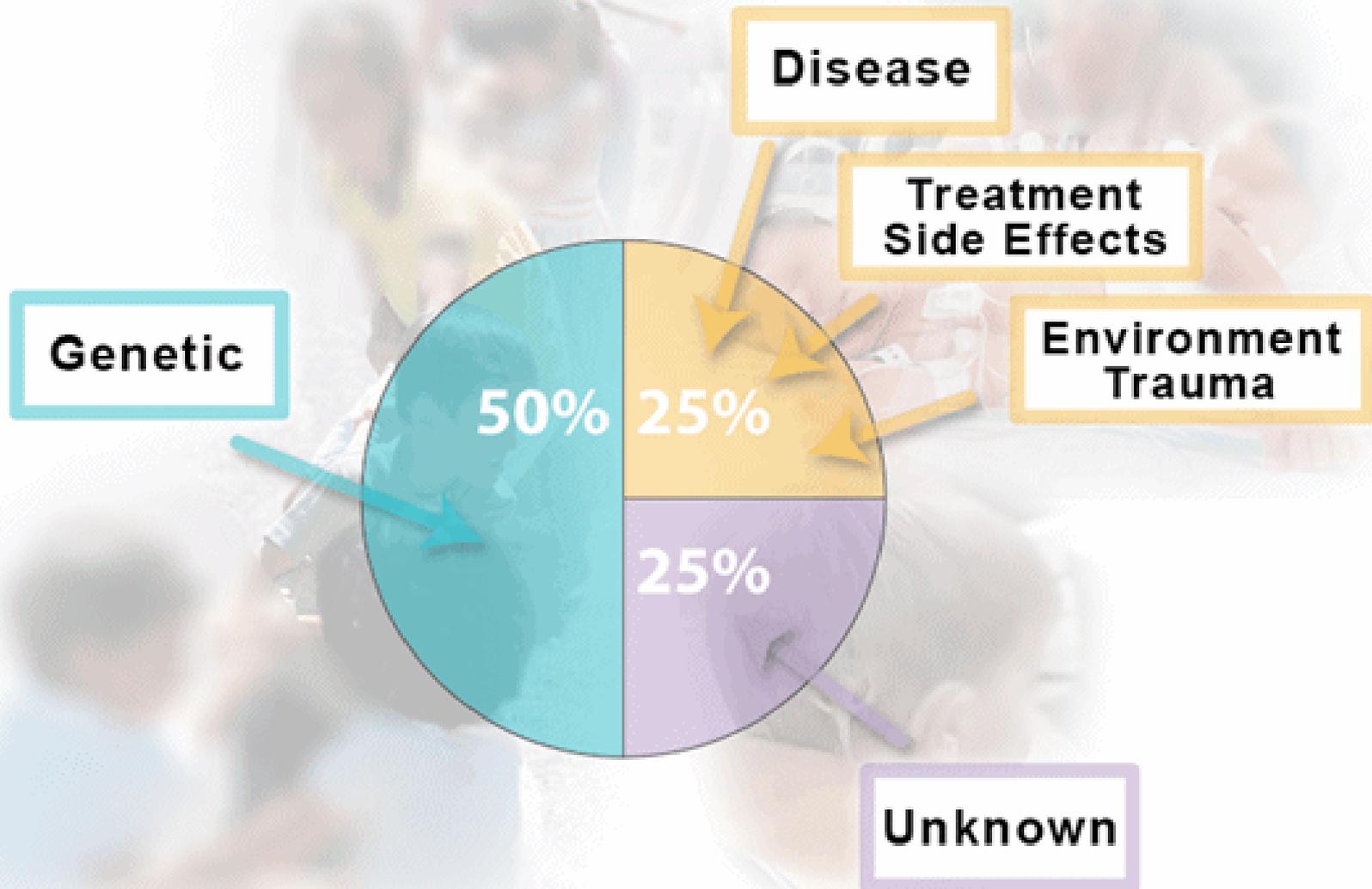


# Hearing loss

- Most common sensory deficit in humans
- 6/1000 children in US born with mild to moderate hearing loss
- 1/1000 children in US born with severe to profound hearing loss
- Additional 1/1000 progress to deafness by adulthood



