

TITLE: Unilateral Sensorineural Hearing Loss

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Although a commonly encountered diagnosis by the otolaryngologist, unilateral SNHL represents a difficult clinical entity for the specialist. Whereas the diagnosis is easily obtained with an audiogram, diagnosis of the etiology and treatment represent the complexity of this clinical situation. Etiology of unilateral SNHL is well defined in otology literature, yet precision in diagnosis eludes even the most thoughtful otolaryngologist. Further complicating workup is the ever growing cost for laboratory and radiology studies. As government and third payer parties begin to look for cost containment strategies, clinicians must begin to look more to complete history, physical, and clinical assessment to guide them in which studies are indicated. Further pressure for cost containment comes from patients that are self pay or have personal medical savings accounts with insurances that have large deductions. The physician must not only be able to explain why laboratory and radiology studies are indicated, but on many occasions may be asked to pick the best use for the patients limited funds.

The following paper outlines three commonly encountered clinical situations, 1) childhood unilateral SNHL of unknown duration, 2) Unilateral inner ear symptoms in the adult, and 3) Sudden sensorineural hearing loss. Each entity is presented from a clinical standpoint. As such, differential diagnosis and workup will be the focus of this discussion. Special attention will be given to cost analysis for the above mentioned reasons. As many of the topics have not been addressed completely in literature, subjective assessment may be included in the paper. The author advises personal assessment prior to incorporating the following conclusions into practice.

Childhood SNHL

Unilateral SNHL in the moderate to profound range correlates with poor academic performance, increased chance in repeating a grade, and behavioral problems. Several studies have shown that intervention prior to 6 months of age results in significant advantages in communication.

Universal infant screening has led to earlier diagnosis of congenital hearing loss. Despite

these efforts, the average age of diagnosis of unilateral SNHL is 8.7 years of age. Although some of the delay is explained by late onset hearing loss, many children function well and elude diagnosis until old enough for traditional audiogram. Bilateral SNHL greater than 50 dB is reported in 1-2/1000. Unilateral SNHL greater than 50 dB is seen in 3/1000. Because of the frequency of diagnosis and the delay in diagnosis, the Joint Committee on Infant Hearing in 1994 has given indications for persistent periodic screening of children at risk as follows: 1) Child with a family history of early onset SNHL, 2) prenatal infection that may lead to SNHL (CMV, rubella, syphilis, toxoplasmosis), 3) Neurofibromatosis II or other neurodegenerative disorders, 4) Persistent pulmonary hypertension in the newborn period.

Evaluation initially begins with suspicion of Sensorineural by a parent or school teacher and is confirmed with behavioral audiometry, ABR or OAE. This leads to consultation with the otolaryngologist. Initial evaluation begins with a history specifically searching for family history, syndromic history, head or noise trauma, exposure to ototoxic drugs or chemicals, prematurity, hyperbilirubinemia, or intrauterine and neonatal infection. Physical exam is usually non-contributory, but a specific search for syndromic features and ear deformity may guide further investigation. Further evaluation with laboratory, radiology, and referral as appropriate soon follow.

Differential Diagnosis

Differential diagnosis of the child with SNHL includes all possible etiologies as seen in the adult with the exception of presbycusis. Most likely is a diagnosis of congenital hearing loss, intrauterine infection, syndromic hearing loss, and intrauterine or neonatal insult.

Genetic hearing loss can be divided into syndromic and nonsyndromic hearing loss. It accounts for 4-11/10,000 of the cases of profound early onset deafness. Genetic basis accounts for 50% of all hearing loss in children. Syndromic hearing loss accounts for 30% of all genetic causes, 80% of which are autosomal recessive, 15% autosomal dominant, and 5% x linked or mitochondrial. Syndromic hearing loss, although less common, allows for cost effective evaluation of other associated conditions. Over 200 syndromes include deafness or hearing impairment. Some of the more common will be discussed here.

