4a. Conferência Latino Americana
de Adaptação Pediátrica

ESPECTRO DA NEUROPATIA AUDITIVA

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São Paulo

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Definições

- Distúrbios auditivos relacionados à disfunção na **sinapse entre as CCI e o Nervo Auditivo**, ou no próprio nervo;
- Evidências clínicas de **funcionamento normal das CCE**;
- **Dificuldades na compreensão da fala**;
- Limiares auditivos para **tons puros** e de **detecção da fala** que podem variar entre a **normalidade** e similares às perdas auditivas **severas**;
- Dificuldades no **processamento auditivo temporal** (mudanças rápidas em um período de tempo)

Características

- **Sem dificuldades** auditivas aparentes, porém com **testes audiológicos alterados**;
- Sem queixas auditivas, porém com **queixa na compreensão da fala;**
- Dificuldades na **presença de ruído**;
- As habilidades auditivas podem se mostrar **flutuantes ou transitórias**;
- Em crianças pode **afetar o desenvolvimento da linguagem e acadêmico**;

Brain (1996), **119**, 741–753

Auditory neuropathy

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Summary

Ten patients presented as children or young adults with hearing impairments that, by behavioural and physiological testing, were compatible with a disorder of the auditory portion of the VIII cranial nerve. Evidence of normal cochlear outer hair cell function was provided by preservation of otoacoustic emissions and cochlear microphonics in all of the patients. Auditory brainstem potentials showed evidence of abnormal auditory pathway function beginning with the VIII nerve: the potentials were absent in nine patients and severely distorted in one patient. Auditory brainstem reflexes (middle ear muscles; crossed suppression of otoacoustic emissions) were absent in all of the tested patients. Behavioural audiometric testing showed a mild to moderate elevation of pure tone threshold in nine patients. The extent of the hearing loss, if due to cochlear receptor damage, should not have resulted in the loss of auditory brainstem potentials. The shape of the pure tone loss varied, being predominantly low frequency in five patients, flat across all frequencies in three patients and predominantly high frequency in two patients. Speech intelligibility was tested in eight patients, and in six was affected out of proportion to what would have been expected if the pure tone loss were of cochlear origin. The patients were otherwise neurologically normal when the hearing impairment was first manifest. Subsequently, eight of these patients developed evidence for a peripheral neuropathy. The neuropathy was hereditary in three and sporadic in five. We suggest that this type of hearing impairment is due to a disorder of auditory nerve function and may have, as one of its causes, a neuropathy of the auditory nerve, occurring either in isolation or as part of a generalized neuropathic process.

Keywords: neural hearing loss; auditory neuropathy

Abbreviations: ABR = auditory brainstem responses; HL = hearing level; nHL = normal hearing level

✓ **Pesquisadores apresentam um grupo de 10 pacientes com resultados audiológicos compatíveis com um distúrbio do VIII Par Craniano**

❖ **Evidências de células ciliadas externas com funcionalidade normal; Presença de EOA e Microfonismo Coclear**

✧ **PEATE com evidências de anormalidade nas vias auditivas; Respostas ausentes ou muito alteradas**

◆ **Audiometria tonal de normal a profunda com diferentes configurações**

➢ **Inteligibilidade de fala imompatível com os resultados da audiometria tonal**

The Laryngoscope
Lippincott Williams & Wilkins, Philadelphia

1988, estudo com 8 crianças

Auditory Neuropathy in Childhood

Karen Jo Doyle, MD, PhD; Yvonne Sininger, PhD; Arnold Starr, MD

Objectives: **Auditory neuropathy is a recently described disorder in which patients demonstrate hearing loss for pure tones, impaired word discrimination out of proportion to pure tone loss, absent or abnormal auditory brainstem responses, and normal outer hair cell function as measured by otoacoustic emissions and cochlear microphonics. We have identified eight pediatric patients having hearing deficits that are most likely due to a neuropathy of the eighth nerve. In this study, the results of audiologic testing performed with these eight children are described.** ***Study Design:*** **Retrospective review of audiologic findings in eight children with auditory neuropathy.** ***Methods.*** **Each subject was tested with pure tone and speech audio-**

INTRODUCTION

A group of pediatric patients has been identified who have hearing loss and absent or severely abnormal auditory brainstem responses (ABRs), yet have normal cochlear function as measured by otoacoustic emissions (OAEs). Starr et al.[1] named this disorder *auditory neuropathy* and studied the auditory abilities of these patients. Starr et al.[2] described an 11-year-old girl who developed progressive hearing loss from the age of 7 years, and who had such great difficulty understanding speech that she was dependent on lip-reading. Her pure-tone audiogram progressed from mild hearing loss to bilateral, moderately severe neurosensory hearing loss with poor

Clinical findings for a group of infants and young children with auditory neuropathy

Clinical Findings for a Group of Children with Auditory Neuropathy

Gary Rance, David E. Beer, Barbara Cone-Wesson, Robert K. Shepherd, Richard C. Dowell, Alison M. King, Field W. Rickards, and Graeme M. Clark

Objective: **To examine the prevalence of auditory neuropathy in a group of infants at risk for hearing impairment and to present an overview of the clinical findings for affected children.**

Design: **Results for 20 subjects who showed repeatable cochlear microphonic potentials in the absence of click-evoked auditory brain stem responses are included in this study. Behavioral and steady state evoked potential thresholds were established in each case. Where possible, otoacoustic emission and speech perception results (unaided and aided) also were obtained.**

Results: **One in 433 (0.23%) of the children in our series had evidence of auditory neuropathy. The audiometric findings for these subjects varied significantly, with behavioral thresholds ranging from normal to profound levels. Discrimination skills were also variable. Approximately half of the subjects showed little understanding, or even awareness, of speech inputs in both the unaided and aided conditions. There were, however, a number of children who could score at significant levels on speech discrimination tasks and who benefited from the provision of amplification.**

Conclusion: **The results suggest that auditory neuropathy is more common in the infant population than previously suspected. The effects of neuropathy on auditory function appear to be idiosyncratic, producing significant variations in both the detection and discrimination of auditory signals. As such, the management of children with this disorder must allow for individual differences.**

(Ear & Hearing 1999;20;238–252)

The use of auditory evoked potential techniques such as the auditory brain stem response (ABR) for the assessment of hearing in young and difficult-to-test children is now well established. A number of normally hearing and hearing-impaired subjects (Gorga, Worthington, Reiland, Beauchaine, & Goldgar, 1985; Hyde, Riko, & Malizia, 1990; Kileny & Magathan, 1987; Picton, Durieux-Smith, & Moran, 1994; Sasama, 1990; Stapells, Gravel, & Martin, 1995; van der Drift, Brocaar, & van Zanten, 1987). As a result, reasonably accurate estimates of hearing level can be made for children who are too immature to cooperate for behavioral audiometry.

However, there have been reports in the literature of isolated cases in which evoked potential threshold levels have been significantly worse than would be expected from the subject's audiogram (Davis & Hirsh, 1979; Hildsheimer, Muchnik, & Rubenstein, 1993; Kraus, Ozdamar, Stein, & Reed, 1984; Lenhardt, 1981; Worthington & Peters, 1980). For example, Kraus and her colleagues identified seven cases in a group of 49 children with absent ABRs who had behavioral thresholds in the normal to moderate hearing loss range. These authors concluded that the inconsistency between behavioral and evoked potential findings in these children may have been the result of dual cochlear and auditory brain stem dysfunction.

The presence of preneural evoked responses such as the cochlear microphonic (CM) potential and otoacoustic emissions (OAEs) in a number of recently reported cases has indicated that ABRs may be absent in children and adults with outer hair cell (OHC) function and reasonable hearing thresholds (Deltenre, Mansbach, Bozet, Clercx, & Hecox, 1997; Sininger, Hood, Starr, Berlin, & Picton, 1995; Starr, Picton, Sininger, Hood, & Berlin, 1996).