## Overview of Causes for Hearing Loss



# **CONGENITAL HEARING LOSS**

## *Infant Hearing Screening*

### **High Risk Register (1972 - 1990)**

Family history

Hyperbilirubinemia

TORCHS infections

Craniofacial Syn

Weight < 1500 gm

Meningitis

Asphyxia / Apgar < 4

Ototoxic Meds

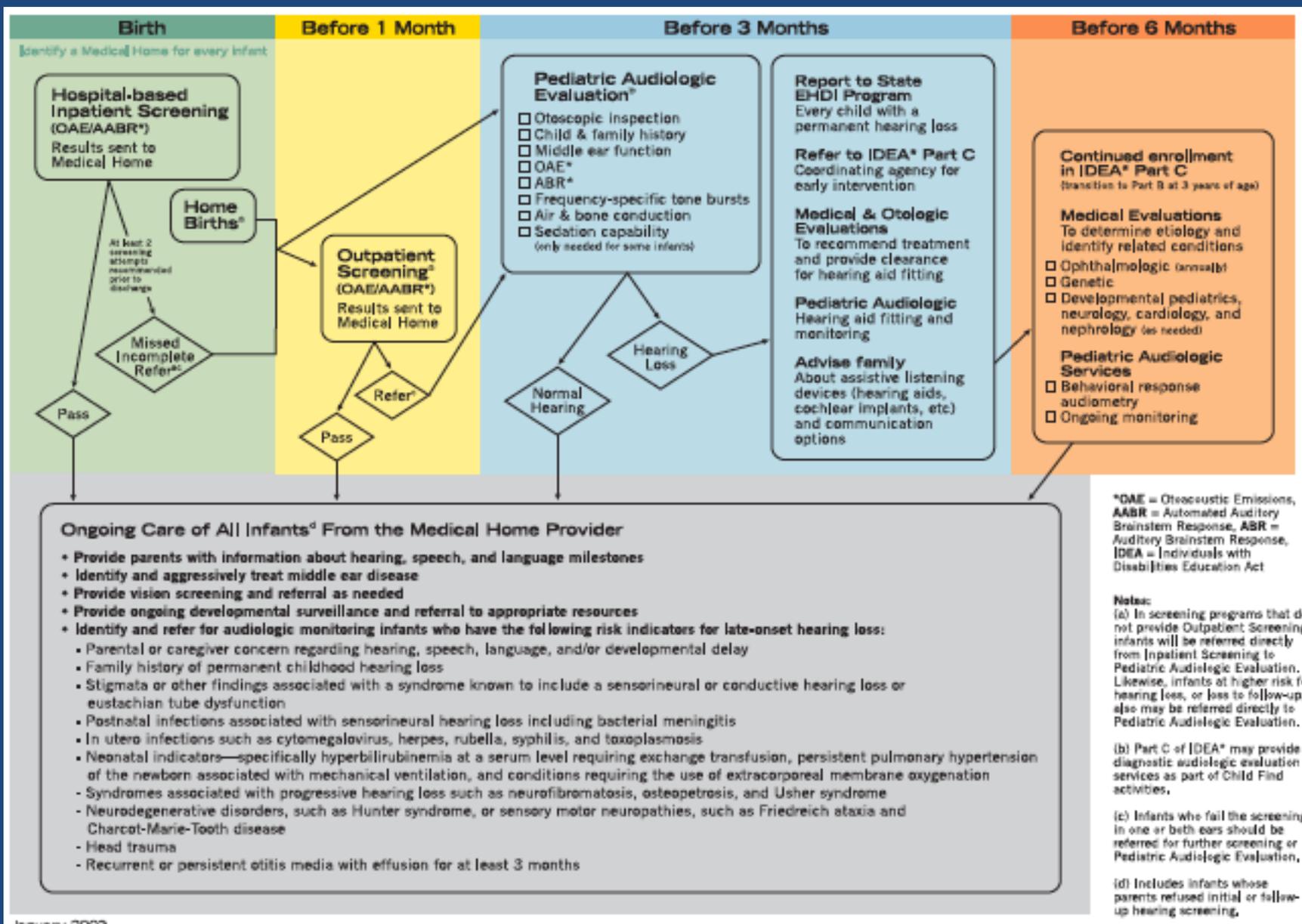
Vent > 2 days

HL Syndromes

# Genetic causes

- Congenital vs delayed
- 70% nonsyndromic
  - 80% Autosomal Recessive
  - 18% Autosomal Dominant
  - 2% Other (X-linked, mitochondrial, chromosomal)
- 30% syndromic



