Original Article

Assessment of Hearing Screening Tests in Pediatric

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Abstract

Background: Congenital hearing loss is one of the prevalent chronic con¬ditions in children. As a result, screening for hearing function is of utmost importance. **Objectives:** This study is aimed at discussing hearing screening in children in general, and newborns in particular to address the questions including why is it done, what tests are used, and the benefits of such processes. **Methodology:** PubMed database was used for articles selection using some keywords. **Conclusion:** These screening tests are getting known even among non-audiologists, and knowledge about such tests will make the clinicians order better tests and offer better care. And these tests helps in identifying the next step in management, especially in high-risk patients. New studies and breakthroughs in the technology will make health care more available, and will encourage screening culture in the society.

Keywords: Hearing screening, congenital hearing deficit, children

Introduction

Congenital hearing loss is one of the most common chronic conditions in children. In developed countries, the prevalence for permanent bilateral hearing loss has been estimated to be 1.33 per 1.000 live births, it is further higher in non-developed countries, with 19 per 1.000 newborns in Africa (sub-Saharan) and up to 24 per 1.000 live births in South Asia [1, 2] .The difference in the prevalence percentage is attributed to the risk factors being more pronounced in non-developed countries, such as decreased gestational age, low birth weight, congenital infections, and genetic factors [3, 4]. Diagnosis of such cases is usually delayed and thus it has major effects and complications on developmental stages and cognition of the child. However, in recent years, the introduction of screening programs for the newborn has been a step forward for the early detection and thus early management of these patients. In this paper, we will review screening programs, the tests used for such screening, benefits of the program and their future as well.

Methodology

PubMed database was used for articles selection using keywords such as Hearing, and Screening. With regard to the inclusion criteria, the articles were selected based on inclusion of one of the mentioned keywords and the exclusion criteria were all other articles which did not have one of these topics as their primary endpoint.

Discussion

Hearing loss present at birth (congenital hearing loss) is the term used to indicate the inability to detect sounds by a person, and can be unilateral or bilateral. Another technical definition is a hearing loss equal to or more than 40 decibels (dB) incomparison to the better hearing ear averaged over frequency ranges for speech recognition ranging from 500 up to 4,000 Hertz (Hz) [5]. The primary mechanism is due to inability to transform the air waves into an electrical impulses or inability to interpret the impulses and/or both.

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How to cite this article: Alhaqbani, M. H. M., Haddadi, M. A. M. Aljahdali, E. S. O., Alandijani, W. A., Alkhudhair, S. I., Alshammari, S. S. et al. Assessment of Hearing Screening Tests in Pediatric. Arch Pharma Pract 2019; 10(3):9-12.

- ### *Hearing loss types and Risk Factors:*

Hearing loss is divided based on the location of the pathology into a conductive and a sensorineural or mixed. Conductive hearing loss is when the outer or middle ear suffers the pathology. In sensorineural hearing loss the parts affected can be the inner ear, auditory nerve and/or central auditory pathway. The last type which is the mixed hearing loss is when the patient has both types of the hearing loss at the same time. The main mechanism behind the conductive hearing loss is the absence of propagation of sound waves through the ear, this can be due to any issues of development in the middle ear, external ear or both, and the second way is via any obstruction of the middle ear caused by effusion even if transient which is seen in otitis media. The second type (sensorineural) can be further dissected into Sensory and Central hearing loss. Sensory hearing loss arise from the hair cells of the inner ear, while Central hearing loss arise from a disorder in the central auditory pathway. Another subtype in this type is Auditory Neuropathy Spectrum Disorder which is a term for pathologies with otoacoustic emissions, along with cochlear microphonic with abnormal auditory brainstem responses. The main pathology in this spectrum is a lesion which can be located in inner hair cells, auditory nerve, and/or auditory pathway. These disorders have a major impact on the patient causing usually an impaired speech discrimination [6, 7].

A lot of risk factors are associated with hearing loss, such as genetic mutations– the most common factor– (e.g. syndromes such as Jervell and Lange Nielsen, Usher, and Alport), congenital infections (e.g. congenital cytomegalovirus – CMV, Zika virus, Toxoplasmosis, and rubella), decrease in weight and gestational age at birth. Other risk factors reported include cranio-facial abnormality, admission to neonatal intensive care unit, some medical interventions such as venous access, and assisted ventilation in NICU, and hospitalization duration of more than 12 days in the NICU [8, 9].

