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Computerized auditory training in an adolescent with neurofibromatosis type 2

Treinamento auditivo computadorizado em uma adolescente portadora de neurofibromatose do tipo 2

Entrenamiento auditivo computarizado en una adolescente portadora de neurofibromatosis tipo 2

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Abstract

Neurofibromatosis type 2 is a genetic disease that affects the Central Nervous System, mainly the auditory pathway, being some of the main symptoms: sensorineural hearing loss, low speech recognition, tinnitus and vestibular dysfunction. This study describes a process of therapeutic intervention through Computerized Auditory Training in an adolescent with medical diagnosis of Neurofibromatosis type 2. This intervention was composed of 16 sessions of AT with a specific software. For this study, the pre and

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post-training evaluation was carried out through the results of basic audiological evaluation, Brainstem Evoked Response Audiometry and evaluation of phonological awareness, as well as analyzing the patient's reports regarding the therapeutic process. The auditory processing behavioral tests were not performed, since the patient presents difficulty in speech comprehension. Patient presented improvement in speech recognition and phonological awareness ability, although the electrophysiological test did not demonstrate such benefits directly in the auditory pathway at the brainstem level, for the click stimulus. Furthermore, the patient's report of improvement in speech comprehension after such intervention could also be highlighted. Thus, the positive effects of auditory training on the improvement of auditory perception could be observed, influencing positively the daily life of the patient.

Keywords: Schwannoma; Auditory Perception; Audiology; Speech Language Pathology and Audiology Therapy; Quality of life; Neurofibromatosis 2.

Resumo

A neurofibromatose do tipo 2 é uma doença genética que afeta o Sistema Nervoso Central, principalmente a via auditiva, sendo alguns dos principais sintomas a perda auditiva neurossensorial, o baixo reconhecimento de fala, zumbido e disfunção vestibular. Relata-se neste estudo um processo de intervenção terapêutica por meio do Treinamento Auditivo Computadorizado em uma adolescente com diagnóstico médico de Neurofibromatose tipo 2. Tal intervenção contou com 16 sessões de TA, realizadas com um software específico. Para o estudo realizou-se a avaliação, pré e pós-treinamento, por meio dos resultados da avaliação audiológica básica, Potencial Evocado Auditivo de Tronco Encefálico e avaliação da consciência fonológica, além de analisar os relatos referentes ao processo terapêutico. Os testes comportamentais de processamento auditivo não foram realizados, pois a paciente apresenta acentuada dificuldade de compreensão de fala. Paciente apresentou melhora no reconhecimento de fala e na habilidade de consciência fonológica, apesar do exame eletrofisiológico da audição não demonstrar tais benefícios diretamente na via auditiva em nível de tronco encefálico, para o estímulo clique. Destaca-se o relato da paciente de melhora na compreensão de fala após tal intervenção. Desse modo, observaram-se os efeitos positivos do treinamento auditivo na melhora da percepção auditiva, influenciando positivamente na vida diária da paciente.

Palavras-chave: Schwannoma; Percepção Auditiva; Audiologia; Fonoaudiologia; Qualidade de vida; Neurofibromatose 2.

Resumen

La neurofibromatosis del tipo 2 es una enfermedad genética que afecta el Sistema Nervioso Central, principalmente la vía auditiva,siendo algunos de los principales síntomas la pérdida auditiva neurosensorial, el bajo reconocimiento de habla, zumbido e disfunción vestibular. En este estudio se relató un proceso de intervención terapéutica por medio de Entrenamiento Auditivo Computarizado en una adolescente diagnosticada con Neurofibromatosis tipo 2. La intervención contó con 16 sesiones de EA, realizadas con un software específico. Para el estudio se realizó la evaluación antes y después del entrenamiento, atreves de los resultados de la evaluación audiológica básica, Potenciales Evocados Auditivos del Tronco Encefálico y evaluación de la conciencia fonológica, además de analizar los relatos relacionados al proceso terapéutico. Los testes comportamentales de procesamiento auditivo no fueron realizados, pues la paciente presenta acentuada dificultad de comprensión de habla. La paciente presentó mejora en el reconocimiento de habla y en la habilidad de conciencia fonológica, por más que el examen electrofisiológico de la audición no tenga demostrado esos beneficios directamente en la vía auditiva en nivel de tronco encefálico, para el estímulo clic. Se destaca el relato de mejorapor la paciente en la comprensión de habla tras la intervención. De esa manera, se observaron los efectos positivos del entrenamiento auditivo en la mejora de la percepción auditiva, influyendo positivamente en la vida diaria de la paciente.