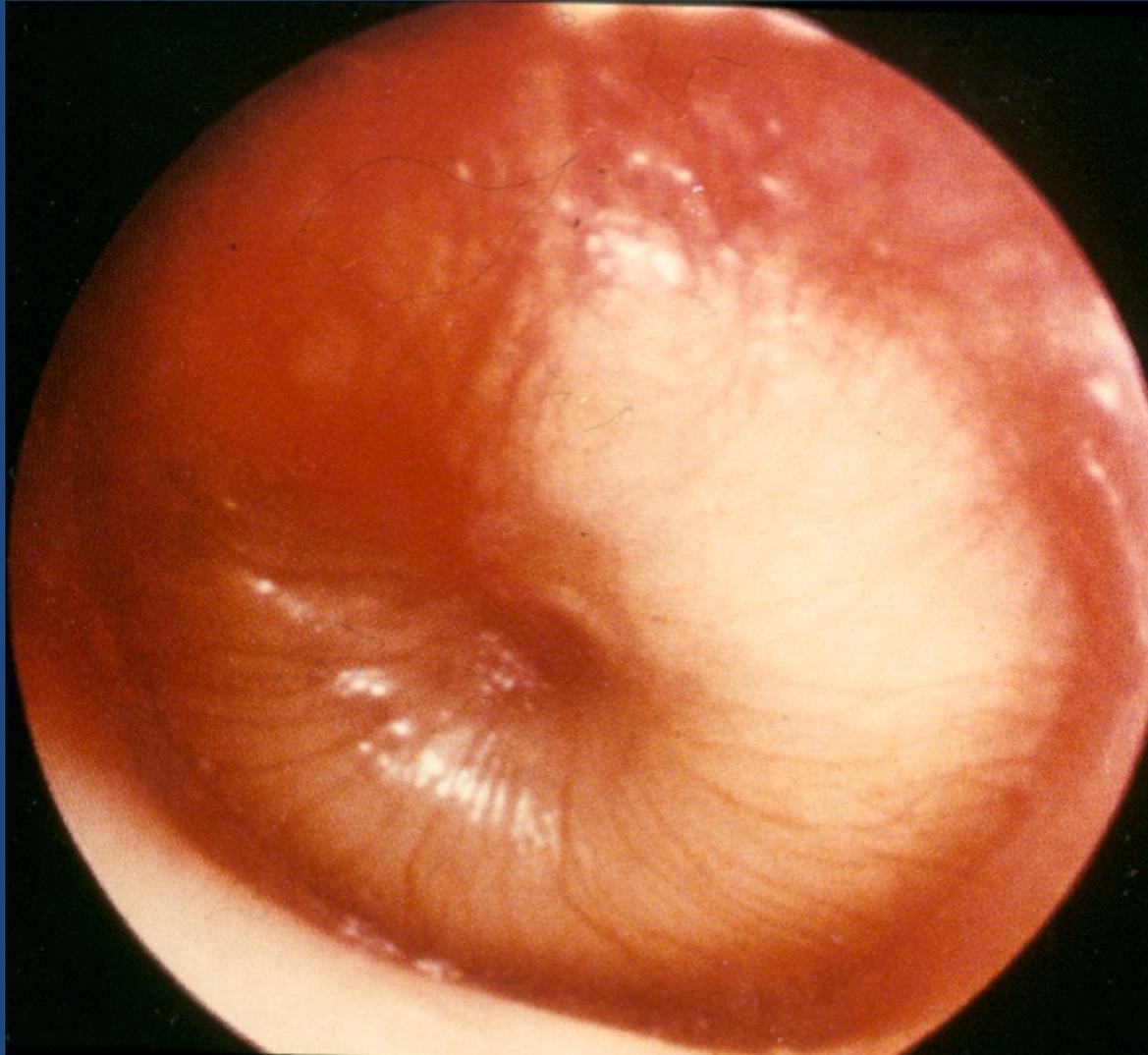


## APPENDIX 2: RISK INDICATORS ASSOCIATED WITH PERMANENT CONGENITAL, DELAYED-ONSET, OR PROGRESSIVE HEARING LOSS IN CHILDHOOD

Risk indicators that are marked with a “§” are of greater concern for delayed-onset hearing loss.

1. Caregiver concern§ regarding hearing, speech, language, or developmental delay.<sup>62</sup>
2. Family history§ of permanent childhood hearing loss.<sup>24,140</sup>
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO,§ assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.<sup>64,131</sup>
4. In utero infections, such as CMV,§ herpes, rubella, syphilis, and toxoplasmosis.<sup>64–67,125,126</sup>
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.<sup>24</sup>
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.<sup>24</sup>
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss,§ such as neurofibromatosis, osteopetrosis, and Usher syndrome<sup>131</sup>; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.<sup>72</sup>
8. Neurodegenerative disorders,§ such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.<sup>131</sup>
9. Culture-positive postnatal infections associated with sensorineural hearing loss,§ including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.<sup>130,131,141</sup>
10. Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization.<sup>127–129</sup>
11. Chemotherapy.§<sup>132</sup>

