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Acute Otitis Media: A Public Health Perspective
Derek Drummond, M.D.
March 18, 1999

Case Presentation
J.D., a previously healthy girl, presented to her pediatrician at 6
months of age with a 3-day history of increasing irritability and fevers. History revealed that she had upper respiratory tract symptoms for 3 days that included nasal congestion, rhinorrhea, dry cough, decreased appetite and low-grade fevers. The parents became concerned the night before presentation when she became very irritable, appeared to be in pain, and vomited once after bottle-feeding. The girl was seen pulling on her ears and was inconsolable prior to arrival at the pediatrician's office. She had no previous medical problems and was not taking any medication other than acetaminophen.

Physical exam demonstrated a 6-month old girl who was crying in her mother's arms. She was alert but not cooperative. She had a temperature of 101.5 F. Examination of the ears revealed a right tympanic membrane that was bulging, erythematous, and didn't move with pneumotoscopy. The nasal mucosa was congested and the patient was mouth breathing. The remainder of the physical exam was unremarkable. She was diagnosed with acute otitis media and prescribed a 10-day course of amoxicillin at 40mg/kg/day. Within 24 hours, her symptoms were improving and she began to feed normally. Follow-up at 2 weeks revealed bilateral middle-ear effusions in a happy, asymptomatic child. The MEE would resolve spontaneously over the next 3 months.

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Acute Otitis Media
A Public Health Perspective

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March, 1999

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The role of immunity in sensorineural hearing loss was first suggested in 1958 by Lenhart. Kikuchi, in 1959, wrote of "sympathetic otitis" whereby surgery on one ear affected hearing in the other. He proposed an autoimmune phenomena as the etiology. In 1961 Beickert, and two years later, Terrayama presented data supporting autoimmunity in experimental guinea pig cochleas. McCabe described 18 patients with bilateral asymmetric hearing loss progressing over weeks to months which responded to steroid therapy. His 1979 paper asserted the importance of a high index of suspicion in these patients since, if diagnosed early, they could be treated and their hearing preserved.

Humoral and cell mediated immunity, the lymphocyte-macrophage system, and the complement cascade work in homeostatic harmony to provide immune protection to the host. B cells are produced in the bone...
marrow and, through antigen stimulation and differentiation into plasma cells, produce specific antibodies. T cells are derived in the thymus and provide regulatory function for B cells, cytotoxic activity, and generate lymphokines. B and T cells also form immunologic memory. Cells of the lymphocyte-macrophage system phagocytose foreign cellular components, process antigen, and produce interferons. The complement cascade amplifies antigen antibody reactions. Chemotactic, anaphylotoxic, opsoninization, and immune adherence functions arise from the complement system. Kinin-like substances are also complement generated.

The inner ear is immunologically active. The endolymphatic sac acts as the afferent limb of inner ear immunity since it can concentrate and primarily synthesize antibody. IgG is the most common antibody produced with IgM, IgA, and secretory component being present in lower concentrations. The distal endolymphatic sac is the site of immunologic activity due to extensive perisaccular lymphatics. Antibody production is independent of serum or cerebrospinal fluid levels. Secondary exposure to antigen in the inner ear induces a more intense response than primary exposure to antigen.

Autoimmunity occurs with loss of homeostatic control in the immune system. Host tissues become recognized as foreign and induce damaging vasculitis and fibrosis. Veldman described a continuum of autoimmunity. On one end, organ specific responses with organ specific autoantibodies and T cells produce tissue alteration (i.e. Hashimoto's thyroiditis). On the opposite end of the spectrum is non-organ specific diseases with circulating non-specific autoantibodies (i.e. systemic lupus erythematosus). In between is organ specific disease with non-specific autoantibodies (ie primary biliary cirrhosis).

Patients with idiopathic autoimmune sensorineural hearing loss present most commonly with bilateral progressive hearing loss. Fifty percent have vestibular signs, and symptomatically progress over weeks to months. Females between the ages of 17 to 42 years represent 65% of the cases reported by Hughes. Twenty percent of Hughes' study later manifested signs of systemic autoimmune disease.

McCabe proposed using ESR, ANA, RF, complement levels, and quantitative immunoglobulin levels as a screening panel for autoimmune inner ear disease in high risk patients. Positive values in any of the screening tests would warrant leukocyte inhibition testing. Hughes classified patients as high risk if they had bilateral and progressive sensorineural hearing loss, no response to conventional therapy, concomitant immune disorders, abnormal screening tests or improvement of hearing with steroid therapy.

Treatment goals in autoimmune inner ear disease include improving speech thresholds to levels treatable with hearing aids in severely affected patients and recovery of hearing to near normal levels in those with mild to moderate losses. Steroids, cytoxan, and plasmapheresis compose the available therapeutic modalities. Hughes advocates high dose (prednisone 20 mg four times daily for 10 days then 10 mg every other day for 3-6 months) steroids as initial treatment. Patients are tapered slowly and restarted if symptoms recur. As initial therapy, McCabe recommends cytoxan (2mg/kg twice daily) combined with steroids (prednisone 30 mg every other day) for 3 weeks. If speech discrimination scores increase by 20% or pure tone average improves by 15 dB, therapy is continued for 3 months. Cyclophosphamide is tapered first followed by steroids. If symptoms recur both drugs are restarted. Three month cycles are
continued until patients can be weaned. No patient required more than 24 months of treatment in McCabe's study. Hughes advises plasmapheresis for those patients unresponsive to steroids and cytoxan after 6 to 8 weeks at the above stated doses. Plasmapheresis theoretically removes unwanted humoral and cellular elements. Treatments are given three times weekly for 2 weeks followed by once weekly for 4 additional weeks.

In summary, otolaryngologists need a high index of suspicion for autoimmune etiologies in patients with sensorineural hearing loss. Ophthalmologic, neurologic and rheumatologic consultations are useful in ruling out systemic vasculitic diseases. Steroids and cyclophosphamide remain the cornerstones of treatment in autoimmune inner ear disease, with reservation of plasmapheresis for refractory cases. If caught early, and with aggressive medical management, hearing stabilization and possible improvement are feasible.

Case Presentation

A 53-year-old white woman was first seen in November 1991 by a private MD, for sore throat, otalgia, scleritis, and temporal headaches. ANA, RF, RPR, and VDRL were all negative at that time. Her ESR was mildly elevated to 56. Magnetic resonance imaging of the head and neck was read as normal. She was treated unsuccessfully for occipital nerve impingement with local steroid injection. Systemic steroids relieved all symptoms until attempted taper when the headaches returned. In December 1991 she was admitted to the Neurology service at The Methodist Hospital with a diagnosis of temporal artery headaches. Further past medical history revealed an episode of temporal headache and pleuritic chest pain 6 months prior to her workup in Beaumont. Neurological and ophthalmologic evaluations revealed no specific anomalies. Lumbar puncture and temporal artery biopsy were without pathological change. VDRL was nonreactive. RF, ANA, and HIV testing were all negative. She had improvement of her headaches with systemic steroids. She was readmitted to Hospital in January 1992 with new onset nausea, vomiting, and sudden hearing loss in her left ear. ESR on admission was 37, urinalysis clear, and blood hematologic assessment showed a very mild iron deficiency anemia. C3, C4, RPR, FTA:ABS, SSA, SSB, RF, ANA, and antineutrophil cytoplasmic antibody were all within normal values. ACE inhibitor level was 8.4 (1.8 to 6.2). Serum protein electrophoresis was consistent with an acute phase response to inflammation. Chest x-ray was normal. Repeat MRI was remarkable only for left middle ear inflammation. Gallium scan showed increased uptake in the auricular region. Electronystagmography was consistent with a left peripheral (nerve or end organ) deficit. Lip biopsy was negative for inflammatory changes. Mastoid and middle ear biopsies obtained after complete mastoidectomy showed no pathologic abnormality. She has been managed with long-term steroid therapy with good control of her vestibular symptoms.
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The most common causes of barotrauma today are from the use of the Self-Contained Underwater Breathing Apparatus (i.e. SCUBA gear), commercial air travel, and from hyperbaric oxygen chambers. In fact, hyperbaric oxygen therapy has been found to produce over a 50% incidence of barotrauma. Well over 50% of the medical problems that are related to barotrauma are referred to an Otolaryngologist. Over 90% of these complaints involve the ear.

The noncompressible middle ear cavity makes the ear susceptible to damage from these ambient pressure changes. Middle ear pressure is governed by a law of physics known as Boyle's Law, which states that at a constant temperature, the volume of a body of gas
is inversely related to the pressure to which it is subjected. Applying this law to diving, demonstrates that if a diver descends 33 feet (or the equivalent of 1 atmosphere of pressure), the ambient pressure will double from 1 atmosphere to 2 atmospheres. This will cause the volume of gas to be cut in half in the middle ear, resulting in a 50% increase in negative middle ear pressure if the eustachian tube is closed. The first 33 feet of descent represents the largest change in the volume of the middle ear while diving or during hyperbaric decompression. A diver must dive to greater than 150 feet in depth to equal the total volume change produced during the first 33 feet. This explains why the majority of otologic diving and hyperbaric injuries occur during shallow dives and not during deep water dives as one might expect. In fact, MEBT and IEBT have been reported to occur in as little as 8 ft of water.

The pressure experienced during air travel is closely regulated by artificial means through the use of pressurized cabins. Commercial air lines maintain a constant pressure differential in the cabin of 8.5 psi above the changing ambient pressure outside the plane. At an altitude of 18,000 feet above sea level, a person in an unpressurized cabin would equilibrate the middle ear to an ambient pressure of 1/2 an atmosphere. Regulating cabin pressure at 8.5 psi above the changing ambient pressures, a passenger will experience a cabin pressure equal to that at sea level (i.e., 1 Atmosphere) while flying at an altitude of 16,000 feet and will require minimal middle ear equilibration. At 40,000 feet, the passenger will have a cabin pressure equal to 7000 feet above sea level. This pressurization is beneficial when a plane descends from a cruising altitude of 40,000 feet to land at sea level. The ambient pressure change experienced by the passenger is decreased by 2/3 since the passenger will be actually experiencing a descent equivalent to a 7000 foot descent. Although pressurized air travel has reduced the potential risks of barotrauma, it is important to keep in mind that only a pressure differential of 80mm Hg is required to close the eustachian tube in normal individuals. A descent from 37,000 ft to 27,000 feet cruising altitude will result in a cabin gauge pressure change equal to 80mm hg, resulting in difficulty with clearing the middle ear.

In order to fully understand the effects that Boyle’s Law has on Eustachian tube function, we will briefly review the normal anatomy and function of the eustachian tube. In the resting state, the eustachian tube is closed. A positive pressure within the nasopharynx, or a contraction of the tensor veli palatini, levator palatini, or the salpingopharyngeus is required to open the eustachian tube when middle ear pressure equalization is attempted. There are three common maneuvers which can be used to aid in middle ear pressure equalization. These are the Valsalva maneuver, the Frenzel maneuver, and the Toynbee maneuver. The Valsalva maneuver opens the eustachian tube by increasing the nasopharyngeal pressure above the middle ear pressure as a result of closing the naries and glottis while increasing intrathoracic and intra-abdominal pressure. This maneuver is the most common cause of barotrauma to the ear if performed too forcefully. The Frenzel maneuver is performed with a low pressure valsalva while contracting the muscles of the pharynx. This forces air into the
eustachian tube without increasing the total intrathoracic pressure. Finally the Toynbee maneuver involves swallowing while pinching the nose, thus creating a negative nasopharyngeal pressure which can force the eustachian tube open. By far, the Frenzel and Toynbee maneuvers are the safest to perform during eustachian tube dysfunction as neither have been associated with barotrauma.

To date, there is no readily available test to identify patients with eustachian tube dysfunction other than a history of difficulty with clearing the middle ear. Factors which would lead to a decrease in eustachian tube function include recent upper respiratory infections, uncontrolled nasal allergy, nasal polyposis, and deviated nasal septum.

The differential diagnosis of the disorders related to barotrauma are damage to the middle and inner ear, inner ear decompression sickness, and alternobaric vertigo.

Middle ear barotrauma, also known as aerotitis media, is due to an inability to equilibrate to ambient pressure changes. The etiology can be located either in the middle ear or external ear. This process also occurs most commonly during the descent as a forced valsalva is attempted, increasing the middle ear pressure and allowing for damage of the tympanic membrane to occur. This process can also occur during ascent if eustachian tube dysfunction causes air trapping, otherwise known as "reverse Squeeze."

The external ear canal can also be a source of middle ear barotrauma if a closed space is created between the outer rim of the concha bowl and the tympanic membrane. The closed space may be due to either a cerumen impaction, ear plugs, or external otitis. As a diver descends, ambient pressure will increase, causing a net negative pressure gradient between the external ear canal obstruction and the tympanic membrane. The obstructing plug is then forced deeper into the external ear, resulting in a tympanic hemorrhage or perforation. The patient typically experiences extreme pain as the descent phase of the dive begins, despite an ability to clear the middle ear.

Edmonds et al from the Australian Diving Medical Center, devised a grading system for middle ear barotrauma. The grading scale is from zero to five. Grade 0 is when a patient experiences symptoms of middle ear barotrauma and no physical findings are present. Grade I is when the presence of tympanic membrane injection can be seen. Grade II has injection as well as hemorrhage within the tympanic membrane. Grade III includes gross hemorrhage and Grade IV includes gross hemotympanum. Grade V includes the presence of a tympanic perforation.

There are three common forms of IEBT: Cochlear Damage resulting in intracochlear and intralabyrinthine hemorrhage, perilymphatic fistula formation, and IEDS secondary to the formation of gas bubbles beneath the round window.
IEBT usually occurs with MEBT, although the absence of MEBT cannot exclude the presence of IEBT. The symptoms of sensory neural hearing loss, vertigo, and or tinnitus should indicate the presence of possible IEBT. The etiology of IEBT has been proposed by Goodhill et al to be secondary to implosive or explosive forces within the cochlea. Goodhill proposed that forces exerted on the cochlea during a forced Valsalva have different effects depending on the patency of the eustachian tube. When the eustachian tube is forced open suddenly, an acute rise in middle ear pressure will result causing an inward bulge of the round window and an outward bulge of the stapes foot plate. If the force is strong enough, implosion of the round window and a secondary outward pull on the stapes footplate may occur.

If the eustachian tube is blocked, a valsalva maneuver will cause an elevation of CSF pressure which will be transmitted through a patent cochlear aqueduct or internal auditory canal causing a rise in the intracochlear pressure. If the difference between the perilymphatic space is sufficiently greater than the middle ear pressure, an explosive rupture of the round or oval window ligament will occur.

Both the implosive and explosive forces generated by a force valsalva are theorized to cause a perilymphatic fistulae or dislocation and rupture of Reissner's membrane, as well as the basilar membrane, the saccule, the utricle, or the semicircular canals. Antonelli and Paparella have studied the temporal bone pathology in scuba diving deaths and confirmed the presence of these pathologic findings. Simmons et al have demonstrated through experimental models that pressure differentials of less than 2 cm of water can cause labyrinthine ruptures.

IEBT secondary to cochlear damage will present with nonfluctuating high frequency sensorineural or mixed hearing loss with or without tinnitus or vertigo. There is typically no progression of symptoms. The treatment of IEBT secondary to damage to the membranous labyrinth and cochlea includes bed rest with head elevation, the use of vasodilators, steroids, histamine, and carbogen, in an effort to decrease inflammatory changes and increase the delivery of oxygen. Parell et al have shown that if proper precautions are taken to maintain proper eustachian tube function, no further deterioration takes place in hearing if a patient returns to diving after experiencing cochlear IEBT.

IEBT secondary to a perilymphatic fistulae typically presents with fluctuating sensorineural or mixed hearing loss, as well as vertigo exacerbated with positional changes, and a sense of constant disequilibrium. On physical exam, a positive Hennebert's sign (the presence of nystagmus when positive and negative pressure is applied to the EAC in the presence of an intact tympanic membrane) has been cited by Thompson and Kohut as a strong positive indicator of a perilymphatic fistula in patients with or without the presence of hearing loss. Healy et al have indicated that a positive
Romberg sign and the presence of positional nystagmus are consistent with the presence of a perilymphatic fistula.

When present, the most common location of the perilymphatic fistula has been demonstrated by Goodhill et al to be at the anterior rim of the oval window in the area of the fistula ante fenestram, which is one of the weakest areas of the otic capsule. The management of patients suspected of having a perilymphatic fistula includes bed rest with head elevation and avoidance of increased intracranial pressure for a variable period of days, with intermittent regular audiograms.

The length of time a patient should be observed before an exploratory tympanotomy is performed is very controversial. In a study by Paparella et al, it was shown that in chinchillas suffering a traumatic round window perforation, all perforations had partially healed after 3 days, and all were completely healed after 9 days. The results of this study form the basis for conservative management of perilymphatic fistulas. Simmons et al recommend that if hearing loss or vestibular symptoms are progressive, or if after 10 days any vestibular symptoms remain, an exploratory tympanotomy should be performed. Parell and Becker also advocate this 10 day observation period. Singleton and Kohut are proponents of a 5 day observation period. Goodhill et al suggest a 48 hour observation period. Pullen et al suggest that the shape of the audiogram should indicate if an exploratory tympanotomy should be performed. He advocates immediate surgery if an audiogram demonstrates a flat shaped total or near total SNHL in the presence of a history of diving or air travel within the past 72 hours. However, he states that if the hearing loss is limited to only the high frequencies, a down-sloping audiogram, a closure of a perilymphatic fistula has not been shown to improve hearing and, thus, surgery is contraindicated unless vertigo is present.

Inner ear decompression sickness (IEDS) is a form of nontraumatic cochlear damage and is the result of gas bubble formations within the inner ear. It is commonly seen after dives to extreme depths using a Helium oxygen mixture as a substitute for nitrogen oxygen mixture, in order to minimize the narcotic effects of nitrogen. IEDS occurs as a diver returns to the surface and, in an attempt to accelerate helium elimination from the tissues, changes from an oxygen-helium mixture to an air mixture. McCormick et al demonstrated that if rapid decompression occurs, inert gas bubbles of helium will form within the microvessels and otic fluids, causing a blockage of the microcirculation and resulting ischemia of the stria vascularis, spiral ligament and semicircular canals. A hypercoagulable state is produced secondary to the activation of factor XII, resulting in further vascular occlusion. As previously stated, the treatment of this disorder is immediate recompression to approximately three atmospheres deeper than the depth at which the symptoms began to occur. Farmer et al have demonstrated a near total return to baseline hearing if recompression is initiated immediately. Proper diagnosis is crucial, as the hyperbaric chamber recompression will aggravate inner ear damage due to perilymphatic fistulas and cochlear barotrauma.
The differentiation between IEDS and IEBT can be made based on the following criteria.

- **Dive profile:**
  1. IEDS is rare in shallow water (only 3 cases reported in less than 100 feet of water),
  2. diving to a depth that is near the limits of the nodecompression zone and not undergoing decompression,
  3. using a HE-02 gas mixture (IEDS),
  4. h/o rapid ascent. (IEBT associated with rapid descent);

- **Time of symptom onset:**
  1. During descent (IEBT),
  2. During ascent (IEDS),
  3. Shortly after decompression (IEDS);

- **Associated symptoms:**
  1. the presence of decompression sickness (only 6 cases in literature of IEDS without systemic decompression sickness),
  2. h/o difficulties in clearing the ear (IEBT),
  3. h/o nasal or sinus problems (IEBT);

- **Physical findings:**
  1. MEB (IEBT),
  2. Central neurologic signs (IEDS).

The last disorder related to barotrauma is the phenomenon known as alternobaric vertigo. This is a syndrome first described by Lundgren in 1965 as vertigo occurring during ascent due to unequal pressure in the right and left middle ear. The duration of the vertigo is usually from a few seconds to minutes and is not associated with hearing loss. In his review of 2053 Swedish divers, Lundgren found a 16.7% incidence of alternobaric vertigo. A review of 526 Australian naval divers by Bayliss cited a 0.4% incidence. Although this process is self-limiting, a diver experiencing alternobaric vertigo while attempting a valsalva maneuver at the surface, should not undergo a dive or air travel.

As people become more active in both air travel and recreational water sports, education regarding the hazards of extreme middle ear pressure changes should be expanded. Patients who are more susceptible to aural barotrauma, either due to lifestyle, upper respiratory infections, or after ear surgery, should take extra precautions to guard against eustachian tube function during air travel or underwater sports or simply avoid these activities when the risks of barotrauma are the greatest.
Case Presentation

A 36-year-old man experienced difficulty clearing his left ear while snorkeling to a depth of 35 feet in Cozumel, Mexico. After attempting a forced valsalva maneuver, the patient reported sudden hearing loss and tinnitus in his left ear that continued to persist. He denied any sensation of dysequilibrium or vertigo. A history of a previous high frequency SNHL was noted before the diving incident. No other past history of middle or inner ear disorders was noted. No history of a recent URI, nasal obstruction, or recent eustachian tube dysfunction was reported. Physical exam demonstrated a Grade III tympanic membrane hemorrhage with an intact drum. The right ear canal, tympanic membrane, and middle ear appeared normal. The Weber exam lateralized to the right ear and ear conduction was greater than bone conduction in the left ear. No evidence of spontaneous nystagmus was noted. Hennebert's sign and Rhomberg sign were negative. Fistula test was negative. An audiogram was obtained demonstrating an 80 dB SNHL on the left and no change in the high frequency loss on the right. An MRI scan demonstrated no abnormalities. The patient was placed on steroids and antibiotics. Carbogen therapy was initiated and two stellate ganglion blocks were performed. A repeat audiogram was obtained one week later that demonstrated improvement in the left SNHL. The patient continued to report no dizziness. The patient was advised to refrain from exertion for 1 month and schedule a follow-up audiogram in six months.

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BELL'S PALSY

July 20, 1995

S. Mark Overholt, M.D.

Bell's palsy is synonymous with idiopathic facial paralysis. It is the most common cause of an acute facial paralysis, and accounts for 75 to 80 percent of all cases. The annual incidence is 20 to 30 per 100,000. It's name is derived from Sir Charles Bell, a surgeon and accomplished artist from Edinburgh, Scotland who lived from 1774 to 1842. In 1821, he published his anatomic diagrams of the course of the facial nerve and its innervation of the facial musculature.

Early treatment strategies for Bell's palsy were primarily surgical. In the 1930's Balance and Duel described decompression of the distal one centimeter of the facial nerve at the stylomastoid foramen. Tumarkin felt that decompression of the
stylovastoid artery was necessary to adequately treat Bell's palsy. By the 1940's the main treatment modality was facial nerve decompression in the mastoid segments. In the 1960's electrical testing allowed refinement of the indications for surgery. Jongkees proposed decompression of the nerve when a difference of greater than 3.5 mAmps was noted by nerve excitability testing when comparing the unaffected to the affected side. Alford further delineated indications and discussed the importance of associated symptoms, the cause of the paralysis, and the experience of the surgeon in deciding to proceed with decompression. Important surgical advances were also developed in the 1960's. These include the middle cranial fossa approach which was initially described by House in 1961, and the total facial nerve decompression described by Pulec in 1966. The 1970's was a transition period for the theoretical treatment approach to a patient with Bell's palsy. With the refinement of electrical testing and the use of steroids, a trend had begun away from surgery. Adour was one of the first neurotologists to state that surgery might not be beneficial. In 1985 May discussed the failure of transmastoid decompression to improve outcome. However, Fisch and Esslen found good results after combined middle cranial fossa and mastoid decompression of the facial nerve in 11 out of 12 patients with Bell's Palsy. The role of surgery remains controversial and indication will be discussed later. By the 1980's and 1990's the treatment had changed to being primarily medical. Coker additionally demonstrated that the electrical evidence of a degenerating facial nerve on electroneuronography was reliable and correlated with histologic evidence of neuronal degeneration.

Several theories exist regarding the cause of Bell's palsy. Most favor a viral inciting event that triggers edema and inflammation in the nerve leading to infarction and nerve damage. Many viruses have been implicated but there is mounting evidence supporting the herpes simplex virus serotype 1 as the etiologic culprit.

Mechanical entrapment in the fallopian canal is the cause of the ischemic injury to the facial nerve. In 1981 Fisch described the "physiologic bottleneck" at the meatal foramen. In his comparative study of adult temporal bone specimens, the average diameter of the fallopian canal was found to be 1.02 to 1.53 mm, except at the meatal foramen where it was 0.68 mm. Histologic examinations have demonstrated hemorrhage and edema in the nerve at this site. Additionally, intraoperative stimulation blockage has been documented to occur in this narrow segment with intact stimulation distally.

Nerve injury is classified according to the schema of Sunderland. This system describes progressively worsening injury to the nerve starting with a neuropraxic injury and ending in neuronotmesis or nerve transection. Five grades of injury are described. The grade of the injury correlates to the final functional recovery of the facial nerve. Clinically facial nerve injury is classified using the House-Brackmann system. Grade I is normal function. Grade II shows slight weakness but no
synkinesis. **Grade III** shows obvious weakness with some mass movement. **Grade IV** has inability to elevate the brow, significant synkinesis, and obvious weakness. **Grade V** has barely perceptible motion, and **grade VI** has no movement.

The differential diagnosis for facial paralysis is broad and must include inflammatory causes such as herpes zoster oticus or sarcoidosis, traumatic injuries such as temporal bone fractures, mastoiditis, cholesteatomas, the Melkersson-Rosenthal syndrome, and primary or metastatic neoplasms of the temporal bone. By far though, idiopathic facial paralysis predominates. It accounts for almost 80% of all cases of facial paralysis.

The epidemiology of Bell's palsy is best described in Peitersen's monograph from the Copenhagen facial nerve study. In over 1000 patients he found there to be no sex predilection. A broad age range was noted, typically from 15 to 60 years old. Rarely did he note bell's palsy in patients less than 15 years of age. Patients presented with a classic constellation of symptoms. There is usually a prodrome of periauricular pain that heralds the onset of facial palsy within 24 hours. The palsy is unilateral and involves all muscle groups. In order to make the diagnosis of Bell's palsy the evaluating clinician must exclude the following: concurrent CNS disease, otologic infection, auricular blebs suggesting the Ramsey-Hunt syndrome, parotid masses, and an occult neoplasm such as a facial neuroma in a patients who has a recurrent palsy.

There is no consensus regarding the work up of a patient with Bell's palsy. Most would include a routine basic audiogram in order to rule out an asymmetric hearing loss, which might suggest another process. Topognostic tests, which include the Schirmer's test of tearing, the salivary flow test popularized by May, and the acoustic reflex test are of historic interest only. While once felt to assist in localizing the nerve injury, we know now that the injury occurs at the meatal foramen. Additionally, they do not offer prognostically valuable information.

Radiographic imaging is reserved for those who have no improvement in there palsy within 6 months and for those who a have recurrent palsy. The MRI is the best study to image the facial nerve. In patients with Bell's palsy gadolinium MRI enhancement of the perigeniculate region is prominent. However, as Schwaber pointed out in 1990, this enhancement does not correlate with outcome. Furthermore, Gribarski showed that 76% of normal subjects will show enhancement of the perigeniculate region with gadolinium.

Electrodiagnostic test are reserved for patients with complete facial paralysis. Available tests include the nerve excitability test(NET), maximal stimulation test(MST), electroneuronography (ENog), and electromyography. The latter is
appropriate when the paralysis has been present for several weeks. In the acute period the first three are most appropriate. NET and the MST are both performed using the Hilger stimulator. The first is a threshold test and the second a supramaximal stimulation test. A criticism of these two tests is that the grading of the response to the stimulus is subjective, and thus may not reliably predict denervation. However, Coker and Fordice demonstrated that a greater than 3.5 mAmp difference side-to-side on NET correlated closely with objective ENog data showing greater than 90% degeneration of the nerve. May showed that in patients who did not have a response to the MST for 10 days, over 85% did not fully recover. Those who did have a response to MST within the first 10 days had excellent recovery. Enog is the most widely used test to evaluate an acute complete facial paralysis. It is an objective reliable test that compares the difference in the amplitude of the compound action potential of the unaffected to affected side of the face. Fisch and Esslen found that when the amplitude of the affected side is less than 10% of the unaffected side the prognosis is poor. Coker correlated the evidence of electrical degeneration with histologic evidence of nerve degeneration.

Treatment of an acute facial paralysis is initially medical. The regimen currently used by most otologists is prednisone 1mg/kg/day tapered over a 10 day period. With the mounting evidence implicating the herpes virus, many additionally recommend acyclovir 800 mg three times a day for 10 days.

Indications for surgery are controversial, but most neurotologists feel that electrical evidence of greater than 90% degeneration or a difference of 3.5mAmp from side-to-side is sufficient to proceed with a decompression. Surgery must be performed as early as possible. After 21 days there is probably no benefit. The decompressive surgery must focus on the meatal foramen. May found that transmastoid decompression alone did not improve outcome, whereas when combining transmastoid and middle cranial fossa decompression Fisch found good outcome in 11 of 12 patients.

Good prognostic variables include presentation with an incomplete palsy, onset of return of facial function within three weeks, and age below 60 years. Peitersen found that 94% of patients who presented with an incomplete palsy progressed to full recovery. If return of facial function was within one week almost 90% fully recovered. Return of function in the second and third weeks yielded full recovery in 83% and 60% respectively. Overall, 85% of patients with Bell's palsy will recover to normal function.

In conclusion, Bell's palsy is the most common cause of an acute facial paralysis. It is unilateral and sudden in onset, often with a prodrome of auricular pain. After excluding other potential causes of a facial paralysis, treatment is with prednisone and acyclovir. Surgery is reserved for those who meet electrodiagnostic criteria or...
are worsening on medical treatment. Decompression must include the site of pathologic compression of the facial nerve, the meatal foramen. Finally, fortunately 85% of patients with Bell's palsy have full return of facial function.

Case Presentation

A white female initially presented at the age of 29, with a 16-day history of a complete right-sided facial paralysis. Complicating this problem was the fact that she was also 34 weeks pregnant. No complaints of associated dizziness or changes in her hearing were reported, but she did note some facial dysesthesias in the distribution of the mandibular branch of the trigeminal nerve. There was no history of recurrent facial edema or a fissured tongue. Physical exam did not show any auricular blebs or parotid masses. An audiogram was normal with the exception of absent right-sided acoustic reflexes. Nerve excitability testing showed a good response at 3.0 milliamps on the unaffected side and no response at 7.0 milliamps on the right. Because of her pregnancy and late presentation she was not offered further treatment. Her recovery was incomplete. In May, she began to notice some unusual movements and spasms in her face. Final recovery left her with good symmetry at rest and excellent eye closure, but she had mild brow ptosis, synkinesis, and gustatory tearing. She was graded according to the House-Brackmann scale as a III out of VI. She subsequently did well with some intermittent dysesthesias of the right face until age 36 when she suffered an attack of Bell's Palsy on the left side of her face. She was treated with prednisone and acyclovir and eventually fully recovered. Approximately two years later she experienced two recurrences of her right-sided facial paralysis. Both episodes were treated with prednisone and acyclovir with recovery to her baseline grade III deficit. She is currently being followed by the neurotology service at The Methodist Hospital to monitor any further recurrence.

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UNUSUAL CEREBELLOPONTINE ANGLE NEOPLASMS

Glenn W. Knox, MD
November 4, 1993

Cerebellopontine angle lesions are the predominant skull base neoplasms that affect the posterior fossa. Although vestibular schwannomas account for the majority of primary neoplasms, a wide array of other lesions must also be considered in the differential diagnosis. In Brackmann and Bartels' series of CPA neoplasms, vestibular schwannoma accounted for over 90%. The remaining primary tumors were meningiomas (3.1%), primary cholesteatomas (2.4%), facial nerve schwannomas (1.2%), other schwannomas (0.2%), and other tumors (1.9%).

Meningiomas represent up to 18% of all intracranial tumors and approximately 3% of CPA tumors. The cells lining the arachnoid villae are the cells of origin. These cells are distributed throughout the intracranial space predominantly in relation to veins and dural sinuses. Meningiomas are benign but
locally aggressive tumors which occur at different anatomic sites in the following order of frequency: parasagittal, falx, convexity, olfactory groove, tuberculum sellae, sphenoid ridge, CPA, tentorium, lateral ventricle, clivus. The gross appearance is typically a globular mass that is firmly adherent to the dura mater, with characteristic speckles scattered throughout the tumor that correspond to the microscopic psammoma bodies. The tumor displaces but does not invade adjacent neural tissue.

**Case Presentation**

A 51-year-old white man noted the onset of vertigo in 1980. This resolved, but returned in the late 1980s, reaching a peak in 1988. His vertigo symptoms had improved since then. He also complained of a multi-year history of mild right-sided tinnitus, which had been variable but more noticeable this year. He complained of a two-year history of mild right-sided hearing loss which had also been more noticeable this year. His major complaint was a sense of fullness in the right ear which he had noticed in January of 1993, which did not improve with medical treatment. Past medical history was noncontributory. Physical examination was unremarkable. A recent audiogram showed a right-sided 40 dB high frequency sensorineural hearing loss with a PB max of 96%. Tympanograms were type A. MRI revealed an intracanalicular mass.

The patient was admitted to TMH and underwent a right excision of cerebellopontine angle mass via the midfossa approach. During surgery, the tumor appeared to be associated with the superior vestibular nerve. The tumor was noted to be tightly adherent to the surrounding structures, was rubbery in consistency, and contained several apparent venous sinuses. The patient recovered uneventfully with useful hearing preserved. Pathology revealed benign fibrous tissue with abundant vessels, consistent with a benign vascular tumor.

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PEDIATRIC CHOLESTEATOMA
A. Paul Vastola, MD
October 7, 1993

The term cholesteatoma is actually a misnomer coined by Johannes Muller in 1838. He described "a layered pearly tumor of fat which was distinguished from other fat tumors by the presence of biliary fat or cholestrin that is interspersed among sheets of polyhedral cells." Aural cholesteatomas are best thought of as skin growing in the middle ear space. Cholesteatomas actually do not contain fat but are composed of an outer matrix which surrounds layers of desquamated epithelium. The matrix is comprised of fully differentiated keratinizing squamous epithelium resting on a collagenous perimatrix. Various theories have been advanced to account for the destructive bone resorption seen commonly with these lesions. These include activation of osteoclastic bone destruction, mechanical pressure necrosis and bone degradation by enzymes such as collagenases and lysozymes.
Cholesteatomas may be classified as either congenital or acquired. Congenital cholesteatomas develop behind an intact tympanic membrane; according to the classic teachings of Derlacki and Clemis there must be no antecedent history of infection or breach of the tympanic membrane. This definition has recently been challenged by Friedman et al who note that approximately 70% of children will have had at least one episode of otitis media. There is little doubt that patients with epidermal rests of cells in the protympanum may also have episodes of AOM. The acquired form of cholesteatoma is much more common. Primary acquired disease arises from a skin lined retraction pocket within which retained keratin debris accumulates. Primary acquired cholesteatoma occurs most commonly in the posterior-superior quadrant of the pars tensa and in the pars flaccida. Secondary acquired cholesteatoma develops from an ingrowth of skin through a tympanic membrane perforation that is then retained within the middle ear, mastoid or both.

The pathogenesis of cholesteatoma growth is still poorly understood as evidenced by the multiple theories currently found in the literature. However, a common denominator appears to be eustachian tubal dysfunction. Bluestone proposes that patients with acquired cholesteatoma have a functional obstruction of the eustachian tube and are thus predisposed to high negative middle ear pressures. The areas of the tympanic membrane most susceptible to these forces are the pars flaccida and posterior-superior regions. The expansion or growth of cholesteatomas are channeled along well defined pathways determined by ligaments and folds. Review of embryologic development is helpful in understanding these pathways. Between the 3rd and 7th months of development the gelatinous tissue of the middle ear space is absorbed. A primitive tympanic cavity develops by growth of an endothelium lined pouch extending from the eustachian tube to the middle ear cleft. Four primary sacs then bud into the cleft. The remnants of these sacs direct the growth of cholesteatoma along predictable pathways in the middle ear. Prussaks space is commonly thought of as the most common point of invasion for primary acquired cholesteatomas. From this space cholesteatomas expand in one of three directions: the posterior route is most commonly seen, and follows the superior incudal space above the incus into the epitympanum. The inferior route follows the inferior incudal space into the mesotympanum. The anterior route from Prussaks space is the route least frequently travelled. The anterior pouch of von Trolsch serves as the avenue for spread into the protympanum.

The presence of cholesteatoma requires surgical intervention unless underlying medical problems contraindicate exposure to general anesthesia. Most cholesteatomas are asymptomatic in their early development; children may come to medical attention with otorrhea but will rarely complain of decreased hearing. Microscopic examination of the ear is imperative in identifying and delineating cholesteatoma. Audiometric evaluation is part of the standard preoperative workup. Computerized tomography may be used to delineate the extent of disease, check the aeration of the mastoid and to help rule out intratemporal and/or intracranial complications.

The primary surgical goal is to achieve a safe, dry ear by removing disease with the preservation of normal anatomy. Improving hearing is a secondary goal. The principles of surgical management are based on the extent of the disease and the presence of complications arising from cholesteatoma. Much debate has emerged in the literature concerning canal wall up procedures vs canal wall down approaches. It is clear that the consensus among otologists over the last several years has been that canal wall up
procedures are indicated in patients with well pneumatized mastoids and adequate middle ear clefts with cholesteatoma limited to the middle ear space or mastoid. Relative contraindications to canal wall up procedures include a sclerotic mastoid, a fistula, an only hearing ear and poor eustachian tubal function. There is no objective measurement of eustachian tubal function though several clinical observations can help predict the ability of the middle ear to ventilate itself. According to Parisier et al the appearance of the pars tensa, the amount of mastoid cellular development and the appearance of middle ear mucosa will reflect the eustachian tubal function and guide the surgical approach. The success of any given approach may be measured in terms of the rate of recidivism. Originally a term applied to criminal behavior, recidivism is the measurement of residual disease plus recurrent cholesteatoma.

Cholesteatoma in children is widely considered to be a more aggressive disease than in the adult population for two major reasons. First, very extensive disease is found more frequently in children compared to adults and second, higher rates of residual and recurrent disease have been documented in the pediatric population. Of note however is the observation that the incidence of complications arising from cholesteatoma is directly related to the duration of the disease and, as such, adults tended to have higher complication rates.

Glasscock in 1981 published a retrospective review of charts comparing rates of recidivism in patients less than 16 years old to those older than 16. Almost 90% of both groups had a canal wall up approach regardless of initial findings. Rates of recidivism were compared and it was found that patients less than 16 had an almost two-fold higher rate and a much shorter interval to recurrence. Based on these findings Glasscock concluded that pediatric cholesteatoma is more aggressive than that seen in adults. There was no statistical analysis presented and no criteria were set forth regarding canal wall up vs canal wall down approaches.

In 1977 Palva published a retrospective evaluation of 65 pediatric patients (<16 years old) matched with 65 adults with cholesteatoma. All patients underwent canal wall down procedures. The duration of disease in the pediatric population was shorter and the number of complications was higher in adults than in children. Operative findings revealed that 65% of pediatric ossicular mechanisms were disrupted compared to 84% in the adult group despite the finding that cholesteatoma was more extensive in the pediatric group. The authors calculated a 5% rate of recidivism in the pediatric group but did not calculate rates in the adult group. Based on the finding that cholesteatoma tended to be more extensive in children and have an overall shorter duration of disease these workers conclude that pediatric cholesteatoma is more aggressive. There is no mention of recurrence rates in adults however.

A study published in 1988 by Parisier illustrates the results of a single surgeon over 15 years. Operative approach was dictated by intraoperative findings with 53% of patients undergoing canal wall down procedures and 30% of patients undergoing canal wall up approaches. The remainder of patients had a tympanotomy for localized disease. Average follow-up was 4 years and rates of recidivism did not differ significantly between the pediatric population and an adult population. Based on these findings, the authors conclude that pediatric cholesteatoma is not a different disease than that seen in adults but that surgical approaches must be individualized to the patient's disease.
A preliminary evaluation of the Baylor experience with pediatric cholesteatoma was performed. There were 26 patients (19 males and 7 females) for a total of 27 operated ears. The age at presentation averaged 8 years (range 2-15 years) with an average follow-up of 4 years (range 3 months to 15 years). The most common presenting symptoms were otorrhea (11/26), hearing loss (4/26) and tympanic membrane perforation (3/26). Five patients had congenital cholesteatoma. Pre-operative and post-operative audiograms were performed in all patients and there was no significant difference in pure tone averages (30dB pre-op compared with 27dB post-op). Fifty-four percent of patients had canal wall up procedures while 27% had canal wall down procedures. Nineteen percent of patients had a tympanotomy approach. Planned second look surgery was performed in 16 of 27 ears. Two patients underwent primary ossicular reconstruction while 9 patients had a secondary ossicular reconstruction. The rate of residual disease was 4/27 (15%) while the rate of recurrent disease was 3/27 (11%) for a recidivism rate of 26%.

In conclusion there remain many questions concerning the pathophysiology of cholesteatoma; whether or not the disease differs in children and adults is still a matter of debate. The next area of inquiry will be on a cellular level. It is clear however, from the above data, that a reasoned individualized approach to cholesteatoma is the best way to achieve low rates of recidivism.

Case Presentations

An eight-year-old white female child presented with a history of chronic eustachian tube dysfunction, numerous episodes of acute otitis media and recent bloody otorrhea from the left ear. Her otologic history dates to the age of three when she had an episode of acute otitis media which was treated with antibiotics and decongestants. The patient subsequently failed a school audiogram and continued to require intermittent courses of oral antibiotics. Subsequent school audiogram failures were attributed to "fluid in the ears." There was no history of facial nerve palsy, vertigo or otologic surgery.

Physical exam was significant for a white mass behind the posterior half of the right tympanic membrane and a central posterior, superior perforation of the left tympanic membrane. There was no evidence of keratin debris although the drum was severely retracted over the malleus. Audiometric evaluation of the right ear demonstrated a slight loss over the lower frequencies and a type C tympanogram. CTT of the temporal bones revealed a soft tissue mass involving the tympanic membrane extending toward the epitympanum without erosion of the scutum.

The child was brought to the operating room where a right tympanomastoidectomy (canal wall up) was performed. Cholesteatoma was found involving the posterior half of the eardrum, extending towards the epitympanum and antrum. The ossicular mechanism was noted to be intact although there was some attenuation of the incudostapedial joint. The cholesteatoma was excised completely, leaving the ossicles intact. A medial graft was incorporated. The patient has done well postoperatively.
Bibliography


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CHORDOMAS OF THE SKULL BASE
Carla M. Giannoni, MD
October 1, 1992

Chordomas are relatively rare neoplasm arising from embryonic notochordal remnants and comprise less than 1% of intracranial neoplasms. They typically occur along the neuraxis, especially at the developmentally more active cranial and caudal ends, notably in the sphenoid-occipital, sacrococcygeal, and vertebral locations. Twenty-five percent to 40% of chordomas occur in the sphenoid-occipital or skull base region. These tumors occur predominately in the 30 to 50 year old age range and show a slight predominance in men.

These tumors can occur extra-axially, in unusual locations, and are then termed primary. They probably arise from ectopic notochordal elements. Chordomas have been described in the nasopharynx, mandible, maxillary sinus, frontal sinus, and, as in our case report, in the petrous apex of the temporal bone.
In 1856 Luschke, working in Virchow's lab, described jelly-like tumors in the region of the clivus of Blumenbach. In 1857 Virchow also examined these tumors and named them "ecchondrosis physaliphora." He believed they arose from the spheno-occipital synchondrosis and were of cartilaginous origin. Muller, in 1858, examined embryonic tissues and, after noting their histologic similarities, postulated these tumors derived from embryonic notochord. In 1895 Ribbert demonstrated the origin of these tumors when he pierced the nucleus pulposus of the vertebral column and found that similar tumors developed. He applied the term "chordoma" to these tumors.

