Unilateral Sensorineural Hearing Loss

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Introduction

• What dilemmas?
  – Increasing number and complexity of tests
  – Increasing costs of medical care
  – What do tests results mean?
  – At what point does cost of testing outweigh value of “rule out” diagnosis?
  – How far do we go to diagnose etiology?
Finical Pressures

- Increasing cost of medical care
- Third party payers pressure to contain cost
- Government pressure to contain costs
- Patient pressure to contain cost
  - Personal Medical savings accounts
  - Higher deductibles
  - Higher co-paying
  - Services not covered
Overview

Diagnosis of the following:

- Unilateral Childhood SNHL of unknown duration
- Sudden sensorineural hearing loss
- Unilateral inner ear complaints
Unilateral Childhood SNHL

• Unilateral SNHL in moderate to profound range correlates with:
  – Poor academic performance
  – Increased chance of repeating a grade
  – Behavioral problems

Intervention prior to 6 months results in advantages in communication
Unilateral Childhood SNHL

- Universal infant screening has led to earlier diagnosis
- Congenital hearing loss may be delayed
- Joint Committee on Infant Hearing guidelines for persistent periodic screening:
  - Child with family history of early onset SNHL
  - Prenatal infection that may lead to SNHL
  - Neurofibromatosis II
  - Persistent pulmonary hypertension
Epidemiology

- Average age of diagnosis unilateral SNHL is 8.7 yo.
- Bilateral SNHL greater than 50dB, 1-2/1000
- Unilateral SNHL greater than 50 dB, 3/1000
- Genetic cause in 50% of individuals
Unilateral Childhood SNHL

- Initial referral because of failed audiogram, parental concerns.
- Confirmed with audiogram, OAE, ABR
- In younger children, otolaryngologist must screen for conductive etiology
Unilateral Childhood SNHL

- Differential diagnosis
  - Syndrome with hearing loss
  - Other genetic cause
  - Meningitis
  - Intrauterine infection
  - Trauma to cochlea, vestibule, VIII
  - Exposure to ototoxic drug
  - Prematurity
  - Autoimmune disease
Unilateral Childhood SNHL

- Differential Diagnosis
  - Hyperbilirubinemia
  - Neurofibromatosis type II
  - Anoxic Brain injury
  - Mumps
  - Neurodegenerative disorder
  - Malignant infiltration
  - Ischemic insult of cochlea
  - Cochlear hydrops
Unilateral Childhood SNHL

• Genetic basis in 50%
  – Over 200 syndromes include deafness or hearing impairment
  – Identification of syndrome allows efficient screening for other anomalies
Unilateral Childhood SNHL

- Usher syndrome
  - Autosomal recessive disorder
  - Retinitis pigmentosa and SNHL
  - Early intervention for future visual and auditory impairment
  - Bilateral cochlear implant should be considered
Unilateral Childhood SNHL

- Waardenburg syndrome
  - SNHL
  - White forelock
  - Multicolored iries
  - Dystropia canthorum
  - Hypertelorism
Unilateral Childhood SNHL

- Jervell and Lange-Nielsen
  - Autosomal dominant
  - Seen in 1.6 to 6 per million
  - Prolonged QT interval and increased risk of sudden death
  - First degree relatives need screening with EKG
Unilateral Childhood SNHL

• Neurofibromatosis Type II
  – Autosomal dominant
  – Bilateral acoustic neuromas
  – Meningiomas
  – Spinal schwannomas
  – Posterior capsular lens opacities
Unilateral Childhood SNHL

- Alports syndrome
  - Autosomal dominant or recessive, or x-linked
  - Slowly progressive bilateral SNHL, can be asymmetrical
  - Progressive renal failure
  - Persistent microscopic hematuria
  - Episodic gross hematuria precipitated by URI
Unilateral Childhood SNHL

- Branchio-oto-renal syndrome
  - 1/40,000 live births
  - 60% with branchial cleft cysts
  - 20% with pure SNHL, majority mixed
  - Shared antigen between the stria vascularis and glomeruli
Unilateral Childhood SNHL

- Noonan’s syndrome
  - 1/2500 live births
  - Webbed neck
  - Pinna abnormalities
  - Short stature
  - 10-15% with SNHL
Unilateral Childhood SNHL

• CHARGE Syndrome
  – Coloboma
  – Heart Defects
  – Atresia choanae
  – Retarded growth
  – Genital hypoplasia
  – Ear abnormalities and deafness
  – Associated with Mondini malformation
Unilateral Childhood SNHL

• Fechner’s syndrome
  – High frequency SNHL
  – Proteinuria
  – Macrothrombocytopenia
  – Ocular disease
  – Extremely rare
Unilateral Childhood SNHL

