RESEARCH PAPER

Targeted Audiological Surveillance Program in Campania, Italy

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Objective: To identify children with postnatal hearing loss, a structured monitoring system is needed. The goal of this study was to describe a targeted surveillance program in Italy to identify children with postnatal hearing loss. **Methods:** Between January, 2013, and December, 2016, all children who received bilateral 'pass' result at the newborn hearing screening, and who were identified as having at least one risk factor, were referred for targeted surveillance. The hospital records of these children were retrieved. **Results:** Among children enrolled, 66 were identified with permanent hearing loss. The most frequent risk factors were family history (35%), prematurity (25.5%), low birthweight (19.2%), severe hyperbilirubinemia (19%), prolonged ventilation (15%) and congenital infection (12.5%). **Conclusions:** An audiological surveillance program in newborns who 'pass' in neonatal screening, but have risk factors, is effective in identifying permanent postnatal hearing disorders.

Keywords: *Hearing screening, Newborn, Postnatal hearing loss, Risk factors.*

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Institution and improvement of universal newborn hearing screening (UNHS) has led to permanent hearing impairments being detected and treated as early as possible. The prevalence of permanent hearing impairment in newborn babies is approximately 0.5-1.5/1000, but it may increase up to 3.5-6/1000 children in school age [2]. The increase is due to the onset of postnatal hearing loss, which is missed in UNHS programs. The Joint committee on infant hearing screening (JCHI) recommends that an appropriate early identification and treatment of permanent hearing disorders, requires audiological surveillance in follow up on all newborns 'pass' but with risk factors for delayed/ progressive and acquired hearing loss [3].

We, herein, report the results of a targeted surveillance program based on selected risk factors in Campania region, Italy.

METHODS

A retrospective analysis of the audiological surveillance activities was performed for the period from January, 2013 to December, 2016. Well babies and neonatal intensive care unit (NICU) babies who received bilateral pass result during the newborn hearing screening and were neonatal intensive care unit one or more risk factor, were referred for targeted surveillance. The mean (SD) age of babies at the time of screening was 4 (2,1) weeks. The data of the present study were drawn from a database which included individual records for each child.

Since 2003, well-babies in Campania region are screened via two stage transient otoacoustic emission (TEOAE): The first in the course of the second or third day of life and the second between 3-4 weeks of age, if a refer result is obtained. TEOAE and automated auditory brainstem response (A-ABR) are reserved for infants under intensive care, prior to discharge. Infants who fail both screenings, either bilaterally or unilaterally, are referred to the nearest pediatric audiology service to perform a comprehensive audiology evaluation. In case of hearing impairment identification, a third level multidisciplinary diagnostic work-up together with appropriate management is provided by the regional reference center (RRC), at the audiology and vestibology unit of the neuroscience department of the university of Naples 'Federico II' [4]. Since 2013, this unit is also in charge of coordination of the audiological surveillance program according to the Position committee on infant hearing screening, 2007.

Children referred for surveillance appointment are accepted according to the following protocol: Audiological assessment in the third level center, every 6 months up to the age of 3 years and then annually up to

the age of 6 years in the presence of risk factors such as cytomegalovirus (CMV) and rubella intrauterine infections; every 6 months up to the age of 3 years, in children affected by syndromes associated with progressive or late-onset hearing loss e.g., Pendred syndrome, distal tubular renal acidosis, Waardenburg syndrome (type II), branchio-otorenal syndrome, Usher syndrome (type II and III), Stickler syndrome, CHARGE syndrome, Down syndrome, Turner syndrome, Alport syndrome, neurodegenerative disorders such as Hunter syndrome, sensory-motor neuropathies such as Friedreich ataxia, Charcot-Marie-Tooth syndrome); every 6 months up to the age of 2 years in case of family history of progressive permanent infant hearing loss, severe asphyxia; at 9-12 months in the case of prolonged ventilation for more than 5 days, craniofacial anomalies including cleft palate and, audiological evaluation at the immediate third level center in every phase of childhood or adolescence in case of chemotherapy, trauma, culture positive infections associated with sensorineural hearing loss, ototoxic drugs, reports from family pediatricians or other health workers and educators, or meningitis.

