the operation is the easy bit, the recovery is the hard bit” is a statement that rings true for such patients and the value of therapy must never be underestimated when managing patient expectations.

Conclusion

The term “professional voice user” now encompasses a broad spectrum of people for whom their occupation is dependant upon having a functional voice, included in this are singers who often have differing expectations with an increased psychological stressors. There is a huge range of underlying pathologies that can be attributable to the voice disorder and often they are multi-aetiological. The multi-disciplinary team is vital to ensure successful treatment and the “voice clinic” where a joint assessment with speech therapist and surgeon working in synergy helps build trust, rapport and confidence in the diagnosis and treatment plan. It is imperative to understand and manage the expectations of the patient relevant to their professional demands and tailor care individually to try to meet these aims.

References

Management of head and neck sarcoma

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Abstract
Head and neck sarcomas are a rare diverse group of neoplasms arising within soft tissues or bones. In this rare tumour type, prospective trials are not feasible. Multimodality treatment plans including surgery, radiation and chemotherapy are often indicated. A multidisciplinary team (MDT) approach involving both sarcoma and head and neck MDTs is recommended for management of these tumours. This article provides an overview of the presentation, pathology and management of these lesions.


Key words
Sarcoma, head and neck cancer, staging, management

Conflict of interest
None

Introduction
Head and neck sarcomas are a rare heterogeneous group of cancers which can arise in the mesenchymal tissues (bones or within the soft and connective tissues). They account for approximately 2% to 15% of all sarcomas, and represent approximately 1% of head and neck malignancies. According to the National Cancer Intelligence Network (NCIN) data, between 1990 and 2010, 476 head and neck sarcoma patients were diagnosed in England, of which 793 (17%) arose in bones of skull and face, and 4003 (83%) within the connective and soft tissue (ratio of approximately 1:5). This was a diverse group with over 30 different histological types. The most common soft tissue sarcomas (STSs) are leiomyosarcoma, liposarcoma (Figure 1) and sarcoma not otherwise specified (NOS). The most common bone sarcomas (BSSs) are osteosarcoma, chondrosarcoma and Ewing’s sarcoma.

Due to the relative rarity of the condition, published data on management and outcomes of head and neck sarcomas are limited, based on small series of patients. Most of these studies report retrospective cohorts managed over a number of decades to obtain a sufficient numbers. Evolution of imaging, surgical technique and adjunctive therapies make conclusions from such small series difficult to interpret.

Guidelines from the British Sarcoma Group (BSG)1, and the European Society for Medical Oncology (ESMO)2 have been published on sarcoma management. However, these are not specific to head and neck sites. In contrast to sarcomas elsewhere, wide surgical margins are more difficult to achieve and the cosmetic and functional impact of major head and neck resections are significant. A multidisciplinary team (MDT) approach involving both sarcoma and head and neck MDTs is recommended for management of these tumours.3

Aetiology
The majority STTs of the head and neck arise sporadically with no obvious cause. However, various familial syndromes, environmental carcinogens and oncogenic viruses along with previous exposure to ionizing radiation have been implicated. In patients with type 1 neurofibromatosis, there is up to a 10% cumulative lifetime risk of developing sarcoma, usually malignant peripheral nerve sheath tumour (MPNST).4 Li-Fraumeni syndrome predisposes the individual to developing STTs, osteosarcoma, pre-menopausal breast cancer, brain tumours and adrenal cortical carcinoma.5 Gardner syndrome, a subtype of familial adenomatous polyposis, has also been linked to STTs.6

Radiation exposure is a recognised risk factor in the development of secondary sarcoma. Overall, osteosarcoma is the most common radiation-induced sarcoma for body sites. In the head and neck region, malignant fibrous histiocytoma is the most common subtypes seen after ionizing radiation exposure.7 The latent period between initial radiation and diagnosis of sarcoma ranges from 9.45 years with a median of 17 years. Chemical agents such as vinyl chloride gas (plastics industry), chlorophenols (sawmill workers) and arsenic (vineyard work) have been implicated as possible causative factors.8 Associations have also been shown between human immunodeficiency virus and human herpesvirus 8 in Kaposi sarcoma9, and for Epstein-Barr virus and smooth muscle tumours in immuno-compromised patients10.

Presentation
Head and neck presentation is dependent on the involved primary site. Lesions arising from the subcutaneous tissues of the face, neck or scalp present with a superficial mass which may be painful. Lesions arising from the upper aero-digestive tract, paranasal sinus and orbit will cause symptoms related to these areas, such as dysphonia, dysphagia, ophthalmology, nasal obstruction, proptosis and diplopia. Cranial nerve deficits may be seen in skull base lesions. The most frequently involved anatomical sites are the superficial neck and parotid, sinonasal tract and visceral spaces of the neck.11

Most head and neck sarcomas present with localised disease. Regional lymph node involvement is unusual although is more frequently associated with specific histological subtypes including rhabdomyosarcomas and epithelioid sarcoma.12 The most common site of distant metastasis is the lungs. Mehendall et al reported that in the absence of lung metastasis, the risk of metastasis to other distant sites is extremely low.13

Investigation
The evaluation of suspected head and neck sarcoma involves cross-sectional imaging and biopsy. Both computed tomography (CT) and magnetic resonance imaging (MRI) provide information regarding loco-regional extension, assessment of tissue composition and the presence of distant disease.14 Although CT provides better bony detail, advantages of MRI include superior soft tissue resolution. In many cases the information provided is complementary. Cross-sectional imaging is best performed before biopsy in order to preserve diagnostic accuracy. Chest CT is recommended to exclude pulmonary metastasis prior to definitive treatment in all cases.15 18F-flurodeoxyglucose positron emission tomography (18F-FDG-PET) is increasingly used in pre-treatment staging although it is currently not considered standard.16

Diagnostic tissue samples can be obtained using fine needle aspiration, core biopsy or open biopsy if required. The biopsy should be planned in a way that the tract can be safely excised at the time of definitive surgery in order to reduce the potential risk of tumour seeding.

Pathology & grading
Histological diagnosis should be based on the 2013 World Health Organisation (WHO) classification of tumours of soft tissue and bone.17 Numerous changes have taken place in STS and BSS classification, predominantly based on the identification of new genetic findings in different tumour types.

A range of outcomes are associated with histological subtypes of sarcoma. For example, all angiosarcomas are considered high grade and should be treated as such. In contrast, many chondrosarcomas are associated with slow growth and low rates of metastasis. Histological subtyping therefore remains critical in predicting the biological behaviour of tumours.

The malignant grade should also be provided in all sarcoma cases for the purpose of prognostication. The “Federation Nationale des Centres de Lutte contre le Cancer” (FNCLCC) grading system is generally used in STS, which distinguishes three malignancy grades based on differentiation, necrosis and mitotic rate.18 Table 1. Again, grading should be performed prior to treatment as neoadjuvant medical therapies may affect these findings and limit the usefulness of grading of a subsequent resection specimen.

The ESMO guidelines do not provide a specific recommendation for BS histological grading. Instead, BSSs are graded according to the cell type and differentiation of the stromal component of the tumour. Less differentiated tumours are given higher grades. Chondroblastic and fibroblastic tumours are usually of lower grade (grade 1 or 2). An absence of cartilaginous lobulation and the presence of spindle cell forms are characteristics of high-grade (grade 3) lesions and are associated with poorer prognosis. Ewing’s sarcoma is always classified as high grade.19

Five-year overall survival is better in low-grade tumour and poor outcome is associated with positive surgical

<table>
<thead>
<tr>
<th>Tumour differentiation</th>
<th>Necrosis (macro and micro)</th>
<th>Mitotic count (p10 high-power fields)</th>
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<tbody>
<tr>
<td>1: Well</td>
<td>0: Absent</td>
<td>1: n&lt;10</td>
</tr>
<tr>
<td>2: Moderate</td>
<td>1: &lt;50%</td>
<td>2: 10-19</td>
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<tr>
<td>3: Poor</td>
<td>2: &gt;50%</td>
<td>3: na&lt;20</td>
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Table 1: Federation Nationale des Centres de Lutte Contre le Cancer histological grading criteria

The sum of the scores of the three criteria determines the grade of malignancy

Grade 1: 2 and 3; Grade 2: 4 and 5; Grade 3: 6, 7 and 8.
chemo-radiation is controversial. Some use neo-adjuvant therapy in order to help achieve a negative surgical margin, whereas others recommend post-operative adjuvant therapy. (Flow chart 1)

**Management - STSs**

The primary treatment modality for STSs is surgery aiming to achieve a wide excision with negative margins. Although there is no consensus regarding margin width, a 2cm margin is considered acceptable. This presents a challenge in head and neck due to the need to preserve critical structures, functional anatomy and in order to minimise cosmetic defects. Hence, head and neck STSs tend to have lower local control rates and worse survival when compared to extremity STSs. Effective treatment of the neck is rarely indicated due to the low rates of nodal disease.

Adjuvant radiotherapy should be considered in cases with intermediate to high grade, lesions >7cm, and for resections with close or positive margins. In addition, recurrence after surgical management alone will be considered for re-resection with post-operative radiotherapy. In low-grade cases, adjuvant radiotherapy may be avoided. However, the complexity of these cases means that each patient should be discussed in a MDT setting. Farhood et al reported that with head and neck STSs, a combined modality approach showed a local control rate of 90% versus 52% in those treated with surgery alone. Similarly, Tran et al published an 87% recurrence-free survival with adjuvant radiation therapy versus 45% with surgery alone. In R1 or R2 resection margins, Barker et al reported an increase in local control from 25% to 54% with adjuvant radiation therapy. These results, while not conclusive, suggest a low threshold for recommending adjuvant radiotherapy is appropriate.

**Management - BSs**

Surgical resection remains the mainstay of treatment for head and neck osteosarcoma. Adjuvant radiotherapy has been associated with improved outcomes for patients with adverse features such as large tumour size, extensive soft tissue infiltration and lymphovascular invasion. However, osteosarcomas are relatively radio-resistant and doses in excess of 60Gy in conventional fractionation are recommended if feasible. In the head and neck region, this is problematic due to close proximity to vital structures and therefore the risk of serious morbidity from treatment. Techniques such as 3D conformal radiotherapy and intensity-modulated radiation therapy could be utilised to overcome these issues. The role of chemotherapy is still ill-defined due to conflicting study results.

Data from Memorial Sloan Kettering Cancer Centre (MSKCC) for head and neck osteosarcoma did not show a local control benefit with adjuvant chemotherapy. However, there is no consensus on the current role of adjuvant chemotherapy. The most commonly used chemotherapeutic drugs have been doxorubicin, dacarbazine and ifosfamide. Published study results have been conflicting. The Sarcoma Meta-analysis Collaboration (SMAC) published a meta-analysis reporting improved local control and metastasis-free survival with adjuvant chemotherapy, but no benefit in overall survival. Hence, adjuvant chemotherapy is not standard treatment in adult-type STSs although some of the sarcomas seen more commonly in the paediatric and adolescent population such as Ewing’s sarcoma, rhabdomyosarcoma of embryonal (Figure 2) and alveolar types are sensitive to these chemotherapeutic drugs.

**Radiotherapy alone may be the only option in management of select STSs where surgical resection is not achievable. However, the control rate for treatment with radiotherapy alone is reported to be approximately 25%-24%. The use of intensity modulated radiation therapy may allow higher doses for better control rate whilst limiting toxicity to normal tissue. As experience with proton therapy increases, the potential advantages of high dose delivery with minimal collateral tissue damage may define a role for this approach in selected cases.**

There is no consensus on the current role of adjuvant chemotherapy. The most commonly used chemotherapeutic drugs have been doxorubicin, dacarbazine and ifosfamide. Published study results have been conflicting. The Sarcoma Meta-analysis Collaboration (SMAC) published a meta-analysis reporting improved local control and metastasis-free survival with adjuvant chemotherapy, but no benefit in overall survival. Hence, adjuvant chemotherapy is not standard treatment in adult-type STSs although some of the sarcomas seen more commonly in the paediatric and adolescent population such as Ewing’s sarcoma, rhabdomyosarcoma of embryonal (Figure 2) and alveolar types are sensitive to these chemotherapeutic drugs.

