Sudden Hearing Loss: Audiological Diagnosis and Management



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Emphasis areas of this presentation:

I. Diagnosis:
   a) types/classes **b)** etiologies
II. Audiological Monitoring
III. Prognostic Parameters
IV. Treatment
V. Post treatment Management:
   a) hearing loss b) tinnitus
VI. Education of Medical Community:
   a) diagnosis b) treatment

Sudden Hearing Loss Symptoms

- Patients may have accompanying:
  - -tinnitus
  - -aural fullness
  - -vertigo
  - -nausea/vomiting

Some Facts about SSNHL

- A rapid loss of hearing
- SSHL can happen to a person all at once or over a period of up to 3 days
- A medical emergency. Occurs within a few hours to three days. 1/3 of patients awaken with HL
- A loss of at least 30 decibels in three sequential frequencies

Natural History

- Spontaneous recovery rate is favorable: 40 to 70%, primarily occurring within 2 weeks post onset
- Spontaneous recovery has been reported in 25% in some other studies
- Recovery rate increases to 50% with early diagnosis and treatment
- Recovery dependent on configuration (upward slope better than flat or downward) and severity of HL (recovery decreases as severity increases)

(Hain, 2006/*ENT Today,* April 2006)

Some Facts about SSHL From NIDCD

- Hearing loss affects only one ear in 9 out of 10 people
- People experience dizziness or tinnitus, or both
- Approximately 4,000 new cases reported each year in the United States
- Happens most often to people between the ages of 30 and 60
- Equal occurrence by gender

Other Criteria

- Usually unilateral. HL mild to severe
- Temporary or permanent
- Represents 1% of all SNHL cases

(Wazen & Ghossani, 2005)

Underlying Factors

- Root causes can be idiopathic or by direct effect
- More than 100 possible causes
- It is rare for a specific cause to be precisely identified
- Only 10 to 15 percent of patients know what caused their loss

Diagnosis
Etiologies

- Infectious diseases
- Trauma/head injury
- Abnormal tissue growth/tumors
- Immunologic diseases such as Cogan's syndrome
- Toxic causes (e.g., snake bites)
- Ototoxicity
- Vascular
- Neurologic

Etiologies



- IDIOPATHIC
- VIRAL complications account for 60% of cases, including the atrophy of the Organ of Corti, shown to be the most common manifestation of viral pathology.

(Koc & Sanisoglu, 2003) & (Schattner et al., 2003)

http://medinfo.ufl.edu/year1/histo/images/h6f.jpg

Etiologies Cont.



- BACTERIAL - cryptococcal
- NEOPLASMS – myeloma (occasionally bilateral), acoustic neuroma
- Multiple Sclerosis
- VASCULAR - cardiopulmonary bypass, sickle cell, vertebrobasilar insufficiency

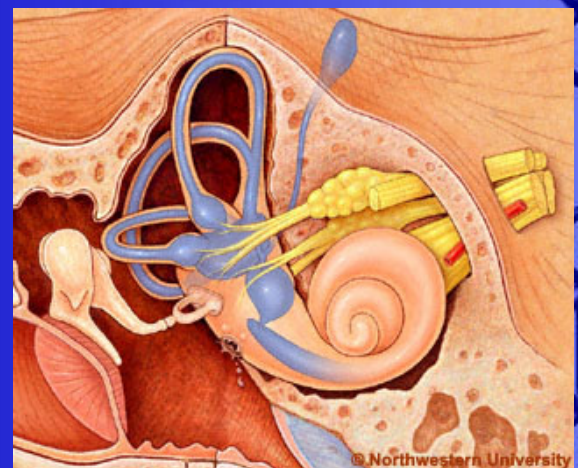


(Plasse et al, 1980) & (Hain, 2006)

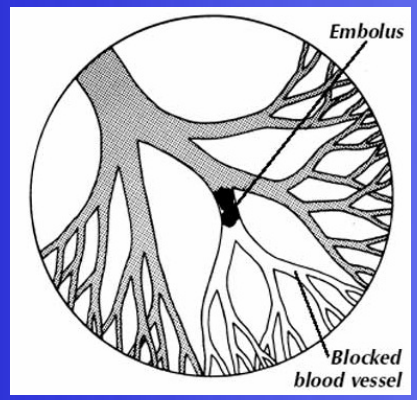
Etiologies

- Vascular occlusion and inner ear membrane rupture also believed to be factors
  - -perilymphatic fistula
  - -cochlear membrane break
  - -barotrauma
  - -Inner ear decompression sickness (Bends)
  - -Large vestibular aqueduct
- Auto-immune: AIED
- Meniere's Disease

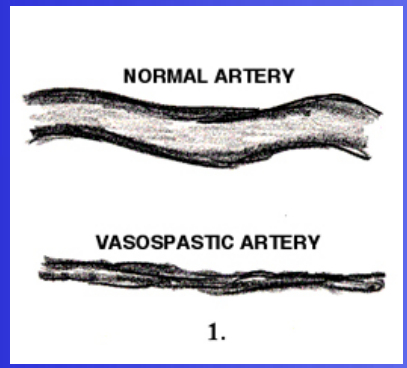
(Hain, 2006) & (Kiris et al, 2003)



Other Causes



- Renal failure and hemodialysis
- Vasospasm, embolism
- Congenital



(Lasisi et al, 2006) & (Hain, 2006)

* Psychogenic Sudden Deafness also has been reported in the literature

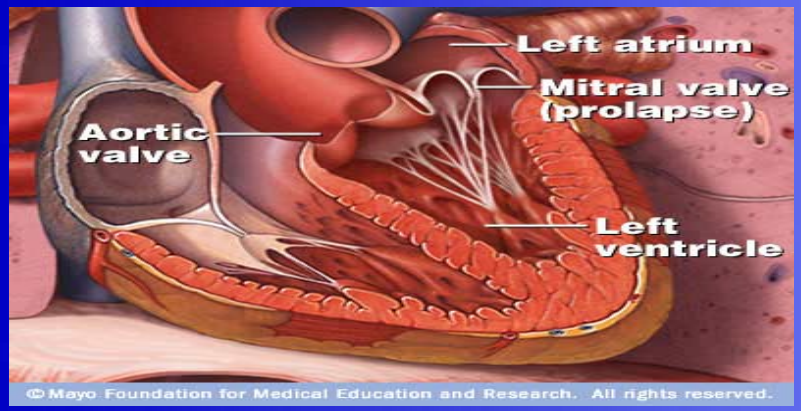
Ban & Jin (2006)

Some common causes of sudden hearing loss seen in ENT offices

(Personal communication, Mark Widick, M.D.)

