LITERATURE REVIEW

Hearing loss in children – literature review and two clinical cases

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ABSTRACT

BACKGROUND. Hearing disorders in new-borns and small children can be difficult to recognize and often go undetected until older age. Speech development is closely related to the hearing ability and, for this reason, hearing disorders during this formative period can have specific consequences in terms of language and personality development.
CASE REPORT. This article presents a brief literature review on this topic and illustrates the general data with two cases. The first case refers to a 4 year-old girl, who was previously diagnosed with retardation and left spastic paresis. Despite physical, mental and hearing difficulties, this child was not enrolled in a kindergarten for children with disabilities, but integrated successfully in a kindergarten for healthy children. The second case refers to a 6 year-old boy previously diagnosed with mental and speech retardation in the Paediatric Neurology Department. The particularity of this case was that the child spoke until 3 years old when he suffered a shock consecutive to a verbal assault.
CONCLUSION. Hearing loss influences speech and language development of infants and young children, and these effects begin within the first 6 months of life. Early diagnosis of hearing loss, coupled with appropriate intervention and management, is necessary for children to reach their full communicative and educational potential. In such cases, multidisciplinary approach (ENT specialist, audiologist, logopedist and psychologist) is recommended.
KEYWORDS: hearing loss, brainstem evoked response audiometry (BERA), auditory steady state response (ASSR).

INTRODUCTION

In the first 6 years of life the human brain develops its acumen for language. By 3 months of age the baby should stir in his/her sleep to nearby conversation. If awake, the baby should quiet to the sound of mother's voice. By 6 months the baby should be able to rouse from his/her sleep at loud noises and turn toward the direction of the sound. By 9 months the child should enjoy playing with a bell or rattle and look up when called by name. The child at this age should be able to utter several different sounds with changing pitch. By 12 months the child should be able to understand a variety of simple words, such as "bye-bye", "no", "yes" and also should imitate some speech sounds and even say two or three words at right times and places, such as "da-da" or "ma-ma".

By 18 months the child should be able to say more than six words and follow simple commands. By 2 years the child should be learning some new words every day. By 3 years the child should be using two and three-word sentences. Family members should be able to understand the child's speech and by the age of 4 years the child should use sentences of four or more words and people outside the family should be able to understand the child's speech[1-3].

Some of the more common syndromes disorders that can result in congenital hearing loss in children are presented in Table 1[4-7].

Sensorineural hearing loss can also result from teratogenic effects of congenital infections on a mother during embryologic development of the fetus. Congenital infections most commonly associated with hearing loss include[8-10]:

- cytomegalovirus (CMV),
- human immunodeficiency virus (HIV),
- rubella,
- syphilis,
- toxoplasmosis.

Improved perinatal medical care has resulted in more surviving infants, but with complications that may include hearing loss. There have been identified a number of factors that are related with a high risk of hearing loss in infants, such as very low birth weight, prematurity, neonatal jaundice[11-15].

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Table 1
Syndromes disorders associated with congenital hearing loss in children

Syndromic disorder	Genes	Clinique symptoms	Scanner
WAARDENBURG	5 genes known	autosomal dominant disorder characterized by lateral displacement of the medial canthi, increased width of the root of the nose, multicoloured iris, white forelock, and mild-to-severe sensorineural hearing loss.	Normal or sometimes abnormal semicircular canals
BRANCHIO-OTO-RENAL	3 genes known	autosomal dominant disorder consisting of branchial clefts, fistulas, and cysts, renal malformation and conductive, sensorineural or mixed hearing loss.	
PENDRED	1 gene	autosomal recessive endocrine metabolism disorder resulting in goiter and congenital, symmetric, moderate to profound sensorineural hearing loss.	Mondini malformation
USHER	A large number of genes	autosomal recessive disorder characterized by congenital sensorineural hearing loss and progressive loss of vision due to retinitis pigmentosa.	Normal

The smaller the age is, the more difficult it is for the doctors to diagnose hearing loss in children (Diagram 1, Diagram 2). There are various audiologic tests used to diagnose hearing loss in infants and children[1,8,16-19].

