

Crianças com encefalopatia crônica não evolutiva: avaliação audiológica e próteses auditivas***

Children with chronic non-progressive encephalopathy: hearing evaluation and hearing aids

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Abstract

Aim: to assess the auditory abilities of children with non-progressive chronic encephalopathy (NPCE), independently of the presence or not of hearing loss, and of the etiology of the encephalopathy; to characterize the benefit of hearing aids in children with NPCE and hearing loss. **Method:** neurologic, otorhinolaryngologic and auditory assessments. Application of the Parent's Evaluation of Aural/Oral Performance of Children (PEACH) protocol. **Results:** out of the 46 assessed children, 22 (48%) presented no hearing loss and 24 (52%) presented some level of sensorineural hearing loss. Regarding the encephalopathy etiology, most of the participants presented ischemic hypoxic encephalopathy followed by infectious process and kernicterus. The results also indicate that 16 (35%) parents suspected that their child had hearing loss; out of this total, 56% had the hearing loss confirmed. Thirty parents (65%) did not have any hearing complaints about their children. For these children the auditory evaluation indicated that 50% presented some level of hearing loss. The PEACH protocol proved to be effective to assess the benefit of hearing aids. **Conclusion:** the results indicate that over half of participants presented hearing loss. No correlation was observed between etiology and complaints of hearing loss. This means that it is not possible to predict hearing loss based on complaints. All children who presented hearing loss benefited from the use of hearing aids.

Key Words: Chronic Encephalopathy; Hearing; Hearing Auxiliary Device.

Resumo

Objetivo: avaliar a capacidade auditiva de crianças com encefalopatias crônicas não evolutivas (ECNE) independentemente de suspeita de perda auditiva e da etiologia e caracterizar o benefício do uso de prótese auditiva em crianças com ECNE que apresentaram perda auditiva. **Método:** avaliação neurológica, otorrinolaringológica e audiológica e aplicação do protocolo *Parent's Evaluation of Aural / Oral Performance of Children* (Peach). **Resultados:** Das 46 crianças avaliadas, encontraram-se 22 (48%) sem perdas e 24 (52%) com algum grau de perda auditiva sensorineural. Quanto às etiologias encontradas nas 46 crianças, a maior porcentagem é de encefalopatia hipóxica isquêmica seguida de processos infecciosos e *kernicterus*. Quanto à suspeita de perda auditiva, nas 16 (35%) crianças cujos pais tiveram suspeita, o percentual de algum grau de perda auditiva foi de 56%, e nas 30 (65%) cujos pais não a tiveram, a avaliação audiológica revelou que 50% apresentaram algum grau de perda auditiva. O protocolo Peach se mostrou um instrumento eficaz para avaliar o benefício da prótese auditiva. **Conclusão:** das crianças avaliadas, mais da metade apresentou perda auditiva, no entanto, não houve relação estatisticamente significativa entre a etiologia e a suspeita de perda auditiva. Assim, consideramos que não é possível prever qualquer perda auditiva a partir da suspeita e recomendamos a avaliação auditiva em todas as crianças com ECNE, pois todas as crianças com perda auditiva examinadas neste estudo revelaram benefícios importantes com o uso da prótese auditiva.

Palavras-Chave: Encefalopatia Crônica; Audição; Auxiliares de Audição.

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Introduction

The chronic non-progressive encephalopathy (NPE) is the non-progressive consequence of a disruption in the central nervous system development during the pre, peri or postnatal period. The deprivation of any stimuli whichever motor, visual or auditory can aggravate the child's limitations, and, similarly, all ways of amplifying the benefits to be received could be applied to its full potential 1,2.

The limitations imposed by hearing deprivation can harm even more children with NPE. Therefore, it is mandatory to identify any grade of hearing loss in order to break the stigma that lack of communication brings on children with multiple deficiency; as so to reduce the insecurities to impact on their communicative/linguistic behavior 3 .

Different authors worldwide conducted an audiologic evaluation on risk children, finding several of them with hearing loss, although not NPE related. Nevertheless, they all recommend the hearing monitoring in this population 4,5,6,7,8,9,10,11. Other authors refer to hearing loss occurrences that may vary from mild to profound (deep) in low weight newborns or premature 12,13,14,15, and several discuss the implications of hearing loss in oral language 1,2,16.

The larger incidence found is on perinatal complications with hypoxic ischemic encephalopathy, commonly known as neonatal anoxia, being this the main cause of diffuse brain damage on newborns 17.

Another important occurrence is related to elevated levels of bilirubin not treated on time, what can possibly lead to an impregnation throughout the central nervous system, called kernicterus 18. Other authors 19,20 reveal that hearing deficiency and NPE are the main sequela from kernicterus, besides the infectious process, with emphasis to bacterial meningitis caused by *Haemophilus Influenzae*, by meningococcal infection, cytomegalovirus and toxoplasmosis.

