Deafness

1 in 1,000 born deaf worldwide
- 0.3% of children under 5 years deaf
1 in 1,000 will develop deafness
500,000 to 750,000 Americans deaf
- No benefit from hearing aids

Two types of hearing loss
- Conductive hearing loss
  - Mechanical deficit in bringing vibrations to inner hear
- Sensory-neural hearing loss
  - Problem converting sound to electrical signal that travels to the brain
Causes of Hearing Loss

Presbycusis

- Gradual loss of hearing as we get older
- 75% of people over 60 have significant hearing loss
- Due to loss of hair cells
  - Especially near the oval window (high frequencies)
- Consonants contain higher frequencies
  - Inability to hear consonants leads to poor speech discrimination
- Test:
  - Speech
  - Speech w/o high frequencies

![Graph showing threshold of audibility](image-url)
Causes of Hearing Loss

Excessive noise, as due to:

- Occupation
  - E.g.: factory noise, pneumatic hammer,…

- Recreation
  - E.g.: shooting range, hunting,…

- Noise exposure
  - E.g.: disco, loud music,…

- Most damaging noise: impulsive
  - E.g. gunshot
Causes of Hearing Loss

Menier’s disease
- High fluid pressure in the inner ear
- Gives a low frequency hearing loss
- Instead of being progressive, it fluctuates
  - Hearing loss may change from day to day
- Also affects balance (gives vertigo)

Otosclerosis
- Excessive growth of bone surrounding middle and inner ear
  - May block stirrup and pinch auditory nerve
- Hereditary; also may develop after childhood measles infection

Sudden hearing loss
- Usually afflicts older adults
- Typically only one ear
- Can be viral or due to vascular accidents

Tinnitus (ringing in the ear)
- Often accompanies hearing loss
  - Nerve fibers to dying hair cells become irritable and discharge
Causes of Hearing Loss

Ear infection (Otitis)

Meningitis

Usher syndrome (also causes blindness)

Conductive hearing loss
  - Damage to eardrum or earbones (usually corrected by surgery)
  - Wax or fluid in middle ear

Autoimmune diseases
  - E.g., rheumatoid arthritis, lupus

Auditory neuropathy
  - Sound enters inner ear normally but transmission from inner ear to brain is impaired
    • May involved damage to hairy cells or faulty connection between hairy cells and auditory nerve

When loss is progressive in only one ear, it may be due to causes beyond the inner ear
  - Acoustic nerve or auditory part of the brain
Hearing Aids

Components:
- Microphone
- Battery-operated amplifier
- A means of transmitting sound to the user
  - Speaker
  - Direct transmission to bones or skull (requires surgical implant)

May selectively amplify high frequencies
- Some have digital equalizers that can be programmed depending on the environment
  - Some are simple devices that only boost high frequencies

Difficult to use with a telephone
- Some accept Direct Audio Input, which allows an external source (e.g. a telephone) to connect directly to the hearing aid
Hearing Aids

Common problems
- Over amplification
- Occlusion effect
  - Hollow sound due to ear canal blockage
- Larsen feedback (whistling)
  - Largely eliminated using digital technology
- Poor speech discrimination in noise

Can only amplify signal - won’t work for deafness

Economic considerations
- Cost (per ear):
  - $800-$1500 (analogic)
  - $1200-$3000 (digital)
  - Cost mostly due to service (fitting etc.)
  - Not covered by Medicare
  - Partly covered by Medicaid
  - Only some insurances cover it
Assistive Listening Devices (ALD)

- Amplified telephones
- Alarms/alerts that emit loud signals, flash and shake bed
- Directional microphones that allow you to hear the person talking to you in a noisy environment
- FM or infrared link from stereo/TV or microphone
  - E.g. in class: teacher speaks to mic, signal transmitted via FM to student’s ALD
    - Reduces problems with reverberation, noise, distance
Telecoils

Telecoil-equipped hearing aids can receive electromagnetic signal via an induction coil

The signal can be generated by:

- **A room loop**: an *induction loop* (wire) surrounding an audience (e.g., in the floor or in the ceiling), connected to the source of sound (e.g., microphone)

- **A neck loop**: a necklace-sized wire loop that can be connected to a radio, TV, some telephones, or an ALD and transmits the signal wirelessly to the coil in the hearing aid

- **A silhouette**: works like a neck loop but it is kept behind the ear
Cochlear Implant

Can be used when the auditory nerve is still working but the inner ear isn’t

- Provides electrical signal directly to the auditory nerve by means of multiple electrodes inserted into the cochlea
- Sound is collected at the ear level and processed by an external module
  - Or via FM, DAI or telecoil from ALD
- Processor splits sound up into different nerve electrical impulses
- Electrical impulses transmitted via external coil to internal coil through the skin
- Electrodes in the cochlea stimulate different auditory nerve fibers
Cochlear Implant

Up to 24 electrodes wound through the cochlea, to stimulate the auditory nerve

- Each electrode stimulates a portion of the cochlea
- The signals transmitted to the electrodes are matched to the corresponding tonotopic organization
  - E.g., for a sound with low frequency, only the electrodes near the apex are stimulated
- Note: there are > 15,000 hair cells

About 100,000 have received an implant so far

- Roughly half adult, half children
- Nearly 3000 with bilateral implant

Need to decide which ear to implant
- The anatomy of the cochlea needs to be intact for the implant
Performance

 Depends on:
  – Quality of technology
  – Cause of hearing impairment
  – Amount of functioning nerve fibers
  – Central processing by the brain

Here is an acoustic simulation of cochlear implant

Transforms from totally deaf to hard of hearing
  – E.g., many can use the telephone

Cost: $45K to $70K (all included)
  – Some of this can be covered by health insurance
Windows of Opportunity

Children: If implanted early enough, a child’s brain can learn to make use of the hearing information

– Otherwise brain used for other sensory modality
  • FDA guideline: 12 months
    – 6 months with special approval
– Can be educated in regular schools
– Most are able to engage in conversation at at or near normal level

No upper age limit
– Better if individual was deaf for a short period of time
  • Otherwise it may be difficult to re-adapt to sound