- ### *Screening:*

Congenital hearing loss diagnosis is usually done in late stages mostly due to its complications, even though it is usually present since birth. Thus, screening program is considered as the optimum approach for early diagnoses in newborn, limiting its complications and easier management. Even though initially it was exclusive for high-risk babies with one of the aforementioned risk factors, with further studies on this subject and accumulation of evidence of the major advantages and benefits of such program on the overall population, the approach was modified. This has a more evident role in developed countries, where such program became almost available in all major hospitals and for almost all newborns. The system usually applies a two-phase screening program using sequential electrophysiological measurements. In newborns, the screening usually measures the otoacoustic emission or the automated brain stem response which is repeated twice. However, even if a baby does not show a response in these tests, they are not labelled with hearing loss until a second test done –preferably before the 3 months age- is repeated. Moreover, passing this test does not rule out progressive, or late onset and/or less severe congenital hearing (less than 40 dB hearing loss). These disorders are almost always not picked up by the initial test, but a follow up testing can easily detect such diseases. The role of the clinician in such cases is to have a high suspicion levels, especially in patients with risk factors and request a follow up testing which may detect the late pathologies in such babies. Babies who do not pass the hearing screening are referred to determine the exact impact of the disease via audiometric assessments (including oto-acoustic, auditory brain stem and audiometry testing), and confirm if both ears are affected or only one ear. Hearing loss ranges from mild between 20 and 40 dB, moderate 41 to 70 dB, severe between 71 and 95 dB, and profound which is more than 96 dB [9].

- ### *Otoacoustic Emission Test:*

Otoacoustic emission (OAE) test is a screening test used to assess hearing loss. It is a low-level sound emitted by the cochlea either spontaneously or evoked by an auditory stimulus. The main goal of this test is to determine the quality of the cochlear status and especially the outer hair cell function. This test includes 4 types; transient otoacoustic emissions (TOAE) or transient evoked otoacoustic emissions (TEOAE) where sounds are emitted in response to an acoustic stimuli of very short duration (clicks or tone-bursts) and is the most commonly used for screening and distortion product otoacoustic emissions (DPOAEs) where sounds are emitted in response to two simultaneous tones of different frequencies which are less commonly used. Other types include spontaneous otoacoustic emissions (SOAE) where sounds are emitted spontaneously without an acoustic stimulus, and sustained-frequency otoacoustic emissions (SFOAEs) where sound is emitted in response to a continuous tone. In TOAE, a probe is inserted deeply into the ear canal of the baby forming a seal like around it, then a stimuli around 84 dB SPL peak equivalent level is introduced, normally it will produce a transient evoked otoacoustic emission if the hearing threshold is 20 dB HL or better. This response is mainly related to the movement of the tympanic membrane backwards and/or forward by the fluid pressure fluctuations originating from inside the cochlea, and the outer hair cells movements is at the center of this all. The mechanism of closing the ear canal with probe increases the sensitivity of picking up this response up to recording below 3 kHz, by preventing the air leakage along these fine movements [10]. The advantages of such tests include high sensitivity to cochlear pathology with a frequency-specific aspect, cheaper compared to auditory brain stem response test and relatively easier to do. However, drawbacks of this test include possible inability to detect pathology if middle ear status is affected (by effusions or debris after birth for example), thus cannot be done in some babies in the first hours of life, and this test usually cannot be used alone to make a diagnoses but rather interpreted along the context of other tests [11].

- ### *Auditory Brain Stem Responses Test:*

Auditory brain stem responses (ABR) is a major screening test used to measure the electrical activity which is produced by reflect neural activity at several discrete points along the auditory pathway once there is an auditory stimuli. Thus, ABR focuses more on the sensorineural component of the hearing loss by detecting neural response to the voices and vibrations. ABR test is done by attaching the patient to electrodes, which are put on his scalp, these electrodes detect electrical current and recording of them is done via computer. The new model which is automated increases the objectivity while reducing the expertise of the tester required and its effect on results, other benefit of ABR test is its ability to measure the auditory sensitivity of the patient. The auditory stimuli are commonly used in the form of click- or tone burst and these are considered as the gold standard for the objective assessment of hearing pediatrics population overall. The obtained thresholds at high frequencies, between 2,000–4,000 Hz, is usually within 10 dB threshold of other tests (behavioral auditory test). However, ABR test is not perfect and has its disadvantages, mainly being costly, time consuming, and in need of an expert to perform the test and interpret the results especially with classic ABR methods. Overall, ABR is effective and sensitive to pathology in both cochlear and retro-cochlear areas; while, OAE is mainly for cochlear area. It can be said that ABR is superior and better for testing than OAE for infant screening. The most common type of sensory hearing impairment in the low-risk patients is of the sensory transmissive type, and this type is detectable via OAEs. This with the economical prospect and availability, makes OAEs the most widely used method in the new-born hearing programs. However, ABR is used for high-risk patients where there is a known risk factor, or high suspicion of neurological damage.

