



Assessment and Management of Auditory Neuropathy Spectrum Disorder (ANSD)

Mary Laing Holland, AuD, CCC-A
Clinical Instructor of Pediatric Audiology
Medical University of South Carolina

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Objective

Evaluate, diagnose, and manage patients with Auditory Neuropathy Spectrum Disorder (ANSD).

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What is Auditory Neuropathy Spectrum Disorder?

- ▶ First identified by A. Starr and colleagues in 1996
- ▶ Term used to describe heterogeneous nature
- ▶ Higher prevalence of SNHL (and ANSD) in newborns discharged from NICU
- ▶ Up to 10% of children with permanent hearing loss show signs of ANSD

(Roush, 2017)
(Rance & Starr, 2017)



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Risk Factors of Auditory Neuropathy Spectrum Disorder

- ▶ Prematurity
- ▶ Low birthweight
- ▶ Hyperbilirubinemia
- ▶ Hypoxia
- ▶ Metabolic Acidosis
- ▶ Exposure to ototoxic drugs
- ▶ Artificial ventilation

(Roush, 2017)
(Rance & Starr, 2017)



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Etiologies of Auditory Neuropathy Spectrum Disorder

- ▶ **Genetic**
 - ▶ Both syndromic and non syndromic
 - ▶ OTOF, PMP22, MPZ, NDRG1
 - ▶ Charcot Marie Tooth (CMT), Friedreich's Ataxia, Mitochondrial Disorders
- ▶ **Prenatal/ Perinatal**
 - ▶ Prematurity, hyperbilirubinemia, low birth weight, etc.
- ▶ **Postnatal**
 - ▶ Measles, mumps, meningitis, CMV, HIV
- ▶ **Cochlear Nerve Deficiency (CND)**



Clinical Findings of Auditory Neuropathy Spectrum Disorder

- ▶ Present otoacoustic emissions (OAEs) **or** present Cochlear Microphonic (CM)
- ▶ Absent or abnormal Auditory Brainstem Response (ABR) at maximum presentation levels
- ▶ Hallmark of ANSD ABR: CM that follows the stimulus polarity when it is reversed



How is it diagnosed?

- ▶ Otoloscopic Examination
- ▶ Immittance Measures
 - ▶ Tympanometry
 - ▶ Acoustic Reflexes
- ▶ Otoacoustic Emissions (OAEs)
- ▶ Auditory Brainstem Response (ABR)
- ▶ Behavioral Testing (when developmentally appropriate)



What results do we expect?

- ▶ Tympanometry/ Acoustic Reflexes
 - ▶ Normal Tympanometry
 - ▶ Abnormal/Absent Acoustic Reflexes
- ▶ Otoacoustic Emissions
 - ▶ Present OAEs
- ▶ Auditory Brainstem Response
 - ▶ Cochlear Microphonic
 - ▶ Abnormal/Absent ABR
- ▶ Behavioral Testing
 - ▶ Results range from normal hearing to profound hearing loss
 - ▶ Of note, unilateral ANSD is commonly associated with cochlear nerve deficiency (thresholds are usually profound)
- ▶ CAEPs
 - ▶ Potentials from auditory cortex (middle latency response, cortical auditory-evoked potentials, mismatch negative, and P300 responses) can sometimes be recorded in affected adults and children



Commonly Reported Clinical Indications

- ▶ Behavioral thresholds in normal hearing to profound hearing loss range
- ▶ Thresholds are typically symmetric
- ▶ Fluctuating or progressive thresholds
- ▶ Speech recognition abilities that are disproportionate to behavioral audiogram
- ▶ Difficulty hearing in noise
- ▶ Impaired temporal processing
- ▶ Speech delay in young children



Initial Management following diagnosis

- ▶ “Recommendations for initial management of children with sensorineural-type hearing loss by Joint Committee on Infant Hearing (2007) are appropriate for infants diagnosed with ANSD” (Rance & Starr, 2017)
 - ▶ Pediatric and developmental evaluation and history
 - ▶ Otologic evaluation with imaging of cochlear and auditory nerves (CT/MRI) and vestibular assessment
 - ▶ Genetics evaluation
 - ▶ Ophthalmologic assessment
 - ▶ Communication development assessment and ongoing evaluation



