

**Case Report:  
AUDITORY NEUROPATHY IN Dr. SOETOMO HOSPITAL**

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**ABSTRACT**

*Auditory neuropathy or auditory dys-synchrony is a rare hearing disorder in which sound enters the inner ear normally but the transmission of signals from the inner ear to the brain is impaired. It can affect people of all ages, from infancy through adulthood. People with auditory neuropathy may have normal hearing, or hearing loss ranging from mild to severe, they always have poor speech-perception abilities, meaning they have trouble understanding speech clearly. Often, speech perception is worse than would be predicted by the degree of hearing loss. In this paper will be reported two cases of auditory neuropathy (8 years old and 5 years old) who were referred to Dr. Soetomo Hospital. Their teachers were questioning about their hearing ability. The diagnosis was determined by behavioural observation audiometry, otoacoustic emission, auditory brainstem response, tympanometry and speech perception testing with live voice. One of the 2 is cases multihandicap with a moderate intelligence quotient (36). The management depends on the patient’s condition, mode of communication using visual input such as sign communication or cued speech and hearing aid usually does not show too much benefit since the first case is multihandicapped and the second case will be evaluated in 3 month for communication. The choice for schooling is different in these cases.*

**Keywords:** auditory neuropathy, diagnosis, management

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**INTRODUCTION**

Auditory neuropathy (AN) is a relatively new diagnosis in the field of audiology and has been the subject of several studies in recent years. Berlin et al (2002), suggested the term auditory dys-synchrony (AD) as a more accurate definition of the disorder for some patients. Whether is called (AN) or (AD), this new diagnosis has been challenging for professionals and parents as they try to identify the most appropriate test battery and management strategies for each patient. The term AD would therefore include both true AN (*i.e.* a true neural abnormality) and other possible underlying mechanisms resulting in neural dys-synchrony, as well as delayed maturation of the lower level auditory pathway. AN is a disorder that affects neural processing

of auditory stimuli that will reduce a child’s ability to understand speech and may affect ability to detect sound to various degrees.

**CASE REPORT**

**Case 1**

An 8-years-old girl was referred to Audiology Department by her teacher for hearing evaluation. Her mother questions about her hearing, since she seems able to hear but seldom answers the question. The history of: prenatal, prenatal and postnatal periods are remarkable. Evaluations of verbal receptive and expressive function were limited.

Table 1. Hearing evaluation results

	Right ear	Left ear
Behavioural Observation Audiometry	minimum response level at 90 dB HL	
Otoacoustic Emission (OAE)	Refer	Pass
Auditory Brainstem Response (ABR)	Wave V at 100 dBHL	Wave V is not detectable at 100 dBHL
Tympanometry	Type A, normal compliance	Type A
Acoustic stapedial reflex, ipsilateral	Absent	absent

Intelligence quotient 36 (moderate mental retarded)

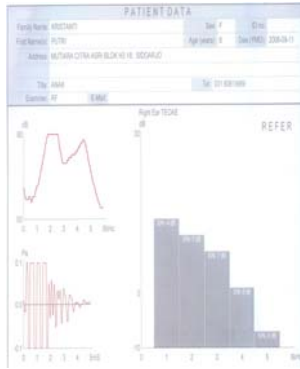


Figure 1. Right OAE

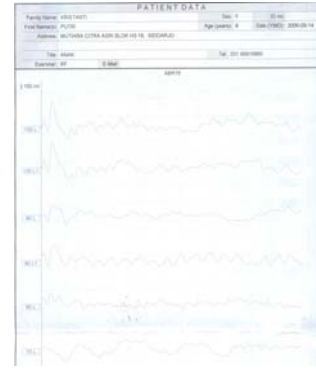


Figure 3. Left ABR



Figure 2. Left OAE

**Case 2**

A 4-years old girl was referred to Audiology Department by ENT specialist from Kalimantan because she does not speak properly at her age. According the her mother, her daughter is able to speak properly in quite environment even with a low intensity of sound such as whispering, however listening ability is poor in a noisy environment.

