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# An Enhanced Audiologic Protocol for Early Identification of Conductive Hearing Loss in Patients with Cleft Palate

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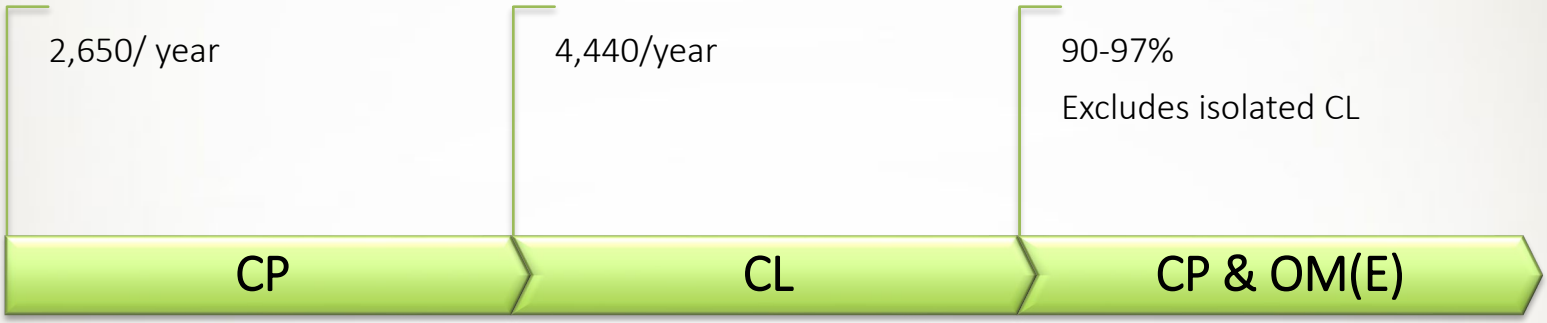
# Brief Overview

- Cleft lip (CL) and/or palate (CP) is a birth defect that occurs in utero
  - Cleft lip forms between 4-7 weeks gestation
  - Cleft palate forms between 6-9 weeks gestation

# CP Statistics




- Eshghi et al.
- Children with CP experience significant vocabulary declines in comparison to their typically developing peers
- Middle ear dysfunction can impact the development of vocabulary skills across all children, not just CP population

# CL/CP Causes & Risk Factors

- Genetics
- Smoking in pregnancy
- Drinking in pregnancy
- Medications
- Epilepsy medications during 1st trimester/1st 3 months
- Diabetes
- Obesity during pregnancy

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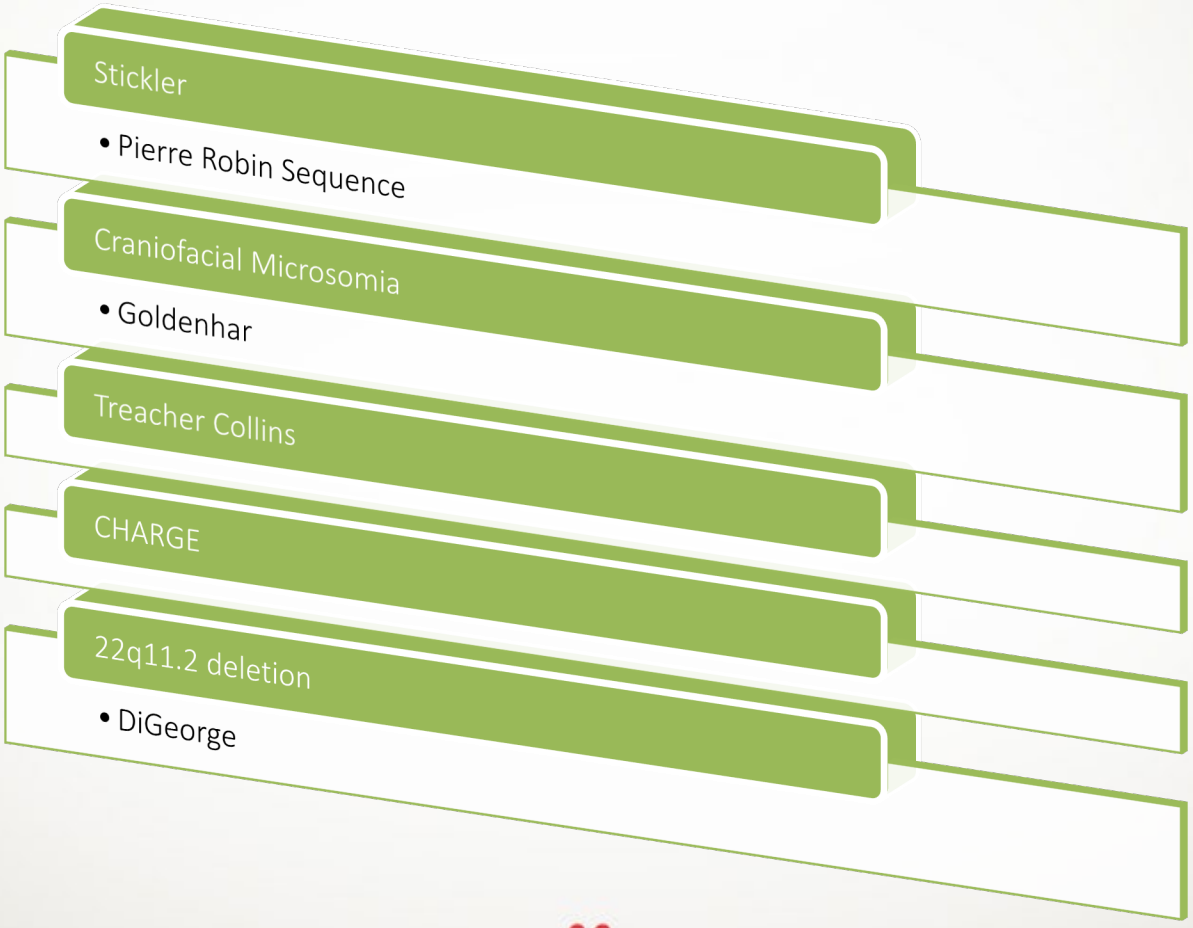
# Genetics:

## Over 400 associated syndromes

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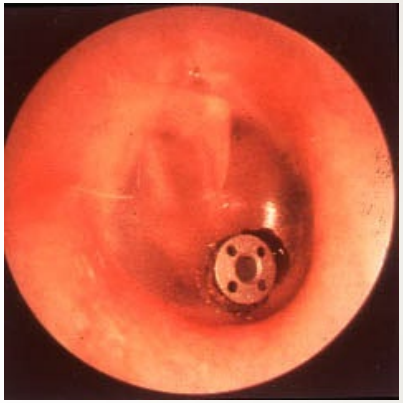
# CP & Otolaryngology

- Initial set of PETs usually placed with palatoplasty (9-12 months)
- > 90% of patients with CL/CP have effusions at time of lip repair ~3 months of age
- Average of ~3 sets per patient
- Decrease in need for PET by age 6-8
- Unable to predict severity of ear problems by initial cleft severity
- Earlier closure of palate not associated with earlier decrease in effusions, though some evidence shows has better overall hearing outcomes
- Some evidence that type of palatoplasty may influence resolution of ear problems



# CP & AUDIOLOGY

- Joint Committee on Infant Hearing (JCIH) Position Statement
  - 1-3-6/1-2-3 screening, audiologic diagnosis, enrollment in early intervention

- Identification of HL and appropriate intervention by 3-6 months of age for maximal audibility is often missed for children with CP

- Audiologist work closely with Otolaryngologist to diagnose, manage, and provide appropriate intervention to children diagnosed with cleft palate

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# CP & AUDIOLOGY

- Current JCIH recs for CP
  - 9 month FU

- *"When present from time of birth, if CHL cannot be medically remediated by 6 mo of age, the child should be considered for hearing aid amplification, communication supports, a referral to early intervention services, even if these services may be short-term. Such interventions are necessary to address and prevent developmental language delays. This type of situation commonly occurs when providing care for infants with cleft palate or Down Syndrome"*

**Table 1**
***Risk Factors for Early Childhood Hearing Loss: Guidelines for Infants who Pass the Newborn Hearing Screen***

