Chapter 23: Stertor, sleep apnoea and velopharyngeal insufficiency

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Stertor

Definition

Stertor may be defined as noisy respiration caused by partial airway obstruction above the level of the larynx. It is a low-pitched, snoring or snuffly sound, in contrast to the higher pitched musical sound of stridor which characterizes partial airway obstruction at or below the level of the larynx. The noise is produced by turbulent airflow through the narrowed airway, setting up vibrations in the surrounding tissues.

Pathophysiology

It is helpful to consider the physics of airflow through a tube in order to understand better the way in which the noise of stertor is produced (Cotton and Reilly, 1983).

The pressure exerted by a gas is equal in all directions, except when it is moving. Linear movement of gas through a tube creates additional pressure in the forward direction and, because of the principle of conservation of energy, there is a corresponding fall in pressure laterally against the walls of the tube. The faster the gas flows, the greater the drop in lateral pressure. This phenomenon is termed the Venturi effect.

When air flows rapidly through a passage that is narrowed, the lateral pressure that is holding the lumen open can drop drastically in accordance with the Venturi effect and cause the walls to collapse and the lumen to close momentarily. Closure of the lumen obstructs the flow of air, the intraluminal pressure returns to normal, and the walls spring open again. This cycle rapidly repeats itself, producing a pattern of intermittent flow with associated turbulence and vibration of the lumen wall. This becomes audible as a noise.

Airway narrowing above the level of the larynx produces the low-pitched sound of stertor. The tissues of the tongue and pharynx are relatively loosely supported and can be quite easily drawn into the airway on inspiration. Inspiration is initiated by movement of the thoracic cage and the diaphragm, producing expansion of the lung parenchyma and bronchi, and drawing in air through the upper air passages, larynx and trachea. Narrowing of the airway results in vigorous inspiratory efforts involving the accessory muscles of respiration, in an attempt to draw in a sufficient volume of air. In order to move an adequate volume of air for the child's respiratory needs, high flow rates through the narrowed segment of airway develop with a consequently powerful Venturi effect, thus constricting the airway further.

Thus stertor is an inspiratory noise. Expiration has the opposite effect on the supralaryngeal regions, blowing the airway open, usually with no associated sound.

The high negative pleural pressures which develop during inspiration against an obstructed airway produce marked indrawing of the soft tissues in the subcostal, intercostal, suprasternal and supraclavicular regions. In the suprasternal notch this retraction or recession
produces the appearance of the trachea plunging down into the chest with each indrawn breath, the so-called tracheal tug.

The respiratory obstruction which produces stertor can lead to hypoxia, hypercapnia, pulmonary oedema, cor pulmonale, vomiting, aspiration pneumonia and occasionally death. Long-term effects include failure to thrive and permanent brain damage (Heaf et al, 1982).

Aetiology

Any condition which restricts the airway in the nose, nasopharynx or oropharynx is liable to produce stertor.

Congenital conditions

Nasal obstruction

The neonate remains an obligate nose breather for the first 2 or 3 months of life (Stool and Houlihan, 1977), and so nasal obstruction in this age-group is an especially serious problem. Total nasal obstruction, for example caused by complete nasal agenesis or bilateral choanal atresia, is immediately life-threatening. Partial nasal obstruction with stertor produces a less dramatic, but nevertheless serious, clinical picture. Feeding becomes slow and difficult, with frequent interruptions as the infant 'comes up for air', and inspissated nasal secretions may produce near-total nasal obstruction with consequent risk of sudden infant death.

Congenital abnormalities of the external nose are very uncommon, but various degrees of nasal dysgenesis can occur and may produce nasal obstruction: these include nasal hypoplasia (sometimes secondary to one of the mandibulofacial dysostoses), varying degrees of congenital occlusion of the anterior nares, and the group of median facial anomalies classified by De Myer, Zeman and Palmer (1963).

Congenital cysts of the nasal cavity may produce obstruction and stertor, depending upon size and location. They include dermoids, nasoalveolar cysts, dentigerous and mucous cysts of the floor of the nose and Jacobson's organ.

Nasal obstruction in the neonatal period may be caused by a swelling of neural origin extending into the nose. This may be a meningocoele (meninges alone), meningoencephalocele (meninges plus brain tissue) or encephalocele (glial tissue with no remaining connection to the brain) (Furstenberg, 1936; Proctor and Proctor, 1979).

Choanal stenosis may produce nasal obstruction with stertorous respiration. In part this is a result of the choanal narrowing itself, and in part caused by secondary stasis of nasal mucus.

Facial skeletal anomalies

Children with craniofacial abnormalities may develop stertor as a result of obstruction in the nose, nasopharynx or oropharynx.
Nasal obstruction occurs, but is seldom severe, in children with unilateral cleft lip and palate. It is caused by a combination of collapse of the ipsilateral alar cartilage and deviation of the nasal septum: the septum is deviated to the contralateral side anteriorly, and to the ipsilateral side posteriorly where it articulates with the malpositioned vomer.

Nasopharyngeal obstruction occurs in Apert's and Crouzon's syndromes as a result of severe posterior displacement of the mid-facial structures and consequent narrowing of the nasopharyngeal airway.

Oropharyngeal obstruction is a feature of glossoptosis, in which there is micrognathia and, as a result, the tongue falls back into the oropharynx. Treacher Collins syndrome is an example of micrognathia in which this may occur. However, the tongue seems to fall back into an obstructing position more readily when there is an associated cleft palate, and the problem is frequently severe in children with the Pierre Robin syndrome where there is both micrognathia and a cleft palate.

**Macroglossia**

The tongue can produce respiratory obstruction if it is much enlarged, even if the mandible is normal. This can occur in Down's syndrome and as part of the organomegaly of Beckwith's syndrome, or it may be isolated and idiopathic.

Enlargement of the tongue can also occur when it becomes involved in a cystic hygroma or haemangioma of the floor of the mouth, although fortunately it is unusual for the swelling to be so gross that respiratory embarrassment ensues.

**Pharyngeal swellings**

A large lingual thyroid mass can produce respiratory obstruction and stertor, as may a lingual thyroglossal duct cyst, or any of the rare congenital tumours of the pharynx. The latter include Tornwaldt's bursa, branchial cleft cysts, teratomata, 'hairy polyps', chordomata, craniopharyngiomata, cystic hygromata and haemangiomata (Parkin and Thomas, 1974).

**Acquired conditions**

**Traumatic**

A septal haematoma may follow birth trauma, accidental injury or assault, or may develop postoperatively following a septoplasty: the ensuing nasal obstruction may be partial or complete. A septal abscess (often an infected septal haematoma) will be more obvious as a cause of stertor because of the associated pain. Facial fractures often compromise the upper airway.