1. Vascular - embolic or kinking of the arterial supply
2. Autoimmune - often not associated with an immune syndrome
3. Perilymphatic fistula - weight training, sneezing, slap injury, penetrating injury into middle ear, airbag deployment, blast...
4. Bubble emboli - Scuba Diving, sudden decompression
5. Viral syndrome - especially reactivation of herpetic virus.
6. Acute bacterial infection
7. Meningitis
8. Complications of cholesteatomas
9. Temporal Bone Fracture
10. Ototoxic antibiotics, Diuretics and Narcotic abuse

Mitral Valve Prolapse and Sudden Hearing Loss



- 349 subjects,
- 86 with ISSNHL and 263 controls
- underwent a 2D-echocardiography.
- Patients with ISSNHL had higher rates of MVP (29.1% vs 2.7%, $p < 0.001$), mitral leaflet thickening (15.1% vs 2.3%, $p < 0.001$), mitral regurgitation (16.3% vs 6.5%, $p = 0.02$) and left atrial enlargement (11.6% vs 3.8%, $p = 0.01$).
- results support the hypothesis that MVP could be one of the etiological factors of ISSNHL.

(Vazquez et al., 2007)

Rare Cases of Sudden Hearing Loss

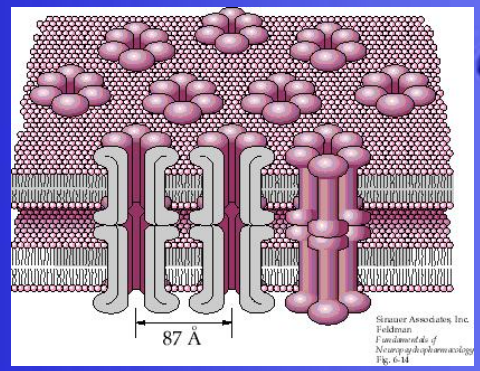
- Bilateral profound SSHL due to Mumps (Unal et al., 1998)
- Focal brain stem infarction has been uncommonly associated with unilateral sensorineural hearing loss (SNHL)

Hearing Loss After Liver Transplantation

- From 521 transplanted patients 16 patients developed severe hearing loss after liver transplantation. Four patients (25%) reported a history of **sudden deafness**
- Hearing loss was attributed to the ototoxic effects of liver transplant medications

Rifai et al. (2005)

Sudden Hearing loss due to Genetic Factors?

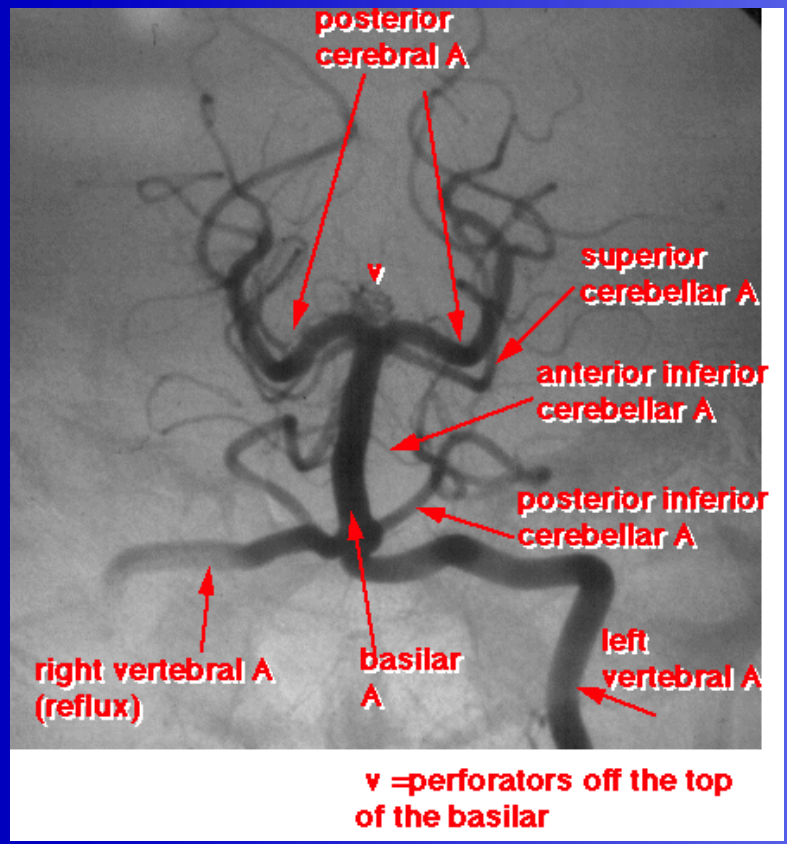


- Hearing loss associated with mutations of the *GJB2*, the gene encoding Connexin 26 (Cx26)
- May result in postnatal ***sudden*** and severe deterioration of the hearing capacity in cases with uncertain age at onset

Orzan & Murgia (2006)

