HISTORY

Name: Mary Madison Age: 3 weeks

Reason for visit: Follow-up evaluation after not passing newborn hearing screening.

Hearing history: Mary did not pass a newborn hearing screening in both ear both during her neonatal stay at UW Medical Center as well as at a rescreening at this clinic on March 31, 2013; she passed the otoacoustic emission screenings in the right ear but did not pass in t

Middle ear history: Mary has a negative history of ear infections.

Family history of childhood hearing loss: Negative.

Medical History: Mary was born at full term (39 weeks) with no complications.

Developmental progress: Mary's parents report that she startles to sounds and awakes to quiet sounds.

Medial Home: Mary lives with her family in Seattle and is followed by her pediatrician, Dr. Sam Smith at Sandpoint Pediatrics.

TEST RESULTS

The BAER and OAE tests were conducted after Mary fell into natural sleep.

Brainstem Auditory Evoked Response

Procedure: The intensity of the stimulus was manipulated to determine the lowest intensity which elicited a detectable response, or threshold of the response. Upon completion of the test, the resulting BAERs were analyzed both in terms of the presence of a response and the latency of waves within each response. BAER threshold values have been corrected to dBeHL values to reflect the relationship between BAER and behavioral thresholds. Both dBnHL and dBeHL values are listed for each intensity level tested.

Normal range: Normal auditory function is defined as BAER thresholds of 0-20 dBeHL.

Results:

Level	Wave V (msec)	Level	Wave V (msec)
Left Ear		Right Ear	
500 Hz		500 Hz	
85 dBeHL (100 dBnHL)**	No response	5 dBeHL (20 dBnHL)	11.70
75 dBeHL (90 dBnHL)**	No response	0 dBeHL (15 dBnHL)	No response
2000 Hz		2000 Hz	
90 dBeHL (95 dBnHL)**	No response	15 dBeHL (20 dBnHL)	8.70
85 dBeHL (90 dBnHL)**	No response	5 dBeHL (10 dBnHL)	No response
4000 Hz		4000 Hz	
95 dBeHL (95 dBnHL)**	No response	10 dBeHL (10 dBnHL)	7.90
90 dBeHL (90 dBnHL)**	No response	5 dBeHL (5 dBnHL)	No response
**measured with			
masking in right ear			
Click			
90 dBnHL at 13/sec	No cochlear microphonic		

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NAME: MADISON, MARY

DOB: 03-25-2013

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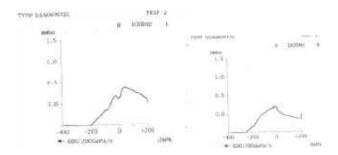
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Immittance and Otoscopy

Procedure: The status of the child's outer and middle ear system was evaluated using tympanometry with a 1000 Hz probe tone. The acoustic reflex/middle ear muscle reflex is a measure of function of the peripheral auditory pathways and was evaluated in each ear using a 1000 Hz probe tone and a broadband stimulus presented ipsilaterally.

Normal range: For tympanometry measures in infants, normal tympanograms are indicated by a positive compliance, but there are no normative values. The normal range of the acoustic reflex/middle ear reflex is at levels of 85 to 105 dB

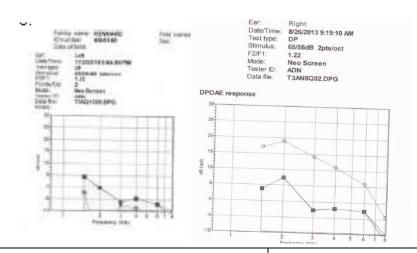
Results: Mary demonstrates normal tympanograms in both ears. Acoustic reflex response threshold is measured at 85 dB in the right ear. Acoustic reflex responses are absent at levels of 85 to 105 dB in the left ear. On otoscopic inspection, the ear canals are noted to be clear bilaterally.



Evoked Otoacoustic Emissions

Procedure: The status of the child's peripheral hearing status was evaluated using both distortion product evoked otoacoustic emissions (DPOAE) and transient evoked otoacoustic emissions (TEOAE). OAEs are acoustic signals generated by the cochlea in response to external auditory stimulation. OAEs are thought to be generated by the outer hair cells within the cochlea and are independent of neural activity. **Normal range:** A normal DPOAE response is indicated by a signal to noise response (SNR) greater than 5 dB and a minimum DP level of -8. A normal TEOAE is indicated by a minimum SNR of 3 dB and an overall reproducibility of greater than 70%. Individuals with normal cochlear function have robust OAEs in response to stimulation from 1000-5000 Hz, whereas individuals with hearing loss greater than 30 dB HL show no OAE in the frequency region of the hearing loss.

