ETIOLOGICAL INVESTIGATION OF DEAFNESS IN NEONATES SCREENED IN A UNIVERSAL NEWBORN HEARING SCREENING PROGRAM

Investigação etiológica da deficiência auditiva em neonatos identificados em um programa de triagem auditiva neonatal universal

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ABSTRACT

Purpose: to describe the results of etiology of deaf in neonates screened in a universal newborn hearing screening program. **Methods:** a descriptive, cross-sectional and prospective study. The study included all newborns diagnosed with hearing loss identified in a universal newborn hearing screening program from August 2003 to December 2006. The etiology of deaf was determined after detailed anamnesis performed by the otorhinolaryngologist; survey of serological tests for toxoplasmosis, rubella, cytomegalovirus, herpes, syphilis and HIV; tomography of the temporal bone and genetic tests. **Results:** 17 neonates were diagnosed with hearing loss in the period studied. 64.7% of cases presented as probable causes prenatal etiology, 29.4% perinatal causes and one child (5.9%) had unknown etiology. Of prenatal causes, 36.4% had confirmed genetic origin and 36.4% presumed etiology of heredity. We confirmed the presence of congenital infections in 18.2% of cases and one child (9%) had craniofacial anomalies as a possible etiology. The degree of hearing loss more frequently observed in the subjects studied was the profound (47.1%). **Conclusion:** the increased occurrence of etiologies in this study was of prenatal origin, followed by perinatal origin.

KEYWORDS: Hearing Loss; /etiology; Screening

INTRODUCTION

The implementation of Universal Newborn Screening (UNHS) Programs is a way to enable early diagnosis of hearing impairments, and its purpose is to minimize the negative effects that this disorder may bring to the child's life[1-3].

The earlier a diagnosis of hearing impairments is obtained, followed by the adaptation of sound amplification devices and by auditory (re) habilitation, the better the results in these children's auditory and language development, since plasticity in the central nervous system is greater during the first year of life [3,4].

Alongside the early diagnosis of deafness and the measures of (re) habilitation, it is important to search for the etiology of the hearing impairment, as recognizing its cause may direct the conduct to be adopted in (re) habilitation. It should be noted that in Brazil there are few etiological studies about hearing impairment in newborns[4].

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Conflict of interest: non-existent

Hearing loss may be genetic or acquired. In this last case, the causes may many times be avoided, such as, for example, infections that occur during pregnancy, meningitis or even the use of ototoxic medication[5].

In newborns and young infants hearing loss may occur due to prenatal causes (genetic inheritance, genetic syndromes, malformations of the inner ear, congenital infections from viruses of rubella, cytomegalovirus, herpes, toxoplasmosis and syphilis, and also by the use of teratogenic substances during pregnancy); perinatal causes (anoxia, prematurity, birth weight below 1500 grams, hyperbilirubinemia, cranial trauma, sound trauma) or postnatal causes (metabolic issues such as hypothyroidism and diabetes, viral infections such as rubella, varicella zoster, influenza, mumps, cytomegalovirus, among others, labyrinthitis and bacterial meningitis, encephalitis and chronic middle ear infections). There are other less frequent causes, such as auto-immune diseases, renal tubular acidosis, neoplasms, cranial trauma, acoustic trauma and use of ototoxic drugs – aminoglycosides, loop diuretics, cisplatin, among others[6] .

Given the diversity of pathologies that may result in deafness, determining the etiology of hearing loss is not an easy task. In spite of the technological advancements, a great part of the cases are still from unknown etiologies. The lack of a longer medical follow-up of these children and of a flowchart about how to proceed in etiological investigation, as well as the fact that some tests, such as image and genetic tests are expensive, may account for the difficulty in establishing the etiology of deafness[7].

It is extremely important to know the etiology of hearing impairment and its occurrence in the population, since many causes may be avoided through Public Health measures. Furthermore, knowing the etiology my aid families and the hearing impaired individuals, as well as the doctor in assessing the prognosis of the hearing loss, and the speech-Language Pathologist and Audiologist in planning auditory (re) habilitation[8].

