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Case reports

Therapy speech impact in quality of life in patients with Machado-Joseph disease

Qualidade de vida em pacientes com doença de Machado-Joseph sob acompanhamento fonoaudiológico para disfagia

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ABSTRACT

The Machado-Joseph disease is a degenerative disease, currently considered the most frequent spinocerebellar ataxia. The degenerative process of the disease affects several regions and functions of the central and/or peripheral nervous system. The dysphagia is one of the symptoms of Machado-Joseph disease, being responsible for the clinical complications and for the aspiration pneumonia, its main cause of death. The goal of this paper is to verify the impact of the speech-language therapy in the quality of life of the Machado-Joseph disease patients. Four patients diagnosed with Machado-Joseph disease attended to the research, three female and one male, with age average in 46,5 years-old (± 18), and complaining about dysphagia. In the first session, all patients answered the protocol of quality of life in dysphagia assessment of Quality of Life in Dysphagia, *SWAL -QOL* were assessed for structural and functional aspects of swallowing and classified according to Functional Intake Scale for Oral (*FOIS*). After six speech therapy sessions conducted a new clinical evaluation of swallowing, again classified according to FOIS and answered the *SWAL -QOL*. Conclusions: After speech therapy, all patients showed better concept in the field fear of eating, food as a burden and fatigue, which can be inferred that there was an improvement in satisfaction with food processes and hence the quality of life accompanied subjects.

Keywords: Quality of Life; Deglutition Disorders; Machado-Joseph Disease; Dysphagia

RESUMO

A doença de Machado-Joseph é uma doença degenerativa e atualmente considerada a ataxia espinocerebelar mais frequente. O processo degenerativo da doença afeta diferentes regiões e funções do sistema nervoso central e/ou periférico. A disfagia é um dos sintomas presentes na doença de Machado-Joseph, sendo responsável pelas complicações clínicas e pela pneumonia aspirativa, sua principal causa de morte. O objetivo deste trabalho é verificar o impacto da terapia fonoaudiológica na qualidade de vida em pacientes com doença de Machado-Joseph. Participaram da pesquisa quatro pacientes diagnosticados com a doença, três do sexo feminino e um do sexo masculino, com média de idade de 46,5 anos (±18) e queixa de disfagia. Na primeira sessão, todos os pacientes responderam ao questionário de Avaliação da Qualidade de Vida em Disfagia, *SWAL-QOL*, foram avaliados quanto aos aspectos estruturais e funcionais da deglutição e classificados de acordo com a Escala Funcional de Ingestão por Via Oral (*FOIS*). Após seis sessões de tratamento fonoaudiológico, realizaram nova avaliação clínica da deglutição, novamente classificados de acordo com a escala FOIS e responderam o *SWAL-QOL*. Conclusões: Após a intervenção fonoaudiológica, todos os pacientes apresentaram melhor conceito nos domínios *medo de alimentar-se*, *alimentação como um fardo* e *fadiga*, podendo-se inferir que houve uma melhora na satisfação com o processo alimentar e, consequentemente, qualidade de vida dos sujeitos acompanhados.

Descritores: Qualidade de Vida; Transtornos da Deglutição; Doença de Machado- Joseph; Disfagia

INTRODUCTION

The Machado-Joseph disease (MJD) is a degenerative disease, which is considered the most frequent spinocerebellar ataxia[1,2]. The MJD was firstly described by Nakano, in 1972, in a family from Massachusetts - EUA, who came from the Azores islands. The clinical variability of this disease allowed that, for several years, it was described as different pathologies. In 1978, it was decided to name the disease, which was called Machado-Joseph. Since then, the diagnostic criteria defined for the MJD started to be internationally accepted and referred. It is known that the disease begins when individuals are about 40 years old. However, there is great variation of beginning, with extremes from five to 70 years old[3]. In Brazil, the only found study estimates prevalence of 1:100.000[4].

The degenerative process of the disease affects different regions and functions of the central and/or peripheral nervous system. For instance, there are the areas which are responsible for motor control of phonoarticulation and deglutition[4-6]. Dysphagia is one symptom of neurodegenerative diseases, responsible for clinical complications, which negatively interfere in patients' quality of life. It may affect the eating pleasure, leading to social isolation[4,5].

The studies about dysphagia in spinocerebellar ataxias are still rare and, although dysphagia in MJD is a common complaint and symptom, it is not a very frequent subject approached by literature. According to some authors, the most common found alterations are associated with oral and pharyngeal phases of swallowing. The frequently described symptoms are related to muscle weakness, dystonia and alteration of motor coordination which, combined with other neuromotor disorders, cause dysarthria and dysphagia. Dysphagia, in MJD patients, presents as main characteristics: gagging and cough, as with liquids as with solid food[6-8].