- Usher syndrome – an autosomal recessive disorder characterized by retinitis pigmentosa and SNHL. Patients with this syndrome require treatment by an ophthalmologist.
- Waardenburg syndrome – characterized by SNHL, pigmentation abnormalities including white forelock, multicolored irises, dystopia canthorum (lateral displacement of the inner canathi), and hypertelorism (lateral displacement of the orbits)
- Jervell and Lange-Nielsen syndrome – autosomal recessive disease. Children with this syndrome have prolonged QT syndrome and congenital SNHL. Seen in 1.6 to 6 / million. Family history of sudden death leads to investigation with EKG. All family members and first degree relatives should be screened with EKG as well.
- Neurofibromatosis Type II – Autosomal dominant syndrome with bilateral acoustic neuromas, meningiomas, spinal schwannomas, and posterior capsular lens opacities.
- Alports syndrome – can be autosomal dominant or recessive, it is most commonly an x-linked disorder. Characterized by slowly progressive bilateral SNHL and progressive renal failure. Persistent microscopic hematuria is a consistent finding. Episodic gross

hematuria precipitated by upper respiratory infection can be elicited in the history of many of these patients.

- Brachio-oto-renal syndrome – found in 1/40,000 live births. 60% of patients have branchial cleft cysts. Shared antigens between the stria vascularis and glomeruli lead to SNHL and renal pathology. 20% of patients have purely sensorineural hearing loss, whereas the majority have mixed hearing loss. Patients commonly have preauricular pits and pinna malformation.
- Noonan's syndrome – seen in 1/2500 live births, patients have a webbed neck, cardiac abnormalities, low set ears, pinna abnormalities, short stature, and in 10-15%, SNHL.
- CHARGE syndrome – Coloboma, Heart defects, Atresia choanae, Retarded growth, Genital hypoplasia, and Ear abnormalities and deafness. Patients with hearing loss have a wedge shaped audiogram. The Mondini malformation is seen in this disorder.
- Fechner's syndrome – a variant of Alports syndrome with high frequency SNHL, proteinuria, macrothrombocytopenia, and ocular disease. This autosomal dominant disease is extremely rare and should only be screened in patients with a family history.
- Pendred syndrome – autosomal recessive disorder. Patients have congenital SNHL, goiter, abnormal perchlorate discharge test, and hypothyroidism. Pendred syndrome is associated with Mondini deformity and a large vestibular aqueduct.

Included in the differential diagnosis for childhood SHNL are environmental insults from infection or exposure to toxins. Intrauterine infections from the TORCHS organisms (Toxoplasmosis, Others, Rubella, Cytomegalovirus, Herpes simplex, Syphilis) cause infectious congenital hearing loss. Meningitis causes SNHL that is most often bilateral. The cochlea may ossify in hearing loss from meningitis over time and cochlear implant, if contemplated, is attempted early. Exposure to ototoxic drugs, either intrauterine or in the neonatal period can cause hearing loss. Maternal drug or alcohol use can cause hearing loss as well. The teratogenicity of accutane (thalidomide) is well documented in literature and can cause hearing loss with multiple other anomalies.

Laboratory and Radiology Testing

Laboratory workup for sensorineural hearing loss in children differs from extensive chemistry, hematology, and function testing to no testing at all, depending on the clinician examining the patient. Below is a list of common tests and the diagnosis that may be elicited from them.

CBC with differential – Drawn to exclude leukemia or lymphoma. Hearing loss from these etiologies is thought to result from a hyperviscosity syndrome leading to ischemic damage to the cochlea, or from infiltration of tumor cells into hearing sensitive areas of the temporal bone. In literature, 1 case of hearing loss as an initial manifestation of leukemia has been reported. This test is highest yield with a history of illness for several weeks, gingival bleeding, bone or joint pain can be elicited.

Platelet studies – Drawn to exclude Fechner syndrome. Because this disorder is extremely rare and will be accompanied by a family history (autosomal dominant), ocular disease, or proteinuria, routine platelets studies are low yield.

ANA, ESR, RF – Drawn to exclude lupus and other autoimmune disorders such as rheumatoid arthritis. These tests are nonspecific. A positive RF is 0.7% sensitive in detecting juvenile RA. These tests are very low yield in children without a history of joint pain or other signs of a systemic autoimmune disorder.

