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Audiologic profile of infants at risk: Experience of a Western Sicily tertiary care centre

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ABSTRACT

Objective: To identify the incidence of sensorineural hearing loss (SNHL) on infant at risk and to classify the degree and type of hearing loss describing the main causes associated in Western Sicily. To compare single TEOAE and combined TEOAE/ABR techniques studying the referral rate, the false-positive and false-negative rates through concordance test (κ coefficient), sensitivity (TPR) and specificity (TNR) for each protocol.

Methods: From January 2010 to June 2011, 412 infants at risk, ranging from 4 to 20 weeks of life, transferred to Audiology Department of Palermo from the births centers of Western Sicily, underwent to audiological assessment with TEOAE, tympanometry and ABR. The following risk factors were studied: family history of SNHL, consanguinity, low birth weight, prematurity, cranio-facial abnormality and syndromes associated to SNHL, respiratory distress, intensive care in excess of 5 days (NICU), pregnant maternal diseases, perinatal sepsis or meningitis, hyperbilirubinemia, ototoxic drugs administration.

Results: Forty-seven infants (11.41%) were diagnosed with SNHL; median corrected age at final audiological diagnosis was 12 weeks. SNHL resulted moderate in 44.68%, severe in 10.64% and profound in 21 cases with a significant difference in family history and NICU infants ($p < 0.0001$). As the number of coexisting risk factors increases, the percentage value of SNHL in infants ($\chi^2 = 12.31$, $p = 0.01$, $r^2 = 0.98$) and the degree of hearing loss ($\chi^2 = 13.40$, $p = 0.0095$, $r = 0.92$) also increase. The study of single TEOAE and combined TEOAE/ABR showed a statistical difference ($\chi^2 = 14.89$, $p < 0.001$) with a low concordance value ($\kappa = 0.87$) confirming the importance of combined techniques for NICU group ($\kappa = 0.86$) where four cases (0.97%) of auditory neuropathy were diagnosed.

Conclusion: This study demonstrates the necessity to implement a neonatal hearing screening program in Western Sicily because of the high percentage of SNHL in infants at risk. Family history of HL is an independent significant risk factor for SNHL easily diagnosed through single TEOAE technique. Combined TEOAE/ABR is the gold standard for NICU babies which are at risk for auditory neuropathy. Coexisting risk factors are an additional risk factor for HL.

1. Introduction

According to world statistics research, hearing loss (HL) is one of the most common congenital anomalies, occurring in ~1–3 cases per 1000 newborn infants in the well-infant population while, in certain "higher risk" populations this value could increase 10- to 50-fold, varying from country to country. Normal hearing (NH) is one of the central nervous system's most essential tools for creating the physiological processes of integration, abstraction and creation of "internal speech", which is a necessary foundation of the thought process; such an early diagnosis and intervention, before six months of age, is effective in allowing children with congenital hearing loss to acquire appropriate cognitive and spoken language skills [1–4]. The potential benefits of early detection can only be realized if an effective newborn hearing-screening program is performed, especially on infant at risk. The 2007 Position Statement from the Joint Committee on Infant Hearing (JCIH) confirmed that neonates who are in Intensive Care Units (ICUs) for >5 days, or who have risk factors and a shorter stay, undergo hearing screening with transient evoked otoacoustic emissions (TEOAEs) and (automated) auditory brainstem response (ABR) testing, that is recommended for screening because of the

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increased occurrence of neural (auditory neuropathy/dyssynchrony) loss in this population, compared with well infants [1]. If the neonates do not pass the tests, then a full diagnostic audiological assessment is to be performed by an experienced audiologist [1].

Risk indicators associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood given in the 2007 Position Statement [1] expand those of older risk registries [5]. Although a family history of hearing loss (syndromic or nonsyndromic), cranio-facial abnormalities, prenatal infections, hyperbilirubinemia requiring exchange transfusion, and culture-positive sepsis remain risk factors, other independent neonatal risk factors have been reported [1]. These risk factors include a need for ventilation, use of oxygen supplementation, respiratory failure, low Apgar scores, acidosis, use of ototoxic drugs including furosemide (especially with high serum creatinine levels), treatment for hypotension, patent ductus arteriosus ligation, hyponatremia, and noise [6–13]. Interaction among risk factors has been shown [11].

Even if an newborn hearing screening (NHS) is implemented in most parts of the developed world as an essential component of neonatal care, in Italy there is no stipulated modality for achieving the goal of early hearing detection in individual regions. In fact, the initiatives are still left to individual hospitals that have activated locale programs based on the collaboration of the single birth centers and the Audiology Sections.

The Department of Audiology of Palermo University represents the main speech and hearing third level centre in Western Sicily; waiting for a NHS sponsored by Regional Health Department, it daily performs an audiological assessment to those infants at risk born in Western Sicily and transferred from NICUs and/or regular nurseries to our specialized area.

