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International consensus

International consensus (ICON) on audiological assessment of hearing loss in children

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ABSTRACT

The prevalence of hearing loss in newborns and infants is estimated between 1 to 3.47 cases per 1000 live births. Early diagnosis and rehabilitation of congenital hearing loss are mandatory in order to achieve a satisfactory linguistic and cognitive development. Without appropriate opportunities to learn language, these children will fall behind their normal hearing peers in communication, cognition, reading and socio-emotional development. After promising results, neonatal screening for hearing loss and audiological evaluation are becoming more extensively carried out. In planning universal neonatal hearing screening programs, transient evoked otoacoustic emissions and auditory brainstem responses are the gold standard for the screening and diagnosis program. However, there is no consensus regarding the use of audiometry and other electrophysiological tests (such as auditory steady-state responses) in current practices. Several screening and audiological assessment procedures have been described and advocated all around the world. But, a systematic scheme of performing diagnosis in the pediatric audiology population is lacking. A consensus conference was held at the International Federation of Oto-rhino-laryngological Societies Congress, in June 2017, to discuss the different current practices and to identify the best neonatal hearing screening and audiological assessment management. This article is intended to provide professionals with recommendations about the "best practice" based on consensus opinion of the session's speakers, and a review of the literature on the efficacy of various assessment options for children with hearing loss.

1. Introduction

The prevalence of hearing loss (HL) in newborns and infants is estimated between 1 to 3.47 cases per 1000 live births. The goal of early HL detection and intervention is to maximize linguistic competence and literacy development for children with hearing impairment [1]. Without appropriate opportunities to learn language, these children will fall behind their normal hearing peers in communication, cognition, reading and socio-emotional development.

The identification of HL through the neonatal hearing screening (NHS) is essential for early intervention. If HL is diagnosed before three months of age, and intervention is initiated before the age of six months, significant changes in cognitive and linguistic development of deaf individuals can be obtained [1]. Several screening and audiological assessment procedures have been described and advocated all around the world. But, a systematic scheme of performing diagnosis in the pediatric audiology population is lacking. The use of automated auditory brainstem response (aABR) and otoacoustic emissions (OAE) is implemented in many countries to allow early identification and timely intervention of babies with HL. In European countries, HL screening consists of either a two-stage OAE testing, or the use OAE as a first step, followed by aABR. There is still no consensus on recommended screening techniques [2–4]. Even in France, the national recommendation of 2014 did not arbitrate and allow either OAEs or aABRs except in NICU [5].

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In case of abnormal hearing screening, an audiological evaluation is required to confirm and characterize the HL, based on several tests. This evaluation includes in particular objective electrophysiological measures (auditory brainstem response [ABR], auditory steady-state response [ASSR]) and/or behavioral methods to estimate hearing thresholds. Recommendations are still differ between countries, and early behavioral audiometry in children younger than 6 months is still controversial [6,7].

2. Setting of the consensus conference

An International Consensus Conference (ICON) was held at the International Federation of Oto-rhino-laryngological Societies Congress, in June 2017, in Paris, France.

The members of the panel were Pr Abdelaziz Raji (Marocco), Pr Hao Wu (China), and the Dr Bernard Wanna (Lebanon). The discussion was led by the moderators, Pr Christophe Vincent and Dr Anne Farinetti (France). Based on a review of the literature, a questionnaire about "audiological assessment of hearing loss in children" was sent to the panelists and their answers were presented and discussed at the conference (Appendix 1).

The objective was to provide professionals with recommendations about the best practice, based on experts' opinion and scientific evidence regarding the efficacy of various assessment options for young children with HL.

The review of the literature used an evidence-based approach to provide balanced and objective classification for making informed decisions about assessment options.

Each article has been assessed using the GRADE scoring system. This rating indicates the amount, general quality and clinical applicability of scientific evidence used for each recommendation, ranged from A to D.

3. Neonatal Hearing Screening

The identification of children with HL through the NHS is essential for early intervention (GRADE D) [1]. To maximize hearing outcomes, it is recommended (1) To screen more than 95% of all newborns by 1 month of age, (2) To perform a comprehensive audiological and medical evaluation at no later than 3 months in case of failed-screening, (3) And to receive appropriate intervention at no later than 6 months of age from health care and educational professionals with expertise in HL and deafness management in infants and young children, in case of confirmed HL (GRADE D) [1].