Data from Memorial Sloan Kettering Cancer Centre (MSKCC) for head and neck osteosarcoma did not show a local control benefit with adjuvant chemotherapy. However, there is no consensus on the current role of adjuvant chemotherapy. The most commonly used chemotherapeutic drugs have been doxorubicin, dacarbazine and ifosfamide. Published study results have been conflicting. The Sarcoma Meta-analysis Collaboration (SMAC) published a meta-analysis reporting improved local control and metastasis-free survival with adjuvant chemotherapy, but no benefit in overall survival. Hence, adjuvant chemotherapy is not standard treatment in adult-type STSs although some of the sarcomas seen more commonly in the paediatric and adolescent population such as Ewing’s sarcoma, rhabdomyosarcoma of embryonal (Figure 2) and alveolar types are sensitive to these chemotherapeutic drugs.

**Management - BSs**

Surgical resection remains the mainstay of treatment for head and neck osteosarcoma. Adjuvant radiotherapy has been associated with improved outcomes for patients with adverse features such as large tumour size, extensive soft tissue infiltration and lymphovascular invasion. However, osteosarcomas are relatively radio-resistant and doses in excess of 60Gy in conventional fractionation are recommended if feasible. In the head and neck region, this is problematic due to close proximity to vital structures and therefore the risk of serious morbidity from treatment. Techniques such as 3D conformal radiotherapy and intensity-modulated radiation therapy could be utilised to overcome these issues. The role of chemotherapy is still ill-defined due to conflicting study results.

Data from Memorial Sloan Kettering Cancer Centre (MSKCC) for head and neck osteosarcoma did not show a local control benefit with adjuvant chemotherapy. However, there is no consensus on the current role of adjuvant chemotherapy. The most commonly used chemotherapeutic drugs have been doxorubicin, dacarbazine and ifosfamide. Published study results have been conflicting. The Sarcoma Meta-analysis Collaboration (SMAC) published a meta-analysis reporting improved local control and metastasis-free survival with adjuvant chemotherapy, but no benefit in overall survival. Hence, adjuvant chemotherapy is not standard treatment in adult-type STSs although some of the sarcomas seen more commonly in the paediatric and adolescent population such as Ewing’s sarcoma, rhabdomyosarcoma of embryonal (Figure 2) and alveolar types are sensitive to these chemotherapeutic drugs.

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demonstrate improved local control or disease-specific survival with the addition of neoadjuvant chemotherapy to conventional management. In contrast, Sniecik et al reported that patients’ overall and disease-free survival were significantly improved by treatment with chemotherapy for those who had complete and incomplete surgical removal of the tumour.

Obtaining negative surgical margins from complete surgical excision has been reported to be crucial not only for local control, but disease-specific survival. Survival rate has been reported to decrease from 75% to 35% in the presence of positive surgical margins or poor functional outcomes. Given the rarity and heterogeneity of this clinical entity, there is necessity for specialist treatment in a multidisciplinary context. In the UK, there are continuous improvements to normalise treatment pathways and standardise data collection for H&N STS, using the sarcoma network to allow meaningful interpretation of outcomes for this challenging patient group.

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17. Laskar S, Basu A, Muckadad MA, D'Cruz A, Pai S, Jambhekar N. Head and neck sarcoma. Wide surgical margins are difficult to achieve, due to close proximity of vital structures, which may compromise outcome. Therefore, adjuvant radiotherapy may be recommended based on adverse tumour or surgical features in order to minimise the risk of local recurrence. Although chemotherapy has a well-defined role in certain tumour subtypes such as rhabdomyosarcoma, its role in adult sarcoma management is currently unclear.

Figure 1: An MRI scan showing liposarcoma of right orbit

Figure 2: An MRI scan showing embryonal rhabdomyosarcoma of the tongue base

Figure 3: A CT scan showing chordoma of the larynx involving the left cricoid cartilage

specific survival rates for 10 and 20 years have been reported at 82% and 68%.

Conclusion

Sarcomas of the head and neck are a rare and diverse group of mesenchymal tumours. The mode of presentation will vary with anatomic subsite and is similar to more common head and neck malignancies. All patients require cross sectional imaging (often both MRI and CT), which is ideally performed prior to biopsy.

The wide varieties of histological subtypes encountered are associated with a spectrum of outcomes, from aggressive lesions such as angiosarcoma to the more indolent behaviour of fibrosarcomas. Further prognostic information can be gained from standardised systems of histological grading.

Surgery remains the main therapeutic modality for head and neck sarcoma. Wide surgical margins are difficult to achieve, due to close proximity of vital structures, which may compromise outcome. Therefore, adjuvant radiotherapy may be recommended based on adverse tumour or surgical features in order to minimise the risk of local recurrence. Although chemotherapy has a well-defined role in certain tumour subtypes such as rhabdomyosarcoma, its role in adult sarcoma management is currently unclear.

Given the rarity and heterogeneity of this clinical entity, there is necessity for specialist treatment in a multidisciplinary context. In the UK, there are continuous improvements to normalise treatment pathways and standardise data collection for H&N STS, using the sarcoma network to allow meaningful interpretation of outcomes for this challenging patient group.
Transoral endoscopic ultrasonic surgery (TOUSS) as an alternative to TORS.

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Abstract
Transoral Ultrasonic Surgery (TOUSS) is a new endoscopic minimally invasive procedure to approach pharyngeal and laryngeal lesions without a robotic platform. The fundamental pillars of this procedure are the Feyh-Kastembauer retractor, the deflecting tip videoendoscope, that allows a high quality image with easy refinement of the endoscopic view, and the ultrasonic scalpel (Thunderbeat). Besides de benign lesions, both early and locally advanced carcinomas are approachable with TOUSS. Many indications have been described, including the TOUSS-Total Laryngectomy technique, and first clinical experience has already been published. Further experience with the technique will show the benefits in terms of functional results and quality of life as well as the cost-eficacy compared with other treatment modalities, especially with other transoral endoscopic procedures that required expensive equipment as transoral robotic surgery (TORS). TOUSS is a low cost procedure that will easily spread the transoral endoscopic surgery to every patient and institution.

Key words
TOUSS, TORS, Thunderbeat, transoral endoscopic surgery.

Disclosure
Dr. Mario Fernandez served as consultant for Olympus Europa SE & Co. KG.

Introduction
There is no doubt about the fact that minimally invasive surgery is the pathway that leads to the future of head and neck surgery. In this sense, transoral laser microsurgery (TOLM) was the most remarkable step forward in the last decades. However, it seems that TOLM has touched the ceiling due to the limitations of the microscope as visualization instrument and the low coagulation capabilities of the laser. On the other hand, considerable progresses are seen in endoscopic head and neck surgery such as nasosinusal and cranial base surgery. The way for transoral endoscopic surgery was opened by Gregory Weinstein on de basis of a robotic procedure, and great advances are evident in the literature. However, the high costs of acquisition, maintenance and disposables, are impairing the spread of the transoral endoscopic philosophy. It is mandatory to design a more affordable procedure to let all surgeons and institutions start with the transoral endoscopic surgery.

This paper describes a novel endoscopic technique, TOUSS – TransOral Ultrasonic Surgery, that combines ultrasonic energy, with high potential for cutting and coagulating, and high definition videoendoscopic imaging. The combination of both technologies allows the same output as robotic surgery for transoral approach of the upper aerodigestive way.

TOUSS set up
After general anesthesia, the intubation is done through the mouth to face posterior or lateral lesions. A nasopharyngeal intubation allows a more confortable procedure in laryngeal and the base of the tongue lesions. The patient is placed in supine position, without any elevation of the shoulders or neck extension. The upper dental arch is protected with a plastic teeth protector. We prefer the Feyh-Kastembauer retractor (FK)
to achieve a proper exposition of the laryngo-opharynx. The FK is fixed with a retractor holder with chest support that allows an easy releasing and refinement of its position during the procedure.7)

The scope holder arm is attached to the left side of the table. The endoscopic view is obtained with a 5 mm videocystoscope that is fitted into the scope holder. Perhaps 3D imaging will demonstrate its advantages in order to a better space positioning, however we have no data related to this point yet. Olympus ENDOYE with the deflecting tip that allows an easy correction of the visualization up to 100º using the joysticks placed at the head the camera. (Figures 1, 2)

The optimal exposition for each patient and each anatomical area should be reached with the correct combination of the appropriate FK blade and its anterior angulation.

In order to get a comfortable procedure and a clean surgical field, the ultrasonic scalpel is mandatory. The ultrasonic scalpel Thunderbeat™ incorporates also a bipolar vessel sealing system that increases the safety of the procedure, especially when vascular structures as lingual or pharyngeal arteries are exposed within the pharynx. The 35cm pistol grip Thunderbeat™ allows stabilization of the instrument in the surgeon’s hand. This consideration is important as there are no structures like access ports to stabilize the instrument. Other grasping laparoscopic forceps are used to manipulate the tissues.

**Surgical technique**

The surgeon is standing up at the head of the patient with the patient positioned at the level of the surgeon’s elbows to get an ergonomic posture.7

One of the key aspects of the procedure is to take time for a correct exposition of the lesion with the FK retractor and the surgical planning of the resection. A lack of surgical planning can lead to the disturbance of the already resected tissue, and the inadequate endoscopic control of the surgical margins that force the surgeon to make a piecemeal resection. This concept is critical at the base of the tongue due to the restrictions of the angle of approach. The resection should start from the superior and lateral mucosal limits. Afterwards the deep resection will release the lesion that will remain attached to the tongue only by the inferior margin. Now the mobility of the specimen will let finish the resection pulling the inferior border of the specimen with the forceps, allowing a vertical section of the inferior limit with the ultrasonic scalpel. (Figure 3, 4, 5).

The coagulation potential of the ultrasonic energy is not necessary at the level of the mucosa. Moreover, the thermal spread to the mucosal surgical margin should be avoided in order to preserve the quality of the specimen. The employ of monopolar tungsten tips to make the initial mucosal incision will let the transection of the mucosa, and the subsequent use of the ultrasonic scalpel at the level of the peripheral margin will avoid its contact while it is activated. This aspect is critical to let the pathologist recognizes the oncological safety of the tumoral resection. The area of artefact in contact with the tumor will create doubts about the safety surgical margin, even with a good oncological resection. However, when there is a sector of normal mucosa between the artefact and the tumor, the artefact can be added to the normal mucosa to determine the safety distance between the tumoral front and the surgical margin. (Figure 6). A long suction cannula should be kept beyond the endoscope tip in order to avoid the smoke overclouds the endoscopic vision when the ultrasonic device is activated.

**Indications**

Regarding the pharynx, oropharyngeal and hypopharyngeal lesions are approachable with TOUSS.7 The approach of posterior and lateral pharyngeal lesions is easy and intuitive. The exposition does not play an important role in such anatomical areas. (Figure 7). The base of the tongue lesions are more dependent on the quality of exposition and the appropriate planning of the resection. It is not uncommon to reach the area of the entrance of the lingual artery at the base of the tongue. The pharyngeal lumen is a septic and wet cavity where the tightness of the vessel sealing line can be jeopardized. In this particular cases, the lingual artery can be easily identified and sealed safely with the Thunderbeat. The bipolar sealing system controls

**Figure 1, 2:** Disposition of FK retractor scope holder and Videolaparoscope. The screen is located at the feet of the patient in an advanced position.

