Audiologic findings in infants with Down syndrome

Achados audiológicos de lactentes com síndrome de Down

Hallazgos audiológicos de lactantes con síndrome de Down

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Abstract

Introduction: *Down syndrome involves morphological characteristic that predispose the arise of hearing alteration. It is crucial for the overall development of the individual an early identification of hearing alteration, and Newborn Hearing Screening would be the first step to early identifying any alteration. However, there are few studies that describe the hearing of this population in the first months of life.* ***Objectives:*** *To evaluate the audiological findings in infants with Down syndrome by Newborn Hearing Screening and audiological evaluation, considering the variables gender, risk factors and gestational age.* ***Methods:*** *Research of experimental basis, descriptive, cross-cut, consisting of children with Down syndrome, who remained in the ICU and/or intermediate care. Hearing screening was performed and, independent of the results, children were referred for audiological diagnosis.* ***Results:***

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In the Newborn Hearing Screening 71,4% of the children failed, there wasn't significant connection of faults with the correlated variables. From the analysis of the auditory test was found normal bilateral hearing in 42,85%, and conductive hearing loss in 57,14%. ***Conclusion:*** *The most of children with Down syndrome fail in auditory screening and in more than half of children, the hearing loss was found, and conductive hearing loss was the most frequent.*

Keywords: *Hearing; Down Syndrome; Triage, Auditory Pathways, Electrophysiology.*

Resumo

Introdução: *A síndrome de Down envolve sinais morfológicos que predispõem o surgimento de alterações auditivas. É fundamental para o desenvolvimento global do indivíduo a identificação precoce de alterações auditivas e a Triagem Auditiva Neonatal é o primeiro passo para que ela ocorra. No entanto, são escassos os estudos que descrevem a audição desta população ainda nos primeiros meses de vida.* ***Objetivo:*** *Avaliar os achados audiológicos de lactentes com síndrome de Down na triagem auditiva neonatal e na avaliação audiológica, considerando-se as variáveis: sexo, indicadores de risco e idade gestacional.* ***Métodos:*** *Pesquisa do tipo experimental, descritiva, transversal, da qual participaram lactentes com síndrome de Down, que permaneceram na UTI e/ou Cuidados Intermediários. Foi realizada triagem auditiva neonatal e, independentemente dos resultados, os lactentes foram encaminhados para investigação audiológica.* ***Resultados:*** *Na triagem auditiva neonatal 71,4% dos lactentes falharam, não houve relação de significância das falhas com as variáveis correlacionadas. A partir da análise conjunta dos testes auditivos verificou-se audição normal bilateral em 42,85% e perda auditiva condutiva em 57,14%.* ***Conclusão****: A maioria dos lactentes com síndrome de Down falha na triagem auditiva e em mais da metade dos lactentes, a alteração auditiva foi encontrada e/ou confirmada, sendo a perda auditiva condutiva a mais frequente.*

Palavras-chave: *Audição; Síndrome de Down; Triagem; Vias Auditivas, Eletrofisiologia.*

Resumen

Introducción: *La Síndrome de Down implica signos morfológicos que predisponen la aparición alteraciónes auditivas. Es crucial para el desarrollo general del individuo la identificación temprana de alteraciónes auditivas y la tría auditiva neonatal es el primer paso que ello ocurra. Sin embargo, son escasos los estudios que describen la audición de esta población dentro de los primeros meses de vida.* ***Objetivo:*** *Evaluar los resultados audiológicos de lactantes con Síndrome de Down en la tría auditiva neonatal y en la evaluación audiológica, teniendo en cuenta las variables sexo, indicadores de riesgo y edad gestacional.* ***Métodos:*** *Estudio de tipo experimental, descriptivo, transversal, del que participaron lactantes con síndrome de Down, que permanecieron en UCI y/o unidad de cuidados intermedios. Se realizó tría auditiva neonatal y independientemente de los resultados, los lactantes fueron encaminados para investigación audiológica.* ***Resultados:*** *En la tría auditiva neonatal 71,4% de los lactentes fallaron, no hubo una relación significativa de fallas con las variables correlacionadas. A partir del análisis conjunto de las pruebas auditivas se encontró audición bilateral normal en el 42,85% y pérdida auditiva conductiva en el 57,14%.* ***Conclusión:*** *La mayoría de los lactantes con sindrome de Down falla en la tría auditiva, y en más de la mitad de los lactantes, la alteración auditiva fue encontrada y/o confirmada, siendo la pérdida auditiva conductiva la más frecuentes.*

