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OTTAWA
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2021



AUGUST 22ND  AUGUST 24TH

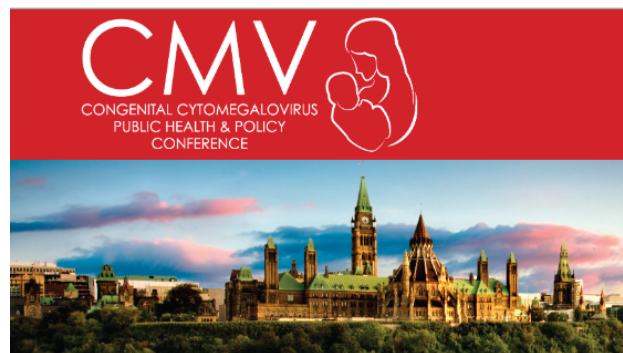
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Parent Story: Jasmine Webster

Hearing and Congenital CMV: Overview of Screening, Diagnosis and Interprofessional Care for Infants and Children

Albert Park, MD
Angela Shoup, PhD



Objective:

- Impact CMV on Hearing
- Screening and diagnostic tests for hearing
- Importance of a team approach
- Surveillance Pearls

Sensorineural Hearing Loss and CMV:

- Most common sequelae from cCMV- 12.6%
- Most common cause of nonhereditary SNHL
- May account up to **20%** pediatric SNHL
- CMV Screening

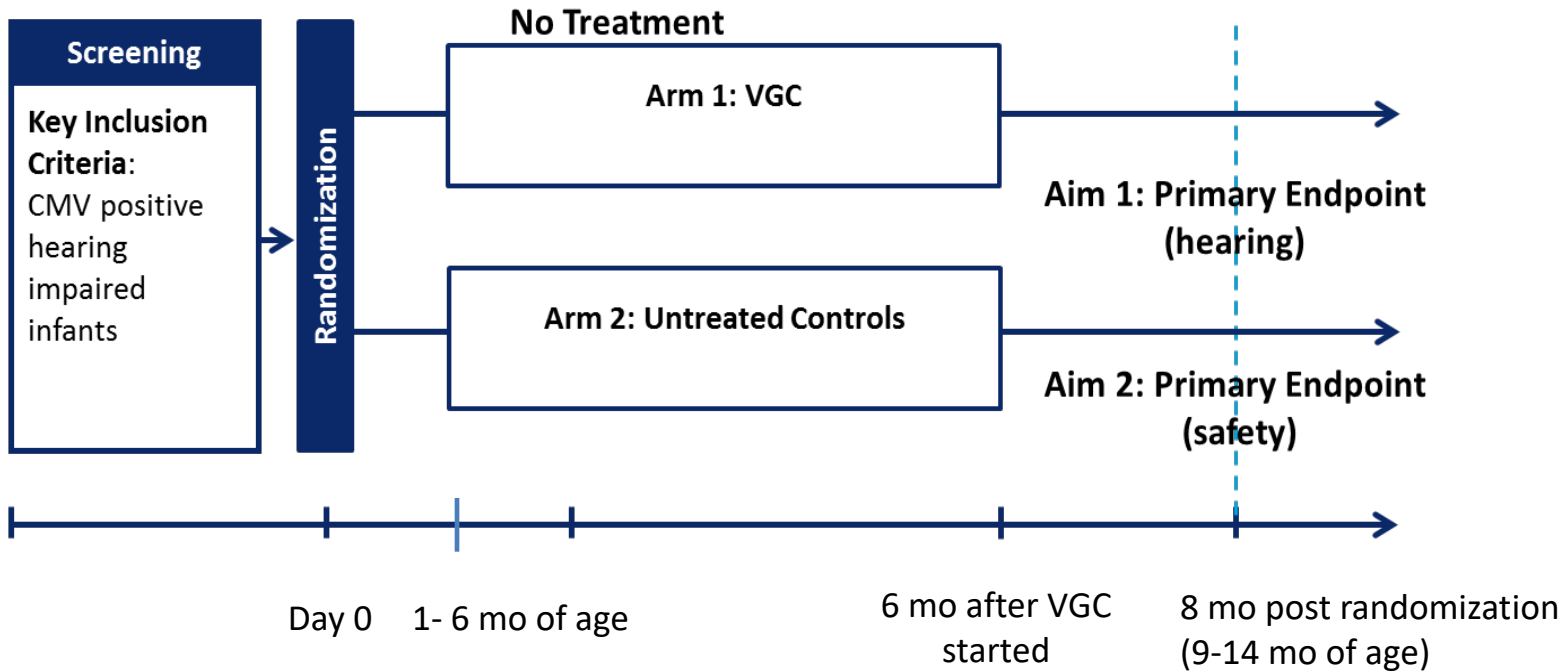
Goderis et al. Pediatrics 2014; Diener et al
Pediatrics 2017

NIH Valganciclovir Ear Trial:

- **Aim 1: Compare the hearing and language outcomes of cCMV-infected with isolated hearing loss treated with VGCV to untreated infants via a multi-institutional double-blinded placebo controlled clinical trial.**
- **Aim 2: To evaluate the safety of antiviral VGCV therapy for cCMV-infected infants with isolated hearing loss.**
- **Aim 3. Evaluate the pharmacokinetics of valganciclovir using pharmacometric modeling to develop a population pK model.**

Study Design:

Asymptomatic cCMV Hearing Impaired Infants Randomized to Valganciclovir and

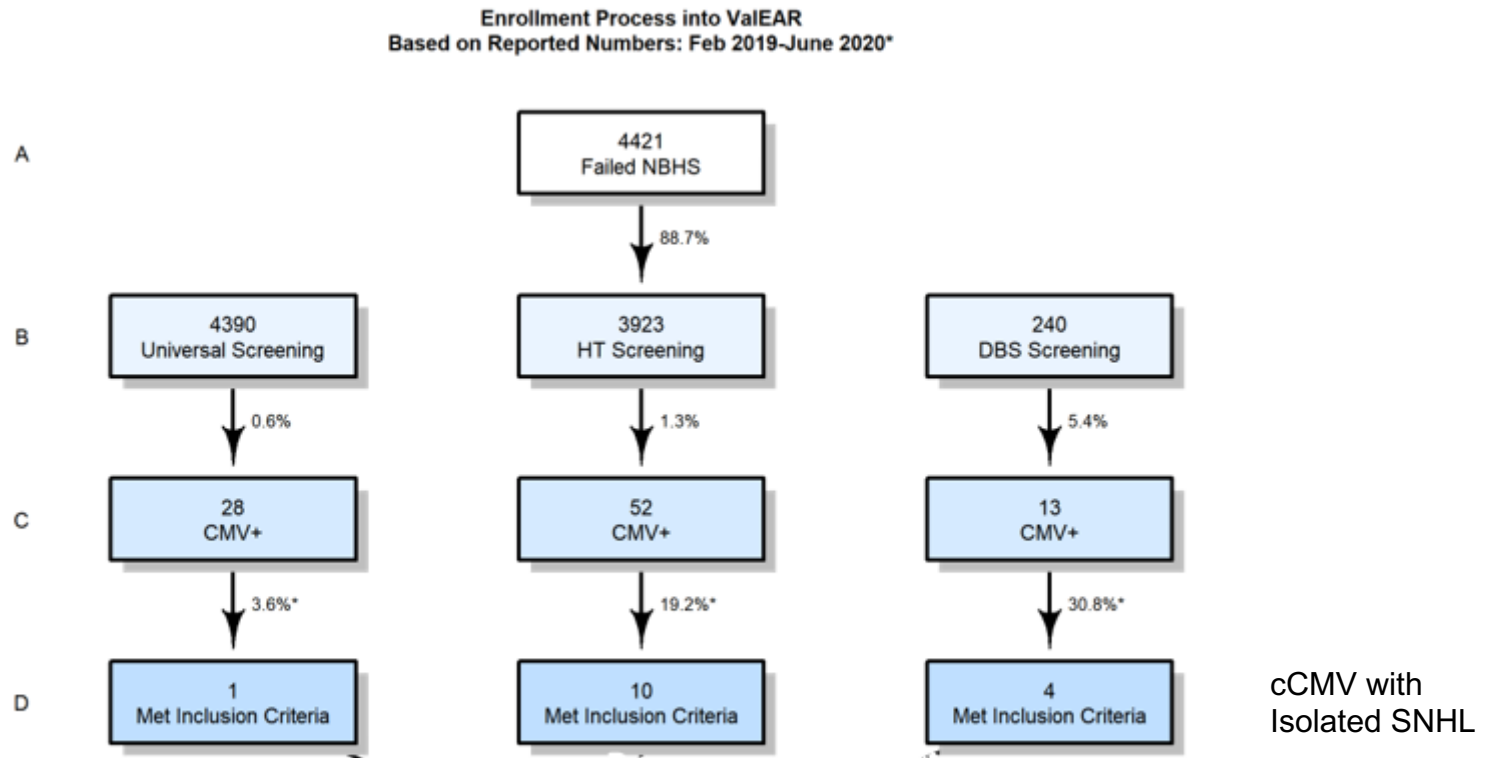


Aim 3: Pharmacokinetics

Over Thirty Institutions Started HT-CMV Screening!



Three Common Screening Approaches:



Any institution or hospital can implement an early CMV screening program

Expanded Targeted Screening Approach:

Failed hearing screen

Maternal CMV infection

SGA

Unexplained hydrops

Intracranial calcifications

Intrabdominal calcifications

Elevated transaminase

Elevated direct bilirubin

Petechial rash

HUS imaging suggestive CMV

Unexplained
thrombocytopenia

Unexplained HSM

Results from Expanded Targeted Screening:

- 754 of 85,553 (0.8%) newborns underwent CMV testing based on expanded targeted screening
- 21 (2.8%)/754 tested positive CMV
- **14 cases sCMV/100k**
- 4.2 cases cCMV/100k – no screening process
- Expect 26 sCMV/100k based on 0.3% incidence and 10% sCMV rate

Sorichetti et al. J Ped 2015; McCrary et al. Outcomes from an Expanded Targeted Early CMV Screening Program. J Peds Inf Dis. In press

Case:

- 2 mo cCMV infant with right moderate and left mild SNHL (isolated SNHL)
- How often should you recheck this child?
- If the child is not cooperative, what frequencies and which ear should be prioritized?

