

CHILD DEVELOPMENT AND REHABILITATION
CENTER

P.O. Box 574, Portland OR 97207-0574

Account No.

Medical Record No. 01-38-94-88

Name Wulf, Matthew A

Birthdate 12/03/1997

CDRC

CLINIC DATE: 03/25/98

CLINIC NAME: METABOLIC CLINIC

DISCIPLINE: PEDIATRIC AUDIOLOGY

REFERRAL SOURCE: Metabolic.

BACKGROUND INFORMATION: Matthew, a 3-1/2-^{month}~~year~~-old, with infantile Refsums (Russlen) syndrome was seen today for audiological assessment. He was born at Emanuel Hospital and did have an otoacoustic emissions hearing screening. The results of this evaluation were consistent with hearing loss. However, parents noted that Matthew seems to be responding to a variety of stimuli in the home environment. They do have an older child who has Refsums (Russlen) syndrome with associated sensorineural hearing loss.

CONDITION OF HEARING: Matthew was evaluated today using informal behavioral observations. During testing, he was wide awake which is not considered to be the optimal state for this assessment. When presented with a 3,000 hertz narrow band noise burst, Matthew showed no orienting response to presentations at 60 or 90 decibels. However, when presented with a 100-decibel tone burst from a tricorn, there was a definitive startle response. Subsequent middle ear study using tympanometry shows normal middle ear pressure and compliance in the right ear and significant reduced mobility in the left. On otoscopy, the right tympanic membrane could be observed while the left canal is significantly more stenotic making visualization difficult.

DIAGNOSTIC IMPRESSIONS: Today's test results allow us to rule out a profound low frequency hearing loss in the better ear. However, based on his risk categories, Matthew is still at risk for high frequency hearing loss as well as progressive hearing loss.

RECOMMENDATIONS: These test results were shared with Mr. and Mrs. Wulf. I recommend that they have an otolaryngology consultation. This was arranged with Dr. Milczuk for April 14. In addition, I do recommend ABR testing to further assess auditory status.



Donald Plapinger, Ed.D.
Pediatric Audiologist

DP:sgr

D: 03/25/98

T: 03/26/98

cc: Henry Milczuk, M.D.
Assistant Professor,
Otolaryngology/Head
and Neck Surgery
and Pediatrics