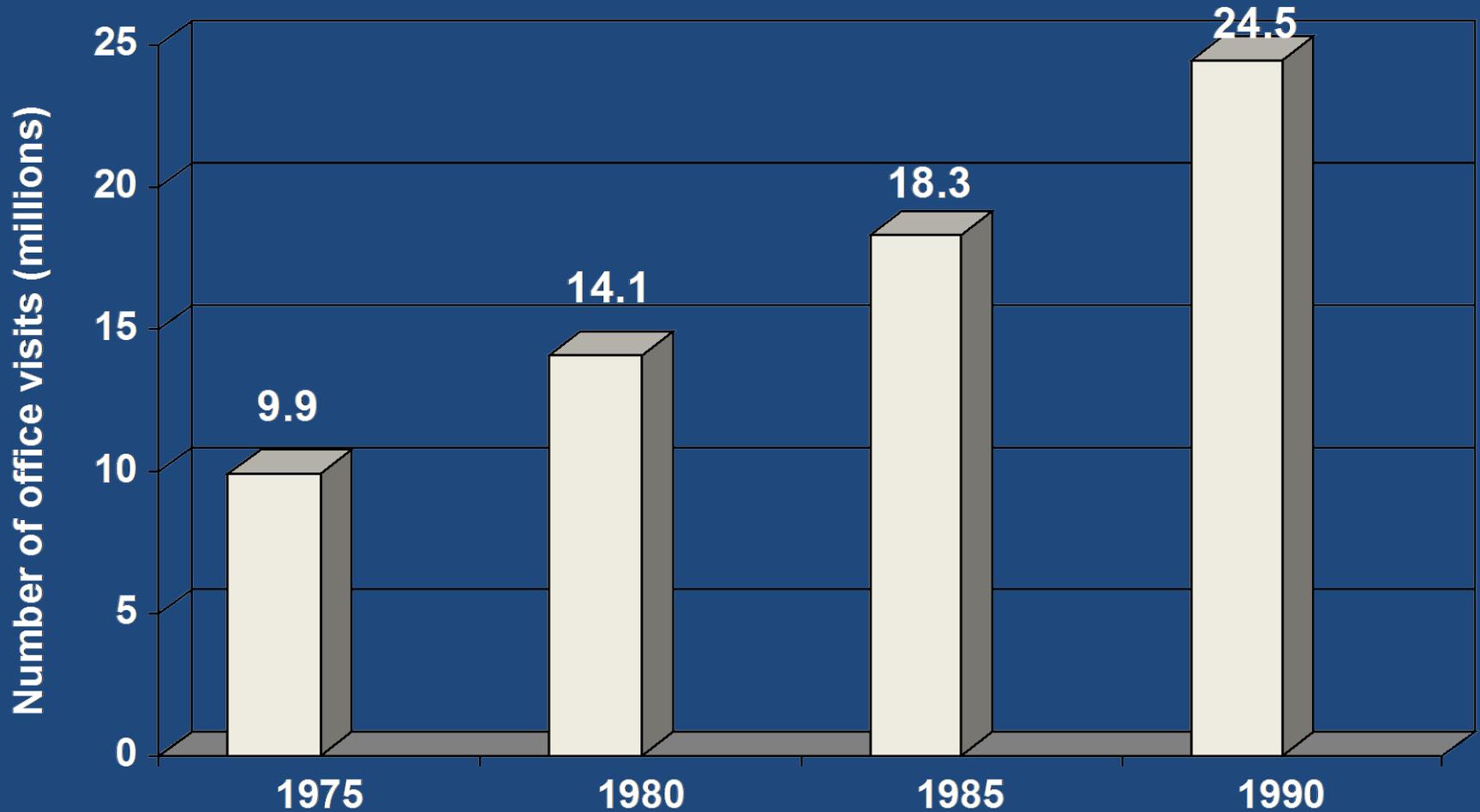
# Acute Otitis Media



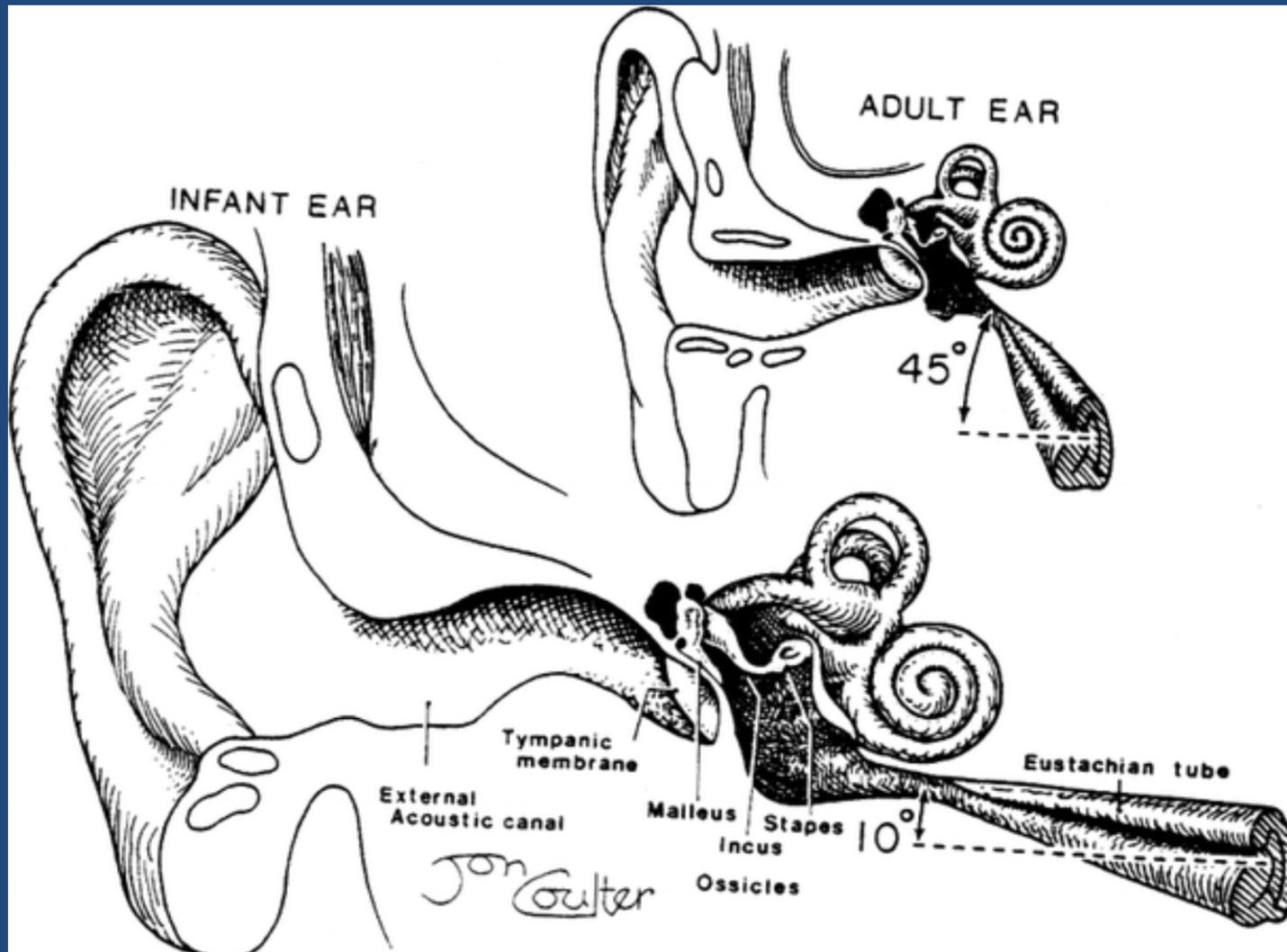
# Otitis media

- Up to 80% children <6 yo will experience at least one infection
- Acute otitis media – typical presentation with otalgia/pulling at ears, irritability and fever, association with URI
- Chronic otitis media – fluid in middle ear for >3mo without signs of acute inflammation
- Within 1<sup>st</sup> year of life, >50% of children will experience otitis media with effusion, most resolving spontaneously