There is no history of any problem during prenatal, prenatal and postnatal periods. Evaluation of verbal receptive and expressive function were good enough, the communication without lips reading (the intensity of sound around 40 dBHL

Table 2. Hearing evaluation results

	Right ear	Left ear
Behavioural Observation Audiometry	minimum response level at 50 dBHL	
Otoacoustic Emission (OAE)	Refer	Refer
Auditory Brainstem Response click	Wave V at 100 dBHL	Wave V at 80 dBHL
A B R tone burst	Wave V at 80 dBHL	Wave V at 80 dBHL
Tympanometry	Type A	Type A
Acoustic stapedial reflex	absent	absent

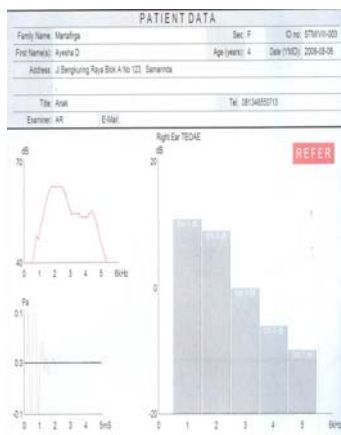


Figure 4. Right OAE

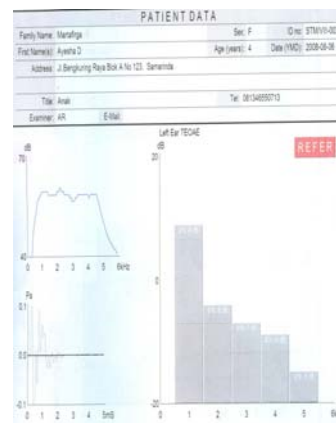


Figure 5. Left OAE

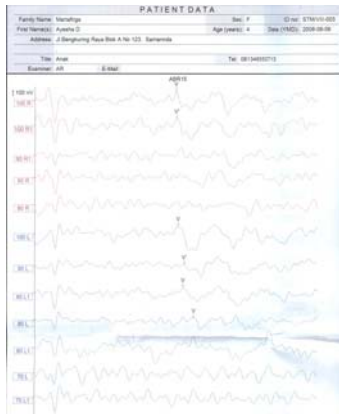


Figure 6. Right ABR

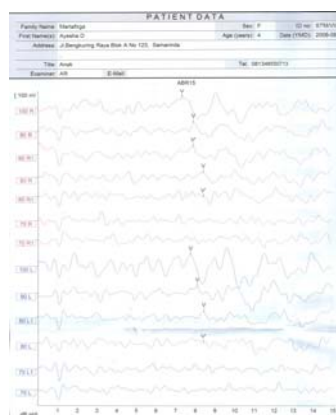


Figure 7. Left ABR

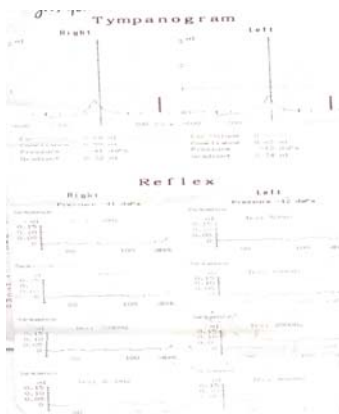


Figure 8. Tympanogram

## DISCUSSION

Singer (2002) estimates that AN occurs in about 1 in 10 children who have hearing loss and severely abnormal ABR i.e. one in ten of those who, because of absent ABR, might at first sight be thought to have

severe/profound sensorineural (cochlear) hearing loss . The true prevalence of AN in the paediatric population with hearing loss, however, is unknown.

Although most AN cases are thought to occur among special care/neonatal intensive care babies, they can occur in the well-baby population and may of course pass the OAE screen. Risk factors for auditory neuropathy are hyperbilirubinaemia, anoxia, hypoxia/prolonged assisted ventilation, extreme prematurity (< 28 weeks), Congenital brain anomalies, demyelinating conditions such as multiple sclerosis, syndromes associated with other peripheral neuropathies (Charcot-Marie-Tooth, Friedrich's Ataxia), genetic factors. Auditory neuropathy has also been reported in cases where it is related to a viral infection or exacerbated by fever. Genetics also play an important role in the aetiology of AN. Families have been identified with siblings with AN. In addition, there are also parents with AN who have children with this disorder (Siniger, 2001; Hood, 2002, Sutton, 2004). Therefore, AN appears to follow both recessive and dominant inheritance patterns (Hood, 2002).

Only 27 % of the patient with AN have no associated medical conditions or family history (Siniger, 2001). However, although these possible risk factors have been observed and information about their occurrence should be obtained, it is crucial to remember that AN is also found in children with no risk factors (Hood, 2002).

These two cases presented there are no risk factors. The site of lesion for AN is not completely understood. However, the characteristics of AN most likely reflect more than a single aetiology, thus, the disorder may more accurately be described as auditory neuropathies (Hood, 1998). There are a number of pathologies that could produce the AN result profile. Some of these include insult specific to the cochlear inner hair cells, abnormality of inner hair cells, auditory nerve fibre synapse, spiral ganglion cell, depleted neuronal populations in the auditory brain stem and demyelination of the auditory nerve (Knox, 2005).