- *Auditory Response Cradle Test:*

Another main test done for screening is auditory response cradle (ARC) which is an automated, micro-processor controlled, objective recording of behavioral changes in patients. This test was to answer one of the major limitations of sensitivity in neonatal tests, which is the inconsistencies in observing and recognition of infants' behavior. ARC gives an 85 dB, high pass noise (2.6– 4.4 kHz) in both ears via closed coupler earphones, and changes in body activity, head turn and head startle. Then a statistical analysis of these variables is done, and subsequently a pass or refer decision is automatically taken. The duration of this test varies between two to ten minutes. However, despite automation, sensitivity is still an issue with for example preterm babies showing 50% sensitivity for identifying severe or profound deafness, and only 20% for moderate ones. This is more evident in NICU babies with up to 50% who passed an ABR screen failing [12].

- *Audiometry:*

Different tests are available for hearing screening in old children, due to more output from the children due to their growth. Visual re-enforcement audiometry is used to test hearing in children between 6 and 24 months of age. In children with adequate neurological and hearing development, a new sound stimuli will provoke a reflex towards the source of the auditory stimuli. However, in some cases, only a skilled audiologists can obtain a reliable data, record the reflex, and make a sound judgment. Another variable of the same test is play audiometry, this one is used in older children mainly between 2 and 4 years of age. The test is conducted by trying to condition them to respond sounds through play activities. After 4 years of age, the standard audiometry which is used in adults is usually done. The test may be carried out with either an air-conduction transducer (e.g. earphone), a bone-conduction transducer, or both of them at the same time. The earphone in air conduction study gives off an already determined frequency auditory stimuli and the patient indicates the hearing of such stimuli with a reply which can be as simple as pressing a button. The same happens in the bone conduction study where instead of an earphone we place an electrode like object on the bone behind the ear usually then vibrate the skull, this stimuli reaches and affect the cochlea immediately, bypassing the external and middle ear. The determination of thresholds (between 250 and 8000 Hertz) for air and bone conductions is critical to establish a clinical proven differentiation between the conductive and sensorineural hearing loss. Thus, the standard audiometry can provide a more detailed analysis of the pathway by getting the status of bone conduction, air conduction and even both of them with frequency analysis of them. Moreover, the visual and play audiometry can be used to give a generalized idea about the integrity of the auditory system as a whole.

- *Pure Tone Test:*

The pure tone test can be done to determine some characteristics in patients with hearing deficiency such as differentiation of the type and determination of the loss extent [13]. Pure tone audiometry can be used as a screening method as well in the pediatrics age group measuring wide speech range frequencies and upper limit of normal hearing ability [14]. The main setback is its limited output and results to the clinician, while it shows a hearing sensitivity, it does not elaborate on auditory processes of signals that the child may face daily [13]. Additionally, the need for a noise cancelled environment and conditions such as tinnitus and/or anatomical anomalies alter the results [15].

The main idea behind screening of older children is discovering any late or progressive congenital hearing loss type. The delay of diagnosis to an average of 2 years and 6 months of age is associated with a lot of consequences in such cases. In most cases, the late diagnosis occurs as a result of parents who do not seek medical help till complications develop. These complications mainly involve delayed speech and language development, rendering them unable to keep up with their pairs, and later on may have a major impact on their learning and their social life as well. However, with early diagnoses, opportunity for early interventions becomes available with hearing preservation or even restoration strategies. Another major point that these tests may be done in ruling out diseases or diagnosing others, especially in disorders such as autism, or neurological delayed

development, or even establishing complications in diseases such as otitis media. The clinicians usually select these tests with differential diagnoses in mind, and rarely as a part of general screening and that is why it is important to understand the importance of such screening programs [16].

Conclusion

The importance of implementing screening program for hearing loss is unparalleled due to its success story in developed countries, and the major benefits that were reported in terms of limiting the complications and all its toll on community. These tests are getting known even among non-audiologists, and knowledge about such tests will make the clinicians choose the best tests and offer better care. Understanding the tests, what do they measure and which one is preferred for which age group are very important factors for clinicians to identify the next step in management, and especially in high-risk patient. Furthermore, implementing these tests will drive the technology used further and with recent breakthroughs, it is getting cheaper and thus more available. These steps will ultimately encourage the screening culture in the society and thus will benefit the population as a whole.

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