**Inflammatory**

The most frequent cause of stertor in children is the common cold. Neonates sometimes become 'snuffly' to the point of significant respiratory difficulty and associated
feeding problems, but older children generally just mouth-breathe and only snore at night. Sinusitis and nasopharyngitis ('adenoiditis') can produce the same pattern in older children.

Tonsillitis rarely causes respiratory embarrassment, except in children with hugely hypertrophic tonsils. Infectious mononucleosis, however, can produce life-threatening airway obstruction as a result of massive inflammatory swelling of all the pharyngeal lymphoid tissue including the tonsils and adenoids, with grossly stertorous respiration.

It is unusual for a peritonsillar abscess to produce a significant degree of respiratory obstruction, but a parapharyngeal or retropharyngeal abscess may so do. Ludwig's angina is generally accompanied by stertorous, difficult breathing owing to the inflammatory swelling in the floor of the mouth, and there is a danger of laryngeal oedema developing with rapidly fatal consequences.

Various types of rhinitis may produce nasal obstruction with mouth breathing, and snoring during sleep, caused by swelling of the nasal mucosa and over-production of mucus. Chronic infective rhinitis ('chronic catarrhal rhinitis') is characterized by a persistent mucopurulent nasal discharge: commonest in the lower socio-economic groups, it may persist until puberty and usually resists all attempts at treatment. Perennial allergic rhinitis in children usually occurs in atopic individuals who also have asthma, and this can compound their respiratory difficulties. Rhinitis medicamentosa, caused by over-use of topical nasal decongestant drops and sprays, is fortunately rare in childhood. Other specific forms of rhinitis are extremely rare, for example diphtheric rhinitis, tuberculosis, rhinoscleroma and leprosy.

**Neoplastic**

The neoplasm that classically produces nasal obstruction and nocturnal stertor in older children and adolescents is the postnasal angiofibroma, which may enlarge to fill the nasopharynx completely. Other tumours may less commonly produce similar symptoms, such as a neuroblastoma, lymphoma or rhabdomyosarcoma.

**Other conditions**

Hypertrophic adenoids are a common cause of nasal obstruction and stertor, and an important cause of the obstructive sleep apnoea syndrome (see below), especially when there are other factors compromising the upper airway such as the cramped nasopharynx of some of the craniofacial anomalies, or the micrognathia of Treacher Collins syndrome.

Nasal polyps in children are usually associated with cystic fibrosis, and there is often a concomitant chronic sinusitis; chronic mucosal swelling and the abnormally thick, tenacious mucus aggravate the obstruction (Baker and Smith, 1970). An antrochoanal polyp may produce stertor during sleep if it is large enough to fill both sides of the nasopharynx.

An important entity in this group of miscellaneous causes of stertor is idiopathic nasal mucosal swelling in the neonate. Many infants have slightly congested nasal mucosa at birth and are 'snuffy', but a very few have severe mucosal swelling with no apparent cause and become stertorous: sometimes the obstruction is so severe that total nasal obstruction ensues
and the infant presents the same picture as a case of bilateral choanal atresia (Mugliston and Mitchell, 1984).

**Iatrogenic**

Nasal obstruction and snoring may result from stenosis of the nasopharyngeal isthmus. This is invariably acquired as a result of surgery: sometimes tonsillectomy and/or adenoidectomy can be responsible, but more usually it is the result of a 'too successful' pharyngoplasty performed for velopharyngeal insufficiency. Nasal and postnasal packing can sometimes cause dramatic stertor and seriously compromise the airway.

**Clinical features**

**Symptoms**

The first priority in assessing a child with stertor is to establish whether there is severe respiratory distress requiring urgent action to secure the airway. If there is no immediate threat to the airway, a careful and systematic history should be obtained.

The nature of the noise should be determined first, as parents, nurses (and indeed other physicians) are often extremely vague on this essential point which gives such a useful indication of the site of obstruction. Having established that the noise is stertor rather than stridor, it will invariably become apparent that it is present on inspiration.

Next, the time of onset should be inquired about. Most of the congenital causes of stertor will present with breathing difficulty from birth, and the obstruction will be immediate and dramatic in those neonates with nasal obstruction owing to their obligatory pattern of nasal respiration. A history of preceding trauma or upper respiratory tract infection may be obtained in the older child who rapidly becomes stertorous, indicating respectively the likelihood of faciomaxillary fractures or an inflammatory condition as the cause. Slowly progressive, increasing stertor suggests a neoplastic cause. The older child who mouth breathes during the day and snores at night may have obstructive sleep apnoea, and specific enquiry should be made about sleep patterns.

In the child with long-standing stertor, aggravating and relieving factors should be sought, especially whether the noise is loudest when asleep or awake, and whether it is related to any particular position.

Direct questioning about associated symptoms should include enquiry about the cry or voice. In supralaryngeal obstruction there is no hoarseness, but the cry is characteristically muffled. There is usually no cough. Feeding difficulties are obvious in the infant with nasal obstruction, who will have to come off the nipple frequently to breathe: sometimes this slow, interrupted feeding pattern is so severe that failure to thrive and gain weight becomes apparent. Other children in whom there are no feeding difficulties may fail to thrive simply because of the increased metabolic demand of breathing against resistance (Heaf et al, 1982).
Signs

The primary objective in the physical examination is to assess the severity of respiratory distress. Decreased activity and responsiveness, pallor, sweating, tachypnoea and recession all indicate severe respiratory distress; cyanosis, bradycardia and hypotension are grave signs that herald respiratory arrest and demand immediate action to restore the airway. Such a child should not be disturbed for additional examination lest respiratory arrest is precipitated, but should instead be immediately transferred to the paediatric intensive care unit or operating theatre for intubation, endoscopic examination and possible tracheostomy. The team involved must include a paediatrician, paediatric anaesthetist and otolaryngologist.

The stable, well-oxygenated child can be examined further in a thorough and systematic way. Attention should first be directed towards assessing in more detail the stertorous noise and establishing whether it is present in the inspiratory or expiratory phases of respiration, or both. An attempt should be made to determine whether the noise is arising from the nose or pharynx by listening at the nose and open mouth, and the chest must be auscultated to ensure there is no associated pulmonary pathology. The child should be placed in various positions to establish whether posture affects the noise: for example, in the Pierre Robin syndrome and other forms of micrognathia or macroglossia the noise is markedly diminished by lying the baby prone with the head extended.

During this general part of the examination, care should be taken to look for any congenital abnormalities.

