International Journal of Pediatric Otorhinolaryngology 82 (2016) 92–97

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Universal newborn hearing screening in Umbria region, Italy

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ARTICLE INFO

Article history:
Received 18 September 2015
Received in revised form 4 January 2016
Accepted 6 January 2016
Available online 14 January 2016

Keywords:
Neonatal screening
Hearing loss
Otoacoustic emissions
Auditory brain stem evoked responses
Italy

ABSTRACT

Objective: In Italy, universal newborn hearing screening (UNHS) was first introduced in some regions from 1997. Umbria Region has launched a UNHS program in all hospitals, which has been implemented throughout the region since July 2010. Before UNHS implementation in Umbria region, the average age of identification of congenital hearing loss was around 32 months of age with an average age of initial amplification treatment at least 2 months later. The coverage rate of newborn screening was only 34.4% in 2006.

The aim of this study was to examine the results of this program and its evolution in the first 2.5 years since implementation in our region.

Methods: Since July 2010, all 11 birth centers and hospitals in Umbria region have been involved in a UNHS program. The screening involves the automated otoacoustic emissions (AOAE) test and automatic auditory brain stem response (AABR) audiometry. The number of screening stages and tests used were different depending on whether the infants had audiological risk factors or not.

Results: A total of 20,841 babies were born in the hospitals involved of whom 20,051 were well born babies (WB), while 790 babies (3.8%) presented identified audiological risk factors (BRF). The overall coverage rate in the study period was 93.8%. The prevalence of hearing loss was 2‰ for WB infants and 4.3% for BRF. Mean age at diagnosis was 5.31 ± 3.95 and 11.28 ± 7.73 months in the WB and BRF groups, respectively.

Conclusions: UNHS has allowed us to substantially increase the coverage rates and decrease the mean age at diagnosis and subsequent treatment. The identification of audiological risk factors is very important for adequate screening and follow-up. However the Joint Committee on Infant Hearing 2007 quality indicators and benchmarks for screening have not yet been fully achieved and there is still scope for some improvement. This could be achieved with a closer cooperation among institutions, parents, pediatricians, and ENT doctors.

1. Introduction

Hearing loss is one of the most common problems in newborn babies. About 1–2 in every 1000 children are born profoundly deaf [1]. Overall permanent congenital hearing loss greater than 25 dB HL in the poorer ear is present in at least three infants per thousand and this prevalence increases with growth [2]. Furthermore, the prevalence of hearing loss is estimated to be between 2.5% and 10% among high-risk infants while infants in neonatal intensive care units (NICU) are 10 to 20 times more likely to have significant hearing loss compared with the healthy population [3,4]. However only half of permanent congenital hearing defects occur in children with risk factors [5]. With regard to the Italian data, in a recent national survey, the prevalence of neonatal sensorineural pre-lingual bilateral hearing impairment >60 dB HL was found to be 0.72 per 1000 inhabitants [6].

Early treatment and rehabilitation are essential to prevent the delayed development of speech and language. Therefore, a child's hearing impairment should be detected as early as possible, so that timely and appropriate interventions can be made [7]. In the absence of a newborn hearing screening program, moderate to severe hearing loss is not identified before a mean age of 2 years and not treated earlier than 40 months, while mild hearing impairment is identified even later, sometimes not until school age [8]. For all these reasons, the most important international

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http://dx.doi.org/10.1016/j.ijporl.2016.01.007
0165-5876/

organizations recommend the implementation of universal newborn hearing screening (UNHS) [9–12].

The Joint Committee on Infant Hearing (JCIH) 2007 quality indicators and benchmarks for screening are as follows: for newborn infants who have completed the screening by 1 month of age, the recommended benchmark is more than 95% (age correction for preterm infants is acceptable). For newborn infants who have failed initial screening and failed any subsequent rescreening before comprehensive audiological evaluation, the recommended benchmark is less than 4%, and for those returning for follow-up, the benchmark is at least 90% [12]. This last benchmark has been increased by JCIH in 2007, since it was 70% in the position statement of 2000 [13]. The detection of hearing loss before 3 months of age and appropriate intervention no later than 6 months of age are required. The JCIH has identified the neonatal audiological risk factors which UNHS programs must take into account in their implementation [12]. This can be seen in Table 1. In fact, in the presence of these risk factors, the so-called "auditory neuropathy" (an audiological entity characterized by normal otoacoustic emissions and altered auditory brainstem response) [14], and progressive and late onset hearing loss are more common [15]. The identification at birth of any of these audiological risks is of fundamental importance since, for the reasons previously described, these children have to be enrolled into a specific screening protocol with subsequent audiological surveillance.