**Figure 3:** Planification of the resection starting on the superior and lateral margins

**Figure 4:** Compression with the blade on the superior aspect of the tongue base will protrude the tissue and a straight access can be achieved

**Figure 5:** After the deep resection, the traction on the inferior aspect of the specimen will face the inferior border of the lesion against the ultrasonic scalpel

**Figure 6:** a: Conventional surgical margin with cold knife; b: Surgical margin with artefact in ultrasonic scalpel. The artefact is in contact with the tumor and no possibility to ensure the safety of the resection; c: Uncertain surgical margin.
Endoscopic vision represents an advantage over the microscopic, as it allows to get closer to the pathology, as well as the possibility to have the whole surgical field on focus. The deflecting tip of new videolaryngoscopes leads to easy and precise refinements of the endoscopic vision which can be done with the joystick at the head of the camera.

Finally, we found in the ultrasonic scalpel the answer to the necessity for a better coagulation tool, especially compared to the laser. It can cut tissue and coagulate up to 6 mm. The ultrasonic scalpel came into head and neck surgery from the hand of Paolo Miccoli as a cutting-coagulation tool for minimally invasive video-assisted thyroidecotomy (MIVAT) and it was the ideal instrument to step forward in developing a new systematic for transoral surgery. Thunderbeat™ incorporates a bipolar vessel sealing system that offers the highest safety in preservation of the vocal fold mobility, intraoperative and postoperative haemorrhagic complications. 

On this basis Transoral Ultrasonic Surgery (TOUSS) was developed as a transoral endoscopic technique, exporting to the head and neck surgery the concepts of laparoscopic surgery. The philosophy, the concept and the technology used in laparoscopic surgery, especially in single-port laparoscopy, are now applied to neck surgery, considering the mouth as a single port.

This concept has been previously developed by Gregory Wenstein since 2005, with the description of transoral robotic surgery (TORS) in an animal model. However, the ultrasonic scalpel is not a bendable tool as a straight shaft is mandatory to transmit the piezoelectric energy to the tip of the instruments, so it is adaptable to a robotic arm but there is no possibility to take advantage of the robotic endorobot. The direct manipulation of the tissue with TOUSS keeps the tactile input, and it represents an additional advantage compared to TORS.

Besides the costs of acquisition and maintenance of a surgical robot, the doubts around the need of such degree of freedom inside the pharynx, makes transoral robotic surgery a questionable solution in short and medium term.

Now TOUSS offers a more powerful setup to reach the transoral removal of lesions in areas where TOLM have bleeding problems and complications. The potential of TOUSS has allowed the transformation of open procedures like total laryngectomy in a transoral non-robotic endoscopic technique. The advantages of TOUSS for the patients are related to the avoidance of open approaches through healthy tissue. This is especially remarkable in radiated patients. In such patients, the avoidance of a mandibulotomy has a positive impact in the patient quality of life.

The robotic equipment is unreachable for most of ENT departments and usually the cost-benefits studies are not encouraging. TORS (transoral robotic surgery) was the first proposal for a transoral and endoscopic approach, but TOUSS is the first one that is feasible and reachable for most institutions. We believe that further cost-efficiency studies of TOUSS compared with TORS will show no doubt about its role in the present and the next future.

Discussion

TOLM has entered our specialty in the 80s and indications have been developed gradually to establish itself as an alternative to open partial laryngeal surgery. The approach of pharyngeal lesions was equally feasible from this microscopic approach. The success of TOLM relies on the possible resection of the lesions without any direct manipulation of the tissue, so it is adaptable to a robotic arm but there is no possibility to take advantage of the robotic endorobot. The direct manipulation of the tissue with TOUSS keeps the tactile input, and it represents an additional advantage compared to TORS.

Besides the costs of acquisition and maintenance of a surgical robot, the doubts around the need of such degree of freedom inside the pharynx, makes transoral robotic surgery a questionable solution in short and medium term.

Minimally invasive surgery has one of its developments fronts in thoracic and abdominal laparoscopic surgery. This systematic has been exported to the neck by Gagner to approach thyroid and parathyroid glands. Gagner’s setup has not been very successful, however it has opened the door to a new concept in neck surgery that is minimally invasive transcutaneous endoscopic surgery, using the endoscope as a visualization tool for neck surgery.

Dysesthesia of the patients, the complications rate and the management of postoperative complications. (Figure 8) The first two patients have been reported, and 6 patients have been already treated with TOUSS-TL. The surgery was complete in all cases and we do not registered any aborted case due to a lack of adequate exposition or intraoperative complication. Only two fistulas were registered, both of them in the two previously radiated patients. Both cases were treated satisfactorily with conservative treatment and no additional surgical treatment was necessary. The benefits in terms of oncological and functional results and quality of life will be reported in the long run.

Figure 8: 10 days postop after a TOUSS Total Laryngectomy. Notice the absence of neck scars and the good shape of the cervical tissues

Transoral Ultrasonic Total Laryngectomy (TOUSS-TL)

Perhaps Transoral Total Laryngectomy is the indication where the big potential of this concept can be better shown. TOUSS-TL consists in the complete removal of the larynx through a combined transoral and trans-stomal endoscopic cervical approach. The surgical steps are already described on the basis of the cadaver lab. The hypothesis of preserving the vascularization of the tissues of the anterior aspect of the neck, avoiding the approach through external neck incisions will improve the neck the amount of energy that it is needed to deliver on the vessel in order to get a safe closure.

Related to the larynx, our current limitation is the procedures that involves the vocal folds. The high coagulation potential of the ultrasonic scalpel is too aggressive and not necessary in the glottic plane. Additionally, in this area, the surgical margin can be as low as 2 mm to ensure the local control. So, there are other energies on more delicate instruments that can preserve much better the remaining normal mucosa.

The TOUSS supraglottic laryngectomy is feasible, but it was necessary. The benefits in terms of oncological and functional results and quality of life will be reported in the long run.

References

Management of occult primary in head and neck

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Abstract

Background. An occult primary tumor metastatic to cervical lymph nodes represents a rare clinical entity posing several diagnostic and therapeutic dilemmas.

Methods. Review of recent literature on diagnostic and therapeutic approaches in this clinical setting.

Results. Besides clinical examination, cross-sectional imaging and ipsilateral tonsillectomy are the mainstay of the diagnostic algorithm. Low-volume neck disease can be treated with either surgery or radiotherapy alone, whereas combined modality treatment is needed in more advanced disease. The rate of emerging mucosal primary tumors is comparable to the rate of metachronous primaries in patients with a known primary tumor which allows, in the majority of cases, treatment of the involved neck only. The impact of the p16/human papilloma virus status of metastatic nodes on treatment decisions and intensity will be defined in current treatment de-escalation clinical trials conducted in patients with a known primary.

Conclusions. After complete diagnostic workout the risk of missing an occult primary is low. Thus, treatment limited to the involved side of the neck is usually justified, with the advantage of lower toxicity and better prospects for salvage treatment when necessary.


Key words
unknown primary tumor; cervical lymph node metastases; squamous cell carcinoma, therapy

Conflict of interest statement
The author declares that he has no conflict of interest.

Introduction
A cancer of unknown primary (CUP) site is defined as the presence of lyphmatic or hematogenous metastases with no primary tumor discovered after rigorous diagnostic work-up. It represents a distinct clinical entity that accounts for up to 5% of patients with solid tumors. The reasons for an unrevealed primary tumor can be numerous and multifactorial, starting with its small size to dormancy or involution as a result of immunologic and apoptotic processes. Recently, the role of senescence, the microenvironment and tumor stem cells in impeding tumor growth has been suggested.

Compared to extranodal involvement, the exclusively nodal presentation of metastases is less frequent. Neck lymph nodes are the most commonly involved nodal group with squamous cell carcinoma (SCC) as the predominant histologic type; adenocarcinoma prevails in other nodal regions. In addition, CUP patients with SCC metastases to the neck nodes are granted a favorable prognosis, particularly when compared to those with metastases of other histologic backgrounds. The following discussion refers to CUP with cervical lymph node metastases of SCC histology.

Clinical presentation
Population-based incidence of CUP metastatic to neck nodes was reported to be around 3% and is influenced by the diagnostic algorithms used to reveal the hidden primary tumor. According to reports of larger series and literature reviews, the typical patient is male in his 6th or 7th decade of life, a heavy smoker and a consumer of alcohol and presents with a level II neck node of N2a classification. A painless neck mass, first observed up to 5 months before setting the diagnosis, is usually reported.

In the era of a global epidemic of human papillomavirus (HPV)-associated oropharyngeal SCC this stereotype might be shifted toward a younger age, different lifestyle orientations and more advanced nodal disease at presentation.

Diagnostic workup
In the patient presented with a mass in the neck and no clinical signs or symptoms of a primary tumor or active
inflammation in the head or neck, the diagnostic procedure is focused on the verification of the mass and the systematic evaluation of upper aerodigestive tract mucosa to identify the primary tumor. Assessment for distant metastases is also an integral part of the diagnostic algorithm in CUP patients. If the mass is located in the lower neck or in the supravacularar region, a primary is usually hidden below the clavicles and diagnostic efforts should be modified accordingly.

Identification of primary tumor (Fig. 1)

Fine-needle aspiration biopsy (FNAB) is the most elegant, safest and most cost-effective way to characterize FNAB is non-diagnostic, or in patients with a high suspicion of lymphoma. Particularly, open biopsy is discouraged due to potential negative consequences (i.e. the risk of local recurrence and complications, systemic dissemination); when performed, it must follow non-invasive diagnostics and must be accomplished by adequate definitive treatment of the neck. Any attempt to identify a primary tumor starts with a detailed clinical examination and fiberoptic endoscopy of upper aerodigestive tract mucosa. With the advent of cross-sectional imaging a better presentation of areas traditionally difficult to examine by surgical endoscopy (e.g. hypopharynx, nasopharynx) was possible which successfully reduced the historically high rate of emerging primaries in these areas. Suspicious imaging findings direct subsequent biopsies: under these conditions Cianchetti et al. reported a primary tumor detection rate of >60% compared to 29% in patients with no suspicious findings on radiographic examinations. Given the sensitivity of modern imaging, the majority of undiscovered primaries are hidden in crypts of tonsillar and lingual tonsils which makes random biopsy sampling obsolete. In particular, this is the case in HPV-associated neck metastases which highlights the importance of p16/HPV status determination in FNAB samples. The same relationship exists between EBV positivity of the involved neck node and the risk of an occult primary in the nasopharynx.

In this context, directed biopsies (by clinical and imaging findings) and an ipsilateral palatine tonsillectomy, followed in cases of an unconfirmed primary with base of tongue resection, are indicated. Deep tonsil biopsy offers a significantly lower likelihood of finding an occult primary compared to a tonsillectomy (11.6% vs. 40.7%). A bilateral procedure seems unnecessary, even if occult bilateral tonsil tumors were reported in up to 23% of cases: also in cases of a known early-stage tonsillar primary tumor treatment is usually unilateral. Removal of the lingual tonsil in a manner comparable to a palatine tonsillectomy didn’t become possible before the development of transoral base of tongue surgery. Combined or sequential (i.e. if no primary is found in the palatine tonsil on frozen sections) removal of both tonsils on the affected side resulted in notable rates of identified occult primaries, approaching 90%, especially in patients with p16/HPV positive neck metastasis. It appears that the implementation of transoral base of tongue surgery into the diagnostic algorithms in CUP is the major contribution to the improved detection rate of occult primaries in these patients.

