

Audiologic Outcomes from a Targeted and Expanded -Targeted cCMV Testing Program: 5-year Northwell Experience

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Abstract (updated)

cCMV is associated with progressive sensorineural hearing loss (SNHL). A hearing-targeted screening program was instituted at Northwell Health hospitals in 2018, where approximately 1% of babies in the U.S. are born each year. This led to increased CMV awareness. Consequently, infants with symptoms other than a failed hearing test, and those born to women with suspected CMV infection, were also tested and referred to ID and Audiology.

Beginning February 2018, 14 babies who failed their newborn hearing screen and were CMV positive on saliva (hearing-targeted cohort) were referred for audiology evaluation. Three were not confirmed cCMV (negative urine), 3 went to outside hospitals with results unavailable to us, and 2 did not show for appointments. Six were seen for diagnostic ABR; 3 were found to have profound sensorineural hearing loss (SNHL) in one ear and one was found to have bilateral asymmetric SNHL. Two children were found to have transient conductive hearing loss (CHL). The average age at initial ABR in this cohort was 7.5 weeks.

An expanded-targeted cohort of 24 infants tested for CMV because of IUGR, microcephaly, abnormal head imaging, or prenatal diagnosis of CMV were referred. One child was found to have progressive SNHL, with onset at 3 years of age, 2 are still being investigated for SNHL, and 3 had transient CHL; no others were found to have SNHL. The average age at ABR/OAE in this cohort was 3.3 months, and 9 months at behavioral evaluation. These infants continue to be monitored due to risk for progressive SNHL.

Additionally, infants with postnatally acquired CMV are being followed. The presentation's goal is to demonstrate that a systematic, ongoing, enhanced-targeted CMV testing program leads to detection and follow-up of infants at risk for SNHL, and increases awareness of CMV that results in more frequent ID and Audiology referrals of at-risk infants.

Background

Our longstanding NBHS program identifies infants with congenital SNHL or transient conductive hearing loss at birth, however, this testing does not identify infants who at baseline are at risk for progressive changes in hearing. It is known that CMV is a leading cause of progressive SNHL. Our Targeted, and subsequently Expanded Targeted CMV testing program instituted in 2018 should identify and allow monitoring of children at risk for a change in hearing to avoid the potential negative effects of undiagnosed SNHL. Up to 43% of infants with cCMV may pass initial NBHS but have progressive hearing loss in infancy or childhood.¹

The CMV testing program was put into practice in February 2018 at all birthing hospitals within the Northwell Health system, in which any baby failing their NBHS in one or both ears was screened for CMV. If a cCMV diagnosis was confirmed by the Division of Infectious Diseases, the baby was referred to Audiology to follow a specific CMV audiology monitoring protocol. Additionally, infants are identified via an enhanced-targeted strategy in which any infant at risk for cCMV as per ID was referred to be followed by the same protocol.

Purpose

Describe the hearing loss and follow-up rates in infants referred after Hearing Targeted testing

Describe the hearing loss and follow-up rates in infants referred after Expanded Targeted testing