The purpose of this study was to describe the results of the etiological investigation of hearing loss conducted in n identified in a UNHS program.

METHODS

This study was approved by the research ethics committee under number 742/2001. All parents and/or caregivers of the involved subjects agreed to their participation upon signing the free consent term. This study is descriptive, cross-sectional and prospective. All of the newborns diagnosed with sensorineural hearing loss identified in a UNHS program conducted in a public hospital in the city of Jundiaí – São Paulo, in the period between August 2003 and December 2006 were included in the study. The children who had unsatisfactory result (fail) in the UNHS and that for any reason did not conclude the audiologic diagnosis process were excluded from the sample.

The method adopted to conduct the UNHS was the recording of the transient evoked otoacoustic emissions combined with the search for the cochleopalpebral reflex. The equipment used to conduct the otoacoustic emissions was the *Echocheck Hand Held ILo OAE Screener* by *Otodynamics*. The search for the cochleopalpebral reflex was performed using the musical instrument *agogo* bell, played in strong intensity.

When the newborn had an unsatisfactory (fail) result in the UNHS, that is, did not obtain the expected responses in any one of the two adopted screening procedures, he was referred for reevaluation in the institution responsible for the conduction of the UNHS. If the unsatisfactory result (fail) persisted, the child was referred for diagnosis, also performed in the institution.

The diagnosis process involved otorhinolaryngological evaluation, the observation of the auditory behavior of the children, acoustic immitance measures conducted using an AZ-7 Interacoustics immitanciometer, the recording of the transient and distortion product evoked otoacoustic emissions using an *ILO 292 DP Echoport, Otodynamics* equipment, as well as the recording of air and bone-conducted brainstem evoked auditory potential using a *Navigator Pro*, *Biologic Systems Corp* equipment.

As newborn's response to behavioral audiometry is not yet reliable, the classification of the severity of haring loss followed Silman and Silvermam's classification[9], but for this the minimum levels of electrophysiological response were used.

The otorhinolaryngological evaluation included a detailed anamnesis that concentrated mainly on the data concerning the risk indicators for hearing loss (RIHL) according to the Joint Committee on Infant Hearing (JCIH)[10]. Even though isolated prematurity is not considered a RIHL by the JCIH[10], an option was made to consider this indicator, since some studies have related prematurity to hearing loss[11,12].

When the presence of hearing impairment was confirmed in the diagnosis process, auditory (re) habilitation began, alongside the etiological investigation. The patients were submitted to laboratorial and image tests. Among the laboratorial exams, serologies for toxoplasmosis, rubella, cytomegalovirus, herpes, syphilis and HIV were conducted. The

image study performed was a tomography of ears and mastoids.

The collection of blood for the genetic study was made in the hospital maternity ward, as it was collected for the neonatal screening tests for congenital and infectious diseases. The genetic exams were conducted at the Genetic Engineering and Molecular Biology Center (CBMEG) of the State University of Campinas (UNICAMP).

Initially, the genetic study investigated the presence of the 35delG mutation in gene *GJB2*. Mutations in gene *GJB2* constitute the main cause of genetic deafness of recessive autosomal inheritance, and 35delG is the most common mutation in many ethnical groups. In case of absence of the 35delG mutation, gene GJB2 was completely sequenced, and mitochondrial mutations A1555G e A827G in gene *MTRNR1* were investigated using molecular biology techniques.

In regard to the etiological investigation, the possible etiologies were classified as prenatal, perinatal or postnatal[6].

The observed data were analyzed descriptively. The significance level was defined as 0.05 (5%) and the confidence intervals were built with 95% of statistical confidence.

RESULTS

In the period from August 2003 to December 2006, of the 8974 newborns submitted to the UNHS, 156 156 (1.41%) were referred for audiologic diagnosis. Of these, 90 (57.7%) were diagnosed with normal hearing or conductive hearing loss; 49 (31.4%), for non-compliance reasons, did not conclude the process of diagnosis, and 17 were diagnosed with hearing impairment (10.9%). Of the total number of subjects identified with hearing loss, 12 were males (70.6%) and 05 (29.4%) females.

