AUDIOLOGICAL EVALUATION
TEST DATE: April 15, 2013

HISTORY

Name: Kevin Klein
Age: 6 weeks
Reason for visit: Follow-up evaluation after not passing newborn hearing screening.
Hearing history: Kevin did not pass a newborn hearing screening using the brainstem auditory evoked response (BAER) on March 5, 2006 during his stay in the UWMC NICU; he did not pass the BAER screening in both ears.
Family history of childhood hearing loss: Negative.
Interim History: Parents report that Kevin is healthy today and has had no illnesses since discharge home.
Middle ear history: Kevin has a negative history of ear infections.
Medical History: Kevin was born at term gestation (39 weeks), 4530 grams, with a neonatal history significant for hyperbilirubinemia treated with exchange transfusions.
Medical home: Kevin lives with his parents in Everett and is followed by Dr. Smith at Everett Clinic.

TEST RESULTS

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

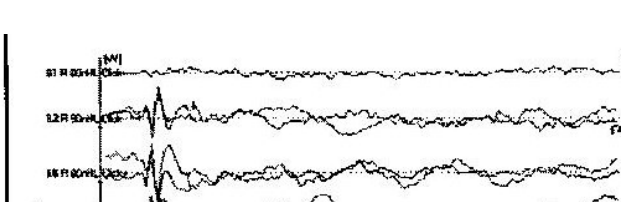
Brainstem Auditory Evoked Response

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 to 20 dBeHL

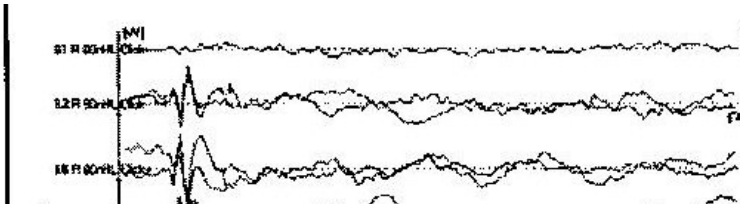
Level	Wave I/ V (msec)		Level	Wave I/ V (msec)
Left Ear			**Right Ear**	
500 Hz			**500 Hz**	
85 dBeHL (100 dBnHL)	**No response**		**85 dBeHL (100 dBnHL)**	**No response**
2000 Hz			**2000 Hz**	
105dBeHL (110 dBnHL)	**No response**		**105dBeHL (110 dBnHL)**	**No response**
Click at 13.3/sec			**Click at 13.3/sec**	
105 dBeHL	**No response****		**105 dBeHL**	**No response****

****BAER testing shows no wave I or wave V responses. However, a cochlear microphonic (CM) response was identified to click stimuli at levels of 80 to 105 dBnHL ; the CM was noted to reverse polarity along with the polarity of the stimulus, and was noted to be absent during a control trial run with the earphone output closed.

Left Ear-Click at 90 dBeHL



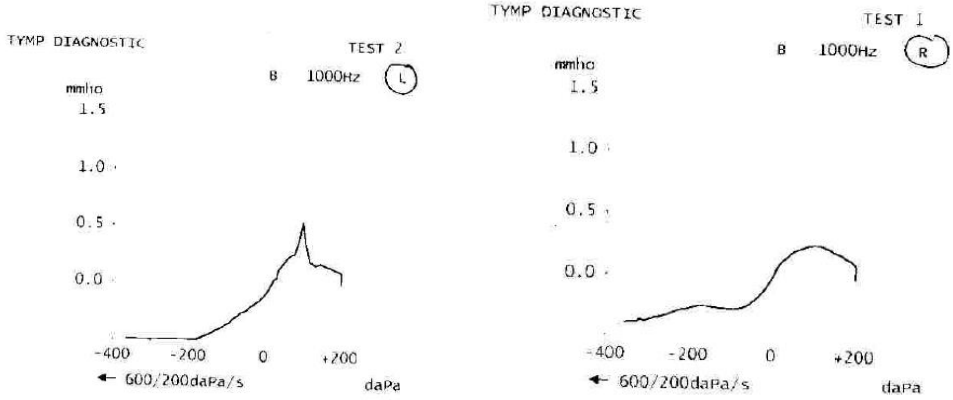
Right Ear-Click at 90 dBeHL



Immittance and Otoscopy

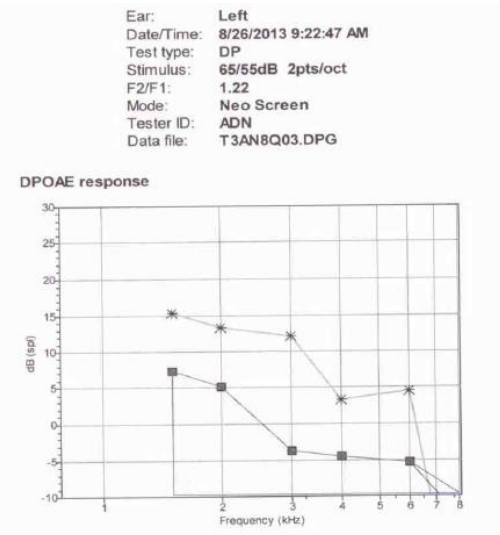
Procedure: The status of the child's outer and middle ear system was evaluated using tympanometry with a 1000 Hz probe tone. The acoustic reflex/middle ear muscle reflex was evaluated in each ear using a 1000 Hz probe tone and a broadband stimulus presented ipsilaterally. On otoscopic inspection, the ear canals were noted to be clear bilaterally.