The embryonic development of the notochord is important in understanding the anatomic distribution of these tumors. The primitive streak forms in the third week of fetal development and forms the notochord. During the fifth week of life, it extends from the coccyx cephalad to the skull base, where it courses through the odontoid process, the posterior sphenoccipital plate and ends at Rathke's pouch. The notochord becomes surrounded by mesodermal tissue which then forms the vertebral column. Ultimately the notochord disappears, but remnants persist in the nucleus pulposus of the intervertebral discs.

Binkhorst et al described seven points of origin of craniocervical chordomas: the dorsum sellae, the Blumenbach's clivus, retropharyngeal region, squama occipalis, nuclei pulposi of cervical vertebrae, and the ligament of the dens.

SYMPTOMS

The clinical presentation depends on the origin and extension of each particular tumor. Delay in diagnosis is common, secondary to the occult nature of the disease and poorly localizing signs and symptoms. A headache arising in the vertex, ipsi-parietal, orbital, or frontal regions is a typical early symptom. This is due to stretching of middle fossa dura and is often severe. Patients may also present with paresthesias or anesthesia of the jaw region, serous otitis media, hearing loss, nasal obstruction/anosmia, syncope and, rarely, vertigo. Recurrent meningitis has also been noted in these patients.

The late symptoms generally depend on the direction of tumor extension. Anterior extension leads to diplopia and ophthalmoplegia, or both. Posterior extension presents as facial or other lower cranial nerve neuropathies.

RADIOGRAPHIC FEATURES

CT findings of an expansile, destructive, lytic lesion with associated soft tissue mass are characteristic of chordomas. However, they are also seen with chondrosarcomas and other similar lesions. Foci of calcification may also be seen. The CT scan is useful in defining the anatomy of bone destruction.

MRI is better than CT for defining the limits of a lesion and any vascular relationships, especially on the T2-weighted images. MRI of chordomas show hyperintensity in T2 and hypointensity in the T1-weighted
images. The T1 images are especially useful in defining any tumor-SCF interfaces.

On arteriogram, an avascular may be seen displacing the basilar artery. Radiographic studies cannot reliably distinguish chordomas from chondrosarcomas and other similar lesions, but can help eliminate other possible diagnoses.

**DIFFERENTIAL DIAGNOSIS**

Benign lesions most frequently occurring at the skull base include: meningiomas, neuromas/schwannomas, glomus tumors, vascular anomalies (e.g., internal carotid aneurysms), congenital cholesteatoma, mucocele, osteomyelitis, eosinophilic granuloma, and cholesterol cysts.

Malignant lesions of this location include primary carcinoma (squamous-, adeno-, acinic, adenoid cystic), metastatic carcinoma (breast, prostatic, renal cell, bronchogenic), rhabdomyosarcoma, nasopharyngeal cancer, lymphoma and chordoma, and other mesenchymal tumors (chondroma, chondrosarcoma, and osteoclastoma).

**HISTOPATHOLOGY**

Four criteria have been used in the histologic diagnosis of chordoma: 1) a lobular arrangement of cells; 2) a tendency of the cells to grow in cords, irregular bands, or pseudoacinar forms; 3) production of abundant intercellular mucinous matrix; and 4) the presence of large physaliphorous cells. A microscopically mixed population of cell types exist: stellate or primordial cells, intermediate cells and the physaliphorous cells. The stellate cell is the only actively proliferating cell. Chordoma cells then proceed through a stage of vacuolization until they reach the characteristic physaliphorous appearance. The cells then progress to destruction and rupture, completing their life cycle.

Immunohistochemical tests have been developed in an effort to aid in the differentiation of these lesions. Chordomas are frequently positive for epithelial antigens - cytokeratin (CK) and epithelial membrane antigen (EMA), and negative for vimentin.

A subtype of chordoma, chondroid chordoma, deserves special mention. Histologically these tumors are a mixture of chordoma, chondroma, and chondrosarcoma.

**PROGNOSIS AND TREATMENT**

The reported average survival for chordomas of the skull base is 4.1 years. There appears to be a better prognosis for chordomas of the nasopharynx and paranasal sinuses. Notably, chondroid chordomas have a reported 15.8 years average survival. The best treatment for these lesions is total surgical excision. Recurrence is the rule and metastases are extremely uncommon; patients usually succumb to local disease.
Radiation therapy has been used in the postoperative care of these patients because of the tumor's usual relentless course to local recurrence and death with current conventional treatments. Fractionated proton radiation therapy is currently advocated for the treatment of chordomas. Because of its higher biological effectiveness, patients can be treated with a higher total equivalent radiation dose.

Case Presentation

A 57-year-old white female presented with a six- to nine-month history of intermittent left temporal headaches. They occurred monthly and usually lasted three days. She had no complaints of visual difficulties, hearing loss, vertigo, hoarseness, or dysphagia. Her past medical, surgical, and family histories were unremarkable. On physical exam she was found to be healthy. All cranial nerves were functionally intact.

An MRI of the brain was done and revealed a signal abnormality of the left petrous apex and adjacent basiocciput with associated enhancement. A CT scan of the head and cervical spine showed a destructive, lytic lesion of the left basiocciput just anterior to the jugular foramen and anterolateral to the foramen magnum with probable erosion of the carotid canal.

A full metastatic workup ensued including routine laboratory work, chest x-ray, mammogram, CT of the abdomen and pelvis, thyroid ultrasound, and serum protein electrophoresis. No significant abnormalities were found. A radionucleotide bone scan showed no additional bony lesions. Four vessel cerebral angiogram was negative for vascular abnormality. Three months later a follow-up CT scan of the head with thin cuts of the temporal bone showed a 1.5 X 2 X 1 cm lytic lesion corresponding with the previously identified lesion, but slightly increased in size. The bony margins of the lesions were irregular and suggestive of an aggressive process. She was then referred for surgical evaluation.

She underwent a joint neurotologic and neurosurgical procedure comprised of a combination left type B infratemporal fossa and temporal fossa approach with biopsy and removal of the tumor. Frozen section revealed the tumor to be a chordoma. The patient tolerated the procedure well and had an uneventful postoperative course. The patient has been referred to Boston for fractionated proton radiation therapy.

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**CONGENITAL AURAL ATRESIA**

January 12, 1995

Alberto D. Fernandez, M.D.

The first mention of attempts to surgically correct congenital aural atresia was made by Thomson in 1843 in England. Although 2 out of the three cases were aborted when a thick atresia plate was encountered, a third record documented what was presumably a tympanic membrane, although it appears that stenosis became a problem postoperatively. In 1883, Kiesselbach attempted an atresia repair on a child that resulted in facial paralysis. It was not until 1947 and the work of Pattee and Ombredonne that a significant improvement in hearing could be obtained with surgery. Pattee reported that fixation of the stapes via the lateral chain to the atresia plate was responsible for these patients' deficits. Ombredonne devised an approach through the mastoid in order to fenestrate the lateral semicircular canal, with reportedly good hearing results.
Using Wullstein and Zollner's tympanoplasty techniques, Bellucci reported a 30 decibel gain in hearing level in 50% of his cases. Shambaugh, in 1967, recommended surgery in unilateral cases only if the cochlear reserve allowed hearing improvement to approach that of the normal ear by 25dB. Several studies by Gill, Naumann and Schuknecht reported their individual experiences adding minor variations to the basic transmastoid approach. With the advent of polytomography and subsequently, computed tomography, finer detail of middle and inner ear structures could be obtained and used to select patients for surgery. Jahrsdoerfer, in 1978, reported the first large series using the anterior approach for congenital aural atresia repair, whereby the middle ear cavity was entered directly via a newly-created external canal without performing a mastoidectomy.

An appreciation of the events that occur during embryogenesis with regard to the inner, middle and external ear is requisite to understanding the trends that occur in atresia.

The inner ear, including all of the components of the membranous labyrinth, develops from an invagination of ectoderm, the otic placode, at approximately 3 week's gestation, and achieves adult dimensions at midterm. Further elaboration on inner ear development and abnormality is the subject of Congenital Sensorineural Hearing Loss.

The middle ear space, including the mastoid air cell system, the Eustachian tube and the inner surface of the tympanic membrane derive from endoderm of the first pharyngeal pouch. Most of these elements complete their development by the 30th week, excepting the mastoid system in which pneumatization can progress well into adulthood. Due to the late development of the mastoid system, several authors have pointed out that patients with atresia who maintain a well-developed mastoid generally manifest greater development of their middle ear space and ossicles, thus making them better surgical candidates.

Ossicular development, which progresses from the 8th through the 26th week, derives from the mesoderm of the first and second branchial arches. The malleus head and body and the short process of the incus arise from the first branchial arch (Meckel's) cartilages. Second branchial (Hyoid) arch derivatives include the manubrium of the malleus, the long process of the incus and the stapes superstructure. The footplate derives from both the second arch and the lamina stapedialis of the otic capsule. Ossification occurs through endochondral bone growth. Release of the ossicles except at their ligamentous attachments occurs by endodermal resorption as it gives rise to the tympanic cavity.

Ectoderm of the first branchial cleft gives rise to the external auditory canal, which develops as an invagination at the site of the future auricle at the fourth gestational week. By the eighth week, a solid core of epithelium arises and extends to the area of the middle ear space, separated from it by a thin layer of mesoderm. At approximately 28 weeks, this core begins to recanalize from medial to lateral until the surface ectoderm is reached giving rise to the external auditory canal. At its medial extent, ectoderm persists as the outermost layer of the TM, with the mesodermal layer reduced to a fibrous sheet interposed between outer ectoderm and inner cuboidal endoderm.

Mesoderm from the tympanic ring serves as the primary impetus for the formation of the tympanic bone.
and the osseous portion of the external auditory canal, starting at about the 12th week. Finally, the auricle is the result of the fusion of six hillocks (His) from the first and second branchial arches, which is usually complete by the 12th week as well. Malformations are graded I through III, with Grade I manifesting as a small, but well-formed auricle; Grade II representing a recognizable auricle with varying degrees of anomaly; and Grade III, which describes a rudimentary appendage or none at all. Auricular anomalies are the subject of Microtia.

Congenital aural atresia has a reported incidence that varies between 1 in 10 to 1 in 20,000. Congenital aural atresia is usually found in isolation and in these instances patterns of inheritance have not been seen. Its association with several known craniofacial syndromes is well known. Aural atresia has been found in association with 22 of the 71 known craniofacial syndromes. The most common of these are Treacher-Collins (mandibulofacial dysostosis), Nager (acrofacial dysostosis), Crouzon's (craniofacial dysostosis), hemifacial microsomia, and the first and second branchial arch syndromes including Goldenhar's (oculoauriculovertebral dysplasia). In unilateral cases, Jafek et al and others have cited an increased incidence of atresia on the right. Bilateral atresia has been reported to occur in approximately 30% of cases.

Establishing a classification system of the congenital ear malformations has been almost as difficult as the operative procedures themselves. Several schemes have been proposed over time, one the first being that proposed by Marx in 1926 for auricular anomalies, which were graded I - III for mild to severe deformities. Altmann in 1955 proposed the first schema based on the temporal bone itself, looking at the status of the canal, tympanic bone, drum and ossicles. Lapchenko in 1967 and Gill in 1969 both set up a four tier scale, examining the degree of middle ear and external canal development, and the presence of ossicular abnormality. Additionally, Gill incorporated into his system, the degree of pneumatization of the mastoid, as this seemed to predict the relative success of operative interventions. Ombredonne and Nager and Colman each tried to establish simplified and clinically practical systems based on Altmann's original scheme, but these were inconsistent at predicting outcome in those cases of major aplasia or group II anomalies. Jahrsdoerfer in 1992 established not so much a classification system, but rather a grading scale as a means selecting patients that would most likely benefit from attempts at repair of their atresias.

This system of grading allows for a quantitative analysis of the temporal bone and those structures that are considered vital to the success of an operation. Whereas prior systems were excellent descriptions of surgical and anatomical findings, they lacked predictive power and were subject to interobserver bias. This scale, which is based on temporal bone CT findings, assesses nine different parameters that are used in making the determination of candidacy for surgery. Of note, the stapes and oval window complex account for 3 out of the 10 points possible in the scale. Other parameters include the middle ear cleft, facial nerve position, status of the ossicles and round window, and pneumatization of the mastoid. Scores of 6 through 10 range from marginal to excellent candidates for surgery while a score of 5 or less usually anticipates a poor outcome.

In general, patients with congenital aural atresia are seen in referral from neonatologists in those cases where gross craniofacial anomalies are present. However, many cases of isolated atresia or even those
with very mild head and neck syndromes may be delayed until later in development and may be referred by pediatricians for either atresia or for problems relating to a stenotic canal. As is usual, a thorough history is important in the initial evaluation of these patients, including questions relating to the gestation of the infant. Specifically, issues of drug utilization, toxic exposures, prior family history of hearing impairment and developmental craniofacial disorders and maternal infections should be probed as possible etiologies.

A thorough physical exam of the head and neck as well as of other organ systems should be performed, especially the spine, extremities and genitourinary systems as these develop concurrently with those of the head and neck. Associated anomalies of the midface and mandible should be noted. Position of the auricle and the degree of external canal development should be checked. The degree of mastoid prominence should be noted as this may give an early insight to the operative potential of the patient. Cranial nerve function, especially that of the facial nerve should be assessed.

An audiologic evaluation is essential in the initial assessment of these patients. A screening ABR should be obtained for several reasons. The first is to establish the presence of a functional inner ear. In very young patients this is accomplished with multichannel air and bone conduction ABR. In cases of bilateral atresia, this form of ABR will also allow one to establish with greater certainty the ear with greater cochlear function by examining the response obtained in wave I, which is indicative of ipsilateral cochlear status only. The second reason for early ABR is that if an intact inner ear is present, the child can be fitted early on with a bone-conducting device. The usual finding is that of moderate to maximal conductive loss, although mild conductive losses are sometimes seen in cases with membranous atresia. A third reason for early ABR is to assess the functional status of the contralateral ear in unilateral atresias. Several cases of sensorineural or mixed losses in the contralateral, normal-appearing ear have been reported and should be ruled out. Having ascertained the functional status of the inner ear, diagnostic imaging is used to assess the degree and nature of the temporal bone deformity.

With CT scanning, an assessment of the ossicular mass, the nature of the atretic plate (whether bony or soft), the position and course of the facial nerve, the degree of external canal development and mastoid pneumatization is possible. The presence of a cholesteatoma, which has a slight predilection for stenotic ears, can also be ruled out. Current recommendations for the timing of CT scan is at approximately 4 years when mastoid pneumatization is most complete. A review of the literature by Cressman et al has shown that approximately 50% of patients seen for this condition are ultimately found to be candidates for reconstruction based on CT scan findings.

The current consensus among those who undertake these procedures is that surgery on cases of either bilateral or unilateral atresia should be deferred until the patient is at least 4 to 6 years of age, with an additional delay to consenting age espoused by others for those cases of unilateral atresia. The need to foster continued mental development and the evolution of speech skills requires that interim bone conduction appliances be provided for these patients with bilateral atresias that have documented cochlear function on ABR. Unilateral atresia patients usually do not require any intervention as long as normal hearing is present in the contralateral ear.
Once the decision to proceed with surgery has been made, a consideration of both the functional and cosmetic aspects of the operation needs to be taken together. Close coordination with the plastic surgeon reconstructing the auricle is needed and usually takes place as a multi-staged procedure. Because of problems relating to healing in a fibrotic, previously-operated area, current practice is to allow the plastic surgeon to operate first with the implantation of an auricular framework of autogenous rib graft in a subcutaneous pocket. Stage II consists of lobule transposition and remnant excision. Correction of the atresia with the creation of a new external meatus, canal, drum and conducting mechanism in continuity with the stapes and inner ear is completed in Stage III along with alignment of the new auricle to the meatus. Stages IV and V usually consist of the creation of a new tragus and the elevation of the auricle off the post-auricular skin.

Contrary to the standard practice of operating on the poorer hearing ear first, as is the case with chronic ear infection, cholesteatoma and otosclerosis, the better hearing ear is operated on first in CAA as this will afford the best chance for an optimal outcome. Two general approaches to the repair of atresia are currently described, both of which commence with postauricular incisions. The first of these is the transmastoid approach that makes use of the familiar landmarks of the tegmen and sinodural angle to approach the mastoid antrum and the bony atresia plate. This dissection is conducted with the aid of a facial nerve monitor, as the course of this nerve is commonly aberrant owing to the anomalous development of the tympanic bone and mastoid. Some surgeons opt to perform incudostapedial dislocation if this is feasible in order to prevent transmitted acoustic trauma to the inner ear with resultant sensorineural loss. Once the plate is removed, a canal wall down mastoidectomy is completed leaving an open cavity.

A second, newer, approach described by Jahrsdoerfer and known as the Anterior Approach involves exposure of the tympanum and ossicular mass by directly removing bone from lateral to medial between the glenoid fossa and the middle cranial fossa dura. Drilling begins just posterior to the condyle, and proceeds medially using the middle fossa dura as a superior guide. An anterior and superior approach presents the least risk in injuring an anomalous facial nerve. This usually leads to an ossicular mass consisting of a fused incus and malleus attached to the atretic plate. Once identified, the atretic plate can be taken down with diamond burrs and curettes, taking care to avoid contact with the ossicular mass. Mobility of the stapes and the status of the oval and round windows is checked. Middle ear reconstruction with a prosthesis is performed if necessary. Otherwise, temporalis fascia and split-thickness skin grafting is performed to create a new eardrum and line the newly-created canal. A meatoplasty completes the procedure and places the canal in continuity with the auricular meatus. Modification of this approach has been described including a canal wall-up approach that allows the surgeon to identify the nerve and atresia plate prior to opening a canal.

The goal of attaining hearing improvement in an ear with a previously moderate to maximal conductive loss varies from persistent maximal conductive loss to cases of near-total closure of the air-bone gap. Although different parameters for successful improvement in hearing have been reported, according to Cressman et al, if one chooses a speech reception threshold of 25 decibels or less, then anywhere from 20 to 80% of patients will have a successful operation. Much of this range hinges on patient selection,
experience and technique. Crabtree, de la Cruz and Glasscock have each reported varying degrees of success with this operation. However, this may reflect the utilization of different parameters and definitions by each of these authors. Jahrsdoerfer reported in 1992 on his results based on patients selected using a 10-point grading system. He reported achieving speech reception thresholds of 25 decibels or less in 75% of 86 cases operated on. Several authors have noted that cases requiring a prosthesis for ossicular reconstruction tend to have greater air-bone gaps postoperatively than those receiving grafts over a mobile ossicular mass.

Complications relating to CAA repair can be divided into intraoperative and postoperative categories. Intraoperative complications include facial nerve injury, sensorineural deafness, and perilymph fistula. Although cited as a potential complication, permanent facial paralysis is exceedingly rare despite its aberrant course through atretic temporal bones. High frequency sensorineural loss secondary to transmitted vibrations has been reported with both approaches. Postoperative or delayed complications for the most part include stenosis and chronic drainage, more common with the transmastoid approach, and graft lateralization that plagues both approaches as well. This can be prevented by the creation of a bony ledge medial to the ossicular mass wherein the graft can be placed, or through the use of a silastic button just lateral to the graft.

Bilateral atresia patients that are either unable or unwilling to undergo surgery have the option of being fitted with a bone-conducting hearing appliance. Both percutaneous and transcutaneous devices have been developed; however, only the transcutaneous version is FDA approved. Older devices that are held to the head by a tight-fitting headband are unsightly and uncomfortable.

Despite the advances that have been made in technique and imaging over the past twenty years, several issues regarding the timing of surgery in general, when to operate on cases of unilateral atresia, and the operability of cases with severe craniofacial malformations remain difficult questions. The issue of when to operate on these patients is a difficult one because many surgeons with considerable experience in this area maintain such varied opinions. Generally speaking, most would agree that the earliest age at which to proceed is at 4 to 5 years of age, thereby allowing time for adequate mastoid and middle ear pneumatization and increasing patient compliance with the postoperative care that is required. Also, most authors state that in cases of unilateral atresia with evidence of cholesteatoma, infection or with very thin atresia plates, surgery should be undertaken earlier. However, there are differing opinions regarding cases of grade II and III unilateral atresia in patients with normal hearing in the other ear. Jahrsdoerfer finds that the benefit of binaural hearing far exceeds the risk of facial nerve injury and other complications. De la Cruz also favors operating earlier on unilateral atresias if a good outcome can be expected based on CT findings. Others, including Crabtree, Bellucci, Glasscock and Fisch, favor waiting until the patient is of consenting age at which point an individually informed decision can be made. All points considered, the decision to proceed with early intervention ultimately depends on the experience of the surgeon and the relative degree of malformation that the individual patient has and with which the surgeon is comfortable.

In summary, congenital aural atresia is a congenital developmental anomaly of the middle ear that manifests with varying degrees of external auditory canal stenosis or atresia, ossicular derangements,
poorly developed mastoid and tympanic cavities and is characterized by a conductive hearing loss that is of a moderate to maximal degree. Heritable forms of CAA are found in association with many of the craniofacial syndromes, but nonheritable, isolated CAA is more frequent. History and physical exam findings are important in the early identification of the problem complemented by audiometric/ABR evaluation in order to allow speech development. CT scanning to assess operative potential is then performed at about 4 years of age, with an approximate yield of 50%. Grading of the patient based on CT findings has been shown to be predictive of outcome. Success rates of as high as 75% have been reported with speech reception thresholds of 25dB or less. Historically, a transmastoid approach has been used, but the problems of stenosis, drainage and long-term care of a mastoid cavity have favored the anterior approach. Issues regarding the timing of unilateral cases are complex and the decision to operate early on in these cases ultimately depends on the experience of the surgeon and the degree of malformation present.

Case Presentation

A 2-year and 9-months-old male, former 36-week premature infant that at birth was noted to have midfacial and mandibular hypoplasia consistent with the diagnosis of Treacher-Collins Syndrome. The family history was negative for relatives with craniofacial or auricular malformations. His past medical history was notable for uncomplicated neonatal jaundice. Medical work-up for congenital cardiac and renal anomalies was negative. On exam, the patient is noted to have malar hypoplasia, absent zygomatic arches, hypoplasia of the mandible with Class III occlusion, and bilateral Grade III microtia with aural atresia. The mastoids, however, are well developed bilaterally. A tracheotomy is present in the neck. The remainder of the physical exam was unremarkable. Upper airway obstruction secondary to severe mandibular hypoplasia necessitated elective tracheotomy in the first week of life. A gastrostomy tube was placed secondary to poor oral intake. At two months of age, he underwent audiometric evaluation consisting of air and bone conduction multichannel ABR. The patient was fitted with a bone-conducting type hearing aid at age four months.

Subsequently, the patient underwent repair of his palatal defect at age two, and most recently underwent bilateral Ilizarov mandibular distraction procedures. After more than two years with his bone-conducting aids, he has acquired comprehensible speech and a vocabulary comparable to that of children with normal hearing.

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CONGENITAL INNER EAR MALFORMATIONS

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The reported incidence of significant sensorineural hearing loss in young children varies from 1:1000 to 1:2000 depending on the population studies. Approximately 20% of patients with congenital sensorineural hearing loss have radiographic anomalies of the inner ear.

The structure of the inner ear consists of a membranous labyrinth surrounded by a bony labyrinth within the petrous temporal bone. The primordium of the membranous labyrinth, which is referred to as the otic placodes, appears in the third week of gestation as thickened planes of surface ectoderm on either side of the developing hindbrain. It differentiates into the otic pit and then fuses to become the otic vesicles, or otocyst, by the fifth week. The otocyst subdivides into two pouches. The ventral portion develops into the cochlear duct and saccule. The dorsal segment is transformed into the endolymphatic sac and duct,
utricle, and semicircular canals. By the 10th week, the adult membranous labyrinth is recognized, and completely developed by week 26. The bony semicircular canals began at the sixth week and are complete by week 22. The superior semicircular canals are completed first, followed by the posterior and finally the lateral. The bony labyrinthine begins in the fourth week as a condensation of mesenchyme and forms a cartilage capsule around the developing membranous labyrinth. Its ossification begins at week 14 and is completed by week 23. Congenital ear malformation results from a defect in the development of the membranous labyrinthine, the osseous labyrinthine, or both.

The malformations limiting the membranous labyrinthine cannot be detected radiographically. Complete labyrinthine dysplasia is very rare, and was first described by Bing and Siebenmann in 1907. It has been associated with Jervell-Lange Nielson and Usher's syndromes. Cochleosaccular dysplasia was first described by Schieber in 1892. It is characterized by a collapse of the cochlear duct and saccule. It is probably the most common form of inner ear pathology in patients with congenital deafness. The Alexander's ear deformity, or the cochlear base turn dysplasia, is related to familial high frequency sensorineural hearing loss.

Jackler has classified cochlear anomalies based on inner ear embryogenesis. The complete labyrinthine aplasia, known as Michel's deformity, represents an early failure in development correlating to the third week of gestation. It is extremely rare. The common cavity deformity represents developmental arrest at the fourth week, and a common cavity of the cochlea and vestibule is formed with internal architecture. Cochlear aplasia results from the arrested development of the cochlea during the fifth week. The cochlea fails to form and appears as a single cavity. The vestibule and semicircular canals may be normal or malformed. Cochlear hypoplasia displays a small, rudimentary cochlea but is associated with a normal or malformed vestibule and semicircular canals. This lesion is due to an arrest at the sixth week of gestation. Incomplete partition deformity, also well known as Mondini's deformity, represents a small cochlea with incomplete or no intrascalar septa. The cochlea is usually flat and has one and one-half turns instead of the normal two and one-half turns. Arrest of maturation at the gestational seventh week may result in the Mondini deformity.

The internal auditory canal (IAC) may be enlarged or stenosed. The narrow IAC can be associated with a failure of the 8th nerve development. Patients with the wide IAC may be predisposed to cerebral spinal fluid (CSF) leaks, resulting in recurrent meningitis.

Patients with the Mondini deformity and other congenital inner ear malformations are at an increased risk for developing recurrent meningitis or perilymphatic fistula. They are predisposed to develop a CSF leak due to the enlarged cochlear aqueduct or an abnormal connection between the internal auditory canal and the membranous labyrinth.

A thorough clinical, audiological and radiological evaluation should be made of all patients suspected of having these deformities. Clinical history should include possible exposure to teratogen during pregnancy, family history of hearing impairment, progression and fluctuation of the hearing loss, and associated vestibular symptom. A routine audiologic evaluation is required. Work-ups can be helpful in determining possible etiology. These include TORCH titers, FTA-ABS, urinalysis, and thyroid function...
tests. High resolution, computed tomography provides excellent visualization of the bony labyrinthise of the inner ear.

Patients are advised to avoid contact sports because of the increased risk of CSF leak following minor head injury. Middle ear infections are treated aggressively because of the increased risk for meningitis. Genetic counseling is provided after a careful analysis of the family history. Hearing rehabilitation, including amplification and especially educational efforts, are also indicated.

Currently, cochlear implantation is indicated for bilaterally profound sensorineural hearing loss without speech discrimination using hearing aids. It is contraindicated in the patient with complete cochlear aplasia and narrow IAC since neural elements for stimulation would be absent. Further study is required to evaluate the long-term benefits of cochlear implantation in the congenital inner malformation.

Case Presentation

A patient was referred to Texas Children’s Hospital at eight months of age with a history of developmental delay and no perception to auditory stimuli. She was a term infant, weighing 7 lbs 11 oz when delivered by Caesarean section due to breech presentation. Her postnatal care was uncomplicated. The pregnancy was only complicated by a urinary tract infection and hypertension during the third trimester. She was recently diagnosed with congenital hypotonia, but was otherwise healthy without any previous ear problems. An ABR demonstrated no response to air or bone conduction click stimuli.

Her family history was only significant for a maternal grandmother with a hearing loss. Physical examination was unremarkable. Titers were negative for cytomegalovirus, rubella, toxoplasma, and herpes simples virus. Thyroid function tests were within normal limits. A high resolution temporal bone CT scan revealed cochlear common cavity deformity.

She was fitted with bilateral behind-the-ear (BTE) hearing aids. Repeat audiogram demonstrated her aided speech detection threshold to be 60 dB, and a 1000 Hz warble tone at 70 dB. She has been involved in a total communication program. Her parents noted significant differences in her sound perception with hearing aids. Currently she has been using a five-word vocabulary.

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Diving Medicine
Brian H. Weeks, M.D.
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There are about 3.4 million certified scuba divers in the United States. And about 300,000 become certified per year. About one-third of all divers experience some diving related medical problem in their careers. The important thing to remember is that even in healthy divers with safe diving profiles, there is a 1% to 5% incidence of decompression sickness, which can be an extremely serious problem.
The most common diving related injury involves middle ear barotrauma. The incidence of vestibular symptoms reaches about 20% among divers in some studies. Most importantly for this talk, as an otolaryngologist we are considered the diving experts. So it is quite probable that sometime in our career we will all see a patient who is a scuba diver.

In literature there are many referrals to breath-hold divers used by kings to recover treasures for their wealth and fame. In the 17th century an open diving bell was documented, which was an air-holding chamber that prolonged possible diving times. In 1810 a copper helmet was designed, which was sealed to a watertight canvas. This allowed air to be pumped down from the surface. Today we have a modification of this original design for very deep hard-hat diving.

In early 1940 Jacques Cousteau designed what was then referred to as the aqua-lung. Today it is known as the scuba device. Scuba is an acronym for self-contained underwater breathing apparatus. These were actually initially used in World War II, but because of their relatively low cost and their reliability, the apparatus was made available to the everyday diver and for recreational purposes.

For those unfamiliar with scuba diving, the basic set up involves a tank, which the diver wears on his back, that delivers pressurized air via a hose and a mouthpiece, which is known as a regulator. The pressure is comparable to the aquatic depth that the diver obtains. A demand regulator either at the tank or at the mouthpiece controls the differential on the pressure from the tank air to the ambient pressure. The duration of the dive is limited to the amount of air in the tank as well as to the physiology of the diver.

The weight of the air at sea level is one atmosphere, so atmospheric pressure is sea level, which is described as one square inch of air for as far as the atmosphere extends. Atmospheric pressure is also recorded as 14.7 lb. per square inch or 760 tore. One atmosphere under water is 33 feet of seawater, which is the equivalent of 34 feet of fresh water. So, there is one-foot difference in the amount of an atmospheric pressure. Probably the most important concept to understand is that when a body is submerged in water, it not only experiences the pressure from the water, but it also experiences the pressure from the air, which is called absolute pressure - atmospheric plus water pressure.

As divers, we refer to the gauge pressure, which is really the pressure that
is important to us and is the pressure that is exerted by the water alone. Just for comparison, 18,000 feet into the atmosphere in an airplane is the equivalent of a half of atmosphere. This gives a perspective as to what we are looking at from a diving standpoint. As you descend to deeper depths, 33 feet, 66 feet, your absolute pressure is the air pressure plus the water pressure.

Basic laws govern scuba diving. Probably the most important law to us as otolaryngologists and divers is Boyle's Law. This law says that at constant temperature, volume and pressure are inversely related. Since we deal with air-filled cavities, this is clearly the most common law relating to us as otolaryngologists. Remember that soft tissue is mostly made of water and is relatively incompressible compared to air-filled spaces. However the sinuses and the middle ears are all affected by pressure changes.

Again, a scuba diver breathes air through his tank, through his glottis, which is delivered to him at one atmosphere pressure. As the diver descends deeper into the ocean, the pressure increases but the lung volume remains constant. However, even at deeper depths, inspiration becomes more difficult because as the density of the molecules changes, it becomes more difficult to breathe the air. As the diver descends, the pressure increases in the lungs because depth pressure increases. However, the volume stays the same. The volume is constant, the density changes and the pressure changes. The fact that the lung volume stays the same is what protects the lungs while we are diving.

Breath-hold diving or free diving is a very common sport especially in Hawaii and along the West Coast. Even snorkeling involves diving using only mask, fins, and a snorkel. Most of these divers are limited to 1-2 atmospheres of gauge pressure in their depth. Again the thing to remember with these divers is that their lung volumes will be changing because they are not using a scuba apparatus to equalize their pressures. At one atmosphere, the lung volume goes down by \( \frac{1}{2} \) and at two atmospheres their lung volume is reduced by \( \frac{1}{3} \) again. Those are important concepts to remember.

The second law is Dalton’s Law, which basically says that in a gas mixture, pressure exerted by each gas is the same as it would exert if it alone occupied the same volume. So, as you increase the total pressure, the relative contributions of each gas in the mixture remain the same. If oxygen is one-third of the mixture at sea level, even if you decrease or descend to three atmospheres pressure, it still would have an equal contribution. The contribution of oxygen triples to the total mixture. This is extremely
important with the biological effects of gas. The partial pressures of each individual gas in the tank determine the tissue diffusion and the amount of gas that is ultimately dissolved in the body tissues and in the blood stream. Remember that oxygen delivery depends on PO2 and not on total gas pressure. A good example to illustrate this is that at sea level the PO2 is 16 tore, which is 21% of 760 tore. At 132 feet of seawater, which is four atmospheres, the PO2 is equivalent to breathing 100% air at sea level. This shows that even though it has the same amount of concentration at that depth and the relative contribution is the same, because the pressure is so much greater, the effects are quite different. This can lead to oxygen toxicity, because breathing the equivalent of 100% oxygen is very difficult and very damaging to the body over long periods of time.

The last law of Physics to consider is Henry's Law, which basically says that given temperature, the mass of the gases dissolved, and the given volume of solvent is proportional to the pressure of the gas with which it is at equilibrium. Basically, this law dictates how much of a gas is absorbed at a given pressure. This is important when we discuss things like nitrogen narcosis and decompression sickness. Again, this is most important when applied with nitrogen solubility in the body tissues during descent and then the relative insolubility when ascent is performed. Nitrogen narcosis is often called The Rapture of the Deep." And was first described by Captain Albert Binky, a seaman. He learned that nitrogen under pressure acts as an anesthetic. At greater depths, there is a greater force driving nitrogen into the tissues. This is an application of Henry's Law.

Divers themselves refer to another law: Martini's Law. This law states for every 50 feet of descent, a diver breathing tank air experiences the equivalent of drinking one martini on an empty stomach. At dives of 100 feet of seawater, or even down to close to 150 feet of seawater, the effect can be like drinking three mixed drinks with no food in your stomach. This can become extremely dangerous especially when the diver needs to make quick decisions and needs to determine exactly what his dive bottom times should be. Commercial divers who dive much deeper than recreational divers, often substitute helium in their gas mixtures for nitrogen in order to limit the anesthetic effect and to allow them to dive deeper without intoxicating effects.

Ears, sinuses, and the neighboring structures are all susceptible to injury during diving. The ear and the Eustachian tube are considered to be the weak links in our body's ability to tolerate diving situations. The nasopharyngeal opening of the Eustachian tube is usually closed. As we all know, it opens with positive nasopharyngeal pressure or by contracture of a
number of muscles: tensor valle of palantini, lavatar palantini, and the palato pharyngeus muscles contract and open the nasopharyngeal orifice of the Eustachian tubes.

In 1937 Armstrong and his colleagues did a very interesting study of something that is probably obvious to us now. Upon descent with divers he found that the Eustachian tube acts as a flutter valve and actually is closed and only opens if the diver equalizes. The Eustachian tube will not open on its own under these pressures to equalize middle ear pressure. It has to be reflexive or voluntary, an act made by the diver himself to ensure that his middle ear pressure is equalized throughout his dive. If pressure is not equalized, the diver experiences what is commonly called a middle ear squeeze, which refers to barotrauma of the middle ear. A Valsalva, which involves holding the nose, closing the mouth and exhaling against a closed glottis, is one of the techniques used to re-inflate or auto-inflate the middle ear and equalize the pressures. If divers have upper respiratory infections, allergies, nasal polyposis or septal deviation, they increase their risk of middle ear squeeze because of their inability to equalize their pressures.

The most common or simple problem that can occur is occlusion of the EAC, which can be related to cerumen, ear plugs, or a hood used in cold water diving. The diver experiences pain during descent because of the pressure build-up due to the occlusion. On examination of these patients, we see congestion of the external auditory canal skin and often of the TM as well, with edema in the canal. There may be rupture of the skin and even a tympanic membrane perforation from this problem, which will cause hemorrhage and severe, severe pain. Treatment is conservative: stay out of the water and do not scuba dive until the area is completely healed.

Probably the most common problem involving the EAC with scuba divers is otitis externa. This is often due to the water exposure or divers drying their ears excessively with cotton after a dive. It is a fairly simple problem, and for a mild case, the patient should use acidified alcohol solution after dives, often called Swim Ear. There are also silicone oil sprays that divers can apply to the EAC skin before dives, providing a protective coat.

The most common problem in the middle ear is barotrauma, also known as aerotitis media or middle ear squeeze. When scuba divers make their descent, the most critical time is within the first atmosphere. It is most important that pressure is equalized properly. If pressures are not equalized, the diver will experience severe otalgia. This can lead to subsequent tympanic membrane rupture. This can actually occur at even just a few feet of water and in pressure differences as low as 5 pounds per
square inch. With a tympanic membrane rupture, there is often otorrhea and the diver frequently experiences dizziness and some form of a mild hearing loss. At surface pressure all pressures are equalized. As a diver descends, the ambient pressure increases and the pressure in the middle ear should be equalized through the Eustachian tubes. However, if there is a block in the Eustachian tubes, the diver can no longer equalize his pressures. If he continues to submerge or to descend, this pressure will increase and eventually the weak link in the system will give, which means that the tympanic membrane will likely rupture. If the diver chooses to return to the surface he can do so. However, he will continue to experience the problem and the situation is often even worsened because small amounts of oxygen are actually absorbed through the middle ear, further increasing the negative pressure differential in the middle ear and causing further pressure. Mucosa edema of the middle ear occurs and there is hemorrhage into the small capillaries. Treatment involves the use of systemic decongestants and mucolytic agents plus or minus an antihistamine for patients experiencing allergy. The otolaryngologist should also instruct the diver to perform periodic auto-inflation if he is not having pain without movement. If continued pain occurs, it usually indicates a middle ear infection. At that time, a decision needs to be made whether the patient would benefit from a myringotomy or PE tube and certainly antibiotics. With a tympanic membrane perforation, the original treatment is the same as always: with oral hygiene and ototopical agents, these usually close nonsurgically. However, tympanoplasty is indicated for persistent perforation.

Divers should be encouraged to concentrate on prevention of these injuries. A nasal decongestant before a dive can help prevent this sort of problem. When these medicines are applied to the nose, the position of the head is very important. The diver should actually have his head down so that the medicine can reach the Eustachian tube ostium more easily. Some divers use systemic decongestants, such as Sudafed, and often add an antihistamine for allergy. It is very important to remember that these should be non-sedating medicines and that no new medicines should be given before a dive unless that have already been tried on the surface. And, as we have all seen before, Afrin can even have rebound hypercongestion, which can lead to nasal obstruction. Because this is a delayed phenomenon, it can happen at depths of maybe 60 or 70 feet below the surface in the middle of the dive. This can lead to what is known as a reverse squeeze. That is expansion of the middle ear air, which can lead to TM distention and perforation, but through the opposite direction - an increase in the middle ear pressure.

Some divers begin Valsalva and auto-inflation in the middle ear before they
even begin their dive. With a very forceful Valsalva, a diver can experience barotrauma. This is related to an increased intrathoracic and abdominal pressure, which ultimately increases the CSF pressure via engorged spinal veins, which can be transmitted to the endolymph, the cochlea, perilymph, and eventually to the round window and oval window membranes. This leads to tinnitus and usually a unilateral high frequency hearing loss.

Another preventative measure is the Frenzel maneuver, which involves holding the nose, closing the glottis and contracting the pharyngeal muscles. This also forces air into the Eustachian tubes. Some people are able to perform this to equalize pressure and don't have to use a Valsalva technique. Other preventive measures include using a feet-first descent. Multiple studies have shown that it is easier to inflate the middle ear with the head up, as this causes less hyperemia of the Eustachian tubes and mucosa. The diver is also less distracted with his head up, and it may help him to remember to continuously auto-inflate his middle ear every 2-3 feet as he makes his descent. An anchor line on the boat should also be used to verify exactly how deep the diver is. If a diver experiences middle ear squeeze, he should ascend a few feet and attempt to auto-inflate, and then begin the redescent. It is also important to have a form-fitting mask, so that nasal compression is easier to perform. Jaw and head movements from side to side are often used to facilitate middle ear inflation.

One of the more uncommon problems seen in divers is what is known as ultinebaric facial paralysis. This is extremely uncommon. This is due to dehiscent portions of the facial nerve, which are vulnerable to barometric trauma. It is usually seen in normal people who descend during a dive, but when they ascend and get to the surface, they have a facial paralysis. This is almost always a transient paralysis and is related to barotrauma. These divers almost always resume normal function of the facial nerve over time.

Inner ear problems in divers are usually related to middle ear barotrauma. As we have discussed, middle ear negative pressure causes a depression of the tympanic membrane. This can be transmitted to the ossicle of the stapes and cause significant and rapid foot plate depression, which can tear the sensitive inner ear membranes and lead to the types of hearing losses we have discussed. The second thing that can occur in the inner ear is sudden pressure equalization, which can cause a rush of air up the Eustachian tube, putting an outward force on the tympanic membranes and the ossicles and pulling the stapes footplate up. This can cause a pressure wave in the inner ear and distort the inner ear membranes. All these membranes are extremely fragile. There are multiple theories as to how the trauma occurs. But it is thought that either Ricener's membrane or the
basilar membranes are involved or the vestibule and semi-circular canals are involved. The shearing force of this rapid fluid movement is transmitted to the labyrinth and cochlea and can cause tears in the membranes and hemorrhage from the small torn vessels.

Round window implosion is a well-described problem in the literature from inner ear trauma during diving. This involves a pull on the stapes footplate and a shock wave through the scala vestibuli to the scala tympani. It causes a bulging inward from a rapid pull on the stapes footplate and it causes an inward pull on the round window membrane, which causes an implosion injury and the leakage of perilymph into the middle ear. A sudden pull on the stapes footplate can also lead to a tear of the annular ligament of the oval window, leading to a crack in the footplate and also leakage. Frequently when divers are having a difficult time auto-inflating, their first inclination is to try harder to equalize the pressure. This can lead to the elevation of the CSF pressure that can be transmitted directly through a patent cochlear aqueduct into the internal meatus, raising intracochlear pressure to dangerous levels. The difference between the middle ear and the perilymph space can cause an explosive outward rupture of the round window membranes. It is the opposite of the implosion injury, an outward injury, which can cause perilymph leakage and a perilymph fistula.

Dr. Healy and his colleagues did a study that looked at 40 cases of perilymph fistula in divers. The patients' chief complaints with this problem were episodic positional vertigo. They did not have as many complaints of hearing loss or tinnitus. These patients all underwent surgical exploration. At exploration, 31 out of 40 of these patients (75% of them) had oval window leaks, 5 of them had round window leaks, and 4 of them had both oval window and round window leaks. None of these patients had a hearing loss for longer than 3 weeks. They regained any significant hearing in those ears. If the suspicion is high for a perilymph fistula, early exploration leads to the best hearing restoration. Healy felt that the most important factors during these procedures were proper magnification, and patience. He commented that after blotting in the middle ear, it often took 5 to 10 minutes for perilymph to build back up or for a leak to be revisualized. In two patients he had to perform bilateral jugular vein compression to enhance the perilymph leakage.