Next the nose should be examined. Assessment of airway patency can be difficult in the infant, but bubbling of mucus in the nasal cavity indicates that there is airflow, as does misting on a silvered surface held at the nares. Examination of the nasal cavities in the infant is best achieved by using an electric otoscope with a suitably-sized speculum, with the child wrapped in a blanket. Choanal patency is then assessed by ensuring that an 8FG catheter will pass through each side of the nose into the pharynx without difficulty. In the older child, the nose can be examined in the conventional way.

Examination of the throat must include an assessment of the size and position of the tongue and lower jaw. The palate must be checked for a cleft and the size of the tonsils judged. If possible, a view of the oropharynx should be obtained to see if there are any masses or swellings distorting the airway: this can be difficult in the infant, and use of the tongue depressor often induces gagging with simultaneous gastro-oesophageal reflux which instantly obscures the view and induces choking which further threatens the already compromised airway. Great care should therefore be taken, and it is best to avoid use of the tongue-depressor if the child has recently had a feed. If the airway seems precarious, and especially if there is the slightest suspicion of acute epiglottitis, no attempt should be made to examine the oropharynx in the clinic or ward: the examination should be delayed until the child is anaesthetized in the operating theatre ready for intubation with anaesthetist, otolaryngologist and pediatrician standing by.
**Investigations**

**Physiological investigations**

The usual regular observations of temperature, pulse, respiratory rate and blood pressure must be charted. The resting respiratory rate is a sensitive index of pulmonary function. Neonates have a rate of about 40 breaths/minute; this diminishes with age so that a 2-year-old child has a rate of about 20/minute, and a 6-year-old child 15/minute. With respiratory distress, tachypnoea is an early and important compensatory mechanism. Associated with this is a rise in pulse rate, bearing in mind that the normal heart rate for a neonate is about 140/minute decreasing to around 80/minute in older children. Bradycardia is a late sign that indicates terminal decompensation, and is accompanied by hypotension (although neonates often develop bradycardia as an immediate response to acute hypoxia).

The single most important laboratory test of respiratory efficiency in the critically ill child is measurement of arterial blood gas levels. The $P_{aO_2}$ indicates the level of oxygen being delivered to the tissues: this too changes with age, normally being about 9.3 kPa (70 mmHg) in the newborn rising to 11.97 kPa (90 mmHg) in the older child. Serious tissue hypoxia results when the $P_{aO_2}$ falls below 6.65 kPa (50 mmHg). The $P_{aCO_2}$ is the best indicator of alveolar ventilation, with a normal range of 3.99-4.66 kPa (30-35 mmHg) in the neonate and 4.66-5.99 kPa (35-45 mmHg) in the older child. However, it must be remembered that $P_{aCO_2}$ is inversely related to respiratory minute volume, and so if the alveolar minute volume doubles the $P_{aCO_2}$ is halved. The pH is a measure of respiratory and metabolic acidosis of alkalosis (range 7.30-7.45).

When serial measurements of arterial blood gases are needed, an indwelling arterial line is necessary. However, a transcutaneous oxygen sensor (ear lobe oximeter) can be invaluable for continuously monitoring tissue oxygenation. Similarly, continuous automatic monitoring of temperature, pulse, blood pressure and ECG can be undertaken if the child's condition is unstable, together with respiratory rate if the patient is intubated. In this situation an intravenous line is mandatory for administration of fluids and drugs.

**Radiological investigations**

In the child without respiratory distress, radiological evaluation of the upper airway is the next step.

In the assessment of stertor, the soft-tissue lateral X-ray of the neck is the most valuable view, and will demonstrate the size of the nasopharyngeal airway relative to the adenoids, and the size of the oropharyngeal airway relative to the tonsil and tongue position, as well as showing any masses distorting the upper airway. However, the film must be of the highest quality to be meaningful; for example a few degrees of rotation will produce misleading appearances by superimposing normal structures on the air shadow. The X-ray must be taken with the head slightly extended during inspiration; flexed expiratory views, especially if the child is crying, produce dramatic distortions of pharyngeal tissues which can bulge enormously in the infant and mimic the appearance of a retropharyngeal abscess or mass.
The high-kV 'Cincinnati' posteroanterior view of the mediastinum enhances the air column in trachea and bronchi by providing improved contrast between air and soft tissues. This view, together with a plain posterosuperior chest film, should be taken to exclude any unsuspected lower airway pathology.

A barium swallow, essential in the examination of a patient with stridor to look for a vascular ring, is not needed for the patient with stertor alone.

Occasionally, other special X-ray studies can be helpful. Xeroradiography shows the air-soft tissue density interface particularly well, and can sometimes by of value in difficult cases, but has the disadvantage of rather high radiation exposure. Hypocycloidal polytomography shows bone destruction well and is used when a neoplasm is suspected, for example a postnasal angiofibroma. For a vascular tumour such as this, angiography is employed in addition.

Computerized tomography (CT) and more recently magnetic resonance imaging (MR) show soft tissues well and can be complementary to polytomography in demonstrating the extent of tumours or other masses. Computerized tomography is especially valuable in the assessment of choanal atresia and stenosis.

**Endoscopy**

Sometimes endoscopic examination is needed to confirm the diagnosis. Under general anaesthesia the nose, oral cavity, nasopharynx, oropharynx and hypopharynx are examined.

An orotracheal tube is employed for the anaesthetic. The nasal cavities are inspected directly using a headlight, and with the aid of straight-ahead (0°) and angled (30° or 70°) Hopkins rod telescopes, or a small-bundle flexible fibreoptic nasoendoscope such as the 2.9 mm Olympus ENG 'P'. A Boyle-Davis gag is then inserted to permit viewing of the oral cavity and pharynx. Examination of the postnasal space may be accomplished with a mirror, or a 120° retrograde telescope passed behind the posterior edge of the soft palate.

In some cases it is difficult to decide whether the respiratory noise is stertor alone, or whether there is also an element of stridor indicating pathology in the larynx or tracheobronchial tree. If such doubt exists, then the endoscopic evaluation must be completed by performing a microlaryngoscopy and bronchoscopy.

At the end of the procedure the endotracheal tube should be removed to permit observation of the airway as the child awakens. This may reveal airway obstruction in the dynamic state (such as pharyngeal collapse that was not apparent when the tube was in place.

**Treatment**

**Medical and conservative measures**

Patients with respiratory distress should normally be nursed in a semi-sitting position, as in this posture the abdominal contents fall away from the diaphragm, allowing it to move
more efficiently and increasing the functional residual capacity of the lungs. However, in some conditions a different position will greatly improve the airway - for example, the prone position is best for children with the Pierre Robin syndrome.

