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Original Article

NEONATAL HEARING SCREENING IN OTORHINOLARYNGOLOGY SERVICE

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ABSTRACT

Introduction: The results of screening programs in hospitals in Brazil studied in isolated studies demonstrate the great difficulty of implementing an effective program that reaches the proposed indicators. **Objectives:** To describe the implementation of the Universal Neonatal Hearing Screening program in the maternity ward and in Otorhinolaryngology service. **Methods:** The risk indicators found, the distribution according to gender, gestational age, age at the time of screening, and the number of failures in the various groups, between July 2014 and December 2015, were detailed. **Results:** Of the 3,155 born alive, 2,224 were screened (70.48% of the total). At baseline, the mean age of the triads was more than double the maximum recommended age (30 days of life). Six months after the introduction of the program, an average age of 20.63 was reached. A 6.3-fold higher probability of failure in children screened before the fifth day was found, and a high abstention rate of patients referred for retest (57%). Of those who attended, 43 failed, giving only 2.41% of failures among the triad. **Conclusion:** The study demonstrated the possibilities and adversities in implementing a TANU program in a service funded by the Brazilian Unified Health System. It was evident after months of the beginning of the program the increase of the index of triads and the improvement in the number of failures.

KEWWORDS: Neonatal screening; Hearing Loss; Risk indicator; Quality indicators in health care; Evaluation of health programs and projects.

INTRODUCTION

The essential characteristic of the human being is language and it is through language that he constitutes himself human. Language is the expression and reception of ideas, knowledge and feelings in a creative, structured, meaningful and interpersonal way. Its purpose is to allow social interaction, expressing what one has in mind, desires and thoughts.^[1]

It is essential that during early childhood, the assessment of auditory maturation and communication markers is carried out. At birth, there are already startled responses to loud sounds, and until the age of three months, the listening child calms down with the sound. Until the age of 8 months you must locate the sound source and start babbling, and until you complete the first year of life you have to produce the first words, which should be at least

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six until the age of 18 months. At the age of two he produces sentences with two words, and at the age of three he is capable of producing sentences. Any loss in any of the markers indicates the need for evaluation by a specialist.

Knowing the etiology of hearing loss allows us to know if something can be done to solve the deficiency or prevent its progression. As with some infectious causes, when treated, they will prevent installation, or at least progression. Knowing the etiology also allows us to understand which modes of communication will be suitable for the child, which may indicate the need to carry out or not the investigation of family members, in addition to contributing to the study of hearing mechanisms.^[2] The drugs used by the mother during pregnancy, and medications administered to the newborn can alter the auditory physiology, causing varying degrees of hearing loss. The best known are the antibiotics of the aminoglycoside class, which alter the mitochondrial mechanism of energy production, in addition to loop diuretics. Furosemide is known to be responsible for temporary and permanent cases of hearing loss. Plasma levels of furosemide greater than 50 mg / L have been associated with sensorineural loss. The incidence of furosemide ototoxicity was estimated at 6% in a small series of patients.^[2]

It is necessary to identify a newborn with hearing impairment in the first month of life, in order to increase the possibility of regeneration of this function. Hearing assessment in neonates should occur even if there are no risk indicators in their history that enhance the probability of this condition having occurred.3 If only those with identified risk factors were assessed, only half of the hearing losses would be diagnosed in time capable of intervention.^[3]

The Joint Committee on Infant Hearing (JCIH) since 2000, and the Brazilian Committee on Hearing Loss in Childhood in 2001, started to recommend hearing screening in the neonatal period, universal, that is, covering all newborns. They consider otoacoustic emissions as the main hearing screening procedure to be used. Children with unsatisfactory results in the screening, must have the confirmation of hearing loss before the third month, and receive the indicated intervention until the sixth month of life.^[4,5]

Since the publication of Law 12,303, of August 2, 2010, it has become mandatory in Brazil to perform OAE free of charge in all hospitals and maternity hospitals in Brazil. The implementation of a TANU program will only make sense if, after its conclusion, the correct diagnosis is reached and the intervention occurs as soon as possible.

This study aims to describe the implementation of the TANU program in the maternity ward and in the service of Otorhinolaryngology. Specific objectives.

1- Assess the rate of live births submitted to TANU.

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2- Characterize the group of newborns (NB) submitted to TANU as to sex, and the presence of IRDA.