Palabras claves: *Audición; Síndrome de Down; Tríaje; Vias auditivas; Electrofisiología.*

Introduction

The anatomical and functional integrity of the peripheral and central auditory system, along with proper stimulation of hearing experiences are crucial to the acquisition and typical language development[1].

Hearing alteration can be identified early in life through the Newborn Hearing Screening. Early identification is essential for the diagnosis and rapid intervention with positive effects on the individual's development.

The first recommendations for the implementation of newborn hearing screening were performed in 1971 in the United States, through the Joint Committee on Infant Hearing (JCIH). In Brazil, the Support Group for Universal Newborn Hearing Screening (GATANU, in Portuguese) was created in 1998, in order to raise awareness of the importance of Universal Auditory Screening. Currently, Federal Law No. 12,303, enacted on August 2nd, 2010, made it mandatory the exam called Evoked Otoacoustic Emissions, free of charges, in all children born on the premises of hospitals and maternities[2,3].

The *JCIH* (2007) and the Multidisciplinary Committee on Hearing Health - COMUSA (2010) suggest several risk factors for congenital hearing loss or late-onset. Among them are the genetic syndromes that usually express hearing loss, including the Down syndrome[2,3] Therefore, infants with Down syndrome need to be submitted to hearing screening and monitoring of the development of hearing and language.

Down syndrome is characterized by the presence of trisomy 21. It may take the form of simple trisomy (95%), translocation (3-4%) or mosaic (1-2%), with an incidence of live births from 1 to each 600 births[4].

Infants with Down syndrome have morphological characteristics that predispose the emergence of conductive hearing loss. They have presented stenosis of the external auditory meatus, short palate, narrowing of the nasopharynx and oropharynx, delayed development of the immune system, and malformations of the Eustachian tube. It is more cylindrical and narrow and may have an abnormal insertion in the nasopharynx. Furthermore, it is also associated with general hypotonia and dysfunction of the tensor muscle of the soft palate which is responsible for opening and closing the Eustachian tube, and together with a lower density cartilage it can result in tube collapse, occurring negative pressure in the area, resulting in development of the fluid in the middle ear and chronic otitis media[5].

The occurrence of otitis media in children is worrying, because even if the hearing loss is mild, with a floating feature, it may interfere with sound stimulation of the central auditory nervous system. In addition, the fluid in the middle ear can cause noise in the cochlea, distorting the sound perception and consequently hindering the sound perception[6].

Hearing loss can compromise the development of listening skills in any child. Children with Down syndrome with intellectual disability due to genetic may have a worsening of language development and their oral expression if they have hearing loss associated with cognitive impairment[4,7].

The literature is consistent as the high incidence of unilateral or bilateral hearing loss in children with Down syndrome, which can vary between 60-90%[8,9]. However, there are few studies that describe the hearing of this population, even in the first months of life. Given the above, the objective of this study was to analyze the audiological findings in infants with Down syndrome found in newborn hearing screening and in audiological diagnosis, considering gender, risk factors and gestational age variables.

Material and methods

This study was approved by the Ethics Research Committee of Unicamp - Campinas Campus, number 112163/2012. The study is cross-sectional, experimental, and descriptive.

The inclusion criteria and the selection of the sample were: infants with Down syndrome born in 2012 in the Center of Integral Attention to Women's Health – CAISM, those that remained in the Intensive Care Unit and/or Intermediate Care, when the responsible has agreed to participate and signed the Free and Clarified Consent Term (FCCT). All infants with these criteria were included in the study, with no exclusion of any subject.