Starr et al. (1996) used the presence of these cochlear responses (OAEs and CMs) and absent or abnormal ABRs in a group of 10 adults and children to identify a disorder that they called "auditory neuropathy" (AN). The term "neuropathy" is used to

Ear & Hearing, 1999

- **Estudo com 20 crianças de maior risco mostra uma prevalência maior do que a esperada em crianças 0,23%**
- **Resultados variados de audiometria e percepção de fala;**
- **Algumas crianças com resultados aceitáveis nos testes de percepção de fala, se beneficiaram do uso de AASI**

Neuroreport:
8 November 1999 - Volume 10 - Issue 16 - p 3429–3435
Auditory and Vestibular Systems

Temporal and speech processing deficits in auditory neuropathy

Zeng, Fan-Gang[1,5]; Oba, Sandy[2]; Garde, Smita[3]; Sininger, Yvonne[2]; Starr, Arnold[4]

Abstract

AUDITORY neuropathy affects the normal synchronous activity in the auditory nerve, without affecting the amplification function in the inner ear. Patients with auditory neuropathy often complain that they can hear sounds, but cannot understand speech. Here we report psychophysical tests indicating that these patients' poor speech recognition is due to a severe impairment in their temporal processing abilities. We also simulate this temporal processing impairment in normally hearing listeners and produce similar speech recognition deficits. This study demonstrates the importance of neural synchrony for auditory perceptions including speech recognition in humans. The results should contribute to better diagnosis and treatment of auditory neuropathy.

- **ENA afeta a sincronia neuronal, sem afetar a função de amplificação da orelha interna.**
- **Relatam ouvir, porém não conseguem compreender a fala.**

- **Testes psicofísicos mostram que o reconhecimento de fala pobre está relacionado a uma Importante alteração no processamento temporal.**
- **Importância da sincronia neuronal para a percepção auditiva, incluindo o reconhecimento da fala.**

THE VARIETIES OF AUDITORY NEUROPATHY

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ABSTRACT

Auditory neuropathy (AN) was initially described as impairment of auditory neural function, with preserved cochlear hair cell function. In this report, 67 patients with audiological and neurophysiological criteria for hearing loss due to auditory neuropathy are described. Reviewing this large body of patients, AN appears to consist of a number of varieties, with different etiologies and sites affected. All varieties share a relatively spared receptor function, and an impaired neural response, with diminished ability to follow fast temporal changes in the stimulus, but different varieties in this general scheme can be distinguished. Analyses of the clinical features indicate that auditory neuropathies vary in several measures including age of onset, presence of peripheral neuropathy, etiology, and behavioral and physiological measures of auditory function. The sites affected along the peripheral auditory pathway may include dysfunction of the outer hair cells, the synapse between hair cell and auditory nerve, and the auditory nerve fibers, with myelin as well as axonal impairments contributing to the disorder.

KEY WORDS

hearing, auditory neuropathy, otoacoustic emissions, hair cells, auditory nerve, brainstem potentials

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2000

- **Estudo com 67 pacientes;**
- **Grande variação**

- **Habilidades para identificar mudanças temporais;**
- **Idade de início;**
- **Presença de patologias periféricas;**
- **Etiologia;**
- **Nas respostas fisiológicas;**
- **Nas respostas à Av. Comportamental;**

- **Locais afetados:**
- **Disfunção das CCE;**
- **Sinapses da CC e Nervo Auditivo;**
- **Fibras do Nervo Auditivo;**
- **Mielinização e Axônios;**

Clinical and Audiological Features in Auditory Neuropathy FREE

Colm Madden, FRCSI; Michael Rutter, FRCS; Lisa Hilbert, MA; John H. Greinwald, Jr, MD; Daniel I. Choo, MD

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Arch Otolaryngol Head Neck Surg. 2002;128(9):1026-1030. doi:10.1001/archotol.128.9.1026.

ABSTRACT

Objective To medically and audiologically characterize a population of children diagnosed as having auditory neuropathy (AN).

Study Design Retrospective medical chart review.

Setting/Subjects We identified 22 patients from a pediatric otology clinic in a tertiary care pediatric hospital setting.

Results A genetic factor in AN is suggested by our identification of 3 families with 2 affected children and 2 other children with family histories that were positive for hearing loss. Clinical features common among our population included a history of hyperbilirubinemia (n = 11 [50%]), prematurity (n = 10 [45%]), ototoxic drug exposure (n = 9 [41%]), family history of hearing loss (n = 8 [36%]), neonatal ventilator dependence (n = 8 [36%]), and cerebral palsy (n = 2 [9%]). Full clinical and audiological data were available for 18 of the 22 children, including otoacoustic emissions, auditory brainstem responses with cochlear microphonics, and age-appropriate audiometric findings. Significantly, 9 of these 18 patients showed improvement in behavioral thresholds over time, indicating that a subset of children with AN may recover useful hearing levels. Also significant was the success of cochlear implantation in 4 children.

Conclusions Management of AN in children requires serial clinical and audiometric evaluations, with a prominent role for behavioral testing. Prematurity, genetics, and hyperbilirubinemia appear to be significant factors in the development of AN; hyperbilirubinemia can be associated with spontaneous improvement of hearing thresholds. For those children not benefiting from amplification or FM systems, cochlear implantation remains a potentially successful method of habilitation.

- **22 crianças**
- **Etiologias do ENA**
- **Fatores Genéticos; História Familiar de DA**
- **Hiperbilirrubinemia; Prematuridade;Drogas ototóxicas; Ventilação Mecânica;**
- **Paralisia Cerebral;**

Non-syndromic recessive auditory neuropathy is the result of mutations in the otoferlin (*OTOF*) gene

R Varga, P M Kelley, B J Keats, A Starr, S M Leal, E Cohn, W J Kimberling

J Med Genet 2003;**40**:45–50

2003, DESCRIÇÃO OTOF
4 famílias com 2 ou mais filhos

It is estimated that about 1 in 500 children are born with a significant hearing loss.[1] Non-syndromic recessive hearing loss (NSRHL) represents a major aetiologic factor in childhood hearing loss since it accounts for approximately 40% of all cases.[2] Many of these genetic forms of hearing loss are indistinguishable with current clinical methods. Even so, more than 12 recessive genes have been identified primarily from large consanguineous pedigrees (see the Hereditary Hearing Loss Homepage http://www.uia.ac.be/dnalab/hhh for an overview).

By definition, non-syndromic suggests a "simple" phenotype limited to hearing loss with no other associated symptoms. However, hearing is a complex process. Since a hearing defect might occur at any place along the auditory pathway, it would seem reasonable to expect to be able to differentiate types of NSRHL based on the location where the auditory process is disrupted. Indeed, new audiological testing strategies now give insight into the point where such defects have occurred.

Pure tone audiometry has been the standard method used to measure hearing threshold but, since it subjectively tests the overall integrity of the auditory pathway, it gives only limited information about where that pathway is failing. The auditory brainstem response (ABR) is an objective measure of the overall auditory transduction process. The otoacoustic emissions (OAEs) test is another objective measure of the auditory pathway, which detects responses of the outer hair cells (OHCs) to environmental sound.[3–6] A good review of auditory tests can be found in Hood.[7]

Some children have a hearing loss based on pure tone audiometry and ABR, but with normal OAEs. This type of hearing loss has been defined as auditory neuropathy (AN).[8] Subjects with AN can have varying degrees of hearing loss with poor speech reception out of proportion to the degree of hearing loss. In contrast to those with non-AN hearing loss, most subjects with AN are not helped by hearing aids. The results of cochlear implantation (CI) have been mixed. Some cases of AN have been helped by CI,[9 10] whereas others have not had such good results.[11 12]

The genetic study of AN is complicated by the fact that the term includes hearing losses with varied aetiologies. AN can

SUBJECTS AND METHODS

The appropriate institutional review boards approved this study and informed consent was obtained from human subjects. We observed four families with two or more children who had sensorineural hearing loss and normal OHC function (fig 1). The degree of hearing loss was determined by standard pure tone audiometry. The scale used to classify the degree of hearing loss is as follows: 0-20 dB HL is normal, 21-40 dB HL is mild, 41-60 dB HL is moderate, 61-80 dB HL is severe, and above 80 dB HL is profound. The status of the auditory pathway was determined using tympanometry, middle ear muscle reflex (MEMR), and ABR. OHC function was measured using distortion product OAE (DPOAE),[3] transient evoked OAE (TEOAE),[18] and/or cochlear microphonics (CM).[19] All subjects diagnosed with hearing loss were examined by an otolaryngologist, a neurologist, and geneticist and underwent testing to rule out syndromic disorders.

