Medical Considerations for Children & Adults when Determining Cochlear Implant Candidacy

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Disclosures

• Consultant for
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  – Advanced Bionics Corporation  AB
  – Applied Genetics Technologies Corporation  agtc

• Research support
  – MED-EL Corporation  MED\textregistered EL
  – Cochlear Corporation  Cochlear
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• Ownership
  – Advanced Cochlear Diagnostics  ACD
Objectives

- Review Pediatric SNHL
  - Clinical algorithm
  - Audiometric testing
  - CMV
  - Genetic testing
  - Imaging
    Normal and malformed anatomy

- Review Adult SNHL
  - Residual hearing
Basic Assumption

Heterogeneous group of disorders
- Variety of causes
- Variety of auditory perceptual abilities
- Individualize treatment
- Expect variable outcomes
- Most children non-syndromic
- Mostly recessive genes

One size does not fit all!

Current algorithm focuses on the level of hearing loss
Causes of SNHL

Prelingual deaf children
1/1000

- Idiopathic 25%
- Nongenetic 25%
- Genetic 50%

Nonsyndromic 70%

- Autosomal recessive 75% – 85%
- Autosomal dominant 15% – 24%
- X-linked 1% – 2%

Syndromic 30%

- DFN1 50%
- Other DFN 50%
Evaluation of Pediatric SNHL

• Complicated clinical algorithm
• Multiple specialties
• Goal: Early hearing decision!

• Problem: CI compromises residual hearing
Clinical Algorithm

- **Make the diagnosis**
  - Single polarity click ABR, OAEs

- **Image early (MRI)**
  - Temporal bone, CNS, cochlear nerve!!

- **Observation until VRA possible**
  ~7-9 months, can vary

- **Behavioral testing**
  - Normal testing: f/u, repeat ABR?
  - Abnormal testing: **amplification**

- **Intensive auditory-based therapy**

- **Failure of speech perception and language**
  - Consider **cochlear implantation**
Timeline

- **Birth**
  - Newborn infant hearing screening
  - Diagnostic ABR (ASSR, OAE)
  - History & Physical
  - Medical Evaluation
  - EKG
  - Imaging
  - Genetic Testing
  - Early intervention Services
  - Auditory-based Therapy
  - Initiation of HA Trial

- **Day 1-7**

- **2-4 Months**

- **6-9 Months**
  - Behavioral audiometric testing
    - Confirmation of Thresholds
  - Auditory-based Therapy
  - Consider CI Evaluation
  - Evaluation of HA Trial

- **11-14 Months**
  - Cochlear Implantation/HA?
  - Auditory-based Therapy
Factors that Delay

**Auditory**
- Delay in diagnosis
- Significant residual hearing
- Fluctuating hearing
- Unreliable or conflicting test results
- ANSD
- Inappropriate amplification

**Speech Development**
- Good progress despite profound HL

**Parental issues**
- Missed appointments
- Don’t wear devices
- No educational buy-in
- Socioeconomic

**Multiple Challenges**
- Cerebral palsy/Autism

**Medical**
- Anatomic uncertainty
  - CN deficiency
  - Severe inner ear malformation
Reynell Developmental Scores

Children with normal hearing
Mean trajectory
- Age <18 mo (n=28)
- Age 18-36 mo (n=48)
- Age >36 mo (n=21)

Children with cochlear implantation
Mean trajectory
- Age <18 mo (n=72)
- Age 18-36 mo (n=64)
- Age >36 mo (n=52)

Comprehension scores

Expression scores
Essence of the Problem

Destroy Residual Hearing

Earlier is Better
Hearing Evaluation

- **ABR**
- **ASSR**
- **OAEs**
- **Behavioral testing**
  VRA, CPA
- **Speech evaluation**
  Surrogate marker of signal quality

- Multi-test evaluation rather than one single test
  - Requires a comprehensive team discussion!
ABR Timeline

• No actionable data
  – Following two attempts
  – Child ≥ 3 months
  Proceed with sedated study!!

• National goal (JCIH): HA fitting by 6 months
  1-3-6 rule, NCH goal: HA fitting by 3 months
  in 2016: average HA fitting of 3.4 months!
Hearing Aid Fitting

- **Exact** science
- Avoid ear **plug**
- Avoid **NIHL**
- Adequate **amplification**
- Regular **checks**
Otologist Issues

• **Surgically** Implantable?
  – Normal or elevated risk?
  – Vestibular compromise?
  – Meningitis

• Once implanted, will it be successful?
  – CN VIII
  – Inner ear anatomy
  – Cognitive function
  – Device retention
    Will the child wear the device?
  – Family buy-in
    Educationally committed parents?
Genetic Testing

• Screening for most common defects
  GJB2, Connexin 26, 30

• Problems
  – Many hearing loss genes
    Financial limitations
  – Genotype/phenotype correlations not always accurate
  – Comprehensive evaluation not replaced
    Imaging, Audio, Speech
CMV-Related Hearing Loss

- **Most common** non-genetic hearing loss
  - > 20% of all congenital HL
  - > 25% of all HL by age 4
- **Wide clinical spectrum**
  - **Severity** of hearing loss
  - **Uni- or bilateral** involvement
  - Often **progressive**
- Difficult diagnosis
- Often hearing loss as the **only symptom**
Imaging

• **CMV** lab testing difficult
  
  **Timing**
  
  – 60% no clinical CMV
  – 70% bilateral
  – 90% profound
  – Vast majority have “non-specific” MRI findings
    • Significantly more common than other HL kids

- Dilated ventricles
- Lissencephaly
- Gyral anomalies
- Paraventricular cysts
- Cerebellar hypoplasia
Treatment of CMV Hearing Loss

- Gancyclovir
  - Treats systemic CMV infection
  - Not approved for hearing loss alone
    …but in clinical trials
  - Potential toxicity
- CMV-Immunoglobulin
- Cochlear implants work in these children!

- Prevention
  - Gancyclovir
    …may prevent HL in multiply involved children
  - Vaccination and newborn CMV screening in trials
    …as universal newborn infant screening administered by governments
Normal Anatomy

- Head of malleus
- Cochlea
- Cochlear aperture
- Body and short process of incus
- Facial nerve
- Lateral SCC
- Vestibule
- IAC

Vestibule, Cochlea, IAC

CN VIII, Cerebellopontine angle cistern
Embryology of Anomalies
Incomplete Partitioning Spectrum
Hypoplastic Malformations
Cystic Anomalies
Adult CI Medical Considerations

• **Aided** speech testing crucial

• Hearing preservation and combined stimulation probably more applicable
  Hybrid, EAS

• Mostly **CT**
  …although **MRI** has been demonstrated to be beneficial in adults
Conclusions

• Pediatric HL requires **large team approach**
• Multiple **professional groups**
  – Physicians
  – (Specialized) Audiologists
  – SLPs, AVTs, Educators
• Early **amplification & proper evaluation**
  *Physician plays a central role*
  – Early cochlear implantation if indicated
• Adults: proper evaluation of **residual hearing!**
Thank you!

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When your child needs a hospital, everything matters.