To summarize, hearing loss and dizziness are indicative of inner ear barotrauma and require immediate treatment. Depending on the etiology, or presumed etiology, treatment involves strict bedrest and convalescence, no coughing, no sneezing, auto inflating or straining. These patients all deserve an audiogram to determine whether this is a middle ear or an inner
ear problem. With inner ear problems with a cochlear etiology, you will see a non-progressive centrally neural hearing loss. And, as I said, with a perilymph fistula one way to distinguish it is that there is often a progressive and fluctuating hearing loss. In the study by Healy, he also found that posturography with EAC pressure changes have a 97% sensitivity for diagnosing a perilymph fistula.

Antonelli and his colleagues did a study looking at 18 temporal bones examined from 11 divers who died from complications during diving, either from rapid ascent or drowning. He found that bleeding into the middle ear and mastoid air cells was nearly universal in all these divers. The most common damage seen was bleeding around Ricener's membrane and the round window membrane. He also found that a number of these divers, usually the ones who had rapid ascent injuries, had rupture of both the utricle and saccule and that tympanic membrane rupture from rapid ascent was far more common in people who dive than had been presumed. The other interesting point was that most of the inner ear damage was not surgically treatable. Other than for the perilymph fistula, most of these injuries do not have surgical treatments.

Lastly, Zannini and his colleagues followed 160 professional divers with serial audiograms. He found that these 160 divers had significantly worse hearing than control divers. The greatest loss was in the group with the longest diving times. He found that their hearing losses were in the highest frequencies, usually in the 8 kilohertz. Divers who reported difficulty auto inflating their middle ears had even worse hearing or the most central hearing loss. Zannini also commented that perilymph fistula demands early surgical exploration and that inner ear barotrauma is managed medically with vasodilators, steroids, histamines, and Carbogen.

There is a different set of problems that divers can experience during descent which involve the sinuses. This is called aerosinusitis or sinus squeeze and involves osteomuculusion and pressure equalization. It is really infrequently an issue in the sinuses but if you have a demitasse mucosa with an upper respiratory infection, an allergy, a polyp occluding the ostium or a septal deviation, this can cause a problem and mimic what happens in the middle ear. You get a negative pressure in the sinuses because of the inability for the pressures to equalize. This causes a vacuum, which can lead to mucosal edema, hemorrhage and severe pain. Management includes sinus irrigation and either endoscopic sinus surgery, if it is a polyp, or septoplasty to straighten the septum.

Tooth squeeze or aerodontalgia is a very uncommon problem. The most
common cause of tooth pain in the diver is from aerosinusitis. But aerodontalgia can be caused by an air pocket underneath the tooth cap or filling. It causes severe pain when the diver descends and the pressure changes. To treat this condition, dental work should be done and the filling or cap should be replaced.

Facemask squeeze is failure to equalize pressure changes in the facemask. It can lead to tissue damage from the pressure differential in the mask along the soft tissues. The most common areas of injury are around the eyeball and the lining of the eyelids where the mucosa is quite sensitive. Exhaling through the nose and into the facemask and equalizing the pressure that way easily prevents facemask squeeze.

Epistaxis is bleeding from the nose into the mask underwater. It is a very common problem. The bleeding is due to pressure changes on Keisselbach's plexus. Oftentimes the diver will not even recognize that he has had a nosebleed. As you move underwater, the selective absorption of color by the blue green seawater makes blood appear blue green. So oftentimes the diver will ascend to the surface and then realize that his mask is full of blood. There is no treatment required. Almost all of these invariably resolve within minutes of reaching the surface just through external compression on the nose.

Alternobaric vertigo is another problem seen in divers. This is transient vestibular dysfunction due to dysequilibrium of pressures in the two middle ears. It can lead to vertiginous feeling and has been seen in up to 15% to 16% of divers in some series. When you are underwater, any sort of incoordination or change in equilibrium can be extremely dangerous. These patients usually have poor auto-inflation and don't usually have any hearing loss or tinnitus. To correct this problem, the diver should reverse the direction of movement, allowing the vestibular system to equilibrate. Another way to correct this involves auto-inflating the middle ear to equalize the pressures.

Finally, unequal caloric stimulation vertigo is a problem seen when cold water enters only one ear (due to either an external canal obstruction or a hood). It is like a caloric stimulation test. The best way to treat this is by removing the obstruction so that both of the ears are sensing the cold water at the same time.

Anxiety is the most common cause of dizziness in the diver. It is actually not due to the vestibular system. There are multiple studies that say that
greater than 50% of all scuba deaths are due to panic or anxiety in the diver. Treatment would include educating yourself properly before you scuba dive. Go through proper certification and never dive alone.

TMJ syndrome is caused by forcefully biting down on the regulator. This is frequently seen in novice divers who become anxious and excited and then bite down. It is treated just like any other treatment of TMJ, with reassurance, heat, NSAIDS and a soft diet.

There are other problems that divers may encounter that are associated with ascent. These are pulmonary rather than pressure accidents and these are very serious problems. These are all caused by holding of the breath during ascent and can be predisposed in patients with obstructive lung disease. At the surface, the pressures are all equal. As we descend, the ambient pressure increases and the lung pressure increases as well. As we are breathing, the pressures all stay equal because of our scuba apparatus. However, if we don't breathe during the ascent, the pressure in the lungs stays high and the pressure on the outside stays low. The lung pressures are higher than ambient pressure and this pressure has to go somewhere.

The first one is mediastinal emphysema, which is caused by local air trapping with escaping alveolar air dissecting along the bronchi into the mediastinum. This can lead to neck and chest discomfort, and shortness of breath. We have all seen what subcutaneous emphysema looks like in the soft tissues of the neck. The treatment for this is 100% oxygen, expectant management and, if the airway becomes an issue, that needs to be treated emergently.

The second thing is pneumothorax, which is caused by air from alveoli rupturing into the pleural cavity. It is sudden and rapid and causes shortness of breath and labored breathing and can actually be life-threatening underwater. The treatment is a chest tube. On a boat, or out in the water when a chest tube is not available, a large bore needle with a condom or a piece of plastic can substitute as a one way air valve to ventilate the air pressures. These patients need to be put on 100% oxygen immediately and, for this type of pneumothorax, patients should undergo chamber recompression.

The most serious problem that occurs in diving is an air embolism, a devastating problem that can lead to neurologic complications. This occurs when alveoli and pulmonary blood vessels rupture and air bubbles get to the blood stream and then track to the body. If these end up in the cerebral
circulation they can coalesce to form larger bubbles and actually entirely block the cerebral circulation. These divers usually experience loss of consciousness after surfacing. Frequently, while still in the water, they have frothy, bloody sputum and complain of chest pain, confusion, and blurry vision. These patients need to be transferred immediately to recompression chambers and frequently need treatment in these chambers intermittently for up to 2 weeks. If you are ever with a diver and suspect air embolism, the diver needs to be transported in the left lateral decubitus position. Air rises, so we want them in the left lateral decubitus to prevent a bubble from blocking the left ventricular outflow track. They also need 100% oxygen. Another issue is air evacuation, since flying in an airplane or at elevation can cause an increased bubble formation in the body. However, if the issue is getting the patient to a recompression chamber as quickly as possible, then the risk is usually warranted, as opposed to taking them by ground.

Not holding the breath, and just breathing normally as the ascent is made will prevent air embolism. Also, make sure that patients with obstructive lung disease do not scuba dive.

Case Presentation

The patient is a 25-year-old male with extensive SCUBA diving experience who was on a dive with friends. The patient was nearing the end of his dive and noted his gauge air pressure to be at 500 psi. He began to surface when, at 50 fsw depth, he begun to breathe 'heavy air', indicating a lack of adequate remaining air in his scuba tank. He emergently descended to his dive buddy at 80 fsw depth and began to share his companion's air. During his emergent re-descent, he noted severe otalgia while failing to equalize his middle ear pressure. Upon reaching the surface, the patient noted left-sided bloody otorrhea and rhinorrhea. He also experienced mild dizziness. On subsequent otolaryngologic exam, the patient had a 25% left tympanic membrane perforation. His dizziness had completely resolved. He was treated with ototopic agents and aural hygiene, and had complete closure of his perforation on 1-month follow-up.

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Eustachian Tube Function and Dysfunction
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The eustachian tube is an 3-4 cm tubular structure which links two of the major areas of interest in our specialty, the nose and the ear. Dysfunction of the eustachian tube causes many common symptoms that present to our clinics, and may have important management implications. This presentation will consists of a review of the history, anatomy and physiology of the eustachian tube, and the role of the eustachian tube in clinical situations.

History

The first description of the eustachian tube is attributed to Alcmaceon of Sparta in
400 BC. It was his belief that the eustachian tube allowed goats to breath through their ears as well as their noses.

In 1562, Bartolomeus Eustachius, the Chair of Anatomy in Rome, published the first detailed description in his thesis Epistola de Auditus organis, accurately describing the structure, course, and relations eustachian tube.

Antonio Valsalva, Professor of Anatomy at Bologna, applied the name "Eustachian Tube" to the pharyngotympanic tube, which was described by Eustachius.

In 1724, around the time of Valsalva, Edme-Gilles Guyot, a postmaster at Versailles, described the technique of eustachian tube catheterization. He succeeded in relieving his own deafness by passing a curved pewter tube through his mouth into the opening.

Since that time many noted otolaryngologists, including Drs. Politzer, Bezold, and Bluestone, have contributed significantly to our understanding of the intricacies of eustachian tube anatomy and function, and management of the various disorders that it is embroiled in.

Development and Anatomy

Understanding the development and anatomy of the eustachian tube provides insight into its role in several pathologic processes. The eustachian tube develops as a persistence of the first pharyngeal pouch. At 10 weeks post conception only the epithelial lining of the lumen has differentiated. Between the 10th and 12th weeks post conception the levator veli palatini and tensor veli palatini muscles develop. At 14 weeks the tensor tympani muscle becomes apparent, cartilage differentiation begins and rugae begin to develop within the tube.

The tube increases in length from 1 mm at 10 weeks post conception to 13 mm at birth. Also, the angle between the eustachian tube and the skull base is 10 degrees at birth.

This is in contrast to the adult length of 35 mm and angle of 45 degrees in adults. Vertical development of the skull, and increases in the angle of the skull base, allow the eustachian tube to reach its adult length and angle by age 7.

Basic Anatomy

In the adult, the eustachian tube can be visualized as two truncated cones attached at their narrowed ends. It runs from the middle ear to the nasopharynx and is
approximately 31-38 mm in length. Its lumen is roughly triangular and has average diameter of 2-3 mm. The lumen is lined by ciliated psuedostratified, columnar epithelium, which sweeps material from the middle ear to the nasopharynx. Mucous glands predominate near the pharyngeal orifice, and there is a gradual change to a mixture of goblet, columnar, and ciliated cells as the middle ear is approached.

The eustachian tube is composed of an osseous and a cartilaginous portion. The osseous eustachian tube or protympanum measures 11 to 14 mm and extends from the anterior and medial portion of the petrous temporal bone. Its orifice is oval shaped, measures 5 x 2 mm and is located above the floor of the middle ear space. When healthy, the osseous portion of the eustachian tube is always patent. The cartilaginous portion measures 20-25 mm and opens into the nasopharynx approximately 10 mm above the plane of the hard palate. The cartilage protrudes into the nasopharynx, and the protruding portion is known as the torus tubarius. The fossa of Rosenmuller is this area in the nasopharynx superior to the torus tubarius.

The cartilaginous portion is composed of one main piece of cartilage and can be accompanied by several accessory cartilages. Its composition and elasticity is similar to that found in the pinna and nose. It is attached at the sphenoid sulcus on the base of skull superiorly and its anteriomedial end is attached to a small tubercle on the posterior edge of the medial pterygoid plate.

**Blood Supply and Innervation**

The blood supply to the eustachian tube is from branches of the internal maxillary artery, ascending pharyngeal artery, and the ascending palatine artery. Sensory and motor innervation of the eustachian tube is provided by a branch from the otic ganglion, sphenopalatine nerve, and the pharyngeal plexus, predominately through branches of the glossopharyngeal nerve. Sympathetic branches reach the eustachian tube from the sphenopalatine ganglion, otic ganglion, glossopharyngeal nerve, petrosal nerves, and the carticotympanic nerve. Parasympathetic innervation is from the tympanic branch of the glossopharyngeal nerve. The multiple nerves innervating the eustachian tube, may be a source for referred pain to other anatomic regions of the head and neck.

**Muscles of the Eustachian Tube**

There are four muscles associated with the eustachian tube. These include the tensor veli palatini, levator veli palatini, salpingopharyngeus, and the tensor tympani.
The **tensor veli palatini** is composed of two distinct bundles of muscle fibers mediolateral to the tube. The lateral bundle takes its origin from the scaphoid fossa, and the lateral osseous ridge of the sulcus tubae for the course of the eustachian tube. It descends anteriorly and lateral and inferiorly, to converge in a tendon which passes around the hamulus and inserts on the posterior border of the horizontal process of the palatine bone and into the palatine aponeurosis of the velum.

The medial most portion of the tensor veli palatini originates on the lateral membranous wall of the eustachian tube and blends with the lateral bundle of the tensor veli palatini. This medial portion of the tensor veli palatini, referred to as the dilator tubae muscle, is probably responsible for active dilation of the eustachian tube by inferolateral displacement of the membranous wall.

The **levator veli palatini** arises from the inferior aspect of the petrous apex, passes inferomedially, paralleling the tubal cartilage, and attaches to the dorsal surface of the soft palate. It is thought to assist in active dilation and provide support.

The **salpingopharyngeus** arises from the medial and inferior portion of the eustachian tube and descends posterior and inferior to blend with the palatopharyngeus muscle. Its physiologic function is undefined.

The **tensor tympani** muscle arises from fibers common to the tensor veli palatini. The tendon of the tensor tympani rounds the cochleaform process and inserts into the manubrium of the malleus. It is not thought to play a role in eustachian tube function.

**Normal Function**

The normal eustachian tube is functionally collapsed at rest, with slight negative pressure present in the middle ear. It opens during swallowing, sneezing, and yawning. The eustachian tube is thought to close through passive reapproximation of the tubal walls by extrinsic forces and recoil of the elastic fibers.

The eustachian tube has three functions: ventilation, drainage, and protection. When the eustachian tube is patent it allows ventilation of the middle ear and equalization of middle ear and atmospheric pressure. It also allows the middle ear to clear unwanted secretions. By staying physiologically obstructed, it protects the middle ear from nasopharyngeal secretions and sound.

Conditions interfering with normal eustachian tube function cover the pathologic spectrum from benign to malignant. Resultant middle ear complications can be the
primary condition that the clinician needs to address, may be a sign of something more serious, or may have implications that will affect the outcome of surgical interventions.

**Eustachian Tube Dysfunction**

Bluestone has classified eustachian tube disorders into obstructive disorders, and disorders of abnormal patency.

Obstructive disorders can be mechanical or functional. Mechanical obstruction can be intrinsic due to intraluminal factors such as mucosal inflammation due to allergy or infection, or extrinsic obstruction resulting in compromise of the lumen. Extrinsic obstruction can be physiologic such as when the patient is supine, or may be caused by a mass lesion such as a neoplasm or an adenoidal mass.

Functional obstruction results from persistent collapse of the eustachian tube due to increased tubal compliance, an abnormal opening mechanism, or both. Functional obstruction is more common in infants and young children, and in many cases can be related to normal or abnormal developmental factors.

**Evaluating Eustachian Tube Function**

There are many methods for evaluating the condition of the eustachian tube, which reflect its deep location and complex physiology.

**IN THE CLINIC**

During the physical examination, otoscopy, pneumatic otoscopy, indirect nasopharyngoscopy, and endoscopy of the nasopharynx can provide clues to the condition of the eustachian tube.

Several maneuvers can be easily performed in clinic that may indicate patency of the eustachian tube. These include the Valsalva test, the Toynbee test, the Politzer test, and eustachian tube catheterization.

The Valsalva test is performed by visual inspection of the tympanic membrane while the eustachian tube and middle ear are inflated by a forced expiration with the mouth closed and the nose occluded by the thumb and forefinger. The test is positive when an intact tympanic membrane is observed moving, or by air heard through a perforated TM. A positive valsalva test only indicates an anatomically patent and probably distensible eustachian tube.
The Politzer test is performed by visual inspection of the tympanic membrane while compressing one naris into which the end of a rubber tube attached to an air bag has been inserted while the opposite naris is compressed with digital pressure. The patient is asked to repeat the letter K or to swallow while air is injected into the nasal cavity. When positive, the overpressure that develops in the nasopharynx is transmitted to the middle ear, and only indicates an anatomically patent ET tube.

Both the Politzer and Valsalva test may be beneficial as a temporary treatment of effusion or high negative middle ear pressure.

The Toynbee Test is performed by visual inspection of the tympanic membrane while the patient swallows with their nose manually occluded. This generates a positive pressure within the nasopharynx, followed by a negative pressure phase and is considered positive when there is an alteration in middle-ear pressure as assessed by pneumatic otoscopy before and after the maneuver. Negative middle-ear pressure or temporary negative middle ear pressure followed by return to ambient pressure after the Toynbee test usually is indicative of normal eustachian tube function. This is in contrast to the Politzer and Valsalva tests which only test patency. The results of this maneuver can often be equivocal, since several studies have shown that a significant portion of normal adults and children can not open their eustachian tubes with this maneuver, and patients with patulous eustachian tube often can not maintain a negative pressure within their middle ears.

Eustachian tube catheterization can be performed, and also can indicate eustachian tube patency.

Radiographic evaluation includes computed tomography, and magnetic resonance imaging. The use of contrast materials to evaluate patency has been described in the past, but is infrequently used today.

IN THE LAB

There are several more complex methods of evaluating eustachian tube function that have been described and most involve the use of manometry, sonometry, of tympanometry. Besides tympanometry most of these tests require complex equipment, and are mainly used in a research setting.

Non-intact Tympanic Membrane Tests

- The Inflation-Deflation test
- Forced Response test
- clearance test
Intact Tympanic membrane tests

- pressure chamber technique
- sonometry
- tympanometry

Clinical Examples Of Eustachian Tube Dysfunction

Here are a small sample of the clinical scenarios where eustachian tube dysfunction is important:

OTITIS MEDIA WITH EFFUSION

Obstruction may result in persistent high negative middle-ear pressure. If pressure equalization does not occur, atelectasis of the tympanic membrane-middle ear, sterile otitis media with effusion, or both can occur. If the negative pressure is overcome, it can aspirate secretions from the nasopharynx resulting in an acute otitis media.

Serous otitis media with effusion can result from either inadequate ventilation of the middle ear or from reflux of unwanted nasopharyngeal secretions into the middle ear. Both types of eustachian tube dysfunction can result in otitis media, abnormal patency and obstruction.

This is common in children and infants probably due to the configuration of their eustachian tube, shorter length, and lower efficiency of their tensor veli palantini.

While serous otitis media is something that many of us treat on a daily basis, Dr. Gacek of Syracuse reminds us, in an article entitled "A Differential Diagnosis of Unilateral Serous Otitis Media", of the potentially serious nature of this condition. Clinicians need to maintain a high index of suspicion, particularly in adults, in unilateral cases, and in persistent or recurrent cases. From Dr. Gacek's article, it is important to remember:

1. The eustachian tube lumen can be obstructed and this is usually from inflammatory, allergic or functional disorders.
2. The nasopharynx is usually obstructed by adenoid hypertrophy, but extensive nasal polyposis, benign neoplasms, and malignant neoplasms also may present in this location.
3. Obstruction may occur from laterally in the infratemporal fossa, by parapharyngeal space masses and neoplasms, or skull base lesions.
4. Medial obstruction from the petrous apex may be caused by solid or cystic lesions, including congenital epidermoids, cholesterol granulomas, neurofibromas, internal carotid artery aneurysms and other rare petrous apex lesions.

5. Effusion of CSF from the middle ear and mastoid caused by temporal bone trauma, surgery, or congenital defects may mimic otitis media with effusion and must be remembered as part of the differential diagnosis.

Dr. Gacek reinforces the importance of a thorough head and neck examination including the nasopharynx, CT scan of the head including the neck, and myringotomy as the minimal workup in any pediatric or adult patient with unilateral recurrent or persistent serous otitis media without an obvious explanation for eustachian tube obstruction.

He also emphasizes that in pediatric patients, the eustachian tube lumen and nasopharynx are the anatomic locations most frequently responsible, but congenital CSF leaks should be suspected in patients with a history of meningitis, or if the fluid after myringotomy resembles CSF.

In adults, all levels should be suspected.

NASOPHARYNGEAL CARCINOMA

Patients with nasopharyngeal carcinoma frequently have complications that relate to their eustachian tube. The frequently present with serous otitis media. Also, high-dose radiation therapy, the treatment for nasopharyngeal carcinoma, causes edema, vasodilation, mucosal damage, and fibrosis of the eustachian tube and middle ear resulting in damage to the middle ear contents and poor middle ear ventilation.

While it seems intuitive that serous otitis media with effusion in patients with nasopharyngeal carcinoma would be caused by mechanical obstruction of the pharyngeal orifice of the eustachian tube, several studies question whether nasopharyngeal tumors, actually obstruct the lumen of the eustachian tube, and instead propose that eustachian tube dysfunction and resulting otitis media with effusion is caused by infiltration of the tensor veli palatini muscle.

With regard to patients after radiation, a study by Hsu, et al, in 1995, showed that 95% of 38 eustachian tubes were patent prior to radiotherapy, 34% where patent at 6 months after radiotherapy, and 60% were patent at 5 years after radiotherapy using the passive opening test. They also showed decreased dynamic function and clearance at six months after radiotherapy and improved at 5 years. They attributed these findings to inflammation caused by radiation rather than tumor obstruction.
Electromyographic evaluation of the tensor veli palatini in patients with nasopharyngeal carcinoma status post radiation indicated neurogenic paralysis.

These authors have also found that in patients treated with ventilation tube insertion for post-irradiation OME tend to develop a chronic draining ear, and deterioration of hearing. They suggest myringotomy, avoidance of ventilating tubes, and frequent local treatment of infections of the nose, sinuses, and nasopharynx to avoid this outcome.

PATULOUS EUSTACHIAN TUBE

Patulous eustachian tubes often present a frustrating problem for patients and clinicians. The incidence is reported to be between 0.3-6.6% of the general population.

Patients with patulous eustachian tubes complain of aural fullness, humming tinnitus, and autophony. They also may hear their own breath sounds, which is known as tympanophonia. The sound is synchronous with nasal respiration and resolves when the patient is supine or when upper respiratory tract inflammation occurs. The sounds may be aggravated by mastication.

Symptoms are usually absent when the patient is supine or relieved when the patient bends forward with the head between the knees. For this reason, patients should not be examined in a supine position. Physical examination may reveal a tympanic membrane that moves during forced breathing through one nostril, and an amorphic sound may be heard using a diagnostic tube in the patient's ear.

The eustachian tube is usually closed, and closure is maintained by the elasticity of its cartilage, mucosal lining, surrounding muscles and fat. Alteration of any of these anatomic components may cause patulous eustachian tubes.

Conditions associated with patulous eustachian tubes include: radiation therapy, hormonal therapy, pregnancy, nasal decongestants, fatigue, stress, and weight loss.

Patulous eustachian tubes in the most severe form may be patent at all times, whereas a less severe form has been reported, where the tube is anatomically closed at rest, but may open easily during exercises or in association with a decrease in peritubal extracellular fluid.

Many patients can be treated with simple reassurance after a thorough history and physical examination. Treatment or removal of underlying factors may reverse the problem. Such as weight gain by patients who have lost weight.
Many medical regimens have been described including agents which produce intraluminal and extraluminal swelling, including: insufflation of boric acid and salicylate powder as described by Bezold, application of nitric acid and phenol, oral administration of saturated solution of potassium iodide (10 drops in juice TID), premarin nasal spray (25 mg in 30 cc NS).

New medications are currently under investigation including a herbal combination being evaluated in Japan, and a medication reported Dr. DiBartolomeo of Santa Barbara, California that is composed of chlorobutanol, benzyl alcohol, diluted hydrochloric acid, and propylene glycol. In the initial report, complete elimination of symptoms was reported by 8 of 10 patients. This formulation was derived from chlorinated pool water based on the observation that several patients had eustachian tube congestion proportional to the frequency of time they spent in a public pool. In letter to the editor in American Journal of Otology, Dr. DiBartolomeo indicated that the medication was held up with the FDA.

In patients who do not improve with medical therapy and who want further treatment, several surgical interventions have been used including electrocauterization of the eustachian tube orifice, peritubal injection with gelfoam, paraffin, avitene, or teflon paste, transposition of the tensor veli palatini muscle medial to the pterygoid hamulus, myringotomy with ventilation tube placement, and insertion of an indwelling catheter and subsequent ventilation tube placement. Catheter placement is through either an anterior tympanomeatal flap or through a myringotomy.

The close anatomic relationship of the eustachian tube and the carotid artery should be noted by clinicians who plan inject materials into the eustachian tube orifice, as injection of telfon paste into the carotid artery has been reported.

**HYPERBARIC OXYGEN THERAPY**

Another clinical situation where proper eustachian tube function is important is in the use of hyperbaric oxygen therapy, particularly in patients who require multiple sessions.

Hyperbaric oxygen therapy involves intermittent inhalation of 100% oxygen under greater than 1 atmosphere of pressure and is being used increasingly in patients with decompression sickness, osteomyelitis, carbon monoxide poisoning, crush injuries, radiation necrosis, and poorly healing wounds. Many of these patients develop otalgia and aural fullness that may be long-standing. Reports in the literature indicate that the incidence of middle ear barotrauma ranges from 5% to
28% of all patients.

Fernau, et al suggest patients should be taught clearing techniques such as the Valsalva or Politzer maneuver, supplemented with topical and/or systemic decongestants, subjected to slower compression rates, or possibly have ventilation tubes placed.

In a study of 33 patients undergoing hyperbaric oxygen therapy by Fernau, et al. in 1992, 82% of patients developed fullness in their ears, 52% developed serous otitis media, and 21% developed otalgia requiring ventilation tubes. Of 11 patients managed with decongestants, 10 patients resolved their effusion and pain and did not require further therapy. 45% of 33 patients had evidence of pre-existing eustachian tube dysfunction using the inflation-deflation test. Of these patients 100% developed aural fullness, 87% developed serous otitis media, and 47% required tympanostomy tubes. Fernau, et al, identified a history of eustachian tube dysfunction as a risk factor for serous otitis media in patients undergoing hyperbaric oxygen therapy.

An article by Presswood, et al, points out that the middle ear complication rate in intubated patients receiving hyperbaric oxygen therapy is 94% compared to 46% of non-intubated patients. (Hemotympanum and otalgia) They state that the use of nasal decongestants in this population is controversial, and probably of no value in patients who are intubated. They recommend ventilation tubes should be placed in their ears prophylactically.

OTHER CLINICAL SITUATIONS

Obviously, there are many more clinical situations that the role of eustachian tube dysfunction is important. Disorders of the eustachian tube present important issues that in diagnosis and management that are faced in daily clinical practice. Despite the large volume of literature on eustachian tube dysfunction, the lack of well-designed prospective studies make the literature difficult to decipher, and these disorders continue to represent some of the most challenging management problems we face as otolaryngologists.

Summary

The eustachian tube is an important anatomic structure that ventilates, protects, and drains the middle ear.

During development the eustachian tube lengthens and the angle between it and the skull base increases from 10 degrees in infancy to 45 degrees in adulthood.
Eustachian tube dysfunction can be caused by mechanical obstruction, which may be intrinsic or extrinsic, by functional obstruction, or by the presence of patulous eustachian tubes.

Otitis media with effusion is a common sequela of eustachian tube dysfunction, but a high index of suspicion must be maintained in adults, in unilateral cases, and in patients with recurrent or persistent disease without an obvious explanation.

Case Presentation

A 54-year-old male was referred to the Bobby R. Alford Department of Otorhinolaryngology and Communicative Sciences for evaluation and management of a right patulous eustachian tube. The patient had previously undergone a right myringotomy and ventilation tube placement without resolution of his symptoms.

He complained of right aural pressure and autophony for over one year. The autophony was particularly bothersome while singing. The patient had a history of high pitched noise exposure while in the military, and wears ear protection while at work. He denied headaches, nausea, vomiting, vertigo, otorrhea, otalgia, ear trauma, or ear surgery. He had no history of recent weight loss, and the remainder of his medical history was unremarkable.

Physical examination revealed his left tympanic membrane to be clear, intact, and mobile. The right tympanic membrane had a scar in the anterior inferior quadrant, but was otherwise unremarkable. The right tympanic membrane did not move with swallowing or Valsalva maneuver. There were no masses noted in the nasopharynx. The rest of physical examination was unremarkable.

The patient had an audiogram which was remarkable for symmetric severe sensorineural hearing loss above 2000 hertz bilaterally. He had Type A tympanograms bilaterally.

The patient was reassured of his condition and Premarin nose drops were started, but the patient failed to respond. The patient was given the option of surgical intervention, and has noted an improvement in symptoms 2 weeks status post eustachian tube obliteration and ventilating tube placement.
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TRAUMATIC INJURIES TO THE FACIAL NERVE
October 12, 1991
Randall S. Zane, M.D.

Traumatic injuries to the facial nerve can be classified by site (extracranial, intratemporal, intracranial) and by the type of trauma (penetrating, non-penetrating, or iatrogenic). The general principles of management remain the same regardless of the location of the injury, but the details vary and will be presented by location.

Blunt trauma to the face rarely causes significant extracranial facial nerve injury. An example is birth trauma, most often the result of a forceps delivery where the exposed facial nerve is compressed against the rigid cervical spine. Penetrating lesions of the facial nerve distal to the parotid gland and medial to the lateral canthus of the eye rarely result in severe disfigurement and most often do not require repair. Repair of the cervical or platysmal division is also usually unnecessary because of the minimal functional
loss that results from transection. Lesions of the main trunk or the temporozygomatic or cervicofacial divisions should be repaired or a major deficit in facial movement will persist. Clean facial lacerations with immediate onset of facial paralysis should be promptly explored. After three days, the severed branch loses its excitability, and that can make its identification tedious and uncertain. If there is gross contamination of the wound requiring debridement, other life threatening injuries where it is not advisable to prolong anesthesia, or if there is a lack of equipment or expertise, an attempt should be made to identify the severed nerve endings and to tag them for later identification. Whenever possible, primary end-to-end anastomosis is ideal and allows the greatest functional return. Interpositional grafting is second best, but a certain percentage of regenerating axons are lost across each anastomosis. For interposition grafting, the greater auricular and sural nerves are popular largely because of the ease in which these sensory nerves are harvested.

Iatrogenic injuries occur most often with extirpation of parotid malignancy where the facial nerve passes through the tumor and must be removed. In this instance, interpositional grafting is the usual procedure. Even when there is a question as to the adequacy of resection, potential long-term survival or the need for postoperative radiotherapy, grafting should be undertaken since it adds only a short time to the operation. The psychological advantages of even incomplete return of function as soon as possible after surgery should not be underestimated. In more routine parotid surgery, the facial nerve should be stimulated near the stylomastoid foramen before wound closure. If brisk facial movement occurs, subsequent facial motion will be satisfactory, even if there is paresis immediately postop. If movement does not occur in all major muscle groups, the nerve should be inspected carefully under magnification for evidence of injury, for example, ligature on the nerve, crush injury, and the like. If a crush injury is suspected it may be necessary to split the sheath to ascertain whether the fascicles are in continuity. If they are not, excision of the damaged segment and repair may be the best course of action.

Temporal bone fractures are a unique form of non-penetrating injury that can cause significant damage to the intratemporal facial nerve. They may be classified as longitudinal or transverse according to the orientation of the fracture planes on high resolution CT scanning, and they may occur in a mixed form. Facial nerve injuries are more common with transverse fractures, but because the incidence of longitudinal fractures is higher, they are seen more frequently as a result of longitudinal fractures.

With longitudinal fractures where the associated hearing loss is conductive in nature, exploration of the geniculate ganglion should be carried out via a middle cranial fossa approach, followed by transmastoid exploration of the tympanic and vertical segments for the occasional injury caused by fractures through the osseous canal. Transverse fractures, by virtue of the associated severe sensorineural hearing loss are explored through transmastoid and translabyrinthine approaches. Injuries in the tympanic and labyrinthine segments are easily accessible for repair via this approach. Lesions including edema, hemorrhage, contusion, and impacted bony spicules are best managed by decompression of the nerve in the fallopian canal. Transection of more than 50% of the nerve requires an interpositional graft to ensure regeneration of an adequate number of motoneurons.
Penetrating injuries of the temporal bone are most often caused by gunshot wounds but can be the result of some stab wounds. If there is any suspicion of a vascular injury of the sigmoid or lateral sinus, jugular vein or carotid system, carotid arteriography is indicated. When surgery is indicated, most can be explored through standard transmastoid approaches. Primary reanastomosis of a severed nerve in the fallopian canal is rarely encountered as an option to restore continuity. Interpositional grafts are frequently necessary using the nerve’s natural self-adhesiveness without the use of suture, and this may be enhanced by the application of autologous fibrin glue. The fallopian canal can be used as a sort of stent to hold the graft in place.

The most common sites of inadvertent injury to the nerve in tympanomastoid surgery are in the middle ear where the tympanic segment of the nerve above the oval window is occasionally dehiscent, and in the mastoid at the second genu where it is susceptible to injury during antrotomy if the usual landmarks including the lateral semicircular canal are distorted or not recognized. If a postoperative paralysis occurs, sufficient time must elapse to ensure that the effects of any local anesthesia have worn off. If the surgeon did not identify the nerve during the operation, exploration is mandatory. If the nerve was identified and the surgeon knows it to be intact, the patient should be followed with electrophysiologic testing. If only postoperative paresis is evident, the patient is followed closely by examination, and if the palsy becomes complete the patient should undergo electrophysiologic testing.

Injury to the intracranial segment of the facial nerve is usually iatrogenic, secondary to tumor removal, but infrequently can result from penetrating injuries or medial transverse temporal bone fracture. Collagen splints have been used for grafting in this location, but results are poor.

As the quality of radiographic studies, primarily high resolution CT scan, has improved, the importance of topognostic testing has fallen off. However, although it is sometimes inaccurate and has not been shown to be of prognostic significance, they can be helpful in determining the proximal extent of injury and should not be forgotten.

In the past, decisions as to surgery traditionally were based on the time of onset of complete paralysis, such that patients with immediate onset following trauma were explored and those with delayed onset were managed conservatively. Electrophysiologic testing has changed the scene quite a bit, since cases of paralysis with an immediate onset have been monitored and have had good spontaneous return of function, while delayed onset has been monitored to progress and lead to suboptimal results. In the first three days, electrical testing takes a back seat to clinical evaluation including the physical examination. Between three days and three weeks, if the patient has visible facial motion, electrical testing is not needed and will be normal if obtained. If there is paralysis, early screening with a Hilger nerve stimulator is used. If the threshold on the involved side stays less than 3.5 mAmmps above the normal side, no further testing is necessary. If the threshold difference goes above 3.5 mAmmps, some authors recommend exploration. However, if ENoG is available to you, it is recommended that serial examinations be performed every one or two days to follow the course of degeneration. Fisch has set up guidelines such
that should degeneration, to greater than 90% of nerve fibers as calculated by ENoG, take place in the first three weeks, the prognosis is poor for spontaneous recovery and that patient should be explored. Anytime after three weeks, there is some controversy regarding the decision to explore, primarily because some nonpenetrating injuries can show spontaneous recovery. Dr. Coker and Dr. Jenkins feel that all penetrating and iatrogenic injuries with evidence of complete degeneration need exploration to document location and cause of the problem. The intact nerve can then be left alone and observed, while the severed nerve is appropriately managed. All blunt injuries outside the stylomastoid foramen can be observed for six to twelve months before intervention.

The most controversy is generated over late exploration for temporal bone fractures with complete paralysis. This is because the natural course of recovery has not been established.

Except by perceptible facial movement or EMG activity, there is no way of knowing whether or not regeneration is occurring across the injury site and that with time facial function could be acceptable. Some argue that bony fragments and fibrosis can impede the spontaneous regeneration at the injury site and that exploration can enhance the opportunity for regeneration. This is a difficult problem to reconcile and should be approached on a case-by-case basis with careful counseling of the patient. Should there be no return of function by twelve to eighteen months post temporal bone fracture, there is little argument that the nerve should be explored with interpositional grafting at the site of injury.

The following management rationale for injuries to the facial nerve is reproduced (by permission) from the award-winning scientific exhibit presented by Dr. Newton Coker and Dr. Herman Jenkins at the 1990 American Academy of Otolaryngology - Head and Neck Surgery meeting in San Diego.

**Case Presentation**

A 54 year old black male was assaulted and stabbed multiple times in the back, shoulders and right face. The single facial wound was one centimeter in length and was vertically oriented 1.5 centimeters anterior to the tragus. Examination revealed complete right peripheral facial paralysis without significant edema or hematoma. Clear saliva was easily expressed from Stenson's duct. On the third day post injury, after stabilization and complete evaluation of his other injuries, he was taken to the operating room for right facial nerve exploration. The main trunk of the facial nerve was identified and found to be transected just proximal to the pes anserinus. All branches of the nerve were distally intact as demonstrated by electrical stimulation of the distal cut edge. A primary neurorrhaphy was carried out under no tension with 10-0 Prolene suture using a perineural technique.
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FACIAL REANIMATION OF THE CHRONICALLY PARALYZED FACE

September 28, 1994
Amy Y. Chen, M.D.

Facial nerve paralysis is a devastating injury that can result in blindness, oral incontinence and social handicaps. The muscles of the face are exquisitely designed for both voluntary and involuntary expression.

The facial nerve emerges from the brainstem via the internal auditory canal and exits the brain through the stylomastoid foramen. At this point, the nerve travels a short distance to the pes where it divides into five motor branches--the temporal, zygomatic, buccal, mandibular and cervical branches.

The nerve is responsible for orchestrating an intimately designed facial musculature system. The facial musculature is capable of maintaining tone as well as expressing both voluntary and involuntary
First, we will discuss the importance of eye protection and methods used to achieve a healthy eye. Paralysis of orbicularis oculi results not only in drying of the cornea but also in poor distribution of tears. The lower lid is not able to contract and disperse the tears across the eye. Furthermore, loss of parasympathetic innervation can result in decreased tear production. This can result in keratitis and rarely, corneal ulceration and cataract formation. Therefore, the primary objective of the surgeon is to protect the eye via surgical and/or nonsurgical methods.

Surgical methods may involve both upper lid procedures such as tarsorraphy, gold weights, and palpebral springs as well as lower lid procedures.

Muscle transposition can be utilized to reanimate the face. Two muscles primarily used are the masseter and the temporalis. The patient must have an intact V for these procedures. Movement of the face can be achieved by instructing the patient to clench the teeth.

Masseter muscle transposition involves harvesting two anterior slips of masseter muscle and transposing it to the upper and lower lip. Advantages include supplementing other reanimation techniques such as XII-VII or temporalis sling and providing static tone for the face. However, a downwards vector may cause the lip to droop.

Temporals muscle transposition is also commonly used. This muscle is used primarily to reanimate the mouth and lower aspect of the face. The temporals muscle is harvested and sewn to upper lip, commissure and lower lip. The key to success is to ensure overcorrection and exaggeration of the lip. Goretex strips can be used to lengthen the muscle so that it can reach the mouth. The defect in the temporalis can be corrected with synthetic insert or a temporoparietal fascial flap. May & Drucker in 1993 reported that the success of temporalis muscle transfer to restore a smile was 80% and to improve mouth function was 96%.

Various neural methods have also been employed to attempt facial reanimation. Primary anastomosis can be utilized effectively in the acute injury. But in the chronic palsy, primarily three techniques are currently used--XII-VII traditional, XII-VII jump, and VII-VII cross face graft. 4-6 months usually pass before the facial muscles begin to show signs of recovery.

The XII-VII traditional anastomosis was first performed in 1903 by Korte. Indications for this procedure include an intact peripheral facial nerve and a nonatrophic facial musculature. However, if the patient has injured the peripheral facial nerve or has atrophied muscles, or if other cranial nerve palsies exist, this procedure is to be avoided. The patient must learn to push his tongue against the teeth when he wishes to smile. Advantages of the procedure are that majority of patients has good movement. However, some patients can develop synkinesis and mass motion as well as atrophy of the tongue.

Another method of XII-VII anastomosis is that of placing an interposition graft between the two, thus
"jumping" from XII-VII. This procedure involves partially severing the XII nerve. One usually uses either the greater auricular nerve or the sural nerve as grafts. The former is preferred since it does not involve another operative exposure. However, the sural nerve can be advantageous for it provides a longer graft as well as larger diameter graft.

For best results, the anastomoses should be performed within one year of surgery. This jump graft procedure can diminish the incidence of tongue atrophy, the problems with synkinesis and mass motion and those with swallowing, mastication and speech. However, patients can have weak facial contractures. Another disadvantage is that the procedure involves two anastomoses rather than one as in the traditional XII-VII.

The incidences of swallowing deficiencies, mastication problems and speech difficulty are greater for XII-VII traditional grafts as compared to XII-VII jump. However, the results of achieving symmetry and excellent facial movement are similar between the two. One is able to reduce the incidence of mass movement with a jump graft.

VII-VII cross face grafts have also been used. Scaramella first described the procedure in 1970. It involves grafting VII from the functional side of the face to the stump on the paralyzed side. Obvious disadvantages include long operative time involving two anastomoses and surgical intrusion of the functional side of the face. Synkinesis and low grade neural input can result in less forceful contracture of the face. This procedure is often used in conjunction with free muscle transfers.

With the advent of microsurgery, it is also possible to transpose free flaps. Harii et al in 1975 were the first to transfer free muscle tissue for facial reanimation. Free muscle transfer provides soft tissue coverage of a defect as well as the opportunity for one to develop voluntary control of the face. This is usually a two stage procedure. The first stage involves anastomosing the nerve graft, usually a cross face graft. 6-12 months later, the free flap is harvested and anastamosed to the site. Commonly used donor sites include the gracilis, serratus anterior, latissimi dorsi, and rectus abdominis. The recipient vessels are typically facial artery and common facial vein.

In summary, the management of the permanently paralyzed face is threefold. First, one must provide protection of the eye. Second, muscle transposition is performed to restore tone to the patient's face. Third, efforts are made to restore facial movement that automatically reflects the breadth and depth of human expression. It is this last goal that remains elusive. However, with these methods, a patient can use the restored movement to express human emotion by practice and adaptation.

**Case Presentation**

A 65-year-old black woman presented with a long history of right-sided hearing loss. Over the past few months, she has had dizziness and unsteadiness in her gait. Past medical history was significant for
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diabetes and hypertension. A hysterectomy was her only surgical history.

Imaging studies revealed a large 5 cm tumor in the cerebellopontine angle consistent with a vestibular schwannoma. Audiogram revealed a profound right sensorineural hearing loss.

Excision of the tumor was performed via a transotic and translabyrinthine approach. The VII nerve was not preserved and therefore the patient developed a right facial paralysis. She did not have other cranial nerve deficits.

Approximately 6 weeks after the initial surgery, the patient was evaluated for facial reanimation. At this time, gold weights were implanted in the right eyelid and a XII-VII hypoglossal-facial nerve interpositional-jump graft procedure was performed. The patient had an uneventful postoperative course and was discharged home two days after surgery.

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THERAPEUTIC OPTIONS FOR TREATMENT OF GLOMUS TUMORS OF THE TEMPORAL BONE

October 26, 1991
Judith J. Owens, M.D.

