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Age at diagnosis of deaf babies: A retrospective analysis highlighting the advantage of newborn hearing screening

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Summary

Objective: Aim of the study was to assess the mean age at diagnosis of bilateral congenital hearing loss in the Audiology and Phoniatry Centre of the University of Turin, pointing out, by North-West Italy experience, the role of the newborn hearing screening in anticipating the age of diagnosis.
Methods: This was a retrospective study. Forty-six congenital deaf babies were reviewed and age at diagnosis was assessed for each, taking in consideration the role of hearing loss risk factors. Eighteen babies (39%) were sent by the centres that participate to the newborn hearing screening program while 28 (61%) came for parental or pediatrician suspicion of hearing loss and for general language delay. Sixteen babies (35%) presented risk factors for hearing loss.
Results: The mean age of identification of severe to profound hearing loss was 20.5 months (S.D. = 15.3) in the whole group; considering the group of 28 babies not screened the mean age was 29.3 months (S.D. = 13.4). This value decreased to 6.8 months (S.D. = 3.6) in the group which underwent screening programme. This difference was statistically significant at Student's *t*-test ($p < 0.001$).

The average ages of diagnosis for healthy versus high risk children were significantly different only in the group of screened babies ($p < 0.05$).
Conclusions: Childhood hearing impairment is one of the most common of congenital disorders, and even though there is a general trend of early identification, in reality

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age of diagnosis is as yet still too late even in developed countries. Our results show that newborn hearing screening could reduce the age at which infants with hearing loss are diagnosed and treated; this would improve speech, language, auditory outcome and the quality of parents and infant life.

1. Introduction

Childhood hearing impairment is one of the most common of congenital disorders. Prevalence is about 1/1000 if the analysis is limited to children with a 50 dB or greater bilateral loss [1].

The need for identifying hearing loss within the first few months of life was advocated in Great Britain more than 50 years ago [2]. Since then, a variety of procedures to identify congenital hearing loss in neonates have been tried [2]. Fortunately, advances in technology have at long last made neonatal identification a realisable goal. In the late 1990s, the development of rapid, low cost screening tests made it feasible to implement screening programs for all newborns for congenital hearing loss during birth hospitalization. The latest approaches, being advocated by many in Europe and in the USA, are the measurement of otoacoustic emissions (OAE) and automated auditory brainstem response (AABR), tests that provide complementary information about the state of the auditory system [3].

It has been demonstrated that newborn hearing screening (NHS) leads to earlier identification and treatment of infants with hearing loss [4,5]. Studies of state wide NHS program in the United States have demonstrated that the mean age of identification of hearing impairment has decreased from 12–24 months before NHS programs were introduced [5], to 3–6 months since their introduction [4]. Moreover the mean age at which infants receive hearing aids has been reduced from 13–16 months before NHS programs began to 5–7 months following their introduction [4]. In a large controlled study comparing in-hospital NHS with no screening, NHS significantly increased the number of infants with hearing loss referred to audiologists by the age of 6 months and increased the probability that infants with moderate and severe hearing loss would be diagnosed by the age of 10 months (57% versus 14%) [6].

For this reason influential groups such as the National Institute of Health, the American Academy of Paediatrics in the USA and the 1998 European Consensus Development Conference attended by representatives from most western European countries have recommended UNHS use [2].

The implementation of screening programs continues to grow. In North-West Italy a NHS has been in operation since January 2002 in 26 neonatology departments. We use a different protocol for newborns without audiological risk (healthy babies) and for those with audiological risk (at risk babies), identified according to the criteria proposed by the Joint Committee on Infant Hearing (2000) [7]. Healthy babies are screened with OAEs while in at risk babies it is suggested to include an AABR test in order to exclude auditory neuropathy [8]. In our program there are four steps for healthy babies, three of which based on transient evoked otoacoustic emission (TEOAEs) recording. The first is carried out before discharge, the second after 15–30 days and the third after 30–60 days; the second and the third are performed in case of "refer" scoring at the previous steps. The fourth step is based on AABR for babies scored as "refer" at the TEOAEs step.

The infants with a "refer" score at AABR underwent a complete audiological evaluation before 6 months of age.

At risk babies are tested with TEOAEs immediately before the discharge; an audiologic questioner is always given to the parents that have to fill in when the baby is 6 months and every 6 months up to 36 months in order to perform a follow up to avoid to loose hearing problem that can be late onset. In case of "refer" at TEOAE, next step is AABR after 20–30 days. Selected babies underwent AABR before 3 months of life even with a "pass" at the first TEOAEs.

Congenital hearing loss has been linked with lifelong deficits in speech and language acquisition, poor academic performance, personal-social and emotional challenges [9]. There are no prospective, controlled studies that directly examine whether newborn hearing screening and earlier intervention result in improved speech, language, or educational development, but several retrospective studies have variously concluded that infants entering treatment programs at younger ages, or infants identified in hospitals with universal screening programs, have better long-term language outcomes [2]. Moreover, the studies demonstrates that intensive early intervention can alter positively the cognitive and developmental outcome of young infants with disabilities thanks to the brain plasticity [10].

The aim of the study was to investigate the outcomes, in terms of age of diagnosis of severe to profound hearing loss, of the universal hearing-screening project that has been started in North-West Italy.