3- Check the age in days of the newborn at the time of the initial TANU test.

4- Identify the IRDA present in the RNs.

5- Check the incidence of IRDA in live births.

6- Identify the failure rate in the TANU according to the age of the first test.

7- Identify the retest test evasion rate and the general failure rate in the TANU.

8- Verify compliance with the TANU quality indicators, according to JCIH and NCHAM.

MATERIALS AND METHODS

This work was approved under protocol number 51356415.2.0000.5514.

The data of newborns born alive and the record of TANU data from July 2014 to December 2015 were recorded, for a total of 18 months, after authorization by the clinical director for the authorization term for the use of data from the medical record. Data on a total of 3,155 newborns were assessed.

The OAE research was performed with the EICO SCAN Etymotic Research device, from MAICO® in the distortion product mode, where four frequencies (2kHz, 3kHz, 4kHz and 5kHz) were analyzed. The newborn was examined on the mother's lap in a quiet environment. An olive of a size proportional to the MAE of the RN was introduced. At the end of the exam, the result was duly noted as: "passed" or "did not pass". The criterion used to consider how "passed" was the presence of an answer in at least three frequencies. Presenting less, it was considered as "did not pass". NBs who failed the first exam were reevaluated after 15 days.

The following data were computed for statistical analysis: gender, identification of the IRDA, when present, performance or not of the TANU, age at the time of the first test, attendance to the retest and result of the retest.

The indicators associated with hearing loss were identified as IRDA, identified by the Joint Committee on Infant Hearing (JCIH) and published in its latest 2007 document.^[6]

Hearing Impairment Risk Indicators (Joint Committee on Infant Hearing - 2007)

 Parents' concern about the child's development, hearing, speech or language
Family history of permanent deafness, starting from childhood, being considered as a risk of heredity. Inbreeding cases should be included in this item
Stay in the ICU for more than five days, or the occurrence of any of the following conditions, regardless of the length of stay in the ICU: extracorporeal ventilation; assisted ventilation; exposure to ototoxic drugs such as aminoglycoside antibiotics and / or loop diuretics; hyperbilirubinemia; severe perinatal anoxia; Neonatal Apgar from 0 to 4 in the first minute, or 0 to 6 in the fifth minute; birth weight less than 1,500 grams;

4. Congenital infections (toxoplasmosis, rubella, cytomegalovirus, herpes, syphilis, HIV)

5. Craniofacial anomalies involving ear and temporal bone

6. Genetic syndromes that usually express hearing impairment (such as Waardenburg, Alport, Pendred,				
among others)				
7. Neurodegenerative disorders (Friedreich's ataxia, Charcot-Marie-Tooth syndrome)				
8. Postnatal bacterial or viral infections such as cytomegalovirus, herpes, measles, chickenpox and				
meningitis				
9. Head trauma				
10. Chemotherapy.				

During the study, compliance with the quality indicators recommended by the 2007 JCIH was also verified.

1. Screening of at least 95% of live births, with the aim of reaching 100%;

2. Complete screenings up to the first month of life;

3. Refer a maximum of 4% of screened newborns to diagnosis;

4. Completion of auditory diagnosis in 90% of neonates referred until the third month of life;

5. Start the use of sound amplification in 95% of infants with confirmed bilateral hearing loss, within one month after diagnosis.

RESULTS

The study was made from the beginning of the implementation of the TANU program. 3,155 children were born in the period between July 2014 and December 2015. Of these, 1,646 (51.49%) were male.

During the 18-month period analyzed by the study, 2,224 newborns (70.5%) underwent TANU. In the first month, only 10.9% of live births were screened, with an evident increase in the rate of patients undergoing TANU monthly, shown in the graph in Figure 1.

In the first month, it was only 10.9%, reaching 94.1% just 4 months after the program was implemented, reaching 100% one year after the beginning.

In the months of December 2014 and January 2015, a significant drop in the trend in the number of children screened was observed, but it soon resumed in the subsequent months.

The newborn's age, in days, at the time of screening was studied month by month, and the mean, standard deviation, mode and median are analyzed in Figure 2.

In the beginning, the average, fashion and median remained high. The median and mode in the first two months were 60 days old, but soon decreased, remaining below 30 days old from the sixth month, and remaining at two days old in the last 8 months studied. The standard deviation remained high throughout the study period, showing the age variance of neonates undergoing TANU.

