Chapter 10: Neurological affections of the pharynx and larynx

David Howard

Neuroanatomy

Neuronal control of the pharynx and larynx begins in the cerebral cortex at the lower part of the precentral gyrus. Additional fibres also arise from some frontal and parietal areas. The majority of fibres descend in the pyramidal tracts, with subsequent partial decussation at the upper border of the medulla to reach the nucleus ambiguus. Most palsies are produced by lesions of the nucleus ambiguus or the vagus nerve trunks and their branches. Lesions can cause unilateral or bilateral paralysis of the pharynx, often associated with palatal paralysis, and/or paralysis of the larynx and cricopharyngeus.

The cranial portion of the accessory nerve contains motor fibres destined for the muscles of the soft palate (except tensor veli palatini), pharynx, and intrinsic larynx.

Glossopharyngeal nerve

The glossopharyngeal nerve is essentially sensory. The nerve is not motor to the palate (it only supplies the stylopharyngeus which cannot be tested clinically) so that when the gag reflex is tested the stimulus is carried by the ninth nerve but the resulting palatal movement is mediated by the tenth nerve.

Vagus nerve

The vagus nerve leaves the cranium in the same sheath of dura as the accessory nerve. The glossopharyngeal nerve lies in front of these. The vagus is swollen during its exit by the superior ganglion cells. Below the jugular foramen the nerve is swollen again for approximately 2.5 cm by the inferior ganglion. This latter ganglion is crossed by the cranial root of the accessory nerve which blends with the vagus below the ganglion. The cells of the ganglion are sensory only.

The vagus nerve descends in the neck within the carotid sheath, between the internal jugular vein and the internal carotid artery, as far as the superior border of the thyroid cartilage and then between the vein and the common carotid artery to the root of the neck. Its subsequent course differs on each side.

Pharyngeal branch

This arises from the upper part of the inferior ganglion and runs between the external and internal carotid arteries to the superior border of the midline constrictor. It forms the pharyngeal plexus along with branches from the sympathetic trunk, glossopharyngeal and external laryngeal nerve.
Superior laryngeal nerve

This arises from the lower portion of the inferior ganglion, and descends on the side wall of the pharynx posterior and then medial to the internal carotid artery. It divides into the internal and external laryngeal nerves.

Internal laryngeal nerve

This descends to the thyrohyoid membrane piercing it above the superior laryngeal artery. It divides and supplies sensory innervation to the supraglottis, valleculae, and posterior surfaces of the arytenoid cartilages. Inferiorly it communicates with ascending branches of the recurrent laryngeal nerve. It also carries afferent fibres from neuromuscular spindles and cricoarytenoid joint receptors (Wyke and Kirschner, 1976).

There are abundant laryngeal receptors, both tactile and pain, providing a high degree of sensitivity particularly of the supraglottic mucosa. The afferent loop of the reflex initiates a bilateral laryngeal reflex. The chemoreceptors in the larynx have unknown functions. The subglottis has a similar receptor, served by the recurrent laryngeal nerve. These subglottic chemoreceptors apparently modify breathing, since some chemical stimuli applied to them initiate slow deep breathing, as does carbon dioxide applied to this part of the tract when it is isolated. The articular joint afferent fibres are particularly well developed, producing a highly sensitive and rapid monitoring response. Some reflexes which control respiration and phonation may arise from stretch receptors in the musculature of the larynx, demonstrable both histologically and by electrophysiology. Debate continues with regard to the exact role of all the sensory receptors and the reader is referred to an excellent contribution by Wyke and Kirschner (1976). Debate also remains as to whether the internal laryngeal nerve is entirely sensory: Williams (1951) described motor fibres innervating the interarytenoids. Certainly no obvious interarytenoid contracture takes place on stimulation of the superior laryngeal nerve.

External laryngeal nerve

This branch descends deep to the superior thyroid artery on the inferior constrictor. It then pierces the muscle, passes around the inferior thyroid tubercle and supplies the cricothyroid muscle. It also gives branches to the pharyngeal plexus and inferior constrictor muscle.

Recurrent laryngeal nerves

The nerve on the right side arises from the vagus in front of the first part of the subclavian artery, winds backwards around the vessel and ascends obliquely behind the common carotid to the side of the trachea. On the right the nerve is commonly in front of the tracheo-oesophageal groove and may even be considerably lateral to the trachea as it reaches the lower pole of the thyroid gland. Near the lower pole both recurrent nerves are always intimately related with the terminal branches of the inferior thyroid artery. Authors differ in reporting the percentage of each variation, but on the right side the nerve occurs with approximately equal incidence behind, in front of, or intermingling with, the terminal branches of the inferior thyroid artery.
On the *left* side the nerve curves around the arch of the aorta and ascends commonly in a more protected position within the tracheo-oesophageal groove. On this side it is most likely to run behind the branches of the inferior thyroid artery, and least likely to pass in front of them.

Both nerves are intimately related to the medial surface of the thyroid lobes and their fascial coverings before passing under the lower border of the inferior constrictor to enter the larynx behind the cricothyroid joint.

As they ascend, each nerve gives branches to the mucous membrane and muscular coat of the oesophagus and trachea. The nerves may divide before entering the larynx. They supply all the muscles of the larynx, except the cricothyroid, and sensation to the mucous membrane below the vocal cords. They also give branches to the inferior constrictor and communicate with the internal laryngeal nerve.

**Aetiology**

It is difficult to produce an exhaustive list of causes of neurological deficits in the pharynx and larynx. The cause of many neural lesions, particularly of the larynx, are unknown.

**Supranuclear lesions**

Cortical lesions producing laryngeal and pharyngeal palsies are rare, and little is known about the effects on the larynx. It requires a bilateral symmetrical lesion of the cortex to produce a pharyngeal or laryngeal palsy, and in such cases respiratory and reflex laryngeal movements may be unaffected. Hemiplegia does not impair vocal cord movements.

**Nuclear lesions**

The nucleus ambiguus may be involved in posteroinferior cerebellar artery thrombosis, tumours of the medulla, bulbar palsy, syringobulbia, motor neuron disease, encephalitis, poliomyelitis, cranial polyneuritis, tabes and rabies. The most common cause of bilateral laryngeal paralysis arising in the nucleus is progressive bulbar palsy or cranial polyneuritis. Before the advent of lead-free pain it was a rare manifestation of lead poisoning. Nuclear lesions usually cause a combined paralysis of the soft palate, pharynx and larynx, but the larynx may be spared if only the superior part of the nucleus is affected. This may produce a palatopharyngeal paralysis - the syndrome of Avellis.

**Posterior fossa and jugular foramen lesions**

These involve the vagus nerves as they emerge from the brainstem and leave the skull. Nerves IX, XI and XII may also be affected and a large number of laryngopharyngopalatal palsies combined with lesions of these nerves have been named. They come under the headings of posterior fossa syndrome and jugular foramen syndrome. Although the many eponyms are of historical interest their use can lead to confusion.
The commonest combinations of associated cranial nerve lesions in the region are:

1. IX, X, XI, in the jugular foramen, Vernet's syndrome;
2. X, XI, Schimdt's syndrome;
3. X, XI, XII, Hughlings Jackson syndrome;
4. IX, X, XI, XII, Collet-Sicard syndrome;
5. IX, X, XI, XII and Horner's syndrome, Villaret's syndrome.

Having established the combination of nerve palsies the presence of an additional Horner's syndrome indicates a lesion outside the skull as the cervical sympathetic ascends to the base of the skull but does not pass through the jugular foramen. Evidence of additional brainstem compression obviously indicates an intracranial lesion.

Lesions in this region may arise from a wide range of diseases. These include skull fractures, primary tumours (particularly glomus tumors, meningiomata) of the temporal bone and postnasal space, metastases, cholesteatoma, extension of infection from the middle ear, jugular bulb thrombophlebitis, tuberculosis, and syphilitic meningitis.

