

## The Difficult to Test Population: Hearing Testing Techniques Using Objective and Behavioral Testing

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## Causes of Hearing Loss

**Approximately 50% associated with genetic disorders**

**70% of these have a recessive cause**

**15% have a dominant cause**

**15% have other forms of inheritance**

## Genetic causes of hearing loss

**There are over 400 identified genetic abnormalities that are associated with hearing loss.**

**The most common non-syndromic cause is due to abnormalities of the connexin-26 gene**

## Dominant Syndromic Causes of Loss

- **Waardenburg Syndrome**
- **Branchio-Oto-Renal (BOR) Syndrome**
- **Neurofibromatosis Type II (NFII)**
- **Stickler Syndrome**
- **Treacher-Collins Syndrome**

## Common Recessive Syndromic Causes of Hearing Loss

- **Usher Syndrome**
- **Alport Syndrome**
- **Jervell and Lange-Nielson Syndrome**
- **Pendred Syndrome**

## Other Genetic Causes

- **Down Syndrome**
- **Goldenhar Syndrome**
- **Angelman Syndrome**
- **Many many others**

### Other Risk Factors for Hearing Loss

- Bacterial meningitis/infections
- Head trauma
- Ototoxic medications
- Recurrent otitis media
- Family history

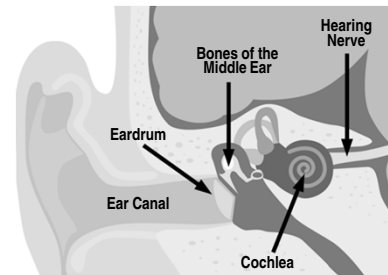
### Other Risk Factors for Hearing Loss

- Low birth weight
- TORCH Complex
- Hyperbilirubinemia
- Low APGAR
- Hypoxia

### Other Risk Factors for Hearing Loss

- Prolonged ventilation

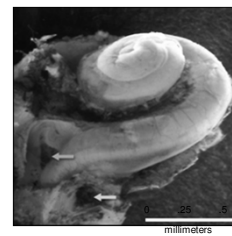
### How We Hear



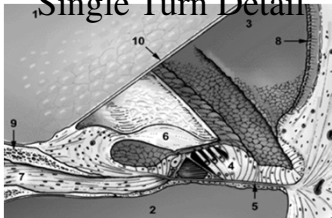
### How We Hear



### Human Cochlea (5 months of gestation).

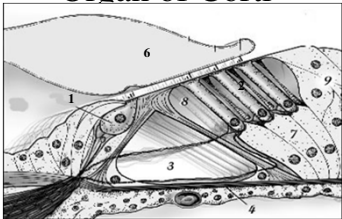


### Single Turn Detail



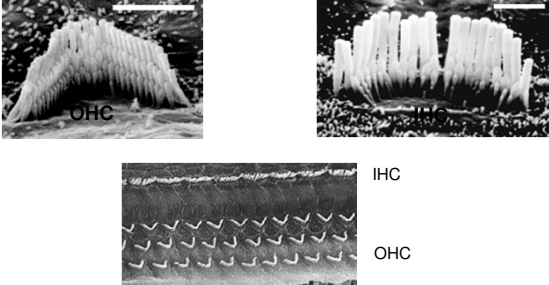
1. Scala Vestibuli	6. Tectorial Membrane
2. Scala Tympani	7. Nerve Fibers
3. Scala Media	8. Stria Vascularis
4. Organ of Corti	9. Spiral Ganglion
5. Basilar Membrane	10. Reissner's Membrane

### Organ of Corti



1. Inner Hair Cells	4. Basilar membrane	7. Deiter's Cells
2. Outer Hair Cells	5. Reticular lamina	8. Nuel's Space
3. Tunnel of Corti	6. Tectorial Membrane	9. Hensen's Cells

### Hair Cells




OHC

IHC

OHC

### Electroacoustic Tests

- Immittance
- Otoacoustic Emissions



### Immittance

- Ear Canal Volume (ECV or PVT)
- Tympanometry
- Static Compliance
- Acoustic Reflex, Decay, & Latency