If necessary, warmed, humidified oxygen should be administered. This is carried out using a blender system that will deliver oxygen in concentrations from 20 to 100%. The concentration is adjusted so that the $P_{aO_2}$ is maintained between 7.98 and 11.97 kPa (60 and 90 mmHg) (6.65-10.64 kPa (50-80 mmHg) in neonates to minimize the risk of retrolental fibroplasia). Delivery is best achieved using a head-box for infants and mist tent or face mask for older children.

Adequate hydration must be maintained with intravenous fluids or tube feeds if oral intake is inadequate. Physiotherapy, to help clear the chest of secretions, should be employed with the great caution in the child with a severely compromised upper airway lest complete obstruction be precipitated.

Immediate measures must be taken to improve the airway if it is compromised to the point of respiratory distress. The method employed depends upon the apparent site of obstruction.

Nasal obstruction does not present a great problem in the older child who will simply mouth breathe. However, the neonate, who is an obligate nose breather, will require an oral airway.

Pharyngeal obstruction is often dramatically relieved by employing a nasopharyngeal airway, and this has been well demonstrated in the Pierre Robin syndrome by Heaf et al (1982).

**Use of the nasopharyngeal airway ('nasal prong')**

A Portex endotracheal tube is employed; for a neonate a 3.0 or 3.5 mm diameter tube is appropriate, and the length required can be estimated from the crown/heel measurement (Heaf et al, 1982). The tube is mounted on a Tunstall connector, passed through the nose and secured with a conventional head band and tapes across the cheeks. Lateral neck radiography is necessary to check the tube position; ideally the tip should be just above the epiglottis. If the tube is too long, choking and vomiting are induced; if it is too short, relief of airway obstruction is not achieved.

To maintain tube patency, regular suction with a catheter is undertaken, preceded by instillation of 0.5 mL of isotonic saline; this is carried out routinely before feeds and at other times as required. The tube is changed every week and the new tube passed through the opposite nostril.

Occasionally vestibulitis develops around the tube, requiring topical antibiotic therapy. Crusting and blockage may sometimes occur and necessitate a tube change. Parents can be trained to manage the tube at home, with weekly visits to the hospital.
A nasopharyngeal airway may be the only treatment required for some stertorous children. Those with the Pierre Robin syndrome, for example, often grow to develop an adequate airway by the age of about 3 months, and use of the 'prong' can then be discontinued. In other children, the nasopharyngeal airway is an invaluable temporary measure, allowing full assessment of their clinical condition to be made prior to more definitive treatment (for example, adenotonsillectomy or tracheostomy).

In some children with severe pharyngeal airway obstruction - for example, caused by a craniofacial anomaly with pharyngeal collapse - a nasopharyngeal airway alone is insufficient, but the addition of continuous positive airway pressure overcomes the residual obstruction by distending the pharynx during inspiration. A ventilator is not required: continuous positive airway pressure may be applied via a special valve, or by underwater immersion of the exhalation arm of a T-piece system.

Endotracheal intubation

In some situations airway management with a nasal prong is inappropriate, and endotracheal intubation is necessary. This is the case with patients with faciomaxillary trauma where the tracheobronchial tree must be protected against inhalation of blood, and in children with massive neoplasms compressing the pharyngeal airway.

Surgery

Surgical treatment of the child with stertor depends upon the aetiology. Most of the conditions causing stertor are discussed in depth elsewhere in this volume, and their surgical management described.

In particular, the principles governing adenotonsillectomy in children with stertor are discussed below under the heading of Obstructive sleep apnoea. However, it is important to add that some children with stertor caused by anatomical restriction of the upper airway, for example some of the craniofacial anomalies, may benefit from tonsillectomy and adenoidectomy, even though hypertrophy of the tonsils and adenoids is not the primary cause of the problem. This is because the flow of air through a tube increases as a function of the fourth power of the radius of the tube in accordance with Poiseuille's formula: thus a small increase in radius is accompanied by a large increase in flow. It is therefore often beneficial to remove the tonsils and adenoids in such a situation, and the consequent improvement in respiratory function may avoid the need for tracheostomy.

The neonate with severe idiopathic nasal mucosal swelling can present difficulties in management. If there is associated rhinorrhea it is important to send nasal swabs for culture and sensitivity to exclude an infective rhinitis. It may be necessary to undertake an examination under anaesthesia in order to exclude absolutely a choanal stenosis. If the cause of the problem is unequivocally established as idiopathic mucosal swelling, topical treatment with nasal drops may be beneficial: 0.5% ephedrine nose drops can be used sparingly, but cause rebound congestion very quickly in infants and so betamethasone drops are more satisfactory, particularly as treatment may have to be continued for several weeks. Occasionally, nasal intubation is the only course. A short nasopharyngeal 'prong' may be used
as described above, but if prolonged intubation is contemplated it is better to insert a pair of Portex nasal tubes under general anaesthesia in the same way as for a case of choanal atresia.

Sleep apnoea

Definition

An apnoea has been defined as cessation of airflow at the level of the nostrils and mouth lasting at least 10 seconds. A sleep apnoea syndrome is diagnosed if, during 7 hours of nocturnal sleep, at least 30 apnoeic episodes are observed in both rapid eye movement (REM) and non-rapid eye movement (NREM) sleep, some of which must appear repetitively in NREM sleep (Guilleminault, van den Hoed and Mitler, 1978). It is frequently more convenient to use an ‘apnoea index’ of the number of apnoeas per sleep-hour: using this notation 5 apnoea/hour or more are needed to diagnose a sleep apnoea syndrome.

Pathological sleep apnoea is classified as central, obstructive or mixed. In central apnoea there are no respiratory movements. In obstructive apnoea there is no airflow despite persistent respiratory effort, with paradoxical movements of chest and abdomen in an attempt to overcome the upper airway obstruction. In mixed apnoea there is initially no airflow or respiratory effort, but after an interval respiratory effort is resumed and eventually re-establishes airflow.

Normal individuals have occasional physiological central apnoeic episodes during REM sleep or at the onset of sleep, but the mean apnoea rate in control subjects is only 0.3 apnoea/hour (women) and 1 apnoea/hour (men) (Guilleminault, van den Hoed and Mitler, 1978).

Normal sleep

Respiration while awake is mainly under voluntary control, but during sleep automatic mechanisms assume greater importance.

At the onset of sleep, respiration is often irregular with short apnoeic episodes. As sleep becomes deeper, breathing becomes more regular. During REM sleep, however, there is decreased muscle tone with diminished responsiveness to hypoxia, hypercapnia and airway obstruction: there is an associated drop in oxygen saturation and a rise in $P_{aCO_2}$, with irregular respiration and occasional apnoeic episodes (Apps, 1983).