**Extracranial lesions**

In the cervical region the list of causes includes penetrating injuries; tumours of the hypopharynx, upper oesophagus, thyroid and parapharyngeal space; enlargement of/or surgery on the regional lymph nodes; and surgery of the thyroid gland.

Arising in the thorax the left recurrent laryngeal nerve has a longer course and is more exposed to compression by an aortic aneurysm, an enlarged left atrium in mitral stenosis, carcinoma of the bronchus, other mediastinal tumours and enlarged mediastinal glands.

The right recurrent laryngeal nerve is more vulnerable to injury during surgery on the thyroid gland because of its more anterior and lateral position at the inferior pole of the gland, rather than being protected in the tracheo-oesophageal groove.

**Paralysis of the palate and pharynx**

**Symptoms**

Unilateral palatal paralysis may not give rise to any symptoms because of compensation by the unparalysed muscles of the opposite side. Direct questioning may reveal slight changes in phonation (especially in professional voice users), snoring, postnasal drip and occasionally a unilateral hearing loss because of eustachian tube dysfunction on the affected side.

Bilateral palatal paralysis prevents the palate from closing off the oropharynx from the nasopharynx and prevents control of airflow through the nose - fluids, and sometimes solids,
thus regurgitating through the nose during deglutition. The voice has a nasal quality (rhinolalia aperta), as is heard in an untreated cleft palate. There is also a tendency to mouth breathing, snoring at night, and mucoid rhinorrhoea.

The patient with a bilateral palatal paralysis can usually swallow sufficient for his needs unless the pharynx is also involved. A combined paralysis is much more serious, particularly if the pharyngeal paralysis is bilateral; the patient cannot swallow and attempts to do so result in spasms of coughing as a result of overspill into the larynx. Soft bulky foods are usually more easily swallowed than liquids and normal solids. Even the patient's ordinary secretions may enter the lower respiratory tract and cause inhalation pneumonia.

In unilateral pharyngeal paralysis the compensation by the opposite constrictors is often efficient, but the patient needs to swallow in a deliberate manner, has bouts of coughing to 'clear' the throat, and may find it easier to sleep on the affected side to prevent laryngeal irritation from pharyngeal secretions.

**Signs**

Unilateral palatal paralysis is detected by examination of the oropharynx. When the patient says 'ah' the palate does not rise on the affected side and the uvula is drawn to the normal side. With a bilateral palatal paralysis the palate remains immobile during phonation. Occasionally it may be difficult to decide whether there is any movement in a bilateral paralysis. The patient cannot whistle or blow up a balloon.

Sensation is tested by touching each side of the palate gently with an orange stick and asking the patient to compare the two. Pain sensation may also be tested, with care, using a long pin. The posterior pharyngeal wall is tested in the same manner. With a unilateral pharyngeal paralysis the pharyngeal reflex is lost on the affected side and the pharyngeal wall droops. In both unilateral and bilateral pharyngeal paralysis pharyngeal secretions collect in the hypopharynx and around the laryngeal inlet and may be clearly seen on indirect laryngoscopy. If the symptoms are strongly suggestive of bilateral pharyngeal paralysis care should be taken on indirect laryngoscopic examination as it may produce sudden aspiration of these pooled secretions.

**Diagnosis**

Rarely, palatal paralysis in children may be referred to the otolaryngologist because the symptoms are believed to be caused by 'adenoids'. The history of enlarged adenoids is very different: the child has nasal obstruction but no nasal regurgitation. The thick nasal intonation caused by enlarged adenoids produces the contrasting speech defect of rhinolalia clause.

Reduced motility, and occasionally fixation, of the palate may be produced locally by scarring following unsatisfactory cleft palate repair, adenoidectomy or tonsillectomy, syphilis, scarlet fever, and tumours.
Investigation

Paralysis of the palate is unlikely to require investigation other than for the underlying cause. Accurate recordings of the degree and type of nasopharyngeal closure with palatal movement have been studied using video recordings obtained via flexible nasoendoscopy. This evaluation is only necessary when palatal or nasopharyngeal surgery is proposed for a long-term and stable palatal paralysis.

Pharyngeal paralysis with varying degrees of dysphagia has been the subject of physical, radiological, endoscopic and electrophysiological investigations. By far the most common in clinical use has been the radiological evaluation of pharyngeal and oesophageal swallowing using radiopaque contrast media such as barium, or the water soluble material gastrografin. Modern cineradiology and cine-fluorography techniques have been further enhanced by the addition of videotape facilities allowing detailed and repeated analysis of each phase of swallowing. The degree of overspill can be seen and cricopharyngeal spasm and notable hold-up clearly shown. This latter feature may help to identify those patients who would benefit from a cricopharyngeal myotomy, but sadly the radiographic appearances and subsequent postmyotomy results do not always correlate.

Pharyngeal pressure measurements, pH monitoring, electromyographic recording and endoscopic examination, although used in research, have yet to prove their worth in most clinical situations.

Treatment

Treatment differs according to the cause and extent of the lesion. Isolated palatal paralysis rarely requires treatment, although troublesome nasal regurgitation may be helped by holding the nose during swallowing or by an upper dental palate with a soft palate extension.

Pharyngeal paralysis requires treatment to provide adequate nutrition and prevention of inhalation pneumonia. Temporary help may be obtained by a fine bore nasogastric tube for feeding and repeated pharyngeal suction to protect the airway. This is unsatisfactory in the long term, and tracheostomy and gastrostomy may be necessary. A cuffed tube is used in the tracheostomy but even this requires regular deflation and may not provide adequate protection against pharyngeal secretions. Under these circumstances total laryngectomy has been used to separate the food and air passages. The neurological deficit can be unpredictable in the long term and spontaneous improvement may occur depending on the aetiology. If this is at all likely, a satisfactory compromise is provided by epiglottopexy (Brookes and McKelvie, 1983) in which the laryngeal inlet is occluded by stitching the epiglottis to the supraglottis via a lateral pharyngotomy leaving a small posterior defect. A valved tracheostomy tube allows speech, while the epiglottopexy prevents any significant overspill. The operation can be reversed if subsequent neurological improvement takes place.
Pharyngeal neurological lesions

*Globus pharyngeus (globus 'hystericus', idiopathic globus)*

This functional disorder is most common in middle-aged women and is associated with a variety of sensations in the pharynx and larynx. Emotion, particularly fear, can cause a marked sensation of a lump, dryness, or 'blockage' of the throat, so it is not surprising that people who are emotionally unstable should refer symptoms to this region. However, the term globus hystericus is misleading as true hysteria is rare and hysterical dysphagia is uncommon even in these patients.

The most common feature in globus pharyngeus is the sensation of a lump in the throat. Often the patient's attention has been drawn to the throat by a previous minor throat infection, a transient incident with food 'sticking' in, or 'catching' the throat, or a relative or friend dying of 'cancer of the throat'. The symptom is most obvious when the patient attempts to swallow his own saliva to 'see if the lump is still there', but there is no true dysphagia and the symptoms often disappear while eating a meal. Repeated attempts at swallowing saliva may lead to aerophagy, with gastric distension and discomfort. The symptoms may have been present for many months and are usually worse if the patient is under any form of stress.

Examination often reveals a notably anxious patient with a pronounced gag reflex, but the pharynx, oral cavity, larynx and neck do not show any evidence of disease.