Glomus tumors arise from paraganglionic chemoreceptor cells and are part of the APUD system of cells of neural crest origin. Paragangliomas of the jugular-tympanic area are the most common tumors involving the middle ear. In 1941, Guild described the distribution of normal glomus bodies within the temporal bone. Then in 1945, Rosenwasser correctly recognized and reported the first case of a tumor arising from these cells. After this report, many new cases were reported and many older cases were reclassified as true glomus tumors.

These neoplasms can occur at any age but the peak incidence is in the fifth decade. Glomus tumors occur four to five times as often in women as in men and there is a predilection for them in the Caucasian race.
The majority of these tumors arise from along the jugular bulb or Jacobson's nerve (85%), while 12% arise from glomus bodies overlying the promontory and three percent have their origin attributed to Arnold's nerve. The most common presenting symptoms include conductive hearing loss and pulsatile tinnitus. Other symptoms may include aural hemorrhage or otorrhea, otalgia and facial palsy. When the tumors enlarge within the jugular foramen neuropathies of IX, X, XI and XII occur. Anterior expansion toward the clivus may produce abducens and trigeminal palsies. Aquino's sign is blanching of the tympanic mass with gentle pressure on the carotid artery. The sign of Brown is the pulsation sign elicited by pneumatic compression and abolished with further compression. These tumors may be multicentric. Association of these tumors with other tumors arising from cells of neural crest origin (MEN I and pheochromocytoma) is not uncommon. This must be considered to avoid the devastating consequences of inducing general anesthesia in a patient with an unrecognized pheochromocytoma. Malignancy occurs in one to three percent of these tumors.

Evaluation includes a careful history and physical examination. High resolution computed tomography provides accurate information about bony destruction and the extent of the disease. Arteriography remains the most useful radiologic modality available for the assessment of a glomus tumor. Prior to angiography, serum catecholamines and urinary vanillylmandelic acid (VMA) levels should be obtained to screen for tumors which secrete these vasoactive compounds.

Therapeutic options for glomus tumors of the temporal bone include surgical excision, radiotherapy, combined therapy and in selected cases observation. Surgical excision offers the only chance for total tumor eradication. Gary Jackson writes that "...surgical management is curative and the treatment of choice." Proponents of radiotherapy argue that radiation provides for the amelioration of symptoms and arrests the progression of the glomus tumors providing a safe and efficacious modality of therapy.

Many surgical approaches have been proposed for the resection of these tumors (see bibliography). No single approach is optimal for all tumors. Jackson and Glasscock proposed guidelines for a planned excision of these tumors. They suggest approaching type I glomus tympanicum tumors via a transcanacl approach, and an extended facial recess approach for type II-IV glomus tympanicum tumors. Type I and II glomus jugulare lesions are treated by standard skull base techniques. Type III and IV tumors require an infratemporal fossa approach. The infratemporal fossa as described by Fisch provides for direct surgical access to the entire length of the intratemporal carotid artery and for control of the great venous sinuses. These infratemporal fossa approaches provide access as follows: Type A provides access to the temporal bone in its infralabyrinthine and apical compartments and its inferior surface. Type B provides access to the clivus and type C provides access to the parasellar region and nasopharynx. Utilizing these skull base approaches it is now possible to perform total extirpation of tumors that were previously considered unresectable. Skull base surgery is not undertaken without certain risks. Mortality rates range from four to seven percent. Reported complications include CSF leaks, new cranial nerve palsies, wound infections, and deafness. The risks of surgery are balanced out by the chance for total tumor eradication. The reported rate of local control for glomus tumors of the temporal bone with surgical treatment from several studies is shown in Table 4.

Although radiotherapy is unable to provide complete tumor eradication, the success of radiotherapy in
controlling the progression of symptoms is quite high. The reported rate of local control with radiation therapy for glomus tumors of the temporal bone from several studies is shown in Table 5. Complications of radiotherapy include osteoradionecrosis, brain necrosis, abscess formation, and radiation induced malignancies.

Case Presentation

A 69-year-old black female had initially sought assistance from a hearing aid dispenser after experiencing several years of slow, progressive left-sided hearing loss. However, a medical referral was made when the presence of a unilateral conductive hearing loss and pulsatile mass on the tympanic membrane were discovered. CT examination of the temporal bones demonstrated an opacification of the left mastoid air cells and middle ear cavity.

She was then referred to Baylor College of Medicine for further evaluation and treatment. Upon questioning, she reported that she could appreciate a rhythmic sound in her left ear that was synchronous with her heart rate. Examination revealed a pulsating tympanic membrane with dilated vasculature that extended onto the skin of the external canal. The sign of Aquino was negative and the Weber lateralized to the left. There were no cranial nerve deficits. Arteriography was obtained. This demonstrated a vascular mass of the left temporal bone supplied by the penetrating vessels of the external carotid artery. These findings were consistent with a glomus tympanicum. The options of surgical resection, radiotherapy and observation were explained to the patient. It was elected to observe the tumor for the time being and to offer her radiotherapy should the tumor enlarge or become symptomatic.

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CONSIDERATIONS IN THE EVALUATION OF THE HEARING IMPAIRED CHILD

Judith J. Owens, MD
March 11, 1993

Approximately 5,000 children are born every year in the United States who will be found to have a significant hearing impairment. The hearing loss may be secondary to congenital or postnatally acquired conditions. Congenital hearing loss is attributed to a defect that the child is born with, either an inherited genetic defect or the result of a prenatally acquired condition. The important non-hereditary causes of congenital hearing loss include drug exposure, prenatal infections (TORCH) and erythroblastosis fetalis.

Three-quarters of childhood hearing impairment is postnatally acquired. Infections, drugs, hyperbilirubinemia, noise exposure, and trauma have all been implicated in contributing to postnatally
acquired hearing loss. In addition to meningitis and sepsis, the important infectious diseases associated with postnatally acquired hearing loss include adenovirus, mumps, chicken pox, hepatitis, EBV, and influenza. Hyperbilirubinemia when greater than 20 mg % will result in damage to the cochlear nucleus.

Although most hereditary disorders appear to follow the rules of Mendelian inheritance, marked variability of gene expression and incomplete penetrance may confound the diagnosis and clinical recognition of many of these disorders. A complete family history is essential to uncover these types of hearing loss. This is especially true for nonsyndromic inherited hearing loss. The hearing loss may be present at birth or may develop later in life. All forms of inheritance, autosomal dominant, autosomal recessive, and X-linked, have been documented in families with nonsyndromal hereditary hearing impairment (HHI).

Hearing loss is a component of over 200 inherited syndromes. Many syndromes in which hearing loss is a significant component were reviewed for this presentation. The major features of some of these syndromes will be reviewed in this abstract.

For convenience these syndromes can be categorized by the association with other morphologic abnormalities. Important syndromes where deafness is associated with craniofacial abnormalities include Treacher-Collins, Goldenhar's, Crouzon's, and Apert's syndromes. All except Goldenhar's (AR) are inherited in an autosomal dominant fashion. Hearing loss in these syndromes is mixed or conductive and may be surgically correctable.

Hearing loss in association with visual loss is a particularly devastating combination. There are four important syndromes with this combination: Usher's, Alstrom's, Cockayne's and Refsum's disease. All are inherited in an autosomal recessive fashion. Retinitis pigmentosa is associated with all except for Cockayne's syndrome. There are four types of Usher's syndrome. Type I is the most severe and is associated with vestibular dysfunction.

Apert's syndrome consists of progressive renal insufficiency, progressive sensorineural hearing loss and various ocular abnormalities. This syndrome demonstrates X-linked dominant inheritance: males are more severely affected and females may have a variable course to their disease because of random X chromosome inactivation.

Jervell and Lange-Nielsen syndrome is the most common syndrome with cardiac dysfunction and hearing loss. There is a conduction defect resulting in a prolonged QT interval on electrocardiogram, and clinically with fainting spells or sudden death. Jervell and Lange-Nielsen is inherited in an autosomal recessive manner.

Pendred's syndrome is the most frequently occurring example of associated deafness and endocrine dysfunction. This disorder results from an error in thyroxin metabolism. A goiter may be clinically palpable and profound sensorineural hearing loss is usually present.
Dermal abnormalities and deafness characterize Waardenburg's syndrome and the LEOPARD syndrome. Clinical features of Waardenburg's include dystopia canthorum, deafness, a high broad nasal root, synophrys, heterochromia irides, and early graying. There are two types of this syndrome characterized by the presence or absence of dystopia canthorum. When dystopia canthorum is absent, but the other features of the syndrome are present, there is a higher incidence of deafness.

Metabolic syndromes, especially the mucopolysaccharidoses, are frequently associated with conductive or mixed hearing loss. Chromosomal abnormalities, specifically trisomy 13 and 18 are often associated with deafness. Because these conditions are lethal in early infancy the deafness is of relatively minor significance. Down's syndrome is often associated with a mixed hearing loss.

Early identification of hearing loss is desirable to optimize rehabilitation. Identification of hearing loss in young children is facilitated by a clinician who looks carefully for a family history of early hearing loss, and who is attuned to the many syndromes associated with hearing impairment. Complete otologic and auditory evaluation of these children is essential.

Case Presentation

An eight-month-old child presented to Texas Children's Hospital for an otologic evaluation. The child was born full term via an uncomplicated vaginal delivery. There was no history of infections, drug or alcohol use during the pregnancy. Both parents are hearing-impaired. The mother reported the etiology of her deafness as "tonsilitis" and the etiology of her husband's hearing loss as "high fever." On physical examination the child demonstrated multiple stigmata of Waardenburg syndrome including dystopia canthorum, heterochromia irides, a broad nasal root, and synophrys or confluence of the eyebrows. The mother has similar features and in addition has a white forelock that is also associated with this syndrome.

Audiologic evaluation revealed no responses to speech, warble tones, or narrow band noise in the sound room. Auditory brainstem response audiometry showed no response to air or bone conducted clicks at equipment levels in either ear. In light of her significant sensitivity loss she was referred for hearing aid evaluation.

Despite the fact that both parents are hearing-impaired and communicate through signing, they were anxious to have the child fitted with hearing aids. The parents were referred for genetic counselling to provide them with a better understanding of the full implications of this disease.

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UPDATE ON HEARING AIDS
Michael G. Stewart, MD
June 3, 1993

Approximately 22 million Americans have some degree of hearing loss. Currently, patients with sensorineural hearing loss represent approximately 90% of hearing aid users. The sales of hearing aids in the United States have grown steadily, from 830,000 units in 1981 to 1,780,000 units in 1992. Behind-the-ear (BTE) aids represent 18.5% of this total, with body and eyeglass aids accounting for only 0.4%; the remaining 81.1% sold are in-the-ear (ITE) aids. This is a distinct change from only ten years ago, when ITE and BTE aids each represented
about 50% of the market.

Electronic hearing aids consist of a microphone, an amplifier, and a receiver. The microphone transduces sound into an electrical signal and sends it to the amplifier. The amplifier increases the amplitude of the electrical signal, and the signal may be further modified by filters and volume or tone controls. The amplified signal is transmitted to the receiver, where it is transduced into sound. When the amount of amplification is expressed in decibels, it is called the gain of the aid.

One complaint of most patients with cochlear loss is a decrease in dynamic range, also called cochlear recruitment. This is because the loudest sounds are still perceived with the same intensity, whereas patients with cochlear loss are unable to hear quiet sounds. Therefore the range between comfortable listening and uncomfortable loudness is much smaller than in normal subjects. Aids must then be able to supply adequate gain to the user without creating sounds which are too loud. Most aids sold today use linear amplifiers, which increase the signal amplitude by a constant amount, regardless of input level. These aids then require some kind of output limiting - such as peak clipping or compression amplification - to differentially amplify soft sounds more than loud sounds.

Earmold design is an important factor in aid design. With BTE type aids, a silicone earmold is used to deliver sound to the ear. With ITE type aids, the aid itself fits into the EAC. In both cases, the aid potentially acts as an earplug and creates the so-called "occlusion effect." Completely blocking the canal results in an increased amplification and perception of low frequencies, especially in the sound of the wearer's own voice. One solution to this problem is venting, or making an opening in the earmold or aid, which attenuates (or decreases) the amplification of low frequencies. However, venting may create a problem of its own: feedback. Feedback occurs when a microphone is placed very near a speaker. The microphone picks up the speaker output, which is amplified and sent to the speaker, which is picked up by the microphone and amplified again, etc. The capacity of the amplifier is quickly overloaded, and the high-pitched sound called feedback results. Feedback is an inherent problem with ITE aids because
the microphone and speaker are on the same piece of equipment. Venting may further decrease the amount of gain possible before the microphone picks up the speaker output and feedback results. Feedback is a less significant problem in BTE aids because the microphone and speaker are physically separated.

There are several types of aids available. Body aids are seldom used today, but have very high gain possible because of the separation of microphone and speaker. Body aids are still used for some patients with profound hearing loss. Eyeglass aids allow the fitting of CROS and BICROS hearing aids. CROS aids are helpful for patients with unaidable loss in one ear only, and BICROS aids are used for a combination of unaidable loss in one ear and aidable loss in the other. In BTE aids, the controls, microphone and amplifier are in the body, and sound is transduced through an earmold. There are three varieties of ITE aids: the full-concha aid, the half-concha aid, and the canal aid. Typically ITE and BTE aids have 65 to 75 dB of gain, and body aids may have 80 dB or higher gain.

The latest technology in hearing aids is the digital aid. In the past, electronic aids have all used analog circuitry, where the sound is transformed into an electrical signal and then further modified. There are natural limitations in the amplifier's ability to amplify the signal without exceeding power limitations or introducing distortion. In digital technology, the electrical waveform is converted into a digital code of a series of ones and zeros. This digital signal is then modified by computer, and transformed back into an electrical signal and sent to the speaker. The advantage of digital processing is that flexibility is much greater. The digital waveform can be transformed in complex ways without loss of precision, and the natural limitations of electronic equipment are not imposed. Specifically, the frequency response can be adjusted more accurately, the output can be limited at any frequency to prevent uncomfortable sound levels, filtering is more accurate, noise is not introduced, and acoustic feedback can be controlled. These modifications result in the more "natural" sound of digital aids, according to many aid users.

Although hearing aids are tested and have published specifications, these are obtained by placing the aid in a standardized device called a 2 cm coupler, and
measuring performance. Real ear testing involves the placement of a tiny probe in the patient's ear canal. The frequency response of the unaided ear canal is measured, then the aid is placed, and the aided frequency response is measured using different sound inputs.

ASSISTIVE LISTENING DEVICES

Many patients have a mild to moderate hearing loss or a narrow band of frequency loss (especially high-frequency loss) that makes hearing aid use difficult. In addition, some patients do not have the manual or mental dexterity to use hearing aids. Assistive listening devices are designed to improve hearing ability in special listening situations - especially if background noise, multiple talkers or large distances are a problem. There are several types of assistive devices.

A hard-wired device uses a microphone close to the source which is directly connected to an amplifier and earphone that the patient wears. The AM radio signal has been used as a wireless system to transmit sound from the source to individual receivers. However, AM signal is limited in power and range, and is very sensitive to electrical disturbances and electronic equipment. Wireless FM systems are much more popular because FM provides a clear signal that is almost immune to outside interference. Finally, infrared light has been used as a signal carrier. Infrared light is invisible, but requires an unobstructed view to the receiver, and is interfered with by some other sources of light.

These assistive listening devices (ALDs) are all based on the principle of improving signal to noise ratio (SN ratio), which means that the desired sound is amplified with respect to surrounding noise. This is very useful for hearing-impaired persons in lecture halls, school, or church, or for such activities as TV viewing with a large group of people. ALDs may also be used in conjunction with a hearing aid.
Case Presentation

A 48-year-old male veteran suffered noise exposure while in the service. He was fitted with a right-side monaural behind-the-ear aid in 1965 while in the military. He has been followed at the Houston Veterans Affairs Medical Center Audiology Clinic since 1972, preferring right ear only hearing aid use. His most recent audiogram shows a stable bilateral moderate to severe sensorineural hearing loss from low to high frequencies. His discrimination was 80% in the right ear, and 70% in the left. Acoustic reflex thresholds were elevated, but consistent with the extent of the sensitivity loss, and tympanometry revealed Type A tympanograms. The patient was recently fitted with bilateral in-the-ear hearing aids, which the patient uses regularly with good results. His real ear testing reveals excellent tuning to the computer-simulated parameters.

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Herpes Zoster Oticus
August 21, 1997
F. Christopher Holsinger, M.D.

History

Herpes zoster has been known since antiquity. Herpes is derived from the Greek meaning “to creep” and zoster “sword belt or girdle.” The Greeks called it “zona” and for centuries thereafter the disease was thought of as a cutaneous process. By the nineteenth century, physicians had begun to question this understanding.
Bright (1831) and Henley (1840) first suggested that herpes zoster represented a cutaneous manifestation of a nerve disease involving the sensory portion of the spinal nerves. Dr. von Barensprung (1861) demonstrated definitely at autopsy inflammatory lesions at the posterior root ganglion in a patient with herpes zoster.

Numerous observers described cases of herpetic eruption at the cephalic extremity due to herpetic infections involving the various cranial nerve ganglia or the upper cervical spinal root ganglia. Tryde gave the first description of cases of herpes zoster associated with facial palsy in 1872.

Head and Campbell published their landmark study in 1900. By correlating autopsy findings and careful, detailed clinical descriptions, these investigators mapped the sensory dermatomes defined by the dorsal root ganglia. Their work laid the foundation for future work and focused the energy and interest of others in this area.

Koerner, writing in the 1904, coined the term “herpes oticus” designating a syndrome that consists of the triad: blisters at the auricle, facial paralysis, and inner ear disturbances.

In 1907, Dr. Hunt, chief of Neurological Sciences at the Cornell University Medical Center, published his hypothesis regarding the etiology of herpes zoster oticus. Rather than presenting a new theoretic construct for disease, Dr. Hunt’s theory advanced the work begun by Drs. Head and Campbell and accurately classified herpes zoster of the head and neck. His paper represented the capstone of nearly a century of medical research and debate in this area. Hunt felt that herpetic eruption and associated symptoms (a viral prodrome, severe otalgia, facial nerve dysfunction, vesicular eruption involving the pinna, and occasionally vestibulocochlear symptoms), which he was the first to describe together as part of a clinical syndrome, were actually the result of, in his words, geniculate ganglionitis.

Hunt analyzed the sensory innervation of the ear and described the “zoster zone” for the geniculate ganglion as comprising a portion of the tympanic membrane, the external auditory canal, the tragus, antitragus, concha, a part of the helix, and a strip of the lobule. Here the typical herpetic lesions of geniculate zoster are most commonly found. The geniculate zone of the auricle is bounded anteriorly by the gasserian zone and posteriorly by the cervical zone, with some overlapping. Hunt classified herpes zoster at the cephalic extremity according to the sensory ganglion involved and the site of the rash into the following groups: (1) geniculate herpes zoster (herpes...
Hunt then further classified the herpes zoster oticus syndrome according to severity of symptoms and the degree of extension: (1) herpes zoster auricularis without neurological signs, (2) herpes zoster oticus with facial palsy, (3) herpes zoster oticus with facial palsy and mild acoustic symptoms (diminished hearing and tinnitus), and (4) herpes zoster oticus with facial palsy and Ménière’s syndrome complex (deafness, tinnitus, vertigo, nystagmus, nausea, and vomiting). Ramsay Hunt described not so much a discrete clinical syndrome but rather a spectrum of herpetic disease, involving the ear and cranial nerves VII and VIII.

To support this classification scheme, he postulated that the major site of the pathologic lesion in herpes zoster oticus was the geniculate ganglion of the facial nerve. Increased pressure of the swollen ganglion on the facial nerve resulted in facial palsy. He further postulated that the etiology of the Ménière’s syndrome complex was either the simultaneous infection of the adjacent ganglions of the eighth nerve or the extension of the inflammatory process from the facial to the auditory nerve by means of nerve fibers that communicate between the two. He believed that this clinical syndrome represented “geniculate ganglionitis.” Unfortunately, he was not able to provide evidence for his theory. With histopathologic studies of autopsy specimens, subsequent investigators demonstrated little, if any, ganglion involvement.

Denny-Brown and his colleagues in 1944 were the first to challenge Hunt’s theory. They examined histopathologic specimens from a patient with auricular and occipital herpes zoster and facial palsy. At autopsy, they found no changes in the geniculate ganglion, but were able to demonstrate other significant findings: a necrotizing ganglionitis of the second cervical ganglion and a patchy motor neuritis of the facial nerve. They concluded “the evidence for geniculate ganglionitis in the Ramsay Hunt syndrome is invalid.”

In time, virology research further clarified these questions. Tissue culture studies published by Drs. Weller and Coons in 1954 confirmed the theory that the virus that causes varicella and herpes zoster is one in the same. In 1965, Hope-Simpson’s novel hypothesis unified thinking on herpes zoster and has laid the foundation for our current understanding. He suggested that the varicella virus lies dormant in the sensory ganglion after the initial infection. In a patient with low circulating antibodies, the virus would reactivate and infection would then manifest itself as herpes zoster,
anywhere along the distribution supplied by that particular ganglion. Indeed, the facial nerve may be affected at any site from the brainstem to the periphery by the herpes virus. What is still not known is what activates the virus after it has lain dormant for so long.

In 1967, Blackley presented histologic evidence to support this emerging notion. He found extensive lymphocytic infiltration along the course of the facial nerve, as well as divisions of the VIII cranial nerve including vestibular and cochlear ganglia. Massive lymphocytic infiltration of the right facial nerve was seen. In addition, there was collapse and disruption of Reissner’s membrane, an increase of round cells in the stria vascularis, and destruction of the organ of Corti near the apical turn. Broad perivascular cuffing of several vessels is seen and marked atrophy of spiral neurones.

**Epidemiology**

A Mayo clinic study estimated the annual incidence of herpes zoster, regardless of site, as 130 cases per 100,000. The attack rate increased dramatically over the age of 60, and 10% this population had identifiable risk factors for decreased cell-mediated immunity including carcinoma, trauma, radiation therapy, or chemotherapy. The increased incidence in the elderly population is explained by a decrease in cellular immune response to varicella-zoster virus with age.

Adour reported the incidence as 5 per 100,000 a year, or one case every 52 minutes. These age-adjusted incidence figures for VZV cranial neuritis with facial paralysis parallel those Hope-Simpson estimated for VZV in the general population in 1965. These figures compare with those for Bell’s palsy: 20 per 100,000 per year. Few studies outside J.R. Hunt’s own assess the incidence of herpes zoster oticus in the population.

**Microbiology: The Herpesvirus family**

Members of the family herpesviridae are found in a wide range of host systems. To date, at least seven different species are known to infect man, including herpes simplex virus (HSV); cytomegalovirus (CMV), Epstein Barr virus (EBV), and varicella zoster (VZV).

Nomenclature is critical. It is important to distinguish VZV as a member of the greater family of herpes viruses. But it is distinct from herpes simplex. This is a common point of confusion.
The varicella-zoster virus (VZV) belongs to the herpes family. It is a double-stranded DNA virus that causes chicken pox (varicella) and zoster infections. Again what distinguishes Varicella from Zoster is the time of presentation. Reactivation of virus stored in sensory ganglia from previous varicella infection results in zoster.

Herpes viruses have an envelope surrounding an icosahedral capsid, approximately 100nm in diameter, which contains the dsDNA genome. When the envelope breaks and collapses away from the capsid, negatively stained virions have a typical "fried-egg" appearance.

**Work Up and Evaluation**

A thorough history and physical examination, a fundamental axiom in medicine, is the most crucial element of the work-up. Diagnosis still hinges on the clinical findings described in Hunt's classification scheme.

**Clinical Presentation**

Frequently, the first symptom is a deep, burning pain in the region of the ear. This is shortly followed in 1 to 4 days by a vesicular eruption of the EAC and concha, or, less frequently, of the face, neck, trunk, palate or fauces. The distribution of the vesicles depends on which sensory afferent fibers are involved by the viral eruption, but all the fibers may be involved, including cranial nerves V, IX, X and the cervical plexus arising from cervical roots II, III, IV. Cranial nerves VII and VIII are almost always both involved. During the acute illness, a varicelliform rash often accompanies the painful vesicular eruption. Facial nerve paralysis, vertigo, and hearing loss are commonly seen.

**Natural History**

The natural history of herpes zoster oticus differs from that of Bell’s palsy in several ways, perhaps reflecting the difference in the behavior of herpes simplex type I and VZV. Bell’s palsy recurs in 10% -12% of cases, but herpes zoster oticus rarely recurs. In addition, the acute phase of the infection, as measured by electrical response and progression, peaks in 5 to 10 days with Bell’s palsy, whereas herpes zoster oticus peaks in days 10 to 14. Finally, 84% of individuals with Bell’s palsy have a satisfactory recovery of function, but only 60% of patients with herpes zoster oticus recover function.
Crabtree (1968) was among the first to suggest that complete facial nerve recovery is less likely following herpes zoster oticus than in other cases of idiopathic facial palsy—despite treatment with high-dose steroids. Ten percent of patients with total facial nerve paralysis and 66 percent of those with partial paralysis recover completely. Recovery is better in those cases in which vesicles appear prior to nerve paralysis. Two percent of patients over age 50 will have severe, while nine percent will have moderate, post herpetic neuralgia.

The timing of the appearance of the vesicular eruption may have prognostic significance. In most cases, eruption and paralysis occur simultaneously. In approximately 25% of cases, the eruption precedes the paralysis, and the likelihood of recovery is higher in this group (Devriese and Moesker, 1988).

**Diagnosis**

The Tzanck prep, which is useful with herpes zoster, located on the truck in more peripheral nerve distribution areas, required toluidine blue staining a scraping from the blister. Occasionally, these blisters are located medially in the ear canal, and are quite small. So, often it is difficult to obtain adequate specimens for the prep. A positive prep demonstrates multinucleated giant cells. Vesicular fluid, when present, can be cultured with human diploid fibroblasts and after 3-5 days multinucleated giant cells within the fibroblast population can confirm clinical diagnosis. These studies, however, require five days or more to produce results. Laboratory confirmation of the diagnosis is based on increasing antibody titers in repeated complement fixation tests. Immunofluorescence of varicella antigen obtained from exfoliated cells from lesions can provide a more expedient verification of clinical suspicion. Often, diagnosis rests alone on the clinical criteria, defined by Dr. Hunt.

**Diagnosis: Immunological Evaluation**

Clinical diagnosis can be confirmed by either viral culture or fluorescence antibody testing using VZV identification reagent (Fluorescein isothiocyanate-conjugated monoclonal anti-VZV; Ortho Diagnostic Systems).

Hadar and colleagues from the Tel-Aviv University Medical Centers designed and tested VZV-specific IgG and IgA antibody titers in serial serum samples of 23 patients with Ramsay Hunt using immunoperoxidase assay. They demonstrated that all patients had VZV-specific IgG.
antibodies, but IgA can be a useful marker in confirming early diagnosis of the disease.

The mechanism of reactivation of VZV in herpes zoster oticus has not been clarified. Although the mechanism involved in general herpes zoster is also unclear, deterioration of cell-mediated immunity is thought to play a specific role as the “trigger” in reactivating the virus. The term "cellular immunity" colloquially refers to the T cell system. In fact, cell-mediated immunity is initiated by lymphokines produced by activated CD4+ T cells, activate macrophages and the precursors of CD8+ cytotoxic T cells. These effector cells then cause cell-mediated immune responses such as delayed- types hypersensitivity and cell-mediated cytotoxicity.

Nucleic acid hybridization and more recently polymerase chain reaction technology have confirmed VZV latency in human sensory dorsal root ganglia.

**Diagnosis: Audiological Evaluation**

In 1976, Byl and Adour were the first to thoroughly review the auditory symptoms and audiological data associated with herpes zoster oticus. They compared auditory symptoms in patient’s with Bell’s palsy and herpes zoster oticus. of 1080 patients with idiopathic facial paralysis, 29% of patients had auditory symptoms, while 37% of 172 patients with HZO had these symptoms. In their series of 1252 patients, 377 patients with facial paralysis had auditory symptoms. However, in only 11 of these 377 patients were abnormal cause-related sensorineural hearing loss documented with audiological testing. All of these patients had been diagnosed with HZO. These authors were the first to recommend a diagnosis in patients with idiopathic facial nerve paralysis and hearing loss—even when the characteristic vesicular eruption was absent. In their series, when recovery of auditory function occurs, a high-frequency sensorineural loss may persist, except in younger patients. Factors that appeared favorable for the recovery of hearing include not being older than 64 years, a mild initial hearing loss, a cochlear pattern of hearing loss, and absence of vertigo.

Wayman et al in 1990 retrospectively reviewed the audiological manifestations of herpes zoster oticus in 186 patients. In their study, active herpes zoster infection was confirmed by a four-fold increase between acute and convalescent complement fixation serum titres of VZV. Audiograms were performed on 152 patients, 82%, not all of whom had auditory symptoms at the time. Seven of these underwent more extensive
evaluation to determine a cochlear or retrocochlear pattern of hearing loss. Patients with a demonstrated cause-related hearing loss were treated with oral prednisone, 60mg/day for six days, then gradually tapered. Of the 152 initial audiograms, 93 or 61% were normal, 29 or 19% were cause-related abnormal, and 30 or 20% were unrelated abnormal examinations. This final group was excluded from their analysis. Vertigo was documented in 8 of 29 of the cause-related hearing loss patients and in only 5 of 93 (5%) of normal hearing patients \( p=0.002 \). Vertigo was more likely with increased severity of hearing loss in the isolated high-frequency hearing loss group. Of the seven patients who underwent more extensive audiological work-up, six had findings consistent with cochlear pathology: elevated SISI, ABLB recruitment pattern, type II Bekesy tracing, with no evidence of tone delay. The seventh patient had severe speech-frequency hearing loss with a speech discrimination score of 18%, type IV Bekesy tracing, evidence of tone delay, and grossly abnormal brainstem evoked response with marked delay or wave V latency, all suggestive of retrocochlear hearing loss. No correlation was demonstrated between severity of facial paralysis and presence of any hearing loss. The incidence of incomplete paralysis was 68% in the normal hearing and 62% in the cause-related abnormal hearing group. Extrapolating their clinical data to the previous histopathologic findings, especially of Blackley et al., these investigators suggested that a cochlear pattern represents inflammation confined to the cochlea while the retrocochlear pattern represents a more profound change involving the entire nerve.

**Diagnosis: Radiological Evaluation**

Magnetic resonance imaging has added a new dimension to the research of herpes zoster oticus and confirmed the histopathologic findings of early investigators. Images are taken with and without the paramagnetic agent Gadolinium as a contrast medium. Gadolinium does not normally cross the blood-brain barrier but this barrier is broken down in the presence of inflammation or edema. This results in increased signal density and enhancement in these areas. Another reason suggested for abnormal enhancement in these situations is venous congestion in the epineurium and perineurium. There is little dispute that the facial nerve enhances on MRI in the majority of patients with acute facial palsy, but the role of MRI in differential diagnosis and prognostic determination is not entirely clear. The majority of published studies refer to MRI of the facial nerve in idiopathic Bell’s facial palsy. It has been shown that even the normal facial nerve shows some mild to moderate enhancement of the geniculate ganglion and the tympanic-mastoid segment. Nearly twenty studies of the use of MRI in evaluation have been published. All the papers reported enhancement of
the facial nerve on MRI scanning in the majority of patients with facial palsy but only three papers suggested that the degree of enhancement or the anatomical level of the facial nerve enhancement had any prognostic significance. Brugel et al (1993) concluded that moderate enhancement in the geniculate ganglion as well as in the labyrinthine segment correlated with a good prognosis with respect to restoration of facial movement while an increased enhancement correlated with poor prognosis. Yanagida et al in 1993 noted that in subjects with Ramsay Hunt syndrome who experience internal auditory symptoms such as vertigo and tinnitus, enhancement was not only in the facial nerve but also in the vestibular and cochlear nerves.

**Treatment: Medical Management**

Introduced in 1977, the antiviral agent acyclovir has dramatically improved the treatment of herpesviridae infections. In the treatment of herpes zoster oticus, acyclovir therapy can be expected to produce House-Brackmann grade I-III recovery in most cases. Consequently, the success of antiviral therapy has greatly diminished enthusiasm for surgical decompression in cases of herpes zoster oticus with facial nerve paralysis.

Acyclovir remains the most widely prescribed and clinically effective antiviral drug available. It is 9-(2-hydroxy-methyl) guanine and a selective inhibitor of the replication of varicella-zoster and both herpes simplex types 1 and 2. It is converted by virus-encoded thymidine kinase to its monophosphate derivative, an event that does not occur to any substantial extent in uninfected cells. Subsequent disphosphorylation and triphosphorylation are then catalyzed by cellular enzymes resulting in acyclovir triphosphate concentrations that are 40 to 100 times higher in VZV-infected cells than uninfected cells. Acyclovir triphosphate inhibits viral DNA synthesis by competing with deoxyguanosine triphosphate as a substrate for viral DNA polymerase. Because acyclovir triphosphate lacks the 3’-hydroxyl group required to elongate the DNA chain, the synthesis of viral DNA is terminated. Furthermore, the viral DNA polymerase is tightly associated with the terminated DNA chain and is functionally inactivated. Viral polymerase has greater affinity for acyclovir triphosphate than does cellular DNA polymerase, resulting in little incorporation of acyclovir into cellular DNA. In vitro, acyclovir is most active against HSV-1, HSV-2, and VZV (average median effective concentrations 0.04, 0.10, and 0.50 µg per mL respectively.) Higher concentrations are required to inhibit replication of the Epstein Barr virus. Cytomegalovirus, which lacks a virus-specific thymidine kinase, is resistant.

Absorption from the GI tract is only 15% - 25% of the ingested dose. In
addition, the blood-brain barrier results in a 50% reduction of circulating acyclovir into cerebrospinal fluid. Thymidine kinase found in VZV has diminished affinity for acyclovir than TK found in HSV. For these reasons, the oral dose is more substantial than the recommended dose for herpes simplex.

A recent randomized prospective clinical study in the NEJM compared 7-day treatment of acyclovir to two other treatment arms: 21-day course of acyclovir with and without prednisone. Neither additional treatment reduced the frequency of post-herpetic neuralgia.

HSV can develop resistance to acyclovir through mutations in the viral gene encoding thymidine kinase, through the generation of thymidine-kinase deficient mutants or through the selection of mutants possessing a thymidine kinase that is unable to phosphorylate acyclovir.

Acyclovir-resistant isolates of VZV have been identified much less frequently than acyclovir-resistant HSV, but they have recently been recovered from bone-marrow transplant patients and AIDS patients. The acyclovir-resistant isolates all had altered or absent thymidine kinase function but remained susceptible to vidarabine and foscarnet. Acyclovir therapy has been associated with very few adverse effects. Renal dysfunction has been reported, especially in patients given large doses of acyclovir by rapid intravenous infusion. This appears to be an uncommon finding and usually reversible. Administering the drug in a slow infusion and ensuring adequate hydration can minimize the risk of administering acyclovir. On the other hand, oral acyclovir even at doses of 800mg five times daily has not been associated with renal dysfunction.

Several limited retrospective studies have been done to examine specifically the role of acyclovir in herpes zoster oticus.

Hall and Kerr from the Royal Victoria Hospital in Belfast reported in a 1985 edition of the Lancet the first use of acyclovir for herpes zoster oticus. They treated seven patients with HZO with a parenteral dose of 5 mg/kg three times per day. Within 3 days these investigators noted a striking improvement in the toxemia associated with this condition. Of the six patients who had total facial paralysis, four recovered completely, one partially recovered, and one patient had no recovery.

Stafford and Welch reported a year later from Newcastle upon Tyne another small series of only 5 patients. Each patient received intravenous
administration of acyclovir (5mg/kg) TID for a minimum of three days, followed by a two-week course of oral acyclovir. High-dose oral steroids (20mg PO QID in four cases; 10mg IVSS dexamethasone QID x 3 days) were prescribed for 5 days in full dosage, tapered gradually over another week. Each patient made a “satisfactory” recovery (4 patients with complete recovery by six months; one with partial recovery).

Dickens et al published their results in 1988. All patients received intravenous acyclovir 10mg/kg every 8 hours over a 7-day hospitalization period. Five of seven patients showed some return of facial function at the time of discharge. Their study suggests that prognosis depends on immediate initiation of therapy, and that multiple cranial nerve involvement did not appear to be a negative prognostic indicator.

Newer antiviral agents are available now for the treatment of herpes zoster. Valaciclovir and Famciclovir are both available in oral preparations and in more convenient thrice-daily dosing regimen.

Valaciclovir is the L-valine ester of acyclovir and essentially an acyclovir prodrug. It is rapidly and almost completely converted to acyclovir in vivo. Valaciclovir requires less frequent dosing than acyclovir due to its superior bioavailability over acyclovir. Acyclovir bioavailability is 3—5 times greater when administered as valaciclovir compared to 800 mg oral doses of acyclovir. Major adverse reactions include nausea (16%-19%), headache (11%-14%), vomiting (4%-9%), diarrhea (4%-6%).

Famciclovir is an oral prodrug for penciclovir and acts through a mechanism that is similar to that of acyclovir. Recommended dose for herpes zoster is 500mg PO TID for 7 days. It also is reported to diminish post-herpetic neuralgias.

Oral steroids have been used in zoster infections since the early 1950's. Some studies advocate the use of steroids along with acyclovir to reduce the incidence of post herpetic neuralgia and to enhance facial nerve recovery. Recovery of labyrinthine dysfunction after pharmacological therapy is not delineated in the current literature.

Case Presentation
A 62-year-old white male presented for consultation to the VAMC Otolaryngology—Head and Neck Surgery service. The patient reported a 3-day history of left facial weakness, left otalgia, and left hearing loss. There was no vertigo, nausea or vomiting, no recent otologic infection, and no recent trauma to the head and neck. On physical examination, a vesicular eruption was noted over the left concha and extending into the left external auditory canal. Grade IV House-Brackmann facial nerve weakness was noted. There was numbness and vesicular eruption along the V2 trigeminal nerve distribution. No nystagmus was elicited. Cranial nerves I-VI, IX, X, XI, XII were grossly intact. A diagnosis of herpes zoster oticus was made. Treatment was begun with oral regimen of acyclovir 800mg five doses per day. Oral prednisone (20mg PO TID) for 5 days was begun and gradually tapered over the next week. The patient was instructed in the use of artificial tears, lacrilube, and nightly application of a left eyepatch. At 3-month follow-up, the vesicular eruption had completely resolved and facial nerve function had returned to normal (House-Brackmann Grade I). There was no post-herpetic neuralgia.

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NEUROTOLOGIC MANIFESTATIONS OF HIV INFECTION
March 24, 1994
Thomas A. Salzer, M.D.

As health care providers we are faced with the prospect of caring for an increasing number of AIDS patients. The disease is no longer limited to specific populations and nearly everyone is at risk. Familiarity with the protean manifestations of AIDS is important for early diagnosis and treatment. AIDS is of particular interest to the Otolaryngologist-Head and Neck surgeon since 40-84% of patients have a symptom or physical finding in the head and neck region at initial presentation. Early diagnosis is essential because prompt initiation of treatment significantly diminishes morbidity and improves both quality and length of life in affected individuals. We will briefly review current statistics relevant to the AIDS epidemic, discuss the classification of HIV associated diseases and review in detail the neurotologic manifestations of HIV infection.
The causative agent for AIDS, HIV, is a retrovirus of the subfamily Lentovirinae. The term retrovirus is used because the virus contains the enzyme reverse transcriptase, with transcribes viral RNA to DNA- the reverse of other viral genetic transcription. HIV appears to be neurotropic and lymphotrophic and preferentially attacks T-helper cells, which are central to the function of the human cell-mediated immune system. Impairment of this system renders the host susceptible to numerous opportunistic infections from viruses, fungi, and protozoas, many of which are native to the oral cavity, pharynx and larynx. Aside from infectious consequences, HIV is also associated with malignancy, degenerative diseases and autoimmune sequelae.

OTOLOGIC-NEUROTOLOGIC MANIFESTATIONS
Otologic complaints are common in AIDS patients and include: hearing loss (62%), otalgia (50%), otorrhea (31%), vertigo (15%) and tinnitus (15%). It is not known if the neurotologic effects seen in AIDS is the effect of HIV alone or a combination of the effects of HIV infection coupled with opportunistic infections and/or possible toxic effects of certain therapeutic agents. The brain may be the site of tumors such as lymphoma or metastatic Kaposi’s sarcoma. The HIV itself may cause aseptic meningitis, subacute encephalitis and isolated cranial nerve neuropathy. The following case reports associate HIV infection with otologic diagnoses:

RAMSAY HUNT SYNDROME
Mishell and Applebaum 1990, reported a case of a 29 y/o homosexual male who was HIV+ with Ramsay Hunt syndrome associated with facial nerve palsy. The patient was treated with intravenous nafcillin and acyclovir.

HERPES ZOSTER MYRINGITIS
Kohan et al 1988 reported two cases of Herpes zoster myringitis which were successfully treated with acyclovir.

KAPOSI'S SARCOMA
KS is a slowly progressive, malignant mesenchymal tumor characterized clinically by red-purple plaques and nodules that may be seen externally or internally. Proliferation of atypical spindle cells within vascular channels is seen on histopathology. KS has been reported to involve the mastoid (Linstrom 1993) and external ear (Morris 1990).

GRADENIGO'S SYNDROME
Linstrom et al 1993 reported a 28 year old male IVDA with otalgia, purulent otorrhea, and sixth and seventh cranial nerve palsy secondary to petrous apicitis due to Aspergillus infection

PROGRESSIVE MULTIFOCAL LEUKENCEPHALOPATHY
Langford and Kuntz 1988 presented two adult patients with AIDS who had multiple cranial neuropathies resulting from progressive multifocal leukencephalopathy. Clinical symptoms included facial paralysis, trigeminal hypeaesthesia, hemianopsia, and deafness. Cranial MRI in both patients showed progressive PML, an unusual demyelinating disease seen in AIDS patients and thought to result from a direct effect
From the Grand Rounds Archive at Baylor of HIV in the cerebral white matter.

PNEUMOCYSTIS CARINII OTITIS / MASTOIDITIS
Pseudomonas carinii is the most common opportunistic pathogen infecting persons with HIV infection, affecting nearly 85% patients at some point of the illness. Otic Pneumocystis typically presents as a unilateral polypoid mass associated with otalgia, hearing loss and occasionally otorrhea. Pneumocystis carinii otitis media and mastoiditis have been. Pneumocystis carinii recovered from aural polyps is occasionally the initial manifestation of HIV infection in patients presenting to otologists for evaluation of otorrhea, otalgia and hearing loss. Pneumocystis carinii of the external canal has been effectively treated with local debridement and a 3-week course of Bactrim.

OPPORTUNISTIC INFECTIONS
Toxoplasmosis is the most common opportunistic infection to effect the CNS (Real, 1988). Kohan reported two such patients which presented with bilateral SNHL. Cryptococcal meningitis is known to be associated with SNHL in up to 25% cases (Real, 1987). Tuberculous meningitis is not infrequent in AIDS patients and is known to be associated with hearing loss.

OTOSYPHILIS
Otosyphilis must always be considered and investigated in a patient with fluctuating, asymmetric or sudden hearing loss. The diagnosis and treatment of syphilitic cochleovestibular dysfunction remains controversial, however the presumptive diagnosis is made in patients with cochleovestibular dysfunction, positive serology for syphilis and no other known etiology for the inner ear disturbance. Smith and Canalis, 1989 found otosyphilis to develop at an accelerated rate from primary syphilis in AIDS patients. All cases arose in patients previously treated for primary syphilis (latent syphilis). They propose that the HIV somehow hastens the development of otosyphilis effectively activating the disease. Furthermore they propose that all patients with otosyphilis should have an HIV test.