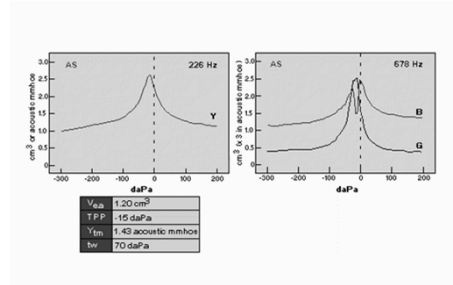
### Ear Canal Volume

- Measure at +200 mmH20
- Provides measure of volume of external ear canal
- Volumes based on age
- Volumes greater than 2.5 suggest:
  - Perforation or
  - Patent PE tube

## Tympanometry

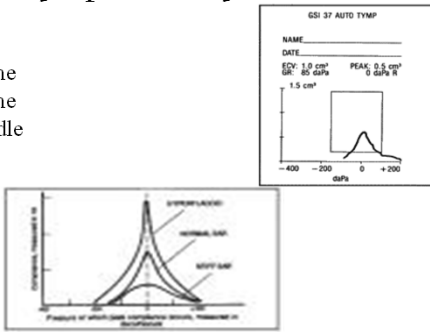
- Often used in conjunction with OAE or acoustic reflex test
- Measures mobility of the eardrum
- Useful for infants and young children

## Tympanograms

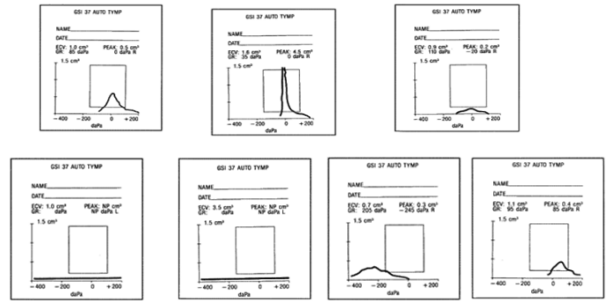


## Tympanometry

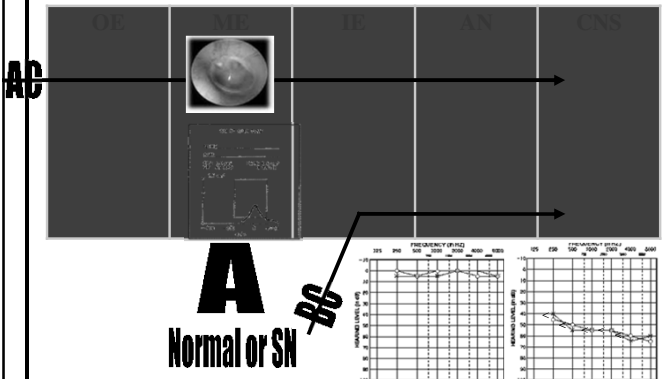
- Objective measure of the function of the TM and middle ear
- 5 or 6 basic shapes