Pathophysiology and aetiology

Obstructive apnoea

Obstructive apnoea is caused by obstruction of the upper airway, and the possible causes are the same as those already discussed in the section on aetiology of stertor. It is this type of sleep apnoea which is most likely to present to the otolaryngologist.

Fluoroscopy and fibreoptic nasoendoscopy have demonstrated that most episodes of obstructive sleep apnoea are caused by pharyngeal collapse in patients in whom there is
already some degree of obstruction of the upper airway (Guilleminault et al, 1978). Diminished pharyngeal muscle tone, especially during REM sleep, precipitates collapse of an already narrowed pharyngeal airway as a result of the Venturi effect already discussed in the section on pathophysiology of stertor. Airway obstruction results in oxygen desaturation and a rise in $P_{acO_2}$, producing increased muscle tone and arousal (often awakening), which terminates the apnoea.

In children, the commonest site of upper airway obstruction is the nasopharynx and oropharynx, caused by large tonsils and adenoids. There is a spectrum of severity in these children, with the fully developed sleep apnoea syndrome at one end of the range; at the other end are children who tend to mouth breathe and snore, but they are often found to have some periods of apnoea when the parents are directly questioned about the possibility.

**Central apnoea**

Central sleep apnoea is caused by an instability in the automatic control of respiration by the respiratory centre in the medulla. This pattern of respiration can be induced in normal subjects when awake by a voluntary period of hyperventilation, when it is termed periodic breathing. When an individual hyperventilates for 2-3 minutes, and then stops and permits his respiration to continue without exerting any voluntary control over it, there is a period of apnoea. This is followed by a few shallow breaths, and then by another period of apnoea, followed again by a few breaths. The apnoea is the result of hypocapnia induced by the period of hyperventilation. During the apnoea, the alveolar $P_{O_2}$ falls and the $P_{CO_2}$ rises. Breathing resumes because of hypoxic stimulation of the peripheral (carotid and aortic body) chemoreceptors before the $CO_2$ level has returned to normal. A few breaths eliminate the hypoxic stimulus, and breathing stops until the alveolar $P_{O_2}$ falls again. Gradually, however, the $P_{CO_2}$ returns to normal, the central (medullary) $CO_2$-driven chemoreceptors come back into play, and normal breathing resumes (Ganong, 1973). Periodic breathing in disease states is called Cheyne-Stokes respiration.

This same imbalance of respiratory control between the central and peripheral chemoreceptors can occur during sleep in some normal individuals at high altitudes, as a result of overbreathing induced by hypoxia. A similar sequence of events is induced by congestive cardiac failure, as a consequence of an imbalance between central and peripheral chemoreceptor responses caused by the prolonged circulation time, and myxoedema can cause central sleep apnoea by the same mechanism.

Neurological lesions in the brainstem or above may produce central apnoea. Some patients with brainstem lesions (for example infarcts) lose automatic control of respiration altogether, and stop breathing when they go to sleep: this is the classical disorder termed Ondine's course after the mythical water nymph whose human suitor was cursed to stop breathing and die if ever he fell asleep.

Other neurological causes in this category include brainstem encephalitis and supratentorial space-occupying lesions. Some patients with central apnoea have no demonstrable neurological or cardiovascular cause, however, and often are found to have a diminished responsiveness to hypoxia and hypercapnia even when they are awake.
Complications of sleep apnoea

Both central and obstructive sleep apnoea are associated with alveolar hypoventilation, resulting in oxygen desaturation and a rise in $P_{a\text{CO}_2}$. Very low levels of oxygen saturation can then produce an increase in pulmonary artery pressure and pulmonary vascular resistance. Eventually, irreversible pulmonary hypertension may develop with consequent cor pulmonale. Those patients with some degree of daytime desaturation as well may develop secondary polycythaemia. Systemic hypertension may also ensue (Shephard, 1984). There is some evidence that sleep apnoea may also play a part in the sudden infant death syndrome (Guilleminault et al, 1984).

There are some individuals who present a combination of the complications outlined above associated with obesity. Stool et al (1977) described the 'chubby puffer' syndrome in three children with obesity, airway obstruction caused by enlarged tonsils and/or adenoids, somnolence and cardiopulmonary disturbance. This seems to be closely related to the classical Pickwickian syndrome (so-called after Joe, the fat boy in The Pickwick Papers) of obesity, hypoventilation, daytime hypersomnolence, cor pulmonale and polycythaemia. The causation of the Pickwickian syndrome is considered to be multifactorial but some patients have obstructive sleep apnoea due to pharyngeal collapse (Sharp, Barroc and Chokroverty, 1980) as well as hypoventilation secondary to the obesity. Any cause of hypoxia in the waking state, such as obesity, will be associated with worse desaturation during sleep (especially REM sleep), and will be compounded by any element of upper airway obstruction.

Clinical features

Symptoms of obstructive sleep apnoea

The cardinal and universal symptom of obstructive sleep apnoea is snoring. Snoring may be described as the 'stridor of the pharynx' and is produced by vibration of the nearly apposed walls of the pharynx and soft palate. The actual site may vary from the level of the velopharyngeal sphincter down to the tongue base and may involve the soft palate predominantly or the pharyngeal walls primarily.

Snoring is a symptom of airway obstruction and progressive obstruction will lead eventually to shutdown of the airway as increasing velocity of inspiratory airflow through the narrowed pharynx creates an irresistible collapsing force on the pharyngeal walls (Venturi effect) (see above: Pathophysiology of stertor). This produces the classical obstructive apnoeic episode which is the second hallmark symptom of obstructive sleep apnoea. The degree of intrinsic airway compromise is of considerable importance in generating these obstructive episodes. Clearly, patients with massive obstructing tonsils and adenoids or narrow pharyngeal dimensions, as in the Treacher Collins syndrome (Shprintzen et al, 1979a), will tend to obstruct with little provocation and the addition of sleep-induced relaxation of the pharyngeal walls completes the embarrassment of the airway.

During periods of apnoea the decrease in oxygen saturation and increasing $P_{\text{CO}_2}$ tends to increase muscle tone and produce arousal. The airway is restored as the patient wakes but frequent waking and disturbed sleep are the consequences. This may result in abnormal sleep movements and restlessness with the patient adopting abnormal positions during sleep. The
patient may wake, cyanotic and gasping for air. A second, but perhaps major, consequence of disturbed sleep is daytime hypersonsmolence. This may become quite extreme in its manifestations, with constant daytime sleepiness in children and adults. The patients tend to fall asleep as soon as their concentration lapses, even when driving a car, with serious effects on concentration and performance at work. There is some evidence that sleep deprivation in children with obstructive sleep apnoea may result in retarded growth and development (Richardson et al, 1980). Feeding is often disrupted both by physical obstruction of the oropharynx and by respiratory difficulties during feeding with a tendency to aspiration.