To evaluate the quality of life (QOL) is a hard task, because it is abstract and subjective. It involves personal, social, professional and emotional aspects. According to the World Health Organization (*WHO*)[9], QOL is defined, up to the present, through several factors, such as: housing, cultural, spiritual and social values, and health. Health is considered one of the most important factors. There are a number of protocols to evaluate QOL, in general terms, for example, the questionnaires *WHOQOL-100*[10], *WHOQOL-Old*[11], *SF-36*[12], etc.

In studies about MJD, it was not found, up to now, specific protocols to evaluate QOL. In relation to dysphagia, a used questionnaire to evaluate QOL is the *SWAL –QOL*[13], translated and adapted to the Brazilian population[14] and validated for application in MJD[6]. The evaluation of QOL related to deglutition allows knowledge about the real impacts of the disease on patients' life. Thus, the purpose of this study is to verify the impact of Speech, Language and Hearing therapy in QOL of patients with MJD.

CASES PRESENTATION

This study is based on a series of four cases with diagnosis of MJD. This research was approved by the ethics committee in research of Hospital de Clínicas de Porto Alegre, number 12-0168. All the patients were informed about the procedures and signed the Free and Clarified Consent Term. It was performed case study of four patients with confirmed diagnosis of MJD, assisted at the Speech, Language and Hearing therapy department of the Physiatry and Rehabilitation Service at a hospital in Porto Alegre, Brazil. The included subjects were patients who took Speech, Language and Hearing therapy from April 2012 to December 2014. All patients had received doctor's forwarding letter to Speech, Language and Hearing evaluation, with no previous Speech, Language and Hearing intervention.

There were three steps:

The first step consisted of application of the SWAL-QOL, which specifically evaluates the impact of swallowing alterations on quality of life, through 44 items, divided in 11 domains[14]. The protocol scores vary from zero, the worst score, to 100, the best score. The 11 domains evaluate: *swallowing as a burden, desire of eating, eating duration, frequency of symptoms, food selection, communication, fear of eating, mental health, behavior, social domain, sleep and fatigue*. The patients respond the questionnaire according to their perception about their daily life, considering dysphagia. The responses are classified in relation to occurrence frequency (always, frequently, sometimes, rarely and never) and degree of agreement about the statement (I totally agree, I agree, I am not sure, I disagree, I totally disagree). So, the *SWAL-QOL* was also used to measure the impact on QOL after Speech, Language and Hearing therapy.

The clinical swallowing evaluation (structural and functional) is performed through the application of a protocol, which was adapted based on swallowing protocols[15-17] frequently used at the Speech, Language

and Hearing department. It is subdivided in anamnesis, evaluation of orofacial structures in relation to mobility, tonicity and sensitivity.

As criteria to evaluate mobility, there were isometric and isotonic movements; to evaluate tonus, it was considered muscular resistance to passive stretching of the myofunctional orofacial structures. To test sensitivity, it was asked the patients to point where it was touched with spatula or cold spoon handle on their face or intraoral region. Their eyes were closed[16]. The patients did not present history of respiratory infection of any type and they were usually able to perform oral eating. It was evaluated the consumption of free and/or enough portions to determine the functional characteristics, including the consumption of, at least, 20 ml of the liquid (filtered water) and of pasty food (yogurt), as well as at least two solid intakes (wafer cookie). The same procedure was performed after the intervention period. The procedure started according to the patients' preference, and they did not receive directions and recommendations, up to the three different consumptions, except if the patient presented some signal or symptom of dysphagia, such as cough and/or gagging.

In the end of the evaluation, the level of dysphagia in MJD patients was classified according to internal evaluation and based on previous studies[15-17]. The classification of the Speech, Language and Hearing diagnosis varied[17] as follows:

- **Mild oropharyngeal dysphagia:** oropharyngeal transit mildly impaired, with no aspiration signals.
- **Moderate oropharyngeal dysphagia:** impaired oropharyngeal transit, signals of penetration/aspiration, but with preserved protection mechanisms.
- **Severe oropharyngeal dysphagia:** impaired oropharyngeal transit, signals of penetration/aspiration, and absence of protection mechanisms.

The Functional Oral Intake Scale (FOIS), translated and adapted to Brazilian population[18], was also used as marker, as it marks through seven specific levels (from zero to seven) the quality and the consistency of the oral food intake. The FOIS levels are:

Level 1: No oral intake

Level 2: Depends on alternative way and presents minimal oral intake of some food or liquid.