Otoacoustic emissions (OAE). The vibrations produced by the biomechanical amplifier of the cochlea, either spontaneously or in response to an acoustic stimulus, are transmitted in a retrograde fashion across the ossicles to the tympanic membrane, which acts like the membrane of a loudspeaker, emitting the vibrations as sound waves into the external ear canal. A sensitive microphone probe inserted into the ear canal can detect these active cochlear vibrations, which are called otoacoustic emissions. OAE are clinically important because they can be used to test the function of the "cochlear amplifier". The emissions reflect the functional integrity of the cochlea, the outer hair cells being a particularly important source of OAEs. The most important application of OAE is for screening cochlear function in newborns, infants and small children, because the majority of hearing disorders in this age group have a cochlear etiology. In the absence of OAEs, additional audiologic tests, such as auditory steady state response (ASSR) and brainstem evoked response audiometry (BERA), should be used.

Auditory steady state response (ASSR) is an objective test used for evaluation of hearing ability in children too young for traditional audiometric testing. The person being tested must be very quiet and still in order to obtain reliable ASSR results. Often, testing is performed under sedation or in natural sleep in case of pediatric patients. Results are obtained by measuring brain activity while the person listens to tones of varying frequency (pitch) and intensity (loudness). The brain activity is recorded using electrodes taped on the forehead and behind each ear. The use of electrodes eliminates the need for active participation of the patient (i.e. pushing a response button every time a tone is activated), the results being detected objectively using statistical formulas that determine the presence or absence of a true response. Similar to traditional audiometric testing, threshold is determined as the lowest level at each frequency at which a response is present. ASSR provides an accurate, frequency-specific estimation of a pure-tone audiogram.

Brainstem evoked response audiometry (BERA) is an electro-physiological test procedure performed in children, especially newborn infants, to check hearing loss or deafness. BERA test is actually a painless and safe procedure. The stimulus, either in the form of clicks or tone pip, is transmitted to the ear via a transducer placed in the insert ear phone or head phone. The wave forms of impulses generated at the level of brain stem are recorded by the electrodes placed over the scalp. When recorded, these impulses contain a series of peaks and troughs. The positive peaks (vortex positive) are referred to by the Roman numerals I - VII. These peaks are considered to originate from the following anatomical sites:

1. Cochlear nerves - waves I and II
2. Cochlear nucleus - wave III
3. Superior olivary complex - wave IV
4. Nuclei of lateral lemniscus - wave V
5. Inferior colliculus - waves VI and VII.

BERA has 90% sensitivity and 80% specificity in identifying cases of acoustic schwannoma. The sensitivity increases proportionally to the size of the tumor.

Behavioural observation audiometry (BOA). This test technique is suited to infants aged less than 7