At the moment, the added value of [18F]fluordeoxyglucose positron emission tomography and/or CT (FDG-PET/CT) in the search for hidden primary tumors in the head and neck seems to be marginal. The main reasons are the detection capability of the procedure which is limited to tumors ≥5 mm, basal uptake of FDG in normal lymphoid tissues, and salivary gland sequestration of FDG that resulted in relatively high false-positive (up to 35%) but also false-negative rates (up to 31%). Thus, a diagnostic biopsy cannot be avoided. Recent meta-analysis confirms these findings and observations: the authors reported a sensitivity and specificity of 0.97 (0.63-0.99) and 0.68 (0.49-0.83), respectively, a diagnostic odd ratio (DOR), positive likelihood ratio (PLR) and negative likelihood ratio (NLR) of 60 (3.2-1137), 3.1 (1.7-5.4) and 0.05 (0.003-0.76), respectively, whereas the primary tumor detection rate was 0.44 (0.31-0.58). Although an absolute cutoff is not defined, in good diagnostic tests the DOR should be >100, PLR >10 and NLR <0.1, which was not the case in this study.

Assessment for distant metastases

For M-staging, FDG-PET/CT proved to be a preferred diagnostic modality. Traditionally, a chest CT scan and abdominal ultrasound or CT are used to exclude systemic dissemination. Other tests (endoscopies, bone scintigraphy with technetium) are performed as clinically indicated. However, according to meta-analysis of studies comparing the diagnostic performance of integrated FDG-PET/CT, FDG-PET alone and/or CT alone for the overall assessment of distant metastases, integrated FDG-PET/CT has superior sensitivity (0.95 vs. 0.85 vs. 0.80) and similar specificity (0.96 vs. 0.95 vs. 0.94) to the other two modalities. When used for the screening of distant metastases in head and neck cancer patients, interobserver agreement in FDG-PET was found to be markedly higher as compared to chest CT. The choice between up-front surgery and radiotherapy is usually biased by institutional practice; however, the advantage of the elimination of a gross tumor burden and potential micrometastases, together with the valuable information about the extent and aggressiveness of the neck disease (more accurate N-classification and recognition of extracapsular tumor spread) provided by the histopathological examination, gives preference to primary surgery in the majority of institutions. With an increasing number of CUP patients with p16/HPV-associated neck metastases and their high sensitivity to radiotherapy and chemotherapeutics this may change.

The type of neck surgery is dictated by the extent of the nodal disease. All 5 neck levels are rarely at risk, thus, selective neck dissection is usually indicated. With regard to radiotherapy, two issues must be addressed: the extent of the radiation volume and the advantage of...
In CUP metastatic to the neck nodes, the addition of systemic therapy to radiation follows the premises set for patients with a known primary SCC. In a definitive and postoperative setting, concurrent/adjuvant radiation modalities aimed to improve neck control in patients at higher risk for neck failure. In view of the increased sensitivity of p16 HPV-associated tumors to standard therapies and superior outcome, the role of concurrent chemoradiation in this subgroup is yet to be defined. The results of currently conducted treatment de-escalation trials will be available in the coming years. Another challenge was posed by questioning the prognostic importance of extracapsular tumor spread in p16 HPV-associated oropharyngeal tumors and by the observation that those with low-volume solitary nodal disease (absent vs. present) provides inferior prognostic information to a more detailed description of extracapsular tumor extent.

At present there is no evidence that concurrent administration of radiotherapy and systemic therapy affects the rate of subsequent systemic dissemination which poses up to a 5% risk for distant failure. The use of effective adjuvant systemic therapy in patients at increased risk for distant metastasis, such as those with high-volume or neck metastases or large volume (N2c/N3) disease. However, none of the induction/adjuvant systemic therapy regimens presently in routine use has the ability to improve the survival of patients with a known primary SCC of the head and neck.

**Early-stage disease**

For low-volume solitary nodal disease (N1) with no signs of extracapsular extension, surgery or radiation alone was found to be equally effective. In view of the excellent outcome in non-smokers with p16 HPV-associated N2a tumors of known origin treated with radiation alone, unimodality treatment seems to be indicated also in patients with stage T2N2a p16 HPV-associated disease. A combination of both modalities does not increase neck control or survival in these patients, only morbidity. Especially in patients directed to radiotherapy, detailed pretreatment imaging examination of the neck is crucial to avoid undertreatment. For the same reason, in surgically treated patients a comprehensive clearing of the nodal regions in the neck and not only a simple excision of an enlarged lymph node are mandatory. Because neck staging and assessment of extracapsular extension are both more accurate after surgery than if evaluation is done only by imaging examinations, a selective neck dissection with its low morbidity and costs (compared to radiotherapy) appears to be the treatment of choice.

**Advanced stage disease**

For patients with determined extracapsular tumor spread and for those with N2 or N3 neck disease, a combined modality approach was used. Balzer et al reported in a systematic review of 18 studies published from 1998 to 2010 that analyzed the treatment outcomes in CUP patients with neck metastases. No statistically significant difference in 5-year survival (52.4% vs. 46.6%, p = 0.05) was found between patients treated with surgery and postoperative radiotherapy/chemoradiation and those who underwent radiation/chemoradiation alone. At the moment it is not known which of the two approaches is more effective and whether the integration of FDG PET in the post-treatment evaluation of the irradiated neck (with surrogates of salvage in less than complete responders) has the potential to shift this balance toward a non-surgical option, particularly in the p16/HPV-positive group.

**Conclusions**

CUP with SCC metastases to the neck nodes constitutes up to 3% of head and neck SCCs. Clinical examination supplemented with cross-sectional imaging and the review of patient history is the primary tool in the risk of missing an occult primary. After comprehensive diagnostics the emergence rate of a mucosal primary equals the risk of a metachronous primary in patients cured of a known primary SCC of the head and neck. Under these conditions, the treatment of the involved neck only is justified in the majority of patients, which also offers the advantage for salvage therapy in case of recurrence or new primary tumor in the head and neck area; it is also less toxic than more extensive treatment. Both primary surgery followed by (chemo)radiation or (chemo)radiation alone seem to be equally effective although the former provides valuable prognostic information that allows better adjustment of overall treatment intensity to the aggressiveness of the disease. The impact of the HPV status of metastatic neck-node biopsy before definitive radiotherapy, alone or followed by surgery, on overall survival or local control remains to be determined.

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Abstract
Fungal sinusitis is heterogenous consisting of both invasive and non-invasive subtypes. The clinical course can range from being indolent (e.g., fungal ball) to rapidly progressive (acute invasive fungal sinusitis). Due to overlap of symptoms with bacterial sinusitis, diagnosis can be delayed, sometimes with fatal consequences in immunocompromised patients. This article aims to give the reader an overview of the various fungal sinusitis subtypes, with particular emphasis on the clinical presentation and imaging subtleties, that would expedite and optimize management.

Key words
Fungal sinusitis, fungal rhinosinusitis, invasive fungal sinusitis, non-invasive fungal sinusitis, granulomatous fungal sinusitis, allergic fungal rhinosinusitis, fungal ball

Invasive fungal sinusitis
Acute Invasive Fungal Rhinosinusitis (AIFR) AIFR is rare but can be rapidly progressive (<4 weeks) resulting in fatality within days. It is associated with anoxia-ischemia and extrinsic extension through a combination of bony destruction, perineural and perivascular spread.

Two main patients are typically at risk: diabetics with ketoacidosis and patients who are immunocompromised (severe neutropenia, haematological malignancy, systemic chemotherapy, steroid therapy, bone marrow transplantation, AIDS). Common pathogens include Aspergillus sp (neutropenic patients) and members of the Mucoraceae (Mucor, Rhizopus, and Absidia). Diabetic patients are particularly at risk from Zygomycetes (Rhizopus, Mucor) as these organisms have an active ketoreductase system and thrive in high glucose acidic conditions.

Mortality rates of at least 50% have been reported. As such, a high degree of clinical suspicion and subtle changes in imaging have to be identified to promptly diagnose and manage this condition.

Clinical Features
The clinical hallmark of AIFR is febrile neutropenia and facial pain with or without nasal congestion and orbital signs. Often however, the symptoms are non-specific (fever, headache, facial pain, rhinorrhea, and diplopia) and can range from being indolent (e.g., fungal ball) to rapidly progressive (acute invasive fungal sinusitis). Due to overlap of symptoms with bacterial sinusitis, diagnosis can be delayed, sometimes with fatal consequences in immunocompromised patients. This article aims to give the reader an overview of the various fungal sinusitis subtypes, with particular emphasis on the clinical presentation and imaging subtleties, that would expedite and optimize management.


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Symptoms and signs occur rapidly, often within hours, reflecting the pattern of invasion into nearby structures. Nasal endoscopy often reveals discolouration/pallor of nasal mucosa progressing to ulceration and tissue necrosis. Spread to the orbit and intracranially can result in proptosis, diplopia, visual loss and neurological deficit (headaches, neurological deficit, seizures and coma). Extension of infection to the cavernous sinus via the orbital apex result in ophthalmoplegia (cranial nerve palsies (headaches, neurological deficit, seizures and coma). Thrombosis of the cavernous section of the carotid artery inevitably results in trigeminal nerve involvement). Thrombosis of the orbital apex result in opthalmoplegia (cranial nerve palsies (headaches, neurological deficit, seizures and coma)). Spread to the orbit and intracranially can result in subarachnoid haemorrhage secondary to thrombosis of the carotid artery /branches or myotic aneurysm from angioinvasion9.

Management

The mainstay of treatment is early aggressive surgical debridement, broad spectrum systemic anti-fungal treatment and reversing the underlying immunosuppression (eg diabetic ketoacidosis or neutropenia. Strict control of diabetes and reversal of neutropenia with granulocyte-colony stimulating factor10 can lead to improved survival). Surgical (endoscopic or open) debridement is carried out until clear, bleeding margins are observed. There is little evidence that radical resection, including orbital exenteration and radical maxillectomy, improves survival11. Indeed, endoscopic resection has been shown to lead to improved survival compared to those who undergoing open surgery12. This may partly be due to the fact that patients undergoing open surgery had far more advanced disease. A multidisciplinary approach involving ophthalmology, maxillofacial and neurosurgical expertise is paramount.

Broad-spectrum anti-fungal therapy eg amphotericin B deoxycholate should be commenced as soon as a diagnosis of AIFS is suspected. Due to its nephrotoxic profile, safer lipid-formulations such as Amphotericin B lipid complex and liposomal amphotericin B have been developed. Fungal cultures are essential in determining sensitivity to anti-fungal agents. Once mucormycosis is ruled out, treatment may be changed to a less toxic azole which is more effective against Aspergillus compared to mucormycosis. Voriconazole is now recommended as first line treatment for invasive aspergillosis of the sinuses by the Infectious Disease Society of America. An alternative azole, posaconazole may be used as a step-down to oral treatment when clinical improvement is seen, to enable long term treatment.

Hyperbaric oxygen therapy (HBOT) has been proposed an adjunct to treatment of AIFS13. HBOT acts directly by increased production of oxygen-based free radicals and indirectly by reversing growth-promoting lactic acidosis and restoration of phagocytes14. To date there is no clear evidence of the efficacy of HBOT in the treatment of AIFS.

Prognosis

In a recent systematic review by Turner et al15, diabetic patients were found to have better prognosis, despite often more aggressive disease, possibly due to the fact that their underlying condition was more easily reversed than other conditions. Patients who have intracranial involvement, or who do not receive surgery as part of their therapy, have a poorer prognosis.

Chronic Invasive Fungal Rhinosinusitis

Chronic invasive fungal rhinosinusitis (CIFRS) is rare. It is characterized by a slowly progressive invasive disease (>3 months). It tends to occur in immunocompetent individuals. It is differentiated from its chronic granulomatous invasive fungal sinusitis (CGIFS) counterpart by a lack of granulomas on histopathology and an association with diabetes mellitus.

Aspergillus spp. (fumigatus) or Mucor spp are the most common causative organisms16. The symptoms often mimic chronic sinusitis which is refractory to standard antibiotic treatment. As the disease advances, proptosis, orbital apex syndrome and neurological deficits may occur.