The hearing prosthesis or Ear Hearing Device (EHD) - is an electronic device that captures the sound signal and converts it into an electric signal which is amplified and retransformed to an acoustic signal to the ear 21,22 .

Therefore, the use of hearing aid in cases of impairment and NPE is to be always recommended when good testing conditions and responses are deemed satisfactory. The multidisciplinary team and family will follow up the progress, advantages and

disadvantages that prosthetization can provide 2,3,23 .

In order to evaluate the benefits of the hearing aid, we conducted the audiologic evaluation in a cabin as well as an evaluation questionnaire. The Parent's Evaluation of Aural/Oral Performance of Children (PEACH) is based on systematic parental observations 24.

The scope of this work study was to evaluate the hearing capacity of children with NPE regardless of the presence of an objective suspicion, and, in cases of hearing loss confirmation, to evaluate the benefit of the hearing aid.

Method

Between February 2006 and February 2007, 90 children were submitted for evaluation; and the established criteria for inclusion was for children with NPE of both genders ranging from 1 to 15 years of age, having the ability to respond to audiologic exams, either with or without suspicion of hearing impairment. 44 children were excluded. 46 children with NPE remained in this study.

As for the procedures, it included neurological, otorrinolaringological and phonoaudiologic evaluations (audiologic evaluation and EHD benefit evaluation).

The audiologic evaluation included: threshold tonal audiometry through air pathway at frequencies of 250 to 8.000 Hz; bone pathway at frequencies of 500, 1000, 2000 and 4000 Hz (applied when the threshold tonal audiometry through air pathway was greater than 20 dBA); speech awareness threshold (SAT) / speech detection threshold (SDT) by visual demonstration or simple pattern word/ command repetition, using the audiometer equipment AC 40-Interacustics and Intera (GN Optometric).

The tympanometry was conducted with the equipment AZ7-R and AZ7 (Interacustics) properly set for frequencies of 500, 1.000, 2.000 and 4.000 Hz.

The classification for the hearing loss grade level was based on the air pathway threshold average at the frequencies of 500, 1.000 and 2.000 Hz 10.

The children identified with hearing loss were submitted to the EHD selection and adaptation process, which has its supply guaranteed by Law GM n° 2.073, of September 28th 2004, by the same institution to have carried out the research: Sorority Santa Casa de Misericórdia de São Paulo.

*Avaliação Aural/Oral do Desempenho da Criança pelos Pais.

The child's ear pre-molding was taken to be sent to the laboratory. After a week, and in possession of the child's molding and the adequate EHD, the adaptation process was initiated. In this first moment the settings were made from the audiologic evaluations as well as from the child's behavior to the amplification. The children would take the EHD home on the same day.

With each child there had been a functional gain (audiologic EHD evaluation on free field in an adequately set cabin environment) and, during a year, phonoaudiologic therapy and weekly family orientation. Before the beginning of the EHD selection and six months after its use, the PEACH questionnaire was applied 24, translated and adapted by the researcher herself in order to evaluate the efficiency of the amplification on the children through a systematic parental observation. The application of the PEACH protocol strictly followed its guidelines, working each issue in an explanatory and enjoyable manner.

Below are the terms developed by the author:

The protocol was developed as an everyday functional performance rating, structured on the systematic parental observation in different situations: six on quiet environment and five on noisy environment, being classified in five scales never: 0 (%), seldom: 1 (25%), sometimes: 2 (50%),

often: 3 (75%) and always: 4 (more than 75%).

Age classification

The children were classified in three groups according to age: from 1 to 4, 5 to 9, and 10 to 14 years old, having the aural / oral performance follow up been conducted by parental observation, as per PEACH protocol guidelines, in quiet (questions 3, 4, 7, 8, 11 and 12) and noise (questions 5, 6, 9, 10 and 13) environments.

Statistic analysis

The data is shown by frequency, percentile and images.

In order to evaluate the correlation between the studied groups' quantitative variables, the chi-square test was used by adopting 5% as a significant level (p).

Resultados

Sample characterization

The age varied from 1 to 15 years old, with an average of 7,5 years, 28 (60,8%) children were of male gender and 18 (39,2%), female.

As for the etiologies found on 46 children, the greater percentile originates from hypoxic-ischemic

CHART 1. Parent's Evaluation of Aural/Oral Performance of Children.