Results:



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ASSESSMENT

Mary demonstrates normal hearing in her right ear and a profound sensorineural hearing loss in her left ear. Specifically, she shows BAER tone pip thresholds at 500 to 4000 Hz at levels of 5 to 15 dBeHL in the right ear and no response at equipment limit levels of 85 to 90 dBeHL from 500 to 4000 Hz in the left ear. Mary demonstrates robust otoacoustic emissions in the right ear consistent with normal auditory function, and, in the left ear, she demonstrates absent otoacoustic emissions, consistent with a significant hearing loss. She shows normal outer and middle ear function in both ears on tympanometry, consistent with a hearing loss in the left ear which is sensorineural in nature. Ipsilateral acoustic reflex responses are present in the right ear and absent in the left ear, consistent with normal auditory function in the right ear and a significant hearing loss in the left ear. She shows absence of a cochlear microphonic BAER response in the left ear along with absent otoacoustic emissions in the left ear, supporting that the hearing loss is sensory in origin and auditory neuropathy has been ruled out. Today's results are consistent with her newborn hearing screenings, suggesting that her hearing loss is congenital. The etiology of Mary's hearing loss is unknown at this time.

Today's physiological tests provide information about Mary's auditory system, but do not indicate how Mary makes use of sound. Information about how Mary responds to a range of frequencies in each ear will be obtained in the future with behavioral testing to tonal stimuli.

RECOMMENDATIONS

Mary's parents, Mandy and Michael, were counseled regarding the diagnosis of a unilateral sensorineural hearing loss and its implications on Mary's access to speech and language. It was discussed that infants with normal hearing in one ear and a significant hearing loss in the other ear, often have adequate hearing for developing normal speech and language skills in early childhood, but are at significant risk for communication delays and academic challenges if the hearing loss is not supported with intervention. Children with unilateral hearing loss often have difficulty hearing in challenging listening settings when background noise is high and have difficulty localizing, or finding the source of sound. Mary may benefit from amplification devices as discussed below. The family was given the "Resource Notebook for Families or Children who are Deaf or Hard of Hearing" notebook published by the WA State Department of Health. As a result of these discussions the following recommendations were made:

Hearing Loss

- 1. It is recommended that Mary's hearing status be monitored closely and that she return to this clinic for continued audiological testing to monitor hearing status and to provide additional information about Mary's responses to sounds with both physiological (BAER/OAE) and behavioral audiological testing when developmentally appropriate. Mary will be seen in this clinic every 3 months for the first year of life and then every 6 months. Monitoring hearing status is important to ensure that Mary maintains normal hearing in the right ear for the development of speech and language skills.
- 2. Mary's parents are instructed to be conscious of positioning Mary so that her right ear is not blocked to sound.
- 3. The etiology of Mary's hearing loss is unknown at this time and her parents are advised to pursue additional testing including an evaluation by an otolaryngologist, a CT/MRI scan of the ear, and genetic testing. For these evaluations, she would be referred to Seattle Children's Otolaryngology Clinic (206-987-2105).

Hearing technology

4. Mary is not a candidate for a traditional hearing aid for the left ear due to the profound degree of hearing loss. As she gets older, she will be a candidate for a hearing aid that transmits sound from the left ear to the right ear and a remote microphone/FM-DM system to help her hear in noisy settings.

Intervention

5. The presence of a unilateral hearing loss does qualify Mary and her family for enrollment in an early intervention program for children with hearing loss and their families. Her parents have been referred to the family resource coordinator in their community to help the family learn about these services. The early intervention programs available in the family's community are described within the notebook given to the family.

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PEDIATRIC AUDIOLOGY/CHDD/UWMC

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cc: parents (Mandy and Michael Madison)
primary care physician (Dr. Smith at Sandpoint Pediatrics)
family resource coordinator (Sandy Duncan-King County)
otolaryngology (Seattle Children's Otolaryngology Clinic)

Sam Smith Graduate Audiology Student Clinician

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