Level 3: Depends on alternative way with consistent via oral of food or liquid

Level 4: Total oral intake of an only consistency

Level 5: Multiple consistency oral intake, but with necessity of special preparation or compensations

Level 6: Multiple consistency oral intake, with no necessity of special preparation or compensations, but with dietary restrictions

Level 7: Oral intake with no restrictions

The second step was the Speech, Language and Hearing therapy, performed for six consecutive weeks. There were six therapy sessions, weekly, with 50 minutes each. The sessions were divided in orofacial myofunctional exercises; direct therapy with liquid and food, as well as recommendations and directions related to safe swallowing habits, including the use of compensatory, cleaning and/or facilitator maneuvers and adaptation to consistencies, if necessary. Individual myofunctional exercises were individually prescribed, according to the found dysfunctions. For example, there were exercises to improve the excursion of the larynx, to increase the movement amplitude or to tonus alterations. The individualized prescription occurs, mainly because of the high degree of MJD pleomorphism, not only in the starting age variability, but also in the presented neurological signs presented by the different patients[2]. The patients received written prescription for daily performance and also assistance every week.

The third and last step occurred after the six sessions. In this moment, all the patients were reevaluated through the same instruments and in the same sequence, as previously described.

So, the inclusion criteria of the four patients with MJD were met.

The sample characterization is described in Table 1.

The statistical analyses were performed through the software SPSS (*Statistical Package for Social Sciences*) version 18.0. First, it was used descriptive analysis of the variables *SWAL-QOL* before and after therapy, presenting average and standard deviation or median and interquartile range, for the quantitative variables, and absolute and relative frequency, for the categorical variables

Table 1. Table of sample characterization of four MJD patients, according to gender, age, time of diagnosis and age of disease beginning

	Absolute values of patients (n=4)	Relative Frequency (%)
Gender		
Female	3	75
Male	1	25
Age		
20-25	1	25
26-30	0	0
31-35	1	25
36-40	2	50
Age of disease beginning		
15-20	1	25
21-25	0	0
26-30	0	0
31-35	3	75
Time of diagnosis		
0-5 years	4	100

RESULTS

In this sample, there were four cases, three female and one male, with age average of 46.5 years old (±18). The patients arrived to Speech, Language and Hearing therapy with the following history:

Patient 1: MJD for three years, family history of the disease (sister and mother), report of falls and unbalance, difficulty in daily life activities, referring seated shower baths, diplopy and pain in the upper limbs during effort. In relation to swallowing, there are episodes of saliva gagging.

Patient 2: MJD for one year, family history of the disease (father and two uncles), report of falls and unbalance, difficulties to perform more complex movements and diplopy. In relation to swallowing, the patient presented complaints about liquid consistency.

Patient 3: for one year, no family history of the disease, no report of other symptoms. In relation to swallowing, the patient presented complaints about liquid consistency.

Patient 4: MJD for three years, family history of the disease (already dead brother and mother), needs help to walk and presents total dependency out of home. Report of eventual dipropy. In relation to swallowing, the patient presented complaints about solid and liquid consistencies.

In relation to clinical swallowing evaluation before intervention, all the patients were diagnosed with mild oropharyngeal dysphagia. After the clinical evaluation, all the patients had the FOIS completed, aiming at monitoring safe food consistencies, as they were all in level 7 (oral intake with no restrictions). After Speech, Language and Hearing intervention, it was performed another clinical swallowing evaluation and it was verified that all the patients maintained the initial diagnosis of mild oropharyngeal dysphagia and FOIS level 7. Only one patient presented diagnosis change to moderate oropharyngeal dysphagia. So, it was necessary diet adaptation during the therapeutic process, for safety. This generated change of FOIS level after therapy to level 5 (multiple consistency oral intake, but with necessity of special preparation or compensations).

The found results in relation to QOL of the four MJD patients are presented in the following table.

Table 2. *SWAL-QOL* score before and after speech, language and hearing therapy

	Pre Therapy	Post Therapy	Pre Therapy	Post Therapy	Pre Therapy	Post Therapy	Pre Therapy	Post Therapy
Patient	1	1	2	2	3	3	4	4
SWAL-QOL domains								
Eating as a burden	75	87,5	50	87,5	50	100	87,5	100
Desire of eating	100	100	100	100	100	100	91,67	100
Eating duration	100	100	100	100	0	37,5	100	50
Frequency of symptoms	80,35	80,35	66,07	69,64	64,29	92,86	78,57	85,71
Food selection	100	100	100	100	100	100	100	100
Communication	50	62,5	50	87,5	62,5	75	75	75
Fear of eating	50	75	31,25	81,25	56,25	75	81,25	87,5
Mental health	85	100	45	100	95	100	100	100
Social	100	100	80	100	100	100	100	100
Sleeping	12,5	87,5	50	75	100	50	100	75
Fatigue	50	83,3	66,67	91,67	83,33	91,67	50	83,33

DISCUSSION

The MJD, also known as spinocerebellar ataxia type 3, is caused by CSG expansions which codify a repetition of the amino acid glutamine in its correspondent protein[19]. It is a neurodegenerative dominant autosomal disease of late beginning, which involves predominantly the cerebellar, pyramidal, extrapyramidal, motor neurons and oculomotor systems. It presents strong phenotypic heterogeneity, but the main pathologic injuries are observed in the spinocerebellar system, as well as in the cerebellar dentate nucleous[2]. As it attacks the parts responsible for motor control, as in phonoarticulation, as in swallowing, Speech, Language and Hearing intervention for dysphagia is essential, because, in the majority of cases, the swallowing difficulty may cause aspirative pneumonia[4].

