THE ROLE OF PERINATAL HEARING SCREENING IN THE NORMAL DEVELOPMENT OF THE INFANT'S LANGUAGE

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Abstract: Detection of hearing loss at birth or in the early childhood is extremely difficult and this affliction tends to represent a major public health issue due to its multiple consequences on human intellectual, social, linguistic, cognitive, emotional and cultural development. The alteration of the physiological properties of neurons in the peripheral and central auditory system is termed plasticity. In order to avoid plasticity, sensory input must be present from birth. Many doctors and educationists are not sufficiently familiar with the signs of hearing loss or its educational impact.[4] That is why congenital deafness must be detected as soon as possible, preferably within the first 3 months in order to insure normal language development of the infant. This makes hearing screening a mandatory procedure in every maternity hospital since congenital deafness is the most frequent condition that can be diagnosed immediately after birth and is also the most common type of sensorineural hearing loss in developed countries (1/750 children develop a potentially debilitating sensorineural hearing loss).

Keywords: congenital, deafness, screening, language, otoacoustic emissions

INTRODUCTION

Hearing loss of variable etiology represents one of the most serious public health issues confronting the world's population. According to data reported in 2001 by the World Health Organization (WHO), over 250 million people (4.2% of the world's population) currently suffer from a form of hearing loss.[6,7,13,14] Of these, congenital hearing loss is particularly important due to its early onset (immediately or shortly after birth) which creates for the neonate an impossibility of acquiring language, thus turning him into a deaf-mute. This particular type of affliction is relatively frequent, with a prevalence reported by different sources in literature as varying between 1-3/1000 newborns[19] and 1/500 newborns[26] but it can increase up to ten times in patients with known risk factors.

Although etiologically heterogeneous, at least 50% of all early onset hearing losses have a genetic cause and of these, the large majority are most probably autosomal recessive (75-80%)[26] and non-syndromic (70%).[19,26,9,16] The rest of 50% is due to environmental factors such as pre- or postnatal infection, birth complications or ototoxic medication.

Approximately 1/1000 children are diagnosed with profound or severe congenital deafness from birth[12] and during early childhood, in the prelingual stage, the prevalence of permanent hypoacusis increases ten times, from 1-3/1000 to 3-5%.

Another 1/1000 children develop hearing loss before reaching adulthood but these forms are usually milder and progressive and have little impact on language development.[12]

SCREENING METHODS

Otoacoustic emissions (OAE) are the most common method for early screening (Transient Evoked Otoacoustic Emissions -TE-OAE and Distortion Product Otoacoustic Emissions – DP-OAE). These are basically sounds generated by vibrations of the cells in the inner ear as a response to acoustic stimulation (clicks or tones) with a microphone in the external ear canal (EAC). They travel backwards, from the internal ear towards the external ear canal where they can be measured with a special probe. The lack of OAEs in a new-born is highly suggestive of congenital sensorineural hearing loss and reflects the status of the peripheral auditory system. This test is extremely quick (1-3 minutes), practical and easy to perform (can be performed by nurses), needs no anesthesia, no special conditions and has no side effects. The equipment is also reasonably priced.

Auditory Brainstem Response (ABR) measures, in addition to the integrity of the inner ear, the auditory pathway. The electrophysiological response of the brainstem to auditory stimulation (microphone in the EAC) is measured by electrodes placed on the scalp. This test takes longer (15-20 minutes) and sometimes requires sedation of the child.

HEARING SCREENING AND LANGUAGE DEVELOPMENT

The loss of hearing (hypoacusis or deafness) is the most frequent congenital pathology we can diagnose immediately after birth which gives us an unusual advantage in modern medicine. Its prevalence increaces in the pre-school and school periods by adding aditional factors such as late onset or late diagnosis. The primary purpose of early diagnosis is increasing the chances for the child to accumulate an adequate vocabulary.[1] Performing a correct diagnosis within the first 6 months followed by appropriate treatment can produce an almost normal (sometimes fully normal) language development.[29,30,31,32]

A number of studies stated that the central auditory system does not mature without acoustical stimulation and the duration of depravation influences the time required for rehabilitation and the level of performance after cochlear implantations. Other studies observed that the rate of maturation of cochlear implanted children is similar to that of normal children but delayed by an amount equal to the duration of auditory depravation.[4] Various entities such as the American Audiology Academy (AAA) developed protocols for hearing screening the neonates population and school-aged children with extension to teenagers in order to improve social and occupational integration of deaf persons.[1]

According to the age of the screening we can perform:[1]

- Neo-natal screening – can be performed for all newborns (universal screening) or just for those who present risk factors (risk group screening) by using OAE or ABR.
- Early childhood screening usually based on Conditioned Play / Response Audiometry,
- Pre-school / school period screening – uses Pure Tone Audiometry.

