Recent Advances in the Pathogenesis, Diagnosis, and Management of Otitis Media

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Otitis media is the most frequent diagnosis made by physicians who care for children. Acute otitis media is usually suppurative or purulent, but serous middle ear effusions may also have an acute onset. Chronic otitis media with effusion has many synonyms, including such terms as secretory, serous, nonsuppurative, and "glue ear". A chronic effusion may be serous, mucoid, or even purulent. In some instances, the eardrum may be retracted or collapsed without a middle ear effusion, which is termed atelectasis of the tympanic membrane, and is the result of persistent or intermittent negative middle ear pressure. It is often difficult to determine from the history and visual inspection of the tympanic membrane the precise type of otitis media present since in most patients, especially infants and young children, the disease is a continuum of the different stages. Some patients may have recurrent acute attacks without an apparent effusion in-between, whereas others may have only chronic otitis media with effusion, and still others may have recurrent acute episodes superimposed on a persistent middle ear effusion. Atelectasis of the tympanic membrane may represent the only pathology in some patients but in others the condition can be present between episodes of otitis media with effusion. Chronic otitis media with perforation and otorrhea is one of the sequelae of acute or chronic otitis media with effusion.

Epidemiology

Infants and young children are at highest risk for the acquisition of otitis media, with the peak prevalence rate occurring between 6 and 36 months, and a lesser peak between 4 and 7 years. One study of 2565 children followed for the first three years of life found that only 29 per cent of infants failed to develop at least one attack of otitis media, whereas about one-third had three or more episodes. In addition, the study showed that after the first episode, 40 per cent of children had a middle ear effusion that persisted for four weeks and 10 per cent had an effusion that was still present at three months. Infants in whom otitis media with effusion develops in the first years of life have an increased risk of recurrent acute or chronic middle ear effusions. The overall prevalence of the disease in children has been estimated to be between 15 and 20 per cent. However, the incidence and prevalence of the disease tend to decrease as a function of age after the age of 6 years. The incidence is higher in males, lower socioeconomic groups, Alaskan natives (Eskimos), American Indians, children with cleft palate and other craniofacial anomalies, and is higher in whites than in blacks. The incidence is also higher in winter and early spring.

Physiology and Pathophysiology of the Eustachian Tube Related to Pathogenesis of Otitis Media

The pathogenesis of otitis media with effusion appears to be related to abnormal function of the eustachian tube. Investigation into the exact nature of this dysfunction requires an understanding of the system constituted by the palate, nasal cavity, nasopharynx, eustachian tube, middle ear, and mastoid air cells. Within this system, the eustachian tube has at least three physiologic functions with respect to the middle ear: equilibration of air...
pressure (ventilation) in the middle ear with atmospheric pressure, protection from nasopharyngeal sound pressure and secretions, and clearance into the nasopharynx of secretions produced within the middle ear.

The protective and clearance functions of the eustachian tube have been assessed by radiographic techniques. Radiopaque material was instilled through the nose of the children in order to assess the protective function by observing the retrograde flow of the medium from the nasopharynx into the eustachian tube. Clearance function was assessed following instillation or insufflation of radiopaque material into the middle ear and observing the flow of the medium down the eustachian tube into the nasopharynx.

The findings of these radiographic studies can be understood if a model of the system is constructed. The eustachian tube, middle ear, and mastoid can be likened to a flask with a long, narrow neck, the mouth of the flask representing the nasopharyngeal end, the narrow neck the isthmus of the eustachian tube, and the bulbous portion the middle ear and mastoid air chamber. When a small amount of liquid is instilled into the mouth of the flask, liquid flow will stop somewhere in the narrow neck, owing to capillarity and to the cushion of air pressure that develops in the chamber of the flask. This basic geometric design is considered to be critical for the protective function of the eustachian tube and middle ear system. Reflux of the liquid into the vessel occurs if a hole is made in the bulbous portion of the flask, since the pressure in the bottom of the flask remains unchanged. This is analogous to the condition in which a perforation of the tympanic membrane or the presence of a tympanostomy tube could allow reflux of nasopharyngeal secretions as a result of loss of the middle ear-mastoid air cushion. The effect of the application of a negative pressure to the bottom of the flask: the liquid is aspirated into the vessel. In the clinical situation represented by the model, high negative middle ear air pressure could lead to the aspiration of nasopharyngeal secretions into the middle ear. The effect of applied positive pressure to the mouth of the flask is also shown: the liquid is insufflated into the vessel. Noseblowing, crying, or closed-nose swallowing could create a high positive nasopharyngeal pressure and result in a similar condition in the human system.

However, one of the major differences between a flask with a rigid neck and a biological tube such as the eustachian tube is that the isthmus (neck) of the human tube is compliant. Application of positive pressure at the mouth of a flask with a compliant neck would distent the neck, enhancing fluid flow into the vessel. The effect of applied negative pressure in a flask with a compliant neck is shown. Liquid flow through the neck would not occur until a negative pressure was slowly applied to the bottom of the flask. In this case, fluid flow would occur even if the neck were collapsed, but if the negative pressure were applied suddenly, temporary locking of the compliant neck would prevent flow of the liquid. Therefore, the speed of application of the negative pressure, as well as the compliance in such a system, would appear to be critical factors in the results obtained. Clinically, aspiration of gas into the middle ear would be possible, since negative middle ear pressure would develop slowly, as the gas was absorbed by the middle ear mucous membrane. However, sudden application of negative middle ear pressure, such as would occur with rapid alterations in atmospheric pressure (as in an airplane during descent, when diving, or when attempting to test the ventilatory function of the eustachian tube), could lock the tube, thus preventing the flow of air. Likewise, sudden application of negative pressure at the nasopharyngeal portion of a highly compliant tube could also result in locking. Negative pressure in the middle ear
would be less likely to functionally obstruct the eustachian tube than negative pressure at the nasopharyngeal end, since the tympanic portion of the tube is a bony funnel. This geometric difference is probably important in the mechanics of passive tubal opening, when a gradient (negative middle ear pressure) is required to open the tube or assist the active opening mechanism.

Certain aspects of the clearance function of fluid from the middle ear into the nasopharynx can be demonstrated by inverting the flask of the model. The liquid trapped in the bulbous portion of the flask does not flow out of the vessel as a result of the relative negative pressure that develops inside the chamber. However, if a hole is made in the vessel, the liquid drains out of the flask, since the suction is broken. Clinically, these conditions occur in cases of middle ear effusion in which pressure is relieved by myringotomy. Insufflation of air into the flask achieves a release of pressure, which may explain the frequent success of politzerization or the Valsalva maneuver in clearing middle ear effusion.

The foregoing description of fluid flow through a flask only presents some of the mechanical aspects of the physiology of the human middle ear system. Other factors that probably affect the flow of liquid and air through the middle ear in the physiologic state would include: (1) the mucociliary transport system of the eustachian tube and middle ear, (2) contraction of the tensor tympani muscle and tympanic membrane movement, (3) active tubal opening mechanisms, and (4) surface tension factors.

The function of the eustachian tube to equilibrate air pressure can be assessed by a modification of the manometric technique. A perforation of the tympanic membrane or a tympanostomy tube must be present, so that middle ear pressure can be directly inflated or deflated employing a pump-manometer system. Figure is a simplified explanation of the combined passive and active function test, when positive pressure is applied to the middle ear - inflation. This test is similar to ascending in an airplane until the eustachian tube passively opens. The test consists of the application of enough positive pressure into the middle ear to force open the eustachian tube. The pressure remaining in the middle ear after passive opening and closing is termed the closing pressure. Further equilibration of pressure is by swallowing - active function - which is the result of contraction of the tensor veli palatini muscle. When the muscle contracts, the lumen of the eustachian tube is opened and air flows down the tube. The pressure can be monitored on a strip chart recorder; the pressure remaining in the middle ear after passive and active function is termed the residual positive pressure.

Figure shows the deflation phase of the study, which is similar to descent in an airplane. Low negative pressure is applied to the middle ear, which is then equilibrated by active function. The pressure remaining in the middle ear after swallowing is termed the residual negative pressure. In certain instances, active function to applied low positive pressure is also assessed. Figure shows that the test is similar to ascent in an airplane, but to an altitude lower than a pressure that would force open the eustachian tube. The patient is asked to swallow in an attempt to equilibrate the pressure by active function.

Figure shows the symbols employed and examples of results obtained in ventilation studies. Example A describes a typical study of a patient with normal function. Following passive opening and closing of the eustachian tube during the inflation phase of the study, the
patient is able to completely equilibrate the remaining positive pressure. Active swallowing also completely equilibrates applied negative pressure - deflation. Example B describes a typical study of a child who had had otitis media with effusion. The eustachian tube passively opens and closes following inflation, but subsequent swallowing fails to equilibrate the residual positive pressure. In the deflation phase of the study, the child is unable to equilibrate negative pressure. Inflation to a pressure below the opening pressure but above the closing pressure cannot be equilibrated by the active swallowing function.

From these studies of fluid flow through a vessel and studies in children and adults, the following physiology of the eustachian tube and stages in the pathogenesis of otitis media with effusion have been postulated. Figure shows that the normal eustachian tube is functionally obstructed or collapsed at rest, with probably a slight negative pressure existing in the middle ear. In ideal tubal function, intermittent active opening of the eustachian tube maintains near-ambient pressures in the middle ear.

It is suspected that, in cases in which active function is inefficient in opening the eustachian tube, functional collapse of the tube persists. The interval between openings would then depend on the establishment of a pressure gradient between the middle ear cavity and the nasopharynx, which passively assists tubal function. Physiologically, this gradient is achieved by the absorption of middle ear gas, which results in progressive negative middle ear pressure. This type of middle ear pressure regulation appears to be quite common in children, as moderate-to-high negative middle ear pressures have been identified by tympanometry in many who are apparently normal.

There appear to be two types of eustachian tube obstruction which would result in an otitis media with effusion: functional and mechanical. Functional obstruction could result from persistent collapse of the eustachian tube owing to increased tubal compliance, or an inadequate active opening mechanism, or both. Functional eustachian tube obstruction is common in infants and younger children, since the amount and stiffness of the cartilage support of the eustachian tube are less than in older children and adults, and since there appear to be marked age differences in the craniofacial base which render the tensor veli palatini muscle less efficient prior to puberty. The other major type of dysfunction of the eustachian tube is abnormal patency. In its extreme form the tube is open even at rest, ie, patulous. Lesser degrees of abnormal patency would result in a semipatulous eustachian tube which would be closed at rest but have low resistance in comparison to the normal tube. Increased patency of the tube may be caused by abnormal tube geometry or to a decrease in the extramural pressure (such as occurs with weight loss, or possibly due to mural or intraluminal factors).

Figure depicts the pathogenesis of otitis media with effusion resulting from eustachian tube dysfunction. Functional obstruction may result in persistent high negative middle ear pressure, and when associated with collapse of the tympanic membrane has been termed atelectasis. If a large bolus of air enters the middle ear from the nasopharynx when there is high negative middle ear pressure, nasopharyngeal secretions could be aspirated into the middle ear and result in an acute bacterial otitis media with effusion.

If middle ear pressure equilibration does not occur, persistent functional eustachian tube obstruction could result in a sterile otitis media with effusion. The occurrence of an otitis
media with effusion at this stage might depend on the degree and duration of the negative pressure, as well as middle ear hypoxia or hypercarbia. Since tubal opening could be possible in a middle ear with an effusion, aspiration of nasopharyngeal secretions might occur, thus creating the clinical condition in which persistent otitis media with effusion and recurrent acute bacterial otitis media with effusion occur together. All infants with unrepaired palatal clefts and many children with repaired palates have otitis media with effusion as a result of functional obstruction of the eustachian tube.

Intrinsic mechanical obstruction of the eustachian tube may be caused by inflammation. Most ears at risk for the development of atelectasis or otitis media with effusion when inflammation is present probably have a significant degree of functional obstruction. An upper respiratory tract infection in such children has been shown to significantly decrease eustachian tube function. Periods of upper respiratory tract infection may then result in either atelectasis of the middle ear, bacterial otitis media with effusion, or a sterile otitis media with effusion owing to swelling of the eustachian tube lumen. The mechanisms are similar to those described for functional obstruction of the eustachian tube. Allergy as a cause of intrinsic mechanical obstruction of the eustachian tube has not been demonstrated.

Extrinsic mechanical obstruction of the eustachian tube may be the result of extrinsic compression by nasopharyngeal tumors or adenoids. Partial obstruction of the eustachian tube may result only in atelectasis of the middle ear or a bacterial otitis media with effusion, but more severe obstruction could result in a sterile otitis media with effusion.

Abnormal Patency of the Eustachian Tube

A patulous eustachian tube usually permits air to flow readily from the nasopharynx into the middle ear, which thus remains well ventilated; but also, unwanted nasopharyngeal secretions can traverse the tube and result in "reflux otitis media". A semipatulous eustachian tube may be obstructed functionally as the result of increased tubal compliance and the middle ear may even have negative pressure, or an effusion, or both. Since the tubal walls are abnormally distensible, nasopharyngeal secretions may readily be insufflated into the middle ear even with modest positive nasopharyngeal pressures, for example, as a result of noseblowing, sneezing, crying, or closed-nose swallowing. If active tubal opening (tensor veli palatini contraction) occurs, resulting in an abnormally patent tube, reflux or insufflation of nasopharyngeal secretions would also be likely. If the eustachian tube has lower resistance than normal but remains functionally obstructed even during attempts at active tubal opening, it is conceivable that nasopharyngeal secretions, rather than air, would more readily enter the middle ear. Native Americans (American Indians) have been shown to have tubal resistances that are lower than those of the average Caucasian. They seem to have an increased incidence of reflux of nasopharyngeal secretion into the middle ear, and frequently suffer from recurrent acute otitis media, which is often associated with perforation and discharge. However, American Indians have a low incidence of cholesteatoma. This type of eustachian tube function and middle ear disease is different from those in individuals who have a cleft palate.
Nasal Obstruction

Nasal obstruction may also be involved in the pathogenesis of otitis media with effusion. Swallowing when the nose is obstructed (due to inflammation or obstructed adenoids) results in an initial positive nasopharyngeal air pressure, followed by a negative pressure phase. When the tube is pliant, positive nasopharyngeal pressure might insufflate infected secretions into the middle ear, especially when the middle ear has a high negative middle ear pressure; or, with negative nasopharyngeal pressure, such a tube could be prevented from opening and be further obstructed functionally (the "Toynbee phenomenon").

Diagnostic Methods

Next to the patient's medical history, the most important diagnostic tool is the otoscopic examination. However, before adequate visualization of the external canal and tympanic membrane can be obtained, obstructing cerumen must be removed from the canal. This can be accomplished with either an otoscope with a surgical head and a wire loop or a blunt cerumen curette, or by gently irrigating the canal with warm water. In the newborn infant the external canal is filled with vernix caseosa which disappears shortly after birth.

Proper assessment of the tympanic membrane and its mobility is accomplished by the use of the pneumatic otoscope in which the diagnostic head has an adequate seal. Physical findings noted during inspection of the tympanic membrane should include position, color, degree of translucency, and mobility. Evaluation of the light reflex is of limited value. The normal tympanic membrane should be in the neutral position, in contrast to a drum that is bulging. The latter condition may be caused by increased middle ear air pressure, or an effusion within the middle ear, or both; the malleus handle and short process are obscured by the bulging drum. Retraction of the tympanic membrane usually indicates the presence of middle ear negative pressure; however, the tympanic membrane may even be severely retracted (presumably because of previous high negative middle ear pressure or inflammation with subsequent fixation of the ossicles and ligaments) without the presence of high negative middle ear pressure. When retraction of the tympanic membrane is present, the short process of the malleus is prominent and the long process is foreshortened. The normal tympanic membrane has a ground-glass appearance; a blue or yellow color usually indicates a middle ear effusion. A red tympanic membrane alone may not be indicative of pathology, since the blood vessels of the drum head may be engorged as the result of crying, sneezing, or blowing the nose. The distinction between translucency and opacification is critical in identification of middle ear disease. The normal tympanic membrane should be translucent, the observer being able to look through the drum and visualize the middle ear landmarks: incudostapedial joint, promontory, round window niche, and frequently the chorda tympani nerve. If a middle ear effusion is present medial to a translucent drum, an air-fluid level or bubbles of air mixed with the fluid may be visible. Inability to visualize the middle ear structures indicates opacification of the drum, which is usually the result of thickening of the tympanic membrane, or a middle ear effusion, or both.

Abnormal middle ear pressure is reflected in the pattern of tympanic membrane mobility when first positive and then negative pressure is applied to the external canal. Pressure is applied by first obtaining an adequate seal between the external auditory canal and the ear speculum, and then by applying slight pressure on the rubber bulb (positive pressure)
followed by release of the bulb (negative pressure). The presence of a liquid, or abnormal pressure (positive or negative), or both, within the middle ear can markedly dampen the movement of the eardrum. When the middle ear has a high negative pressure, the tympanic membrane will not move to applied positive pressure but will move outward with applied negative pressure if the applied pressure exceeds the middle ear negative pressure. Conversely, when high middle ear positive pressure is present, the drum will not move to applied negative pressure but will move to applied positive pressure as long as there is air within the middle ear. The tympanic membrane will not move to applied positive or negative pressure when the middle ear-mastoid cavity is completely filled with a liquid. When available, the otomicroscope may be helpful in the evaluation of the difficult-to-assess tympanic membrane.

The most significant advance in the identification of middle ear disease is the use of the **electroacoustic impedance bridge** with which a **tympanogram** can be obtained. To perform tympanometry, a small probe is inserted into the external auditory canal. A tone of fixed characteristics is presented via the probe, and the compliance (mobility) of the tympanic membrane is measured electronically while the external canal pressure is artificially varied. The patterns obtained are highly diagnostic in distinguishing a normal ear from one in which a middle ear effusion is present. It is especially beneficial for the following: as an aid in diagnosis when otoscopy is equivocal or difficult to perform; for objective determination of the middle ear pressure; as an aid in teaching otoscopy; and for screening for ear disease. Tympanometry also may be helpful in identifying middle ear disease other than effusion, such as disarticulation or fixation of the ossicular chain.

However, tympanometry does not assess hearing; **audiometry** measures this function. Usually, in patients over the age of two years behavioral audiometry, which is a subjective assessment of hearing, is possible. In the young infant or in children who are difficult to test, objective audiometry is necessary (auditory brainstem response audiometry or the acoustic reflex obtained with an electroacoustic impedance bridge).

**Aspiration of the middle ear** is the most definitive method of verifying the presence and type of a middle ear effusion. Diagnostic tympanocentesis may be performed by inserting, through the inferior portion of the tympanic membrane, an 18-gauge spinal needle attached to a syringe. Culturing the ear canal and cleansing of the canal with alcohol should precede the procedure; the canal culture is helpful in determining whether organisms cultured are contaminants from the canal or pathogens from the middle ear. **Microbiologic studies** of the aspirate constitute an invaluable diagnostic aid to the clinician. Indications for tympanocentesis include the following:

1. Otitis media in children who are critically ill.
2. Unsatisfactory response to antimicrobial therapy.
3. Onset of otitis media in a child who is receiving an antimicrobial agent.
5. Otitis media in the neonate, the very young infant, or in the immunologically deficient patient, in each of whom an unusual organism may be suspected.
When therapeutic drainage is required, a myringotomy should be performed and the incision should be large enough to allow for adequately drainage and aeration of the middle ear. Myringotomy should always be performed when a suppurative complication is present and may also be helpful following tympanocentesis for the indications listed above. In addition, a myringotomy frequently will be beneficial for a child who has unusually severe otalgia, either when initially examined or at any time during the course of the disease.

Roentgenographic assessment of the ear and temporal bone is frequently helpful in diagnosis and management; polytomography is more helpful in certain instances than conventional radiographs.

When the tympanic membrane is not intact (as a result of perforation or insertion of a tympanostomy tube), assessment of the ventilatory function of the eustachian tube by pressure-flow studies may be an additional aid in the diagnostic workup of a child with middle ear disease.

**Acute Otitis Media With Effusion**

In the classic description of this condition, a child who has an upper respiratory infection for several days suddenly develops otalgia, fever, and hearing loss. Examination with the pneumatic otoscope reveals a hyperemic, opaque, bulging tympanic membrane that has poor mobility. Purulent otorrhea may be present. However, earache and fever are not invariable concomitants of infection. Because of the variability of symptoms, an otoscopic examination should always be included in the evaluation of infants and children; those who have diminished or absent mobility and opacification of the tympanic membrane should be suspected of having a bacterial otitis media with effusion. Middle ear infection must be ruled out in any child with a "fever of undetermined origin". When the diagnosis of acute otitis media with effusion is in doubt, or when determination of the causative agent is desirable, aspiration of the middle ear should be performed.

Rational therapy for acute otitis media with effusion depends upon knowledge of the bacterial cause of the disease. The bacteria that have been cultured from middle ear effusions in children with acute otitis media have been shown to be the same found in the nasopharynx. *Streptococcus pneumoniae* has been cultured from approximately 40 per cent of the effusions and is the most common causative agent in all age groups. *Haemophilus influenzae* causes about 20 per cent of cases. This proportion declines with increasing age but *H. influenzae* is still significant in all age groups. Recently, there has been an increasing percentage of *H. influenzae* strains, 15 to 30 per cent, that have been beta-lactamase producing and, therefore, ampicillin-resistant. Group A beta-hemolytic streptococcus and *Staphylococcus aureus* account for 7 and 2 per cent, respectively. In about 25 per cent of effusions, no bacteria are cultured. In neonates, approximately 20 per cent of effusions may contain gram-negative enteric bacilli.

In patients with classic signs and symptoms of acute otitis media, antimicrobial therapy is the treatment of choice. Since the clinician rarely is certain of the causative organism before starting therapy for otitis media, ampicillin is the single most useful drug, and will usually be effective against the four most commonly encountered bacteria. Oral ampicillin, 50 to 100 mg/kg/24 hr, in four divided doses for 10 to 14 days, is recommended. Amoxicillin,
20 to 40 mg/kg/24 hr, is probably equally effective and can be given in three divided doses. If the patient is allergic to the penicillins, then a combination of oral erythromycin, 50 mg/kg/24 hr, and triple sulfonamides, 100 mg/kg/24 hr (or sulfisoxazole, 150 mg/kg/24 hr), in four divided doses, is a suitable alternative. The combination of trimethoprim and sulfamethoxazole, 8 to 40 mg/kg/24 hr in two divided doses, also can initially be given to penicillin-sensitive individuals, but its effectiveness in the treatment of acute otitis media caused by *Streptococcus pyogenes* is uncertain. A new cephalosporin, cefaclor, 40 mg/kg/24 hr, in three divided doses, appears to be a promising new antimicrobial agent for otitis media since it is effective against the common pathogens causing acute otitis media.

Additional supportive therapy, including analgesics, antipyretics, and local heat, will usually be helpful. In some instances, meperidine hydrochloride may also be required for sedation. The efficacy of antihistamines and decongestants in the treatment of acute otitis media has not been proven.

If the patient continues to have appreciable pain or persistent fever, or both, after 24 to 48 hours, tympanocentesis/myringotomy should be performed as a diagnostic and therapeutic procedure. At this stage the presence of an effusion alone does not constitute a clinical failure. In patients with unusually severe earache, myringotomy may be performed initially in order to provide immediate relief.

When an unusual organism is cultured from a middle ear aspirate, sensitivity testing will help in the choice of antimicrobial agents. An example of this situation is an acute otitis media caused by *H. influenzae* that is resistant to ampicillin. When this occurs or when the patient fails to improve clinically after the initial treatment with ampicillin or amoxicillin and a tympanocentesis/myringotomy is not performed, the initial antimicrobial should be changed since an ampicillin-resistant *H. influenzae* should be suspected. Erythromycin in combination with a sulfonamide, or trimethoprim-sulfamethoxazole, or cefaclor are appropriate choices at present.

All patients should be reevaluated approximately two weeks after the institution of treatment. At this time some patients will have had complete resolution of the middle ear effusion but in others complete clearing of the effusion may take six weeks or longer. Within two to three months the tympanic membrane should be entirely normal. If complete resolution has occurred and the episode represents the only known attack, the patient may be discharged. However, periodic follow-up is indicated for patients who have had recurrent episodes.

**Recurrent Acute Otitis Media**

It is not uncommon for children, especially infants, to have recurrent bouts of acute otitis media. Some children have an acute episode with almost every respiratory tract infection, have more or less dramatic symptoms, respond well to therapy, and improve with advancing age. Others are more difficult, in that they have persistent middle ear effusion and suffer recurrent episodes of acute otitis media with effusion superimposed on the chronic disorder. The child with recurrent acute otitis media with effusion that completely clears between episodes may be managed as previously outlined. However, if the bouts are frequent and close together, further treatment, similar to that described for patients with chronic otitis media with effusion, is indicated. In many of these children, the underlying cause is not
evident but myringotomy with insertion of middle ear ventilation tubes is frequently helpful. Prophylactic antimicrobials (a daily dose of ampicillin or sulfonamides) have been advocated as an alternative to myringotomy and ventilating tubes in children with recurrent acute otitis media with effusion who are free of effusion between attacks. The efficacy of myringotomy with tympanostomy tube insertion and of chemoprophylaxis is as poorly established as is the usefulness of other forms of prevention, such as hyposensitization and adenoidectomy.

**Chronic Otitis Media With Effusion**

Chronic middle ear effusions may be thin (serous), thick (mucoid), or purulent. Pneumatic otoscopy will frequently reveal either a retracted or convex tympanic membrane. The membrane is usually opaque, but when it is translucent an air-fluid level or air bubbles may be seen and an amber or sometimes bluish fluid may be apparent in the middle ear. The mobility of the eardrum is almost always impaired. Occasionally, even when the middle ear is free of effusion, the tympanic membrane will be retracted and its mobility impaired. This finding usually indicates the presence of negative middle ear air pressure, which, when extreme, is termed "atelectasis of the tympanic membrane"; it may be accompanied by the same symptoms usually associated with otitis media with effusion. In both conditions, auditory acuity is usually decreased, and although systemic symptoms are usually absent, there may be behavioural disturbances owing to the child's inability to communicate adequately. A feeling of fullness in the ear, tinnitus, and even vertigo may be present. Audiometry may be helpful in establishing the diagnosis but is not a reliable indicator, because some patients, even with thick middle ear effusions, have fairly good hearing. Tympanometry is a more reliable diagnostic tool. A patient with chronic otitis media with effusion who has not received prior antimicrobial therapy should be treated initially as a case of acute otitis media with effusion, since bacteria are frequently present.

A study was conducted of 274 children who had recurrent acute or chronic otitis media with effusion. Of the ears with effusion 45 per cent were found to contain bacteria, and 11 per cent bacteria what were "probable pathogens" (S. pneumoniae, H. influenzae, and S. pyogenes). Bacteria were also found in 40 per cent of the ears without effusions. The type of organism found did not vary with the age of the patient studied or the season of the year. Pathogens have also been aspirated from young infants with chronic effusions. However, the efficacy of antimicrobials, corticosteroids, decongestants, and antihistamines for chronic otitis media with effusion has not been proven. Occasionally, attempts at middle ear inflation by Valsalva's or Politzer's method are successful.

If the effusion persists for eight weeks or longer, or if there have been frequent recurrences of episodes of acute otitis media with effusion, the patient requires further evaluation. Several avenues of investigation are open: a search for respiratory allergy may prove fruitful; a lateral roentgenogram of the nasopharynx may reveal adenoid tissue obstructing the nose and nasopharynx; immunologic studies may be of value if other organs are involved (the lung, for example). More thorough physical examination may reveal abnormalities, such as submucous cleft palate or a tumour of the nasopharynx, that require definitive management.

For those patients in whom medical management has failed, myringotomy with aspiration of the middle ear fluid is indicated. Frequently, insertion of a ventilation tube may
be necessary to allow the middle ear mucous membrane to return to normal and to prevent subsequent accumulation of effusion. Myringotomy and insertion of ventilation tubes may also be helpful in patients with atelectasis of the middle ear when significant symptoms - pain, hearing loss, vertigo, or tinnitus - are present. Ventilation tubes should be used to prevent permanent structural damage and cholesteatoma if a deep retraction pocket develops in the posterosuperior quadrant or in the attic (pars flaccida) portion of the tympanic membrane. Occasionally, troublesome otorrhea develops after the insertion of tympanostomy tubes. This can usually be treated successfully with ear drops containing neomycin, polymyxin, or colistin with hydrocortisone. Since these medications may be ototoxic, some physicians advocate the use of systemic antibiotics without the aural drops. In most children otitis media with effusion is usually self-limiting and will improve with advancing age, but in selected cases allergic hyposensitization and adenoidectomy may be beneficial; however, the efficacy of these methods of management has not been established. Tonsillectomy (in the absence of documented recurrent tonsillitis) does not seem to alter the course of otitis of any type and should not be performed alone or in conjunction with adenoidectomy for these conditions.

Since otitis media with effusion is universal in the infant with an unrepaired cleft palate and very common following surgical repair of the palate, tympanostomy tubes should be inserted when a chronic effusion is present to prevent the complications and sequelae of otitis media.

**Complications and Sequelae of Otitis Media**

Today, the intracranial suppurative complications of otitis media with effusion are relatively uncommon except in neglected cases. However, the complications and sequelae that occur within the aural cavity and adjacent structures of the temporal bone are more common, and awareness of their possible existence is essential in management of children with otitis media with effusion. Even though many of these less serious conditions may not threaten life (as when there is an intracranial extension of the disease), the quality of life may be severely affected, making prevention imperative.

**Nonsuppurative Complications and Sequelae**

The nonsuppurative complications and sequelae of otitis media with effusion are hearing loss, adhesive otitis media, tympanosclerosis, and ossicular discontinuity.

*Hearing loss* is by far the most prevalent complication and morbid outcome of otitis media with effusion, and may be caused by one or more of the intra-aural complications or sequelae. To a varying degree, fluctuating or persistent loss of hearing is always associated with acute or chronic otitis media with effusion. The presence of high negative pressure within the middle ear (atelectasis), in the absence of an effusion, can also be associated with a significant hearing loss. The audiogram usually reveals a mild to moderate conductive loss. However, there may be a sensorineural component, generally attributed to the effect of increased tension and stiffness of the round window membrane. This hearing loss is usually reversible with resolution of the effusion, but permanent conductive hearing loss can result from irreversible changes secondary to recurrent acute or chronic inflammation, such as adhesive otitis, tympanosclerosis, or ossicular discontinuity. Irreparable sensorineural loss may also occur, presumably as the result of spread of infection through the round or oval window.
membrane. Audiometry can be reliably performed in children over three years of age, but children under three years are the group at highest risk for effusions and associated hearing loss and in these patients standard audiometric assessment is difficult to perform reliably. Whenever an otitis media with effusion is diagnosed clinically or by tympanometry, there is a concurrent hearing loss. The relation between persistent or episodic hearing loss and impairment in the cognitive, language, and emotional development of children has been reported. However, the degree and duration of the hearing loss required to produce such deficits have not been defined.

Adhesive otitis media is the result of a healing reaction following inflammation of the middle ear. The mucous membrane is thickened by proliferation of fibrous tissue, which frequently impairs the movement of the ossicles, resulting in a conductive hearing loss. The condition is irreversible, making prevention of middle ear inflammation the most effective means of reducing the incidence of this condition, since medical and surgical treatment after adhesive otitis media is present has been disappointing.

Tympanosclerosis is characterized by the presence of whitish plaques in the tympanic membrane and nodular deposits in the submucosal layers of the middle ear. Pathologically there is hyalinization with deposition of calcium and phosphate crystals. Conductive hearing loss may result from embedment of the ossicles in the deposits. As in adhesive otitis media, a prerequisite seems to be chronic middle ear inflammation. Prevention has been the only successful means of controlling this disease.

Ossicular discontinuity is the result of rarefying osteitis secondary to chronic middle ear inflammation. The most common ossicle involved is the long process of the incus but the crural arch of the stapes, body of the incus, or manubrium of the malleus may also be eroded. The conductive hearing loss resulting from this complication frequently can be corrected surgically.

Suppurative Complications and Sequelae

The intratemporal suppurative complications and sequelae secondary to otitis media are perforation of the tympanic membrane, acquired cholesteatoma, mastoiditis, facial paralysis, and suppurative labyrinthitis.

Perforation of the tympanic membrane most frequently occurs with spontaneous rupture of the central portion of the eardrum during a bout of acute otitis media with effusion. Persistent purulent otorrhea may follow, in which instance a culture should be obtained, if possible, from the middle ear, and appropriate antimicrobials administered accordingly. In addition, antibiotic-cortisone otic medication may be helpful. Healing of the tympanic membrane frequently follows cessation of the suppurative process. A central perforation that fails to heal spontaneously and which is associated with a dry middle ear and good eustachian tube function may be amenable to surgical closure of the perforation with a graft - tympanoplasty. However, if the otorrhea persists, or if the drainage seems to be coming from an apparent posterosuperior or attic (pars flaccida) perforation, then a cholesteatoma should be suspected. Chronic suppurative otitis media with mastoiditis is represented by a perforation of the tympanic membrane or a cholesteatoma in which there is a persistent or episodic
purulent discharge. The most common pathogenic organisms are the gram-negative bacilli, such as *Bacillus proteus* and *Pseudomonas aeruginosa*. Aural polyps, which appear as red friable masses, may protrude through one of these perforations and may indicate the presence of a cholesteatoma. Treatment consists of active medical or surgical management, or both.

*Acquired cholesteatoma* is a sac-like structure lined by keratinized, stratified, squamous epithelium with accumulation of desquamating epithelium or keratin within the middle ear. Clinically, white, shiny greasy debris accompanied by a foul-smelling discharge can be observed in either the posterosuperior or an attic portion of the tympanic membrane and is a sequela of otitis media or atelectasis, or both. Tympanomastoid surgery is indicated, and if it is delayed the disease can invade and destroy other structures of the temporal bone and spread to the intracranial cavity.

*Mastoiditis* or inflammation of the mastoid air cell system frequently accompanies acute and chronic otitis media with effusion. Radiographic examination of this condition reveals a cloudy mastoid. The process is usually reversible as the effusion resolves with appropriate medical management. Occasionally, a severe acute otitis media with effusion is accompanied by mastoiditis in which there is pain, tenderness, edema, and erythema of the postauricular area. The pinna is displaced inferiorly and anteriorly, and swelling or sagging of the posterior-superior canal wall may also be present; this is the stage of mastoid periostitis. It is treated with immediate tympanocentesis / myringotomy and systemic ampicillin, with possible later adjustment of medication according to the antibiotic susceptibility of the organism. If the condition progresses to the stage of rarefying osteitis, the infectious process may break through the cortex of the mastoid to form a subperiosteal abscess. The infection may also break through the mastoid tip into the neck (Bezold's abscess) or fistulize into the external ear canal. At this stage, when osteitis is present, mastoid surgery is required to prevent further intratemporal or intracranial complications. *Petrositis* may result from acute or chronic infections of the pneumatized apical and perilabyrinthine cells of the temporal bone. The triad consisting of otitis media with effusion, paralysis of the homolateral external rectus muscle, and pain in the homolateral orbit or retro-orbital area, constitutes *Gradenigo syndrome*.

*Facial paralysis* secondary to involvement of facial nerve may occur during an episode of acute otitis media with effusion. Exposure of facial nerve may occur during an episode of acute otitis media with effusion. Exposure of the nerve caused by a congenital bony dehiscence within the middle ear is thought to be pathogenic mechanism. When it occurs as an isolated complication, a myringotomy should be performed and parenteral antibiotics administered. The paralysis will usually improve rapidly without requiring further surgery (ie, facial nerve decompression). Mastoidectomy is not indicated unless mastoid osteitis is present as demonstrated by the clinical and roentgenographic appearance of the mastoid. However, immediate surgical intervention is indicated when a facial paralysis develops in a child who had chronic suppurative otitis media with or without cholesteatoma.

*Suppurative labyrinthitis* may occur during an episode of acute otitis media with effusion from the direct invasion of bacteria through the round or oval windows. When chronic suppurative otitis media is present the infection may penetrate the windows or enter through a pathologic fistula of the bony horizontal semicircular canal. Signs and symptoms
include vertigo, nystagmus, tinnitus, hearing loss, nausea, and vomiting. Treatment consists of intense parenteral antimicrobials; however, surgical labyrinthectomy may be indicated to prevent spread to the intracranial cavity.

**Intracranial suppurative complications** of acute and chronic otitis media are meningitis, focal encephalitis, brain abscess, sinus thrombophlebitis, extradural abscess, subdural abscess, and otic hydrocephalus. Today, these complications occur more often in association with chronic suppurative otitis and mastoiditis, with or without cholesteatoma, than with acute otitis media with effusion. Infection can spread from the middle ear and mastoid to the intracranial structures by one of the following routes: vascular channels (osteothrombophlebitis), direct extension (osteitis), or preformed pathways, for instance, round window, previous skull fracture, and congenital or surgically acquired bony dehiscences. Clinically, any child who has an acute or chronic otitis media and who develops one or more of the following signs or symptoms, especially while receiving medical treatment, should be suspected of having a suppurative intracranial complication: persistent headache, severe otalgia, onset of fever, nausea, or vomiting. The following signs and symptoms demand an intensive search for an intracranial complication: stiff neck, focal seizures, ataxia, blurred vision, hemiplegia, intention tremor, papilledema, diplopia, past-pointing, dysdiadochokinesia, aphasia, or hemianopsia. Conversely, children with intracranial infection (recurrent meningitis or brain abscess) must have middle ear - mastoid disease ruled out as the origin of the disease. The life-threatening complications of middle ear disease in children are relatively uncommon. Our goal should be to reduce these complications still further by effective management of acute and chronic otitis media with effusion.

**Summary**

1. Otitis media is one of the most common diseases of childhood.

2. Pathogenesis is related to eustachian tube dysfunction.

3. Etiology is primarily bacterial (*S. pneumoniae*, 40 per cent; *H. influenzae*, 20 per cent). Bacteria are also present in chronic otitis media with effusion ("secretory otitis").

4. *H. influenzae* is present in all age groups, and 15 to 30 per cent are ampicillin-resistant.

5. Diagnosis is by pneumatic otoscopy, or tympanometry, or both.

6. Tympanocentesis and/or myringotomy is important diagnostic-therapeutic procedure in selected patients.

7. Ampicillin (or amoxicillin) is initial therapy of choice.

8. Erythromycin and sulfonamide, trimethoprim-sulfamethoxazole, or cefaclor is recommended for those who have poor clinical response to initial antimicrobial therapy.
9. Efficacy is yet to be shown for antimicrobial prophylaxis, decongestants, antihistamines, myringotomy and tympanostomy tubes, and adenoidectomy with or without tonsillectomy.

10. Attendant conductive hearing loss is probably related to abnormalities in cognition, language, and learning.
Assessment of Hearing

Thomas J. Fria


The assessment of hearing in infants and children is usually performed by an audiologist to achieve three main objectives: the identification of infants and children "at risk" for hearing impairment; the assessment of the nature and degree of such impairment; and the management of impaired children. This article focuses mainly on assessment and management, but a comment on identification is necessary since this area relies on the active participation of the primary care physician.

Identification of Hearing Impairment

The primary care physician often is the first to make contact with children who present with conditions that place them at risk for hearing loss, and can play a vital role in identifying children who require further testing to rule out hearing loss. A family history of childhood deafness, elevated bilirubin levels (greater than 20 mg per 100 mL of serum), maternal intrauterine infections (such as rubella, cytomegalovirus, herpes, and toxoplasmosis), craniofacial anomalies, low birth weight (less than 1500 gm), or neonatal asphyxia represent justifiable reasons for ruling out hearing loss. Approximately 1 in 1000 newborn babies has a hearing impairment, but the incidence is 30 to 40 times greater in newborn infants presenting with risk conditions.

Parental suspicion of hearing loss in a child should not be taken lightly; it may be the earliest indication of significant impairment in the child; in a child who has one or several risk conditions, it mandates referral for in-depth audiologic assessment. Hearing assessment must not be postponed because the child appears to be "too young" to test. There are adequate behavioral and nonbehavioral techniques to rule out hearing loss in any child, regardless of age.

Behavioral Assessment Techniques

The assessment of hearing with behavioral techniques involves the observation of the child's behavioral response to a given auditory stimulus. The nature of this behavioral response is age-dependent. Newborn and young infants respond typically with nonvolitional behavior, but older infants and children can respond with more volition. Consequently, the response repertoire of the child will dictate the most appropriate assessment technique.

Behavioral Observation Audiometry

For the purposes of this discussion an infant is defined as a child whose functional age is less than 24 months. The most appropriate assessment technique for infants is behavioral observation audiometry (BOA). As the name implies, BOA is a procedure whereby the examiner presents a stimulus sound and observes the associated response. Slightly modified versions of BOA that incorporate classic conditioning principles have been used successfully in 6 to 24 months old infants. BOA assesses the responses that are inherent to the infant: the
MORO, aural palpebral, arousal, and cessation reflexes. These reflexes are strongest in the infant who is less than five months of age.

The Moro reflex is a generalized motor response sometimes called the "startle reflex". In infants with normal hearing the startle reflex can be observed in response to rather intense (80 to 85 dB) stimulus sound. The aural palpebral or "eye blink" reflex may consist of either a rapid squint or blink in response to intense sound. In infants with normal hearing it requires a 105 to 115 dB stimulus. The arousal and cessation reflexes are somewhat self-explanatory; the sleeping infant is aroused and the restless infant is quieted by the presentation of an intense sound stimulus. Careful consideration should be given to the baby's baseline activity prior to stimulus presentation because the initial value of this activity can preclude a response from an infant who truly has normal hearing.

At approximately five months of age, the infant will begin to reflexively seek the source of sound (the "orientation" reflex or the "localization" response). This response begins as a rudimentary head turn in the horizontal plane and matures to a full eye and head turn in all planes by nine months of age. The localization response can extend the use of BOA to the assessment of older infants; moreover, this response is the basis for the conditioning modifications of BOA mentioned earlier. Conditioning modifications of BOA are best suited for assessing 12 to 24 month old infants, but successful results have been reported in the assessment of 5 month old infants.

The most familiar example of BOA is the cursory hearing screening that the primary care physician may use in the office setting. Many physicians have favorite noisemakers or other sound-producing devices (such as a hand clap) to use in such situations, and this form of BOA can influence the physician's decision to refer the child for further testing. An apparently normal response to noise-makers has reduced validity when accompanied by persistent parental suspicion of hearing loss. Noisemakers cannot detect hearing loss limited to the middle to higher frequencies, and if the baby has residual hearing he may respond to the device in an apparently normal fashion. A referral for further testing is justified whenever parental suspicion persists.

The level of background noise and the frequency and intensity of the auditory stimulus are controlled in the audiology center. Testing takes place in a sound-treated test booth with the child positioned on the parent's lap between two loudspeakers. The audiologist presents various auditory stimuli (voice, noise bands, frequency modulated pure tones or warble tones, and sometimes music and recorded environmental sounds) from an adjoining control room and observes the child's responses through a connecting window.

The intensity level of the auditory stimulus necessary to elicit an observable response will vary with the developmental level of the child. Wilber has compiled normative data from several studies on expected intensity level of auditory response in infants. The intensity level required to just elicit a response, regardless of stimulus type, decreases with increasing age. Responsivity is consistently better to the more meaningful speech stimulus than to the less familiar warble tones or noise bands.
**Conditioned Responses**

Reflexive responses begin to diminish in strength between 2 and 4 months of age, and the child becomes more responsive to softer and more meaningful sound such as the human voice. At 4 to 5 months of age the "localization" response begins to emerge, initially consisting of rudimentary attempts to locate the sound source, and, by 6 to 9 months of age, consisting of a full head and eye turn to the source of sound.

The localization response makes it considerably easier to evaluate infants who should have this capability with BOA. Moreover, certain conditioning techniques have been devised to strengthen this response in order to assess the baby's hearing. These techniques also involve the presentation of a stimulus sound and the observation of the associated behavioral response, but the response is strengthened or conditioned by using a reinforcer.

In 1961, Suzuki and Ogiba suggested the use of a blinking light located near the loudspeaker to reward the "orientation" or localization response in young infants. This technique is commonly referred to as conditioning orientation reflex audiometry (CORA).

Visual reinforcement audiometry (VRA) is a refinement of CORA that uses an interesting visual stimulus (such as an animated toy) to reward the child's localization response. Localization behavior is established during the conditioning phase of VRA, then the intensity level of the test stimulus is decreased until the child no longer responds. Some researchers have reported successful threshold estimation with VRA for babies as young as 5 months of age.

Tangible reinforcement operant conditioning audiometry (TROCA) is another conditioning procedure that uses a tangible reinforcer (such as raisins or other edibles) to reward the child for pressing a bar in response to an auditory stimulus. A special apparatus is used to dispense the reward when the bar is pressed. Visual reinforcer operant conditioning audiometry (VROCA) is a variant of the TROCA procedure which uses a visual reinforcer instead of a tangible reinforcer to reward the bar-press response.

Although operant conditioning techniques have been found to be most successful in 13 to 20 months old infants, they have also been used successfully in infants less than 12 months of age and have been shown to have particular value in the assessment of severely retarded children.

**Play Audiometry**

Once the child attains a developmental level of 2.5 years he is capable of voluntary responses to sound. Conventional audiometric techniques can be used, but they must be modified to be more interesting to the young child. The situation is structured to make listening a game, and the child actually responds by "playing" the game. For this reason the technique is often called "play" audiometry. This method can be used to assess hearing levels for both speech and pure tone stimuli (air and bone conduction). A game used frequently is to have the child drop a block into a box each time the tone is heard. With a short practice session, the child can be taught to respond appropriately as stimulus frequency and intensity are varied.
Hearing levels for speech stimuli can be obtained by using a picture or object identification task in which the child is requested to pick up or point to the item named by the examiner. Several picture identification tests (such as the Word Intelligibility by Picture Identification (WIPI) test) have been standardized for assessing speech discrimination ability in the young child or the child with limited speech and language skills.

**Conventional Audiometry**

The child 5 years and older can learn to perform for audiologic testing in a conventional manner. Conventional testing is conducted in a sound-treated test room with stimuli presented monaurally through earphones. Routine procedures include pure tone testing, speech reception threshold, and speech discrimination testing.

Pure tone testing is designed to assess the child's hearing threshold for the pure tone frequencies most important for understanding speech (250 to 8000 Hz in octave steps). The pure tone signals are presented through earphones (air conduction), then through a bone vibrator positioned on the mastoid (bone conduction). The patient is instructed to respond by raising a hand or pressing a button each time the tone is heard.

Speech reception threshold testing is designed to assess the child's threshold for simple speech material such as familiar two-syllable words with equal stress on each syllable (spondaic words). The child is instructed to repeat the words heard.

In addition to threshold level tests, a suprathreshold speech discrimination test is administered to determine the competency of the child's understanding for conversational level speech. Standardized lists of monosyllabic words serve as stimulus items, and the child is instructed to repeat the words heard. For younger children, pediatric word lists have been devised (for example, the PBK series).

Regardless of the developmental age of a child, some information can be obtained concerning auditory function using behavioral audiometric techniques. Modification of conventional audiometric methods provides the means to evaluate children who are very young or difficult to test. In view of the various techniques available, audiologic assessment should never be deferred because a child is too young or too difficult to test.

**Nonbehavioral Assessment Techniques**

The results of behavioral assessment techniques are unreliable in certain infants and children because they require the overt response of the child. Nonbehavioral techniques rely instead on the responses of the autonomic nerves system or on the electric responses of the child's auditory nervous system.

**Autonomic Responses**

The tests that measure autonomic responses to sound include respiration and cardiac audiometry. Respiration audiometry relies on measurable changes in the pattern and rate of respiration associated with an acoustic stimulus. The test involves the comparison of post stimulus respiration rate to the individual's base rate. Two popular methods of detecting
respiratory changes are: the use of a temperature-sensitive thermistor placed within 1 cm of
the subject's nostril which monitors the amount and duration of change in air pressure via a
change in temperature; and monitoring changes in the diameter of the rib cage on inspiration
and expiration by a special device strapped to the thorax which is so sensitive that the
slightest respiratory movement is registered. Recent studies have shown that respiratory
audiometry can estimate hearing sensitivity for both air and bone conducted stimuli. The
technique appears to work best on infants less than 12 months of age. However, the influence
of various procedural and subject related variables is not clearly defined and awaits further
investigation.

Cardiac audiometry involves the monitoring of changes in heart rate in response to
auditory stimulation. The method requires a source of auditory stimulation (stimulus
generator), EKG electrodes, and a recording system that identifies each heart beat and
measures the interval between successive beats. A simple electrocardiogram can identify each
beat but more involved instrumentation, such as a strip chart recorder that displays the
distribution of heart beats over time or a cartiotocometer, is required to measure the interval
between beats.

Several variables influence the success of cardiac audiometry. For example, the change
in heart rate is significantly dependent on the age and the state of arousal of the subject.
Schulman found that normal term babies show an acceleration in heart rate following the
onset of an acoustic stimulus when awake but a deceleration in rate when asleap. Premature
babies, however, show only an acceleration in heart rate until about 35 weeks' gestational age,
when the response pattern begins to look more like that of a term baby. At a developmental
age of 3 months, the cardiac response is more characteristic of the decelerative adult pattern.
In addition, respiration can influence the heart rate by causing a spontaneous variation in heart
beat; with inspiration the heart beat rises and with expiration the heart beat falls.

Although cardiac audiometry may provide a valid index of hearing in adults and older
children, its applicability to the neonatal population remains uncertain. Gerber, Mulac, and
Swain found that the parameters of the cardiac response to sound vary considerably between
and within infant subjects. The maturational influences or the response remains uncertain, and
the optimal acoustic stimulus has yet to be defined.

Auditory Electric Responses

Auditory electric response audiometry monitors stimulus-related changes in the
electrical activity of the auditory pathway from the cochlea to the auditory cortex. These
auditory "evoked potentials" (AEPs) comprise component responses that occur at different
times following the stimulus onset; early components correspond to cochlea and eighth cranial
nerve activity (electrocochleography), and later components reflect successively higher levels
in the auditory tract.

With the exception of electrocochleography (which requires a needle electrode passed
through the tympanic membrane), AEPs can be recorded from the scalp with noninvasive
electrodes. The activity recorded in this fashion is filtered, amplified, then analyzed by a
special purpose computer.
In our experience, the response component best suited for infants and children is called the auditory brain stem response (ABR). It consists of five to seven positive waves that correspond approximately to major sections of the auditory pathway. The ABR occurs in the first 10 msec following the onset of a brief acoustic stimulus such as a click.

The threshold of the ABR is determined by reducing stimulus intensity to a level that just elicits a repeatable response. For a click stimulus, this threshold provides a reasonable approximation of hearing for the frequency range from 1000 to 4000 Hz. Other types of stimuli can be used to estimate hearing in lower frequencies, but the reliability and validity of these estimates remain uncertain.

The ABR is a valuable adjunct to the assessment of hearing in infants and children. Its use is reserved to either establish a baseline response in children who cannot be tested behaviorally or to clarify the interpretation of ambiguous results of behavioral assessment. The test is noninvasive, and the response itself is immune to the effects of varying mental state and/or sleep (either natural or induced). The test takes approximately one hour for a complete evaluation. One critical requirement is that the child must lie very still to avoid obliteration of the response by muscle artifact. This means that most infants and children below six years of age require sedation for the ABR test. This requirement, in addition to the need for experienced personnel to interpret the results, confines the use of the ABR to major medical centers devoted to pediatric assessment.

Assessment of Middle Ear Function

Acoustic Impedance

Middle ear disease may result in an abnormal increase (or decrease) in the resistance a sound encounters at the tympanic membrane. This resistance is a complex interaction of the properties of mass and stiffness of the middle ear system and is more appropriately called "impedance".

A reasonable approximation of this acoustic impedance can be gained by delivering a sound to the ear and measuring the degree to which this sound is reflected back away from the tympanic membrane. If a great deal of the energy is reflected, the impedance at the membrane is high; conversely, if very little energy is reflected, impedance is low. High impedance can correspond to the middle ear abnormalities, such as otitis media with effusion, that are likely to increase impedance. The sound situation may correspond to conditions that abnormally reduce impedance, such as ossicular discontinuity.

Consequently the measurement of acoustic impedance at the plane of the tympanic membrane permits certain inferences to be made about the condition of the middle ear. The instrumentation used for such measurement is shown diagrammatically. A "probe" tip with three small apertures is sealed in the external auditory meatus. Each aperture serves a separate function. The incident sound or "probe" tone is delivered through one, the second leads to a microphone, and the third provides an avenue for varying air pressure in the meatus. The most familiar name for this device is an acoustic impedance "bridge". An approximation of the absolute acoustic impedance for a given ear is obtained by "balancing" the bridge. In other words, a reference level of sound energy built into the bridge is varied until it matches that
reflected from the tympanic membrane, and the corresponding impedance value (in ohms or cubic centimeters of equivalent volume) is read from the device.

**Tympanometry**

Tympanometry is a measure of "relative" acoustic impedance because it monitors the change in impedance associated with the variation of an air pressure load on the tympanic membrane. A positive air pressure (for example, 200 mm H2O) is applied to the tympanic membrane through the probe tip aperture, then the pressure is released gradually toward atmospheric pressure and reapplied in a negative direction to as much as - 600 mm H2O. The associated change in impedance can be observed on a meter or displayed on an X-Y plotter or strip chart recorder with the abscissa and ordinate respectively representing air pressure and relative impedance. The trace itself is called the tympanogram.

The tympanogram will peak when the air pressure on both sides of the tympanic membrane is approximately the same. Consequently, the position of the peak on the abscissa represents the air pressure in the middle ear. The height of the peak denotes the relative impedance at the tympanic membrane corresponding to the air pressure in the middle ear; sometimes this is called tympanic membrane compliance. The shape or gradient of the peak reflects the responsiveness of the tympanic membrane; a smoother or rounded peak denotes a more sluggish responsiveness to pressure changes than a sharp, well-defined peak. These three features of the tympanogram - peak position, height, and gradient - contribute useful information in the clinical setting.