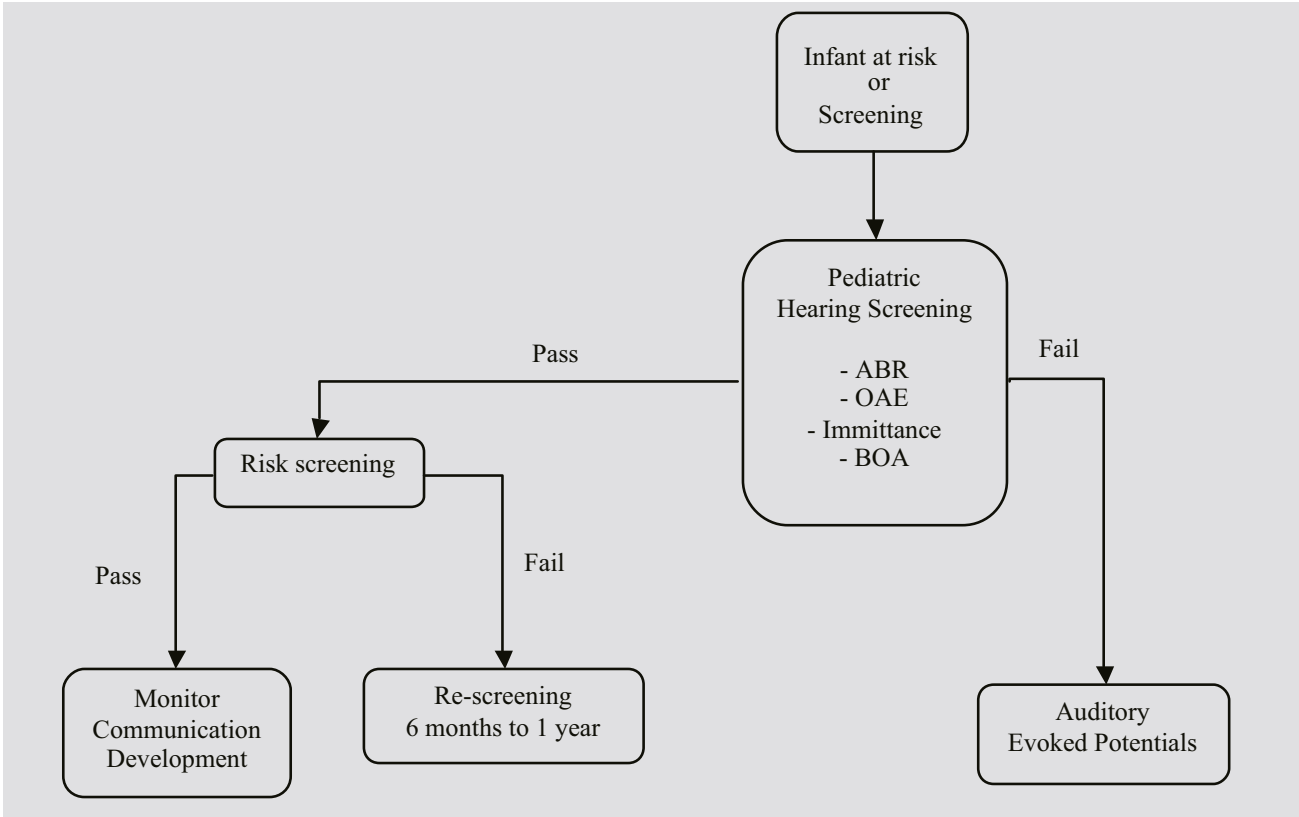

Diagram1 Hearing consultation model for infants aged 0-6 months. The model begins with screening, followed by re-screening and then by assessment of hearing sensitivity of those who do not pass the screening (according to Brad A. Stach[8]).

