Cochlear Implantation for Sensorineural Hearing Loss

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Northeast Ohio Consortium for Children with Cochlear Implants
Objectives

- Discuss the various degrees of hearing loss
- Review indications for cochlear implantation
- Describe the cochlear implant device
- Outline the surgical steps in cochlear implantation
- Discuss postoperative management
- Future trends

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Etiology for sensorineural hearing loss

- Meningitis
- Medications
- Genetic - Ushers, Waardenberg
- Trauma
- Congenital inner ear deformities
- Autoimmune
- Post-natal - NICU, ECMO, anoxia
- Aging-Presbyacusis
Hearing loss in the United States

- Affects approximately 2 million people
- 1:1000 live births
- Potential disadvantages
  - Loss of occupational and educational opportunities
  - Loss of social opportunity
  - Isolation
  - Depression
  - Personal danger

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Newborn screening

- Screening is now universal in Ohio, effective in June 2004
- All newborns have their hearing screened prior to hospital discharge
- Previously only “high-risk” infants were screened, missing nearly one-half of children with significant hearing loss
Newborn screening

- Conducted with automated ABR
- Babies who fail their initial screen are re-screened prior to discharge
- Babies who fail their re-screen in both ears are scheduled for a diagnostic ABR and OAE evaluation prior to discharge
Newborn screening

- Children who are identified with hearing loss and enrolled in an appropriate rehabilitation program by the age of six months develop age appropriate language in early childhood and maintain these skills at least through entry in to school.

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Anatomy

1. Sound waves
2. Ear canal
3. Eardrum
4. Middle ear
5. Cochlea
6. Hearing nerve
Histopathology of SNHL

- Advanced hearing loss related to hair cell loss with reserve of viable nerve fibers
- Average hair cell counts of 14,000 in deafness (normal 35,000 to 40,000)
- 65% of surviving hair cells are found between 6 to 22 mm from the round window
- 10,000 neurons with 3,000 apically necessary for speech discrimination
Normal histology
Damaged hair cells
What is a cochlear implant?

- Converts sound energy to electrical energy
- Microphone
- External speech processor
- Signal transfer hardware
- Subdermal receiver
- Implantable electrode
How the implant works
History of cochlear implants

• 1950’s Djourno & Eyries
  – Direct stimulation of the auditory nerve with an electrode

• 1960’s House & Urban
  – Developed a single electrode cochlear implant for stimulation of postlingually deaf adult
History of cochlear implants

- 1970’s
  - Research showed that intraco cochlear electrodes could produce sound awareness via spiral ganglion cell stimulation despite deterioration of the organ of Corti
History of cochlear implants

• 1980’s
  – First successful implantation of 3M-House single-channel device and Nucleus 22 multi-channel device (tonotopic stimulation)

• 1990’s
  – FDA approval of Nucleus 22 channel device for children—once language acquisition in pre-lingual children was proven

• 150,000 cases performed to date worldwide
Indications for cochlear implantation

• Infants (12 - 24 mos.)
  – Profound bilateral SNHL
  – No benefit from trial of hearing aids
  – No medical contraindications
  – High motivation and appropriate expectations from family
  – Earlier implantation if SHNL secondary to meningitis
Indications for cochlear implantation

- Older Children (25 mos. - 17 yrs.)
  - Severe-to-profound bilateral SNHL
  - No benefit from trial of hearing aids
  - Word recognition scores less than $\leq 30\%$ in best-aided condition
Indications for cochlear implantation

Adults

- Prelinguistic or postlinguistic onset of severe to profound hearing loss
- HINT Sentence recognition
  - ≤ 50% implanted ear
  - ≤ 60% nonimplanted ear or binaurally
- No medical contraindications
- A desire to be part of the hearing world
Ethics of cochlear implantation

• From the medical point of view, CI is an effective treatment option for a severe disability
• However, to some deaf activists, it represents unwanted technology that threatens their way of life
• Most viable ethical and moral arguments have resolved once pre-lingual patients shown to develop normal language skills
• Cultural arguments still exist in Deaf community
Preoperative planning

• Evaluation by otolaryngologist
• Audiology & speech pathology involvement
• Developmental psychologist
• Genetic testing - Connexin 26
• CT temporal bone
  – Cochlear abnormalities
  – Middle ear/mastoid inflammation
  – Position of jugular bulb/carotid
  – Internal auditory canal width
Cochlear implant systems

