

Risk Factors for Congenital Hearing Loss: Which Are the Most Relevant?

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ABSTRACT

Introduction: Congenital hearing loss is a multifactorial disease that affects mostly premature newborns exposed to certain risk factors. The recent widespread introduction of newborn hearing screening in Tîrgu Mureş, Romania prompted the need for a multidisciplinary centralized study on the subject. **Case series presentation:** We collected data from 340 neonates born in 2014 who had undergone neonatal hearing screening with otoacoustic emissions. Our focus group consisted of 137 neonates with a gestational age of less than 37 weeks. We collected data on the pregnancy, birth, and perinatal period, and then analyzed the impact of infections, treatments — with or without ototoxic potential, hypoxia, mechanical ventilation, intrauterine malnutrition, and the presence of malformations. Premature neonates showed a significantly higher number of REFER results than those with a normal gestational age. The rates of low birthweight, head circumference, and infant length; low Apgar scores; hypoxia in the perinatal period; the presence of placental pathology, mechanical ventilation, and perinatal infections; and the concomitant potentially ototoxic treatment were significantly higher in the REFER group. The age of the mother, bilirubin levels, and oxytocin use during birth did not prove to be relevant. **Conclusions:** Known risk factors can cause hearing loss in a considerable number of newborns and mandate a multidisciplinary approach to the problem at hand. The referral of these patients to an ENT specialist and their correct management according to an individualized hearing recovery plan is crucial

Keywords: congenital hearing loss, risk factors, neonatal hearing screening, otoacoustic emissions

INTRODUCTION

Congenital hearing loss affects 2–3 out of 1000 newborns. If left unnoticed and untreated, it may cause serious delays in the speech and cognitive development of the child.¹ Hearing loss can be classified as conductive, sensorineural, or mixed. Conductive hearing loss involves any pathology that limits sound reaching the inner ear such as meatal atresia, ossicular chain malformation, acute or chronic otitis media with effusion, or tympanic membrane perforation. In the case of sensorineural hearing loss, the inner ear or the auditory nerve is affected. Mixed hearing loss is a combination of sensorineural and conductive hearing loss. Sensorineural hearing loss that occurs at or shortly after

birth can have hereditary or non-hereditary causes. The latter include intrauterine TORCH infection (toxoplasmosis, cytomegalovirus, rubella, and herpes), meningitis, treatment with ototoxic or teratogenic medication during pregnancy (aminoglycosides, loop diuretics, isotretinoin, or antimalarials), prematurity, and hypoxia in the perinatal period. Hereditary causes can be classified as syndromic (Alport, Norrie, Usher, Pendred, Waardenburg, Jervell, and Lange-Nielsen) or non-syndromic (autosomal dominant, autosomal recessive, x-linked, or mitochondrial), and may be present at birth, show signs of progression, or manifest later in life, when the child is older.²⁻⁷

Treatment options vary according to the underlying cause. Certain types of conductive hearing loss (e.g., otitis media with effusion, cholesteatoma etc.) may be amenable by medical and surgical treatment (adenotomy, ventilation tube insertion, tympanoplasty, bone-anchored hearing aid etc.), while in the case of sensorineural hearing loss, treatment options include hearing aids or cochlear implantation, depending on the degree of hearing loss. Cochlear implantation is indicated for severe and profound neurosensory hearing loss.⁸⁻¹⁰

Screening for congenital hearing loss in newborns represents the first and most important step in the evaluation of this condition.¹¹ Otoacoustic emissions and auditory brainstem response audiometry are used for such screening purposes, the latter often being the preferred method in order to avoid overlooking auditory neuropathy.¹² Screening can be generalized or targeted. Generalized screening means each and every newborn undergoes the screening procedure, while targeted screening indicates that only those with certain known risk factors are screened. Those affected should undergo ENT, audiological, and genetic examination.¹³

Diagnostic audiological investigation methods include otoacoustic emissions, auditory brainstem response, and auditory steady-state response.¹⁴ Otoacoustic emission (OAE) tests are an objective hearing investigation method meant to determine cochlear status, more specifically hair cell function. A normally functioning cochlea both receives sounds and also produces low-intensity sounds called OAEs. These sounds are most likely produced by the contraction of the outer hair cells in the cochlea. With the help of supersensitive microphones, these sounds can be measured and recorded. There are four main types of otoacoustic emissions: spontaneous otoacoustic emissions (SOAEs), transient otoacoustic emissions (TOAEs) or transient evoked otoacoustic emissions (TEOAEs), distortion product otoacoustic emissions (DPOAEs), and sustained-frequency otoacoustic emis-

sions (SFOAEs).¹⁵ Auditory brainstem response (ABR) and auditory steady state response (ASSR) audiometry are objective neurologic investigation methods of auditory brainstem function in response to sound. A brief click or tone pip is emitted from an insert earphone, and the elicited electrical signal is measured by electrodes, typically placed on the forehead, mastoid process, and the occipital region.¹⁶ The purpose of our study was to look for correlations between potential risk factors for congenital hearing loss and neonatal hearing screening results in a population where no such study had ever previously been performed, in the hope of raising awareness about congenital hearing loss.