Initially, the medical records of the infants were consulted to confirm the diagnosis, identification data record, birth conditions and risk factors for hearing loss present in their clinical history.

Then the hearing screening was applied at hospital discharge period; the shortest period of stay was three days and the longest was 20 days,

using the Brainstem Auditory Evoked Response - BAER test, with the Madsen Accuscreen-Otometrics equipment. The acoustic stimulus was the click type. Electrodes were glued to the skin and headphones were inserted in the infant's external auditory meatus. It was considered that the infant PASSED the test when presenting response to the 35 dB NA stimulus bilaterally. The 40 dB and 45 dB NA intensities were studied.

Subsequently, all screened infants, both those who have passed or failed the test were scheduled for audiological evaluation at the Laboratory of Audiology of Center for Studies and Research in Rehabilitation Prof. Gabriel Porto, School of Medical Sciences, University of Campinas (CEPRE/FCM/UNICAMP). This conduct was guided by the high failure of hearing screening rate, presence of risk factors for hearing loss of late and/or progressive onset, as well as frequent conductive hearing loss in Down syndrome due to its morphology, justifying the concern in monitoring aspects of the infant's middle ear.

The assessment after hospital discharge was established by anamnesis, Transient Otoacoustic Emissions (TOAE), Imitanciometry and BAEP mode diagnosis. Infants' age in the evaluation varied from one to 10 months, some rescheduling was needed because of the difficulty in completing the assessment at the scheduled day, because the infant should be in natural sleep at the time of evaluation.

Anamnesis was made of questions to parents or caregivers about the family history of hearing loss, concern with the development of the infant, position and feeding type, presence or absence of gastroesophageal reflux and otalgy. In addition, reaction to sounds such as attention and location to the sound source was researched.

Surveys of transient otoacoustic emissions were made with the ILO 292 USB Otodynamics equipment, in order to assess cochlear function, specifically the outer hair cells. The criteria adopted for TOAE normality had overall reproducibility higher or equal to 50%, wave stability higher or equal to 70%, signal/noise ratio by frequency band higher or equal to 5 dB to the 1000 Hz frequency and higher or equal to 7 dB for the other frequencies, with obligatory in at least three frequency bands[10].

Tympanometry, through the use of 1000 Hz probe tone, Impedance Audiometer AT235h equipment, from Interacoustics, was performed for evaluation of the tympanic membrane mobility and functional conditions of the middle ear. The normal curve criterion was type A, with pressure from -100 to + 50 BP and volume from 0.33 to 0.80 Ml[11].

Brainstem Auditory Evoked Response-BAER, Eclipse Interacoustics equipment, was used to stimulate the click type. During the test the infants were in natural sleep. To consider normal examination the integrity of the auditory pathway was evaluated by collecting two responses to 80 dB HL, verifying the reproducibility between the traces and the absolute latencies of waves I, III and V, and interpeaks I-III, I-V and III-V. The electrophysiologic threshold was also investigated by stimulating at 60, 40 and 30 dB HL. The study using the same equipment for standardization of full-term and premature[12] infants' responses was adopted as a reference for normality criteria.

From analysis combined of the tests, the child's hearing was classified in normal or hearing loss. Infants with hearing loss were referred to the ENT examination at the Clinical Hospital of Unicamp.

All infants, for presenting Risk Indicators for the development of late hearing loss, were referred to the Hearing Monitoring at CEPRE (Center for Studies and Research in Rehabilitation) for monitoring up to two years old.

The results were analyzed using descriptive analysis of frequency and measures of central tendency and dispersion (mean, median and standard deviation) and inferential analysis ($p<0.05$).

Results

The results are presented in the order of hearing tests performance. Newborn hearing screening was performed before hospital discharge of infants, in this study before the first month of life, and other tests were applied as outpatient until the first year of life.

In 2012, 477 neonates received assistance in ICU/Intermediate Care for more than 48 hours. Of this, 362 had hearing screening, 54 failed in at least one ear. Of the infants who remained in the ICU/Intermediate Care in that year, seven were diagnosed with Down syndrome (1.46%), which corresponds to the incidence of 1:68 hospitalized, three (42.8%) were female and four (57.2%) were male. The mean maternal age, gestational age time (Capurro), weight and length of hospitalization of these children were, respectively: 38 years; 37.4 ±

2 weeks (featuring 6 term births and 1 preterm), 3085g and 7.1 ± 6.8 days.