It is advisable to investigate these patients by a blood count to exclude anaemia, and by a barium swallow which may show obvious cricopharyngeal spasm without any other abnormality, but the role of this finding in the pathogenesis of the problem remains unclear. The spasm may be associated with lesions at the lower end of the oesophagus such as hiatus hernia, carcinoma or achalasia. There is no doubt that many globus patients can be demonstrated to have a hiatus hernia and it is postulated that the lower oesophageal pathology causes reflex vagal stimulation and subsequent alteration in the tone of the hypopharyngeal musculature, particularly the cricopharyngeus. However, many patients have hiatus hernias without globus type symptoms and research has not yet explained the cause of globus pharyngeus.

If symptoms persist direct rigid, or flexible endoscopy may be indicated as these methods are more likely to detect early organic disease, particularly of the postcricoid area, than radiology.

If no organic disease is shown by adequate investigation the patients are often rendered symptom free by reassurance and explanation. Occasionally, an anxiolytic, such as diazepam, may help troublesome symptoms but this type of medication is only required for a small proportion of patients. Speech therapists may have a useful part to play, particularly if there is an associated functional dysphonia. Rarely, the more emotionally crippled patient requires psychiatric referral.
Cricopharyngeal spasm and pharyngeal pouch

These pharyngeal disorders almost certainly have a neurogenic basis but are more fully discussed in Chapter 14.

Glossopharyngeal neuralgia

Apart from the distribution of the pain, glossopharyngeal neuralgia resembles the much commoner condition of trigeminal neuralgia. The pain occurs in brief agonizing stabs which may be of great severity. They usually start in relation to the tonsil and radiate down the side of the neck, in front of the ear and to the back of the mandible. Very rarely the pain may begin deep in the ear.

The attacks are usually precipitated by swallowing or by protruding the tongue, but when the ear is the main site external stimulation of the ear or skin may provoke an attack. Similar, but more continuous pain may be caused by tonsil or pharyngeal neoplasm and this must be carefully excluded.

When the throat is the main site of pain, some relief may be obtained by direct application of cocaine to the lateral pharyngeal wall and posterior third of the tongue. Carbamazepine (Tegretol), given in increasing dosage up to 200 mg four times daily, may control the attacks and additional relief may be obtained with sedatives. If these measures prove unsuccessful operation on the nerve may be necessary. It is always wise to regard the neuralgia as evidence of an underlying lesion until proved otherwise by events or by surgical exploration. The list of findings at surgical exploration includes a long styloid process, aberrant vessels coursing over the nerve, unsuspected neurofibromata and cholesteatomata. Skull base and lateral X-rays may be useful and selected cases may require computerized tomographic (CT) scanning.

The nerve may be approached via the tonsillar fossa, skull base, or intracranially. Wilson approached the nerve through the tonsillar fossa where it lies on the stylopharyngeus (Wilson and McAlpine, 1946). It may be avulsed or divided, but adequate safe dissection to the base of the skull is not possible by this route and only symptoms referable to the throat are relieved. A long styloid process may be fractured or partially removed during the same procedure (after tonsillectomy) but the vogue for this procedure seems to be declining.

Approach to the nerve at the skull base is difficult owing to its relatively small size, depth and relation to other important structures. It must be avulsed to remove the jugular and petrosal ganglia otherwise the connections with the tympanic plexus remain intact and any ear symptoms continue.

Injection of the nerve with alcohol at the point of emergence from the skull base can be undertaken but this is a difficult and hazardous procedure which has not gained wide acceptance.

The most reliable results are obtained with modern neurosurgical techniques involving craniotomy and division of the nerve fibres soon after emergence from the medulla.
Additional division of the upper two rootlets of the vagus nerve containing auricular and pharyngeal branches may be necessary.

**Herpes zoster**

Rarely, the neurotrophic varicella zoster virus may affect the distribution of the ninth and tenth cranial nerves. The vesicular eruption usually affects the palate, the pharynx and the laryngeal inlet on one side. The pharyngeal lesions may be isolated or be accompanied by other eruptions on the auricle or the anterior pillar of the fauces such as occur in geniculate herpes (Ramsey Hunt's syndrome).

The eruption is usually preceded by general malaise, fever and anorexia, particularly in the elderly. Herpes zoster may occur at any age, but most patients are over 50. Initially the throat is sore for a few hours before typical vesicles appear. These break down to form shallow ulcers, and they may be accompanied by severe pain. They heal without scarring but intractable pain may persist for many years in elderly patients. Local treatment with an antiseptic mouthwash keeps the pharynx clean, and local analgesics are useful before meals. Systemic steroids have been advocated but are of no proven value. Strong analgesics such as pethidine may be required in the acute stages but are contraindicated for persistent postherpetic pain. Carbamazepine (Tegretol) is rarely useful in postherpetic pain and nerve division or injection often provides only temporary relief. A combination of chlorpromazine and dihydrocodeine can be given for long periods and antidepressants may be required to treat the depression which may accompany the persistent pain.

**Diphtheria**

This disease is caused by the Klebs-Loeffler bacillus, *Corynebacterium diphtheriae*. It produces a membranous exudate at the initial site of infection which is later followed by distant toxic effects, of which polyneuropathy and circulatory failure are the most important.

It has a worldwide distribution but only isolated outbreaks occur in the UK following the introduction of active immunization. Because of the rarity at the present time the first cases of recent outbreaks have been missed and the subject deserves careful consideration.

Diphtheria is most common in the 2-10-year-old age groups and is spread by droplet infection from carriers and patients. Active immunization has been followed by the disappearance of carriers.

**Pathology**

The organisms remain at the site of infection and do not become invasive. The powerful exotoxin initially causes epithelial necrosis, followed by an inflammatory reaction with the necrosis forming the characteristic membrane. The membrane is 'false' as it consists of invaded and necrotic layers of mucosa and is not an exudate. The membrane is adherent and is difficult to remove. The corynebacteria are present at the margin of the membrane and swabs should be taken from this area.
The neurological symptoms are associated with fatty degeneration of the myelin sheaths of involved nerves with a consequent slowing of nerve conduction velocity. The fauces are the commonest site of diphtheric infection and as absorption of toxins is more rapid than from the other sites (nose, nasopharynx, larynx), faucial diphtheria is usually associated with most toxaemia. Palatal paralysis is attributed to the ascent of toxin from the common faucial site of infection to the medulla. (In cutaneous infection local ascent of the nerves by the toxin is responsible for the local development of paralysis.) Paralysis of accomodation, generalized polyneuropathy and myocardial damage are the consequences of dissemination of the toxin via the blood stream.

**Symptoms and signs**

Although infection can also occur in the nose, nasopharynx and larynx, the faucial site is much the commonest and paralysis of the palate is usually the earliest neurological symptom. The onset of faucial diphtheria is usually insidious: the child becomes quiet and anorexic but rarely complains of a sore throat. Lassitude and general malaise are associated with a normal temperature or mild pyrexia seldom above 38.4°C (101°F). Initially the membrane is absent, but may become extensive within 24 hours. It usually begins on one or both tonsils and spreads on to the fauces, uvula and palate. It may vary from cream to yellow to grey, glistens, and bleeds with removal. Paradoxically it is usually thinner and less well defined in the more severe cases but covers a wider area of the pharynx. Other sites of membrane formation, notably the nasopharynx, must always be checked. Fetor is striking and characteristic and there is usually a cervical adenitis. Early signs of toxicity are marked pallor, drowsiness, vomiting and tachycardia.

Palatal paralysis, usually bilateral, may occur within a few days but is commonest during the second or third week. It leads to regurgitation of fluids through the nose and a nasal voice. The palatal reflex is absent. Paralysis of accomodation usually develops in the third or fourth week and generalized neuropathy between 5 and 7 weeks after infection. This generalized neuropathy (which is not necessarily preceded by palatal and eye palsies) involves paralysis of the pharynx, intrinsic muscles of the larynx and the diaphragm. It is a serious complication because dysphagia may be complete and there is risk of aspiration pneumonia and respiratory failure.