A characteristic of auditory neuropathy is difficulty hearing in noise. Greater difficulties hearing in competing noise than behavioural audiogram would suggest, other features indicative of auditory processing difficulties (Krauss, 2001; Roush, 2007).

In the second case, her mother complains that the patient can not speaking in noise, this is characteristic of auditory neuropathy.

How is auditory neuropathy diagnosed? To diagnose AN use a combination of several methods. These

include tests of otoacoustic emissions (OAE), auditory brainstem response (ABR) and the other tests. Other tests may also be used as part of a more comprehensive evaluation of an individual's hearing and speech-perception abilities (Hood, 1998; Knox, 2005; Sutton, 2004).

Otoacoustic emissions (OAE): An OAE test uses a small, very sensitive microphone inserted into the ear canal to monitor the faint sounds produced by the outer hair cells in response to stimulation by a series of clicks. This test measures the integrity of the outer hair cells of the cochlea and cochlear function. Cochlear microphonics (CM) tests the function of the cochlea similarly.

Auditory brainstem responses (ABR): This test uses scalp electrodes to measure electrical activity in response to a click sound. An ABR test monitors brain wave activity in response to sound using electrodes that are placed on the person's head and ears. Abnormal results of ABR testing indicate that the hearing nerve, as well as the brainstem nuclei, may not correctly process the sounds.

ABR and OAE testing are painless and can be used for newborn babies and infants as well as older children and adults.

Pure tone audiogram testing is a graphic plot of a patient's thresholds of auditory sensitivity for pure tone (sine wave) stimuli. It does not test a patient's ability to process sound. This test shows only the patient's ability to hear sounds or tones.

Speech audiometry use spoken words and sentences rather than pure tones. Tests are designed to assess sensitivity (threshold) or understanding (intelligibility) of speech.

Acoustic reflex (AR) measures the contraction of the stapedius muscle in the middle ear. Deviation from the normal threshold on AR testing indicates potential abnormalities of the hearing nerve and auditory system.

Criteria for the diagnosis of AN are as follows (1) normal outer hair cell function as determined by OAE or CM; (2) absent or severely abnormal ABR test results at maximal stimulus (100 dBnHL); (3) absent or elevated acoustic stapedial reflex thresholds.

Suspect AN in older child or adult with the following audiologic findings: Pure tone thresholds are abnormal. The entire range of abnormalities, from near-normal to profound, may be seen. A more severe loss is usually displayed in the lower frequency thresholds. The

audiogram findings may vary some, but the overall milieu usually remains unchanged. Younger children who are not cooperative, the examination performed by Behavioural Observation Audiometry.

Poor speech discrimination scores are out of proportion with the level of loss suspected based on the pure tone average.

Table 3. The hearing evaluation test results

Test	Outcome
Otoacoustic emission	Normal
Auditory Brainstem Response	Absent (or severely abnormal)
Pure Tone Audiogram / Behavioural Observation Audiometry	Normal to severe-profound hearing loss (Sometimes can be asymmetric)
Acoustic Stapedial Reflexes :	
- Ipsilateral	Absent
- Contralateral	Absent
Speech recognition : - In quiet	Variable
- In noise	Generally poor

The majority of practicing audiologists agree that OAE should be present in AN patients. However, 6.3% disagree and believe OAE should not be present. Some patients with AN lose their OAE over time but this does not appear to be related to any change in the audiogram for AN patients (Knox, 2005).

While most cases of AN identified to present are bilateral, sometimes few patients have unilateral AN (asymmetric). 75.3% of the audiologists agreed that unilateral AN can exist, however 24.7% did not believe unilateral AN was possible. Unilateral AN/AD patients display normal auditory function in one ear, while there are consistent patterns of AN in the other ear (Hood, 2004).

Podwall et al (2002) reported a child with unilateral AN, an 11-year-old boy, who was referred for an audiologic evaluation after failing his annual hearing screenings in his left ear. At age 7 years (when OAE were not available at this particular facility), the patient was diagnosed with normal hearing sensitivity for the right ear and a severe to profound sensorineural hearing loss for the left ear by pure tone audiometry testing. At age 9 years, OAE were performed and the emissions were present bilaterally, however the presence of OAE in the left ear was unexpected. At age 10 years, the auditory evoked potentials testing were elicited at 80-90 dBHL. Tympanometry revealed normal middle ear compliance bilaterally, while ipsilateral acoustic reflexes were present in the right ear and absent in the left ear. This

case is unique as the presence of OAE with profound hearing loss, absent acoustic reflexes, and absent brainstem auditory evoked response testing suggests the presence of AN only in his left ear. This is significant because there have not been many cases of unilateral AN documented in previous literature.

