THE IMPORTANCE OF THE EARLY DIAGNOSIS IN THE CONGENITAL HEARING LOSS – CASE PRESENTATION

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Abstract

The congenital hypo auditory hearing means hearing loss present at birth, due to genetics or non-genetics.

According to the data provided by the National Institute of Deafness and other Communication Disorders (NIDCD), hearing loss is met at 1-3/1000 live healthy infants, but the prevalence grows to 10-50 times to infants with risk. [1,2,3] The purpose of the neonatal auditory screening is to select from the general population, the infants with risk of having hearing loss, given that out of the 10 infants with congenital hypo auditory hearing 9 come from parents who do not have hearing problems. [4]

The article presents three clinical cases of congenital hypo auditory hearing diagnosed in families with or without a history of hearingloss, with or without risk factors for hearing loss associates. Thus, it aims to attract attention to the importance of the neonatal hearing loss in the general population and not only in the presence of the risk factors.

Key words: congenital hearing loss, risk factors, universal neonatal auditory screening

Introduction

Deafness, if it is bilateral especially, trains a sensory deprived the more prejudiced as it occurs in children. If it remains undetected, even an average decrease of unilateral hearing may have negative consequences. [5] Therefore, to minimize the consequences of a deficiency, the hearing loss should be detected early, so that by the age of 3 months to be established the diagnosis and up to 6 months to initiate an appropriate treatment. [6]

The sensory deprivation consequences are even more dramatic as the hearing loss is larger and as occurs earlier.

The early detection and the management of the hearing loss in infants are essential for the normal development of speech, language, literacy and cognitive capacity.

They demonstrated that the early treatment improves the speech, language and intellectual performance of the children, regardless of the initial level of the hearing loss.

Presentation of case

Case I. S.P. new born sweep of 2700 g. female, in the urban environment, from gemelar pregnancy of 38 weeks, with physiological evolution, without suffering at birth; APGAR 8/9.

The result of the auditory screening carried out by OEA to hospital discharge was REFER, and the retest performed at a month from birth had a negative result: REFER.

The auditory testing of the twin sister -6/7-APGAR has not revealed pathological changes: PASS.

The heredo-collateral antecedents have not revealed historically of hearing loss in family, it probably being the reason of the family negation who did not want at the time in question the continuation of the investigations.

The somatic development was normal according to the stages of age, with the first steps made around the age of 11 months and the articulation of the first words at about 18 months.

Around the age of 2 years, the parents have noted that the girl does not react to sounds and have requested a consult ENT, which not revealed pathological changes. On 25.05.2011 (at the age of 3 years and 1 month) for the diagnosis of chronic adenoiditis; bilateral serous otitis they carried out the adenoidectomy and bilateral transtympanal drainage.

The plant evolution in the terms of purchase of the language and the reaction to sounds caused an auditory evaluation by an ABR (Auditory Brainstem Response) and ASSR (Auditory Steady State Responses) on 12.08.2911-at the age of 3 years and 4 months.

After the clinical exam, family investigation, interdisciplinary consulting (Pediatrics) and the performed investigations (less the genetic ones) has established the diagnosis of the bilateral average sensorineural hearing loss right ear and severe left ear.

They recommended bilateral auditory prosthesis, logopedical recovery and ENT periodical revaluation.

Case II. V.I. The newborn premature female from rural areas, from a 28 weeks pregnancy with birth weight of 900 g, APGAR 2/3 with asphyxia during birth and bouts of apnea during neo-natal period. She received hyperbaric for 10 days, remaining hospitalized in the intensive care unit for a month and the total hospital stay duration being of three months.

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The auditory testing performed initially by OEA and then by ABR (08.09.2008, three months after the birth) resulted: REFER-REFER.

The heredo-collateral antecedents have not revealed historically of hearing loss in family.

They recommended the continuation of the auditory objective investigations but have not been presented at this stage of diagnosis and could not be seen. The investigation made thereafter established that presents a normal somatic development, but without can quantifying the hearing loss (if it exists).

Case III. G.E. new born mature 2900 g, male, from urban area, derived from the physiological pregnancy of 39 weeks with physiological evolution, birth-cranial spontaneous presentation, without suffering at birth; APGAR 9/10.

The heredo-collateral antecedents revealed historically of hypo auditory hearing in family: mother-congenital hearing loss, father acquired hearing loss, and grandfather paternal- hearing loss probably acquired.

The auditory screening performed on 25.08.2008 by OEA and then to a month by OEA and ABR raised the suspicion of a hearing loss (REFER-REFER).

On 04.10.2011 (at the age of 3 years and 2 months) is performed the auditory evaluation balance by OEA and the tympanometric measures, ABR and ASSR and they establish the diagnosis of the bilateral severe sensorineural hearing loss.

They recommend bilateral auditory prosthesis, auditory recovery and ENT revaluation (to the cochlear implant).

Discussions

1. The importance of the early identifying of the hearing loss is long time accepted, but only in the recent years it has put in place a national identification program of the hearing loss even from birth (program where also Oradea enters).

They use two types of tests:

The first test was developed in England and carries the name of acoustic oto-emissions (OEA) that evaluates the function of the inner ear; it takes a few minutes and is performed during the natural sleep. If the test result is "REFER" does not mean that the babe shows hearing loss (may be restless or can present still liquid in the external auditory conduct during the childbirth, the result being not conclusive if there is a pathology of the external ear). If OEA are missing, the new infant has a neuro-sensory hearing loss at least 30 dB.

If also the second test by OEA does not get a positive response, the second type of test is to be done: AABR- (automated auditory brainstem response). This test reflects the function of the conductive ways up to the level of the brain stem and is carried out during the natural sleep - the average long 5-30 min.

The most effective screening is based on the pairing of the two objective tests, this being mandatory in the infants who require inpatient in ATI (neonatal intensive care unit) for more than 5 days. 6,7

2. Initially, since 1975, the J.C.I.H. (The Joint Committee of Infant Hearing) recommended that the neonatal screening for hearing loss to be carried out by ABR to all children with the risk factors present. But only 50% of children which present hearing loss present one or more risk factors (as seen in the above cases), so, even if the screening program would work perfectly, about 50% of children with hearing loss would not be detected. In addition, through the program of screening of all the risk factors would be very difficult that the parents to be recalled for diagnostic evaluation. Incidentally, the first reaction and (most anticipated) of the parents is denial.

According to the J.C.I.H. Year 2007 for the children detected with permanent hearing loss must be performed at least once an assessment by ABR until the age of 3 years within the complete auditory evaluation. 6

For the children who pass the neonatal screening but present risk factors must be performed at least once an auditory evaluation between the months 24-30, repeated for the children with the cytomegalovirus infection, syndromes associated with hearing loss progressive, neuro-vegetative disorders, trauma or any other positive cultures for postnatal infections, for the children with history of hearing loss or who had chemotherapy or ECMO (extracorporeal membrane oxygenation). 6

3. The early identification and intervention can prevent the severe psychosocial, educational and linguistic repercussions. Ideally, new birth babe screening must be completed at the age of one month (NCHAM) and diagnosed before the age of three months to be enlisted in the program of early intervention until the age of 6 months. 6,8

4. There are researches that have compared the children with hearing loss who were early detected and when that occurred before the age of 6 months with the children intervened over the age of 6 months. In time, they noticed that the children that were early detected are 1-2 years ahead of those identified later in language, mental development and social attitude. 9

Thus, in the long term, we can make serious savings for the education of the hypo auditory hearing children hearing early tracked down.

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