Diagnosis

The clinical work-up (nasendoscopy, imaging and biopsy) is identical to AIFS. There can be nasal congestion and polyposis and evidence of fungal invasion on histology. Imaging features relatively similar to the acute counterpart except for the pattern of intranasal calcification on CT. In the AIFS, intranasal calcifications show a fine punctate appearance whilst in CIFS, as more calcium metabolites are deposited in the fungal mass, a more dense and coarse appearance is witnessed17. Localised erosion of the sinus walls are seen with extension into adjacent tissues, orbit and intracranial compartments.

The treatment of CIFRS is similar to AIFS i.e a combination of surgery followed by antifungals. Systemic and topical antifungals18 should be started until cultures exclude Mucor species. Azole drugs such as voriconazole19 and itraconazole are promising alternatives as they are effective via the oral route and are therefore easier and cheaper to administer for longer term treatment but are less effective against Mucor sp. A prospective randomised unblinded study compared Amphotericin B (conventional or liposomal) and itraconazole in the management of CIFRS and found that both were equally efficacious20.

Chronic Granulomatous Invasive Fungal Sinusitis

Chronic granulomatous invasive fungal sinusitis is rarely seen in the West and is more common in the North Africa, the Middle East and Asia. Aspergillus flavus is the most common causative fungus. It follows an indolent path and may be found in both immunocompetent and immunodeficient patients. Imaging features are nonspecific and similar to other forms of invasive fungal sinusitis. It is defined by the presence of non-caseating granulomas with Langhans type giant cells and fungal hyphae, although it may co-exist with other types of fungal sinusitis21.

It is treated by surgery followed by antifungals. Antifungals like voriconazole instead of amphotericin may be used as the disease is caused by Aspergillus flavus. Indeed, Rupa et al22 recommends postoperative treatment with either oral itraconazole or voriconazole for disease with limited extension and oral voriconazole for advanced disease extending to the brain. Amphotericin B was not recommended as first line therapy for CGIFS.

Non-invasive fungal sinusitis

Allergic Fungal Rhinosinusitis

Allergic fungal rhinosinusitis (AFRS) is the most common form of fungal sinusitis. This condition shares similar histopathologic features with allergic bronchopulmonary aspergillosis (ABPA) and was first reported by Safirstein1 in 1976.

AFRS consists of a non-invasive collection of eosinophilic mucus resulting from a type I hypersensitive delayed allergic response to inhaled mould spores which colonize the sinus cavity. The fungi most commonly identified in the eosinophilic mucin include Alternaria, Bipolaris, Cladosporium, Curvularia, Drechslera and Helminthosporium from the dematiaceous family and Aspergillus species23. The importance of type I hypersensitivities in AFRS remains controversial as there are alternative hypotheses including antibody-independent pathways and local sinonasal IgE production24. The typical patient with AFS is younger and atopic and immunocompetent. It also more frequent in warm, humid environments and lower socioeconomic status25.

Clinical Features

Symptoms mimic chronic rhinosinusitis which is refractory to medical treatment. They present often with unilateral, but sometimes bilateral nasal congestion, post nasal drip,
Many experts use Bent and Kuhn criteria for diagnosis of AFRS. Patients may also present with signs and symptoms of bony erosion and involvement of the orbit, including proptosis and telecanthus.

Diagnosis

Many experts use Bent and Kuhn criteria for diagnosis of AFRS (table 1). These criteria consist of both major and minor criteria, and all five major criteria must be met in order for a diagnosis to be made. The presence of minor criteria supports the diagnosis of AFR. The amount of fungal hyphae can be variable and sparse, therefore even in its absence, the presence of allergic mucin is virtually pathognomonic for AFRS.

<table>
<thead>
<tr>
<th>Table 1. Bent and Kuhn Diagnostic Criteria</th>
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<tr>
<td><strong>Major</strong></td>
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<td>- Type I hypersensitivity (history, skin test or in vitro testing)</td>
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<tr>
<td>- Nasal polyposis</td>
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<tr>
<td>- Characteristic CT findings</td>
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<tr>
<td>- Eosinophilic mucin without invasion</td>
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<td>- Positive fungal stain</td>
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<tr>
<td><strong>Minor</strong></td>
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<tr>
<td>- Asthma</td>
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<tr>
<td>- Unilateral disease</td>
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<tr>
<td>- Radiological bone erosion</td>
</tr>
<tr>
<td>- Fungal cultures</td>
</tr>
<tr>
<td>- Serum eosinophilia</td>
</tr>
<tr>
<td>- Charcot-Leyden crystals</td>
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<td>- (eosinophil degradation products)</td>
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CT imaging typically shows characteristic pansinusitis, hyperattenuation of the intrasinus contents (representing thick allergic mucin) surrounded by lower attenuation hyperplastic mucosa. The double density sign may be seen in affected sinuses i.e heterogeneous signal intensity due to the deposition of heavy metals within the mucin (Figure 2). Sinus walls are often expanded, remodelled or eroded due to the expansive nature of mucin and its propensity to incite a local inflammatory response (Figure 3). The MR imaging signal is variable, ranging from isointense to hypointense signal to signal void on T1- and T2-weighted images, depending on the contents of the sinus. Therefore, MRI should always be interpreted in the presence of CT; on T2-weighted images, the high metal concentration within proteinaceous allergic fungal mucin may show up as a signal void (Figure 4), mimicking the appearance of a normally aerated sinus. Both T1 and T2 weighted images show peripheral enhancement.

Management

The mainstay of AFRS treatment remains surgery though adjunctive medical management is critical for optimal outcomes.

In 2014 Gan et al published an evidence-based approach for the postoperative medical management of AFRS. The review concluded that postoperative systemic and standard topical nasal steroids are recommended; oral antifungals, and immunotherapy are options in cases of refractory AFRS; and did not provide recommendations for topical antifungals and leukotriene modulators due to lack of evidence. Recently, AFRS which is refractory to surgery and conventional medical treatment has been shown to be responsive to anti-IgE antibody, omalizumab.

Fungal Ball

A fungal ball consists of sequestered fungal hyphal elements within a sinus without allergic mucin, invasive or granulomatous changes. It is distinct from saprophytic fungal infection which corresponds to fungal spores found on crusts and mucus in the nose. Fungal balls are typically found in immunocompetent individuals. Aspergillus fumigatus is the most commonly implicated pathogen. Previous dental treatment and radiotherapy are sometimes implicated in the development of fungal ball.

Clinical Features

Fungal balls commonly affect the the maxillary sinus followed by the sphenoid sinus. Patients may be completely asymptomatic with the diagnosis made incidentally on imaging. Other symptoms include headache or facial pain, post-nasal drip and cacosmia. Fungal balls affecting the sphenoid sinus may present with retro-orbital pain at the vertex.

Diagnosis

CT scanning is the imaging of choice for suspected fungal balls. Five CT features are commonly found, including a heterogeneous soft tissue density in a single unilateral sinus, absence of an air-fluid level, erosion of the inner wall of the sinus, sclerosis of the lateral sinus wall and the presence of calcification. In particular, the presence of erosion of the inner wall of the sinus and the presence of calcification have a positive predictive value in the diagnosis of fungal balls of 94.6% and 93.2% respectively.
The presence of calcification is thought to be due to the deposition of calcium salts within the fungal ball. MRI scanning is not usually necessary, but may be used as an adjunct to CT imaging to differentiate mucosal swelling or mucus retention from a fungal ball. On MRI scanning, fungal balls are often hypointense compared to the underlying mucosa. The treatment of fungal balls is surgical with no adjunctive therapies usually necessary. Functional endoscopic sinus surgery is employed to create a wider opening of the affected sinus ostium. All fungal material should be removed and the sinus thoroughly irrigated with saline. Biopsies are taken from the underlying mucosa to rule out invasion. Any dental filling material present in the sinus should also be removed. In immunocompetent patients there is no role for the use of systemic antifungal agents.

Management
The treatment of fungal balls is surgical with no adjunctive medical treatment usually necessary. Functional endoscopic sinus surgery is employed to create a wider opening of the affected sinus ostium. All fungal material should be removed and the sinus thoroughly irrigated with saline. Biopsies are taken from the underlying mucosa to rule out invasion. Any dental filling material present in the sinus should also be removed. In immunocompetent patients there is no role for the use of systemic antifungal agents.

Conclusion
Fungal sinusitis symptoms can overlap with bacterial or viral rhinosinusitis. An important appreciation of the clinical presentation and radiological findings is vital for timely diagnosis, improving patient outcomes and limiting morbidity. A multidisciplinary team approach engaging various surgical and medical disciplines should be encouraged in the management of more complex cases of fungal sinusitis.

References

Odontogenic causes of maxillary sinusitis and their management

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Abstract

Odontogenic maxillary sinusitis is an uncommon but important disease entity and is often recognised late. The increasing use of computed tomography (CT) in the assessment of unilateral sinus symptoms has improved the ability to recognise and manage this group of conditions. Here we discuss the details of a number of dental causes of maxillary sinus disease. A description of the journey of a tooth is used to demonstrate how certain pathologies occur and how they may lead to disease within this specific region. Management options are also discussed which often require close collaboration with dental and oral/maxillofacial colleagues.


Key words

Maxillary sinus, sinusitis, odontogenic

Introduction

Odontogenic sinusitis accounts for up to 10%-12% of all cases of maxillary sinusitis. A simple look at the relevant anatomy explains the relationship, with the floor of the maxillary sinus formed by the alveolar process of the maxilla (Figure 1). The formation and continuing integrity of this alveolar bone is intimately linked to the development and maintenance of normal, healthy dentition.

A wide range of dental problems, both primary and iatrogenic, can compromise the normal functioning of the maxillary sinus. A disruption in the normal mucusociliary pathway created by the ciliated pseudostratified columnar epithelium within the maxillary sinus can lead to mucus stasis, overgrowth of organisms and sinus mucosal inflammation. There is a two-fold increase in maxillary sinus disease in patients with periodontal disease and this relates to the close proximity of teeth to the maxillary sinus. All maxillary teeth have the potential for causing problems in the sinusosal cavities but the first and second permanent molars are those most commonly involved in maxillary sinusitis due to their root morphology and positioning. Less commonly, the maxillary second premolars and third molars can be involved. A dental cause for maxillary sinus disease should be considered in those patients with symptoms of unilateral maxillary sinusitis with a history of odontogenic infection, dento-alveolar surgery or in those patients resistant to conventional sinusitis therapy. The most common cause of odontogenic sinusitis is iatrogenic and this accounts for over half of cases reported in the literature.

The tooth journey

Knowing the basics of odontogenesis and the journey of a tooth from embryonic cells to an erupted, functional structure will give a greater understanding of the origin and behaviour of odontogenic pathology relevant to the maxillary sinus.

The tooth germ is essentially a collection of cells derived from the first pharyngeal arch and neural crest. It is connected to the oral cavity via an in-growth of oral ectoderm – the dental lamina and is organised into three main parts:

- the enamel organ - gives rise to ameloblasts which produce enamel and also the Hertwig Epithelial Root Sheath which determines the shape of the tooth roots
- the dental papilla - produces odontoblasts which form dentine
- the dental follicle - produces cementoblasts, osteoblasts and fibroblasts which give rise to cementum, the periodontal ligament and alveolar bone which are the supporting structures of a tooth

Tooth development progresses through various cell production and organisation stages which include the bud, cap and bell stages, then the formation of the dental hard tissues and the formation of the tooth supporting structures. Each of the cell layers and stages of odontogenesis can give insight into the origin of future odontogenic pathology relevant to maxillary sinusitis. In addition to normal odontogenesis, teeth must follow particular eruption pathways to become functioning, healthy oral structures. Impacted teeth are common and can be associated with various pathological processes affecting underlying bone and adjacent structures.