The index of suspicion should be high in the homosexual population because of the high incidence of syphilis in this group. 35-55% patients with AIDS have a past history of other sexually transmitted diseases including syphilis. Otosyphilis is well recognized as a cause of otologic disease in patients with AIDS (Morris and Prasad, 1990).

Gleich et al identified three factors associated with hearing improvement in patients with otosyphilis treated with intravenous penicillin and corticosteroids. In there study 31% patients with otosyphilis experienced hearing improvement, tinnitus decreased in 85% and vertigo improved in 86%. Specific factors associated with hearing improvement include: hearing loss present less than 5 years, fluctuating hearing, and age less than 60 years. Improvement was unrelated to the severity of the loss or previous treatment.

CENTRAL NERVOUS SYSTEM
HIV is frequently accompanied by neurological complications which are observed in 40% to 75% of patients during the course of the disease. De la Monte discovered CNS involvement in more than 90% of
HIV infected patients at autopsy. Neurological findings are detectable in approximately one third of AIDS patients. At autopsy however, up to of 75% subjects reveal neuropathological changes related to HIV infection. The most common neurologic syndrome seen in AIDS patients is subacute encephalitis. Early stages are characterized by subtle cognitive changes that may progress to dementia in several weeks or months. The HIV appears to be directly involved in both clinical and pathological changes in the CNS. Neurotologic workup may reveal central vestibular and auditory dysfunction. ENG findings show ataxic pursuit and optokinetic nystagmus with total loss of caloric excitability. ABR indicate absolute and interpeak latencies and synthetic sentence identification test yield reduced scores. At autopsy the AIDS virus was found in mononuclear and multinuclear giant cells in the cortical and subcortical gray matter, cerebral and cerebellar white matter and throughout the brain stem.

Hausler compared the incidence of peripheral and central auditory and vestibular disorders occurring in different stages of HIV infection. The results of audiological, vestibular and electrophysiologic tests performed on symptomatic and asymptomatic HIV-positive homosexual males with age matched seronegative homosexual males were compared. 57% of symptomatic and 45% off asymptomatic HIV positive patients had substantial abnormalities in comparison to minor abnormalities detected in 12% seronegative patients. The results suggested a high incidence of neurotologic disorders in HIV positive patients and that subclinical involvement of the auditory and vestibular system is common.

The frequency of electrophysiologic abnormalities in asymptomatic patients indicates subclinical involvement of the Central nervous system.

SENSORINEURAL HEARING LOSS
The incidence of SNHL in patients with HIV ranges from 23 - 49% . Kohan reported seven patients with persistent SNHL. In three instances the hearing loss could be traced to the use of well-known ototoxic drugs used in the treatment of AIDS. The remaining four patients were divided among CNS toxoplasmosis and cryptococcal meningitis, CNS toxoplasmosis alone, tuberculous meningitis and leutic otitis. For example, acyclovir may cause vertigo, trimethoprim-sulfamethoxazole may cause vertigo and tinnitus, and azidothymidine may lead to vertigo and hearing loss.

The characteristics of the SNHL in patients with HIV are not uniformly defined. The degree of hearing loss, range of loss and even incidence appear variable. Hearing loss in the high frequencies is more commonly reported than in the low frequencies. Delayed latencies of brainstem auditory-evoked responses have been observed in patients with HIV, suggesting neuropathies of the central auditory and vestibular region.

Boccellari found an association between immune suppression in HIV patients and neurophysiologic measures. Asymptomatic subjects without evidence of immune suppression do not appear to be at greater risk for neurophysiological impairment than HIV-negative subjects. The HIV-positive individuals with evidence of immunosuppression, however, appear to have an increased likelihood of central conduction time slowing. In contrast, studies by Pagano suggest that subclinical involvement of the upper brain stem occurs in HIV infection and that brainstem auditory evoked potential abnormalities could be the direct
result of the HIV effect on central nervous system structures.

Case Presentation

A 35-year-old man with human immunodeficiency virus infection diagnosed seven years ago was hospitalized for disseminated histoplasmosis which served as his AIDS-defining illness. During that admission he was also found to have Mycobacterium avium-intracellulare septicemia and bilateral external otitis secondary to Pseudomonas aeruginosa. CT scan of the temporal bones and gallium scan failed to reveal bony involvement. The otitis externa was treated with local debridement, culture specific intravenous antibiotics and otic drops. The patient experienced near total resolution of his symptoms with decreased otalgia, erythema and edema. Post treatment cultures were negative. Two months later he presented to the Otolaryngology service for evaluation and treatment of recurrent otitis externa which had progressed despite oral ciprofloxacin and Cortisporin otic drops. Examination revealed severe external otitis with erythema, edema and ulceration limited to the concha cavum and a right paranasal ulcer. The possibility of drug reaction was entertained and initial improvement was seen with withdrawal of the otic drops, however symptoms worsened thereafter prompting tissue biopsy of the ear and paranasal region for histology and culture. Histologic evaluation revealed multiple rralport inclusion bodies consistent with Herpes simplex infection. Audiometric evaluation was normal. The patient's condition improved dramatically on intravenous acyclovir.

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INFECTIOUS INTRACRANIAL COMPLICATIONS OF SUPPURATIVE EAR DISEASE

Andrew L. de Jong, MD
March 18, 1993

The existence and treatment of the intracranial complications of suppurative ear disease can be traced to the European Neolithic period. It is well-known that prehistoric man performed trephining, or the surgical opening of the skull with primitive instruments, for such infections. Until the 1940s the outlook for these patients remained grim, with a mortality of 75%. However, the introduction of penicillin by Fleming drastically changed this, decreasing mortality to 10%.

There are four well-known infectious otitic intracranial complications:
1. Epidural abscess
2. Subdural abscess
3. Meningitis
4. Brain abscess

The signs and symptoms of these complications can often be difficult to detect initially. Impending complications should be suspected when: 1) otologic infection recurs two to three weeks after initial treatment; 2) fetid aural discharge is present; 3) headache or subtle mental status changes in the presence of otologic disease; and 4) otalgia in the setting of chronic ear disease. These warning signs should prompt further investigation.

The most useful initial study is computerized tomography (CT) with contrast. If the CD findings are equivocal and the index of suspicion remains high, this should be followed by magnetic resonance imaging (MRI) with and without contrast. Therapy is tailored to each specific complication but should include intravenous antibiotics, medical management of elevated cerebrospinal fluid pressure, and neurosurgical consultation. Once the patient is neurologically stable, attention should be focused on definitive otologic intervention.

The pathogenesis, microbiology, and treatment of each otitic intracranial complication were discussed in detail.

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Case Presentation

A 20-year-old white woman with an extensive otologic history including left mastoidectomy in 1984 with revision in 1991, presented to another hospital with a five-day history of right-sided fetid aural discharge, otalgia, headache, and fever. She had been placed on oral antibiotics three days earlier by a private physician. On admission she was noted to be lethargic with neck rigidity, vomiting, and photophobia. Her aural exam was significant for the discharge and a posterior, superior deep retraction pocket. CT scanning revealed right mastoiditis and probable posterior fossa bony erosion. A lumbar puncture was consistent with meningitis, with the CSF fluid growing staphylococcus, proteus, and pseudomonas species.

The patient was transferred to BTGH on broad spectrum antibiotics. A right modified radical mastoidectomy with removal of a cholesteatoma was performed. A bony defect was found over the lateral sinus, but the sinus was patent. Postoperatively, DM had persistent headaches and neck pain. She developed somnolence, ataxia, and dysdiadochokinesia. CT and MRI imaging showed an infratentorial subdural empyema. A suboccipital craniotomy was performed by the neurosurgical team with removal of a sterile empyema. DM then made steady progress with only mild residual ataxia.
Bibliography


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Although seldom encountered in the practice of modern otology, lateral sinus thrombosis (LST) and other intracranial complications of otitis media still occur. The classic picture of this disease is often modified by prior antibiotic treatment, making the diagnosis and management difficult.

Infective thrombosis of the lateral sinus was first described in 1826 by Hooper. This disease was universally fatal until surgical intervention was established in 1888 by Lane. Despite later advances in surgical techniques mortality remained at nearly 50% until the introduction of antibiotics. Prior to the advent of antibiotic therapy, the mortality of all intracranial complications was extraordinarily high. In a study of autopsy statistics at LA County Hospital, it was found that before the introduction of antibiotics approximately 25:1,000 deaths were due to an intracranial complication of otitis media. The death rate
from these complications dropped 90% after the introduction of antibiotics.

The intracranial complications of otitis media include purulent meningitis, extradural or peridural abscess, LST, brain abscess and otitic hydrocephalus. Respiratory mucosa, intact boney walls and protective granulations provide natural defense barriers within the middle ear; complications occur when these are overcome. The spread of infection through the natural defenses can occur by osteothrombosis, bone erosion and when present along preformed pathways.

Classic symptoms of LST include a "picket fence" fever pattern; chills; progressive anemia (especially with beta-hemolytic strep); and, symptoms of septic emboli, headache and papilledema may indicate extension to involve the cavernous sinus. The Toby-Ayer test is measured by monitoring the CSF pressure during a lumbar puncture. No increase in CSF pressure during external compression of the internal jugular vein on the affected side, and an exaggerated response on the patent side, is suggestive of LST.

Since the introduction of antibiotics, some authors have noted that a high percentage of cases are due to chronic rather than acute cases of otitis media; however this finding has not been consistent in all reports. Teenagers and young adults are more commonly affected in modern reports whereas younger children were reported in higher numbers in earlier series. In the pre-antibiotic era streptococcus and staphylococcus were reported as causing the majority of cases of LST, recent reports have included anaerobic and gram negative organisms as well.

The diagnostic procedure of choice is MRI with MR angiography. The thrombus can be identified by its signal intensity on MRI and the flow void in the affected sinus is clearly documented on MR angiography.

Treatment is always surgical removal of the infected thrombosis in addition to broad spectrum antibiotic coverage. Once a highly controversial issue, ligation of the internal jugular vein is seldom necessary. In the majority of recent cases, anticoagulation has not been found to be necessary. However, it has been advocated by Shambaugh and may be indicated in selected cases.

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**Case Presentation**

A 6-year-old Hispanic male presented with no prior history of systemic illness or otologic disease. He was seen at Ben Taub General Hospital with a five day history of upper respiratory infection, headache, and bilateral otorrhea. Two days earlier swelling and tenderness had developed along the left sternocleidomastoid muscle. On arrival, his temperature was 101°F and white blood cell count was 18,300; he was alert and appropriate, but irritable and uncooperative. A CT scan was obtained that showed bilateral opacification of the mastoid air cells, and an abscess in the left upper neck. That night he was taken to the OR for incision and drainage of the neck abscess and complete otologic examination.
There was marked swelling of the left external auditory canal which precluded visualization of the left tympanic membrane. The right external auditory canal was also swollen, but the tympanic membrane could be visualized. The tympanic membrane was found to be thickened, a myringotomy was performed, purulent fluid was aspirated from the middle ear cleft and a ventilating tube was left in place. He was initially treated with Ceftriaxone (Rocephin) and Cortisporin otic drops. Cultures obtained from the right tympanic aspiration and the left neck abscess grew *Streptococcus pneumoniae* and a *Pseudomonas* grew from the left external canal specimen. Antibiotic therapy was changed to ticarcillin and clavulanate (Timentin). However, despite this, he continued to spike fevers to 101 and 102°F. On April 2, 1992 a second CT scan with thin cuts through the temporal bones was then obtained. This study suggested thrombosis of the left lateral sinus. While arrangements were being made for surgical decompression, an MR angiogram was performed. This confirmed vascular flow through the right sigmoid sinus and jugular vein, but no flow through the left lateral sinus or jugular vein. A few hours later, he underwent a left complete mastoidectomy with incision of the lateral sinus and removal of an obstructing thrombus.

Postoperatively, his antibiotics were changed to Ceftazidime (Fortaz), Amikacin and Penicillin G. Anticoagulation therapy was instituted for two weeks following surgical decompression. Subsequent MRA studies showed the development of collateral flow around the left sigmoid sinus and internal jugular vein. Additionally, reduced flow in the right system persisted. On April 21, 1992, he underwent right mastoidectomy. An MRA obtained on April 30, 1992 demonstrated unobstructed flow through the right venous system and no evidence of recanalization through the left sigmoid sinus or jugular vein.

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ACUTE MASTOIDITIS

February 3, 1994
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Acute mastoiditis is rarely seen today. Prior to the discovery of antibiotics, acute mastoiditis was the most common complication of acute otitis media and often resulted in death. The incidence has dropped significantly with the advent of antibiotics. Likewise, the frequency of mastoidectomy for this condition has decreased ten fold, down from 20% in 1938 to 2.8% in 1948 with a 90% decrease in mortality rate. Acute mastoiditis is a natural extension of acute otitis media. At the onset of infection, acute inflammation of the middle ear also involves inflammation of the mastoid air cells, which is not associated with bony resorption and must be differentiated from clinically significant mastoiditis.

Temporal bone development is integral in the ability to develop this disease. The degree of pneumatization varies greatly in temporal bones. Infection, heredity, ventilation, environment, and...
nutrition all play a role in the pneumatization process. Inflammation that develops is easily passed through these contiguous pneumatized regions. Not only can inflammation spread to these pneumatized regions, but it can spread to adjacent areas.

The evolution of acute mastoiditis begins when the mucosal lining of the pneumatized cells become inflamed and produce an exudate. Serosanguinous fluid eventually becomes mucopurulent. Spontaneous perforation of the tympanic membrane or myringotomy would halt the process at this point. However, 1-5% of these cases go on to the next phase. The cellular walls of the pneumatized cells then become demineralized due to: increased osteoclastic activity, pressure of the purulent exudate on the thin bony septae, and ischemia of the septae secondary to reduced blood flow. As the bony septae breakdown small abscess cavities form leading to coalescence. Finally the coalescent cells form an empyema, or pus under pressure, which then escapes to surrounding areas. Treatment of acute mastoiditis depends on the pathologic stage at which it is encountered.

The bacteriology of acute mastoiditis surprisingly differs from that of acute otitis media. The causative organism in acute otitis media are usually S pneumo or H Flu. However, acute mastoiditis is more commonly due to Group A Beta-Hemolytic Strep and S pneumo with rare involvement by H. Flu. Subacute and chronic disease is usually attributed to S. Aureus and gram negative rods such as E. Coli, Proteus and Pseudomonas.

The signs and symptoms of acute mastoiditis mimic severe acute suppurative otitis media; the disease entities are distinguished by the duration of symptoms. When the symptoms persist or recur after several weeks of acute otitis media, they point toward development of a coalescent process within the mastoid. The most common symptoms are otorrhea and otalgia. Subperiosteal abscess is noted by a fluctuant mass with overlying edema and erythema. This process produces displacement of the ear downward, outward, and forward. Sagging of the posterosuperior meatal wall occurs secondary to thickening of the periosteum overlying the bone in the area of the antrum. The tympanic membrane can simply appear normal, thickened or can demonstrate a small central perforation. Neurologic changes may be seen with intracranial complications. Perforation of the mastoid tip along the medial aspect of the SCM through the incisura mastoidea produces a deep abscess in the neck known as a Bezold's abscess.

Acute mastoiditis is one of the 10 basic complications of acute otitis media. Some worth noting are listed here. Petrositis is indicated by Gradenigo triad -- acute or subacute otitis media, retro-orbital pain and abducens palsy. Labyrinthitis due to suppurative ear disease is potentially fatal with spread to the cerebrospinal fluid producing meningitis. Presence of an extradural abscess is best evaluated by CT scan but must be excluded intraoperatively by visualizing normal dura through thin intact bone. Sigmoid sinus thrombosis may be asymptomatic or associated with toxemia or septic emboli. Griesinger's sign may be encountered and is noted by the presence of edema erythema of the posterior aspect of the mastoid process associated with mastoid emissary vein thrombosis. The Tobey - Ayer or Queckenstedt test is usually of historic significance and shows a rise in CSF pressure with occlusion of the normal IJ and NO change in CSF pressure with occlusion of the thrombosed vein. Brain abscess formation begins with cerebritis and should be closely monitored if suspected with CT scan every 1-2 weeks.
Evaluation of the patient with acute mastoiditis begins with the history and physical examination. The majority of the diagnosis is based on clinical judgment. Laboratory evaluation reveals a leukocytosis and elevated erythrocyte sedimentation rate. Mastoid radiographs are characteristic and will show cloudiness of the mastoid air cells associated with fuzziness of the bony partitions. Although helpful in the diagnosis, most agree that mastoid x-rays are not helpful in determining whether mastoid surgery is necessary. CT scan of the temporal bones is helpful in evaluation of concomitant intracranial complications as well as discerning any anatomical variations preoperatively. Chest x-ray is useful to rule out infiltrates in patients with septicemia and possible embolic phenomena secondary to lateral sinus thrombosis. Tympanocentesis is performed for guidance of antibiotic therapy.

Acute mastoiditis may be associated with periostitis, osteitis, or may be masked. This differentiation is key in the management of the patient. Acute mastoiditis with periostitis can be described as infection in the mastoid air cells which spreads to cause inflammation of the overlying periosteum. The route of infection may be through direct spread through a defect in the mastoid cortex or more commonly through venous channels, a.k.a. mastoid emissary veins. Periostitis should not be confused with subperiosteal abscess which requires surgical intervention.

Acute mastoiditis with osteitis is also known as coalescent mastoiditis. Less than 15% of patients with periostitis will progress to coalescence. This coalescence may spread in several directions. The spread of the pus anteriorly to the middle ear via the aditus ad antrum producing spontaneous resolution. If the pus spreads to the soft tissues lying anteromedially a Bezold's abscess may form. The infection may also spread lateral to produce a subperiosteal abscess. The spread of pus medially to the petrous air cells causes petrositis. Posterior spread to the occipital bone may result in osteomyelitis of the calvarium a.k.a. Citelli abscess.

Masked mastoiditis has an insidious course, owing to the use of broad spectrum antibiotics to treat middle ear disease. The apparent resolution of symptoms of acute otitis media "masks" the development of mastoiditis and leads to the presentation with intracranial complications. The presence of pathology may only be apparent on careful neurologic examination. Diagnosis is usually made on CT of the temporal bones which demonstrates temporal bone pathology in conjunction with the accompanying intracranial complication.

Acute mastoiditis can also occur in conjunction with chronic ear disease. It has been the teaching in the past that acute mastoiditis only occurred in well pneumatized mastoids in which the thin bony trabeculae are easily broken down. Chronic ear disease has long been associated with a sclerotic "cue - ball" like mastoid which is less susceptible to demineralization. However, several studies note the presence of cholesteatoma in patients with acute mastoiditis.

Antibiotic therapy is guided by the patients history. If the history is otherwise uncomplicated by ear disease or protracted episode of otitis media, the infecting organism is probably S. pneumo or S. pyogenes which is best treated by ampicillin. If the patient has a protracted course of otitis media then coverage for S. Aureus and gram negative organisms is necessary with a penicillinase resistant penicillin.

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and an aminoglycoside or single antibiotic coverage with a cephalosporin. Anaerobic coverage should be added to the above therapy when suspected.

There exists controversy over the management of this disease. All studies on acute mastoiditis are retrospective and conclusions drawn regarding therapy are limited. There have been no prospective or controlled studies on the surgical and medical management of the disease. This is limited by the relative rarity of the disease in the current day. Options for treatment are antibiotics alone, antibiotics plus myringotomy, Incision and drainage of abscess, and mastoidectomy. If periostitis is present tympanocentesis followed by myringotomy should be performed with placement on intravenous antibiotics directed toward gram stain results. It is preferable to insert pressure equalization tubes for drainage over a longer period of time. This periosteal involvement usually resolves within 24 to 48 hours after initiation of therapy. If clinical deterioration occurs, manifest by persistent fever and otalgia, or development of subperiosteal abscess, then further surgical intervention is indicated. Analysis of the literature reveals approximately 57% of cases are able to be treated in this manner.

Indications for surgery include acute mastoiditis with subperiosteal abscess, acute mastoiditis not responsive after 24 to 48 hours of intravenous antibiotics and myringotomy, and intracranial complications with evidence of mastoid coalescence. These indications were derived from the current literature.

Some have managed the disease with simple I & D of the abscess which does not afford decompression and drainage of the underlying mastoid. There have been no studies to evaluate the efficacy of this mode of therapy versus mastoidectomy. Most reported cases of simple I & D have been anecdotal. 60% of the cases reported in the literature have come to mastoidectomy after I & D.

The procedure of choice is a complete simple mastoidectomy. Simple mastoidectomy is an emergency procedure and should be performed on a clinically stable patient with control of sepsis before undergoing anesthesia. Mastoidectomy may be performed at a later date if the patient is unstable after drainage of intracranial process. The goal of surgery is to effect drainage. Establishment of good communication between the middle ear and mastoid is achieved by removing the edema and granulation in the aditus ad antrum. External drainage is necessary with a drain in the mastoid cavity to drain the antrum, and a pressure equalization tube inserted to ventilate the middle ear space.

In conclusion, acute mastoiditis is a diagnosis based on clinical acumen. When acute mastoiditis is suspected a careful history and physical examination should be ascertained to evaluate for intratemporal or intracranial complications. The presence of acute mastoiditis does not exclude the presence of underlying chronic ear disease, and should be taken into account in the evaluation and treatment of the patient. The presence of subperiosteal abscess or progression of symptoms on the above therapy warrants surgical intervention. Simple mastoidectomy is the surgery of choice. Prospective controlled studies are necessary to firmly identify indications for medical versus surgical therapy.
Case Presentation

An 11-year-old boy with a history of chronic otitis media with effusion presented with a 10-day history of fever, right otalgia, and a right, dull occipital headache. The headache was not associated with photophobia. A CT scan of the head revealed a right parieto-occipital epidural abscess. Physical examination revealed a young, alert boy with a temperature of 102°F. Examination of his right ear revealed a thickened, but intact tympanic membrane and middle ear effusion. Otorragic edema, erythema, tenderness, and fluctuance was evident. The remainder of the physical examination, including a neurological exam, was normal. White blood cell count was 18.7 cells/mm3 with a left shift. CT scan of the temporal bones revealed soft tissue changes within the middle ear and mastoid and an overlying subperiosteal abscess and possible lateral sinus thrombosis.

The patient was taken to the operating room and underwent bur hole drainage and irrigation of a right parieto-occipital epidural abscess by the neurosurgical team followed by a right simple mastoidectomy, exploration of the lateral sinus, insertion of a pressure equalization tube, and Penrose drainage of the mastoid cavity. Findings included coalescent mastoiditis and no evidence of lateral sinus thrombus. Intraoperative cultures grew microaerophilic streptococcus; the antibiotic regimen was changed to Cefotaxime and Penicillin G. The right temporoparietal cerebritis showed no progression to brain abscess and no neurologic sequelae. Drains were discontinued when drainage decreased. The patient was discharged on the thirteenth postoperative day in good condition to continue intravenous antibiotics for a total course of 6 weeks. Weekly CT scan of the head demonstrated slow resolve of the cerebritis. The patient has had no further episodes of otitis media and is currently doing well in the sixth grade.

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Although the underlying etiology of Meniere's syndrome is unknown, a consistent histopathologic finding is hydrops (dilatation) of the endolymphatic spaces. The hydrops presumably results from a malfunction of the resorptive function of the endolymphatic sac. The classic constellation of symptoms includes: fluctuating hearing loss, episodic vertigo, tinnitus, and a sensation of fullness in the ear. These symptoms, however, do not necessarily develop simultaneously and many patients do not develop them at all. Subcategories of Meniere's syndrome describe these other conditions; for instance, cochlear hydrops (fluctuating hearing loss alone) or vestibular hydrops (vestibular symptoms without hearing loss).

In most patients, Meniere's syndrome is ultimately self-limited; over time the patient suffers deterioration of hearing and a gradual subsiding of the episodic dizzy spells. This evolution, however, may require 1 to
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2 decades. In the interim, the patient's lifestyle may be severely impaired.

A few patients cannot be adequately managed by medical means alone and surgical intervention must be considered. The surgical procedures for Meniere's disease may be categorized as those designed to alter the function of the vestibular system and those that ablate the vestibular system either with or without preservation of hearing.

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**Case Presentation**

A 58-year-old white female noted the onset of fluctuating right hearing loss, right tinnitus and vertigo eight years prior to presentation. For the first 5 years her symptoms were controlled with sodium restriction and diuretics. Three years prior to presentation she began to experience intense episodic vertigo associated with nausea and vomiting. Two years prior to presentation she underwent a right endolymphatic shunt procedure with some relief of her symptoms. After a 6-month period the vertigo and dizziness returned. She underwent a revision of the endolymphatic shunt 1 year prior to presentation, again followed by initial relief and then recurrence of symptoms. She was referred to The Methodist Hospital for further evaluation. Preoperative evaluation showed predominantly low-frequency sensorineural hearing loss in the right ear, with a PTA of 45 and a PBmax of 40%. Left ear showed a PTA of 10 and PBmax of 100%. ENG showed right unilateral weakness. ABR was normal bilaterally. The patient underwent a right middle cranial fossa vestibular nerve section. Postoperatively the patient did well and has been free of vertiginous episodes.

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Introduction

Tympanoplasty without mastoidectomy is performed to control infection through eradication of disease and to reconstruct the sound conducting mechanism. Tympanic membrane grafting may or may not be required. This presentation focuses on tympanoplasty types I through III and ossiculoplasty. It is not a detailed discussion of otitis media, mastoidectomy or stapedectomy.

Anatomy

Horst Wullstein (1956) said, "The tympanic membrane has two functions, sound pressure transformation
for the oval window and sound protection of the round window." The outer ear functions to capture air pressure waves. The middle ear functions to convert air pressure waves efficiently into endolymphatic fluid waves, and the inner ear (cochlea) serves to convert fluid waves into nerve impulses. This sound-transforming mechanism includes the ossicles whose lever action adds 3 dB to the sound level at the oval window. The hydraulic principle explains how the difference in the surface area of the tympanic membrane and the stapedial footplate increases the hearing level by 27 dB. The tympanic membrane also serves to protect the round window from sound by efficiently conducting the pressure waves to the oval window and delaying the arrival of pressure waves to the round window.

Middle ear pathology associated with perforation or a conductive deficit includes otitis media, otitis externa, granulation tissue and trauma including myringotomy. Other disorders can interfere with the ventilatory or conducting function of the middle ear without producing perforations. These include tympanosclerosis, otosclerosis, congenital cholesteatoma, and Eustachian tube dysfunction.

The Eustachian tube, or pharyngotympanic tube, originates in the middle ear and extends into the nasopharynx. It is normally closed, but opens for 0.1 to 0.2 seconds during swallowing to allow air to move between the nasopharynx and middle ear to equalize the pressure across the tympanic membrane. There is no doubt that Eustachian tube dysfunction can lead to negative pressure in the middle ear and retraction of the tympanic membrane. But, we cannot distinguish which patients have retractions from previous tubal dysfunction and which patients have persistent tubal dysfunction and will tend to reform retractions after surgery.

**Preoperative Evaluation**

The examiner should make note of otorrhea, hearing loss, otalgia, vertigo and facial weakness when evaluating a patient with middle ear pathology. A hearing loss of 30 dB or more may represent ossicular disruption. Progressive loss with no obvious middle ear pathology on exam may represent tympanosclerosis or otosclerosis.

A microscopic exam should be performed and pneumatic otoscopy used to assess the mobility of the tympanic membrane and the malleus. A Fistula Test can be performed if there is a history of dizziness or a marginal perforation.

**Surgical Technique**

The particular technique that will best repair the tympanic membrane is impossible to determine preoperatively with confidence. The final decision regarding medial or lateral placement of the graft or the ossiculoplasty to be performed must be reserved until the ear has been examined under anesthesia.

Wullstein created a classification scheme in 1956 identifying five basic types of tympanoplasty. Type I is myringoplasty. The Type II tympanoplasty is for tympanic membrane perforations with erosion of the malleus. It involves grafting on to the incus or the remains of the malleus. The Type III tympanoplasty is
indicated for destruction of the lateral ossicles, but with an intact and mobile stapes. It involves placing a
graft onto the stapes and providing protection for the round window. The Type IV tympanoplasty is used
for ossicular destruction including destruction of all or part of the stapes arch. It involves placing a graft
onto or around a mobile stapes footplate. The resulting middle ear consists of the hypotympanum and the
Eustachian tube orifice only. The impedance matching system is abandoned. The Type V tympanoplasty
is used when the footplate of the stapes is fixed. The Paparella modification Type Va with fenestration of
the horizontal canal, has largely been abandoned in favor of Type Vb with stapedectomy. Farrior (1971)
proposed subclassifications based on the resultant ossicular/ prosthesis arrangement.

There are two basic approaches to tympanoplasty without mastoidectomy. The retroauricular and the
transaural approaches are equally successful, but most surgeons choose a retroauricular approach to
anterior perforations.

Tympanic membrane grafting can be accomplished by medial or lateral grafting. Medial grafting, also
known as the underlay technique, involves creating a tympanomeatal flap via a canal incision and
elevation of the annulus and tympanic membrane. Graft material is then secured between the tympanic
membrane and a bed of Gelfoam placed in the middle ear. Lateral grafting can be performed with a
variety of materials. One technique uses an overlay graft of temporalis fascia and free canal skin. This
produces a thick, well-vascularized graft that will clean itself by epithelial migration, but care must be
taken to maintain the acute angle between the drum and the canal anteriorly, which is essential for a good
hearing result. Recurrent retraction pockets can be prevented for the most part, by cartilage grafting and
scutumplasty.

Graft Materials

Materials used for tympanic membrane repair include autologous grafts (from the same person),
homografts (from other humans), xenografts (from animals), or allografts (synthetic materials). Rambo
(1958) described the use of autologous muscle in a procedure he called a musculoplasty. Fat patch
myringoplasty is an autologous tissue technique. Autologous temporalis fascia and canal skin are used
for medial and lateral grafting. Replacing the tympanic membrane with cartilage prevents subsequent
retractions, but obviously shielding large portions of the drum with cartilage causes a substantial hearing
loss. A composite cartilage-perichondrium graft taken from the tragus was evaluated by Poe and Gadre
(1993), who found that reinforcing the posterior quadrants and pars flaccida with cartilage did prevent
retractions.

Homograft materials used in tympanoplasty have included dura, tympanic membranes, ossicles, corneas
and banked cadaveric materials such as tympanic membranes with attached mallei, including sclera,
infant dura, and nasal septal cartilage. One potential drawback of homografts is the theoretical risk of
transmitting pathogens such as HIV. The fixation process inactivates viral particles and the remaining
material is organic but non-viable. The grafts are implants not transplants. Yet, the fear of litigation has
decreased the number of hospitals willing to bank organic materials. Other potential disadvantages
include the additional time required to sculpt and the cost of tissue banking.
Allograft membranes incorporating wire, metal, ceramic, and plastic, have been largely unsuccessful according to Campbell (1990). The disadvantage of using allografts is their tendency for extrusion.

**Ossiculoplasty**

The ossicles may become fixed or lose continuity. They may be eroded by a mass or inflammatory process, or they may be congenitally malformed. Any of these can prevent the transmission of sound to the inner ear.

Austin, in 1971, devised a system for classifying ossicular defects. He reasoned that loss of the malleus head or neck was not important to reconstruction, and that the incus would be mobilized for reconstruction. So, the condition of the handle of the malleus and the stapes superstructure is the relevant issue in ossiculoplasty. He classified the four possible defects as types A through D, and noted that most commonly both the incus and stapes are present. The next most common situation is to have an isolated defect in the stapes arch. This scheme can aid in planning reconstruction.

Since 1964, homograft ossicles have been used to reconstruct the ME mechanism. The incus could be prepared in two ways. The notched incus with short process can connect the malleus to the capitulum of the stapes. The notched incus with long process can connect the malleus to the stapes footplate when the stapes superstructure was missing or removed. In 1986, hydroxylapatite prostheses became available in two lengths as an incus or incus-stapes substitute.

Several maneuvers are key to the success of an ossiculoplasty. Special attention should be given to dislocating the incudostapedial joint, ensuring mobility of the footplate, removing mucosa from the malleus handle, and popping the prosthesis in place. Malleus head fixation is addressed by dislocation of the I-S joint followed by dislocation of the I-M joint and removal of the malleus head. Erosion of the manubrium of the malleus can be addressed by reconstruction with a homograft tympanic membrane and malleus or use of a PORP (malleus-incus) prosthesis. If the malleus handle and the stapes arch are absent, a TORP (malleus-incus-stapes) can be used.

There are a wide variety of prostheses available to the surgeon. The stapes prostheses are either wire loops or pistons. Some of the pistons have a cup rather than a crook to hold the incus. The incus prostheses come as struts or struts with side arms. Incus-stapes prostheses come in more than 9 types. They are struts or pistons of stainless steel alone or together with a polymer.

PORP's (partial ossicular reconstruction prostheses) are for lateral chain reconstruction in the absence of the malleus handle. They have a broad lateral platform designed to minimize point pressure on the TM to resist extrusion. There are 13 TORP's (total ossicular reconstruction prostheses), available for use as malleus-incus-stapes substitutes. Ossicular prostheses range in price from about $39.00 to $263.00, and are constructed of stainless steel, platinum, tantalum, polyethylene, Plasti-pore, fluoroplastic, and hydroxylapatite.
Staging Tympanoplasty

In 1991, James Sheehy reviewed the current thinking regarding staging of tympanoplasty in badly diseased ears. Staged operations have been preferred since the mid-1960's at the House Institute and elsewhere. The first operation would eliminate disease and aerate the middle ear cleft, and the second would reconstruct the sound pressure transfer mechanism. There are three requirements for a successful functional result in tympanoplasty: 1) a tympanic membrane; 2) an air-filled, mucosa-lined middle ear space; and, 3) a secure connection between the vibrating tympanic membrane and the inner ear fluids.

The need for staging is determined in part, by the second of these: a mucosa lined middle ear space. If there is minimal mucosal disease at the time of the initial operation, there is no need for staging and the conductive mechanism may be reconstructed immediately. Often large areas of mucosa are diseased. Infection must be controlled before healthy mucosa can regenerate. Edematous mucosa and uninfected granulations are reversible. Mucosa should not be stripped if there is any possibility of rejuvenation, because denuded areas are prone to formation of synechiae or adhesions, scarring or granulation. Such scarring can impair drum and ossicular mobility. Another obstacle to the regeneration of the mucosal lining is the presence of residual squamous epithelium in the middle ear. Remnants from a cholesteatoma or epithelium migrating medially through a perforation may take root on denuded bone in the middle ear.

If removal of the middle ear disease denudes large areas of the medial wall, staging is necessary to prevent fibrosis of the middle ear cleft. Staging may also enhance the hearing result by allowing the new drumhead to "settle" before ossiculoplasty, thereby reducing the risk of lateralization (Donaldson and Snow, 1992).

Some believe that although staged procedures may produce slightly better hearing result, they are often unnecessary. This is because many patients with extensive middle ear disease heal amazingly well after the first procedure. The inability to predict who will heal well leads some to reconstruct the conductive mechanism in a single procedure hoping for a good result. In this way, those who do well are spared a second operation (Wehrs, Schuknecht, Austin, Gacek, Jansen, and Shea).

Middle Ear Stents

Exogenous and synthetic materials have been used to prevent adhesions within the middle ear after resection of mucosa for disease. Rambo (1961) recognized this problem and began filling the middle ear cleft with paraffin. In the mid-1960's, thin plastic sheets of polyethylene, Teflon and silicone rubber (silastic) were used. Occasionally, this sheeting was pushed laterally by a bed of scar tissue and extruded. This led to the use of thicker pieces of 0.04 inch silicone rubber or 0.3mm Supramid.

More recently, biodegradable substances such as Gelfilm and Gelfoam (purified gelatin matrix) have been used. They do not require removal at a later time and will not extrude. The disadvantage comes when the material is absorbed and no longer stents the middle ear. Some do not stent the middle ear space at all, but rather avoid adhesions by leaving the perforation ungrafted until the second stage.
Donaldson and Snow (1992), in a prospective study of 71 patients found a better long-term hearing result in patients who had mastoidectomies and silastic implants as a part of their first stage procedure. Some suggest that these measures improve middle ear ventilation by increasing the "air-cushion" available to resist eustachian reflux.

Surgical Indications and Contraindications

The indications for surgery are conductive hearing loss due to TM perforation or ossicular dysfunction; chronic or recurrent otitis media and recurrent otitis media due to contamination through a perforated tympanic membrane; progressive hearing loss due to chronic ME pathology; perforation or hearing loss persistent for more than three months due to trauma, infection or surgery; and, the inability to bathe or participate in water sports safely due to perforation of the TM. Normal hearing is not a contraindication to surgery in the presence of disease, because secretions and granulations may close a perforation or protect the round window thus improving a conductive deficit, or a cholesteatoma might conduct sound as a functional portion of the ossicular chain.

Glasscock (1976) listed four absolute contraindications and seven relative contraindications to tympanoplasty. Absolute contraindications are uncontrolled cholesteatoma, malignant tumors, unusual infections, and complications of chronic ear disease such as meningitis, brain abscess or sinus thrombosis. The relative contraindications include Eustachian tube insufficiency, uncooperative patients, a dead ear, a better hearing or only hearing ear, elderly patients, young children and cases of repeated failure.

He stated that the factors considered in the surgeon's decision to close the ear or leave it open constitute the indications or contraindications for tympanoplasty. He stressed that tympanoplasty has been controversial from the start because filling an infected cavity with foreign material and then closing it off to the outside, flies in the face of classical surgical training.

Much has been made of the presence or absence of discharge in the ear at the time of surgery. Several studies found that the secretion type (dry vs. mucoid or purulent) in the middle ear at the time of surgery had no effect on the final success rate for myringoplasty. Poor eustachian function as evidenced by contralateral effusion or atelectasis predicts poorer results (60% success).

Post-Operative Care

One goal of post-operative care is to keep the patient comfortable. Infection is generally prevented by antibiotic-soaked canal and middle ear packing and host defenses. In order for the graft to heal it must remain in contact with organic matrix or stroma, be free from infection, and not experience shearing forces or excessive tension. Aural hygiene is aimed at achieving this environment. Maneuvers that change the transtympanic pressure are forbidden, such as sneezing with the mouth shut, using a straw to
Complications

Possible complications of tympanoplasty include: failure of the graft to heal resulting in residual or recurrent perforation, lateralization of a graft, canal stenosis, scarring or adhesions in the middle ear, perilymph fistula and hearing loss, erosion or extrusion of a prosthesis, dislocation of a prosthesis, facial nerve or chorda tympani injury.

Other problems may or may not be surgical complications, such as recurrence of cholesteatoma or atelectasis of the middle ear space with retraction of the TM. The patient should be informed of these preoperatively, and understand the importance of aural hygiene and follow-up care.

Pediatric Tympanoplasty

Timing of repair in the pediatric population is very controversial. Glasscock (1976) gave young age as a relative contraindication to tympanoplasty because children under three or four are prone to upper respiratory infections and otitis media. Koch et al (1990) reported an 81% success rate for children age 8 and older, but only a 30% success rate in younger patients. They concluded that tympanoplasty before age 8 results in a high rate of failure because of poor Eustachian tube function and frequent URI's. Smyth (1992) agreed, noting that patients less than 10 years old had a higher failure rate for myringoplasty than older children. This was independent of secretion type, perforation site, and graft material.

Yet, others such as Lau and Tos (1986) found no significant difference in outcome between the 2 to 7 age group and those children ages 8 to 14. They suggested that early operation may prevent progression of ossicular chain resorption. Ophir et al (1987) reported a 79% overall success rate, and their success in younger children (5-8) was comparable to the rate for older children. They concluded that myringoplasty had a good chance of success at any age. Kessler et al (1994) reviewed the results of 209 myringoplasties and concluded that even in young patients (2-6 years) myringoplasty has a high success rate (75-94%), and that age alone could not be considered a contraindication to surgery.

Surgical Outcome

In his 1992 Toynbee Memorial Lecture, Gordon Smyth of Belfast stated that any hope for cost-effective health care reform required urgent self-audit and the provision of more realistic advice to patients. He took exception to the generally quoted success rate for myringoplasty of 90%. When the success rate is corrected for the length of follow-up, using Survival Life-table Analysis, the success rate for maintaining a healed TM diminished to 81% at 11 years. He also suggested that the success of ossiculoplasty should be measured as the effect on binaural hearing, rather than the air-bone gap on the operated side. This is because the operated ear must reach an air conduction level of 30 dB at speech frequencies or be within 15 dB of the other ear for the patient to benefit (Belfast rule of thumb) (Smyth and Peterson, 1985). Smyth warned that, "Fiction and fact need untangling, otherwise surgeons are little better than gossips."
Summary

In summary, tympanoplasty without mastoidectomy is performed to eradicate middle ear disease and reconstruct the conductive hearing mechanism. Priorities in tympanoplasty are to prevent recurrence, to improve hearing and to minimize ear after care.

Though most would agree that myringoplasty has a high success rate and that staged operations optimize the hearing result, controversy abounds in other areas. Specifically we need to standardize outcome measures. To say that a tympanoplasty failed or succeeded in a given case is meaningless if our criteria are different. 1) It seems reasonable to use post-operative sensorineural hearing level rather than the pre-operative level to calculate closure of the "air-bone gap." 2) When reporting pure tone average, "air-bone gaps," these results should be tabulated in increments of 5 dB. 3) We should agree to report effusions or retractions as numbers without lumping them into surgical failures.

We need studies with longer follow-up times. This is difficult for three reasons. 1) The population is highly mobile in this country. 2) Patients no longer choose which specialist they will see or when they will see him. Attempts to control medical costs may arbitrarily limit follow-up for asymptomatic individuals. 3) Patients who are doing well may not see the benefit of follow-up, and conversely, patients with poor results may blame their surgeon and seek referral to a different otolaryngologist. The solution to this problem lies in improved physician to physician communication. Pre-operative patient counseling that stresses the importance of follow-up may help as well.

We need more studies that limit the variability of the population by focusing on a single type of procedure or pathology. The solution to this problem lies in cooperative studies to increase the population in each study.

Finally, we need improved means of measuring Eustachian tube function. It may be necessary to measure small gradual pressure changes on an ambulatory basis, like esophageal pH monitoring, in order to gain clinically relevant information.

Case Presentation

A 20-year-old man with a history of chronic otitis media as a child, requiring the placement of tympanostomy tubes on three separate occasions, presented with a left myringotomy that failed to heal after the last set of tympanostomy tubes. He developed a chronic perforation but rejected surgical repair in favor of expectant management and meticulous aural hygiene. He has had no ear infections in the last year. He has no complaints of decreased hearing, otorrhea, tinnitus, dizziness, or ear pain.

Recently, he returned for follow-up examination and requested a tympanoplasty so that he could swim,
bathe and exercise without concern for ear protection.

He has no family history of chronic middle ear disease. He has seasonal allergies, but otherwise has no medical problems. He takes no medications and has no drug allergies.

On physical examination he has a 3cm x 3cm hemangioma of the soft palate on the right. Examination of the left ear reveals a normal external auditory canal; the tympanic membrane has a 30% posterior marginal perforation with ragged edges and an adjacent posterior-central monomer. The middle ear mucosa appears normal. The long process of the incus and stapes appear normal.

He underwent left lateral graft tympanoplasty via a retroauricular approach. The middle ear exploration revealed no pathology. A type I tympanoplasty was performed using a temporalis fascia and free canal skin overlay technique.