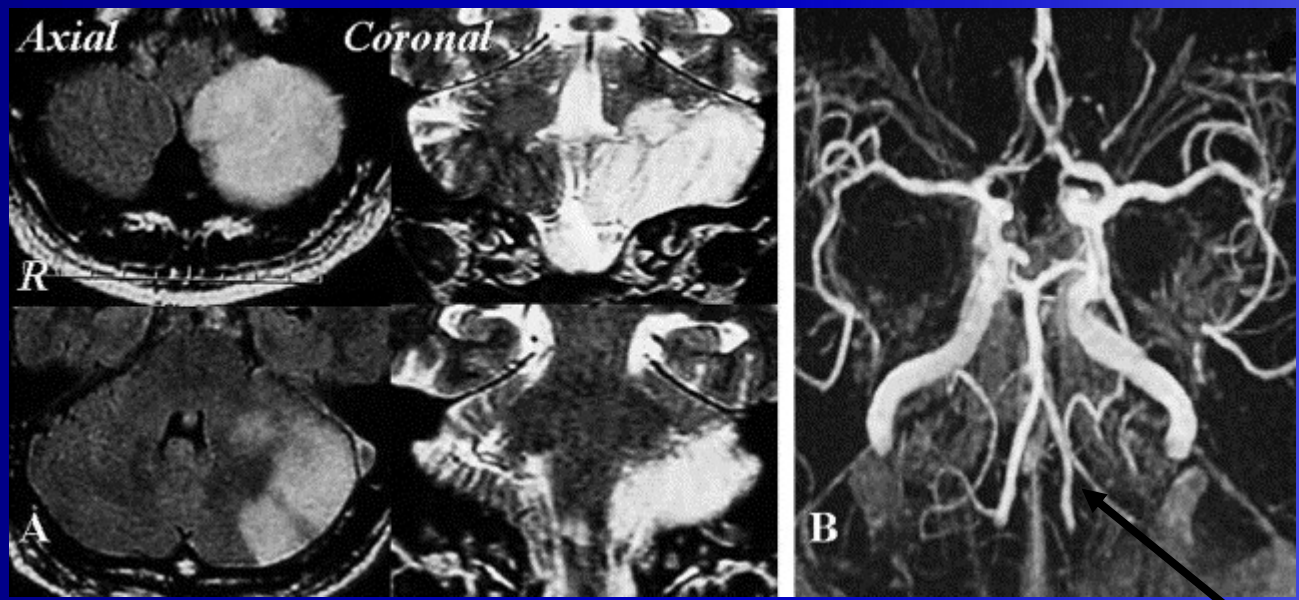
Sudden Hearing loss Vertebrobasilar ischemia (VBI)

- Vertebro-basilar system supplies most of the auditory system, including the inner ears



http://www.upstate.edu/neurosurgery/resources/med_students/angio/annapbas.GIF

Anterior and posterior inferior cerebellar artery infarction with **sudden deafness** and vertigo

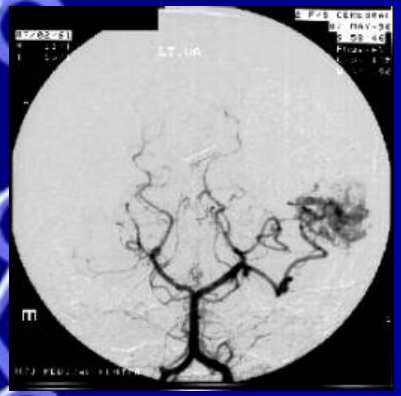


MRA

Left VA

Note narrower VA on the left. Patient had transient vertigo plus sudden deafness

Murakami et al. (2006)

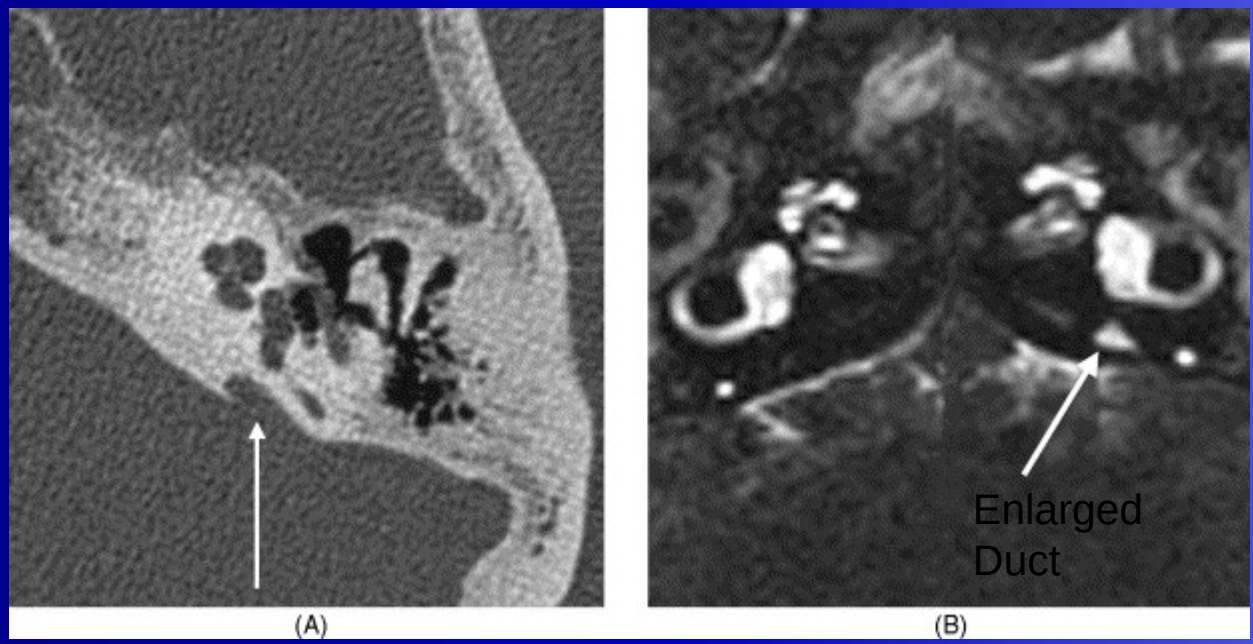


Vertebrobasilar Ischemia

- From 364 consecutive cases of VBI diagnosed by clinical features and brain MRI, 29 patients were identified as having **sudden deafness** as a symptom of VBI
- The inner ear is vulnerable to low blood flow within the vertebrobasilar system. Inner ear has a complete absence of collateral circulation. It also has a very high-energy metabolism which makes it vulnerable to vascular insults.