Even when teeth develop normally and erupt into anatomically correct positions, they enter a relatively hostile environment. A host of microorganisms exist in the oral cavity, deleterious to both the calcified tissues of the teeth themselves and the tooth supporting tissues. In addition, a variety of non-microbial threats such as mechanical abrasion/attrition/trauama and chemical agents also exist. The resultant pathology itself can impact upon the maxillary sinus, as can the treatment modalities used by dentists/surgeons to eliminate the pathology.

We discuss the common odontogenic causes of sinusitis below and their relationship to this journey. These can be categorised into three main categories; benign/malignant pathology, infective/inflammatory causes and iatrogenic causes.

A. Benign and malignant pathology

Ectopic teeth

Impacted teeth are relatively common occurrences, especially involving third molars, maxillary canines and maxillary second premolars. Normal eruption is interrupted or impeded resulting in abnormal tooth positioning. This is especially true for late erupting teeth or those with long eruption pathways, such as the maxillary canine. Impacted teeth can remain dormant and cause no problems but any associated cystic change or surgical treatment to remove them can result in maxillary sinus disease.

Ectopic teeth/supernumeraries/odontomes are only seen rarely in non-dentate areas such as the maxillary sinus. They are commonly only identified during imaging for investigation into the cause for any consequential maxillary sinusitis. When secondary pathology exists, early surgical intervention is recommended for removal of the tooth and any associated cyst which may require open or combined endonasal and oral approaches.

Odontogenic cysts

The lining of these cysts are all derived from the remnants of the tooth-forming organ and can be subdivided into developmental and inflammatory types. These lesions can cause significant bony destruction and the tissues surrounding them can become inflamed/infected and lead to secondary maxillary sinusitis.

Radicular cyst

- The most common odontogenic cyst, accounting for more than 65% of all such lesions
Dentigerous cyst

- Originates in the follicular tissues overlying the crown of unerupted teeth
- Lining is supported by a fibrovascular capsule free from inflammatory cell infiltration
- True dentigerous cysts are attached to the amelocemental sheath and is supported by a chronically inflamed fibrous capsule.
- Enlarge slowly and generally do not grow to very large dimensions (Figures 2 and 3).
- Treatment involves either endodontic therapy or removal of the non-vital tooth and enucleation of the cyst lining.

Radiologically can have a multi-loculated appearance depending on a number of complex factors, this can range from small OACs which may close spontaneously but any ascending sinus inflammation. Direct communication between the odontogenic (or otherwise) inflammatory process and the sinus lining influences the mucociliary pathways within the maxillary antrum, leading to mucus stasis, bacterial colonisation and a proliferating inflammatory process. Most infective cases involve a polymicrobial aerobic-anaerobic infection with gram negative anaerobes particularly prominent. Other cases involve anaerobic streptococci, gram-negative bacilli, and Enterobacteriaceae.10

Periapical pathology

In periodontal disease the initial site of bacterial colonisation is the gingival sulcus (the space between the tooth and gingiva) rather than within the root canal system. The inflammatory process begins as gingivitis which is not associated with alveolar bone loss but depending on a number of complex factors, this can progress to periodontal disease which causes bone resorption and apical migration of the disease process.

The thickness of the maxillary floor may influence the chance of inflammatory tooth disease leading to

Malignant odontogenic tumours, according to the WHO classification, consist of:

- Malignant Ameloblastoma
- Primary Intraosseous Carcinoma
- Other carcinoma arising from odontogenic epithelium, including from odontogenic cysts

These are rare lesions but again have potential to impact on the maxillary sinus.

B. Infective and inflammatory causes

Bacterial infection

Oral infective/inflammatory causes of maxillary sinusitis include dental periapical pathology, advanced periodontal disease, secondary infection of (non-inflammatory) odontogenic cysts and rarely Actinomycosis, a subacute/chronic bacterial infection usually caused by Actinomyces israelii.

Granulations and an oro-antral communication can be seen into the antrum during oral surgical procedures. In some instances this may result in no associated sinus pathology
but the majority require surgical removal. Simple radiographs can be helpful in locating these displaced fragments but often CT or cone beam CT imaging is required. Surgical approaches to retrieve displaced roots can be trans-alveolar or via a Caldwell-Luc approach intra-orally or via an endonasal approach which requires a wide antrostomy to gain access (Figures 8 and 9).

**Implants**

Dental implants require osseo-integration within sufficient healthy bone to be successful. Insufficient planning and poor surgical technique can lead to protrusion of a dental implant into the maxillary sinus and this in turn can act as a nidus of infection. The placement of dental implants is increasingly being facilitated by sinus-lift procedures. A variety of foreign bodies/materials can enter the sinus itself then a wide antrostomy and retrieval may be possible but sometimes these require an oral, combined and rarely an open approach.

**Conclusions**

The majority of causes for odontogenic maxillary sinusitis are secondary to dentally related procedures. Occasionally a primary lesion of the teeth may lead to a source of maxillary sinusitis. Radiological investigations are increasingly identifying the causes of this disease entity and aid with the planning for surgical management, particularly for the approach required. A broad awareness of local anatomy and pathology with a multidisciplinary team approach to treatment is the best way to successfully manage this potentially refractory type of sinusitis.

**References**


**Figure 5:** Buccal mucosal advancement flap with buccal fat pad to obliterate the oro-antral communication

**Figure 6:** Large oro-antral fistula

**Figure 7:** Dental panoramic radiograph of an oro-antral fistula

**Figure 8:** Left periapical cyst of left upper 2nd molar pre-operatively

**Figure 9:** Post-operative findings after Caldwell Luc approach for cyst removal and tooth extraction. Maxillary flap repair of defect. Residual sinus disease present
Body dysmorphic disorder and aesthetic treatment

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Body Dysmorphic Disorder, BDD, is a distressing and potentially disabling condition characterised by a preoccupation with imagined or slight physical defects in appearance, this results in time consuming rituals causing significant psychological distress or impairment in personal, social and occupational functioning.

Common areas of concern are skin, face, hair, body build and breasts. Patients most common negative beliefs are that their skin is discoloured or a body part is deformed or flawed or that the size and shape of their body parts are not correct or there is an asymmetry present, for example in the eyes or nose. As a result, to improve or disguise these perceived flaws most patients engage in ritualised behaviours such as staring in the mirror, skin picking, hair pulling, excessive make up, manicure or elaborate rituals such as grooming routines, feeling the body part to test for smoothness, size and flaws, constant mirror checking, seeking reassurance from friends and family and shopping extensively for doctors for aesthetic or surgical treatments. However, avoiding mirrors and bright lights and public places may also be a feature of Body Dysmorphic Disorder.

The main questions seem to be where does this normal concern with looking as good as possible and becoming preoccupied with looks change into becoming pathological? What are the implications for aesthetic practitioners and surgeons? Should such patients be treated by performing aesthetic and surgical procedures to rectify perceived cosmetic imperfections? This is a relevant question not only for those routinely performing cosmetic procedures, such as rhinoplasty, but for all doctors since these patients will present to varying degrees in many clinical settings.

Human beings have always been concerned with their appearance, and being content helps develop self-esteem, confidence and a place in family and society. Not having this emotional peace with one’s own body and appearance may lead to significant psychological and social problems. Historically, Thersites was said to be the ugliest Greek in the Trojan War and from an ancient Greek point of view an ugly soul must inhabit an ugly body. Dysmorphia is a term was derived from the Greek word ‘dysmoria,’ meaning misshapen or ugly. Excessive concerns with physical deformity has been known in the past as ‘Quasimodo Complex’.

Sigmund Freud’s Wolfman became famous and he was later described by Brunowick1: “He neglected his daily life and work because he was engrossed, to the exclusion of all else, in the state of his nose. On the street he looked at himself in every shop window; he carried a pocket mirror, which he took out every few minutes. First he would powder his nose; a moment later he would inspect it and remove the powder. He would then examine the pores, to see if they were enlarging, to catch the hole, as it were, in its moment of growth and development. Then he would again powder his nose, put away the mirror, and a moment later begin the process anew. His life was centered on the little mirror in his pocket, and his fate depended on what it revealed or was about to reveal.”

The prevalence of BDD varies, for example, a German population study gave a prevalence of 1.7%. However, the prevalence of Body Dysmorphic Disorder in cosmetic surgery settings and dermatology clinics suggest that the disorder is much more common in these populations, with a prevalence of between 3 and 10%, it affects men and women equally, however women seek treatment for BDD more frequently than men, cultural factors may also play a part. In one study involving aesthetic rhinoplasty candidates 24.5% fulfilled the DSM IV criteria for BDD4.

Unfortunately, Body Dysmorphic Disorder is poorly diagnosed in psychiatric settings, so it follows that it is less frequently diagnosed in aesthetic clinics and surgical settings. Although the diagnosis is often missed, it is easy to make.

The picture may be complicated, however, by co-existing conditions, for example depression is present in 80 – 90% of patients with BDD and over one third suffer from social phobia amongst other psychiatric diagnoses. However, it seems that the social phobia onset was typically before that of Body Dysmorphic Disorder and not caused by concerns about appearance1. Although BDD is considered to be in the spectrum of Obsessive Compulsive Disorders, it is interesting to note that patients with BDD do not get relief from their anxiety when they perform checking rituals, such as mirror checking, in fact these may increase the sense of despair, unlike in Obsessive Compulsive Disorders. Despair may be accompanied by feelings of self-loathing, guilt, shame, embarrassment and fear of being judged.

Frequency of perceived defects:

- Skin (73%)
- Hair (56%)
- Nose (17%)
- Weight (22%)
- Adipose tissue (22%)
- Breasts/pectorals/nipples (21%)
- Ears (20%)
- Eyelids (20%)
- Teeth (20%)
- Face size/shape (12%)
- Lips (12%)
- Buttocks (12%)
- Chin (11%)
- Eyebrows (11%)
- Hips (11%)
- Ears (9%)
- Arms/wrists (9%)

- Waist (9%)
- Genitals (8%)
- Cheeks/cheekbones (8%)
- Calves (8%)
- Height (7%)
- Head size(6%)

Most doctors have experienced patients expressing concerns about their appearance at some stage. Hence such concerns are very common and cultural factors may play a significant part, as well as sex, demographics and social subcultures. However, as in much of psychiatry, where there is significant impact on personal, social and occupational functioning or the degree of preoccupation, often proposed as one hour a day, despite the perceived defects or flaws being very slight or not observable to others then the concern and preoccupation reaches significant proportions in terms of psychological morbidity.

The relevance of Body Dysmorphic Disorder in aesthetic or surgical settings is important. Perhaps the most important thing to bear in mind is that most patients will not reveal their symptoms. Very few reported these symptoms voluntarily to their psychiatrist although these were a significant factor towards their suicidal, perhaps due to embarrassment or fear of being judged. Many may even believe that they are fundamentally unacceptable and therefore unlovable. This often leads to high levels of social isolation and poor social support for sufferers of BDD and this must be a contributing factor to the fact that up to 80% of patients with BDD have experienced suicidal thoughts.

If the diagnosis is missed the consequences can be significant. It is unlikely that a person suffering from Body Dysmorphic Disorder will be satisfied with any aesthetic intervention or any number of cosmetic surgical procedures. Insight can vary in patients with BDD, which is fewer in number to poor or absent insight to delusional, which are the majority, about three quarters of all patients. There is, of course, a continuum of preoccupations and there are many patients who are concerned about their appearance and do not suffer from BDD because they are not preoccupied or distressed or dysfunctional; when these patients seek aesthetic treatment and cosmetic surgical intervention to improve their looks, confidence and self-esteem, and these interventions appropriately performed can change someone’s quality of life.
In other patients with BDD, however, this can lead to multiple surgery and even attempts at self-body modifications, some attempts may include self-mutilation. All of these attempts usually mean that the patient’s flaws and defects in their perception are not improved and very rarely is quality of life improved.