Results

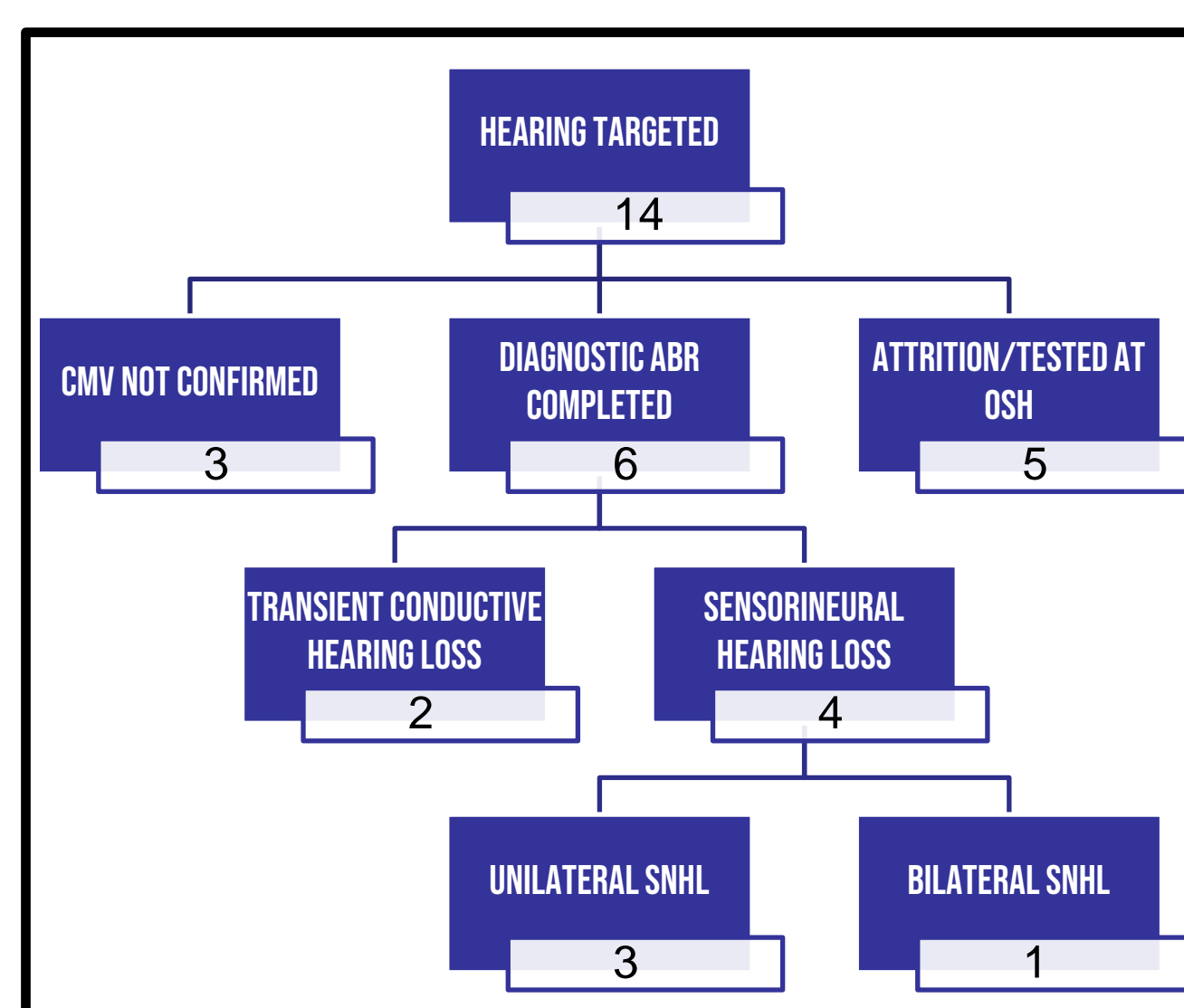
Cohorts studied

Hearing Targeted Cohort of 14 infants referred for audiology monitoring protocol after failed newborn hearing screen and saliva PCR positive (Figure 1)

Expanded Targeted Cohort of 24 infants referred for audiology monitoring protocol after saliva or urine PCR positive in work up for IUGR, microcephaly, abnormal head imaging, or prenatal diagnosis of CMV (Figure 2)

Routine SNHL Cohort of 20 CMV negative infants diagnosed with hearing loss at birth in one or both ears, and followed by routine audiology monitoring protocol, for comparison with both the cCMV groups:

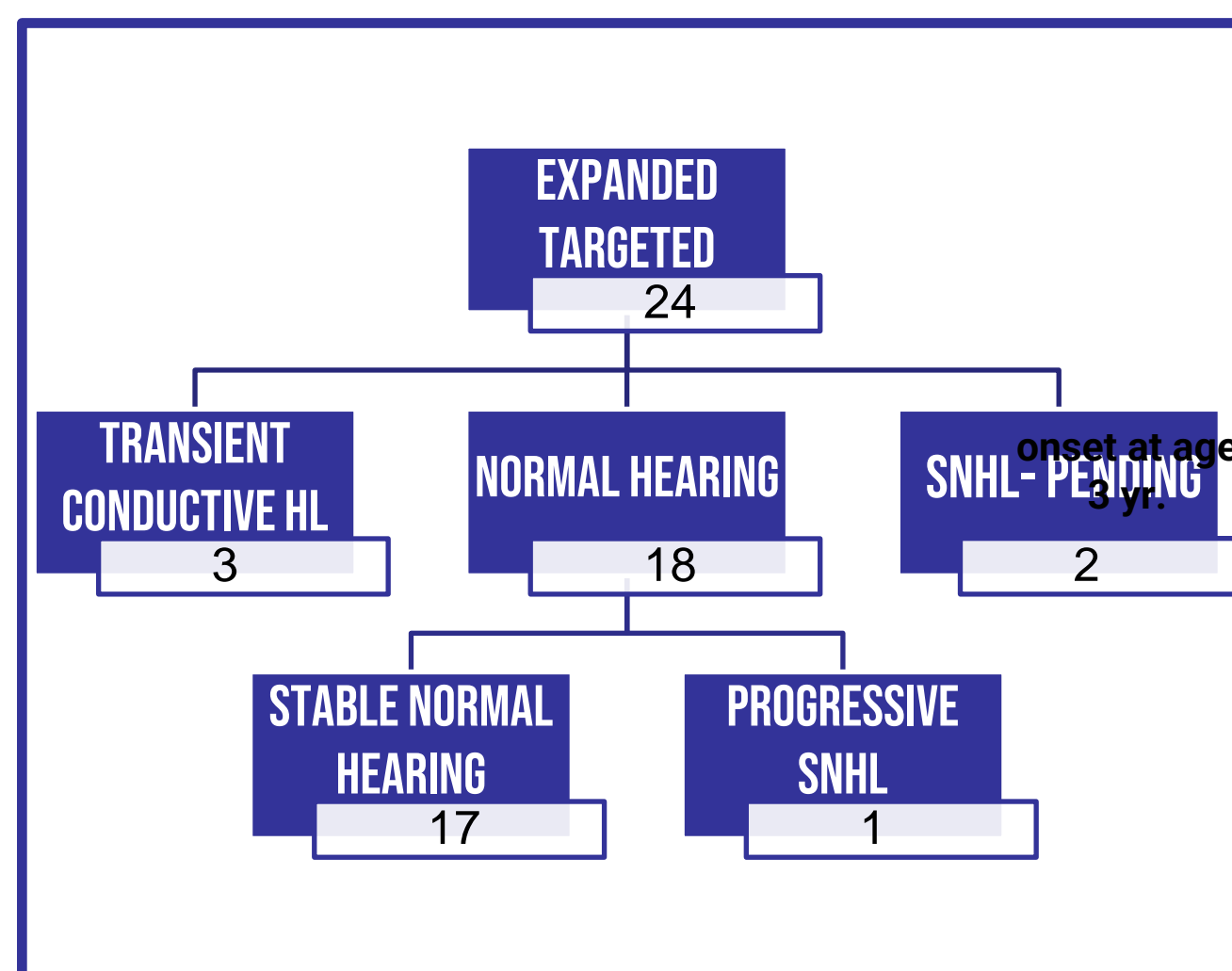
A. Compliance rate with follow-up visits (Figure 3) B. Laterality of hearing loss (Figure 4)



Average age at initial ABR = 7.5 weeks

Completely asymptomatic	2
Microcephaly, SNHL	1
SNHL only	3

Figure 1



Average age at ABR/OAE = 3.3 months
Average age at behavioral evaluation = 9 months

Completely asymptomatic	6
Completely asymptomatic, developed SNHL age 3	1
Asymptomatic, possible SGA	4
Asymptomatic, possible microcephaly	1
SNHL only, pending confirmation	1
SNHL pending confirmation, CNS symptomatic	1
Symptomatic, premature VLBW	7
Late referral, possibility of pCMV	3