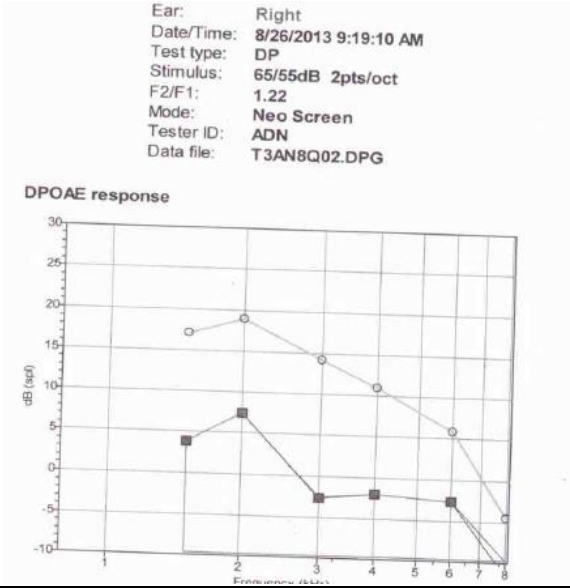
Normal range: For tympanometry measures in infants, normal tympanograms are indicated by a positive compliance, but there are no normative values. The normal range of the acoustic reflex/middle ear reflex is at levels of 65 to 90 dBHL for a broadband stimulus.
Results: Acoustic reflex responses recorded ipsilaterally to a broadband noise stimulus are absent at levels of 65 to 90 dBHL in both ears.



Evoked Otoacoustic Emissions

Procedure: The status of the child's peripheral hearing status was evaluated using both distortion product evoked otoacoustic emissions (OAE). 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 DP OAE response is indicated by a signal to noise response (SNR) greater than 5 dB and a minimum DP level of -8. 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.





ASSESSMENT

Kevin demonstrates auditory neuropathy spectrum disorder (ANSD) in both ears. Specifically, Kevin shows no repeatable BAER responses at equipment limit levels of 90 to 105 dBeHL in both ears. He has cochlear microphonic responses in both ears to click stimuli at 90 dBeHL, consistent with cochlear function in both ears. Kevin demonstrates robust otoacoustic emissions in both ears, also consistent with normal cochlear function, as indicated by DPOAE responses which demonstrate passing criteria from 1500 to 8000 Hz in both ears. He shows normal tympanograms, consistent with normal outer/middle ear function in both ears. Kevin shows absent acoustic reflexes in both ears, consistent with abnormal function of the auditory pathways. Today's results are consistent with the results of his initial newborn hearing screening six weeks ago, indicating no change in his hearing status. The etiology is likely associated with Kevin's severe neonatal hyperbilirubinemia.

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

RECOMMENDATIONS

Kevin's parents, Kate and Karl, were counseled regarding the diagnosis of auditory neuropathy spectrum disorder (ANSD) and its implications on Kevin's speech and language development, and education. It was discussed that children with ANSD show a range of behavioral responses to sound, ranging from near normal responses to no responses to sound. The decision to provide an individual child with amplification is determined by behavioral testing when the child is developmentally able to provide reliable responses. However, if there are concerns that it will be some time before reliable measures can be obtained, then a trial with low gain hearing aids may be appropriate. Children with ANSD benefit from early intervention services to enhance communication skills through parent support and education. The family was given the "Resource Notebook for Families of Children Who are Deaf or Hard of Hearing" 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 Kevin's hearing and status of his auditory system be monitored for any change in function and to provide additional information about his responses to sounds with behavioral audiological testing when developmentally appropriate: Kevin will be seen in this clinic every 3 months for the next year.

2. The etiology of Kevin's hearing loss is thought to be associated with neonatal hyperbilirubinemia, however, additional testing could be conducted including an evaluation by an otolaryngologist, a CT scan or MRI of the auditory system, and genetic testing. For these evaluations, Kevin is referred to Seattle Children's Otolaryngology Clinic (206-987-2105).

Hearing Technology

3. Kevin's candidacy for hearing aids will be determined by behavioral hearing testing starting at 6 months developmental age.

Intervention

4. Referral to an early intervention program for children with hearing impairment and their families. The family was given information about early intervention services available in their county of Snohomish. They have been advised to find out about all the programs available so that they can choose a program that is a good fit for their family and their goals for their child.
5. The family has been referred to a family resource coordinator in their community to assist the family with in early intervention resources.

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cc: parents (Kate and Karl Klein)
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family resource coordinator (Sandy Smith-King County)
otolaryngologist (Seattle Children's Otolaryngology)