Methods:

- Asymptomatic cCMV patients with isolated SNHL were selected for the study*
 - 2 populations: congenital/early-onset or delayed-onset
- Audiologic assessments (ABR, behavioral audiometry, and tympanometry) were conducted by trained audiologists blinded to CMV status
- Hearing loss by ABR defined as >25 dB for click or any tone burst frequency at 1, 2, or 4 kHz
- Hearing loss by behavioral testing was defined as >20 dB at 1, 2, or 4 kHz
- SNHL was analyzed by laterality, ie better- and poorer-hearing ear
- Hearing change by ear was analyzed at 12 months and 18 years of age
- Clinically significant worsening in hearing >10 dB in click ABR, >15 dB in 1 frequency, or >10 dB change in 2+ frequencies

Torrecillas V et al. Should you Follow the Better-Hearing Ear for Congenital Cytomegalovirus Infection and Isolated Sensorineural Hearing Loss? Otolaryngology- HNS Journal, 2019.

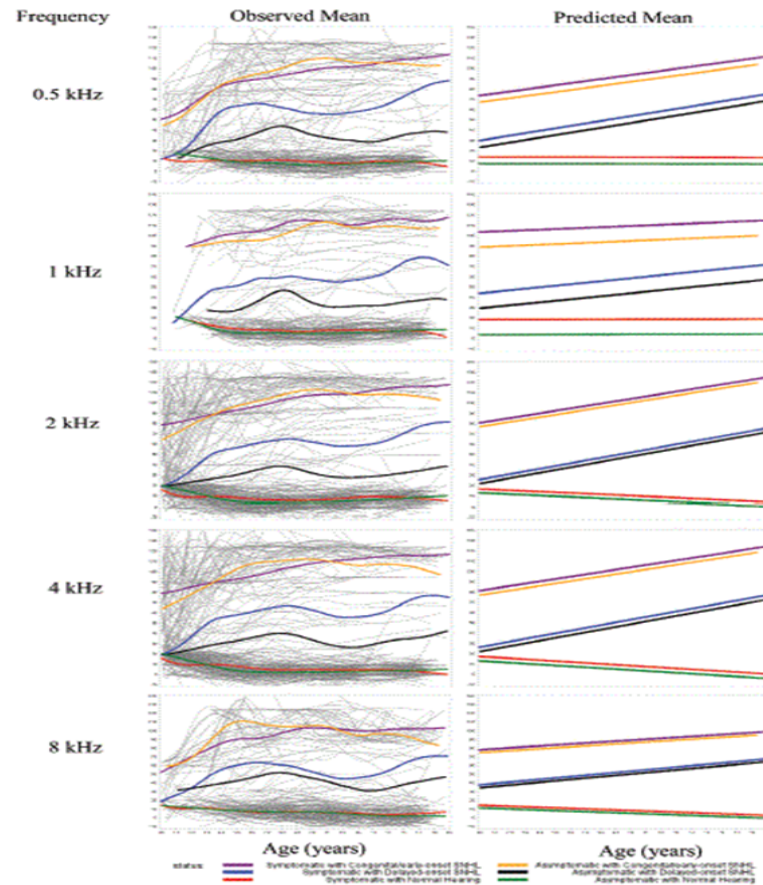
Results:

- Most congenital/early-onset SNHL patients had worsening hearing thresholds in the **poorer-hearing ear** and no change in the better-hearing ear by 12 months of age.
- The poorer-hearing ear worsened earlier and more precipitously than the better-hearing ear in most patients with congenital/early-onset SNHL **through childhood**.
- Delayed-onset SNHL patients also demonstrated earlier and more precipitous worsening hearing in the **poorer-hearing ear**, however, the overall progression was slower and the degree of loss was less severe.

Torrecillas V et al. Should you Follow the Better-Hearing Ear for Congenital Cytomegalovirus Infection and Isolated Sensorineural Hearing Loss? Otolaryngology- HNS Journal, 2019.

Results:

- Same database to evaluate hearing loss trajectory for symptomatic and asymptomatic cCMV infected children
- n=96 symptomatic and n=92 asymptomatic
- Severity worsened for all cCMV children at **ALL** frequencies tested (0.5, 1, 2 and 4 kHz)



Recommendations:

- Monitoring individual hearing thresholds in **both** ears is important for appropriate interventions and future evaluation and efficacy of antiviral treatment.
- All frequencies will worsen if progression occurs. Can use DPOAE for surveillance.
- Surveillance should continue through adolescence
- Utah: every 3 mo x 3 years then every 6 mo thru 6 years of age
- Would then recommend at least annually afterwards

Conclusion:

- Hearing most common sequelae from CMV
- Any child can undergo hearing testing regardless of age
- Long-term follow-up important
- Need to test both ears

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The National Technical Resource Center for
Early Hearing Detection and Intervention (EHDI) Programs