In Italy, UNHS was first introduced in some regions from 1997. The nationwide coverage rates of newborn hearing screening were 29.3% in 2003 and 48.4% in 2006 [8].

Before UNHS implementation in the Umbria region, the average age of identification of hearing loss in children was around 32 months of age with an average age of initial amplification treatment at least 2 months later [16]. The coverage rate of newborn hearing screening in Umbria was only 34.4% in 2006 [8].

Umbria Region, with its DGR (Regional Council Resolution) n°789 dated 21 May 2007, has launched a UNHS program in all hospitals, which has been implemented throughout Umbria since July 2010, to establish a program with high coverage, low referral rate, high follow-up rate, and early intervention. In fact, Umbria is one of five regions that currently apply UNHS in Italy.

The aim of this study is to examine the results of this program and its evolution in the first 2.5 years since its implementation in our region.

Table 1
Audiological risk factors for neonatal hearing loss according to the JCIH position statement of 2007.

- *Caregiver concern* regarding hearing, speech, language, or developmental delay
- *Family history* of permanent childhood hearing loss
- *Neonatal intensive care* of more than 5 days or any of the following regardless of length of stay: *extracorporeal membrane oxygenation (ECMO)*, assisted ventilation, exposure to *ototoxic medications* (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix), and *hyperbilirubinemia* that requires exchange transfusion
- *In utero infections*, such as cytomegalovirus (CMV), herpes, rubella, syphilis, and toxoplasmosis
- *Craniofacial anomalies*, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
- *Physical findings*, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss
- *Syndromes* associated with hearing loss or progressive or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes, including Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson
- *Neurodegenerative disorders*, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome
- *Culture-positive postnatal infections* associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis
- *Head trauma*, especially basal skull/temporal bone fracture that requires hospitalization.
- *Chemotherapy*

2. Materials and methods

Since July 2010, all 11 birth centers and hospitals in Umbria region have been involved in a UNHS program. All children born between July 2010 and December 2012 and screened in the hearing screening program, both well born babies (WB) and babies born with identified audiological risk factors (BRF), were included in the study.

The screening involves the automated otoacoustic emissions (AOAE) test and automatic auditory brain stem response (AABR) audiometry. The number of screening stages and tests used were different depending on whether the infants had audiological risk factors or not.

2.1. Well born babies (WB)

The AOAE tests were performed 24–36 h after birth by dedicated trained personnel (1st stage). With a negative test in both ears (*pass*), the screening was considered completed. The same test was repeated at 1 month of age at the same birth center if the baby had failed the first hearing test (*refer*) (2nd stage). If the neonate also failed the second test, the infant was referred to a second level diagnostic hospital (Perugia or Terni) for further investigation before the age of 3 months, to carry out both AOAE and AABR audiometry (3rd stage). Infants with *refer* results were sent to a specialized center (Perugia) for confirmation of a diagnosis of permanent congenital hearing loss (PCHL), to obtain an etiopathogenetic diagnosis, to define the entity of hearing loss, and to initiate treatment with hearing aids or implanted prosthetic devices (4th stage). The screening protocol in WB babies can be seen in Fig. 1.

2.2. Babies with audiological risk factors (BRF)

The AOAE tests and AABR audiometry were performed as soon as possible after birth when the general conditions allowed it, by dedicated trained personnel (1st stage). Performing AABR in all infants born with audiological risk factors is necessary to identify the auditory neuropathy. With a negative test in both ears (*pass*), the screening was considered completed. Infants with *refer* results

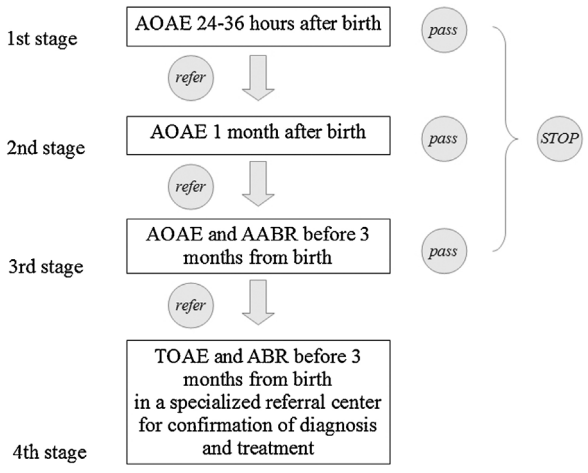

Fig. 1. UNHS protocol for well born babies (WB).