**Diagnosis**

Any membranous throat condition should be regarded with suspicion particularly if it is difficult to remove and associated with a low grade fever but a relative tachycardia. Infectious mononucleosis can produce a grey membrane and oedema in the throat identical to diphtheria and is the commonest infection to cause real difficulty in diagnosis. However, other signs such as generalized lymphadenopathy, splenomegaly and the finding of abnormal mononuclear cells in the peripheral blood help to distinguish the condition. Agranulocytosis and acute leukaemia may produce lesions of the throat resembling diphtheria.

Quinsy, Vincent's angina, thrush and herpes zoster should be readily distinguishable.

Nose and throat swabs should be taken in every suspected case, but negative cultures do not exclude diphtheria and bacteriological confirmation of virulent diphtheria may take 3
or 4 days. Paralytic complications increase in frequency and extent with increasing delay of treatment by antitoxin, and this therapy must never be withheld while awaiting bacteriological confirmation of the suspected clinical diagnosis. Once toxin is fixed in heart muscle or peripheral nerves it is not affected by antitoxin which can only neutralize circulating toxin.

**Treatment**

The importance of rest in this condition cannot be overemphasized. All patients should be nursed flat in isolation, often for many weeks. They should be mobilized gradually and only when clinical and electrocardiographic evidence of myocarditis is absent. The old fashioned adage of 'a pillow a week' until the patient is sitting up remains a good regimen. As complete recovery can ensure in many cases, it is tragic when death results from too early and too great an exertion because of impatience.

The dose of antitoxin required depends on the site of the disease, the extent and duration of the membrane. It varies from 20,000 units intramuscularly to 200,000 units intravenously. A repeat dose of antitoxin is unnecessary if the correct initial assessment is made but can be given after a 2-3 day delay if the membrane continues to spread. Penicillin should be given to all cases to limit the spread of infection. It does not affect any preformed toxin and is in no way a substitute for antitoxin. Intravenous injections of hydrocortisone and 1:1,000 adrenaline drawn up into syringes must be available when administering antitoxin as it is a horse-serum preparation. A history must be taken of previous injections and allergy.

Palatal paralysis usually calls for no treatment other than that dictated by the general condition of the patient, but when the pharynx is affected a nasogastric tube must be passed and left in situ to provide feeding and to lessen the risk of inhalation pneumonia. A sucker may be used to remove pharyngeal secretions, but in severe cases a tracheostomy with a cuffed tube may be necessary. If both respiratory and pharyngeal muscles are affected intermittent positive pressure ventilation can be carried out via the tracheostomy.

Although paralysis recovers spontaneously, the restoration of movements is said to be hastened by exercises, such as whistling and blowing up balloons.

**Prognosis**

This depends on the virulence of the infecting organism, the position and extent of the membrane, and the delay in administering antitoxin. Death from diphtheria during the first week is caused by circulatory failure or laryngeal obstruction. Myocarditis may prove fatal in the second or third weeks and respiratory failure is responsible for most later deaths. Complete recovery without sequelae is usual but may take up to 6 months in severe cases. Most patients are out of danger within 10 weeks. Active immunization with triple vaccine, commencing at 4 months of age, usually prevents or modifies diphtheria but fatal attacks may still occur.

Most convalescent and long-term carriers can be treated with a 7-day course of erythromycin. Tonsillectomy is rarely required.
Acute anterior poliomyelitis

This acute viral disease is characterized by local or widespread muscular paralysis resulting from destruction of anterior horn cells in the spinal cord and corresponding cells in the nuclei of the cranial nerves. It is of particular interest to the otolaryngologist because paralysis of the pharynx and larynx occur in the bulbar type of disease, with involvement of the brainstem nuclei.

Aetiology

This disease is also known as infantile paralysis and in areas of the world where the virus is uncontrolled and sanitation poor, young children are still the main victims. However, in developed countries there has been a shift of the age of onset towards young adults. In the UK and many other countries which have had active immunization campaigns, poliomyelitis occurs only sporadically and the classical midsummer and autumn epidemics are of historical interest only.

The neurotropic virus exists in three known types which have different antigenic properties. It is transmitted by pharyngeal secretions and food contaminated by virus excreted in the faeces. The virus enters the body via the nasopharynx or gastrointestinal tract and has an incubation period of 7-14 days. Axonal spread occurs, but the virus is also bloodborne and this is the main route of transmission to the central nervous system. Aycock and Luther (1929) indicated that the virus easily gained entry via the raw tonsillar bed after tonsillectomy. There is also good evidence that physical exertion during the stage of incubation predisposes to increased paralysis, particularly in those muscle groups used in the exercise. Paralysis may also develop in limbs into which injections are given. All these factors are obviously best avoided during an epidemic. Detailed reviews of the virology and epidemiology are available (Cohen, 1969; Drouhet, Debre and Celers, 1970).

Pathology

The virus is often widespread throughout the brain and spinal cord but has a particular affinity for motor nuclei of cranial nerves and the anterior horn motor cells. Depending on the severity of the attack these cells necrose, usually within the first few days of the disease, following which no further destruction occurs. Other cells show evidence of damage by loss of Nissl granules but these are presumably capable of recovery. Necrosed cells are removed by neurophages, and examination of the spinal cord many years after the acute episode may show almost complete loss of anterior horn cells in the affected areas. Approximately one-third of cells have to be destroyed to induce clinical signs of paralysis. Paralysed muscle undergoes severe neurogenic atrophy.

Symptoms and signs

There are three possible distinct phases of this illness. Entirely subclinical infections and trivial illnesses lasting a few days with headache, diarrhoea and sore throat occur in most cases. The illness aborts before the paralytic stage and these apparently healthy individuals greatly outnumber paralytic cases and probably provide the greatest source of infection. Depending on factors such as the type and virulence of the virus, a variable proportion of
patients who recover from the initial mild illness enter a second stage of meningitic involvement, the so-called pre-paralytic stage, 2-3 days after recovery from the first. Headache and general malaise return, are far more severe and are accompanied by lumbar and limb pain, and cervical rigidity. In children delirium and convulsions may occur. This group of symptoms resembles that of other forms of viral meningitis.

Finally, a small group of patients continues into the third, paralytic phase. Unfortunately, some adults who develop paralysis do not go through two clearly defined preceding phases. Prediction of the extent of paralysis is difficult but severe limb pain and loss of tendon reflexes suggest a bad prognosis. Rest in bed at the first sign of an attack is essential.

In most patients maximum paralysis is reached within 3 days and occurs in a random manner. Spinal paralysis may affect the respiratory muscles and early recognition is important. A useful simple test is to ask the patient to count as far as possible with a single breath, a number below 15 suggests serious respiratory insufficiency.

Brainstem damage is serious, swallowing becomes impossible and pharyngeal secretions are aspirated. Combined bulbar and respiratory paralysis is still often fatal despite the best treatment.

Fortunately, except in severe cases, only some of the muscles originally affected remain paralysed. Recovery begins after about a week, and may continue for over 3 months.

**Treatment**

Barrier nursing is essential. Both bulbar paralysis and respiratory paralysis may require the attention of the otolaryngologist. Respiratory embarrassment caused by accumulation and aspiration of pharyngeal secretions must be distinguished from true paralysis of the muscles of respiration. Respiratory weakness requires artificial respiration. If there is no bulbar palsy a tracheostomy is not necessary if a negative pressure cuirass or cabinet respiratory is used. These respirators once widely used with good results, require critical day-to-day management with a high level of skill on an appropriate unit. Nowadays they have been largely replaced by intermittent positive pressure ventilation via a naso- or oral endotracheal tube, or through a cuffed tracheostomy tube. The advantage of intermittent positive pressure ventilation is that, if bulbar paralysis is also present, the cuffed intratracheal tube prevents the aspiration of pharyngeal secretions, food and vomit. Nasogastric feeding is obviously important in patients with bulbar paralysis. These patients require detailed management of blood gases, bronchial secretions, secondary infection, nasogastric feeding, care of the skin, bladder, bowels, and satisfactory communication. These many factors are best undertaken by an experienced team on an intensive care unit.