Key points

- Approximately 1 in 500 children has hearing loss, with non-syndromic recessive hearing loss (NSRHL) being the most common. We studied four families with a unique type of NSRHL called non-syndromic recessive auditory neuropathy (NSRAN). Contrary to most hearing losses, the auditory pathway up to and including the cochlear outer hair cells functions normally in these NSRAN subjects, suggesting that the lesion is in the inner hair cells or more central. Most subjects with NSRAN and other types of auditory neuropathy (AN), including syndromic and non-genetic cases, have speech perception disproportionately poorer than the degree of hearing loss and disappointing results with hearing aids. To elucidate the type of hearing loss, we searched for the causative gene in four families.
- Audiological studies confirmed the presence of AN in four NSRHL families. A genome wide linkage study was carried out and the otoferlin gene (*OTOF*) was

MENTAL RETARDATION AND DEVELOPMENTAL DISABILITIES
RESEARCH REVIEWS 9: 225–231 (2003)

AUDITORY NEUROPATHY/DYS-SYNCHRONY: DIAGNOSIS AND MANAGEMENT

2003, TERMINOLOGIA NEUROPATIA AUDITIVA/ DESSINCRONIA AUDITIVA

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Auditory brainstem responses (ABRs) and otoacoustic emissions (OAEs) are objective measures of auditory function, but are not hearing tests. Normal OAEs reflect normal cochlear outer hair cell function, and an ABR indicates a synchronous neural response. It is quite possible for a patient to have normal OAEs but absent or grossly abnormal ABR and a behavioral audiogram that is inconsistent with either test. These patients, who may constitute as much as 10% of the diagnosed deaf population, have auditory neuropathy/dys-synchrony (AN/AD). To diagnose AN/AD accurately, ABRs are obtained in response to condensation and rarefaction clicks to distinguish cochlear microphonics (CM) from neural responses. Appropriate management is confounded by variation among patients and changes in auditory function in some patients over time. Recommendations for management include visual language exposure through methods such as American Sign Language (ASL), Cued Speech, or baby signs, and closely following patients.

MRDD Research Reviews 2003;9:225–231.

Key Words: auditory neuropathy/dys-synchrony (AN/AD); auditory brainstem response (ABR); otoacoustic emissions (OAEs); cochlear microphonic (CM); middle-ear muscle reflexes (MEMRs)

INTRODUCTION AND ORIENTATION—ABR AND OTOACOUSTIC EMISSIONS ARE *NOT* INFALLIBLE HEARING TESTS

The casual reader of auditory screening literature might be led to believe that otoacoustic emissions (OAEs) and auditory brainstem responses (ABRs) are in and of themselves objective tests of hearing. *They are not.* When normal OAEs are present, they reflect normal outer hair cells in the cochlea and imply a normal middle ear. However, some very deaf patients have normal otoacoustic emissions but have either absent inner hair cells or compromised neural synchrony. This should not be interpreted as a brainstem or brain disorder but more peripherally as part of a disruption of the inner hair cell nerve fiber junction or the nerve trunk itself [Starr et al., 1996; Amatuzzi et al., 2001].

Similarly, a patient may have an absent ABR and *not* be vention are not appropriate. Absent ABRs and absent OAEs in the presence of normal tympanometry (confirming a normal middle ear system) strongly indicate peripheral hearing impairment requiring intervention. Absent ABRs with present otoacoustic emissions, normal tympanometry, and absent middle-ear muscle reflexes (MEMRs) strongly suggest auditory neuropathy/dys-synchrony (AN/AD). ABRs should be obtained separately for positive and negative polarity clicks; comparison of these responses is useful in identifying the cochlear microphonic and separating it from the neural response [Berlin et al., 1998].

We have seen a few of these patients develop hearing, speech, and language normally who would never have been identified had their initial screening not included an ABR. Some develop into normal hearing and speaking adults who show little trouble other than difficulty hearing in noise; they are sometimes later mis-diagnosed as having "Central Auditory Processing Disorders" or even Attention Deficit Disorders because their ability to attend to signals in noise is inordinately poor. Some others act and live a Deaf life. The majority fall somewhere in between, showing distinct auditory problems but also showing periods of sporadic hearing or having difficulty in hearing far beyond that predicted by their pure tone audiograms.

DEFINITION OF AUDITORY NEUROPATHY/ DYS-SYNCHRONY

When ABRs and middle-ear muscle reflexes are absent, but otoacoustic emissions are present (or have been at one time), the patient is at great risk for auditory neuropathy/dys-synchrony. Data from such patients are summarized in Figures 1, 2, and 3. The rationale for the additional term is expanded upon in another publication [Berlin et al., 2001] but is reviewed briefly here. The name "auditory neuropathy" implies a confirmed pathology of the VIIIth N., when in fact evidence supports multiple etiologies and multiple locations ranging from the inner

Terminologia

- Neuropatia Auditiva, Starr – 1996
- Dessincronia Auditiva, Berlin et al. – 2001; Rapin e Gravel – 2003
- Espectro da Neuropatia Auditiva (ENA) ou Distúrbios do Espectro da Neuropatia Auditiva (DENA), Como, NHS – 2008
- Diretrizes (Guidelines for Identification and management of Infants and Young Children with Auditory Neuropaty Spectrum Disorder, Northern, J - editor) – Hayes e Sininger (2008)

Guidelines for

Identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder

The Children's Hospital
Bill Daniels Center for Children's Hearing

Guidelines Development Conference
at NHS 2008, Como, Italy

Northern, J - editor – Hayes e Sininger (2008)

Tipos

- Tipo I – Pré-sinaptico
- Tipo II – Pós-sinaptico
- **Starr et al. – 2004**

- Local de lesão
- Nervo auditivo ou CCI e sinapses
- **Distúrbio do Nervo Auditivo**
- **Distúrbio Auditivo sináptico**
- **Starr et al. - 2008**

Terminologia

- Continuar utilizando “Neuropatia Auditiva” já reconhecido por pacientes, familiares e profissionais;
- Habilidades auditivas e de linguagem com grande variação de sintomas;
- O termo **"Espectro”** poderia expandir o conceito do distúrbio em termos de locais acometidos, além do próprio nervo auditivo;