The nature of the illness in BDD may fluctuate and there may be periods of fairly normal functioning. Many people with BDD have particular difficulties with photographs and avoid family events and photographs, which only seem to confirm their beliefs. It is commonly believed that media influences affect illnesses such as eating disorders it may also be the case that by emphasizing the necessity of aesthetic beauty, these influences may also contribute to Body Dysmorphic Disorder in a similar way.

It is possible to screen with screening tests, however sometimes a few simple questions may be sufficient to alert the physician that further inquiry, screening or referral may be appropriate. Many patients (and some doctors) do not realise that BDD is treatable and in fact researchers have shown that patients want their clinician to ask them about BDD symptoms, so if done in a sensitive manner, this should not be any more difficult than other routine questions a clinician asks in practice.

For example: Some people worry a lot about their appearance. Do you worry a lot about the way you look and wish that you could think about it less? If this receives a positive answer, then follow up questions to clarify the extent and nature of bodily concerns can be asked. ‘Are you worried about the way you look?’ For example, questions may be asked about skin, acne, scars, hair, shape and size of your nose, jaw, lips, stomach, hips, or any other body part. ‘How has this problem affected your life?’ On an average day, how much time do you usually spend thinking about how you look? (Add up all the time in total a day).

The COPS Questionnaire contains 9 pertinent questions to which graded responses are possible.

1. How often do you deliberately check your features?
2. Do you feel that your features are particularly ugly?
3. To what extent do you feel your features will distress you?
4. How often do your features currently lead you to avoid situations or activities?
5. To what extent do your features currently occupy your mind? Is there anything you do about it and is it hard to stop thinking about it?
6. If you have a partner, to what extent do your features currently have an effect on your partner, for example, affect their eating, enjoyment of food?
7. To what extent do your features currently interfere with your ability to work or study, or your role as a homemaker?
8. To what extent do you worry about your features?
9. To what extent do you worry about your social life? (with other people, e.g. parties, pubs, clubs, outings, visits, home entertainment).
10. To what extent, do you feel that your appearance is the most important aspect of who you are?

Although therapeutic interventions in terms of medications and psychological treatments are possible, BDD is often a chronic illness and even with specialist intervention and treatment many relapses may occur and the prognosis for complete recovery is poor.

In terms of treatment of BDD, Cognitive Behaviour Therapy, CBT, is considered to be the treatment of choice, and pharmacological therapy has been based around antidepressants mainly Selective Serotonin Reuptake Inhibitors, SSRI, which are needed in higher doses and for longer durations than usual. However, on discontinuation relapse rates are high.

The UK NICE guidelines for OCD and Body Dysmorphic Disorder (National Collaborating Centre for Mental Health 2005) make use of a graduated approach to treatment, for mild cases self-help books with guidance are recommended. Some evidence from a meta-analysis exists for both psychological and SSRI treatment. Moderate cases therefore should be offered CBT or a SSRI and a combination of both should be offered to severe cases.

In terms of patients with BDD seeking treatment from doctors, the most commonly visited doctors were dermatologists, followed by patients seeking surgical rhinoplasty, and treatment for acne and surgical intervention did not improve the BDD. Surgical or cosmetic intervention can result in the patient transferring concern to another flaw or further increasing focus on the surgically altered one, which may never be seen as ‘beautiful’, with requests for further surgery that can become a vicious cycle, causing significant problems for the patient and doctor.

The relevance of BDD in aesthetic treatments and cosmetic surgery is clear. These patients present commonly and are on a spectrum, men are just as likely as women to have sought cosmetic surgery and suicidality may increase when cosmetic surgery is denied and 25–30% of patients with Body Dysmorphic Disorder in a psychiatric clinic have had a history of attempted suicide. Many patients with poor insight seek cosmetic treatments as an alternative to psychological or psychiatric treatment. Insight is not a significant factor in terms of aesthetic treatment; the outcome is likely to be poor even in those patients with relatively good insight since they may continue to see their defect or flaw as still ugly after treatment or surgery.

It is possible, with judicious selection, to greatly improve the quality of life of patients requesting aesthetic or surgical interventions since, of course, most people who attend aesthetic clinics or request surgical intervention are not suffering from BDD. However, it is important to realise that BDD is common in such settings, has significant implications in terms of dissatisfaction and it is not in the patients’ best interest that aesthetic interventions are offered to them.

In conclusion, in view of the prevalence of BDD, a case can be made for a screening questionnaire to be carried out on all patients requesting aesthetic interventions. It is interesting to note that BDD occurs on a spectrum and the milder end of the spectrum may be more difficult to detect in aesthetic or surgical clinics. However, by asking a few simple screening questions, which should be part of routine consultation prior to agreeing to undertake any aesthetic procedure it is relatively easy to screen for BDD. If positive answers are forthcoming from the patient, then advice can be given in a sensitive and empathetic manner and referral should be offered to appropriate psychiatric services.

This simple and easy process, which takes a few minutes during initial consultation may prevent significant heartache in the future and would probably be in the best interest of the doctor and certainly be in the best interest of the patient who may be suffering from Body Dysmorphic Disorder.

References
Haemorrhagic hereditary telangiectasia in ENT

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Abstract

Hereditary haemorrhagic telangiectasia (HHT) or Osler-Weber-Rendu disease is a rare autosomal dominant condition characterised by arterio-venous malformations (AVM). These can affect the mucous membranes, but also the lungs, brain and liver. Epistaxis is a common symptom, so ENT often sees these patients and there are recommendations as to which of these AVMs to screen for. The management consists of trying to reduce the frequency, intensity and/or duration of epistaxis so as to improve the patient’s quality of life and their need for transfusions, as no cure exists. Systemic medications such as bevacizumab, tranexamic acid and tamoxifen have a role. Surgery in the form of coagulating vessels and reducing the number of telangiectasia also helps, but these will usually need to be repeated. Young’s procedure still remains the definitive operation for problematic symptomatic patients.

Introduction

Hereditary haemorrhagic telangiectasia (HHT) or Osler-Weber-Rendu disease is an autosomal dominant condition with high penetrance and variable phenotype. A recent population based UK study demonstrated prevalence as 1 in 9400 individuals, with women more predominantly affected1. It is characterised by arteriovenous malformations (AVM) – both at the capillary level forming telangiectasias and as vascular elements. Additional prolonged bleeding due to lack of the contractile elements.

Diagnosis of patients with HHT is made using the Curacao criteria with the core symptoms and signs being epistaxis, telangiectasias at characteristic sites, diagnosis of visceral AVMs and 1st degree relative with the condition (see Figure 1). The involvement of the ENT clinician in these patients is related to their epistaxis, which in some patients can be difficult to manage. A recent study demonstrated ENT involvement in 79% of patients with HHT but instigation of screening from ENT departments is very low6. Awareness of the systemic manifestations of the disease by the clinician is paramount, as targeted screening is necessary to identify systemic AVMs early to avoid significant morbidity or even mortality.

This review will explore the current evidence for the management of epistaxis in these patients in addition to the screening issues that face them with respect to the systemic aspect of the condition.

Diagnosis and screening

In 2009, the International HHT Foundation published guidelines on the diagnosis and management of the condition on best available evidence, in addition to consensus from international HHT experts4. The subsequent publication of new data in the field regarding management of systemic AVMs has resulted in re-evaluation of these guidelines, especially within the context of a UK healthcare system. Taking each AVM in turn we have evaluated the literature to provide current appropriate guideline for patients with HHT:

Pulmonary AVM

The need to identify a pulmonary AVM early is paramount because if untreated, these can result in cerebrovascular accidents, cerebral abscesses and even worse, death5. The International Guidelines had already determined that outcomes following embolization of pulmonary AVMs were highly successful with a good safety profile7. Since the publication further evidence of the use of the AMPLATZER plug into larger malformation has added further weight to the argument for embolisation in these patients and thus screening in all HHT patients for the detection of such an AVM8. In terms of the screening tool of choice, the guidelines recommend a transthoracic bubble echo, which has limited availability in the UK due to being operator dependent. Given that high resolution CT is much more accessible and correctly identifies pulmonary AVMs when they are at treatable size9, this would be the recommended screening investigation of choice if transthoracic bubble echo is unavailable.

Cerebral AVM

The guidelines published in 2009 stated that cerebral AVMs should be screened for using MRI. The recommendation was on the basis that approximately 23% of HHT patients harboured an AVM and awareness of one would enable treatment to prevent a potential cerebral bleed10. HHT patients under 45 years old have a 1.4-3% per annum per patient lifetime risk of a haemorrhagic stroke11, but further data may suggest that this risk is lower for HHT patients than those without HHT12,13. More recent data has suggested that the prevalence of a cerebral AVM in the HHT population is lower than originally demonstrated, 7.7% for AVM and cerebral aneurysms in 2.4%14. They were largely supratentorial (85%) and all were less than 3cm. The Brain Vascular Malformation Consortium HHT Investigator Group has also demonstrated no statistically significant differences in the types of cerebral AVMs and the genotype of the patient15. They identified 3 major types; capillary vascular malformations (commonest – 61%), arterio-venous malformations and arterio-venous fistulae. Multiplicity of lesions was common, as was the largely superficial nature of the lesions. Previous studies have also shown that haemorrhages from the commonest type (capillary) have not been observed16. Despite this, a large multicentre blinded RCT trial examined treatment options for cerebral AVMs – medical versus surgical17. The trial was stopped early due to inordinately high morbidity and mortality in the surgical arm of the trial. This therefore poses the question of should patients be screened for something when there is debate over the treatment, or lack of, for it? Granted, the likelihood of a detected AVM is small and supratentorial and therefore more amenable and accessible to treatments with higher success rates. But the converse argument of the low risk of these small AVMs bleeding in the first instance vs the risk of surgical intervention is also to be considered. This is a dilemma and one which should be discussed open and honestly with patients. At the Hammersmith in London, one of two National HHT centres, it is now their practice not to routinely screen patients for cerebral AVM.

Gastrointestinal telangiectasia

Patients with symptomatic gastrointestinal AVMs present with melaena and anaemia, which may be out of context from their usual epistaxis. The standard investigation for these patients is with upper endoscopy however, emerging evidence suggests that the endoscope, in HHT patients can actually trigger significant bleeding18. Capsule endoscopy appears to be a safer alternative, resulting in fewer bleeds, if the option is available19. Recent data has offered argon plasma coagulation (APC) as an effective treatment for treating the gastrointestinal vascular malformations in addition to emerging case reports on the efficacy of Bevacizumab in severe gastrointestinal bleeding20. In patients who are asymptomatic with respect to gastro-intestinal AVMs, the International Guidelines screening advice of yearly haemoglobin checks in patients above 35...
A VMs are limited – the morbidity and mortality of gene9. Although there are recognised genes that cause the families should undergo genetic screening for the causative possible or definite HHT on Curaçao criteria and their This is an area that is somewhat more difficult to navigate. Genetic screening

management of patients in the acute setting at times and more This will in part be down to education of patients on how frequency and/or severity of their episodes of epistaxis.

aim to improve the patient’s quality of life by reducing the As there is no cure at present for this condition, treatments

telangiectasia), tranexamic acid (stabilises blood clot) or squamous metaplasia of mucosa, hence protecting

found to improve frequency of epistaxis when given other problems45. Other options that have been tried are silastic splints insertion and closing the nostril by taping46. However, these need more evidence to support their use.

Reducing bleeding in general Embolisation and arterial ligation can be loosely classified in this section as they aim to reduce the blood supply to the offending telangiectasia. These techniques are really reserved for the acute setting. There are a few articles about radiological embolization, and the consensus is that it rarely completely stops the bleeding, but does reduce the symptoms for a while. There is nothing in the literature about elective vessel ligation alone for this condition.

