



Proposal of application of alternative communication for patient with neurodegenerative disease and its impact on quality of life: case report

Proposta de aplicação da comunicação alternativa para paciente com doença neurodegenerativa e seu impacto na qualidade vida: relato de caso

Propuesta de aplicación de comunicación alternativa para paciente com enfermedad neurodegenerativa y su impacto en la calidad de vida: caso clínico

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Abstract

This is a case report of a 51-year-old male, with complete college education, favorable socioeconomic status, diagnosed in 1999 with Friedreich's Ataxia. He arrives at the speech therapy clinic, with an emphasis on assisting adults with degenerative diseases, under the guidance of the genetics team at the service of the same hospital. Speech examination examines moderate to severe oropharyngeal dysphagia and severe dysarthria. Dysphagia is rehabilitated via private home care at the patient's option and in the outpatient clinic with the objective of improving quality of life, a proposal for the application of augmentative and/or alternative communication was created to develop the communication skills of the

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patient who was no longer expressing himself. Two evaluations (pre and post therapy) and four therapeutic intervention sessions were carried out to learn and implement the alternative communication board. At the end of the therapeutic process, there was low adherence to the use of augmentative and / or alternative communication, even with the self-perception of the unintelligibility of his speech, using the folder restricted to speech therapy. Both the patient and his companions reported that even after several attempts there was a denial of the use of alternative communication. Although there were few sessions, there was no impact on the patient's quality of life after using augmentative and/or alternative communication.

Keywords: Friedreich's ataxia; Dysarthria; Adult, Communication

Resumo

Trata-se de um relato de caso de um indivíduo do sexo masculino com 51 anos, nível superior completo, nível socioeconômico favorável, diagnosticado em 1999 com Ataxia de Friedreich. Chega ao ambulatório de Fonoaudiologia, com ênfase no atendimento de adultos com doenças degenerativas, sob encaminhamento da equipe de genética do serviço do mesmo hospital. Ao exame fonoaudiológico diagnostica-se uma disfagia orofaríngea de moderada a grave e uma disartria grave. A disfagia é reabilitada via *home care* particular por opção do paciente, e no ambulatório, com objetivo de melhora da qualidade de vida criou-se uma proposta de aplicação da comunicação aumentativa e/ou alternativa para o desenvolvimento das habilidades de comunicação do paciente que já não estava mais se expressando. Foram realizadas duas avaliações (pré e pós terapia) e quatro sessões de intervenção terapêutica para o aprendizado e implementação da prancha de comunicação alternativa. Ao término do processo terapêutico verificou-se baixa adesão ao uso da comunicação aumentativa e/ou alternativa, mesmo com a auto-percepção da ininteligibilidade da sua fala, utilizando a pasta restrita ao atendimento fonoaudiológico. Tanto o paciente quanto seus acompanhantes referiram que mesmo após várias tentativas houve negação ao uso da comunicação alternativa. Embora tenham sido poucas sessões, não houve impacto da qualidade de vida do paciente após uso da comunicação aumentativa e/ou alternativa.

Palavras-chave: Ataxia de Friedreich; Disartria; Adulto, Comunicação

Resumen

Este es un informe del caso de un hombre de 51 años, con educación universitaria completa, estatus socioeconómico favorable, diagnosticado en 1999 con ataxia de Friedreich. Llega a la clínica de terapia del habla, con énfasis en ayudar a adultos con enfermedades degenerativas, bajo la guía del equipo de genética al servicio del mismo hospital. El examen notas la disfagia orofaríngea moderada a severa y la disartria severa. La disfagia se rehabilita mediante atención domiciliaria privada a elección del paciente y en la clínica ambulatoria con el objetivo de mejorar la calidad de vida, se creó una propuesta para la aplicación de comunicación aumentativa y/o alternativa para desarrollar las habilidades de comunicación del paciente que ya no era más expresándose a sí mismos. Se realizaron dos evaluaciones (pre y post terapia) y cuatro sesiones de intervención terapéutica para aprender e implementar el tablero de comunicación alternativo. Al final del proceso terapéutico, hubo una baja adherencia al uso de comunicación aumentativa y/o alternativa, incluso con la autopercepción de la ininteligibilidad de su discurso, usando la carpeta restringida a la terapia del habla. Tanto el paciente como sus compañeros informaron que incluso después de varios intentos hubo una negación del uso de comunicación alternativa. Aunque hubo pocas sesiones, no hubo impacto en la calidad de vida del paciente después de usar comunicación aumentativa y/o alternativa.

