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Prelingual sensorineural hearing loss and infants at risk: Western Sicily report

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ABSTRACT

Objective: To evaluate independent etiologic factor associated with sensorineural hearing loss (SNHL) in newborn at risk; to study the role of their interaction especially in NICU infants who present often multiple risk factors for SNHL.

Methods: The main risk factors for SNHL reported by JCIH 2007 were evaluated on 508 infant at risk ranging from 4 to 20 weeks of life, transferred to the Audiology Department of Palermo from the main births centers of Western Sicily. After a global audiological assessment, performed with TEOAE, tympanometry and ABR, the prevalence and the effect of risk factors was statistically studied through univariate and multivariate analysis on the total population (normal and deaf subjects).

Results: Fifty-one infants (10.03%) were diagnosed with SNHL (45 bilateral and 6 monolateral) with a mean hearing threshold of 87.39 ± 28.25 dB HL; from logistic regression analysis family history of hearing impairment (HI) and TORCH infections resulted independent significant risk factors ($P < 0.00001$ and $P = 0.024$ respectively). High SNHL percentages were evidenced also in NICU babies, due to the various pathologies and risk factors presented by these infants, and among newborns who suffered from hyperbilirubinemia requiring exchange transfusion (11.97% and 9.52% respectively). Craniofacial abnormalities (CFA) and syndromes associated to HI showed an important relationship ($P < 0.00001$) with conductive hearing loss (CHL). Multiple regression analysis of the variation in SNHL among NICU infants evidenced an increased risk for SNHL of 21.24% and of 19.33% respectively in preterm infants and in case of hyperbilirubinemia if respiratory distress is concomitant with these risk factors. It was also observed an higher risk of SNHL (99.66%) in case of coexistence of prematurity and hyperbilirubinemia. Finally among infants with very low birth weight (VLBW) it was evidenced a statistically difference between the mean weight of SNHL infants respect to NHL newborns ($P = 0.048$).

Conclusion: The high SNHL prevalence (10.03%) in our cohort underlines how infants at risk are more susceptible to suffer from SNHL; in particular NICU newborns have a 33% greater chance of developing SNHL, because of the presence of multiple risk factors (*or* = 1.33) and their interaction. As the number of coexisting risk factors increases, the prevalence of SNHL also increases ($r^2 = 0.93$).

1. Introduction

Occurring 1–2 infants every 1000 births [1,2], prelingual sensorineural hearing loss (SNHL) is more frequent than other congenital pathologies routinely screened at birth like phenylketonuria (1:15,000 newborns) and hypothyroidism (1:4500 newborns). This value can raise 10- to 50-fold in presence of risk conditions associated with SNHL, particularly among NICU babies.

Hearing impaired children present more difficulty developing verbal skills, language, learning and speech when compared with their normal hearing peers. Auditory deprivation influences also cognitive and affective development of infants making consequences in their interpersonal relationships [3,4].

So, promoting a system to early detect hearing impairment in the first months of life can prevent the severe consequences of hearing loss. This objective can be achieved by implementing a neonatal hearing screening extended to the entire population or at least to infants at risk as suggested by Joint Committee on Infant Hearing (JCIH) in the Position Statement of 2007 [5]. It is constituted by older risk registries [6] expanded with new independent neonatal risk factors like need for ventilation, use of oxygen supplementation, respiratory failure, low Apgar scores, acidosis, use of ototoxic drugs, treatment for hypotension, patent

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ductus arteriosus ligation, hyponatremia, and noise [7–13]. Interaction among risk factors has been evidenced [11].

Nevertheless a large diffusion of newborn hearing screening (NHS) in most parts of the developed world as an essential instrument of neonatal care, in Italy there is no stipulated modality to reach the objective of SNHL early detection in individual regions. It obviously leads to a ~30% of undiscovered SNHL with a medical diagnosis made later than 6th month of age. In Western Sicily, the tertiary Speech and Hearing Center of the University of Palermo with the collaboration of the main birth centers, developed a local program to early identify SNHL on newborns at risk.

The main purpose of this work was, studying the prevalence of SNHL on infants at risk in Western Sicily, to describe the distribution of risk factors associated with SNHL analyzing their role in the development of deafness and the effects of their interaction also through a multivariate analysis of the commonest risk factors inside the population. Understanding the joint effect of these risk factors could help to identify those newborns who necessitate within few days of life of a complete audiological assessment respect to an hearing screening.