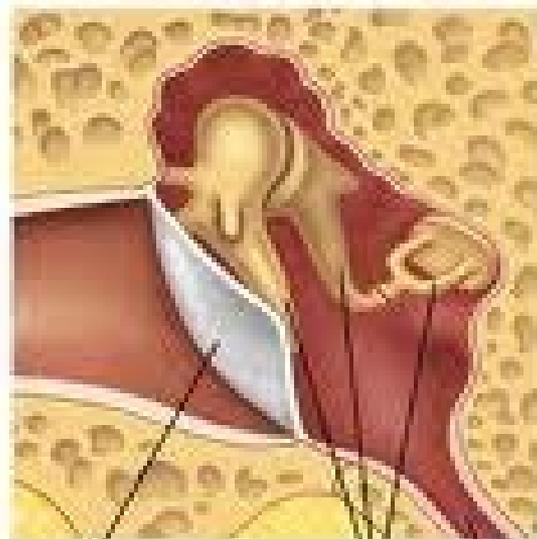
# Office Visits For Otitis Media



# Adults vs. Children



**Normal middle ear**



Ear drum

Auditory bones

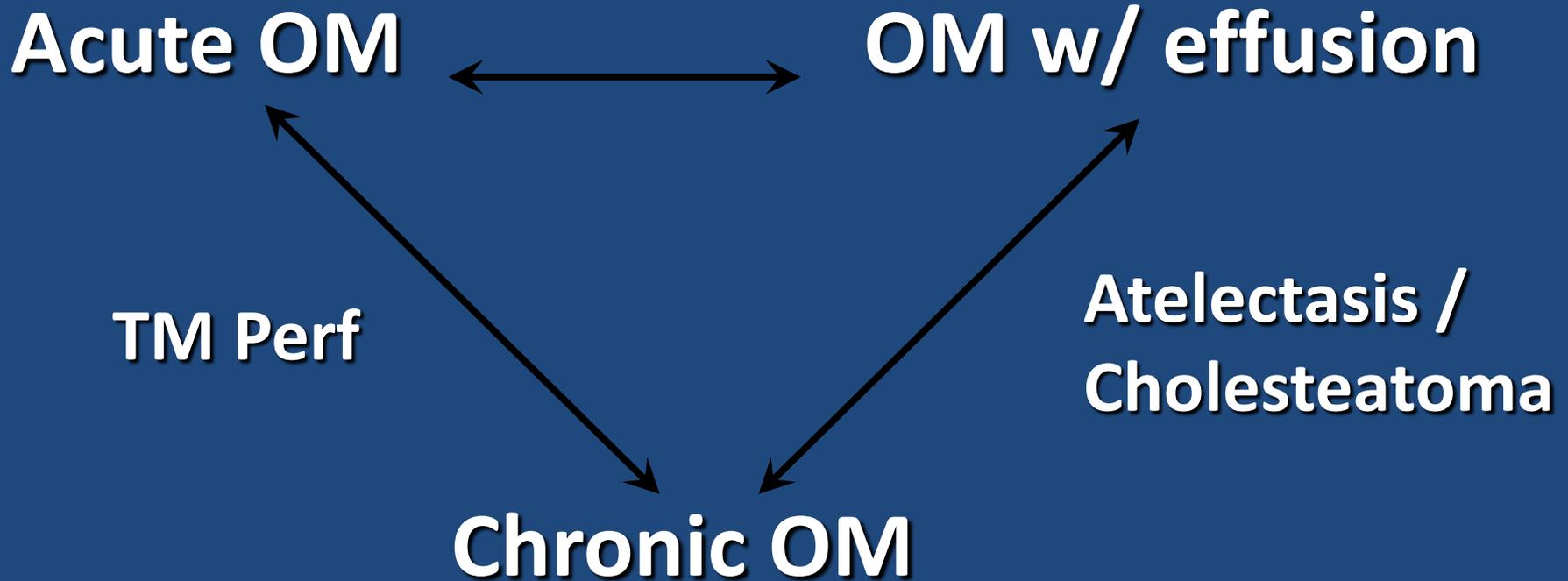
Eustachian tube

**Otitis media**



Infected fluid  
in middle ear

# Classification of Otitis Media



- Acute (< 3 weeks duration)
- Subacute (3-12 weeks duration)
- Chronic (>12 weeks duration)

- Recurrent acute otitis media (4+ episodes/yr or 3+ episodes/6mo)
- Otitis media with effusion (fluid behind middle ear)
- Suppurative otitis media (presence of purulent otorrhea)

# “A Tale of Two Diseases”

- Acute OM
  - Younger than 3 yrs
  - Exam – often normal btw infection
  - Hearing – often normal
- OM w/ effusion
  - Older than 3 yrs
  - Exam – Middle ear effusion
  - Conductive Hearing Loss

# Appropriate Antibiotics

- Uncomplicated AOM:
  - High dose amoxicillin (80 mg/kg)
  - Avoid sulfanamides, macrolides and low dose suppression
- Persistent or recurrent AOM:
  - amoxicillin (80 mg/kg) / clavulanate
  - cefuroxime (30mg/kg)
  - ceftriaxone (3 injections)