Audiological Management

- ▶ Hearing Aids
- ▶ Cochlear Implant
- ▶ Assistive Listening Devices
 - ▶ Remote Microphone/ FM System (personal or sound field)
 - ▶ Beneficial for milder cases of ANSD
- ▶ Factors that might influence outcomes (Roush, 2017):
 - ▶ Age at diagnosis
 - ▶ Age at hearing aid fitting
 - ▶ Consistency of hearing aid use
 - ▶ Quality and intensity of early intervention
 - ▶ Degree of parental involvement
 - ▶ Additional developmental and medical conditions

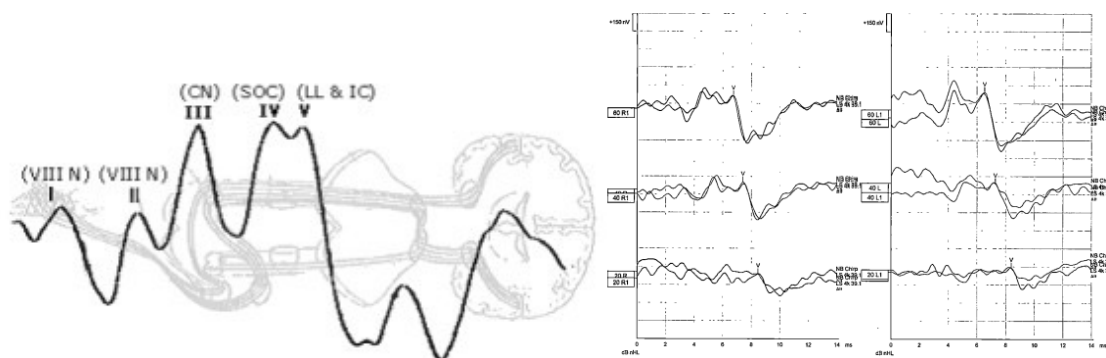


Multidisciplinary Management

- ▶ Multidisciplinary approach
 - ▶ Audiologist
 - ▶ Otolaryngologist
 - ▶ Speech Language Pathologist
 - ▶ Educational Audiologist
 - ▶ Ophthalmologist
 - ▶ Primary Care Physician
 - ▶ Neurologist
 - ▶ Genetics



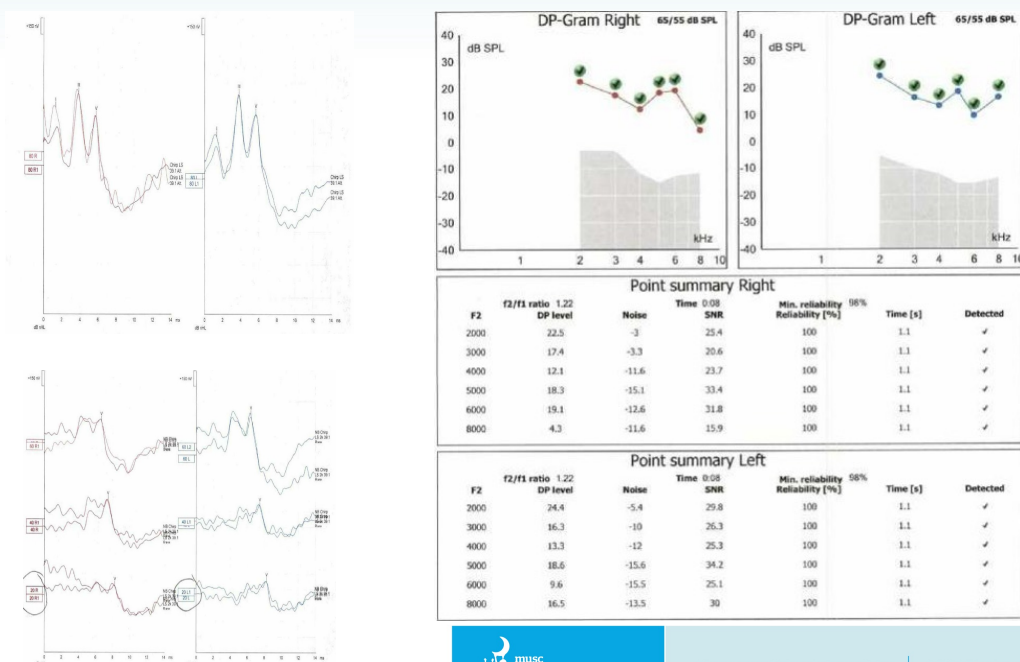
Interpreting Auditory Brainstem Response