It is also thought that nocturnal enuresis may be exacerbated by obstructive sleep apnoea (Richardson et al, 1980), although hard evidence for these rather soft symptoms is difficult to come by.

**Symptoms of central sleep apnoea**

There may be few symptoms in patients with central apnoea. Hypersonsmolence is unusual. The patient may be aware of stopping breathing at night; awakening in a 'panic or choking attack' or aware of being unable to breathe.

**Signs of obstructive sleep apnoea**

The great difficulty in diagnosing sleep apnoea is that there may be few, if any, physical signs during the day. However, in the child, a poor nasal airway with constant mouth breathing is usual. The upper lip is often coated with nasal mucus and daytime respiration may be noisy with variable stertor present. The child may be underweight and undersized for his age and there is some evidence that chronic respiratory obstruction is largely responsible for this delay in general development (Richardson et al, 1980), together with real physical problems in swallowing.

Oral examination may show 'midline tonsils' which seem to occupy the oropharynx completely, and nasal airflow may be minimal on objective testing. Physical signs may dramatically increase if the child is asleep, with obvious signs of respiratory embarrassment, sweating, stertor and tracheal 'tug' with intercostal recession.

In the adult, obesity is often present and may be substantial with patients weighing 150 kg plus. The patients may be hypersonmolent and have difficulty concentrating and staying awake during the taking of a medical history. Systemic and pulmonary hypertension may be present. Obstructing pathology in the upper airway in adults is not common, but a diligent search should be made for obstructing nasal pathology - polyps etc - and the association of obstructive sleep apnoea with skull base abnormalities and syndromes has already been discussed.

**Signs of central sleep apnoea**

Central sleep apnoea is very much less common than sleep apnoea of the obstructive type. Patients may present with signs germane to the causal pathology in the central nervous system, that is, patients who have had encephalitis affecting the brainstem, or patients with lesions in the pons, midbrain or above would manifest signs of other neurological disorders.
Alternatively, patients may present with signs relating to disruption of their sleep pattern. Hypersomnolence, irritability and, paradoxically, insomnia may be present. However, a proportion of patients with central sleep apnoea have little in the way of signs of neurological or cardiac disease and diagnosis will depend on the history and subsequent positive identification with a sleep study.

**Investigations**

**Sleep studies**

Although the history and examination may be highly suggestive of sleep apnoea, observation of the patient during sleep is definitely required if the presence of sleep disordered breathing is to be recognized and quantified. A *polysomnograph* or full sleep study is a detailed examination during sleep, with monitoring of sleep stage (EEG, EMG and eye movements recordings), chest and abdominal movements (for paradoxical movements during efforts to respire), and transcutaneous monitoring of oxygen saturation with measurement of nasal or oral airflow and continuous ECG recording. Such a study is detailed and expensive and the authors believe an initial screening or ‘mini-sleep study’, with observation during sleep and ear lobe oximeter monitoring of oxygen saturation, with an ECG strip and chest X-ray, is useful in identifying those patients who require a full study, and in most children it is sufficient to document the need for adenotonsillectomy without proceeding further. There is no doubt that if major surgery is being contemplated a full sleep study is warranted, and as appropriate treatments are developed for obstructive apnoea, such as uvulopalatopharyngoplasty or tracheostomy, this will increasingly be the case. However, in children a screening study is probably more appropriate unless there are doubts about the site of obstruction.

**Radiology**

A plain lateral X-ray of the postnasal space and upper airway is extremely useful in identifying and documenting adenoidal obstruction of the nasopharynx and tonsillar obstruction of the oropharynx. It also shows the position of the lower jaw and tongue base in addition to documenting the airway.

Computerized tomographic scans of the pharyngeal airway in patients with obstructive sleep apnoea (Bohlman et al, 1983) have shown significantly reduced dimensions of the airway at naso- and oropharyngeal level. This is really of research interest only and it is not appropriate to perform CT scanning routinely.

Chest radiography should be performed to exclude cardiopathy and right heart failure.

**Nasoendoscopy**

The use of the flexible fiberoptic endoscope to examine the physical state and dynamics of the upper airway is extremely valuable in documenting both physical and functional airway obstruction. The Olympus ENG ‘P’ flexible laryngoscope is ideal in having a 2.9 mm bundle which allows examination of the upper airway in the smallest infants. The nasal cavity, postnasal space, velopharyngeal sphincter and hypopharynx and larynx are examined in turn and, if no obvious physical obstruction is seen, a reverse Valsalva or Müller
manoeuvre can be performed to try to document areas of functional obstruction. It may even be necessary to perform this examination under light nitrous oxide anaesthesia without intubation to identify pharyngeal airway dysfunction in children (Southall et al, 1986).

*Treatment*

**Medical**

Medication has little to offer other than in central apnoea when acetazolamide may be useful in improving respiratory function, by providing central respiratory stimulation.

**Continuous positive airway pressure**

Continuous positive airway pressure can be helpful in patients with a tendency to pharyngeal airway collapse during sleep. This type of dysfunction can be minimized by increasing the pressure of the inspired air. This requires either a plastic pressure chamber to fit over the head, or alternatively a face mask. There are problems in maintaining treatment in this way, particularly in children, but more major surgical intervention may be avoided (Editorial, 1986).

**Surgical**

The major complications of obstructive sleep apnoea are potentially reversible, although end-stage cor pulmonale is not; therefore identifiable obstructive lesions in the upper airway should be dealt with surgically as soon as possible. This usually involves simple adenotonsillectomy in the paediatric group, but obstruction may occur at any site from the nasal cavity to the larynx and careful evaluation is required to reveal the site of obstruction (Kravath, Pollak and Borowiecki, 1977).

**Tracheostomy**

Some children and adults with pharyngeal airway dysfunction experience major nocturnal arterial desaturation (to around 50% P_{O_2}) and may show major cardiac dysrhythmias. These patients represent a high risk group and may well progress to cardiac failure or to life-threatening dysrhythmias (Southall et al, 1986). Tracheostomy is indicated in these unusually severely affected cases and is rapidly effective in reversing the gross cardiorespiratory abnormalities described.

Surgical modification of the major site of pharyngeal airway shutdown is possible and interest has centered on surgical modification of the velopharyngeal sphincter by uvulopalato pharyngoplasty (UPP) as described by Fujita et al (1981). This operation has been performed on quite large numbers of adults and is successful in relieving obstructive sleep apnoea in between 50 and 70% of cases, depending on the criteria involved in selecting the patients (Sher et al, 1985). The operation has not been applied in the paediatric group, where lymphoid obstruction of the upper airway is invariably the rule.
**Premedication**

Patients with the obstructive sleep apnoea syndrome are extremely sensitive to sedation. Heavy premedication prior to surgery may cause a marked deterioration in the patient's respiratory status and even produce a respiratory arrest. The same strictures apply to postoperative sedation and both should be avoided in patients with obstructive sleep apnoea. Anaesthetists involved in treating these patients should be alerted to the dangers of over-sedation.