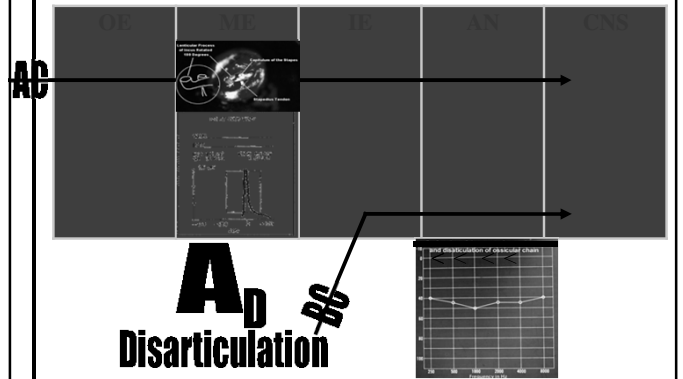
## Tympanogram Types



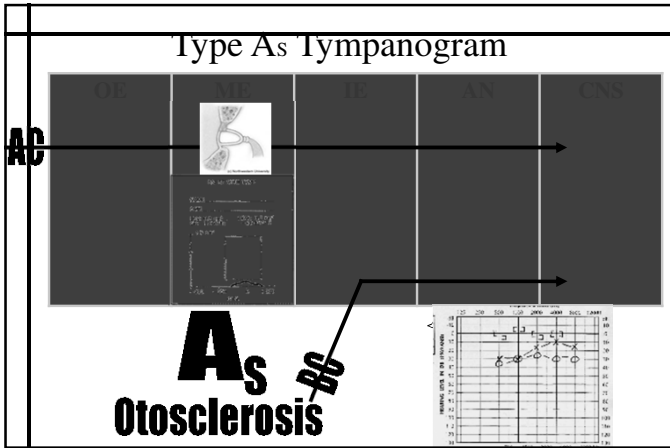
## Type A Tympanogram



## Type Ad Tympanogram



### Type A<sub>s</sub> Tympanogram



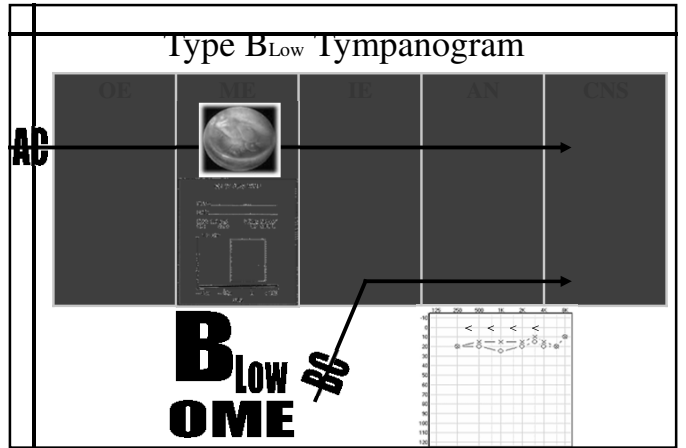
AC

**A<sub>s</sub>**  
**Otosclerosis**

SS

This slide illustrates a Type A<sub>s</sub> Tympanogram. It features a central image of a tympanogram showing a very sharp peak at 226 Hz, indicating a stiff middle ear. To the right is a corresponding graph with a single, narrow peak. Below the graph is a schematic diagram of the ear with an arrow pointing to the ossicles, and a small inset graph showing the peak. The text 'AC' is on the left, and 'SS' is written diagonally. The main diagnosis is 'A<sub>s</sub> Otosclerosis'.

### Type B<sub>Low</sub> Tympanogram



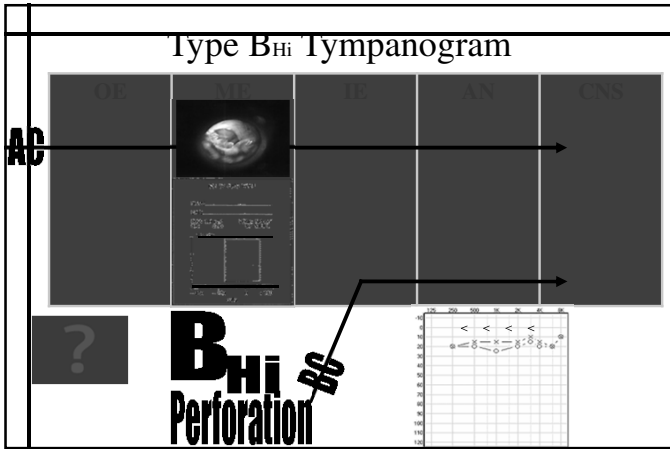
AC

**B<sub>Low</sub>**  
**OME**

SS

This slide illustrates a Type B<sub>Low</sub> Tympanogram. The central image shows a flat line across the frequency range, indicating no middle ear movement. The graph to the right shows a flat line. Below is a schematic diagram of the ear with an arrow pointing to the ossicles and a small inset graph showing a flat line. The text 'AC' is on the left, and 'SS' is written diagonally. The main diagnosis is 'B<sub>Low</sub> OME'.

### Type B<sub>Hi</sub> Tympanogram



AC

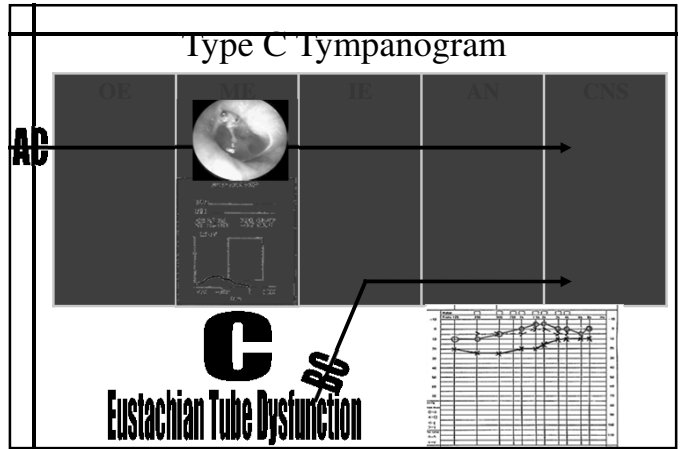
**B<sub>Hi</sub>**  
**Perforation**

?