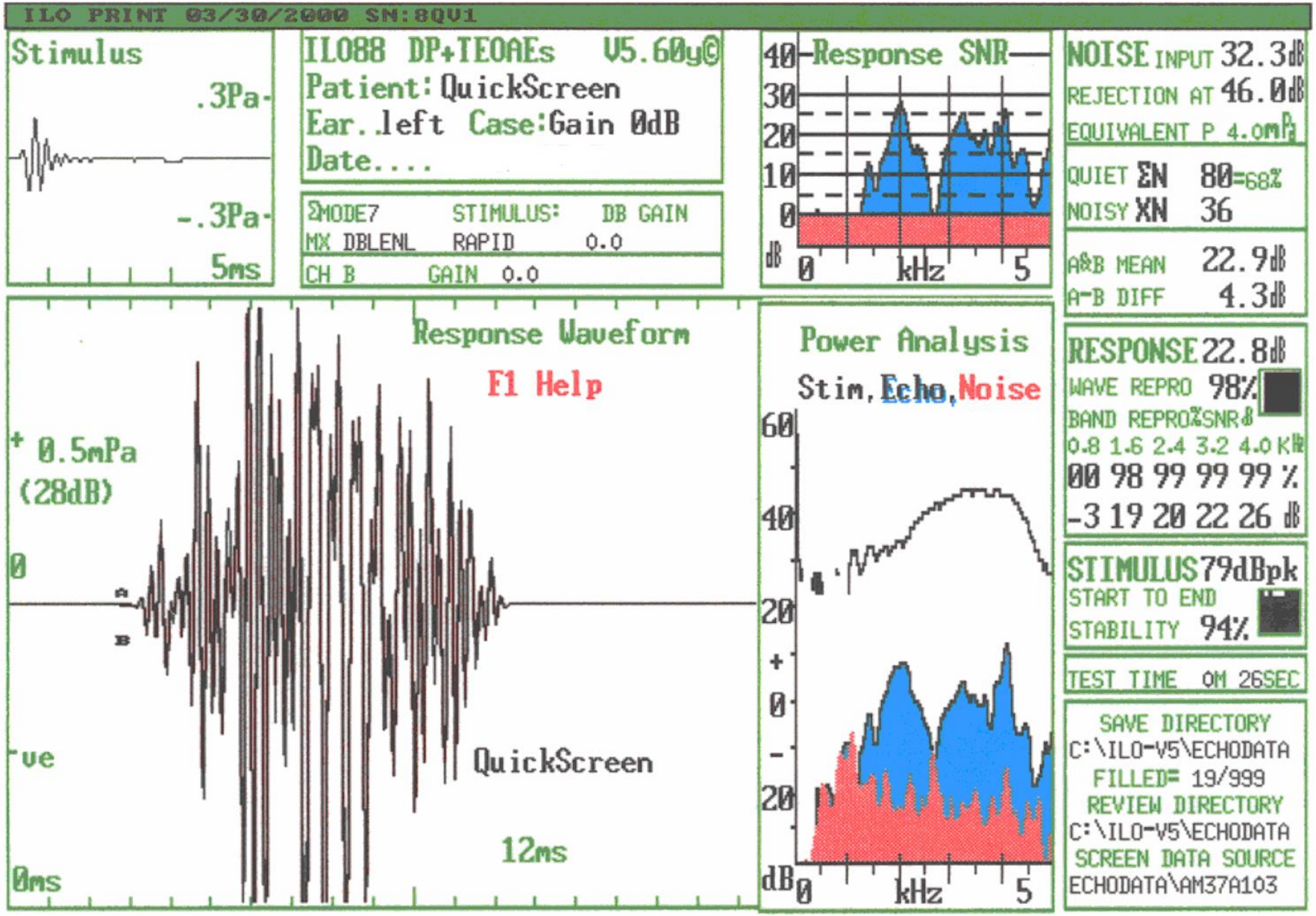
Desafios

- Cada indivíduo com o ENA deve ser considerado de **forma única**, devido à grande variação de dificuldades relatadas;
- Os testes audiológicos **não podem predizer** o grau de dificuldade apresentado por cada indivíduo;
- A **intervenção** aplicada às PANS nem sempre se adequa ao indivíduo com ENA;

DIAGNÓSTICO
Bateria de Testes

- Emissões Otoacústicas Evocadas (EOA)
- EOA-T e/ou EOA-PD –
- EOA podem estar ausentes pela presença de alterações de orelha média, principalmente em crianças;

- Pesquisa de presença/ausência de Microfonismo Coclear (MC)
- PEATE/BERA com cliques em 80-90 dBnNA
- Estímulos com polaridade invertidas (condensado e rarefeito)
- Comparar com os resultados obtidos com os tubos do fone de inserção clipados.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Date....
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MX DBLENL RAPID 0.0
CH B GAIN 0.0
Response SNR
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20
10
0
dB
0
kHz
5
NOISE INPUT 32.3dB
REJECTION AT 46.0dB
EQUIVALENT P 4.0mPa
QUIET ΣN 80=68%
NOISY XN 36
A&B MEAN 22.9dB
A-B DIFF 4.3dB
Response Waveform
F1 Help
+ 0.5mPa
(28dB)
0
-ve
QuickScreen
12ms
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Power Analysis
Stim, Echo, Noise
60
40
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kHz
5
RESPONSE 22.8dB
WAVE REPRO 98%
BAND REPRO%SNRdB
0.8 1.6 2.4 3.2 4.0 KHz
00 98 99 99 99 %
-3 19 20 22 26 dB
STIMULUS 79dBpk
START TO END
STABILITY 94%
TEST TIME 0M 26SEC
SAVE DIRECTORY
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FILLED= 19/999
REVIEW DIRECTORY
C:\ILO-V5\ECHODATA
SCREEN DATA SOURCE
ECHODATA\AM37A103

v5 | -ILO92(F)-Otoacoustic Distortion Product Analyser-4/17/00-OTUdynamics

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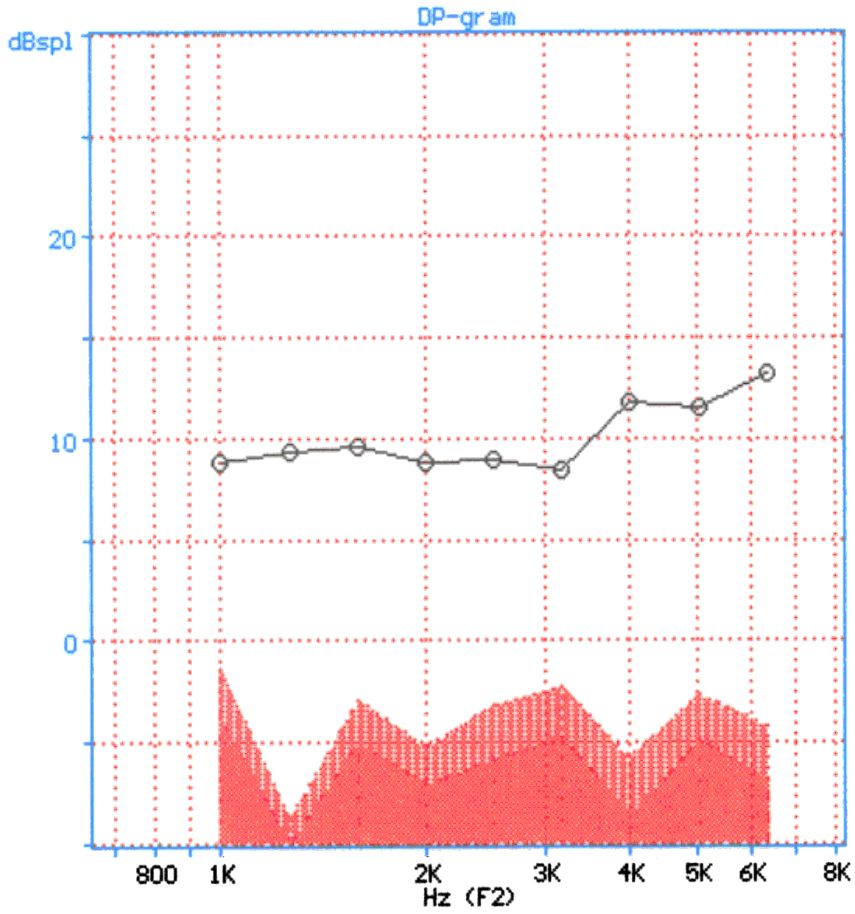
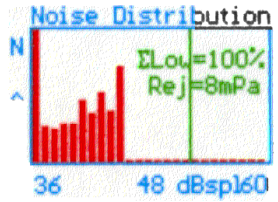
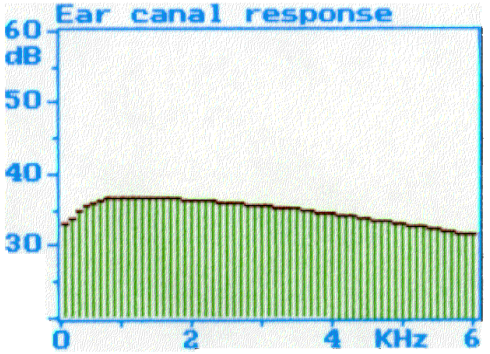