**The Acoustic Middle Ear Muscle Reflex**

The stapedius and tensor tympanic muscles contract reflexibly in both ears, then an intense acoustic stimulus is presented to either ear. This contraction is called the acoustic reflex and it results in an increase in acoustic impedance at the tympanic membrane. Consequently the acoustic reflex can be detected by monitoring impedance just prior to and during adequate acoustic stimulation. The sudden increase in impedance coincident with the stimulus represents the contraction of the middle ear muscles. Reflex tests can be conducted by placing the probe tip in the meatus and stimulating either the same ear or the contralateral ear.

It is likely that middle ear disease will obviate detection of the acoustic reflex. Since the condition has already influenced the acoustic impedance, further increases in impedance secondary to muscle contraction may not be detectable. Moderate conductive and severe sensorineural hearing loss may also result in a failure to detect the acoustic reflex.

**Clinical Interpretation of Impedance Data**

Tympanometry and, in certain instances, acoustic reflex tests provide an essential adjustment to audiologic assessment of infants and children. The information provided can clarify the clinical picture by excluding or confirming middle ear disease.

Tympanometry is ideally suited for the office situation: the test is non-invasive, rapid, and can be administered by minimally trained personnel. However, the equipment is relatively
expensive, and many commercially available versions of the basic impedance bridge have not been validated. This can put the clinician at a disadvantage, especially when interpreting the data obtained.

Table 2 lists several variants of the tympanogram and the pressured middle ear conditions associated with each variant. The variants represent changes in peak position, height, and gradient in various combinations. Consequently, simplified interpretation can be based on these tympanogram features. Jerger and Paradise et al have classified tympanograms into distinct categories to further simplify interpretation.

Table 2. The Relationship between Tympanogram Variations and the Condition of the Tympanic Membrane and/or Middle Ear

<table>
<thead>
<tr>
<th>Pressure</th>
<th>Mobility</th>
<th>Presumed tympanic membrane / middle ear condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Normal</td>
<td>Low</td>
<td>Middle ear effusion, and/or thickened tympanic membrane, and/or ossicular fixation</td>
</tr>
<tr>
<td>Normal</td>
<td>High</td>
<td>Flaccid tympanic membrane or ossicular discontinuity</td>
</tr>
<tr>
<td>Negative</td>
<td>Normal</td>
<td>High negative pressure with or without middle ear effusion</td>
</tr>
<tr>
<td>Negative</td>
<td>Low</td>
<td>Middle ear effusion, and/or thickened tympanic membrane, and/or ossicular fixation</td>
</tr>
<tr>
<td>Negative</td>
<td>High</td>
<td>Flaccid tympanic membrane and high negative pressure (or ossicular discontinuity and high negative pressure)</td>
</tr>
<tr>
<td>Positive</td>
<td>Normal</td>
<td>High positive pressure with or without middle ear effusion.</td>
</tr>
</tbody>
</table>

Because otitis media significantly alters the tympanogram, tympanometry is very sensitive to its presence. The majority of ears with otitis media with effusion (OME) present with tympanograms that have a rounded peak of reduced height and are elevated in the negative pressure range. The absence of a peak is another common finding.

The validity of the tympanogram depends on the age of the child. The technique is less valid in infants less than seven months of age, presumably because of the excessive ear canal distensibility in these babies.

Management

The nature of hearing impairment can be conductive, sensorineural, mixed (conductive and sensorineural), or central. The degree of loss can range from minimal to profound, and the impairment can be unilateral or bilateral. The onset of loss can be prelingual (before speech is acquired) or postlingual. Successful management is a function of all these factors in addition to other handicaps the hearing impaired child may sustain.

Unilateral Hearing Loss

The child with unilateral hearing loss, either conductive or sensorineural, will have difficulty in at least two listening situations: localizing the source of auditory signal, and hearing in the presence of noise. Localization ability can be enhanced by training the child
to take advantage of other sensory avenues, especially vision; discrimination between foreground signals and background sounds can be improved by structured practice in listening in the midst of increasingly interfering background sound. Classroom and family adjustments include placement of the child with his good ear toward the primary source of instruction and close to the speaker to that background sounds are at a minimum, and teching the child to "stage-manage" himself so that his good ear is close to the speaker.

If hearing loss in the poorer ear can be improved with amplification, then a hearing aid may enhance binaural hearing. If not, a hearing aid that provides a contralateral routing of signals (CROS) may be helpful. CROS hearing aids have been helpful especially to individuals who have a sudden onset of hearing loss in one ear.

Because parents, and even the unilaterally handicapped child himself, may be concerned that the normal ear will acquire a hearing loss, judicious medical and audiological monitoring is imperative. Excessive exposure to noise (amplified music) and sudden changes in pressure (scuba diving) should be avoided. The detection and treatment of recurring ear infections and the maintenance of good general health are equally important.

**Bilateral Hearing Loss**

Conductive Loss. A hearing threshold elevation caused by middle ear disease, usually OME, is probably the most common problem encountered by the pediatric otolaryngologist. Hearing levels may range from 0 to 40 dB. Usually the child is not a candidate for a hearing aid unless the reduction in hearing sensitivity is expected to persist or if hearing fluctuates drastically during treatment. Amplification can be provided, especially to the child who is developing language and speech, until his hearing returns to normal.

In general, the child with a conductive loss merely requires louder auditory signals. By knowing the degree of the child's hearing loss, parents can inform the classroom teacher of the child's needs and can convey the child's need for special classroom consideration while the middle ear disease is active. The teacher should know that hearing may fluctuate and that there will be times when the child may need extra help in hearing instructions given to the class. The teacher will find that communication is enhanced if he or she gets the child's attention before giving an instruction, speaks clearly and distinctly in giving instructions, confirms that the child did understand the instruction accurately, does not talk while facing the blackboard, and rephrases the instruction, rather than merely repeating it, if the child does misunderstand. The child should know that he or she may miss what the teacher says. The child needs help to pay strict attention to the teacher's instruction. It is his responsibility to get the message. Parents and teachers must be careful, however, not to exaggerate the handicap a child may experience from mild reduction of hearing sensitivity. Usually, the brighter the youngster, the less handicap a mild conductive hearing reduction will impose.

When a child has a reduction in hearing sensitivity caused by a congenital malformation of the middle ear and sustains a conductive hearing impairment that is not amenable to surgery, amplification should be recommended. If the ears are dry, an air conduction hearing aid can be used. The aid can be either an ear-level or a body worn instrument. If the hearing reduction is 65 dB or less, the hearing aid can correct the loudness reduction in the ear in which it is worn. This leaves the child with only a unilateral hearing
loss, and he can be treated as having essentially normal hearing for learning. Binaural amplification is another alternative. The classroom and parental adjustment suggested earlier is appropriate for this child, too. If the ears are not dry or if the child has congenital atresia, a bone-conduction receiver worn with a body hearing aid can be recommended.

**Sensorineural Loss.** Children with a sensorineural hearing loss require more extensive management. Their problem is not only elevated hearing threshold, but distortion of sound so that intelligibility of speech may be reduced.

The management process for these children begins with parent education. Quite often, parents are more prone to accept the use of a hearing aid when they recognize that their child's hearing loss cannot be corrected medically. The more severe the hearing reduction, the more amenable parents are to the use of amplification.

Once parents have accepted the reality of irreversible hearing loss, they often grasp at the hope that the hearing aid will "correct" the serious impairment. However, hearing aids to not correct severe sensorineural hearing problems. The audiologist and hearing therapist must use considerable skill in helping parents and the hearing impaired child to adjust to the benefits and limitations of a hearing aid. The nature and extent of hearing loss, intellectual factors, and parental acceptance and support influence the success of amplification. Many well planned programs to provide hearing aid orientation are available to children, parents, and teachers.

**Amplification**

A personal hearing aid is a miniature, sound-reproducing system comprised of a microphone, amplifier, and receiver. The microphone picks up the sounds in the environment and changes them from acoustic to electric signals. The amplifier boosts the strength of the signals, and the receiver converts the amplified electric signals back into sound waves and sends them into the ear. An ear mold serves to focus the amplified sound into the ear.

**Body hearing aids** have a microphone, amplifier, and battery in one package from which a cord carries the electrical signal to the receiver worn at the ear (air conduction receiver) attached to a custom-made earmold which hold the receiver as close to the ear as possible. The cord may be attached to a small vibrator (bone conduction receiver) worn behind the ear and held on the head with a headband. Body worn hearing aids generally provide more amplification and are suitable for those with severe and profound hearing reduction or for infants whose ears are too tiny to hold an ear-level instrument.

**Ear-level hearing aids.** sometimes called over-the-ear or post-auricular aids, have the microphone, amplifier, receiver, and battery in a single package. The sound from the receiver is delivered to the ear by means of a tube which is attached to the custom-made earmold worn in the ear. Ear-level hearing aids are by far the most frequently used instruments by both adults and children.

**In-the-ear hearing aids.** All parts of the aid are built into the custom earmold or are in modular form and can be plugged into the mold. Generally, in-the-ear aids have less gain than ear-level instruments and are somewhat more noticeable than pictured in the
advertisements. They are generally inappropriate for children because of the need to have the earmold re-made as the ear grows.

Sensorineural hearing impairment causes both a reduction in loudness of incoming auditory signals and distorted and blurred audition. Hearing aids increase the loudness and to some extent can provide selective amplification in the frequency range of the hearing reduction. Because hearing aids are small, they cannot provide the high-fidelity associated with sound systems generally, and they certainly cannot provide "normal" hearing. Also, the ear with distortion cannot be perfectly "corrected" with a hearing aid; the user must learn to accommodate to distortion and rely on other sensory avenues (speech reading, environmental clues, facial expressions, and body language) for accurate reception of sound messages.

The hearing aid user with a sensorineural loss often finds that the device cannot distinguish useful from distracting sounds (signal versus noise). Consequently, the user needs time and usually training to psychologically "tune in" an important auditory signal and "tune out" unwanted noise.

The user with bilateral hearing loss who wears one aid has all the problems associated with unilateral hearing loss. Two hearing aids may provide truly binaural hearing which makes both localization and the speech-to-noise ratio more natural. Unfortunately, financial considerations can determine whether a child is to have one or two aids. There are children whose hearing impairment in each ear is so different that two aids cannot provide true stereophonic hearing. A competent audiologist will advise parents of the type of aid or aids best suited to the child's special needs. The audiologist may lend the child a second hearing aid for trial and instruct the parents on what to look for to determine the benefit of a second aid and warrant its purchase.

**Education**

Education for the hearing impaired child entails more than listening to sound. It includes the development of both receptive language (learning what is said) and expressive language (learning to express needs, thoughts, and ideas). In addition, it includes speech development or correction so the child can be understood by others. In general, severe hearing reduction has its greatest effect on language and speech. A hard-of-hearing person is one who, generally with the use of a hearing aid, has residual hearing sufficient to enable successful processing of linguistic information through audition. A deaf person is one whose hearing loss precludes successful processing of linguistic information through audition, with or without a hearing aid. Deaf and hard-of-hearing children are on quite different ends of a continuum and, depending on the hearing impairment, a variety of educational aids may be required.

Educational management is also affected by age of onset, nature of hearing loss, intellectual status, presence of other handicaps, and educational resources in the community, as well as parental understanding, expectancies, and ability to participate in the child's education.

When a significant hearing loss is identified in infancy, the child's home is the school setting and the parents are his teachers. A special educator trained to work with very young children helps parents to learn to communicate with their hearing impaired child and helps
the child begin to use his residual hearing with amplification. The special educator plans an individual education program (IEP) for the child; parents are required to be involved with planning this program (Education for All Handicapped Children Act, Public Law 94-142).

Nursery programs for hearing impaired children are located in large metropolitan areas in which there are enough children to constitute a class. Many infant stimulation programs designed for developmentally delayed youngsters have personnel trained to work with hearing impaired children. It seems best, however, that a severely hearing impaired child attend a school designed for his educational needs rather than a program for children with widely different problems.

Special schools are available in large communities for hearing impaired children who are two years of age and can use full-time special education. If the child lives too far from such programs, he may need to attend a residential school. All states have at least one residential school for the deaf. Parents should visit all existing facilities and judge them on the basis of class size (a school's classes for deaf children should not exceed seven youngsters), the availability of supervision, a sufficient number of classes to allow a homogeneous grouping of pupils and teacher training to teach hearing impaired children. The choice of school often must be based on geography. Yet, with the current right-to-education movement, many small school districts are developing classes for hearing impaired children. Parents should be warned, however, that it is a disservice to a child to place him in an inferior educational program in order to keep him close to home.

The child who is hard-of-hearing and who, with the aid of amplification can learn language and speech will probably have educational resources within the public school system. These may include a self-contained classroom for the youngster who cannot be placed in regular classes, or a special resource room to which the child goes for part of his day with a special teacher, while the rest of the day is spent in regular classes with his normal hearing peers. The special teacher tutors him academically so that he can compete with the children in regular classes. Some hearing impaired children can manage in regular schools with the help of a hearing therapist who sees the child once or twice a week for academic tutoring, speech-reading training, auditory training, and speech correction. The therapist can serve as a consultant to the regular classroom teacher, helping the teacher to accommodate the hearing impaired child in the classroom.

Hearing impaired children who have other handicaps, such as mental retardation, specific learning disabilities, or severe visual handicaps, require more than the special education designed for the hearing impaired child. Placement in a class is often determined by the child's most severe problem. The children most likely to be integrated into regular classrooms, even though they require supportive help, are those whose parents can participate in the child's education, and those youngsters whose language facility, including reading, is not seriously abnormal.

**Educational Methods For Deaf Children**

There are differences of opinion among educators as to how to educate the hearing impaired child, especially the deaf child. Should he be taught with an oral approach - learning to lipread, talk, and use his residual hearing? Or should he learn manual communication skills
sign language and finger spelling? The communication mode used by the school which the child attends will determine how he is to be educated. If a child cannot learn orally, a "total communication" (simultaneous oral-manual approach) may be feasible. No approach is a panacea for the difficult job of language education for a deaf child. The audiologist must inform parents of the conflict of approaches and make recommendations based on the child's hearing levels, language aptitude, and the parents' wishes.

Parents of Hearing Handicapped Children

The physician and audiologist have the unique responsibility to teach parents who, in turn, teach their hearing impaired child, about why he has a hearing handicap, the benefits of hearing aids, how the aids work, how to listen, how to take advantage of visual clues, and how to stage-manage himself for ease of communication. However, the most important goal is to help the parents and their child to maintain good mental health. The child needs to know that he is important. Parents need help to deal with their guilt feelings, grieving, lack of self-confidence in facing the realities of an irreversibly hearing handicapped child, frustration at well-meaning friends and family who give free advice that may not be helpful, and disappointments with professionals and hearing aids that cannot "cure" the problem. Being part of a parent group may be helpful. Well-informed and knowledgeable parents provide the handicapped child with the support and self-esteem he needs to feel that he is not the cause of his parents' grief. The goal of the physician is to constantly search for ways to meet the needs of parents who will in turn, provide a healthier climate in which their hearing impaired child can grow and learn.
Sinusitis and its Complications in the Pediatric Patient

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Criteria for the diagnosis and specific therapy of acute sinusitis have not been developed for the pediatric age group. How then, does one distinguish a viral upper respiratory infection (requiring at most symptomatic treatment) from secondary bacterial infection of the sinuses (probably requiring specific antimicrobial therapy)? The continuity of the nasal and sinus membranes would seem to favor a simultaneous inflammatory response when a viral infection or offending allergen is present. Factors that prevent or obstruct the drainage of respiratory secretions are probably important in determining the likelihood of bacterial superinfection.

Knowledge of the embryology, anatomy, and physiology of the sinuses is essential to understanding the onset and progression of clinical sinus infection.

Embryology and Anatomy

All of the paranasal sinuses develop as outpouching of the nasal chamber with varying extensions into their respective bony vaults. The openings or ostia of each sinus differ in size and configuration. These differences contribute to the relative susceptibility of each sinus to inflammatory and infectious process. The general architecture of the nasal chamber and nasopharynx and the dynamic aspects of facial growth also contribute to the relative risk for involvement of each sinus.

The nasal chamber is formed by cartilage anteriorly and bone posteriorly. The alar cartilages and the caudal end of the septum form the entry way into the nose. They then give way laterally to the portion formed by the medial wall of the maxillary and ethmoid bone. The medial wall of the nasal chamber is the septum; the floor of the nose is formed by the palatal bone, and the superior border of the nasal cavity is the perpendicular plate of the ethmoid bone. Posteriorly this chamber opens into the nasopharynx. Along the lateral wall of the nasal chamber are three shelf-like structures - the inferior, middle and superior turbinates. Beneath each turbinate is the corresponding meatus. The frontal, maxillary, and anterior ethmoid sinuses open in the middle meatus; the sphenoid and posterior ethmoid cells open high in the nasal vault into the superior meatus.

The maxillary sinus develops early in the second trimester of fetal life as a lateral outpouching in the posterior aspect of the middle meatus. At birth it is a slit-like structure with its long axis parallel to the attachment of the inferior turbinate and its floor barely below that. The sinus cavity grows in width and height. Laterally, it has reached the infraorbital foramen by age one and passes beneath it by age two. Ultimately at full size the lateral border of the maxillary sinus will reach the lateral orbital rim. The position of the floor of the sinus is determined by the eruption of the dentition. The sinus floor remains above the nasal floor until age eight or nine. In adults, the maxillary sinus floor can be as much as 5 to 7 mm below the nasal floor depending on the dental anatomy. The connecting route between the maxillary sinus and the nose is the primary maxillary ostium, a duct, often almost a
centimeter long. Accessory maxillary ostia have also been described resulting from
dehiscences of the nasal wall; these are generally not observed before age four. Infrequently
one can also find septae in the maxillary sinus, resulting in separate compartments rather than
a single large cavity. The volume of the fully developed maxillary sinus is approximately 12
to 15 mL.

The ethmoid sinus develops in the fourth month of gestation. It is not a single large
cavity but a grouping of individual cells, 3 to 15 in number, each with their own opening, or
ostium. Aeration of the ethmoid cells is variable leading to a honeycomb radiographic
appearance. The cells are small anteriorly and large in the posterior group.

The boundaries of the ethmoid labyrinth include the anterior cranial fossa superiorly,
the sphenoid sinus posteriorly, the orbit laterally, the nasal chamber and the middle turbinate
inferomedially, and the lacrimal bone anteriorly. The walls of the ethmoid labyrinth are thin,
especially in the lateral aspect bordering on the orbit. The lateral wall of the ethmoid sinus
is referred to as the lamina papryacea (“paper wall”). Purulent infection may spread by direct
extension from the ethmoid sinus through natural dehiscences in the bone to involve the orbit.

The variability of frontal sinus development is well known. In adults, 80 per cent will
have bilateral but often asymmetric frontal sinuses, 1 to 4 per cent will have agenesis of the
frontal sinuses, and the remainder will have unilateral hypoplasia. The frontal sinuses develop
either from an anterior ethmoid cell or directly from an anterosuperior evagination of the
nasal chamber. The position of the frontal sinus is supraorbital after age four but is not
distinguishable from the ethmoid sinus until 6 to 8 years of age. After that, it progresses for
another 8 to 10 years before reaching full adult size. Depending on the particular cell in the
frontal recess or anterior ethmoid sinus which develops into the frontal sinus, the conduit
between sinus and nasal cavity will be either a short and wide ostium or a long and narrow
nasofrontal duct.

Although the sphenoid sinus occupies a strategic position in the base of the skull, its
slow growth and relative isolation preserve it from frequent infection. Isolated involvement
of the sphenoid sinuses is uncommon; however, they may be involved as part of a
pansinusitis.

**Physiology**

Airflow through the nose allows for warming, humidification, and filtering of inspired
gases. This flow is affected by septal deflections, turbinate enlargement, and obstructing
masses such as polyps or adenoids.

The mucous membrane that lines the sinus cavities is pseudostratified ciliated
columnar epithelium with goblet cells and submucosal glands. It is continuous with the
respiratory epithelium that lines the nasal cavity. The mucus blanket secreted by the goblet
and submucosal cells entraps particulate antigens. As the sinus cilia beat toward the ostia, the
mucus and entrapped material are expelled into the nose. Interference with ciliary activity or
obstruction of the sinus ostia will result in retained secretions.
Ostial size and configuration are important determinants of drainage. Although the maxillary sinus ostium is relatively large, there may be a 1 cm isthmus connecting the nasal cavity and maxillary sinus. In addition, the position of the maxillary sinus ostium high on the medial border of the sinus cavity prevents gravitational drainage of secretions. The drainage of each ethmoid cell independently into the either the middle or superior meatus is aided by gravity. However, the minute openings of the individual ostia are easily obstructed. The frontal sinus opening can be either large with a short course or narrow with a tortuous nasofrontal duct through the anterior ethmoid complex.

Obstruction of the sinus ostia occurs in acute sinusitis because of generalized hyperemia, edema, and copious mucus. If secondary bacterial infection supervenes, there is an increase in acute inflammatory cells and superficial erosion of the lining epithelium. Measurements of oxygen tension in the maxillary sinus secretions have been uniformly low during acute infection. In addition, the pH is also reduced and carbon dioxide content increased. This may contribute to ciliary dysfunction and impaired phagocytic activity.

**Signs and Symptoms**

Common symptoms of sinusitis in adults are facial pain, headache, and fever. In children over five years of age, sinusitis is signaled in a similar fashion to that in adults. These older children may complain of headache (retro- or supra-orbital pain) and facial pain or swelling, usually with a history of a recent upper respiratory infection. In the younger child, there are often less specific complaints. Nasal symptoms (purulent discharge and congestion) and cough, when they are protracted (> 7 days) or unusually severe, may indicate sinus infection. The characteristics of the cough are variable - dry or wet, night or day. A history of malodorous breath in the absence of pharyngitis or dental decay may also be a clue to the diagnosis of acute sinusitis in the preschooler.

Physical examination may contribute to the diagnosis of acute sinusitis in the older child but is rarely an aid in the younger child. The swelling and erythema of the nasal mucosa and turbinates are not specific. Cervical adenopathy is usually not impressive, fever is variable, and transillumination is difficult both to perform and interpret. If mucopurulent discharge is seen draining from the middle meatus or if facial swelling or tenderness is appreciated overlying the ethmoid, maxillary, or frontal sinuses, then sinusitis is a probable diagnosis.

**Diagnostic Methods**

When the history or physical examination suggest the diagnosis of acute sinusitis, several techniques are available to confirm or support the clinical impression.

**Transillumination**

Transillumination may be helpful in diagnosing inflammation of the maxillary or frontal sinuses. Two methods exist for assessing transillumination of the maxillary sinus. In adults, transillumination has received mixed reviews as a diagnostic aid in sinusitis. The technique is helpful if light transmission is normal or absent. "Reduced" transmission or "dull" transillumination are assessments that correlate poorly with clinical disease. The accuracy of
transillumination has not been evaluated in children. The increased thickness of both the soft tissue and bony vault in youngsters may limit the clinical usefulness of transillumination.

**Radiography**

In adults with acute maxillary sinusitis, the reliability of radiographic evaluation as a diagnostic tool has been repeatedly demonstrated. The standard radiographic views are an anteroposterior, occipitomental, and lateral projection of the skull. A reduction in the air content of the sinus cavity (semi or complete opacification), accumulation of fluid, or thickening of the mucous membranes are the radiographic changes that are appreciated in acute inflammation.

As a rule, and with only rare exceptions, radiographically normal sinuses can be expected to be fluid free. Completely opaque sinuses will have free fluid or pus by aspiration in 80 to 88 per cent of patients. In sinuses with thickened mucous membranes but central aeration, fluid is present approximately 50 per cent of the time. In general, the greater the mucous membrane thickening, the more likely that fluid is present. Diagnostic accuracy in predicting fluid can be increased in patients with mucous membrane thickening by doing an occipitomental projection in lateral position with the affected side downward.

In chronic maxillary sinusitis, the most frequent radiographic finding is thickening of the mucous membrane; retention cysts and polyps are occasionally seen.

In children, the accuracy of sinus radiographs as a measure of sinus infection has not been validated. Caution concerning radiographic interpretation has been advised by Caffey. He comments that the redundant sinus mucosa and even tears in infancy may produce radiographically opaque sinuses, which are not infected. Similarly, he cautions that asymmetry in facial bone or sinus development and/or overlying soft tissue swelling can produce differences in the apparent aeration of the sinuses and lead to misinterpretation of the radiographs. Other investigators have cast doubt on the diagnostic specificity of radiographs in acute sinus disease in children by demonstrating a similar prevalence of radiographic abnormalities in the sinuses of healthy children, as compared with the sinuses of children with suspected sinus infection. Those who defend the diagnostic value of radiographs in children with acute maxillary sinusitis have failed to document the infection by sinus aspiration. A study correlating clinical signs and symptoms of acute sinusitis with radiography and sinus aspirates is essential to establish the credibility of radiographic findings in the pediatric age group.

**Ultrasonography**

Two recent reports have evaluated ultrasonography as a diagnostic aid in maxillary sinusitis. The advantages of ultrasonography versus radiography are the use of non-ionizing radiation and better ability to discriminate between mucosal thickening and retained secretions. In a study of adult patients, Mann compared radiography, sinoscopy, and ultrasonography. Ultrasonography was found to be particularly useful in predicting the presence of fluid in patients whose radiographs showed partial or complete opacification. Conformity between the findings at ultrasound and irrigation was observed in 90 per cent of patients. Similarly, a study of 61 children ages 3 to 12 compared ultrasonography to
radiography and sinus aspiration. The agreement between irrigation findings and A-mode ultrasound was 96 per cent. Use of sinus ultrasonography at the Children's Hospital of Pittsburgh has revealed some technical and diagnostic difficulties. More experience will be required to accurately assess the value of ultrasonography in the diagnosis of sinusitis, particularly in children less than three years of age.

**Sinus Aspiration**

A positive culture of an aspirate of sinus secretions is the sine qua non for diagnosing bacterial sinusitis. Although by no means a routine procedure, aspiration of the maxillary sinus - the most accessible of the sinuses - can be easily accomplished in an outpatient setting with minimal discomfort to the patient. Puncture is best performed by the transantral route with the needle directed beneath the inferior turbinate through the lateral nasal wall. This route for aspiration is preferred in order to avoid injury and permanent damage to the natural ostium. If the patient is unusually apprehensive or too young to cooperate, a short-acting narcotic agent can be used for sedation.

Careful sterilization of the puncture site is essential to prevent contamination by nasal flora. Ten per cent cocaine applied intranasally will achieve mucosal anesthesia and antisepsis. Lidocaine should be injected into the submucosa at the site of the actual puncture. Secretions obtained by aspiration should be submitted for cell count, gram stain, culture and sensitivity. A bacterial colony count will assure that the culture results reflect actual sinus infection rather than contamination; counts of $10^4$ or greater colony forming units per mL give a high degree of assurance of in situ infection. Alternatively a gram stain preparation of sinus secretions may be performed. Bacteria in low colony count (likely contaminants) will not be seen on smear.

Indications for sinus aspiration in patients with suspected sinusitis include: clinical unresponsiveness to conventional therapy; sinus disease in an immunosuppressed patient; severe symptoms such as headache or facial pain; and life-threatening disease at the time of clinical presentation.

**Microbiology**

The use of various techniques to obtain, transport, and culture maxillary sinus secretions has resulted in differing and often contradictory reports regarding the microbiology of sinusitis. Failure to describe the patient population studied or eliminate patients with partial antimicrobial therapy further complicates comparison of various investigations. However, a knowledge of the bacteriology of secretions obtained directly from the maxillary sinus by needle aspiration (with careful avoidance of contamination from mucosal surfaces) is a necessary guide to specific antimicrobial therapy.

The role of anaerobic bacteria as pathogens in sinusitis has only recently been examined with adequate attention to anaerobic transport and culture techniques. Poor drainage of the inflamed sinus results in a lower pH and oxygen pressure, thereby providing an excellent environment for the growth of anaerobic bacteria. However the growth of anaerobic bacteria may be impaired in sinus secretions obtained by irrigation since irrigation raises the oxygen pressure and dilutes bacterial titers. Finally, few studies have looked for viral agents
as a cause of sinus infection despite evidence that viruses alone may produce acute sinus disease.

**Acute Sinusitis**

Two studies, performed 15 and 30 years ago, using specimens obtained by aspiration of free fluid from the maxillary sinus of adults, showed a predominance of *Streptococcus pneumoniae* and *Haemophilus influenzae* in patients with acute or chronic sinus infections. The routine comparison and excellent correlation of gram stains of the fluid with culture results strengthens the validity of these investigations, although quantitation of the bacteria was not done. More recent reports echoed these results and also noted a significant minority of infections to be caused by anaerobes, again in both acute and chronic cases. The most convincing data reported, in two elegant studies with careful attention to bacteriologic technique, show nontypable *H. influenzae* and *S. pneumoniae* to be the most commonly found pathogens, accounting for approximately 65 per cent of all significant bacterial strains recovered. Other bacteria implicated include *Neisseria* species, *Streptococcus pyogenes* (group A), and alpha hemolytic streptococcus. Mixed infection with heavy growth of two bacteria was occasionally found, although most cultures grew only a single organism. Viruses were recovered from 11 of the 70 positive specimens; there were six isolates of rhinovirus, three of influenza A, and two of parainfluenza virus. Five of these 11 specimens also had significant growth of bacteria.

A recent study performed in children with acute maxillary sinusitis has shown the bacteriology of sinus secretions to be similar to that found in adults. The predominant organisms include *S. pneumoniae*, *H. influenzae* (nontypable), and *N. catarrhalis*. Several viral isolates including adenovirus and parainfluenzae were recovered.

**Chronic Sinusitis**

Three studies performed with adequate microbiologic methods have examined the bacteriology of chronic sinusitis in adults. Anaerobic bacteria were the predominant pathogens in two of the studies, but *H. influenzae* and *Streptococcus viridans* predominated in the third. One of these studies also found a light growth of *Staphylococcus aureus* in a significant number of specimens. These discrepancies may be explained by differences in the subjects studied: some had chronic sinus disease for many years; others, more likely, had acute exacerbations of chronic conditions.

The single report of probable chronic sinus infection in children reviewed the bacteriology of sinus irrigations in patients undergoing myringotomy and tube placement. Unfortunately the patient population was poorly described and neither quantitation of bacteria nor gram stains of secretions were performed. The report suggests that *H. influenzae* may be the most frequent bacteria recovered in chronic maxillary sinus infection in youngsters.

**Surface Cultures**

It would be desirable to culture the nose, throat, or nasopharynx in patients with acute sinusitis if the predominant flora isolated from these surface cultures was predictive of the
bacteria recovered from the sinus secretions. A particularly rigorous study in adults demonstrates that there is no correlation between cultures obtained from the anterior nares or nasal vestibule and cultures of the sinus aspirate in acute maxillary sinusitis. Similarly, in a study of pediatric patients with acute maxillary sinus infection, a comparison of the predominant flora in nasopharyngeal and throat cultures (processed semiquantitatively) with the bacteria isolated from maxillary sinus secretions showed no correlation. Comparison of results of cultures obtained from the middlemeatus (with a nasal speculum) and those obtained from the sinus secretions shows a better correlation than the studies previously cited; however, the authors state that "in an individual patient with sinusitis it is more reliable to base therapy on the results of previous bacteriologic investigations than on the individual bacterial findings in the nose". We agree with this statement and cannot recommend the use of surface cultures as a guide to the bacteriology and therapy of acute sinusitis.

**Treatment**

Therapy for acute maxillary sinusitis in the preantibiotic era consisted of sinus aspiration and irrigations. The current availability of numerous antimicrobial agents, to which the bacteria recovered from sinus secretions are susceptible, prompts consideration of antimicrobials in lieu of multiple irrigation procedures in the treatment of sinus infection. The objectives of antimicrobial therapy of acute sinus infection are sterilization of the sinus secretions, prevention of suppurative orbital and intracranial complications, achievement of a rapid clinical cure, and prevention of chronic sinus disease.

Conflicting reports appear in the literature regarding efficacy of antimicrobials in the treatment of acute sinus infection in adults as judged by radiographic resolution and findings at subsequent irrigation procedures. An array of antimicrobial agents and varying dosage schedules make comparisons of different studies difficult and discrepancies hard to explain. However, several points emerge: (1) Appropriate antimicrobials eradicate susceptible microorganisms in sinus secretions whereas inappropriate agents fail to do so. (2) In order to accomplish sterilization of the sinus secretions, a level of antimicrobial agent exceeding the minimum inhibitory concentration of the infecting microorganism must be present in the sinus secretions. (3) In some instances in which adequate antimicrobial levels within sinus secretions are documented, sterilization of secretions is still not accomplished. This observation points to the importance of local defense mechanisms (such as ciliary activity and phagocytosis) which may be impaired in the altered environment within purulent sinus secretions (decreased partial pressure of oxygen, increased carbon dioxide pressure, and decreased pH). Therefore, irrigation and drainage of sinus secretions may be required in some patient. (4) There does appear to be a decrease in the serious suppurative orbital and intracranial complications of paranasal sinus disease consequent to the use of systemic antimicrobials. (5) No controlled prospective studies comparing treatment regimens have been performed in the pediatric age group.

**Antimicrobials**

Until the results of controlled, prospective studies comparing treatment regimens are available, medical therapy with an antimicrobial agent is recommended in children diagnosed to have acute maxillary sinusitis. The relative frequency of the various bacterial agents suggests that ampicillin (100 mg/kg/day in four divided doses), or amoxicillin (40 mg/kg/day
in three divided doses) are appropriate agents. In areas in which ampicillin-resistant organisms
are prevalent or when the patient is allergic to penicillin, or when there has been an apparent
antibiotic failure, several alternative regimens are available. The combination agent
sulfamethoxazole-trimethoprim (prescribed on the basis of 40 and 8 mg/kg/day respectively
divided into two doses) has been shown to be efficacious in acute maxillary sinusitis in adults.
It is important to remember, however, that this agent may be ineffective in patients with
group A streptococcal infections. The new oral cephalosporin cefaclor (prescribed at 40
mg/kg/day in three divided doses) may be another substitute agent. Likewise, the combination
of erythromycin-sulfasoxazole (prescribed on the basis of 50 and 150 mg/kg/day in four
divided doses) is also suitable.

**Decongestants and Antihistamines**

The effectiveness of antihistamines or decongestants or combination antihistamine-
decongestants applied topically (by inhalation) or administered by mouth in patients with
acute or chronic sinus infection has not been adequately studied. Limited investigation of
specific agents in clinical rhinitis have shown that some produce a decrease in nasal
resistance. However, in the one study performed on patients with sinorhinitis, oral
phenylpropanolamine did not significantly increase the size of the maxillary ostium. The
effectiveness of any of these agents on the ultimate course of the respiratory illness or the
incidence of complications has not been examined.

**Complications**

**Orbital Cellulitis**

Orbital cellulitis is the most frequent serious complication of acute sinusitis and
despite antimicrobial therapy is a potentially life-threatening infection. The orbit is susceptible
to contiguous infection from the paranasal sinuses because it is bordered on three sides by the
sinuses. The orbital cavity is separated from the ethmoid air cells only by the thin laminapapyracea,
and natural bony dehiscences are commonly found. In addition, the ophthalmic
venous system has no valves; consequently, the extensive venous and lymphoid
communications between the face, nasal cavity, pterygoid region, and the sinuses allow flow
in either direction. These channels also connect directly with the cavernous sinus and
intracranial venous system.

There has been much confusion in the literature concerning infections of or about the
eye because of the frequent interchange of the terms inflammatory edema, periorbital,
preseptal, and orbital cellulitis. The terms periorbital and preseptal refer to infection or
inflammation outside or anterior to the orbital contents. The septum is a connective tissue
reflection of periosteum which inserts into the eyelids and provides an anatomic barrier
protecting the orbit. Orbital cellulitis implies infection within or involving the bony confines
of the orbit. Inflammatory edema is tissue congestion caused by impaired venous drainage.

The child who appears with a "swollen eye" presents a difficult problem in differential
diagnosis for the clinician. Sinus infection is a major predisposing cause of a swollen eye but
there are other entities to be considered (Table 1). Infected periorbital lacerations,
conjunctivities, dacryocystitis, systemic or contact allergy, seborrheic or eczematoid
dermatitis, and nasal vestibular infections may cause swelling about the eye. A last important category of infections are those cases of *H. influenza* type B (HIB) periorbital or so-called preseptal cellulitis. These HIB infections usually occur in children less than two or three years of age. They are characterized by an abrupt onset, rapid progression, and systemic toxicity. The children have high fever and often the periorbital tissue has a violaceous, almost hemorrhagic discoloration. The area is markedly swollen and tender. The texture of the skin is altered and there is induration of the subcutaneous tissue. Examination of the globe will show no proptosis and intact extraocular movements. HIB is frequently recovered from the culture of the blood or tissue aspirate. Because of the young age of most of these patients, interpretation of sinus radiographs is difficult. Since most *H. influenzae* recovered from sinus aspirates are nontypable, the relationship of this acute bacteremic HIB infection to sinusitis is unclear and we prefer to categorize it separately. These children should receive antimicrobial therapy appropriate for potentially invasive HIB disease.

**Table 1.** Differential Diagnosis of "Swollen Eye" (Excluding Paranasal Sinus-Related Problems)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Periorbital laceration</td>
<td>Trauma should be obvious. Cellulitis is most often caused by group A streptococci or coagulase-positive staphylococci</td>
</tr>
<tr>
<td>or abrasion</td>
<td></td>
</tr>
<tr>
<td>Insect bite</td>
<td>The site of inoculation is usually evident. The surrounding area is nontender but often pruritic.</td>
</tr>
<tr>
<td>Allergy</td>
<td>Systemic allergy is usually not strictly unilateral. Contact allergy does not produce local or systemic signs of infection.</td>
</tr>
<tr>
<td>Conjunctivitis</td>
<td>The primary site of involvement is the conjunctiva, not the surrounding soft tissue. Slower progression; no systemic toxicity.</td>
</tr>
<tr>
<td>Dacrocystitis</td>
<td>Originates in medial lower lid. May progress to moderately severe cellulitis. Radiographic evidence of sinusitis is usually absent.</td>
</tr>
<tr>
<td>Dermatitis</td>
<td>Seborrheic or eczematoid skin changes apparent.</td>
</tr>
<tr>
<td>Nasal vestibular cellulitis</td>
<td>Cellulitis begins about nasal vestibular area and produces cellulitis edema of the lower eye lid prior to development of cellulitis in the upper lid or orbit.</td>
</tr>
<tr>
<td>Preseptal or periorbital</td>
<td>Intense infection in preseptal area usually caused by <em>H. influenzae</em> type b. There is often a violaceous or hemorrhagic appearance to the tense, indurated tissue about the eye.</td>
</tr>
</tbody>
</table>

**Classification and Treatment**

Once the diagnosis of an orbital complication of sinusitis has been clarified, the severity of involvement must be judged. The classification most frequently used in establishing the severity of the orbital cellulitis is shown in Table 2 (modified from Chandler). It is essential to establish the severity of the cellulitis clinically so that appropriate decisions can be made regarding specific therapy and the need for surgical drainage. The first stage of the Chandler classification, and the complication most often seen clinically, is inflammatory edema. This is not an actual infection of the orbit but rather swelling caused by impedance of the local venous drainage. As such, it is a warning of a potentially serious infection within the sinus. This inflammatory edema has also been referred to as "periorbital cellulitis". However, inflammatory edema in which the swelling is soft, nontender, with no induration
and only minimal cutaneous changes can be distinguished from true periorbital cellulitis caused by HIB.

**Table 2. Classification of Orbital Cellulitis**

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I Inflammatory Edema</td>
<td>Inflammatory edema beginning in medial or lateral upper eyelid; usually nontender with only minimal skin changes. No induration, visual impairment, or limitation of extraocular movements.</td>
</tr>
<tr>
<td>II Orbital Cellulitis</td>
<td>Edema of orbital contents with varying degrees of proptosis, chemosis, limitation of extraocular movement and/or visual loss.</td>
</tr>
<tr>
<td>III Subperiosteal Abscess</td>
<td>Proptosis down and out with signs of orbital cellulitis (usually severe). Abscess beneath the periosteum of the ethmoid, frontal, or maxillary bone (in that order of frequency).</td>
</tr>
<tr>
<td>IV Orbital Abscess</td>
<td>Abscess within the fat or muscle cone in the posterior orbit. Severe chemosis and proptosis; complete ophthalmoplegia and moderate to severe visual loss present (globe displaced forward or down and out).</td>
</tr>
<tr>
<td>V Cavernous Sinus Thrombosis</td>
<td>Proptosis, globe fixation, severe loss of visual acuity, prostration, signs of meningitis, progresses to proptosis, chemosis, and visual loss in contralateral eye.</td>
</tr>
</tbody>
</table>

Infection from the paranasal sinuses may actually spread into the orbit. This is signaled by intense pain, swelling, and fever accompanied by proptosis, limitation of extraocular movement, and impaired vision. If the infection infiltrates the orbit directly there will be a diffuse cellulitis. Alternatively, there may be a more local accumulation of pus - a subperiosteal or orbital abscess.

If the infection has progressed beyond stage I, then intravenous antimicrobials and hospitalization are mandatory. Cultures of the blood and sinus aspirate should be performed aerobically and anaerobically. A lumbar puncture may be indicated in addition to studies to identify intracranial suppurative complications. Decompression will be required if there is a subperiosteal or orbital abscess: however, orbital cellulitis may respond to antimicrobials without surgical intervention. The choice of antimicrobials should be guided by knowledge of the usual bacteriology of acute sinusitis. If the gram stained smear of the sinus aspirate or abscess drainage shows unsuspected organisms, appropriate antimicrobials should be added until the results of the cultures are known.

**Diagnostic Aids**

In most instances the cause of the swollen eye can be established by history and physical examination. When sinusitis is the underlying problem, sinus radiographs will be abnormal. The common radiographic patterns are partial or complete opacification, mucous membrane thickening, or an air-fluid level. Usually the ethmoid and maxillary sinuses are involved simultaneously; however, there may be a pansinusitis involving the ethmoid, maxillary, frontal, and sphenoid sinuses. Occasionally overlying soft tissue swelling may
confuse the interpretation of the radiographs. If there is diffuse swelling about the eye including the bridge of the nose, the underlying sinus may appear hazy, even without in situ inflammation. However, there will usually be either ipsilateral or contralateral clouding of the sinuses in areas without overlying soft tissue swelling; this should help to clarify the diagnosis.

Ultrasound scanning on the B mode is occasionally helpful in detecting abscesses in the posterior orbit. However, computerized tomography (CT) is especially useful in defining and localizing the extent of abscesses in the posterior orbit. Simultaneously, a detailed examination of the sinuses can also be obtained by doing additional views on CT. CT should be reserved for patients in whom abscesses are suspected or when orbital cellulitis has not responded, as expected, to medical therapy.

**Outcome**

Frank orbital complications of sinusitis are fortunately rare. If the diagnosis and appropriate therapy are not carried out promptly, vision may be lost. Severe neurologic sequelae or death may follow cavernous sinus thrombosis.

**Intracranial Complications of Acute Sinusitis in Children**

Intracranial extension of infection is the second most common complication of acute sinus disease. Although the incidence of suppurative intracranial complications in patients with sinusitis is unknown, paranasal sinusitis is the source of 35 to 65 per cent of subdural empyemas.

**Pathology and Pathogenesis**

Infection may enter the intracranial compartment by two routes. Direct extension may occur through necrotic areas of osteomyelitis in the posterior wall of the frontal sinus. The underlying dura becomes thickened with inflammatory exudate, and a heavy layer of granulation tissue develops on its outer surface, forming an extradural empyema. Although the dura is generally resistant to infection, bacterial penetration may take place along the course of the small vessels that traverse its thickness. This results in a subdural empyema which then excites an intense inflammatory reaction in the subjacent arachnoid. This direct route of intracranial extension is more commonly associated with chronic otitis infection than with sinusitis.

An alternative route of intracranial bacterial entry is provided by the valveless venous network which interconnects the intracranial venous system and the vasculature of the sinus mucosa. Thrombophlebitis originaing in the mucosal veins progressively involves the emissary veins of the skull, the dural venous sinuses, the subdural veins, and finally the cerebral veins. By this mode of spread, the subdural space may be selectively infected without contamination of the intermediary structure; that is, a subdural empyema can exist without evidence of extradural pus or osteomyelitis. Intracranial extension of the infection by the venous rute is common in paranasal sinus disease, especially in its acute phase or during an acute exacerbation of chronic inflammation.
Further intracranial spread of infection depends on the competence of the arachnoid as a barrier to bacterial invasion. Despite its thinness, the adult arachnoid is relatively impermeable to infecting agents, and bacterial meningitis only rarely complicates subdural empyema. In infants, however, infection may be transmitted freely across a presumably immature arachnoid, and the incidence of bacterial meningitis was 75 per cent in one series of subdural empyema.

Although subarachnoid infection secondary to subdural empyema is uncommon in young children and adults, extensive cortical thrombophlebitis is a frequent complication. The involved gyri are edematous and hyperemic, often showing small foci of infarction. Sometimes septic thrombosis of a major dural sinus occurs, resulting in massive bilateral cerebral edema and hemorrhagic infarction. This explains the appearance of seizures, focal neurologic deficits, and increased intracranial pressure in patients with only a seemingly insignificant amount of subdural pus. Inasmuch as cortical thrombophlebitis is less common in extradural empyema, focal signs are much rarer in this entity.

The mechanism of brain infection is more obscure. In the majority of abscesses secondary to sinus disease or subdural suppuration, the brain is adherent to the patch of inflamed dura. It is likely that an initial focus of ischemia or necrosis caused by cortical venous occlusion provides an ideal environment for the growth of anaerobic and microaerophilic organisms. Infection is then carried deeper into the white matter by the penetrating cerebral vessels. The resulting area of cerebritis then undergoes central liquefaction while the perimeter is gradually surrounded by a capsule made up of an inner layer of granulation tissue, a middle layer of collagen, and an outermost shell of glial cells. A "mature" abscess capsule takes two to three weeks to toughen. The fibroblasts responsible for capsule formation come from vessel walls. Since the deeper white matter is not as vascular as the cortex, the abscess wall near the ventricle is thinnest, and ventricular discharge of the purulent contents is not uncommon.

Four distinct sites of subdural empyema can be distinguished. The pus (1) may spread diffusely over the frontal-parietal convexity; (2) may be loculated into focal pockets anywhere over the hemisphere, but predominantly over the frontal pole and the occipital cortex (probably due to gravity in the supine patient); (3) may be interhemispheric (parafalcine), or (4) may be under the tentorium in the posterior fossa. Parafalcine suppuration often involves the opposite side by extending underneath the falx; the parafalcine space is also the favored site of recurrent infection.

**Clinical Features**

Many authors have noted a preponderance of males over females when reviewing suppurative intracranial complications of sinusitis. The peak age is between 10 and 20 years, although younger children are not immune. Four groups of symptoms and signs may be recognized:

*Signs of pansinusitis.* About 50 to 60 percent of patients with subdural empyema secondary to sinusitis present with signs and symptoms of acute frontal sinusitis or an acute exacerbation of a chronic pansinusitis. There is usually low grade fever, malaise, and frontal headache, often accompanied by marked forehead tenderness. Occasionally, subperiosteal pus
overlying the anterior wall of the frontal sinus may result in dramatic epicranial edema and a painful fluctuance (Pott's puffy tumour).

**Signs of increased intracranial pressure.** The initial headache worsens despite prolonged treatment with analgesics and oral antibiotics. Vomiting becomes intractable and the level of consciousness deteriorates gradually. This is usually a result of early cerebral edema in the area adjacent to the extradural or subdural pus. With an isolated extradural collection, cortical involvement is less severe and the patient usually remains alert. With subdural empyema, however, stupor and coma may supervene rapidly.

**Signs of meningeal irritation.** During the stage of depressed sensorium, there is usually evidence of meningeal irritation including nuchal rigidity and photophobia. This reflects an intense inflammatory response in the leptomeninges in contact with a subdural abscess rather than true septic leptomeningitis. Since signs of leptomeningeal inflammation are uncommon in pure extradural suppuration, a subdural accumulation should be suspected if protracted symptoms of fever and headache are accompanied by prominent signs of meningeal irritation.

**Focal neurologic deficits.** Focal neurologic deficits are usually caused by a combination of local brain compression and cortical venous thrombosis and infarction. A frontoparietal convexity subdural empyema causes contralateral brachiofacial weakness, contralateral conjugate gaze palsy, and expressive dysphasia. Lower limb involvement is usually late. Focal seizures involving the arm and face occur in over 60 per cent of patients with dorsolateral subdural empyema. With a parafalcine empyema, jacksonian seizures often begin in the foot and march upward to include the trunk, the arm, and finally the face. Weakness also primarily affects the lower extremity with sparing of speech and facial motor functions. Bilateral parafalcine collections may present with paraplegia simulating thoracic spinal cord compression.

In the terminal stage, the patient is comatose, hemiplegic, has evidence of generalized and meningeal sepsis, and shows signs of uncal or tonsillar herniation.

**Diagnostic Methods**

If the diagnosis of intracranial suppuration is suspected from clinical examination, a lumbar puncture should be deferred to avoid brain herniation. The usual cerebrospinal fluid findings associated with subdural or parenchymal abscesses consist of an elevated protein, pleocytosis with a variable neutrophil count, a normal glucose, and a sterile culture ("parameningeal" focus of infection).

Plain radiographs of the skull show evidence of pansinusitis in over 60 per cent of patients. Frontoethmoidal osteomyelitis is seen only when the sinusitis is chronic. In young children, suture diastasis may be the first clue to a raised intracranial pressure.

Both electroencephalography (EEG) and radionuclide brain scan are too nonspecific to have any diagnostic value. Although the EEG may demonstrate focal slowing and decreased amplitude, and radionuclide scan shows increased uptake over an area of empyema, 50 per cent of patients in several recent seris of subdural empyema have had normal EEGs.
and radionuclide scans. Moreover, it is impossible, on radionuclide scans alone, to distinguish subdural empyema from subdural hematoma, infarction, cerebritis, and brain abscess.

The typical angiographic findings of a subdural empyema are displacement of the cortical vessels from the inner table of the skull and a midline shift of the anterior cerebral vessels. Inward displacement of the superior sagittal sinus indicates the presence of an extradural collection. With a parafalcine lesion, the proximal portion of the anterior cerebral artery is shifted to the opposite side, whereas the distal portion is displaced to the ipsilateral side away from the falk (the "S" sign). A hypertrophied falcine artery may also be seen. However, small interhemispheric and subfrontal empyemas unable to produce significant vascular displacement have been missed with angiography, and it may be difficult to identify a coexisting parenchymal abscess when extracerebral vascular distortion is prominent.

CT is now recognized to be the most definitive test for the diagnosis of intracranial infection. It is relatively noninvasive, is able to define even small purulent collections with exact localization, and minimizes the risk of missing a concomitant brain abscess or bilateral empyemas. A parenchymal abscess shows up as a lucent center with an intensely enhancing capsule and surrounding edema. With extracerebral pus, the remarkable degree of underlying cerebral edema and midline shift distinguishes it from a subdural or an extradural hematoma.

Treatment

Treatment of the intracranial complications of sinus infection requires antimicrobials, drainage, and excellent supportive care. There is evidence that preoperative meningitic doses of antibiotics may improve survival. Since the predominant organisms isolated from subdural empyema secondary to sinusitis include anaerobic and microaerophilic streptococci, nongroup A streptococci, _Staphylococcus aureus_, and a mixture of Proteus and other gram-negative rods, the initial antibiotic regimen prior to culture and sensitivity results should be a combination of penicillin G, a penicillinase-resistant penicillin, and chloramphenicol. The recovered pus may be sterile owing to partial treatment given before the diagnosis made.

Hyperosmolar agents should be given if intracranial hypertension threatens brain herniation. Systemic steroid is prescribed with caution because of its theoretical ability to suppress granulocytic and immune functions. Anticonvulsants should be given prophylactically to protect against a 79 per cent incidence of associated seizures.

Extradural and subdural empyemas should be dealt with by a generous craniotomy. The entire collection of pus can be evacuated and the area profusely irrigated with bacitracin solution under direct vision, and, with a judiciously fashioned flap, the opposite parafalcine space can be explored. Following drainage of an extradural abscess, the dura should be opened to rule out an underlying subdural empyema, since even a well-enhanced CT scan cannot distinguish between a purely extradural empyema and one with an associated subdural empyema. Extradural and subdural drains are left in place for 3 to 5 days for continuous drainage and intermittent antibiotic irrigation. In most cases of subdural empyema, the underlying brain is so swollen that the bone flap must be left out for external decompression. All osteomyelitic bone must be debrided. The frontal sinus is opened widely, its contents exenterated, and its cavity drained.
An underlying brain abscess is best handled by initial intracapsular evacuation and catheter drainage. This minimized iatrogenically induced deficits associated with radical excision of deep-seated lesions located in eloquent areas of the brain, and reduces the problem of multiloculation commonly encountered in repeated percutaneous aspiration.

Postoperatively, the patient should be maintained on intravenous antibiotics for a minimum of 2 to 3 weeks. Intermittent antibiotic irrigation of the abscess or empyema cavities can be done through the catheters until their removal in 3 to 5 days. The shrinking of the abscess can be followed accurately by serial CT scans.

Despite modern diagnostic and surgical capabilities, the mortality associated with subdural empyema and brain abscess remains over 25 per cent. Causes of death and permanent morbidity are related to delayed diagnosis, recurrent suppuration, missed concomitant parenchymal abscesses, extensive cortical and dural sinus thrombophlebitis, and fulminant bacterial meningitis in infants. Early diagnosis remains the most effective way for improving survival.
Allergic Rhinitis

James Fagin, Roger Friedman, Philip Fireman


Allergic rhinitis is the most common of all allergic disorders, affecting over 20 million people in the USA. Because it is not life-threatening and its symptoms may not be incapacitating, allergic rhinitis may be ignored by the pediatrician. Yet this frequent illness causes significant morbidity which results in the expenditure of many millions of dollars in health care and the loss of millions of school and working days. The symptoms may be episodic or perennial; when symptoms recur annually during certain months, the syndrome is called seasonal allergic rhinitis, and when symptoms occur throughout the year, the syndrome is called perennial allergic rhinitis. Typically, seasonal allergic rhinitis does not develop until after the patient has been sensitized by two or more pollen seasons. Seasonal allergic rhinitis is frequently referred to as "hay fever" or "summer cold". These descriptive terms are misleading and should be discarded because fever is not a symptom associated with allergic rhinitis, and hay or the common cold virus are not incriminated in the etiology of this syndrome.