Lee and Baloh (2005)

Sudden Hearing Loss and large endolymphatic duct and sac syndrome (LEDS)



Koesling, Rasinski &Amaya (2005)

Enlarged endolymphatic duct and sac

Sudden sensorineural hearing loss as the initial manifestation of acute leukemia

- Hemorrhage into the cochlea can result in acute sensorineural hearing loss in leukemic patients



Harada, Namiki & Kawabata (2000)

Bilateral Sudden SNHL due to cancer

- **Cases have been reported following metastasis of melanoma. This patient has Leptomeningeal carcinomatosis with sudden onset bilateral sensorineural hearing loss. Leptomeninges is the combination of the two inner layers of meningese (**arachnoid and pia mater).

Jeffs, Lee and Wong (2006) AND Wagemakers et al. (2005)



Gadolinium-enhanced T1-weighted MRI images demonstrating abnormal thickening and enhancement of the vestibulocochlear nerves within the

internal acoustic meati bilaterally (arrows).

Jeffs, Lee and Wong (2006)

Diagnosis

- AUDIOMETRY: pure-tone, speech, and immittance. ABR (nerve fx), and OAE (cochlear fx)
- MIDDLE AND LATE EVOKED POTENTIALS: to monitor neural synchrony associated with tinnitus

(Wazen & Ghossani, 2003)

Diagnosis

- Tinnitus: pitch & loudness matching, MML, RI
- Head and Neck test. Romberg Tandem test
- ENG for those with dizziness or vertigo
- Rotary chair
- Imaging: MRI with contrast to diagnose CPA lesions. CT scans for select patients

(Hain, 2006), (Choudhury et al, 2005), & (Eggermont & Roberts, 2004)

Audiologic Management
DPOAE Evaluation

DPOAE as a Prognostic Factor for Idiopathic Sudden Deafness

- DPOAE was employed to predict treatment outcomes.
- DPOAE amplitude was a significant prognostic indicator.

Chao (2006)

Prognosis of SSHL

- The duration of symptoms before treatment and the severity of hearing loss may predict a recovery
- Presence of vertigo may worsen an outcome in SSHL. Vestibulopathy classification often a valid predictor of hearing recovery
- Patient age does not seem an important prognostic factor in SSHL

Pajor, Durko & Gryczyski (2003)
& Koc and Sanisgolu (2003)

Factors that affect prognosis of SSHL

- Age, ENG findings, flat/descending shapes, or total **deafness** in the initial audiogram, late identification of wave V during the follow-up, worse initial PTA thresholds, and delayed patients' presentation were significantly correlated to poorer hearing outcome.

Xenellis et al. (2004)

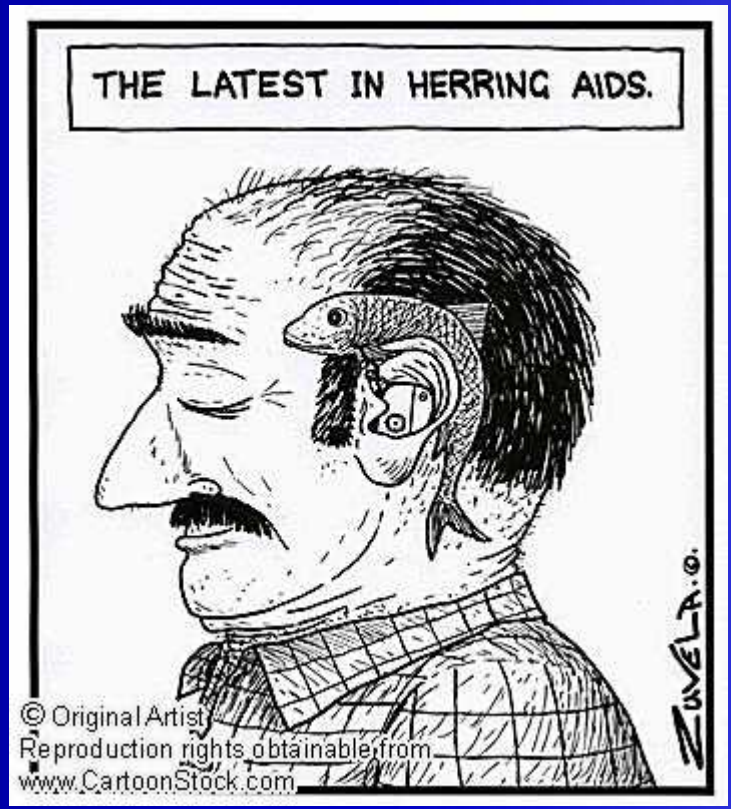
Recovery Parameters

- Comparison of PTA between first and final visits (Hearing Gain).
- Does not account for HL severity
- Ratio of hearing gain to first hearing level
- Recovery rate aka Rate of improvement (%). One formula can be:

  Rate of Improvement (%)= initial thresholds-final thresholds X100

(Penido et al, 2005) & (Koc & Sanisoglu, 2003)

Another Etiology



SSNHL Diagnosis

- Medical history: consideration of vascular, metabolic, and neurological events
- Hematologic examination
- Urinalysis
- Serologic testing
- Histopathologic testing
- Microbiologic testing

(Koc & Sanisoglu, 2003)



Diagnosis

Tests:

- FTA-Abs for syphilis
- ANA, Rheumatoid factor, ESR for autoimmune diseases
- Coagulation profile (INR, PTT, clotting time) for coagulopathy
- CBC and differential for infection
- TSH for thyroid disease
- Fasting blood glucose for diabetes
- Cholesterol, triglycerides for hyperlipidemia

