Pediatric Hearing Loss

Moderator
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Panelists
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Objectives

• Identify causes of pediatric hearing loss
• Understand universal hearing screening
• Understand the role of CMV and hearing loss
• Understand the work up of type of different types of hearing loss

Case 1
3 month old male

Ex 25 week premie
Exchange transfusion
Cardiac surgery
Referred both ears
Panel

- Work up of micro premies with hearing loss
- When to consider implants
- Should other diagnoses be worked up
Case 2

• 4yo AA female
• Sudden hearing loss
• Failed but then passed newborn hearing screen
• History of airplane flight
• “Can’t Hear” in PMD
Case 2

- Labs drawn
- High dose steroids
- MRI/CT
- Timing of repeat audio?

Imaging

- What imaging do you obtain?
- CT vs MRI
  - CT better for bone, EVA
  - MRI with no radiation exposure in infant
  - Both obtained for pre cochlear implant

Sudden Hearing Loss in Children

- Not a lot of data
- High dose steroids
- Must consider Enlarged Vestibular Aqueduct Syndrome
Steroid use

- Use in pediatric population
- Length of time
- Lab to check

Audio in one week
Case 3
Newborn failed hearing screen

• 36 week male, history negative
• referred in left ear at birth and at two 2 weeks
CMV positive

- Start valgancyclovir
- Refer to genetics
- Refer to infectious disease
- amplification

Protocol for referred newborn hearing screen

- 1:3:6 rule
  - Screening by one month old
  - Diagnosis by 3 months
  - Intervention by 6 months
- CMV needs to be treated with the 4-6 weeks

CMV

- Congenital vs acquired
  - Test for IgG, Ig M,
- Testing
  - Urine vs saliva
- Treatment
  - Oral valgancyclovir
    - May have bone marrow suppression
CMW hearing loss

- MRI of Brain
- Infectious Disease Consult
- Stigmata include brain calcifications, rash,
  – Ocular disease

valgancyclovir

- Oral
- 6 weeks to 6 month treatment plan
- Lab check for bone marrow suppression
- Can halt progression and/or lead to some recovery

Case 4

- Newborn with failed hearing screen
Components of hearing loss screen

- Vary at institutions
- CHW initially with screen targeting Connexin GJB 2,6, SLC64A
- Now GSPMC Hearing Loss Advanced Sequencing Panel with 157 genes

Testing

- EKG
- Ophthalmology exam
- Cbc
- Genetic screen
- Imaging
Common Syndromes

• Jevell Lange Nielsen
• Usher’s Syndrome
• Alport Syndrome
• Pendred’s Syndrome

Timing of tests

• When do you image
• Early eye exam
• EKG

Case 5

• 5 yo healthy male
• Failed school screen
• Audio shows unilateral mild hearing loss
Case 5

- Not a lot of data
- Advocate hearing aid use may be of limited benefit
- Important to monitor for progression

Case 6

- 12 month old with unilateral profound hearing loss
• Most likely to have positive finding on imaging
  • Eye exam
  • Genetic work up

Case 7
• 5 year old with failed hearing screen
• Failed but then passes hearing screen
• History of poor speech
• Audio shows bilateral moderate sensorineural hearing loss
Imaging shows bilateral enlarged vestibular aqueducts

- Proceed with other testing
- Counseling
- Consideration for cochlear implant
Enlarged vestibular aqueduct

• Did well with hearing aides
• At age 8, in MVA and progressed to bilateral profound loss
• Underwent bilateral cochlear implants

Summary

– Follow the 3:6:12 rule for hearing screening in newborns
– Consider CMV early in newborn hearing loss
– Imaging options
– Full testing with moderate to profound losses
– Tailor other testing options for mild and unilateral losses