Relevant Financial Relationships

• All authors are employed by Mass Eye and Ear

• There are no financial conflicts of interest
Cytomeglovirus (CMV) is the most common infectious cause of birth defects in the United States, affecting 1/200 infants

- 10-15% symptomatic Congenital CMV (cCMV)
  » Includes microcephaly, IUGR, jaundice, cerebral palsy, seizures, failure to thrive, cognitive delays, learning issues, vision and hearing loss.

- 85-90% asymptomatic cCMV (including isolated hearing loss)
  » 15% of asymptomatic cCMV babies will develop hearing loss
In the United States

• No consensus on how to treat asymptomatic cCMV (isolated hearing loss)

• Individual hospitals are developing their own protocols
cCMV – Treatment with Valganciclovir

- Symptomatic CMV, automatically treated with Valganciclovir (6 mos)
  - To improve neural transmissions

- Asymptomatic CMV (isolated hearing loss)
  - Parents are given information about the antiviral treatment and it is their decision to proceed or not
  - 6 week course to start. If baby is doing well, continue for 6 months
Mass Eye and Ear/Mass General Hospital

cCMV Protocol for Isolated SNHL

• MEE oversees the newborn hearing screening program at MGH
  – Since 2014, at MGH, every baby that refers on the hearing screening is automatically screened for CMV
  – If the baby is CMV positive
    • Threshold ABR and Otology/Infectious Disease within 2 weeks. Treatment with Valganciclovir is initiated ASAP.
Case 1  *20 years ago*
Prior to routine antiviral treatment

Age 5 months, first ABR

Age 17 months, fit with hearing aid right ear
Case 1  *20 years ago*

- 27 months - both ears decreased

2.5 years – profound bilateral
Case 1 *20 years ago*

History

- Documented speech and language delay related to history of hearing loss.
- 3 years: Cochlear implant for the right ear.
- 7 years: Cochlear implant for the left ear.
- Patient is now attending college and continues to receive excellent benefit from her bilateral cochlear implants.
  - Last testing in 2017
    - CNC (binaural CI) – 86% whole word, 94% phonemic
Case 2  *6 years ago*

Left ear refer on NBHS, cCMV+
ABR – 2 weeks old, started antivirals

8 months – ended treatment
Hearing improved bilaterally
Case 2 *6 years ago*

Ages 14 – 28 months, stable hearing

Age 3 years – left ear decrease, 2nd round of antivirals
Case 2 *6 years ago*

Age 3.5 years- HA fit to right ear, Rx: CI left
Word recognition: CNC-72% right; ESP Cat 2 left

Age 4-5 years, hearing stable.
CNC-80% right; ESP category 2 left.
Uses right hearing aid; left CI recommended
Case 2 *6 years ago*

5 years - CNC right ear: 78% to 2% in 3 months. ESP Cat 2 on the left ear.

- Age 5.3 years, received CI for the left ear
- Continues to use hearing aid in the right ear.
- Speech and language development has been on target. Currently, attending a mainstreamed preschool class with support from teacher of the deaf.
- Good benefit from the left CI.
  - CNC – 70% whole word, 87% phonemic
- CI for the right ear is currently being recommended.
Case 3  *2 years ago*

- Right ear refer on NBHS
- ABR at 9 days old

* 13 days old – started antivirals
* ABR at 4m – left ear improved
Case 3 *2 years ago*

- 9 mos – stable hearing

* 18 mos – stable hearing
Case 3  *2 years ago*

- 21 mos – decreased hearing both

- Cochlear implant for the right ear is scheduled for 7/30/2019.

- Discussed hearing aid for the left ear.

- Monitoring audiograms- every three months or if a change is suspected.

- Continue with Early Intervention.
  - Speech and language development is currently on target.
Case 3  *2 years ago*

- 26 mos.
- Mother stated concern about decreased hearing.
- Tested on 7/3 and on 7/8
- Left hearing aid will be fit on 7/15
- Right CI is scheduled for 7/30/2019
Discussion

• Prior to routine anti-viral treatment for asymptomatic cCMV babies:

  – Rapid progression of hearing loss, severe to profound by age three, noted for our small population.

  – Hearing was not monitored as closely, adversely affecting speech/language development.
Discussion

• With anti-viral treatment for asymptomatic cCMV babies:

  – Prolonged periods of normal hearing in at least one ear allows consistent access to speech sounds.

  – Frequent monitoring is needed. Testing every 2 months.
Conclusions

• GOAL: CMV testing should be incorporated into Universal Newborn Screening Program protocols.
  – Massachusetts is working towards universal CMV screening

• Prompt fitting of amplification is indicated in cases of unilateral hearing loss.

• Earlier implantation of poorer ear should be considered