**Prevention**

Salk introduced the first vaccine by killing the three virus strains with formalin and injecting the preparation systemically. The value of this preparation became well established in the 1950s but it did not prevent colonization of the gut by wild virus and an immune patient could still be a carrier. It has therefore been superseded by the oral administration of
an attenuated live vaccine of the Sabin type, given as one or two drops on a sugar lump. Immunity appears to last for at least 3 years and in children and young adults booster doses are needed at intervals of a few years. There can be no doubt that in countries where a sustained vaccination programme has been pursued paralytic poliomyelitis has been virtually abolished but it is important that standards are not relaxed allowing the problem to grow again (Paul, 1971).

Rabies

This viral disease of mammals is usually transmitted to man by the bite of a dog with infected saliva. Rabies was eradicated from the UK 80 years ago but remains enzootic in foxes in mainland Europe. It has been estimated to cause 15,000 deaths a year in other parts of the world. 'Furious' rabies is characterized by intense arousal and hydrophobia associated with inspiratory muscle and pharyngeal spasm. 'Dumb' rabies presents as an ascending paralysis.

Pathology

The virus enters via a bite and is transmitted along nerve trunks in both directions to infect many organs including salivary and lacrimal glands. Brain and spinal cord show ganglion cell degeneration and marked perineural and perivascular round cell infiltration.

Symptoms and signs

The incubation period varies widely from 10 days to years (usually 1-2 months depending on the distance of the infected bite from the central nervous system. The first general symptoms are depression, apprehension and insomnia. Then follows hydrophobia, the classical sign of rabies: attempts to drink water produce gross spasm of the pharynx, larynx and respiratory muscles. This spreads to all the muscles of the body producing opisthotonus. At its height even the sound or thought of water will elicit the spasm. Other signs are hypersalivation, dysphagia, cranial nerve palsies, cardiac arrhythmias and severe psychiatric disturbance. Patients sink into a coma after a few days and even with intensive care only a few patients survive proven rabies.

The condition must be distinguished from tetanus which has a much shorter incubation period of 2-3 weeks. Trismus is the early symptom and pharyngeal spasm is absent. In hysteria true pharyngeal spasm is absent and the problem is amenable to drugs and suggestions.

Prophylaxis and treatment

Animal bites must be thoroughly cleaned, and the wound left open. The indications for vaccine treatment are clear (WHO Committee, 1966) and it should begin as early as possible after the bite. The newer inactivated vaccine given in human diploid strain COI38 has been shown to be safe and effective (Wiktor, Plotkin and Grella, 1973; Aoki et al, 1975). Hyperimmune gamma globulin is now given as a rule with vaccine.
Patients with established rabies require heavy sedation to control spasm and suffering. Total paralysis with curare and artificial respiration offer the only hope.

**Myasthenia gravis**

This disease occurs in all ages and all races but is twice as common in women as men. It is usually seen in young adults and may present to the otolaryngologist when the bulbar muscles are involved. Current evidence suggests it is an autoimmune disease with damage to the acetylcholine receptors in the motor end-plate. Any group of muscles may be affected and the disease tends to remit and relapse. There is abnormal muscle fatiguability, sometimes confined to an isolated group of muscles. The extraocular muscles are those most commonly involved. If it is restricted to the bulbar muscles there is dysarthria, dysphagia, regurgitation of fluid through the nose, and movements of the tongue, palate and pharynx are decreased. The symptoms typically appear in the evening when the patient is tired, particularly towards the end of a meal. They improve after a night's rest. If the patient is asked to count aloud the voice becomes less distinct and more nasal.

Diagnosis depends on the typical clinical picture, the evaluation of the effects of intravenous edrophonium (Tensilon test), modern electromyographic measurements, or the detection of antiacetylcholine receptor antibodies in the blood.

The standard symptomatic treatment for myasthenia gravis is still neostigmine or pyridostigmine, the dosage of which is steadily increased until the maximal effect is obtained. This does not affect the underlying natural history. Immunosuppressive treatment with prednisone, with or without azathioprine (Imuran) may be required. Plasmapheresis has been life saving.

Most remissions occur in the first few years of the disease but unfortunately relapses are common. Most deaths also occur in the early years of the disease. Neonatal myasthenia is occasionally seen in infants of affected mothers but usually recovers in a few weeks.

Thymic enlargement is often found in young patients and may be caused by a thymoma, some of which are malignant. The place of thymectomy remains debatable. The results are best in young women with a short and severe history, but both sexes may benefit and operation is indicated if there is rapid deterioration with optimum medication. If a thymoma is present radiotherapy may be used before surgery. Only one-third of patients with thymoma survive beyond 5 years.

Myasthenia is closely related to thyrotoxicosis and less commonly with sarcoidosis, diabetes mellitus, rheumatoid arthritis and systemic lupus erythematosus. Accumulating evidence suggests that the disease has several different clinical, immunological, and genetic forms (Feltkamp et al, 1974; Fritze et al, 1974).

**Motor neuron disease (progressive bulbar palsy)**

This is a disease of middle age or later in which degeneration affects motor neurons in the anterior horns of the spinal cord, certain somatic motor nuclei of cranial nerves, and in the cerebral cortex. Both upper and lower motor neurons are affected and symptomatology
is diverse. The cause is, as yet, unknown. Pathological fasciculation can occur in any situation where some motor neurons degenerate and others persist, but it is most common in motor neuron disease.

The most common presentation is wasting of the muscles of the hand and upper girdle, but another common presentation, of interest to the otolaryngologist, is progressive bulbar palsy. This is more common in women causing dysphagia and dysarthria. It is often combined with upper motor neuron signs and evidence of pseudobulbar palsy. If weakness begins with bulbar palsy it usually spreads to the shoulders and arms.

The classical sign of the bulbar presentation is fasciculation and wasting of the tongue as a result of degeneration of the hypoglossal nuclei. This sign is unreliable in a protruded tongue, and it must be elicited with the tongue on the floor of the mouth. Differential diagnosis from myasthenia gravis is rarely difficult.

Dysphagia with aspiration is the most dangerous symptom and the course of the disease is more rapid if this is the initial symptom.

No treatment has any effect on the course of this relentless and depressing disease. Cricopharyngeal myotomy has its proponents, but does not always help the dysphagia and any improvement is usually short lived. The knowledge of a fatal creeping paralysis can lead to a complete collapse of morale, and requires careful handling.

**Palatal myoclonus**

In this condition there are rhythmical movements of the soft palate, occurring 60-180 times per minute (palate nystagmus). They develop insidiously, on one or both sides, and interfere with sleep, swallowing and respiration. They persist during sleep and may be inhibited initially by voluntary control. It appears to be a disorder of the olivocerebellar modulatory projection on the rostral brainstem and is seen in multiple sclerosis and brainstem infarction.

It may occur in association with myoclonus of the pharynx, larynx, eyes and diaphragm. The palatal myoclonus may be audible as a clicking sound and is sometimes abolished by anticonvulsants.

**Laryngeal neurological lesions**

**Functional disorders**

Many patients of a type similar to those with globus pharyngeus present with disorders of laryngeal sensation or voice production. These are related to social upsets, anxiety, smoking and vocal misuse. Following exclusion of organic disease local measures, reassurance, and advice from a speech therapist are usually successful.
'Spastic' larynx

Paralysis of the larynx may be caused by peripheral or central damage. As mentioned previously the latter is rare at a cortical level and the lesions are 'spastic'. This must not be confused with the so-called 'spastic' dysphonia which has been discussed at length in recent years. Spastic dysphonia is a rare bizarre dysphonia, possibly hysterical in origin, with overaction of the interarytenoid muscle against which the patient produces a markedly interrupted flow of speech. Improvement in some of these patients may be obtained by cutting the recurrent laryngeal nerve.