• Pendred syndrome
  – Autosomal recessive
  – Congenital SNHL
  – Goiter
  – Hypothyroidism (varies)
  – Abnormal perchlorate discharge test
  – Associated with Mondini and Large Vestibular aqueduct
Unilateral Childhood SNHL

• Other causes
  – TORCHS (Toxoplasmosis, Others, Rubella, Cytomegalovirus, Herpes simplex, Syphilis)
  – Meningitis
  – Ototoxic drug exposure
  – Maternal drug alcohol use
  – Maternal use of teratogenic drug (thalidomide)
Diagnostic Testing

- CBC with Differential
- Platelet studies
- ANA, ESR, RF
- BUN, Creatinine, Urinalysis
- Serum glucose
- Thyroid function tests, Perchlorate test
- RPR, TTPA
- GJB2 (Connexin 26)
- EKG
- CT, MRI
Diagnostic Testing

- CBC with differential
  - Used to screen for leukemia or lymphoma
  - Hearing loss due to hyperviscosity or temporal bone infiltration
  - 1 case of leukemia with hearing loss as initial manifestation in literature
  - Low yield
Diagnostic Testing

- Platelet studies
  - Drawn to exclude Fechner syndrome
  - Patients with family history, ocular disease, history of diagnosed proteinuria should be screened
  - Test is low yield
Diagnostic Testing

- ANA, ESR, RF
  - Drawn to screen for autoimmune disorders, Lupus, Cogan's, juvenile rheumatoid arthritis
  - Test nonspecific
  - Positive RF is 0.7% sensitive in detecting juvenile RA
  - Testing without history of joint pain or other signs of systemic autoimmune disorders is low yield
Diagnostic Testing

• BUN, Creatinine, Urinalysis
  – Used to screen for concurrent kidney disease such as Alport’s disease
  – Should be drawn with history of gross hematuria, family history of kidney disease, family history of slowly progressive hearing loss
  – Routine screening is low yield
Diagnostic Testing

• Serum glucose
  – Used to screen for Alston syndrome (Impaired glucose tolerance, retinal degeneration, neurosensory deafness, acanthosis nigricans, hepatic dysfunction)
  – Only 50 cases reported since 1959
  – Very low yield
Diagnostic Testing

- Thyroid function testing
  - Drawn to screen for hypothyroidism and Pendred syndrome
  - Hearing loss as a sole symptom of hypothyroidism is very rare
  - Pendred syndrome not always associated with hypothyroidism. Perchlorate testing test of choice
  - Testing highest yield in children with goiter, signs of hypothyroidism, large vestibular aqueduct or Mondini malformation
Diagnostic Testing

- **RPR, TTPA**
  - Drawn to look for syphilis.
  - Hearing loss with tertiary syphilis usually associated with other manifestations of the disease, but can be sole manifestation
  - History of maternal syphilis exposure, signs of tertiary syphilis make testing higher yield
  - Testing low yield
Diagnostic Testing

- EKG
  - Used to screen for prolonged QT interval in Jervall and Lange-Nielson syndrome
  - Test highest yield with family history of childhood death or syncope
  - Low yield
Diagnostic Testing

• **GJB2 gene (Connexin 26)**
  - Responsible for as much as 50% of autosomal recessive nonsyndromic hearing loss
  - Use most important in genetic counseling for parents of hearing impaired children
  - Homozygous children have a 25% chance of having siblings with hearing loss. Negative or heterozygous results gives siblings 14% chance
CT and MRI

- Used to scan for inner ear malformations
- Mondini malformation associated with perilymphatic fistulas and recurrent meningitis
- Large vestibular aqueduct associated with hearing loss as a result of minor head trauma
- MRI used for screening for acoustic neuroma in children with neurofibromatosis type II
Mafong et al, Laryngoscope 2002

- Retrospective chart review 114 children with SNHL
- ANA, RF, ESR, CBC, Platelet studies, BUN, Creatinine, Urinalysis, Serum Glucose, FT4, TSH, T3, FTA-ABS, RPR, EKG, CT scans reviewed
Mafong et al, Laryngoscope 2002

- EKG positive in 1 of 15 patients tested.
- ESR and ANA were positive in 22% of patients. No correlation with clinical disease. Occasional nonspecific abnormalities in CBC.
- All other laboratory testing negative.
Mafong et al, Laryngoscope 2002

- CT scan
  - 39% with abnormality
  - Large vestibular aqueduct in 13%
  - Cochlear dysplasia in 7%
  - MRI added to diagnosis in 4, one of which related to hearing loss (fistulous connection from IAC to temporal bone)
Mafong et al, Laryngoscope 2002

- Authors concluded that routine laboratory evaluation should be reconsidered given its low diagnostic yield
- They supported routine use of EKG and CT scan.
Conclusions