Treatment



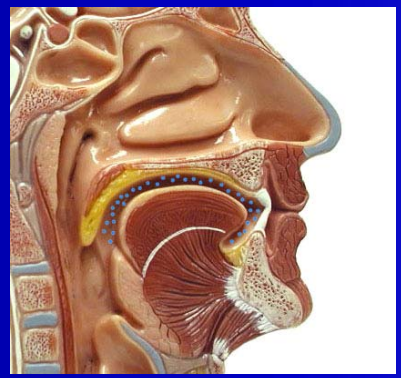
- The most common therapy for SSHL, especially in cases with an unknown (idiopathic) cause, is steroidal administration
- Corticosteroids usually work to reduce inflammation and decrease swelling
- Steroidal treatment helps some SSHL patients who also have conditions that affect the immune system

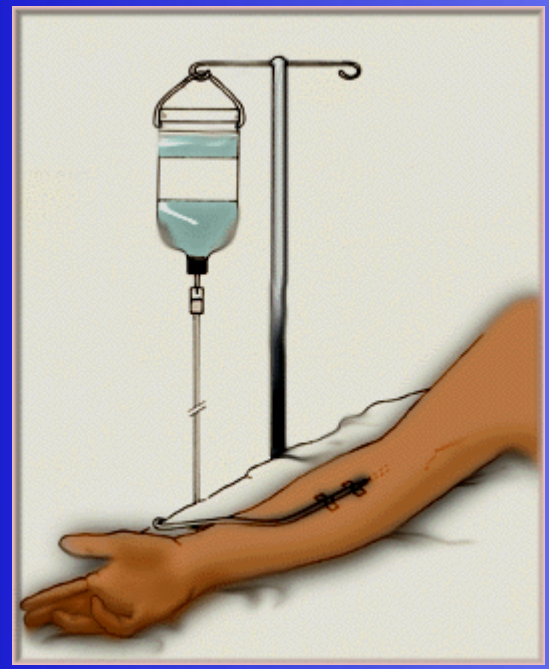
Corticosteroid Treatment

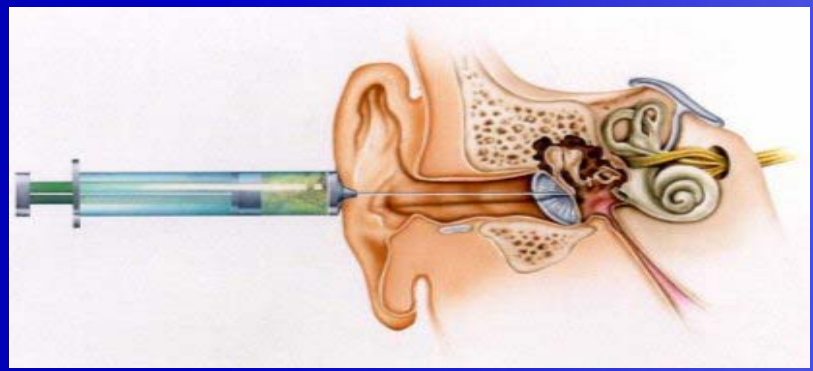
- Prednisolone shown to sufficiently increase potassium secretion of stria vascularis (be aware of differences between *prednisone* and *prednisolone*)
- Endocochlear potential restoration?
- Only class of drug with demonstrated efficacy in clinical trials

Routes of Administration

- P.O.
- I.V.
- Intratympanic or transtympanic







Oral Course

- Recommendation: Oral prednisone 1mg per kg per day for 1 month, with gradual taper per patient response
- Higher dose prednisone necessary per patient relapse, with a cytotoxic drug such as methotrexate (up to 15mg per week) , if relapse continues.
- Cytoxan added to regimen if previous therapy ineffective.

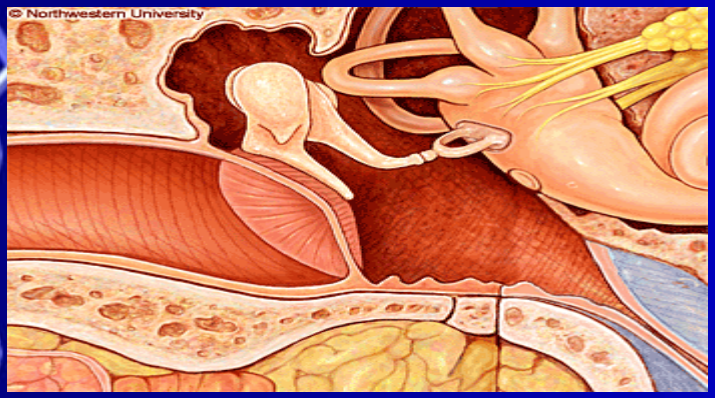
Intravenous

- Methylprednisolone, 1mg per kg per day (1-2 weeks)
- Pentoxifilline, 200mg per day (1-2 weeks)

Drug Therapy

- If patient shows no improvement after 10 days of systemic therapy other options are available!





Transtympanic

- Methylprednisolone, 40mg daily, perfused via round window through the TM.
- Local anaethesia concurrently administered
- Patient requested to keep head tilted contralateral to affected ear for 30 minutes
- In addition, an antibiotic can be prescribed (prophylaxis)

Transtympanic Route

Transtympanic administration:

-relatively easy

-systemic (oral, I.V. routes) side effects/toxicity not a factor

-higher concentration in perilymph

-does not cause functional or morphologic alterations in the ear

-methylprednisolone has best absorption profile

(Dallan et al, 2006)

Transtympanic vs. Oral Steroidal Therapy

- Currently, there are no published studies which compare these two approaches. There are some studies in progress funded as clinical trials by NIH, which will be available in 2009.

