

USH Talks

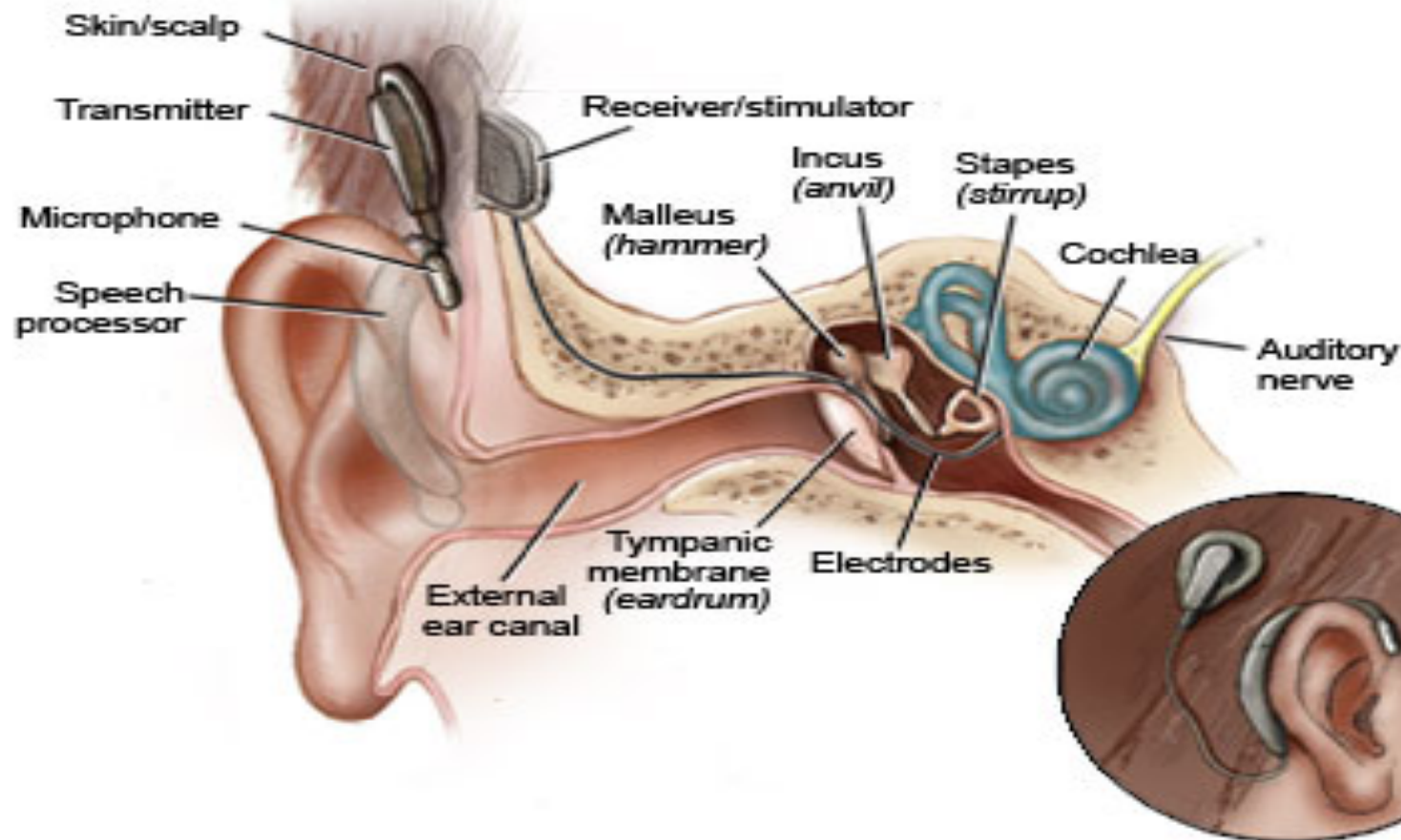
Usher Syndrome and Cochlear Implants

Margaret A. Kenna, MD, MPH

What is a cochlear Implant?

- It is not a hearing aid
- Provides auditory sensation by stimulating surviving neuronal elements in the inner ear
- Supports development/maintenance of spoken language
- Provides environmental awareness

Cochlear Implant



Possible Communication Options for a Deaf Child

- Spoken language
- Manual communication - ASL, SEE
- Combination spoken and manual
- One form of communication does not necessarily mean you cannot use the other

Candidates for Cochlear Implant

- Age 12 months or older; can be earlier
- Bilateral profound sensorineural hearing loss
- Pre-lingually deaf children
- Post-lingually deaf children and adults
- Hard of hearing adults and children (severe to profound)

Are people with Usher Syndrome good candidates for cochlear implants?

- People with Usher Syndrome are good candidates
 - if they meet the hearing loss requirements
 - If they (or their parents if they are children) want a cochlear implant
 - If they want to use spoken language as a communication modality, and they have the capacity to learn it

Which patients with Usher Syndrome are candidates for a cochlear implant?

- Patients with Usher Syndrome type 1, who often are born with profound hearing loss, are most likely to be candidates during early childhood
- Patients with Usher Syndrome who may start out with more hearing but progressively lose their hearing, may be candidates as older children or adults.

Have people with Usher Syndrome received cochlear implants?

- Yes, many have, and many have received lots of benefit
- Even if the patient receives a cochlear implant, especially if they are a child, they will need speech and language therapy
- Although most patients do well, there is a wide range of possible outcomes in terms of spoken language development

Reasons why people with Usher Syndrome may not be good candidates

- There are many reasons why a patient may or may not be a good candidate for a cochlear implant
- Each patient's situation is different, and should be discussed with their cochlear implant team

Reasons why people with Usher Syndrome may not be good candidates

- They are older children or adults who have mainly used sign language their whole life, have not worn or used hearing aids effectively, and have not developed spoken language
- There are medical reasons that could make the surgery risky, or the outcomes after the surgery less predictable

Special considerations for cochlear implants in Usher Syndrome

- There may be effects on balance
- There is some early evidence that patients with Usher Syndrome type 1 using their cochlear implants may actually have better balance

Summary

- Patients with Usher Syndrome may be good candidates for cochlear implants
- Making a definite diagnosis of Usher Syndrome may help in the decision making about a cochlear implant
- Patients and their families should talk to others with cochlear implants, or who decided not to have a cochlear implant, as part of the decision making process

Thank you!!!