The prevalence of allergic rhinitis in the general population is considered to be about 10 per cent, with the peak incidence in the post-adolescent teenage child. Broder et al in the study of a well-defined population in Tecumseh, Michigan, found the prevalence of allergic rhinitis to increase from less than 1 per cent in infancy to 15 to 16 per cent after adolescence.

The term allergy was introduced by a pediatrician, Clemens von Pirquet, in 1906 to designate the host's altered reactivity to an antigen; the end result could be helpful or harmful to the host. The term allergy as commonly used in contemporary clinical practice indicates an adverse reaction. In this context, allergic rhinitis is best defined as the adverse pathophysiologic response of the nose and adjacent organs which results from the interaction of allergen with antibody in a host sensitized by prior exposure to that allergen.

**Symptoms and Signs**

Initial symptoms in seasonal allergic rhinitis progress from frequent sneezing and nasal pruritus to rhinorrhea and finally to nasal obstruction. These symptoms vary considerably from season to season and may also differ markedly at various times of night and day. Patients not only have nasal pruritus but also itching of the eyes, throat and ears. Many children constantly rub the nose with the hand or arm in an effort to relieve the nasal itch and perhaps to improve the nasal obstruction. Other children may press the palm of the hand upward against the nose in an "allergic salute", which often leads to the development of a transverse nasal crease. The patient with nasal obstruction will be a constant mouth breather and snoring will be a prominent nighttime symptom. Patients may report generalized malaise, irritability, and fatigue; these symptoms are often difficult to differentiate from the side effects of antihistamines frequently used for symptomatic therapy. The pattern of symptoms frequently distinguishes seasonal from perennial allergic rhinitis, especially in areas with obvious seasonal climatic changes.
With development of the allergic reaction, clear nasal secretions will be evident and the nasal mucous membranes will become edematous without much erythema. The mucosa appear boggy and blue-gray. The turbinates may appear to be congested and swollen. If nasal obstruction is present, it may be necessary to shrink the mucosa with a vasoconstrictor in order to document the absence of nasal polyps, which are relatively uncommon in allergic rhinitis, occurring in less than 0.5 per cent of patients. Patients, particularly children, with allergic rhinitis and significant nasal obstruction and venous congestion, may also demonstrate edema and darkening of the tissues beneath the eyes, the so-called "allergic shiners". The conjunctiva may also demonstrate a lymphoid follicular pattern with a cobblestone appearance.

**Etiology**

The development of allergic rhinitis requires two conditions: atopic familial predisposition to develop allergy, and exposure of the sensitized patient to the allergen. Inhalants are the principal allergens responsible for allergic rhinitis. These microscopic airborne particles include pollens, mold spores, animal danders, and environmental dust, either house or occupational. Seasonal allergic rhinitis is primarily induced by pollens from the germination of nonflowering vegetation. In the temperate climates, the most important are tree pollens in the spring, grass pollens in the late spring and early summer, and ragweed pollens in the late summer and early fall. In warm climates, mold spores may be airborne year round, but in climates in which snow and freezing occur in the winter months, airborne mold spores are present intermittently during the spring, summer, and fall until there is significant frost. In patients with perennial allergic rhinitis, mold spores may be a significant inhalant allergen indoors along with epidermal animal danders and house dust. The principal allergen in house dust has not been identified but may be attributed in part to the house dust mite, dermatophagoides. Food allergens are less important in the etiology of allergic rhinitis, but cannot be ignored, especially in young children.

Despite several extensive retrospective family and twin studies, there is no agreement as to the hereditary pattern in allergic diseases. Most investigators believe that several genetic loci are involved in the expression of allergic disease and that inheritance is multifactorial. Recent immunologic studies have isolated some of these genetic influences. Animal studies have shown that synthesis of specific antibodies to well characterized antigens is controlled in part by immune response (Ir) genes which are linked to the major tissue histocompatibility locus (HLA). The studies by Levine and co-workers have suggested that ragweed allergic rhinitis and immune responses to purified ragweed antigen E were linked to a particular HLA haplotype in successive generations of allergic families. Marsh et al reported a significant correlation between haploptotype HL-A7 and increased IgE antibodies, to a low molecular-weight purified ragweed antigen (Ra5) in a group of patients with allergic rhinitis who were sensitive to this small portion of the ragweed allergen.

**Immunopathophysiology**

Allergic rhinitis, along with allergic asthma and allergic urticaria, is described immunologically as an immediate hypersensitivity syndrome and is mediated in large part by immunoglobulin E (IgE) antibodies. The properties of IgE compared with the other serum and secretory immunoglobulins are shown in Table 1. IgE is normally present in minute quantities
compared with the serum immunoglobulins IgG, IgA and IgM. The IgE antibodies are synthesized after allergen challenge largely by plasma cells that are located in lymphoid tissues adjacent to mucosal membranes. These IgE antibodies passively sensitize the membranes of tissue mast cells and circulating blood basophils. The exact nature of the binding of IgE to mast cell and basophil cell membranes is not known, but it involves the Fc portion of the IgE molecule and an appropriate receptor in the cell surface. The combination of allergen and its specific IgE antibody results in a sequence of energy-dependent enzyme reactions with alteration of the mast cell membrane. This initiates the release and synthesis of the specific pharmacologic mediators of the IgE immediate hypersensitivity reaction. These mediators include histamine, the slow-reacting substance of anaphylaxis (SRS-A), eosinophil chemotactic factor (ECF-A), platele aggregation factor (PAF), and other kinins and vasoactive substances that cause the increased vascular permeability, local edema, and increased eosinophil-laden secretions seen in patients with allergic rhinitis.

Table 1. Properties of Human Serum and Secretory Immunoglobulins

<table>
<thead>
<tr>
<th></th>
<th>IgG</th>
<th>IgA</th>
<th>S-IgA</th>
<th>IgM</th>
<th>IgD</th>
<th>IgE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adult serum concentration (mg/mL)</td>
<td>10</td>
<td>2</td>
<td>-</td>
<td>1.5</td>
<td>0.03</td>
<td>0.0002</td>
</tr>
<tr>
<td>Antibody activity</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>?</td>
<td>+</td>
</tr>
<tr>
<td>Neutralization (viral, toxin)</td>
<td>+</td>
<td>±</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Anaphylactic (histamine release)</td>
<td>±</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Blocking antibody</td>
<td>+</td>
<td>±?</td>
<td>±?</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Maternal-fetal transfer</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Present in secretions</td>
<td>±</td>
<td>+</td>
<td>++</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Fix to mast cells (homocytotropic)</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Classic complement activation</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Alternate complement pathway</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>+</td>
</tr>
</tbody>
</table>

Histologic examination of the nasal mucosa will demonstrate distended goblet cells in the presence of enlarged, congested mucous glands. The tissues are infiltrated with eosinophils and with lymphoid cells that have a paucity of neutrophils. The intracellular spaces are enlarged, and the basement membrane is thickened. Mast cells are also present in the mucosal tissues, but the significance of their relative numbers has not been defined.

Using airway resistance studies and quantitative pollen challenges, Connell has shown that a larger dose of allergen was required to increase resistance in the nasal mucosa that had remained unchallenged than was required to obtain the same effect after a week of daily exposures. He also demonstrated that, in patients sensitized to several pollens, repeated challenges with one allergen conditioned the nasal mucosa to react to lower dose of the second allergen than would have been needed if given singly. This priming phenomenon could well account for the persistence of symptoms experienced by many patients toward the end of the pollen season, in spite of decreased exposure to allergen. It is also thought that this priming effect favors an increase in responsiveness to nonspecific stimuli such as changes in humidity and temperature.
Laboratory Studies

A variety of laboratory studies are useful to the clinician in establishing a diagnosis of allergic disease and in providing a guide to specific therapy. Several of these tests are enumerated in Table 2.

**Table 2. Laboratory Studies Helpful in Diagnosis of Allergic Disease**

<table>
<thead>
<tr>
<th>Nonspecific Studies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eosinophils</td>
</tr>
<tr>
<td>Blood (&gt; 600 eosinophils per mm$^3$)</td>
</tr>
<tr>
<td>Secretions (nasal, sputum, conjunctival)</td>
</tr>
<tr>
<td>Total serum IgE (age-related)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Specific Antigen-Antibody Studies</th>
</tr>
</thead>
<tbody>
<tr>
<td>In vivo (provocative) tests</td>
</tr>
<tr>
<td>Skin tests (20 min wheal and flare)</td>
</tr>
<tr>
<td>Scratch</td>
</tr>
<tr>
<td>Prick</td>
</tr>
<tr>
<td>Intracutaneous</td>
</tr>
<tr>
<td>Nasal inhalation challenge</td>
</tr>
<tr>
<td>Bronchial inhalation challenge</td>
</tr>
</tbody>
</table>

| In vitro serum tests              |
| RAST (radioallergosorbent)        |

The nasal secretions of patients with allergic rhinitis usually contain increased numbers of eosinophils. It is difficult to quantify nasal eosinophilia accurately. Greater than 3 per cent eosinophils seen on a nasal smear is considered to be an increase. Eosinophilia may not be present in patients who have not been exposed to specific allergens recently or in patients with a superimposed infection. Steroids can significantly reduce eosinophilia but antihistamine therapy has no significant effect on nasal eosinophils. Documentation of nasal eosinophilia is a simple, inexpensive procedure that can be performed easily in an office setting.

In certain patients it may be helpful to confirm the clinical impression of allergic rhinitis with documentation of specific IgE antibodies by in vivo skin testing or in vitro serum radioallergosorbent (RAST) testing. Skin testing with the suspected allergens is mandatory in all patients prior to initiation of immunotherapy. Clinicians should be selective in the use of allergens for skin testing and should employ only common allergens of potential clinical importance in their patients. To avoid false-negative skin tests, antihistamines should be withheld for 24 to 48 hours before skin tests are performed. Prick skin testing may be more reliable than intradermal skin testing; the specifics of such testing are outlined in standard allergy textbooks. Food skin testing should be reserved for patients who present diagnostic problems, with intermittent or perennial symptomatology. The major problems with skin testing, especially for food allergens, have been the lack of potency, stability, and purity of the allergen solution. The crude, undefined composition of allergens often produces false-positive reactions secondary to an irritating effect on the skin. Great care must be used in interpreting the results of food skin testing because there is often a discrepancy between the production of clinical symptoms after ingestion of the food and positive skin reactions to foods.
As mentioned earlier, the in vitro RAST for assessing the presence of serum IgE antibodies to various allergens has recently been employed as a diagnostic aid in allergic rhinitis. For certain allergens, RAST has been shown to be as reliable as skin tests; it is also more specific, although less sensitive, than skin tests. At present, skin testing is felt to be a more useful tool than RAST for the diagnosis of allergic disease. It is generally accepted that skin testing is less expensive, more sensitive, and technically easier to perform. Furthermore, there is a wider range of antigens available for skin testing, and the results may be interpreted immediately. RAST is indicated in the management of the allergic patient with generalized dermatitis and in the young infant in whom skin tests would be difficult to perform. Table 3 summarizes the relative advantages of RAST and skin tests in diagnosing allergic conditions.

Table 3. Relative Advantages of RAST Test and Skin Test in Diagnosis of Allergic Conditions

<table>
<thead>
<tr>
<th>Skin Test</th>
<th>RAST Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Less expensive</td>
<td>No patient risk</td>
</tr>
<tr>
<td>More sensitive</td>
<td>Specific and quantitative</td>
</tr>
<tr>
<td>More antigens available</td>
<td>Not affected by drugs</td>
</tr>
<tr>
<td>Results available promptly</td>
<td>Antigen stability</td>
</tr>
<tr>
<td>Technically easier</td>
<td>Patient convenience</td>
</tr>
<tr>
<td>May detect non-IgE mediated</td>
<td>Useful (dermatographism, widespread</td>
</tr>
<tr>
<td></td>
<td>dermatitis)</td>
</tr>
<tr>
<td>Good correlation with history</td>
<td>Good correlation with history and</td>
</tr>
<tr>
<td>and RAST tests</td>
<td>skin tests.</td>
</tr>
</tbody>
</table>

Because of the recent observations that IgE antibodies may be present in nasal secretions but not evident by skin testing or the presence of serum IgE antibodies, a nasal provocation test occasionally may be useful in assessing a patient with a negative skin test who is suspected of reacting to a particular allergen.

The in vitro cytotoxic leukocyte test for foods and other allergens has been advocated by some, but its usefulness as a laboratory test has not been confirmed in controlled studies by other investigators. Also, recent studies have employed the use of leukocyte inhibition factors and lymphocyte transformation to investigate the possible role of food allergy in patients with symptoms of allergic rhinitis but these tests have not yet been documented to be clinically relevant.

Differential Diagnosis

Children who present with rhinorrhea and nasal obstruction may have symptoms not only because of allergy, but also as the result of infections, foreign bodies, structural changes, pregnancy, drug reactions, neoplasms, or other nonallergic causes. The common cold virus is the most frequent cause of upper respiratory infection (URI), and at the outset a viral URI, with its symptoms of clear, watery rhinorrhea and sneezing, resembles allergic rhinitis. Redness of the nasal mucosa is characteristic of a URI and usually distinguishes it from allergic rhinitis. Nasal infections can be superimposed on allergic rhinitis.
<table>
<thead>
<tr>
<th></th>
<th>Allergic</th>
<th>Nonallergic</th>
<th>Nonallergic</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>ENR</td>
<td>Vasomotor</td>
<td></td>
</tr>
<tr>
<td>Usual onset</td>
<td>Child</td>
<td>Child</td>
<td>Adult</td>
</tr>
<tr>
<td>Family history of</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>allergy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Collateral allergy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Symptoms</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sneezing</td>
<td>Frequent</td>
<td>Occasional</td>
<td>Occasional</td>
</tr>
<tr>
<td>Itching</td>
<td>Common</td>
<td>Unusual</td>
<td>Unusual</td>
</tr>
<tr>
<td>Rhinorrhea</td>
<td>Profuse</td>
<td>Profuse</td>
<td></td>
</tr>
<tr>
<td>Congestion</td>
<td>Moderate</td>
<td>Moderate to marked</td>
<td>Moderate to marked</td>
</tr>
<tr>
<td>Physical examination</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Edema</td>
<td>Moderate to marked</td>
<td>Moderate</td>
<td>Moderate</td>
</tr>
<tr>
<td>Secretions</td>
<td>Moderate</td>
<td>Watery</td>
<td>Mucoaid to watery</td>
</tr>
<tr>
<td>Nasal eosinophilia</td>
<td>Watery</td>
<td>Common</td>
<td>Occasional</td>
</tr>
<tr>
<td>Allergic evaluation</td>
<td>Common</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Skin tests</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IgE antibodies</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Therapeutic response</td>
<td>Positive</td>
<td>Coincidental</td>
<td>Coincidental</td>
</tr>
<tr>
<td>Antihistamines</td>
<td>Positive</td>
<td>Coincidental</td>
<td>Coincidental</td>
</tr>
<tr>
<td>Decongestants</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Corticosteroids</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cromolyn</td>
<td>Good</td>
<td>Fair</td>
<td>Poor to fair</td>
</tr>
<tr>
<td>Immunotherapy</td>
<td>Fair</td>
<td>Fair</td>
<td>Poor to fair</td>
</tr>
<tr>
<td></td>
<td>Good</td>
<td>Good</td>
<td>Poor to fair</td>
</tr>
<tr>
<td></td>
<td>Fair</td>
<td>Unknown</td>
<td>Poor</td>
</tr>
<tr>
<td></td>
<td>Good</td>
<td>None</td>
<td>None</td>
</tr>
</tbody>
</table>

ENR = eosinophilic nonallergic rhinitis.

The above conditions can usually be differentiated from allergic rhinitis, but the separation of perennial allergic from perennial nonallergic rhinitis is often difficult. Nonallergic rhinitis can be classified as either eosinophilic nonallergic rhinitis (ENR) or vasomotor rhinitis (Table 4). ENR may occur in children, but is more common in adults. It stimulates the perennial type of allergic rhinitis, but no immunologic etiology can be implicated. In this disorder, the edematous mucous membranes are often pale and eosinophilia is always present, but the usual methods of detecting a specific allergen and its mediating antibodies fail to establish a specific cause. Vasomotor rhinitis is a nonallergic form of persistent nasal disease usually seen in older children and adults, which also is manifested by watery rhinorrhea and nasal obstruction. The patient reports over-responsiveness of the nose to minimal changes in air temperature, odors, and often to change in the position of the head. These patients appear to have unusual awareness of symptoms and their complaints are disproportionate to the magnitude of their symptoms. It is important to delineate patients with ENR and vasomotor rhinitis from patients with allergic disease because of their different responses to therapy. Immunotherapy is not to be used in patients with nonallergic disease,
and drug therapy with antihistamine decongestants control symptoms inconsistently. Recent evidence suggests that ENR, unlike vasomotor rhinitis, may improve significantly with either topical or systemic corticosteroids (unpublished data by authors).

**Therapy**

Successful therapy of allergic rhinitis involves three primary considerations: identification and avoidance of the specific allergens, pharmacologic management, and immunotherapy.

**Identification and Avoidance.** Complete avoidance of causative allergens is the best therapy for allergic disease because without exposure to allergens the allergic reaction will not take place. Elimination of exposure to an animal dander by elimination of the animal from the household, or elimination of a food allergen from the diet, may provide complete or partial relief of symptoms. Measures to control house dust, especially in the bedroom, can be effective treatment for certain patients. These measures include providing rubberized or plastic airtight enclosures for mattresses and box springs, the use of synthetic bedding fabrics, and the removal of stuffed toys, stuffed furniture, heavy drapery, and dust catchers, such as book shelves and record cabinets, from the bedroom. Electrostatic precipitrons can be installed in central forced-air heating and cooling systems, and these can substantially reduce not only house dust but pollens and other airborne particles. Because single-room electrostatic precipitron units are less effective and may generate irritating ozone, they are not recommended.

**Pharmacologic Management.** If the patient cannot completely avoid the allergen, symptoms can be controlled with drugs in many cases. Antihistamines are preferred for treating mild to moderate allergic rhinitis. Several groups of antihistamines differ in chemical structure and in action. Since the effectiveness of one group may diminish after several months or years of use, an antihistamine of another group may then be efficacious clinically. Therefore, the clinician should become familiar with the use of one or more antihistamines in each of the listed groupings (Table 5). When nasal obstruction by secretions is a prominent symptom, an alpha-adrenergic decongestant, such as phenylephrine, phenylpropanolamine, or pseudoephedrine, should be used individually or in combination with an antihistamine. Topical nasal alpha-adrenergic vasoconstrictors usually provide prompt symptomatic relief but should not be used for more than several days. After 7 to 10 days of using a topical decongestant many patients develop so-called rebound vasodilatation and, at times, habituation. It is necessary to discontinue nose drops in order to relieve this "rhinitis medicamentosa".

If symptoms cannot be controlled with antihistamines, decongestants, and avoidance of allergens, several clinicians have suggested corticosteroid therapy. In children, the risk-benefit ratio of treating even severe allergic rhinitis with oral or parenteral corticosteroids is sufficiently high to contraindicate their use. However, several topical corticosteroids, especially for short-term seasonal use, might prove to be useful in the future. Beclomethasone, a poorly absorbed and rapidly metabolized topical corticosteroid, is currently enjoying considerable popularity in Europe for the treatment of allergic rhinitis without severe or systemic side effects. Cromolyn sodium, which inhibits the release of mediators from mast cells, is another topical aerosol pharmacologic agent that is available in Europe but not yet available for the treatment of allergic rhinitis in the USA. Although European investigators
have found the drug to be effective in 75 per cent of patients treated, such high efficacy has not been reported in a recent study of patients with allergic rhinitis in the USA.

**Table 5. Oral Antihistamines**

<table>
<thead>
<tr>
<th>Trade Name</th>
<th>Oral Preparations</th>
<th>Dose (per 24h)</th>
<th>Max Sin Dos mg</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Ehylendiamines</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tripelennamine Pyribenzamine hydrochloride</td>
<td>Tablets, 25 and 50 mg; delayed 5 mg/kg action, 50 and 100 mg</td>
<td>50</td>
<td></td>
</tr>
<tr>
<td>Tripelennamine Pyribenzamine citrate</td>
<td>Elixir, 37.5 mg/5 mL</td>
<td>50</td>
<td></td>
</tr>
<tr>
<td><strong>Ethanolamines</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Diphenhydramine Benadryl hydrochloride</td>
<td>Capsules, 25 and 50 mg; elixir 12.5 5 mg/kg mg/5 mL</td>
<td>50</td>
<td></td>
</tr>
<tr>
<td>Carbinoxamine Clistin maleate</td>
<td>Tablets, 4 mg</td>
<td>0.4 mg/kg</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>Timed release tablets, 8 and 12 mg</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Elixir, 4 mg/5 mL</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Alkylamines</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chlorpheniramine Chlor-Trimeton maleate</td>
<td>Tablets, 4 mg</td>
<td>0.35 mg/kg</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>Timed release tables, 8 and 12 mg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Brompheniramine Dimetane maleate</td>
<td>Tablets, 4 mg</td>
<td>0.5 mg/kg</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>Timed release tablets 8 and 12 mg</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Triprolidine</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Actidil hydrochloride</td>
<td>Tablets, 2.5 mg</td>
<td>0.2 mg/kg</td>
<td>2.5</td>
</tr>
<tr>
<td></td>
<td>Syrup, 12.5 mg/5 mL</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Piperazines</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cyclizine Marezine hydrochloride</td>
<td>Tablets 50 mg</td>
<td>3 mg/kg</td>
<td>50</td>
</tr>
<tr>
<td><strong>Piperidines</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cyproheptadine Periactin hydrochloride</td>
<td>Tablets 4 mg</td>
<td>0.25 mg/kg</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>Syrup, 2 mg/5 mL</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Azatadine maleate Optimine</td>
<td>Tablets, 1 mg</td>
<td>0.1 mg/kg</td>
<td>22 div doses</td>
</tr>
<tr>
<td><strong>Phenothiazines</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Promethazine Phenergan hydrochloride</td>
<td>Tablets, 12.5, 25, and 50 mg</td>
<td>0.5 mg/kg</td>
<td>50</td>
</tr>
<tr>
<td></td>
<td>Syrup, 6.25 and 25 mg/5 m</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Methdilazine Tacaryl</td>
<td>Tablets, chewable, 3.6 mg</td>
<td>0.3 mg/kg</td>
<td>8</td>
</tr>
<tr>
<td></td>
<td>Tablets, 8 mg</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Syrup, 5 mg/5 mL</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Immunotherapy.** If symptomatic drug therapy and avoidance cannot adequately control symptoms or if drugs inadvertently provoke significant side effects, immunotherapy (hyposensitization) with allergen solutions may be indicated. The specifics of this therapy are discussed in greater detail in one of several allergy texts. Before proceeding with immunotherapy, the physician should institute a comprehensive investigation of the causative factors, and the patient’s history of symptoms should be closely correlated with the presence of specific IgE antibodies, determined preferably by skin test results or by an in vitro RAST. In several double-blind studies, immunotherapy or hyposensitization injections with solutions
of pollen have been shown to be effective in reducing the symptoms of allergic rhinitis. Immunotherapy may be expected to provide significant clinical improvement in 80 to 90 per cent of patients with pollen-induced allergic rhinitis. If improvement is not obtained after a 2 year trial of immunotherapy, the patients should be reevaluated and discontinuation of immunotherapy should be considered. Duration of immunotherapy injections in patients who achieve clinical benefits depends on the patient's overall clinical response. In the presence of clinical improvement, patients should be given the opportunity to stop the immunotherapy after approximately 5 years of injections. It has been claimed that immunotherapy in children for seasonal allergic rhinitis may reduce their chances of developing pollen-induced asthma but this report is open to question and has never been confirmed.

The mechanism by which immunotherapy promotes clinical improvement has not been precisely delineated. Laboratory evidence has shown that immunotherapy results in the development of IgG blocking antibodies, a decrease in specific IgE antibodies, and a reduction in leukocyte histamine release. Immunotherapy has also been shown to modulate T cell function by inducing the generation of antigen-specific T suppressor lymphocytes. The clinical correlation of these studies with efficacy of immunotherapy may in the future provide the clinician with means to better manage the patient with allergic rhinitis.
Immotile Cilia Syndrome

Gilbert A. Friday, Eduardo J. Yunis, Rocco M. Agostini


Immotility of the cilia in the respiratory tract has recently been described in individuals with chronic respiratory disease. Changes in the normal configuration of cilia are observable at the ultrastructural level by electron microscopy. In patients with Kartagener's syndrome (situs inversus, bronchiectasis, and sinusitis), there is a defect in mucociliary transport owing to functionally ineffective ciliary movement which has been called "immotile cilia" syndrome or, as suggested more recently, "dyskinetic cilia" syndrome. The dynein arms, which are normally found attached to the microtubular doublets, are absent or deficient (short "spur"). Dynein arms contain the ciliary adenosine triphosphatase protein dynein. Adenosine triphosphate induces active sliding of adjacent microtubular doublets resulting in ciliary movement. Other abnormalities described in patients with immotile cilia syndrome include absence of radial spokes even in the presence of dynein arms, transposition of ciliary microtubules, and loss of parallelism of the plane of arrangement of the central pairs of microtubules in adjacent cilia (normally within 25°).

Ciliated epithelia are present in the respiratory tract, the paranasal sinuses, the eustachian tubes, the ependyma lining of the ventricles of the brain and spinal cord, and the oviducts. Human sperm tails are modified cilia with identical ultrastructure and are subject to the same ciliary deficiencies. When occurring in sperm tails, sterility has been noted, but no sterility problems have been noted in women with the same cilia defects. Kartagener's syndrome appears to be of autosomal recessive inheritance. It is assumed that during early embryonic life, ciliary beats in the growing embryo determine the type of laterality. When ciliary movements are absent, laterality may develop fortuitously, thus affecting a situs inversus in about half the affected individuals.

Recurrent episodes of bronchitis with segmental atelectasis, sinusitis, nasal polyposis, otitis, and mastoiditis have all been noted in individuals with immotile cilia. The lining of the bronchial tree and nasopharynx with its communicating chambers (sinuses and middle ears) are normally covered with a fine carpet of mucus. This mucus is swept along by ciliary movement of the pharynx, then swallowed or expelled. Defective ciliary movement or dysfunction is seen in the absence of significant defects in immunoglobulins, complement, phagocytic granulocytes, and alveolar macrophages associated with recurrent and chronic infection.

Laboratory Studies

Children with histories of recurrent bronchitis, sinusitis, polyps, otitis media, and in some cases of bronchiectasis in which immunologic studies have been found to be normal, should be investigated for ciliary abnormalities. Immunologic studies should include evaluation of immunoglobulins, both quantitatively and qualitatively, assessment of the cellular immune system, complement screening, and evaluation of the phagocytic system. In some individuals, allergy evaluation is appropriate.
Nasal and bronchial biopsies and, in some instances, biopsies of the middle ear and sinus areas can be performed to demonstrate ciliary abnormalities. With a curette, biopsies of the mucosa of the inferior and middle conchae may be taken and examined within a few minutes by phase contrast microscopy. The nasal mucosa may show only a few areas with recognizable cilia, and no motile cilia may be seen. Electron microscopy studies of biopsies of ciliated respiratory mucosa (or sperm samples) can provide confirmation of the diagnosis of immotile cilia syndrome, but an adequate sample is necessary. Between 50 and 100 cilia with proper orientation in cross-sectioned profile should be studied before a diagnosis of immotile cilia syndrome is rendered. Dynein arms may not be visible on all of the outer doublets of normal cilia because of variations in orientation and fixation. Other ultrastructural findings may include cilia with multiple axonemes; an addition, deletion, or disorientation of an outer doublet; and ballooning of the outer ciliary membrane. However, these have not been shown to be diagnostic for immotile cilia syndrome.

**Differential Diagnosis**

The immotile cilia syndrome should be considered in the differential diagnosis of children with chronic upper and lower respiratory infections. Chronic bronchitis and bronchiectasis, although disruptive to respiratory epithelia, do not alter normal ciliary beating and function.

Mucociliary dysfunction can be seen in asthma. Tracheobronchial secretions may be difficult to expectorate and may contribute to bronchial obstruction. Impairment of mucociliary transport mechanisms have been noted, but the pathogenesis is still poorly understood. Elaboration of chemical mediators in the lungs seems to depress mucociliary function. The clinical pattern of acute bronchospasm should be helpful clinically, and ciliary ultrastructure will be found to be normal by electron microscopy.

Patients with cystic fibrosis have normal mucociliary clearance and ciliary ultrastructure. These patients tend to have more severe bronchial obstruction than patients with immotile cilia and have more discomfort from chronic rhinitis, sinusitis, and otitis media.

Various immunologic abnormalities must be carefully excluded by appropriate studies. Children with cellular immune abnormalities, complement defects, and phagocytic defects will also present differential diagnostic problems comparable to patients with immotile cilia.

**Therapy**

Since children with immotile cilia have difficulty with mucociliary transport and clearance, efforts can be made to help them clear their respiratory mucosa. Postural drainage and physical therapy may be helpful to prevent pooling of secretions. This should be attempted three or four times per day during episodes of lower respiratory tract infections. Administration of a bronchodilator prior to postural drainage probably will be of little benefit since bronchospasm is not a primary feature of this syndrome. On the other hand, when there is combined nonallergic asthma or even significant change in pulmonary function with bronchodilators, it can be helpful to maintain the individual on therapeutic levels of theophylline.
Since motile cilia are a primary defense mechanism in the sinuses, chronic sinusitis is likely to develop. The lower respiratory tract will tend to have chronic retention of mucus and inhaled material causing bronchi to dilate to a bronchiectatic condition. Preventive measures should include immunization against respiratory pathogens such as *Bordetella pertussis*, rubeola, influenza, and *Streptococcus pneumoniae*. Early detection of bacterial infection by culture must be available. Appropriate antibiotic therapy is, of course, indicated on an intermittent or even at times on a daily basis. Chronic sinusitis and bronchiectasis may require continual antibiotic therapy, rotating antibiotics to avoid the emergence of resistant organisms. Usually, antibiotics that are effective against Haemophilus species as well as pneumococci, including amoxicillin, ampicillin, or a combination of trimethoprim and sulfamethoxazole or cefaclor, are effective. Recurrent respiratory infections tend to cause these children to lose many days of school and separate them from their peers, resulting in significant educational and developmental problems. Ciliary immotility is compatible with a fairly normal life and in this age of antibiotics with a fairly normal life span. It will, however, likely lead to chronic bronchitis and eventually to obstructive changes in the airways.
Neck Masses in Children: Adenopathy and Malignant Disease

Basil J. Zitelli


Neck masses in children are a frequent occurrence and may tax the clinical acumen of even the most astute physician. The differential diagnosis is extensive, but generally includes congenital lesions, lymphadenopathy, and malignant masses. Although the fear of malignancy accompanies virtually every child with a neck mass, cancer of the head and neck comprises only about 15 per cent of such masses in children admitted to the hospital. A logical method of evaluation is needed to prevent becoming lost in a quagmire of possibilities. By reviewing anatomy and mode of presentation, and obtaining a detailed history and careful physical examination, the physician may often arrive at a proper diagnosis on clinical grounds and confirm it with appropriate laboratory examination. In the following section, congenital lesions of the neck, lymphadenopathy, and common malignant masses will be reviewed.

Surface anatomy of the neck is reviewed. The neck is bounded superiorly by the lower margin of the mandible, mastoids, and superior nuchal lines, and inferiorly by the clavicles and a line through the spinous process of the seventh cervical vertebra. Two cervical triangles are formed by the sternocleidomastoid muscle medially and the midline of the neck anteriorly and posteriorly. These triangles are important in the physical diagnosis of neck masses.

Since the differential diagnosis of a cervical mass is lengthy, review of certain historical and physical characteristics may lead the clinician in the appropriate direction. Lesions present at birth suggest congenital cysts or anomalies. Congenital cysts, however, may present later if they become infected and may be easily confused with regional lymphadenitis. Inflammation may be associated with congenital lesions as well as lymphadenopathy, and its presence does not allow one necessarily to predict the benignity of the lesion. Masses that enlarge slowly over several months generally are benign whereas rapidly enlarging masses, particularly nontender, matted nodes, suggest malignancy. Inflamed masses that are painful when eating suggest sialadenitis.

Physical characteristics point to possible diagnoses as well. Diffuse, soft, spongy masses may be vascular malformations, such as cystic hygromas or hemangiomas. Cystic hygromas may transilluminate, whereas a bluish hue over the mass suggests a hemangioma. A midline mass that retracts while swallowing classically describes a thyroglossal duct cyst. The neonate who presents with a head tilt may have a fibrous tumor in the sternocleidomastoid muscle, implying congenital muscular torticollis. Generalized adenopathy is a sign of systemic illness, and often other signs and symptoms will lead to the diagnosis. Chronic localized inflammation, however, may indicate mycobacterial adenitis.

In addition to physical characteristics, location of the cervical mass is an important detail. Excluding thyroid nodules, most masses anterior to the sternocleidomastoid muscle are benign. Thyroid nodules in children, however, should be suspected to be malignant until proven otherwise. Other malignancies are likely to be found as a single mass in the posterior triangle or as multiple masses extending across into both anterior and posterior triangles. It must be emphasized that while this is generally true, there are exceptions. Supraclavicular
adenopathy strongly suggests mediastinal disease, and granulomatous or lymphomatous disease should be sought assiduously. Masses along the anterior border of the sternocleidomastoid muscle, particularly if associated with a fistula, are most likely to be branchial cleft abnormalities.

**Lymphadenopathy**

Palpable cervical lymph nodes are a common finding in any pediatric practice, and are often a normal finding. It is estimated that there are greater than 500 lymph nodes in the body, ranging in size from less than 1 mm to 1 to 2 cm. The amount of lymphoid tissue is age dependent, with proliferation until puberty when lymphoid mass is twice adult values. Generally lymph nodes are easily palpable in children, with cervical, axillary, and inguinal nodes being particularly easy to palpate. In addition, the spleen may normally be palpated in 14 per cent of newborn infants and in 7 per cent of children less than 10 years of age. A thymic shadow, commonly seen in infancy, is generally not radiographically apparent, however, after three years of age. Regional drainage areas of cervical lymph nodes are demonstrated.

Lymph nodes may enlarge through cellular proliferation intrinsic to the lymph node or by proliferation and invasion of cells normally extrinsic to the node. Stimulation of intrinsic lymphocytes may occur by various antigens, hyperthyroidism, and lymphomas. Histiocytes may proliferate in lipid storage diseases such as Gaucher's disease, or in histiocytosis X, or in the benign condition sinus histiocytosis. Nodal enlargement by cells extrinsic to lymph nodes occurs by invasion of polymorphonuclear leukocytes during bacterial and fungal adenitis, and by invasion of malignant cells into nodes in metastatic tumors or leukemia.

The evaluation of adenopathy properly employs knowledge of normal lymphoid development, a careful history, and detailed physical examination. The clinician should look specifically for location, shape, size, consistency, symmetry, mobility, signs of inflammation, suppuration, and overlying skin discoloration. Examination for more widespread or systemic disease should be completed. Overall, however, it may be helpful to divide the approach to cervical lymphadenopathy into regional or generalized lymphadenopathy.

**Regional Adenopathy**

Occipital nodes are commonly enlarged in afflictions of the scalp as in infections and seborrheic dermatitis. They may be enlarged in conditions that produce generalized adenopathy as well, or in specific viral illnesses such as rubella. They are normally palpable in about 5 per cent of children.

Preauricular nodes drain the temporal region and conjunctival sac and are commonly involved in "oculoglandular" syndromes, Parinaud's syndrome, or conjunctivitis and ipsilateral preauricular lymphadenopathy. Chlamydial conjunctivitis, adenovirus, cat scratch, and tularemia have been implicated as infectious etiologies, as have syphilis and tuberculosis. In addition, Hodgkin's disease may present with preauricular adenopathy. The clinician must rule out branchial cleft cysts and parotid inflammation as causes for swelling in this area.
Submaxillary and submental nodes enlarge with infections of the teeth, lips, and gums. Herpetic stomatitis, dental abscesses, and occasionally dry, chronically cracked lips may be associated with submaxillary or submental adenopathy. Salivary gland enlargement, particularly in cystic fibrosis, may be confused with enlarged lymph nodes. Over 90 per cent of children with cystic fibrosis over 2 years of age will have submandibular salivary gland enlargement.

By far, the most frequently involved groups of nodes include the superior and inferior deep cervical nodes, posterior cervical nodes, and superficial cervical nodes. These nodes enlarge with infections and enlargement may persist for long periods of time after the infection. One report demonstrated persistent adenopathy for 10 months after a herpes infection. They may persist singly or multiply. The inferior deep cervicals drain a wide area including the entire head and neck, arm, thorax, lung, and mediastinum. Hence, these nodes may be affected by a large number of problems affecting a wide anatomic area.

Viral upper respiratory tract infection is the most common cause of enlargement, producing discrete, minimally tender, oval, soft nodes. Bacterial lymphadenitis usually produces systemic toxicity with unilateral or bilateral, swollen, acutely tender, sometimes fluctuant nodes with overlying warmth and erythema. Streptococcus pyogenes and Staphylococcus aureus have been isolated from lymph node aspirates. The staphylococcus is often penicillin resistant, and antibiotic therapy would require the use of a semisynthetic penicillin such as dicloxacillin. Rarely, Hemophilus influenzae and anaerobic bacteria have been isolated from lymph node aspirates as well. Mycobacterial adenitis will be discussed in detail in another article in this issue.

Cat-scratch disease may cause regional nonbacterial, tender lymphadenitis. A brief period of headache, malaise, and fever usually follows a scratch from a cat, or rarely a dog or monkey bite. A primary pustule or papule forms and may persist for 2 to 7 weeks. Tender lymphadenopathy develops during the initial two weeks and may persist for 2 to 3 months. Suppuration occurs in 10 to 25 per cent of patients. Node pathology may range from reticulum cell hyperplasia to granuloma and abscess formation. Diagnosis is confirmed when three of the four following criteria are met: a primary lesion or history of animal contact or scratch; a sterile node aspirate with laboratory data excluding other possibilities; a lymph node biopsy consistent with cat-scratch disease; and a positive skin test to cat-scratch antigen. This antigen is not commercially available, however, and recently there has been some question as to its safety. In the majority of patients, no active intervention is necessary.

Infectious mononucleosis, or Epstein-Barr virus (EBV) infection, may produce localized cervical adenopathy or generalized adenopathy. Fever, malaise, tonsillopharyngitis, and hepatosplenomegaly may be associated with the illness in older children. Infants and young children often have atypical disease with silent or mild upper respiratory tract infections. Diagnosis is made by demonstrating a positive heterophil test. Young patients, however, may not have heterophil agglutinating antibodies. Twenty-seven to 91 per cent of 2 to 5 year old patients with EBV infection will be heterophil positive; 53 to 94 per cent of 6 to 10 year old children and 100 per cent of older children will be positive. Definitive diagnosis can be made by measuring specific EBV antibody titers. Definitive diagnosis can be made by measuring specific EBV antibody titers. Nearly half of children with
mononucleosis-like illnesses will be EBV negative. Cytomegalovirus infection, toxoplasmosis, and adenovirus infection have been identified in patients with clinically similar illnesses.

Sinus histiocytosis is a syndrome of massive, bilateral, painless cervical adenopathy usually occurring in blacks. Occasionally other node groups are involved. Fever, leukocytosis with neutrophilia, elevated erythrocyte sedimentation rate, and hypergammaglobulinemia are other common features. This syndrome is benign and is characterized by persistent adenopathy for months to years and gradual resolution of the adenopathy. Occasional, extranodal sites such as the orbit may be involved. Histologically the lymph nodes show pericapsular fibrosis, dilated sinuses with intrasinusoidal histiocytes, and frequent plasma cells. The etiology is unknown, and the disease may recur.

Kawasaki's disease is characterized by acute nonpurulent swelling of cervical lymph nodes, fever lasting one to two weeks, conjunctivitis, dryness and fissuring of the lips, and rash of the trunk and extremities leading to membranous desquamation from the fingertips. Often there are elevations in the acute phase reactants, leukocytosis, thrombocytosis and urinary sediment abnormalities. Approximately 2 per cent of patients die of myocardial infarction owing to coronary artery aneurysms. The etiology is known and treatment with steroids and/or aspirin is controversial.

Generalized Adenopathy

Generalized lymphadenopathy occurs when nodes of two noncontiguous regions are involved in a pathologic process. Zuelzer and Kaplan provide an excellent review of generalized adenopathy. Systemic viral infections may cause generalized nodal enlargement, and illnesses that produce exanthems, such as rubella and scarlet fever, produce generalized lymphadenopathy as well. Bacterial infections including tuberculosis, syphilis, brucellosis, and typhoid fever also have been implicated. Other infections including histoplasmosis and toxoplasmosis may cause illness similar to EBV infection including adenopathy. In some immunodeficiency states, particularly chronic granulomatous disease and immune-mediated illness, generalized lymph node enlargement may be a prominent physical finding. In the acute stages of systemic lupus erythematosus and juvenile rheumatoid arthritis, two thirds of patients may have generalized adenopathy. Autoimmune hemolytic anemia occasionally has been associated with such adenopathy as to resemble lymphoma. The adenopathy of serum sickness generally resolves as the other major manifestations disappear. In lipid storage diseases such as Gaucher's syndrome, hepatosplenomegaly is present in addition to adenopathy. Sarcoidosis in children may be characterized by generalized adenopathy; the chest x-ray is usually abnormal, exhibiting hilar adenopathy, parenchymal fibrosis, or mottling. Chronic phenytoin administration has been linked with lymphoma-like syndromes with generalized enlargement of lymph nodes. Malignant conditions in children such as acute leukemia, histiocytosis, and metastatic neuroblastoma may have diffuse nodal enlargement. In general, however, all of these illnesses usually have other distinguishing signs or symptoms to aid in diagnosis.

Malignant Tumors

Of seven children admitted with neck masses, one will have a malignant mass. The tumors usually are derived from mesenchyme. Lymphoid tumors tend to predominate, the cell
type with Hodgkin's disease and lymphosarcoma comprising 55 per cent of malignant head
and neck tumors. Rhabdomyosarcoma is the most frequent solid tumor of the head and neck
in children. Fibrosarcoma, neurofibrosarcoma, thyroid neoplasms, and neuroblastoma are less
common.

Although lymphosarcoma occurs approximately twice as frequently as Hodgkin's
disease in general, Hodgkin's disease presents twice as frequently (80 per cent) as
lymphosarcoma (40 per cent) with a neck mass. Hence, Hodgkin's disease and lymphosarcoma
occur with about equal frequency when presenting as a neck mass. Hodgkin's disease
generally will present as a painless, slowly enlarging, unilateral (80 per cent), firm, nontender
node located in the upper one-third of the neck. When lower neck masses are found, as in
supraclavicular adenopathy, mediastinal involvement is frequent. Six per cent of Hodgkin's
disease may present as preauricular adenopathy simulating parotid swelling. Hodgkin's disease
generally affects children over 5 years of age and, in contrast to lymphosarcoma, fewer
extranodal sites are involved. Lymphosarcoma, on the other had, is more frequent in the
younger patient. Extranodal sites, such as the tonsils, are four times more frequent in
lymphosarcoma than in Hodgkin's disease. The nodes are described as a rubbery, discrete,
painless mass.

Rhabdomyosarcoma is the most frequent solid tumor of the head and neck, comprising
approximately 10 per cent of malignant head and neck tumors. It presents as a painless mass
virtually at any site, and its accompanying symptoms depend upon the organ of encroachment.
The nasopharynx, middle ear, mastoid, and orbit are frequent sites of involvement.

Fibrosarcoma and neurofibrosarcomas may present as a painless mass arising from the
cheek, jaw, nose, or sinuses. They make up about 5 per cent of malignant tumors in the head
and neck. These tumors have a low tendency to metastasize.

Thyroid masses in children should be suspected of being malignant, particularly if
there is a history of irradiation to the head or neck. Generally, midline or slightly lateral
masses are in close proximity to the thyroid gland so that diagnosis is not very difficult. Most
of these tumors are of the medullary or mixed papillary and follicular type.

Although neuroblastoma is the most common solid tumor in childhood overall, it
ranked sixth in frequency in malignant tumors of the head and neck. Primary neuroblastoma
of the neck occurred in nine of 178 cases and generally could be distinguished from
metastatic neuroblastoma by the early onset of neurologic signs as Horner's syndrome. In
contrast, metastatic neuroblastoma usually spread to lymph nodes, and neurologic signs
depended on growth of the node to impinge on neural structures. This occurred relatively late.

Age is a factor in the type of tumor found, as alluded to above. Children under 6 years
tend to have neuroblastoma, most frequently followed by lymphosarcoma, rhabdomyosarcoma,
and Hodgkin's disease. Children from 7 to 13 years have Hodgkin's disease and
lymphosarcoma with almost equal frequency, with thyroid cancer and rhabdomyosarcomas
following. The primary tumor of the head and neck in adolescents is Hodgkin's disease.

Evaluation of the child with a malignant neck mass requires a careful general
examinatin with special attention given to the reticuloendothelial system. In addition, since
one of every six malignant neck masses has an associated tumor of the nasopharynx, a thorough examination of the ear, nose, and throat is mandatory. Routine laboratory studies as well as vanillylmandelic acid (VMA) spot test and possibly bone marrow examination are also indicated. Lymph node biopsy may give a definitive histologic diagnosis. Should a lymph node be interpreted as nondiagnostic, however, patients should be followed until the adenopathy resolves or a diagnosis becomes apparent. Kissane and Gephardt reported on 37 patients with initial nondiagnostic lymph node biopsies. Six patients died within the five to 20 year follow-up. Four died of probable immune deficiency and thrombocytopenia and two died of malignant disease. Similarly, Lake and Oski found 17 per cent of 41 initial nondiagnostic node biopsies to have noticeable disease on subsequent node biopsy. Hence, follow-up of these patients is imperative.
Cervical Lymphadenitis Caused by Atypical Mycobacteria

Eward W. Saitz


Mycobacteria can cause a variety of human diseases, of which tuberculosis is the most familiar to clinicians in this country. Tuberculosis is caused by the organisms *M. tuberculosis* and *M. bovis*, or by *M. africanum* as reported from only the western coast of Africa. Although uncommon in the USA, leprosy is prevalent in other parts of the world and is caused by *M. leprae*. Atypical mycobacteria can generally be thought of as mycobacteria other than the species named above. The term "atypical" was first used in the 1950s when it was apparent that some mycobacterial isolates from clinical specimens were neither *M. tuberculosis* nor *M. bovis*. Other descriptive terms have included "anonymous" or "unclassified" but these terms are now inappropriate, since the mycobacteria have been given species names, and serotypes have been described for some. "Opportunistic" had been suggested as a term, because a predisposing condition was felt necessary for disease to occur in humans. Although this may be true for some adult pulmonary diseases, it does not account for most pediatric infections. Also, the names mycobacteria other than tuberculosis (MOTT) and nontuberculous mycobacteria (NTM) have been suggested. However, the term atypical is used in this article because it is more commonly recognized by clinicians, and infections from these organisms differ in pathogenesis and treatment from those caused by *M. tuberculosis*. In addition, the literature concerning these infections is listed in the *Index Medicus* under the subject-heading "Mycobacterium Infections, Atypical".

The anatomic sites commonly affected by these organisms differ in adults and children. Usually, adults have chronic pulmonary infections, whereas children have chronic lymph node infections. The specific topic of this article is lymph node infections in children that occur in the neck and face. However, infections have also been reported in axillary, epitrochlear, and inguinal nodes. In addition, skin, soft tissue, skeletal, lung, genitourinary, meningeal, and disseminated infections have occurred.

**Descriptive Terminology**

Approximately 20 years ago, Runyon proposed a preliminary classification of atypical mycobacteria that was based on growth characteristics and the ability to form pigment. This classification includes four groups. Group I, the photochromogens, do not produce pigment if grown in the dark but become bright yellow after exposure to light. Group II, the scotochromogens, produce a bright orange pigment whether grown in the light or dark. Group III, the nonphotochromogens, do not produce pigment when grown either in the light or dark. Group IV, the rapid growers, produce visible growth within several days. (Most mycobacteria require several weeks to produce visible colonies.) Currently, these mycobacteria are more precisely identified by species names, such as *M. scrofulaceum*, although most species generally fit into one of Runyon's original groups.
Epidemiologic Aspects

Atypical mycobacteria are frequently found in soil, water, milk, and house dust; they can also be isolated from domestic and wild animals, and from birds. Some studies preceded species identification and, therefore, the isolates were described only by the Runyon group. It is now known, however, that all species within each group are not pathogenic for humans. For example, both *M. scrofulaceum* and *M. gordonae* can be categorized as Runyon Group II. Although *M. gordonae* have been recovered from soil, they are unlikely to be recovered from clinical specimens such as pus that has been obtained from infected lymph nodes.

Chapman isolated atypical mycobacteria of Groups II, III, and IV from raw and pasteurized milk, and found that the number of isolates obtained in winter was greater than that found in spring and summer. Similar seasonal variations were found in isolations from human material. In another study, the majority of children with cervical lymphadenopathy from atypical mycobacteria presented for treatment during the winter and spring.

Recognition of the large number of people apparently infected with atypical mycobacteria came with the Navy recruit skin testing studies conducted by Edwards and associates. Recruits were skin tested upon entry to the service with a standard tuberculin test (PPD-S) and also with PPD-B, which was prepared from a Battey strain of atypical mycobacteria that was isolated from a patient in the Battey State Hospital in Rome, Georgia. Of recruits who originally resided in the southeastern USA, 60 to 75 per cent had reactions to PPD-B greater than 2 mm, whereas only 17 to 24 per cent of recruits from the Northwest had similar reactions. These results show an interesting geographic variation and indicate a large number of people have been infected with some type of atypical mycobacteria.

Although disease might be expected to occur more often, some evidence indicates it is unlikely to occur even with prolonged exposure to the organisms. For example, an adult with pulmonary disease had persistently positive sputum for *M. scrofulaceum*, and none of his children became infected. Also, atypical mycobacterial lymphadenitis has been reported infrequently in the same family. Only five pairs of siblings were found in Lincoln's review of 477 cases of adenitis, and it was felt that their infections could be related to a common source rather than to familial spread. Furthermore, lymphadenitis rarely progresses to widespread disease. Disseminated disease has been reported, but these children have usually presented with widespread disease when first seen. Also, they often had preexisting diseases or altered immune systems. One child with cervical lymphadenopathy died a year after diagnosis with disseminated disease caused by a Group II organism identified as Gause. However, this case was peculiar because the child had been treated three years previously with isoniazid and streptomycin for pulmonary tuberculosis. Therefore, the ability of atypical mycobacteria to cause disease seems relatively limited and, in the case of lymphadenitis, the likelihood of progression to disseminated disease is remote.

Suspected Pathogenesis

Atypical mycobacterial lymphadenitis occurs predominantly in children between one and three years of age. Chapman proposed that infection may be related to ingestion of organisms in mild, and entry via skin breaks associated with eruption of the primary teeth. Although reports of oral infections can be found in the literature, they concern much older
patients. Rice reported an 18-year-old man with a retropharyngeal abscess and cervical lymphadenitis. Cultures from both sites grew mycobacteria identified as Battey bacilli. Schuit described a primary mycobacterial infection of the pharynx in a seven-year-old girl. Cultures from both the pharyngeal mass and the cervical node grew *M. intracellulare*, and biopsies from both areas showed non-caseating granulomas with acid-fast bacilli.

**Clinical Presentation**

Most cases of atypical mycobacterial lymphadenitis have been reported in children between one and five years of age; however, as mentioned previously, the predominant age affected is between one and three years. The disease is rare in children less than seven months, or more than 12 years, of age. The lymph nodes enlarge suddenly, but only one side of the neck or face is usually involved. The child does not appear to be sick at the time, although there may be a history of respiratory infection during the previous weeks. While some mild pain and tenderness may be present, there are no marked symptoms such as those seen with acute pyogenic infections. Several nodes often enlarge in one location and initially they may appear to be a single mass. In some cases, the mass is quite large. The enlarged nodes usually occur near the angle of the jaw, but occasionally involve the preauricular area. They are found less frequently in the submental area and along the sternocleidomastoid muscle. Supraclavicular involvement is unusual and unless associated with suspected pulmonary tuberculosis probably should indicate the need for immediate biopsy.

Early in the clinical course, the skin becomes adherent to the underlying tissues at the peripherly of the mass. Also, increased vascularity is commonly seen in the same location. Within several months, the nodes become fluctuant. (Some observers have felt that this is a more rapid evolution than is usually seen in infections with *M. tuberculosis*.) In addition, the overlying skin usually changes in color from pink to purplish-red. Although the skin appears erythematous, it is not warm to the touch. This rather distinctive color change is followed by thinning and a parchment-like appearance of the skin, and then by spontaneous drainage from the underlying node. While these skin changes and the development of fluctuation seem characteristic of mycobacterial infections, they are not specific in distinguishing atypical mycobacterial infections from those caused by *M. tuberculosis*. Also, some children develop fluctuation without any skin changes; this may erroneously suggest other diagnoses.

**Diagnosis**

Children of the above-mentioned ages who have chronic unilateral lymph node enlargement should be suspected of having atypical mycobacterial infections, particularly if the nodes are located around the angle of the jaw or in the preauricular area. The presence or development of characteristic skin changes and fluctuation of the mass would further support this clinical impression. The children appear healthy and there is absence of disease elsewhere in the body. Generalized lymphadenopathy and abdominal organomegaly are not found, and chest radiographs are usually normal. Also, there is no history of recent contact with an adult who has suspected or proven infection with *M. tuberculosis*.