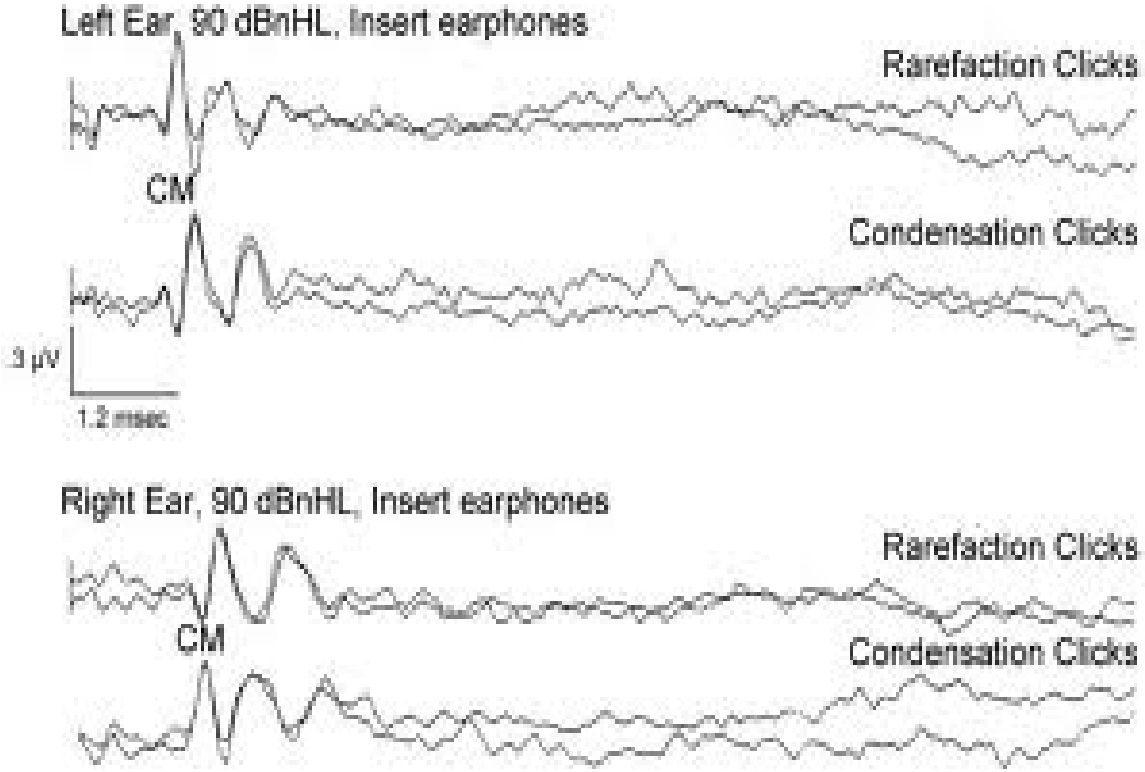
Patient record data

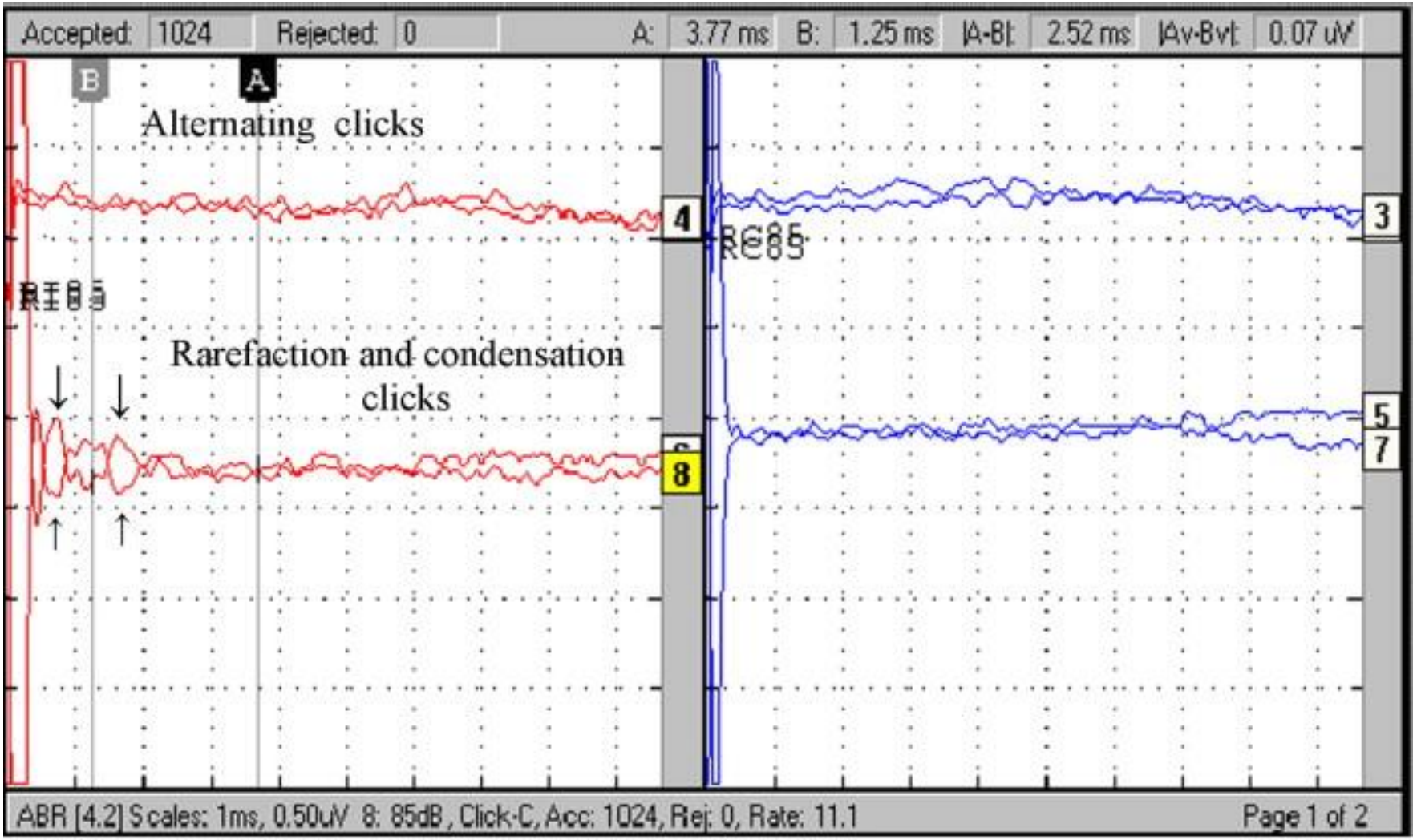
Name: Example of Normal

Ear:

No.: 123

Note:


Left Ear, 90 dBnHL, Insert earphones
Rarefaction Clicks
CM
Condensation Clicks
3 µV
1.2 msec
Right Ear, 90 dBnHL, Insert earphones
Rarefaction Clicks
CM
Condensation Clicks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B: 1.25 ms
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B
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Alternating clicks
4
3
Rarefaction and condensation clicks
8
5
7
ABR [4.2] Scales: 1ms, 0.50uV 8: 85dB, Click-C, Acc: 1024, Rej: 0, Rate: 11.1
Page 1 of 2
0 ms
12ms
1.50 uV

(A) (B)

PEATE/BERA

- Ausência de respostas; sem registro de ondas I, III e V;
- Presença de ondas com tempo de latência precoces (ondas I ou III); MC?
- Sincronia pobre com atraso de tempo de latência da onda V;

ENA x Perda Descendentes

- Diagnóstico diferencial
- Ambos podem ter MC;
- Ambos podem ter EOA alteradas ou parcialmente presentes;
- PEATE/BERA alterado
- **Complementar PEATE com FE**