Most of the children diagnosed with hearing impairment (70.6%) had at least one RIHL, while only 05 (29.4%) had no RIHL (p=0.016).

The distribution of the relative frequency of the RIHL was studied. It should be noted that several children had more than one RIHL and that the percent was calculated for a total of 12 children, those who had RIHL (Table 1).

Table 1 – Distribution of the risk indicators for hearing impairment

Risk Indicator	N	%
Craniofacial Anomaly	1	8.3%
Anoxia	3	25.0%
Family History	3	25.0%
Cytomegalovirus	1	8.3%
Consanguinity	3	25.0%
Hyperbilirubinemia	1	8.3%
Ototoxics	1	8.3%
Prematurity	4	33.3%
Syphilis	1	8.3%
ICU	7	58.3%

Distribution of relative frequency

The most frequent RIHL was a stay in the neonatal ICU for more than 48 hours, observed in 7 cases in this study (58.3%).

In regard to hearing thresholds, profound hearing loss was most frequently observed in the studied subjects (47.1%) (Table 2). Since the degree of hearing loss may vary among the ears of the same subject, both ears were considered, and thus the percentages were calculated considering a total number of 34.

Table 2 – Distribution of the degree of hearing loss

Degree of loss	N	%
normal	3	8.8%
mild	2	5.9%
moderate	5	14.7%
severe	8	23.5%
profound	16	47.1%

Distribution of relative frequency

As far as the etiological investigation of hearing loss, after the battery of conducted tests, 64.7% of the studied children had prenatal causes as a possible etiology, 29.4% had perinatal causes and 5.9% or, one child, had unknown etiology. Among prenatal causes, 36.4% were of confirmed genetic origin and 36.4% had presumed hereditary etiology. The presence of congenital infections was confirmed in 18.2% of the cases and one child (9%) had a craniofacial anomaly as presumed etiology.

The results of the etiological investigation and the degree of hearing loss observed for each subject are shown in Table 3.

DISCUSSION

The higher occurrence of deafness in male children observed in this study (70.6%) is in agreement with national and international literature, that shows that hearing impairment is more frequent among males when compared to females[13,14].

A great part of the newborns diagnosed with hearing impairment (70.6%) had at least one RIHL. The RIHL that were most frequently observed in this study were staying in the neonatal ICU for longer than 48 hours (58.3%), followed by prematurity (33.3%). The differences in classification of the RIHL among the studies make it difficult to establish direct comparisons. However many studies show a high occurrence of the RIHL related to an ICU stay in their results[15,16]. This finding is expected as it is estimated that, in Brazil, three to four children for every 1000 are born deaf, a number that increases to two to four children in every 100 newborns when they stay in the Intensive Care Unit (ICU)[4].

Prematurity is a RIHL that usually appears associated to low birth weight, making it difficult to completely separate both RIHL. Premature newborns usually have low birth weight in addition to several other complications that may result in hearing loss[16,17].

The highest occurrence of etiologies observed in this study was of prenatal origin (64.7%). There were no cases of postnatal etiologies observed, which is expected, given that the studied subjects were newborns and young infants. The presence of congenital infections was confirmed in two patients (18.2%) where one was a case of syphilis and the other a case of cytomegalovirus.

Billings[7] observed the occurrence of 1.4% of congenital infections in 211 patients and compares this finding to other studies that had obtained indexes varying from 16.2 to 18.1%. Butugan[17] describes an occurrence of 5.84% of rubella during pregnancy, 1.95% of toxoplasmosis and 0.65% of cytomegalovirus.

In a prospective study with 14.021 newborns in the United States, 74 patients had congenital cytomegalovirus. Of these, 22% had hearing loss[18].