Previously, skin test antigens prepared from various atypical mycobacteria were available from the Centers for Disease Control in Atlanta, Georgia. When tested with these antigens, children with atypical mycobacterial infections reacted to the atypical PPDs and had
larger reactions to them than to PPDs prepared from *M. tuberculosis*. Therefore, it was usually possible to distinguish between atypical mycobacterial infections and those caused by *M. tuberculosis* on the basis of the skin test reactions. Also, the skin tests were quite helpful in suggesting a mycobacterial etiology, rather than a malignant process, before skin changes or fluctuation occurred. Unfortunately, these antigens presently are not available because they are being evaluated in bioassay studies and tests of clinical usefulness.

Children with atypical mycobacterial infections may also react to standard tuberculin tests because of the cross reactivity that is known to occur. When tested with a 5 TU dose of PPD-tuberculin, these children tend to have reactions of less than 10 mm induration. (Conventionally, reactions of 10 mm or greater are believed to indicate infection with *M. tuberculosis*.) Surprisingly, a moderate number of children will have no reaction to PPD-tuberculin (5 TU). However, this lack of reactivity should not be used as a criterion to rule out the possibility of atypical mycobacterial disease. It is known that some patients infected with *M. tuberculosis* also show no reaction to conventional tuberculin tests.

Disturbingly, a few children with atypical mycobacterial infections have large reactions to PPD-tuberculin (5 TU) early in the course of their disease, and this may incorrectly suggest a diagnosis of *M. tuberculosis*. Schuit has shown that when these children are retested several months later, the size of their PPD-tuberculin reactions decreases considerably. Therefore, a child who is suspected of having an atypical infection, and whose reaction to PPD-tuberculin (5 TU) is large, should be retested several months later to see if the size of the reaction decreases.

While the above characteristics will presumptively identify children with atypical mycobacterial lymphadenitis, the only definitive proof is isolation of the infecting mycobacterium. This degree of diagnostic certainty, however, may not be necessary in all cases. Material can be obtained by needle aspiration, incision and drainage, or by surgical biopsy of the involved node. Mycobacteria require several months to grow, and in most reported series approximately one-half of the cultures that were obtained demonstrated no growth. Acid-fast bacilli may also be seen on smears from pus or in tissue sections, but the morphology of the commonly isolated atypical mycobacteria is not distinctive from the morphology of *M. tuberculosis*. Although some investigators have felt that the histopathology of atypical infections was distinguishable from infections caused by *M. tuberculosis*, Reid concluded that the features were not specific enough to enable such a differentiation.

**Treatment**

The predominant species of mycobacteria isolated from cervical lymph nodes have been either *M. scrophulaceum* or members of the *M. avium-intracellulare* complex (Runyon Groups II and III respectively). Both show marked resistance in vitro to the usual antimycobacterial drugs. Early reports from Dallas, Texas mentioned the frequent isolation of *M. kansasii* (Runyon Group I). While this species may respond to rifampin, the number of *M. kansasii* isolates in Dallas has decreased markedly in recent years for unknown reasons, and *M. kansasii* have been reported infrequently in other series. Because of the factors of drug resistance of the common species, the low risk of human-to-human spread, and the remove
chance of dissemination, antimycobacterial drugs are felt to be unnecessary. Good results without the use of such drugs have been reported by several investigators.

Surgical therapy of these infections has included needle aspiration, incision and drainage, incision and curettage, and total excision. A recent review of the management of atypical mycobacterial lymphadenitis favors total excision, but we believe that an alternative "nonexcisional" approach, which will be described later, has some advantages.

**Long-Term Effects**

Most children with atypical mycobacterial infections have not been followed for long periods of time, and therefore the development of late sequelae, such as recrudescence in adult life, is unknown. One problem has occurred, however, when these children are tuberculin-tested at the time of entering school. Their previous atypical mycobacterial infections are often forgotten, and because of cross reactivity to the tuberculin test, they are mistakenly thought to have tuberculosis.

**Pittsburgh Experience**

From 1967 to the present, the author has seen major children from this geographic area who were suspected of having atypical mycobacterial lymphadenitis. Of these children, a group with the following characteristics was selected. The location of the node was in the neck or face; the culture was positive for either *M. scrofulaceum* or members of the *M. avium-intracellulare* complex; the results of skin tests with atypical antigens were available; and the children resided in the western Pennsylvania area. Twenty-seven children satisfied all four conditions and these children had not been previously reported.

During the same period, two additional children were seen who had similar species of atypical mycobacteria isolated from a different location. Both had inguinal node infections, one from *M. scrofulaceum* and the other from *M. avium-intracellulare* complex. Two other children had cervical nodes from which *M. fortuitum* was isolated. This organism is usually associated with injections or trauma, and in one case it was known that the child was cut in the face by an exploding soft-drink bottle. Because of the pathogenesis of *M. fortuitum* appears to be different, both of these children were excluded from this report. Also, no isolates of *M. kansasii* were obtained from nodes in any location.

The infections usually involved a group of nodes in only one anatomic location. The cervical group of nodes, ie, those near the angle of the jaw and the submandibular area, were most commonly affected. In five children, however, both the cervical and the preauricular nodes were involved. Except for one child, all infections were unilateral. There was more consistency in the locations associated with *M. scrofulaceum* infections that those caused by the *M. avium-intracellulare* complex. Infections in nodes along the mid-to-lower portions of the sternocleidomastoid muscle and in the submental area were found only with the latter species.

The ages are given in terms of estimated onset of the enlarged nodes as determined by history. Most infections had their onset in children between one and three years of age (67
per cent). The youngest child was 13 months, the oldest 10 years, 7 months. Almost equal numbers of males and females were affected, but considerably more infections were seen in white children. In the majority of children, the onset was during the first six months of the year. This seasonal variation was most apparent with the *M. scrofulaceum* isolates.

All 27 children were skin tested with a 5 TU dose of PPD-B, G, and Y. (These antigens were originally supplied by the Centers for Disease Control in Atlanta, Georgia.) All children reacted to one or more of the tests, and the mean sizes of the reactions in terms of induration were 13 mm, 14 mm, and 8 mm, respectively. Twenty-five children were simultaneously tested with PPD-tuberculin (5 TU). In all 25 patients, the degree of induration from one of the atypical PPD tests exceeded the PPD-tuberculin by at least 6 mm. (The atypical tests do not distinguish among the various species of atypical mycobacteria.) However, in 13 children (52 per cent) there was no induration to PPD-tuberculin. If these children had not been tested with the atypical skin tests, it may have been falsely assumed that they did not have a mycobacterial infection. Some referring physicians mentioned that their patients had reactions to tuberculin screening tests and more recently we have added a Tine test. In five children recently seen with positive cultures who had no induration to PPD-tuberculin, the Tine test caused some induration in all of them. The reactions were usually small (1 to 2 mm) but there was definite induration. Therefore, in the absence of atypical PPD skin tests, the dual use of a Tine test and PPD-tuberculin may be more helpful in suggesting a diagnosis of an atypical mycobacterial infection. (Whether other screening tests would work equally well is unknown.)

The total duration of these infections varies from approximately 9 to 15 months. Generally, the skin changes occur within the first several months. Initially, the border of the mass becomes pink and an increased number of small blood vessels appear at the periphery. As the color progresses to a characteristic purplish-red, the skin develops a parchment-like appearance and begins to peel. The node is usually fluctuant at this time and spontaneous drainage may occur. Early in the course, the skin becomes attached to the outer edges of the node, and the dimpling of the skin that occurs later is most likely related to this early inflammatory response. If the node spontaneously ruptures, or if surgical drainage is performed, there may be continued drainage for a period of months. The enlarged node then gradually regresses and the opening in the skin closes over. Finally, the skin color returns to normal.

Although the literature suggests that these nodes should be excised, we have found other approaches to be satisfactory. Because of the chronic inflammation, the nodes are densely adherent to the surrounding tissues, and therefore dissection is quite difficult. Injury to the mandibular branch of the facial nerve can occur following surgical excision; in one series, skin breakdown followed the excision of preauricular nodes in two of five children. Once the skin is involved, a moderate sized area of excision may be necessary, and it is not surprising that breakdown occurs, particularly in the preauricular area. The "nonexcisional" methods that have been used include needle aspiration, incision and drainage, and incision and curettage. The specific method chosen depends on the stage of the disease and the extent of the fluctuant area. We feel it is safer to revise the scar and skin contracture later, if necessary, rather than to widely excise the lesion and risk a problem of skin breakdown or facial nerve injury, even though these risks are relatively small. Also, these children have not been treated with antituberculous drugs unless the diagnosis was uncertain initially.
Of the 27 children reported here, only six had other surgical procedures. Five children had biopsies and one child had a partial excision of her mass. In all six, another disease (usually malignant) was suspected preoperatively, and in five, skin testing with atypical PPDs had not yet been performed. Skin testing was suggestive of atypical mycobacterial disease in the sixth child, but because the mass had not regressed, a biopsy was performed.

No known complications have resulted from this nonexcisional approach. In some children, however, drainage from the affected nodes has persisted for two to four months. The parents and children have accepted this prolonged course when it is explained that the drainage is not contagious and that the lesion will eventually heal. Photographs of other children with healed lesions have been used to illustrate the end results. Interestingly, nonexcisional procedures such as incision and manual expression of the pus, or incision with enzyme treatment, had been proposed in the past to treat tuberculous lymphadenitis caused by \textit{M. tuberculosis}.

Dissemination of disease following lymph node infections has not been observed. Two cases, however, occurred in one family. A younger sister had a partial excision for a suspected parotid tumor and within two months her older brother developed an infection on the same side of his neck. \textit{M. scrofulaceum} was obtained from the cultures of both children. In another child a mass developed in the same location six years after her infection with \textit{M. scrofulaceum}. A recurrent mycobacterial infection was suspected but never proven. The surgeon described the node as acutely inflamed, but cultures for bacteria (including anaerobic organisms), yeast, fungi, and mycobacteria were sterile. The histopathologic findings were interpreted as chronic inflammation. Interestingly, a radiograph of the patient's neck showed bilateral calcification of the nodes at the time of readmission. The calcification was more marked on the left side, which was the site of the original infection.

During this same 13 year period, there were no isolates of \textit{M. tuberculosis} from nodes in the neck and face as determined from review of the bacteriology records at Children's Hospital of Pittsburgh. However, \textit{M. tuberculosis} was recovered from an inguinal node of one child and an axillary node of another. These results vary considerable from a study in North Carolina where \textit{M. tuberculosis} was recovered from a large number of children.

Factors to consider in the differential diagnosis of chronic unilateral cervical lymphadenitis in the anatomic locations described previously vary depending on the local experience. In our area, the factors primarily include atypical mycobacteria, malignant disease, and nonspecific lymphadenopathy. In other areas, infections with \textit{M. tuberculosis} should be included. Unfortunately, it was easier to distinguish between infections from atypical mycobacteria and those from \textit{M. tuberculosis} when skin tests for atypical mycobacteria were available. Hopefully, these antigens will again be available in the USA.

\textbf{Summary}

Atypical mycobacterial lymphadenitis is basically a benign disease. Although children between one and five years of age are commonly affected, the predominant occurrence is in those from one to three years old. The children are frequently brought to the physician because of chronic unilateral lymph node enlargement. Most often, the nodes are located near...
the angle of the jaw, but they may also be found in the preauricular area. The overlying skin usually becomes discolored within several months of onset, and the nodes become fluctuant. Some data are available to support the hypothesis that a primary oral source is responsible for these infections. Mycobacteria identified as *M. scrofulaceum* and those belonging to the *M. avium-intracellulare* complex are the common species that have been isolated from infected nodes. Infections tend to occur more frequently during the first six months of the year.

Although currently not available, atypical PPD skin tests were useful in distinguishing atypical mycobacterial infections from those caused by *M. tuberculosis*. These skin tests were also quite helpful in suggesting a mycobacterial infection, rather than a malignant process, before skin changes occurred or the nodes become fluctuant. Skin tests with standard PPD-tuberculin (5 TU) may result in intermediate sized reactions, ie, less than 10 mm of induration. However, some children with culture-proven atypical mycobacterial infections either have no reactions, or large reactions, to the same tests.

Because of the low risk of communicability, in vitro drug resistance of the commonly isolated species, and the unlikelihood of spread within the host, antimycobial drugs are not required to treat these infections. Although the literature favors surgical excision of the infected nodes, an alternative nonexcisional approach has some advantages and seems to provide good long-term results.

Finally, it is important to remind parents that children who have such infections may react to tuberculin tests later in life. This is particularly important in order to prevent unnecessary concern or inappropriate treatment for suspected tuberculosis, when these children are tuberculin-tested at the time of school entry.
Neck Masses of Congenital Origin

Lois A. Pounds


Palpation of the neck of infants and children is an important part of the pediatric physical examination. The general pediatrician is usually searching for lymph nodes associated with upper respiratory infections or infections of the face or scalp; the pediatric endocrinologist is evaluating the thyroid gland; the pediatric oncologist is looking for tumor, either primary or metastatic; the pediatric cardiologist is feeling for carotid pulsations; and so forth, down the line of subspecialities. Any or all may come across the three major congenital masses in the neck, and while these are not serious problems in childhood, they can be diagnostically confusing. A knowledge of the embryology and anatomy of these masses is helpful in making an accurate diagnosis. The three lesions to be discussed are thyroglossal duct cysts, branchial cleft cysts and cystic hygroma. Of the benign congenital masses in the neck, 72 per cent will be cysts of the thyroglossal duct, 24 per cent will be branchial cleft cysts, and 4 per cent will be cystic hygroma.

Thyroglossal Duct Cysts

In the third week of embryonic life the thyroid anlage begins to develop on the floor of the pharynx at the base of the tongue. As the embryo grows, the developing thyroid gland descends in the neck, maintaining its connection to the tongue by a narrow canal, which is the thyroglossal duct. At the tongue end of the duct, the epithelial cells differentiate into tongue mucosa and persist as the foramen cecum of the tongue. At the thyroid end, the duct epithelium is induced to form thyroid tissue and, in some individuals, a pyramidal lobe of the thyroid gland. The mid-portion of the duct persists as a microscopic thread of undifferentiated epithelial cells tracking from the foramen cecum, through the hyoid bone periosteum or the bone itself, to the thyroid gland. For unknown reasons, this undifferentiated thread of cells may become active later in life and differentiate into columnar, ciliated, or squamous epithelium or into glandular tissue (sebaceous, salivary, or thyroid). When the cells are activated they produce a midline mass which is almost always cystic and contains mucoid material. Occasionally the cyst or another part of the duct will create a sinus tract to the skin at or just lateral to the midline. Anatomically these masses may be found anywhere from the foramen cecum to the thyroid gland, but most are adjacent to the hyoid bone.

Clinically the mass is usually observed by the mother, who may report that it is intermittently present. In fact, these cysts are best visualized when the neck is hyperextended and the usual small ones are not visible when the head is in neutral position. The majority of these are 1 to 2 cm in diameter, but some are 5 to 6 cm in diameter on presentation. Thyroglossal cysts are smoothly rounded with well defined margins and are slightly movable. Because of the thyroglossal duct attachments, the cyst will rise with tongue protrusion and move with swallowing. The cysts are nontender unless there is inflammation of the cyst or an associated sinus tract. They do not transilluminate well because of the overlying fascia of the neck. The sinus openings may be anywhere from the suprasternal notch to the hyoid bone. Careful palpation may enable the tract of the duct to be felt from the opening in the skin to the hyoid bone. The opening may show heaped up edges secondary to low grade chronic
inflammation. Drops of clear or cloudy mucus may escape from the sinus. If the sinus is present, the diagnosis is clear. If there is only the mass of the thyroglossal cyst, observation over several weeks will differentiate it from an inflammatory lesion. Other tumors that may be found in this region are lipomas and dermoid cysts. Lipomas are more lobulated and softer than thyroglossal duct cysts, whereas dermoids are attached to the skin and move with it. Rarely the thyroid gland itself will not descend to its normal position and will be found in the region of the hyoid bone. The absence of a thyroid gland in the normal position should suggest that the upper mass is the thyroid gland.

Treatment is probably not necessary for cysts less than 6 mm in diameter because they are asymptomatic and have no morbidity. The large ones and/or those that have been infected should be surgically excided. Sistrunk, in 1920, described the definitive surgical procedure. This includes excision of the cyst, the central 1 cm of the hyoid bone, and the tract of the duct up to the foramen cecum. Leaving behind remnants of the duct invites recurrence of either the cyst or the sinus tract.

**Branchial Cleft Cysts**

In the early embryo the branchial arches, branchial clefts, and pharyngeal pouches are the primitive structures from which the muscles, bones, and ligaments of the face and neck and their nerve and blood supplies will develop. Embryologists and surgeons have determined that the source of most cysts of branchial origin arise from remnants of the second branchial cleft. This cleft is really a groove separating the second and third branchial arches. Remnants of this cleft would lie anterior to the sternocleidomastoid muscle, between the internal and external carotid arteries and on a line between the tonsillar fossa and the junction of the middle and lower thirds of the sternocleidomastoid. In fact, when fistulas are found these course from the sternocleidomastoid to the posterior tonsillar pillar passing between the internal and external carotids. The cysts are lined by squamous or columnar epithelium and usually have lymphoid tissue adjacent to them. The sinuses are lined by stratified squamous, ciliated, or columnar epithelium and these tracts may have lymphoid tissue in the walls.

Clinically, the branchial sinuses present as a 2 to 3 mm slit anterior to the lower third of the sternocleidomastoid. They can be identified in the newborn infant. The importance of identifying the sinus is obvious as it is a portal of entry for pathogenic organisms. The cysts of branchial origin appear at any age and usually are opposite the middle third of the sternocleidomastoid. The commonest age at presentation is in the early school age child. The presence of a rounded mass in the position described, which is nontender and only slightly movable, makes branchial cleft cyst a good possibility. If the patient is seen in the context of an acute upper respiratory infection, the mass may be mistaken for an enlarged lymph node. Indeed because of adjacent lymphoid tissue, the cyst may enlarge concomitant with the lymph nodes. Observation that the lymph nodes decrease in size after the infection, while the mass persists, should differentiate this lesion.

Treatment is surgical removal of all the remnant structures. In the newborn infant with a sinus tract, it is not advisable to explore the neck until the structures are large enough for appropriate dissection. The parents should be instructed in good skin care to prevent infection of the tract, and any infection that occurs should be promptly treated. In the older child with a cyst, the surgery is elective and should be done in those with visible masses.
Cystic Hygroma

These masses of lymphatic tissue appear to arise from the lymphatic sacs, which develop either as a direct bud from the jugular vein, or from mesenchymal tissue which coalesces and ramifies and then acquires venous connections. These lymphatic sacs appear to develop outpouchings that are then pinched off or sequestered to give rise to a cyst. The cysts have been found in the groin and axilla but most commonly occur in the neck. These lymphatic cysts have thin walls with sparse fine vessel vascularity, and are lined with endothelial cells. Narrow outgrowths of endothelial cells from the cyst wall can then insinuate themselves between muscle bundles, nerve fibers, or other tissues, produce lymph-like fluid, and enlarge in that manner.

Most extensions communicate with the main cyst cavity but there can be multiloculated cysts. Cystic hygroma is not a lymphatic malignancy, but it can damage surrounding tissue by pressure and can become large enough to compromise the airway or interfere with swallowing.

Most cystic hygromas (90 per cent) are found in the posterior cervical triangle behind the sternocleidomastoid muscle in the supraclavicular fossa. These can extend toward the midline, up toward the angle of the mandible, or out toward the crest of the shoulder. The mass may be quite large in the first few weeks of life. In Gross's series, 65 per cent were present at birth, 80 per cent discovered in the first year, and 90 per cent were found before the second birthday. The mass is usually soft, compressible, and may be smoothly rounded or slightly lobulated. It transilluminates well which confirms its cystic character. Occasionally any acute upper respiratory infection will trigger a sudden increase in size of the cyst, possibly because the normal lymphatic channels of the region become inflamed and partially obstructed. It appears that this obstruction causes lymph stasis which distends the cyst. Even though anastomotic connections to normal lymph channels are not easily demonstrated pathologically, these cases suggest that they do exist.

In the past, attempts to treat these cysts included aspiration, injection of sclerosing agents, radium therapy, and X-irradiation. Watchful waiting for spontaneous regression has also been recommended. With modern anesthesiology and pediatric surgical techniques, the safest and most satisfactory outcomes are from surgical dissection of the whole lesion. In the large infiltrating masses, the pediatrician should realize that this is not "shelling out" a clear cut mass, but is careful dissection through planes of the neck to get all the ramifications of the cyst. Occasionally, these procedures need to be staged when the mass is very large.
Swallowing Disorders in Infancy

Stanley E. Fisher, Michael Painter, Gregory Milmoe


Disorders of deglutition are frequently encountered in pediatric patients, especially in association with neurologic deficits. These swallowing abnormalities may lead to chronic lung disease and to marked malnutrition, thereby posing difficult and often frustrating problems of long-term management. This article provides an overview of swallowing disorders in infancy. More detailed information may be found elsewhere. As will become evident, therapy of swallowing dysfunction is often complex and symptomatic. Much is yet to be done to delineate the underlying pathophysiology of pediatric swallowing disorders.

Development of Swallowing

The ability to swallow begins during the second trimester of fetal life. Normal fetal swallowing may contribute to the complex maternal-fetal regulation of amniotic fluid volume. For example, mothers who deliver infants with esophageal atresia often have polyhydramnios.

The suck involves labial and buccal constriction as well as initial elevation followed by posterior thrust of the tongue. In premature infants, the suck, or oral phase of swallowing, is usually weak and readily subject to fatigue. In healthy term babies, the oral phase is adequate for sustained extraction of liquid nutrient through a nipple. However, because of tongue movement, intake of solids is usually difficult, as the nutrients are forced up against the hard palate and back out through the lips. By 3 to 4 months of age, most infants can handle solid foods with a modicum of facility. In North American society, solids are sometimes introduced as early as 2 weeks, although exclusive formula or breast feeding is usually an adequate source of nutrition during the first 6 to 12 months of life.

After the suck, the bolus of food passes to the posterior tongue and is propelled into the pharynx. With pharyngeal constriction, the upper esophageal sphincter (UES) relaxes, the cricoid cartilage moves forward, and the sphincter contracts, forcing the bolus into the esophagus. This aspect of swallowing usually takes less than one second and is well developed in term infants.

Normal Neurophysiology of Swallowing

Swallowing may be conveniently divided into three phases: oral (delivery of food into the mouth), pharyngeal (passage of food through the pharynx with elevation of the soft palate and closure of the epiglottis), and upper esophageal (relaxation and contraction of the UES). The neurologic coordination of swallowing is primarily a brain stem function involving several cranial nerves and a swallowing center located in the reticular substance of both the medulla oblongata and pons.

The coordination of suck and the oral phase of swallowing is mediated through the cranial nerves, which supply the facial structures essential for swallowing. Motor supply of the muscles of facial expression is through the seventh cranial nerve and sensation is through
the fifth cranial nerve. Sensation in the palate and uvula is mediated through branches of the
ninth cranial nerve. The motor supply of the tensors of the palate is through ninth and tenth
cranial nerves. The twelfth cranial nerve is responsible for supplying motor function to the
tongue, while taste derives from the seventh, ninth, and tenth cranial nerves. Interruption of
either the sensory or motor aspects of oral and early pharyngeal phase innervation will lead
to a poor suck and failure to propel food into the pharynx. Also, the nasal passages will not
be properly sealed off by the soft palate, and nasal regurgitation of food may occur.

The bundles of muscles surrounding the pharynx interlace to form virtually a
continuous sheet, although these muscles can be divided into superior, middle, and inferior
constrictors. The UES consists primarily of the cricopharyngeus muscle. Primarily the vagus,
trigeminal, spinal accessory, and glossopharyngeal nerves are involved in the pharyngeal and
upper esophageal phases of deglutition. Functionally and anatomically, the pharynx has a
rather rich supply of sensory nerves that are connected to the central nervous system by the
glossopharyngeal nerve. Innervation of the UES is predominantly through the vagus nerve,
although there are contributions from the ninth and eleventh cranial nerves. The activity of
the pharyngeal component of swallowing is totally dependent on normal central nervous
system function, as loss of either the brain stem swallowing center or the motor nuclei of the
cranial nerves produces loss of pharyngeal contraction. The UES and upper portions of the
esophageal body are only partially dependent on the central nervous system. The swallowing
center is not essential for the esophageal phase of swallowing.

Classification and Presentation of Swallowing Disorders

In infancy, swallowing disorders may present as isolated feeding difficulties but most
commonly are seen in infants with multiple problems. The most frequent underlying problem
is either neurologic or myopathic. Central nervous system injury, such as intrauterine
infection, vascular accidents in the newborn period, hydrocephalus, congenital porencephaly,
or tumor, may cause swallowing difficulty. Neurologic or myopathic disorders of the mouth,
pharynx, or esophagus may also impair swallowing. A list of such disorders is found in Table
1 and is reviewed by Painter.

Disorders of the Oral Phase

Difficulty with the oral phase usually presents as a "poor suck". primary dysfunction
of the oral phase may be the result of an anatomic abnormality such as cleft lip and palate
or micrognathia. Premature infants have decreased suck strength and stamina, which may
resolve within a few days.

Secondary problems with sucking are often related to neurologic deficits. Infants with
severe cortical deficiency are usually lethargic and cannot sustain a good suck. There may
also be cranial nerve damage which results in poor tongue function (twelfth nerve) or
decreased buccal strength (seventh nerve). Seventh nerve damage may also present as
unilateral salivary drooling. Finally, neurologic deficits that impair swallowing may be
aggravated or induced by extensive neurosurgical procedures such as tumor removal.
Table 1. Neurologic and Myopathic Disorders of the Mouth, Pharynx, and Esophagus

**Neurologic Disorders Involving the Mouth**

Lingual, gingival, glossal, and buccal mucosae
- Ulcerative nodular and inflammatory lesions
  - Ataxia telangiectasia
  - Chédiak-Higashi disease
  - Behçet's disease
  - Herpes simplex
  - ECHO virus
  - Histoplasmosis
  - Blastomycosis
  - Actinomycosis
  - Vitamin deficiency
  - Toxins

Pigmentary and vascular lesions
- Adrenoleukodystrophy
- Sturge-Weber disease
- Hereditary hemorrhagic telangiectasia
- Fabry's disease
- Tangier disease

Neurologic disorders involving the teeth and gingivae
- Neurofibromatosis
- Lesch-Nyhan syndrome
- Parathyroid disorders
- Mucopolysaccharide disorders
- Tuberous sclerosis
- Melkersson's syndrome
- Familial dysautonomia
- Klippel-Feil syndrome
- Duane's syndrome
- Encephalocoeles
- Chromosomal disorders

Neurologic Disorders Involving the Pharynx and Esophagus

- Infectious mononucleosis
- Brain stem lesions
- Myotonic dystrophy
- Oculopharyngeal dystrophy
- Myasthenia gravis
- Neonatal
- Juvenile
- Dermatomyositis
- Cricopharyngeal achalasia
- Cerebral palsy.

**Disorders of the Pharyngeal Phase**

Abnormalities of pharyngeal function typically lead to feeding difficulties characterized by coughing, gagging, nasopharyngeal regurgitation, failure to gain weight, or respiratory distress secondary to aspiration. Emesis may accompany the coughing and gagging, requiring differentiation from primary spontaneous gastroesophageal reflux.

An anatomic abnormality such as cleft palate or weak, inadequate closure of the soft palate may contribute to pharyngeal phase dysfunction. However, our experience has been that
feeble pharyngeal function (weak pharyngeal contraction) or incoordination of pharyngeal closure is more common.

Patients with inadequate or incoordinated pharyngeal contraction will show initial nonpropulsive pharyngeal activity on barium swallow. The bolus will stop abruptly at the upper esophagus. However, the x-ray findings may be confused with true cricopharyngeal achalasia because the UES has relaxed and contracted before the pharynx finally propels a bolus of barium toward the esophagus. Such pharyngeal-cricopharyngeal (P-CP) incoordination can be differentiated from true cricopharyngeal achalasia by manometric studies. In most cases, P-CP incoordination and feeble pharyngeal function are of neurologic or myopathic origin.

Permanent or transient pharyngeal dysfunction may also be traumatic or iatrogenic. Specifically, a foreign body may lodge in the hypopharynx resulting in pharyngeal edema with painful, inadequate pharyngeal contraction. When a foreign body is suspected, airway and pharyngeal endoscopy is mandatory. We have found that the most dangerous pharyngeal foreign body is the small battery found in 35 mm cameras. These batteries cause severe chemical injury within minutes of lodging in the pharynx. Similarly, trauma to the pharynx, such as ingestion of a caustic substance or falling on a sharp object held in the mouth, may lead to sterile or purulent swelling of the pharynx with subsequent pharyngeal dysfunction. Again, endoscopy is often indicated in these patients. Finally, neurosurgical procedures that involve the brain stem may iatrogenically alter pharyngeal contraction.

Disorders of the Upper Esophageal Phase

Failure of relaxation of the UES and lack of coordination between the pharyngeal and esophageal phases are the two most common swallowing disorders we see. Patients present with essentially the same symptoms as those with pharyngeal phase dysfunction and, after barium swallow, are often lumped under the diagnosis of cricopharyngeal achalasia. Although some infants appear to have no underlying cause of UES dysfunction, most have neurologic disorders. Occasionally, the patients have vocal cord paralysis and present with stridor as well as swallowing difficulty. Both manometric and endoscopic procedures are often necessary, in addition to x-ray evaluation, to define the nature and extent of apparent UES dysfunction.

Disorders That Simulate Swallowing Disorders

In some of our neurologically impaired patients, disordered esophageal motility with lack of peristalsis has led to regurgitation during swallowing. This can result in symptoms typical of pharyngeal or upper esophageal phase dysfunction, yet manometric studies show normal P-CP coordination with UES relaxation. Also, we have cared for infants who were initially thought to have swallowing disorders because of coughing and gagging during feeding; yet x-ray, manometric, and pH monitoring studies revealed only spontaneous gastroesophageal reflux, and the patients have responded to appropriate therapy. Finally, vascular anomalies, such as a vascular ring, which partially obstruct the trachea and/or esophagus, may present as feeding difficulty with or without stridor. Both x-ray and endoscopic procedures are necessary to identify these abnormalities.
Evaluation of Swallowing Disorders

When historical and physical (especially neurologic) findings suggest a possible swallowing disorder, clinical and laboratory evaluation is required to confirm the diagnosis.

Clinical Evaluation

Careful clinical observation is essential in the evaluation of patients with swallowing disorders. Such observations should be performed initially by the physician. Also, at most children's medical centers, including Children's Hospital of Pittsburgh, nurse specialists concentrate on infants with feeding problems. Their clinical impression of the nature of the feeding difficulty often aids the physician in directing laboratory evaluation. For example, when the history suggests psychosocial deprivation or rumination as a possible etiology, the careful attention of the nurse specialist, working with the mother as well as the infant, frequently reveals no swallowing difficulty at all. Conversely, the nurse specialist can readily identify poor sucking, nasopharyngeal regurgitation, postprandial emesis, and other conditions, and call the findings to the physician's attention.

The other aspect of clinical evaluation involves nutrition. Nutritional rehabilitation is essential to the continuing care of infants with swallowing disorders. It is necessary to assess the nutritional status of the patient, as the degree of nutritional deficiency may determine the urgency of surgical intervention (for example, central hyperalimentation or feeding gastrostomy). Initial assessment involves use of growth grids to estimate growth velocity, estimation of adipose tissue (skin fold thickness), and blood tests (hemoglobin, albumin, folate, ferritin). More importantly, continued assessment of nutritional status is required in hospital as well as after discharge. This should be done in conjunction with personnel experienced in nutritional management, such as a member of the hospital nutrition division. Nutritional monitoring should continue throughout infancy, especially after surgical procedures have been performed to alleviate the swallowing disorder or to permit introduction of adequate calories.

Radiology

The barium swallow and TV-tape esophagram remain the primary laboratory diagnostic tools in the evaluation of swallowing dysfunction. The radiologic findings in swallowing disorders are covered elsewhere in this issue.

Manometry

Manometry is useful in further characterization of swallowing dysfunction following radiologic evaluation. In particular, it is possible to distinguish between isolated cricopharyngeal achalasia and P-CP incoordination. Both conditions may appear the same on barium swallow, including the presence of the so-called cricopharyngeal bar. Manometry can also quantitate sufficiency of such and pharyngeal contraction. Finally, esophageal function can be evaluated and manometry plus pH monitoring can help to rule out spontaneous gastroesophageal reflux.
At the Children's Hospital of Pittsburgh, we use the Honeywell (5808C) recording system (Honeywell, Denver, Co) with a probe containing three solid-state pressure measuring devices set 2.5 cm apart (Konigsberg Instruments, Pasadena, Ca). The solid-state transducers are of low compliance with rapid response and can be oriented in an anterior-posterior position. These are important characteristics for the evaluation of UES dynamics.

Infants under 6 months of age usually are not sedated; older infants and children are sedated with 2 mg per kg each of meperidine and secobarbital. After the probe is passed through a pacifier into the throat, it is advanced so that the transducers all lie within the stomach. The probe is then slowly pulled back so that the location and resting pressure of the lower esophageal sphincter can be determined. Following this, with the three transducers in the midesophagus, the infant is stimulated to swallow by gentle tugging on the pacifier or injection of 1 mL of normal saline into the mouth. The presence or absence of esophageal peristalsis is thereby determined. Finally, the probes are withdrawn so that one sets in the pharynx, one in the upper esophageal sphincter, and the last in the upper esophagus. Swallowing activity is usually spontaneous at this point of the examination. With the probe in this position, the pharyngeal and upper esophageal phases of swallowing are readily evaluated. After completion of manometric studies, the acid reflux test is used to test for spontaneous gastroesophageal reflux.

**Endoscopy**

Direct visualization of the mouth, nasopharynx, oropharynx, esophagus, and upper airway may be accomplished by endoscopy with flexible or rigid instruments. Such an inspection gives a view of the structures but no indication of their function. However, endoscopy may provide more accurate information when trauma or obstruction is a major consideration. Rigid endoscopy has advantages in terms of depth of field and ease of foreign body removal. Flexible techniques allow air insufflation to distent a viscus so as to see the entire luminal surface. Osteophytes and other vertebral anomalies may present a greater problem for rigid endoscopes than for flexible ones. Infants with airway compromise sometimes do better with the narrower rigid esophagoscope than with the flexible one (6 mm versus 9 mm diameter). Esophagoscopy should be performed after manometry, as the procedure may be traumatic enough to temporarily alter manometric readings of swallowing.

**Therapy**

Treatment of children with swallowing disorders is directed at eventual amelioration of the underlying problem and at improvement of the patient's nutritional status. One wants to avoid the consequence of continued malnutrition in patients who are at a stage of active development. In the less severe cases, this would mean adaptation of the feeding pattern to obtain maximal intake and avoid aspiration. Position during and after feeding, volume of each feeding, consistency of the food, use of high caloric dietary supplements, feeding tubes, and different shapes and sizes of nipples must be individualized. This requires careful observation by both the physician and nurse specialist.

For the more severely affected child, either gastrostomy or intravenous hyperalimentation may be necessary. With gastrostomy, the patient can more easily be managed at home, although a few institutions have been successful with home
hyperalimentation in infants. It has been our experience that some patients with isolated swallowing problems may actually outgrow their dysfunction with time. Thus, gastrostomy allows the patient sufficient time for "maturation" of swallowing function while permitting adequate nutrition. This may be illustrated by the case of a 5 month old infant with cricopharyngeal achalasia that resolved spontaneously at 9 months of age. The infant originally was hospitalized because of choking, vomiting, and aspiration pneumonia. On x-ray, barium was aspirated into the lungs although cricopharyngeal achalasia was not appreciated. Esophageal manometry reveal cricopharyngeal achalasia with inadequate relaxation of the UES as well as P-CP incoordination. It was elected to delay any surgical procedure on the cricopharyngeus, and a gastrostomy was performed. Four months later, the patient as slowly advanced on oral feeding and by 10 months of age was on a normal oral diet. No further evidence of choking or aspiration pneumonia occurred.

In addition to the use of gastrostomy to provide the patient with adequate nutrition, cricopharyngeal myotomy has been recommended as a definitive treatment for swallowing dysfunction. The efficacy of this procedure depends on making the UES patulous. This would be theoretically beneficial for a patient with normal oral and pharyngeal phases of swallowing but isolated failure of relaxatin of the cricopharyngeus. It might also aid an individual with normal oral and upper esophageal phases but feeble pharyngeal function. At this point, there have been insufficient studies to indicate the value of cricopharyngeal myotomy. In addition, it is not possible to identify the ideal patient for such a procedure. Finally, the possibility of the patient outgrowing the problem must always be kept in mind when considering surgery early in the course of illness.

**Summary**

It should be evident that more needs to be learned about swallowing disorders in infants, especially since current modes of therapy are usually nonspecific and sometimes unsatisfactory. Specifically, there is a need to be able to identify infants who will "outgrow" their problem versus those in whom a permanent disability exists which requires more aggressive surgical intervention. Finally, it cannot be overemphasized that nutritional assessment and management is critical to the care of infants with moderate to severe swallowing problems.
Home Care of the Child with a Tracheotomy Tube

Sarah Foster, Dorothy Hoskins


Over the past decade, the number of children being discharged from the Children's Hospital of Pittsburgh with a tracheotomy tube in place has risen sharply. The home care of the child with a tracheotomy tube, once believed to be an impossible task, has now become almost routine. The excellent survival rate of these children at home results from the motivation of parents and the extensive training that they receive before the discharge of their child from the hospital. Table 1 lists the diagnoses most frequently encountered in children in whom long-term tracheotomy tubes are placed.

Table 1. Diagnoses Made in 136 Children with Long-Term Tracheotomy Tubes (Children's Hospital of Pittsburgh, 1968 to 1980)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>No</th>
<th>Total (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vocal cord paralysis</td>
<td>19</td>
<td>14</td>
</tr>
<tr>
<td>Croup</td>
<td>17</td>
<td>12.5</td>
</tr>
<tr>
<td>Hemangioma</td>
<td>12</td>
<td>8.8</td>
</tr>
<tr>
<td>Subglottic stenosis</td>
<td>11</td>
<td>8</td>
</tr>
<tr>
<td>Surgical procedure*</td>
<td>8</td>
<td>5.9</td>
</tr>
<tr>
<td>Brain damage**</td>
<td>8</td>
<td>5.9</td>
</tr>
<tr>
<td>Down's syndrome***</td>
<td>7</td>
<td>5.2</td>
</tr>
<tr>
<td>Brain tumor</td>
<td>6</td>
<td>4.4</td>
</tr>
<tr>
<td>Stridor</td>
<td>3</td>
<td>2.2</td>
</tr>
<tr>
<td>Juvenile papilloma</td>
<td>3</td>
<td>2.2</td>
</tr>
<tr>
<td>Prematurity****</td>
<td>3</td>
<td>2.2</td>
</tr>
<tr>
<td>Pierre Robin syndrome</td>
<td>2</td>
<td>1.5</td>
</tr>
<tr>
<td>Guillain-Barré syndrome</td>
<td>1</td>
<td>0.7</td>
</tr>
<tr>
<td>Cystic fibrosis</td>
<td>1</td>
<td>0.7</td>
</tr>
<tr>
<td>Miscellaneous*****</td>
<td>35</td>
<td>25.7</td>
</tr>
</tbody>
</table>

* Includes such procedures as cardiac surgery and tracheoesophageal fistula.
** Includes brain damage received from such sources as battered child syndrome and near drowning.
*** Patients underwent cardiac surgery.
**** Premature infants had long intubations for lung disease, and other surgical conditions.
***** Remaining diagnoses include automobile accidents, tumors, muscular dystrophy, accidents at home, and so forth.

The role of the home care coordinator begins once the family has been informed by the physician that a tracheotomy will be necessary. Support is offered to the family, and the groundwork is laid for future teaching. The age of the child, the capabilities of the parents, the social situation and lifestyle of the family, and fears and concerns that family members may have about home care should all be considered in planning an approach to parent
teaching. Parents will be apprehensive and question their ability to care for the child. They may have questions about the length of time the tube will be in place and the possibility of unwittingly doing something to harm their child.

The home care coordinator should also be sensitive to complications or other medical conditions that may present special problems in care, especially of infants. The baby who must undergo a tracheotomy is often separated from the mother soon after birth and may require a long stay in the hospital. Mothers may experience feelings of guilt or mourning in giving birth to a baby with a defect. If this is the mother's first child, she will need to learn routine baby care in addition to the special tracheotomy care. As soon as she is physically able to do so, the mother should be encouraged to become involved in the baby's care.

A tracheotomy tube in an older child, who can make his needs known to others and who has achieved some independence, is usually not as stressful to parents, especially after the child resumes normal play and daily activities. Sometimes stress can be relieved by introducing parents to other parents who have children with tracheotomy tubes. The parents who are experienced in home care can offer support and encouragement as well as, perhaps, helpful suggestions for care.

After the first 48 hours post-tracheotomy, and if there are no complicating factors, parents can begin to become involved in the care of the child. Holding and feeding the child helps to reestablish parental bonds. Under the supervision of hospital personnel, parents learn to care for the child and his new way of breathing. Knowing when to suction the child is a concern of parents. This judgment is developed as the parent spends time caring for the child in the hospital under our supervision. We encourage parents to spend at least one night with the child before discharge from the hospital. A diagram is used to explain to parents the alteration in the child's breathing and the temporary inability of the child to talk or make noise. However, in all other respects, including eating, the child is normal.

When the tract in the neck is well formed, parents can be taught to change the tube. The tube should be changed at least weekly to avoid occlusion by dried mucus or mucous plug, or whenever there is any sign of respiratory distress. Although initially this task produces the most anxiety in parents, once they have accomplished it successfully they are much more at ease.

A practical, clean rather than sterile procedure has been developed at the Children's Hospital of Pittsburgh for suctioning in the home, and no infections resulting from this procedure have been reported. The home care coordinator should explain the necessary equipment and technique to the parents. The equipment includes: a suction machine, tracheotomy tubes and suction catheters which are washed with soap and water and reused, a cool mist humidifier to be used especially when the child sleeps, cotton twill tape for ties, a dropper bottle to instill saline, and DeLee traps for suctioning away from home. The suction machine can be rented or purchased at a medical supply store; humidifiers are available in most drugstores; and tracheotomy tubes, catheters, and twill tape are available through the hospital. The cannula of the tube is often shortened for small infants, and the outer cuff on the Shiley tube is sometimes shortened to allow forward movement of the head, especially for infants with neuromuscular problems. The end of the cannula is then smoothed with an acrylic bur drill in the dental department.
Parents are encouraged to treat their child as normally as possible in order to allow normal growth and development, and to use discipline appropriate to the level of the child. Playing with such toys as swings and bicycles should not be forbidden if the child is at an age to enjoy them. Talking to and playing with the child just as for any other child will stimulate normal lingual development; as the child grows, and if no other complications develop, he will follow the usual pattern: first babbling, making noises, then learning to say words. Three of our children go to school and several babies have been camping!

However, developmental delays may occur in infants with vocal cord paralysis or other neurologic problems. Home care planning for these infants should include referral to an infant stimulation program. Because such children often stay in hospital for longer periods of time, the program usually begins in the hospital and involves the parents, who learn a variety of techniques to help their child.

Infants with bronchopulmonary dysplasia and a tracheotomy require extra moisture to the trachea (which is supplied by a nebulizer setup), frequent chest physical therapy and suctioning, and possibly oxygen, an apnea monitor, and an Ambu bag. With diligent care, parents can accomplish satisfying results with these infants at home. These infants also require continued involvement with an infant stimulation/OT, PT program at home.

Some parents quickly learn home care techniques and are anxious to take their child home, whereas others require a longer learning period. The particular situation of the family may require the help of a social worker, and the home care coordinator should make arrangements for this supplemental assistance. Parents should be taught to prevent or manage emergencies at home, such as occlusion caused by a mucous plug, accidental extubation, bleeding, or excessive dryness in the trachea.

Home care of the child with a tracheotomy tube shortens the hospital stay, allowing the child to return to his home environment and a normal lifestyle sooner. The intensive teaching provided by the home care coordinator makes this possible. Until a repeat endoscopy determines that the tracheotomy tube can be safely removed, the home care coordinator provides continuity of care for the family and joins with them in solving problems that may arise in home care. Often practical solutions are found that can then be passed on to other families with similar problems. Home care of the child with a tracheotomy tube has proved to be a valuable and a viable alternative to prolonged hospitalization.
Acute Upper Airway Obstruction: Croup and Epiglottitis


In few pediatric conditions are the benefits of optimal management and the risks of inappropriate action so clear as they are in acute upper airway obstruction. Infants and small children are uniquely susceptible to these disorders as a consequence of several anatomic and physiologic factors, and though severity of obstruction and rapidity of progression vary depending upon cause and time of presentation, precipitous decompensation to complete obstruction and/or respiratory failure is an ever present possibility.

Critical management decisions depend upon rapid assessment of the approximate level of the airway affected and of the degree of respiratory distress. In view of the difficulties of anesthetic delivery and securing the compromised airway of a small child, the services of a team of physicians experienced in such management are required in severe cases. Finally, all measures must be carried out in a manner that does not upset the child in order to avoid exacerbating his condition. Appreciation of these factors, greater understanding of the various distinct disorders, and advances in techniques of medical and surgical care have substantially reduced resultant morbidity and mortality. At our institution we have found that in addition to having a skilled team and advanced facilities, the use of a standard protocol greatly facilitates management.

With the goal of sharing practical information concerning approach and management, this article is devoted to presentation of an overview of the clinical features of acute upper airway disorders followed by more detailed discussions of croup and epiglottitis, two of the most common and important of these conditions. Emphasis is on providing guidelines for assessment, initial intervention, and airway stabilization and management, which we have found useful and successful.

General Approach to Acute Upper Airway Obstruction

Level of Airway Involvement

All forms of acute upper airway obstruction tend to present with stridor, retractions that are primarily suprasternal and subcostal (with mild distress), and mild to moderate increases in respiratory rate and heart rate. Beyond this there is considerable variability among the conditions. From a management standpoint, the major causes can be divided conveniently into two main categories: those involving supraglottic structures (severe tonsillitis with adenoidal enlargement, peritonsillar and retropharyngeal abscesses, epiglottitis/supraglottitis), and those affecting subglottic areas (croup, foreign bdy aspiration, and angioedema). Table 1 depicts the major similarities and differences between the two categories in terms of clinical signs and symptoms. The mode of onset and the age of the patient can also help to distinguish among etiologies (Table 2).
Severity of Respiratory Distress

Since studies such as blood gases and x-rays can prove hazardous, less precise clinical means of determining severity of distress must be used. As a general rule, stridor tends to worsen as obstruction increases; however, when airway compromise becomes severe, air entry is so diminished that audible sounds decrease. Cyanosis does not become evident until the \( \text{PO}_2 \) is less than 40, and thus is a late sign. Anxiety or restlessness and tachycardia at rest are early indicators of hypoxia. Bearing the above in mind, the signs enumerated in Table 3 can be used to help roughly gauge severity. If the child has severe symptoms in any one category, he must be considered to be in severe distress and treated accordingly.

Table 1. Clinical Features of Acute Upper Airway Disorders

<table>
<thead>
<tr>
<th>Supraglottic Disorders</th>
<th>Subglottic Disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stridor</td>
<td>Quiet and wet</td>
</tr>
<tr>
<td>Voice alteration</td>
<td>Muffled</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>+</td>
</tr>
<tr>
<td>Postural preference</td>
<td>+</td>
</tr>
<tr>
<td>Barky cough</td>
<td>-</td>
</tr>
<tr>
<td>Fever</td>
<td>+</td>
</tr>
<tr>
<td>Toxicity</td>
<td>+</td>
</tr>
<tr>
<td>Trismus</td>
<td>+ usually in quinsy</td>
</tr>
<tr>
<td>Facial Edema</td>
<td>-</td>
</tr>
</tbody>
</table>

Assessment Techniques

The major errors in initial management of upper airway problems are underestimation of distress, overzealous examination, and performance of laboratory studies that disturb the child, thereby worsening his condition and defeating therapeutic goals. Every effort should be made to keep the patient calm, to maintain his sense of security, and not to separate him from his parents. Parents should be encouraged to hold the child or sit next to him and assist as needed. The child should be allowed to maintain the position in which he is most comfortable, and at no time be restrained. Fast movements that might be seen as threatening should be avoided. If supraglottic disease is likely or distress is moderate to severe, laboratory studies should be withheld and visualization of the mouth and airway should be deferred until this can be done under controlled conditions by a team skilled in establishing an airway.

History

A brief chronology of the illness with questions concerning mode of onset and signs and symptoms is needed, along with information about antecedent illness and events. Past history of cardiorespiratory and allergic disorders is also important.
Table 2. Additional Features of Acute Upper Airway Disorders

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Mode of Onset of Respiratory Distress</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe tonsillitis</td>
<td>Gradual</td>
</tr>
<tr>
<td>Peritonsillar abscess</td>
<td>Sudden raise in temperature, toxicity and distress with unilateral throat pain, following earlier tonsillitis</td>
</tr>
<tr>
<td>Retropharyngeal abscess</td>
<td>Sudden raise in temperature, toxicity and distress after preceding URI or pharyngitis</td>
</tr>
<tr>
<td>Epiglottitis</td>
<td>Very acute onset of high temperature with rapid progression of dysphagia and distress in previously well child</td>
</tr>
<tr>
<td>Croup</td>
<td>Sudden onset (usually at night) of loud stridor and barky cough after preceding URI</td>
</tr>
<tr>
<td>Foreign body aspiration</td>
<td>Sudden choking episode while eating nuts, carrots, or chewing on small object, followed by onset of distress either immediately or, more typically, following a silent period of a few hours</td>
</tr>
<tr>
<td>Angioedema</td>
<td>Sudden onset shortly after eating, bee sting, or environ. exposure.</td>
</tr>
</tbody>
</table>

Table 3. Estimation of Severity of Respiratory Distress

<table>
<thead>
<tr>
<th>Color</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Retractions</td>
<td>Normal</td>
<td>Normal</td>
<td>Pale, dusky or cyanotic</td>
</tr>
<tr>
<td>Air entry reduction</td>
<td>Normal</td>
<td>Moderate</td>
<td>S e v e r e a n d generalized with use of accessory muscles</td>
</tr>
<tr>
<td>State of consciousness</td>
<td>Normal</td>
<td>Moderate</td>
<td>Severe</td>
</tr>
<tr>
<td></td>
<td>restless when disturbed</td>
<td>Anxious, restless when undisturbed</td>
<td>L e t h a r g i c , depressed.</td>
</tr>
</tbody>
</table>
Physical Examination

In addition to the attention to emotional needs described above, the practice of doing most of the examination from a distance minimize disturbance and enables more reliable observation by avoiding the increases in heart rate, respiratory rate, and retractions produced by crying. The parent is asked to remove the child's shirt, and the following are rapidly assessed from a few feet away: general appearance, color, respiratory rate, audible sounds, retractions and use of accessory muscles, drooling, and state of consciousness. Thereafter, the patient can be approached slowly in order to check the pulse and auscultate the chest. If these efforts provoke upset, they should be abandoned, as pulse will be unreliable and auscultatory findings altered. If the child has fever and sore throat without evidence of respiratory distress and without drooling or if drooling is present without respiratory distress and history is not typical for epiglottitis, a gentle attempt to examine the mouth may be made.

Note: Lethargy and depression, especially when accompanied by pallor and/or cyanosis, are ominous signs. In the absence of a skilled team, do not attempt more than the above examination if these signs are present, if the child is drooling, or if he demonstrates postural preference. Most particularly, do not attempt to examine the mouth, as this may precipitate laryngeal spasm, massive vagal discharge, and cardiorespiratory arrest.

Following rapid assessment, the approximate degree of distress and level of involvement can be determined, enabling the physician to select appropriate initial therapeutic measures (see protocols below).

Guidelines for Management if Patient Is Initially Seen in Office or Hospital Without Necessary Personnel and Facilities

Assess as described above.

If signs of supraglottic disease or moderate to severe subglottic disease:

Provide with oxygen, allowing parent to hold the mask.
Arrange transport by ambulance.
If child is small, have parent carry him to the ambulance.
Allow parent to hold or sit next to child in transit.
Let patient maintain position in which he is most comfortable, without restraints.
Transport with personnel capable of advanced life support.
Speed and use of siren should depend on child's tolerance.

Note: If the child should suffer a respiratory arrest in transit, brief suctioning of the pharynx followed by bag and mask ventilation with oxygen can be life-saving. Force of ventilation should be gauged to adequately expand the patient's chest. Attempting to bypass the obstruction using a large bore needle inserted through the cricothyroid membrane has been found unsuccessful.
Protocol for Initial Hospital Management of Acute Upper Airway Obstruction

Team members: Otolaryngology resident, ICU fellow, and anesthesiologist (all skilled at intubation), pediatric resident, and respiratory therapist.

1. Triage nurse rapidly assesses the child's status on arrival in the emergency room and takes him to a treatment room.

2. Room assignment:

   Moderate or severe distress of Signs of supraglottic disease --&gt; Critical Care Room (Equipped for advanced cardiopulmonary resuscitation).

   Mild distress with Subglottic symptoms --&gt; Observation / Intermediate care.

3. Critical care or intermediate care nurse asks parent to remove the patient's shirt and provides humidified oxygen.

   Note: If the patient is under 5 or evinces fear of the mask, it is given to the parent to hold as close as the child will tolerate.

4. As this is being done, the clerk summons the pediatric resident who comes immediately and assesses as described above.

5. As the pediatrician evaluates, the nurse prepares equipment that may be needed for direct visualization and intubation.

6. Action following initial assessment:

   a. Signs of supraglottic disease, severe subglottic disease, possible foreign body aspiration or angioedema --&gt; Team is paged to emergency room immediately.

   b. Mild to moderate croup --&gt; Trial of mist and medical therapy (see section on croup).

7. When the team arrive, the patient is rapidly assessed with a view to measures needed to secure the airway.

   Note: If the patient is in severe distress, direct examination and all studies are deferred until after an artificial airway has been established in the operating room.

5. Possible disposition

   a. To operating room if intubation, tracheotomy, or other surgery is urgently required.

   b. To intensive care unit or ward depending on condition for observation and therapy if operative intervention is not required or is best deferred.
c. Trial of mist and/or racemic epinephrine for croup with disposition dependent upon response (see section on croup).

d. Administration of epinephrine, benadryl, and steroids for angioedema (with team present).