BUN, Creatinine, Urinalysis – used to look for concurrent kidney disease such as in Alport's syndrome, a disease of slowly progressive bilateral hearing loss and progressive renal failure. Episodic gross hematuria precipitated by UTI's can usually be elicited from the history. Persistent microscopic hematuria is a consistent finding that leads to the diagnosis with simple urinalysis. BUN and creatinine are used as confirmatory tests when a urinalysis is positive. This test is low yield without a family history of slowly progressive hearing loss or kidney disease or a history of gross hematuria.

Glucose – Used for screening for Alström syndrome characterized by obesity, impaired glucose tolerance with insulin resistance, retinal degeneration, neurosensory deafness, acanthosis nigricans, hepatic dysfunction, and other endocrine abnormalities. Only 50 cases have been reported since 1959. Because of its rarity and unusual presentation with SNHL only, glucose measurements in asymptomatic individuals is very low yield.

Thyroid function tests – Drawn to look for cretinism and Pendred syndrome. Hypothyroidism in children significant enough to cause hearing loss is usually accompanied by cold intolerance, weight gain, and other symptoms of cretinism. Pendred syndrome is more appropriately diagnosed with an abnormal perchlorate discharge test as the thyroid function test can be normal. Children with goiter, signs and symptoms of hypothyroidism, evidence of large vestibular aqueduct or Mondini deformity should have a thyroid function test. Further suspicion and a negative test may warrant a perchlorate discharge test.

RPR, TTPA – Drawn to look for syphilis. Known as the great imitator. The association of SNHL and syphilis is well established. This test is low yield as SNHL as the only presentation of hearing loss is rare. Any history of maternal syphilis exposure, or signs of tertiary syphilis would justify use of this test.

EKG – Looking for prolonged QT interval in Jervall and Lange-Nielson syndrome. This test is highest yield with a family history of sudden childhood death because of cardiac disease or a history of syncope.

GJB2 gene (Connexin 26) – responsible for as much as 50% of autosomal recessive nonsyndromic hearing loss. Carrier rate has been described as high as 3% in the Midwest. GJB2 is found in 35% of moderate to profound SNHL. This test is high yield. Its use is most important for genetic counseling, as children who are homozygous for the gene have a 25% chance of siblings having hearing loss, whereas a negative or heterozygous result gives siblings a 14% chance of hearing loss.

CT Scan – Multiple abnormalities can be seen in the temporal bone, cochlea and semicircular canals. The Michel aplasia is a near complete agenesis of the petrous portion of the temporal bone likely secondary to an insult in the 3rd week of fetal development. These children are not candidates for cochlear implant. Mondini aplasia is seen when only the basal turn of the

cochlea develops, likely secondary to arrest of development in the 6th week of fetal development. It is associated with perilymphatic fistulas and recurrent meningitis. Controversy surrounds cochlear implantation in these children and amplification is usually recommended. Large vestibular aqueduct is described as any vestibular aqueduct greater than 1.5 mm in diameter. It has been associated with fluctuating and progressive SNHL, typically bilateral. It is also associated with vertigo, unsteady gait and Pendred syndrome. Children with large vestibular aqueduct syndrome are at risk for progressive hearing loss after minor head trauma.

MRI – Many of the above abnormalities can be seen with the MRI. MRI is not warranted to screen for acoustic neuroma as this diagnosis is exceedingly rare in the pediatric population. The exception to this is in the child with neurofibromatosis type II.