(ENT Today, 2006)

Side Effects of Steroidal Treatment

- Immune system compromised
- Anxiety
- Low tolerance
- In patients with diabetes/osteoporosis, complications may arise

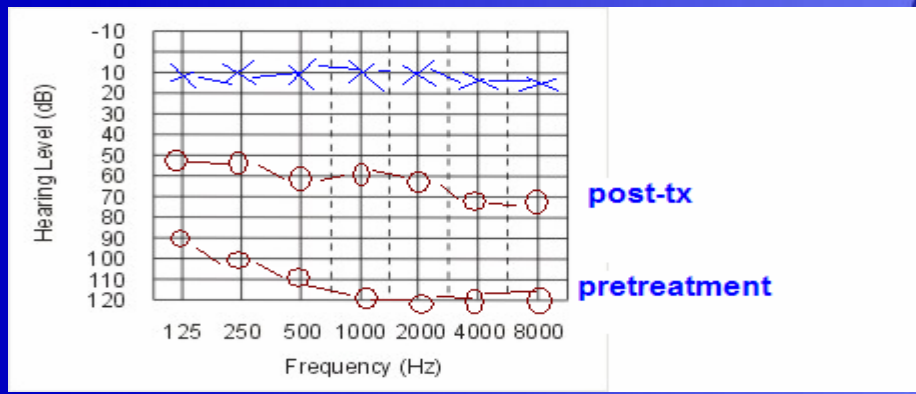
Alternate Drug Therapies

- Diuretics
- Vasodilators
- Antivirals
- Hemodilutors



A Case for Hydergine

- Hydergine (ergyloid mesylate) has also been prescribed for SSHL
- A patient with unilateral profound loss was able to recover to a moderate level after use of this medication. Spontaneous recovery cannot be ruled out, however.



Other Treatment options

Carbogen inhalation:

- Carbogen, a mixture of oxygen (95%) and carbon dioxide (5%), is thought to assist with air and blood flow inside the ear
- Shown to increase peripheral tissue oxygenation, with minimal CO2 retention or vasoconstriction

(Koc & Sanisoglu, 2003)

Other Treatment Options

- Gingko Biloba?
- Exploratory tympanotomy (to R/O perilymphatic Fistula)

(Kiris, et al, 2003) & (Hain, 2006)

“Shotgun”

- Multiple therapies: steroids, diuretics, antihistamines, and carbogen therapy employed simultaneously
- Significant differences not found in recovery compared to placebo

(Wilkins et al, 1987)

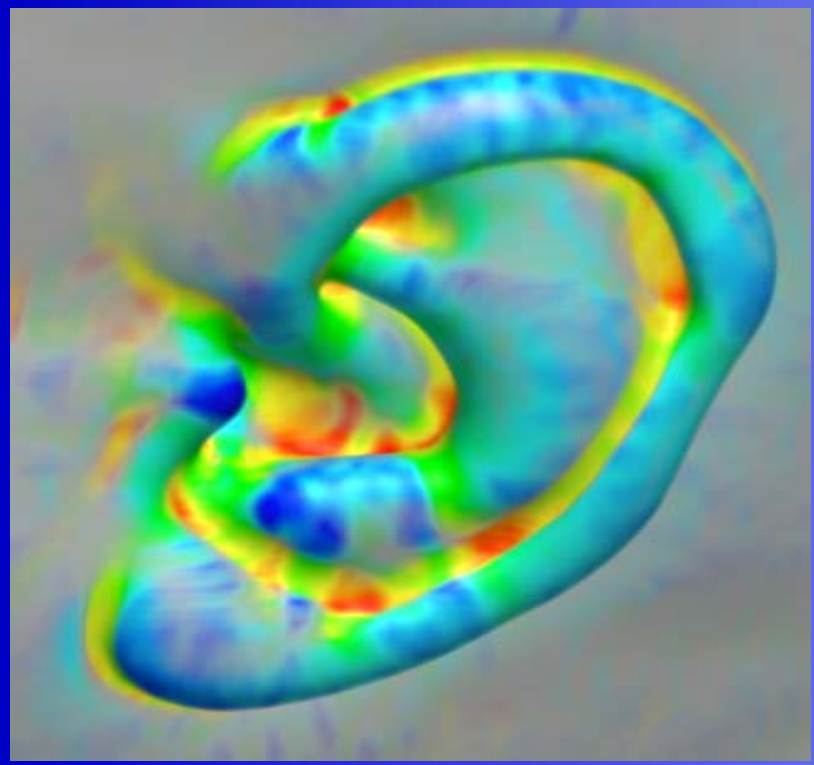
Lifestyle Modifications

- Low sodium diet
- Avoidance or conservative use of stimulants, tobacco, and alcohol
- Limited noise exposure
- Restricted physical exertion

Treatment

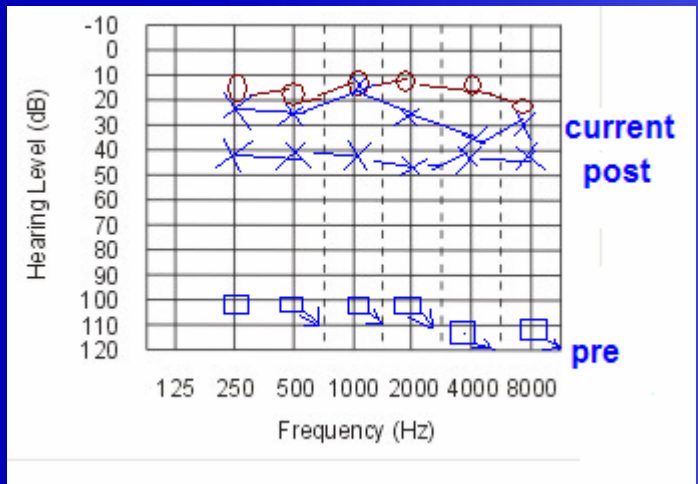
- Tinnitus management
- Sound generators (Broad band or narrow band/freq. spec)
- Masking with cochlear implant?
- TRT
- Behavioral Cognitive Tx.
- Amplification
- Aural rehabilitation

(Vernon, 2000) & (Henry, et al, 2005)



Case Presentation

- Male, 40 y.o., diagnosed with SSHL and tinnitus, AS. MRI negative for CPA space-occupying lesion. Cardiologic evaluation supported mild mitral valve prolapse (MVP)
- Oral corticosteroid therapy followed by two episodes of intratympanic treatment
- Patient's hearing improved to 25 dB HL within three months of treatment. Patient is currently receiving tinnitus management and uses a tinnitus device.