Note: The parent is allowed to accompany the child to his destination and is separated from him only upon entering the operating suite.

9. Laboratory studies

a. X-rays may be obtained in cases of croup to assess degree of subglottic narrowing, in suspected epiglottitis in which direct visualization reveals a normal epiglottis, when diagnosis is unclear, and in cases of foreign body aspiration, provided distress is not severe and patient is accompanied by the parent and a physician. Portable films done in the emergency room are preferred.

b. Blood work and cultures are done only after the airway is felt to be secure. The same applies for starting an intravenous line.

Croup

The croup syndrome is rather unique in that it can cover the spectrum of minimal to life-threatening symptoms, and tends to have a highly variable course. Few diseases have generated greater emotion and less factual information regarding treatment; racemic epinephrine, steroids, intubation versus tracheostomy are just a few areas of controversy. Interpretation of research findings has been difficult because of major design flaws in most studies. Furthermore, the fluctuation in symptomatology typical of the syndrome and the number of individual factors that appear to affect outcome (anatomic and physiologic features unique to small children, age, sex, atopy, reactivity of lower airways, underlying illness, nutritional status, and emotional state of patient and parents) have made research design difficult and have complicated attempts to assess response to therapy. This review attempts to place our current view of croup into historical perspective and also presents our own approach based on the limited data available.

Historical Background

Prior to World War II, diseases such as diphtheria, epiglottitis, and infectious croup were considered together in discussions of upper airway obstruction. Since that time microbiologic and clinical research has allowed more appropriate separation. In 1941, Sinclair identified *Haemophilus influenzae* type b as the etiologic agent of acute supraglottitis, and Rabe in the late 1940s defined acute laryngotracheobronchitis as a separate, presumably viral illness. In 1974, Bucan et al identified parainfluenza viruses 1 and 3 and influenza virus as the primary etiologic agents of the disease. Additional epidemiologic studies in this country have confirmed that the largest outbreaks of croup tend to occur in the autumn of even numbered years and correlate with parainfluenza I virus activity.
Clinical Entities

Subsequent to separation from supraglottic disorders, the croup syndrome (involving edema and/or inflammation primarily in the subglottic area) has been subdivided into at least three different disorders: viral or infectious croup, bacterial tracheitis, and spasmodic croup. The clinical picture of viral croup is well known to all modern pediatricians: mild upper respiratory tract symptoms for a few days, then sudden onset of harsh "barking" cough with varying degrees of inspiratory stridor, waxing and waning course over several days (with symptoms usually worse at night), followed by full recovery in most patients without treatment; some patients need to be hospitalized and a few require intensive care and intubation or tracheostomy.

Earlier discussions of laryngotracheobronchitis are at variance with the current picture. In the 1940s Orton and others described a form of croup characterized by extreme toxicity and included autopsied cases with thick, mucopurulent material found throughout the upper airways. Secondary pyrogenic infection was strongly suspected. The cases of "bacterial tracheitis" recently reported by Jones et al strikingly resemble these earlier case histories. Their patients had a viral croup prodrome but then developed high fever, toxicity, and airway obstruction requiring intubation and suctioning. Purulent material, laden with bacteria, was found below the glottis. Over the past 18 months we have seen several patients resembling those with bacterial tracheitis. Typical viral isolates were obtained in two patients, suggesting that this entity may actually represent the reemergence of severe viral laryngotracheobronchitis, a possibility first mentioned by Cherry. Further studies with careful attempts at viral and bacterial isolation will be necessary in order to determine whether the bacterial infection is primary or secondary in these cases.

Spasmodic croup is another, and probably clinically distinct, entity. Patients have no fever and minimal, if any, prodrome. Stridor develops almost instantaneously, usually at night after one or two brief coughs. Mist, cold air, or emesis can bring relief just as quickly. This illness may recur and may rarely, as we have seen, cause complete airway obstruction requiring intubation. The pathogenesis and relation to atopy are unknown but endoscopy in some patients has revealed edema and pallor rather than the inflammation and crusting characteristic of infectious croup. Table 4 offers a short comparison of the croup entities.

Management

Croup includes a number of disorders with different courses. Even if viral croup alone is considered, each seasonal epidemic may be secondary to a different virus and have variable severity. Over the past few years we have seen tracheostomy rates vary from four or five per season to several times this number. Bearing this and the complexities introduced by individual factors in mind, we will review current therapeutic modalities and our own clinical approach.

Initial recognition of croup and its separation from supraglottitis and foreign body aspiration are vital. Although the clinical picture is usually quite distinct, when doubt exists we evaluate and stabilize in the emergency room, and then either obtain posteroanterior and lateral x-rays of the neck if distress is mild, or bring the patient to the operating room for endoscopy under general anesthesia if distress is moderate to severe. We feel strongly that it
is better for an occasional child with croup to undergo controlled laryngeal inspection than to run the risk of precipitating a cardiorespiratory arrest in a child with unrecognized supraglottitis.

**Table 4. Croup Syndrome - Comparison of Major Entities**

<table>
<thead>
<tr>
<th></th>
<th>Viral Croup</th>
<th>Bacterial Tracheitis</th>
<th>Spasmodic Croup</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>&lt; 3yrs (mean 21 mo)</td>
<td>1 mo-6 yrs (N=6)</td>
<td>Childhood</td>
</tr>
<tr>
<td><strong>Etiology</strong></td>
<td>Viral</td>
<td>Prior URI</td>
<td>Atopy (?)</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>Prior URI</td>
<td>Rapidly progressive,</td>
<td>Sudden, usually</td>
</tr>
<tr>
<td></td>
<td></td>
<td>toxicity, airway obstr</td>
<td></td>
</tr>
<tr>
<td><strong>Course</strong></td>
<td>Waxing and waning, usually worse at night, generally mild</td>
<td>E d e m a , inflammation, pus below cricoid</td>
<td>Rapid resolution</td>
</tr>
<tr>
<td><strong>Endoscopic Findings</strong></td>
<td>E d e m a , inflammation, and crusting, primarily in subglottic area</td>
<td>Artificial airway, antibiotics</td>
<td>Subglottic pallor and edema</td>
</tr>
<tr>
<td><strong>Management</strong></td>
<td>See text and Table 6</td>
<td>Spontaneous resolution and induced emesis.</td>
<td>Self-limited, induced emesis.</td>
</tr>
</tbody>
</table>

Patients with typical croup are evaluated clinically using a croup scoring system (Table 5) modified from Taussig by Davis and Galvis. This enables assessment of the approximate degree of distress not only initially, but also over time, when repeated at intervals. It is also useful in determining response to short-term modes of therapy such as mist and racemic epinephrine. Finally, the score or pattern of scores can be used to help determine disposition (Table 6).

**Humidification and Oxygen**

Humidification of inspired gases to prevent drying of secretions makes sense in croup therapy, as droplets are actually delivered to at least a portion of the diseased area. Several studies show improvement in croup scores when saline (by intermittent positive pressure breathing (IPPB)) and mist therapy ("croup room") are used alone. We usually use Plexiglas hoods for infants in they fit properly and are tolerated, and croup tents for larger preschool age children who are too young to tolerate use of a mask. In addition to humidification, adequate hydration by oral and intravenous routes is emphasized in order to further decrease the likelihood of inspissation of secretions.
Table 5. Subjective Assessment of Clinical Severity of Croup

<table>
<thead>
<tr>
<th></th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stridor</td>
<td>None</td>
<td>Mild</td>
<td>Moderate</td>
<td>Severe on inspiration and expirations or none with markedly decreased air entry</td>
</tr>
<tr>
<td>Retraction</td>
<td>None</td>
<td>Mild</td>
<td>Moderate</td>
<td>Severe, marked use of accessory muscles</td>
</tr>
<tr>
<td>Air entry</td>
<td>Normal</td>
<td>Mild decrease</td>
<td>Moderate decrease</td>
<td>Marked decrease</td>
</tr>
<tr>
<td>Color</td>
<td>Normal</td>
<td>Normal (0-score)</td>
<td>Normal (0-score)</td>
<td>Dusky or cyanotic</td>
</tr>
<tr>
<td>Level of Normal conscious.</td>
<td>Restless when disturbed</td>
<td>Anxious, agitated; restless when undisturbed</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 6. Guidelines for Disposition of Cases of Croup

**Mild Cases (Score 4-5 or Less)**

Child is given trial of mist therapy and usually improves significantly. He is then sent home with recommendations for observation, use of a vaporizer, emphasis on oral fluids, and antipyretics as needed. Symptoms of increasing distress (such as increased stridor, retractions, and anxiety at rest) are described so that parent has a clear idea of when to return to hospital.

**Mild to Moderate Cases (Score of 5-6)**

Disposition depends largely on child's response to mist. Patient's age, time of day, distance from hospital, and parent's condition are also taken into consideration. Older children who present at night, respond well to mist, and live nearby are probably best managed at home. Infants, children with known increased risk factors (history of prior intubation, known subglottic stenosis), those presenting during the day time, and those who live a great distance away are considered for admission.

**Moderate Cases (Score of 7-8)**

Patient is admitted unless he is older, lives nearby, and has an outstanding response to mist. Otherwise he is given racemic epinephrine in the emergency room prior to transfer to ward or intensive care unit.

**Severe Cases (Score > 8, or Child Has Any One Sign in Severe Category)**

Team is mobilized as per protocol and racemic epinephrine is given as soon as possible. Older patients who respond well to this may be admitted to the ward for close
observation; younger patients and older children who have a fair response are admitted to the intensive care unit; those who have a poor response, or are tiring or cyanotic are admitted directly to the operating room for establishment of an artificial airway.

Oxygen use stems from the work of Newth who found hypoxemia as the predominant blood gas abnormality in croup and showed the correlation between oxygenation and respiratory rate. Although the authors recommended frequent blood gas determinations, we generally refrain from these outside an intensive care setting. Progressive airway obstruction should be observed clinically as the need for intubation or tracheostomy may precede a rise in pCO₂.

**Racemic Epinephrine**

Studies of postintubation croup and large uncontrolled studies of croup syndrome suggested that racemic epinephrine had dramatic effects on airway edema and obstruction. The first small controlled study indicated that mist rather than the drug was most important in therapy. A larger study by Westley showed that racemic epinephrine (delivered by IPPB) provided better temporary relief than saline (IPPB) for patients who failed to respond to initial mist therapy. Most observers believe that the drug works locally as a vasoconstrictor to decrease edema. Although racemic epinephrine (2.25 per cent) has been utilized in most studies, L-epinephrine (1 per cent) should work equally well, as it contains only the active isomer. Our present routine is to use racemic epinephrine (0.5 mL per 4 mL of normal saline) only on patients who are being admitted or are hospitalized as rebound may occur, though usually only to the croup score prior to therapy. The patient's response is noted using the croup scoring system. The need for increasingly frequent treatments with lesser degrees of improvement over time may indicate the need for an artificial airway. Thirty minutes is a minimum time between treatments. We generally use inhalation alone with a mask and nebulizer in struggling patients, reserving IPPB therapy for more cooperative children. As noted by Barker, IPPB may provide better entry of particulate material into the airway if there is tracheal collapse during inspiration.

**Steroids**

Perhaps the greatest controversy in the treatment of croup centers upon the role of steroids. Their known anti-inflammatory effects, particularly their actions to stabilize lysosomal membranes and to diminish capillary dilatation and permeability, have stimulated much interest in their application and a great deal of research. A recent article carefully analyzes the studies to date and finds significant methodologic problems with each, raising questions about validity of results.

A detailed analysis of each study is beyond the scope of this article, but several points may be made. Steroids may have beneficial effects in certain types of croup, but may also be potentially harmful, as in bacterial tracheitis. The most beneficial results from steroids appear to have been obtained when a single high dose was given early in the course; there are no data to support numerous repetitive doses.

Table reviews the controlled studies of croup; dosages have been converted to dexamethasone activity for a 10 kg child. While opinion on the use of steroids for croup is
divided at our institution, most clinicians use 1 to 1.5 mg/kg of dexamethasone (maximum 30 mg) intramuscularly as early as possible in the course of the illness.

Airway Management

Provision of an artificial airway is necessary in some patients with viral croup and certainly in most patients with bacterial superinfection. Tracheostomy was for many years the procedure of choice, as initial studies suggested that nasotracheal intubation might cause serious airway injury. However, a number of more recent studies suggest that carefully done nasotracheal intubation provides effective treatment for croup and may actually reduce morbidity when compared to tracheostomy. As there is currently no prospective, controlled study of this question, we have designed a protocol to be initiated in the near future. Until that time severe croup is treated by tracheostomy at our institution.

Several factors point to the need for airway support: patient fatigue, cyanosis, worsening obstruction (croup score), decreasing response to racemic epinephrine (both in terms of response to treatment and duration of interval between treatments), and toxicity with evidence of superinfection. As time is necessary to arrange for controlled intubation or tracheostomy, a worsening course must be appreciated promptly.

Epiglottitis

Epiglottitis (supraglottitis) is one of the most distinctive and dramatic diseases of childhood. The clinical course is fulminant and includes unique inflammatory and obstructive manifestations which interfere with swallowing, vocalization, and respiration. The disease is almost always caused by a single organism, Haemophilus influenzae type b. With early recognition and careful airway management, the once high case fatality rate can be reduced nearly to zero. This section described the principal clinical features and treatment of this condition, with special emphasis on early diagnosis and initial management. Much of the information for this report comes from our extensive experience with Haemophilus epiglottitis at Children’s Hospital of Pittsburgh, and a review of the medical records of 100 consecutive bacteremic cases successfully treated at this institution during the past nine years.

Clinical Picture

Epiglottitis is the term commonly used for an inflammatory condition that involves not only the epiglottis but also adjacent tissues, including the ventricular bands, arytenoids, and aryepiglottic folds. These structures may be involved in varying degrees, but the process typically does not extend down to the vocal cords or subglottic tissues. This distinctive supraglottic pathology accounts for the unique symptomatology and particularly for differences between the condition and that of laryngotracheobronchitis or croup. Stridor and respiratory distress may occur in both, but sore throat and dysphagia suggest epiglottitis whereas hoarseness and cough are more characteristic of croup. Epiglottitis also tends to occur in older children; the average age for our patients was 3.1 years (range seven months to 10 years), and more than one third were four years of age or older (see Tables 1 and 2).

The clinical presentation of our 100 cases is summarized in Table 7. The various manifestations are grouped by usual order of appearance and anatomic region involved. For
example, fever and sore throat were generally followed by swallowing difficulty, and this
soon progressed to respiratory distress. The timing of complaints is related to the first medical
visit, whether in a physician's office or in a hospital emergency room. This distinction might
be clarified by an example: respiratory distress was present in 56 patients prior to the first
medical visit but was noted in 67 by the time of hospital admission.

The earliest manifestation was often the sudden onset of high fever. Fever was also
the most common complaint and was noted in 88 of the 100 patients; fever with sore throat
was observed in 46 patients. It is therefore not surprising that 11 children were originally
thought to have pharyngitis. Four of the 11, however, also had one or more of the following
associated problems: drooling, dysphagia, voice change, or preference for a sitting position.

The second group of complaints included various manifestations of swallowing
difficulty. These were the most prominent complaints for many patients and were nearly
always associated with fever (sometimes with sore throat). Excessive drooling was the most
frequent of these complaints (noted in 49), and tended to occur several hours after the fever
was first noted. Review of the medical records sent from other hospitals at the time of transfer
of patients with epiglottitis showed that problems with swallowing were not always
recognized or deemed important enough to record, probably accounting for mistaken initial
diagnosis in several cases.

Table 7. Frequency of Early Complaints from 100 Consecutive Patients with
Epiglottitis (Supraglottitis) at Children's Hospital of Pittsburgh

<table>
<thead>
<tr>
<th>Complaint</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sore throat and feverishness (16 hours)*</td>
<td>46</td>
</tr>
<tr>
<td>Swallowing difficulties (one or more)</td>
<td>69</td>
</tr>
<tr>
<td>Excessive drooling (8 hours)</td>
<td>49</td>
</tr>
<tr>
<td>True dysphagia</td>
<td>27</td>
</tr>
<tr>
<td>Refusal to drink or eat</td>
<td>21</td>
</tr>
<tr>
<td>Respiratory or vocal problems (one or more)</td>
<td>84</td>
</tr>
<tr>
<td>Respiratory distress (5 hours)</td>
<td>56</td>
</tr>
<tr>
<td>Stridor or noisy breathing</td>
<td>30</td>
</tr>
<tr>
<td>Change in voice</td>
<td>28</td>
</tr>
<tr>
<td>Preference for sitting</td>
<td>16</td>
</tr>
<tr>
<td>Cough</td>
<td>6</td>
</tr>
</tbody>
</table>

* Time in parenthesis represents the mean duration of the three most common complaints
prior to the first medical examination.

Respiratory and vocal manifestations formed the third and most common group of
complaints. An indication of the rapidity of the course of this infection is the fact that the
onset of respiratory distress usually occurred within 12 hours of the first clinical
manifestation. Our children with epiglottitis were often reluctant to speak or had a muffled
voice, quite unlike the hoarse voice of croup. Stridor, if present, was softer than that of croup
and cough was infrequent (present in only six). The children with epiglottitis usually appeared
quite ill and often assumed a characteristic position during the later stages of the illness.
There was refusal to lie down and a preference for a sitting position with head forward, neck
extended, and mouth open. The child often drooled and there was an anxious expression. We
feel that it is important to stress these features since misdiagnoses continue to occur. Five of our patients were initially diagnosed as having croup despite the fact that three had a history of excessive drooling, and two of these three preferred the sitting position for easier breathing.

**Associated Problems**

All of our patients had been in good health prior to the onset of epiglottitis. Acute otitis media, however, was found in 14 children during the initial examination. A previous publication reported the frequent occurrence of cervical adenitis and pneumonia with epiglottitis, but we found only three children with enlarged, tender cervical lymph nodes and another five with probable pneumonia, two with pleural effusion. Nearly half of our patients had abnormal chest radiographs as some time during their hospital stay, but they probably represented atelectasis in most cases. Six of our children had clinical and radiographic changes consistent with pulmonary edema following intubation. There were no examples of other major manifestations of *H. influenzae* type b infection (meningitis or arthritis), and this is in agreement with other recent studies in which these conditions were specifically mentioned. Nine of our children had spinal fluid examination, with normal results in each case. Meningitis has rarely been associated with epiglottitis, possibly because of genetic differences in susceptibility to the two disorders; however there is one Scandinavian report noting this association in four of 25 cases of epiglottitis.

**Assessment and Initial Management**

Not infrequently parents call on the telephone for advice concerning a child with respiratory distress. In such situations, after taking a brief history, experienced physicians have found that either having the parent imitate the child's noisy respirations or listening to the child breathe over the telephone can enable them to assess the degree of distress and probable diagnosis with a fair degree of accuracy.

Guidelines for assessment and initial management, whether the child is first seen in the office or a hospital with the necessary facilities, have been described in the introductory section of this article. The importance of minimizing disturbance and avoiding attempts at visualization of the epiglottis in the absence of a skilled team cannot be overemphasized. There were five instances of respiratory arrest in our series. Three occurred at the time of initial examination, and one occurred in a child who had been strapped down on a stretcher for ambulance transport.

The clinical picture and course are usually so characteristic as to make diagnostic studies such as x-rays and cultures unnecessary prior to securing the airway. Furthermore, the risk of precipitating complete obstruction while doing them generally contraindicates them. There are exceptions, however. Occasionally a child will present with a history and signs suggestive of epiglottitis, but with minimal signs of obstruction and little drooling, or has fever, sore throat, and mild drooling with an atypical history. In such cases visualization may be attempted and may reveal pharyngeal infection with no epiglottic swelling or other pathologic findings may be evident. In the former instance, a portable lateral neck x-ray can be extremely helpful, often demonstrating swelling of the aryepiglottic folds and ventricular bands with sparing of the epiglottis. Whether this supraglottitis represents an earlier phase or a milder form of the disease is unclear, but the cause is also *H. influenzae* type b, and
recognition is important because progression of obstruction will occur without appropriate treatment.

Airway Stabilization

The most important aspect of management, of course, is to maintain or establish an adequate airway. If decompensation occurs before the patient reaches the hospital, ventilation with bag and mask or even mouth to mouth resuscitation can overcome laryngeal spasm and prove life-saving. Once the diagnosis is apparent and the patient has reached the hospital, our current choice of procedure is nasotracheal intubation, rather than tracheostomy, to avoid surgery and the somewhat longer period of recovery. Early elective tracheostomy should be employed if personnel experienced in nasotracheal intubation and its aftercare are not available. Steroids and antibiotics with bag and mask ventilation have been successfully utilized without intubation in mild or even moderately severe cases, but should never be attempted without excellent monitoring facilities and constant attention by personnel skilled in airway management.

Antibiotics

Once the airway is secure, antibiotic management should include parenteral therapy with a drug effective against *H. influenzae*. High doses of ampicillin (200 mg/kg/day) have been employed in most cases at this institution. With the appearance of ampicillin-resistant strains, chloramphenicol has been suggested as additional initial therapy. Since maintenance of an adequate airway is the crucial aspect of therapy for epiglottitis, and since meningitis and other major medical complications are rare, we generally prefer to avoid this potentially toxic drug and treat with ampicillin alone. Chloramphenicol was added in several patients with persistent fever or a report of ampicillin resistance. Three of our five patients with ampicillin-resistant *H. influenzae* were treated successfully with ampicillin as the sole antibiotic, and the other two children were already afebrile by the time chloramphenicol was added. Similar results have been reported by others. Intravenous therapy is continued until the child has defervesced, been extubated, and is able to take and retain oral fluids. Thereafter oral therapy is given to complete a seven day course.

Anesthetic Management and Intubation

Securing the airway of a child suffering from acute epiglottitis or severe croup challenges even the most experienced anesthesiologist. Success depends on the close communication and collaboration of a well organized team following an established protocol.

Preparation

Since complete obstruction of the airway can occur at any time either prior to or during induction, the anesthesiologist must be prepared for all possible events. An array of nasotracheal tubes and laryngoscope blades should be available in the operating suite. Nasotracheal tubes are preferred because they are easier to fasten securely and enable provision of better oral hygiene. Table 8 demonstrates the guides used in our institution for selecting the appropriate sized tube. An endotracheal tube at least 1 mm internal diameter smaller than normal should be ready for use. Generally, the appropriate sized tube can be used
when intubating children with epiglottitis. For croup, however, the initial tube tried should be
0.5 mm internal diameter smaller, as the subglottic narrowing produced by this disease often
necessitates use of a tube much smaller in size than that which would otherwise be
appropriate. Although nasotracheal intubation is usually successful, an oral tube with stylet
in place should also be prepared in the event of loss of airway. Laryngoscope blade sizes are
as follows: premature - premature (0); infant (newborn to 3 mos) - Miller 1; 3 mos to 3 yrs -
No 1.5; child 3 yrs to 9 yrs - No 2; child over 9 yrs - No 3.

Because all patients should be presumed to have full stomachs and therefore to be at
risk for regurgitation and aspiration, a large suction catheter (minimum size 14 French) must
be included in the preparation.

Prior to the start of anesthetic induction the otolaryngologist must be prepared to
perform an emergency tracheostomy if it should prove necessary. A rigid bronchoscope,
completely assembled and functional, must also be available in the operating suite.

Table 8. Approximate Sizes of Endotracheal Tubes

<table>
<thead>
<tr>
<th>Age</th>
<th>Internal Diameter (mm)</th>
<th>French</th>
</tr>
</thead>
<tbody>
<tr>
<td>Premature</td>
<td>3.0</td>
<td>12</td>
</tr>
<tr>
<td>Newborn</td>
<td>3.0-3.5</td>
<td>12-14</td>
</tr>
<tr>
<td>Infant</td>
<td>3.5-4.0</td>
<td>14-16</td>
</tr>
<tr>
<td>10-12 mos</td>
<td>4.5</td>
<td>18</td>
</tr>
<tr>
<td>13-24 mos</td>
<td>5.0</td>
<td>20</td>
</tr>
<tr>
<td>2-3 yrs</td>
<td>5.5</td>
<td>22</td>
</tr>
<tr>
<td>4-5 yrs</td>
<td>6.0</td>
<td>24</td>
</tr>
<tr>
<td>6-7 yrs</td>
<td>6.5</td>
<td>26</td>
</tr>
<tr>
<td>8-9 yrs</td>
<td>7.0</td>
<td>28</td>
</tr>
<tr>
<td>10 yrs and up</td>
<td>7.0 (cuffed)</td>
<td>28</td>
</tr>
</tbody>
</table>

Finally, preparation of the patient is important whether or not he appears to be
interested in his surroundings. The parent is allowed to accompany the child to the entrance
of the operating suite. In transit from the emergency room, the anesthesiologist talks to the
patient constantly in calm, soothing, hypnotic tones, providing reassurance and explaining in
simple terms what is happening and why. This running conversation is continued until
induction has occurred. In addition, movements are slow and deliberate, and the child's
positional preference is honored to further minimize disturbance.

Anesthetic Delivery

Since most children who suffer from epiglottitis or croup prefer an upright position,
anesthesia should be delivered with the child in this position. The anesthetic technique of
choice in this institution is that of a spontaneous inhalation induction using halothane and 100
per cent oxygen. An intravenous technique using thiopentone and succinylcholine may be
used, but should be attempted only by anesthetists experienced with problems of the
compromised airway. Nitrous oxide will hasten the induction, but with the risk of diffusion
hypoxia its use is not justified until the airway has been secured. While the induction is
proceeding, it is usually possible to insert a No 20 or 22 Teflon intravenous catheter to provide ready access for medications and to enable adequate hydration.

**Intubation**

Children with epiglottitis are treated with nasotracheal intubation. It has been our experience and that of others that with the majority of these patients it is possible to assist and control ventilation manually, using a bag and mask, prior to insertion of the nasotracheal tube. However, should this become impossible, it is important to attempt an oral intubation using the prepared oral tube. If this is unsuccessful, a rigid bronchoscope should be tried. Only when these attempts fail should an emergency tracheostomy be performed. If manual ventilation is possible, a depolarizing muscle relaxant, such as succinylcholine, can be used to facilitate intubation once the patient is anesthetized. Otherwise it is safer to avoid using muscle relaxants. Following insertion, the nasotracheal tube is held in place while tincture of benzoin is applied from ear to ear, over the lips, and over the tube segment just outside the nostril. After this has dried, strips of half-inch adhesive tape are applied over the benzoin and wrapped around the tube from both above and below.

For patients with croup, oral intubation is performed first. Then a tracheostomy is performed over the endotracheal tube. Following tracheostomy, direct laryngoscopy and bronchoscopy are performed (see article on tracheostomy).

**Laboratory Studies and Medication**

Once the airway is secured, blood can be drawn and cultures obtained while the patient is still anesthetized. An initial dose of antibiotic is administered to patients with epiglottitis. Since it is our opinion that these children require sedation in the intensive care unit (ICU) during the period of intubation and observation, an initial intravenous injection of morphine sulphate, 0.1 mg/kg, is also given in the operating room. This has been found to minimize the agitation and thrashing that commonly occur during transfer from operating room to ICU.

**Intensive Care Unit Management**

Children who have undergone intubation or tracheostomy for epiglottitis or croup require the facilities of an ICU for monitoring and treatment. Because tracheostomy care is discussed in the preceding article, management of the intubated patient with epiglottitis will be stressed here. Upon arrival in the ICU, the chest should be auscultated to insure that both lungs are being ventilated, and the position of the tube should be confirmed by chest radiographs. The tip of the tube should always be about mid-trachea. It is prudent, once it has been established that the tube is appropriately placed, to mark it at the level of the nostril. Thus, if the tube is inadvertently withdrawn or advanced, this will be recognized quickly.

**Monitoring**

In all patients with artificial airways, the electrocardiogram should be displayed on an oscilloscope continuously so that arrhythmias, which may be early signs of blood gas abnormalities, may be easily detected. Blood pressure must be monitored accurately. Body
temperature is checked at intervals, and should hyperthermia develop, it is treated with antipyretics. Serial determination of arterial blood gases is important in the first few hours since pulmonary complications may develop which may necessitate supplemental oxygen and assisted ventilation. Intermittent samples can be obtained with the use of 25 gauge needles, but if frequent blood samples are required the radial artery should be cannulated.

**Supervision and Sedation**

Despite use of measures to restrict movement such as elbow restraints, constant nursing observing is necessary to keep children from accidentally removing the tubes. Heavy sedation is also helpful. It obviates the fear, stress, and anxiety produced by the ICU environment, and by separation from the parents (especially in the preschool age child); in reducing agitation and movement, sedation minimizes the risk of trauma to the glottis and subglottic areas that can result from the to-and-fro motion of the endotracheal tube. Effective sedation can be accomplished by alternating morphine and diazepam, 0.1 to 0.2 mg/kg each, intravenously every four hours. Occasionally a very restless child may require more frequent doses during the early course of management. If this depresses respirations, use of assisted ventilation is then necessary to avoid hypoventilation. It is, however, crucial to rule out hypoxia as the source of agitation before increasing sedation.

**Respiratory Care Program**

A few principles and steps are essential in caring for patients with artificial airways. Adequate humidification must be provided to avoid blockage of the airway by inspissated secretions. A minimum of 2 cm of water of continuous positive airway pressure must be applied to the airway to maintain a normal functional residual capacity, and to prevent closure of small airways. One to two minutes prior to endotracheal suctioning, the lungs should be hyperinflated with 100 per cent oxygen. Gentle bagging is required to promote adequate ventilation past the mucus without driving it deeper into the distal airways. Using the single-glove sterile technique, 1 to 5 ml of sterile saline without preservative (depending on the child’s size) should be instilled into the endotracheal tube to facilitate removal of secretions. The child should be bagged again and a vibratory squeeze should be applied. This is done by placing one hand over each side of the chest and simultaneously vibrating and compressing it after a manually accomplished deep inspiration. With repetitive vibratory squeezes, the gas present in the lung forces secretions up and out of small airways into the major bronchi where they can be reached by the suction catheter. The catheter should have a diameter less than 50 per cent of the diameter of the airway, and have side and end holes. When inserted, it should pass the tip of the endotracheal tube. A suction pressure of about 15 mm Hg is recommended. As the catheter is withdrawn rapidly, a twisting motion should be applied to enhance the removal of secretions that may be clinging to the bronchial mucosa. This procedure is repeated every two to four hours, or more often if the tracheobronchial secretions are abundant.

**Postintubation Course**

Segmental atelectasis is frequently observed in patients in whom an artificial airway is used. However, in children with epiglottitis, it may be related to the child’s primary disease. The incidence of this complication is about 25 per cent, and it is most likely related to
inspissation of secretions. Poor fluid intake secondary to severe dysphagia, the increase in insensible water loss that accompanies high fever, and inadequate coughing also contribute. In some instances this problem has been confused with pneumonitis; however, sequential radiographic examination will demonstrate rapid clearing of the atelectasis owing to intubation and tracheobronchial suctioning, usually before the endotracheal tube is removed.

The second most frequent complication of epiglottitis is pneumonia. In our experience, this develops in about 8 per cent of all patients admitted with epiglottitis.

Pulmonary edema associated with relief of airway obstruction has been described recently. In our institution it has been observed in about 7 per cent of the cases admitted. It should be anticipated in those children in whom airway obstruction has been severe and prolonged, often necessitating emergency insertion of an artificial airway. In these patients, bypassing the obstruction often fails to bring about dramatic improvement of their respiratory distress. Major physiologic changes, such as hypoxemia and massive sympathetic discharge, have been postulated to contribute to this type of pulmonary edema. It is likely that the extremely high transpulmonary pressure generated during inspiration with a severely obstructed airway increases the pulmonary blood volume markedly. The obstruction provides some degree of protection from this by decreasing venous return to the thorax during exhalation. When an artificial airway is inserted and the obstruction bypassed, a sudden increase in venous return to the central circulation occurs, along with a marked increase in the intravascular hydrostatic pressure. At this point true pulmonary edema develops. Prevention of this situation must begin the moment the airway is inserted and involves the application of 4 to 5 cm of water of continuous positive airway pressure to the airway, which can be discontinued gradually over the next 24 to 36 hours. If severe pulmonary edema develops, assisted ventilation should be instituted immediately, fluids restricted, diuretics given as indicated, and measurement of the pulmonary artery capillary wedge pressure considered.

**Extubation**

The duration of intubation is often more related to the availability of the operating room than to the patient's clinical course. The great majority of children can be extubated safely 36 hours after intubation. In our institution extubation occurs between 36 and 72 hours, with a mean of 48 hours. Prior to this it is advisable to examine the epiglottis at the bedside by direct laryngoscopy to determine if the swelling has decreased significantly. If this is so, the child is taken to the operating room for decannulation and complete endoscopy. It is wise to discontinue sedation at least 6 to 8 hours prior to extubation so that the child is fully awake after the anesthetic for the endoscopy wears off, and is thus able to maintain his protective reflexes.

Postextubation subglottic edema, if present, is usually detected during endoscopy. In this situation, as soon as the endotracheal tube is removed, nebulized 2 per cent racemic epinephrine diluted with sterile saline to a total volume of 3.5 mL is delivered by face mask or IPPB for 15 minutes. Additional doses of racemic epinephrine are indicated if postextubation airway obstruction persists. If the response to racemic epinephrine is not long-lasting and the obstruction becomes severe, reintubation with a tube smaller than the one used originally is indicated. A course of dexamethasone, 0.5 to 1 mg/kg every 6 hours for 24
hours, should be given before attempting extubation again. Fortunately, reintubation is rarely necessary.

Following successful decannulation, the patient is given mist by mask or face tent and is transferred to a ward for observation for an additional one to two days.
Tonsillectomy and Adenoidectomy

Jack L. Paradise


No practice involving health care for children has excited more heated controversy among health professionals than has surgical removal of the tonsils and adenoids. Long the most common major operation carried out on children, tonsillectomy and adenoidectomy (T&A) continues to draw professional, legislative, and lay attention as a treatment for which benefits in relation to costs and risks have never been adequately assessed, and for which indications remain largely ill-defined. Nonetheless, the continuing high rate of performance of T&A, despite some recent decline, attests to its strong hold on the minds of many physicians and parents as a treatment of importance and value. Annual expenditures for tonsil and adenoid surgery in the USA probably exceed one-half billion dollars.

Although T&A is often thought of and carried out as a single, combined operation, each of the two components - tonsillectomy and adenoidectomy - requires individual attention when considering indications for operation. As of present writing, convincing evidence is lacking that T&A, in the conditions for which they are usually undertaken, either are or are not superior in efficiency to conservative management. Unfortunately, the few reported trials of T&A have as a group been inadequate and inconclusive. In order to arrive at rational indications for tonsil and adenoid surgery, groups of children must first be defined who have particular symptom complexes severe enough to justify particular operations; in those groups the efficacy of the operations must then be tested by means of an integrated group of randomized, controlled trials; and finally, if the operations prove efficacious, their overall impact must be assessed as critically as possible in relation to their risks and costs. It was with this frame of reference that the current Children's Hospital of Pittsburgh study was designed and undertaken.

The Children's Hospital of Pittsburgh Study

Goals, Design, and Methodology

The Children's Hospital of Pittsburgh study, instituted on a pilot basis in 1971, supported since 1973 by the National Institute of Child Health and Human Development, and still in progress, addresses a number of questions involving both the natural history of presumably tonsil- and adenoid-related problems and the results of tonsil and adenoid surgery. In particular, the study focuses on the efficacy of tonsillectomy in reducing the frequency and severity of episodes of pharyngitis, the efficacy of adenoidectomy in reducing the frequency and severity of episodes of otitis media, and the effect of adenoidectomy on the course of nasal obstruction that is caused by large adenoids.

Salient characteristics of the study include the following:

1. A team of individuals is employed specifically for the work of the study; virtually all historical accounts are obtained and virtually all examinations conducted by study personnel.
2. On entry, each subject receives independent evaluations by a pediatrician and an otolaryngologist.

3. Tonsillectomy and adenoidectomy are treated as separate procedures.

4. Criteria for entering the clinical trials of tonsillectomy and of adenoidectomy are specified (Tables 1 and 2).

5. Criteria are specified (Table 3) for exclusion from randomization (ie, when indications for operation appear compelling).

6. Standardized systems are used for quantifying or rating relevant clinical findings and diagnoses.

7. A continuing process of testing the inter-observer reliability of study staff members is maintained.

8. Assessment of middle ear status is based on combined otoscopic, tympanometric, and audiometric examinations.

9. Standardized cephalometric radiographs are used in assessing adenoid size.

10. Each subject receives a screening allergy/immunology evaluation that includes both skin tests and determination of serum immunoglobulin levels.

11. Standardized dental and orthodontic observations concerning each subject are recorded by a pedodontist.

12. Follow-up of each subject is carried out by direct clinical examination at six-week intervals and also at the time of intercurrent respiratory illness, and is supplemented by bi-weekly, standardized telephone inquiries.

**Preliminary Findings**

*Limitations of Undocumented Histories of Recurrent Throat Infection.* Many of the children referred to the study as potential candidates for the tonsillectomy trial have histories of recurrent episodes of throat infection that appear to meet all the entry standards listed in Table 1 except documentation. These children are admitted to the study and followed prospectively; if at least two observed episodes of throat infection then develop with patterns of frequency and clinical features that match or exceed those described in their presenting histories, they become eligible for the tonsillectomy trial. To date, a large majority of such children have failed to develop frequent or severe episodes of throat infection. From this experience it is reasonable to conclude that *undocumented* histories of recurrent throat infection do not validly forecast subsequent experience and hence do not constitute an adequate basis for subjecting children to tonsillectomy.
Table 1. Children’s Hospital of Pittsburgh T&A Study: Criteria for Entering Controlled, Randomized Trial of Tonsillectomy

1. Recurrent throat infection
   a. At least three episodes in each of three years, or five episodes in each of two years, or seven episodes in one year; and
   b. Each episode must have been characterized by one or more of the following:
      - Oral temperature 38.3°C or higher
      - Enlarged (> 2 cm) or tender, anterior cervical lymph nodes
      - Tonsillar exudate
      - Positive culture for group A beta-hemolytic streptococcus
   c. Apparently adequate antibiotic therapy must have been administered for proven or suspected streptococcal episodes; and
   d. Each episode must have been confirmed by examination and its qualifying features described in a clinical record at the time of occurrence
or, 2. Peritonsillar abscess
or, 3. Chronic (minimum 6 months) tonsillitis, persisting despite appropriate antimicrobial therapy
or, 4. Nonurgent obstructive symptoms if tonsils very large, including
   a. stertorous or mouth breathing, with or without episodes of obstructive sleep apnea
   b. muffled, "hot potato" voice if child is at least 6 years old
or, 5. Chronic (minimum 6 months) enlargement (> 2 cm) or tenderness of anterior cervical lymph nodes, persisting despite appropriate antibiotic therapy.

Table 2. Children’s Hospital of Pittsburgh T&A Study: Criteria for Entering Controlled, Randomized Trial of Adenoidectomy

1. Recurrent suppurative or nonsuppurative otitis media, if myringotomy and insertion of tympanostomy tube have been performed at least once previously
or, 2. Persistent nasal obstruction
   a. manifested by stertorous or mouth breathing, with or without episodes of obstructive sleep apnea, and by hyponasal speech, and
   b. accompanied by roentgenographic evidence of adenoid hypertrophy, and
   c. apparently not due to allergy
or, 3. Chronic sinusitis or nasopharyngitis
   a. accompanied by both clinical and roentgenographic evidence of adenoid hypertrophy, and
   b. apparently not due to allergy, and
   c. persisting despite appropriate antimicrobial and other medical therapy.
Table 3. Children’s Hospital of Pittsburgh T&A Study: Criteria for Exclusion from Controlled, Randomized Trial and for Prompt Surgical Intervention

<table>
<thead>
<tr>
<th>Indication</th>
<th>Tonsillectomy</th>
<th>Adenoidectomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Upper airway obstruction apparently due to large tonsils or adenoids or both, and accompanied by evidence of alveolar hypoventilation or cor pulmonale</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Consisten interference with swallowing apparently due to tonsilar enlargement</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Severe nasal obstruction due to adenoidal enlargement, resulting in apparent discomfort in breathing</td>
<td></td>
<td>X</td>
</tr>
</tbody>
</table>

The Clinical Trial of Tonsillectomy. Preliminary findings to date indicate that in children with recurrent episodes of throat infection that meet the entry criteria shown in Table 1, tonsillectomy is indeed effective in reducing morbidity from throat infection over a period of at least two years. On the other hand, it is noteworthy that the absolute morbidity from throat infection experienced by control subjects has in general not been extreme: about half the control subjects have thus far each experienced fewer than three episodes of throat infection per year, and almost two-thirds of these episodes have been rated clinically as mild.

With the regard to the efficacy of tonsillectomy for peritonsillar abscess, chronic (as distinct from recurrent acute) tonsillitis, and chronic isolated cervical lymphadenitis, the number of patients with these conditions admitted to the study have been too few to warrant drawing conclusions.

Tonsillectomy, when carried out for obstructive symptoms unequivocally attributable to large tonsils, has consistently afforded complete relief; in children not operated on, data currently available are insufficient to indicate the extent to which such symptoms - and the underlying tonsillar hypertrophy - may subside spontaneously.

The Clinical Trial of Adenoidectomy for Otitis Media. In order to define a population at high risk for development of otitis media, the protocol of the current study specifies that, as a prerequisite to entering the trial of adenoidectomy on the basis of recurrent otitis media, a child must have received myringotomy and tympanostomy tube insertion in the past and must subsequently have experienced at least one episode of either suppurative or non-suppurative otitis media (Table 2).

Preliminary data show that adenoidectomy by no means eliminates the problem of recurrent otitis media in such children, but it remains uncertain whether adenoidectomy somewhat reduces the rate, severity, or duration of recurrent episodes. The question is particularly complex because of the large number of variables that may potentially influence outcome. A large number of study subjects will be required in order to reach firm conclusions.
**The Clinical Trial of Adenoidectomy for Nasal Obstruction.** Children are admitted to the clinical trial of adenoidectomy for nasal obstruction only if the obstruction is appreciable (Table 2) and can be shown with reasonable certainty to be caused wholly or mainly by large adenoids. Such children who have received adenoidectomy appear to be experiencing almost uniformly excellent results persisting for at least two years. In the control group, some spontaneous improvement has developed within the first year or two in some of the patients, but complete resolution of nasal obstruction has developed in relatively few. The children providing these data have been mainly five and six years old, and information regarding children either younger or older is as yet inadequate for analysis.

It is important to consider whether the significantly greater relief of nasal obstruction achieved in the operated group resulted in benefits to the children (Table 4) that offset the cost and risks of the operation. In an effort to test the development of one such benefit - improved nasal function - we assessed olfaction in a group of study children using varying concentrations of phenylethyl alcohol, a rose-like odorant. We found that children rated as having no nasal obstruction showed almost uniformly good olfactory function, whereas most of those with severe obstruction showed poor function.

Other hoped-for outcomes of adenoidectomy remain to be evaluated. For example, certain orthodontists have attributed presumably abnormal growth patterns (long and narrow faces, low tongue placement, narrow upper jaws, steep mandibles, and open anterior bites) to large adenoids and have advocated surgery to prevent or ameliorate the so-called adenoid facies. One study claimed improvement in dentofacial measurements following adenoidectomy but failed to include unoperated control subjects. Comparative analysis of the standardized cephalometric roentgenographs obtained periodically from adenoidectomy and control subjects in the Pittsburgh study may help to clarify this question. Outcomes of adenoidectomy such as an increased sense of comfort in breathing and improved facial appearance are essentially subjective and not readily measurable.

**Table 4. Costs and Potential Benefits of Tonsillectomy or Adenoidectomy or Both**

<table>
<thead>
<tr>
<th>Costs</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Currently (September, 1981) $ 1500 at Children’s Hospital of Pittsburgh</td>
</tr>
<tr>
<td>2. Risk of anesthetic accidents:</td>
</tr>
<tr>
<td>malignant hyperthermia</td>
</tr>
<tr>
<td>cardiac arrhythmia</td>
</tr>
<tr>
<td>vocal cord trauma</td>
</tr>
<tr>
<td>aspiration with resulting bronchopulmonary obstruction or infection</td>
</tr>
<tr>
<td>3. Risk of miscellaneous surgical or postoperative complications:</td>
</tr>
<tr>
<td>hemorrhage</td>
</tr>
<tr>
<td>airway obstruction due to edema of tongue, palate, or nasopharynx, or to retropharyngeal insufficiency</td>
</tr>
<tr>
<td>otitis media</td>
</tr>
<tr>
<td>emotional upset</td>
</tr>
<tr>
<td>4. Unknown risks</td>
</tr>
</tbody>
</table>
Potential Benefits If Efficacious

1. Reduction in frequency of ENT illness:
   - discomfort
   - inconvenience
   - school absence
   - parental anxiety
   - work missed by parents
   - costs of doctor visits and drugs
2. Reduction in nasal obstruction, with improved:
   - respiratory function
   - morbidity
   - comfort
   - sleep
   - craniofacial growth and development
   - appearance
3. Reduction in hearing impairment
4. Improved growth and overall well-being
5. Reduction in long-term parental anxiety.

Currently Acceptable Surgical Indications and Surgical Decision Making

Notwithstanding the large remaining areas of uncertainty about the efficacy of tonsillectomy and of adenoidectomy, decisions about surgery must currently be made on the basis of information now available. Operation is clearly indicated in a small number of severely affected children and clearly not indicated in the great majority of children in whom the degree of tonsil- or adenoid-related illness or disability is insubstantial. Between these extremes lies an intermediate group concerning whom there is room for legitimate controversy. The uncertainty regarding these latter children is of two general categories: (1) the tonsil or adenoid surgery is of uncertain efficacy, as in the case of recurrent otitis media; or (2) efficacy appears to be reasonably well established, as in the case of adenoidectomy for nasal obstruction secondary to large adenoids, but in the patient at hand clinical severity, prognosis, or both may be in doubt. For example, it may not be possible to determine satisfactorily whether (a) a child is affected severely enough to justify an operation; (b) a child is so severely affected as to make surgery mandatory rather than optional; or (c) a child is so likely to soon "outgrow" the condition as to render operation of only short-term value.

Definite Surgical Indications

Operation is clearly indicated in unusual circumstances in which massive hypertrophy of tonsils, adenoids, or both results in unquestioned dysphagia, in extreme discomfort in breathing, or, even more extremely, in alveolar hypoventilation or cor pulmonale (Table 3).

Alveolar hypoventilation is a difficult diagnosis to confirm, short of the development of clinical, roentgenographic, or electrocardiographic evidence of cor pulmonale. The measurement of blood pO$_2$ and pCO$_2$ both when awake and during sleep, a procedure that currently cannot be accomplished non-invasively, is required. Extremely stertorous breathing when awake and frequent episodes of obstructive apnea during sleep should lead to a high
index of suspicion of the presence of alveolar hypoventilation. It appears likely that the condition, while unusual, may be more common than is generally appreciated.

On the other hand, even in children with symptoms or signs of appreciable obstruction, surgery should not be an immediate and automatic alternative. Seemingly dramatic obstructive manifestations, even when long-standing, may be caused by edema that accompanies relatively inapparent infection, rather than to fixed structural changes. Such obstructive symptoms may sometimes lessen considerably with vigorous antimicrobial treatment. Accordingly, a trial of an appropriate antimicrobial agent is often advisable before deciding whether surgery is mandatory or even reasonably indicated.

**Uncertain but Reasonable Surgical Indications**

Although the conditions in Tables 1 and 2 are still being tested, they appear to constitute reasonable indications for tonsillectomy and adenoidectomy, respectively. For all of these conditions the common denominator is an arbitrarily defined minimum degree of severity, frequency, or duration. Equally reasonable, however, might be modifications *within the spirit* of these minimum standards. For example, in the case of frequently recurring or persistent episodes of recurrent otitis media, it appears acceptable (1) not to require prior tympanostomy tube placement before considering adenoidectomy, or (2) to include tonsillectomy if adenoidectomy were being embarked upon as a treatment option.

However, the fact that a child meets the above criteria should not necessarily lead to a decision in favor of surgery. Each decision should be individualized. For example, in a child with recurrent tonsillopharyngitis, additional factors that might influence the decision include accessibility of health care, school achievement and progress, parental tolerance of illness, other family stresses, and the relative out-of-pocket costs to the family of medical and surgical management. One study argues in favor of tonsillectomy for the patient with rheumatic heart disease who has large tonsils and in whom antistreptococcal prophylaxis cannot be maintained with confidence.

Regarding the question of using surgical criteria less stringent that those listed in Tables 1 and 2 (for example, the criteria listed by a committee of the American Academy of Pediatrics), the purpose of these criteria is to define a *minimally acceptable* rather than an *optimal* standard of care. Recognizing that these criteria are arbitrary, it nonetheless appears that relaxing them would rarely achieve appreciable benefit, whereas without them many children might undergo unnecessary operations. The Pittsburgh study should eventually provide information regarding the expected illness experience of the children who fall just short of meeting these criteria, since such children also are followed in the study prospectively. Should their prognosis prove in general to be benign, relaxing the criteria would hardly appear to be appropriate. If, on the other hand, these children are found to experience appreciable tonsil- or adenoid-related illness, further study of the efficacy of surgery should be done.

Halitosis caused by the accumulation of debris in tonsillar crypts may justify tonsillectomy in those unusual circumstances in which more conservative measures, such as gargling or pharyngeal douche, prove ineffective.
Contraindications to Tonsil and Adenoid Surgery

Contraindications to surgery include velopharyngeal, hematologic, immunologic, and infectious conditions.

A number of abnormal conditions that result in velopharyngeal insufficiency, such as overt cleft of the palate, submucous or covert cleft of the palate, neurologic or neuromuscular abnormalities leading to impaired palatal function, and an unusually capacious pharynx, are contraindications to adenoidectomy. In each of these conditions the presenting complaint is likely to be hypernasality, a symptom that the unwary observer may fail to distinguish from hyponasality. If adenoidectomy is undertaken to improve the "nasal" speech of such children, the symptoms may worsen markedly, since the adenoids had been serving to help fill the relative velopharyngeal void and thus to facilitate normal speech production. Suspicion of a submucous cleft of the palate should be aroused by observing a bifid uvula or widening and attenuation of the median raphe of the soft palate. The diagnosis may be further ascertained by palpating along the junction of the hard and soft palates, where a V-shaped midline notch, rather than the normal rounded curve, is strongly suggestive of a submucous cleft. This examination should be performed on all children, with or without hypernasality, for whom adenoidectomy is being considered. Irrespective of the findings, if hypernasality secondary to velopharyngeal insufficiency is suspected, it is advisable to refer the patient to an individual or a team skilled in the evaluation and management of cleft palate.

Hematologic contraindications to tonsil or adenoid surgery consist of anemia and disorders of hemostasis. Surgery should not be undertaken if the hemoglobin concentration is less than 10 gm per dL or the hematocrit less than 30 per cent. When surgery is being considered, careful inquiry should always be made about a family or past history of unusual bleeding or bruising, as certain rare hemostatic disorders may not be detectable with readily available tests. Routine preoperative studies should include measurements of the hemoglobin or hematocrit, prothrombin time, and partial thromboplastin time, and an estimate of the platelets, usually from a stained blood smear.

In the view of some clinicians, the existence of frank respiratory allergy that has not been treated for at least six months constitutes a contraindication to tonsil or adenoid surgery unless urgent, obstructive symptoms are present. The opinion that tonsil or adenoid surgery in allergic children may precipitate the development of asthma has not been tested in clinical studies. Certainly in children without urgent obstructive symptoms who have both upper respiratory allergy and large tonsils or adenoids, a reasonable trial of anti-allergic management as a precursor to considering surgery appears to be prudent.

Tonsillectomy or adenoidectomy should not be undertaken in patients with local infection unless urgent obstructive symptoms are present, appropriate, prolonged antimicrobial treatment has been maintained unsuccessfully, or, in the view of some, a peritonsillar abscess is present. Ordinarily, an interval of at least three weeks following an episode of acute infection will allow for general recuperation and reduce the risk of operative hemorrhage.
Adverse Effects of Tonsil and Adenoid Surgery: Real and Potential

Physicians who recommend tonsil or adenoid surgery must weigh the possibility of adverse consequences ranging from death, to nonfatal direct and indirect anesthetic and surgical complications, to hypothetical interference with immunologic defense mechanisms. Unfortunately, accurate statistics regarding mortality and morbidity in large patient populations are not available.

The death of a child as a consequence of tonsil or adenoid surgery is especially tragic if, as is usually the case, the operation was elective. Fatality rates have been variously reported during the past 25 years as ranging from 1 per 1000 to 1 per 27,000 patients, but the validity of these reports is open to question. Except for a probably irreducible minimum of anesthesia-related deaths (the anesthesia-related mortality rate unadjusted for age was recently reported to be 1 per 14,000 patients), death as a result of tonsil or adenoid surgery should be entirely preventable.

The possible nonfatal complications of tonsil or adenoid surgery are summarized in Table 4; velopharyngeal insufficiency and emotional upsets are discussed in detail above and below, respectively. The risk of hemorrhage can be minimized by avoiding operation during or immediately following episodes of infection, by careful attention to surgical technique, and by avoiding the use of aspirin for relief of postoperative pain. Nonetheless, either primary or secondary hemorrhage is bound to occur in some cases, and transfusion will occasionally be required. Data reported from a large number of hospitals surveyed during 1965 indicate a transfusion rate of 0.4 per cent. Otitis media has been reported as a not infrequent postoperative complication, but it is not clear that the risk is higher than in comparable patients not operated upon.

Whether tonsil or adenoid surgery imposes immunologic risks of any practical consequence remains uncertain. The heightened risk of poliomyelitis that was an important deterrent to surgery before the advent of an effective vaccine, and for which an immunologic basis has more recently been elucidated, is no longer of practical concern. The concern that tonsillectomy might predispose to the development of Hodgkin's disease has apparently been dispelled by more recent epidemiologic investigations. It is possible that removal of the immunologically active tonsils and adenoids will later undermine the patient's resistance to disease of some sort, but the likelihood appears to be small.