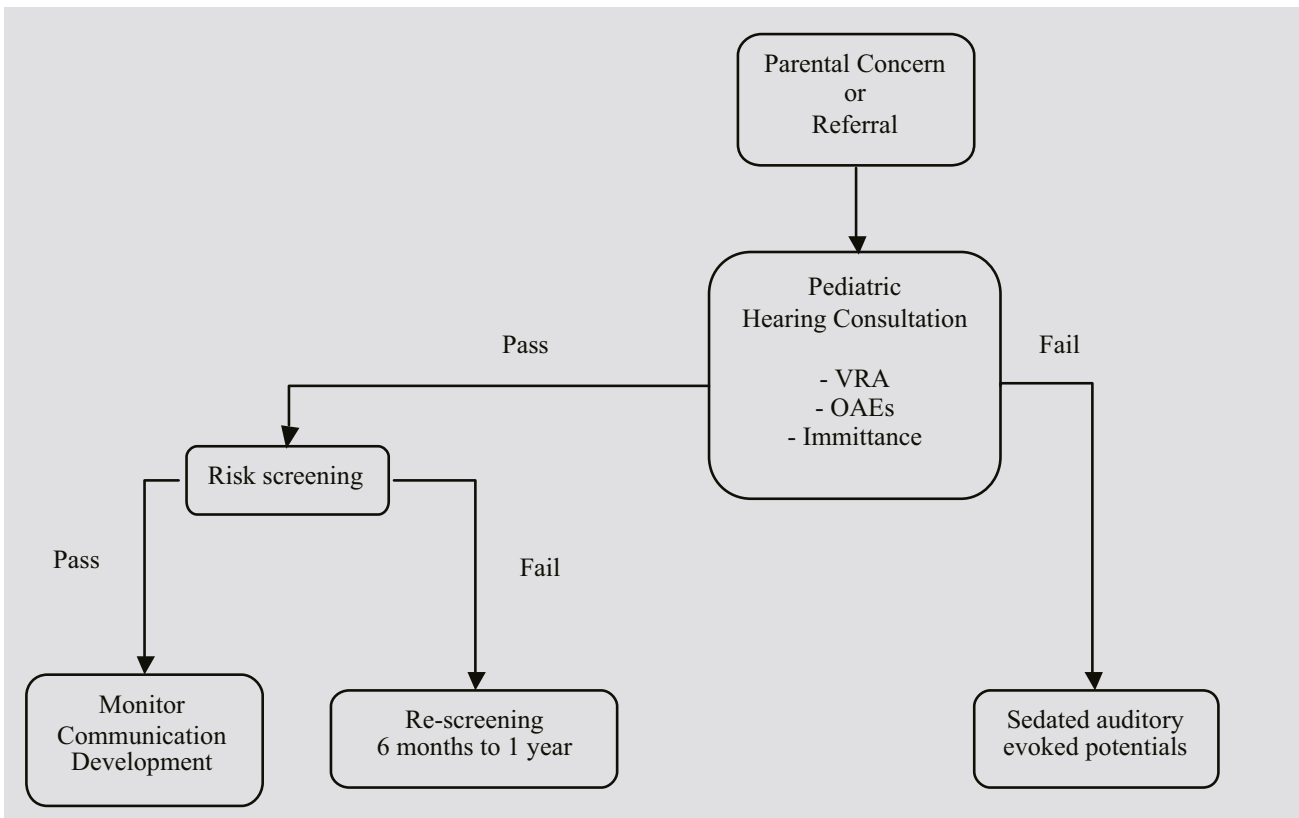

Diagram 2 A paediatric consultation model for assessing children aged 6 months to 2 years. The model begins with screening, followed by assessment of middle-ear function and hearing sensitivity in those who do not pass the screening (according to Brad A. Stach[8]).

months. During the test, one audiologist makes a sound, making sure that the child cannot see him/her, while a second audiologist watches for any change in the child's behaviour (e.g. sudden reflexive movement, eye blinks or changes in sucking patterns). The type of sound is recorded together with its intensity and the nature of the behaviour change.

Visual reinforcement audiometry (VRA) is an audiometric test used in paediatric assessment, in which an appropriate response to a signal presentation, such as a head turn toward the speaker, is rewarded by the activation of a light or lighted toy.

Tympanometry is not a test for hearing sensitivity, but a procedure that can evaluate the eardrum compliance, the middle ear pressure and air volume, when a soft sound and air pressure are introduced in the ear canal. It's helpful in identifying middle ear problems in children, such as Eustachian tube dysfunction or middle ear effusion[20-23].

CASE REPORT

Case 1

A 4-year-old girl was referred to our ENT Department with psychomotor retardation, left spastic paresis and a suspicion of bilateral mild-to-severe sensorineural hearing loss. The hearing loss appeared to be caused by the cytomegalic virus (CMV) or the cytomegalic inclusion disease, a viral infection usually transmitted in utero. There was no family history of hearing loss and no other significant medical history.

The otoendoscopic examination revealed a normal tympanic membrane in both ears.

For a concrete evaluation of the auditory function, we proceeded to perform a BERA (brainstem evoked response audiometry) examination, whose results are showed in Figure 1. The absence of I - V waves was observed on both ears, these changes being suggestive for cophosis or a profound neural hearing loss.

The second test we performed, for confirming the initial diagnosis, was an auditory steady-state response (ASSR) (Figure 2). The examination result sustained the diagnosis of bilateral cophosis.

