Association Between Craniofacial Anomalies and NBHS Fail Rate

Ruth S. Marin PhD[1], Kaitlyn Sheapp, AuD[2,3], Larry Medwetsky PhD[3]
Adventist HealthCare White Oak Medical Center[1], Adventist HealthCare Shady Grove Medical Center[2], Gallaudet University[3]

Introduction

Craniofacial anomalies (CFA) are one of the leading causes of hearing loss in newborns, however, the literature does not describe the incidence based on specific disorders.
Awareness of the prevalence of specific CFA and their associated risks for hearing loss can help guide:

- development of evidence-based practice regarding detection and documentation of risk factors at birth
- appropriate recommendations for follow-up testing and monitoring.

Methods

Records were reviewed for 39,813 infants born at Adventist Healthcare (AHC) Shady Grove Medical Center and AHC White Oak Medical Center between January 1, 2014, and December 31, 2019, to determine the association between the presence of craniofacial anomalies and newborn hearing screening (NBHS) fail rates. This project was approved by the IRB at AHC and Gallaudet University.

Results

- Infants with craniofacial anomalies are at greater risk for failing the newborn hearing screening
- Overall fail rate was .74% for all newborns with or without a CFA
- Fail rate for all newborns with CFA was 4.4%
- Atresia/microtia yielded the highest NBHS fail rate
- Preauricular sinuses/tags were the most common CFA but yielded the lowest NBHS fail rate

Selected References

American Speech-Language-Hearing Association (ASHA) (n.d). Newborn Hearing Screening. (Practice Portal). Retrieved August 18, 2020 from www.asha.org/Practice-Portal/Professional-Issues/Newborn-Hearing-Screening.

Hutt, N., & Rhodes, C. (2008). Post-natal hearing loss in universal neonatal hearing screening communities: Current limitations and future directions. *Journal of Paediatrics and Child Health*, 44(3), 87-91.

Joint Committee on Infant Hearing. (2019). Year 2019 position statement: Principles and guidelines for early hearing detection and intervention programs. *Journal of Early Hearing Detection and Intervention*, 4(2), 1-44.

Joint Committee on Infant Hearing. (2007). Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics*, 120(4), 898-921.

Scan for full reference list

Results (continued)

Table 1. Prevalence of Specific CFAs at Birth and Associated NBHS Fail Rate

Category of CFA	Total % of CFA (n=819)*	NBHS Fail Rate (n=36)
Syndrome associated with hearing loss	9.4% (77)	18.18% (14)*
Ear sinuses (ear pits)/ tags	82.25% (672)	0.74% (5)
Atresia/Microtia	1.47% (12)	100% (12)
Malformed Ears, other	1.35% (11)	36.36% (4)
Skull Malformations	0.61% (5)	20% (1)
Cleft Lip/Palate	4.16% (34)	5.8% (2)*
Other	0.98% (8)	0.00% (0)

*Two babies with multiple anomalies were included in both categories

Table 2. Prevalence of Unilateral vs. Bilateral Preauricular Sinuses/Tags and Associated NBHS Fail Rate

Category of CFA	Prevalence (n=)	NBHS Fail Rate
Unilateral Preauricular Tag	30.95% (208)	0.96% (2)
Unilateral Preauricular Sinus	51.34% (345)	0.29% (1)
Bilateral Preauricular Tags	2.68% (18)	0.00% (0)
Bilateral Preauricular Sinuses	14.43% (97)	0.21% (2)
Both Preauricular Sinuses & Tags (unilateral or bilateral)	0.61% (4)	0.00% (0)

Discussion

- Our overall NBHS fail rate (.74%) was lower than the national fail rate (4%, ASHA, ND) possibly due to our 2-step screening protocol
- The NBHS fail rate for babies with CFA (4.4%) is six times higher than our overall fail rate
- For CFA, the NBHS fail rate varies from 0-100%, confirming the need to delineate the CFA category into specific disorders
- Preauricular ear sinuses/tags have such a low incidence of NBHS failure that they should not be included for follow-up unless there are other features associated with hearing loss or syndromes
- Of the newborns that were ultimately diagnosed with hearing loss (n=15), 44.2% had a CFA

Conclusions

- Infants with CFA were 6x more likely to fail the NBHS when compared to the fail rate for all infants
- Audiological follow-up and monitoring is not warranted for infants with preauricular sinuses and tags unless the infant exhibits other features associated with a syndrome that has an associated risk for hearing loss
- Because the NBHS fail rates in this study varied greatly for the different CFA, further research should be completed to determine if these findings can be replicated. If so, we recommend that the Joint Committee on Infant Hearing (JCIH) update:
  - the list of risk factors for hearing loss to delineate the current CFA category into subgroups
  - recommendations for each specific CFA, as infants in this risk factor group should be followed based on their specific anomaly rather than as homogeneous group