Besides Down syndrome, it was observed that the infants also had other risk factors for hearing loss present in their clinical history, as follows: ICU stay for more than 5 days, the use of mechanical ventilation, prematurity, use of ototoxic medication and consanguinity. The predominant risk factor in five (71.4%) infants was the use of mechanical ventilation.

The description of infants with Down syndrome who have passed or failed the newborn hearing screening is exposed in Table 1.

Table 1. Infants with Down Syndrome, in relation to the presence or absence of response in newborn hearing screening, considering gender, gestational age and number of risk indicators variables

	HEARING SCREENING				
	Passed		Failed		p-value
	N	(%)	N	(%)	
GENDER					1.00
Male	1	25.0	3	75.0	
Female	1	33.3	2	66.7	
RI Number					1.00
0	1	50.0	1	50.0	
1	0	0.0	1	100.0	
2	1	50.0	1	50.0	
3	0	0.0	2	100.0	
GA					1.00
PTNB	0	0.0	1	100.0	
FNB	2	33.3	4	66.7	

Risk indicators (RI); Gestational age (GA); Preterm newborn (PTNB); Full-term newborn (FNB); Fisher's exact test

In the anamnesis, prior to the audiological evaluation, in the questions related to hearing loss in the family history, six (85.7%) parents and/or caregivers reported no history, and one (14.3%) could not inform, as the infant was in a shelter. Regarding the concern about the development of the child in matters relating to hearing and language, two (28.6%) parents and/or caregivers reported concern in both aspects, one (14.3%) in relation to language and four (57.1%) had no concerns about it.

The type of feeding reported by three (42.8%) parents and/or caregivers was natural, three (42.8%) reported bottle feeding and one (14.3%) by both methods. Sitting, as feeding position, was reported by all parents and/or caregivers. The gastroesophageal reflux was reported by four (57.1%) parents and/or caregivers seven (100%) reported that the infant has never shown any reaction to suggest otalgy to the valuation date after the first month of life.

Regarding the reaction to sound, six (85.7%) family members and/or caregivers reported that the infant wakes up in the presence of high noise and seven (100%) reported that there is no crying in the presence of loud noises. Regarding the sound location, six (85.7%) had observed that the infant seeks sound that is out of their sight and seven (100%) reported attention to the voice.

Tables 2 and 3 show the results obtained after hospital discharge, hearing assessment, Transient Otoacoustic Emissions tests and Imitanciometry, respectively.

Table 2. Infants with Down Syndrome, in relation to the presence or absence of transient otoacoustic emissions, considering gender, gestational age and number of risk indicators variables

	TOAE				
	Presence		Absence		
	N	(%)	N	(%)*	p-value
GENDER					1.00
Male	1	25.0	3	75.0	
Female	0	0.0	3	100.0	
RI Number					1.00
0	1	50.0	1	50.0	
1	0	0.0	1	100.0	
2	0	0.0	2	100.0	
3	0	0.0	2	100.0	
GA					1.00
PTNB	0	0.0	1	100.0	
FNB	1	16.7	5	83.3	

*= absence rate; Transient Otoacoustic Emissions (TOAE); Risk indicators (RI); Gestational age (GA); Preterm newborn (PTNB); Full-term newborn (FNB); Fisher's exact test

Table 3. Infants with Down Syndrome, in relation to the tympanometry curve, considering gender, gestational age and number of risk indicators variables

	Curve type A		Curve type B		
	N	(%)	N	(%)	p-value
GENDER					1.00
Male	2	50.0	2	50.0	
Female	1	33.3	2	66.7	
RI Number					0.43
0	2	100.0	0	0.0	
1	0	0.0	1	100.0	
2	0	0.0	2	100.0	
3	1	50.0	1	50.0	
GA					1.00
PTNB	0	0.0	1	100.0	
FNB	3	50.0	3	50.0	

Risk indicators (RI); Gestational age (GA); Preterm newborn (PTNB); Full-term newborn (FNB); Fisher's exact test

In BAER, only 12 ears were considered for statistical analysis, corresponding to the bilateral analysis of six infants, excluding one infant. The exclusion in this test occurred because the evaluation was done in only one ear. In preparation of the contralateral ear, the infant woke up and there was no return to the test completion.