| | Risk Factor Classification | Recommended Diagnostic Follow-up | Monitoring Frequency |
|---|---|---|---|
| | Perinatal | | |
| 1 | Family history* of early, progressive, or delayed onset permanent childhood hearing loss | by 9 months | Based on etiology of family hearing loss and caregiver concern |
| 2 | Neonatal intensive care of more than 5 days | by 9 months | As per concerns of on-going surveillance of hearing skills and speech milestones |
| 3 | Hyperbilirubinemia with exchange transfusion regardless of length of stay | by 9 months | |
| 4 | Aminoglycoside administration for more than 5 days** | by 9 months | |
| 5 | Asphyxia or Hypoxic Ischemic Encephalopathy | by 9 months | |
| 6 | Extracorporeal membrane oxygenation (ECMO)* | No later than 3 months after occurrence | Every 12 months to school age or at shorter intervals based on concerns of parent or provider |
| 7 | In utero infections, such as herpes, rubella, syphilis, and toxoplasmosis | by 9 months | As per concerns of on-going surveillance |
| | In utero infection with cytomegalovirus (CMV)* | No later than 3 months after occurrence | Every 12 months to age 3 or at shorter intervals based on parent/provider concerns |
| | Mother + Zika and infant with no laboratory evidence & no clinical findings | standard | As per AAP (2017) Periodicity schedule |
| | Mother + Zika and infant with laboratory evidence of Zika + clinical findings<br>Mother + Zika and infant with laboratory evidence of Zika - clinical findings | AABR by 1 month<br>AABR by 1 month | ABR by 4-6 months or VRA by 9 months<br>ABR by 4-6 months<br>Monitor as per AAP [illegible] Periodicity schedule (Adebanjo et al., 2017) |
| 8 | Certain birth conditions or findings:<br>• Craniofacial malformations including microtia/atresia, ear dysplasia, oral facial clefting, white forelock, and microphthalmia<br>• Congenital microcephaly, congenital or acquired hydrocephalus<br>• Temporal bone abnormalities | by 9 months | As per concerns of on-going surveillance of hearing skills and speech milestones |
| 9 | Over 400 syndrome[illegible] been identified with atypical hearing thresholds***. For more information, visit the Hereditary Hearing Loss website (Van Camp & Smith, 2016) | by 9 months | According to [illegible] history of syndrome or concerns |
| | Perinatal or Postnatal | | |
| 10 | Culture-positive infections associated with sensorineural hearing loss***, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis or encephalitis | No later than 3 months after occurrence | Every 12 months to school age or at shorter intervals based on concerns of parent or provider |
| 11 | Events associated with hearing loss:<br>• Significant head trauma especially basal skull/temporal bone fractures<br>• Chemotherapy | No later than 3 months after occurrence | According to findings and or continued concerns |
| 12 | Caregiver concern**** regarding hearing, speech, language, developmental delay and or developmental regression | Immediate referral | According to findings and or continued concerns |

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- **Objective:** Characterize onset and prevalence of CHL in pediatric pts. w/ CP prior to palatoplasty
- **Design:** Retrospective cohort study
- **Setting:** Multidisciplinary cleft and craniofacial clinic at a tertiary care center
- **Patients:** Pts w/ CP who received pre-op w/u
  - Exclusion: Patients with bilat permanent hearing loss, expiration prior to palatoplasty, or no pre-op data
- **Main Outcome Measures:** Age of identification of CHL in patients after implementation of enhanced protocol

*Original Article*

ACPA American Cleft Palate Craniofacial Association

The Cleft Palate Craniofacial Journal 1-6

DOI: 10.1177/10556656231178437
journals.sagepub.com/home/cpc

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## An Enhanced Audiologic Protocol for Early Identification of Conductive Hearing Loss in Patients with Cleft Palate

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**Abstract**

*Objective:* To characterize the onset and prevalence of conductive hearing loss (CHL) in pediatric patients with cleft palate (CP) prior to palatoplasty with an enhanced audiologic protocol.

*Design:* Retrospective cohort study

*Setting:* Multidisciplinary cleft and craniofacial clinic at a tertiary care center.

*Patients:* Patients with CP who received audiologic workup pre-operatively. Patients with bilateral permanent hearing loss, expiration prior to palatoplasty, or no pre-operative data were excluded.

*Interventions:* Patients with CP born February 2019 to November 2019 who passed newborn hearing screening (NBHS) received audiologic testing at 9 months of age (standard protocol). Patients born December 2019 to September 2020 underwent testing prior to 9 months of age (enhanced protocol).

*Main Outcome Measures::* Age of identification of CHL in patients after implementation of the enhanced audiologic protocol.

*Results:* The number of patients who passed their NBHS in the standard protocol (n = 14, 54%) and the enhanced protocol (n = 25, 66%) did not differ. Infants who passed their NBHS, but demonstrated hearing loss on subsequent audiologic testing did not differ between enhanced (n = 25, 66%) and standard cohort (n = 14, 54%). Of patients who passed NBHS in the enhanced protocol, 48% (n = 12) had CHL identified by 3 months, and 20% (n = 5) by 6 months of age. With the enhanced protocol, patients who did not undergo additional testing post NBHS significantly dropped from 44.9% (n = 22) to 4.2% (n = 2) ($P < .0001$).