Modern studies based on questionnaires for parents have invalidated the theory which stated that a deaf child can be diagnosed by his every day behavior. Also, research shows that up to 50% of children that have major hypoacusis-generated educational deficiencies by the age of 9, have initially passed a neo-natal screening test[8] and 9-10/1000 children will present permanent auditory impairment in one or both ears before they reach school age.[23,28]

The American Pediatrics Academy (AAP) recommends neo-natal screening as well as early childhood and school age screening although in the U.S.A. the neo-natal screening tests up to 95% of all neonates through programs such as EHDI (Early Hearing Detection and Intervention). Current information suggests a higher diagnosis rate at the school age compared to neonates (in the U.S., 10-20% of all hypoacusis cases during childhood cannot be identified by neo-natal screening due to late onset of the disease. A similar study in the UK has shown that for every 10 children diagnosed with permanent bilateral deafness by neo-natal screening there are 5-9 children who will develop a similar affliction by the age of 9.[8]

In order to facilitate screening, the WHO developed 10 principles for early screening (Table 1)

Table 1– WHO principles of screening[1]

1. The condition sought should be an important health problem.
2. There should be an accepted treatment for patients with recognized disease.
3. Facilities for diagnosis and treatment should be available.
4. There should be a recognizable early or latent symptomatic stage.
5. There should be a suitable test or examination.
6. The test should be acceptable to the population.
7. The natural history of the condition, including development from latent to declared disease, should be adequately understood.
8. There should be an agreed policy on whom to treat patients.
9. The cost of case finding (including diagnosis and treatment of patients diagnosed) should be economically balanced in relations to possible expenditure on medical care as a whole.
10. Case finding should be a continuous process and not a *once and for all* project.

In conclusion we can state that an estimated prevalence of permanent hypoacusis of 3/1000 newborns can increase to 9-10/1000 children of school age,[28] and that a permanent or transient hearing loss affects more that 14% of all school children.

EDUCATIONAL IMPACT OF SCREENING

Historically speaking, undiagnosed hypoacusis has affected the performance of countless generations of school children, limiting their options for higher education and ultimately restricting their professional evolution.[10] Children with severe or profound bilateral hearing loss reach the 4th grade mean performance levels for reading and writing at the age of 17-18, which pleads for early detection through universal screening.[11] Of all those who reach higher education, 70% give up before graduating.[27] The total unemployment rate for persons aged 16-64, with sensory disabilities is 60%.

The class-room represents an environment based on verbalization in which a correct transmission and reception of sound is essential for the learning process.[25] A diminishing of hearing, be it permanent or fluctuant, makes voice reception more difficult, especially in a class-room with all its echoes, noises and remote sound sources.[5] The behavioral effects can be often subtle and resemble those of ADHD, learning problems or mental retardation. Children with hearing loss often show:

- Difficulties in processing verbally or auditory transmitted information.
- Frequent request for repeating the information.
- Fatigue from listening.
- Inadequate answers to simple questions.

- Isolation from colleagues.
- Difficult reading.
- Difficulties with written or spoken language.
- Easily frustrated.

SCREENING TARGET GROUPS

a) Neo-natal screening

Extremely useful since congenital deafness, although relatively frequent, is not readily apparent for doctors or parents immediately after birth.

- Risk group screening – only for subjects that have clear risk factors for developing congenital deafness (prematurity, gestational age, VLBW, ELBW, low Apgar score, congenital infection, ototoxic treatment, intraventricular hemorrhage, respiratory disstres, neonatal hypoxia, mechanical ventilation, NICU admission, neonatal hypotension, hypoxic-ischemic perinatal encephalopathy, prolongued jaundice, cranio-facial anomalies and other congenital anomalies.). It is mandatory that these patients be tested. This type of screening is currently performed in most developed European countries and is responsible for around 50% of all diagnosis.[21]
- Universal screening – represents hearing testing of all neonates and can diagnose up to 80% of all congenital deafness cases. Testing takes place during the first 2 – 3 days and can be repeated some weeks or months after. This kind of screening is performed in the U.S.A.

b) Early childhood

The smallest alteration of hearing during the newborn period can lead to significant delay in language development and this represents a clue towards early diagnosis of a minimal hypoacusis.[18] Children with minimal or moderate hypoacusis can pass the neo-natal screening and many of those who do not pass have no follow-up screening during the next years. We must not forget that a minimal deafness can represent the onset of a progressive deafness that will worsen in time.