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OSTEODYSTROPHIES OF THE MIDDLE EAR AND TEMPORAL BONE

February 9, 1995
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Five major osteodystrophies that affect the middle ear and temporal bone are presented: Paget's Disease, Osteogenesis Imperfecta, Osteopetrosis, Fibrous Dysplasia, and Osteitis Fibrosa Cystica. The epidemiology, pathophysiology, clinical features, histopathology, diagnosis, treatment and prognosis of these five disorders are discussed.

Paget's Disease

The classic description of a patient with Paget's disease is that of a deaf old man with a bent back, bowed legs, curved tibia, a large skull and a wobbling gait. However, these advanced changes are not frequently seen. Czerny (1873) was first to describe this disease, but it was Sir James Paget (1877) who correlated
and described the clinical and pathological features of this disease. Paget's disease is a chronic, progressive disorder and is believed to be AD or X-linked. The disease primarily affects older people and commonly presents in 6th decade. There is evidence for a viral etiology, possibly a slow virus. Diagnosis is based on history, exam, radiographic and laboratory findings. Frequently, the alkaline phosphatase is often grossly increased; the acid phosphatase is often elevated - prostate cancer should be ruled out. The calcium is usually normal. Demineralization of bone is seen radiographically, such as the well-known osteoporosis circumscripta and cotton-wool patches of the skull. The histology characteristically shows osteoclastic bone resorption and a mosaic pattern of new, lamellar bone interspersed with marrow spaces that become obliterated with fibrous tissue. Typically, pagetoid bone and microfractures are seen in the temporal bone.

Symptoms are mainly due to the excess osteoid and resulting soft, easily deformed bones. Hearing loss occurs in 5% - 44%: mixed, CHL or SNHL. CHL component is very rare due to ossicular fixation and surgery is not recommended. Treatment consists of symptomatic relief. Salmon calcitonin can promote healing of osteolytic lesions, and stabilize hearing. Prognosis is generally good; however, there is a 20-fold risk for developing osteosarcoma in the pagetoid lesions and these malignancies are very aggressive lesions and portend a poor prognosis.

**Osteogenesis Imperfecta**

Osteogenesis Imperfecta results from a defect in the biosynthesis of the alpha 1 or alpha 2 chains of type I collagen. This results in bone fragility with multiple fractures, bone deformities, thin skin, and capillary fragility with increased bleeding tendency. Histopathologically, those with the most severe form possess thin, immature cortical bone with small, irregular spicules of bone, decreased osteoid matrix and increased fibrous tissue and vascular spaces. A thin, fragile stapes is typical. Those with less severe forms have hypercellular, nonlamellar (immature) bone which fails to mature; there are large remnants of cartilage in the endochondral bone and delay in ossification of the endochondral layer. Radiographically this disorder appears similar to osteoporosis with demineralization / deossification of bone and sometimes with bowing of long bones, thinning of the cortices and exuberant calus formation at fractures. In the skull, embryonic microfusion lines or "wormian bone" is seen. A "halo sign" of demineralization of the otic capsule, similar to otosclerosis, may be found.

Previously in two groups (congenita and tarda), they are now grouped in four main categories: 1. blue sclera and an associated CHL; 2. most severe form; 3. blue sclera and an associated CHL, more severe than 1, and associated with skeletal deformities; and, 4. similar to 1, but associated with grey sclera and normal hearing or SNHL. Severe SNHL occurs in 40% of cases and correlates with gray or normal sclera. CHL reflects structural changes in the ossicles and usually correlates with patient with blue sclera. The CHL may be due to soft or missing stapes crural arch, due to otosclerosis or due to a combination of the two. Otosclerosis commonly occurs in association with OI and the histologic independence of these two disorders has been clearly demonstrated. Otosclerosis in these patients tends to be more aggressive and there is a higher incidence of SNHL and window obliteration. Medical treatment with calcium, vitamin D, calcitonin, and anabolic steroids have not been shown to be helpful. Treatment is aimed at function; bracing and orthopedic stabilization is provided as needed, and physical therapy to maintain
good range of motion and muscular tone (e.g. swimming) is encouraged. Stapedectomy may yield good results for CHL.

Osteopetrosis

Osteopetrosis is due to defective osteoclast function with failure of normal bone resorption, while normal osteoblastic bone formation continues with excess deposition of mineralized osteoid and cartilage and displacement of marrow spaces. Radiographically, one sees increased density of all bones with loss of marrow spaces and obliteration of mastoid air cells and paranasal sinuses. Later, the periosteal bone becomes sclerotic without marrow spaces, resulting in dense, brittle bones and anemia. In infants, the temporal bone is primarily dense, calcified cartilage; the stapes exists in fetal form; dehiscence of VII is a consistent finding.

Patients with the "tarda" form may be asymptomatic or they may have syndactyly, easy fracturing of bones, bone pain, osteomyelitis, and skull thickening, and, possibly, narrowing of foramina of optic, trigeminal, facial, and auditory cranial nerves. CHL is due to encroachment of bone on the middle ear space with entrapment or ankylosis of the ossicles. Those with the congenital form die at an early age from anemia or sepsis (none live more than 20 years). Bone marrow transplantation from HLA-identical sibs has been used successfully with reversal of anemia & osteosclerosis.

Fibrous Dysplasia

Fibrous dysplasia usually presents in the 2nd or 3rd decades and is a chronic, slowly progressive, but not self-limiting disease of unknown cause and without a known genetic predisposition. It is pathologically and radiologically similar to osteitis fibrosa cystic. Monostic, polyostic and syndromal (McCune-Albright) types are described. Histopathology shows replacement of normal cancellous bone by a fibrous stroma arranged in a whorled pattern with "C or Y shapes" of immature, woven bone with scattered islands of cartilage. The marrow is replaced with fibrous tissue. There is active simultaneous osteoblastic and osteoclastic activity. Radiographic studies show thickening +/- sclerosis of bones, especially of the femur, tibia, ribs, jaw and skull; the paranasal sinuses may be obliterated. Variable amounts of the irregularly arranged spicules of bone lead to the "ground glass" appearance on x-ray. The typically unilateral distribution may resemble malignant growth and biopsy may be indicated for confirmation of the diagnosis. Temporal bone involvement is unusual, but it can cause SNHL, vertigo & VII palsy due to erosion. CHL due to EAC stenosis or cholesteatoma can also occur.

Major otologic findings include: unilateral CHL; swelling of mastoid or temporal regions; SNHL (erosion otic capsule); stenosis of EAC; cholesteatoma due to stenosis; pain (associated with cholesteatoma); vertigo (erosion otic capsule) or VII paralysis (erosion fallopian canal). Partial surgical removal can be performed for functionally or cosmetically deforming masses. XRT is contraindicated in the treatment of FD because a 0.4% transformation rate to osteo-, chondro-, or fibro-sarcomas (400 time the de novo risk) has been observed.
Osteitis Fibrosa Cystica (von Recklinghausen's disease)

This disease is due to hyperparathyroidism and the resultant hypercalcemia and loss of bone calcium. Elevated calcium (96% of patient) and elevated radioimmunoassay for iPTH (carboxyl end) provide the diagnosis. The cause is usually a parathyroid adenoma (85-90% cases), occasionally associated with MEN I syndrome. It may also be due to chief cell hyperplasia. (MEN I or Wermer's syndrome consists of hyperparathyroidism, pancreatic islet cell tumors and pituitary tumors.) The hypercalcemia caused by parathyroid hormone influenced release of calcium from bone causes a clinical symptom complex of "Bones, Stones, Moans, Groans, and Hypertones." This refers to the complications of hypercalcemia that are observed: skeletal deformities; muscle weakness and wasting; bone pain and fractures; anemia; renal stones (nephrocalcinosis); abdominal pain, ulcer disease and pancreatitis; and impaired mentation, depression, hyperreflexia, and rarely coma.

Histopathology shows tongues of fibrous tissue with leading osteoclasts; the resulting abnormal bone is demineralized and made up of loosely arranged trabeculae of varying sizes interspersed with marrow spaces containing fibrous tissue. Cystic degeneration of fibrous tissue may occur and lead to the formation of brown tumors. Temporal bone shows similar changes and some cases of SNHL have been observed. Radiographic studies show subperiostal erosions ("pepper-pot skull"), chondrocalcinosis and nephrocalcinosis. Neck exploration are generally curative with restoration of normal calcium balance. Surgery should be entertained especially in those patients that exhibit one of the following five criteria: Ca >11.0, x-ray evidence for metabolic bone disease, presence of a complication of hypercalcemia, active nephrolithiasis, or decreasing renal function.

Case Presentation

A 72-year-old Hispanic man presented in April 1992 with right hip and back pain. A routine lumbar spine series revealed marked degenerative changes of the lumbar spine with narrowing of the intervertebral spaces and some increased bone densities of the right ilium and lumbar spine. The findings were consistent with degenerative change of the spine or possibly Paget's disease. Further evaluation was significant for an elevated alkaline phosphatase and normal levels of calcium, ionized calcium, phosphorous, magnesium and PSA. A 24-hour urine calcium collection was significantly elevated (416 mg/24 hr). He was seen by the Endocrinology service and started on Indomethacin and Didronel with partial pain relief; salmon calcitonin was then begun with good alleviation of his symptoms. With treatment, his alkaline phosphatase levels decreased to the high normal range. He did well, and in May of 1993 a taper of his calcitonin was commenced. Unfortunately, in November 1993 he re-presented with a compression fracture of L5. He was restarted on calcitonin with good response and subsequently re-tapered off calcitonin over six months.

The patient was referred to the Otolaryngology service in September 1994 with a complaint of hearing loss of several years' duration. The loss was greater in the left ear than the right. He also noted mild
episodic dizziness over the previous two months. Routine audiometric evaluation revealed a right downsloping sensorineural hearing loss and a left mixed hearing loss with a significant conductive component. Tympanograms were normal. Laboratory evaluation showed an alkaline phosphatase again elevated (115) and he was restarted on calcitonin with relief of his pain and dizziness. The patient has been referred for amplification and is currently doing well on a tapering dose of calcitonin.

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OTOTOXICITY
Andrew T. Lyos, MD
August 20, 1992

Ototoxicity is defined as the tendency of certain therapeutic agents to cause functional impairment and cellular degeneration of the inner ear and of the eighth cranial nerve. Ototoxicity may be reversible or irreversible. Ototoxicity is differentiated from neurotoxicity where the site of action is central to the eighth cranial nerve.

The aminoglycosides are a class of broad-spectrum antibiotics capable of producing profound, irreversible sensorineural hearing loss (SNHL). Morphologic studies demonstrate that the inner row of outer hair cells (OHCs) of the basal turn of the cochlea are affected first, followed by the other two rows of OHCs and the inner hair cells. Prospective studies demonstrate an incidence ranging from 4% to 24%. Clinically, patients frequently develop symptoms following the cessation of therapy. SNHL is primarily
high frequency and may be unilateral. Risk factors for aminoglycoside ototoxicity include: therapy lasting more than seven days, elevated serum levels, prior exposure to aminoglycosides, noise exposure, high daily dose, and use in neonates.

Loop diuretics, including furosemide and ethacrynic acid, are capable of producing SNHL which is frequently reversible. The site of action is thought to be the stria vascularis, with inhibition of potassium chloride co-transport. Risk factors include renal insufficiency, use in neonates, intravenous administration, and co-administration with aminoglycosides.

Cis-platinum is a relatively new cytoreductive agent used to treat a variety of malignancies. The mechanism of ototoxic action is uncertain; however, it produces a loss of OHCs in the basal turn of the cochlea in a fashion similar to the aminoglycosides. Morphological changes have not been noted in the vestibular organs. Clinical studies demonstrate an irreversible ototoxicity incidence ranging from 9% to 91%, depending on the criteria used and the cumulative dose. Symptoms are frequently present at the onset of measurable hearing loss. Risk factors for ototoxicity include renal insufficiency, intravenous bolus, co-administration with aminoglycosides, and increased cumulative doses. An audiogram should be obtained at the onset of therapy, before each successive dose, and with the onset of symptoms.

Salycilates have been used for more than 100 years and have long been known to produce reversible hearing loss associated with tinnitus. The mechanism of action is thought to involve vasoconstriction of capillaries in the stria vascularis. Symptoms typically resolve within 24 hours following cessation of therapy; however, cases of irreversible SNHL and tinnitus have been reported.

Following a single dose in sensitive individuals or, more typically, after a prolonged course, the antimalarial agent quinine can produce the symptom complex of hearing loss, tinnitus, vertigo, nausea, and vomiting (cinchonism). Symptoms resolve following cessation of therapy. However, several cases of irreversible SNHL and tinnitus have been reported.

Monitoring for ototoxicity should be individualized to the drug and to the patient. The early symptoms seen with some drugs, such as the salycilates and quinine, obviate the need for formal monitoring. The profound, irreversible effects seen with drugs such as the aminoglycosides and cis-platinum require much more formal monitoring.

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**Case Presentation**

A 64-year-old man was initially referred to the Otolaryngology service for evaluation prior to initiation of aminoglycoside therapy. He had no prior otologic history, including surgery, noise exposure, or ototoxic drugs. Past medical history was significant for a right total knee replacement seven months prior to consultation. His postoperative course was complicated by wound dehiscence and osteomyelitis. He underwent removal of the hardware and debridement. Cultures grew *Serratia marcescens*. The Infectious
Disease service was consulted and a six-week course of gentamycin was initiated.

A baseline audiogram was obtained which demonstrated a mild sensitivity loss on the right with normal sensitivity on the left to 1 kHz, at which point he had a severe sensorineural loss. An audiogram performed eight days later demonstrated no change in sensitivity; however, the PB performance dropped from 95% to 76% on the right, and from 70% to 28% on the left. The patient denied symptoms of hearing loss, tinnitus, or vertigo at that time. However, he noticed progressive difficulty understanding speech during the next several days. On physical examination, the tympanic membranes were clear and there was no evidence of nystagmus.

The dose of gentamycin was decreased and serum levels were monitored closely. He completed five weeks of therapy prior to discontinuation due to elevation of his blood urea nitrogen and creatinine. Serial audiograms performed weekly during the course of therapy, as well as those performed during the following five years, have failed to demonstrate any changes in sensitivity or speech understanding.

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OTOTOXICITY
April 25, 1996
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Aminoglycoside ototoxicity is associated with the total daily dose, duration of treatment, previous treatment with aminoglycosides, and renal disease. The ototoxicity has a delayed onset (months to years later). Aminoglycosides enter the fluids of the inner ear via the bloodstream or via the round window with topical application. The drug is cleared more slowly from perilymph and endolymph than from serum, hence the once-daily dosing theory to reduce ototoxicity. A review of 29 clinical trials (Barclay et al, 1994) found that if the same total daily dose of drug is given with a once-daily regimen as opposed to the traditional 2-4 doses/day, the efficacy of antibiotic activity was the same, but there tended to be less ototoxicity.
Symptoms of aminoglycoside ototoxicity include irreversible high-frequency sensorineural hearing loss, decreased otoacoustic emissions, and disequilibrium. With further toxicity, this can lead to complete deafness and/or oscillopsia. Studies of the organ of corti reveal widespread loss of outer hair cells. Potential mechanisms of toxicity include:

1. Aminoglycosides binding to the cell membrane and blocking transmembrane signaling systems.
2. Inhibition of the enzyme, ornithine decarboxylase, leading to decreased polyamine synthesis (Henley, 1994).
3. An unknown drug metabolite may be responsible for the ototoxicity. (Schacht, 1993).

Briefly stated, topical antimicrobials (particularly Cortisporin) are severely ototoxic in guinea pigs (Barlow et al, 1994). However, there is no evidence for toxicity of topical agents in humans. There has never been a controlled study that demonstrates hearing loss, and there has never even been a case report of hearing loss after treatment with a topical antimicrobial.

Salicylates and NSAID's cause reversible tinnitus and hearing loss. Also decreased otoacoustic emissions have been noted. Theories of salicylate ototoxicity include:

1. Vasoostriction of vessels in stria vascularis, leading to decreased cochlear blood flow (Didier et al, 1993).
2. Reversible ultrastructural changes in outer hair cells that correlate with decreased electromotility (Dieler et al, 1991).

In summary, aminoglycosides cause permanent structural damage to hair cells, and lead to permanent hearing loss and disequilibrium. Salicylates cause reversible changes in hair cells or the cochlea, and lead to reversible symptoms.

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**Case Presentation**

A 41-year-old female presented with complaints of high-pitched ringing in her ears for one week. She stated the sound was worse in quiet environments, and was particularly noticeable when trying to fall asleep. She had no complaints of hearing loss, aural fullness, unsteadiness, or vertiginous symptoms. She had no history of ear infections or ear surgery. Family history was negative. There was no history of autoimmune disease, skin rashes, or arthritis. The patient denied use of diuretics, intravenous antibiotics, or chemotherapy medications. She did state that she had
been taking 3 aspirin tablets 3 to 4 times per day for stress headaches recently.

Physical examination, including complete neuro-otologic exam, was unremarkable. A preliminary diagnosis of salicylate ototoxicity was made, and discontinuing the use of aspirin was recommended to see if her tinnitus resolved. In addition, a neurology appointment was made for evaluation of her headaches. After one week the patient returned for follow-up and stated that her tinnitus, as well as her headache, had resolved. She had no further complaints and was discharged from clinic.

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Acute Otitis Externa is an infection of the skin of the cartilaginous portion of the ear canal. Contributing factors include moisture, canal occlusion, local trauma, and allergic disease. These factors lead to a loss of the protective wax layer, causing edema of the squamous epithelium with plugging of the glandular secretory ducts. Consequent scratching induces local trauma, allowing bacteria to invade through the skin, leading to inflammation and production of exudate.

The pathogens in acute otitis externa are *pseudomonas* (41%), *peptostreptococcus* (22%), *Staph. aureus* (15%), and *Bacteroides* (11%). The differential diagnosis includes malignant external otitis, chronic external otitis, furunculosis, otomycosis,
herpes zoster oticus, bullous external otitis, granular external otitis, chondritis, cellulitis, and eczematoid dermatitis.

Malignant external otitis is a complication of acute otitis externa. The disease starts as a local infection of the external auditory canal and spreads through the fissures of Santorini (in cartilage of ear canal) towards the parotid gland and mastoid. This can lead to facial nerve paralysis. As the disease progresses, there is spread along the skull base to the jugular foramen (paralysis of CN IX, X, and XI), and finally to the hypoglossal canal (paralysis of CN XII).

Symptoms are similar to acute otitis externa, however, the main difference is that the disease is not responsive to several weeks of conventional local therapy. On physical exam, there may be granulation tissue present in the ear canal, cranial nerve dysfunction, or a palpable bony defect in the anterior wall of the ear canal. Risk factors include diabetes, elderly, and immunocompromised status. The pathogen in malignant external otitis is nearly always *Pseudomonas* (99.2%).

Relevant laboratory studies include an erythrocyte sedimentation rate (ESR) and a fasting glucose level. A bone scan is important to demonstrate osteomyelitis. It will remain positive for years after a full recovery, so it is not useful in following response to therapy. A gallium scan will demonstrate active inflammation in either soft tissue or bone, and it will return to normal if the disease is effectively treated. A CT scan is useful to determine bony erosion and soft tissue involvement, however its sensitivity for detecting osteomyelitis is low (30%).

Therapy for malignant external otitis includes meticulous control of blood glucose, local debridement of granulation tissue, and possibly hyperbaric oxygen (Davis et al., 1992). Traditional antibiotic therapy is 6-8 weeks of IV drugs (usually an anti-pseudomonal penicillin or cephalosporin, and an aminoglycoside). Recent studies have demonstrated that oral ciprofloxacin has been 96.4% successful in treating mild to moderate malignant external otitis (Gehanno, 1994). In severe disease, IV therapy is recommended initially, and then a long course of oral ciprofloxacin is needed. Therapy is continued until the gallium scan is clear.

**Case Presentation**

A 59-year-old female was referred from the General Medicine Clinic for a 1 month history of bilateral ear pain and otorrhea. She had been treated with Cortisporin otic drops without benefit for several weeks. Her past history was unremarkable. She had no history of diabetes or immunosuppressed states. Physical exam revealed the
left ear canal to be erythematous and tender. There was scant mucoid discharge and no granulations. The right ear canal was also erythematous, swollen, and tender. There was abundant mucoid discharge, and granulation tissue was present at the cartilaginous/bony junction posteriorly. Both tympanic membranes were normal. The rest of the exam was unremarkable. Laboratory studies revealed a slightly elevated WBC of 12.7, a greatly elevated ESR of 92, a normal random blood glucose of 100, and a slightly elevated glycosylated Hgb of 7.4.

The patient was admitted to the hospital with the diagnosis of malignant external otitis. Her ears were debrided and cultures were sent, which eventually grew *Pseudomonas aeruginosa*. A CT scan done on admission showed soft tissue thickening around the ear canals, but was otherwise normal. A technetium bone scan was negative, and a gallium scan was positive for bilateral temporal inflammation. The patient was placed on ceftazidime and gentamicin otic drops. Her ear canals were regularly debrided. Twice daily blood glucose checks during hospitalization were all normal. After 10 days, her ear canals had no granulation tissue or discharge, and she was discharged to home on ciprofloxacin. In follow-up, she has been seen in at 2 weeks and 4 weeks post-discharge and has a normal exam. She is still taking ciprofloxacin.

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PATHOLOGY AND PATHOGENESIS OF OTITIS MEDIA
April 21, 1994
Philip A. Matorin, M.D.

Pathology

Understanding the pathology and pathogenesis of otitis media (OM) is important in predicting the prognosis and sequelae of the disease. In this way treatment can be tailored to individual needs.

There have been several attempts to gain a consensus definition of OM. In 1980, the Ad Hoc Committee on the Classification of Otitis Media defined OM based on the temporal sequence: Acute < 3 weeks; Subacute 4-8 weeks; Chronic >9 weeks. In 1989, the Task Force of the 4th International Symposium on Otitis Media published a working classification. Acute Suppurative OM refers to an identifiable infection of the middle ear of short duration and sudden onset. Secretory OM refers to the presence of a middle ear
effusion without acute signs or symptoms. Chronic Suppurative OM refers to a chronic middle ear discharge through a perforated tympanic membrane.

Much of our understanding of the pathology of OM comes from experimental animal models and from temporal bone preparations. After Eustachian tube obstruction in the cat and chinchilla, edema hyperemia, subepithelial hemorrhage with PMN infiltration are noted, modeling acute Suppurative OM. In addition mucoperiosteal thickening and osteitis can be seen. Mild metaplasia from squamous to cuboidal epithelium is noted.

Continued Eustachian tube obstruction models secretory OM. Early transudation of serum into the subepithelial space is seen within 18-24 hours. More metaplasia is seen with increased numbers of pseudostratified, ciliated columnar epithelial cells, and goblet cells. Resorption of water is thought to promote thickening of the effusion converting thin serous fluid into a thick mucoid effusion. In temporal bone preparations, thickening and fibrosis of the tensor tympanic is seen. This is felt to be important in the development of spasm and contracture of the muscle resulting in retraction and fixation of the tympanic membrane.

In chronic OM models, columnar metaplasia is greatest, fibroblastic density is increased. A lymphoid infiltrate is noted with polypoid changes present in the mucosa. Osteitis and osteoneogenesis are also commonly noted. Friable immature granulation tissue with neovascularization and fibroblastic proliferation is seen. Mature granulation with subepithelial fibrosis and decreased vascularity are found in long standing disease. The sequelae of COM including tympanosclerosis, cholesterol granuloma, and cholesteatoma can also be encountered.

Paparella's group has proposed that OM exists as a continuum of mucoperiosteal disease. Epidemiologic evidence comes from studies showing the overlap and progression of disease from acute suppurative OM to secretory OM or chronic OM. One out of 5 cases of Acute OM is superimposed on a chronic mucoid OM. Temporal bone histology also demonstrates the continuum of mucosal disease. Lastly, experimental animal models of Eustachian tube obstruction followed longitudinally have demonstrated the gradual change from suppurative, to secretory, to chronic histopathology.

Pathogenesis

Since the time of Politzer, the Eustachian tube has been identified as central to the pathogenesis of OM. For OM to occur, three conditions must be met. Bacteria must adhere to the nasopharyngeal mucosa. The bacteria must enter the Eustachian tube and finally, the bacteria must be able to multiply in the middle ear environment.

Pathogenic bacteria bind to cell surface receptors in a highly specific fashion. The adhesin receptor binding mechanism prevents colonizing bacteria from being swept away by the mechanical cleansing action in the nasopharynx. Adhesion through a receptor mechanism is suggested by the fact that adherence can be saturated by an increase in bacterial concentration. In general, the more adherent an
organism is to the nasopharyngeal mucosa, the more likely it will be a successful pathogen. In children, the adenoid bed is an important source of pathogenic bacteria. In addition, Influenza A can cause destruction of the normal tubal mucosa promoting bacterial entry. One study showed a 5-fold increase in the rate of pneumococcal OM in chinchillas infected with the virus.

The Eustachian tube itself is cartilaginous in its lower 2/3 portion. It is normally in the closed position. The main dialator of the tube is the tensor veli palatini innervated by the mandibular division of the trigeminal nerve. Comparing the infant to the adult tube, the Eustachian tube is shorter (13mm vs 31-38mm), shallower (10 deg. vs 45 deg from horizontal), and has abundant goblet cells. These anatomic features make the pediatric Eustachian tube much more likely to become dysfunctional.

Eustachian tube function can be divided into ventilation, drainage, and protection from nasopharyngeal secretions. Bluestone has described 4 functional abnormalities radiographically by instilling contrast material into the nasopharynx. The abnormalities include retrograde obstruction, abnormal distensibility, middle ear reflux, and retrograde obstruction with abnormal distensibility. The patulous Eustachian tube predisposes to middle ear reflux. The abnormally compliant Eustachian tube could result in middle ear reflux with slight increases in nasopharyngeal pressure. On the other hand, rapid changes in pressure could cause locking of the tube and functional obstruction.

The ventilatory function of the Eustachian tube is also affected by the gaseous equilibrium of the middle ear space. Large rapid changes can occur because the middle ear space is a relatively small pocket of gas surrounded by vascular mucosa. Gas is exchanged between the middle ear cavity and the mucosa. Recent evidence suggests that the diffusion gradient is due to the nitrogen partial pressure gradient.

Anatomically, neoplasms, cleft palate, and large adenoids can cause obstruction of the Eustachian tube. In children, it appears that adenoid size does not correlate with the incidence of OM. Adenoids act as a source of pathogenic bacteria rather than anatomic obstruction in the pathogenesis of OM.

The final stage in the development of OM is bacterial replication in the middle ear space. Therefore, the immunology of OM has generated great interest. In human studies, IgG is the predominant immunoglobulin involved. Several clinical series have shown lower levels of IgG2 in otitis prone children. These children have greater susceptibility to non-typeable H.Flu. Higher levels of interleukin-1 have been found in both younger children and those with cleft palate with Eustachian tube dysfunction and OM. Increased levels of specific hydrolase activity and higher levels of lipoxygenase products have also been found in otitis media effusions.

Case Presentation

An 11-month-old full term white male child presented with a history of otitis media for the past 5 months. He completed six courses of several different antibiotics without resolution. Physical exam
showed dull, slightly retracted tympanic membranes and no other abnormalities. Visual reinforcement audiometry showed normal sensitivity for age. Acoustic immittance measurements showed type C tympanograms. In view of the persistence of the effusion, the patient underwent bilateral myringotomy and placement of pressure equalization tubes. Thin viscous fluid was aspirated from both middle ear cavities. One month postoperative assessment revealed that the child was doing fine with well-placed, patent PE tubes.

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The diagnosis and management of perilymphatic fistulas
Troy A. Callender, MD
October 21, 1993

A perilymphatic fistula is an abnormal communication between the inner ear fluids and the middle ear. Perilymphatic fistulas can be classified into four broad categories: iatrogenic, traumatic, spontaneous, and congenital. Symptoms of perilymphatic fistula are variable and include sensorineural hearing loss, vertigo, dysequilibrium, unsteadiness, motion intolerance, tinnitus and aural fullness. Simmons in 1968 first postulated that sensorineural hearing loss could result from intracochlear membrane breaks. In 1971 Goodhill stated that the labyrinth is hydrostatically loaded with intimate relations to hydrodynamic forces in the carotid arterial system, intracranial venous-sinus systems and the CSF pressure gradients in the subarachnoid space. He proposed that the membranous labyrinth could be ruptured by an explosive route via the external auditory canal and eustachian tube and concluded that membrane ruptures could occur...
throughout the membranous labyrinth with both acoustic and/or vestibular system sequelae. Simmons later expounded on his original theory and proposed the "double membrane break theory" in the development of sensorineural hearing loss. In this theory he postulated that the second membrane break was the result of a pressure gradient created by the first break, profound sensorineural hearing loss resulted from the mixing of perilymph and endolymph, healing of intracochlear breaks halts the mixing of fluid and the widespread hearing loss disappears resulting in hearing loss at the specific site where local tissue damage occurred along the membrane.

The diagnosis of perilymphatic fistula is very difficult to make since there is no diagnostic test for this disorder. The diagnosis depends upon a high degree of clinical suspicion and ruling out other possible causes for the symptoms. In addition to the hearing loss, vestibular dysfunction, tinnitus and aural fullness, patients may also have recruitment, the Tullio phenomenon and a positive fistula test. Evaluation of these patients should include a careful history and physical examination, audiogram, electronystagmography, fistula test, imaging to rule out an intracranial tumor and exclusion of other possible causes for the symptoms (i.e., autoimmune diseases, vascular diseases, infectious diseases and endolymphatic hydrops).

The treatment of suspected perilymphatic fistulas is controversial as are most aspects of this disorder and consists of conservative medical therapy or surgical exploration and closure of the fistula. Medical therapy consists of strict bed rest, head elevation, and refraining from all strenuous activity. Some authors advocate this treatment for all cases of suspected perilymphatic fistula if the patient presents early in the course of the disease, whereas others use it to treat patients with symptoms consistent with a perilymphatic fistula but no antecedent history of trauma or surgery. After 5 to 7 days, the patients are re-evaluated and if symptoms persist, surgical exploration is undertaken. Surgical treatment consists of a middle ear exploration and careful examination of the oval and round window niches, looking for crystal clear fluid that persistently wells up after careful suctioning. Special maneuvers such as trendelenburg, internal jugular vein compression and valsalva may be required to adequately visualize a fistula. In addition to fistulas of the oval and round windows, fistulas can also develop from microfissures around the oval and round windows as well as from Hyrtl's fissure and the fissula ante fenestram. Most authors agree that whether a fistula is found or not, a tissue seal with fat, fascia, perichondrium or loose areolar tissue should be made around both the oval and round windows since there is a possibility that the fistula is only intermittently patent or is too small to be visualized. Some authors feel that surgery as an initial form of therapy is indicated in all patients with symptoms for greater than one month, whereas others would proceed only if there is a clear antecedent history of trauma or surgery associated with the typical symptoms of perilymphatic fistula. There are several large studies reporting the results of surgical treatment of perilymphatic fistulas. The percentage of patients with fistulas found surgically ranges from 24% to 93%. Vestibular symptoms had the best response to surgical treatment, with 60% to 90% of cases improving. Hearing loss responded much less favorably except in fluctuating or progressive sensorineural hearing loss in which case surgery would often stabilize or slightly improve the hearing. Postoperative management consists of bed rest, head elevation and no straining for the first 5 days, followed by 4 to 6 weeks of light, non-strenuous activity. The objective assessment of treatment results awaits the development of an objective test for diagnosing perilymphatic fistulas.
Case Presentation

A 59-year-old white man presented to the VAMC Otolaryngology Clinic 4 days after he developed sudden right hearing loss. The hearing loss developed shortly after he sneezed and was associated with a headache. He denied vestibular symptoms, otalgia, fever or a history of otologic surgery. His past medical history was unremarkable except for left hearing loss of probable sudden onset in 1976 for which he was extensively evaluated while in the Navy. Physical examination was unremarkable and significant laboratory findings included a ESR of 3 mm/hr, rheumatoid factor < 30, negative ANA, nonreactive RPR and mildly elevated liver function tests. Audiogram obtained on admission revealed a profound SNHL on the left with no word discrimination and moderate to severe mixed hearing loss on the right with 35% word discrimination. Tympanograms were type A.

The patient was admitted to the VAMC hospital for bed rest and treated with carbogen, intravenous steroids and diuretics. MRI scan of the brain, posturography, and fistula test were all negative. Because the patient's history was suggestive for a perilymphatic fistula, exploratory tympanotomy was performed on the second hospital day. At surgery, clear fluid was noted to collect around the oval window niche consistent with a perilymphatic fistula. Both windows were sealed with loose areolar tissue and the patient recovered unremarkably. A repeat audiogram obtained 2 months after surgery revealed a 10 dB improvement in the PTA and a 10% improvement in speech discrimination score on the right side.

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LESIONS OF THE PETROUS APEX  
August 4, 1994  
Philip A. Matorin, M.D.

The petrous apex is the most inaccessible portion of the temporal bone. The petrous apex can be involved in congenital, infectious, inflammatory, and neoplastic processes. Since direct examination is not possible, careful attention to the often subtle signs and symptoms, as well as keeping a high index of suspicion, is necessary to diagnose these entities. The true incidence of these rare lesions is difficult to estimate, but the House group has reported that acoustic neuromas are about 30 times more common in their practice.

The petrous apex is a truncated pyramid forming the medial portion of the temporal bone. The base is bounded by the bony labyrinth and the carotid artery anteriorly. The superior surface is the middle cranial fossa, Meckel's Cave, and the ascending carotid artery. The posterior surface is the posterior cranial fossa.
and Dorello's canal transmitting CNVI. The inferior surface is the jugular bulb and the inferior petrosal sinus. The petrous apex is marrow filled in 84%, pneumatized in 9%, and sclerotic in 7%.

Clinical presentations can range from subtle to dramatic. Involvement of the Eustachian tube can result in aural fullness or otitis media. Compression at the foramen ovale can cause V3 paresthesia or numbness. Stretching of the dura can cause headache of eye pain. In addition, ophthalmoplegia, tinnitus, vertigo, hearing loss, and facial paralysis can occur.

CT and MRI have revolutionized the preoperative work-up of petrous apex lesions. An accurate differential diagnosis is critical in that it may influence the surgical approach for specific lesions. The two modalities may provide complimentary information based on density, bone erosion, contrast enhancement, and imaging characteristics with T1 and T2 weighting.

Several benign anatomic variants may produce radiographic abnormalities that can mimic pathologic conditions. Thirty to thirty-five percent of petrous apices are pneumatized and 6.8% may have asymmetric pneumatization that can be demonstrated by CT. On MRI, the less pneumatized, marrow-filled apex would appear bright on T1 and T2, mimicking a lesion. A giant petrous air cell can cause distortion of the IAC appearing similar to an expansile lesion. Lastly, simple effusions, mucoceles, and mucus retention cysts of the petrous apex appear as soft tissue density with preservation of the air cell septae. Expansile mucoceles may have eroded margins. On MRI, these appear low on T1 and bright on T2.

Cholesterol granuloma is the most common lesion of the petrous apex. It is approximately 10 times more common than cholesteatoma and 40 times more common than mucocele. The cyst wall lacks the keratinizing squamous epithelium of cholesteatoma. On CT, the lesion appears as a homogeneous soft tissue ovoid lesion posterior to the horizontal carotid canal. On MRI, it appears bright on T1 and T2 without enhancement with gadolinium. The surgical treatment is re-establishment of aeration of the apex by using a silicone drain into the middle ear or mastoid.

Cholesteatoma of the petrous apex can be congenital or acquired. Acquired cholesteatomas are rare and occur by extension from the middle ear space along the supralabyrinthian air cell system along the anterior epitympanic space. Facial nerve dysfunction occurs in 20% - 50% of cases. On CT, cholesteatoma appears as a smooth, expansile lesion hypodense to brain without contrast enhancement. On MRI, it appears as low intensity on T1 and hyperintense on T2, similar to CSF. The translabyrinthian-transcochlear and the middle fossa approaches are used most often for extirpation of these lesions.

Metastatic lesions of the temporal bone are uncommon. However, the petrous apex and clivus are two of the most common sites for metastasis in the head and neck. The most common primary sites are the breast, lung, kidney, stomach, prostate, and larynx. Metastases to the skull base in isolation are uncommon and infraclavicular metastases are usually present. A bone scan should be performed when metastasis is suspected. Most tumors are erosive, but some stimulate sclerosis from osteoblastic activity. The MRI appearance is variable depending on the type of lesion.
Other lesions of the petrous apex that should be in the differential include: petrous apicitis usually due to pseudomonas and rarely TB; intrapetrous carotid artery aneurysm, chondroma/chondrosarcoma, meningioma, schwannoma, and chordoma.

## Case Presentation

A 64-year-old white lady presented with a nine-month history of asymmetric left-sided hearing loss of sudden onset. The hearing loss was accompanied by tinnitus. She denied headache, vertigo, and otalgia. She denied any significant medical or surgical history and had no history of otologic disease, nor previous otologic surgery. She did have a history of left Bell's Palsy one year before presentation, which had resolved completely. She admitted having a 75 pack/year smoking history. Audiometry revealed an asymmetric, moderate to profound, left sensorineural hearing loss, absent acoustic reflexes and significantly depressed speech audiometry. CT scanning of the temporal bones showed a well defined, lobulated, expansile lesion of the left petrous apex. By MRI, the lesion was bright on T1 and T2. CT scanning of the chest and a screening mammogram were negative. The petrous apex was entered after complete mastoidectomy via the retrofacial air cell tract. A cyst filled with a brown exudate consistent with a cholesterol granuloma was drained into the mastoid using a pediatric feeding tube. Two months post operatively, she was asymptomatic, doing well, and had minimal change in her hearing.

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Causes of Failure and Complication in Surgery for Otosclerosis

Mas Takashima, M.D.

November 11, 1999

The restoration of hearing in patients with conductive hearing loss secondary to otosclerosis has, as some may say, been the most important development in otology. A number of techniques for opening the oval window and reconstructing
the ossicular chain have been developed since Rosen first mobilized the stapes in 1953. Excellent hearing improvement is possible with most of the techniques in use today. However, a significant number of complications or failures continue to compromise the expected hearing results and necessitate revision surgery. The best chance of success is at the time of the initial procedure, with revision surgery successful in less than 80% of cases. It is the purpose of this grand rounds to review the most common pitfalls incurred in stapedectomy, as well as to report surgical complications of this procedure.

The pertinent anatomy includes the tympanic membrane, the anterior and posterior crus of the stapes, the capitulum, the long process of the incus, the tendon and stapedius muscle, the footplate of the stapes, the tympanic segment of the facial nerve, the cochlea, and the labyrinth.

The earliest surgical attempts to improve hearing loss associated with stapes fixation was by Kessler in 1876. In this procedure, he incised the posterior portion of the tympanic membrane, separated the incus stapedial joint, removed the bony canal wall when necessary and attempted to mobilize the stapes by applying pressure to the capitulum in various directions. If the stapes could not be mobilized, it was removed. The interest in stapes surgery abruptly stopped in 1900 when at the International Congress, Politzer and Sutton and other prominent otologists declared these operations were useless and dangerous. It is surmised that this statement was prompted by large numbers of complications that had gone unreported.

Next, the area of surgical fistulization began with the work of Barany and Holmgren. They fistulized the superior semicircular canal in otosclerotic ears, allowing the dura to rest against the membranous labyrinth. Hearing improvement was fruitfully noted with this procedure. Sordell studied with Holmgren and devised a three-stage operation in which the skin-covered horizontal canal fistula was used to improve hearing. In 1938, Lempert simplified this to a one-stage operation. After being modified in 1945, this operation became the standard for the next seven years. The next major advancement occurred in 1952 when Rosen accidentally mobilized the stapes during palpation on the ossicular chain in a fenestration operation. The patient noticed remarkable, immediate hearing improvement, which lasted for years. One year later, when mobilization had become the standard of care, Rosen reported hearing improvement to at least the 30 decibel level in 22% of 211 operations he performed. In 1954, Shambaugh applied the operating microscope to stapes mobilization. This paved the way for Shea to revive the stapedectomy with the modifications of an intact ossicular chain in a covered oval window. To date, multiple modifications of this procedure have been performed using various types of prostheses and grafts.

Otosclerosis is the primary focal spongiform disease of the labyrinthine capsule.
Valsalva first described this pathologic process underlying otosclerosis in 1735, from the autopsy of a deaf patient. Today, there is still some controversy regarding the etiology and pathogenesis of otosclerosis. Briefly, otosclerosis, otherwise known as otospongiosis, is thought to be a familial autosomal dominant disorder with a variable penetrance of 25% to 40%. The mechanism has recently been suggested to be an autoimmune reaction to cartilaginous remnants and collagen fiber type II. This is thought to stimulate an inflammatory reaction in lysosomal proteases in and around the otospongiotic foci.

For a successful surgical outcome, one must begin planning treatment with the initial physical examination. This begins with an overall observation of the patient. The age of the patient alone should not be a contraindication to performing surgery. In patients less than 16 years of age, there is a greater chance that the conductive impairment is secondary to congenital anomalies rather than otosclerosis. Likewise, very active diffuse obliteratorive otosclerosis may be found in a young patient, predisposing to a higher incidence of complications. Yet surgery should be considered in the young because hearing acuity is essential during the formative years of development. Patients older than 75 years are still surgical candidates if they are in good health. Hearing acuity becomes more important as other senses decline with age. In order to prepare for surgery and to prevent failures and complications, infections and anatomic variations must also be considered. The otologic evaluation may reveal dermatological conditions of the external ear canal that will predispose to postoperative infections. These include chronic external otitis, trauma from hearing aides or self-inflicted injury with cotton-tipped swabs. Likewise, one may see a small, crooked stenotic ear canal that, until corrected, will restrict surgical movement. Small exostosis usually does not impair access to the mesotympanum and may be left untouched. Even larger exostosis, which is limited to the anterior canal wall, usually does not interfere with the surgical approach. When the removal of the large posterior exostosis is necessary, it may be impossible to preserve a satisfactory tympanomeatal skin flap. In such cases, it is best to remove the exostosis first and delay the stapedectomy.

Brahe Pedersen and Felding have postulated a connection between the influenza viral infection and unexplained sensorineural hearing loss immediately after stapes surgery. Although without evidence, this cannot be proven, it would seem prudent to avoid operation when respiratory viral infections are prevalent, especially in the patient, the patient's family or the surgeon.

The diagnosis of otosclerosis should be confirmed only after other reasons for progressive hearing loss have been ruled out. The most common differential diagnoses of conductive losses are malleus fixation, serous otitis media, ankylosis of the malleus incus joint, Waardenburg's syndrome, Paget's disease, tympanosclerosis and progressive lysis to the long process of the incus.
Tympanometry provides the most useful method to exclude most of these cases, particularly serous otitis media and ossicular fixation. Clinical history is of special value to rule out systemic disease such as Paget's disease, osteogenesis imperfecta or ankylosing rheumatoid arthritis.

Many of the common anatomic variations, congenital anomalies and pathologic conditions are not known until seen during surgery. The surgeon must be well trained and experienced, first in recognizing these malformations, and second, in skillfully correcting them. A systematic examination of the unusual should be done. A superiorly located jugular bulb may come into juxtaposition with a tympanic annulus and in this position is vulnerable to injury during elevation of the tympanic meatal flap. For this reason, the elevation of the tympanic annulus inferiorly should not be performed with both strobos of the elevator. Tears of the jugular bulb, of course, result in profuse bleeding and constitute an alarming, although not serious, complication. Elevating the head of the operating room table and packing the area with Gelfoam may control the bleeding. If the bleeding is readily controlled, the operation may be completed. If the tear is large and the bleeding is difficult to control, the procedure should be terminated.