*Conclusion:* Even with passed NBHS, CHL is still present for infants with CP pre-operatively. Earlier and more frequent testing for this population is recommended.

**Keywords**

hearing loss, cleft palate, early communication

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# Patient Characteristics

- No statistical significance btw groups

**Table 1.** Patient Characteristics.

| | Standard protocol (N = 26) | Enhanced protocol (N = 38) | *P* Value |
|---|---|---|---|
| Sex | | | .257 |
| Female | 46% (N = 12) | 61% (N = 23) | |
| Male | 54% (N = 14) | 39% (N = 15) | |
| Race | | | .381 |
| Asian | 4% (N = 1) | 3% (N = 1) | |
| Black | 8% (N = 2) | 3% (N = 1) | |
| Other | 19% (N = 5) | 15% (N = 6) | |
| White | 69% (N = 18) | 79% (N = 30) | |
| Insurance | | | .359 |
| Private | 12% (N = 3) | 13% (N = 5) | |
| Public | 85% (N = 22) | 79% (N = 30) | |
| Other (ie, Military) | 0% (N = 0) | 8% (N = 3) | |
| Self-Pay/No Insurance | 4% (N = 1) | 0% (N = 0) | |
| Cleft Type | | | .276 |
| Complete | 50% (N = 13) | 34% (N = 13) | |
| Incomplete | 46% (N = 12) | 50% (N = 19) | |
| Submucous | 4% (N = 1) | 16% (N = 6) | |
| Associated genetic syndrome | | | .602 |
| Present | 23% (N = 6) | 29% (N = 11) | |
| Absent | 77% (N = 20) | 71% (N = 27) | |
| Prematurity | | | .769 |
| ≤36 Weeks | 27% (N = 7) | 24% (N = 9) | |
| >37 Weeks | 73% (N = 19) | 76% (N = 29) | |
| Neonatal intensive care unit Stay | | | .100 |
| >5 Days | 58% (N = 15) | 37% (N = 14) | |
| None or ≤5 Days | 42% (N = 11) | 63% (N = 24) | |
| Newborn hearing screening | | | .336 |
| Pass | 54% (N = 14) | 66% (N = 25) | |
| Fail | 46% (N = 12) | 34% (N = 13) | |

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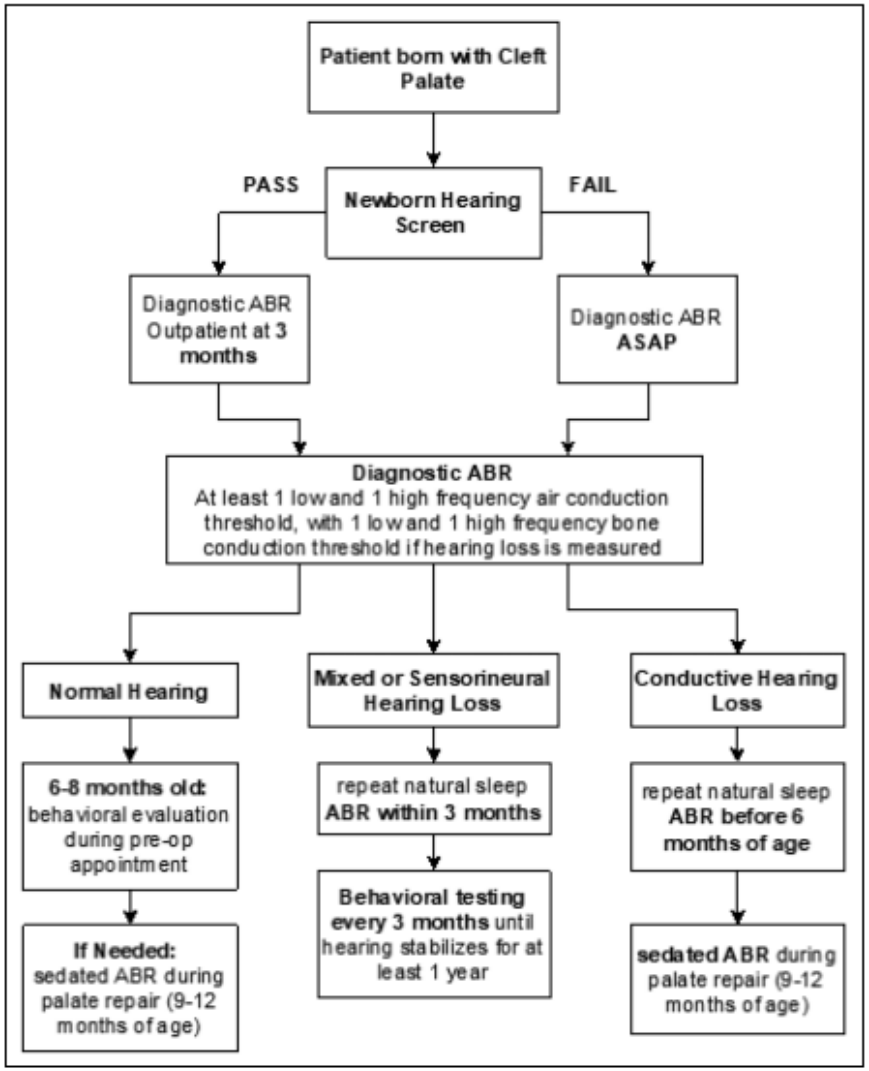