Palabras clave: Ataxia de Friedreich; Disartria; Adulto, Comunicación

Introduction

Friedreich's ataxia (FRDA) is a pathology with an autosomal recessive, progressive mode of inheritance, with an inability to walk autonomously 10 to 20 years after the onset of symptoms. It appears classically around puberty, rarely in early childhood and, in some cases, later¹. It is multisystemic and the main neurological characteristics include difficulty in progressive gait, limb ataxia, weakness, instability of ocular fixation and sensory loss and deep reflex, in relation to endocrinology, from 20 to 40% have diabetes mellitus; in cardiologic patients, 66% have ventricular hypertrophy, which causes dilated cardiomyopathy and cardiac conduction disorders. In addition, non-motor symptoms are quite common, among these, depression, fatigue and mood disorders^{2,3}.

Dysarthria is one of the main clinical signs, they may be absent initially and show greater progression after loss of ambulation, although diadochokinesis becomes noticeable early, due to pyramidal degeneration, causing slowness more than irregular movement, starting to impair intelligibility^{4,5}.

The FRDA speech and voice phenotype has been characterized by reduced intelligibility resulting from altered prosody and severe articulatory inaccuracy⁶, as well as altered nasality (predominantly hypernasality)⁷. The detailed auditory perceptual analysis showed that individuals with FRDA have difficulty in controlling pitch and loudness, have increased sentence time, production of inaccurate consonants and high tension phonation⁸. No evidence was found in the indexed literature on any treatment to improve speech in cases of FRDA⁹. In these cases, it is known that there are great difficulties in daily activities and in communication skills, leading to low quality of life and social isolation¹⁰.

Neurodegenerative diseases progressively affect the change in the possibility of oral communication by individuals², and consequently, in quality of life. Communicative aspects, food, physical appearance, strategy to cope with social symptoms, need adequate support¹¹. The evaluation of the quality of life related to the health of these individuals has been increasingly used to provide alternatives that assist this patient within the health-disease process¹².

Augmentative and / or alternative communication (AAC) has been presented as an important area of clinical practice and research, which allows individuals who have limitations or absence of speech, to communicate effectively¹². Bandeira and collaborators (2011)¹² developed a study that implemented the use of AAC boards in 30 adult and elderly patients hospitalized between 20 and 70 years of age, unable to communicate through speech, observed an improvement in quality of life, as well as the importance and the need for communication between patient-staff and patient-family.

The hypothesis of this study is that the difficulty of progressive oral communication could develop and improve the patient's communicative skills, thus improving quality of life. Given this, this work aims to present a proposal for the application of augmentative and / or alternative communication for a patient with neurodegenerative disease, as a way to improve the quality of life.

Case report

This work was approved by the Research Ethics Committee of the responsible institution under n° 2019-0789. The research was carried out between July and August 2019 and was explained to the subject involved (and their guardians), who, after consent, signed the Informed Consent Form.

Four sessions were held in order to apply the CCA proposal and its impact on quality of life. For that, prior to the beginning of the sessions, the WHOQOL - OLD¹³ quality of life questionnaire was applied. The questionnaire consists of 24 questions and their answers vary from 1 to 5, 1 being nothing and 5 extremely, and it is divided into 6 categories (Functioning of the Sensory, Autonomy, Past, Present and Future Activities, Social Participation, Death and Dying and Intimacy). The questionnaire scores from 1 to 100 (best quality of life index), the higher the value, the higher the patient's quality of life. The questionnaire was proposed as a marker of improvement in quality of life before and after intervention.

G.B.T, male, born on 12/20/1967, 51 years old, complete higher education, favorable socioeconomic status, diagnosed in 1999 with Friedreich's ataxia (350 repetitions GAA 9q13-q21.2 intron 1 gene X25). He suffered a traffic accident in 2009 presenting traumatic brain injury with few motor sequelae and without interference with linguistic



functions. The medical clinical examination shows a wheelchair restriction, important postural instability, squarewavejerks, cervical hypotonia with head falling forward, significant atrophy of the interossei, tenar region, hypotenar, strength III-IV distally in the upper limbs (worse than extensor movements); grade III proximally, force abolished on dorsiflexion and bilateral plantar flexion and depression.