Of the total of 2,224 newborns submitted to TANU, 2,019 (90.78%) did not have IRDA and 205 (9.22%) had some factor, distributed according to Table 1.

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The main risk factor identified was the use of ototoxic drugs, with 59 children (28.78% of all patients with IRDA and 2.65% of all screened). The second most prevalent IRDA was the low rate of apgar at birth, in a total of 30 patients (14.36% of the IRDA and 1.35% of the screened), followed by low birth weight, with 24 patients (11.71% of the IRDA and 1.08% of the screened) and then by the congenital infection, with 22 patients (10.37% of the IRDA and 0.99% among the screened). ICU admission, the presence of craniofacial anomaly and parental consanguinity were responsible for less than 1% of the total number of screened, each. There was only one case of neonatal jaundice at the level of exsanguineotransfusion. Of the 205 patients with IRDA, 96 were male and 109 were female.

Congenital infections were found in 22 (9.22%) of the patients with IRDA, distributed as shown in Figure 3, in four infections, the most prevalent being congenital syphilis, found in 11 evaluated patients.

Of the 2019 patients without IRDA who were screened 281 (13.92%) failed, a rate very similar to 40 of the 205 (19.51%) patients with IRDA. There was also no significant difference between genders in relation to the failure rate, as well as a homogeneous distribution between genders regarding the presence of IRDA, seen in figures 4 and 5.

The failure rate had a big difference between the two different age groups at the time of screening. Figure 6 shows this difference, showing the highest chance of failures in the group of children screened for up to five days of life.

There was no record of age at the time of screening 18 NBs, of the remaining 2206, 1025 (46.46%) were screened within 5 days of life, and 1,181 (53.54%) after the fifth day of life. Of all patients screened up to the fifth day, 45.07% failed. Those screened after the fifth day of life, only 7.11% failed. There was a 6.3 times greater probability of failure in children screened before the fifth day.

Of all those submitted to TANU, 2019 did not have any risk factor, of these, 420 failed the first test and needed a retest, but only 182 (43%) attended. Of these, 43 (a total of only 2.41% of the screened) failed the retest and needed to go to the diagnostic stage. Distributed according to Figure 7.

The average age of newborns screened at the service was within the quality parameter, before 30 days of life. The coverage of the program maintained coverage rates within the acceptable limit for quality only in the months of July and August 2015, remaining below 95% throughout the remaining period. Only 43 neonates from the group Without IRDA were referred for diagnosis, a total of only 2.41% of the screened. The other two quality parameters evaluated in this study were not achieved: neither the conclusion of the auditory diagnosis in 90% of the newborns referred until the third month of life, nor the sound amplification in 95% of the infants with confirmed bilateral hearing losses, within the within one month of diagnosis.

ILLUSTRATIONS



Figure 1: Graph of live births submitted to TANU.



Figure 2: Age chart at the time of screening (in days).



Figure 3: Distribution of the incidence of congenital infections.



Figure 4: Graph of groups with IRDA and without IRDA - Passed x Failed.



Figure 5: Distribution by gender in failed and passed.



Figure 6: Failure rate of <or equal to 5 days and> 5 days.



Figure 7: stratification of newborns submitted to TANU.

Table 1: Distribution of IRDA.

Distribution of IRDA found in the study						
	Ν	Relationship with IRDA	Relationship with Submitted			
Use of Ototoxic	59	28,78%	2,65%			
Apgar below 4 in the 1st min or below 6 in the 5th	30	14,63%	1,35%			
Weight less than 1,500 g	24	11,71%	1,08%			
Congenital Infection	22	10,73%	0,99%			
ICU stay for more than 5 days	15	7,32%	0,67%			
Craniofacial anomaly	11	5,37%	0,49%			
Parental inbreeding	7	3,41 %	0,31%			
Family History of Hearing Impairment	3	1,46%	0,13%			
Jaundice at the level of exsanguineotransfusion	1	0,49%	0,04%			
Total	205	100,00%	9,22%			

DISCUSSION

It was estimated by the Brazilian Institute of Geography and Statistics (IBGE), in the 2010 census that, at that time, Brazil had 190,755,799 inhabitants, spread over a territorial area of 8.5 million square meters. According to data from this same census, about 9.7 million Brazilians had hearing loss. There are more than 5,000 municipalities, with great geographical, demographic, social, cultural and economic differences. This picture allows us to infer that the implementation of an auditory health system, like any health care, in a country like Brazil, is a challenge.^[4,7,8]

