## 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

Human Cochlea (5 months of gestation).
**Single Turn Detail**

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

**Organ of Corti**

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

**Hair Cells**

- Inner Hair Cells (IHC)  
- Outer Hair Cells (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

A Normal or SN

AD Disarticulation
Type A<sub>s</sub> Tympanogram

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

Type B<sub>Low</sub> Tympanogram

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

Type B<sub>Hi</sub> Tympanogram

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

Type C Tympanogram

**C**
Eustachian Tube Dysfunction

**Static Compliance**

Acceptable range: .2 to 2.5

- Flaccid: disarticulation, flaccid TM, etc.
- Normal mobility
- Stiff: otosclerosis fluid, tympanosclerosis, etc.

**Acoustic Reflex Testing**

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

![Distortion-Product OAEs](image)

### Transient Evoked OAEs

![Transient Evoked OAEs](image)

### Transient OAEs

![Transient OAEs](image)

### 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

<table>
<thead>
<tr>
<th>Tympanogram</th>
<th>Acoustic Reflexes</th>
<th>OAE</th>
<th>Normal peripheral and lower brainstem function (possible APD) normal hearing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type A</td>
<td>Normal or Depressed</td>
<td>OAE Absent or Elevated</td>
<td>Cochlear loss, outer hair cell loss, ABR normal, hearing aids beneficial</td>
</tr>
<tr>
<td>Normal Range</td>
<td>Normal or Depressed</td>
<td>OAE Absent or Elevated</td>
<td>Auditory Neuropathy/Auditory Dys-synchrony</td>
</tr>
<tr>
<td>Absent</td>
<td>Normal or Depressed</td>
<td>OAE Absent</td>
<td>Severe or profound inner ear loss (occasionally otosclerosis)</td>
</tr>
<tr>
<td>Absent</td>
<td>Absent</td>
<td>OAE Absent</td>
<td>Conductive or mixed loss (possible severe/profound loss)</td>
</tr>
</tbody>
</table>

Evoked Potentials

- EcochG
- ABR
- 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