# Otitis Media Prevention

## Control risk factors

- Day care
- Smoking

## Appropriate (selective) use of Antibiotics

## Non antibiotic prophylaxis

- Xylitol gum?
- Eustachian tube surfactant?
- Gene studies

## Vaccines

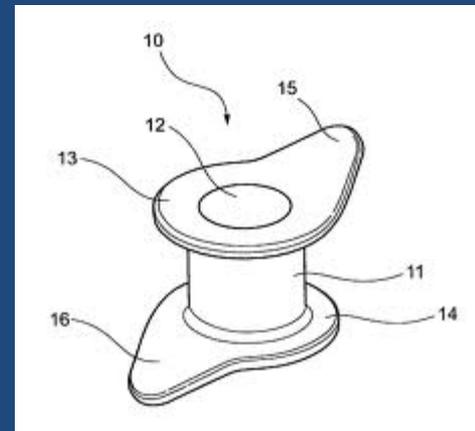
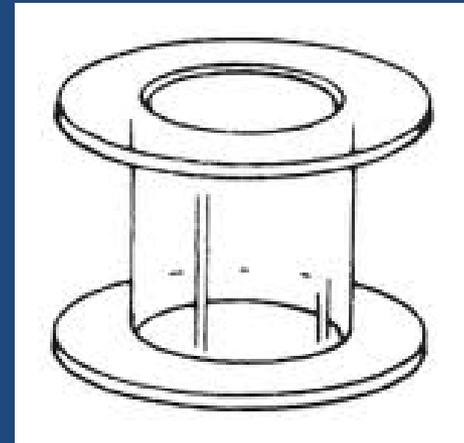
## Surgery – tympanostomy tubes



A small incision is made in the tympanic membrane



Tube inserted to drain fluid



# Technological advances in the treatment of hearing loss

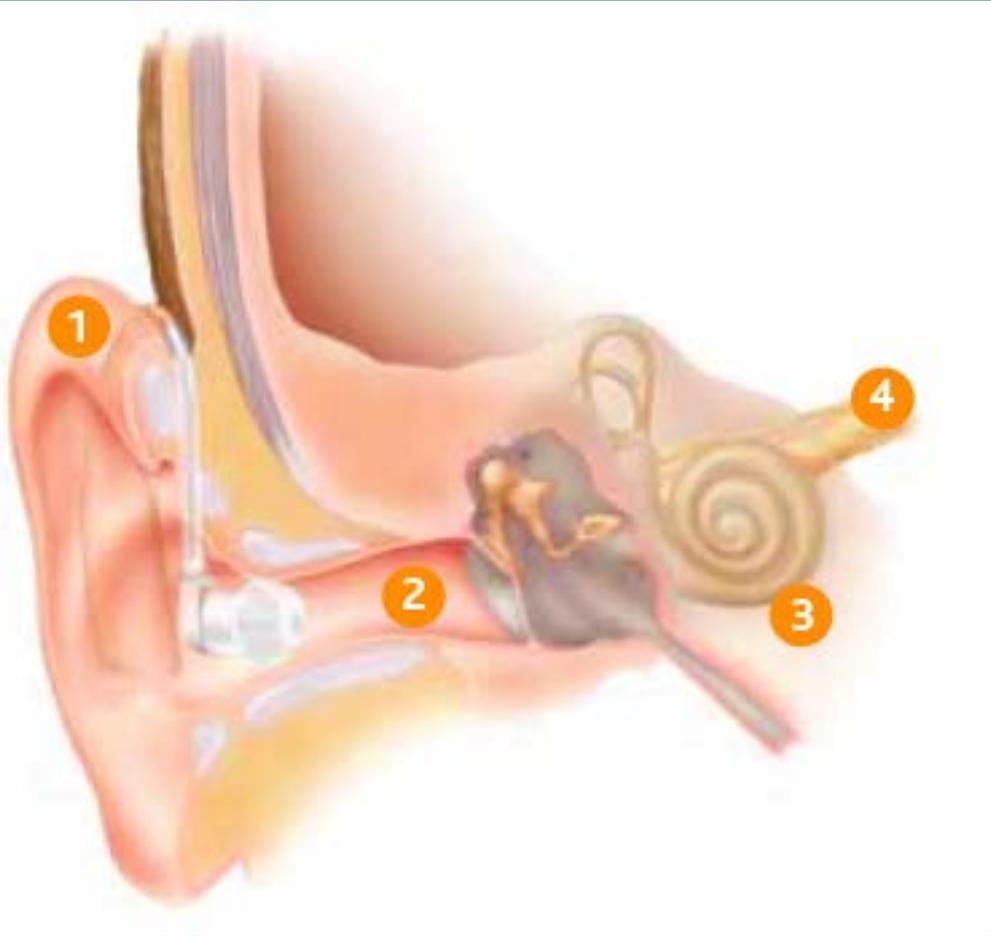
- Hearing aids
- Bone-anchored (bone conduction) hearing aids
- Cochlear implants



# Hearing aids

- Amplifies sound that is being transmitted to the middle ear
- Multiple styles dependent on degree of hearing loss and patient preference
- Can also be used to treat tinnitus
- Components
  - Microphone to pick up sound
  - Amplifier circuitry
  - Receiver (loudspeaker) to deliver sound to ear
  - On/off switch and batteries

# Hearing aids



- Microphone picks up signal → amplifier
- Receiver sends amplified sound down ear canal
- Motion transmitted to cochlea via ossicles and eardrums
- Cochlea sends signal to brain