Palabras claves: Schwannoma; Percepción Auditiva; Audiología; Fonoaudiología; Calidad de vida; Neurofibromatosis 2.

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Introduction

Neurofibromatosis (NF) is a genetic disease, whose manifestation and severity vary in each patient, with multiple clinical manifestations, the main symptoms being: sensorineural hearing loss; low speech recognition and understanding, especially in noisy environments; tinnitus and vestibular dysfunction[1,2]. It differs from Meniere's disease because it presents continuous symptoms and not only in crises. This genetic disease can be classified as NF type 1 or type 2, and the latter affects the central nervous system and is characterized by the presence of the Schwannoma of the VIII cranial pair, usually bilateral[3].

The VIII Schwannoma is thus named for proliferating in Schwann cells, and is also known as acoustic neurinoma. It affects the cerebellar angle, which is located in the posterior cerebellar fossa, where portions of the cranial nerves V, VII, VIII, IX, X and XI[1] are found. It is characterized by being a benign, slow-growing tumor that affects the internal auditory canal, evolving into the pontine cistern. In more advanced stages, it can compress the cerebellum and even the brainstem[3].

The diagnosis of this pathology is carried out through electrophysiological and behavioral audiological tests - such as tone-pure threshold audiometry, percentage speech recognition index, stapedian acoustic reflexes and brainstem auditory evoked potentials - in addition to vestibular exams such as Electronystagmography, imaging tests, such as the magnetic resonance of the skull and computed tomography of the temporal bone, indicated by the doctor in charge[3].

Concerning hearing, there are often discrepant values between the degree of hearing loss and the speech recognition, as well as a decrease in the abilities of auditory perception for speech sounds and their acoustic particularities, causing limitations in relation to social, cultural and emotional. It is also possible to find cases of sudden hearing loss or auricular fullness associated with floating loss, beyond the alterations already mentioned in the electrophysiological tests of hearing[3].

The treatment should consider the time of tumor diagnosis and the particularities of each case - such as size and location, for example -, and surgery is still a very useful resource for cases of Schwannoma in VIII pair[3].

Both types of NF may be associated with changes in auditory abilities, especially speech comprehension, and auditory processing disorder (APD) that can result from a post partum event (neurological trauma or infections) [4,5].

For this disorder, auditory training (AT) may be a therapeutic resource to promote changes in neural bases and auditory behavior, favoring brain plasticity and thus, improvement in the hearing abilities of the subjects[6].

Regarding the low speech recognition of the subjects diagnosed with Schwannoma, and the difficulties in processing auditory information due to impairment in the VIII pair, it was hypothesized that auditory training could be indicated in a subject with Neurofibromatosis type 2, aiding in the treatment and especially in speech comprehension, both in the pre and post operative periods.

Therefore, the objective of this study was to describe the effects of computerized auditory training in an adolescent diagnosed with Neurofibromatosis type 2.

Case report

Case study developed at the Audiology and Auditory Processing Outpatient Clinic of a Higher Education Institution, from March to June 2016. The subject of this research signed the Free and Informed Consent Term (FICT), respecting the ethical issues proposed by the Resolution 466/2012 of the National Health Council. The study was approved by the Ethics Committee in Research of the mentioned institution, under the number 43171715.0.0000.5346.

An 18-year-old female diagnosed with Neurofibromatosis type 2 participated of this study. The doctor in charge requested Pure tone threshold audiometry, Immittanciometry and Brainstem Auditory Evoked Potential (BAEP) tests, since the patient present complaints of hearing loss and difficulty in understanding speech.