He arrives at the speech therapy clinic, with an emphasis on assisting adults with degenerative diseases, under the guidance of the genetics team at the service of the same hospital. Speech-language examination examines moderate to severe oropharyngeal dysphagia and severe dysarthria. G.B.T, accompanied by his mother and caregiver, reports being under speech therapy for swallowing rehabilitation three times a week, but without speech therapy. His mother complains, mainly, about the child's communication difficulties and the decrease in social interaction, in which the patient chose not to maintain verbal contact with people outside the family nucleus due to the communication difficulties. The patient continued with his external activities, such as attending the club and going to restaurants under the care of the caregiver. G.B.T reports progressive worsening in speech with poor intelligibility, emotionally interfering in the relationship between family members. When he arrives at the clinic, the patient has cognition within the normal range for age and education and in cases of need of expressing himself, he uses his cell phone / computer restricting his communication to isolated words and from that moment on, the possibility thought was AAC. Due to the clinical conditions resulting from the patient's basic pathology², and the lack of scientific evidence of speech rehabilitation in these cases⁹, the AAC implementation strategies were discussed with the mother and patient since the patient is still very responsive, but has a great communication restriction.

A proposal for the application of the AAC was planned, within the model of care recommended by the outpatient service of the service in question, with 4 visits being made. The frequency of visits occurred once a week, totaling one month of intervention with the patient.

- Session 1: Assessment and Basic Needs

Due to the changes identified regarding the impact of the difficulty of oral communication in activities of daily living and in interpersonal relationship with family members, a questionnaire on quality of life in the elderly (WHOQOL - OLD)¹³ was applied, with a final score of 59 points. We opted to use a quality of life questionnaire in the elderly because it is a validated questionnaire and at the same time simpler than the WHOQOL, due to the patient's conditions, which although he is not considered to be elderly, may have decreased performance in very long activities. The planning of the construction of the therapeutic board (on paper) was initiated, since, due to the motor difficulty in the use of technologies, a low-tech model was chosen, and it was used through pointing. During the evaluation, all the required occupational strategies and needs were investigated, in addition, the main images to be used for the basic activities of daily living were planned together with the patient, mother and caregiver, including: going to the bathroom, feeling cold, feeling hot, feeling pain, knowing the time, getting ready in the chair, getting up, going to sleep, showering, taking water, feeling hungry, taking the medicine, reaching the cell phone.

- Session 2: Guidelines for using the AAC board

Two clipboards with the previously established images were delivered. During 30 minutes of care, the patient, caregiver and mother talked about the importance of social interaction, communication and how the AAC could happen in a supplementary way, due to the great difficulty of communicative intelligibility. Still in attendance, some simple phrases were built using images and communication skills were explored, and communication between patient and family members was encouraged, showing the situations in which the AAC could facilitate the understanding and wishes of the patient. In care, both the mother and the caregiver were instructed on the use of the boards.

- Session 3: Expanding communicative possibilities

During the therapeutic sessions, the patient's portfolio was expanded, adding more activities of daily living (taking medicine, watching television,

reading the newspaper, going to the bank, going to the mall, going for a walk), his favorite foods and fruits, feelings (happy, hopeful, angry, sad, angry, tired), clothing, colors, animals, some objects, among others. In addition, in all consultations, the importance of communication was resumed for improving the quality of life, the possibility of social interaction, autonomy, adequate care. The potentialities were discussed in view of all the restrictions imposed by the clinical condition, with the intention of enabling an improvement in the patient's quality of life, enabling non-verbal expression, thus reducing social isolation. In addition, the instructions were always passed on to the mother and caregiver about the importance of using the board, encouraging use at home and in everyday situations of social interaction, when they do not understand their wishes showing the folder, always leaving it close, to establish dialogue and encourage use, as well as conversational strategies with storytelling, stories supported by images.