(Hall, 1992)

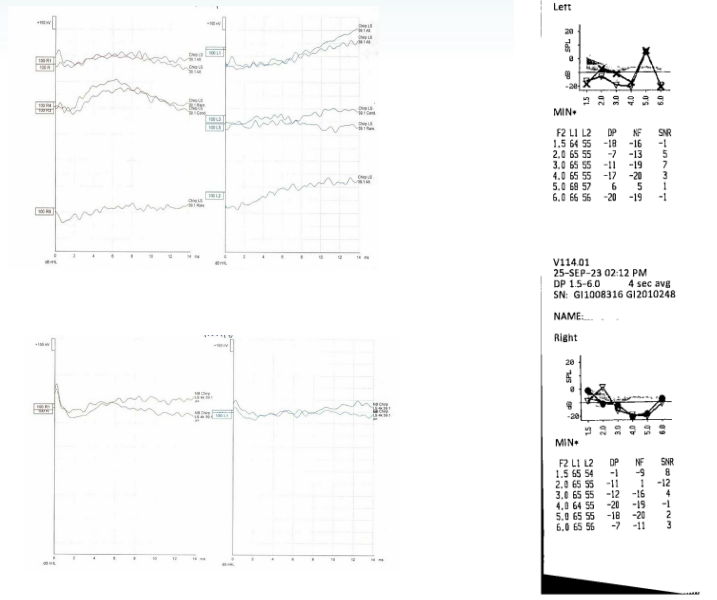
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Normal Hearing Sensitivity



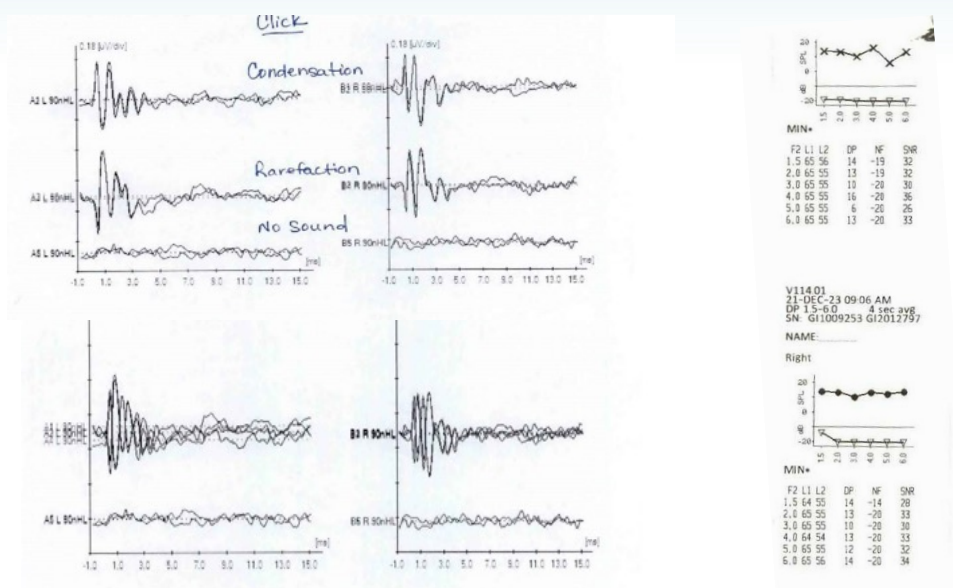
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Profound Sensorineural Hearing Loss



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Auditory Neuropathy Spectrum Disorder



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	Case Example

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History

- ▶ Born at 37 weeks gestation
- ▶ Normal pregnancy
- ▶ Complications: respiratory distress and right pre-auricular skin tag
- ▶ 3 week NICU stay: mechanical ventilation for > 10 days, seizures and persistently elevated direct bilirubin levels of unknown etiology. Did not receive exchange transfusions.
- ▶ MRI: periventricular white matter hemorrhages
- ▶ ALGO ABR Screening: Failed bilaterally x3 prior to discharge
- ▶ Age at discharge: 3 weeks