Determining why a child has a hearing impairment is a complex task, which most often is not done in an isolated consultation. It is vital to know which parts of the auditory system are functioning correctly and take into account that the underlying explanation may not be in the common group of causes of hearing loss.^[2]

TANU is part of a set of actions within the scope of secondary health care, in order to perform the early detection of hearing loss and enable the early

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intervention of such a significant condition. Neonatal hearing loss is the most prevalent disease at birth, and if not detected, it will impair speech acquisition, hinder language and cognitive development. Its prevalence is estimated at three per thousand live births. This prevalence increases even more when considering newborns who required intensive care, reaching four out of every 100 births. This prevalence is high when compared to other diseases that can be screened in the neonatal period, such as phenylketonuria, with an incidence of 1: 10,000, or sickle cell anemia, with two cases for every ten thousand births.

There are several causes of hearing loss due to maternal infections during pregnancy. All children of mothers with a diagnosed infection need further investigation. The most common congenital viral infection and the most common cause of non-genetic hearing loss is that caused by cytolomegavirus (CMV). About 1 in 150 children is born with congenital CMV infection. Due to the potentially delayed presentation of CMV-related hearing loss, TANU tests may be normal.^[2, 9] Since the

introduction of the rubella vaccine in 1969, the number of cases in developed countries decreased dramatically.

Infection of congenital rubella involving the inner ear results in bilateral sensorineural hearing loss, which normally affects the mid frequencies more than the low and high frequencies. The incidence of sensorineural loss in congenital HSV infection is rare.10 The end result of congenital syphilis in the temporal bone is endolymphatic hydrop.^[2] Most newborns with congenital toxoplasmosis are asymptomatic, although CNS involvement, visual, and other systems are possible. The prevalence of toxoplasmosis associated with hearing loss in infected newborns ranges from 0% to 26%.

In 1999, Yoshinaga-Itano, in a review of intervention studies with American children, revealed that hearing loss undergoing treatment before the age of six months allows the child to develop language normally. Therefore, early diagnosis and intervention provide better conditions for emotional development and social interaction.^[10, 11]

The implementation of TANU was challenging. The evaluation of the program follows quality criteria proposed by the international quality services of reference in hearing health, such as the JCIH and the NCHAM, in addition to being in accordance with the national document of the Guidelines for Attention of Newborn Hearing Screening. The proposed quality criteria are used as tools to control the effectiveness of the programs implemented.

Throughout the work, there was a correct registration of all IRDA, with a check of incidence, which remained above the value of the population average.

In the period analyzed by the study, 2,224 newborns (71.5%) underwent UAN. A value considered low for NCHAM and JCIH if the percentage was analyzed separately. However, by dissecting the study, it can be seen, over time, an evident increase in the rate of patients undergoing TANU monthly. In the first month, only 10.9% were submitted, which reached 94.1% just 4 months after its implantation. This reveals the improvement in logistics and the integration of the pediatric, speech therapy and otorhinolaryngology teams.

Even with a team responsible for reduced screening, composed only of a speech therapist who works 40 hours a week during business hours, optimal levels of coverage were obtained, in the last six months of the study reaching levels above 90%, but which can only be reached by the assistance of the otorhinolaryngology team.

The age of the newborn at the time of screening is an important measure of the quality of the service provided since the anticipation of the audiological diagnosis is

desirable so that the intervention is as early and effective as possible. However, we found in our study that extreme precocity (before five days of life) is related to a higher failure rate in patients with or without a risk factor.

There is an important relationship between early screening and the rate of unnecessary referrals to diagnosis. The changes in the external and middle ear are closely related to false positives (or false "Failed") in the screening, such as vernix in the EAC, amniotic fluid in the middle ear, and even gastroesophageal reflux. A discussion should be carried out, and other studies carried out, in order to find the ideal age for screening, since high failure rates are not desired. The JCIH and NCHAM recommend TANU before hospital discharge, but this model has been associated with higher rates of test failure, especially in Brazilian hospital systems.^[12]

Syphilis is a health problem that, in the Brazilian scenario, was under control. But in recent years, it has proved to be an increasing problem. This fact was again evidenced by our study, which showed a considerable prevalence of congenital syphilis.