A Basic Audiological Assessment was performed, which involved visual inspection o external auditory meatus, in order to verify the presence of some external ear impairment with the Standard Mark II otoscope. Pure tone threshold audiometry to determine auditory thresholds by air conduction in the frequencies from 0.25 to 8 kHz and by bone conduction in the frequencies from 0.5 to 4 kHz, in an acoustic booth (ANSI S3.1-1991 level of environmental noise). In order to deter-

mine the Speech Recognition Threshold (SRT) and Speech Recognition Percentage Index (SRI) were used dissyllable words due to the patient's low understanding in monosyllabic words. These procedures were performed in the Interacoustics audiometer, model AD229. Immittanciometry, composed of tympanometry and investigation of contralateral acoustic reflexes, was also performed in the frequencies of 500, 1000, 2000 and 4000 Hz, to evaluate the integrity of the middle ear, as well as the central verification through the reflex arc, with the Interacoustics equipment, model AT235. Classification of auditory thresholds was used by the World Health Organization (1997).

The results of the audiological pre-training corroborated the patient's complaint, evidencing mild sensorineural hearing loss, type A tympanometric curve and absence of contralateral acoustic reflexes of both ears. In the logoaudiometry, only 12% of SRI was observed, bilaterally, at adequate and comfortable intensity.

Brainstem Auditory Evoked Potential (BAEP) was also performed in order to evaluate the integrity of the auditory pathway at the Brainstem level. For the Record of the BAEP, the Smart-EP module of the brand Intelligent Hearing Systems®, two channels was used. Adolescent skin hygiene was performed with NUPREP® brand with gauze and abrasive paste to allow impedance of the electrodes with the skin. The four electrodes were fixed with electrolytic conductive paste, the active electrode placed on the forehead (Fz), the ground (Fpz) laterally on the forehead, and the reference electrodes on the left mastoid (T3) and right mastoid (T4), according to the International 10/20 standard. Insertion heads were used to present the stimuli and the impedance of the electrodes was analyzed before the start of the examination (below 3 ohms). The BAEP record was monoaural; it was initiated with the click stimulus at the intensity of 80 dB nHL. Then, the responses were recorded at the intensities of 90 and 99 dB nHL. The plot was reproducible to analyze the presence/absence of the waves. Regarding the path acquisition parameters, the 2048 click stimuli, of 100 µs duration, with rarefied polarity were used, the stimulus presentation rate was 27.7 stimuli per second, 100-3000Hz filters were used and a maximum of 10% of artifacts were accepted. The absence of waves I, III and V was observed in the maximum intensity of the equipment, right and left.

After analyzing the results, the patient was referred to therapy at the Auditory Training Laboratory of the institution in question. Due to the performance presented in the speech recognition test, it was not possible to perform the auditory processing behavioral battery. However, knowing the benefits of auditory training, such as the improvement of neuronal plasticity, auditory abilities and consequently of auditory behaviors, this model of therapeutic intervention was chosen, aiming at minimizing the complaints and difficulties reported in the anamnesis, in a pre operative period[5,6]. The medical conduct was also the referral for the surgery of removal of the neurinoma and tumors, because it was a public hospital, the subject in question waited for the procedure. Thus, performing AT was a way of providing an immediate therapeutic approach.

Before starting the therapy itself, a phonological awareness test was performed to evaluate the subject's ability to operate the language as an object through phonological processing at the syllabic and phonemic levels. The results were analyzed by sum of the points in each level, which showed 53 hits in a total of 70 possibilities, 34 in the syllabic level and 19 in the phonemic level.

After this step, the computerized auditory training protocol was followed, using the Active Listening software[7], taking into account the individual characteristics of the patient. This program consists of 12 tasks with the aim of improving auditory processing skills, being binaural interaction, figure-ground, temporal resolution, temporal standard, discrimination, binaural integration and separation. At the start of the program, a registration and calibration is performed, in order to meet the individual characteristics of the patients. Therapy sessions follow a hierarchy of tasks, working different listening skills at different levels of complexity each session, with the possibility of inserting competitive noise in order to make training difficult. There were 16 sessions, twice a week, with 30 minutes each. From these, 12 sessions were assigned the software tasks, and the other four sessions were assigned to the specific abilities provided by the software - binaural interaction, figure-ground and auditory discrimination - in which the patient presented greater difficulty[7]. The Sony MDR-ZX100 supra-headset was used. One activity per session was performed according to the following order of presentation[8] (Table 1)