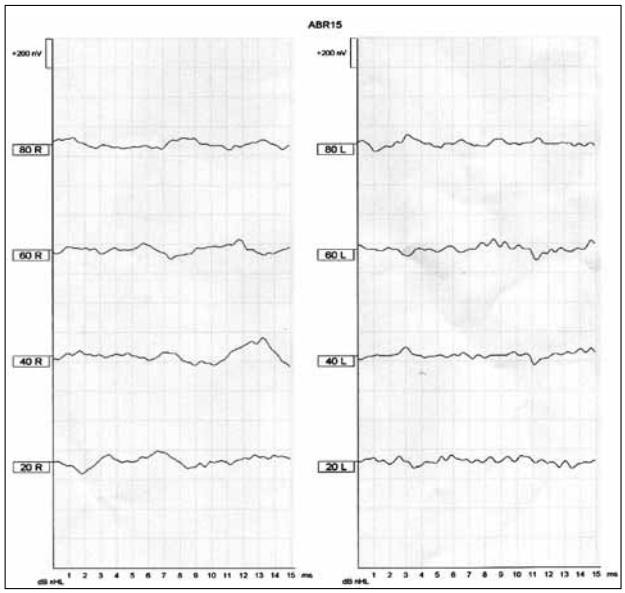

Figure 1 BERA results – absence of I-V waves on both ears

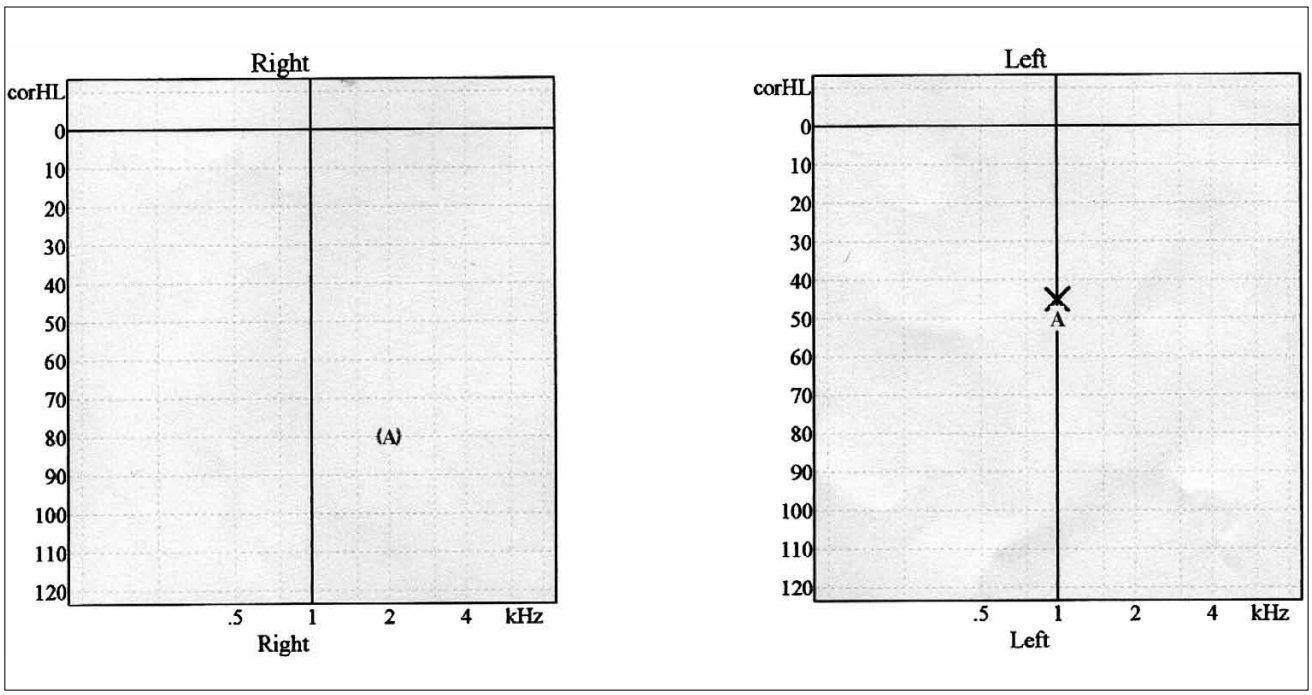

Figure 2 ASSR results – bilateral cophosis

Despite physical, mental and hearing difficulties, this child was not enrolled in a kindergarten for children with disabilities but successfully integrated in a kindergarten for normal children. This child is not likely to develop speech and language normally and she may be at risk for academic achievement problems. The parents were provided with information about the nature of the disorder and the strategies that can be used for achieving an almost normal evolution.

This is a case where multidisciplinary approach (audiologist, logopedist and psychologist) is mandatory.

Case 2

We present the case of a 6-year-old boy diagnosed with mental retardation and language disabilities. This child spoke until 3 years old, normally for his age, when he suffered a shock after a verbal assault. It should be noted that the boy's mother was known with mental retardation.

The CT scan examination revealed no recent changes in vascular space and no tumoral processes.

The otoscopic examination revealed a diffusely hyperemic tympanic membrane on both ears, with the absence of the cone of light.