Figure 2

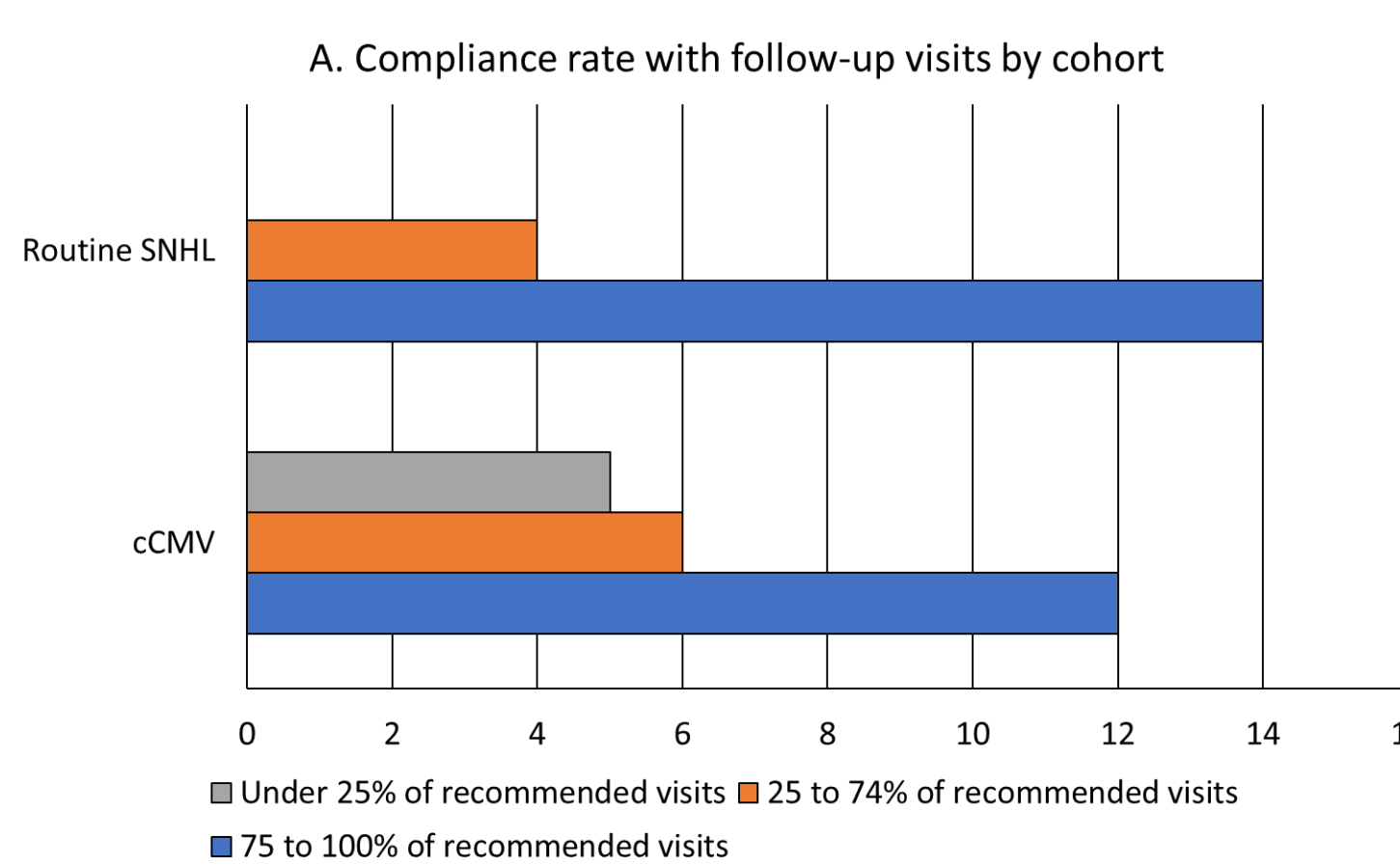


Figure 3

Follow-up visit compliance for cCMV infants is fairly high but would benefit from enhanced administrative resources