Brainstem paralysis of the larynx is, by contrast with cortical paralysis, flaccid.

Laryngeal spasm

This rather ill-defined condition is rarely neural and in young children is usually infective in nature. In the past, poor living conditions, rickets, whooping cough, upper respiratory tract sepsis, poliomyelitis, tetanus, and rabies have all been involved. Nowadays these are all rare in the UK.

Adults may suffer from 'choking' attacks with considerable panic and functional overlay. If these episodes are severe and there is concomitant vagal activity, so-called 'laryngeal vertigo' and a frank fainting episode may occur. These episodes may arise at night in individuals with marked oesophageal reflux and cricopharyngeal incompetence from any cause. It is presumed that a small amount of stomach content is aspirated into the larynx and the patient awakes with a 'choking' attack. This is an unusual disorder and other commoner causes of nocturnal dyspnoea should be excluded.

Superior laryngeal nerve palsy

Paralysis of this nerve whether on one or both sides is often clinically unrecognized and it has been somewhat neglected in laryngeal studies.

In unilateral superior laryngeal nerve paralysis, the voice is not severely affected and compensation occurs quickly. The disability to a professional voice user, in particular a singer, may, however, be significant. This form of paralysis is more readily recognized on examination because of the asymmetric tilt and shift of the larynx and the ipsilateral, slowly bowed, and flabby cord. Deprived of one of its tensors the affected cord also appears longer. Arynoid movements are unimpaired. The voice sometimes fails to regain its former strength even though its quality returns.

Bilateral superior laryngeal nerve palsy is even more rarely recognized. The symmetry of the larynx at rest and during phonation makes the presence of this combined paralysis difficult to detect. The absence of anterior tilt allows the epiglottis to hang more over the endolarynx and the slightly flaccid, bowed and hyperaemic cords are more difficult to see. The bowed cords allow excess leakage of air during phonation and the voice is lower, weaker, breathy and lacks inflection. With good compensation the speaking voice returns to normal but the singing voice is severely compromised.
The detection of both these paralyses has been aided by modern fibreoptic laryngeal endoscopes, stroboscopy, and video recording (Howard and Lund, 1986).

Superior laryngeal nerve palsy occurs in a significant number of patients undergoing thyroid surgery and a detailed knowledge of the variable anatomy of this nerve is essential. Dissection and division of the superior pole vessels immediately adjacent to the capsule of the upper pole of the thyroid gland will lessen the chances of damage. Trauma to the neck, particularly from road traffic accidents, can cause superior laryngeal nerve palsy.

**Recurrent laryngeal nerve palsy**

The eventual static position and appearance of a paralysed vocal cord will depend on the degree and permanency of denervation and the degree of associated muscle atrophy and fibrosis. Semon (1881) and his contemporaries, and otolaryngologists up to this day have failed to recognize these variables, and produced an unnecessary amount of disagreement and controversy. Indeed accurate recording of the position of a paralysed vocal cord is difficult because only 3-6 mm separate the median, paramedian and so-called 'cadaveric' positions. Therefore, many cases are reported inaccurately and this may help to explain the apparent inconsistencies between the various theories.

The 'cadaveric' position does not always occur after death and is best replaced by the term 'lateral'. By far the most satisfactory method of describing the position of the paralysed cord is to state clearly, in millimetres, the distance it lies from the median. If detailed follow-up and evaluation is to be undertaken for research and publication, then endoscopic photographic documentation should be used to ensure accuracy of long-term measurements.

It is also best to describe the paralysis, whether unilateral or bilateral, in terms of abductor or adductor paralysis, that is the direction of movement which the cord cannot make.

**Semon's law**

Semon's law is of historical interest only. The weight of neurohistological and neurophysiological evidence against it is now overwhelming, and it plays no part in modern laryngology. Briefly, Semon proposed that the motor fibres innervating the adductor and abductor muscles lay in separate bundles in the recurrent laryngeal nerve and that they had difference susceptibilities to an advancing lesion of the nerve, giving rise first to an abductor paralysis with the cord in the median position, and then adductor paralysis so that the cord came to rest in the lateral (cadaveric) position. There are many clinical inconsistencies to this, in addition to neurohistological and neurophysiological evidence that this is not the case. The reader is referred to the excellent account by Wyke and Kirschner (1976) for further study.

**Wagner and Grossman theory**

Wagner (1890) and Grossman (1897) proposed a theory which has had more recent experimental confirmation by Arnold (1962) and Dedo (1970). In the absence of cricoarytenoid joint fixation, an immobile cord in the paramedian position has a total pure unilateral recurrent nerve paralysis, and an immobile vocal cord in the lateral (cadaveric)
position has a combined paralysis of superior and recurrent nerves (the adductive action of cricothyroid is lost).

However, clinically it is not uncommon to see patients with intrathoracic lesions (which should produce a pure recurrent palsy) with the paralysed cord in the lateral (cadaveric) position. Purported explanations for this are stretching of the nerve by the intrathoracic lesion thus pulling the vagus down from the skull base and injuring the superior laryngeal nerve; and possible retrograde atrophy of the vagus to the nucleus ambiguus. Damage to the laryngeal nerves produces loss of sensation as well as motility. This may produce an incompetent laryngeal sphincter and possible life-threatening aspiration.

No doubt controversy with regard to the above considerations will continue but, from a practical point of view, the management of a patient with a vocal cord palsy depends on the aetiology of the lesion and on the defect it causes. In the case of a unilateral cord palsy the patient may obtain excellent vocal compensation without treatment, only the timbre of the voice being altered. 'Idiopathic' defects may recover up to 3 years after onset and it is necessary to wait a minimum of 6 months to see if recovery occurs in a paralysed cord before undertaking any definitive treatment. It is also wise to regard the diagnosis as 'unknown' rather than idiopathic for at least 18 months after onset.

Aetiology

Stell and Maran (1978) reviewed five large series of paralysis of the recurrent laryngeal nerve in the literature. The following groups were discernible.

**Malignant disease**

This accounted for 25% of cases, one-half being caused by carcinoma of the lung.

**Surgical trauma**

This was responsible for 20% of cases with surgical procedures on the lung, heart, oesophagus and mediastinum now outnumbering those produced by thyroidectomy.

**Idiopathic**

In 13% no cause could be found although virus infections such as influenza and infectious mononucleosis have been suggested as aetiological agents. In this idiopathic group all patients who are smokers should be considered to have carcinoma of the lung until proved otherwise.

**Inflammatory**

Another group accounting for 13% of cases with pulmonary tuberculosis still being the major cause.
Non-surgical trauma

In 11% of cases the cause was stretching of the nerve by enlargement of the left atrium, aortic aneurysm, or neck trauma.

Neurological

A proven neurological cause was found in 7%, that is cerebrovascular disease, Parkinson's disease, multiple sclerosis, head injury, alcoholic and diabetic neuropathies.

Miscellaneous

The remaining 11% of cases did not fit neatly into any of the above categories and they included a wide range of illnesses such as rheumatoid arthritis, haemolytic anaemia, syphilis, and collagen diseases.

Owing to its long course the left recurrent laryngeal nerve is affected in approximately three-quarters of the cases and the right in about 15%, the remainder being bilateral.