As one would expect, performing all of the above tests on every child presenting to the clinic would produce large costs for both the patient and third party payers. The question for the clinician is when to order which tests. In a study by Mafong et al, a retrospective analysis of 114 children with SNHL was performed to evaluate the effectiveness of various testing. ANA, RF, ESR, CBC, Platelet count, BUN, Creatinine, Urinalysis, Glucose, FT4, TSH, T3, FTA-ABS, RPR, EKG, and CT scan results were reviewed in 114 patients. None of the blood tests led to the diagnosis of the etiology of hearing loss. Although ESR, and ANA were abnormal in several patients, rheumatology evaluation elicited no diagnosis. CBC showed mild anemia in some children. This was not treated or thought to be the cause of hearing loss. An EKG was found to be positive in 1 of 15 leading to the diagnosis of Jervall and Lange-Nielsen syndrome. CT was performed in 76 patients, CT and MRI in 14 patients, and 7 had MRI alone. Overall, 39% of cases showed some abnormality. Large vestibular aqueduct was seen in 13% of imaging studies. Cochlear dysplasia was seen in 7% of cases. MRI findings distinct from CT were seen in 4 patients, 1 of which may have been related to the hearing loss (fistulous connection from IAC to the temporal bone). The authors concluded that routine laboratory evaluation should be reconsidered given its low diagnostic yield. They also supported routine EKG and CT scan.

Based on the above information the following guide to evaluation of the child with SNHL can be used:

If the history reveals an etiology that needs no further treatment (meningitis, congenital viral infection, previous trauma with documented temporal bone involvement, prolonged exposure to ototoxic drugs, cerebral palsy or other anoxic brain injury, or history of prolonged severe hyperbilirubinemia, no further testing is necessary. If no etiology can be detected, the author recommends the following.

Due to the chance of sudden childhood death that could otherwise be avoided by a simple and inexpensive test, EKG should likely be performed in all children with SNHL. Although congenital syphilis will usually manifest other of the classical stigmata such as interstitial keratitis, Hutchinson's teeth (notched incisors), mulberry molars, bilateral painless knee effusions, nasal septal perforations, or saddle nose deformity, SNHL can be the only symptom. As such, and due to the fact that syphilis is a treatable and potentially fatal disease, an RPR is warranted in workup of this disease. TPPA should be reserved for confirmation. CT scan of the temporal bone is warranted due to the high incidence of abnormalities. In addition, if a large vestibular aqueduct is discovered, patient counseling can be used to prevent future hearing loss

from minor trauma. This information can also be used in preoperative evaluation for cochlear implantation if hearing loss is bilateral. GJB2 screening is useful for genetic counseling. Parents should be counseled that this test will only be useful they would avoid children depending on future likelihood of a deaf child. As counseling is best done by a geneticist, referral is suggested. Finally, an audiogram at 6 months and 1 year should be scheduled with a follow-up clinic visit. If the child develops progressive hearing loss in one ear or gait or balance disturbances, MRI is warranted. In addition to the above tests the following may need to be ordered, but should not be ordered routinely. The following history and physical findings will help focus the workup.

- fever or illness for more than 3 weeks, gingival bleeding bone or joint pain, signs of autoimmune disease (Order CBC with Diff for leukemia/lymphoma, ANA, ESR, RF for JRA or lupus/other autoimmune)
- family history of progressive early onset hearing loss in father/mother and/or grandfather/mother (Urinalysis for Alports, genetics consult for syndrome evaluation)
- family history of progressive vision loss (ophthalmology consult for Usher syndrome or other auditory visual syndromes)
- history of hematuria or family history of kidney failure (urinalysis, BUN, creatinine for Alport's syndrome)
- thyroid goiter, signs of hypothyroidism,, Mondini malformation or large vestibular aqueduct by CT scan (Thyroid function tests and perchlorate test for Pendred syndrome)
- history of frequent urination, excessive thirst (Glucose for Alstron syndrome).
- history of progressive hearing loss, gait or vestibular symptoms, focal neurological symptoms (MRI of brain and IAC's)
- family history of neurofibromatosis or history of neurofibroma, meningioma, glioma, schwannoma, or juvenile posterior subcapsular lenticular opacity (MRI of brain and IAC's)

This system allows for a blend of cost effective analysis and comprehensive screening.