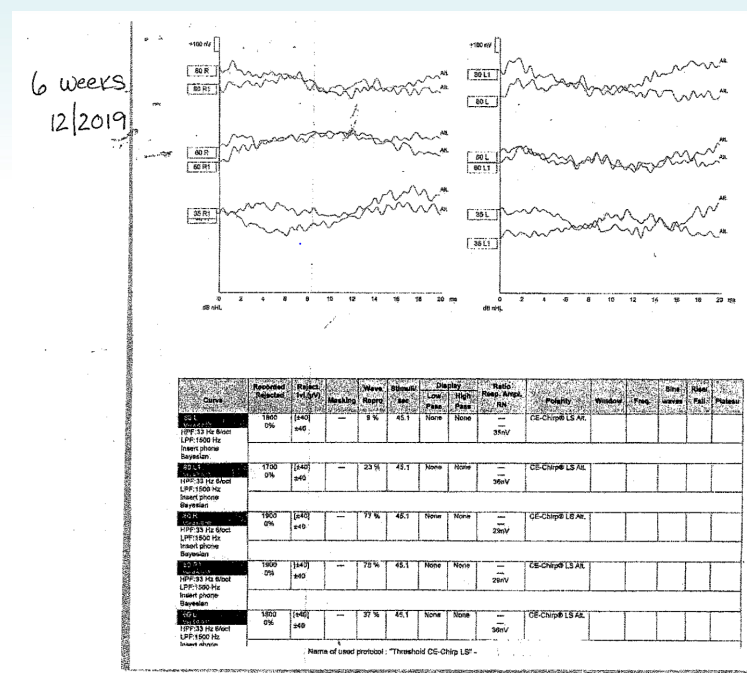
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Diagnosis

- ▶ Failed outpatient ABR rescreening at 6 weeks of age
- ▶ Returned one month later for sleep deprived ABR
 - ▶ Skin tag had been removed
 - ▶ Parent report: Startles to loud sounds at home
 - ▶ Results: ABR interpreted as “moderate hearing loss bilaterally with clear middle ears”
- ▶ Recommendations:
 - ▶ Amplification (appropriate referrals sent and earmold impressions were made)
 - ▶ Fitting was not complete due to unknown reasons



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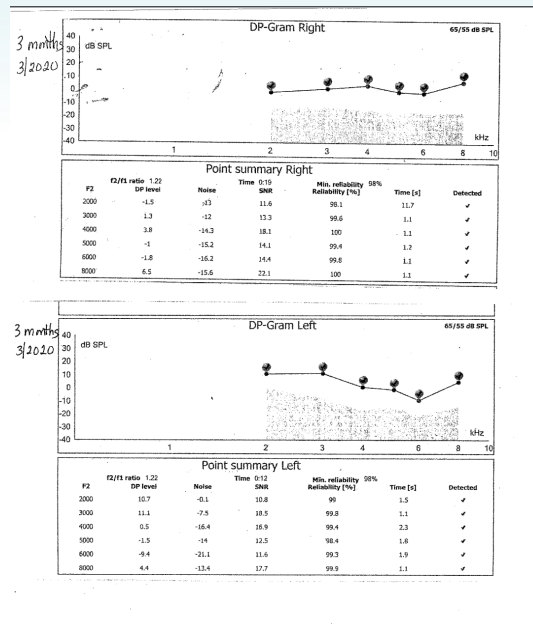


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Diagnosis

- ▶ Seen through ENT clinic
 - ▶ Present DPOAEs bilaterally
 - ▶ Concern for ANSD
 - ▶ Recommendation: Repeat ABR
- ▶ Two Days Later: Repeat natural sleep ABR
 - ▶ Present DPOAEs bilateral
 - ▶ Abnormal ABR
 - ▶ Present Cochlear Microphonic
 - ▶ Recommendation:
 - ▶ Behavioral testing when patient can sit unsupported
 - ▶ Amplification no longer recommended as no threshold information could be obtained through ABR

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Evaluation/Behavioral Testing

- ▶ **1st Attempt**
 - ▶ 7 months old, responding to sounds at home per parent report
 - ▶ Speech Awareness Threshold: 25 dB HL in sound field
 - ▶ Threshold of 35 dB HL at 500 Hz in sound field
- ▶ **2nd Attempt**
 - ▶ 8 months old, responding to sounds at home per parent report
 - ▶ Speech Awareness Threshold: 35 dB HL in sound field
 - ▶ Testing attempted using insert earphones, but patient did not tolerate
- ▶ **3rd Attempt**
 - ▶ 9 months old, responding to sounds at home per parent report
 - ▶ Speech Awareness Threshold: 30 dB HL in left ear using insert earphone
 - ▶ Poor reliability for pure tone testing as patient was observed looking around the room