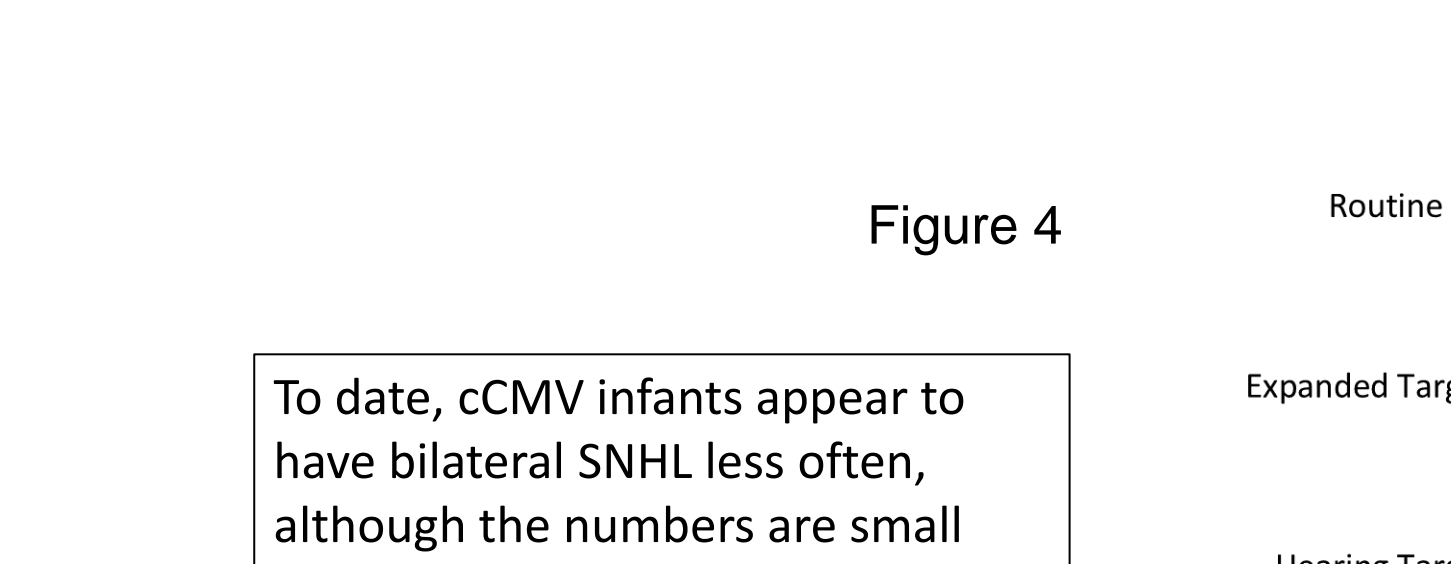


Figure 4

To date, cCMV infants appear to have bilateral SNHL less often, although the numbers are small

Methods

Routine two-step newborn hearing screening (NBHS) measures are performed for all babies; initial screening completed via OAE using Otodynamics Otocheck DP followed by AABR using Natus Algo 3i or 7i if OAE screening was failed in one or both ears. Both methods are noninvasive and easily performed in neonates by trained screening technicians prior to hospital discharge.

Hearing Targeted: If one or both ears do not pass the AABR screening, CMV screening via urine sample or buccal swab is completed and the baby is referred for outpatient diagnostic evaluation via ABR.

Expanded Targeted: Any babies diagnosed with cCMV regardless of NBHS result are referred for audiology evaluation and monitoring.

All diagnostic ABR testing is completed by licensed audiologists at the LIJ Hearing and Speech Center at Northwell Health using the Vivonic Integrity ABR system.

Audiologic Monitoring Protocol for Infants and Children Diagnosed with cCMV

1. Initial ABR evaluation in natural sleep prior to four weeks of age or immediately following cCMV diagnosis
2. Repeat ABR evaluation to confirm results if hearing loss is identified and routine fitting of amplification
3. Repeat ABR evaluation at 4-5 months of age
4. Behavioral audiology evaluation at 7 months of age, including TEOAE and immittance testing
5. Routine behavioral audiology evaluation every 3 months until age 3, followed by behavioral audiology evaluation every 6 months until age 6 and annually thereafter
6. Consider ABR under sedation if frequency and ear specific results cannot be obtained over two visits, or there is concern for a change in hearing

Discussion

In 2018 we instituted a system-wide (all 10 birthing hospitals), successful, targeted/expanded hearing testing program for all newborns. The Expanded Targeted cohort is a result of increased awareness of cCMV, as a collateral benefit of the Hearing Targeted program. Babies with mild cCMV symptoms are being diagnosed more commonly, and referred to Audiology for long-term follow-up, which decreases the risk of subsequent poorer outcomes in speech, language, education, social functioning, cognitive abilities and overall quality of life.²

Four of 11 babies with confirmed cCMV in the Hearing Targeted cohort had sensorineural hearing loss. Only 1 (2 are pending confirmation) of 24 babies with confirmed cCMV in the Expanded Targeted cohort had sensorineural hearing loss.

Prompt referral and coordination of appointments between Pediatric ID and the Hearing and Speech Center has proved effective in capturing and monitoring children at risk for hearing loss due to cCMV. This is evident from the high follow-up rate, which is similar to the rate of follow-up in the pediatric hearing-impaired population without cCMV. Follow up in the cCMV cohort has improved over time due to increased knowledge of the protocol among audiologists, otolaryngologists, neonatologists and Infectious Disease physicians.

As we continue to monitor the effectiveness of our cCMV hearing monitoring program, we are discussing means of ensuring follow-up through 6 years of age in those cCMV babies who passed their newborn hearing screen.

Acknowledgements

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References

1. American Academy of Pediatrics (1999). Newborn and Infant Hearing Loss: Detection and Intervention. Task Force on Newborn and Infant Hearing. Pediatrics, 103, 527-530.
2. Lieu J, Kenna M, Anne S, Davidson W. Hearing Loss in Children: A Review. JAMA. 2020;324(21):2195-2205.