SS

This slide illustrates a Type B<sub>Hi</sub> Tympanogram. The central image shows a flat line, but with a question mark in a box to the left. The graph to the right shows a flat line. Below is a schematic diagram of the ear with an arrow pointing to the ossicles and a small inset graph showing a flat line. The text 'AC' is on the left, and 'SS' is written diagonally. The main diagnosis is 'B<sub>Hi</sub> Perforation'.

### Type C Tympanogram



AC

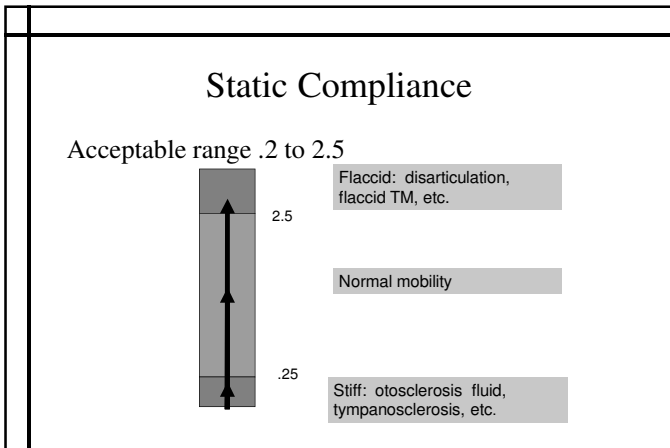
**C**  
**Eustachian Tube Dysfunction**

SS

This slide illustrates a Type C Tympanogram. The central image shows a flat line, but with a question mark in a box to the left. The graph to the right shows a flat line. Below is a schematic diagram of the ear with an arrow pointing to the ossicles and a small inset graph showing a flat line. The text 'AC' is on the left, and 'SS' is written diagonally. The main diagnosis is 'C Eustachian Tube Dysfunction'.

### Static Compliance

Acceptable range .2 to 2.5



2.5

Flaccid: disarticulation, flaccid TM, etc.

Normal mobility

.25

Stiff: otosclerosis fluid, tympanosclerosis, etc.

This diagram shows a vertical scale for static compliance. The scale ranges from 0 at the bottom to 2.5 at the top. A horizontal bar labeled 'Normal mobility' is positioned between approximately 0.25 and 2.5. A horizontal bar labeled 'Flaccid: disarticulation, flaccid TM, etc.' is positioned above 2.5. A horizontal bar labeled 'Stiff: otosclerosis fluid, tympanosclerosis, etc.' is positioned below 0.25. A vertical double-headed arrow spans from 0.25 to 2.5.

### Acoustic Reflex Testing

- Measures contraction of the stapedius muscle in the middle ear
- Useful for infants and young children

### ART

### Acoustic Reflexes

### Otoacoustic Emissions

David Kemp discovered OAEs

- Acoustic energy produced by the cochlea and recorded in the external auditory canal
- Most likely energy produced by outer hair motility and possibly outer hair cell cilia
- Objective test
  - DPOAE
  - TEOAE

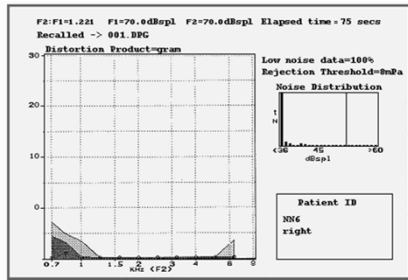
### Otoacoustic Emissions (OAE)

- Measure sound produced by hair cells in the cochlea
- Ideal for those who remain quiet during the testing