PRESENÇA DE MICROFONISMO COCLEAR NO PEATE-CLIQUE: DIAGNÓSTICO DIFERENCIAL ENTRE ESPECTRO DA NEUROPATIA AUDITIVA E PERDAS AUDITIVAS COCLEARES DESCENDENTES EM CRIANÇAS

Presence of cochlear microphonics in click-ABR: differential diagnosis between auditory neuropathy spectrum disorder and steeply sloping cochlear hearing loss in children

Gabriela Ribeiro Ivo Rodrigues [1], **Silvia Nápole Fichino** [2], **Dóris Ruthy Lewis** [3]

RESUMO

Tema: diagnóstico diferencial entre espectro da neuropatia auditiva e perdas auditivas cocleares descendentes em crianças com presença de microfonismo coclear no PEATE-clique. **Procedimentos:** este relato de caso descreve os resultados da avaliação audiológica de duas crianças atendidas no Centro "Audição na Criança" da Divisão de Educação e Reabilitação dos Distúrbios da Comunicação da Pontifícia Universidade Católica de São Paulo (CeAC/DERDIC/PUCSP) que apresentaram microfonismo coclear no registro do PEATE-clique. As crianças foram submetidas às avaliações utilizando-se o PEATE-clique, o registro das emissões otoacústicas e a avaliação audiológica tonal, com a técnica da Audiometria de Reforço Visual. **Resultados:** as avaliações comportamental, eletroacústica e eletrofisiológica revelaram que as crianças apresentam perda auditiva sensorioneural (coclear) com configuração descendente, de modo que a presença do microfonismo coclear no registro do PEATE-clique era provavelmente gerada pela preservação da cóclea nas frequências baixas. **Conclusão:** os casos apresentados mostram que na ausência das emissões otoacústicas e presença do microfonismo coclear, não se deve interpretar isoladamente cada exame, para que não ocorram equívocos no diagnóstico, que pode ser confundido com o Espectro da Neuropatia Auditiva. O microfonismo coclear pode aparecer em outras condições, tais como em perdas auditivas cocleares descendentes.

DESCRITORES: Potenciais Evocados; Perda Auditiva Neurossensorial; Cóclea

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Conflito de interesses: inexistente

INTRODUÇÃO

Os potenciais evocados auditivos de tronco encefálico com o estímulo clique (PEATE-clique) vêm sendo amplamente utilizados na avaliação da função auditiva em neonatos e crianças [1-4]. Na suspeita do Espectro da Neuropatia Auditiva (ENA) os PEATE-clique são realizados com polaridades de estímulo condensado e rarefeito, para pesquisa do microfonismo coclear (MC) [5].