Symptoms and signs

Evaluation begins with listening to the patient's voice as he gives the history. A faint whisper suggests a functional adductor paralysis, a forced whisper an organic adductor paralysis. A voice which tires with use suggests a unilateral abductor paralysis and stridor and aspiration occur with a bilateral abductor paralysis. The patient's age and occupation are of paramount importance.

Additional symptoms in the upper respiratory and gastrointestinal tracts, and the rest of the head and neck are obviously of relevance and the patient's past medical history and smoking habits are of particular importance.

Investigations

Haematological tests

A full blood count, erythrocyte sedimentation rate and serology are useful screening tests, but other indices such as viral studies and blood glucose tests are rarely useful.

Radiology

Plain X-rays and tomograms of the chest, and a good submentovertical skull base view yield the best results. Further plain X-rays of the nasopharynx, neck and petrous bones are occasionally useful. A barium swallow and thyroid scan may be required if the cause remains undetected. Computerized tomographic scanning is becoming increasingly available but is rarely indicated in these investigations.
**Endoscopy**

Panendoscopy with rigid and flexible instruments may be used to elucidate or confirm the diagnosis and the causative lesions. Particular points of note are the need for biopsy of the fossa of Rosenmüller and the bronchial carina even if these appear normal. This is particularly necessary in those patients whose history and radiology have not suggested a cause. The affected arytenoids must be palpated to distinguish vocal cord paralysis from cricoarytenoid fixation.

**Treatment**

Evaluation of these patients will show them to belong to one of the four following groups. The treatment may vary even within the same group depending on the general state of the patient and the aetiology of the lesion. Many patients will be helped by speech therapy, with or without associated surgery.

*Unilateral abductor paralysis*

This is an important group to consider first because speech therapy may be the only treatment necessary. The single palsied cord lies in the paramedian position and any initial hoarseness may well disappear as the unaffected cord compensates. When the left cord is involved, the aetiology is commonly that of carcinoma of the lung and presentation with the vocal cord palsy means that the carcinoma is already inoperable.

If the dysphonia persists and is distressing to the patient then treatment is by Teflon injection. The voice, although reasonable, tends to tire with repeated use, and professional voice users are particularly likely to request treatment. Teflon injection is most likely to produce a good result when the cord is in the paramedian position but the amount used is critical and the technique is more fully described later. The post-injection voice is rarely entirely 'normal', although the volume and quality are improved. It is important that the patient with a paralysed cord is advised before surgery that, although there is a good chance of improving the voice, it will not necessarily return to its previous 'normal' level.

*Unilateral adductor paralysis*

The flaccid palsied cord lies in the lateral position and gives rise to a weak husky voice sometimes no more than a whisper. In addition, as this lesion is most commonly the result of damage to the vagus or both superior and recurrent laryngeal nerves, the laryngeal sphincter is incompetent, part of the larynx insensitive, and consequently aspiration may occur.

Both the type and timing of treatment depend on the aetiology of the unilateral adductor paralysis. If the cause is a carcinoma, particularly of the bronchus, then the patients appreciate the improvement of voice and ability to cough that can be obtained with a prompt Teflon injection. Many of these patients with carcinoma will have only a few months to live and delay to await any possible laryngeal compensation is unwarranted. There is also increasing support for immediate injection of palsied cords resulting from major thoracic operations, thus enabling the patient to cough satisfactorily during the postoperative period.
Teflon injection for an adductor paralysis gives overall poorer results than for abductor lesions. This is a result of the difficulty of closing the posterior part of the glottis between the arytenoids where there will always be some air wastage. Overspill may be helped by injection but the results are more difficult to predict and require experience and care on the part of the surgeon.

Unilateral adductor paralysis not caused by carcinoma requires a waiting period of at least 6 months to allow for compensation or evidence of recovery. Speech therapy should be given during this time. If the unilateral adductor paralysis is a result of laryngeal trauma there is usually considerable scarring, particularly of the thyroarytenoid muscle and it may not be possible to displace the cord medially by means of a Teflon injection. In order to alter the position of the scarred cord from lateral to medial a number of alternative procedures have been advocated. The best results are obtained using a laryngofissure approach and moving the paralysed cord medially by inserting either muscle or cartilage between it and the adjacent thyroid lamina. To achieve a similar result, needles of nasal septal cartilage may also be inserted endoscopically using microlaryngoscopic techniques. This latter procedure is technically more difficult but avoids the complications occasionally associated with laryngofissure in an already scarred larynx. A reversed type cordopexy has been used fixing the cord in the midline via an external approach.

Bilateral abductor paralysis

This lesion is usually the result of damage to both recurrent laryngeal nerves at thyroidectomy. Other treatable causes are rare. The cords lie in the paramedian position and the voice is good but the degree of stridor is very variable. In the acute situation stridor may be life threatening and a tracheostomy required. However, if the lesion develops more slowly and the patient is relatively inactive there may be little or no stridor. Nevertheless, at some point, usually associated with an upper respiratory tract infection, all these patients will develop stridor. If the lesion is diagnosed soon after thyroidectomy immediate re-exploration of the neck is indicated. This is not so much to re-anastomose sectioned nerves (which gives overall poor results), but more in the hope of finding the nerves caught in a ligature. Removal of this often allows good long-term recovery. The author advocates a positive approach to post-thyroidectomy paralysis, but re-exploration of the nerves is pointless after delay of more than 6 months because of motor end-plate degeneration and fibrosis of laryngeal muscles.

The choice of treatment for patients with established bilateral abductor paralysis is wide and numerous operations have been described. No operation should be attempted until at least 6 months after the onset of paralysis, thus allowing for any possibility of spontaneous recovery. If there is recovery of any movement in one cord then possible operations should be undertaken only on the other cord.

When considering treatment it is important to remember the basic point that the patient has a good voice but poor airway. Any operative procedure on the cord to improve the airway will decrease the quality of voice and on occasions fail to improve the airway. Many of these patients will require or present to the laryngologist with a permanent tracheostomy. If this is fitted with a speaking valve they have the excellent situation of a good airway and a good voice. The only disadvantage to this being the actual wearing of the tube which some patients are unable to accept. A permanent speaking valve tracheostomy is usually the best choice in
the professional voice user. It is the author's experience that patients who have become used to wearing such a tube will decline further surgical intervention and they should not be persuaded into undergoing any of the following numerous procedures.

**Arytenoidectomy**

This procedure was first used by veterinary surgeons in the 19th century on race horses with unilateral cord palsies. Ivanoff (1913) undertook the first human arytenoidectomy. King (1939) described an extralaryngeal approach to mobilize the arytenoid and affected cord laterally and attach it to a severed omohyoid muscle and the thyroid ala. Woodman (1946) modified this procedure by excising the arytenoid and suturing the residual vocal process to the inferior cornua of the thyroid cartilage. This latter operation became popular during the following 30 years but is associated with variable results of voice and airway and complications of infection and stenosis.

Arytenoidectomy can also be accomplished via a laryngofissure or lateral thyrotomy approach, combined with lateral cord mobilization and fixation. The thyrotomy approach allows good access to the arytenoid and a submucosal dissection.

Endolaryngeal arytenoidectomy was first described by Thornell in 1949 and, like the other procedures, gives variable results. It could be a difficult operation to perform with microsurgical instruments but can now be carried out bloodlessly and precisely using the CO₂ laser. It may be combined with partial or complete cordectomy using the laser and is now the operation of choice in the author's unit.

**Cordectomy**

Since the early part of this century various open and endoscopic methods of cordectomy have been advocated but variable results and, in particular, granulation formation have made surgeons wary of the procedure. With the advent of the CO₂ laser the endoscopic operation is precise and quick to perform and may be combined with arytenoidectomy. Removal of the posterior half of one vocal cord and the arytenoid gives a good compromise of voice and airway. The operation is completed in a haemostatic field and is associated with minimal postoperative oedema and granulation. The in-patient stay is very short and more tissue can easily be removed at a second endoscopy if necessary. The operation can be undertaken in those patients who have a compromised airway but who have so far managed without a tracheostomy.