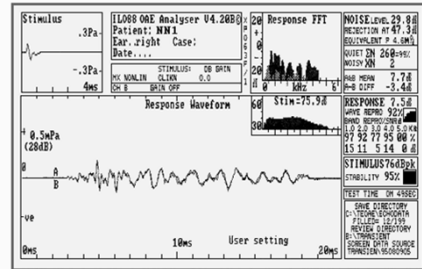
### DPOAE

### Distortion-Product OAEs

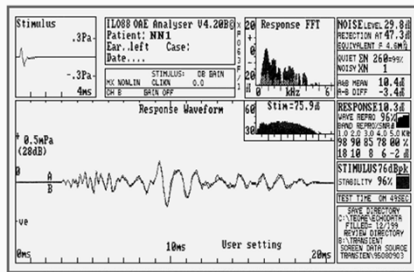
### Distortion-Product OAEs



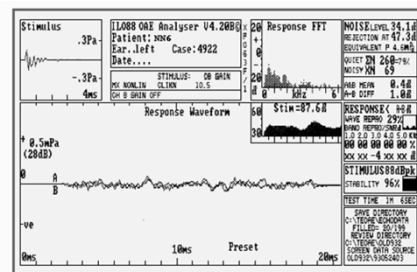
### Transient Evoked OAEs



### Transient OAEs



### Transient OAEs



### What Can Audiology Tell Us?

- Type of loss
  - Conductive
  - Sensorineural



### What Can Audiology Tell Us?

- Severity of loss:
  - Mild: hears conversation; may need amplification
  - Moderate: develops speech and language but requires amplification
  - Severe: requires amplification, auditory management and professional assistance
  - Profound: requires significant support, amplification and, in some cases, cochlear implants

## Types of Hearing Loss

- **Conductive**
  - Middle or outer ear
  - Persistent middle ear infections
  - Fluctuating hearing levels
- **Sensorineural**
  - Inner ear and beyond
  - Cochlear hair cell dysfunction
  - Filter and distort sound

## Conductive vs. Sensorineural

- **Conductive**
  - Middle ear dysfunction
  - Some treatable with medication
- **Sensorineural**
  - Pathology in the inner ear, the cochlea, or the 8th cranial nerve
  - No medical treatment
  - Cochlear implant appropriate for some

## Electroacoustic Triage Trio

- Tympanogram
- Acoustic Reflex
- Otoacoustic Emissions

## Triage Trio

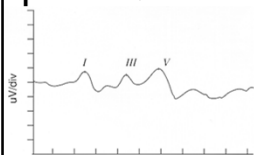
<b>Tympanogram</b> Type A	<b>Acoustic Reflexes</b> Normal	<b>OAE</b> Normal	<b>Normal</b> peripheral and lower brainstem function (possible APD) normal hearing
<b>Tympanogram</b> Type A	<b>Acoustic Reflexes</b> Normal Range Normative Data	<b>OAE</b> Absent or Depressed	<b>Cochlear</b> loss, outer hair cell loss, ABR normal, hearing aids beneficial
<b>Tympanogram</b> Type A	<b>Acoustic Reflexes</b> Absent or Elevated	<b>OAE</b> Present	<b>Auditory Neuropathy/</b> Auditory Dys-synchrony
<b>Tympanogram</b> Type A	<b>Acoustic Reflexes</b> Absent	<b>OAE</b> Absent	<b>Severe or profound inner ear loss</b> (occasionally otosclerosis)
<b>Tympanogram</b> Not Type A	<b>Acoustic Reflexes</b> Absent	<b>OAE</b> Absent	<b>Conductive or mixed loss</b> (possible severe/profound loss)

## Evoked Potentials

- EcochG
- ABR
- Middle Latency
- Late Response



Happy baby all wired up for ABR test

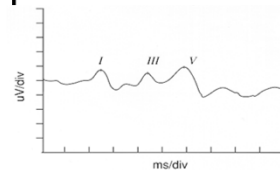


## Evoked Potentials

- EcochG
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- Middle Latency
- Late Response



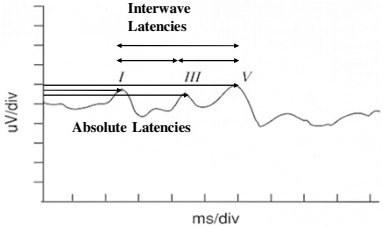
Happy baby all wired up for ABR test





### ABR Wave

- I Auditory Nerve
- II Auditory Nerve
- III Cochlear Nuclei
- IV SOC
- V ???



### Behavioral Techniques

- BOA
- VRA
- COR
- Speech testing compared to pure tones
- CPA