Diagnosis of Acoustic Neuroma

Another unique problem for the otolaryngologist is the adult patient with unilateral inner ear symptoms. The incidence of acoustic neuroma in the general population is 12 per million per year. MRI today can detect tumors as small as 3mm. The gold standard for diagnosis of CP angle pathology is now MRI of the IAC's with gadolinium. The challenge to the otolaryngologist is in the decision of which patients need an MRI. An alternative to diagnosis of the acoustic neuroma is the ABR. ABR's are used as a screen for acoustic neuromas by some clinicians. The main advantage of ABR is decreased costs. MRI is generally 5 times as expensive as ABR. The disadvantage of ABR is sensitivity. Several studies have been done on the sensitivity of ABR. Wilson et al in 1992 found sensitivity of ABR to be 85%, but only 67% in tumors of the small category. Chandrasekhar et al in 1995 found an overall sensitivity of 92%, 83% for small tumors. Gordon and Cohen in 1995 found an overall sensitivity of 88%, 69% for small tumors. All of the above authors recommended MRI for patients with histories highly suggestive of acoustic neuroma (Unilateral decrease in word recognition, asymmetric SNHL) and ABR for patients with low suspicion of acoustic neuroma (presbycusis and slight

asymmetric SNHL, vertigo of short duration). Ruckershern et al in 1996 did a prospective study of ABR and MRI in detection of acoustic neuroma. 47 patients were identified over 3 years. ABR had a sensitivity of 63% and a positive predictive values of 26%.

The question then becomes, which patients need an MRI and which patients can have screening with ABR. A study was done by Robinette and Bauch et al which tried to answer the question, how much more expensive is MRI? They did a retrospective review and identified 95 patients with acoustic neuromas. They were divided into 3 groups based on the following factors: High risk – all of the following: asymmetric hearing loss, tinnitus, and greater than 30% decrease of word recognition in the affected side, Intermediate risk – sudden sensorineural hearing loss or unexplained persistent tinnitus, Low risk – isolated vertigo or historically explained intermittent tinnitus or historically explained unilateral hearing loss. Using the probability of acoustic neuroma in each group (30%, 5%, 1%), the sensitivity of ABR in detecting tumors in based on size of the tumor (100%, 93%, 82%), and a positive predictive value for ABR (12%), they calculated the cost difference for screening with ABR and MRI for positive results verses MRI initially. In addition, they estimated the number of tumors that would be missed if ABR alone was used. In the high risk group, they estimated that ABR would save \$40,000 with no missed tumors. In the intermediate group, they estimated a savings of \$900,000, but the clinician would miss 4 out of 900 patients screened with ABR. In the low risk group, \$1.7 million dollars would be saved at the cost of missing 1 out of 1600 patients screened. They recommended clinical decision making with consideration of the considerable savings in the intermediate and low risk groups.

With the above figures in mind, the clinician can make recommendations based on historical and audiographic information. All patients with unilateral SNHL of unknown etiology greater than 20 DB, word discrimination decrease of greater than 30% of the asymptomatic side, or unilateral persistent tinnitus with vertiginous symptoms should be screened with MRI as it is most sensitive, and savings with ABR screening is minimal in this group. 0.6 to 2% of cases of sudden unilateral SNHL screened with MRI with reveal acoustic neuroma. In addition, 10% of patients with acoustic neuroma report a history of sudden sensorineural hearing loss. For these reasons, idiopathic unilateral SNHL should be screened with MRI. For all other patients (historically explained unilateral tinnitus or hearing loss or isolated vertigo) ABR as a screening tool is a reasonable alternative. Discussion with patients over cost verses risk is warranted in these cases. In addition, patients with limited funds may ask for alternative testing possibilities. ABR seems to be a reasonable alternative for patients with financial issues that do not fall in the high risk category. Patients in the high risk group will have minimal savings that do not justify the decrease in sensitivity.

Sudden sensorineural hearing loss

Sudden sensorineural hearing loss creates a diagnostic dilemma for the clinician. Although uncommon, an otolaryngologist may see several patients per year, depending on the referral base of his practice. The incidence has been estimated between 5 and 20 per 100,000. It is most common between the ages of 40 and 54, and female to male incidences are equal. It is defined as a loss of at least 30 dB in 3 contiguous frequencies over a time course of 72 hours or fewer. The natural history of SSNHL favors resolution of symptoms in a significant number of cases. Approximately 65% of patients diagnosed with SSNHL recover within 20 dB of the

affected ear or greater than 50% of the initial hearing loss, even without treatment. The challenge in studying treatment for this disease centers on differentiating therapeutic response from spontaneous resolution. In addition, several tests have been proposed for acute evaluation of SSNHL, most based on prevalent theories and current disease models. Several of the more popular theories are discussed below.