**Velopharyngeal insufficiency**

**Definition**

Velopharyngeal insufficiency is defined as inadequate closure of the palatopharyngeal sphincter, resulting in nasal escape or rhinolalia aperta and a tendency to nasal regurgitation of fluids and food.

**Incidence**

**Cleft palate and variants**

Gross velopharyngeal insufficiency occurs in patients with unrepaired palatal clefts - such cases are increasingly rare in the developed world. Most children with a complete cleft lip and palate undergo lip repair at 3 months or 5 kg and palatal repair at 12-18 months. However, primary repair of a cleft palate fails to secure palatopharyngeal competence in 20-30% of cases (Krause, Tharp and Morris, 1976).

**Submucous cleft palate**

This condition classically consists of a bifid uvula, diastasis of the palatal muscles with separation in the midline and a 'zona pellucida', and a notched posterior border to the hard palate. The defect may be obvious to the examiner or rather subtle and more difficult to spot. A bifid uvula alone occurs in 1.2% of the population and is a microform of cleft palate. The cleft palate deformity may be regarded as a continuum of defects extending from a complete cleft of the primary and secondary palate down to simple bifidity of the uvula. This latter finding is a visible marker pointing to possible abnormalities of the intrinsic (musculus uvulae) and extrinsic muscles of the palate, which may not be obvious on casual inspection. About 20% of these patients will have velopharyngeal insufficiency because of their anatomical deficiency and adenoidectomy will tend to unmask velopharyngeal insufficiency in the remainder and must be avoided (Croft, Shprintzen and Ruben, 1981).

**The occult submucous cleft palate**

This entity was described by Kaplan (1975) and Croft et al (1978). These are patients with intrinsic muscular abnormalities in the soft palate with absence of the musculus uvulae bulge on the palatal dorsum as seen nasoendoscopically (Croft et al, 1978), and abnormal insertion of the levator 'sling' into the posterior border of the hard palate. These patients may only develop velopharyngeal insufficiency after a provocation to the palatopharyngeal
sphincter mechanism as in adenoidectomy. There are only occasional reports of velopharyngeal insufficiency developing after adenoidal regression in adolescence in these patients (Mason and Warren, 1980). Kaplan (1975) also noted that some of these patients have abnormal facial characteristics, related to mesodermal deficiencies, and these facial characteristics include:

1. maxillary hypoplasia 75%
2. lip deformity at the vermilion border 75%
3. drooping of the oral commissure 25%
4. alveolar arch abnormalities (suppressed lateral incisors) 5%
5. external ear abnormalities 10%

**Congenital velopharyngeal insufficiency**

This condition, present from birth, results in velopharyngeal insufficiency for speech, and is caused by disproportion between the soft palate and nasopharynx. The literature contains many references to ideal functional length of soft palate and depth of nasopharynx (Calnan, 1971), and there is no doubt that some individuals do have this problem. However, it seems that at least some of the cases diagnosed in the past as velopharyngeal disproportion were probably cases of the occult submucous cleft palate (Croft, Shprintzen and Ruben, 1981) which had gone unrecognized.

**Palatal neuromuscular dysfunction**

These patients have severe velopharyngeal incompetence which can be extremely difficult to manage. The morphology of soft palate and pharynx is normal but usually there is gross impairment of velar motion and related pharyngeal wall activity. In a recent series of 120 cases of velopharyngeal insufficiency (Croft, Shprintzen and Ruben, 1981), 17 (14%) had neurological problems including the Arnold-Chiari malformation, myotonic dystrophy, pseudobulbar palsy and isolated pharyngeal paralysis. The degree of velopharyngeal insufficiency may be very severe and it is extremely difficult to help these patients surgically, because of the paucity of function in the palate and related pharynx.

**Postoperative velopharyngeal insufficiency: temporary dysfunction**

Although emphasis has been already placed on the potential for adenoidectomy to cause postoperative velopharyngeal insufficiency, it, in fact, invariably 'unmasks' an already pre-existing anatomical abnormality in the velopharyngeal sphincter mechanism. The operations which would create velopharyngeal insufficiency are: resection for palatal tumours, palatal damage following 'guillotine' tonsillectomy (which is really of historical interest only); and the new operation for snoring and sleep apnoea, uvulopalatopharyngoplasty. This latter operation involves removal of the tonsils and reduction of the palatoglossus and palatopharyngeus muscles, distal soft palate and uvula. It is a procedure which is only being performed in adults at present. The operation requires careful preoperative assessment as it may lead to velopharyngeal insufficiency. Some series report transient rhinolalia aperta and nasal regurgitation in about 10% of patients following surgery (Sher et al, 1985), and this would match the present authors' experience. Fortunately most of the patients appear to regain velopharyngeal competence with the passage of time.
Pathology

The palatopharyngeal sphincter mechanism consists of palatal muscles - extrinsic and intrinsic - and the pharyngeal wall muscles. The pharyngeal wall muscles which may influence velopharyngeal closure consist of the superior constrictor, the upper fibres of palatopharyngeus and the variably present salpingopharyngeus muscle. There is a difference of opinion about the major muscle component of lateral pharyngeal wall motion; some would feel that levator palatini acts by moving the salpingopharyngeal fold medially on contraction (Isshiki, Harita and Kawano, 1985), whereas others believe that the superior constrictor muscle performs this role. The superior constrictor muscle certainly acts in producing limited 'sphincteric' contraction of the posterior pharyngeal wall in both normal subjects and patients with cleft palate (Croft, Shprintzen and Rakoff, 1981). However, these fibres may, in fact, be the most rostral fibers of palatopharyngeus which encircle the pharynx within the confines of the superior constrictor muscle. The prime elevator and mover of the soft palate is the levator palatini which elevates the velum upwards and backwards towards the posterior pharyngeal walls. The musculus uvulae contracts during valving, increasing the dorsal mass of the soft palate and producing the so-called 'levator eminence'. The palate makes contact with the surrounding lateral and posterior pharyngeal walls which are variably dynamic. A recent endoscopic study in 80 normal subjects and 120 patients with velopharyngeal pathology (Croft, Shprintzen and Rakoff, 1981) showed surprising variability of valving patterns with the main closure pattern being coronal (mainly palatal), sagittal (mainly lateral wall), circular, and circular with Passavant's ridge. Interestingly, active posterior wall contraction occurs in 20% of the normal population studied and appears to be more than a 'compensatory' mechanism. Furthermore, much the same spread of attempted closure patterns could be ascertained in the group of individuals with incompetent sphincters.