Conclusion HHT is rare but problematic condition and patients can be difficult to manage. Screening for related AVMs and genetic testing of family members should be considered and discussed with patients, as there are implications of the potential findings. Simple emollients work well for patients, but can be considered the next step in the management along with considering systemic medications. Year’s procedure still appears to be the most definitive treatment but less restrictive alternative

References
Endoscopic vidian neurectomy for intractable vasomotor rhinitis

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Abstract

Vasomotor rhinitis (VMR) is the most common form of non-allergic rhinitis. Vidian neurectomy was described five decades ago for intractable vasomotor rhinitis, however, in the pre-endoscopic era it was not popular due to limited access and significant complications. The advent of endoscopic sinus surgery has allowed surgical approaches for VMR to improve. This article provides a brief overview on the management of VMR and discusses the various endoscopic surgical approaches to vidian neurectomy.


Key words

vasomotor rhinitis, vidian neurectomy

Introduction

VMR is a form of non-allergic, non-infectious rhinitis. Studies have established that 14 million people in the United States are affected by VMR and that this disease costs $2-3 billion annually.1 VMR is an upper respiratory hyper-responsive to non-specific triggers like exposure to cold, strong smells, changes in air temperature, humidity, alcohol ingestion or tobacco smoke. It is difficult to differentiate non-allergic rhinitis from allergic rhinitis and post infectious rhinitis as all three forms nasal mucosa has hypersensitive vasomotor responses.

The mainstay of treatment is conservative management like any form of rhinitis focusing on symptom control and improving quality of life. This includes avoidance of stimulants, use of nasal anticholinergic nasal sprays along with topical nasal steroids. Surgery is indicated only in patients with severe symptoms who fail to respond to medical therapy.

Vidian neurectomy is advocated for the management of intractable VMR. This procedure was first described by Golding-Wood in the 1960s. It became unpopular because of difficulty in accessing the vidian nerve and the associated surgical morbidity, mainly from the approach, including bleeding, poor visualisation, difficulty in accessing and identifying the vidian nerve, damage to maxillary, maxillary veins, and infraorbital hypostasis.

However, recently endoscopic vidian neurectomy has gained popularity. The advances in endoscopic techniques have overcome the inherent risks of conventional vidian neurectomy, which has revolutionised the technique, improved access with better visualisation and risk of complications.

In this article we describe the anatomy of the vidian nerve, current surgical approaches for vasomotor rhinitis and discuss the associated complications with each of these procedures.

Surgical anatomy of vidian nerve

The vidian nerve is formed by the fusion of the greater superficial petrosal nerve and deep petrosal nerve which runs in the pterygoid (vidian) canal to enter into the pterygo-palatine fossa (PPF). The deep petrosal nerve carries sympathetic fibres which originate from the internal carotid artery whereas the greater superficial
Lee et al. classified the PC into three types based on CT (PC) courses in the floor of sphenoid sinus. The pterygoid (vidian) canal is located inferomedial to foramen rotundum on the floor of the sphenoid bone. The pterygoid (vidian) canal transmits the vidian nerve, ganglion, and the sympathetic component of the vidian sphenopalatine ganglion.

The vidian nerve passes through the ganglion. The parasympathetic fibres synapse with cell bodies of the ganglion, and the sympathetic component of the vidian nerve passes through the ganglion. The pterygoid (vidian) canal transmits the vidian nerve and vessels. The anterior opening of the pterygoid (vidian) canal is located inferomedial to the foramen rotundum on the upper medial part of the anterior surface of the pterygoid process of the sphenoid bone. The pterygoid (vidian) canal then courses in the floor of the sphenoid sinus.

Lee et al. classified the PC into three types based on CT findings. The pterygoid canal protruding completely within sphenoid sinus - type 1 (Fig 1); the pterygoid (vidian) canal is on the floor of the sinus or partially protruding into the sphenoid sinus - type 2 (Fig 2); The PC is completely embedded in the sphenoid corpus - type 3 (Fig 3). Azer et al. reported that type 2 PC is found most commonly (54%), followed by type 3 VC (36%) and type 1 (10%).

There are huge anatomical variations of PC from person to person and from one side to the other therefore it is very important to evaluate and analyse the anatomical variations on CT pre operatively.

Surgical approaches to vidian neurectomy:

Golding-Wood first described vidian neurectomy (VN) for VMR in 1961. Later, a transeptal approach to the pterygopalatine fossa (PPF) and VN was described by El-Guindy. Due to difficult access and significant complications, these procedures were abandoned.

The recent advances in endoscopic sinus surgery has revitalized an interest in delineating the anatomy of the vidian nerve and PPF and new endonasal approaches to this area began to surface.

Endoscopic vidian neurectomy (EVN) There are various approaches described in literature.

1. Pterygopalatine fossa (PPF) approach Robinson and Wormald described a technique of EVN where a U-shaped incision is made over the sphenopalatine foramen (SPF) and a mucosal flap is raised up and over the sphenoid face. The bone from SPF to the anterior face of sphenoid is drilled down up to peristomeum. The peristomeum is incised to open into PPF and the vidian nerve is identified in the PPF and is cut. The sphenopalatine artery is cauterized early on to prevent bleeding.

2. Transsphenoidal/Intrasphenoidal Approach Su et al. introduced an antegrade transsphenoidal approach with two subtypes. The subtypes depend on how much (or how little) the pterygoid (vidian) canal protrudes into the sphenoid cavity above the sinus floor. A generous sphenoidotomy at the sphenoid ostium is performed in both types. The first subtype is intrasphenoidal approach which consists of de-roofing the superior aspect of the bony vidian canal in the floor of the sphenoid sinus followed by isolating a length of the nerve to then divide it (Fig 4). This approach can only be used for those well-pneumatized sphenoid sinuses in which the pterygoid (vidian) canal is thin and well isolated from the surrounding bone of the sinus floor and walls. This anatomic variation is uncommon.

Second subtype is transsphenoidal approach, where the bone of the anterior wall of the sphenoid is taken down, and this bony removal is carried out laterally to the sphenoid process of the palate bone. The sphenoid process is resected until the pterygoid (vidian) canal can be identified. A curved probe can be used to “hook” the vidian nerve, and once isolated, the nerve is then divided.

Endoscopic posterior nasal neurectomy

This procedure involves transection of the posterior nasal nerve (PNN) for the treatment of intractable VMR. PNN is a nerve ramus from the pterygopalatine ganglion innervating the nasal mucosa. This nerve supplies only nasal mucosa therefore transection of PNN avoids the postoperative complication of dry eyes. Palatal numbness may also be avoided, because the posterior nasal nerve is not located as close to the maxillary nerve as is the vidian nerve. Two separate techniques for PNN transection have been described. The first, referred to as the trans turbinate approach, is typically performed in combination with submucosal resection (SMR) of the inferior turbinate. From the incision in the turbinate, the mucosa of the middle meatus/lateral nasal wall is elevated and the peristomeum cut and elevated until the sphenopalatine foramen (SPF) is visualized. The nerve bundle identified as the posterior nasal nerve courses from the SPF toward the inferior turbinate. This is isolated and cut, or when it cannot be isolated from the sphenopalatine artery, the structures are separated with a bout of laser energy. This nerveBalanced with a second technique uses a transnasal approach similar to that described for transnasal endoscopic sphenopalatine artery ligation and begins with a vertical incision made in the middle meatus roughly 5 mm anterior to the lateral attachment of the middle turbinate. A mucoperiosteal flap is elevated posteriorly to the cribo ethmoidalis and then above and below this point until the SPF is identified. The posterior nasal nerve is identified along with the sphenopalatine artery (SPA) and divided.

Complications of vidian neurectomy

The most common complication after VN is postoperative xerophthalmia (dry eyes). This occurs in 23–100% of the patients. Some authors considered xerophthalmia as a confirmatory sign for vidian neurectomy. These effects are

![Figure 1: Type 1 (vidian nerve is protruding in sphenoid sinus cavity)](image1)

![Figure 2: Type 2 (vidian nerve on the floor of sinus)](image2)

![Figure 3: Type 3 (vidian nerve in the corpus of sphenoid sinus)](image3)

![Figure 4: Intra operative image showing dissection of vidian nerve and vidian nerve is shown lifted using a ball probe)](image4)
permanent numbness were reported.11-13 No long-term complications from xerophthalmia were reported. Nasal crusting or dryness occurred in 15-28% of patients.14,15

Check, palate and/or gingival numbness was reported to occur in 3-22% of patients.8,15,16 All of the numbness resolved within 1 week to 12 months and no cases of permanent numbness were reported.11,13

From recent systematic reviews on surgical management of VMR, it is evident that EVN is effective in controlling symptoms of refractory VMR in most patients and its effects are maintained over a period of 2-5 yrs. EVN is associated with less morbidity than traditional transantral approach and they are transient.15,17

Discussion

First-line treatment of VMR is medical and in the form of topical therapies including intranasal corticosteroids, intranasal antihistamines, and intranasal ipratropium bromide. Surgical intervention should only be considered in the event that aggressive medical therapy has failed to control a patient’s symptoms. A variety of surgical options for medically refractory VMR have been described. Postoperative xerophthalmia is significantly more likely to occur when using the intraphenoidal or transphenoidal approach compared with the PPF approach. Alternatively, check-palatal numbness is significantly more likely to occur after the PPF approach than the intra- or transphenoidal approach. Fortunately, any complications experienced after EVN are self-limiting and will most likely resolve on their own. The choice of approach should be based on the patient’s anatomy and the surgeon’s experience. If the patient’s anatomy is favorable for intraphenoidal identification and resection of the vidian nerve, this approach should be used above others because it is considerably less invasive. This anatomic configuration of VMR, it is evident that EVN is effective in controlling symptoms of retractable VMR in most patients and its effects are maintained over a period of 2-5 yrs. EVN is associated with less morbidity than traditional transantral approach and they are transient.15,17

References

Giant Parathyroid Adenomas: A Minimal Invasive Parathyroidectomy Approach to 15 cases

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Abstract

Introduction
We present a case series of 15 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 > 3.5g. They have a greater pre-operative calcium and parathyroid hormone level.

Methods:
A case note review of 15 giant adenomas operated on at a single institution by the senior author was performed (2006-2015). There were 7 males and 8 females, with a median age of 60.8 years. Data was collected on patient demographics, symptoms, biochemistry, ultrasound and sestamibi results, operation outcomes, complications and histopathology.

Discussion:
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 3.5g – 20g with a mean of 6.33g. Histology confirmed all the glands to be benign. All patients had a complication-free postoperative period.

Results:

Hybrid Tracheostomy: a safe, effective technique with elimination of waiting times for emergency theatre?

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Introduction:
Percutaneous tracheostomy gained popularity as it can be performed by intensivists, on the unit without transfer to an operating theatre.

Surgical tracheostomy is reserved for patients in whom a percutaneous tracheostomy is deemed unsuitable.

From our experience, patients have prolonged delays due to lack of a suitable emergency theatre slot and availability of ENT personnel.

In order to streamline the surgical tracheostomy provision, we developed a hybrid tracheostomy technique.

This paper describes the technique, its advantages and reviews the outcomes of our small pilot study.

Methods:
A minimally invasive technique of hybrid tracheostomy combines the open approach of a surgical tracheostomy, to allow direct visualisation of the trachea and deal with any bleeding or anatomical difficulties, followed by...
percutaneous tracheostomy technique for insertion of the tracheostomy tube.

**Results:**
A total of 9 patients have undergone the hybrid tracheostomy as part of our pilot study.

It avoids transfer of the critically ill patient from the ICU to the operating theatre

It is a shorter procedure than a percutaneous tracheostomy (Range 5 – 10 minutes).

The time between referral to tracheostomy being performed was reduced as the waiting period for an emergency theatre slot was eliminated. (Waiting period range was 8 – 96 hours).

There were no reported intra-operative or post-operative complications.

**Conclusion:**
This is a safe technique performed under direct vision, and allows effective insertion of a tracheostomy tube. It reduced waiting times and can be scheduled for a convenient time.

We would recommend that other departments consider the implementation of this technique.

**References:**
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