Vascular compromise – The cochlea derives its blood supply from the Labyrinthine artery. It is a unique structure in the head and neck because of the lack of collateral blood flow. This leads to the possibility of cochlear ischemia through a variety of mechanisms. Some of the more popular theories are arterial thrombosis, emboli, vasospasm, or hyperviscosity syndromes.

Intracochlear membrane rupture or perilymph fistula – The cochlea relies on the resting potential between the endolymph and perilymph to produce action potentials that allow transformation of mechanical energy to electrical energy. Intracochlear membrane rupture was proposed in the 1960's as a possible cause of SSNHL. In addition, perilymph fistulas can cause fluctuating hearing loss and vertigo, and spontaneous fistula remains in the differential diagnosis.

Viral infection of the labyrinth – Viral seroconversion of patients with SSNHL is seen higher than in controls. In addition, histology studies have shown changes similar to viral infections in cadaver studies. For this reason, Herpes infection of the labyrinth remains a popular theory.

Autoimmune inner ear disease – several authors have shown that patients with SNHL have antibodies to specific inner ear proteins. Autoimmune diseases such as Cogan's syndrome and SLE, among others, have been associated with SNHL. Response to steroids has strengthened this theory.

Diagnostic testing

An audiogram should be performed on all patients with sudden hearing loss. Serial testing provides documentation of the progression or resolution of the hearing loss and response to treatment. Other tests commonly ordered by otolaryngologist are listed below:

- 1) CBC – Screening for leukemia/lymphoma
- 2) ESR, ANA, RF – Screening for autoimmune disorders (Cogan's, Wegner's, polyarteritis nodosa, temporal arteritis, Burger's, SLE)
- 3) Glucose
- 4) T3, T4, TSH – Hypothyroidism
- 5) PT, PTT – Hemorrhagic inner ear disorders
- 6) RPR, TTPA- syphilis
- 7) HIV
- 8) Lymes titer
- 9) Cholesterol/Triglycerides – vascular thrombosis causing inner ear ischemia
- 10) Anti-hsp 70 or 68 KD heat shock protein (autoimmune inner ear disease)
- 11) MRI – Screen for acoustic neuroma or central pathology

An analysis of the above test is warranted. As discussed above, complete history,

physical, and clinical judgment can limit the cost to the patient while still allowing accurate diagnosis.

Leukemia and lymphoma are rare causes of SNHL as discussed earlier. Unless gingival bleeding, bone pain, or lymphadenopathy is present, CBC is not warranted. Autoimmune disease probably plays an important role in SNHL. The chances of systemic autoimmune disease presenting solely as SNHL is rare. Literature supporting an inner ear autoimmune disease is growing. As the treatment of choice is steroids, testing for an autoimmune component would be higher yield after initial response to a short course of steroids and subsequent failure with taper of the steroids. Even in this situation, ESR, ANA, and RF are likely low yield. Cold intolerance, weight gain, and decreased energy would likely be present if hypothyroidism was the cause of hearing loss. As such, routine screening is considered low yield. Hemorrhage as a cause of sudden SNHL is likely a rare event. Even rarer would be abnormalities in PT, PTT, and INR causing hemorrhage. A history of anticoagulant use, not including daily aspirin, may increase the yield of this test. Without this history or other signs of a bleeding disorder, this test would be low yield.