The audiological evaluation is done as per the age group of children. Test battery is click-auditory brainstem responses (ABR), transient evoked oto-acoustic emissions (TEOE), tympanometry at 3-9 months; TEOE, tympanometry, visual reinforcement audiometry at 9-12 months; and visual reinforcement audiometry, conditioned play audiometry and tympanometry at 3.5 years.

Degrees of hearing loss is based on the Bureau International for Audiophonology (BIAP) [5] classification viz normal (< 20 dB HL), mild (21-40 dB HL), moderate (41-70 dB HL), severe (71-90 dB HL) and profound (> 90 dB HL).

All families of children confirmed with hearing loss were offered a genetic evaluation and counselling. The evaluation, included a review of family history of specific genetic disorders or syndromes, genetic testing for gene mutations such as *GJB2*, *GJB6* (connexin-26 and 30), and syndromes commonly associated with early-onset hearing loss.

Statistical analyses: Descriptive statistics were used, and Pearson chi-squared analyses were performed in order to identify variables of significance.

RESULTS

The flow of all children referred to the targeted surveillance program is shown in **Fig. 1**. A total of 165416 children were eligible for UNHS (158386 'well babies' and 7030 'high risk'), of which 2752 (1.6%) children had a 'refer' result and underwent a comprehensive audiology evaluation. Another 2340 children had a 'pass' result, but with at least one risk factor and were referred for audiological surveillance program. Thus the recorded rate was 1.41% (2340/165416) in the period under investigation. With regards to individual risk factors, the largest proportion of referrals were generated from family history (35%), low birthweight (19.2%), prematurity (25.5%), severe hyperbilirubinemia (19%), prolonged ventilation (15%) and congenital infections (12.5%) (**Table I**).

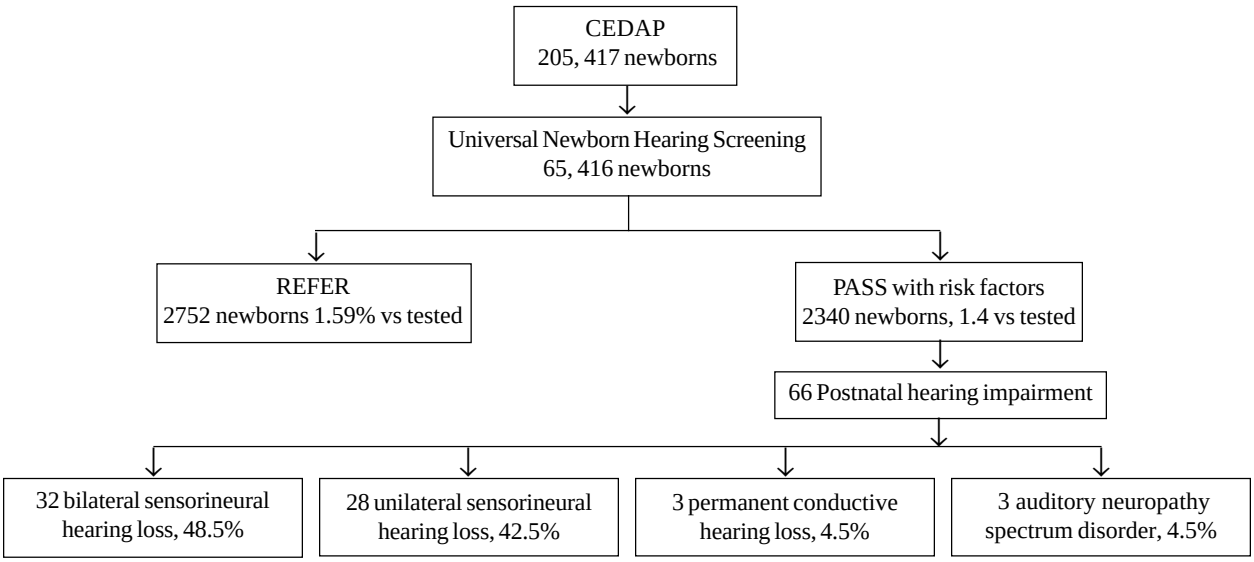

Fig. 1 *Results of the audiological surveillance program.*

WHAT THIS STUDY ADDS?

- We provide the results of a targeted surveillance program in Italy using a risk-factor list, providing information on prevalence of risk factors and characteristics of postnatal hearing loss.