- Session 4: Re-evaluation and application of protocol

At the end of the learning process and therapeutic implementation, the patient's reassessment was also carried out. G.B.T considered his communication very bad, but he used the folder only in speech therapy. Both the patient and his companions reported that even after several attempts to use the board, there was difficulty in accepting the use of the AAC by the patient, although he mentioned benefit. In the service he followed the guidelines, but at home he did not incorporate the use of the folder. WHOQOL - OLD13 was reapplied with a score increase to 62 points. Despite the slight improvement in the quality of life questionnaire, the patient did not incorporate the use of AAC in his daily life. After discussing the case and weekly care with a view to swallowing in home care, the elderly maintain a link to the clinic for management, reassessments and support every two months.

Discussion

The use of symbols in communication does not prevent the emergence or maintenance of orality¹⁴, which can increase the communication of the patient with difficulties of expression, and being considered a possibility in cases where the patient can no longer communicate verbally. In

the reported case, the patient presented speech, with very impaired intelligibility, with progressive worsening and speech-language diagnosis of severe dysarthria. It is observed that with the progression of the underlying pathology, communication has become increasingly compromised. Assistive technology - in this case realized through the AAC board - must be understood as an aid that can promote the expansion of a deficient functional skill, and enable communication¹⁵. The patient under study did not incorporate the use of AAC in his routine, some hypotheses were considered, since the patient was able to increase his communication possibilities, among these: the beginning of a new form of communication even when speech was found harmed and the patient used to this situation, another issue could be the mother's advanced age (main interlocutor).

In one study, the implementation of the use of alternative communication boards in a sample of adults revealed an impact on quality of life in individuals who were unable to communicate through speech¹¹. Effects on quality of life were observed in a therapeutic group of patients with different types of aphasia, from the implementation of AAC resources¹⁶. The early introduction and use of AAC in cases of dysarthria or apraxia can be beneficial, since conventional treatments in more severe cases may have limited results. AAC becomes a support for the oral, reading and writing of patients¹⁷.

The AAC's approach is often associated with decontextualized tasks recognition, naming and / or training point of symbols¹⁸. The investigation of processes of signifying the desires of patients with neurodegenerative diseases, who progressively lose their communicative skills must be worked in a contextualized and discursively oriented way. The professionals who live with these people must be attentive to their verbal and non-verbal communication, which are often identified through psychosomatic complaints¹⁹.

Despite the limitations of this study, in our population, no similar articles were found in the literature, although dysarthria is always described in cases of FRDA. AAC should be a strategy to be implemented in similar cases, since there is no scientific proof of therapeutic efficacy for speech, but a progressive decline. Despite the benefit, to date, no experimental and / or observational studies have been found indicating treatment efficacy in the speech of patients with FRDA⁹. It is a proposal

for possible rehabilitation, with its difficulties and benefits, but we have to focus on evidence-based therapeutic methods for this population.

The report presented is a resource of light technology that makes it possible to maintain human communication, seeking quality of life. Since these patients are usually young and lose their ability to communicate very early, it is important that intervention strategies for the continuity of communication are promoted²⁰. Although the patient did not show a change in quality of life after using AAC, early AAC interventions have shown better results²¹. Although there are many challenges to be studied in clinical practice, more research needs to be carried out to develop rehabilitation protocols.

A Brazilian study used expanded alternative communication in 13 patients with Amyotrophic Lateral Sclerosis, although focused on assessing quality of life in communication, the results were positive²¹. The beginning of the intervention, as well as the support of the partners, is one of the main facts for a successful implementation²². Although in the case of this patient the disease affected the speech and the motor part, some functions must be rethought due to the results obtained in this study. One of the greatest difficulties in patients with neurodegenerative disease has been the time to start, which may be one of the issues due to the low adherence to AAC, as well as the little impact on quality of life²³. As much as the AAC has a common practice in speech therapy, its implementation must undergo individual adaptations focused on the patient's age, education, as well as on their underlying disease²⁴.

This article allows speech therapists to reflect, as clinical, scientific and social challenges are launched with each new patient we receive at the speech therapy clinic, but they must always be seen within an evidence-based practice.

Conclusion

In this case report of a patient diagnosed with FRDA, despite the fact that communication is increasingly unintelligible, 4 speech therapy sessions aimed at implementing the use of AAC were not effective for the use of the device and did not impact the patient's quality of life.

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