Current Study

- Retrospective study in progress: ten patients with unilateral persistent sudden hearing loss and tinnitus were asked to complete follow-up questionnaires
- Five chose amplification to manage tinnitus. Use of hearing aids can result in habituation to tinnitus by means of an environmental sound gain increase

(Danesh & Andreassen, 2007) & (Henry et al., 2005)

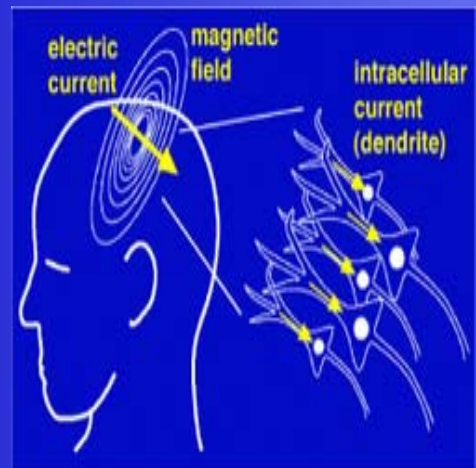
The importance of audiologic management in persistent hearing loss after medical management of SSHL

Studies of the auditory evoked magnetic fields have shown activity in multimodal association areas in patients with sudden unilateral hearing loss (Makela, 1997). This encourages the neural plasticity school of thought.

Cortical Plasticity & SSHL

- Scalp magnetoencephalography (MEG) used by Vasama and Makela to examine cortical plasticity
- Auditory evoked magnetic field recorded up to 5 years post onset of SSHL
- Magnetic field distribution pinpointed location of brain activity
- Auditory system reorganization shown

(Neuman, 2005)



Cortical Plasticity & SSHL

- LATE ONSET AUDITORY DEPRIVATION-monaural amplification and subsequent poor performance of speech recognition. This indicates the importance of amplification in cases with unilateral hearing loss.
- HEARING AID ACCLIMITIZATION-is the time necessary to improve auditory performance. Patients with unilateral SSNHL and hearing aids should be given time for this.
- Electrophysiologic measures may be able to determine how cortical plasticity affects the patient's success with amplification. Evoked potentials may be able to quantify the use of hearing aids as catalysts for brain activity pattern change (e.g., change in amplitude and latency of LAEPs)

(Neuman, 2005)

Future Treatments

- Gene therapy
- Antiapoptic agents
- Autoprotective drugs: AM-111 (treats cochlear stress)
- Antioxidants

ENT Today, Dec. 2006

Primary Care Physician Education

- Surveys for investigation on how PCPs diagnose SSNHL
- Results will allow audiologist to effectively collaborate with physicians to design protocols