2. Materials and methods

From January 2010 to December 2011, all infants transferred from the main births centers of Western Sicily to the Department of Audiology of the University of Palermo, were assessed for the presence of risk factors associated with permanent congenital, delayed-onset or progressive hearing loss; the study group consisted of 527 infants, 298 males and 229 females, ranging from 4 to 20 weeks of life at the moment of the first appointment. After ethical Committee approval, the study protocol was fully explained to patients or their guardians, and written informed consent was obtained from each patient. Out of 527 patients, 519 (98.48%) accepted to participate in this study but 11 infants were lost to follow-up monitoring. The final response rate resulted 96.39% corresponding to 508 infants. Through the discharge letter released by the birth centers and through a questionnaire answered by the parents the following risk factors were researched, making a distinction between prenatal and perinatal risk factors (JCIH): in the first group, family history of permanent childhood hearing impairment, consanguinity, pregnant maternal infection (TORCH) and drugs exposition during pregnancy; in the second group, premature birth (gestational age $\leq$ 37 weeks), intensive care in excess of 5 days, respiratory distress, hyperbilirubinemia requiring exchange transfusion, very low birth weight (<1500 g, VLBW), cranio-facial abnormality (CFA) and syndromes associated to hearing impairment (HI), perinatal infections (like sepsis and meningitis), ototoxic drugs administration (furosemide, dexamethason, vancomycin, gentamycin and tobramycin), acidosis, hyponatremia, head trauma. A specific analysis of the interaction between risk factors was performed among NICU infants, who are a subcohort characterized by exposition to multiple risk conditions that increase the probability to develop hearing loss.

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 same qualified bio-medical staff evaluated all infants with ABR, TEOAE and tympanometry measurement. ABR measurements were recorded through AMPLAID mk22 auditory evoked potentials system in a soundproof room; all children were in natural sleep or in calm conditions throughout the assessment. Both ears were sequentially tested. 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 100 dB HL. 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 recorded. An infant was considered to have passed the ABR test if a replicable wave V response was present at 30 dB HL in both the ears while SNHL 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 (CHL) was estimated [14]. TEOAEs and tympanometry measurement, when available, were used to confirm the diagnosis of CHL. Particularly, TEOAEs were evaluated 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 instead was performed with an Interacoustics AT 235H system using the standard settings and a 1 kHz probe-frequency and an air pressure range of −400 to −100 mmH_2O with automatic recording.

The parents of an infant with suspecting 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. Statistical analysis was conducted with Matlab® computer programme; χ^2 test, odds ratio (*or*) and/or exact test of Fisher test, linear regression (*r* value) were used, following usual conditions of application. ANOVA test and multivariate analysis were performed in NICU subcohort. Significance was set at 0.05.

3. Result

Five hundred and eight infants, ranging from 4 to 20 weeks of life, underwent global audiologic assessment at our department because of the presence of one or more risk factors associated with hearing impairment. With a male:female ratio of 1.37, the females were 214 (42.13%) and the males were 294 (57.87%).

Hearing loss was detected in 97 children (56 male and 41 female) but only in 51 cases (10.03%), a diagnosis of SNHL was made. With 32 males and 19 females suffering from SNHL, the male:female ratio was 1.68; no statistically significant difference in prevalence of SNHL among sex (χ^2 = 0.55, P = 0.45, *or* = 1.25) was observed. Forty five subjects (88.23%), for a total of 90 ears, were diagnosed with bilateral SNHL; a symmetric HL (inter-aural threshold difference < 30 dB HL) was evidenced in the 91.11% corresponding to 41 cases (80.39% of the total SNHL newborns), while in the 8.89%, corresponding to 4 cases, this SNHL was asymmetric (inter-aural threshold difference 30 $\geq$ dB HL). Finally, in 6 infants (11.76%), with a response threshold in the best hearing ear $\leq$ 40 dB HL, an unilateral SNHL was detected. Among the 96 ears suffering from SNHL, our study evidenced an hearing threshold mean value of 88.47 ± 28.28 dB HL (median 100 dB HL) for the 47 left ears, of 86.4 ± 28.48 dB HL (median 100 dB HL) for the 49 right ears, of 87.39 ± 28.25 dB HL (median 100 dB HL) for both ears without any difference between left and right ears (t = 0.35, f.d. = 94,