The results of the present study revealed only two cases of congenital infections, in agreement with the findings of a systematic review that shows that congenital infections are decreasing in more recent studies, possibly because the prenatal care concerning these diseases has become more effective[19]. While Brazilian studies[14,20] still report a great number of cases of deafness due to rubella, the present study did not find any cases of congenital rubella.

Among the prenatal causes observed, 36.4% were of confirmed genetic origin and 36.4% had presumed hereditary etiology. Of the cases with presumed hereditary etiology, three patients had Family history of hearing loss and consanguinity was present in one of the cases. Consanguinity is considered a hereditarity[4]. Literature shows variations ranging from 1.95 to 33% of occurrence of hereditary deafness[7,17,21,22]. There are reports of 70% of occurrence in the rural area of Pakistan, or even 44% in Saudi Arabia, and it should be taken into account that in these countries, consanguinity is mainly related to cultural issues of these societies[22].

Hereditary deafness is classified according to the occurrence of clinical findings as syndromic and non-syndromic. In the present study, an occurrence of 36.4% of non-syndromic cases was verified. Mustafá et al.[23] report that about 60% of the cases of sensorineural hearing loss may be attributed

Table 3 – Probable etiology and degree of hearing loss observed in the studied subjects

Subject	Probable etiology	Degree of hearing loss
1	Prenatal cause (consanguinity)	**RE:** severe **LE:** profound
2	Perinatal cause (ICU and anoxia)	**RE:** severe **LE:** moderate
3	Prenatal cause (genetic: homozygosis 35delG)	**RE:** profound **LE:** profound
4	Perinatal cause (ICU, prematurity and anoxia)	**RE:** severe **LE:** moderate
5	Perinatal cause (ICU) mutation A827G	**RE:** normal **LE:** moderate
6	Perinatal cause (ICU, prematurity and anoxia)	**RE:** profound **LE:** profound
7	Prenatal cause (family history)	**RE:** profound **LE:** profound
8	Prenatal cause (genetic: homozygosis 35delG)	**RE:** profound **LE:** profound
9	Prenatal cause (syphilis)	**RE:** mild **LE:** moderate
10	Prenatal cause (craniofacial anomaly)	**RE:** mild **LE:** moderate
11	Prenatal cause (cytomegalovirus) mutation A827G	**RE:** profound **LE:** profound
12	Prenatal cause (family history)	**RE:** severe **LE:** normal
13	Prenatal cause (genetic: homozygosis 35delG)	**RE:** profound **LE:** profound
14	Prenatal cause (family history) mutation A827G	**RE:** severe **LE:** severe
15	Perinatal cause (ICU, prematurity, hyperbilirubinemia)	**RE:** severe **LE:** severe
16	Unknown	**RE:** normal **LE:** profound
17	Prenatal cause (genetic: homozygosis 35delG)	**RE:** profound **LE:** profound

Key: ICU – Intensive Care Unit; RE – right ear; LE – left ear.

to genetic factors, and the remaining 40% have several different etiologies. Among genetic causes, the syndromic hereditary forms account for 30% of the cases of hearing impairment in children, and the non-syndromic forms are the most prevalent in the hearing impaired population, present in about 70% of the cases.

In this study, all cases with confirmed genetic etiology had the 35delG mutation, in the *GJB2* gene, in homozygosis. This mutation is the main responsible for non-syndromic genetic hearing loss. This type of mutation is usually associated with severe/profound hearing loss, which may be observed in all the cases with this etiology[14,18].

A possible explanation for a great number of homozygotic patients for the 35delG deletion is the ethnic contribution of the studied population that is mainly of Italian descent. Mutations in the *GJB2* occur very frequently in this ethnic group[18,19].

The mitochondrial mutation A827G in gene *MTRNR1* was observed in three cases. This mutation is related to the susceptibility of hearing loss associated to the use of ototoxic medication. In these three cases, however, there was no history of

exposure to aminoglycosides. The A827G mutation is apparently frequent in Brazil, and there are not many studies that mention its association to aminoglycosides, resulting in hearing loss[20]. The A827G alteration is possibly a neutral polymorphism, and would thus not be responsible for hearing loss in these cases.