Preparation for Hospitalization and Surgery, and Care in the Hospital

Children who are to undergo surgery should be prepared for the experience well in advance. Parents should describe the expected course of events in as much detail and as frankly as possible, commensurate with the child's ability to comprehend. Children should be told that they will experience a certain amount of discomfort but that every effort will be made by hospital personnel to minimize it. Many hospitals permit advance visits so that children may see the facilities and equipment to be used and become acquainted with some of the personnel. Coloring or story books can also be helpful in the familiarization process. Once admitted to the hospital, children should have free and unlimited access to parents or parent surrogates, and parental rooming-in should be encouraged, especially for children younger than school age. One or both parents should remain with the child during the period
immediately preceding the trip to the operating room and should be at the bedside when the child returns. Careful preparation and kind, thoughtful management of the entire process of hospitalization and surgery should virtually eliminate the risk of untoward psychological consequences in previously well-adjusted children. For the child who is emotionally disturbed, the same general principles apply but, in addition, specialized professional advice may be appropriate to minimize the risk of neurotic misinterpretation of the operative event by the child.
The Role of Pediatric Nurse Practitioners in the Tonsil and Adenoid Study

Ruth Z. Bachman, Kathleen M. DiGaudio, Margaret T. Menninger


Numerous reports in the literature support the ability of the nurse practitioner to perform effectively the skills of history-taking, physical examination, assessment of illness, and counseling in hospital, clinic and community settings. This article describes the role of the pediatric nurse practitioner (PNP) and the implementation of these skills in yet another clinical setting, the clinical research project. The authors address the role of the PNP in the context of the research protocol, the nurse-patient interaction, and the relationship of the PNP with other health-care professionals. The authors are research associates involved in a long-term prospective study of indications for tonsillectomy and adenoidectomy (T&A) at the Children's Hospital of Pittsburgh.

The primary responsibility of the PNP in the T&A Study is to carry out the day-to-day clinical protocol. Initially, the PNP reviews information that is submitted by referring health professionals in order to determine eligibility for enrollment into the study. Children who meet the criteria undergo an extensive initial evaluation including comprehensive history, examination of ear, nose, throat, and chest, and a battery of diagnostic studies. Pediatricians and otolaryngologists who are members of the study team examine the patient at the initial visit, and the pediatrician reexamines the child at least every six months thereafter. Follow-up visits with the PNP are scheduled at six-week intervals and whenever an otolaryngologic illness is suspected. At each visit, the PNP reviews the patient's course since entry into the study, evaluates his current status, including how this fits into the overall study framework, and, within certain limits, undertakes management according to study protocol. Pediatricians and otolaryngologists are always available for consultation, and one or more physicians involved in the study evaluate children who are found or suspected to have illness of more than a mild degree. Whenever surgical decisions are considered, the PNP organizes and presents the relevant data for joint review by a pediatrician and an otolaryngologist. The members of the study team then evaluate the patient and together decide whether protocol criteria for surgery have been met.

Interobserver reliability in the study is ensured by the utilization of standardized systems for quantifying, rating, and recording observations. Independent findings by two or more examiners at any one visit are systemically compared and analyzed, thus maintaining consistency and replicability of findings.

Genuine case and consideration of the patient is as important in clinical research as it is in any other area of nursing practice. The roles of the PNP as listener, health educator, and patient advocate in no way interfere with the duties of managing the study protocol. On the contrary, the study runs more smoothly when there is a strong nurse-patient relationship. The parents are more motivated to provide accurate histories, the child cooperates more fully in physical examinations and other procedures, and there are fewer missed appointments. A recent review of the clinic "no show" rate found it to be only 12 per cent. As rapport develops between the PNP and her patients, she is often the first to learn of the general medical and psychosocial concerns of the families. Ear, nose, and throat (ENT) problems as
well as routine health and developmental issues may be discussed during clinic or telephone consultations. Families are encouraged to maintain close ties with their primary health care provider and are referred to this caretaker or to an appropriate special facility if problems unrelated to the T&A study arise.

Hospitalization and surgery are often threatening experiences for children and their families. For children scheduled to have a tonsillectomy or adenoidectomy, anticipatory guidance concerning hospitalization is important. To help families to develop realistic expectations regarding the hospital experience, the PNP describes hospital routines and surgical procedures and explains postoperative care both in hospital and at home. Visits by the PNP during hospitalization provide an opportunity for the family to discuss feelings and ask questions, and for the PNP to reinforce postoperative instructions.

A collaborative relationship among the members of the research team is critical to the progress of a large clinical research project. Regular staff meetings provide opportunities for open discussion of clinical or administrative issues affecting the project. For example, if there are unanticipated problems with routine procedures or standardized working definitions, the staff works together to arrive at solutions to the problems. In managing the care of the patients, the PNP in the T&A Study also collaborates with a variety of health care professionals apart from the research team. Community pediatricians and physicians, school nurses, audiologists, speech pathologists, allergists, hospital house officers, social workers, and hospital staff nurses may all be involved in the care of the study patient at some time. This type of collaborative relationship among professionals results in a reciprocal learning experience. The PNP also functions as a clinical instructor for medical students and PNP students, who are regularly assigned to the T&A Clinic as part of their formal education. These students learn about the clinical and research issues involved as well as about pneumatic otoscopy and other examination skills.

Participating in and helping to plan the conduct of a clinical research project that involves direct patient care is not only challenging, but also requires dedication to the principles of quality health care. The authors find this setting highly favorable for both facilitating mutually satisfying nurse-patient relationships and providing opportunities for counseling, teaching, and learning.
Sleep Disorders Associated with Upper Airway Obstruction in Children

Ellen M. Mandel, Charles F. Reynolds


Acute upper airway obstruction is a well-recognized, easily diagnosed medical emergency. Chronic persistent or intermittent upper airway obstruction, however, may not be obvious on routine physical examination of the awake child, but its consequences may be just as severe. Obstruction may increase in the supine position or be further accentuated by sleep. Sleep disorders with associated ventilatory disturbance are not uncommon in these children and may lead to a wide variety of physical and behavioral disorders. Some children progress to florid heart failure and eventual death if the diagnosis is delayed. With the aid of modern technology, we are now able to recognize the process at its earlier stages, but we are just beginning to realize the great impact of this problem.

Etiology and Pathophysiology

In children the most common cause of chronic airway obstruction and sleep-related obstructive phenomena is hypertrophy of the nasopharyngeal lymphoid tissue, specifically the adenoids and tonsils. However, obstruction at any site along the upper airway may cause similar disturbances, as seen with glottic web, vocal cord paralysis, nasal septal deviation, and the Pierre-Robin syndrome. Cinefluoroscopy during sleep in children with obstructive sleep apnea shows that the soft palate falls backward against the posterior pharynx, the tongue moves posteriorly, and the lateral walls of the hypopharynx approximate medially during inspiration. The child gasps in an effort to overcome the obstruction, arousing partially, with loud snoring marking the resumption of air flow. In children without a history of sleep-related problems, the hypopharynx remains patent during sleep. The fact that not all children with large tonsils and/or adenoids exhibit sleep disturbances suggests that central nervous system control of respiration and muscle tone may play a large part in this problem.

Laboratory Findings in Children With Sleep Apnea

With the increased awareness of the importance and diverse effects of sleep disorders, special laboratories have been established to monitor electrographically physiologic variables during sleep, a process called polysomnography. The specific approach varies somewhat between laboratories but often includes continuous monitoring of the EEG, EKG, respiratory effort by chest and/or abdominal strain gauges, or an endoesophageal pressure transducer), nasal airflow, extraocular movements, chin muscle tone, and arterial oxygen saturation. Audiovisual monitors may also be included. With this information the occurrence, duration, and type of apneic episodes can be noted as well as any accompanying changes in heart rate or rhythm, and arterial oxygen saturation. The amount of time spent in the various sleep stages can also be quantitated, and movements and behaviors during sleep may be noted. These studies are important in diagnosing the nature of the sleep disorder (such as central or obstructive apnea) and the severity of the problem. Using polysomnography, Guilleminault has defined the respiratory irregularities during sleep. A respiratory pause is defined as
cessation of airflow for less than 10 seconds; an *apneic episode* is a cessation of airflow for more than 10 seconds. A *sleep apnea syndrome* is defined as 30 episodes or more of apnea during a seven-hour sleep period. These definitions, however, vary with different investigators, and the boundary between normality and abnormality is not yet well defined.

Apnea is of several types, depending on its etiology. *Central apnea* is the cessation of breathing owing to the failure of the respiratory center to initiate a respiration. This is recorded by absence of chest movement or change in endoesophageal pressure, and the lack of nasal and oral airflow. *Obstructive apnea* is unsuccessful airflow despite a respiratory effort, as seen by increased chest wall or abdominal movements and increased negative endoesophageal pressure. *Mixed apnea* is a combination of these two types, being the absence of respiratory efforts followed by unsuccessful respiratory efforts. In another study by Guilleminault eight children diagnosed as having a sleep apnea syndrome were monitored, and a range of 78 to 816 apneic periods was noted during one nocturnal sleep period. Obstructive apnea was noted in 83 per cent of these episodes, with the remainder equally divided between central and mixed episodes. Eliaschar reported a series of 14 children with symptoms of moderate to severe upper airway obstruction. On polysomnographic monitoring, five patients had only central, obstructive, and mixed episodes. However, even "normal" children were found to have some apneic periods. Seven children without upper airway obstruction had an average of 21 central apneic episodes per night. Carskadon et al looked at 22 normal children in which an average of 18 central respiratory pauses per night were found, half of these being less than 10 seconds in duration. In the eight children with predominantly obstructive apnea described by Guilleminault, each apneic episode was accompanied by oxygen desaturation, at times down to 40 mm Hg. The oxygen saturation value, as determined by ear oximetry, depended mainly on the length of the apneic period, and was generally lowest following obstructive apnea, as compared with the central and mixed types. Tilkian suggests that this phenomenon may be caused not only by hypoventilation but by ventilation-perfusion mismatch in the lungs during respiratory obstruction.

To assess more accurately the degree of hypoventilation experienced by children with a history of sleep apnea, arterial blood gas measurement has been employed. In some severely affected children increased pCO₂ and decreased pO₂ may be found even in blood samples from the awake child. In many children, however, an indwelling arterial catheter must be inserted in order to compare blood gas values in the awake child often having normal values with those values when the child is sleeping and presumably experiencing the greatest obstruction.

Hemodynamic changes caused by airway obstruction are also seen. Radiographs of the chest may reveal cardiomegaly and there may be changes of right-sided hypertrophy on an electrocardiogram. Cardiac catheterization data are mostly limited to those children who have presented with cardiac complications, usually frank congestive heart failure. Features of cor pulmonale are found, such as right heart enlargement with elevated right-sided pressures and pulmonary hypertension. In some, evidence of left ventricular dysfunction is also apparent. When relief of the airway obstruction is obtained during catheterization, the abnormal values are seen to normalize. Abnormalities of systemic blood pressure are also seen in patients with airway obstruction. Guilleminault reported that systemic hypertension occurred in five of the
eight patients in his series. Blood pressure returned to normal following relief of the airway obstruction. In adults with predominantly obstructive sleep apnea, the normal pattern of decreased systemic arterial pressure during sleep did not occur and cyclic elevations in systemic arterial pressures began with the apneic episode. This was attributed to hypoxemia triggering sympathetic discharge and peripheral vasoconstriction. Disturbances in cardiac rate and rhythm have also been noted. Guilleminault reported marked sinus arrhythmias associated with sleep apneic episodes. Massumi describes a child with bradycardia in association with obstructive episodes.

Clinical Picture of Children With Sleep Apnea

Several behavioral aberrations have been noted in patients with sleep apnea. Excessive daytime sleepiness is quite often the presenting complaint, and may range from lethargy and increased napping to frank "sleep attacks" resembling narcoleptic episodes. However, in contrast to narcoleptic patients, these patients do not feel refreshed or rested after these sleep periods. During these daytime sleep attacks, apneic episodes may be observed. These sleep attacks may be secondary to both disordered nocturnal sleep and to CNS effects of cardiorespiratory dysfunction. Paradoxically, these children may appear hyperactive, as they move around trying to avoid falling asleep. The effects on school performance and mental functioning may be marked, with the children variously diagnosed as hyperactive, learning disabled, and in some cases mentally retarded. It is not known how much of a role the repetitive hypoxemia plays.

Nighttime behavior is also affected. In Guilleminault's series of eight cases of sleep apnea syndrome, three children fought going to sleep in their own rooms, and two of the children reported hypnagogic hallucinations. When children with the sleep apnea syndrome do fall asleep, loud snoring is reported nearly universally. Rather than lying supine, some of these children may require several pillows or may even prefer to sleep in a sitting position. Depending on the severity of the obstruction, sleep, as monitored by EEG, becomes more disturbed, with microarousals at the end of apneic episodes just prior to resumption of respiration. Parents have described their children's sleep as very restless, with abnormally increased body movement, sometimes accompanied by somniloquism and somnambulism. Enuresis occurring in previously night-dry children has also been reported to occur in patients with sleep apnea. Guilleminault reported the recurrence of enuresis in seven of the eight children in that series. It was noted that these enuretic episodes occurred all during the night, in contrast to the occurrence of primary enuresis mainly in the first third of the night (during slow-wave sleep).

The sleep apnea syndrome has been associated with both overweight and underweight conditions, and occurs in children of normal weight. In the series of eight children reported by Guilleminault et al five were underweight and two were overweight. Daytime somnolence was present in both groups. Stool et al described three children with recent weight gain and obesity, airway obstruction, and somnolence. These children experienced some degree of airway obstruction prior to their weight gain, which in turn seemed to increase their symptoms. After surgical relief of the obstruction, two children lost weight; the third was lost to follow-up. Menashe also reported one child who became obese during the illness but lost weight postoperatively.
Diagnosis

History and physical examination are the first steps in evaluating the degree and cause of chronic airway obstruction leading to sleep disorders. A detailed description of the child's sleeping habits must be obtained, noting excessive daytime sleepiness, sound or restless sleep, snoring, respiratory pauses or apneic periods, associated cyanosis and diaphoresis, position during sleep, nightmares, enuresis, and peculiar behaviors. Other associated symptoms include morning headaches, weight changes, dysphagia, behavior and mood changes, and declining school performance.

The physical examination of the awake patient may provide helpful clues to the source of the obstruction. Mouthbreathing, with or without noisy respirations at rest, may be noted. The nose must be examined for conditions and lesions which may be contributing to the obstruction, such as signs of allergy, infectious rhinorrhea, polyps, and anatomic problems such as a deviated septum. Direct or indirect visualization of the adenoidal mass may be possible in some children. Examination of the throat includes noting tonsillar size in relation to the size of the oropharynx. Speech qualities should also be noted, especially hyponasal speech suggesting nasal obstruction, and a "hot potato" voice suggesting a large amount of tissue in the oropharynx. The chest and heart should be auscultated for signs of cor pulmonale, including rales, a prominent P2 heart sound, and a gallop rhythm. Palpation of the abdomen for an enlarged liver or spleen may aid in this diagnosis as will edema of the extremities. Measurement of blood pressure is also a necessary part of the examination.

In addition to routine physical examination, patients should be examined while asleep or at least supine and relaxed. If the child is subject to excessive sleepiness, he or she may unwittingly demonstrate this problem during the course of the history and physical examination.

Radiographic studies may also be helpful. A cephalometric lateral neck radiograph can demonstrate the relationship between the size of the adenoid mass and the space in which it is contained. A small amount of radiopaque contrast material can be instilled into the nose to further demonstrate blockage of the nasal choanae. Cinefluoroscopy has been advocated as a noninvasive technique to demonstrate occlusion of the pharyngeal and hypopharyngeal airway during sleep. A chest radiograph is important in looking for evidence of cor pulmonale.

Polysomnographic monitoring is useful in documenting sleep disturbances and their relationship to airway obstruction. In the absence of a formal sleep laboratory, individual components of the study may be approximated. Respiratory and cardiac monitors may be used; audio and video tapes of the sleeping and awake patient may be compared. Arterial blood gas determinations employing an indwelling arterial catheter may be obtained. However, these are momentary values, and arterial catheters are not without their risks and complications. Noninvasive methods of continuous measuring of PaO2, PaCO2 and oxygen saturation employing cutaneously applied oxygen and carbon dioxide electrodes and ear-oximeters are currently being developed and used to obtain these values. Cardiac catheterization is performed when indicated. Kravath et al have advocated the placement of a nasopharyngeal tube in patients with suspected airway obstruction, and comparison of
polygraphic studies and blood gas values before and after tube placement as a method of diagnosis.

**Treatment**

In most children, symptoms of the sleep apnea syndrome are relieved by elimination of the upper airway obstruction, usually by adenoidectomy with or without tonsillectomy. In some cases, previously adenoidectomized children will display the syndrome secondary to tonsillar hypertrophy alone. In some children, however, adenoidectomy and tonsillectomy are not sufficient, suggesting a greater CNS role in failure to maintain upper airway patency, and improvement is noted only after an airway is established by use of a nasopharyngeal tube or tracheotomy. A tracheotomy valve may be used, which can be closed during the day and opened only at night. In well selected cases tracheotomy essentially reverses all aspects of the sleep apnea syndrome. In children with other sites of airway obstruction, treatment must be aimed at the specific problem.

**Summary**

Improved case identification of children with upper airway obstruction during sleep should result if physicians are aware of such signs and symptoms as excessive daytime sleepiness, loud snoring, restless sleep, recurrent nocturnal enuresis, systemic and pulmonary hypertension, undergrowth or obesity, and cor pulmonale. Furthermore, partial airway obstruction during wakefulness may be a risk factor for the development of sleep apneas or hypopneas. In suspected cases, polysomnography is a useful method for confirming and quantitating the type (central, obstructive, or mixed) and extent of ventilatory disturbance during sleep and its functional significance (such as arterial oxyhemoglobin desaturation or cardiac arrhythmia). Other methods may be employed to yield similar data.

There seem to be at least two groups of children reported in the literature, those in whom there is a specific surgically correctable lesion (such as adenotonsillar hypertrophy) versus those who eventually need tracheotomy because of collapse of upper airway musculature during sleep. In the latter group of children, it is necessary to hypothesize an additional defect in the CNS regulation of respiration during sleep.

Further research is necessary to define the boundary between normal and abnormal breathing during sleep, and to understand more thoroughly the effects of intermittent hypoventilation on daytime functioning.
Radiologic Imaging in Otorhinolaryngology

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Technical Aspects and Advances

The radiographic technique is foremost among the methods of radiologic imaging, despite the potential biologic hazard of ionizing radiation in children. Irradiation can be decreased by close adherence to diagnostic indications, thus precluding unnecessary examinations. However, for specific clinical problems, many of which exist in otorhinolaryngology, the radiographic technique is the only available method of evaluation.

Effective and noninjurious immobilization of the child is important as the first step in the special effort that must be made to eliminate unnecessary irradiation. Collimation of the x-ray beam is another. Recent technologic advances in radiologic imaging have provided additional means for reducing irradiation. For the past five years we have been using a high speed film screen system (the Kodak Lanex rare earth screen with Ortho G film) for most pediatric roentgenographic examinations. This green sensitive film with rare earth screen system decreases irradiation by almost half, as compared to the radiation needed for a par speed film system. The use of carbon fiber for radiographic tables and screens, although expensive, offers another substantial decrease in radiation exposure.

Video fluoroscopic evaluation is often mandatory in evaluating the upper airway, swallowing function, and phonation. We use a low-dose video fluoroscopic system in which only 30 to 40 mrad/min are required to obtain fluoroscopic images of satisfactory diagnostic quality in small children. This videotape fluoroscopic recording system offers a substantial reduction in irradiation by eliminating the need for repetitive fluoroscopy for diagnostic evaluation or for teaching.

Recent advances in computed tomography (CT) which are especially important for pediatric patients include reduction in radiation exposure and shorter scanning times. The images of 1.5 mm sections of temporal bones obtained by specially designed CT modifications have better resolution than those obtained by conventional thin section tomography.

Because no ionizing radiation is involved, ultrasound is uniquely useful in pediatric imaging and improvements are ongoing. Recently, ultrasound has been widely accepted as an important method for evaluating intracranial lesions in young infants.

Radionuclide imaging in otorhinolaryngology is limited, but can be used to evaluate neoplasms or inflammatory lesions affecting bone. Radiation dosage to the patient is relatively low in these procedures.

Close collaboration between clinicians and radiologists is necessary for the optimal choice of available imaging modalities.
Airway Obstruction

In this discussion, obstruction of the airway is defined as an impediment to ingress or egress of air occurring between the mouth or nose and the main bronchi. Airway obstruction may be congenital or acquired.

Congenital Obstruction

Congenital airway problems amenable to radiologic diagnosis are choanal atresia, Pierre-Robin anomaalad, and congenital stridor. The Pierre-Robin anomaalad and bilateral choanal atresia are apt to cause life-threatening respiratory distress from birth. A lateral radiograph of the nasopharynx in the Pierre-Robin anomaalad will suffice to confirm mandibular hypoplasia and to show the degree of airway occlusion caused by glossoptosis. Choanal atresia is confirmed by demonstrating that a small amount of sterile propylidione oil suspension (Dionsil Oily) instilled into the nasal cavity does not pass into the posterior nasopharynx. This procedure is preferably done under video fluoroscopic control with a horizontal x-ray beam, raising the table to an upright position, and placing the infant supine on the footboard. Choanography is facilitated by removing excess secretions and instilling a few drops of 0.25 or 0.5 per cent phenylephrine hydrochloride before injecting contrast medium.

Floppy aryepiglottic folds, misleadingly known as "laryngomalacia", is by far the most common cause of neonatal stridor. Fluoroscopy in the lateral view may show characteristic infolding of the epiglottis and aryepiglottic folds on inspiration, but a radiologic diagnosis can only be made if the infant exhibits stridor during video fluoroscopy.

Radiographs may show vocal cord paralysis, tracheal stenosis, laryngeal web, or a soft tissue mass causing respiratory distress in the newborn. Supraglottic and glottic masses include ectopic thyroid and thyroglossal, branchial cleft, glottic, and aryepiglottic cysts. Hemangioma and mucocele are characteristically subglottic in location.

Acquired Obstruction

Most acquired airway obstructions occur in the hypopharynx or larynx and produce stridor. Croup (laryngotracheobronchitis) and acute epiglottitis are most common. If the degree of obstruction is not of such severity that immediate endotracheal intubation is necessary, anteroposterior (AP) and lateral radiographic finding in croup is the *steeple sign*, a tapered narrowing of the subglottic portion of the trachea in the AP view. The caliber of the trachea in the lateral view may not be obviously diminished. In bacterial tracheitis ("membranous croup") radiographs may show concentric tracheal narrowing and irregular filling defects within the tracheal lumen.

A radiographic finding common to all lesions that impede the entry of air at or below the epiglottic level is ballooning of the hypopharynx. Marked ballooning reflects a more severe degree of obstruction and often is absent when obstruction is mild.

Acute bacterial epiglottitis causes a broadening of the epiglottic shadow in the lateral view of the neck. Involvement of the aryepiglottic folds and surrounding tissues has prompted
the suggestion that *supraglottitis* may be a more precise term. Other causes of epiglottic swelling include traumatic hematoma, angioneurotic edema, and caustic burns.

Laryngeal papilloma, the most common laryngeal tumor of childhood, may be visible on radiographs. Pulmonary spread of papillomas has been reported.

An aspirated foreign body lodging in the hypopharynx, larynx, or upper trachea may produce stridor. Metallic objects of heavy density are easily seen on plain radiographs. Objects of plastic, aluminum, or vegetable matter, similar in density to soft tissue, may be visible when surrounded by air. Very small objects, especially those of soft tissue density, may not be distinguishable with conventional radiographic technique. Xeroradiography may show objects that escape detection on conventional radiographs.

A foreign body lodged in the distal trachea or bronchus may produce wheezing rather than stridor. An esophageal foreign body large enough to compress the adjacent trachea also may cause respiratory distress. Incomplete occlusion of a bronchus most often produces a ball-valve type of obstruction, permitting air to enter but not to exit the distal segments. This produces air trapping (obstructive emphysema) in the involved lobe or segment, or in the entire lung if the occlusion involves a main bronchus. Complete bronchial occlusion eventually causes atelectasis.

When severe, obstructive emphysema produces obvious hyperinflation in the involved portion of the lung on a full-inspiratory frontal radiograph of the chest. The cardiomedialntal structures are displaced toward the contralateral side and the volume of the contralateral normal lung appears diminished. These findings are sometimes misinterpreted as representing atelectasis involving the unobstructed lung. If air trapping is not severe the findings may be subtle. A radiograph obtained in full expiration may unmask obstructive emphysema that is not apparent on the inspiratory radiograph. An expiratory radiograph is easily obtained in a cooperative child, but in a younger child or one who is breathing rapidly it may be necessary to produce forced expiration by pressing upon the child's abdomen with a lead-gloved hand during the radiographic exposure.

Video fluoroscopy of the chest is more sensitive for detecting minimal unilateral air trapping than are plain inspiratory-expiratory radiographs. Video fluoroscopic findings of obstructive emphysema are displacement of the mediastinum toward the normal lung during expiration, returning toward the midline on inspiration; diminished excursion of the hemidiaphragm on the abnormal side; and hyperinflation of the obstructed portion of the lung during expiration. The fluoroscopic findings may be very subtle, with slight displacement of the mediastinum apparent only when the child is crying or sobbing. In our experience, use of right and left decubitus views to detect obstructive emphysema is less satisfactory than video fluoroscopy.

If a foreign body lodged above the carina or in the esophagus produces ball-valve tracheal obstruction, chest fluoroscopy will confirm only that there is ineffective emptying of both lungs, indistinguishable from diffuse air trapping of bronchospasm.
**Radiographic Evaluation of Upper Airway Obstruction**

Frontal and lateral views of the chest and neck are essential baseline radiographs for evaluating upper airway obstruction. The neck can be included in the frontal view of the chest if the child's head is positioned so that the chin does not obscure the upper airway. A lateral view of the chest, with shoulders abducted and arms held behind the patient (Pancoast position), also affords a clear view of the neck in the lateral projection, although in older children we prefer a separate lateral view centered on the pharynx. If there is great urgency because of severe respiratory distress a lateral view alone (often obtained in the emergency department with portable radiographic equipment) will serve to confirm or exclude epiglottitis or the presence of a radiopaque foreign body in the upper airway.

Joseph et al devised a radiographic technique using high kilovoltage, filtration, and magnification to overcome the usual difficulty visualizing the trachea superimposed on the cervical spine in the AP view. Their results were impressive, but we have had no experience with the technique. Xeroradiography or tomography may be useful when conventional radiographs have not satisfactorily shown the larynx or trachea. However, both of these methods result in greater radiation exposure than conventional radiographs and therefore are not used routinely.

With any of these radiographic techniques careful attention must be paid to good positioning. Poor inspiration or flexion of the neck may cause normal buckling of the infant's trachea. Slight rotation of the neck in the lateral view may simulate epiglottic swelling. The normal omega-shaped infantile epiglottis also may be confused with epiglottic swelling.

Evaluation of the larynx with radiopaque contrast medium is rarely indicated in children. Satisfactory radiographic detail of the laryngeal structures can usually be obtained, when necessary, by thin section tomography or CT using the normally present air as the contrast medium.

The chest radiograph is a valuable adjunct in the child with airway obstruction. Pulmonary edema may occur concurrently with the obstruction or may develop after insertion of an airway to relieve obstruction (see related discussion by Davis et al elsewhere in this issue).

**Disorders of Swallowing**

**Pharyngeal Incoordination**

Pharyngeal incoordination refers to any functional disturbance of propulsion of ingested material from the mouth into the esophagus. Clinical manifestations include nasal regurgitation of feedings and choking, coughing, or gagging. Abnormal function of the cricopharyngeus muscle, the pharyngoesophageal constrictor, is the underlying problem in many of these patients. The cricopharyngeus may relax sluggishly when the bolus reaches it, as in dysautonomia, or the cricopharyngeus may be hypertonic throughout swallowing. Whether this abnormal tone is called "achalasia" or "spasm" depends upon the manometric pressure pattern.
The video fluoroscopic findings in cricopharyngeal spasm or achalasia are specific and easily recognized. Swallowing of barium is observed in the lateral view. When the swallowed bolus reaches the cricopharyngeus the sphincter fails to open completely and part of the bolus is retained in the hypopharynx. The retained portion of the bolus may be regurgitated into the mouth or nose, may be aspirated into the trachea, or may simply pool in the hypopharynx despite repeated swallowing efforts by the child.

The key finding is an abnormal constriction at the level of the pharyngoesophageal sphincter as the bolus passes from the hypopharynx into the esophagus. This can be a relatively long segment of concentric narrowing or a short, transverse, bar-like posterior indentation on the barium column. Giedion and Nolte observed in some infants a prominent cricopharyngeus impression that did not cause obstruction and this they regarded as a normal physiologic variation.

Fatigue aspiration is a phenomenon in which otherwise normal infants may begin to aspirate toward the end of feedings. The pathophysiology is unknown. Esophagographic confirmation requires that the quantity of barium given be sufficient to simulate a normal feeding.

Swallowing disorders are best studied radiographically by recording the examination on videotape, which permits repeated slow-motion observation of swallowing without subjecting the child to excessive fluoroscopic irradiation. We also use a videodisc that records onto 15 still-frames per second and can be replayed for evaluation at slower frame rates or as single frames.

Cervical Esophageal Disorders

Many of the problems involving the intrathoracic portion of the esophagus that produce swallowing disturbances are, for want of space, outside the scope of this communication. Laryngotracheoesophageal cleft and pharyngeal diverticulum are two uncommon congenital lesions that pose considerable diagnostic difficulty. Pharyngeal diverticulum may simulate esophageal atresia clinically but can be demonstrated by an esophagogram. It is likely that some reported "congenital" diverticula actually were unrecognized traumatic pseudodiverticula. Laryngotracheoesophageal cleft both clinically and radiographically mimics H-type tracheoesophageal fistula. Barium inundating the trachea through the cleft makes esophagography difficult and somewhat hazardous. The endoscopist hardly fares better, many clefts having been missed on laryngoscopy. Felman and Talbert recommend esophagography with an endotracheal tube in place.

By far the most common acquired lesion of the esophagus is an impacted foreign body. Whenever an ingested foreign body is suspected we obtain radiographs to include the entire alimentary tract from mouth to rectum. If these do not reveal an opaque foreign body, and signs and symptoms persist, an esophagogram is performed. The three sites where esophageal foreign bodies most often lodge are the thoracic inlet, the indentation on the esophagus produced by the left main bronchus, and the diaphragmatic hiatus. Sharp objects often catch in the pyriform sinuses. Some radiologists use an inflated Foley cather technique to extract blunt foreign bodies from the esophagus, but our surgeons prefer endoscopic extraction.
Acquired stricture of the esophagus may result from ingested caustics or from peptic reflux esophagitis. Food or other swallowed objects impacting at a stricture can acutely obstruct the esophagus. The location and extent of strictures, and the effect of therapeutic esophageal dilatation, are well evaluated by barium esophagography. We use water-soluble contrast media only when esophageal perforation is suspected.

A traumatic pseudodiverticulum results from laceration of the posterior pharyngeal wall, often by instrumentation such as forceful passage of a nasogastric tube. There may be an associated cricopharyngeal spasm identical to neurogenic cricopharyngeal achalasia.

Retropharyngeal abscess widens the prevertebral soft tissues in the lateral view of the nasopharynx. Either flexion of the neck or the expiratory phase of respiration may produce considerable forward bulging of the prevertebral soft tissues in the normal infant. An equivocal finding may be resolved by repeating the radiograph with careful attention to good inspiration and extension of the neck, or by having the child swallow a small amount of barium under video fluoroscopic observation.

The barium esophagogram is the key to radiographic evaluation of swallowing disorders, regardless of the suspected cause. The single exception is a radiopaque foreign body identified on plain films, for which an esophagogram is superfluous. However, if a foreign body has lodged in an unusual location it is prudent following removal of the object to obtain an esophagogram which may reveal an unsuspected stricture.

**Soft Tissue Masses in the Neck**

Whether or not to evaluate soft tissue swelling in the neck with radiography is a decision that must be based on clinical circumstances. In most instances of superficial soft tissue inflammation or lymphadenopathy there is little to be gained from plain radiographs. However, with soft tissue swellings or masses of less certain etiology, or those obstructing the airway by extrinsic compression, radiographs and other imaging modalities may give useful information. The displacement of normal air-filled structures in frontal and lateral radiographs of the neck may indicate the extent of soft tissue masses. Contrast medium given by mouth will show if the cervical esophagus or hypopharyngeal structures are displaced. Cystic hygroma, ectopic goiter, teratoma, lymphadenopathy, and neoplasms may produce radiographic widening of the prevertebral soft tissues indistinguishable from retropharyngeal abscess.

Computed tomography (CT) with intravenous contrast enhancement can indicate the vascularity of a lesion as well as demonstrate fat content or calcification that may not be apparent on plain radiographs. CT has proven useful for evaluating both laryngeal disorders and neck masses.

Most soft tissue masses in the neck are accessible to ultrasound, and state-of-the-art instrumentation affords superb anatomic definition. Ultrasound is used widely for the evaluation of thyroid enlargement or nodules, but we believe that this modality is underutilized for cervical masses. Ultrasound delineates the size and cystic or solid nature of a mass without the potential radiation hazard of CT. Ultrasound might become valuable as the initial imaging procedure for cervical masses as it often is for abdominal masses.
An enlarged salivary gland is commonly subjected to radiographic assessment. Plain radiographs may show a salivary duct stone. Sialography, the injection of oily contrast medium into a salivary duct, can confirm a clinical diagnosis of sialadenitis or can outline a salivary gland tumour. Ultrasound of the salivary gland is potentially useful.

**Complications of Tracheostomy: Radiographic Evaluation**

Tracheostomy, one of the most ancient of operative procedures, has undergone dramatic improvements in technique as well as in cannula design and materials. Although the incidence of acute complications has decreased considerably in recent years, a substantial risk of late complications still exists following tracheostomy in infants and children. The combined incidence of tracheal stenosis and granuloma after tracheostomy ranges from 1.9 to 4.3 per vrn, but in a recent prospective study the incidence of tracheostomy-induced complications was 26 per cent.

After decannulation slight irregularities of the trachea can persist normally for about 4 weeks in infants and for as long as over a year in young children. Tracheal stenosis and granuloma can occur almost any time after tracheostomy; granulomas can grow slowly over a number of years. These obstructive lesions occur most commonly at two specific sites: the upper margins of the stoma and that place in the trachea where the distal tip of the tracheostomy tube rubs. Granuloma occurs most often at the superior anterior margin of the stoma. After decannulation, periodic follow-up radiographic evaluation is essential to confirm normal healing without complications.

The lateral radiographic view usually shows the findings to better advantage, although the AP view is also needed for complete evaluation. High kilovoltage (140 KVP) technique may improve visualization in the AP view. Xeroradiography can be of additional value because of its edge enhancement effect if the conventional radiographs show equivocal findings. Tracheobronchography is rarely indicated, and can best be done with tantalum powder for which one needs a license from the US Food and Drug Administration. Posttracheostomy tracheomalacia may occur with or without tracheal stenosis or granuloma. Video fluoroscopy is the best method to evaluate tracheomalacia. We suspect that the incidence of tracheomalacia is at least as high as that of the obstructive lesions, but the true incidence of segmental tracheomalacia following tracheostomy has not been adequately documented.

**Voice Disorders**

Video-velopharyngography (VVPG), a fluoroscopic examination recorded on videotape with simultaneous voice recording, is a most useful method of evaluating patients with speech problems. Adequate coordination of complex motions of the tongue, soft palate, and lateral pharyngeal walls is essential for normal speech. Closure of the velopharyngeal portal during phonation is the result of two types of motion: midsagittal velopharyngeal contact by elevation and posterior extension of the soft palate, and medial motion of localized regions of both lateral pharyngeal walls.

The VVPG is performed by a radiologist in close collaboration with the speech pathologist. Before starting, the VVPG is explained to and rehearsed with the patient, whose
full cooperation is essential in this examination. The standard procedure and observations are as follows: (1) Lateral view: the patient is positioned erect and gross anatomy of the facial bones and nasopharyngeal air passage are examined. (2) Injection of radiopaque contrast material: a barium mixture (approximately 120 volume per cent) is used to coat the pharyngeal mucosa. About 3 to 4 mL of the barium mixture is injected into each nostril with hyperextension of the neck to facilitate coating. (3) Frontal views: the patient is examined erect with the chin slightly up. Medial movement of the lateral pharyngeal walls is examined. Symmetric approximation of the upper part of the lateral pharyngeal walls occurs in normal children. During swallowing of the pharynx completely contracts. (4) Lateral view: the size and configuration of barium-coated adenoids and soft palate are well examined in this projection. Complete closure of the velopharyngeal portal should occur during phonation of "P", "K", and "S" sounds. However, the medial motion of lateral pharyngeal walls may contribute to velopharyngeal closure even when velopharyngeal closure is incomplete in this projection. (5) Lateral hyperextension view: in this projection the velopharyngeal gap, if present, may be more pronounced. Passavant's ridge, a local protrusion of a portion of the posterior pharyngeal wall, can be seen well in this position. (6) Basilar view: the patient is placed prone and the neck and head are hyperextended to obtain a tangential projection at the level of velopharyngeal closure. Either a circular or an oval shaped velopharyngeal approximation may be observed depending upon which motion, transverse or sagittal, is mainly responsible for its closure. This view is the most important for evaluation of patients with cleft palate who have had pharyngeal flap surgery, because the velopharyngeal openings are lateral to the flap and are demonstrated only in this projection.

The consultative discussion might include, in addition to the radiologist and speech pathologist, the pediatrician, the plastic surgeon, and the otolaryngologist.

Radiographic Evaluation of the Paranasal Sinuses

The paranasal sinuses (PNS) are pneumatized cavities which communicate with the nasal cavities and are lined by respiratory epithelium. The maxillary and ethmoidal sinuses, although small, are fully developed at birth. By three years of age the maxillary sinuses may be as large as 2 mL in volume and radiologic evaluation is feasible. The frontal sinuses begin their ascent into the frontal bone at two years, reach the nasion at three years, and become radiographically visible at six years of age. They are not fully developed until adolescence. The frontal sinuses may be absent or hypoplastic in a few normal children. The sphenoidal sinuses begin to pneumatize between two to six years and are fully developed at adolescence. Like the frontal sinuses, the sphenoidal sinuses are commonly asymmetric and septated.

The conventional radiographic examination of the PNS is the simplest, least expensive, and most practical examination. It includes the frontal (Caldwell), occipitomental (Waters), and lateral erect views. The basal (submentovertex, axial) view is done on selected patients whose ethmoidal or sphenoidal sinuses are equivocally abnormal in the routine views. A reverse Waters view is performed in children younger than five years. The techniques for all these views are described by Darling. Limitation of space does not allow a detailed description of normal radiographic anatomy.

The interpretation of PNS radiographs in children younger than three years is limited by the small size, nonpneumatization, redundancy of mucosal lining, or presence of tears in
the maxillary sinuses. These limitations are compounded by superimposition of roentgen images. Thin section tomography (TST) is a radiographic method which allows examination of a thin body section. This modality has tremendously enhanced knowledge of both normal and abnormal roentgen anatomy and supplements the features that are seen on the plain radiographs. TST is closely monitored by a radiologist who should know the clinical problem prior to the examination. Leaded eyeglasses are used on all patients (whenever possible) to reduce eye lens irradiation. The indications for TST are: (1) to further define an obscure abnormal finding suspected on plain radiographs; (2) to define the extent of injury in multiple facial fractures; (3) to evaluate congenital anomalies; (4) to determine the size and extent of a tumor in relation to adjacent structures; and (5) to evaluate postoperative changes, tumor recurrence, and efficacy of treatment.

The radiographic signs of PNS disease include variable degrees of opacification, mucosal thickening, air-fluid levels, bony erosion, mass lesion (solid or cystic), foreign body, calcification, and fracture. Meaningful interpretation of these roentgen findings rests on correlation with the clinical signs and symptoms.

**Congenital Anomalies**

Congenital anomalies of the PNS are usually associated with the first and second branchial arch syndromes (Treacher-Collins, cleft palate, and so on). Isolated hypoplasia or agenesis of the maxillary or ethmoidal sinuses are uncommon and can mimic mucosal thickening or opacification on the radiographs. Aplasia of sphenoidal sinuses is extremely rare. When pneumatization of the sphenoidal sinuses is not clearly shown on routine radiographs in patients with nonspecific intracranial symptoms, TST is indicated.

**Infections**

Sinusitis, the most common PNS disease in children, is an inflammation of the sinus mucosal lining which is continuous with the nasal cavity. The maxillary and ethmoidal sinuses are the most commonly involved, singly or in combination. The radiographic findings of variable degrees of opacification, air-fluid levels, or mucosal thickening are not specific for sinusitis. Correlation with the clinical findings is necessary for a definitive diagnosis.

The maxillary sinuses are closely related to dentition. Unerupted molars can mimic masses, partial opacification, or air-fluid levels in the antra. In older children, maxillary sinusitis may result from extension of an alveolar abscess. Orthopanographic examination is an excellent diagnostic aid.

Homogeneous opacification of the maxillary and ethmoidal sinuses is an almost constant finding in patients with cystic fibrosis. This may be caused by tenacious secretions, mucosal redundancy, infection, or by polyps obstructing the ostia.

Acute ethmoidal sinusitis is a major cause of periorbital cellulitis and orbital abscess. The plain radiographic finding of diffuse opacification of the ethmoidal and/or maxillary sinuses associated with the characteristic clinical findings (lid edema, proptosis, ophthalmoplegia, and abnormal vision) are sufficient to make the diagnosis. However, TST
and CT are extremely helpful in demonstrating the site and extent of subperiosteal abscess or extension into the intracranial structures.

Acute frontal sinusitis, more commonly seen in older children, is an important disease because of the high incidence of intracranial extension. The mucoperiosteal borders and air and fluid content should be carefully evaluated. CT is the method of choice to evaluate intracranial extension.

Isolated infection of the sphenoidal sinuses is rare but sphenoidal involvement in pansinusitis is not uncommon. The vital structures around the sphenoidal sinus make early diagnosis of sphenoidal sinus disease extremely important. Air-fluid levels in the sphenoidal sinuses may occur in infection, posttraumatic hemorrhage, or from accumulated secretions during prolonged recumbency.

Overgrowth of the paranasal sinuses is a common finding in patients with mental retardation. It is also seen after ventriculoperitoneal shunting. Unilateral overgrowth of the sinuses is a characteristic finding in patients with Dyke-Davidoff syndrome (unilateral cerebral atrophy with ipsilateral thickening of diploic space, hypertrophy of PNS, and elevation of petrous bone).

**Tumors**

A soft tissue mass in a PNS may be either a polyp or a mucous retention cyst. A polyp may obstruct the ostium or cause bony erosion. TST is indicated for evaluation of the extent of bony abnormality before surgery. Polyps occur in 10 to 30 per cent of patients with cystic fibrosis. Mucous retention cysts are very common in children. These cysts frequently occur at the floor and occasionally at the roof of a maxillary sinus. In the Waters view retention cysts appear as semicircular shadows of increased density without associated bony destruction.

A primary neoplasm of the PNS is extremely rare in children. Malignant neoplasms, such as lymphoepithelioma, rhabdomyosarcoma, and lymphosarcoma may invade the sinuses. The plain radiographic findings include a soft tissue mass, erosion of adjacent bone, and sclerosis. TST and CT usually define the size of the neoplasm and the extent of bone destruction. Nonmalignant diseases such as anterior meningocele, Wegener's granulomatosis, mucormycosis, and juvenile nasopharyngeal angiofibroma can invade the PNS and deform or destroy their walls.

**Fractures**

Maxillofacial trauma in children differs from that in adults. Unerupted and mixed dentition gives stability and elasticity to the facial bones, thus greater force is required to fracture these bones. Multiple fractures should be carefully looked for.

Blowout fracture is a result of a blow to the orbit transmitting force to the globe and increasing intraorbital pressure. The weakest point in the orbital wall is the floor medial to the infraorbital foramen. Fracture at this point may permit herniation of the orbital contents into the maxillary sinus. Clinical signs are ecchymosis, diplopia, and ophthalmoplegia. The
plain radiographic findings are a soft tissue mass in the antral roof, haziness of the maxillary sinus owing to overlying soft tissue swelling or hemorrhage into the sinus, and fracture. Tomography is usually done to define a fracture when one is not clearly shown on plain radiographs.

The Le Fort classification of multiple facial fractures is very useful. Plain radiographic examination is limited because of marked soft tissue swelling or hematoma. TST defines the fractures and displacement of fragments.

Maxillary Sinus Ultrasound

A few reports have indicated that A-mode ultrasound is useful for detecting the presence of fluid in the maxillary sinuses. An investigation currently underway will correlate clinical, radiographic, and sonographic findings with the bacteriologic data from sinus aspirates in children suspected of having maxillary sinusitis (see discussion by Wald et al elsewhere in this issue). In our first 44 subjects, sinuses that were completely opaque on radiographs have invariably produced an abnormal ultrasound pattern but we have not yet established any other meaningful correlations. Several pitfalls in sinus sonography have surfaced. A few patients with mucosal thickening on radiographs and positive aspirates have had no discernible sonographic abnormality. Technical sources of error abound and we have had difficulty distinguishing normal from abnormal ultrasound patterns in young children. Any conclusions regarding the value of ultrasound for evaluating children's maxillary sinuses must await further experience with the method.

Radiologic Evaluation of Hearing Disorders and Related Problems

Congenital Anomalies

Detailed clinical evaluation and various audiologic tests are supplemented by TST. In preoperative evaluation of congenital abnormalities patency of the external auditory canal, the size of the tympanic cavity, integrity of the ossicular chain, course of the facial nerve canal, and presence of the internal ear structures are necessary information. Fitz and Harwood-Nash listed 20 syndromes associated with temporal bone anomalies. Membranous anomalies of the inner ear cannot be identified on radiographs. The type of hearing loss (conductive, neurosensory, or combined) is important information to the radiologist evaluating the radiographic findings.

Infections

Infection is the most common cause of acquired hearing loss in children. Serous otitis media, acute suppurative otitis media, and mastoiditis may produce abnormal findings on plain radiographs (Towne, Laws, Stenvers, and lateral nasopharyngeal views). The degree of mastoid cell pneumatization, presence of bony erosion in the epitympanic space, and size of the adenoids are important findings. Enlarged adenoids can obstruct the upper airway and eustachian tube. Obstruction of the latter can cause serous otitis media. The anterior and inferior margins of the adenoidal mass are well outlined by air on a lateral nasopharyngeal radiograph. Adenoidal-nasopharyngeal (AN) ratio is a simple and accurate method to objectively evaluate adenoid size. An AN ratio above 0.80 indicates enlarged adenoids.
Chronic otitis media with secondarily acquired cholesteatoma is best evaluated with TST. The characteristic findings are destruction of the attic spur (scutum) and lateral attic wall, haziness (with or without soft tissue mass) in the tympanic cavity, and destruction or displacement of the ossicles. In chronic ear infections, complications such as brain abscess, otitic meningitis, Gradenigo syndrome, and dural sinus thrombosis are evaluated with TST or CT scan. Repeated episodes of meningitis may be caused by a fistula in the oval window, CSF leak caused by trauma, or chronic mastoiditis.

**Trauma**

Clinically, fractures involving the temporal bones present with bleeding from the ears, CSF otorrhea, hearing loss, facial nerve paralysis, or Battle's sign. The two kinds of temporal bone fractures are longitudinal (85 to 90 per cent) and transverse (10 to 15 per cent). Longitudinal fractures lie parallel to the petrous bone and involve the ossicles and walls of the external auditory canal and tympanic cavity. These fractures are best seen in the lateral tomogram. Transverse fractures lie transversely through the axis of the petrous bone and usually result from a direct blow to the occiput and severe trauma to the anterior and middle cranial fossae. The internal ear structures bear the brunt of temporal bone trauma and are best seen in the frontal tomogram.

The plain radiographic findings are opaque mastoid air cells, diastasis of the lambdoid or occipitomastoid sutures, and a fracture line. TST in two projections is indicated. In the frontal tomogram decreased air content in the external auditory canal and tympanic cavity, ossicular chain disruption, or radiolucent defects in the tegmen tympani or inner ear structures indicate presence of a fracture. In the lateral tomogram, fracture of the external auditory canal wall, ossicular disruption, and opacity or soft tissue mass in the tympanic cavity are characteristic findings. When facial nerve paralysis is present the facial nerve canal should be evaluated in both views.

**Tumors**

Deafness caused by temporal bone tumors is rare in children. Acoustic neuroma, histiocytosis X, rhabdomyosarcoma, and epidermoid cyst are the four major tumors that affect the temporal bone. Definitive diagnosis of these lesions rests on histologic examination. The plain radiographic findings of acoustic neuroma include asymmetry (difference of more than 2 mm in height) of the internal auditory canal (IAC), erosion of the IAC's porus acusticus, and sclerosis around the IAC. A small extracanalicular acoustic neuroma may produce subtle changes around the porus acusticus. Thin section tomography and/or CT increase the diagnostic accuracy. The findings on CT are mass of increased density in the cerebellopontine angle, obliteration and displacement of the fourth ventricle, and widened cisternal angle. More sophisticated CT equipment may obviate the use of invasive procedures such as arteriography, posterior fossa myelography, and amipaque CT cisternography in the future.

Eosinophilic granuloma affects the temporal bone in 25 per cent of the cases. On plain radiographs a radiolucent defect with no significant reactive sclerosis is seen as an isolated lesion in the petrous bone or as part of multiple skull lesions. Similar lesions may involve the remainder of the skeleton.
Fifty per cent of embryonal rhabdomyosarcomas are located in the head and neck. Only 7 per cent of these affect the ear. This neoplasm commonly masquerades as chronic otitis media, inflammatory polyp, or granulation tissue in the external or middle ear. Plain radiographs may be normal. TST shows mass effect and bony erosion, and CT shows a contrast-enhancing mass.

Epidermoid cysts are commonly located in the midline but those that are laterally located involve the base of the brain or temporal bone. These are round masses or cysts lined by epithelial tissues. On plain radiography, they appear as round radiolucent defects with sclerotic borders. CT is valuable in demonstrating extension of the cyst into the intracranial structures. The major CT finding is a mass that does not enhance with intravenous contrast medium.
Subglottic Stenosis

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Subglottic stenosis refers to a narrowing of the space bounded inferiorly by the inferior margin of the cricoid cartilage and superiorly by the inferior surface of the vocal cord. This space is the narrowest portion of the upper airway, the lumenal diameter in a full-term newborn baby being 4.5 to 5 mm. This key area is predisposed to injury because of certain inherent anatomic features: it is a circular structure surrounded by a rigid, intact, cartilaginous ring abutting the spinal column that is exposed to external trauma. In addition to these factors, this area is the narrowest portion of the airway lined by respiratory epithelium that is prone to mechanical trauma and submucosa of loose connective tissue (which is capable of edematous swelling), thus rendering it susceptible to internal (endolaryngeal) trauma.

Congenital Subglottic Stenosis

Subglottic stenosis is considered to be congenital in the absence of a history of endotracheal intubation or other apparent cause. It is the most common laryngeal disorder that might produce serious airway obstruction requiring tracheotomy in infants under one year of age. Congenital laryngotracheal anomalies, such as tracheoesophageal fistula, tracheal stenosis, and vocal cord paralysis, occur in 7 per cent to 10 per cent of patients with subglottic stenosis, and congenital anomalies not related to the larynx and trachea have been reported in 10 per cent of these patients.

The narrowing of the lumen may be caused by both cartilaginous and soft tissue deformities. Cartilaginous ring has been described to be unusually small in diameter or elliptical in shape; in rare cases, the first tracheal ring is "trapped" or "telescoped" inside the anterior cricoid arch. The soft tissue abnormality results from thickness of the submucosa that is caused by either connective tissue or by hyperplastic dilated mucous glands. The narrowing is circumferential or could occupy only part of the ring. The symptoms and their severity, and age at onset are clearly related to the amount of narrowing of the subglottic space. In severe cases, respiratory distress associated with stridor will be present at birth; this was true for half of the subjects in the largest series of patients with congenital subglottic stenosis reported by Hollinger. Milder cases become evident during the first weeks or months of life, presenting with either a prolonged episode of croup or recurrent croup. Laryngeal inflammation can precipitate severe respiratory distress since the limiting cricoid cartilage only permits inward swelling of tissues at the expense of the airway. Thus an airway already compromised by congenital subglottic stenosis will become inadequate by the slight edema associated with a viral infection. Ten per cent of these children will have recurrent pneumonitis. A barking or brassy cough may be the only symptom of minimal subglottic stenosis. The most important sign, if present, and that in no way is pathognomonic, is stridor, which is often both inspiratory and expiratory, and of low pitch.

The diagnosis of congenital subglottic stenosis is strongly suggested by the history and clinical presentation, assuming that a high index of suspicion exists. Lateral and anteroposterior neck films, laryngeal tomograms, and lateral xeroradiograms may provide
further evidence, but the ultimate proof is the endoscopic diagnosis. Subglottic stenosis is confirmed in a full-term newborn infant if a 3 mm infant bronchoscope (having an outer diameter of 4 mm by 4.5 mm) cannot pass readily through the subglottic region. A complete evaluation of the tracheobronchial tree is mandatory to rule out other concomitant breathing anomalies.

The management of congenital subglottic stenosis is based on the experience that children outgrow the disorder. In our experience, less than half of the patients will require tracheostomy as an alternative airway, most of which will be decannulated within two years. Only a small number will require surgical correction. The smallest tracheostomy tube that will permit adequate ventilation should be used in order to allow preservation of the function of the larynx and to permit the child to phonate. With the newly developed tracheostomy tube, adequate care can be administered at home after the parents receive appropriate instructions. The mortality rate in this group of patients, which is directly associated with tracheostomy, is still about 2 per cent. Periodic endoscopic dilatations of the stenotic area appear to help, but no control studies have been reported.