Evaluation

- ▶ Family switched to another AuD provider.
- ▶ 9 months old and receiving physical therapy and is in the process of scheduling speech therapy.
- ▶ Family reports that patient can hear and responds with head turns to sounds and voices at home
- ▶ **4th Attempt:**
 - ▶ Tympanometry:
 - ▶ Type A (Left): Too noisy for interpretation (Right)
 - ▶ Acoustic Reflexes
 - ▶ Absent AR at 1000 Hz (Left)
 - ▶ Speech Awareness Threshold: 20 dB HL
 - ▶ Threshold at 25 dB HL at 1000 Hz in the sound field



Evaluation

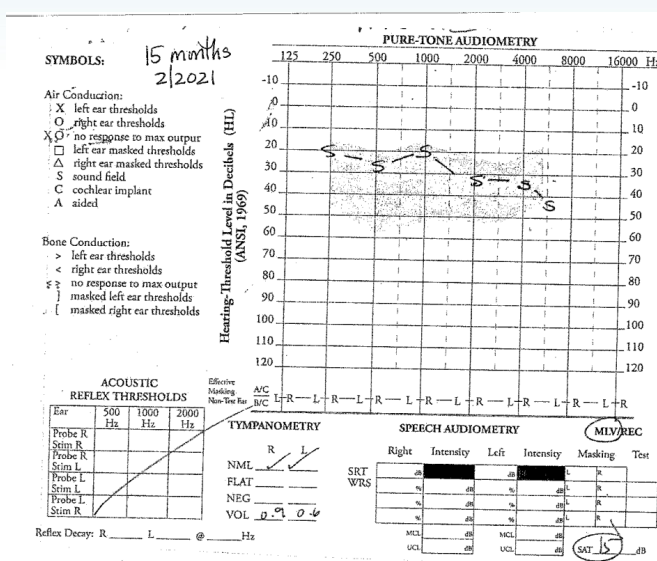
- ▶ Patient was then seen every two weeks for the next three months for behavioral testing
- ▶ The following was obtained:

Ear	500 Hz	1000 Hz	2000 Hz	3000 Hz	4000 Hz	6000 Hz
Right	15	15	20	20	40	DNT
Left	25	25	40	DNT	40	40

- ▶ Patient often did not tolerate inserts, so testing was often completed in sound field
- ▶ Sedated ABR completed in conjunction with PE tube placement at 22 months of age.
 - ▶ Results:
 - ▶ No response at 100 dBnHL using 1000 Hz CE-Chirps
 - ▶ Cochlear Microphonic present using single polarity Chirp stimuli

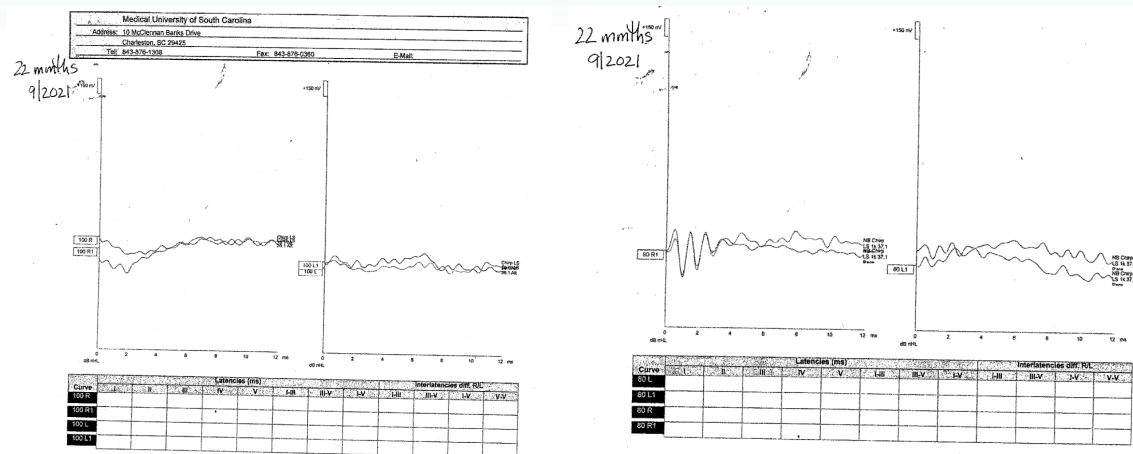
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Behavioral Audiometry – 15 months