Tympanometry results, shown in Figure 3, revealed a middle-ear disorder, characterized by flat, Type B tympanogram and absent stapedial reflex bilaterally. These results indicated an increment in the mass of the middle-ear mechanism, a result that is equal to the presence of effusion in the middle-ear.

ASSR evaluation performed in this particular case (Figure 4) indicated a mild hearing loss of the right ear and a medium hearing loss for the left ear. Correlating these findings with the tympanometry results, a sensorineural hearing loss was excluded.

The particularity of the case is that the child spoke until 3 years old. After the verbal assault he was exposed to, the parents noticed the behavioural change.

According to our clinical and paraclinical evaluation, the primary diagnose was otitis media with effusion, the child being a perfect candidate for surgical treatment – bilateral myringotomy and tympanostomy tube placement. After this procedure, patient's hearing is likely to improve up to normal. The child is capable of acquiring normal speech, but he will need psychological counselling and logopedic intervention.

The patient will be re-evaluated periodically, especially during the early academic years.

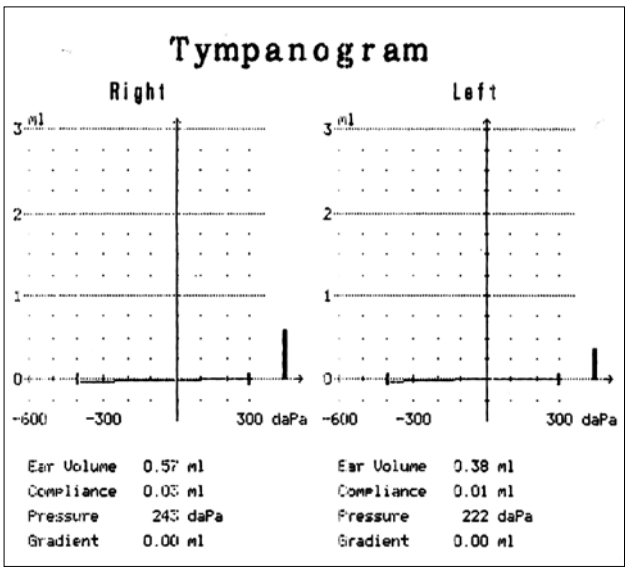

Figure 3 Tympanometry results – bilateral type B tympanogram characteristic for middle ear effusion