There was integrity of the auditory pathway to the brainstem, with the stimulus of 80dB in all evaluated infants. Table 4 shows the findings of the latencies and inter-latencies. The mean found for electrophysiologic thresholds were 35 dB HL in the right ear and 36.6 dB HL in the left ear.

Table 4. Infants with Down Syndrome, in relation to results of baer obtained from the bilateral evaluation (n = 12)

	Right ear			Left ear			
Measure evaluated	**Mean (ms)**	**Standard deviation**	**Median**	**Mean (ms)**	**Standard deviation**	**Median**	**p-value**
Wave I - 80dBHL	1.23	0.12	1.22	1.63	0.66	1.27	0.36
Wave III - 80dBHL	3.91	0.24	3.92	3.99	0.45	3.90	0.55
Wave V - 80dBHL	5.92	0.24	5.90	5.89	0.50	5.72	0.54
Wave V - 60dBHL	6.41	0.34	6.50	6.52	0.55	6.33	0.63
Wave V - 40dBHL	7.50	0.27	7.40	7.31	0.67	7.23	0.50
Wave V - 30dBHL	8.26	0.17	8.30	7.58	0.33	7.70	0.14
Interpeak I-III	2.44	0.26	2.54	2.18	0.38	2.17	0.32
Interpeak III-V	2.01	0.30	2.00	1.91	0.09	1.90	0.71
Interpeak V-I	4.45	0.37	4.52	4.08	0.32	4.12	0.18

Student's t test paired

From the combined analysis of the applied hearing tests, it was found that three (42.8%) of the infants had normal bilateral hearing and four (57.1%) had conductive hearing loss, at least unilateral (Table 5).

Speech therapy conduct after evaluating these children was forwarded to the ENT clinic of all children who had some type of hearing loss and forwarded to the semiannual hearing monitoring up to their 2nd year of life, held in CEPRE.

Table 5. Infants with Down Syndrome, in relation to newborn hearing screening and audiologic diagnosis results

Neonate	**Gender**	**RI**	**RE**	**LE**	**RE**	**LE**	**RE**	**LE**	**RE**	**LE**	**Type of Hearing Loss**
			HS		**TOAE**		**TC**		**ET**		
1	F	1	P	P	AS	AS	B	B	----	40	Conductive
2	F	2	F	F	AS	AS	AA	AA	30	30	Normal
3	M	3	F	F	AS	AS	B	B	40	40	Conductive
4	M	0	F	P	AS	PS	AA	AA	30	30	Normal
5	M	0	P	P	AS	AS	AA	AA	30	30	Normal
6	M	1	F	F	AS	AS	B	B	40	50	Conductive
7	F	1	F	F	AS	AS	B	B	40	40	Conductive

Risk indicators (RI); Hearing screening (HS); Transient Otoacoustic Emissions (TOAE); Tympanometry Curve (TC); electrophysiologic threshold (ET); Passed (P); Failed (F); Absence (AS); Present (PS)

Discussion

The incidence of infants with Down syndrome in the ICU and/or Intermediate Care, in the twelve--month period described in this study was high (1:68). There is 1 case for every 600/800 births in Brazil, with an average of 8,000 births per year[13]. However, there are no records of incidence of this population in the ICU. The high incidence of infants with Down syndrome present in this study may be due to the fact that the sample is originated from a hospital considered the largest hospital for women's health in the state of São Paulo, reference to pregnancy or risk deliveries to the metropolitan region of Campinas[14], thus presenting appropriate structure for the care of women in advanced gestational age and with Down syndrome diagnosed even in the prenatal period.