The aim of this study was to identify the incidence of permanent childhood hearing impairment (PCHI) on population at risk; to classify the degree and type of hearing loss describing the main causes associated with SNHL in infants at risk in Western Sicily focusing on the importance of a suitable and adequate hearing assessment especially for those populations at risk for permanent hearing impairment. The final purpose of this study was to investigate the referral rate and the false-positive and false-negative rates for the single TEOAE technique and the combination of TEOAE/ABR technique in infant at risk; comparing two screening protocols, we attempted to find the better technique that could give high sensitivity (the proportion of children without SNHL who have a negative test, i.e. are correctly identified by the screening protocol) and acceptable specificity (the proportion of children without SNHL who have a negative test, i.e. are correctly identified by the screening protocol) for hearing screening/diagnosis of infants at risk.

2. Materials and methods

This study was carried out by the Department of Audiology, University of Palermo and investigated all infants transferred from the births centers of Western Sicily, with the purpose to identify risk indicators for permanent congenital, delayed-onset, or progressive sensorineural hearing loss, from January 2010 to June 2011; the population consisted of 424 infants, 248 males and 176 females, ranging from 4 to 20 weeks of life at the time of first appointment. Following Ethical Committee approval, the study protocol was fully explained to parents, and written consent was obtained for each child. Out of 424 patients, 419 (98.82%) parents accepted the participation of their child to the study. However, as seven infants were lost to follow-up, the final response rate amounted to 412 patients (97.16%). The following characteristics were determined from the medical record of patients through a specific questionnaire answered by the mothers about the presence of: family history of permanent childhood hearing impairment (syndromic or nonsyndromic), consanguinity, low birth weight (<1500 g), cranio-facial abnormality (CFA) and syndromes associated to HL, physical findings such as white forelock, respiratory distress (IRDS), intensive care in excess of 5 days, pregnant maternal infection (TORCH), culture proven sepsis, culture proven or clinically suspected meningitis, cerebral bleeding, cerebral infarction, hyperbilirubinemia requiring phototherapy, ototoxic drugs administration (furosemide, dexamethason, vancomycin, gentamycin and tobramycin), acidosis, treatment for hypotension, patent ductus arteriosus ligation, hyponatremia.

An experienced audiologist and otorhinolaryngologist examined the condition of the external auditory canal and tympanic membrane with otoscopy, and nose, throat, head and face in search of ear anomalies and syndromic features related to hearing impairment.

The audiological assessment was performed by the same qualified bio-medical staff and consisted of ABR, TEOAE and tympanometry measurement. ABR measurements were recorded in a soundproof room; all children were in natural sleep or in calm conditions throughout the assessment. Both ears were sequentially tested. AMPLAID mk22 auditory evoked potentials system was used for testing the infants. After adequate preparation of skin, recording silver electrodes were attached to upper forehead (recording electrode), the ipsilateral mastoid process (reference electrode) and contralateral mastoid process (ground electrode). Thus the Fpz-M1-M2 electrode montage was used for recording the ABR. The acoustic stimuli consisted of unfiltered full square wave pulses of 100 μs duration and with alternating polarity. The clicks were delivered monaurally by a hand-held TDH-49 headphone, at a rate of 21/s. The analysis time was 15 ms. The recording bandwidth for click threshold determination was 100–2500 Hz. The electrode and inter electrode impedance were ensured to be below 5 kHz and 2 kHz respectively. Each run consisted of summing the responses to 2000 clicks. Click stimuli were presented starting at a level of 90 dB nHL. With step sizes of 10 dB the level was decreased until no response was found. The response threshold was estimated by the lowest level at which a response was found. An infant was considered to have passed the ABR test if a replicable wave V response (response present on at least two identical sound stimulation levels) was present at 30 dB nHL in both the ears while sensorineural hearing loss was defined as elevated ABR response thresholds (≥40 dB) in one or both ears. Moreover, the absolute latencies and interpeak intervals as well as the response thresholds were recorded. Experienced clinical specialists interpreted the ABR response waves. The response latencies in milliseconds were obtained by establishing the peak of the wave and reading out the digitally displayed time. From the latency intensity curves the level of conductive hearing loss was estimated. This has been described in the literature as a valid method to identify a conductive hearing loss [14]. TEOAE and tympanometry measurement were used to confirm the diagnosis of conductive hearing loss when available. In particular, the first one was performed using the Otodynamics ILO 288 USB II system with the standard settings; the stimulus level was set to 84 dB SPL, a number of 260 averages was used. Tympanometry was performed through Amplaid 766, with a probe frequency of 220 Hz and an air pressure range of −400 to −100 mmH_2O with automatic recording.

The parents of an infant suspected of hearing impairment were informed of the results of the initial test and received recommendations to return for a follow up evaluation after 3 weeks. If the results were confirmed the children were scheduled for early intervention programs (including hearing aids and rehabilitation) and auditory follow-up evaluation.

Statistical analysis was conducted with Matlab® computer programme; χ^2 test, odds ratio (*or*) and/or exact test of Fisher test were used, following usual conditions of application. Significance