In addition to the average age of newborns screened at the service being within the quality parameter, before 30 days of life, the program's coverage maintained notable coverage rates. But it was possible to reach a minimum of 95% only twice in the analyzed period. There was a high abstention rate in the retest, of 57%, a fact that needs to be corrected with some rescue measure. Of the 182 who attended the retest, only 43 failed again, which corresponds to a rate of 2.41% of all neonates who completed the program, a value well below the maximum required under the quality criteria, of 4%. Due to the lack of a program for infants with hearing loss, it was not possible to conclude the diagnosis of the screened neonates. There was also no reference and, therefore, the neonates were not amplified.

In addition to completing screening properly, a hearing health program must be aware of postnatal losses. Several factors can contribute to hearing problems that occur after birth. Although congenital hearing loss affects 3 in 1,000 births, at least the same number of children acquire hearing loss during the first months of life. As with congenital hearing loss, late-onset hearing loss can be classified based on the location of the problem (conductive, sensorineural or mixed). Although there are few solutions for reversing congenital hearing loss, the prognosis for some of the causes acquired in the postnatal period is often more encouraging. Function return is sometimes an option. The search for the determination of congenital or acquired hearing loss is important and should be investigated. The delay in recognizing congenital genetic etiologies was often attributed to parents who were not observant or professionals rushed in the diagnosis.^[2]

There is a lack of standardization of the program in association with computerized systems, which is a quality criterion of the program. In addition to demonstrating the TAN profile, we recommend with this study that the quality of the TANU program be improved and monitored according to the quality parameters proposed by NCHAM. The next stage will be to implement a system of analysis of all identifiable parameters for the evaluation of the program in order to reach all quality criteria.

CONCLUSION

71.55% of live births were submitted to TANU from July 2014 to December 2015. There was a progressive increase in the monthly index, reaching 94.10% just four months after its introduction, with a period of decline that was soon recovered, maintaining a monthly pattern above 90% one year after the beginning of the program, frequently reaching the percentage of 100% coverage.

1625 male and 1528 female neonates were born at the time. In the program, 1066 male and 1204 female infants were screened, therefore 65.6% and 78.8% respectively.

In the month the program started, the average age of screened infants was 79.09 days of life, reducing the time to acceptable values six months after the introduction of the program, with an average age of 20.63 days of life. The fashion and median of the last seven months was 2 days of life, indicating the precocity of screening in the service.

Of those screened, 205 (13.25%) had one or more risk factors. The following IRDA were identified during the assessment.

The incidence of each IRDA is summarized in table 1. The main risk factor identified was the use of ototoxic drugs, with 59 children (28.78% of all patients with IRDA and 2.65% of all screened). The second most prevalent IRDA was the low rate of apgar at birth, in a total of 30 patients (14.36% of the IRDA and 1.35% of the screened), followed by low birth weight, with 24 patients (11.71% of the IRDA and 1.08% of the screened) and then by the congenital infection, with 22 patients (10.37% of the IRDA and 0.99% among the screened). ICU admission, the presence of craniofacial anomaly and parental consanguinity were responsible for less than 1% of the total number of screened, each. There was only one case of neonatal jaundice at the level of exsanguineotransfusion. Congenital infections were found in 22 (9.22%) of the patients with IRDA, distributed as shown in Figure 3, in four infections, the most prevalent being congenital syphilis, found in 11 evaluated patients.

Of all patients screened up to the fifth day, 462 (45.07%) failed. Of those screened after the fifth day of life, only 84 (7.11%) of the 1,097 failed. A 6.3 times greater probability of failure in children screened before the fifth day.

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Of the 420 patients without IRDA who failed the first test and needed a retest, 238 (57%) did not attend. Of the 182 who underwent the retest, 43 (a total of only 2.41% of those screened) failed the retest, and needed to go to the diagnostic stage.

It was possible to fully meet two of the five quality criteria, screening before the first month of life and a referral rate for diagnosis of less than 4% of the screened. In addition, the coverage ratio was partially covered, reaching a minimum of 95% in just two months. The other two quality parameters evaluated in this study were not achieved: neither the conclusion of the auditory diagnosis in 90% of the newborns referred until the third month of life, nor the sound amplification in 95% of the infants with confirmed bilateral hearing losses, within the within one month of diagnosis.

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