**Figure 1.** Enhanced audiologic protocol.

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# Results

- The n of patients who passed NBHS at birth did not differ btw standard vs. enhanced protocol
- Infants who passed NBHS but demonstrated HL on subsequent audiologic testing did not differ between the two protocols.
  - *Please note w/ less referrals to audiology in the standard protocol, HL was ID'ed later (~9 months) vs. than those ID'ed at 3 & 6 months in enhanced protocol
- **Of patients who passed NBHS in the enhanced protocol:**
  - **48% (n=12) had CHL ID'ed by 3 months of age**
  - **20% (n=5) had CHL ID'ed by 6 months of age**
    - ***68% of patients w/ HL prior to 9 mo's of age***
- W/ standard protocol, 45 % (n+22) did not undergo pre-operative testing
- W/ enhanced protocol, only 4% did not undergo pre-operative testing (p<.0001)

*The Cleft Palate Craniofacial Journal 0(0)*

**Table 3.** Hearing Loss with Passed NBHS.

| Hearing loss in patients who passed NBHS | | |
|---|---|---|
| Passed NBHS | Standard (n = 14)/26 | Enhanced (n = 25)/38 |
| Hearing loss present | 57% (n = 8) | 68% (n = 17) |
| No hearing loss measured | 43% (n = 6) | 32% (n = 8) |

**Table 2.** Hearing Loss Severity.

| Severity | Standard protocol (N = 26) | Enhanced protocol (N = 38) |
|---|---|---|
| Mild | 38% (N = 10) | 29% (N = 11) |
| Mild-moderate | 19% (N = 5) | 29% (N = 11) |
| Moderate | 15% (N = 4) | 8% (N = 3) |
| Moderately-severe | 0% (N = 0) | 5% (N = 2) |
| Unclassified* | 4% (N = 1) | 5% (N = 2) |
| Normal | 23% (N = 6) | 24% (N = 9) |

*Represents absent DPOAE results when unable to obtain behavioral information.

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# Conclusion

- Significant improvement in early detection of HL
- CHL was prevalent in cohorts well before 9-month JCIH recommendation (for passed NBHS)
- Increased detection of HL was primarily among patients who passed NBHS
  - Failed NBHS = testing sooner
- Severity mostly mild / mild-moderate
  - Speech & language development
    - ***not** structural articulation disorders associated w/ CP
  - Psychoeducational & Psychosocial Development
- *Proposal: Earlier routine audiologic evaluation in this population, regardless of NBHS result at birth.*

# Limitations

- Small sample size
  - Retrospective data
  - Single institution study
  - Replication of data w/ higher n to confirm true incidence of HL for CP population
- Tertiary referral center
  - May have more complexities
  - May have higher comorbidity conditions vs. patients treated at lower acuity centers
  - *Genetics syndromes in this group of CP patient were similar to global estimates of non-tertiary referral centers.
- Limited sleep state
- Missed appointments / age out of sleep state by next apt date