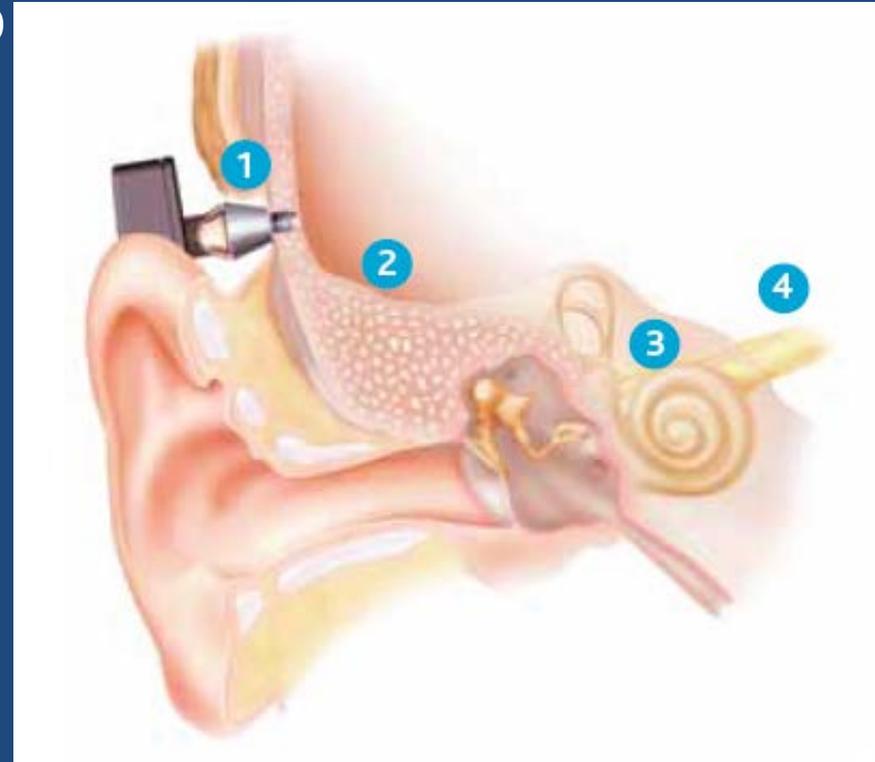
# Bone-anchored hearing aids



- Indications
  - Bilateral conductive hearing loss
  - Unilateral sensorineural hearing loss
  - Mixed hearing loss

# Bone-anchored hearing aids

- Device conducts sound to bone-anchored implant
- Bone transmits sound to the cochlea directly, bypassing external canal and middle ear ossicles
- Sound processed by the cochlea
- Impulses sent to brain to be processed as sound



# BAHA results

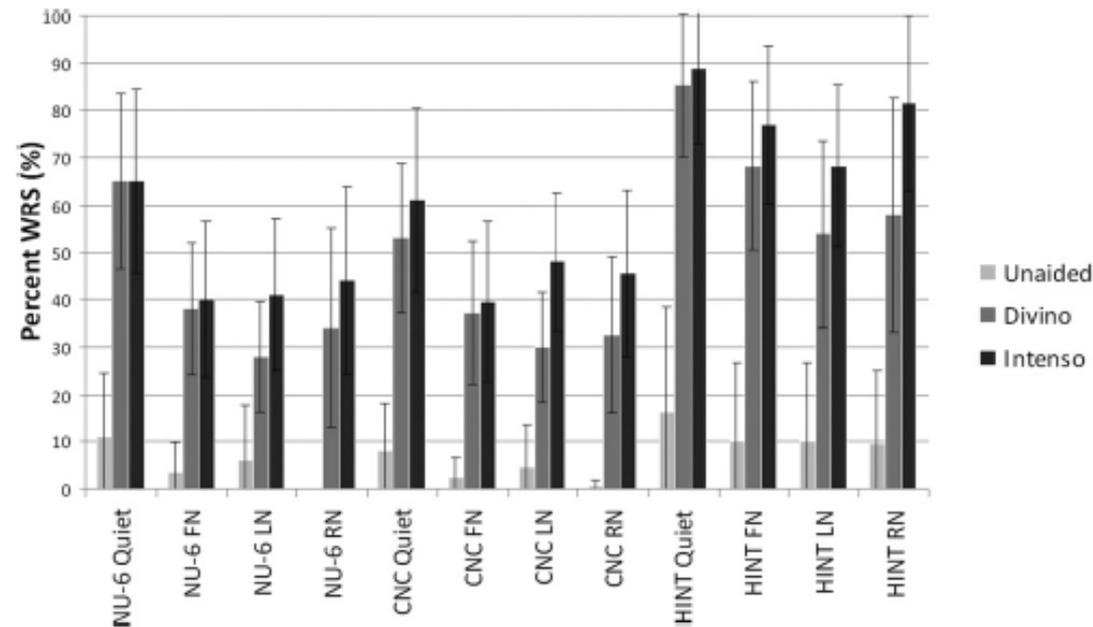
- Even in the most difficult to aid condition (unilateral deafness with moderate hearing loss in the better ear), subjects had better speech understanding and quality of life scores

**Glasgow Benefit Inventory (GBI) scores**

	All patients n = 23	Divino n = 9	Intenso n = 14
Total response score	18	20	17
General subscale	26	28	25
Social support	28	31	26
Physical health	12	13	12
Patients w/improved QOL	21 (91%)	9 (100%)	12 (86%)
Patients w/neutral or negative QOL	2 (9%)	0 (0%)	2 (7%)
Patients would recommend procedure	21 (91%)	8 (89%)	13 (93%)