Acquired Subglottic Stenosis

The wide variety of mechanisms that potentially produce subglottic stenosis is often surprising to those unfamiliar with this subject. Although it may be caused by chemical or thermal inhalation, benign or malignant neoplasm, or even connective tissue disorders such as lupus erythematosus, there has been a definite increase in the incidence of chronic subglottic stenosis in children caused by blunt external laryngeal trauma. This is partly the result of an increasing number of vehicular accidents in which the extended neck (caused by the acceleration/deceleration events involved in car accidents) hits the dashboard or the front seat, resulting in closed laryngeal fractures. The other major cause of external blunt trauma to the larynx is the "clothesline" injury: a child who is riding a minibike or trailbike can be hit in the anterior neck by a branch or a clothesline, sustaining a laryngeal fracture that may later result in subglottic stenosis. Such injuries may require emergency care but occasionally may present later with progressive upper respiratory obstruction caused by subglottic (and/or laryngeal) stenosis.

Endotracheal Intubation

By far, however, the leading cause of chronic subglottic stenosis in children is prolonged endotracheal intubation. This noninvasive modality, first advocated by McDonald and Stocks in 1965 to provide an airway and as an adjunct to artificial ventilation, is being used with increasing frequency in the management of the critically ill neonate and child. The reported incidence of chronic subglottic stenosis following prolonged endotracheal intubation in neonates and infants ranged from 12 to 20 per cent in the late 1960s and early 1970s, to from 4 to 8.5 per cent at present. Five per cent of neonates intubated for respiratory distress syndrome have been reported to have chronic subglottic stenosis. The actual incidence is probably higher since these data include only patients who had survived their primary illness. All reports also include only patients who demonstrated subglottic stenosis while still hospitalized or shortly thereafter, but fail to report an extended follow-up which might unveil subglottic insufficiency only during a "cold" or following a surgical procedure requiring a short endotracheal intubation. Thus, as neonatal and infant survival is improved in the newly
developed neonatal and pediatric intensive care units, more cases of subglottic stenosis secondary to prolonged endotracheal intubation are expected to be recognized.

The patient may require endotracheal intubation to ensure an adequate airway for weeks and sometimes months, resulting in continuous trauma to the larynx that may ultimately result in chronic subglottic stenosis. Several factors contributing to this trauma have been elucidated: (1) direct trauma to the mucous membrane by traumatic intubation or by repeated intubations; (2) the piston action of the respirator and the abrasive action of the tube against the mucosa during swallowing or in restless patients; (3) indirect trauma to the mucosa and cartilage from the continuous pressure of a large tube or the inflated cuff, causing ischemic necrosis of the mucous membrane and cartilage; (4) superimposed bacterial infection compounding the mechanical trauma; (5) chemical irritation of tubes made from rubber or plastic other than polyvinyl chloride, as well as toxic residues of sterilization with ethylene oxide gas; and (6) prolonged periods of intubation.

Development of Stenosis

The histopathologic sequence in the development of stenosis in neonates and children begins with mucosal edema and hemorrhage, followed by ulceration of the mucosa. With time, infection, or both, the ulceration becomes more extensive, penetrating into the perichondrium and cartilage and causing perichondritis and frank chondritis. Healing may occur by epithelial growth in the early stage with various degrees of subepithelial fibrosis (even without removal of the tube) or by exuberant granulation tissue, which indicates injury to the cricoid cartilage, its nutrient perichondrium, or both. Frank necrosis of the cricoid may then occur in severe cases, resulting in loss of cartilaginous support and collapse of the upper airway. This stage of injury with its proliferative phase may result in "soft" stenosis and a compromised airway, and is usually managed by tracheotomy. The reparative process may resolve following the establishment of an adequate upper airway, or may progress to the stage of "hard" cicatricial stenosis, which is generally but not inevitably circumferential. This end stage of subglottic injury is responsible for the permanent, crippling airway obstruction that requires long-term by-pass with tracheotomy.

The development sequence of subglottic stenosis secondary to endotracheal intubation in human beings has been reconstructed by the examination of a large number of autopsy specimens of neonates, children, and adults. However, it is obvious that although these studies contributed significant insight into the process, they have not precisely defined the role of many variables that affect the development of chronic subglottic stenosis. All factors being equal, the duration of intubation is crucial indeed; however, even the definition of prolonged intubation differs in the opinion of several authors, ranging from 48 hours to six days. Hawkins has shown that the neonatal cricoid cartilage is hypercellular with a scant, gel-like matrix rendering the cartilage more pliable and yielding to pressure. With growth, the matrix increases and becomes less hydrated, more fibrous, more rigid, and considerably more susceptible to pressure-related injury. This is substantiated by the fact that neonates can tolerate intubation for a longer period of time (measured in weeks) compared with older infants and children (measured in days). In spite of these favorable conditions, signs of subglottic injury in the larynges of 15 of 16 neonates who were intubated for six days or longer were found at autopsy.
The role of infection in the development of a stenosing lesion, which is simultaneously a destructive and a reparative process, was recently investigated by Saski et al using dogs. They concluded that tracheotomy-related infection of a mucosal injury of the subglottic lesion can progress to chondritis (and later stenosis) that may be prevented by the administration of systemic antibiotics or by meticulous stomal care. These experimental findings are in agreement with those of Strong and McGovern et al, who reported that subglottic stenosis is twice as common in children when intubation is followed by tracheotomy. The latter studies, although retrospective and uncontrolled, were substantiated and may have far-reaching consequences in the management of pediatric respiratory insufficiency, since tracheotomy is almost always carried out when extubation is difficult or when, using a vague or arbitrary definition of time span, intubation is considered to be too prolonged.

Air hunger is almost a unique feature of subglottic stenosis; it is manifested by low-pitched stridor that is inspiratory but could be biphasic. Although hoarseness is not a symptom of the pure subglottic lesion, a weak voice, caused by the small expiratory volume reaching the vocal chords, may be present.

Although the radiation dosage is relatively high, xeroradiography is the current radiologic technique of choice in the evaluation of chronic airway disease in the young child, as the tissue-air interface is better defined than in tomography. The best technique is the endoscopic evaluation, although it is still deficient in respect to defining the thickness of the stenosis, its extent, and the adequacy of airway diameter.

**Management of Chronic Subglottic Stenosis**

The management of chronic subglottic stenosis secondary to prolonged endotracheal intubation is both controversial and frustrating, as manifested by the wide spectrum of therapeutic procedures proposed in the last decade, all of which have been far from ideal, especially in children. The basic principle in treating this form of airway obstruction is to bypass the obstruction with tracheotomy, then launch a therapeutic attack on the stenotic area. The method of treating stenosis with dilatation has been applied to acquired subglottic stenosis, since the results of this therapeutic modality in congenital subglottic stenosis were encouraging. The application of this modality to acquired stenosis secondary to intubation appears to be costly. Fearon and Cotton reported a 24 per cent mortality rate from causes directly related to tracheotomy and 26 per cent of the children in their series still had a tracheotomy several years later. Similar results have been reported by Hollinger et al, who had the largest series of patients with acquired subglottic stenosis: 11 per cent died and 28 per cent still had a tracheotomy after several years of treatment. A negative impression regarding the value of dilatation is also conveyed by others; however, it appears that in mild cases early recognition of the possibility of developing stenosis (granulations, ulcerations, or difficult extubation) improves the chances that dilatation will be successful.

Since the basic principle in treating chronic subglottic stenosis is to control and reduce fibrosis, systemic administration of steroids has been tried, with varying results: Skolnick believes that steroids are not efficacious, while Hawkins employed them successfully after every dilatation. Intraliesional injection of steroids has been reported by several authors. This method was used in a small number of patients "early" in the disease process with conflicting results; it requires weekly injections under general anesthesia for about three months.
Successful laser excision of granulations, subglottic webs, and severe cicatricial stenosis in a small number of children has recently been reported. It has been our experience in a small number of children that laser excision is useful in thin, web-like, not extensive narrowing of the subglottic stenosis; in severe cases, this treatment modality alone cannot establish an adequate airway. If this method proves successful in a controlled study, it will offer many advantages: (1) precise control, (2) protection of surrounding structures with preservation of anatomy and juncture, (3) preservation of the airway without tracheotomy in some patients, (4) rapid, excellent healing with minimal stricture formation or functional disturbance, (5) the possibility of repeated surgery without trauma and edema, and (6) superior cost effectiveness.

Several other surgical methods have been described for treatment of the stenosis in its cicatricial phase. Prolonged stenting for at least six weeks was successful in six of seven children between the ages of two and nine years, with one death related to the management. All patients in this series appear to have sustained trauma to the larynx secondary to intubation, although this was not specifically stated.

When laryngotracheal cartilages are split with stenting or lumen maintainers, the lower thyroid cartilage as well as several tracheal rings are divided anteriorly; the cricoid is split anteriorly and, sometimes, posteriorly. This allows expansion of the stenotic area (with or without "coring out" the stenosis and with or without grafting) that is maintained by either a stent or by homologous bone (hyoid), cartilage grafts, and composite flaps. These methods, with minor variations, have yielded an airway that is 60 to 90 per cent "adequate" so that decannulation was possible two to four months after the operation. In one series in which decannulation was successful in 70 per cent of patients who underwent an operation, a roughly comparable group of patients was not operated on; however, most reports have lacked a control group. In addition, the studies have been retrospective and have not provided a clear definition of the disease or information (such as extent, thickness, and duration) on the condition of the stenosis.

**Future Goals**

In spite of the voluminous literature on acquired subglottic stenosis, the basic sequential development of this injury and reparative process has not been delineated. Is perichondrium or cartilage injury essential to trigger the cicatricial stenosis, and how important is infection and/or tracheotomy? What is the contribution of the general hemodynamic status of the child to the presumed ischemic necrosis of the cricoid?

Regarding the precise diagnosis of the vertical extent of the stenosis, which is crucial to the treatment and outcome, we still fall short of the optimum. As yet no satisfactory method exists to determine the character of the keystone of the problem, the cricoid ring, except by exposing it through an external surgical approach.

There is an urgent need for a unified method to report pretreatment and treatment outcomes and to develop objective methods for demonstrating the subglottic area and the stenosis. A clinical tool to measure the fundamental status of this area will certainly facilitate this objectivity and will help to define a true subglottic stenosis.
These and many other questions cannot be answered in a clinical setting because of the large number of confounding variables. Thus, the development of an animal model of chronic subglottic stenosis secondary to prolonged intubation should be a top priority.
Vascular Abnormalities

Sang C. Park


Although extrinsic compression of the trachea or a bronchus resulting from an abnormal vascular structure is uncommon, it is an important cause of serious, persistent respiratory problems in infancy. Since the aorta, brachiocephalic vessels, and pulmonary arteries are closely related to the trachea and bronchi, minor aberrations of these vascular structures may produce extrinsic compression of the respiratory tract. A number of different anatomical vascular abnormalities may cause airway compromise. During the past 10 years, 26 patients with various vascular abnormalities has surgical intervention to alleviate airway obstruction at the Children's Hospital of Pittsburgh. Twenty-one were caused by aortic arch anomalies and the remaining 5 were related to pulmonary arterial anomalies (Table 1).

**Table 1. Surgical Experiences with Vascular Abnormalities at the Children's Hospital of Pittsburgh (1969 to 1980)**

<table>
<thead>
<tr>
<th>Type of Anomalies</th>
<th>No of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Double aortic arch</td>
<td>13</td>
</tr>
<tr>
<td>Both arches patent</td>
<td>9</td>
</tr>
<tr>
<td>One arch atretic</td>
<td>4</td>
</tr>
<tr>
<td>Aberrant innominate artery</td>
<td>4</td>
</tr>
<tr>
<td>Right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum</td>
<td>3</td>
</tr>
<tr>
<td>Left aortic arch with right descending aorta and right ligamentum arteriosum</td>
<td>1</td>
</tr>
<tr>
<td>Distal origin of the left pulmonary artery</td>
<td>2</td>
</tr>
<tr>
<td>Tetralogy of Fallot with absent pulmonic valve</td>
<td>3</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>26</strong></td>
</tr>
</tbody>
</table>

**Symptomatology**

The clinical manifestations of these vascular abnormalities may vary considerably depending on the type of lesion and severity of encroachment of trachea, bronchi, or esophagus. The most common presentation is the picture of airway obstruction, mainly stridor, wheezing, or excessive secretion. In the newborn period respiratory symptoms are usually mild and rarely recognized. However, symptoms usually become apparent during the first few months of life. These patients often have a history of recurrent respiratory embarrassment, or infection, requiring frequent medical attention. Some have been referred for evaluation of a possible allergic problem such as asthma. Unless there is severe airway compromise, symptoms usually vary from time to time. During sleep and quiet moments some patients show little sign or respiratory difficulty. However, their symptoms are frequently exacerbated by crying or exertion. Upper respiratory infection also may trigger further compromise of the airway as a result of inflammatory changes in the tracheobronchial lumen.
Although dysphagia with solid food ingestion may be seen in some patients with large and/or tight retroesophageal vascular compression, it is a rather uncommon manifestation in the pediatric age group. Some patients may have excessive oropharyngeal secretions that result in frequent aspiration pneumonia. Despite chronic respiratory difficulties, the majority of patients with vascular abnormalities maintain an adequate weight gain. Only those with frequent pulmonary infection and/or feeding problems may have problems of failure to thrive and poor development.

**Types of Vascular Abnormalities**

Double aortic arch is the most common vascular abnormality requiring operation. In this condition, the trachea and esophagus are completely encircled by the bilateral aortic arches owing to persistence of both right and left embryologic fourth aortic arches. Both aortic arches may be patent, or may be atretic and remaining as a fibrotic cord. In either condition, it usually causes marked tracheal compression with severe respiratory symptoms in early infancy.

In patients with a right aortic arch, an aberrant left subclavian artery and left ductus or ligamentum arteriosum is a rather common abnormality. This often forms a loose vascular ring, and thus a relatively small portion of the patients with this anomaly have significant encroachment upon the trachea or esophagus which requires surgical intervention. In some patients with this anomaly, significant dysphagia may develop when solid food is added. We have also seen a case of rare aortic arch anomaly consisting of a left aortic arch and right descending aorta with a right ligamentum arteriosum which caused tracheal compression.

Pulmonary sling (distal origin of the left pulmonary artery or vascular sling) is a rare but serious congenital anomaly in which the left pulmonary artery arises abnormally from the right pulmonary artery. It then takes a course between the trachea and esophagus, resulting in severe encroachment of the proximal right main bronchus. Patients with this condition usually develop respiratory symptoms early in infancy and manifest persistent wheezing and stridor.

Various congenital cardiac lesions, primarily those with a large left to right shunt and a dilated pulmonary artery, may cause external compression of the trachea and bronchial tree and the resultant symptoms of airway obstruction. Of particular interest, patients with tetralogy of Fallot and absent pulmonary valve usually have a markedly dilated pulmonary artery which frequently causes severe airway obstructive symptoms.

Another important cause of airway obstruction by a vascular structure is an aberrant innominate artery. Some patients in whom origin of the innominate artery is from the aortic arch distal to the midline and posteriorly oriented may have compression along the anterior aspect of the trachea. On occasion, the innominate artery may arise abnormally from the aortic arch as a common trunk with the left carotid artery. This anomaly may also cause anterior compression of the trachea. Although clinical symptomatology is quite variable, the tracheal compression can be severe enough to cause apneic spells and even necrosis of the trachea. According to a large series from the Hospital for Sick Children in Toronto, only a small percentage of patients with this abnormality require surgical intervention and the majority of patients improve spontaneously with age.
Diagnostic Approach

Although there are a number of causes of chronic stridor in infancy and childhood, the possibility of a vascular abnormality causing extrinsic airway compression should be kept in mind in the differential diagnosis. A chest roentgenogram with barium filling the esophagus is the simplest and yet the most informative procedure in the evaluation of patients who have airway symptoms and are suspected of having a vascular abnormality. Since most patients with respiratory problems often have already had chest roentgenograms, careful review of the films may be helpful initially to evaluate tracheal or bronchial abnormalities such as an abnormal shift or constriction. If dysphagia and aspiration problems coexist, a barium esophagram with simultaneous fluoroscopic examination is indicated. However, a simple four view cardiac series is often sufficient to evaluate this condition. The radiographic studies facilitate determining the situs of the aortic arch. An abnormal indentation of the esophagus caused by an abnormal vessel is also readily recognized. Most aortic arch anomalies causing vascular rings have a posterior esophageal indentation. An anterior esophageal indentation is typical of a pulmonary vascular sling but on rare occasions an aberrant subclavian artery may also pass between the trachea and esophagus. Patients with aberrant innominate artery usually have a normal barium esophagram. However, in these patients an anterior indentation of the trachea 1 to 2 cm above the tracheal bifurcation can be seen on the lateral view of chest and neck roentgenograms.

Although an abnormal esophagram frequently provides diagnostic information as to the presence of a possible anatomic derangement, several different anomalies may show similar esophagographic findings. Therefore precise anatomic diagnosis cannot be certain without angiographic studies. Tonki et al have advocated simultaneous angiographic and barium esophagram studies during the catheterization to evaluate the anatomic relation between the blood vessels and the esophagus. We have found that leaving a small radiopaque nastogastric line, such as a Swan Ganz catheter, in the esophagus during the study is helpful to determine spatial relationships since the trachea is located just anterior to this esophageal line. At the termination of the study, the balloon can be inflated with contrast material and slowly withdrawn from the stomach to the oropharynx to evaluate the site and severity of esophageal narrowing. A cine-fluoroscopic record of this catheter withdrawal is made to correlate the site of esophageal narrowing with angiographic information.

Certain vascular structures such as an atretic aortic arch segment or a ligamentum arteriosum cannot be visualized by angiography. If tenting or distortion of the brachiocephalic vessels or a blind aortic diverticulum in the proximal portion of the descending aorta is observed, then an atretic aortic arch and/or ligamentum on the contralateral side of the descending aorta (and thus a possible vascular ring) should be suspected.

Biplane angiography with multiple views using selective injection in the vessel suspected of causing compromise is essential for adequate evaluation of these abnormalities. Although the morbidity of bronchoscopy in the hands of a skilled otolaryngologist is insignificant, this procedure should be reserved for patients with special indications or diagnostic problems since it might exacerbate symptoms in patients with a compromised airway. For instance, bronchoscopy in the patient with an aberrant innominate artery is helpful to demonstrate a discrete pulsatile compression on the anterior aspect of the trachea, 1 to 2
cm above the carina. Tracheobronchographic studies in these patients should be discouraged in view of their associated high morbidity.

Management

Simple existence of a vascular ring is not an indication for surgical intervention. However, when airway compromise from an abnormal vascular structure is significant, surgical intervention should be undertaken without delay. A double aortic arch invariably requires surgery in early infancy, and in these patients the smaller or atretic arch should be divided. In patients with a right aortic arch, aberrant left subclavian artery, and left ductus (or ligamentum) arteriosus, division of the ductus usually relieves airway symptoms. Occasionally division of the aberrant subclavian artery is necessary to alleviate dysphagia. In the case of distal origin of the left pulmonary artery, the left pulmonary artery is divided at its origin from the right pulmonary artery and anastomosed to the main pulmonary artery using cardiopulmonary bypass. An aberrant innominate artery that causes respiratory compromise can usually be suspended against the sternum with a satisfactory postoperative result.

Postoperative recovery in these patients is considerably variable. Although prompt symptomatic improvement may occur shortly after surgery, persistent respiratory symptoms are not unusual in many patients. Some patients may have such significant damage or distortion of the tracheobronchial tree owing to chronic compression by an abnormal vascular structure that severe respiratory symptoms may persist postoperatively. In some of these patients severe associated tracheomalacia may require a tracheal graft with autologous rib in order to alleviate life-threatening respiratory problems.

The prognosis for survival is generally good in most patients with aortic arch anomalies. However some patients, especially those with distal origin of the left pulmonary artery, may succumb before the diagnosis is made. Patients with this anomaly frequently have associated cardiac as well as noncardiac and additional tracheal abnormalities, and as a result their prognosis is poor.

Increased awareness of these vascular abnormalities as a cause of respiratory problems in infants and children will prompt appropriate diagnostic and surgical management and even further improve the morbidity and mortality.
Structure and Function of the Child's Vocal Tract

The pediatric vocal tract is different from its postpubescent counterpart. The upper airway in the child is characterized by its own set of structural parameters. The section briefly reviews the characteristics of the pediatric larynx and pharynx that make it unique, the functional consequences of these, and the brief discussion of the phonation and resonation process.

Larynx and Pharynx

The larynx in the child is not simply a "small" adult larynx. It is basically different in shape and extralaryngeal relationships. The neonatal thyroid cartilage is best described as a gradually curving semicircle, the two laminae meeting at an angle of about 130 degrees. The cricoid cartilage is quasi-oval in shape. The cricoid lamina tilts posteriorly, such that the subglottic airway resembles a funnel. Thus, the diameter of the immediately subglottic airway is less than that of the supraglottic in about 92 per cent of the population. Fishman and Parkley recently reported that about 8 per cent of the infant specimens studied were found to have a tracheal diameter less than that of the cricoid. The subglottic tube extends posteriorly and inferiorly in relation to the larynx, as well. The narrow internal cricoid and the direction that it takes are two points that should be remembered in attempting to intubate a neonate. The most significant difference in the neonatal arytenoid cartilage is that it is far less mobile than that of the adult owing to its grossly different shape.

At birth, the vocal cords are about 3 mm in length. They almost double in length through the first year of life. In the neonate, the cords appear somewhat concave from above because of the extension of the vocal process of the arytenoid cartilage almost halfway into the muscular cord. The bowed condition of the cord and the fact that about half of the cord is rigid, reduce the vibratory capacity of the structure and limit its use as a phonatory organ.

The most basic difference observed in the infant vocal tract is the high position in the neck maintained by the larynx. The cricoid cartilage lies opposite the third cervical vertebra, with the larynx tucked within the hyoid bone, which itself is high in the neck. This allows for the maintenance of a two tube system whereby the infant can breathe during extended periods of sucking. The position of the larynx in the infant makes it an excellent respiratory organ but a poor phonatory one. The reduction in vertical dimensions of the supraglottic pharynx to the level of the oral cavity makes this area a poor resonating cavity and prevents the cephalad laryngeal excursion necessary for the phonation of high pitched sounds.

Oral Cavity

The second significant difference in the infant's vocal tract is the relationship between the hard palate and the cranial base. The neonate's osseous palate is located in a more superior
position than that of the adult. Thus, the velum, too, is found more cephalad. At this stage, the levator veli palatini runs laterally rather than superolaterally, acting as a tensor rather than as an elevator of the palate. The position of the velum and the orientation of the extrinsic muscular elevator limit the capacity of the velum to function effectively in its role in phonation. The tongue is also limited in its ability to function as an articulator owing to its structure and position in the neonate. The tongue is located entirely within the oral cavity at this early stage, having no pharyngeal portion. In addition, it is short anteriorly-posteriorly and wide laterally, making it a clumsy organ for articulation. Also, in that the tongue fills the oral cavity in the infant, the capacity of the cavity to act as a resonator is severely reduced.

Thus, the structure of the vocal tract in the infant is more suited to respiration than phonation. However, the system is certainly capable of producing voice, grossly no doubt, but sound nevertheless.

Function of the Larynx in Phonation

In addition to being at a less than mature stage of development, the phonatory mechanism of the child is not subject to as fine motor control as that of the adult. Thus, the voice changes one may hear in a child are often more gross and more dramatic than those heard in the adult. Important to the production of voice is the ability to maintain a threshold subglottal pressure that is relatively constant during an utterance. This ability is one that improves with physical growth and language development. The production of sound is explained by the interaction of the elastic properties of contracted muscle and aerodynamic forces (see Borden and Harris for a more detailed review). The sound produced at the level of the vocal cords (the fundamental frequency) is subsequently modified by the pharynx at all levels (resonation) and finally altered by the shape of the oral cavity (articulation) to produce speech sounds.

Pitch changes are accomplished by shifts in the length, thickness, mass, and isometric tension of the vocal cords. Increased length and decreased thickness, hence decreased mass and increased isometric tension, within the body of the cords will result in the production of a higher pitch. A sudden increase in expiratory force, subglottal pressure, can also lead to the production of a high pitch voice, usually as a pitch break. Discrete pitch change is a learned behavior. Thus, the infant does not have the exacting motor control at this early stage to produce the wide range of pitches observed in the adult.

Intensity and pitch changes often interact in the untrained voice, certainly that which describes the young child. Intensity change by itself if accomplished by increasing subglottal pressure, resulting in a greater sinusoidal displacement of the vocal cords. Ability to produce a loud voice independent of pitch shifts will also improve with increased motor control.

Thus, although the pediatric larynx is structurally different from its adult counterpart, the organ works in a similar fashion to that of the adult. With improving competence in motor control and changes in structure with development, the child begins to approximate the variety of laryngeal postures necessary for human communication of thought and feeling.
Common Pediatric Voice Disorders

Deviant voices are generally tolerated and frequently prized in our culture. The freckle-faced, raspy-voiced kid in the television commercial warms our hearts, and the tough-guy with a voice that speaks of cigarettes and whiskey at three o'clock in the morning readily becomes a movie folk hero. Thus, the clinician is hard-pressed to determine which deviant voices require treatment. The physician may base this determination on pathologic changes in the vocal cords; the speech-language pathologist may have to rely on less objective determinants. A deviant voice may be considered in need of treatment if it meets one of the following three criteria: calls attention to itself, interferes with communication, or causes the speaker to be unhappy.

Incidence reports of voice disorders in children range from below 1 per cent to more than 20 per cent of the population. The majority of surveys indicate that approximately 6 to 9 per cent of children in the USA demonstrate a voice disorder. Of the many voice disorders which have been described, those most likely to appear in a pediatric practice fall into three basic categories: disorders of vocal quality, disorders of pitch, and disorders of resonance. This system of categorization is arbitrary and by no means exclusive. Quite frequently the disorders overlap. The production and the perception of voice are multidimensional phenomena, and it is unlikely that a voice will be perceived to be defective along only one dimension.

The most common pediatric disorder of voice quality is that of hoarseness. This quality is sometimes referred to as roughness or huskiness, and it usually implies a voice which is low pitched, strained, and grating. It may also contain aspects of harshness, such as raspiness and a crackling quality. Pitch range may be narrow. Breathiness may also be present, together with inappropriate variations in audibility. When no organic disorder is present, this problem is essentially caused by excessive muscle tension, sometimes referred to as vocal hyperfunction, vocal abuse, or vocal misuse. This type of voice is commonly heard with laryngitis, and may frequently occur after periods of excessive vocal use such as vociferous participation in sporting events. It is probably only worthy of concern if it persists beyond a few weeks, or if it occurs frequently. Hoarse patients tend to be intense people who may present with behavior problems.

Voice pitch may be considered deviant if it is higher or lower than is expected in the culture for the child's age and sex. Other problems of pitch include excessive pitch breaks and very narrow pitch ranges. Disorders of pitch may reflect organic disorders, or they may result from vocal misuse and/or personality problems.

Disorders of resonance typically refer to problems of hypernasality or hyponasality. Some nasal resonance may be found in all speech. In the English language, the m, n, and ng are nasalized. One frequently hears slightly nasalized production of vowels that precede or follow those three consonant sounds; if this is not offensive, it may be considered normal. When inappropriate or excessive nasal resonance becomes objectionable, a problem of resonance is present. Hypernasality may also be present on other consonants such as s, k, z, and g, which may be nasally emitted with a characteristic snort. Such problems are frequently associated with velopharyngeal dysfunction, though they may be primarily a result of defective functional articulation. Hyponasality is a lack of nasal resonance on the m, n, and
ng sounds. The patient may sound as if he has a severe cold, and may appear to be substituting b, d, and g for the nasal consonants. Hyponasality frequently is the result of an obstruction in the nasal pathway.

In evaluating a child with a voice disorder, the pediatrician should consider the possibility that the deviant voice is an index symptom suggesting a broader physical and/or emotional problem. Changes in physical and emotional states should be explored, along with a history of past and present stresses. Careful monitoring of the voice symptom over time should be undertaken. Referral for laryngological evaluation and consultation with a certified speech-language pathologist should be considered if the problem persists for more than a few weeks.

**Evaluation by Speech-Language Pathologist**

The speech-language pathologist is likely to engage in an evaluation of a pediatric voice disorder only in conjunction with an evaluation by an otolaryngologist. The speech-language pathologist brings to this evaluation unique and complex skills that require intensive training on the graduate level. Although the evaluation will vary in relation to the age of the patient, the chief complaint, and the results of the laryngological investigation, certain basic elements are likely to pertain. An evaluation generally involves a thorough history taking, a series of observations, and a series of tests.

The history will likely include general information regarding the patient and the patient's family, a detailed history of the complaint, a supplementary medical history, a history of the patient's voice usage, and possibly a psychosocial history. Information gathered in such a history will be used not only in the formulation of a concept of etiology, but also in planning for a treatment procedure that is tailored to the individual and the family.

The speech-language pathologist will observe the patient in both structured and unstructured situations. It will be important to determine the patient's effectiveness as a communicator, his habitual vocal usage, his strategies for dealing with his voice problem, and other vocal habits that may affect voice, such as excessive talking, excessive shouting, coughing and throat clearing.

The formal tests available to the speech-language pathologist in the evaluation of voice are numerous and varied. The evaluator is likely to develop a voice profile for the patient, and may include specific evaluation of pitch and loudness, quality, phonation time, breathing, resonance, articulation, vocal habits, and others. In addition, the speech-language pathologist is likely to attempt some brief therapy or "stimulability examination" in order to obtain some indication as to how the patient may respond to therapeutic approaches.

Results of the evaluation will be considered in conjunction with the findings of other professionals who may be involved with the patient. Recommendations may be made for voice therapy, or for voice therapy in coordination with other treatment, on the basis of the results of all evaluations and the statements of the child and family regarding their motivation and attitudes toward treatment.
Treatment

Voice therapy is usually considered the treatment of choice under the following circumstances: to determine if a laryngeal disorder can be alleviated through voice therapy; for adaptation to congenital or acquired anomalies; and for nonorganic cases. Laryngeal problems that are likely to respond favorably to voice therapy include vocal nodules, vocal fold thickening, polypoid conditions, and chronic nonspecific laryngitis. This is especially true in patients in whom vocal misuse or abuse is felt to be the cause of the problem. Surgical removal of vocal nodules is usually postponed until the patient has had a significant trial with voice therapy. A trial period of voice therapy cannot be expected to produce significant results unless that period is longer than three months, with sessions taking place at least once a week. The use of complete vocal rest is controversial; it is a rare child who can tolerate complete vocal rest for as long as 14 days. The stress that may be created for a child and his family when any length of vocal rest is prescribed may in itself contraindicate such a procedure. Whispering is never helpful.

The treatment of voice disorders by the speech-language pathologist will vary greatly as a result of the training of the professional, the philosophical basis of the therapeutic approach, the age and disorder of the patient, the time available for the treatment, and the like. The therapy is likely to involve an approach to helping the young patient understand the nature of the voice disorder, the importance of voice improvement, and the nature of the therapeutic steps that must be taken by the child. The therapist is likely to engage both the child and the family in environmental manipulation, which may include reduction of shouting in the home, reduction of shouting at play, appropriate amounts of rest, and attention to other aspects of vocal hygiene.

A significant portion of the therapy will be devoted to symptom management. The patient is likely to be taught to discriminate between appropriate and inappropriate voice production, to produce voice correctly, and to habituate the new and more desirable vocal patterns. Space does not permit a discussion of the multitudinous methods by which these objectives may be obtained. The therapist must be knowledgeable in the areas of anatomy and physiology of voice production, techniques of therapy, and above all, human nature.

Personality factors are frequently believed to play a significant role in the development of vocal disorders. Therefore, vocal psychotherapy is often undertaken as an integral part of the management of voice disorders. This aspect of the treatment may be undertaken by a specifically trained speech-language pathologist, or in conjunction with a psychiatrist or psychologist. In many cases, it is the quality of the vocal psychotherapy which will determine the outcome of the treatment.

It is important that the physician and the speech-language pathologist engage in periodic objective reassessment of the patient's condition. In the early stages of treatment, it is wise to undertake reassessment at three months intervals. Decisions regarding termination should be made in a collaborative manner, and periodic reevaluation should be done by all members of the voice team. An approach to the treatment of voice disorders which combines the skills of the physician and the speech-language pathologist should serve to produce optimal results for the pediatric patient.
Infections of the Head and Neck

Kenneth E. Shuit, Jonas T. Johnson


Cellulitis and abscess of the potential spaces of the neck, deep to the superficial fascia, have become known as deep neck infections. These infections merit special consideration because of the life-threatening complications that may result if treatment is delayed or inadequate. In the preantibiotic era we have been led to believe that deep neck infections were frequently encountered. More recently, deep neck infections have been less common and knowledge about them is generally obtained from textbooks or anecdotal accounts. Deep neck infections continue to be important, albeit relatively unusual, problems. Furthermore, these clinical situations offer circumstances in medicine in which astute judgment and timely intervention continue to be life-saving.

Anatomic Consideration

Connective tissue envelopes all the viscera of the neck and, in fact, a fascial plane simply represents the condensation of such connective tissue. Infection and abscess formation may cause pressure, and the limits of these infections are defined by the fascial planes of greatest resistance.

A layer of fascia envelops the spinal column and paraspinal muscles. This fascia attaches to the transverse process of the vertebrae laterally, then splits into a prevertebral and alar layer. This forms a potential space known as the prevertebral space. The prevertebral space runs from the base of the skull to the diaphragm and, under normal circumstances, it is empty. Most reported infections of the prevertebral space come from direct extension of osteomyelitis of the vertebrae. Immediately anterior to the prevertebral space, and behind the pharyngeal wall, lies the retropharyngeal space. This space extends from the base of the skull to approximately the level of the tracheal bifurcation. It is for this potential connection with the mediastinum that it is called the "danger space". Lymphatic glands of the retropharyngeal space drain primarily the nasopharynx, sinuses, and posterior nose.

The parapharyngeal space is adjacent to the retropharyngeal space. This cone shaped space is bounded by the skull superiorly and the hyoid bone inferiorly. The lateral border is the mandible and parotid gland, while medially, it is bounded by the pharyngeal fascia. It is because of the relatively nondistensible superior, posterior, and lateral boundaries of the parapharyngeal space that abscess formation often first produces bulging into the pharynx.

The parapharyngeal space must be distinguished from the site of the peritonsillar abscess which lies medial to the pharyngeal fascia immediately under the capsule of the tonsil. The parapharyngeal space is traversed by the carotid artery, jugular vein, sympathetic chain, as well as cranial nerves IX, X, XI, and XII. The carotid sheath pierces the parapharyngeal space at the hyoid bone before continuing into the mediastinum. The carotid...
sheath thus has been termed the "Lincoln highway" of the body. The parapharyngeal space contains lymph nodes that drain primarily the oral cavity and oropharynx.

More anteriorly lies the submandibular space between the skin and the floor of the mouth. This space is divided into a sublingual and submental portion by the mylohyoid muscle. There is relatively free communication between the two portions of the space along the posterior edge of the mylohyoid muscle. However, the primary area of presentation may vary depending on abscess pressure. That is, if the mass is primarily sublingual there may be edema and retraction of the tongue, with eventual airway compromise (Ludwig's angina), whereas a submental (inframylohyoid) infection may present primarily as a mass in the neck.

Pathogenesis

Deep infections of the head and neck such as those discussed in this article frequently are related to various types of trauma or focal infection sustained to the oral cavity or its contents. For example, Taffel, in a survey of 45 cases of Ludwig's angina, reported that only 3 (7 per cent) appeared to have no antecedent cause, whereas the remainder (93 per cent) were preceded by diverse events such as recent lower molar extraction (42 per cent), toothache and associated apical abscess (27 per cent), trauma to the mouth floor (4 per cent), previous submaxillary adenitis (4 per cent), compound fractures of the mandible (4 per cent), and sore throat (9 per cent). The role of dental infections or procedures in the development of Ludwig's angina seems important, especially conditions involving the second and third molars, possibly associated with the position of the root apices and the relatively thin cortex of the mandible in this region, both giving dental infections ready access into the submaxillary space.

Similarly, the pathogenesis of retropharyngeal abscesses is most likely associated with extension of infection from the pharynx to lymph nodes in this anatomical space which are numerous and large in young children and usually atrophy prior to adulthood. Importantly, infection of the adjacent fascial plane, the prevertebral space, however, is often associated with osteomyelitis of the cervical vertebrae with pyogenic bacteria or mycobacteria rather than with suppurative processes of the nasopharynx.

Finally, peritonsillar abscesses are most frequently associated with preceding pharyngotonsillitis. Other factors that encourage the development of suppuration in some patients and limit the infection to the tonsil itself in others are unknown.

The microbiology of infections of the fascial spaces of the face and neck reflect the endogenous flora of the mouth. The mouth is not a uniform cavity with respect to its microbial contents and can be affected by a variety of factors including age, diet, nutritional status, dentition, the presence of periodontal disease, and the use of antimicrobial therapy. Despite the differences that these factors can bring about in the oral flora, it is important to emphasize that anaerobic bacteria play a large and important part in the bacteriology of the oral cavity. Several authors, for example, have determined that there are approximately $2.7 \times 10^{11}$ microorganisms per gram of debris found in the gingival crevice and that approximately 90 per cent are anaerobic bacteria (Bacteroides, Peptostreptococcus, Veillonella,
Fusobacterium). Of importance in relating normal mouth flora to the pathogenesis of deep neck infections is the observation that these mouth organisms are found transiently in the blood after tooth extractions and in the infections described in this article.

**Clinical Presentation**

Although different anatomic spaces are involved in these infections, the presenting symptoms are frequently similar. Of great importance is the possibility of rapidly developing respiratory compromise.

**Ludwig's Angina**

This infection of the submandibular space most often begins as a cellulitis which causes edema, induration, tenderness, and elevation of the floor of the mouth. The infection may progress to edema of the tongue but principally is characterized by a brawny, tense swelling that is maximum in the region of the submaxillary gland (bull-neck). The process may be unilateral or involve both sides of the neck. Development of fluctuance is rare and the lymph nodes and salivary glands usually are not involved. The infection may progress to the point that speech is difficult or impossible. Drooling because of pain on swallowing is common. Respiratory distress is a late but most serious sequela and is characterized by stridor and in some cases by cyanosis.

Diagnosis of this infection may be difficult because of its rarity and because of its confusion with other infections such as localized abscesses of the salivary glands or cervical adenitis. The importance of accurate and rapid diagnosis of this infection cannot be overemphasized because of the potential for serious complications. The absence of preceding mouth trauma does not eliminate the possibility of the development of Ludwig's angina.

**Peritonsillar and Parapharyngeal Abscesses**

Focal infections of this region usually begin with unremarkable pharyngitis that progresses despite antibiotic treatment. The most striking feature of peritonsillar abscess is a unilateral medial displacement of the affected tonsil; this feature is often less obvious in the case of parapharyngeal abscesses. In both infections, however, examination of the pharynx may be difficult because of trismus, but adequate physical examination reveals a toxic appearing patient who may be drooling because of the extreme pain associated with these abscesses. The patient's voice may have a quality that, because of the mass effect, is different from the hoarseness associated with vocal cord lesions.

It can be difficult to differentiate between a peritonsillar and parapharyngeal abscess. In both cases, the patient will have medial displacement of the tonsil; however, with a parapharyngeal mass, the critical observer will note that the whole lateral pharyngeal wall is involved, not just the tonsil. Trismus, dysphagia, and drooling are common to both conditions. The key distinction is involvement of the neck in parapharyngeal abscess. This is reflected in diffuse brawny induration often with spasm of the muscles causing stiff neck. Fluctuance is
unusual because of the overlying heavy muscles (sternocleidomastoid). This is to be
distinguished from multiple tender but discrete nodes often palpable in peritonsillar abscess.

<table>
<thead>
<tr>
<th>Peritonsillar</th>
<th>Parapharyngeal</th>
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<tbody>
<tr>
<td>Tonsil pushed medially</td>
<td>Pharyngeal wall pushed medially</td>
</tr>
<tr>
<td>Discrete cervical nodes</td>
<td>Diffuse brawny edema</td>
</tr>
<tr>
<td>No stiff neck</td>
<td>Torticollis.</td>
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**Retropharyngeal and Prevertebral Abscesses**

Although the pathogenesis of these infective processes may be different, their clinical
presentation is similar, with posterior pharyngeal pain and dysphagia that may lead to
drooling. Irritation of the paravertebral ligaments may produce meningismus as well. On
physical examination, a definite pharyngeal mass may be appreciated. Lateral roentgenograms
of the neck will show an increase in the space between the anterior wall of the cervical spine
and the pharyngeal wall. This interval has been measured for children of different ages and
actual calculation may be helpful in doubtful cases.

**Treatment**

All patients with deep neck infections must be hospitalized for observation and
treatment. Basic considerations such as rehydration by intravenous fluids and fever control
can be met. Blood and the suspected sites of infections should be cultured and high dose
intravenous antibiotics should be started. Drugs suitable for the suspected organisms should
be empirically begun because of the seriousness of these infections, which dictates against
withholding drugs until cultures are available. The adequacy of airway must be ascertained
and followed closely. The next consideration in management is the stage of the infection. The
infection may simply be cellulitis initially, and, as such, may respond entirely to management
with fluids and antibiotics.

Although most of the microorganisms that cause these infections are sensitive to
penicillin (streptococci and anaerobes), deep neck infections associated with staphylococci and
gram-negative bacilli have also been reported, causing some authors to suggest broader
coverage than penicillin alone until culture identification and sensitivity testing have been
completed. Thus, alternatives such as the combination of ampicillin and a semisynthetic
penicillin or one of the newer cephalosporins may be a rational initial choice. In all cases, the
drug should be administered intravenously and in large doses until the focal signs of infection
have largely disappeared. Reportedly, 10 to 15 per cent of patients will recover on antibiotics
alone.

If the infection progresses to abscess, it must be recognized so that drainage may be
instituted. The failure to improve objectively over a 24 hour period is an important indication
for drainage. Retropharyngeal and parapharyngeal abscesses do not ordinarily develop
fluctuance, because of their anatomic position. Fluctuance should not be expected even with a
severe abscess.
In every case in which drainage is necessary, the basic tenets of surgical management must be respected, namely, adequate visualization and protection of vital structures, airway protection, and control of hemorrhage. Most healthy, young adults can tolerate incision and drainage of a peritonsillar abscess under local anesthesia while sitting up. Retropharyngeal abscess may often be effectively drained intraorally; however, airway management by either intubation or less often, a tracheostomy, is necessary. The complex anatomy of the parapharyngeal space requires general, endotracheal anesthesia and an external approach. Ludwig's angina and submental abscesses may be drained either intraorally or externally depending upon the relationship to the mylohyoid muscle.

Complications

Should cellulitis progress to abscess there may be increasing pain, corresponding to increasing abscess pressure. This in turn may cause the abscess to "point" either into the pharynx or to track along one of the previously mentioned anatomic planes into other anatomic areas such as the mediastinum. It is to avoid these problems and to obtain controlled drainage and resolution of infection that surgery is necessary. The presence of suppurative in and about the carotid sheath may also result in septic thrombophlebitis or the internal jugular vein or erosion of the carotid artery system. Paralysis of any of the adjacent major nerve trunks occurs on occasion as well. Although these complications are unusual, it behooves the physician managing a patient with a deep neck infection to be familiar with and observe for them.

These complications, should they ensue, require special techniques such as mediastinal drainage, ligation and excision of the thrombosed jugular vein, or ligation of the eroded carotid artery, all of which are beyond the scope of this presentation.

Conclusion

Cellulitis and abscess of the deep structures of the neck, although rare, are important clinical entities. Effective treatment requires early diagnosis and the institution of appropriate therapy. Under these circumstances, many cases of cellulitis may be aborted prior to abscess formation. The failure to improve or the progression of symptoms on therapy is an indication for surgical intervention in order to prevent the potentially dire effects of a complicated deep neck infection.
Genetics of Congenital Deafness

Mark W. Steele


From 10 to 26 per cent of cases with hearing loss are known to be congenital, that is, present at birth or by early childhood; approximately 52 per cent of these are genetically caused. Although these include rare instances of conductive hearing loss consequent to osseous malformations of the middle ear (such as in branchial arch syndrome), the term congenital deafness in this article refers only to profound, sensorineural hearing loss that is irreversible and bilateral. Estimates of the incidence of genetic congenital deafness in Great Britain, Japan, Germany, and the USA are 1 in 2000 to 1 in 6000 live births.

Some General Principles of Genetics

A specific gene can be inherited from the previous generation or can arise de novo in an individual owing to spontaneous mutation (in the latter, no prior family history of the gene would be likely). Such new mutations can be passed on to the next generation. Specific genes, then, may not always be inherited, but they are heritable. The genetic makeup of an individual is the genotype, the effect of which is the phenotype. The genes are distributed among 23 pairs of chromosomes. One pair of chromosomes, the X and Y, contain some genes determining primary sex differentiation. Females have 2 X chromosomes; males have one X and one Y chromosome. The remaining 22 pairs of chromosomes are the autosomes. Since chromosomes exist in pairs, all genes exist in pairs except those on the X and Y chromosome in males. The X and Y chromosomes are not similar (homologous) genetically or morphologically. A particular gene at each gene locus can exist in several slightly different molecular forms called alleles. Respecting any one pair of genes, the alleles can be identical (homozygous) or not (heterozygous). By the reproductive process (meiosis), one member of each gene pair is passed on from each parent to its offspring.

Autosomal Inheritance

Consider a single pair of genes on an autosome where there are two possible alleles: A and a. Then, respecting this gene pair, an individual can have one of three possible genotypes: AA, Aa, or aa. Suppose that the phenotype AA and Aa are both designated "A" (that is, identical) while the phenotype of aa (designated "a") is different. Then, the effect of allele A dominates that of allele a (the latter is said to be recessive, that is, its phenotype is seen only when homozygous). Respecting such a "simple mendelian trait", there are six possible matings in the population.

First, assume that the dominant allele, A, causes congenital deafness and that the recessive allele, a, results in normal hearing. Then all the offspring of matings type 1, 2, and 3 would be deaf. However, in human beings one rarely finds homozygotes for abnormal autosomal dominant genes (AA in this illustration). Probably such genotypes are lethal to the fetus in utero. Consequently, respecting autosomal dominant deafness, one can assume with confidence that the genotype is heterozygous (Aa), and that the only possible mating types are 4, 5, and 6. Deaf offspring would result only from mating types 4 and 5. The relative
frequency of the three mating types will depend on the frequencies of alleles A and a in the population and the randomness of the matings. For deafness, mating is not random, since about 85 to 90 per cent of congenitally deaf individuals have a congenitally deaf mate. Consequently, mating type 4 would be over-represented compared to a randomly mating population. However, the observed ratio of the phenotype "A" (deaf) to "a" (normal hearing) in offspring from mating type 4 would be 2:1 rather than 3:1 since AA fetuses would not be born. The risk of deaf offspring for mating type 5 would be 50 per cent at each conception.

Next, consider the reverse situation in which the dominant allele, A, causes normal hearing while the recessive allele, a, causes congenital deafness. In this situation, all six mating types are possible but deaf offspring would result only from mating types 4, 5, and 6. Mating type 6 would produce only deaf offspring while the risk of deaf offspring at each conception from mating types 4 and 5 would be 25 per cent and 50 per cent respectively. However, again the frequency of mating type 6 would be over-represented in the population since the congenitally deaf tend to marry each other.

**X-linked Inheritance**

Since genes on the X chromosome have no mate on the Y chromosome, all X-linked genes act as dominants in the male, that is, they express themselves in the phenotype. In the XX female, however, X-linked genes are all paired just as autosomal genes are all paired. Consequently, only in the female can X-linked genes act as dominants or recessives. The hallmark of X-linked inheritance is that a male cannot pass his X-linked genes on to his sons since a male gives only his Y chromosome to his sons. Conversely, a male passes all his X-linked genes on to his daughters to whom he gives his one X chromosome.

Consider an X-linked gene with two possible alleles, D and d, where D is dominant and produces normal hearing and d is recessive and produces congenital deafness (remember, the alternatives dominant and recessive here apply only to the female). Note that none of these three mating types produces deaf daughters and only mating type 3 would produce deaf (X⁺Y) sons (50 per cent risk at each male conception).

**More Complex Matings**

The genetic etiology of congenital deafness is quite heterogeneous. Five or six distinct autosomal recessive traits (such as Pendred and Usher syndromes) account for about 3 per cent of congenital deafness and probably many more rare traits, a few of which may be found in any particular deaf population. Each deafness trait presumably results from homozygosity of an abnormal recessive allele at a different specific gene locus. Likewise, there are several distinct X-linked recessive and autosomal (and possibly X-linked) dominant genes that can result in congenital deafness. Any one individual, then, can bear genes for deafness at several different gene loci which can result in complex mating types. Numerous other complex matings are possible. Of the three examples, type 1 is the most common. This is fortunate since the mating does not result in deaf offspring. Mating types 2 and 3 are more rare but are frequent enough to be considered in genetic counseling, particularly if there is consanguinity. Matings type 2 and 3 illustrate "pseudodominance", that is, although each deafness results from homozygosity of a specific autosomal recessive gene, the risk for deaf offspring from the matings is 50 per cent at each conception - the same risk as for an autosomal dominant
gene for deafness (the risk of deafness is 50 per cent whether caused by the recessive allele, a, or the dominant allele, A).

Genetic Syndromes With Congenital Deafness

Fraser found about 50 per cent of cases of congenital deafness to be genetically determined. Autosomal recessive inheritance was the most common (33 per cent), autosomal dominant inheritance was next (15 per cent), and X-linked recessive inheritance was rarest (about 2 per cent). Among Fraser's autosomal recessive cases, the single most common diagnosis was "clinically undifferentiated" (26 per cent); next were Pendred syndrome, deafness with goiter (5 per cent); Usher syndrome, deafness with retinitis pigmentosa (1.2 per cent); and cardioauditory syndrome of Jervell and Lange-Nielsen (0.5 per cent). The remaining 0.3 per cent of autosomal recessive cases consisted of congenital deafness as part of various rare syndromes of congenital abnormalities.

Among the autosomal dominant cases, again the single most common diagnosis was "clinically undifferentiated" (11 per cent). Auditory pigmentary syndromes, such as Waardenburg syndrome, were next most common (4 per cent). Rare entities, such as Alport Syndrome (deafness with nephritis) or deafness with earpits or with optic atrophy, comprised less than 1 per cent of the cases.

All of the 2 per cent of X-linked recessive cases of congenital deafness were diagnosed as clinically undifferentiated. However, congenital deafness is associated with well known X-linked recessive syndromes such as cutaneous albinism. About 0.6 per cent of the congenitally deaf in Fraser's survey (1.2 per cent of the girls) were diagnosed as Wildervanck's syndrome (congenital deafness with abducs palsy, retraction of eyeball, and Klippel-Feil deformity) and female. The genetic etiology of this condition is probably heterogeneous.

Genetic Counseling

Genetic counseling for congenital deafness must be a team effort including, among others, primary care physicians, otolaryngologists, audiologists, and geneticists. Precise genetic counseling is facilitated most by a specific diagnosis and a detailed family history; both often depend most on the efforts of others prior to the formal genetic consultation. For example, if the geneticist sees a deaf couple who both have Usher syndrome, the couple can be counseled with confidence that there is a 100 per cent risk for deaf children. On the other hand, if the diagnosis for one deaf spouse is Usher syndrome and for the other is Pendred syndrome, the couple can be advised with confidence that their risk for a congenitally deaf child is about the same as that of the general population, less than 1 in 1000. Unfortunately, for about 40 per cent of individuals the diagnosis is congenital deafness, clinically undifferentiated, genetically determined (the latter often being decided only by exclusion). Approaches to genetic counseling for these individuals can be illustrated by looking at three different types of matings: when congenital deafness is found in both spouses, only one spouse, or neither spouse.
Both Spouses Congenitally Deaf

This is the most common situation since approximately 85 per cent of congenitally deaf persons marry each other. The risk of these couples for congenitally deaf children ranges from that of the general population to 100 per cent. This range can be narrowed if there are offspring whose hearing status is known; but aside from eliminating the extremes, the range of the risk may still remain broad in any one particular case. Fraser estimates that the chance of both spouses being autosomal recessive homozygotes at the same gene locus (mating type 6 with 100 per cent risk for deaf children) is less than 2 to 3 per cent. However, the chance of this increases significantly if the spouses are consanguineous or if both come from a small isolated population which harbors a particular form of autosomal recessive deafness (such as the Jews of Britain).

Even when the deafness is clinically undifferentiated, the family pedigree may provide sufficient information for precise genetic counseling. Evidence for autosomal dominant or X-linked recessive inheritance of the deafness should be searched for carefully. Unfortunately, lack of such evidence does not completely eliminate these possibilities since one deaf partner or the other could represent a new mutation for deafness. Fraser found among clinically undifferentiated deaf persons, new mutants in 10/22 X-linked recessive cases and 127/143 autosomal dominant cases. Another complication is that dominant genes do not always express themselves when present (penetrate), that is, actually produce their phenotype in the heterozygote. The average penetrance of dominant genes for clinically undifferentiated congenital deafness is about 67 per cent; that is, if an individual has the gene, the chance of that individual being congenitally deaf is only 67 per cent, not 100 per cent. Another complication in pedigree analysis is the phenomenon of "pseudodominance".

It should be clear that genetic counseling of a clinically undifferentiated deaf couple often cannot be precise in regard to risk, and the couple should understand that reality. When precise risk calculations are not feasible, empirical risks can be helpful. According to Fraser, the average empirical risk for a clinically undifferentiated congenitally deaf, nonconsanguineous couple having a congenitally deaf child is 10 per cent.