The location of the facial nerve should be determined before proceeding with removal of the stapes. Usually, the facial nerve lies superior to the oval window, which permits the surgeon to acquire a complete view of the footplate. In this position, the lateral surface of the nerve is usually covered by bone, although the inferior surface may be partially dehiscent. When the nerve overhangs the footplate, there is frequently a lateral as well as an inferior dehiscence to the fallopian aqueduct. Dehiscence of the facial nerve is not uncommon, and in about 0.5% of middle ears there is a sizable dehiscence, so that the nerve bulges down and obscures the arch and footplate. Even in these cases, for the experienced surgeon, it is often possible to remove the footplate, although somewhat blindly, and the prosthesis can be contoured so as to adapt to the overhanging facial nerve.

Otosclerosis of the round window niche is of no significance unless the round window opened is completely closed. When obliteration of the niche is severe, only a dimple may mark its location. From a retrospective study of 30,567 stapedectomies, this has been shown to occur in 300 patients or 1% of cases. In this situation, the mucosa should be elevated in an attempt to determine that the closure is complete. When it is not possible to determine with certainly whether the window is blocked, the stapedectomy should be performed. Patients with round window closure tend to have an early onset of hearing loss, frequently beginning in childhood or in adolescence, similar to the hearing losses associated with obliterative otosclerosis. However, these patients develop a more severe mixed hearing loss than those patients with disease limited to the oval window. A family history can be obtained from 50% of the patients with otosclerosis. Shea's series
showed 78% of patients who developed round window closure had a strong family history consistent with otosclerosis.

The stapedial artery, which is present in the early stage of embryonic life and persists in many lower animals, is usually absent in the human ear. A pulsating vessel nearly filling the crural arch will identify a persistent stapedial artery. In some cases, the stapes can be removed and the prosthesis introduced, but in 0.2% of the operations it is not feasible to proceed with the surgery. Rupture of the vessel could result in profuse hemorrhage and is to be meticulously avoided. A small artery on the footplate is a consistent finding and is not to be confused with the persistent stapedial artery.

Until the advent of stapedectomy surgery, the high incidence of malleus ankylosis had not been recognized. Malleus ankylosis occurs with a disease entity unrelated to, but sometimes associated with, otosclerosis of the oval window. A bridge of lamellar bone extending from the anterior part of the head of the malleus to the anterior epitympanic wall fixes the head of the malleus. Presumably, this bridge of bone results from failure of the malleus to completely separate from the epitympanic wall during embryonic development. The otologic history, often secured in retrospect, usually reveals hearing loss in childhood that becomes worse in adult life. Such cases represent malleus ankylosis with subsequent development of otosclerosis. When there is a history of hearing loss since early childhood without progression, the case may represent pure malleus fixation without oval window otosclerosis. If there is a history of hearing loss dating from childhood or if the hearing loss is unilateral, the preoperative evaluation could certainly include tests for malleus mobility. Should examination with the pneumatic otoscope be inconclusive, mobility should be determined with direct instrumental manipulation with the aid of magnification. If there is fixation, it is necessary to perform an atticotomy and widely expose the head of the malleus and the body of the incus. Proper treatment will consist of amputation of the head of the malleus followed by ossicular reconstruction.

Congenital anomalies of the stapes and incus are usually suspected because of a history of hearing problems from early childhood. The anomalies may range from simple fixation of the footplate due to incomplete resolution of the mesenchymal tissue during embryologic development, to the absence of the stapes in infancy. The stapes may be small and articulate with a short, underdeveloped long process of the incus. The crural arch may contain bone or may be replaced by a single bony strut. The floor of the oval window niche may consist of a solid layer of bone without evidence of a footplate or the oval window niche may be completely missing. The correction of simple congenital stapes fixation may be the same as for fixation due to otosclerosis. Corrective surgery for anomalies is attended with a higher incidence of injury to the inner ear because of frequent associations of
anomalies to the membranous labyrinth. For example, the utriculosaccular deck may be located in the oval window area and may be vulnerable to injury. These patients should be counseled regarding the greater risk of sensorineural hearing loss. In very rare cases, the otosclerotic foci may not only obliterate the oval window niche, but may also extend into the vestibule. This has been shown to occur in 7%-11% of the cases in which stapedectomies failed. When removing the bone, the surgeon will find that the growth extends into the vestibule beyond the footplate level. This is best managed using a slow rotation per minute microdrill to create a 0.8-mm diameter cylinder. In this situation, laser creates a potential risk to the patient because of heat transfer to the perilymph of the vestibule and subsequently to the utriculosaccular.

The gusher is a dramatic complication to stapes surgery. The smallest control hole in the footplate can produce a perilymph leak so profuse that it fills the middle ear and internal auditory canal within seconds. The existence of this abnormality is suggested by an upward sloping air conduction audiogram with best responses at 2 kilohertz. This complication is more common in ears with congenital fixation of the footplate. Management of the stapes gusher is the same as management for a CSF leak. Once the surgeon notes the onset of fluid leaking from the aperture in the stapes footplate, leakage may be controlled with a small piece of Surgicel. A graft should be placed over the leaking footplate with the completion of the stapedectomy. A lumbar drain will now effect a diversion of the CSF thereby decreasing the pressure of the purulence. The head of the patient's bed is elevated and daily fluid intake is restricted. The flow of CSF may last for several days and a sensorineural hearing loss is likely to occur. Clearly, if such ears can be identified, the operation is contraindicated.

Next, we will move on to surgical trauma. Tears of the tympanic meatal flap that may occur in the following locations. First, a linear tear or buttonhole perforation may occur in the skin flap. Usually, these tears require no repair. However, when replacing a flap at the completion of the procedure, care must be taken to avoid enfolding the margins of the tear. The skin flap may separate from the tympanic annulus as well. This usually occurs in an inferior location and is due to failure to elevate the tympanic annulus from the sulcus. Another cause of a tear of this type is failure to achieve total elevation of skin flap from incision-to-incision. Usually no repair is required if separation occurs inferiorly. However, if it occurs superiorly, grafting may be necessary.

Subluxation and luxation of the incus consists of a tear in the capsule of the incudomalleolar joint, but with a sufficiently intact capsule to maintain the incus in its normal anatomical position. Although the long process of the incus will be excessively mobile, the operation may be completed and functional results may be satisfactory. Luxation of the incus is due to a complete disruption of the
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Incudomalleolar joint and demands that the removal of the incus and the use of a malleus over-window prosthesis. Attempts to replace and maintain the incus in its original position are usually not successful. The incus may be accidentally dislocating during curetting of the bony annulus and during manipulations of the oval window. The long process of the incus may be accidentally displaced when withdrawing the instrument from the oval window niche. Fractures of the long process of the incus are an unusual complication, but may happen during the wire tightening procedure. If the fracture appears near the tip of the long process, the wire may still be placed on the stump. If this is not possible, the procedure must be altered.

Injury to the facial nerve during stapedectomy is uncommon, but there may be a direct injury to the nerve in the facial canal by careless use of the instruments, touching with the drill or heating by a drill if the tip is not cool. An interesting remark made by Schuknecht states that direct instrumental injury to the facial nerve is extremely rare and has only occurred once during his numerous stapedectomies. Since the risk to the facial nerve injury is almost nil, Schuknecht considered that the possibility of its occurrence need not be mentioned to the patient before operation.

During a stapedectomy operation, it is frequently necessary to displace the chorda tympani nerve to gain adequate exposure. There is controversy concerning the advisability of stretching the nerve or cutting it. Approximately one-fourth of patients complain of ageusia in the postoperative period. Chorda tympani nerve dysfunction is usually transient and fewer than 5% of patients experience permanent deficits. It should be appreciated that if the nerve is cut, it will produce permanent loss of sensation of taste on the anterior two-thirds of the tongue on the same side. No reinnervation of the taste buds can take place, either from the chorda tympani nerve on the opposite side or the posterior third of the tongue supplied by the glossopharyngeal nerve. Permanent loss of the chorda tympani innervation results in atrophy of the taste receptors. The dorsum of the tongue becomes smooth and pale. The patient may not complain of the loss of taste if one nerve is cut, as long as the tongue retains sensation for 66% of the surface. If the nerve is stretched during operation, a persistent abnormal sensation in the tongue may occur, described by the patient as salty or metallic. Bilateral loss of the chorda tympani nerve produces marked symptoms in the majority of patients. In addition to the loss of taste, there is loss of secretum motor supply to the submandibular and sublingual glands that produces an uncomfortably dry mouth. Deficits include a diminution of taste sensation or a dry mouth. Revision causes cases to deserve special attention.

Sensorineural hearing loss is perhaps the most disappointing and devastating complication for both the surgeon and patient. Complete sensorineural hearing loss can occur even in the most meticulously and appropriately performed stapes
procedure. Most commonly, however, are complications due to surgical trauma. Intraoperative studies performed in stapedectomy indicate that acoustic trauma in drilling, excessive movement of the stapes producing a hydraulic effect, rupture of the membranous inner ear, rapid loss in perilymph that may lead to damage at the membranous structures in the vestibule, footplate fragments or bone death in the vestibule, and the floating footplate are the great offenders for vestibular and presumably cochlear damage. The reported incidence of dead ears after operation varies from 0.5% to 4%. It is important to appreciate that these are figures from a series of operations performed by expert surgeons with a special interest in stapedectomy. Results achieved by the occasional operator are not reported and it is likely that the incidence of cochlear damage is much higher. Mawson has shown that in a series of 1000 operations, there was a hearing loss at 4% in the first 50 operations, 2% hearing loss after the next 50 operations, no loss in the next 500 and 0.25% of the remaining 400 operations.

An attempt at removal of the floating footplate may result in a dead ear and if this complication is encountered it must be dealt with correctly. If the footplate is visible, it may be possible to remove it by manipulation and extraction with a fine hook, or a small drill hole may be made at the margin of the oval window and a fine hook used to remove it. If the footplate cannot be removed without excessive manipulation, it should be left in place and a soft tissue graft placed over the oval window and the operation abandoned.

The depressed footplate may result from incorrect attempts to remove a floating footplate or may occur during attempts to carry out a total stapedectomy. Causse documented this at an incidence of 0.16%. No attempt should be made to retrieve the submerged footplate by instrument as this may cause severe cochlear damage. Roche et al have described a method of dealing with this complication that is not damaging to the contents of the vestibule. Drops of blood are poured laterally into the vestibule and when the vestibule is full of blood and a clot is formed, the clot is removed by a lateral application of the sucker and the footplate coated with the clot is usually removed from the oval window. It is of paramount importance for the surgeon to understand the relationship of the stapes footplate to the vestibular contents. Almost all patients have a minimal safe distance of approximately 1-mm between the medial surface of the footplate and the utricle and saccule. Penetration of the vestibule by more than 1 mm with the footplate instruments or prosthesis may perforate the utricle and saccule inducing sensorineural hearing loss, vertigo or both. In patients with endolymphatic hydrops the saccule may extend to contact the medial surface of the footplate, thereby placing these patients at a higher risk for complications from this procedure.

Concerning postoperative complications, prosthesis displacement is the most common postoperative complication, accounting for 50-70% of revision surgeries. The hearing deterioration may be acute, as the prosthesis becomes dislodged or
chronic as it gradually displaces out of position. This complication may be encountered many years after stapes surgery. The diagnosis is definitely made at reoperation, but may be suspected based on tuning fork and audiometric tests.

Prosthesis loosening due to erosion or notching of the incus and prosthesis displacement due to necrosis of the incus also occurs. With removal of the stapes arch, the blood supply to the long process of the incus is divided as it courses over the incudostapedial joint. Collateral vessels exist from the body of the incus and maintain the viability of the remaining bone. Circumferential overtightening of the prosthesis can remove the blood supply to the tip of the incus.

The signs and symptoms of perilymphatic fistula were first described by Lewis in 1961, and this complication, which at one time was thought to be unusual, is now accepted as being one of the more common complications of stapedectomy. The surgeon creates a fistula at every stapedectomy operation and relies on the natural process of healing or in some techniques a graft of soft tissue, to seal the opening that has been made. In most operations, there is enough surgical trauma to the oval window mucoperiosteum to lead to the production of an inflammatory repair envelope around the prosthesis, sealing the opening into the oval window. With symptoms of a perilymphatic fistula or fluctuating hearing loss, tinnitus, a feeling of fullness in the ear and vertigo, there is no doubt that a small fistula remained after many stapedectomy operations with incomplete closure of air bone gap, but the hearing result may be acceptable to the patient. Although a perilymph fistula usually leads to sensorineural hearing loss, this is not always the case and a persistent conductive loss after an operation should warn the surgeon that there might be a fistula. A revision operation to close it would be the best line of treatment. The fistula may be primary, dating from the time of operation when there is a failure of the oval window to seal, or it may be secondary, where it can appear many months or even years after the original operation. Hearing tests carried out soon after the onset of fistula will show findings similar to those seen in labyrinthine hydrops. For example, puretone sensorineural hearing loss and low frequency initially, followed by a flat loss that fluctuates. There may be recruitment as well. Vestibular tests such as the Hallpike caloric test, EMG, and fistula test with a pneumatic otoscope may all be helpful, but have been found to be negative in 20% of the cases. Welder described a newer technique, diagnosis by radioactive Indian-111 that is injected into the lumbar subarachnoid space. The demonstration of an increased radioactivity in the nasopharyngeal secretions strongly supports the diagnosis of a fistula.

The treatment of a perilymphatic fistula is a tympanotomy at the earliest possible moment with an attempt to close the fistula. When the leak is detected, the fistulous track is excised and the prosthesis removed with great care. The opening of the vestibule is covered with a soft tissue graft that is held in place by another
prosthesis. Unless early treatment is instituted, the chances of restoration or improvement of hearing are small. In some cases troublesome dysequilibrium may remain. It is imperative that the surgeon is fully aware of these complications and realizes that some techniques are safer than others. The use of Gelfoam to seal the oval window in stapedectomy produces a very thin membrane and has caused the highest incidence of fistula formation. The perilymphatic fistula is the only complication of stapedectomy that is dangerous, and although the risk of meningitis is small, death from meningitis after this operation has been reported.

As with the perilymphatic fistula, postoperative granulomas can also be a cause of dysequilibrium, vertigo or progressive sensorineural hearing loss. Granuloma formation is seen in approximately 1:100 cases of revision stapedectomy. Foreign body reaction is a suspected etiology of postoperative granuloma formation. Glove, starch and Teflon have all been implicated. For this reason, direct pullout prosthesis contact should be avoided. It may also be prudent to rinse the prosthesis prior to its insertion. The condition usually manifests between the 5th and 15th postoperative day and is characterized symptomatically by hearing loss after an initial hearing gain or a sensation of unsteadiness. Associated with the hearing loss are loss of speech discrimination and a sensation of fullness in the ear. Examination reveals an edematous, thickened and hypervascular skin flap as well as dullness and reddening at the posterior part of the tympanic membrane. Audiometric studies show a combined sensorineural and conductive hearing loss that is worse in the higher frequency that is also associated with decreased speech discrimination. High-dose steroids may decrease the inflammatory response and its effects on the inner ear. Emergency surgical intervention is imperative. Surgical exploration reveals a granulomatous mass extending from the oval window niche to completely envelop the prosthesis and the long process of the incus. In about half of the cases the granuloma extends into the vestibule. The granuloma, including the portion within the vestibule, must be removed in its entirety along with the prosthesis. They may also be vaporized with laser after the prosthesis has been removed.

Cholesteatoma following stapedectomy is a rare complication. Mawson reported only one post-stapedectomy cholesteatoma in a review of nearly 2000 cases. Only five other cases have been previously reported in the literature. Proposed mechanisms for formation include prosthesis extrusion, the presence of a squamous epithelium in the fascial graft, inversion of the tympanomeatal flap and a marginal perforation associated with a disruptive annulus.

Diplacusis occurs in approximately one-third of the patients. There may be a variation of distortion of sound, especially music and the human voice, of which patients frequently complain. For the first few weeks after the operation most people complain of pure tones appearing higher in pitch when compared with the unoperated side. It is seldom problematic and usually fades by six weeks
postoperatively. Almost all patients have some degree of phonophobia postoperatively as well. Reassurance is adequate treatment. A subset of this has been documented as an abnormal, emotional response. In a 1993 article in Laryngoscope, Lloyd Stores describes such a case:

"This is the most curious and unexpected phenomena that I have ever encountered. They brought this girl in that I had performed the stapedectomy a few days prior. Honestly, her mouth was open and fixed and she had a glassy stare. From the history, it turns out that her symptoms developed almost immediately after I took the first packing out of her ear canal. The mother-in-law was a talker and she continued to talk about how useless her daughter-in-law was. The daughter-in-law, my patient, had not been hearing all of this for years and she went into a catatonic trance that was unbelievable. She really freaked."

In conclusion, we have come a long way from the original attempts at mobilization of the stapes by Kessel, as resurrected by Rosen, which led to the modern day stapedectomy performed by Shea. A survey of articles in the last 30 years concerning the complications of stapedectomy indicates that although there are some differences over the relative importance attributed to a certain problem, there is general agreement about the direction in which new research should be directed. The situation is well summarized by Dr. Shea in his 40-year report on over 14,449 stapedectomies, where he makes the observation that although the original objections to stapedectomy have all but disappeared, it is now increasingly necessarily to concentrate on the problems arising from the operation itself.

**Case Report**

I.K. is a 36-year-old Indian male who presents to the BTGH clinic with a several year history of progressively worsening hearing loss in his left ear. The patient has previously undergone an unknown middle ear procedure on the right ear, performed in another state for hearing loss. Of note is that the patient's mother and father also have had some degree of hearing loss. An audiogram obtained on March 23, 1999 revealed a 40-50 decibel conductive hearing loss in the left ear with a pure tone average of 67 decibels. On physical exam, the patient had an unremarkable head and neck examination except for his conductive hearing loss. The patient was taken to the operating room on April 13, 1999. Upon elevation of the patient's tympanomeatal flap, there was a narrowed oval window niche. The facial nerve was in the fallopian canal without dehiscence. The stapes was fixed with diffuse otosclerosis of the footplate. A 0.8 mm diamond drill was then used to thin the thickened footplate. The footplate was then perforated and a 4.0 mm Fisch stapes piston was placed.
Postoperatively, the patient did well, and during the first 15 days, he stated that his hearing was markedly improved. On post operative day 16, the patient experienced a sudden decrease of hearing in that ear. There were no associated symptoms of vertigo, otorrhea or ear pain. A repeat audiogram was performed which revealed similar results as the preoperative test. The patient was thus taken to the operating room again on June 17, 1999, for a middle ear exploration, where it was noticed that the oval window was once again obliterated. It was demonstrated that the prosthesis was no longer sitting in the oval window, but was surrounded and riding on top of a thickened and obliterated footplate. A re-drill out of the oval window was performed, and the vestibule was entered carefully with a sharp pick. A longer 4.5mm Fisch prosthesis was then used. Postoperatively, the patient was started on sodium fluoride. Unfortunately, the patient never returned for a postoperative visit.

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The history of stapedectomy is one of near discovery, condemnation, reintroduction and refinement. Prior to the twentieth century poor lighting and magnification, nonstandardized audiometry, and a lack of understanding of the middle ear conduction mechanism plagued early researchers. Procedures concentrated on either partial or total removal of the tympanic membrane and ossicles. Mobilization of the stapes, described by Miot and later by Blake, would eventually be condemned by the otologic world. Siebenmann, in 1900, capped the early era of stapes surgery by stating, "Clinical experience teaches that all endeavors at mobilization of the stapes in otosclerosis are not only useless but often harmful."

Between 1900 and 1952 attempts at fenestration of the horizontal semicircular canal came to the spotlight. Results were reported as excellent but temporary. Microscopy and rotating drills were
introduced. In 1929 the American Otologic Society commissioned a committee to compile a resume of all literature concerning otosclerosis up to 1928. The result included a 700 page, two volume document covering histology of the otic capsule, heredity, audiology, and treatment of otosclerosis.

Successful surgery for hearing restoration truly began with Lempert's fenestration operation. Rosen, a student of Lempert, serendipitously discovered that footplate mobilization improved hearing immediately in a patient with otosclerosis. He continued to refine his mobilization procedure until Shea, his student, reopened the era of stapedectomy in 1956. Shea developed the modern technique of footplate mobilization, soft tissue grafting of the oval window, and ossicular replacement.

Shea is also credited with the first stapedotomy, performed on a young girl in 1960. A 6 millimeter stapedotomy with piston insertion and soft tissue packing provides equally improved hearing with less risk for damage to inner ear structures. Higher frequency hearing is preserved better with stapedotomy. Robinson summed up the stapedectomy/stapedotomy debate best by declaring the "best" procedure is the operation which provides the individual surgeon with a predictable and safe outcome along with improved hearing to levels consistent with current otologic standards.

Depletion of the pool of patients with otosclerosis occurred during the "golden age" of stapes surgery throughout the sixty's and early seventies. Increased numbers of trained otolaryngologists and improved hearing amplification further contribute to an overall limited exposure in modern practice and residency. Stapedectomy is the second least performed procedure in American residency, as shown by Harris and Osborne in 1990.

The issue of training residents to do successful stapedectomy first appeared in the literature in 1983. Chandler and Rodriguez-Torro revealed a 62% closure of the air-bone gap to within 10 dB in resident cases, well below the 90-95% standard expected of practicing otologists. Five studies of results from training programs have since revealed results ranging from 64%-82% closure to within 10 Db in training programs. Of interest are the results of Shapira, et al. Their residents, in Israel, did stapedectomy in all four years of training. Results during the first two years of residency compared with American programs. During the second two years, however, results approached and often equaled those of staff. These findings further emphasized the critical importance of adequate numbers of procedures to insure acquisition of competence.

Solutions to the problem of numbers of cases include increased observation of otologic procedures with correlation in the temporal bone laboratory. Residents should display competence in surgery for chronic middle ear disease prior to attempting stapedectomy. Finally, all procedures should be supervised by senior otologic staff and one consistent technique used. For physicians in practice, batching stapedectomies into groups with review of needed skills in the temporal bone lab followed by consecutive case completion and review of results could provide valuable feedback in regard to outcome and maintenance of required skills.

In summary, stapedectomy has a fascinating history. Had Rosen not been open-minded to his first
accidental mobilization, more delay in technique development was inevitable. The best procedure is surgeon dependent varying with individual skills and experience. Otosclerosis is now at a steady incidence making increased work in the temporal bone laboratory mandatory for both the acquisition and maintenance of required skills in footplate manipulation.

**Case Presentation**

A 68-year-old white male presented with a fifteen year history of progressive hearing loss. He had the most difficulty hearing in small group conversations. He denied tinnitus, dizziness, otalgia, or chronic middle ear infections. He did admit to a long history of loud noise exposure. Past surgical and medical histories were unremarkable. Weber exam showed localization to the right. Audiometry revealed normal to moderate sensorineural and severe to moderate conductive loss in the right ear with absence of acoustic reflexes. The left ear had mild to moderate sensorineural loss with normal acoustic immittance measures. Speech intelligibility scores were within the normal range bilaterally. The patient underwent right middle ear exploration, stapedectomy, with perichondrial grafting over the oval window and insertion of a standard well Robinson prosthesis.

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Sudden sensorineural hearing loss is a confusing and controversial issue in otolaryngology that most of us will encounter in our practice. The impact of the loss of hearing on the patient can be catastrophic and most physicians consider it a medical emergency. On the other hand, it usually becomes frustrating for the physician, as the cause is most often not obvious and the treatment empiric and poorly understood.

There is no accepted definition of the problem, no standard method for evaluating these patients, no agreement on what the usual etiology is, no easy way to stage the loss and no clearly effective treatment. Some authors feel strongly that sudden sensorineural hearing loss is just a symptom and not a disease entity. The most useful definition of sudden sensorineural hearing loss is an idiopathic loss greater than 30 Db in three contiguous frequencies and that occurs in less than 3 days. Most sudden losses occur...
within minutes to several hours, and about 1/3 of these patients awaken in the morning with a hearing loss. Approximately 1/2 note imbalance or vertigo and the intensity of vertigo correlates in general with the degree of hearing loss. Byl reported an incidence of between 5-20 cases per 100,000 population per year.

Doug Mattox and Blair Simmons at Stanford reported a 5 year prospective study involving 166 patients, looking at the natural history of the process. They found that males and females were equally affected with a mean age of 46 years. There was no seasonal or geographic clustering. They noted that 65% of patients showed some degree of recovery without any medical or surgical intervention at all. The likelihood of recovery was influenced by the shape of the audiogram - all but one patient with an upward sloping audiogram had complete or good recovery and all but two patients with a severely downsloping audiogram had a fair or poor recovery. Other poor prognostic indicators that are generally agreed upon in the literature include sensitivity loss greater than 90 Db in the mid-frequencies, advanced age, and the presence of vertigo. In his triologic thesis, Dr. Frederick Byl compiled data from 225 patients over an eight year period forming a prognostic table. He also noted the direct relationship between the severity of the initial hearing loss and potential recovery and recorded several modifying factors. The study confirmed that the sooner a patient is seen, the better the prognosis for recovery.

Unfortunately, there is no better understanding of the etiology of sudden sensorineural hearing loss than there was 1 or 2 decades ago. It is certain that there is no single etiology - the loss of hearing is just the final outcome for many potential insults to the inner ear. The four most accepted hypothetical etiologies are viral infections, vascular insult, labyrinthine membrane rupture, and autoimmune disease.

Although viral infection of the inner ear is an attractive theoretical explanation for sudden hearing loss, the supporting evidence is not conclusive and consists of 4 types:

1) coincident symptoms suggesting coincident viral infection;

2) serological data showing coincident viral infection;

3) histopathological studies showing similarities between known viral infections and cases of sudden hearing loss;

4) extrapolations from the degeneration of the inner ear seen in viral disease such as the prenatal rubella syndrome.

Uncontrolled studies from Van Dishoeck, Jaffe, and Schuknecht reported "flu-like" symptoms in 20 - 60% of patients suffering idiopathic sudden sensorineural hearing loss. However, a study by Rowson and Hinchcliffe in 1975 found the same symptoms of upper respiratory tract infection to be present in approximately 40% of the general population. Veltri et al looked at 77 unmatched patients with sudden sensorineural hearing loss and found a conversion rate of 65%. Multiple viruses were implicated including influenza A and B, rubeola, rubella, mumps, herpes simplex, and CMV. Wilson and coworkers
studied 122 patients, 63% of whom seroconverted compared with 40% of controls. Other studies from Morrison and Booth as well as Rowson and Hinchcliffe failed to demonstrate a viral titer rise in sudden hearing loss patients. The most consistent evidence supporting viral etiology is the histopathologic findings of varying degrees of atrophy of the organ of Corti, stria vascularis, and tectorial membrane with variable loss of the neuronal population as shown by Schuknecht and others. Perhaps the strongest evidence for the involvement of viruses in hearing loss comes from the use of immunofluorescent antigen studies such as those performed by Davis and Johnson, demonstrating the ability of rubeola and mumps to infect the inner ears of animal models.

A vascular cause for sudden hearing loss is attractive because it is logically consistent with immediate onset of symptoms and because there are established models of acute hearing loss secondary to vascular occlusion in hypercoagulable states such as leukemia and sickle cell disease. However, most of the experimental and clinical evidence casts serious doubt on a vascular etiology. One would expect that older patients with known peripheral vascular disease would be the ones most often afflicted, but in fact, most patients are younger and have no stigmata of systemic vascular disease. Perlman and other investigators have demonstrated permanent loss of the cochlear microphonic and action potential occurs after only 30 minutes of ischemia. The histopathology of ears affected by sudden hearing loss also differs significantly from temporal bones examined after experimental vascular occlusion. Permanent obstruction leads to marked degeneration of the neurons followed by fibrous and osseous proliferation within the inner ear, changes that are not seen typically in sudden sensorineural hearing loss.

Intracochlear membrane breaks were proposed as a cause of sudden hearing loss by Simmons in 1968, but the evidence is only coincidental. Schuknecht and Donovan studied 12 temporal bones of patients afflicted with sudden sensorineural hearing loss, and found no evidence of Reissner's or basilar membrane rupture.

There are several known autoimmune diseases that are epidemiologically associated with sudden sensorineural hearing loss, including Cogan's syndrome, systemic lupus erythematosus, and temporal arteritis. Some of the most convincing evidence has come out of the University of Tennessee by Yoo and coworkers who showed that monoclonal antibodies specific for type II collagen can activate an immunologic response in the inner ear of the rat leading to sensorineural hearing loss as documented by ABR testing. They were able to demonstrate perivascular inflammation and fibrosis and degeneration of the spiral ganglion on histopathology, and using immunofluorescent techniques they saw immune complexes in the otic capsule.

Unfortunately, the lack of scientific understanding of the etiology or etiologies of the problem limits its treatment to simple empiricism. Every proposed treatment has study to support it, but corroborative data from independent researchers is usually lacking. Perhaps the fatal flaw in most of those studies is that a single treatment is applied to a symptom that undoubtedly has multiple etiologies.

Wilson and his colleagues did a double blind study of 67 patients with sudden sensorineural hearing loss. 33 were treated with oral steroids and 34 received placebo. 52 additional subjects served as controls. 76%
of patients who had hearing loss greater than 90 Db failed to recover regardless of whether or not they received steroids. 78% of the steroid treated patients with hearing loss less than 90 Db improved, compared with only 38% of matched untreated patients. There have been a few other studies supporting these findings. However, Dr. Wilson returned to his lab and several years later published a report failing to demonstrate improved recoveries in patients with proven viral antibody seroconversions treated with steroids.

The use of diuretics is sort of borrowed from their use in the treatment of Meniere's disease. The use of heparin and warfarin is borrowed from their use in cerebral vascular occlusion in which they seem to promote better collateral circulation. Dextran is an example of an effective plasma expander that is useful in reducing red cell sludging and improving microcirculation. Stellate ganglion blocks produce Horner's syndrome with vasodilation and increased blood flow and is done by making an injection lateral and inferior to the thyroid gland to the C-7 vertebra, and then backing off slightly to the area of the ganglion and injecting Lidocaine. Haug, Draper and Haug demonstrated a 70% chance for a patient to recover a 10 Db pure tone gain and 10 dB discrimination improvement with this procedure compared to a 20% chance of a similar gain if not treated with stellate ganglion blocks. They also noted that the most successful cases were treated in the first 20 weeks after symptoms appeared. Some of the vasodilators commonly employed include nylidrin hydrochloride, papaverine hydrochloride, nicotinic acid, histamine, atropine, and carbogen. Despite the many studies over the years looking at various vasodilators, there has been no adequately controlled study that supports the use of vasodilators in sudden hearing loss. Suga and Snow in their 1969 report actually demonstrated experimentally that vasodilators could in fact reduce cochlear blood flow by causing shunting. Professor Fisch performed several elegant studies and concluded that carbogen was the safest and most effective.

"Shotgun" therapy includes as many potentially helpful treatment regimens as can be safely combined. However, there are very few studies looking at the efficacy of shotgun therapy and those that do exist obviously have so many variables that they are impossible to control for.

The workup and shotgun regimen recommended by the faculty here at Baylor is attached. This treatment protocol was put together based in part on the experience of the faculty and in part on the information available in the literature, but again there is no scientific data to show that it is any more effective than other treatment protocols or even expectant observation.

In summary, sudden sensorineural hearing loss remains a poorly understood and frustrating problem for the otolaryngologist. More rational treatment will probably only follow the elucidation of the specific cause or causes of this disease process.

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**Case Presentation**

A 55-year-old healthy white male smoker with no previous otologic history, noted the acute loss of...
hearing in his right ear during his usual morning activities. He denied strenuous activity stating that he had finished brushing his teeth and began to put on his clothes when he suddenly noted a crescendo high pitched tone in his right ear and inability to hear. There were no other associated symptoms of dizziness or visual change. After several hours without improvement, he presented to the emergency room and an otolaryngology consult was obtained. Head and neck examination including otoscopy was completely normal. An audiogram revealed a severe primarily sensorineural sensitivity loss on the right. Laboratory studies including SMA-20, CBC, thyroid function tests, and urinalysis were within normal limits. Syphilis serology was non-reactive and erythrocyte sedimentation rate was minimally elevated at 15 mm/hr. The veteran was admitted and placed on the Baylor sudden sensorineural hearing loss protocol which will be described.

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Sudden sensorineural hearing loss (SHL) is one of the most perplexing and controversial unsolved mysteries in otolaryngology. SHL has no universally accepted definition, but one useful definition is an idiopathic hearing loss of sensorineural origin, greater than 30 dB in three contiguous frequencies that occurs in less than three days. Most patients report onset of hearing loss within minutes to several hours. About one-third of patients will awaken with the loss; others may discover the problem when they try to use the telephone, or may describe a brief period of fluctuating hearing before the loss. About 50% of patients complain of concomitant unsteadiness or vertigo.

Most studies find no seasonal, geographic, ethnic, racial or sexual predilection for SHL. The right and left ears appear equally vulnerable. In a series of 225 cases, Byl in 1984 noted bilateral SHL in 2% of
patients. Reported overall incidence of SHL ranges from 5% to 20% per 100,000 persons per year. The mean overall age for SHL is 46 years, according to a large series by Mattox and Simmons.

Sudden hearing loss is most likely a symptom rather than a discrete disease, merely representing the end result of many insults to the inner ear. Known etiologies of sudden hearing loss include multiple sclerosis, Cogan's syndrome, Buerger's disease, macroglobulinemia, leukemia, sickle cell disease, polycythemia, syphilis, bacterial infection, mumps, ototoxic drugs, metastatic tumors, trauma, and lupus. Ten percent of patients with Meniere's disease and up to 10% of acoustic neuroma patients will present with sudden hearing loss. In patients with sudden hearing loss, all of these known etiologies should be considered and ruled out before the most common "diagnosis" - idiopathic sudden hearing loss (SHL) - is accepted. Among patients with SHL, four etiologic theories remain pre-eminent: viral, vascular, membrane rupture, and auto-immune.

Viruses have long been suspected as etiologic agents in SHL. Wilson et al, in 1983, studied 122 patients with SHL in the Boston area, and documented 63% viral seroconversion, compared with 40% of controls. Conversion rates were statistically significantly higher for mumps, rubeola, varicella-zoster, influenza, and CMV. Mumps virus was cultured from the perilymph of a patient with SHL by Westmore in 1979, and CMV has been cultured from the perilymph of a congenitally infected infant. Davis, Davis and Johnston, and Shimokata et al documented viral infection of the inner ear in animal models with CMV, mumps, and rubeola, respectively. However, SHL has not successfully been induced in an animal model using viruses. Schuknecht and Donovan in 1986 studied the temporal bone pathology in twelve ears with SHL and reviewed the literature for ten others; comparing the histopathologic findings in these SHL patients with findings from patients with known viral labyrinthitis, they found similar degenerative changes: atrophy of the organ of Corti, tectorial membrane, and stria vascularis in most cases. They concluded that viral cochleitis was the most probable cause of SHL.

Simmons in 1968 proposed a double membrane break theory for SHL. Goodhill et al in 1973 discovered perilymph fistulas in three patients with sudden hearing loss, thus establishing round and oval window breaks as accepted etiologies for sudden hearing loss. Intracochlear membrane breaks, with and without round or oval window breaks, as a cause for SHL, have been poorly supported clinically and histopathologically. Most patients with SHL do not provide a history suggestive of implosive or explosive forces on the membranous labyrinth; Schuknecht and Donovan found no active or healed membrane breaks in the twelve temporal bones they examined.

A vascular etiology for SHL is logically appealing, since sudden onset is suggestive of a vascular event. Histopathologic findings after permanent occlusion of the labyrinthine vasculature include necrosis of the membranous labyrinth followed by ossification and fibrosis - these findings are not consistent with those seen in most cases of SHL. Furthermore, temporary occlusion of the internal auditory artery was studied by Perlman et al in 1959, and these workers found irreversible loss of cochlear function after about one hour. They found that the hair cells, ganglion cells, and spiral limbus were the structures most frequently affected. These histopathologic findings are at odds with those from SHL ears studied. The irreversibility of the loss is also inconsistent with the reversibility of SHL in most cases. Finally, SHL typically strikes
relatively young patients without systemic manifestation of vascular disease. This population seems unlikely to suffer isolated labyrinthine vascular compromise.

In recent years autoimmune ear disease has been added to the theoretical etiologic triad of vascular, viral, and membrane rupture. Lehnhardt in 1958 first theorized that bilateral hearing disorders could result from autoimmune reactions directed against inner ear antigens. McCabe in 1979 first proposed the clinical entity of autoimmune sensorineural hearing loss. Yoo et al in 1983 induced autoimmune sensorineural hearing loss in rats by immunization with type II collagen, and demonstrated the presence of monoclonal antibody to type II collagen within the otic capsule. Harris in 1983 demonstrated the full competence of an independent inner ear immune system, capable of producing its own antibodies. In further work reported this year, Drs. Yamanobe and Harris isolated five autoantibodies to inner ear-specific antigens in patients with idiopathic sensorineural hearing loss. No study has yet detected inner ear-specific autoantibodies in SHL, and no study has proven the development of SHL in animal models using such antibodies. The role of autoimmunity in SHL has yet to be clarified.

Given the lack of understanding of the etiology or etiologies of SHL, treatment regimens have of necessity been empirical. One of the most commonly used treatments is vasodilator therapy. Many agents have been tried, but one of the most popular has been carbogen, which is a gaseous mixture of 5% CO2 in 95% O2. Ugo Fisch in 1983 compared carbogen therapy with papaverine and low molecular weight dextran therapy in SHL patients. He found no significant short term difference in hearing results, but discovered significantly better hearing in carbogen-treated SHL patients after one year.

Wilson et al in 1980 reported a double-blinded study of 67 SHL patients treated with steroids or placebo, and included an additional 52 controls who received no treatment. They found that patients with isolated midfrequency losses recovered without regard to therapy, and that 76% of patients with losses greater than 90 dB failed to recover regardless of therapy. However, in 74 patients with hearing loss less than 90 Db not limited to the midfrequencies, 78% treated with steroids improved, compared with 38% of untreated patients. Subsequent studies have confirmed the benefit of steroids in selected patients. This is the only controlled study showing conclusive superiority of any treatment for SHL over no treatment.

Other treatment regimens have included diuretics, anticoagulants, plasma expanders, and diatrizoate meglumine, or Hypaque, an intravenous contrast dye. Controlled studies have not been performed to show a benefit from any of them. Stellate ganglion block is an invasive technique used in SHL therapy. Ganglion block causes vasodilation and subsequent increased blood flow through sympathetic blockade. Haug et al from Houston reported in 1976 that 70% of 56 patients with SHL treated by a series of blocks experienced greater than or equal to 10 dB pure tone average improvement and over 10% gain in speech discrimination scores, versus only 20% of 20 patients treated by other means. Because this procedure involves deep injection of anesthetic in the low neck, it carries obvious risk of lung injury and injury to vital neural and vascular structures. Many centers, including Baylor, use a so-called "shotgun" approach to SHL treatment, in an attempt to address all theoretical etiologies. Wilkins et al in 1987 evaluated one exhaustive shotgun regimen of dextran, histamine, Hypaque, diuretics, steroids, vasodilators, and carbogen, but found the results no different from those for no treatment.
It is generally agreed that spontaneous recovery is common in SHL, usually occurring within two weeks of onset. Mattox and Simmons stated that 65% of all SHL patients will have spontaneous recovery of functional hearing without treatment. Others have estimated that about one-third have spontaneous return to normal hearing, and another one-third have return to functional hearing with a residual deficit. Obviously, the ideal treatment protocol would improve on these numbers. Only steroid treatment, as evaluated by Wilson et al, in 1980, has been proven superior to no treatment, and this success came only in patients with moderate loss across many frequencies. Byl in 1984 reported a prospective study of 225 SHL patients. He found the most important factors influencing recovery to be severity of initial loss, degree of vertigo, and time from onset to initial visit. All of these factors were inversely related to degree of recovery. Other facts identified by Byl and others include patient age, shape of initial audiogram, and ESR. Byl found that patients under 15 years of age and over 60 recovered less well than their counterparts within these two age limits. Most researchers agree that SHL patients with upsloping and midfrequency loss audiograms recover the best, while those with downsloping initial audiograms fare the worst. Elevated ESR has been shown to be a negative prognostic sign. Tinnitus, present in 70% to 85% of SHL patients, has no prognostic value.

Sudden sensorineural hearing loss remains a poorly understood and maddening problem for the otolaryngologist. Prognostic predictors include audiogram shape, severity of loss, patient age, presence of vestibular symptoms, and duration of symptoms. Viral and autoimmune mechanisms are currently the best etiologic candidates, but the heterogeneity of patients and findings makes the discovery of a single pathologic pathway for all SHL unlikely.

Case Presentation

A fifty-two-year-old Latin American man presented to Ben Taub General Hospital with a twelve hour history of sudden hearing loss in the left ear. The patient had been watching television when he felt a pop in his left ear, followed by tinnitus and subjective hearing loss. He denied dizziness, vertigo, nausea, or vomiting, and gave no history of straining, trauma, or barotrauma. He also denied any history of ototoxic drugs or otologic surgery. Hearing was subjectively normal bilaterally prior to this episode. Past medical history was remarkable only for insulin-dependent diabetes mellitus, and social history was negative for drinking, smoking, and drug use. Physical examination was unremarkable except for tuning fork tests indicating a left sensorineural hearing loss. The patient had a negative fistula test and no spontaneous nystagmus. Laboratory studies were all within normal limits, including an ESR of 2 and negative syphilis serologies. Admission audiogram showed a pure tone average of 73 dB on the left and 8 dB on the right, and speech understanding depressed to 32% on the left, normal on the right. Tympanograms were Type A bilaterally. The patient was admitted and put to bedrest with a low salt diet. He underwent stellate ganglion blocks times three, and was started on a prednisone taper from 80 mg and given carbogen treatments. He experienced subjective improvement in his hearing on day three, and repeat audiogram on day four showed pure tone average of 42 on the left, with speech discrimination of 60%. The patient was discharged on day 5 on prednisone taper and low salt ADA diet, with light activity. Outpatient MRI of
the head was normal. At follow-up two months later, the patient reported subjective return of normal hearing. Audiogram showed pure tone average of 12 dB on the left, 8 dB on the right, with 100% speech discrimination bilaterally.

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TEMPORAL BONE FRACTURES
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April 2, 1992

Temporal bone fractures may cause a variety of signs and symptoms. These include hearing loss, vertigo, facial paralysis, cerebrospinal fluid (CSF) otorhinorrhea, tympanic membrane (TM) perforation, hemotympanum, canal wall laceration, and Battle's sign. Adults and children are similarly affected but several series have found that the complication rate is lower in children.

Fractures of the temporal bone are commonly classified based on the relationship of the fracture line to the long axis of the temporal bone. Longitudinal fractures are the most common accounting for 80% of the temporal bone fractures. This fracture extends along the length of the temporal bone. Transverse fractures extend directly across the petrous bone and make up about 10% of temporal bone fractures. Mixed fractures display some characteristics of each.
Longitudinal fractures usually present with classic findings of laceration of the ear canal, tympanic membrane perforation, ossicular disruption, facial paralysis, and hearing loss. The hearing loss is predominately conductive but may have a sensorineural component as well. Facial paralysis occurs in about 10-20% of longitudinal fractures. These fractures result from trauma directed laterally over the temporal area. Conversely, transverse fractures of the temporal bone show hearing loss, hemotympanum, facial paralysis, and vertigo. Facial nerve paralysis occurs about 50% of the time. Transverse fractures result from trauma to the anterior and posterior aspects of the head.