• EKG – sudden childhood death
• CT – High incidence of abnormality
  – Large vestibular aqueduct – avoid contact sports
  – Used for preoperative information should bilateral SNHL develop
  – Mondini malformation lead to further testing
Conclusion

• Syphilis
  – Rare without classical stigmata (interstitial keratitis, Hutchinson’s teeth, mulberry molars, bilateral painless knee effusions, nasal septal perforation, saddle nose deformity)
  – Simple treatment, potentially fatal
  – Recommend testing with RPR, confirmation with TTPA
Conclusion

• **GJB2 (Connexin 26)**
  - Screening only if result will affect future childhood planning
  - Genetics consult warranted for counseling of results
Conclusion

- Fever or illness more than 3 weeks, gingival bleeding, bone or joint pain, signs of autoimmune disease, order CBC and ANA, ESR, RF

- Family history of progressive early onset hearing loss in first or second degree relative, order urinalysis, genetics consult
Conclusion

- Family history of progressive vision loss or visual complaints, order ophthalmology consult
- History of hematuria or family history of kidney failure, order urinalysis. If urinalysis positive order BUN, creatinine
- Thyroid goiter, signs of hypothyroidism, Mondini malformation, large vestibular aqueduct by CT scan, order thyroid function test, consider perchlorate test
Conclusion

- History of frequent urination, excessive thirst, order serum glucose
- History of progressive hearing loss, gait or vestibular symptoms, focal neurological symptoms, order MRI of brain and IAC’s
- History of neurofibroma, meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacity or family history of Neurofibromatosis type II, order MRI of brain and IAC’s.
Sudden Sensorineural Hearing Loss

- Incidence estimated between 5 and 20 per 100,000
- Most common between ages of 40 and 54
- Loss of at least 30dB in 3 contiguous frequencies in 72 hours or less
- 65% diagnosed will spontaneously recover within 20dB or greater than 50% of total loss
Sudden Sensorineural Hearing Loss

- **Etiology**
  - Vascular compromise from hyperviscosity, embolic event, vasospasm.
  - Intracochlear membrane rupture or perilymph fistulas
  - Viral infection
  - Autoimmune inner ear disease, systemic autoimmune disease (Cogan’s, Wegner’s, polyarteritis nodosa, temporal arteritis, Berger's, SLE)
Sudden Sensorineural Hearing Loss

• Diagnostic testing
  – CBC
  – ESR, ANA, RF
  – Serum Glucose
  – T3, T4, TSH
  – PT, PTT
  – RPR, TTPA
Sudden Sensorineural Hearing Loss

• Diagnostic testing
  – HIV
  – Lyme titer
  – Cholesterol/Triglycerides
  – Anti-hsp 70 (68KD heat shock protein)
  – MRI
Sudden Sensorineural Hearing Loss

- CBC
- ESR, ANA, RF
- Serum Glucose
- Thyroid function studies
- RPR, TTPA
Sudden Sensorineural Hearing Loss

- **PT, PTT**
  - Used to look for hemorrhagic etiology of SSNHL
  - Patients likely to have other manifestations of coagulopathy
  - Higher yield if patient currently on an anticoagulant
Sudden Sensorineural Hearing Loss

- HIV
  - Shown to decrease hearing in up to 30% of patients with active infection compared to controls.
  - Literature shows 3 cases of SSNHL with positive HIV
  - Low yield
  - HIV treatment early improves life expectancy
Sudden Sensorineural Hearing Loss

- Lyme titer
  - No literature case reports of Lyme disease as an etiology
  - Recent history of tick exposure
  - Low yield
Sudden Sensorineural Hearing Loss

- Gagnebin 2000 (French study)
  - Analyzed screening HIV, Lyme, syphilis
  - 102 charts reviewed
  - Two patients with positive Lyme titers, not responding to treatment
  - All HIV negative
  - Latent syphilis without signs of neurosyphilis in one patient
  - Concluded screening is low yield without history
Sudden Sensorineural Hearing Loss

- Anti-hsp70 (68KD heat shock protein)
  - Thought to be a marker of autoimmune inner ear etiology
  - Early studies have shown steroid responsiveness in patients with positive tests
  - Samuelsson screened 27 with SSNHL and 100 controls. 19% and 14% respectively (not significant)
  - Yeom in 2003 tested for anti-hsp 70 in 20 patients with rapidly progressive SNHL and 20 controls. No significant difference
Sudden Sensorineural Hearing Loss

- Anti-hsp70 (68KD heat shock protein)
  - Testing is high yield
  - Question as to validity of results
  - Question of utility of positive results
  (most patients treated with steroids)
Sudden Sensorineural Hearing Loss

- Cholesterol and Triglycerides
  - Labyrinthine artery atherosclerosis as an etiology
  - Friedrich – 49 patients with neurootologic symptoms. Increased LDL and LDL/HDL ratio as compared to controls
  - Nuti – No significant difference in cholesterol and LDL/HDL ratio with controls
Sudden Sensorineural Hearing Loss