Syphilis has been shown to cause SNHL. As discussed above, syphilis is likely a very rare cause of sudden SNHL if not accompanied by other manifestations, but due to the potentially fatal outcome of the disease and simple treatment regiment, screening is warranted. A French study by Gagnebin et al performed in 2000 analyzed the utility of screening for HIV, Limes disease, and Syphilis. They tested 102 patients with Sudden SNHL. Two of the patients had positive Lymes titers, both of which were not responsive to treatment. One was found to have latent syphilis but no signs of neurosyphilis. All HIV tests were negative. The author concluded screening for infectious causes of sudden SNHL was low yield. The literature also reveals 3 cases of positive HIV in cases of sudden SNHL. Several studies have tested the hearing in HIV and shown decreased hearing in up to 30% of patients. In addition, treatment regiments for HIV are improving and life expectancy of infected individuals is increasing. Argument for or against routine screening could be made, but all patients considered high risk for HIV should be screened (IVDU, male homosexual, multiple sexual partners, history of previous venereal disease). No reports of sudden sensorineural hearing loss as a sole manifestation of Lymes disease could be found. Routine screening is considered low yield.

Much has been written about the 68kD heat shock protein and screening for antibodies to it (Anti-hsp 70) for diagnosis of autoimmune sudden SNHL. Samuelsson et al screened 27 patients and 100 healthy controls for Anti-hsp 70 antibodies and found an incidence of 19% and 14% respectively. Yeom et al in 2003 tested for the anti-hsp 70 antibody in 20 patients with rapidly progressive sensorineural hearing loss and 20 control patients without hearing loss. They found no significant difference in the incidence of heat shock protein antibodies and controls and questioned the use of this test in the diagnosis of AISNHL. Other studies have shown a higher incidence of steroid responsiveness in patients with positive tests. Much study is needed in this area to assess the utility of this test. Testing for the heat shock protein would be highest yield in patients responsive to steroids with remission after steroid taper.

Screening for cholesterol and triglycerides has gained recent popularity. Theory revolves around atherosclerosis and ischemic cochlear disease as an etiology. Friedrich et al studied 49 patients with neuro-otologic symptoms and tested serum cholesterol and triglycerides. They

found increased LDL and a higher LDL/HDL ratio as compared to controls. Nuti et al studied total cholesterol and LDL/HDL ratio in their group of patients and found no significant difference from controls. Ullrich et al tested lipids and triglycerides in 24 patients with sudden SNHL and found no difference from controls. Kojima et al studied 12 subjects with sudden SNHL in whom the event was at least 1 month prior to entering the study. Patients with total cholesterol levels greater than 230 mg/dL or greater were treated with diet and administration of lipid lowering drugs. Significant improvement in hearing in the 125 – 2000 Hz range was found in the treatment group. The American collage of physicians guidelines for 2004 recommends screening for lipid abnormalities in men aged 35 to 65 years and women aged 45 to 65 years and using only a total cholesterol level. As such, total cholesterol is warranted in patients presenting sudden SNHL in the above age range. Testing out of this age range is considered low yield.

Acoustic neuroma has long been associated with sudden SNHL. Patients with acoustic neuroma present with sudden SNHL 10% of the time. Numbers as high as 2.5% of all patients with sudden SNHL have acoustic neuroma. Aronzon et al treated patients with sudden SNHL and MRI proven acoustic neuroma with high dose of steroids. He found improvement in hearing in all patients. The above numbers warrant screening with MRI for all patients with acoustic neuroma. MRI has the additional benefit of screening for most central pathology.

With the above arguments in mind the author recommends the following in patients presenting with sudden SNHL:

- All patients screened with RPR. Positive results confirmed with TTPA.
- MRI in all patients presenting with SSNHL, regardless of response to steroids.
- Total cholesterol in men aged 35 to 65 years and women aged 45 to 65 years if no cholesterol testing in the last year.
- HIV in all patients with high risk of STD's, +/- for screening in all patients.
- ESR, ANA, RF in patients with response to steroids and relapse after steroid taper.
- CBC, Thyroid function tests, PT, PTT, Lymes titers based on history and physical exam findings only.
- 68 kD protein in research settings, +/- screening after response to steroids with relapse after steroid taper.

Conclusion

As medical science evolves and the number of diagnostic tests expands, the physician will be asked more and more to evaluate the medical necessity and cost benefit of diagnostic testing. Research projects based on cost analysis will become more important as the focus of medicine shifts towards cost containment. A balance between cost containment and diagnostic accuracy must be struck by the modern day physician to best serve the health of the community.

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