**Reinnervation procedures**

In 1927 Colledge and Ballance demonstrated in monkeys, baboons and a single human case that bilateral recurrent nerve damage could be repaired by anastomosis of the phrenic nerve to the recurrent laryngeal nerve. This resulted in abduction of the cords in quiet respiration with an adequate airway. Crumley (1982) has demonstrated split phrenic nerve to recurrent nerve anastomosis giving respiratory abduction of the vocal cord while preserving diaphragmatic motion. This procedure deserves further evaluation but the nerve-muscle pedicle technique first reported in humans by Tucker in 1976 has not produced good results in other surgeon's hands. This latter technique involves transferring a portion of sternohyoid
muscle with its nerve supply from the ansa cervicalis into the posterior cricoarytenoid muscle to try to produce reinnervation and movement.

**Bilateral adductor paralysis**

Apparent bilateral adductor paralysis may occur in patients with psychiatric disturbances. There may be a severe dysphonia but the commonly used term 'hysterical aphonya' is often inappropriate. The milder cases may be considerably helped by speech therapy particularly if the underlying problems (usually social) can be resolved. A few patients may need psychiatric referral and recurrence of symptoms is not uncommon.

Organic disease producing this bilateral lesion is fortunately rare but is usually a serious central nervous system disease or neoplastic process involving the medulla, skull base or upper neck. With both cords in the lateral position these patients are not only aphonie but are unable to cough well and have incoordinated swallowing. This laryngeal incompetence results in life-threatening aspiration in a very short time. Initial management involves a tracheostomy with a cuffed tube and a nasogastric tube for feeding. Further management is usually required depending on the aetiology. Total laryngectomy is the only sure way to protect the lungs but may not be undertaken if there is a possibility of neurological improvement. Epiglottopexy (Brookes and McKelvie, 1983) is the procedure of choice in neurological problems where subsequent neurological improvement may occur.

Other methods of management have been advocated such as Teflon injection of the cords and supraglottis, closure of the glottis by suturing the cords, and cricopharyngeal myotomy. The results of these other methods are variable and often poor and the author does not recommend them in a complete bilateral adductor palsy.

**Teflon injection in vocal cord paralysis**

The injection of Teflon paste into the paralysed vocal cord is now firmly established as an effective treatment with minimal complications. Indications for its use and the correct timing of surgery have already been considered in the previous section on vocal cord palsy.

In 1911, Brünings successfully treated patients by injecting paraffin into the laterally placed vocal cord. A modification of the injection syringe he described is still the instrument of choice today. Subsequent to his description, injected paraffin in other parts of the body was found to cause embolization and paraffinoma and its use was abandoned. Over 40 years later Arnold (1955) revived the technique using cartilage particles as the injection material. Following this a variety of substances such as bone paste, silicone, tantalum oxide powder, tantalum and Teflon were evaluated. Teflon paste emerged as the material of choice. Its permanence, low tissue reactivity and lack of carcinogenicity in over 20 years of use are well documented. The reader is referred to the excellent review of the characteristics of Teflon paste by Montgomery (1979). After injection the Teflon particles are walled off by foreign body reaction and the suspending glycerol is slowly absorbed. Initial accompanying oedema abates and an immediate good postoperative voice may deteriorate a little. A second injection may be necessary and even with experienced surgeons approximately 20% of patients will require this. A firm rubbery mass persists at the injection site as the paste migrates minimally, but it does not appear in lymph nodes and appears to be almost totally inert.
Several techniques have been used to evaluate the functional results of the injection such as aerodynamic studies, high speed photography of the vocal cords, video recordings of the vocal cords, voice spectrographs and phonation times. However, the simplest and most useful measure is simply to record the talking and singing voice of the patient pre- and postoperatively. The injection should produce a clearer, stronger voice (and decrease aspiration if present) without significant impairment of the patient’s airway.

Method

The injection can be performed under local or general anaesthesia and each method has its advantages and disadvantages together with surgeons who strongly advocate each method. The final choice will depend on the patient, his or her overall condition, and the local methods of practice and facilities. It is preferable for the surgeons to be familiar with both techniques.

Dedo, Urrea and Lawson (1973) gave an excellent account of the method used under local anaesthesia with results in 135 patients. This technique is used with the patient in a sitting position with the larynx and pharynx anaesthetized by sprays and droplets of cocaine. The surgeon views the larynx by indirect laryngoscopy and injects Teflon from a Brünings syringe into the paralysed vocal cord. Following withdrawal of the needle the patient can be asked to phonate and the quality of the voice checked. Further injection can be undertaken if necessary. This method undoubtedly gives good results but is not tolerated well by all patients and, indeed, is refused by some. The ability to assess the right amount of Teflon paste should not be too highly regarded since oedema of the surrounding cord occurs rapidly within 2-3 minutes and unless the injections can be completed in this time the true depot size is unpredictable. Subsequent resolution and glycerol absorption also occur in postoperative weeks.

The method is of course most useful in patients whose overall condition contraindicates general anaesthesia. Recently Ward, Hanson and Abemayor (1985) described a transcutaneous technique, injecting the cord via a needle placed through the anterior neck into the larynx, in a patient with severe trismus. The result was checked using a flexible fibreoptic telescope passed into the pharynx and attached to a videocamera.

The major criticism of the technique under general anaesthesia has been the likelihood of over-injection. However, this rarely occurs when the basic requirements are observed. These are that the other cord should be fully functional and the injected cord should not be pushed right up to the midline. The author's preference is for the following technique. Under general anaesthesia with fine catheter intubation or Venturi ventilation a Dedo laryngoscope is placed with accompanying Loewy suspension. A Brünings syringe is loaded with Polytet Teflon paste and care taken to ensure that the paste has filled the attached needle and passes easily. The first injection is made in the posterolateral corner of the middle third of the vocal cord just anterior to the vocal process. With each click of the syringe rachet 0.1 mL of paste is deposited and usually 0.3 to 0.4 mL is required to push the cord medially to lie close to the midline. It is most important to realize that paste is compressible and is not all delivered immediately the handle is squeezed. Therefore several seconds should be allowed to elapse following each click so that all the paste is extruded and the cord accurately assessed. The injection should cause a gentle fusiform enlargement and care should be taken to avoid
placing the paste in Reinke's space, too far laterally into the ventricular floor, or too deeply into the subglottis. A second injection is usually required in the anterolateral corner of the middle third of the vocal cord, that is the midpoint of the membranous cord. Because of the firm adherence of the mucous membrane to the vocal process it is impossible to inject posteriorly to close any gap between the arytenoids.

An intramuscular injection of dexamethasone 8 mg is given at the time of the procedure to minimize oedema but routine antibiotics are unnecessary. The majority of these operations can be carried out on a day-case basis, but a postoperative overnight stay may be indicated by the general condition of the patient.

**Complications**

The morbidity from this procedure is small but in all large series there is an incidence of about one in 100 cases requiring tracheostomy. The patient should be warned of this possibility preoperatively. Tracheostomy may be required because of acute progressive oedema which usually responds promptly to steroids, antibiotics and humidification allowing subsequent decannulation after only a few days. Less often a hemilaryngitis will occur 7-10 days postoperatively and requires antibiotic treatment.

Over-injection or injection into the subglottis both produce airway impairment but should be rare if care is taken. The author has successfully treated this complication by removal of the injected Teflon and any necessary additional tissue with a CO₂ laser.

The minimal soreness and foreign body sensation experienced by many patients subsides in a few days and an improvement in the voice should be obtained in over 90% of patients. As stated previously, the best results are in those with a pure unilateral recurrent nerve lesions.