	Never 0%	Seldom 25%	Some times 50%	Often 75%	Always >75%
1. Child's use of hearing aids / cochlear implant					
2. Is your child upset by loud sounds					
PEACH items nº (scale). Item					
3. (Q) Respond to name in quiet					
4. (Q) Follow verbal instruction in quiet					
5. (N) Respond to name in noise					
6. (N) Follow verbal instruction in noise					
7. (Q) Follow story read aloud					
8. (Q) Participate in conversations in quiet					
9. (N) Participate in conversation in noise					
10. (N). Participate in conversation in public transport					
11. (Q) Recognise voice of familiar persons					
12. (Q) Converse on the phone					
13 (N). Recognise sounds in environment					
(Q) Quiet sub-scale score: (3+4+7+8+11+12)/24*100					
(N) Noise sub-scale score: (5+6+9+10+13)/20*100					
Total PEACH score: overall /44*100					

encephalopathy perinatal on 27 children, followed by infectious process on 9, kernicterus on 6, hypoxic-ischemic encephalopathy prenatal on 3 and malformation on 1.

As for cerebral paralysis, a greater concentration of the hemiparesis type was found in a total of 26 children, followed by spastic diplegic type on 18 and atetosis on 2.

From the 46 children evaluated by threshold tonal audiometry through air pathway, 22 (48%) were found without loss (classified as "normal") and 24 (52%) at some degree of sensory-neural hearing loss. Out of these 24, 10 children showed bilateral deep grade, 7 severe, 5 moderately severe, 1 moderate, and 1 mild.

Out of 26 children, the speech awareness threshold (SAT) was obtained by a 3-syllabus repetition, and on 5 children, by simple order command. The speech detection threshold (SDT) was conducted on 15 (33%) children with severe and deep sensory-neural hearing loss.

On the tympanometric evaluation, 100% of the children showed curve type A. In all children that did not show hearing loss, a detection of present answers from the stirrup muscle was made possible; enabling this evaluation to rule out any compromise to the middle ear. The 24 children that did not show reflexes to the stirrup muscle evaluation tested positive for hearing loss.

We were able to observe from a total of 46 children with complain of hearing loss - despite there had been no record of such complain registered by the parents of 30 (65%) of these children - the audiologic evaluation revealed 50% of them showed some level of hearing loss. Among the 16 (35%) children with parents having suspected of hearing loss, the percentile showing some level of impairment was 56% (9 children). There was no statistically significant association between the complain of loss and the hearing loss (p=0,4612).

Table 1 shows the percentile and the quantity of children with and without complain related to the level of hearing loss. The no complain group (30 children) offered 50% with hearing impairment distributed in mild loss (3,3%), moderately severe (13,3%), severe (20%) and profound (13,3%). On the 16 children with complain it was found 56% with hearing loss, being the greater percentage (37,5%) on children at deep grade loss. There was no difference between the groups in relation to the level of loss.

Table 2 relates the two groups to the etiology. The data indicates a greater percentage to the hypoxic-ischemic encephalopathy perinatal etiology on both groups. On the group without

complain the second greater percentage found was kernicterus (16%), followed by infectious process (13,3%), hypoxic-ischemic encephalopathy prenatal (10%) and malformation in only 3,3% of the cases. On the group having registered a complain, the second greater percentage is related to infectious process (31,3%), followed by kernicterus found in only one case. There was no difference between groups in relation to the etiology.

The EHD benefit evaluation utilizing the PEACH protocol showed the distribution of children from the following age groups: 1-4; 5-9 and 10-14 years old in response to the PEACH protocol on a silent and noisy environment, both before and after the use of the EHD. An enhancement on the aural/oral performance was verified in all groups according to parental observation.

TABLE 1. Distribution of group patients with and without complain by level of hearing loss.

Level of hearing loss	No complain		With complain	
	N	%	N	%
Normal	15	50	7	43,8
Mild	1	3,3	0	0
Moderate	0	0	1	6,3
Moderate/severe	4	13,3	1	6,3
Severe	6	20	1	6,3
Profound (deep)	4	13,3	6	37,5
Total	30	100	16	100

p = 0,215

TABLE 2. Distribution of patients by group etiology.

Etiology	No complain		with complain	
	N	%	N	%
Hypoxic-ischemic encephalopathy perinatal	17	56,7	11	62,5
Hypoxic-ischemic encephalopathy prenatal	3	10	0	0
Kernicterus	5	16,5	1	6,3
Malformation	1	3,3	0	0
Infectious process	4	13,3	5	31,3
Total	30	100	16	100

p = 0,311

Discussion

The evaluation of the hearing capacity on children with NPE, regardless of hearing loss suspicion, is important because the therapeutic modality indicated can vary according to the type of loss classification, and such classification will depend on where the lesion took place to cause the impairment, as well as other singularities demanded by the NPE itself.