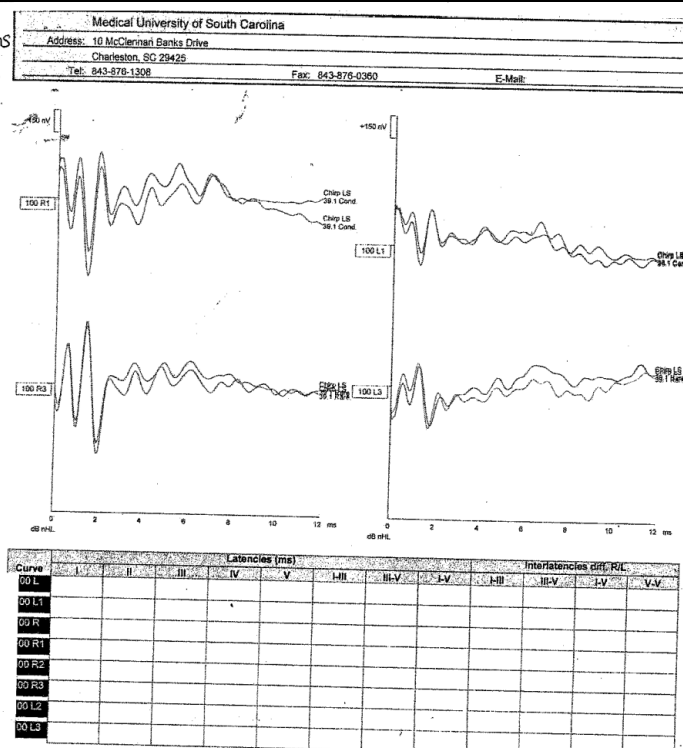
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Sedated ABR- 22 months of age



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22 months
9/2021



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Evaluation – 2 years 1 month old

- ▶ Audiogram was obtained 3 months after sedated ABR
- ▶ Parents reported:
 - ▶ Patient not acting like himself
 - ▶ Speech Regression
 - ▶ Receiving PT and ST through
- ▶ ENT Exam
 - ▶ Left Ear: Draining
 - ▶ Right Ear: PE tube clogged
- ▶ The following results were obtained in sound field:

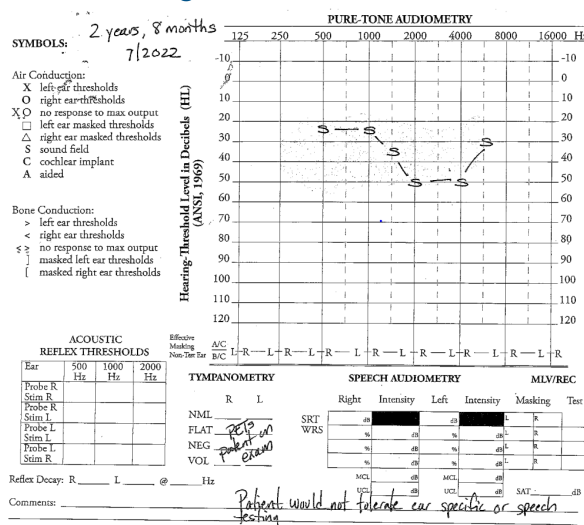
250 Hz	500 Hz	1000 Hz	2000 Hz	4000 Hz	6000 Hz	SAT
30	35	40	DNT	40	50	DNT

- ▶ ENT Recommendations:
 - ▶ Developmental Pediatrics referral (parents declined)

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Evaluation – 2 years 8 months old

- ▶ Behavioral thresholds consistently demonstrated mild/moderate hearing loss.



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Recommendations/Management

- ▶ Audiologist Recommendations:
 - ▶ Hearing Aid Trial
- ▶ Approval for hearing aids obtained through Babynet
- ▶ The hearing aids were never fit.
- ▶ Patient has not been seen for audiometric testing since prior to his third birthday.
- ▶ He was seen for ear tube check at 3.5 years of age.
- ▶ At visit, parents denied concerns for hearing. Audiological follow up was recommended, but patient has not followed up.



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