- Nucleus 24 Freedom
- Med-El (Medical Electronic) Cochlear Implant
- Clarion 90-K
Nucleus Freedom Contour Advance

- Cochlear, Ltd. In Sydney, Australia
- Highlights:
  - Rounder – easier to implant
  - Ease of electrode insertion
  - Strong Titanium case
  - Smaller – less drilling
  - Removable Magnet MRI compatible up to 1.5 tesla
  - Self curling, protective of delicate structures
Nucleus Freedom Contour Advance

• Self-curling electrode array
• 22 half-band electrodes
• Adjacent to the inner wall of the cochlea
• Cast *pre-curved* to regain pre-designed shape & size
Clarion

- Advanced Bionics of California
- Clarion CII Bionic Ear
  - Platinum digital sound processor
  - CII BTE digital microprocessor
Med-El cochlear implant

- Manufactured in Innsbruck, Austria
- Offers three types of cochlear implants:
  - Standard array
  - Split electrode array
  - Compressed array
“This is not brain surgery”
Incisions for cochlear implant
Planning the incision
Incision exposing bone
Mastoidectomy
Cochleostomy
Cochleostomy
Drilling the well for the implant
Core design concept

The electrode array is easily inserted with the help of a malleable stylet, as shown below:

1. Stylet holds the array straight prior to insertion.

2. Stylet is removed after insertion.

3. Array curls.
Electrode array in cochlea
Stylet withdrawal
CUT THE BOVIE!!!!
Electrode sewn into bone
Post operative skull film
Surgical considerations

- Cochlear dysplasia
  - Variable postoperative speech perception results
  - Risk for intraoperative complications
- Aberrant facial nerve
- Intracochlear ossification
Outcomes

• More than 95% of Deaf children implanted at 1 year obtain open-set speech understanding....Normal language

• Earlier implantation results in greater gains in speech perception

• Approximately 70% of post-lingual deaf CI patients may use the telephone
Cost of cochlear implant?
Cost of cochlear implant?
Post-operative expectations

- Post-operative anesthesia care
- Home going concerns
- Questions families may ask
Wound care

- Mastoid dressing remains for ~2-3 days
- Patient may shower once dressing is removed
- Absorbable sutures
Questions from families

• When will the patient be able to hear?
  – The implant is activated 3-4 weeks after surgery by the audiologist

• What type of activities are allowed?
  – No heavy lifting 2 weeks after surgery
  – Precautions during contact sports – helmet
  – Remove CI for water sports, showering
Questions from families

• Is it OK to have an MRI?
  – Only if there is a removable magnet such as Nucleus 24 device

• Should there be precautions for future surgery?
  – *Monopolar* cautery should NOT be used
  – Biopolar instruments may be used but keep away ~1 cm from implant
Potential complications

- Wound complications
  - Infection
  - Hematoma
  - Wound dehiscence
  - Skin flap
  - Necrosis
- Facial nerve injury
- Exposed electrode
- Device failure
- Perilymph fistula
- Meningitis

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Meningitis

• US Food and Drug Administration identified a possible association between cochlear implants and bacterial meningitis in 52 people worldwide
  - Ages 21 months to 72 years
  - Onset < 24 hours to > 5 yrs after implantation
  - Majority of organisms S. pneumoniae
Meningitis

• Etiology
  – Nidus to a local infection in patients with bacteremia
  – Preoperative otitis media
  – Immunodeficiencies
  – CSF leak

• Immediate workup and treatment

• Vaccination
Future Developments

- Smaller devices
- Totally implantable cochlear implant
- “Short implants” for high frequency hearing loss
- Bilateral cochlear implantation
- Stem cell work
- Gene therapy
Electric-Acoustic Stimulation (EAS)

CAUTION: Investigational device. Limited by law to investigational use
Indication Areas for HA, CI and EAS

- No amplification necessary
- Hearing aid
- EAS indication
- Relative CI indication

N = 23521

Courtesy W. Gstöttner, J. Kiefer, Frankfurt

• CI indication
New FLEX eas Electrode

- Improved mechanical flexibility
- Requires less insertion force leads to less insertion trauma
- Thin front-end with 5 single contacts 1.9mm contact spacing
Insertion Depth in EAS?