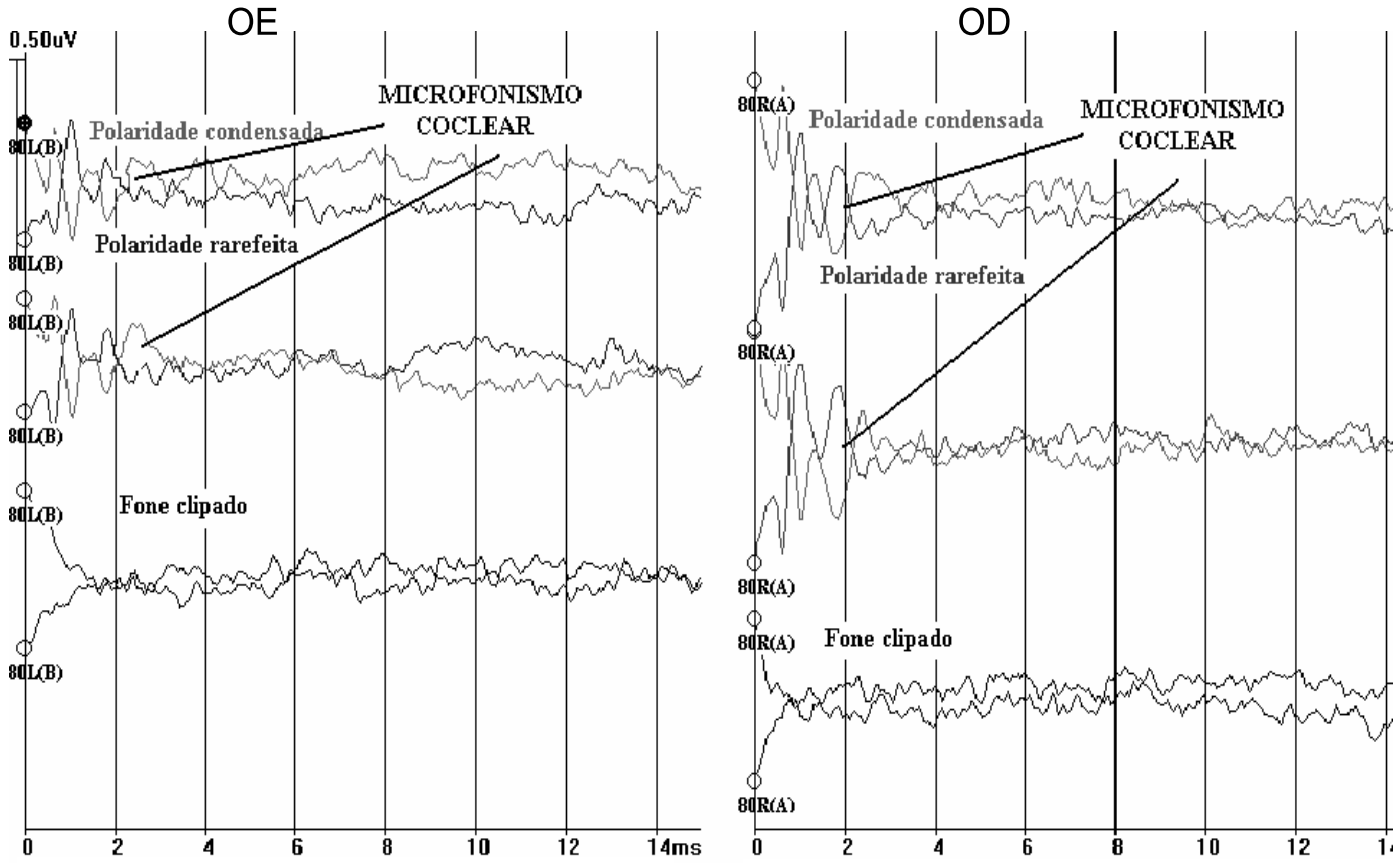

Figura 1 – Resultado do registro do PEATE-clique do caso 1

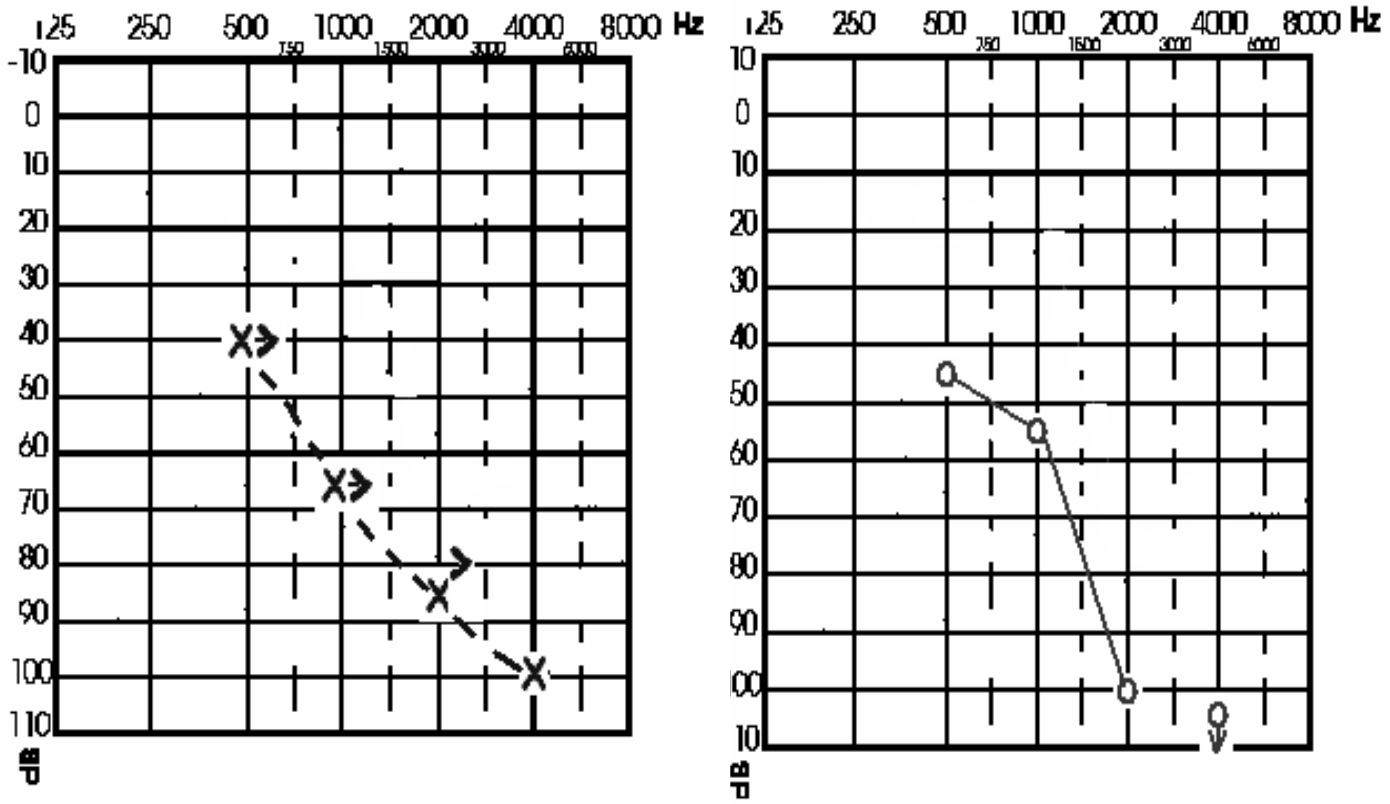

Figura 3 – Audiograma do caso 1

Testes Adicionais

- Pesquisa dos Reflexos Acústicos;
- Ausentes ou alterados;

- Para crianças abaixo de 6 meses de idade, tom teste de 1000Hz;

Testes Adicionais

- Pesquisa da Supressão das EOA;
- Pequena aplicação na clínica audiológica;
- Pode ser um complemeno em indivíduos com EOA presentes;

Supressão EOA

- Pesquisa em 60 dBNPS
- Cliques lineares
- Com e sem presença de ruído (white noise)
- Ruído de 60 dB
- Sistema Olivo-Coclear Medial (OCM)

Avaliações Necessárias

- Avaliação ORL
- Avaliação Audiológica
- Avaliação de Desenvolvimento
- Estudo Genético
- Avaliação Oftalmológica
- Avaliação Neurológica
- Avaliação de Linguagem

Amplificação

- O PEATE/BERA não deve ser o parâmetro para a amplificação;
- Resultados da Audiometria Tonal, Detecção e Reconhecimento de Fala, e Observação do Comportamento Auditivo devem dar suporte aos ajustes da amplificação;
- Monitoramento devido à flutuação;
- Uso do FM pode contribuir;

Ear & Hearing:
June 2002 - Volume 23 - Issue 3 - pp 239-253
Article

Speech Perception and Cortical Event Related Potentials in Children with Auditory Neuropathy

Rance, Gary; Cone-Wesson, Barbara; Wunderlich, Julia; Dowell, Richard

Abstract

Objectives: 1) To investigate the unaided and aided speech perception abilities of children with auditory neuropathy (AN) and to compare their performance to children with sensorineural hearing loss. 2) To establish whether cortical event related potentials (ERPs) could be recorded in children with AN, and to determine the relationship between the presence of these responses and speech perception.

Design: Unaided and aided speech perception assessments (PBK words), and cortical-ERP testing was carried out in a group of 18 children with AN. Data also were obtained from a cohort of age and hearing level matched children with sensorineural hearing loss.

Results: The speech perception performance of the 15 children with AN able to complete a PBK-word assessment, fell into two distinct categories. The children either showed no open-set speech perception ability (7/15 cases), or performance levels similar to their sensorineural counterparts (8/15 cases). Approximately 50% of children with AN showed ERPs of normal latency, amplitude and morphology. In all cases, response presence (at normal latencies) was consistent with reasonable speech perception ability, and response absence was consistent with negligible speech perception.

Conclusions: In approximately 50% of children with auditory neuropathy, the provision of amplification results in significant open-set speech perception improvements. The results confirm the previously published reports that speech perception ability cannot be reliably estimated from the behavioral audiogram in children with AN. Obligatory ERP test results may offer a means of predicting perceptual skills in newly diagnosed youngsters as the presence of ERPs (with age-appropriate latency and morphology) was correlated with significant open set speech perception abilities and amplification benefit. The absence of the ERP in contrast, indicated profound hearing disability evidenced by profound hearing loss and/or extremely poor speech perception.

2002, Ear & Hearing

- **18 crianças – ENA**
- **Habilidades de percepção da fala com sem AASI;**
- **Cortical Event Related Potentials (ERPs) em ENA;**

- **50% das crianças – ASSI e ERPs**

The Laryngoscope
Lippincott Williams & Wilkins, Philadelphia

1999, 1 criança IC

Cochlear Implantation in Auditory Neuropathy

Richard T. Miyamoto, MD; Karen Iler Kirk, PhD; Julia Renshaw, MA; Debra Hussain, MA

Objective: **Auditory neuropathy is a recently described clinical entity characterized by sensorineural hearing loss in which the auditory evoked potential (ABR) is absent but otoacoustic emissions are present. This suggests a central locus for the associated hearing loss. In this study the results observed in a child with auditory neuropathy who received a cochlear implant are presented and compared with those of a matched group of children who were recipients of implants.** ***Methods:*** **A single-subject, repeated-measures design, evaluating closed-set and open-set word recognition abilities was used to assess the subject and a control group of matched children with implants who had also experienced a progressive sensorineural hearing loss.** ***Results:*** **The subject demonstrated improvements in**

tailed accounting of the clinical course of a child with Friedreich's ataxia who received a Nucleus 22-channel cochlear implant (Cochlear Corp., Englewood, CO) and compare his progress to a matched group of pediatric cochlear implant recipients who experienced progressive sensorineural hearing loss resulting in profound hearing loss.

METHODS

Subject

J.G., a 4-year old boy was refe
because of progressive hearing loss a
periencing progressive visual loss. Ir
a mild hearing loss in the right ear

The Laryngoscope
Lippincott Williams & Wilkins, Inc., Philadelphia

2001, 5 crianças IC

Cochlear Implants in Five Cases of Auditory Neuropathy: Postoperative Findings and Progress

Jon K. Shallop, PhD; Ann Peterson, MA; George W. Facer, MD; Lee B. Fabry, MA; Colin L. W. Driscoll, MD

Objectives: **To review our experiences with some of the preoperative and postoperative findings in five children who were diagnosed with auditory neuropathy and were provided with cochlear implants. We describe changes in auditory function, which enabled these children to have significant improvement in their hearing and communication skills.** ***Study Design:*** **Pre- and postoperatively, these children received complete medical examinations at Mayo Clinic, including related consultations in audiology, pediatrics, neurology, medical genetics, otolaryngology, psychology, speech pathology, and radiology.** ***Methods:*** **These children typically had additional medical and audiological examinations at more than one medical center. The hearing assessments of these children included appropriate behavioral au-**

Mayo Clinic Rochester have not had any postoperative medical or cochlear implant device complications. All of the children have shown significant improvements in their sound detection, speech perception abilities and communication skills. All of the children have shown evidence of good NRT results. All but case D (who was not tested) showed evidence of good postoperative EABR results. Otoacoustic emissions typically remained in the non-operated ear but, as expected, they are now absent in the operated ear. ***Conclusion:*** **Our experiences with cochlear implantation for children diagnosed with auditory neuropathy have been very positive. The five children we have implanted have not had any complications postoperatively, and each child has shown improved listening and communication skills**

Ear & Hearing:
August 2006 - Volume 27 - Issue 4 - pp 399-408
doi: 10.1097/01.aud.0000224100.30525.ab
Research Articles

Auditory Neuropathy Characteristics in Children with Cochlear Nerve Deficiency

Buchman, Craig A.; Roush, Patricia A.; Teagle, Holly F. B.; Brown, Carolyn J.; Zdanski, Carlton J.; Grose, John H.