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Table 1. Proposed activities, stimulated auditory ability, gnostic process involved and brief explanation of tasks

Task	Stimulated auditory ability	Gnostic process	Function
Follow the flute	Temporal organization (Duration pattern) Attention and Memory	Non verbal gnosis	Long and short sounds were reported and the subject was asked to reproduce the same listening sequence. Easy and medium levels with three sounds presentation, difficult with four sounds and insane with five sounds.
Follow the piano	Temporal pattern (Frequency pattern) Attention and Memory	Non verbal gnosis	Sounds of different intensities (low and high pitch) were presented and the subject was asked to reproduce the same sequence heard. Easy and medium levels with three sounds presentation, difficult with four sounds and insane with five sounds.
How many intervals	Temporal resolution Attention	Decoding	Sounds and intervals were presented and, in this activity, the subject, whenever he realized the interval, had to click on a number or, at the end of the sequence, only on the number corresponding to the total of the intervals listened to.
How many sounds	Temporal organization Attention	Non verbal gnosis	Sounds of musical instruments were presented in different sequences and the subject was asked to say how many times he heard the sound.
What sound heard	Detection and discrimination Attention	Ordination	Two verbal sounds and a question regarding what was heard were presented (the words were the same or different).
Follow the sequence	Association Auditory memory for non verbal sounds Attention	Organization	Sounds of animals were presented. The subject should memorize and organize them in the order they requested. Easy and medium levels: alphabetize the names of the animals heard, reverse those names, say only the third ear, etc. For this activity, the subject had visual aid of an alphabet trail. Difficult and insane levels: a story was heard and, at the end, a question was asked that the subject should interpret.
Hearing and attention	Recognition Hearing closure Attention	Decoding	Two words were presented in an auditory and written form and the subject had to respond as requested in the statement (Do the words rhyme? Do words begin with the same sound? Do words have the same number of syllables? Etc). Difficult and insane levels: the written and listened words were different.
Hit the bull's-eye.	Binaural separation Attention	Coding	Two words were presented at the same time, one in each ear, and the subject was asked to identify the target word and indicate which side was presented. The target word could be said before or after the presentation.
Left Right	Binaural integration	Coding	Words were presented, both on one side and now on the other, and the subject had to identify the words and from which side each one came out. Easy level: one word on each side, medium level: two, difficult level: three and insane: four.
Binaural	Localization Attention Memory	Decoding	Sounds of musical instruments were presented, sometimes on one side, sometimes on the other, far, near, and the subject had to identify from which location each sound came out. Easy and medium levels: the answer could be given while the stimuli were being heard. Medium and insane levels: the response was given after the presentation of the sequence, requesting memorization.

Task	Stimulated auditory ability	Gnostic process	Function
Take IF you can (Bonus track)	Attention	Coding	A tray of fruit was presented and the subject should click only on the apple that was moving. Easy and medium levels: only apple on the tray changing speed or changing size. Difficult level: other fruits were added and the child should only get the apple. Insane level: different fruits on the tray and customers' requests were heard; as requested by the statement, the subject should click on the fruit that was blinking or the fruit auditively requested
Follow the rhythm (Bonus track)	Attention	Non verbal gnosis	Different musical rhythms were presented and the subject could choose which one he wanted to play; should listen to the music and click on the colored cards that were falling.

Source Melo et al, 2016.

Afterwards the intervention period, all exams applied in the evaluation were reapplied in order to analyze the effect of auditory training. A SRI of 32% to the right and 28% to the left was observed in logoaudiometry. In the BAEP, the absence of waves I, III and V was observed. In the evaluation of phonological awareness, a score of 63 points was obtained for a total of 70, being 38 for the syllabic level and 29 for the phonemic level.

Discussion

It is known the importance of medical findings and the poor prognosis regarding hearing preservation, after neuromuscular removal in many cases of type 2 neurofibromatosis. Especially, the focus of this case report was to describe the effects of computerized AT on an adolescent diagnosed with this pathology, so the AT was a palliative therapeutic approach, prior to the surgical intervention, and try to minimize the patient's complaints regarding speech comprehension more quickly.