A Simple Test

The “Hum” or humming Test

-similar to a Weber test

- helps to distinguish sudden hearing loss from more benign cases

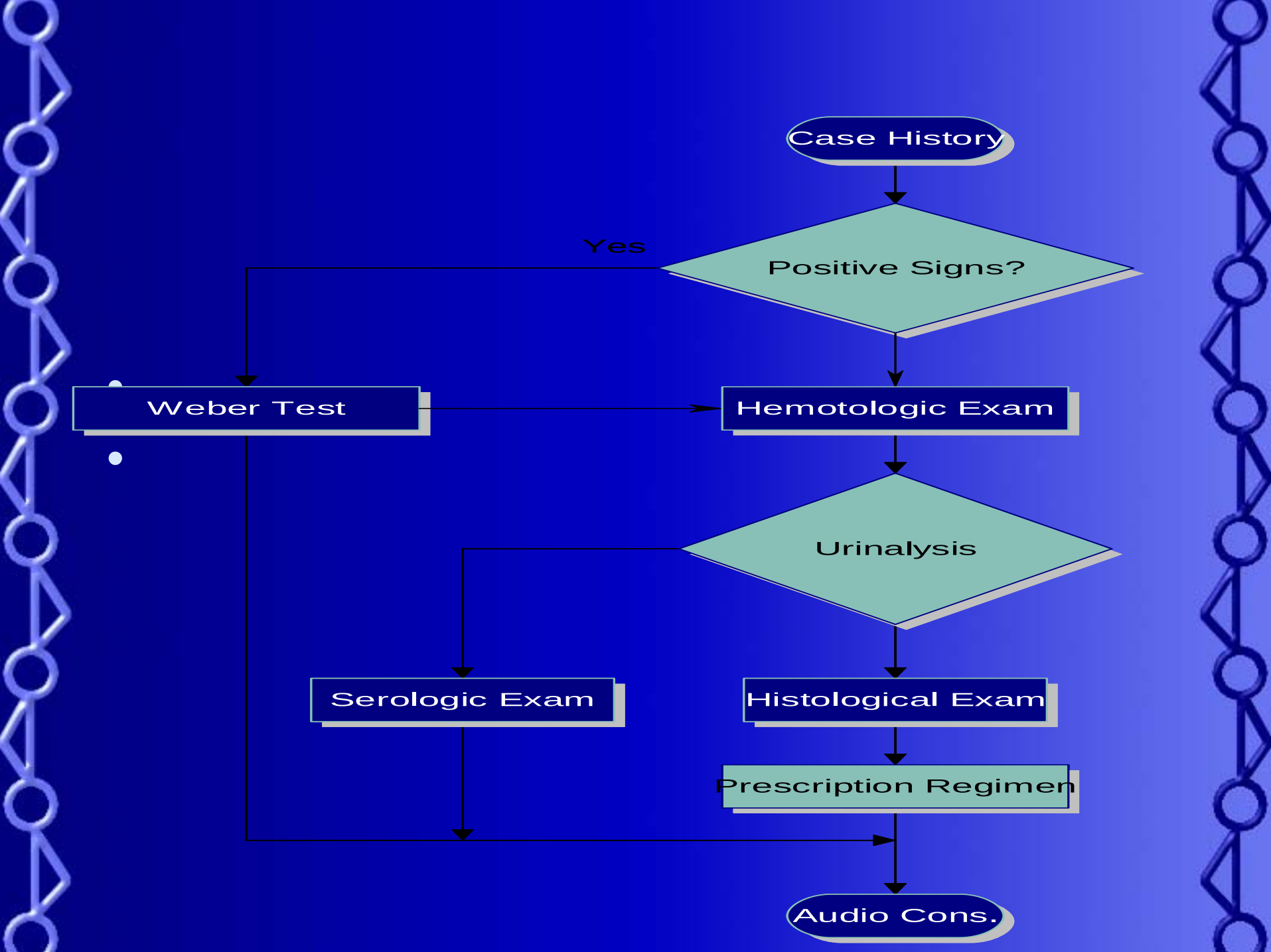
-can be done over telephone

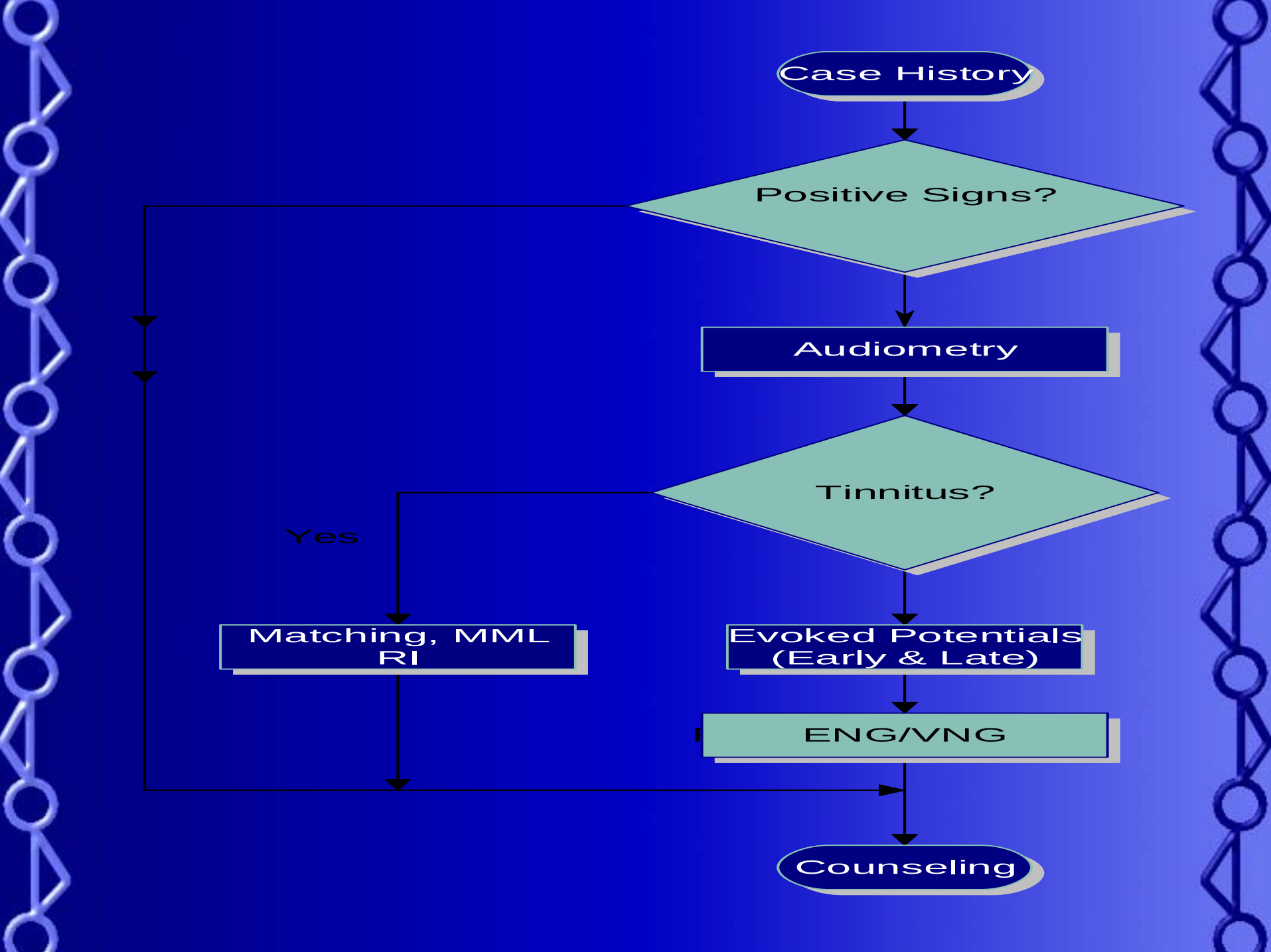
1. Patient hears voice in blocked ear? Possibly cerumen or other conductive cause

2. Voice heard in good ear? Suspect nerve loss

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- Eva Orzan and Alessandra Murgia (006)


Case History
Positive Signs?
Yes
Weber Test
Hemotologic Exam
Urinalysis
Serologic Exam
Histological Exam
Prescription Regimen
Audio Cons.


Case History
Positive Signs?
Audiometry
Tinnitus?
Yes
Matching, MML
RI
Evoked Potentials
(Early & Late)
ENG/VNG
Counseling