Through the targeted surveillance program, a total of 66 (0.4%) children were identified with postnatal hearing loss - the most frequent risk factors were family history (26%), and prematurity (15%). Other less common risk factors reported were cleft palate (2 cases) and Pendred syndrome (1 case). Assisted prolonged ventilation (P=0.003), family history (P=0.003), cranio-facial anomalies (P=0.01), and congenital infections (P=0.02) were the significant risk factors for postnatal hearing loss.

Postnatal hearing loss exhibited the following types: 32 (48.5%) bilateral sensorineural hearing loss, 28 (42.5%) unilateral sensorineural hearing loss, and 3 (4.5%) each with permanent conductive hearing loss and auditory neuropathy spectrum disorder (ANSD). The degree of hearing loss was as follows: mild, 26 (39%); moderate, 10 (15%); severe, 6 (9%); and profound, 24 (37%). The mean (SD) age at diagnosis was 9.7 (7.8) months.

Table I Characteristics of Children in Targeted Follow-up Group and With Postnatal Hearing Loss

Characteristics	*Targeted follow-up group, n=2340*	*Postnatal hearing loss*
Male gender	1077 (46)	32 (48)
Number of risk factors		
1	1570 (67.1)	-
2	262 (11.2)	-
3	407 (17.4)	-
4	29 (1.2)	-
5	72 (3.1)	-
Type of risk factors		
Family history	819 (35)	17 (26)
Prematurity	596 (25.5)	10 (15)
Low birthweight	449 (19.2)	9 (13.6)
Hyperbirilubinemia	444 (19)	7 (10.6)
Prolonged ventilation	351 (15)	5 (7.6)
Congenital infections	292 (12.5)	5 (7.6)
Neonatal asphyxia	187 (8)	4(6)
Craniofacial anomalies	140 (6)	2 (2.5)
Syndromes	112 (4.8)	1 (1.5)
Pediatricians/ caregivers reporting	70 (3)	0
Bacterial meningitis	5 (0.2)	0 (0)

DISCUSSION

Out region's surveillance program aims to detect postnatal and progressive hearing loss to avoid after-effects due to late diagnosis [6,7]. In the present work, we have used the list of risk factors proposed by JCIH in 2007 replacing the item 'entry in NICU' with prematurity (<37 weeks) and low birthweight (<2500g), in order to avoid too many referrals in the program for these risk factor and to make the follow-up protocol more feasible, effective and selective, and extracorporeal membrane oxygenation (ECMO) was replaced with severe asphyxia [8,9]. We also redefined the audiological protocol proposed in relation to both the timing for each risk factor and the methodology by age group, as described above.

Our data confirm the increase of permanent hearing disorders in the postnatal period; which, in our evaluation, reaches a rate of 2.6%. In the analyzed sample, a prevalence of permanent hearing impairment of a mild degree is evident. The immediate identification is particularly relevant because of the negative impact on the linguistic and curricular outcomes of this hearing loss [10]. In contrast to Beswick, et al. [11], where neonatal asphyxia was the primary cause of postnatal hearing loss, we found family history, congenital infections, and prolonged mechanical ventilation as significant risk factors.

The epidemiological and clinical aspects of the hearing impairment identified in the postnatal period in the current study supports the need and effectiveness of audiological surveillance during early childhood [6,7,12]. Audiological surveillance allowed us to identify not only cases of progressive hearing loss that probably arose in the postnatal period, but also congenital forms that had avoided the neonatal auditory screening, as the high presence of mild forms exhibits. The significant risk factors identified in this study need further evaluation in other regions and different populations.

Disclaimer: The views in this article are those of the authors and do not necessarily represent the official views of the Disability Research and Dissemination Center or the Centers for Disease Control and Prevention.

Ethical clearance: University of Naples 'Federico II' Ethics Committee; No. 56/18, dated March 26, 2018.

Contributors: RM: conceptualized and designed the study and drafted components of the initial and final manuscript and had a major role in the written manuscript as submitted; AF, EM:

participated in the conceptualization and design of the study, oversaw the collection of the data; EB: conducted the statistical analyses; CM, CL, GA: participated in the review of the literature, assisted in data collection, drafted sections of the initial manuscript, and participated in editing of the final manuscript as submitted; ME: conducted the initial literature review; FT: supplied critical background material for the study, and critically reviewed the manuscript. All authors approved the final manuscript as submitted.

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