The findings of this study corroborate with the literature when claiming that auditory training is effective in improving auditory perception, since it involves a set of tasks related to language, cognition and auditory abilities [9,10]. These are adressed to the activation of the auditory system and associated systems, in order to improve the auditory functions originated by the central nervous system, especially in the base etiology of this study, which compromises the elemental structures for sound processing at central level as the auditory nerve[11].

Only one study was carried out in the literature that involved AT and NF[11], in which eight sessions of Acustically Controlled Auditory Training were performed once a week, lasting 50 minutes each in patients with NF1 and APD. The authors concluded that such intervention was effective for the improvement of the auditory closure abilities, figure-ground and temporal resolution, including the benefits obtained after the AT remained after one year of its end in the study population. Although it was performed in another type of NF, it is possible to point out that such data corroborate with those of this study, since after the intervention, evolution was observed in all the findings; in the SRI the patient presented improvement in speech recognition after training auditory perception, demonstrating an improvement in the perception/discrimination of speech sounds. Studies show that such intervention is capable of improving figure-ground and auditory closure skills [11,12]. It is known that the auditory skills required for the SRI test is the same for such cited abilities, in this way, the same mechanisms of processing of these stimuli are used, asserting the findings of this study due to these data.

Likewise, in relation to phonological awareness, the patient presented a higher performance in relation to pre-training, being more evident at the phonemic level. This suggests that the skills in manipulating and operating the speech sounds were improved, demonstrating that the patient was more attentively listening, especially to the requests of the researcher. These findings corroborate with the literature when affirming that phonological awareness is sensitive to AT, since the discrimination of syllables and phonemes is constantly stimulated, as

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well as the structure of the sounds of the language, favoring phonological processing[9].

The absence of waves in the BAEP suggests that the stimuli presented were not effective in driving and triggering responses of the central structures evaluated by this potential, with click stimulus. This fact suggests the electrophysiological changes were not influenced by the number of sessions proposed, that is, the intervention period was not enough to promote a significant neuronal plasticity in the register of this potential. The data agree with the findings of another study, which analyzed the Long-Latency Auditory Evoked Potential (LLEP) in the elderly after such intervention[10]. On the other hand, it diverges from another study that reveals better results, referring to the latency and amplitude of the waves evaluated, for both the BAEP and the LLEP, and the latter was not evaluated in the present study[13]. At the time of the care of this patient, LLEP was not available, so a possible limitation of the present study is pointed out here.

Another hypothesis that justifies the absence of waves in the BAEP can only be due to the underlying etiology, since this by promoting damage to the central structures, such as the VIII cranial nerve - which is evaluated by wave I of this potential - does not allow the response and stimulation to the higher brainstem structures, such as the superior olivary complex and the lateral lemniscus, evaluated respectively by waves III and V[13]. It is then assumed that with the surgical intervention to remove the tumor, there could be the presence of waves, even though these were not morphologically adequate.

Despite the subjectivity, the patient's report endorses what the findings of this study revealed; the intervention was significant for her daily life activities, since there was improvement in speech comprehension, allowing a higher quality of life before surgical intervention. Furthermore, the patient reported interest in continuing the sessions after the removal of the tumors, because she realized that the AT brought benefits to her communication and consequently quality of life. The issue of quality of life in patients with NF2[14] has already been pointed out with some concern, as the significant clinical symptoms of this pathology bring important reflexes to their lives.

It should be emphasized that such intervention was complementary to the medical treatment and in this specific case, it was an alternative for the rehabilitation of the communication, while the adolescent studied awaited the surgical procedure previously indicated by the doctor in charge.

Other approaches to intervention in cases of Neurofibromatosis type 2 are cited in the literature, such as the Brainstem Auditory Implant[15] after tumors removal surgery, when there is a great loss of hearing and the use of new effective drug therapies for patients [16,17] with NF2, which function as a vascular endothelial growth inhibitor reducing schwannoma growth and tumors in some patients. However, this study hypothesized that auditory training would have a positive effect on patient communication and this materialized, evidencing the importance of new studies with more robust sample arrangements to prove this hypothesis.

Conclusion

Positive effects of auditory training were found in this case of NF2, since it allowed an improvement in the auditory perception capable of influencing the subject's quality of life. It is also suggested that studies are carried out in this same perspective, whereas no specific research with auditory training in this pathology was found in the databases.

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