In the first case, possibility were (1) this case might lose the right OAE over time (left OAE pass), according to Knox (2004) that some patients will lose their OAE over time; (2) unilateral, because of her IQ (36) so she did not cooperative, BOA test can not show the patient's ability to hear sounds or tones; (3) bilateral asymmetric.

The treatment of patients with auditory neuropathy starts with the parents. The key issues in management are information and support for the family (particularly in view of the uncertainties around prognosis), the need for repeated audiological and communication assessment, and decisions about hearing aids or other intervention. Children with AN need reassessment at frequent intervals to determine whether the problem is due to maturation or not, and because of potential fluctuations in auditory function. Children with AN are at risk for communication difficulties and need to be monitored accordingly. The management of the child with AN requires a multidisciplinary team approach which may involve some or all of the following audiological physician, ENT/otolaryngology, pediatrician, neurologist, geneticist, speech-language therapist, teacher of the deaf, and whole member of families (Knox, 2005; Roush, 2007).

Intervention involved were (1) conventional hearing aids The decision on whether to aid can only be based on behavioural results and observations from families regarding the child's responses to sound. Hearing aid benefit should be determined. Benefit is not determined based only on aided detection thresholds, but on the development of speech perception skills. Monitoring the child's hearing aid fitting is very important.; (2) FM systems. FM systems (with or without personal hearing aids) have been found to be beneficial for children with AN.; and (3) cochlear implants.

Communication Methods (Knox, 2005; Roush, 2007): (a) language development is critical, work closely with speech/language pathologists, educators; (b) visual communication methods (cued speech/sign language) are necessary for language development; (c) Auditory Verbal Therapy by itself is ineffective, it will be performed after cochlear implantation.

In this first case, the management are as follows: (1) she will be observed for 3 months at school, learning for communication by cued speech; (2) until now, hearing

aid still considered or it's better if she use assistive listening device (FM); (3) schooling in SLB C because her IQ is 36; (4) evaluated in 3 months.

Management for the second case were (1) re-evaluation on hearing and speech development in 6 months, because her communication is good enough; (2) learning for communication supported by visual cues and it's better to reduce noisy surrounding listening situation; (3) hearing aid is not necessary until know, except there is a hearing impairment in the future.

## CONCLUSIONS

Auditory neuropathy is identified by appropriate physiological measures of auditory function. Although variation among patients is considerable, a common characteristic is an absence of neural synchrony, as shown by absent ABR and evidence of outer hair cell function, reflected by the present of OAE.

Auditory neuropathy has an unpredictable course, and the disorder is manifest in a unique way for each patient affected. In this cases management is depend on the condition and each patient has difference management.

## REFERENCES

- Berlin C, Li L, Hood L, et al, 2002. Auditory Neuropathy/Dys-Synchrony after the diagnosis, then what? *Seminars in Hearing*. 23(3), 209-214.
- Hood L, 1998. Auditory neuropathy: What is it and what can we do about it? *The Hearing Journal*. 51(8), 10-18
- Hood L, 2002. Auditory neuropathy/auditory dys-synchrony : New insights. *The Hearing Journal*. 55(2), 10-18
- Knox EN, 2005. Diagnosis and management of auditory neuropathy / dys-synchroni. A capstone project submitted in partial fulfilment of requirement for the degree of Doctor of Audiology. Washington.
- Krauss N, 2001. Auditory Neuropathy : An Historical and Current Perspective. In Sininger Y & Starr A. Eds, *Auditory Neuropathy: A new perspective on hearing disorders*. San Diego, 1 - 10
- Podwall A, Podwall D, Lamendola P, et al, 2002. Unilateral Hearing Loss. *Journal of Child Neurology*. 17(4), 306-309
- Roush P A, 2007. Differential Assessment and Management of Auditory Neuropathy/Auditory Dys-synchrony in Infants and Young Children. August 4, Power point presentation.
- Sininger Y, Oba S, 2001. Patients with auditory neuropathy : Who are they and what can they hear? In

: Sininger Y, Starr A. Eds. Auditory Neuropathy : A new perspective on hearing disorders. San Diego, 15-36.  
Sininger Y, 2002. Identification of Auditory Neuropathy in Infants and Children. Seminars in Hearing. 23(3), 193-200.

Sutton, 2004. Newborn Hearing Screening and Assessment. Assessment and Management of Auditory Neuropathy / Auditory Dys-synchrony. A Recommended Protocol.