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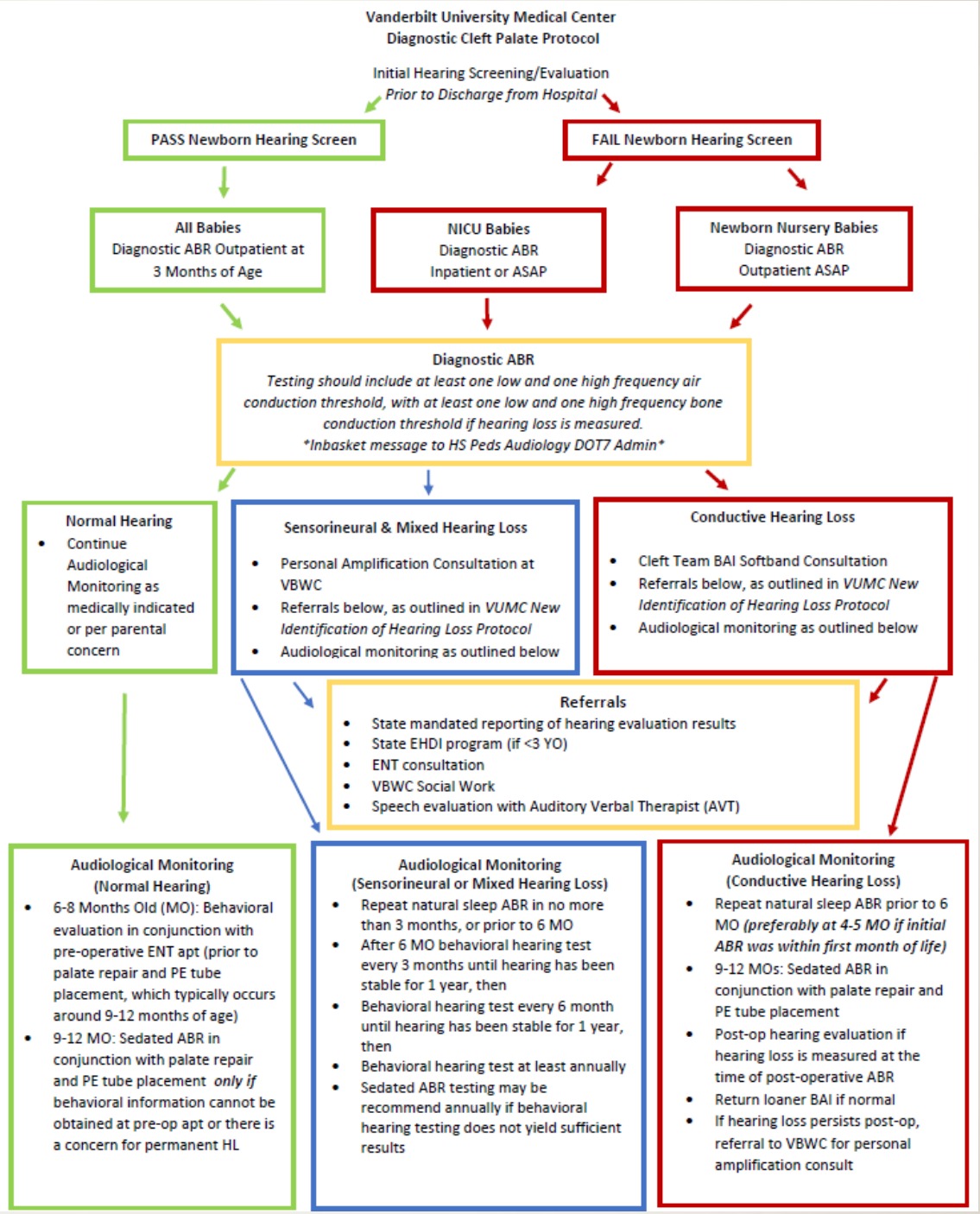
# VUMC HL Interventions



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# Citations


- Ellis EW, Smetak MR, Alving-Trinh A, Golinko M, Phillips JD, Belcher RH. An Enhanced Audiologic Protocol for Early Identification of Conductive Hearing Loss in Patients with Cleft Palate. The Cleft Palate Craniofacial Journal. 2023;0(0). doi:10.1177/10556656231178437

- Dhillon RS. The middle ear in cleft palate children Pre and post palatal closure. J R Soc Med. 1988;81(12):710-713. doi:10.1177/014107688808101209

- Eshghi, M., Adatorwovor, R., Preisser, J. S., Crais, E. R., Zajac, D. J., Department of Biostatistics, & Department of Dental Ecology. (2018, December 22). Vocabulary Growth From 18 to 24 Months of Age in Children With and Without Repaired Cleft Palate. Retrieved September 29, 2019, from https://pubs.asha.org/doi/pdf/10.1044/2019_JSLHR-L-18-0207.

- (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. DOI: 10.15142/fptk-b748

- Young NM, Tharpe AM. Current perspectives on childhood hearing loss. Otolaryngol Clin North Am. 2021;54(6):xv–xvii. doi:10.1016/j.otc.2021.08.011


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