Genetics and deafness

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Deafness definition

- Person who has little or no functional hearing and depends upon visual rather than auditory communication

<table>
<thead>
<tr>
<th>Degree of deafness</th>
<th>dB heard</th>
<th>Hearing ability</th>
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<tbody>
<tr>
<td>Profound</td>
<td>95 dB</td>
<td>Lip reading required for understanding speech but may benefit from hearing aids</td>
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<tr>
<td>Severe</td>
<td>70-94 dB</td>
<td>Require lip reading even with hearing aids</td>
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<tr>
<td>Moderate</td>
<td>40-69 dB</td>
<td>Difficulty following speech without a hearing aids</td>
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<tr>
<td>Mild</td>
<td>21-39 dB</td>
<td>Difficulty following speech in noisy situations</td>
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Deafness: Quick Facts

• Deafness is most common sensory deficit in the United states
• Affects over 28 million Americans
• 1:1000 infants prelingual deafness
• 1:1000 infants prelingual hearing loss
• 1:1000 deaf before adulthood
Causes of Deafness:

- 60% of childhood hearing loss is genetic
  - Syndromic
  - Nonsyndromic
- 40% of childhood hearing loss is caused by infectious/environmental factors
  - Prenatal infections (CMV, toxoplasmosis, etc)
  - Meningitis
  - Low birth weight/prematurity
  - Ototoxic medications
  - Mechanical ventilation
  - Trauma
Types of genetic hearing loss

- Syndromic (30%)
- Non-syndromic autosomal recessive (56%)
- Non-syndromic autosomal dominant (12%)
- Non-syndromic X-linked, mito, others (2%)
Modes of inheritance for non-syndromic hearing loss

**Autosomal Dominant**
DFNA – 12%

**X-linked**
DFN – 1%

**Autosomal Recessive**
DFNB – 56%

**Mitochondrial**
1%
Why is deafness so common?

There are so many genes involved in hearing.