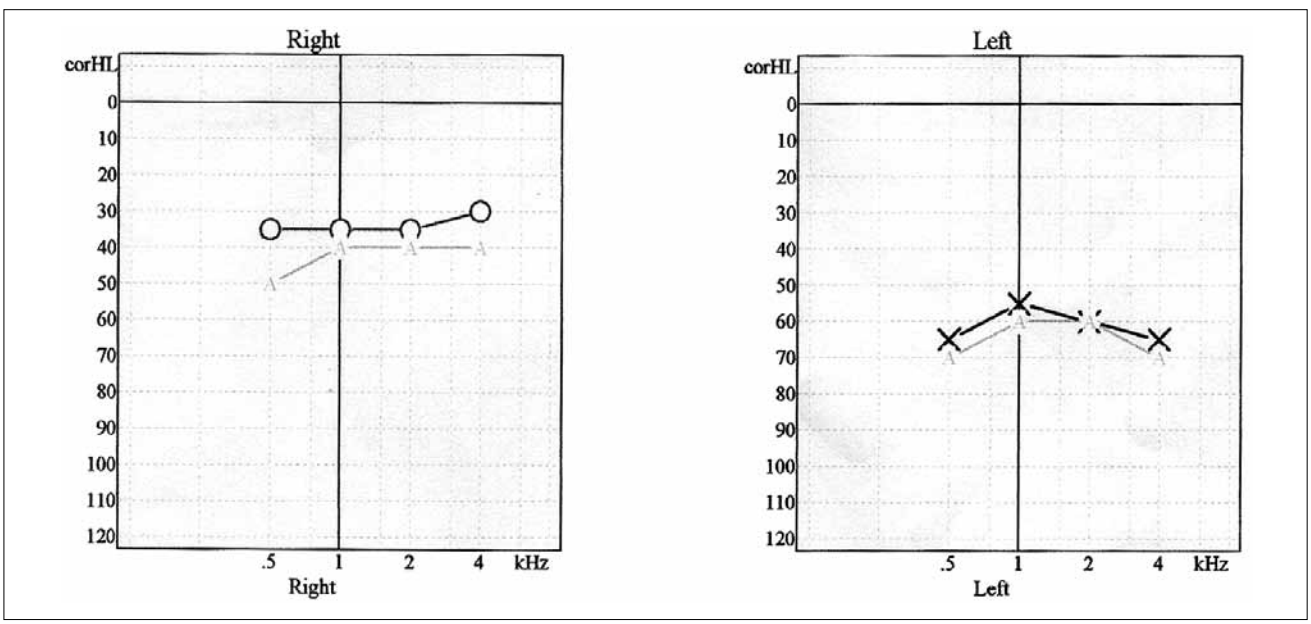

Figure 4 ASSR evaluation - mild hearing loss of the right ear and medium hearing loss of the left ear

DISCUSSIONS

Hearing loss in children is a serious concern because it interferes with speech and language development. Hearing deficit among school age children contributes to poor academic performance, as a child's brain develops in response to the amount of sensory stimulation it receives[24]. In addition to and related to communication disabilities, the consequences of hearing loss in children include psychosocial problems such as frustration, irritability, anxiety and even depression.

There are many causes which can lead to hearing problems among paediatric population: external ear and middle ear diseases, auditory nerve pathology, different syndromes and malformations, trauma etc. Negative emotions like stressed situations at which children are exposed or on-going abuses and even neglecting parents can cause hearing loss.

Erwin D. Riedner and Paul L. Efros[25] reported cases of children with hearing deficit after emotional abuse, similar with our case. Emotional trauma inhibits the child's ability of understanding and his/her creativity. These cases suggest that children are able to develop emotional strategies to protect themselves, one of these being speech refusal. In these cases, multidisciplinary approach (audiologist, logopedist and psychologist) is a must.

It is very important for children diagnosed with hearing loss at screening to be referred for clinical testing and later integrated in a specialized education system[26-28]. Sign language can be a useful tool when other methods are impossible to be applied.

CONCLUSIONS

Children with hearing loss are born with the same basic capacities for learning and language as normal children. They can reach language potential as a normal child if they receive appropriate educational programs and support as soon as possible.

Children hearing screening is important for determining the normal or abnormal auditory function, in order to identify infants who have significant hearing loss.

A team of ENT specialist, audiologist, logopedist and psychotherapist is needed in all those children with hearing loss. Early diagnosis of hearing loss, coupled with appropriate intervention and management, is necessary for children to reach their full communicative and educational potential.

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