- **Cholesterol and Triglycerides**
  - Ullrich – tested lipids and triglycerides in 24 patients with SSNHL. No significant difference from controls
  - Kojima – 12 patients with SSNHL
    - Event was at least 1 month prior study
    - Patients with total cholesterol greater than 230mg/dL treated with diet and medications
    - Significant improvement in 125-2000 Hz
Sudden Sensorineural Hearing Loss

• Cholesterol and Triglycerides
  – American College of Physicians recommends screening for lipid abnormalities in men 35-65 and women 45-65 years of age using total cholesterol level only.
  – Cholesterol or triglyceride levels as an etiology of SSNHL likely low probability
  – Testing in this age range is high yield
Sudden Sensorineural Hearing Loss

- MRI
  - Used to screen for acoustic neuroma
  - Patients with acoustic neuroma present with SSNHL 10% of the time
  - As high as 2.5% of all patients with SSNHL have an acoustic neuroma
Sudden Sensorineural Hearing Loss

• MRI
  – Able to screen for acoustic neuroma and other causes of hearing loss
  – Aronzon – treated patients with SSNHL and MRI proven acoustic neuroma with high dose of steroids
    • Improvement in hearing of all patients
    • Response to steroids does not exclude acoustic neuroma
Conclusions

• All patients should be screened with RPR, confirmed with TTPA (treatable, life threatening, low cost)

• MRI in all patients, regardless of response to steroids

• Total cholesterol in men aged 35-65, women aged 45-65 if no testing in the last year
Conclusions

• HIV testing in all patients with high risk of STD, +/- in all patients

• ESR, ANA, RF in patients with history and physical findings of autoimmune etiology, response to steroid, but relapse after taper
Sudden Sensorineural Hearing Loss

- CBC, Thyroid function tests, PT, PTT, Lyme titers based on history or physical exam findings only
- 68 KD protein in research settings, +/- in response to steroids with relapse after steroid taper
Acoustic Neuroma

- 12 per million per year
- MRI can detect tumors as small as 3mm
- Gold standard for diagnosis is MRI of IAC’s with gadolinium
- Test cost at this institution is $3200
Acoustic Neuroma

- **ABR**
  - Alternative to MRI for diagnosis
  - MRI generally 5 times more expensive than ABR
  - Decreased sensitivity
  - Cost at this institution is $500
Acoustic Neuroma

• Sensitivity of ABR
  – Chandrasekhar 1995 – sensitivity of 92%, 83% for small tumors
  – Gordon 1995 – sensitivity 88%, 69% for small tumors
  – Ruckershern 1996 – sensitivity of 63%, PPV 26%
Acoustic Neuroma

• Robinette and Bauch
  – Retrospective review to identify 95 patients with acoustic neuromas
  – Divided into 3 groups
    • High risk – asymmetric hearing loss of greater than 20 DB over three frequencies, greater than 30% decrease of word recognition
    • Intermediate risk – SSNHL or unexplained persistent unilateral tinnitus
    • Low risk – isolated vertigo or historically explained intermittent tinnitus or historically explained SNHL
Acoustic Neuroma

- Used probability of acoustic in each group as (30%, 5%, 1%)
- Sensitivity of detecting tumors with ABR based on size (100% large, 93% medium, 82% small)
- Used PPV of 12% in ABR
Acoustic Neuroma

• Calculated cost of MRI for patients in each group
• Calculated cost of ABR, and MRI for all ABR’s suggestive of acoustic neuroma
• Calculated cost difference of two groups based on risk from history and physical
• Calculated number acoustic neuromas missed if first screened with ABR
Acoustic Neuroma

- High risk - $40,000, no missed tumors
- Intermediate risk - $900,000, clinician would miss 4 tumors in 900 patients screened
- Low risk – $1.7 million, 1 tumor missed out of 1600 screened
- Authors recommended clinical decision making with consideration of cost savings in intermediate to low risk groups
Conclusion

• MRI in:
  - All patients with unilateral SNHL greater than 20dB difference from unaffected side not explained by history
  - Word discrimination difference of 30% or greater from asymptomatic side
Conclusion

• MRI
  – Unilateral persistent tinnitus not explained by history
  – Persistent vertigo
  – Unilateral sudden SNHL, regardless of response to steroids
Conclusions

• ABR vs. MRI
  – Historically explained unilateral tinnitus
  – Historically explained hearing loss
  – Isolated vertigo
Summary

- Number of diagnostic tests expanding
- Physicians asked more and more to evaluate medical necessity and cost benefit of diagnostic testing
- Research projects based on cost analysis
- Balance of cost containment and diagnostic accuracy