Children with no neo-natal screening, those with neo-natal screening and suspicion of congenital deafness but with no follow-up screening or those with a late onset of the disease will be diagnosed too late in order to prevent serious language development problems [17]

An early childhood screening (0-3 years) will diagnose with permanent hearing loss approximately 2/1000 children and another 6-7/1000 will be diagnosed later on in life.[3]

The Joint Committee of Infant Hearing (JCIH) is currently trying to implement a global methodology for screening at the ages of 9, 18 and 24-30 months and/or whenever a suspicion arises for doctors or parents.

c) Pre-school period screening

The purpose of this screening is to diagnose children who evaded a neo-natal screening or had no follow-up. A second purpose is the diagnosis of late onset disease. Until school age, 6-7/1000 children will develop permanent hypoacusis.[3]

d) School age screening

This type of screening is for children that have evaded the screening system all together up to this age, had no follow-up after a previous diagnosis or have late onset disease. These cases are easier to identify since they enter an educational system and show evident difficulties in learning and adapting. This screening targets typically the following groups.[1]

- All students in specific grades (for instance all students in the 4th grade, nation-wide).
- Referral students – referred by teachers or parents for obvious learning difficulties.
- All new students enrolling for the first time in the school system.

e) Targeted grade levels

Screening the children population at early ages is essential in early diagnosis of hypoacusis and normal development of language but even developed countries such as the U.S.A. or the UK have no specific research for specifying the ages or grades where screening will be most efficient. Some pediatrics societies recommend screening at the school ages of 4,5,6,8 and 10 years.[22] Further studies have identified a considerably larger number of cases in the 2nd grade compared to the 1st grade which would recommend other screenings after school enrollment and even far beyond elementary school.[23]

- Pre-school age screening (kindergarten or 1st grade enrollment age) will identify less than ¼ - ½ of all deaf students.[1]
- Screening at the ages recommended by AAP (4,5,6,8 and 10 years), more specifically at kindergarten level, pre-school level, 1st, 3rd and 5th grade will identify more than ½ but less than ¾ of all previously missed cases.
- Approximately 90% of all new cases will be diagnosed by screening beyond the 3rd grade level.
- Screening in the 5th, 6th, and 7th grade has very similar results to screening in the 7th and 9th grade.
- Even though screening in the 7th grade has very similar results to screening in the 9th grade, the first one is recommended in order to gain 2 extra years of treatment and intellectual development.
- In order to diagnose approximately 70% of all previously missed cases, screening must be performed at least at the levels of 1st, 3rd, 5th, 7th and 9th grade.
- The trend for diagnosis of new cases decreases towards the 1st, 2nd and 3rd grade and increases in the 5th grade, suggesting a possible higher prevalence of hearing loss in the upper-elementary school.
- In addition to the minimum grades mentioned above, more new cases will be diagnosed in the 2nd grade rather than in other secondary grade.

CONCLUSIONS

The neuro-linguistic development of a newborn requires auditory stimulation within the first two years of life and deficits during this period are almost impossible to recover. For this reason, early diagnosis of a congenital hearing loss is of outmost importance. If such a diagnosis is possible, the children will be able to develop normal language and will no longer require special education.

The hearing screening for newborns is based on automated tests (OAE and ABR) that have good sensitivity and specificity (80-90%) in order to raise the suspicion of a deaf child. The main advantage of OAE is that it does not require specialy trained personnel and can be performed very soon after birth. It also has no side-effects, takes very little time (a test requires 25-330 seconds), is cheap and requires no sedation. ABR on the other hand is more expensive, takes longer (15-20 minutes), requires sedation but at the present moment represents a golden standard for congenital deafness diagnosis.

Universal screeing is a goal that, unfortunately, has not been reached even by developed countries of the E.U. but which, once instated, could allow early diagnosis of congenital hearing loss. We must also consider the usefullness of subsequent screening since some children can show a late onset of the disease. The screening in early childhood, at the pre-school and school age and even for teenagers can insure an optimal neurological, lingustic and educational development for all children.

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