In addition to having Down syndrome, five infants (71.4%) had other risk indicators for hearing loss, reinforcing the need for auditory monitoring of this population. In the sample, the predominant risk indicator was the use of mechanical ventila-

tion. In cases of Down syndrome, the prevalence of abnormal respiration can reach to 80%[15], suggesting that the use of ventilation is sometimes necessary to these children. Respiratory changes of infants in this sample may also have contributed to the high incidence of ICU/Intermediate Care hospitalization.

The newborn hearing screening, which was the first procedure performed in this study, was applied in all infants with Down syndrome. Of these infants, five (71.4%) failed to at least one ear. There was no significant relation between these findings and the correlated variables: gender, risk indicators and gestational age (Table 1).

In the same period, 355 infants without Down syndrome underwent newborn hearing screening and at least one ear failed among 47 ears, corresponding to 13.23%, indicating a percentage of greater failure in infants with Down syndrome.

The data obtained in the anamnesis, conducted after hospital discharge, in the same day scheduled for hearing evaluation, showed that parents and/or caregivers showed reduced concern with aspects related to the development of hearing and language, which may be related to infants' reaction to wake up and/or cry in the presence of loud noises, to seek sound out of sight and to show attention to the voice, as well as unawareness of the damage that the hearing may result in child development, especially in the development of oral language.

Questions about feeding position were considered necessary in the anamnesis because of the importance of knowing and conduct guidelines regarding the relation of anatomy of the Eustachian tube, breastfeeding and otitis. However, all caregivers have held breastfeeding in the proper posture.

Some parents and/or caregivers had concerns with aspects that involve risk of death, such as heart, respiratory and digestive. It should be noted that this phase is also a moment to accept the child with Down syndrome, with feelings varying from doubts, uncertainties and insecurities. Thus, the conditions of hearing loss may go unnoticed by caregivers and infants are not evaluated early. In a previous study with 106 children with Down syndrome, aged between 3 and 14, whose objective was to evaluate ENT change, a common characteristic of this syndrome, 35.8% of children had never been evaluated by ENT specialist, and among those who had already been evaluated more than half (64.2%) had never been to audiological evaluation[16].

Regarding the hearing, parents and/or caregivers reported that they did not notice behavioral changes such as irritation and/or crying to nurse, related to the occurrence of hearing loss and/or otalgy on their children after hospital discharge period until the return for complete hearing evaluation. However, at the time of completion the evaluation/diagnosis, two children showed discomfort when handling the ear, being recorded type B curve in both cases and they were referred for medical evaluation. The reactions of infants to high intensity sounds, such as waking up in the presence of intense noise, sound location and attention to the voice, observed by parents and/or caregivers in most infants, end up being a picture of auditory responses that may mask mild hearing loss.

Regarding the Transient Otoacoustic Emissions (TOAE), it was absent in six cases (85.7%) (Table 2). Only one infant (14.3%) that failed the hearing screening initially obtained presence of otoacoustic emissions, at three months of age. In two infants (28.6%) the absence of otoacoustic emissions showed false-positive, and these infants were aged between four and nine months. This false-positive occurrence is justified in the literature because the transient otoacoustic emissions may be absent when there is a change of external and middle ear structures, even with normal function of outer hair cells[10]. In Down syndrome, many children have stenosis of the external auditory meatus, which may be one of the reasons that led to TOAE absence even after several attempts to sealing the external auditory meatus with multiple eartips available in the equipment. In addition to these factors that can lead to no record of TOAE, they may indicate a hearing loss, because in ears where hearing thresholds are greater than 30 dB HL, or there is external ear compromise or middle ear, are no longer observed TOAE[17]. Given these findings, the importance of the audiological diagnosis is emphasized based on the agreement between the results of all tests instead of a single procedure.

The infant who had the presence of transient otoacoustic emissions at three months of age, compatible with the other tests in the diagnosis and that initially failed the newborn hearing screening, may suggest that there is a change of conductive matter, caused by impairment of the external and/

or medium ear over the study period, which was solved before the reassessment period[18].