Currently, two physiological procedures are recommended as NHS for early detection of HL: evoked OAE and aABR.

3.1. What procedure should we use?

3.1.1. OAEs

Evoked OAE are low-level sounds primarily generated by the outer hair cells in the cochlea and are recorded in the external auditory canal after stimulation. This procedure is fast and inexpensive, has a high rate of false-positive results (middle ear effusion) and false-negative results (auditory neural spectrum disorders).

These emissions are usually classified according to the generating stimulus:

- transient-evoked (TEOAEs) are evoked using a click (broad frequency range) (CEOAEs) or tone burst (brief duration pure tone) (TBOAEs) stimulus at one level of 80 dB SPL;
- stimulus frequency OAEs (SFOAEs) are measured during the application of a pure tone stimulus, and detected by the vectorial difference between the stimulus waveform and the recorded waveform;
- distortion product OAEs (DPOAEs) are evoked using a pair of primary tones $f1$ and $f2$ with particular intensity (usually one level of L1 and L2 65/55 dB SPL at least at four frequencies).

TEOAEs are present in preterm and full-term infants (range from 82 to 100%), and are thus theorically feasible from 30th week after conception (GRADE B) [8,9], (GRADE C) [10]. However, it is recommended to use the aABR to not miss ANSD.

The most useful clinically OEA are the TEOAEs and the DPOAEs (GRADE D) [1]. There are numerous differences between both methods, which are important to help us decide to perform either TEOAE or DPOAE.

3.1.1.1. Hearing level thresholds detection (GRADE B) [11]. When TEOAEs are present, hearing thresholds equal to or better than 20 dB HL would be predicted in case of hearing loss related to endocochlear dysfunction with outer hair cells dysfunction. TEOAEs are inevitably absent in cases with sensorineural hearing loss exceeding 40 dB HL or in cases of middle ear pathology. A mild hearing loss with thresholds ranging from 25 to 35 dB HL is considered in the zone of uncertainty, where the interpretation of TEOAEs is not clear.

3.1.1.2. Zone of uncertainty (GRADE C) [11]. The zone of uncertainty is wider in DPOAE recordings than in TEOAEs, ranging from 25 to 50–60 dB HL. This zone of uncertainty could explain the risk of higher rate of false-negative in case of use of DPOAEs instead of TEOAEs.

3.1.1.3. Frequency range. TEOAEs are most effective in sampling cochlear function in the mid-frequency region (1000 to 2000 Hz), and CEOAEs are almost as frequency-specific as TBOAEs (GRADE C) [12,13]. On the other hand, DPOAEs can be measured over a broad range of frequencies, they perform better than TEOAEs at 4000 Hz or more, but are not accurate predictors of hearing status at lower frequencies. DPOAEs are superior to TEOAEs at frequencies above 2 kHz (GRADE A) [14–16]. In conclusion, neither OAEs nor DPOAEs can show clear superiority.

3.1.1.4. Influence of SNR. CEOAEs (Chirp) has a relatively high false-positive rate, often due to infant physiological and background noise adversely affecting the emission recording, leading to a "refer" screening result, especially for the low frequencies (below 1000 Hz). In attempt to reduce these false-positive screening, TBOAEs (Tone Burst) may elicit a greater signal to noise ratio than CEOAEs.

In this way, the introduction of combined CEOAE and TBOAE protocols may assist in the reduction of "refer" results, and hence the false-positive rates of UNHS programs.

- In conclusion, there is no recommendations showing the superiority of TEOAEs over DPOAEs in hearing screening protocols (GRADE B) [8], (GRADE C) [10].

3.1.2. Automated ABRs

It is important to use different screening protocols according to the term of the child in order not to miss neural disorders.

OAEs reflect the status of peripheral auditory system extending to the cochlear outer hair cells (OHC). In contrast, automated ABRs (aABRs) reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway. According to the JCIH recommendations in 2007, both techniques can be used to

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