Clinical features

Symptoms

Velopharyngeal insufficiency is always accompanied by nasal escape or rhinolalia aperta during speech. Reduced intelligibility of speech results from disruption of consonant production. The inability to generate high intraoral pressures creates serious disturbances with fricatives such as /s/ and /z/ and plosives such as /p/ and /b/. These distortions may be accompanied by snorting caused by excessive nasal airway turbulence and 'glottal stops' to try to reduce nasal escape. Extra vocal effort places strain on the larynx and there is an increased incidence of vocal cord pathology in patients with velopharyngeal insufficiency (McWilliams, Lavorato and Bluestone, 1973). Gross velopharyngeal insufficiency may also be accompanied by nasal regurgitation of fluids and food during swallowing.

Signs

Facial

When velopharyngeal insufficiency occurs, a careful search for stigmata of cleft palate or its microforms is required with particular attention to the facies and upper jaw, with palpation of the hard and soft palate. Detection of nasal airflow during connected speech may be straightforward but, in marginal cases, a nasal escape indicator may be helpful. This is a
piece of polystyrene foam inside a U-shaped small-bore glass tube. When the tube is connected to the nose, the polystyrene agitates in the presence of abnormal nasal airflow. The use of a small laryngeal mirror or nasal occlusion is less satisfactory.

**Otological**

Patients with long-standing velopharyngeal insufficiency have been shown to have an increased incidence of middle ear disease. The incidence (approximately 50%) approaches that seen in the cleft palate population (Heller et al, 1975).

**Speech assessment**

It is possible to confuse the difference between hyper- and hyponasality. In hyponasality the nasal resonance normally heard in connected speech is missing, there being reduced or absent airflow through the nose. This abnormality or 'rhinolalia clausa' may sound a little like hypernasality with constant and excessive nasal resonance in some cases. Obviously, such a confusion might lead to the wrong treatment being tried and the authors have seen adenoids removed in children with hypernasality in the belief that they really required an increased nasal airway. This confusion may occur in subtle cases and an expert speech therapy opinion should always be sought before resorting to surgery. It is also dangerous to perform adenoidectomy in children who have had major problems requiring speech therapy in the past. A regression may occur and velopharyngeal insufficiency may be precipitated.

**Investigations**

**Endoscopy**

**Oral endoscopy**

Viewing and assessing function of the velopharyngeal sphincter perorally became popular during the 1960s, but has the major disadvantage of not allowing assessment during connected speech.

**Nasoendoscopy**

An excellent view of the velopharyngeal isthmus can be obtained pernasally and there is no impediment to connected speech (Pigott, 1969). Rigid or fibreoptic flexible instruments are now routinely used to establish the degree, site and severity of velopharyngeal insufficiency. This is most important in treatment planning and in subsequent follow-up.

**Radiology**

**Radiographs**

Plain lateral X-rays of the soft palate in phonation provide very limited information about the state of the velopharyngeal sphincter, but will give information about velar elevation and posterior wall motion.
**Multiview videofluoroscopy**

This certainly provides the most comprehensive and functional view of the velopharyngeal sphincter in motion (Skolnick, 1970). The velum and pharyngeal walls are coated with barium via a nasal dropper and the patient examined in lateral, frontal, base and oblique projections. Simultaneous sound recordings are taken and radiation exposure is significantly lower than with cine-radiography. An excellent view of the various component movements contributing to velopharyngeal closure is readily obtained.

**Audiometry/tympanometry**

Approximately 50% of the patients will have middle ear pathology and conductive deafness. Appropriate investigation will include full audiometric assessment.

**Treatment**

**Medical/conservative**

**Speech therapy**

Speech therapy is a vital part of therapy both in its own right and in patients with inconsistent velopharyngeal insufficiency who may well learn to achieve velopharyngeal closure in sustained speech. Even if surgery is required, speech therapy is a vital part of treatment, both pre- and postoperatively.

**Biofeedback**

Patients with inconsistent velopharyngeal insufficiency who are capable of closure for isolated speech tasks can be helped by nasoendoscopy plus video monitoring to show up the problem and to teach remedial techniques.

**Obturators**

If there are major contraindications to surgery then prosthetic speech appliances can be constructed and mounted on a dental plate to fit between the palate and pharyngeal walls. These can be difficult to fit and are little used.

**Surgical**

**Cleft palate repair**

Repair is usually accomplished between the ages of 12 and 18 months. The major techniques are the Von Langenbeck repair and V-Y retroposition or the Wardill-Kilner technique. These are described in Chapter 21.
**Pharyngoplasty**

About one in four patients who have primary surgery for the correction of palatal cleft will fail to achieve velopharyngeal competence and most will require further surgery if normal or near normal speech is to be achieved. Although prosthetic management is a possibility, most centres would opt for surgery if there are no contraindications.

**Posterior wall augmentation**

This technique of narrowing the velopharyngeal port by positioning a non-toxic substance in the posterior pharyngeal wall is long established. Current interest centres on cartilage, Teflon paste or collagen. The authors' experience is that the method can only succeed if the gap in the velopharyngeal sphincter is very small (4 mm), and the vertical positioning of the implant is vital in relation to the variable height of velopharyngeal sphincter closure. Nasoendoscopic control is ideal in securing the correct site and amount of Teflon to be injected (Lewy, Cole and Wepman, 1965).

**Pharyngeal flap ± palatal pushback**

Although described over 100 years ago, the pharyngeal flap has only come into general use during the past 30 years. The method consists of elevating a unipedicle soft tissue flap from the posterior pharyngeal wall and suturing and 'setting' it into the deficient soft palate. The flap may be either superiorly or inferiorly based, but most surgeons now prefer the former. The flap provides an organic soft tissue obturator for the velopharyngeal sphincter (Shprintzen et al, 1979b). Complete velopharyngeal closure depends on lateral wall movement medially to contact the flap, thus closing off the lateral airways or 'ports'.

Success rates following pharyngeal flap procedures vary from 33 to 100% (Bernstein, 1967; Shprintzen et al, 1979b), but speech is invariably improved.

**Sphincter-pharyngoplasty**

Sphincter-pharyngoplasty or the Orticochea-Hynes pharyngoplasty is essentially a transfer of the palatopharyngeus muscles into a higher plane in the posterior pharyngeal wall, uniting the two muscles in a sphincteric manner in the plane of velopharyngeal closure. The operation is useful in those patients whose velopharyngeal insufficiency and sphincter gap is modest.