A previous study evaluated 46 children with a mean age of 6, all of them had stenosis of the external auditory meatus in different degrees and impaction of cerumen, and after their removal, only eight children had tympanic membrane with normal appearance, and the study of otoacoustic emissions by distortion product indicated that these were significantly lower compared to the control group[19], which may suggest that the audiological findings of otoacoustic emissions in that population are due to their anatomical characteristics.

In the evaluation of the middle ear through tympanometry, type A curve was found in three (42.8%) and type B curve in four (57.1%) children (Table 3). These findings were performed with 1000 Hz probe. The literature suggests that the 1000 Hz probe has more specificity and sensitivity compared to the 226 Hz probe in a study with children with down syndrome[20].

In our study, the means of the electrophysiological thresholds were 35 dB HL for the right ear and 36.6 dB HL for the left ear. Similar results were found in hearing evaluations of 25 infants with Down syndrome who had an electrophysiologic threshold mean of 37.5 dB HL[21]. Latencies and inter-latencies found in this population (Table 4) had no disagreements of what was expected for children full-term born without the presence of risk indicators found in the literature[12].

Considering the combined analysis of the tests, with Tympanometry and BAER as differential tests to determine the presence or absence of hearing impairment, three infants (42.8%) had normal results in the diagnosis, one of which had passed the newborn hearing screening and two had failed.

In four infants (57.1%) conductive hearing loss was confirmed. This result was consistent with the failure in newborn hearing screening of three infants (42.8%), and incompatible with an infant (14.3%) that initially passed the newborn hearing screening for 35 dB HL stimulus, and later, at 4 months old received absence of transient otoacoustic emissions, type B tympanometry curve and electrophysiologic threshold at 40 dB HL in the diagnosis, suggesting conductive hearing loss and immediate need of medical evaluation. Medical evaluation was performed and there was placement of ventilation tube.

Conductive hearing loss found in this study proved to be incident even in the first months of life in this population, which emphasizes the importance of newborn hearing screening and audiological monitoring since the early identification and early intervention can be decisive factors in the evolution and prognosis of children with impaired hearing; the period between birth to six months of age is the most effective for the stimulation of the brain and formation of auditory pathways[18].

In a retrospective study of ENT manifestations in Down syndrome, two patients, of a total of 11, passed the assessment of otoacoustic emissions. The nine patients that failed were evaluated with BAER and Imitanciometry. In four individuals the electrophysiologic threshold obtained by BAER was high, between 40-60 dB HL, suggestive of conductive hearing loss. In imitanciometry results serous otitis was found in six cases and four patients had mixed hearing loss[7].

A previous study reported that Down syndrome was present in 4 of the 12 conductive cases of high-risk infants who failed the newborn hearing screening. In one of the conductive unilateral cases there was malformation of the outer ear and the external auditory meatus [22].

The results of this study showed that hearing losses were mild, supporting the literature which states that in most cases of hearing loss the cases are mild to moderate, and may have as causes the increase of wax in the external auditory meatus, accumulation of secretion in the middle ear, frequent ear infections and abnormal format of the ossicles located in the middle ear[8,23]. In addition, the authors report that despite being common the existence of chronic otitis media in children, there is not always clinical manifestations, which emphasizes the importance of more detailed examinations and auditory monitoring, since the occurrence of otitis media has consequences in childhood, especially in relation to language acquisition. The main changes referred to in the literature are: the occurrence of phonetic and speech articulation errors, as well as the difficulty in understanding the lecture[24].

The health professionals concern with the otitis media effects justify the need for tympanocentesis surgeries with insertion of ventilation tube, the most common procedure performed in children of various countries[24].

The occurrence of conductive changes in infants of this study and false-positive in TOAE

results reinforce the need for health professionals to clarify caregivers the possibility of occurrence of these failures and the need for diagnosis and systematic monitoring of these children hearing, as well as hearing relations with the development of language.

Conclusion

In this study most infants with Down syndrome failed the hearing screening, and in more than half of infants hearing loss was found and/or confirmed; conductive hearing loss was the most frequent.

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