• How deep should the electrode be inserted for EAS?
• Deep insertion, covering both the functioning and the non-functioning region of the cochlea?
• Insertion depth covering most of the non-functioning region (abt. 18-20 mm)?
• Insertion only in the straight part of the basal region of the cochlea (abt. 10 mm)?
• Insertion only in the very basal part or only extra-cochlear stimulation (6 mm / extra-cochlear)?
Clinical Experience
Frankfurt Pilot Study

- Residual hearing was preserved in 18/21 patients
  - 13 (0-10 dB)
  - 5 (11-20 dB)
  - 3 hearing loss
Results

Pre- and Postop thresholds for subjects where at least partial hearing was achieved

Gstöttner, Adunka, 2004
Results

Average speech discrimination scores in EAS subjects

Gstöttner, Adunka, 2004
EAS – External Devices

- Two independent devices
- Different number / sizes of batteries
- Different battery life-time
- ITE hearing aids at their limits
Design of a Combined EAS-Device

- Combined power supply
- Combined input signal
- Independent compression (AGC)
- Independent fitting
Combined EAS-Device

- Simplified handling
- Increased user comfort
- Higher amplification than ITE HA
- Flexibility to adapt to EAS learning process
MED-EL EAS Clinical Studies

- www.hearingpreservation.com
- Frankfurt study (completed) - single-site to explore feasibility, surgical techniques and electrodes
- EAS Europe Multi-Center study (n=30) (ongoing) – to verify efficacy of electric/acoustic stimulation product for EU market
- EAS US Multi-Center IDE Clinical Trial (IDE FDA approved) – to establish efficacy/safety for US market
Fully Implantable Cochlear Implant

• Technically: much more complex than conventional cochlear implants

• Surgically: complex but not prohibitively complex
Fully Implantable
Microphone Design Options

• Acoustic attenuation of the signal as low as possible
• Microphone located under the skin
• An ear-canal microphone
Batteries

- **Design:** 1 day per charge
  - Cycle life as long as possible
  - Charging period as short as possible – patient will wear a small external charger for less than one hour a day
  - Batteries should not lose significant capacity over time
Bilateral

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US Adult Bilateral Cochlear Implantation Study

N=19 at 12 months

13 showed all 3 effects (bilateral advantage)

Head shadow (19), squelch (15) and summation effects (17) were significant
New Adult Bilateral Studies

- Loudness summation and AGC settings effects
- Extension of SPECT scan study with bilaterally implanted patients
- Evoked potentials in bilaterally implanted patients
- Psychophysical measures
Pediatric Bilateral Study

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Status

• Bilateral vs unilateral performance longitudinally
• Speech/language perception and production over time
• Localization
• Speech in noise
Drug Delivery Systems

Not currently available
Developments in Inner Ear Biology

- New drugs/biological treatments
- Genetics
- Stem cell implantation
- Study of the process of apoptosis
Drug Delivery Systems

- **Goals:**
  - Preserve spiral ganglion cells
  - Prevent apoptosis
  - Reduce inflammation and tissue growth
  - Maintain residual hearing

- **Method:**
  - Combine inner ear drug delivery with electrical stimulation in CI patients
The interface between drug delivery and the cochlear implant

- C. Jolly
- G. Reetz
- F. Béal
- F. Aattendorf
- W-D. Baumgartner
- J. Miller

Not currently available
Implant System concept

catheter

Implant housing

refillable pump

connector

2 twin outlets

electrode contacts

refillable pump

catheter

Implant housing

connector

2 twin outlets

electrode contacts

Drug delivery channel

Spiral ganglion cells

Scala vestibuli

Scala tympani

Electrode
Cross section

- Wire
- Contact
- Drug delivery channel
- Silicone
- Double outlets
ELECTRODE DESIGN
(Based on FLEX Technology)

outlets

8 mm

central channel

outlets
Modular system is primed and connected intra-operatively.

- Electrode array
- Implant
- Catheter and needle
- Pump

Pump and catheter are to be removed after therapy.

Embedded micro-port and septum.
Conclusions

- Neurotrophic factors preserve spiral ganglion cells and reduce hearing threshold in animal studies.
- A fluid-based drug delivery system is technically feasible.
- Modelling of drug diffusion shows that with two outlets, we achieve a more uniform drug concentration in the cochlea.
- At 0.5 µl/hr, pressure increase is negligible.
- Fluid-based drug delivery offers more flexibility than surface coating and can be terminated.
Summary

• Cochlear implants are an appropriate sensory aid in deaf children and adults who receive minimal benefit from conventional amplification
• Future and ongoing research is promising
Thank You!