Abstract

Objective: To describe a group of children exhibiting electrophysiologic responses characteristic of auditory neuropathy (AN) who were subsequently identified as having absent or small cochlear nerves (i.e., cochlear nerve deficiency).

Design: A retrospective review of the clinical records, audiological testing results, and magnetic resonance imaging (MRI) studies. Fifty-one of 65 children with AN characteristics on auditory brain stem response (ABR) testing had MRI available for review. Nine (18%) of these 51 children with ABR characteristic of AN have been identified as having small (*N* = 2; 4%) or absent (*N* = 7; 14%) cochlear nerves on MRI.

Results: Of the nine children with cochlear nerve deficiency, five (56%) were affected unilaterally and four (44%) bilaterally. Eight of nine presented after failing a newborn infant hearing screening, whereas one presented at 3 yr of age. On diagnostic ABR testing, all 9 children (9 of 13 affected ears; 69%) had evidence of a cochlear microphonic (CM) and absent neural responses in at least one ear. In the unilateral cases, AN characteristics were detected in all affected ears. In bilateral cases, at least one of the ears in each child demonstrated the AN phenotype, whereas the contralateral ear had no CM identified. Only one ear with cochlear nerve deficiency had present otoacoustic emissions as measured by distortion-product otoacoustic emissions. In children with appropriate available behavioral testing results, all ears without cochlear nerves were identified as having a profound hearing loss. Only 4 (31%) of the 13 ears with cochlear nerve deficiency had a small internal auditory canal on MRI.

Conclusions: Children with cochlear nerve deficiency can present with electrophysiologic evidence of AN. These children frequently refer on newborn screening examinations that use ABR-based testing methods. Similar to other causes of AN, diagnostic ABR testing will show a CM with absent neural responses. Given that 9 (18%) of 51 children with available MRI and electrophysiologic characteristics of AN in our program have been identified as having cochlear nerve deficiency makes this a relatively common diagnosis. These findings suggest that MRI is indicated for all children diagnosed with AN. Moreover, electrophysiologic evidence of unilateral AN in association with a profound hearing loss should make the clinician highly suspicious for this problem. Although children with cochlear nerve deficiency who have a small nerve may benefit from cochlear implantation or amplification, these interventions are obviously contraindicated in children with completely absent cochlear

2006,

◆ AUSÊNCIA OU DEFICIÊNCIA NO NERVO COCLEAR

UNIVERSIDADE DE SÃO PAULO
INSTITUTO DE PSICOLOGIA

ANA CLAUDIA DE FREITAS MARTINHO

Neuropatia Auditiva/Dessincronia Auditiva em crianças usuárias de implante coclear

2007

informa
healthcare

Original Article

International Journal of Audiology 2010; 49: 30–43

Multi-site diagnosis and management of 260 patients with Auditory Neuropathy/ Dys-synchrony (Auditory Neuropathy Spectrum Disorder*)

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Key Words

Auditory neuropathy
Auditory dys-synchrony
Auditory neuropathy spectrum disorder

Abstract

Test results and management data are summarized for 260 patients with diagnoses of Auditory Neuropathy Spectrum Disorder (ANSD). Hearing aids were tried in 85 of these patients, and 49 patients tried cochlear implants. Approximately 15% reported some benefit from hearing aids for language learning, while improvement in speech comprehension and language acquisition was reported in 85% of patients who were implanted. Approximately 5% (13/260) of the total population developed normal speech and language without intervention. Patients were diagnosed at our laboratory (n=66) or referred from other sites (n=194), and all showed absent/grossly abnormal auditory brainstem responses (ABR), often 'ringing' cochlear microphonics, and the presence or history of otoacoustic emissions. Etiologies and co-existing conditions included genetic (n=41), peripheral neuropathies (n=20), perinatal jaundice and/or anoxia and/or prematurity (n=74). These patients comprise 10% or more of hearing impaired patients; their language acquisition trajectories are generally unpredictable from their audiograms.

Sumario

Se resumen los resultados de las pruebas y los datos del tratamiento de 260 pacientes con diagnóstico de Espectro de desórdenes de la Neuropatía Auditiva (ANSD). En 85 de estos pacientes se probó el uso de auxiliares auditivos y 49 pacientes recibieron un implante coclear. Aproximadamente 15% reportaron algún beneficio con los auxiliares auditivos para la adquisición del lenguaje mientras que el 85% de los que recibieron un implante reportaron una mejoría en la comprensión y la adquisición del lenguaje. Aproximadamente 5% (13/260) de la población total desarrolló lenguaje normal sin intervención. Los paciente fueron diagnosticados en nuestro laboratorio (n=66) o referidos de algún otro lado (n=194) y todos mostraron ausencia o anormalidad importante de los potenciales evocados (ABR), frecuentemente con una microfónica coclear "timbrante" y con presencia o historia de emisiones otoacústicas. La etiología o las condiciones co-existentes incluidas fueron: genéticas (n=41), neuropatías periféricas (n=20), ictericia perinatal y/o anoxia y/o prematurez (n=74). Estos pacientes representan 10% o más de los pacientes con hipoacusia; su trayectoria en el proceso de adquisición del lenguaje es generalmente impredecible a partir de sus audiogramas.

- **260 PACIENTES;**
- **85 AASI**
- **49 IC**
- **5% AUDIÇÃO NORMAL**
- **15% RELATARAM BENEFÍCIOS COM AASI**
- **85% RELATARAM BENEFÍCIOS NA**
- **PERCEPÇÃO DA FALA COM IC**

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Speech perception and cortical auditory evoked potentials in cochlear implant users with auditory neuropathy spectrum disorders

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ABSTRACT

Objective: To characterize the P_1 component of long latency auditory evoked potentials (LLAEPs) in cochlear implant users with auditory neuropathy spectrum disorder (ANSD) and determine firstly whether they correlate with speech perception performance and secondly whether they correlate with other variables related to cochlear implant use.
Methods: This study was conducted at the Center for Audiological Research at the University of São Paulo. The sample included 14 pediatric (4–11 years of age) cochlear implant users with ANSD, of both sexes, with profound prelingual hearing loss. Patients with hypoplasia or agenesis of the auditory nerve were excluded from the study. LLAEPs produced in response to speech stimuli were recorded using a Smart EP USB Jr. system. The subjects' speech perception was evaluated using tests 5 and 6 of the Glendonald Auditory Screening Procedure (GASP).
Results: The P_1 component was detected in 12/14 (85.7%) children with ANSD. Latency of the P_1 component correlated with duration of sensorial hearing deprivation ($^*p = 0.007$, $r = 0.7278$), but not with duration of cochlear implant use. An analysis of groups assigned according to GASP performance (*k*-means clustering) revealed that aspects of prior central auditory system development reflected in the P_1 component are related to behavioral auditory skills.
Conclusions: In children with ANSD using cochlear implants, the P_1 component can serve as a marker of central auditory cortical development and a predictor of the implanted child's speech perception performance.

TANU

- Protocolos diferenciados para neonatos com e sem IRDA;
- EOA para crianças sem IRDA;
- PEATE/BERA para crianças com IRDA;
- IRDA – Maior probabiidade de ENA
- Reinternação – nova triagem

OBRIGADA!!

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