CASE SERIES PRESENTATION

We collected data from 340 neonates, born in 2014 at the County Emergency Clinical Hospital of Țirgu Mureș, Romania, who had undergone neonatal hearing screening with otoacoustic emissions using a Natus Echo-Screen device. Both male and female candidates were randomly selected, and according to the principle of generalized screening, every newborn underwent neonatal hearing screening using TEOAE (1.5–3.5 kHz) and DPOAE (2–4 kHz). The accumulated data was analyzed with descriptive statistics in SPSS software. In each case the mother signed the official consent that medical information could potentially be used in later studies.

The screening was performed at least 12 hours after birth, as close as possible to the discharge, and both ears were screened individually. The initial screen consisted of a maximum of two attempts on each ear. In the case of a REFER result, the patient was scheduled for a re-screen using evoked otoacoustic emissions. The re-screen was usually performed prior to the age of 1 month. Both ears were re-screened individually with a maximum of two attempts on each ear. Although a REFER result upon re-screening does not necessarily mean that the infant has congenital hearing loss (inadequately performed test, meconium in the external ear canal etc.), patients with a REFER result upon re-screen were referred for an ENT and diagnostic audiological evaluation.

We included 137 premature neonates in our study and collected data about the pregnancy, birth, and perinatal period. We analyzed the impact of infections before, during, and shortly after birth, treatments with ototoxic potential, perinatal hypoxia, placental pathology, mechanical ventilation, and possible correlations between gestational age, Apgar score, the need for care at the neonatal intensive care unit, administration of oxytocin during labor, and

TABLE 1. Study results

	PASS	REFER in one ear	REFER in both ears
Mean birthweight (g)	2760.87	1914.35	1895.71
Mean fetal length (cm)	50.02	42.91	45.29
Mean head circumference (cm)	32.17	29.84	29.43
Mean gestational age (weeks)	37.30	34.13	33.64
Mean Apgar score at 1 minute	8.60	6.81	7.92
Mean Apgar score at 5 minutes	9.31	7.90	8.62
Mean bilirubin level (mg/dL)	7.40	6.47	5.57

hyperbilirubinemia. We found there was no significant difference among genders ($p = 0.208$). Head circumference ($p = 0.005$), fetal length ($p < 0.001$), and birthweight ($p < 0.001$) were significantly lower in the REFER group. Gestational age was significantly lower in the REFER group ($p = 0.001$). The age of the mother did not prove to be statistically significant ($p = 0.123$). Those with uni- or bilateral REFER after re-screening had significantly lower Apgar scores at 1 and 5 minutes after birth ($p = 0.001$). Neonates affected by placental pathology (nuchal cord, placental abruption, chorioamnionitis etc.) had a REFER result in 22.9% of cases, while those without placental pathology had a REFER result in 9.2% of cases, this representing a statistically significant difference ($p = 0.016$). Oxytocin use during labor did not prove to be significant ($p = 0.140$). Those affected by perinatal hypoxia had a REFER result in 25% of cases (17.2% in one ear, 7.8% in both ears), while those who were unaffected had a REFER rate of 7.9% (4.5% in one ear, 3.4% in both ears), the result proving to be statistically significant ($p < 0.001$). Hyperbilirubinemia did not prove to be a statistically significant risk factor. In our study those in the REFER group actually had lower (statistically insignificant) levels of bilirubin than those in the PASS group (Table 1). According to several large-scale studies on the subject, hyperbilirubinemia is generally accepted to be an important risk factor for congenital hearing loss.^{17,18} Among those who did not receive any potentially ototoxic medication, 94.6% had a PASS result and 5.4% had a REFER result (3.6% for one ear, 1.8% for both ears), while among those who received potentially ototoxic medication (aminoglycosides and loop diuretics), only 76.9% had a PASS result and 23.1% had a REFER result (13.9% for one ear, 9.3% for both ears), the difference proving to be statistically significant ($p < 0.001$).

A systematic literature review of 40 articles on the subject has found similar results.^{19,20} Neonates admitted to the neonatal intensive care unit had a PASS rate of 82.4%, while those who were not admitted there had a PASS rate of 91.6%, the difference proving to be statistically signifi-

cant ($p = 0.048$). Neonates requiring mechanical ventilation had a PASS rate of 71.7%, while for those without the need for mechanical ventilation the PASS rate was 91.5%, the difference proving to be statistically significant ($p < 0.001$). Neonates with perinatal infections had a REFER rate of 23.9%, while for those who did not the REFER rate was 5%, the difference proving to be statistically significant ($p < 0.001$). We encountered congenital birth defects with possible association with hearing loss in three distinct cases. We had REFER results in all three cases.

CONCLUSIONS

Known risk factors can cause hearing loss in a considerable number of newborns and mandate a multidisciplinary approach to the problem at hand, ideally by a team consisting of otolaryngologists, audiologists, geneticists, speech pathologists, and educational specialists. The age of the mother, oxytocin use during labor, and bilirubin levels did not prove to be a risk factor, while low birthweight, head circumference, length, gestational age, use of potentially ototoxic medication, perinatal infections, the presence of placental pathology, perinatal hypoxia, neonatal intensive care, and mechanical ventilation all represent risk factors. While the Apgar score is not a risk factor in itself, a low Apgar score must direct our attention to the possibility of congenital hearing loss. The percentage of patients lost in the follow-up procedure, in our opinion, can be lowered by carefully informing the parents of the problem at hand. False negative and false positive screening results both represent serious issues that need to be addressed. The timely referral of these patients to an ENT specialist and their correct management according to an individualized hearing recovery plan is crucial.

CONFLICT OF INTEREST

The authors would like to state that there is no conflict of interests and that no funding was received.

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