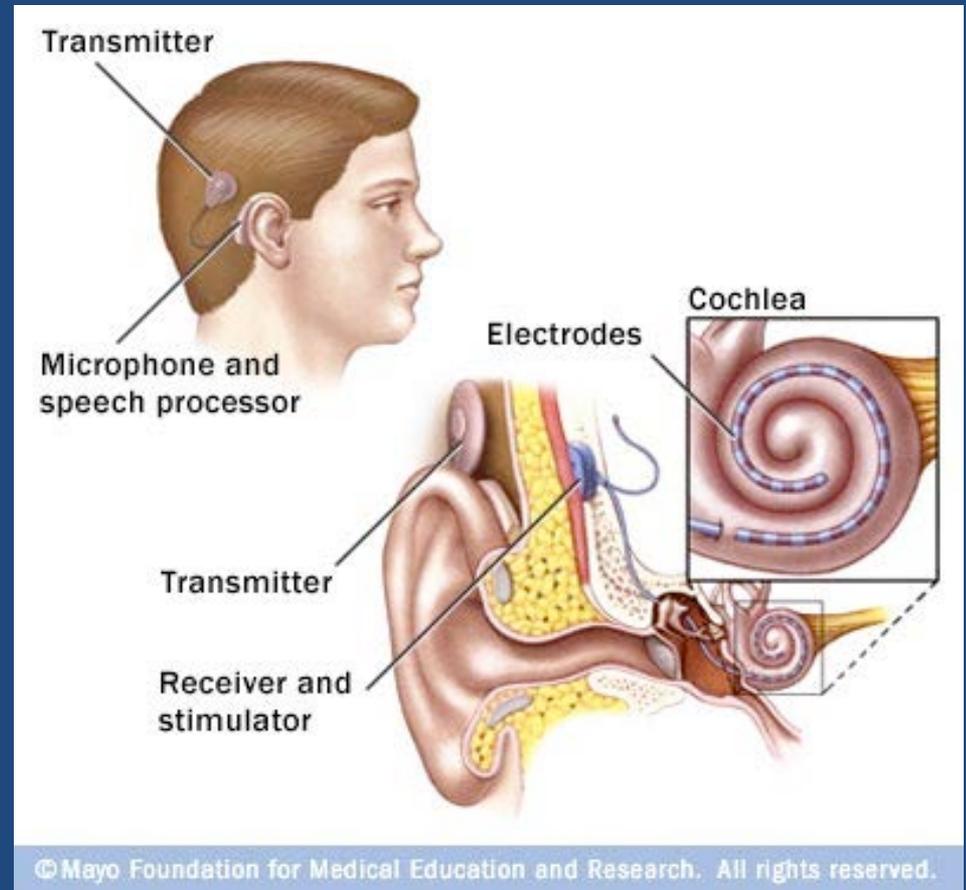
*QOL*, quality of life.

GBI scores range from -100 to +100, with -100 representing maximal negative benefit and +100 representing maximal positive benefit.



# Cochlear implants

- Electronic devices that convert acoustic sounds into electrical pulses that stimulate the auditory nerve directly.



# Cochlear implant candidacy

**TABLE 31-1 -- Criteria for Cochlear Implantation**

	1985	1990	1998	Current
Age	Adults	Adults; children (>2 yr)	Adults; children (>18 mo)	Adults; children (>12 mo)
Onset of hearing loss	Postlingual	Postlingual adults; pre- and postlingual children	Adults and children pre- and postlingual	Adults and children pre- and postlingual
Degree of hearing loss	Profound	Profound	Adults—severe-profound; children—profound	>2 yr old—moderate to profound; <2 yr old—profound
Adult open-set sentences	0%	0%	<40%	<50% in implanted ear; <60% contralateral ear
Pediatric speech scores	NA	0% open-set	<20% (MLNT/LNT); lack of auditory progress	<30% (MLNT/LNT); lack of auditory progress

# What's Changed in CI Selection Criteria?

- Adults
    - Moderate to Moderate-severe-to-profound bilateral SNHL
    - HINT sentence recognition
      - $\leq 50\%$  implanted ear
      - $\leq 60\%$  nonimplanted ear or binaurally
  - Infants (12 – 24 mos.)
    - Profound bilateral SNHL
    - Plateau in development of auditory skills\*
  - Older Children (25 mos. – 17 yrs.)
    - Severe-to-profound bilateral SNHL
    - Plateau in development of auditory skills\*
    - MLNT or LNT word recognition  $< 30\%$  in best-aided condition
- \*given history of appropriate intervention*

# Candidacy for Cochlear Implant

## Best to Worst:

- Post-lingual deafened adult
- Post-lingual deafened child
- Pre-lingual deaf child
- Pre-lingual deaf adult

New evidence show excellent results for early implantation of pre-lingual deaf children

# CI outcomes

- 1988 – First NIH consensus statement – suggested that multichannel implants more likely to be effective than single-channel implants, and 1/20 patients could carry a normal conversation without lip reading
- 1995 – Second NIH consensus statement – “A majority of these individuals with the latest speech processors for their implants will score above 80 percent correct on high-context sentences, even without visual cues.”
- 2008 – Gifford et al. -- >25% of CI patients achieve 100% scores on standard sentence material, need for more difficult material to assess patient performance.

# Summary

- Hearing loss in children is treatable.
- Early intervention is key in giving the child the best outcome possible.
- Identifying risk factors and associations may help in the treatment of the hearing loss
- If there is ever a concern regarding hearing, a referral to audiology for a hearing test and subsequent referral to an otolaryngologist may be warranted.