One Spouse Congenitally Deaf

In this less common situation, the risk for a congenitally deaf child ranges from that of the general population to 50 per cent. If the family history is not informative and the deafness of the spouse is clinically undifferentiated, the genetic counseling again is difficult and imprecise. If the deafness is autosomal dominant, the couple's risk for a deaf child still may be less than 50 per cent owing to low penetrance. If the deafness is autosomal recessive, the risk still may be more than that of the general population because of pseudodominance (mating type 3). However, in the absence of consanguinity the incidence of pseudodominance in such matings is only about 1 per cent. If the husband is the deaf spouse, X-linked recessive inheritance must be considered as a cause, although it is less likely than autosomal dominance or recessiveness. If X-linked recessive, however, the increased risk for deaf children would not be for the couple but rather for their daughter's sons (mating 2 and 3). All in all, this couple's empirical risk for a deaf child is about 5 per cent. Their recurrent risk (the risk subsequent to the birth of a deaf child), however, would approach 50 per cent at each conception depending on penetrance.
Neither Spouse Congenitally Deaf

In most instances, these couples seek genetic counseling after the birth of a congenitally deaf child. If the congenital deafness is clinically undifferentiated and there is no informative family history, the risk of recurrence ranges from that of the general population to about 25 per cent. The lower extreme would apply if the child's deafness resulted from a new dominant mutation. The upper extreme would represent recessive inheritance (mating type 4 or, if the deaf child is a male, mating type 3). Unless the parents are consanguineous, the mean empirical recurrent risk is 10 per cent. This empirical recurrent risk is inversely proportional to the birth order of the first deaf child. If the first deaf child is the first born, the recurrent risk is 12.5 per cent; if the second born, it is 10 per cent; if the third born, it is 7.5 per cent; and if the fourth born or higher, the recurrent risk is 5 per cent. After the birth of the second deaf child, the recurrent risk would approach about 25 per cent at each conception.
Audiologic Aspects of Learning and Behavior

Craig B. Liden


Over the past decade, issues of child development and behavior have moved into a prominent position in the mainstream of general pediatric care. This has been characterized by an expanded definition of health to include not only "quantity of life" but also "quality of life" for the child. Efficient learning and appropriate behavior have become significant measures of outcome of high quality primary health care for the child. Contemporary societal demands have substantially contributed to this trend. Most prominent in this regard has been the passage of Public Law 94-142, the Education for All Handicapped Children Act. This law mandates a role for the physician in the identification, assessment, and development of individualized educational programs for all handicapped children. National figures suggest that 10 to 20 per cent of all children belong to this category (for example, those who are learning disabled, emotionally disturbed, mentally retarded, physically handicapped, or sensory impaired). As a consequence of increased awareness of the interrelationship of educational and health factors, educational professionals and parents have increasingly turned to pediatricians, general practitioners, and otolaryngologists among others for consultation when problems arise in a child's learning or behavior. This frequently takes the form of asking the physician to provide a simple etiology, such as hearing loss, to explain the problem.

For the most part, physicians have been poorly prepared to meet this demand. The absence of a unifying theory of development and behavior and the lack of a universally understood and accepted taxonomy of problems has placed several methodologic contraints on research and clinical activity in this area. Attempts to rigidly apply the traditional "medical model", with its focus on defining etiology, to issues of learning and behavioral failure has generally been unrewarding. Furthermore, by its very nature development (and thereby learning) implies change over time. Similarly, behavior varies depending on the context in which it occurs. Therefore, by definition, a certain degree of ambiguity is a part of all learning and behavioural problems. This inherent uncertainty is magnified when data are reconstructed retrospectively as is often the case when assessing a learning or behavioura problem. Failure to acknowledge these factors when assessing and managing children with learning and behavioral problems may result in applying simplistic solutions to complex problems.

These issues have been clearly demonstrated with respect to audiologic factors and their relationship to learning and behavioral function. Recent studies have suggested that chronic or recurrent otitis media during early life is associated with impairments of language or cognitive development or with poor academic achievement. Others have inferred that this relationship has a cause and effect nature. A critical review of the literature reveals that all reported studies suffer from major limitations in design or method. Despite these weaknesses, some professionals have called for mass screening programs to detect middle ear effusion or have advocated aggressive treatment measures upon the recognition of the presence of middle ear effusion.
In order for physicians to define an appropriate role in responding to specific questions about the contribution of audiologic factors to learning and behavioral failure, it is necessary for them to have an understanding of the relationship of these factors to other aspects of learning and behavior and an awareness of basic principles of child development. This article synthesizes these basic scientific facts into guidelines for decision making for the clinician.

**Components of Learning and Behavior**

A variety of theories of child development and behavior exist in the contemporary literature which are, for the most part, poorly integrated. Theoreticians from a variety of disciplines have evolved their own terminologies, which are rarely interchangeable, to describe development and behavior. A functional model of these components is presented here as a working foundation for the clinician, and is not intended to be definitive or all inclusive.

The elements of a single learning episode or behavior can be broken down into four basic components: intake systems, information processing, output systems, and neurobehavioral functions.

1. **Intake Systems.** The first component refers to the integrity of sensory systems bringing information or stimuli into the central nervous system. Most important in this regard is auditory and visual acuity. Objective procedures and criteria exist to define the adequacy of these systems.

2. **Information Processing.** This component involves the processing of information which enters the central nervous system. Basic scientific knowledge about this phenomenon is sparse as the discrete processes postulated to be involved occur exclusively within the central nervous system. There is no direct way to describe and characterize these processes. Rather, it is necessary to make indirect inferences about them based on observations of responses to stimuli presented to the central nervous system (CNS) through intact sensory systems.

The various stages of information processing can be viewed as having a sequential relationship to one another. *Perception* entails the initial discrimination of information presented to the CNS through the sensory systems in terms of such parameters as form, space, position, sound, pattern, and sequence. *Integration* includes the interpretation of the pattern or sequence of information, the melding of information input through differing sensory systems, the retrieval of past information, and the formation of ideas. The next component, *encoding*, involves output processing including the selection of mode of output.

Other terminology frequently used to describe these components of information processing includes auditory discrimination, auditory processing, receptive language (comprehension), expressive language (word retrieval), auditory sequential memory, visual perception, spatial organization, visual sequential memory, higher order conceptualization and immediate recall, and short and long memory. A bevy of standardized language, psychological, and educational tests has been developed to attempt to describe strengths and weaknesses of these components in the individual child.
3. **Output Systems.** This level includes the mobilization and integration of motor output systems which result in a demonstrable act of learning or behavior. This includes visual motor integration, articulation, and body movement and control. Information from this act and its consequences feed back into the system through sensory intake systems.

4. **Neurobehavioral Functions.** Each of the three previous components are influenced and modulated by a variety of neurobehavioral functions. *Neuromaturation* can be arbitrarily defined as the underlying neurologic maturity and organization of the developing child. In this sense, it can be postulated to include such things as state of myelinization, degree of integration between discrete systems, and efficiency of neuroregulatory processes. Clinically, neuromaturation may be measured through observing the presence of so-called primitive reflexes and soft neurologic signs. *Temperament* refers to those presumed intrinsic behavioral characteristics that influence a child's interaction with the environment. Nine components of temperament have been described. In general, children belong to one of three temperamental types (easy, difficult, or slow to warm up) based on ratings in these categories. Recent studies suggest a relationship between certain temperamental ratings and learning and behavioral problems. Currently, temperament can only be measured through subjective rating scales that are susceptible to observer bias. Although the concept of *attention* is encompassed in several of the categories of temperamental function, it warrants special consideration. Attention refers to more than attention span or vigilance and includes the child's state or level of arousal, cognitive tempo (balance between impulsivity/reflectivity), purposeful focusing and filtering of distractions, and monitoring (quality control) of information attended to. Like temperament, attention and its component parts are most frequently subjectively assessed in a clinical setting.

Although these components of learning and behavior may not bear a linear relationship to one another as implied in the graphic model presented here, it is important to note that inefficiencies in any one component may have overflow or cascading effects on other components. The individual components are reciprocally dependent on each other in order to ensure efficient functioning.

**Principles of Development and Behavior**

From a review of the contemporary literature on child development, it is possible to synthesize a set of philosophical premises upon which an assessment protocol to address school failure and its possible audiologic components can be based. These premises include the following:

1. **Multiplicity of factors influencing development.** A wide variety of factors significantly impact on a child's development. These include, but are not limited to, biological, psychological, economic, sociological, and educational risk factors.

2. **Continuum of casualty.** These developmental risk factors do not necessarily impact on an individual child in an "all or none" fashion. Furthermore, children may respond differentially to the same risk factors. Outcomes of exposure to risk factors characteristically vary along a continuum ranging from a full expression of a physical or behavioral problem,
to partial expression, to no expression at all. Limitations in our current state of knowledge preclude accurate prediction of a child's specific response to a given exposure.

3. **Transactional scheme of development.** A child's functional status at any point in time is the product of a series of transactions between constitutional factors (intrinsic qualities) and environmental factors and life events. These reciprocal interactions are constantly evolving and are unique for each child. The vast majority of learning and behavioral problems are not simply the result of a unidimensional progression of a single risk factor operating in a cause-effect manner. Rather, research over the past decade has shown that learning and behavioral dysfunctions more often result from reciprocal interactions over time between constitutional and environmental factors. A truly comprehensive assessment of a child's development can only be achieved when the reciprocally interactive relationships between various parameters of constitutional and environmental development are addressed.

4. **Age interacts with risk factors.** The level of dysfunction associated with exposure to a potential risk factor varies as a function of age of occurrence, duration, and intensity.

5. **Manifest dysfunction interacts with environmental demands.** The expression of a dysfunction (ranging along a continuum of casualty) varies as a function of the performance demands made by the environment. Environmental demands vary according to age-related expectancies, culture, socioeconomic level, and the presence of family life stress events. Therefore, dysfunction in childhood (learning failure or behavioral problems) can be seen as the product of constitutional predispositions of a child that have interacted with environmental influences. The intrinsic predispositions may be present at birth but manifest themselves only when the environment requires efficient performance.

6. **Behavioral commonality of heterogeneous underlying weakness.** There is a commonality to the behavioral manifestations of a wide variety of underlying developmental or behavioral weaknesses. That is, a child with a conductive hearing loss, one with a receptive language problem, or one with an attention deficit disorder may all clinically be seen as having a short attention span and being highly distractible. Only a comprehensive assessment process can delineate underlying differences which may require vastly different management strategies.

**Clinical Applications**

**Common diagnostic process applied to learning and behavioral dysfunctions.** Utilizing the model of components of development and behavior and integrating the theoretical framework presented previously, it is possible to define a common diagnostic process that can be applied to a spectrum of learning and behavioral complaints. Although satisfactory approaches to these problems by nature require multidisciplinary input, this flow diagram provides a framework for decision making for the clinician. In this sense, it provides a guideline for defining the diagnostic elements necessary for comprehensive assessment and management of these problems. Similarly, for the consultant asked to attend to potential audioligic aspects of a problem, it reinforces the necessity for a comprehensive approach in order to avoid premature diagnostic closure or the onset of treatment protocols that address only an isolated component of a complex process.
The vast majority of learning and behavioral problems present to the physician as the result of a disparity between an expectation of efficiency and actual performance in any life sphere: home, school, or medical. Owing to the commonality of presenting manifestations of a wide variety of underlying dysfunctions, all learning or behavioral problems warrant some level of execution of this diagnostic process, the depth and intensity varying with the severity of the problem. Whether the physician is the first professional contacted or is brought in later as a consultant, he should ensure that each component of this process has been executed in order to generate a comprehensive management plan. The components of this common diagnostic process are as follows:

Historical data collection - the child. This should be a survey of potential risk factors including familial, pregnancy, perinatal, early health, delayed developmental milestones, temperamental dysfunctions, and somatic dysfunction such as enuresis or encopresis.

Physical assessment. At themost basic level this should include a functionally relevant physical examination including a traditional neurologic examination and basic sensory acuity screening.

Neurodevelopmental assessment. This should include an assessment of the components of development and behavior described previously (information processing, output systems, and neurobehavioral functions). A variety of assessment batteries are available to define strengths and weaknesses in the following areas of function: gross and fine motor skills, sequencing, retention, visual/perceptual, auditory/language, neuromaturation, temperament, and attention.

Psychosocial assessment. This should include an assessment of bonding/attachment dysfunction, disordered parent expectations, associated behavioral dysfunction, diminished self-esteem, management/discipline difficulties, inadequate support systems, and life stress events.

Psychoeducational assessment. This should include a history of past educational difficulties, manifest performance failure on psychoeducational testing, disordered school expectations, inadequate educational programs, and attention inefficiency.

Case formulation. With learning and behavioral problems, case formulation should entail a description of strengths and weaknesses with respect to constitutional medical, behavioral, and developmental function; environmental strengths and weaknesses; and the transactions between constitutional and environmental factors. The goals of this process are to define etiology if possible, focus the need for further evaluation or services, and serve as the basis for re-ordering expectations and setting treatment objectives.

Addressing the audiologic question. When a child is referred to the clinician for an evaluation of potential audiologic contributors to a learning or behavioral problem, the same diagnostic process should be applied in a limited fashion. Clearly, the physician needs to answer the specific medical question posed and to judge whether a particular medical factor directly contributes to the presenting behavioral or learning problem. Clearly, sensory deficits, like other medical factors, frequently do occur in children with learning or behavioral
problems; however, they rarely provide a simple answer to the problem. Therefore, the clinician must also search for other variables that may be additional significant contributors (such as temperament dysfunction or chronic inattention). If these are present, it is important to ascertain whether a comprehensive diagnostic approach has been undertaken. A referral to a multidisciplinary team (through the school or other setting) is indicated when this approach has not been taken.

**Screening and early intervention.** Although most professionals agree that early identification and intervention are the ideal with respect to successful outcome of learning and behavioral problems, the clinicians must avoid the temptation to prematurely transfer scientific knowledge to the clinical domain. This is particularly relevant to audiologic aspects of these problems. Most of our basic scientific knowledge in this area is just evolving. Therefore, in the primary care or consultative setting when faced with a child with an audiologic deficit, such as otitis media with conductive hearing loss, the clinician must strike a critical middle ground. The medical problem needs to be addressed in a rigorous fashion. Its potential relationship to learning and behavioral dysfunction should not alter or influence either the kind or intensity of treatment unless clearly indicated. Rather, this possible association should be acknowledged by the application of a comprehensive data collection process. This means screening such children longitudinally for associated delayed developmental attainment, temperamental dysfunction, school performance failure, attention inefficiency, somatic dysfunction, and behavioral dysfunction and referring these children for more comprehensive assessment and treatment when such factors are present. To do otherwise runs serious risks for the individual child and the family in terms of the damage of self-fulfilling prophecies and for society as a whole in terms of costly interventional procedures that ultimately may do little good. To truly address the needs of children, this phenomenon, which has characterized the fields of child development and education over the past three decades around a variety of medical factors, must be avoided.

**Therapeutic implications.** The efficacy of treatment of learning and behavioral problems is directly dependent on precisely describing the strengths and weaknesses and their interactions which are generated by the diagnostic process. By their nature, these problems require comprehensive treatment plans. For a given child this may mean medical treatment (such as resolution of a sensory deficit or treatment with a stimulant medication for attention inefficiency), behavioral treatment (such as behavioral management guidelines, behavior modification, or psychotherapy), and educational treatment (such as learning disabilities tutoring, language therapy, or remedial reading). Furthermore, success in one treatment modality is rarely independent of others. Therefore, it is imperative for clinicians not to view their roles as providers of an isolated answer and cure, but rather as important team members in a comprehensive process. Although parents and schools often desire such a neatly packaged answer, it rarely can be provided and can be a disservice to the child if it precipitates the withholding of other necessary services.

**Summary**

To effectively deal with learning and behavioral problems, physicians must acquire a new knowledge base and interact meaningfully with professionals from a variety of disciplines. They must be aware that audiologic factors are only one part of a complex matrix of components contributing to learning and behavior. Although the exact nature of these
components is still being defined, their interrelationship can be hypothesized. An individual can overcome the uncertainties and avoid inappropriate diagnoses in this field by applying a comprehensive, descriptive diagnostic process. With a broad understanding of the transactional model of learning and behavior, physicians can ensure that audiologic problems are defined and addressed appropriately. By acknowledging this approach in the identification, assessment, and management of children with learning and behavioral dysfunction, the physician can become a powerful advocate for these children.
Symptoms of interest to the otorhinolaryngologist are often of interest to the psychiatrist as well, and proper care of the patient requires the care of both specialists. Complaints about children, "I don't think he hears well enough", "he doesn't speak well for his age", "she has too many sore throats", "she has quite suddenly lost her voice", or symptoms such as "sinus headaches", hoarseness, dizziness, tinnitus, and difficult swallowing may have a psychological basis as often as an organic one. It is, of course, important that physicians think not in terms of either/or, but rather how much of the problem is psychosocial and how much is in the biological, physiological sphere and the interaction of these systems. Proper medical care today requires the physician to approach the patient's complaints with a biopsychosocial model of clinical investigation.

Additionally in pediatric patients the aspect of development must be considered. The importance of visual, oral, and auditory input in the human infant's growth and development is well known. Distortions in development occur when there is interference in the ability of the infant to respond to its environment or in the environment's ability to respond properly to the infant's needs. The fit between the infant and its environment, whether good or poor, will determine its development.

For instance, infants with esophageal atresia, when gastrostomy is necessary and interference with normal feeding practices occurs, show profound failure to develop gross motor skills in contrast to normal progress in visual and fine motor skills. It seems that "the physical art of oral feeding provides an important stimulus and organizing force for a variety of developmental accomplishments".

A surgical colleague with great experience feels that some of his best surgical repairs of cleft palate have not resulted in good speech, whereas other children with less than ideal repair have had very good speech. He attributes this to other factors in the child's environment, chiefly family attitudes and parent-child interaction, and spends a great deal of time talking with parents about their feelings about the child as part of the preoperative preparation.

Thus an examination of a child with symptoms or complaints referable to otolaryngologic care require not only attention to the ears, nose, and throat and their anatomy and physiologic function, but also to the child's function as a developing human being, interacting in his family with parents and siblings, and in the community and school with his peers.

**Psychology of the Pediatric Patient**

Psychological and social growth and development, as well as physical growth, is very rapid in children. Chronologic age is not necessarily the equivalent of psychological age, and
some children are able to behave in a more mature fashion than others. Also, the child's caretaking environment needs assessment, for parents vary in their abilities.

The three-month-old infant with croup requires special attention because of the small size of the larynx and the fact that even a small degree of edema may be life-threatening. Equal attention must be paid to the attitude of his caretakers. The more experienced, and the less frightened and anxious the mother and/or father or nurse, the more reliable will be the reports on the child's condition and his care during the critical episode. The infant whose parents are unduly anxious and unable to tolerate the anxiety of an ill infant may need to be hospitalized; whereas the more experienced parent, with a child equally ill, may be able to care for the child at home.

Separation Anxiety

As infants approach the age of 8 to 10 months, separation anxiety becomes apparent, in which the child is more acutely aware of who his caretakers are. The younger infant can be equally well cared for by his nurse or mother. However, after 8 to 10 months of age, and particularly as the child reaches the second year of life, separation from the parent is tolerated poorly. This means that the child between 8 months and 3 to 4 years who is to undergo elective procedures, such as a myringotomy, the placing of tubes in the ear, or even an examination of the ears and throat, will tolerate these procedures with greater equanimity in the presence of people he knows and trusts, provided that they are able to deal with their own anxiety and not complicate the situation for the child and/or medical specialist. Obviously hospitalization of very young children requires regard for parents as well.

By the time the child reaches school age, he is usually able to tolerate care by strangers without upset. He has learned that adults, teachers in the school, can be as helpful and trustworthy as his own parents, physicians, policemen, storekeepers, and others. Rather than being frightened by adults or strangers, the school age child is more concerned with his body integrity and is worried about "the shots" and "being hurt".

The adolescent has greater stability and confidence in others outside the home, but has concern about his own ability to deal with stressful tasks. Fear of the loss of control, "saying something" or "doing something" while under anesthesia, may be more difficult for the adolescent that any type of surgical manipulation.

Therefore, the otolaryngologist dealing with different age groups will have to deal with each of them somewhat differently and develop individualized strategies for them and for the different families in which they live.

Pain and Restraint

The otolaryngologist who deals with children is much like the pediatrician or pediatric dentist in that the young patients with whom they work often are of an age at which developmentally they are not able to cooperate for examination. Long ago, Dr C. A. Aldrich, an eminent pediatrician, in attempting to find a stimulus that would cause an infant to cry, and yet not wishing to inflict pain on the infant, came upon the use of restraint. Simply holding an infant's foot so that he cannot move will usually cause him to protest and cry.
Human beings resent restriction of movement, and children, normally active, resent "holding still". It takes a fair amount of psychological stability in order for a child to cooperate with even simple medical examinations, such as mouth or throat examination.

At the same time physicians dealing with children should be aware that restraint is not all bad, and indeed, has an anxiety relieving aspect to it. The nurse who says to the six-year-old boy, "Johnny, I know you want the doctor to draw the blood sample and get this test out of the way as quickly as possible, so I will help you hold your arm still for him", is well aware of the fact that the healthy part of the child does wish to hold his arm still. The frightened and anxious part of the child would like to run from the room and keep his arm as far away from the physician's needle as possible, behavior that can be seen as noncooperative.

Explaining to the child or to the parent what is necessary and how it will be approached also relieves anxiety. For example, the explanation to the parent might be that the examination of the child's ear reveals a bulging eardrum, that the pressure is causing the earache, and treatment requires a lancing of the drum. Such an incision will be painful but short-lived. Though sedation or anesthesia could be given, restraint by wrapping of the child in a sheet, with parents involved and present, will be less dangerous and more efficient. Involving parents in the examination of their children helps the child be less anxious, and the average parent is relieved to be helpful to the physician and their child. Occasionally parents may abdicate this assistance to the physician's nurse or other helper, but they appreciate knowing that the physician would involve them if they were willing.

Not all children have had the same experiences in growing up, and some children are more anxious than others. Not all parents are the same, and some are more anxious and frightened about medical care than others. A few questions such as, "What kind of a child is Sarah? Does she get frightened easily? How does she behave with strangers? Do you think she will be able to do what I have suggested?, and talking to even very young children, are helpful. Telling a two-year-old child, "I'm going to look in your ear with my light. Let me know if you want me to stop", helps the child to feel that things are not out of his control and will help him to tolerate better the anxiety of an examination. Obviously, the physician who wishes to work quickly will not be able to do so with the pediatric population. It takes more time and energy to deal with children and their families. Children cannot be rushed, are not sophisticated, and quickly make a medical office or surgical suite acutely aware of their displeasure. The tolerant unhurried physician has a sedative effect in his very manner on both children and their parents. The physician's feelings, whether anxiety or anger, are easily communicated, and though the adult patient may "put up with" a physician's insensitive and abrupt manner, the pediatric patient and the child's parents will not.

**Otolaryngologic Syndromes of Psychiatric Interest**

A child's failure to develop language ability is a relatively common complaint. Although mental retardation or hearing deficit are usually first thought of in a differential diagnosis, more commonly the failure is secondary to a lack of stimulation of the child and parental dysfunction. This may range from the severe problems presented by the autistic or symbiotic psychotic child to that of the overprotected, immature child with babyish speech.
Deafness and blindness may be on a hysterical basis, even in a very young child. Abt described a three-year-old child with hysterical blindness. Some of the most difficult diagnostic problems are the differentiation of whether the child's auditory apparatus is intact or whether the child is not responding at a cortical level. The autistic child may appear to be deaf by all standards but the most sophisticated of audiologic testing, and even then the examiner may have doubts.

Lye Burns

It is a tribute to pediatrics and to otorhinolaryngology that lye burns of the esophagus have decreased as a result of public education efforts and education of manufacturers. Still, young children in less than protective environments will ingest poisons and foreign bodies and require otorhinolaryngologic attention. Such children and families also require attention to their psychosocial environment in order to prevent recidivism. Children requiring esophageal dilatation, whether for congenital stenosis or chemical stricture, may also profit from psychiatric consultation.

Case Example. A four-year-old girl suffering from severe esophageal stricture caused by lye burns was uncooperative and required anesthesia for dilatations. With psychological support by a clinical nurse specialist (pediatric nurse with postgraduate training in child development) and involvement of the mother in the preparation of the child and care over a six-month period, the child was able to be dilated without anesthesia, and eventually to be dilated by her mother at home.

Esophageal Stenosis

Case Example. Beth (BD, Nov 1965) had a history of being an occasional vomiter until beginning feeding of solid foods at six months. She had increasing difficulty and by 22 months of age would gag, tear, and vomit projectily after eating solids. She could swallow milk, baby foods, and soups. After vomiting she would smile and be comfortable.

At the time of her first hospitalization at age 23 months she was in the 10th percentile for height and 50th percentile for weight. She had iron deficiency anemia with a hemoglobin of 7.0 gm and a hematocrit of 29. An esophagogram performed in November, 1967 revealed "a well-defined area of stenosis of the esophagus at the junction of the distal and middle third; the findings clearly indicate congenital stenosis of the esophagus".

After treatment of the anemia and esophageal motility studies, esophageal dilatation under anesthesia was begun. Dilatations were done March 27, April 10, April 25, and May 18, 1968. During the May, 1968 hospitalization psychiatric consultation was requested because of the concern about frequent dilatations and anesthetic risk. It was the psychiatrist's opinion, based on previous experience, that if the family could be helped to be less anxious and more relaxed, the esophageal function could be improved. A parent of another child handled in similar fashion agreed to talk with Beth's parents, the psychiatrist, and a pediatric nurse specialist (graduate training in child development). Follow-up care and counseling were offered, and a home visit was planned. This mode of treatment was agreed to by the otolaryngologist. At the home visit following discharge, the child and family seemed to be coping well.
There was no further contact from the summer of 1968 until a follow-up call was made to Beth's mother in the spring of 1980. She reported, "It was such a help to talk to that mother! The next day I decided to give Beth bacon and eggs and she did just fine and has continued to do so. She does have some trouble occasionally if she doesn't chew food well. If she eats too big a piece to swallow, she may have to vomit, and if she gets upset she also may not be able to swallow".

Beth has coped with her parents' divorce subsequently when she was 8 years, the mother's remarriage, and the birth of a brother at age 12 years. Today at 14.5 years of age, she is described as an active adolescent, good student, cheerleader, physically healthy, and "doing just fine". The mother did not wish for her daughter to have further medical contact or x-ray examinations of the esophagus.

**Seal Bark Cough**

Seal bark cough is a clinical syndrome that can be diagnosed "from the doorway". These children, usually of latency age and boys, have a characteristic barking, explosive cough. We have seen this cough in children we feel are compulsive and whose parents have high expectations, and regard the cough as a tic. Reassurance of parents and child that the etiology is primarily psychological rather than organic, and that the cough is a response to a tense environment, usually causes symptoms to disappear. The underlying personality, however, remains.

**Hoarseness**

Likewise hoarseness, with or without vocal nodules, is often associated with psychological tension.

*Case Example.* A five-year-old girl, the second of three children, having brothers ages seven and four, was seen with a complaint of "hoarse voice and immature speech". Laryngoscopy revealed bilateral thickening of the cords. The child was seen in psychiatric consultation as "the angriest little girl I've ever seen". Her parents were not very giving or sensitive to this child's needs. In therapy over a matter of months she was able to express her anger more appropriately and to ventilate her feelings. Her parents also were able to modify their behavior to some degree and the child's voice returned to normal quality.

**Aphonia**

Aphonia on a hysterical basis usually occurs in the adolescent female.

*Case Example.* Susan, 12, had a history of voice strain at an athletic meet 4.5 months prior to being seen and then "lost her voice". Seen by an otorhinolaryngologist several times for inflammation of the vocal cords, she was treated with medication. Though on a final visit the vocal cords found to be "clean and mobile" with no structural abnormality, the hoarseness persisted.

Susan was the fourth of five children with two older sisters and an older and younger brother. Her mother had died during childbirth, and she and siblings had been raised by her
mother's sister and her husband, though it was reported that she believed her aunt to be her natural mother in spite of different surnames.

Susan's mother-aunt had a very hoarse voice, much like Susan's, saw the girl as "happy, active, evil", and seemed impatient with the girl's inability to talk.

This girl related well and talked easily, but in a loud whisper. She was a striging, bright girl, more concerned than she needed to be about performing well. She had many friends, male and female, and tended to deny negative feelings about anyone or anything.

It was explained to her that her symptoms could be explained on the basis of psychological tension and difficulty in expressing feelings. This patient regained her normal voice one week after the diagnostic interview. Whether this was a "flight into health" or a transitory developmental conflict could not be determined on the basis of the limited evaluation.

**Summary**

Diagnosis and treatment of children with complaints or symptoms referable to the ears, nose, and throat require not only an examination of those organs, but also of the child as a developing psychological, social being, living in a family and interacting with a psychosocial environment. Once the physician approaches these problems with investigation of the various symptoms involved and some understanding of child development, management is clearer and more effective.
The Interaction Between Dentistry and Otolaryngology

M. M. Nazif, Richard C. Ruffalo


The oral cavity, including the dentition, has a close anatomic and physiologic relationship with the pharynx, the maxillary sinus, major and minor salivary glands, the nasal cavity, and the ears. This relationship necessitates a great deal of interaction and cooperation between dentists and otolaryngologists. This article reviews major areas of interaction in an effort to enhance the overall quality of care of pediatric patients.

Otalgia Dentalis

The ear receives its sensory nerve supply from the trigeminal, vagus, and glossopharyngeal nerves, and from branches of the upper cervical roots. Therefore, in the absence of local signs of otitis media, the possibility of referred pain should be considered. Such pain can originate in the oral cavity or in the temporomandibular joint. Dental pain can be referred through the trigeminal nerve to the ear. The cause of referred pain may be a pulpitis or a periapical abscess or, in younger patients, teething that is accompanied by significant gingival irritation or pericoronitis. Conversely, true otitis media can present solely as dental pain. In the absence of oral signs of dental disease, the possibility of otalgia secondary to otitis media should be considered.

The Maxillary Sinus

The relationship of the maxillary sinus to the dentition changes significantly with age. During childhood, the maxillary sinus enlarges as the posterior teeth form and erupt. With the extension of the sinus floor downward, the apices of the maxillary teeth eventually intrude into the sinus cavity with only a very thin, bony lamella separating the teeth from the sinus. This "mature" relationship is reached at approximately 15 years of age. The presence of dental periapical infection may lead to the elimination of any bony barriers between the dentition and the sinus, possibly facilitating direct bacterial invasion of the sinus cavity. Necrotic tissues may be forced into the maxillary sinus during root canal treatment of maxillary posterior teeth. The extraction of maxillary posterior teeth with advanced periapical lesions can create an antral-oral fistula. Occasionally, root tips of maxillary teeth are forced either partially or completely into the maxillary sinus during dental surgery. Foreign bodies can also be forced into the maxillary sinus through an antral-oral fistula. Most sinusitis of dental origin occurs in patients over 15 years of age and is seldom seen in a preschool population. Killey and Kay reviewed a series of 362 antral-oral fistulae and found only nine in patients between birth and 15 years of age. True sinusitis can also present as pain in the premolars and molars. Multiple toothaches involving posterior maxillary teeth with little or no evidence of caries or large restorations should suggest a diagnosis of maxillary sinusitis.
Nasal Cavity

Oronasal Fistula

An oral fistula may occur as a complication of the surgical removal of an impacted tooth in the palate or the premaxilla, or following the improper removal of palatal tori. Such a fistula may also exist in patients with cleft palate and may persist following palatal closure. Surgical closure of such a defect is the treatment of choice.

Intranasal Teeth

Intranasal teeth may be single or multiple, primary, permanent, or supernumerary. The cause of this pattern of ectopic eruption is not clear. Intranasal teeth may be asymptomatic and thus be found only on routine clinical or radiographic examination. Symptoms that may be associated with intranasal teeth include midfacial pain, nasal congestion or obstruction, headache, rhinitis caseosa, fever, epistaxis, rhinorrhea, or oronasal fistula. Surgical removal of nasal teeth usually alleviates these related symptoms.

Emergency Care

The majority of pediatric dental and maxillofacial emergencies result from either trauma, infection, or burns.

Traumatic Injuries

Trauma to the dentition or other oral structures is often associated with facial injuries such as nasal fractures, midfacial fractures, and fractures of the anterior nasal spine or other adjacent structures. Conversely, all patients with facial trauma should receive a thorough oral evaluation, since the presence of an intraoral injury may complicate recovery or compromise the airway.

History and Examination

A concise history is the first and most important step in making an accurate diagnosis, and should include information concerning the circumstances of the accident (when, how, and where it occurred) as well as past medical history and the postinjury status of the patient. A careful clinical examination that determines the nature and extent of injury will help the physician to choose the necessary course of action and diagnostic procedures.

Careful examination of the dentition, gingiva, oral mucosa, and facial bones is extremely important in the presence of a perioral injury. Such an examination should include (1) the dentition, to rule out crown or root fractures, excessive mobility, alveolar fractures, lateral or vertical displacement, or avulsions. This is best achieved by a thorough visual inspection of all existing teeth and by palpation of suspicious areas; (2) the gingival and oral mucosa, to rule out any soft tissue injury, possible jaw fractures, or impactions of foreign bodies; (3) facial and jaw bones, to rule out possible fractures; and (4) dental occlusion, to determine any trauma-related abnormal relationships.
Certain symptoms, such as tenderness, trismus, hemorrhage from the nose or the ear, deviation on opening or closing the mouth, gingival tears, or the presence of cloudy sinuses, necessitate further detailed evaluation.

**Soft Tissue Injuries**

A variety of soft tissue injuries, including lacerations, abrasions, contusions, puncture wounds, and avulsions, can occur. Degloving injuries may also occur with or without associated bony fractures. The first step is usually to cleanse the injured area of blood clots and debris in order to determine the extent, depth, and degree of vascular involvement. Some intraoral lacerations do not require suturing. Bleeding usually subsides spontaneously, and healing proceeds satisfactorily. Small lacerations with well approximated margins belong to this group.

Large lacerations, through-and-through lacerations, and lacerations associated with extensive, recurrent, or uncontrolled bleeding require careful assessment. Soft palate lacerations require a thorough pharyngeal inspection. The possibility of foreign body entrapment, immediate or delayed vascular injury, or formation of pharyngeal abscesses should be seriously considered.

Lacerations involving the labial frenum of infants are quite common and usually cause parents a great degree of alarm. Under normal conditions, these lacerations should not be sutured. Restriction of manipulation and a carefully planned soft diet for 24 hours should suffice.

Management of bites and avulsion wounds requires appropriate wound care, followed by careful debridement and primary closure whenever possible. Intraoral avulsions are best treated with either gingival grafts, if necessary, or left to heal by secondary intention. The administration of antibiotics and tetanus prophylaxis are indicated whatever course of action is taken.

**Injuries to the Teeth**

It has been reported that close to 50 per cent of children sustain some type of injury to their teeth. Occasionally, dental injuries are overlooked in the presence of the more obvious extraoral soft tissue injury.

**Crown Fractures.** The dentin or the pulp becomes exposed in crown fractures. The treatment of choice involves the application of calcium hydroxide to the exposed surface in order to promote pulpal healing, and should be performed as soon as possible. Occasionally, dental fragments become embedded into adjacent tissues, or may be swallowed or aspirated. Appropriate examination of adjacent tissues, particularly the lips, and an x-ray examination of the chest may be required.

**Root Fractures.** Involved teeth may exhibit increased mobility. The final diagnosis can only be determined with dental radiographs. Differential diagnosis should include alveolar fractures. Neither alveolar nor root fractures are usually visible on skull films. Alveolar
fractures should be suspected whenever gingival tears exist and several teeth exhibit synchronized mobility.

**Displaced Teeth.** Early treatment of displaced teeth tends to improve the prognosis significantly. Therefore, it is recommended that an attempt be made to reposition laterally displaced teeth with gentle thumb-index finger-gauze pressure as early as possible. In most instances, splinting is required for a period of 4 to 8 weeks after the displaced teeth have been repositioned.

**Avulstion (Total Loss).** Avulsion of primary teeth only requires proper care of the resulting soft tissue injury. An avulsion of a permanent tooth may require reimplantation. The prognosis of reimplantation is directly related to the length of time the tooth remains outside the socket. Therefore, it is recommended that an attempt be made to replace the avulsed tooth into the socket after careful cleansing of the area. Gentle pressure can be maintained on the tooth with gauze until the patient is ready for splinting. If immediate reimplantation is impossible, the tooth should be placed in normal saline or a moist towel and transported to the nearest qualified dentist or emergency room. Regular follow-up care and root canal therapy are usually required.

**Burns**

Burns involving the oral cavity usually heal with contracture and scarring. Mucous membranes are more susceptible to burns because of their low resistance. Available dental techniques using special splints to keep anatomic structures in proper relationship before scar formation occurs can prevent or greatly minimize the formation of contractures.

**Facial Bone Fractures**

The incidence of such fractures in pediatric populations is relatively low. The most common are nasal fractures, followed by fractures of the mandible. Most of these injuries result from trauma; however, other factors, such as child abuse or delivery by forceps, should be considered.

**Mandibular Fractures**

The vast majority of mandibular fractures occur in the weakest parts of the mandible, which are the condylar necks and the areas of intrabony, developing permanent tooth buds. The presence of a mandibular fracture is signalled by signs such as ecchymosis, swelling, chin or facial lacerations or abrasions, limitation of jaw movements, deviations, or malocclusion. A final diagnosis is made after radiographic examination. Special techniques that utilize dental films or tomography are indicated in certain areas.

**Midface Fractures**

This type of fracture is not very common in children. In 1901, LeFort divided midfacial fractures into three groups: LeFort I, which primarily involves the maxilla; LeFort II, which primarily involves the maxilla and the nasal complex; and LeFort III, which involves the whole midface, separating it from the cranium. Most of these fractures require
a cooperative effort and a thorough and detailed examination to ensure proper care; diagnosis can be challenging and frequently requires tomography. Such fractures should be suspected with the presence of malocclusion, swelling, ecchymosis, cloudy sinuses, or segmental mobility of the maxilla. Ophthalmologists, otolaryngologists, and dentists should work jointly to avoid the problems which could otherwise complicate these fractures.

Neck Masses of Dental Origin

Acute Localized Adenopathy

These are swellings of various sizes usually associated with the regional lymph nodes. Although the majority of such cases are idiopathic, a dental cause should be ruled out. The primary site can be a dental infection from advanced caries, trauma, gingivitis, or pericoronitis. In infants and school children, pericoronitis that is associated with the eruption of teeth should be carefully considered. Most of these swellings appear to occur between the ages of 10 and 12 years. Typically, the swellings occur near the angle of the mandible, in the submental or submandibular spaces. Most of the reported cases have been caused by streptococci or staphylococci. Early diagnosis and prompt treatment may help to avoid the need for incision and drainage.

Acute Odontogenic Infections

These enlarging masses are often associated with trismus, tenderness, fever, and malaise. When superficial, the abscesses may be fluctuant; if they are deeper to the cervical fascia or beneath the periosteum of the mandible, they will be firmer. The spread of infection is determined by gravity and anatomic barriers. An enlarging submental, sublingual, or submandibular abscess may cause an airway obstruction or may extend into the mediastinum. Such odontogenic infections are relatively common in children because of the high incidence of caries and the wide marrow spaces that allow the rapid spread of infection. The majority of upper neck abscesses are dental in origin; therefore, it is essential to identify the primary site. The offending tooth is usually very tender to percussion, and the surrounding gingival tissues are edematous and tender to palpation. Failure to identify the primary site can result in recurrence and the possible development of an extraoral sinus tract. Again, this phenomenon is more frequently seen in children.

Other uncommon infections of the head and neck, such as osteomyelitis, actinomycosis, or acid fast infections, can present clinically as a swelling that is similar to the more common odontogenic abscess.

Osteomyelitis of the Mandible

Osteomyelitis of the mandible in children is relatively rare. One such case of odontogenic etiology has been reported in which Bacteroides fusiformis was recovered. Garré’s osteomyelitis, a focal growth thickening of the periosteum with a peripheral reactive bone formation, may occur in the mandible of children. The most common cause is a mandibular first permanent molar that is abscessed. In most cases of osteomyelitis, the erythrocyte sedimentation rate (ESR) is elevated. The radiographic features of a lytic process
and periosteal elevation may be present, although severe acute osteomyelitis is not always associated with the radiographic stage of bone scans.

Treatment is dependent on the results of a culture and sensitivity tests. The prognosis is dependent on the onset of treatment. Purulent infection should be treated surgically, and nonpurulent infection should be treated with antibiotics that are administered parenterally. After a reduction in clinical signs and a decrease in the ESR, oral antibiotics can be administered for three weeks or longer until all the clinical signs have disappeared and the ESR is less than 20 mm per hr. For chronic osteomyelitis, the minimal regimen for oral antibiotic treatment is six months.

**Actinomycosis**

Actinomycosis is a chronic granulomatous disease caused by a group of actinomyces. The most common of these organisms in humans are *A. israelii* and *A. bovis*. Actinomycotic infection advanced by contiguous spread, disregarding tissue planes. Cervicofacial actinomycosis is the most common form of this disease. The organisms are normally present in the oral cavity and may enter through gingivae, necrotic teeth, extraction sites, or trauma. They spread into adjacent tissues and cause abscesses and the formation of fistulae and fibrous scar tissue. An interesting feature is the presence of the typical yellow spicules of sulfur granules in the pus. The prognosis associated with cervicofacial actinomycosis is generally good. The treatment of choice is penicillin given in high doses intravenously for up to six weeks, followed by oral penicillin for three to six months.

**Dental Care for the Immunosuppressed Patient**

The survival of patients with neoplasms during childhood has shown a steady improvement over the past 25 years largely because of chemotherapy. Many chemotherapeutic agents are now available, and new ones are being introduced rapidly. These drugs exhibit various degrees of toxicity. Patients often exhibit leukopenia, thrombocytopenia, anemia, or other symptoms. Major complications associated with the use of immunosuppressant drugs include the following.

**Infection.** Infection is a serious problem in immunosuppressed patients, as the host defenses are impaired and the antibody responses are inhibited. The oral cavity is particularly vulnerable because several drugs can cause disruption of intact integument or the everpresent array of oral flora, and the possible disruption of oral mucosa by mechanical means. Such breaks can open pathways for localized invasion by bacteria, viruses, or fungi. This invasion may become life-threatening unless it is treated early and aggressively. Careful and frequent monitoring of the oral status is recommended. Early dental evaluation that includes radiographs, assessment of oral hygiene, and elimination of all foci of infection is very important. Gentle manipulation of oral mucosa during routine oral hygiene is advised. The emphasis should be placed on prevention and thorough, regular evaluation. Most dental care can be safely provided during stages of remission. The use of prophylactic antibiotics should be carefully considered, especially if the procedures that are planned can cause bacteremia.

**Hemorrhage.** In most instances, spontaneous gingival or postsurgical bleeding results from the effects of the disease process itself or the chemotherapeutic agents on the number
and function of platelets. Therefore, thrombocytopenia should be suspected in such patients, and the bleeding tendencies of the patient must be evaluated before any dental surgical procedure is planned. A complete blood count, prothrombin time, partial thromboplastin time, and a platelet count should be obtained. Platelet transfusion should be ordered before dental extractions or an oral surgical procedure is performed. Elective procedures should be delayed until the patient is in remission.

Local Irritation. Several drugs currently in use can cause variable degrees of ulceration within the oral cavity. Such ulcers must be distinguished from surface changes by infection. Symptomatic treatment, such as frequent warm saline rinses, chloroseptic mouthwashes, or topical anesthetic lotions, are indicated to make the patient comfortable. Some degree of diet control may be required in severe cases.

Dental Care of Patients Receiving Radiation Therapy

Therapeutic doses of radiation to the head and neck have several major side effects, which include the following.

Xerostomia. Xerostomia can be demonstrated within two weeks following the initiation of radiation therapy. This condition is progressive and can lead to complete xerostomia in severe cases. The saliva may become more viscous. With the reduction in flow of saliva and its increased viscosity, the ability of saliva to lubricate and to act as an efficient buffer solution is drastically impaired. As a result, patients develop heavy dental plaque, which in turn initiates a rapid destruction of the dentition.

Vigorous oral hygiene, dietary restriction of frequent carbohydrate consumption, daily rinsing with a fluoride mouthwash, weekly application of neutral sodium fluoride gel, and a monthly professional evaluation may prevent radiation-related caries in most instances.

Infections. Certain changes in the bacterial and fungal oral flora develop as a result of xerostomia in patients who receive radiation therapy. Such changes lead to the overgrowth of opportunistic organisms such as Candida albicans. Should such an infection occur, culture and sensitivity tests should be obtained and appropriate treatment administered.

Osteoradionecrosis. This is the most serious complication of radiation therapy. Osteonecrosis is the result of a reduced blood supply and the decreased healing capacity of the irradiated tissues. The probability of developing radionecrosis is directly related to the dose of radiation given. If extractions are required, they should be performed prior to the initiation of radiation therapy. If postradiation caries cause dental infection and teeth must be extracted, an aggressive intravenous antibiotic prophylaxis for the duration of hospitalization, which is followed by high doses of oral antibiotics, may prevent most complications.

Loss of Taste. This complication is usually self-limiting, and taste sensation returns within one year. Most complications can either be prevented or can be reduced to a reasonable level. All patients who are scheduled for radiation therapy of the head and neck should receive a dental examination, have all necessary dental care completed, and should begin a strict oral hygiene program prior to the initiation of radiotherapy. Shielding of the
salivary glands and other bony structures should be attempted, if this would not interfere with the primary objectives of radiation therapy.
Infectious mononucleosis is frequently encountered in pediatric practice. Symptoms of malaise, fever, fatigue with sore throat, cervical adenopathy, and splenomegaly are well known manifestations of the disease. The otolaryngologist usually has minimal contact with these patients. Most present with symptoms that are managed by their pediatricians and only a small percentage of those affected children are hospitalized.

A review of admissions at Children's Hospital of Pittsburgh between 1970 and 1980 revealed 61 children whose presenting symptoms of infectious mononucleosis warranted hospitalization. Thirty-three (55 per cent) presented with otolaryngologic complaints (Table 1). One-third of those children admitted had some degree of airway obstruction. The remaining presentations were peritonsillar cellulitis (1), facial paralysis (1), periorbital cellulitis (3), epistaxis (2), suppurative lymphadenitis (1), external otitis (1), and sinusitis (2). The reason for admission of the remainder is shown in Table 2.

Infectious mononucleosis has been known as a disease of young adults, usually affecting those between 15 and 35 years of age. The review of our series shows equal distribution in children ≤ 5 years (19), 6 to 10 years (21), and ≥ 10 years (21). There was no sex predilection and 79 per cent were Caucasian. Sixty-seven per cent had splenomegaly which concurred with the incidence of other series. Ten to 15 per cent of these children are known to have group A beta-hemolytic streptococcal throat culture. Our finding is similar with 18 per cent. Thirty-nine children had white blood counts greater than 10,000 per cu mm. A lymphocytosis with atypical cells occurred in 78 per cent.

The following cases illustrate otolaryngologic presentations of infectious mononucleosis that are not frequently seen by the pediatrician. A recognition of these various chief complaints and treatment modalities is important in managing this disease.

Case 1: Airway Obstruction

A three-year-old black girl presented with a four-day history of sore throat and progressive dysphagia. Examination revealed +4 hypertrophied tonsils with exudate, inspiratory stridor, and significant airway obstruction. There was marked cervical adenopathy and the tip of the spleen was palpable. The child appeared toxic and dehydrated. The white blood count was 19,200 with a lymphocytosis. A "monospot" was positive. The lateral x-ray film of the neck confirmed adenotonsillar enlargement and airway obstruction. A throat culture was negative for group A beta-hemolytic streptococcus.

Because of imminent obstruction, a nasopharyngeal airway was placed without difficulty and secured. An intravenous line was begun and dexamethasone (Decadron) was given intravenously as a bolus (1 mg per kg) with a maximum dosage of 10 mg. One-half the initial dose was then given every six hours for four doses. The airway was removed after 24 hours without problem. Antibiotics were not administered. Oral intake gradually returned to
normal and the child was discharged on the fourth day. Follow-up was uneventful.

**Discussion**

Waldeyer's ring is the mass of lymphoid tissue that encircles the oropharynx and nasopharynx: the palatine and lingual tonsils, adenoids, and lateral pharyngeal bands. Airway obstruction in children with infectious mononucleosis is caused by hypertrophy of this tissue. Most children present with nasal obstruction with snoring, mouth breathing, and inspiratory stridor. Tonsillar enlargement can obstruct the oral airway. Many patients have dysphagia and are dehydrated. Significant cervical adenopathy can cause poor neck mobility.

Twenty-two children presented with airway obstruction requiring an otolaryngologic consultation: 13 were ≤ 5 years of age, 5 ranged from 6 to 10 years of age, and 4 were ≥ 10 years of age. Fifty-nine per cent were less than five years of age. Management varied over the years. However, intravenous steroids have been used more frequently since 1975. In no instance was a tonsillectomy performed. One tracheotomy was done in 1974 on a nine-year-old Caucasian girl who presented with airway obstruction.

Numerous modes of therapy have been proposed for treating airway obstruction in infectious mononucleosis. Tracheostomy was first suggested as a means of intervention in 1949 by Jones and Jones. This received considerable support in the years following. The procedure is not always straightforward though, as described by Lee in 1959. He reported a 16-year-old Caucasian girl who presented with progressive airway obstruction. A tracheostomy was performed under local anaesthesia, but because the patient could not lie still, the airway became obstructed and the patient had a cardiac arrest. The procedure was completed under general anaesthesia.

Tonsillectomy has been supported by those who feel that the disadvantages of emergency tracheostomy in children outweigh the advantages. This treatment modality was first reported in 1956 by Ranta of Finland. Since then, it has been a popular way of managing airway obstruction in infectious mononucleosis. The problems with this operation and general anaesthesia should not be viewed lightly in these children. The tonsils are usually acutely infected and friable. Liver function abnormalities and prolonged coagulation times have been reported. Consequently, bleeding may become a major issue.

Yeager reported performing a tonsillectomy on a 4.5-year-old Caucasian girl with infectious mononucleosis in 1964. During induction, a rubber airway was passed because of labored respirations and these were relieved. The operation proceeded with a blood loss of 150 to 200 mL. It was necessary to place suture ligatures in both tonsillar fossa. This case supports tonsillectomy in airway obstruction in patients with infectious mononucleosis. However, it illustrates the potential for compromise of the patient because of the significant blood loss seen with acutely infected tonsils. In addition, the airway may be successfully managed by use of a nasopharyngeal airway alone.

While our method of managing these children is more conservative, we feel that it is safe and still solves the problem of impending airway obstruction. Initially, these children receive intravenous hydration with total fluid losses being taken into account. Decadron, 1 mg per kg, with a maximum dose of 10 mg, is given initially. One-half of the loading dose is
then given every six hours for 48 hours. A soft rubber nasopharyngeal airway is placed and secured. All tubers are removed within 48 hours. Several children were maintained on doses of oral prednisone for a few days following the tube removal at the discretion of the attending physician. None of these children has returned to Children's Hospital with mononucleosis, airway obstruction, or recurrent tonsillitis. This review describes our success in managing acute airway obstruction in these patients without subjecting them to the increased risks of anesthesia and surgery. We propose that this regimen is a safe and reasonable way of treating acute airway obstruction in the pediatric patient with infectious mononucleosis.

Case 2: Facial Paralysis

A 2.5-year-old Caucasian girl presented to Children's Hospital of Pittsburgh with a three-day history of fever, sore throat, and bilateral otalgia. One day prior to admission, her mother noticed that the right side of her face was not moving well. Examination confirmed bilateral otitis media and right facial peripheral paralysis. Splenomegaly and hepatomegaly were present. Tonsils were +3 enlarged with exudate and there was cervical adenopathy. A "monospot" was positive. White blood cell count was 10,400 with a lymphocytosis. The child was begun on prednisone, 2 mg per kg per day, and ampicillin, 250 mg orally every six hours. A myringotomy was performed. The paralysis began to resolve within 24 hours and the child was completely well by the end of the week.

Discussion

Infectious mononucleosis has been associated with dysfunction of all the cranial nerves causing anosmia, parosmia, unilateral deafness, and facial paralysis most often. The relationship with facial paralysis was first described by Osell in 1937. In otitis media, it is felt that the facial nerve becomes involved secondary to swelling and compression in the fallopian canal. Complete recovery in the young patient with a viral illness is common and is related to the length of time it takes for function to begin to return.

Myringotomy is frequently used for treatment of facial nerve paralysis in otitis media. Steroids are controversial in the management of facial paralysis with a viral or idiopathic cause but were employed here with apparently good results. Patients should be followed with facial nerve stimulation and watched carefully for signs of degeneration. Because the facial nerve is frequently involved in viral infections, other causes such as mumps, chickenpox, herpes zoster, influenza, and polio should be considered. The course is one of rapid onset with slow but usually complete resolution. No topographic studies have been performed in these patients, but the course appears similar to that of Bell's palsy.

Case 3: Epistaxis

A five-year-old Caucasian girl presented to Children's Hospital of Pittsburgh with a left epistaxis of several hours' duration. She was taking erythromycin for an upper respiratory infection. Examination revealed +3 tonsillar hypertrophy, and a palpable spleen tip. White blood cell count was 8,700 with a lymphocytosis. "Monospot" was positive. A platelet count on admission was 20,000 but fell to 4,000 by the next day. A bone marrow confirmed the diagnosis of idiopathic thrombocytopenic purpura. The nose was packed with Surgicel and
bleeding was controlled. The platelet count returned to normal over the following week. No further intervention was necessary.

**Discussion**

Infectious mononucleosis has been associated with autoimmune hemolytic anemia and viral etiologies have been proposed for idiopathic thrombocytopenic purpura. Here, the low platelet count secondary to the idiopathic thrombocytopenic purpura was the obvious cause of the epistaxis. Cautery, nasal packing, or other routine ways of managing nosebleeds cannot overcome the basic coagulation defect. Therefore, the manipulation and trauma of these procedures must be minimized. Packing with an absorbable coagulant such as Oxycel or Surgicel will usually suffice until the underlying hematologic defect can be corrected. The child's activity should be kept to a minimum and anything that increases circulatory pressure such as leaning over, straining, nose blowing, and sneezing should be avoided.

**Conclusion**

We have described the significant, potentially life-threatening complications of infectious mononucleosis for which the pediatrician may require the assistance of the otolaryngologist. Recognizing the various presentations of infectious mononucleosis is of particular importance. Our approach to the pediatric airway is conservative, safe, and avoids operative intervention. Management of facial paralysis and epistaxis involves treatment of the underlying disease.