Recently a new classification system was proposed by Ghorayeb and Yeakley at The University of Texas, Houston. They studied 150 temporal bone fractures radiographically using CT scan with three-dimensional reconstruction. Based on their findings they proposed a new classification system. They identified an oblique fracture type that had originally been described by Voss. In their review this was the most common fracture type, occurring in 74.7% of the temporal bone fractures in this series. Other fracture types encountered in this series include transverse, mixed, longitudinal, and fractures confined to the petrous apex. They differentiate the oblique and longitudinal fractures based on the orientation of the fracture line on the external aspect of the temporal bone. The oblique fracture crosses the external canal in a horizontal plane and then extends upward obliquely toward the middle fossa. The fracture misses the otic capsule and may extend toward the petrous apex where the fracture line may extend to the foramen lacerum. Conversely, the longitudinal fracture line is oriented in a more vertical plane.

Histopathologic examination of temporal bone fractures reveals several patterns of injury. As previously mentioned, longitudinal fractures extend anterior to the otic capsule and do not cause direct injury to the otic capsule. In transverse fractures the otic capsule may be completely disrupted leading to complete loss of audiovestibular function. Fractures of the otic capsule do not heal, but may have fibrous union and new bone formation. This lack of healing may lead to infectious complications and the development of labyrinthitis ossificans. This is an important consideration because of the potential for cochlear implantation in patients with bilateral temporal bone fractures and profound deafness. Loss of audiovestibular function results from concussion of the inner ear as well. This may occur in patients with longitudinal fractures or may occur in patients without a temporal bone fracture. As demonstrated experimentally in cats by Schuknecht, high frequency hearing loss may occur from the transfer of vibratory energy to the cochlea. Lindsay described the changes noted in concussion and found the predominate injury to be hemorrhage into the audiovestibular elements. It is thought that hemorrhage induces a hyperplastic inflammatory response which may lead to degeneration of neural elements, fibrosis, and eventual ossification.

The examination of a patient with suspected temporal bone fracture begins with a history and physical exam. Often times the initial assessment occurs in the emergency room where the patient is also being evaluated for other more life threatening injuries as well. History about the mechanism of injury should be obtained along with any audiovestibular symptoms. The physical exam begins with a general examination of the patient. The head and neck should be examined carefully, looking for the signs and symptoms of temporal bone fractures. The facial nerve should be carefully examined and any evidence of weakness documented. When the patient's condition improves audiogram and CT scan of the temporal
bones should be performed with further workup directed by any subsequent complications. These complications include hearing loss, vertigo, facial nerve injury, and cerebrospinal fluid otorrhea.

Conductive and sensorineural hearing loss can result from temporal bone fractures. Classically, conductive loss is associated with longitudinal fractures, and sensorineural loss with transverse fractures. But each may have components of both. The etiology of the conductive hearing loss may result from tympanic membrane perforation, hemotympanum, and ossicular disruption. Persistent conductive hearing loss suggests an ossicular disruption. Middle ear injury with ossicular derangement was initially reported at the time of autopsy by Keleman, and was noted by Thornburn in 1956. Shortly thereafter Hough described this as well, and later published his experience with 31 patients. He found that the most commonly damaged ossicle is the incus. This is thought to result from its relative lack of support compared to the other ossicles; and its location between the other two ossicles, which subjects it to torsional forces. Treatment of these injuries requires middle ear exploration and appropriate ossicular reconstruction.

Sensorineural hearing loss may result from disruption of the membranous labyrinth or concussion. Treatment options for traumatic sensorineural hearing loss are limited. For the majority of patients the only option is rehabilitative. This consists of hearing aids and cochlear implantation for patients with profound bilateral hearing loss.

Another sequelae of temporal bone fractures is vertigo. The incidence varies between different studies with a incidence of 78% by Tuohimaa, while Griffiths reports only 24%. The mechanism of the vertigo may vary and can result from a variety of causes which may be central or peripheral. Central causes are related to injury to the brainstem and result from dysfunction of the vestibular nuclei. There are a number of peripheral causes, the most common being benign positional vertigo. Patients that have associated vertigo with a fluctuating hearing loss should be suspected of having a perilymph fistula. Disruption of the labyrinth may lead to a unilateral weakness that causes vertigo. Another cause of vertigo that has been reported is endolymphatic hydrops. Rivzi describes a patient who sustained a transverse temporal bone fracture that left the otic capsule intact. This patient developed Meniere's-type symptoms and several months later died of unrelated causes. On examination of the temporal bones the fracture line was noted to cross the vestibular aqueduct. Since the majority of these patients' symptoms resolve within 12 months, initial treatment is conservative. Patients who have persistent incapacitating symptoms may require vestibular nerve section or labyrinthectomy.

Temporal bone fractures may result in facial nerve paralysis. Approximately 20% of longitudinal fractures and 50% of transverse fractures result in a facial palsy. This derives from edema, intraneural hemorrhage, bony fragment impingement and dehiscence of the nerve. In a review of 15 cases of longitudinal temporal bone fractures at Baylor, Coker et al found the perigeniculate region to be the most common site of injury. This injury occurred there about 93.3% of the time. Surgical findings noted neural edema present in 93%, intraneural hemorrhage in 40%, bony fragment impingement in 33%, and one patient with dehiscent geniculate ganglion and dehiscent proximal tympanic segment. In this same
review, three patients with transverse temporal bone fractures underwent surgical exploration and they were noted to have injury to the labyrinthine and tympanic segments of the facial nerve.

The evaluation of patients' facial paralysis from temporal bone fractures begins with early diagnosis. Patients with complete paralysis should be followed up with electrical testing. Several tests are available, with nerve excitability test, ENOG, and EMG being the most commonly used. Audiogram is also important since hearing status influences the choice of surgical approach. Initial treatment is conservative, consisting of eye protection and steroids. Facial nerve exploration and decompression is indicated for patients who show significant neural degeneration. For patients with serviceable hearing, decompression by a middle cranial fossa/transmastoid approach is best. For those without hearing a translabyrinthine/transmastoid approach is used.

CSF otorrhea may result from temporal bone fractures, although the incidence is relatively low. In a review of 1,185 patients with skull fractures Raaf found that 3.6% presented with CSF otorrhea. Of these, 97% closed spontaneously. Of the patients with otorrhea 7.8% developed meningitis, with one patient dying. Patients with longitudinal fractures usually develop CSF leaks from the tegmen where the fracture line perforates the dura. Transverse fractures are more likely to cross the vestibule and leakage may occur through the defect. Also, since the TM may be intact they are more likely to present with CSF otorhinorrhea. In most cases identification of the fluid is obvious. In questionable cases the most specific test available is electrophoretic identification of the B2 fraction of transferrin, which is specific for CSF. For persistent leaks localization may be attempted with CT scan with metrizamide.

Since the majority of these leaks close spontaneously management is conservative. Several studies have failed to show any benefit from prophylactic antibiotics, and, while it remains controversial, they are probably not indicated. Initial management includes elevating the head and avoiding elevations of intracranial pressure. If this fails to resolve the leak the next step in management is insertion of lumbar drain.Leaks that persist, despite these measures, require surgical closure. While the period of waiting is controversial, surgical closure should be considered for leaks that have persisted for two weeks despite conservative therapy. The surgical approach is based on the suspected location and status of hearing. If the patient has good hearing and the defect is suspected in the tegmen, a middle fossa approach may be used. For patients with no hearing, obliteration via a translabyrinthine approach may be used.

In a review of 82 temporal bone fractures in 75 patients Ghorayeb et al presented several other more unusual complications of temporal bone fractures. These include abducens nerve palsy which may be bilateral or unilateral, trigeminal paralysis, and aseptic sigmoid sinus thrombosis. In their review the incidence of abducens palsy was 6.67%. The abducens nerve has the longest intracranial course of any cranial nerve and is intimately associated with the facial nerve and the trigeminal nerve. It appears the nerve is most vulnerable as it enters Dorello's canal at the end of the petrous apex. Because of the complex anatomy of this nerve the exact mechanism of injury is unclear and may result from stretching and contusion alone. Patients present with an inability to abduct the eye. Many of these injuries are partial and may recover. Treatment consists of alternately patching one eye and if the injury fails to recover, corrective surgery may be attempted.
Ghorayeb et al found an incidence of trigeminal paralysis in 2.67% of the cases. This injury also occurs near the end of the petrous apex where the trigeminal ganglion sits in Meckel's Cave. Patients present with hypesthesias and paresthesia in the trigeminal distribution. They may also have weakness of the muscles of mastication. No therapy is available for this injury.

Sigmoid sinus thrombosis was found in one patient in this series. Patients may be asymptomatic or symptoms of increased intracranial pressure may occur. These include headache and retinal changes. The diagnosis is made with CT scan which shows occlusion of the sigmoid sinus with soft tissue. Clinically the diagnosis is made with the Tobey-Ayer Test, which consists of measuring changes in the CSF pressure with compression and release of the jugular veins. For septic sigmoid thrombophlebitis the treatment of choice is exploration of the sinus via a mastoid approach. For aseptic sigmoid sinus thrombosis Ghorayeb et al caution against exploration in debilitated patients, but advocate exploration as part of any other ear procedure. The patient presented in this review underwent facial nerve decompression and at that time the sinus was explored.

Another rare complication of temporal bone fractures is cholesteatoma formation. This occurs in fractures that involve the external canal. With the disruption of the bony external canal small amounts of epithelium may be trapped. Many years later these patients may present with large and extensive cholesteatomas.

Due to the complex anatomy, temporal bone fractures may cause a variety of disorders. Often the patients have sustained other more life threatening injuries that take precedence over the evaluation of temporal bone fracture. Early identification of the fracture, along with careful observation in the recovery period, is necessary for proper treatment.

Case Presentation

A 21-year-old black male presented to the Emergency Room after suffering blunt trauma to the head. He reportedly was hit with a vase in the right temporal area and lost consciousness at the scene. He complained of decreased hearing and bleeding from the right ear, but denied any vertigo. Physical exam of the right ear revealed a posterior external canal wall laceration and a hemotympanum. The facial nerve was intact. CT scan of the head and temporal bone was obtained which revealed a longitudinal fracture of the right temporal bone. He was admitted to the Neurosurgery Service for observation. On the second day of hospitalization he developed a complete right facial paralysis and he was transferred to the Otolaryngology Service. Audiogram showed a mild conductive hearing loss in the right ear, and acoustic reflexes showed a probe effect in the right ear. He was started on steroids and received local care for the right eye. He was followed daily with nerve excitability tests with the side to side difference remaining less than 2mA. On the seventh day after the onset of the paralysis, nerve excitability testing revealed a greater than 3.5 mA difference between sides. ENOG revealed greater than 95% degeneration of the facial nerve. He subsequently underwent a combined middle cranial fossa/transmastoid exploration and
decompression of the facial nerve. At the time of surgery a bony spicule was found impinging on the facial nerve at the geniculate ganglion. The facial nerve was intact but edematous in the labyrinthine and tympanic segments. The postoperative course was uneventful and the patient was discharged to home on the sixth postoperative day.

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 TEMPORAL BONE GUNSHOT WOUNDS: 
EVALUATION AND MANAGEMENT 
Douglas D. Backous, MD 
August 5, 1993 

With the significant increase in handgun availability since the 1960s, treatment of gunshot injuries is no longer limited to major trauma centers. Five thousand Americans under the age of 20 die annually from gunshot wounds in the United States alone, with many more victims surviving with significant injury. The easy availability of guns is seen in rural as well as in metropolitan areas across the country.

Wound ballistics is the study of the effects of missiles striking tissues. Kinetic energy is directly proportional to the mass of the projectile and proportional to the square of the velocity at which the bullet is traveling. The most efficient missile enters tissues and imparts the majority of its kinetic energy to
surrounding structures without exiting the victim. Traditionally, gunshot injuries could be classified as civilian or military. Civilian injuries were inflicted with low velocity weapons (90-210 m/sec) while military wounds typically were caused by high velocity weapons (610 m/sec and up). Due to medium to high velocity weapons appearing on the streets, more civilians are suffering from military-type injuries.

Gunshot injuries to the temporal bone affect intracranial, vascular, middle or inner ear, cranial nerve, and external canal structures. Being the toughest bone in the body, the temporal bone deflects missiles, often resulting in hidden injuries. Early intracranial injuries include skull fractures, subdural hematoma, foreign bodies, pneumocephalus and cerebrospinal fluid leaks. Temporal lobe abscess, meningitis, aphasia, hemianopsia, and hemiplegia are late intracranial complications. Venous injuries to the transverse and sigmoid sinuses, jugular system and cavernous sinus have been reported. Acute transection of internal or external carotid arteries is usually obvious while pseudoaneurysms and carotid-cavernous fistulae can develop later during the recovery period. Tympanic membrane perforations, ossicular disruption, labyrinthine destruction, and mixed longitudinal and transverse temporal bone fractures are the most commonly associated middle ear injuries. The majority of victims suffer acoustic injury inflicted either by direct damage from the bullet path or, more commonly, from blast effects. Fifty percent of victims will have facial nerve involvement with transections by far outweighing contusions. The vagus, hypoglossal, and abducens nerves are the next most frequently damaged cranial nerves. Bony and cartilaginous fractures, cicatricial scarring, temporomandibular joint displacement, canal cholesteatoma, and chronic bony infections can occur after gunshot injury involving the external auditory canal.

Initial assessment includes a standard trauma workup with attention directed to life-threatening problems first. General and neurosurgical clearance is mandatory prior to thorough head and neck and otologic examination. Evaluation of the ear should be done with sterile instruments to avoid further soft tissue contamination. Computed tomography of the temporal bones is ordered once the patient is stable. Carotid and vertebral arteriograms should be obtained at the slightest suspicion of potential vascular involvement. Venous phases of these studies are important. Audiometric testing documents hearing at baseline and prior to any surgical manipulation of the otic structures and aids in selecting approaches to the middle ear and facial nerve. Vestibular testing may be obtained to document the nature and location of lesions in symptomatic patients and is useful to follow vestibular compensation.

Patients are first stabilized from life-threatening injuries. Acute care for intracranial injuries includes intravenous steroids and osmotic diuretics. Antibiotic prophylaxis is key, especially if dural injury is suspected. Lumbar drainage and craniotomy should be employed as indicated. Embolization, packing at the skull base, and vessel ligation in the neck are options for vascular injuries. Mastoidectomy and tympanoplasty should be considered whenever remaining anatomy permits and residual hearing is documented. Debridement of the external auditory canal, labyrinthectomy and temporal bone obliteration should be used depending on the extent of destruction within the temporal bone and depending on the status of remaining critical structures.

Presentation with a complete facial nerve paralysis signals a transection and should be managed aggressively. Exploration of the wound to localize damaged nerve endings is best done before 72 hours
when distal nerve segments have not undergone degeneration. Exploration is often delayed while other injuries are treated. Exploration is then best completed in the subsequent 6 to 12 months. The best results are accomplished with end-to-end anastomosis. Due to local destruction from blast effects, end-to-end anastomosis is frequently not feasible mandating interposition grafting techniques. The great auricular nerve provides 5 cm to 7 cm of graft material while the sural nerve affords up to 30 cm of donor tissue. Delayed loss of facial nerve function most likely results from nerve edema or contusion and decompression suffices as definitive operative management. House-Brackmann grade 4 recovery or worse is expected with interposition grafting while up to grade 3 recovery has been documented from decompressions. Local eye care is mandatory to avoid exposure keratitis while nerve recovery progresses. Other cranial nerve injuries are managed symptomatically. Long-term follow-up is required since many of these patients will need secondary facial reanimation procedures.

In conclusion, multispecialty assessment is required for patients with gunshot wounds involving the temporal bone. Life-threatening injuries are addressed first, followed by a thorough head and neck workup. Care must be taken to avoid missing hidden injuries induced secondary to blast effects or due to deflection of the missile off of the otic capsule. Indications for surgical exploration of the temporal bone include facial nerve paralysis, vascular injury, cerebrospinal leakage, chronic otitis media, conductive hearing loss and to debride necrotic tissue.

Case Presentations

A 44-year-old black male presented to the shock room at Ben Taub General Hospital with a single gunshot wound to the left preauricular area. On admission, he was awake and alert, with a complete left facial nerve paralysis. He had significant tissue loss on the left side of his face and in the left tonsillar area. After airway stabilization by endotracheal intubation, emergent computerized tomographic scanning of the head was negative for intracranial pathology and four vessel arteriograms showed no vascular injury. A barium swallow revealed no extravasation into the pharynx or esophagus. He was extubated after 48 hours. The left external auditory canal was remarkable for bloody discharge and squamous debris, with the Weber lateralizing to the right. The remaining cranial nerves were intact. No other injuries were found after thorough physical examination. The patient had swallowed the bullet that was originally presumed to be resting in his posterior oropharynx. Temporal bone CT scans revealed a comminuted left mastoid tip and fractured external auditory canal and temporomandibular joint. Audiogram showed no responses in the left ear and a moderate sensorineural loss on the right. No response could be elicited by nerve excitability testing on the left side. The patient was discharged after 11 days in the hospital. He was readmitted seven days later, at which time he underwent left radical mastoidectomy, complete facial nerve decompression, sural nerve grafting and meatoplasty. The patient did well postoperatively and has been followed regularly in clinic. He has a House-Brackmann grade VI recovery and is now, at two years postrepair, being considered for a secondary procedure to improve his facial tone and to gain left eye closure.
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TUBERCULOUS OTITIS MEDIA
Troy Callender, MD
November 12, 1992

Despite tremendous advances in the treatment and prevention of tuberculosis (TB) it remains one of the most common, lethal infectious diseases in the United States. Worldwide, TB is the single most important bacterial infection, with more than 7 million active cases. Fortunately the ear is rarely infected by mycobacterium tuberculosis; current incidence is estimated to be between 0.9% and 0.04%. The tubercle bacillus can spread to the middle ear by several routes. The most common route is hematogenous. Other routes of infection include regurgitation of the tubercle bacillus through the eustachian tube, through previously existing tympanic membrane perforations; and by direct extension from a nasopharyngeal site of infection. TB can also be transmitted congenitally and is associated with a high incidence of ear involvement. However, congenital TB is extremely rare and hardly ever presents with isolated ear involvement.
Classically, tuberculous otitis media is described as having an insidious onset with painless otorrhea, multiple tympanic membrane perforations, abundant pale granulation tissue in the middle ear, early severe hearing loss out of proportion to clinical findings, and bone necrosis. However, the current literature indicates most patients actually present with a single perforation. Indications of tuberculous otitis media include hearing loss out of proportion to clinical findings, failure to respond to the usual medical therapy, post-mastoidectomy recurrence of granulation tissue, slow wound healing, persistent otorrhea, and the formation of bony sequestra.

The evaluation of these patients should include a history of contact to active tuberculosis, placement of a PPD skin test with control, culture and stain of ear drainage for AFB, biopsy of granulation tissue for histology and culture, and evaluation of the immune system if indicated. All patients should be evaluated with a chest x-ray, urinalysis, sputum for AFB culture and smear, gastric aspirates in children, and lumbar puncture if CNS involvement is suspected or if the patient has miliary TB. On examination of the ear, the tympanic membrane will appear dull and thickened with dilated vessels on the surface early and will later develop perforations from liquefication of caseous tubercles. There will also be abundant granulation tissue in the middle ear. Late complications include facial paralysis, labyrinthitis, postauricular fistulae, subperiosteal abscess, petrous apicitis, and intracranial extension of infection. Radiologic evaluation of the temporal bone cannot differentiate TB from non-specific infections. A well-pneumatized mastoid with chronic otitis media is suggestive of tuberculous otitis media but not diagnostic, as these cases can also have sclerotic and destructive mastoid lesions.

The differential diagnosis of tuberculous otitis media includes fungal infections, Wegener's granulomatosis, midline granuloma, sarcoidosis, syphilis, necrotizing otitis externa, atypical mycobacterial infections, and histiocytosis X.

The treatment of tuberculous otitis media is primarily medical and should include a 6- to 9-month course of Isoniazid, Rifampin, and Pyrazinamide. Indications for surgical intervention include the late complications of tuberculous otitis media; cases unresponsive to medical therapy; extensive disease with bone sequestra or necrotic bone; and reconstruction of the tympanic membrane and ossicular chain after the middle ear disease has been eradicated.

**Case Presentation**

A 10-month-old black female, developed persistent otitis media at 4 months, which failed to respond to multiple courses of antibiotics. On referral to the Pediatric Otolaryngology Service, examination was remarkable for bilateral dull, thickened, erythematous tympanic membranes with poor mobility on insufflation. Myringotomy on June 22, 1992 revealed granulation tissue filling the middle ear cleft bilaterally. Post-operatively she was improved but had persistent scant otorrhea in spite of outpatient medical therapy. A CT scan of the temporal bones was obtained and showed opacification but no destructive lesions. The patient was returned to the OR on September 23, 1992. Middle ear tissue was
obtained for routine AFB, fungal cultures, and histologic examination, and her ventilation tubes were replaced. She was admitted and a pediatric infectious disease consultation was obtained.

Additional historical data obtained at that time revealed the patient to have had close contact with a relative with active tuberculosis. Complete evaluation during this admission was remarkable for elevated total IgG and IgG1, + ELISA HIV with an indeterminate western blot, miliary CXR pattern consistent with TB, CSF analysis showing 62 WBC's, 2 RBC's, 82% monos, a glucose of 36 mg/dl and protein of 38 mg/dl consistent with tuberculous meningitis. A CT scan of the brain revealed tuberculomas in the posterior fossa and a PPD skin test was positive with 19 mm to 20 mm of induration. ABR was compatible with a mild conductive loss.

Treatment was instituted with isoniazid, rifampin, pyrazinamide, and kanamycin. She was also treated with oral prednisone. She had a dramatic otologic response with resolution of granulation tissue over 2 to 3 weeks. Except for occasional fever she was otherwise well. She was kept in the hospital for 4 weeks to insure compliant therapy and is being closely followed as an outpatient. Final cultures at eight weeks show no AFB growth.

**Bibliography**


TYMPANOSTOMY TUBES
February 23, 1995
Warren Morgan, M.D.

Otitis media is the most common diagnosis in children, affecting two thirds of all children by the age of two years. Recurrent otitis media or otitis media with effusion may be treated with tympanostomy tubes, and approximately 700,000 children undergo this procedure each year. In an article that appeared in JAMA, Kleinman and colleagues studied the medical appropriateness of tympanostomy tube insertion in children under the age of 16. After developing clinical indicators, they retrospectively reviewed a large group of patients. They felt that preoperative indicators for tympanostomy insertion were inappropriate for 25% and equivocal for 33% of the patients reviewed. Despite several biases in the study, this paper received a lot of national attention, increasing parental concern about tympanostomy tubes. In addition, the increase in cost concerns related to health care reform has lead to increased scrutiny as well.
Tympanostomy tubes are indicated for chronic otitis media with effusion, recurrent acute otitis media, tympanic membrane atelectasis, and complications of acute otitis media in children. A recent health care panel convened by the US government recommended tympanostomy tubes as a treatment option for otitis media with effusion that persists for three months with associated hearing loss. If the effusion persists four to six months despite other treatments, then tympanostomy tubes should be inserted at that time. Recurrent acute otitis media (5-6 episodes/year) with persistent middle ear effusion is best treated with tympanostomy tubes. Children with recurrent otitis media without persistent effusion may be treated with antibiotic prophylaxis or tympanostomy tubes. Children who fail antibiotic prophylaxis are candidates for tympanostomy tubes.

No guidelines exist for preoperative audiologic testing in normal children undergoing tympanostomy tube insertion. Preoperative testing may identify undetected hearing loss and document hearing for medicolegal reasons. Manning et al reviewed the audiologic results of 600 children undergoing preoperative audiologic testing prior to tympanostomy tube insertion at Children's Medical Center in Dallas, Texas.

They identified 6 patients (1%) with previously undetected sensorineural hearing loss. Each patient in this group had risk factors for hearing loss. Also, because the study was done in a tertiary children's hospital, the rate of hearing loss was higher than in the community. Despite these factors, they still recommended screening of all patients.

A vast array of tympanostomy tubes have been designed. Tympanostomy tubes have been made out of Teflon, silicone, polyethylene, stainless steel, titanium, and even gold plated steel. Karlan et al performed a laboratory and clinical study comparing silicone and Teflon tubes and found the Teflon to be superior. Metal compounds such as stainless steel, titanium, and gold have been considered good compounds for tympanostomy tube construction. Several studies have failed to show a significant advantage over synthetic tubes.

The shape and design of the tympanostomy tube is more important in determining its function. Large inner flanges increase the duration of middle ear ventilation. Tympanostomy tubes with a large inner diameter and short length are less likely to occlude but may allow easier passage of water. These characteristics must be considered during the selection of tympanostomy tubes.

Tympanostomy tubes may be arbitrarily divided based on the duration of ventilation. Short term tubes are shaped like grommets or bobbins and have and the average duration of intubation is about 10 months. The tubes are easy to insert, have a low rate of obstruction, and low rate of permanent perforation. Long term ventilation tubes are typically T-shaped and have the opposite characteristics. The average duration of ventilation is around 30 months. Both groups of tubes have been modified to provide intermediate characteristics.

Other operative factors may be important in tympanostomy tube insertion. In the past, several authors have felt that ear canal preparation was important prior to tympanostomy tube insertion. But Baldwin and Aland found no advantage with iodine preparation of the external canal prior to tympanostomy tube
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insertion. Myringotomy direction and location have been thought to influence the duration of middle ear ventilation and subsequent healing of the tympanic membrane. Although some authors have reported that intubation is longer when the tube is inserted in the anterior superior quadrant, a study by April et al found no difference between the anterior superior and anterior inferior quadrants. Likewise, myringotomy direction had no effect in a study by Guttenplan et al.

Tympanostomy tubes may be inserted under local or general anesthesia. Some otolaryngologists have reported successful tympanostomy tube insertion using iontophoresis for local anesthesia in cooperative children over the age of six. Summerfield and White described insertion of tympanostomy tubes in 103 patients with an age range of 3-12 years using EMLA cream for local anesthesia. EMLA is a mixture of lignocaine and prilocaine in a high concentration that is absorbed across the epidermis providing local anesthesia. They were successful in 93.2% of the patients, with all failures occurring in children less than four years. Many children undergo the tympanostomy tube insertion under general anesthesia. Markowitz-Spence et al reviewed the anesthetic complications in a series of 510 children undergoing general anesthesia for tympanostomy tubes at the Children's Hospital of Buffalo. No complications were reported in 423 (82.3%). Minor complications without long-term sequelae occurred in the other patients.

Postoperative topical antibiotic prophylaxis after tympanostomy tubes remains controversial. Many otolaryngologists use otic drops to prevent blockage of the tube by blood or discharge and to prevent postoperative otorrhea, despite the theoretical risk of ototoxicity. Garcia et al performed a meta analysis of the five previous studies on the use of antibiotic drops and found a reduction in otorrhea by 85 %. They recommend continued use of topical antimicrobial prophylaxis in high risk cases with mucoid or purulent effusions. For routine cases without effusion and normal middle ear mucosa, they do not advocate routine use.

Water may pass through a tympanostomy tube and cause a middle ear infection. Consequently, most otolaryngologists instruct parents to take precautions to avoid water exposure. This usually includes the use of ear plugs during bathing or water sports, while others forbid water sports. Paradoxically, children with ear tubes who swim do not have an increased rate of otorrhea, as shown in nine different studies.

For middle ear contamination to occur through a tympanostomy tube, water must reach the tympanic membrane and then cross the tympanostomy tube into the middle ear. Middle ear contamination with bath water is much more irritating than other solutions. Studies suggest that middle ear contamination without immersion underware occurs rarely.

Occasionally, a tympanostomy tube will persist beyond the desired period of middle ear ventilation. The otolaryngologist must then decide at what time to remove the tube. Based on an increased rate of complications in tubes that persist for three years or longer, most tubes are removed at 2-3 years. No other tests or criteria exist to determine when they should be removed.

Tympanostomy tubes may have significant complications. Melelland reviewed his series of 307 patients under the age of 10 years who had been treated with tympanostomy tubes and followed for a minimum of 1 year. The most common complication was otorrhea in 34.5% of patients. This include postoperative and
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delayed otorrhea with chronic otorrhea occurring in 5.5% of patients. Tympanic membrane perforation occurred in 6.2% of patients, but only 2.8% had chronic perforation that required surgical closure. Persistent tympanostomy tube and cholesteatoma occurred in 0.2% of patients. Several other studies have been done on the overall rate of complications, showing similar rates.

Long term complications following myringotomy and tubes was reported by Ben-Ami et al for Afula, Israel. They retrospectively reviewed and examined 64 patients 10 years after having myringotomy and tubes. In their review, 77.9% of patients were treated with one procedure, and the remainder required multiple procedures. Two patients had perforations on follow-up. Myringosclerosis was found in 42.3% of tympanic membranes with only 17.8% of the patients having myringosclerosis in the area of the previous tympanostomy tube. Only two patients (1.9%) developed long term otorrhea.

Otorrhea, the most common complication, occurs in around 20% of all patients with tympanostomy tubes. Postoperative otorrhea is most likely related to the presence of purulent fluid or inflamed middle ear mucosa. Delayed otorrhea occurs due to another episode of acute otitis media, which may result from either external contamination through the tube or from nasopharyngeal reflux. Management of post tympanostomy otorrhea usually begins with ototopical agents and antibiotics. If this fails to clear the infection, then a culture is taken to identify any resistant organisms and guide further therapy. Sometimes if all therapy fails, removal of the tympanostomy tube is required.

Chronic tympanic membrane perforation remains problematic with tympanostomy tubes. The rate of perforation varies widely in reported series and is heavily influenced by tympanostomy tube type and duration of intubation. In a number of studies, the rate of chronic perforation with short term grommet type tube is less than 5%, while with long term tympanostomy tubes the rate of perforation is around 15-20%. The longer the period of intubation, the greater the risk of persistent perforation. This factor should be considered during selection of tympanostomy tubes.

Tympanosclerosis, or more accurately myringosclerosis, occurs in around 40-50% of children with tympanostomy tubes. The incidence of tympanosclerosis in chronic otitis media not treated with tympanostomy tubes is around 10%. The etiology of the tympanosclerosis is unclear and is related to both the disease process and the tympanostomy tube. In several studies the myringosclerosis does not cause an appreciable difference in hearing.

The development of cholesteatoma with tympanostomy tubes is a relatively rare occurrence, reported in less than 1% in most series. Cholesteatoma may result from squamous debris being trapped in the middle ear around the tympanostomy tube. In some series, patients developed cholesteatoma in the attic area away from the tube site.

In conclusion, tympanostomy tubes are an important tool in the treatment of chronic otitis media in children. Successful management requires the use of appropriate indications and the proper selection of tube type.
Case Presentation

A 10-month-old female infant was referred to the Otolaryngology Clinic for recurrent otitis media for which she had been treated six times with antibiotics. Each episode responded to antibiotics, but she would develop another episode shortly after completion of antibiotic therapy. She was not routinely exposed to cigarette smoke, and was not in day care. The remainder of her history was normal. Her physical examination was remarkable for bilateral middle ear effusions. After discussing treatment options with the family, they elected to proceed with insertion of tympanostomy tubes. Preoperative audiologic evaluation was normal. She underwent bilateral myringotomies with tympanostomy tube insertion under general anesthesia. At the time of surgery she had thick middle ear effusions. Postoperatively she received a 3-day course of Cortisporin otic solution. She recovered well and has had no further episodes of otitis media.

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A patient presenting with vague disequilibrium and occasional episodic vertigo is a common occurrence in the general otolaryngology clinic. Typical etiologies include status post head injury and the vestibulopathy of the elderly. These patients often have mild progressive symptoms without spontaneous resolution. Many patients, over time, will have resolution of their symptoms, but for the patient who continues to have symptoms, treatment is frequently required. Traditional treatment methods have relied upon medication such as Antivert® and Valium® and perhaps Cawthorne type exercises in an attempt to encourage central nervous system adaptation and rehabilitation. I will briefly describe vestibular physiology, as it relates to compensation, and discuss the design and efficacy of a program of vestibular rehabilitation.
The function of the vestibular system is to transduce head acceleration into a signal the brain can interpret. This information is used by the central nervous system (CNS) for gaze stabilization on the retina and posture control. Transduction is performed for rotational motion by the semicircular canals and for linear motion by the otolith organs. A spontaneous discharge rate is present in the nerve fibers arising from these sense organs and rotation or displacement is excitatory in one direction and inhibitory in the opposite direction. Any difference between the discharge rates of the bilateral symmetrical vestibular apparatus is perceived as motion.

After a unilateral vestibular insult, there is persistent asymmetry in the vestibular nerve discharge rates leading to the incorrect sensation of vertigo and motion. These static symptoms, present in the absence of motion, quickly resolve with restoration of symmetrical discharge rates in the vestibular nuclei. Symptoms brought about by head motion (dynamic) take longer to compensate and the remaining vestibular organ as well as the visual system and the somatosensory network work in concert to maintain posture and balance. In most cases by reliance on these and other inputs the CNS can adapt to the unilateral loss of input and near normal function in all aspects of daily living can be attained. The mechanisms for the static and dynamic aspects of vestibular compensation to a peripheral deficit are largely unknown. Several mechanisms have been proposed: commissural pathways between nuclei of the vestibular system, reactive synaptogenesis, cerebellum nuclei modifications, and denervation supersensitivity. The most likely mechanism for resolution of static symptoms is, however, that neurons of the vestibular nuclei have a spontaneous pacemaker activity which takes over after input is eliminated from the peripheral organ. The compensation mechanism is affected by external manipulations which are of therapeutic value. It is known that sedative medications may prolong the time course and decrease the extent of compensation while stimulants such as amphetamines accelerate compensation. Other external factors can also influence the compensation process. Visual input is important in dynamic compensation and animals deprived of visual input do not manifest dynamic compensation although this has no effect on static deficits. Somatosensory and visual dysfunction impair dynamic compensation. The best example is the diabetic with retinal and somatosensory disease who suffers a new onset vestibular deficit. Because of the existence of dysfunction in all three limbs of the posture and balance system, compensation will likely be very minimal. Exercise is important in vestibular compensation in that immobility has been demonstrated to prevent dynamic compensation from occurring.

The importance of exercise in vestibular rehabilitation is not new, having first been described by Cawthorne and Cooksey in 1946. They noted that patients who exercised early after a unilateral ablation did better and they did so faster than patients who did not exercise. This clinical observation in support of modern experimental evidence provides the basis for rehabilitation; CNS compensation is facilitated by exercises with visual and somatosensory stimulation. The first step in a rehabilitation program is an assessment which is typically performed by a physical or occupational therapist. The patient is questioned about symptoms and an objective clinical assessment of vestibular function is made by physical examination, rotatory function, and posturography. A specific, tailored program is developed to address the particular deficits. The treatment strategy relies on the following factors: balance retraining for postural control mechanisms; eye and head coordination with progressively more difficult visual tracking tests; habituation therapy; and a general conditioning program. Indications for rehabilitation therapy include patients with: movement induced dizziness or poor compensation after a unilateral
vestibular injury; status post head injury; the elderly patient with vague disequilibrium; and poor compensation or recurrent symptoms after surgery.

Several large series have been published recently detailing results from vestibular rehabilitation programs. Horak and Shemway reported a prospective study of twenty-five subjects with a peripheral vestibular disorder and symptoms greater than six months duration. These patients were divided into three treatment groups. The first group underwent a tailored program of twice weekly outpatient exercises. The second group was given a regiment of general (Cawthorne) exercises. Finally, a third group was given medications such as Antivert® and Valium® only. The results are encouraging in that the patients who were on medication alone showed no objective improvement on posturography scores, but in the patients who had a tailored vestibular exercise regiment, there was a statistically significant increase in the posturography scores indicating improvement in compensation. A subjective dizziness index also showed significant improvement with the vestibular exercise program while no improvement was seen with the general exercise program and approximately 50% improvement with the medication alone group.

A prospective study from Shepard et al at the University of Michigan of 98 patients with diagnoses including peripheral, central, and mixed etiologies. These patients participated in a tailored program with twice daily home therapy lasting ten to fifteen weeks. Results showed a reduction in symptoms scores in 87% with complete resolution of symptoms in 36%. Patients with head injury accompanied by postural problems, but with normal visual and somatosensory systems, and those with secondary gain did the worst. Patients with episodic motion dysfunction had the best prognosis for improvement with this protocol.

In summary, vestibular rehabilitation is a important treatment modality for patients with vestibular dysfunction who are not surgical candidates. An increasing proportion of our society, the elderly, are often prime candidates for vestibular rehabilitation therapy. It appears that patients with peripheral vertigo with classical symptoms, especially of an episodic variety will definitely benefit from a vestibular rehabilitation program but also patients with a vague disequilibrium and probable central etiology for this condition may also benefit from vestibular rehabilitation therapy.

Case Presentation

A 39-year-old female had benign paroxysmal positional nystagmus, vertigo, and imbalance secondary to a mild blow to the head. She fell while downhill ski racing and hit her head (no loss of consciousness) but was able to continue her activities within several minutes. She was previously healthy, with no significant medical problems, led an active life, and continued employment as a nurse. Symptoms of vertigo, nausea, and disequilibrium progressed over two weeks post-injury and persisted for ten months.
Neurologic examination was normal; CT and MRI were negative. Tests of horizontal vestibulo-ocular reflex control (caloric and rotary chair) were within normal limits. On ENG testing, the patient demonstrated a directionally fixed vertical oblique nystagmus and vertigo in the right Hallpike position. All other positional tests were normal. Symptoms progressed to the point of interfering with her work and she sought medical treatment. She received outpatient therapy twice a week for six weeks, and twice daily performed home exercises targeted at a variety of postural, gaze, and motion perception disabilities. In addition, she participated in a low-impact aerobics program several times a week. After six weeks of therapy, repeat platform posturography revealed that she had normal body sway during all six of the sensory organization tests and her complaints of dizziness had significantly decreased. She still had mild complaints (intensity two on a scale of zero to ten) in 12 positions. She continued with her exercises, and within six months she was vertigo-free in all positions and has remained so.

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VESTIBULAR NEURITIS

July 14, 1994
Douglas D. Backous, M.D.

Vestibular neuritis refers to a disorder of the vestibular system without an associated auditory deficit or other disease of the central nervous system. It is a unilateral peripheral disorder, primarily affecting patients in their third and fourth decades, which is associated with ipsilateral caloric weakness. Except for persistent unsteadiness, symptoms usually resolve by 3-6 months. The episode of vertigo usually consists of a series of attacks within a 10 day period. Of the patients in Coats' series, 38.2% had an antecedent infection, 20% with sinusitis, and 61% had spontaneous nystagmus. Besides the unilateral caloric weakness, electronystagmography reveals a directional preponderance and beating nystagmus away from the affected side. Vestibular neuritis has also been referred to in the literature as vestibular neuronitis, epidemic vertigo, acute labyrinthitis, vestibular paralysis, and vestibular neuropathy.
A multiple attack variant of vestibular neuritis occurs in a younger age group and consists of recurrent attacks over months to years. Symptoms of unsteadiness and disequilibrium have shorter duration, and patients have less severe caloric weakness. Preceding upper respiratory illness is more commonly reported in this subset of patients.

The differential diagnosis of vestibular neuritis includes Meniere's disease, vestibular schwannoma, perilymphatic fistula, cerebellar infarction, multiple sclerosis, disequilibrium of aging, basilar artery insufficiency, vestibular atelectasis, and metastatic carcinoma. Meniere's disease (endolymphatic hydrops) patients describe tinnitus, fluctuating hearing loss, and frequent episodes of vertigo of duration limited to hours. Caloric responses are normal early in the course of endolymphatic hydrops. Tinnitus and hearing losses are much more common in the presentation of vestibular schwannoma than is vertigo. Patients can have signs of brainstem compression and often have other deficits in cranial nerves, primarily V and VII. The majority of perilymphatic fistula patients describe "popping" while straining as a precipitating event to their symptoms. Vertigo is often positional with fluctuating hearing loss and normal caloric testing. Cerebellar infarction can be caused by hemorrhagic or ischemic events in the distribution of the anterior inferior or posterior inferior cerebellar arteries. These patients report symptoms consistent with the single attack variant of vestibular neuritis. Multiple sclerosis is a demyelinating disease of mainly white matter. Early symptoms include blurred vision, vertigo, cranial nerve palsies, and awkward extremity use. Nystagmus, scanning speech, and intention tremors occur later. There is no sex predilection and patients are typically in their third to fourth decades of life when diagnosed. This disease has an average duration of 20 years from discovery until eventual death. Findings of oligoclonal banding in CSF and white matter plaques on MRI scanning help establish the diagnosis. Disequilibrium of aging is due to degenerative changes seen in the vestibular labyrinth and to interference with fluid motion within labyrinthine sense organs. Sudden head movements exacerbate the vertigo. Basilar artery insufficiency is caused by occlusion of the vestibular branch of the internal auditory artery. This most often occurs in an older age group of people who also have high frequency hearing loss. Additional neurotologic findings are not uncommon. Collapse of vestibular membranous structures either primarily or as a component of another process such as endolymphatic hydrops is described as vestibular atelectasis. The most likely etiologies include viral and degenerative changes seen in aging patients. The principle symptom is unsteadiness with head movement that is not amenable to medical management but is not severe enough to warrant surgical therapy. The symptoms associated with metastatic carcinoma to the temporal bone depend on where the metastasis occurs and can include vertigo or unsteadiness.

Work-up of patients with vestibular neuritis begins with thorough history and physical examination. Audiometry and vestibular testing are the cornerstones to diagnosis with imaging and laboratory studies being guided by findings on examination. Bedside examination includes Doll's eye test, head shaking nystagmus, dynamic visual acuity, caloric testing, rotational testing, past pointing, Romberg and Fukuda tests, and tandem walking.

Treatment is primarily supportive with reassurance that symptoms will resolve in most cases. Vestibular suppressants are reserved for patients with severe symptomatology. Medical labyrinthine ablation or surgical therapy are rarely indicated.
In conclusion, the characteristic symptom complex of unilateral vestibular weakness without auditory or other central nervous system pathology comprises vestibular neuritis. Vestibular nerve atrophy or degeneration may occur but vascular etiologies are unlikely. Treatment is primarily conservative and the majority of patients will recover in 3-6 months. Refractory cases should be investigated for other etiologies.

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**Case Presentation**

A 65-year-old white man presented to the Otolaryngology clinic with a three-day history of severe vertigo. He reported nausea and vomiting for the first two days of his illness but successfully ate breakfast on the day he was seen in clinic. He denied hearing loss and tinnitus. His past medical and surgical histories were unremarkable. He had no previous exposure to ototoxic drugs and denied further neurologic symptoms. Physical examination revealed an obviously uncomfortable white male in a wheelchair. Otologic examination was without abnormality. Weber testing with a 512 Hz tuning fork was to midline. Romberg and Fukuda testing indicated right-sided pathology. Other than a crisp left beating nystagmus, cranial nerve examination was normal. In light of the nystagmus and severe vertigo, the Dix-Hallpike maneuver was not done. Audiometry revealed a symmetrical mild-to-moderate high frequency hearing loss. Acoustic reflexes were mildly elevated bilaterally with adequate speech understanding. Type A tympanograms were present bilaterally. The patient was treated with valium, m q 8 hours. He returned to the clinic 7 days later, able to walk and with persistent unsteadiness. Electronystagmography performed one week later revealed 30% unilateral weakness on the right with a 55% directional preponderance to the right. Rotatory testing revealed a right peripheral disorder. On return clinic follow-up he had a residual feeling of unsteadiness but no vertigo.

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