The determination of the relation between the hearing deficiency and the NPE etiology favors the global diagnostic. Our findings indicate a greater percentage to children with etiology classified as hypoxic-ischemic encephalopathy perinatal for NPE, as well as for NPE associated to a hearing deficiency, according to 7,13,14,15.

Another NPE etiology associated to hearing loss is kernicterus. The concern is due to the descendent audiologic pattern of individuals. As the child may hear the phonemes of lower frequencies (most deep), and depending on the social communication stimuli of exposure, parents could believe the child only lacks attention without suspecting of hearing impairment. That is verified in our work - of 6 children with the kernicterus etiology^{18,19,20}, all showed hearing loss, but only one had a case of hearing loss under parental suspicion.

In our study, the neurological and sensorial implications derived from infectious processes consisted 37, 5% of the cases (7 meningitis, 1 citomegalovirus and 1 toxoplasmosis) the same was found in^{1,2,16}.

The audiologic^{4,7,8,13, 14}, findings the same of us and show it possible to quantify the hearing loss through subjective exams, and along with the imitancymetry and the OAEs, to produce a differential diagnostic of the sensory-neural hearing losses.

The results of audiologic evaluations have implications on the EHD choice, as well as on its planning and communication strategies. The issue of comfort related to the amplification process is one of the concerns when considering prosthetizing a child with NPE, given that motor, cognitive and emotional limitations are important in the therapeutic intervention.

Therefore, the prescription to the EHD input and output depends on the loss configuration; however, it is the parental observation on the everyday use of the device that will allow the audiologist to adequately adjust it to each child with respect to their personal hearing discomfort boundries. It is important to consider that the time of prosthesis use does not pose as a limiting factor, but as an aspect that will serve to better accommodate the children upon their new condition and we agree with^{21,22}.

When thinking of prosthetizing, the first step is work up to the maximum the use of residual hearing, which one of the objectives is to enhance the development of hearing capacity, and consequently, the linguistic capacity. Nevertheless, it is important to highlight that EHD is only a part of the work and not the solution of hearing problems to agree to^{3,23}.

The children's adapting time to the EHD varied between one week to three months; it means that some may have undergone its daily use since the beginning and others only after several weeks (usually in case of severe hearing loss). We believe the principles of this process to be motivation and expectancy, and not as much the level of hearing loss or motor compromise of the studied children, for many with severe loss have adapted to the EHD in a very short period of time, even when sign language was applied. It seems as the factors that contribute to the success of the therapeutic process - which includes adaptation - create the transformation of real expectancy and continuous motivation, so much as by the parents, as by the therapists.

We have established these age groups (1 to 4; 5 to 9 and 10 to 14 years old) for believing that on children with NPE showing some level of hearing loss, the expectancy of answers could make a difference on communicative behavior. Therefore, the goal was not to evaluate which child specifically showed the benefit isolatably while in a silent or noisy environment, but to present the age group set as a whole, and its benefits to the amplification.

As a result, we were able to notice the benefit on silent environments to be greater than the one applied on a noisy environment, thus improving the signal-noise relation established by the EHD.

Subsequently, it is important to highlight this is a work study based on stimulation and orientation designed to enhance the hearing of these children, as well as to favor their global development. We know the EHD alone does not guarantee the aimed goal for the audiologist or the family of these children. The therapeutic work is constant and intense, and without this process it would not be possible to evaluate this set of information, given that on each session, the possibility for exchanging the child's living experiences between the therapist and the family was made possible, thus contributing to a differentiated approach capable of answering to their special needs.

It is known that parents with NPE children spend most of the time at a constant pilgrimage to different professionals, but on the other hand, they may be lacking to actually see them, by carrying out such an automated behavior. Therefore, the application of the PEACH protocol makes it possible to interact in a closer manner, thus helping to build a better involvement between parents and children. On the same hand, it was exactly such an approximation that led to uncertainties pertaining to the possible deafness of the child, as the continuous presence of parents has functioned as a block from seeing the children beyond their motor

hindering.

As deafness is intimately connected to communication, and once the later is compromised by neurological damage, it seems as not possible to actually see a hearing problem, given that deafness - which is not visible, unlike, for example, a motor compromise - goes unnoticed by the parents and many health professionals, that will instead try to care for what is in evidence. Although in cases that neurological compromising is severe, health professionals must be cautious; nonetheless, it is equally accurate that these cases be treated singularly; because once faced with several impossibilities, having the possibility to hear can be

a way of keeping an important contact. We; however, insist for this to be a particular given situation.

Conclusion

There is no doubt the closing stages of this study do not bring the subject to a conclusion, given that singularities of each child and each family, the regular monitoring of the EHD programming, the parents' doubts, moreover when related to hearing and EHD benefits, constantly vary, demanding always new initiatives and research. Be as it may, the benefits owed to the EHD use is notorious on all studied patients.

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