Still among the possible etiologies classified as prenatal, one subject (9%) had craniofacial anomaly. Literature reports a variation from 3.25% to 7.0% of hearing loss resulting from this etiology[7,22,23].

Among the studied children, 29.4% had perinatal causes as the possible etiology. Perinatal causes are extremely common in this population. Some of the causes are neonatal anoxia, prematurity, hyperbilirubinemia, low birth weight, ventricular hemorrhage and long ICU stays [24,25]. A Brazilian study has shown that some perinatal factors such as low birth weight, anoxia, hyperbilirubinemia and mechanical ventilation were significant for hearing loss[26].

Literature shows an occurrence of 9.6 to 14% of perinatal causes as probable etiologies for hearing impairment[19,24,25].

The high occurrence of perinatal causes in this study is possibly due to the fact that the hospital where the UNHS was conducted is a reference in the care of high-risk newborns for other cities that are not equipped with a NICU. Thus, the maternity ward of the hospital receives the most complex cases in its area.

Among the studied children, one (5.9%) remained with an unknown etiology. The undetermined causes of deafness are still quite frequent in studies. Literature shows a variation ranging from 15% to 40%[5,7,17,19,24,25,27-29].

A possible explanation for such a low number of unknown etiologies lies in the fact that all examinations could be performed, including the genetic study. The decrease in unknown causes, mainly in more recent studies derives from the increase in the use of the molecular genetic tests that are currently available[19,25].

It should also be considered that, among the cases with unknown etiologies, there may be cases of underdiagnosed congenital infections or more rare genetic disorders that are still not identified by molecular tests[25].

Faced with the difficulties that are frequently found in the field of child audiologic diagnosis, the knowledge brought by this study may contribute to a better understanding of the etiologies of deafness in Brazil, so that prevention measures may be designed based on its results.

CONCLUSION

A total of 17 subjects were identified with hearing impairment in the period that the study was conducted. In these the highest occurrence of etiologies was those of prenatal origin, followed by those of perinatal origin and one case remains with an unknown etiology.

Among the prenatal causes, 36.4% were of confirmed genetic origin and 36.4% had presumed hereditary etiology. The presence of congenital infections was confirmed in 18.2% of the cases and one subject (9%) had craniofacial anomaly as the probable etiology.

RESUMO

Objetivo: descrever os resultados da investigação etiológica da deficiência auditiva realizada em neonatos rastreados em um programa de triagem auditiva neonatal universal. **Métodos:** estudo descritivo, transversal e prospectivo. Foram incluídos no estudo todos os neonatos diagnosticados com deficiência auditiva identificados em um programa de triagem auditiva neonatal universal no período de agosto de 2003 a dezembro de 2006. A provável etiologia da deficiência auditiva foi determinada após anamnese detalhada realizada pelo médico otorrinolaringologista; pesquisa das sorologias para toxoplasmose, rubéola, citomegalovírus, herpes, sífilis e HIV; tomografia dos ossos temporais e exames genéticos. **Resultados**: foram diagnosticados 17 sujeitos com deficiência auditiva no período estudado. 64.7% dos casos estudados apresentaram como provável etiologia causas pré-natais, 29.4% causas peri-natais e um sujeito (5,9%) apresentou etiologia desconhecida. Das causas pré-natais, 36.4% tiveram origem genética confirmada e 36.4% etiologia presumida de hereditariedade. Foi confirmada a presença de infecções congênitas em 18.2% dos casos e um sujeito (9%) apresentou anomalia craniofacial como provável etiologia. O grau de perda auditiva mais frequente observado nos sujeitos estudados foi o profundo (47,1%). **Conclusão:** a maior ocorrência de etiologias observada neste estudo foram as de origem pré-natal, seguida das de origem peri-natal.

DESCRITORES: Perda Auditiva; /etiologia; Triagem

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