Regardless of the presence of some disease, but because of the increase of life expectation, it has been observed greater concern on maintaining quality along the years. In the presence of some disorders, specifically in relation to Speech, Language and Hearing aspects, rehabilitation should not be disconnected from aspects related to QOL.

In the present study, considering the functional disorders, following the degenerative ataxias, it was confirmed, from the results, that dysphagia compromises individuals' QOL, but with the differences in each case[2,6]. It highlighted the importance of evaluation and intervention, according to the found impairment level. The QOL instruments are protocols which help individuals to perceive the problem. From them, the *SWAL-QOL* is the only instrument which broadly evaluates dysphagia, regardless of the ethiology[10,13], and it is also validated for MJD[5]. In this disease, dysphagia is one of the symptoms which interfere in QOL, because it is related to severe complications, such as malnutrition, dehydration and aspirative pneumonia[6]. The QOL evaluation, considering Speech, Language and Hearing for dysphagia, would allow the evaluation of the rehabilitation effects in several stages of treatment. Thus, the use of specific instruments such as the *SWAL-QOL* would allow people to know, in a measurable way, the real impact of dysphagia in QOL and, so, to direct and to maximize the treatment proposal and rehabilitation.

In the analysis of the present sample, it was possible to verify that, from the evaluated items, the four patients presented improvements in the domains eating as a burden, fear of eating and fatigue. These dominions were reported as the ones which affect the most the dysphagic individuals[13], because eating, seen as a burden, may reflect the patients' fear of eating, mostly because they do not present conditions to do it and/or because of the consumption of a consistency which causes muscle stasis or fatigue. Thus, a better result of these domains positively implies MJD patients' QOL.

The domain food selection was the only one which did not present alterations, possibly because all the individuals of the sample presented FOIS 7, that is, without necessity of food change. Nevertheless, a previous study[13] verified that the patients who presented some type of food or consistency restriction demonstrated more fear of eating. Besides, they described the

swallowing problem as the most difficult one, with more impact on mental health.

In the research which should validate the SWAL-QOL in MJD patients, it is described that there was clarity in the differentiation between patients with the disease and control group. Only the domains food selection and duration did not present significant difference. It was stated about the relative psychometric validity to apply it with this disease, and it was necessary studies with patients who present the disease for a longer time. It was also mentioned the correlation of this results with other QOL questionnaires[5].

In a study performed with dysphagic patients, after encephalic vascular accident, significant alterations were verified in relation of *time of eating, fear of eating, mental* and *social health and fatigue* after evaluation through the *SWAL-QOL*, referring impairments which are similar to the findings of the present study, considering QOL[20]. In the analysis of each patient, individually, the findings aim at each patient's peculiarities and at the relation that they present in the presence of the disease and its complications.

Patient 01 presented improvement in six of the eleven evaluated domains and, in the other domains, there was stability, with no worsening. Patient 02 presented improvement in eight of the eleven evaluated domains. In the domains fear of eating and fatigue, the improvement was more impacting. Patient 03 presented improvement in six of the eleven domains. Eating as a burden and frequency of symptoms impacted the most positively the patient's self-perception. The dominion sleep presented worsening for patient 03 and patient 04. However, it is known that sleeping disorders are commonly reported by patients with neurodegenerative diseases, especially by patients with MJD[21].

Patient 04 reported worsening in the sleeping and eating duration domains. There was worsening related to dysphagia, with alteration from mild dysphagia, in the beginning of therapy, to moderate, along the supervision. Some studies[4,5] observe that the incidence of moderate dysphagia in this population is higher than severe dysphagia, but there are cases in which, even after years of disease progression, there may not be dysphagia. From the eleven evaluated domains, patient 04 presented improvements in four of them.

From the four supervised patients, three of them did not present dysphagia alterations, maintaining the mild level of dysphagia and FOIS level 7. However, patient 04 presented alteration of dysphagia level and change in FOIS from 07 to 05. This change may have happened because this patient had precocious beginning of disease and had started Speech, Language and Hearing therapy with manifested signs of the disease, in a more advanced stage, compared to the other studied patients. Although this patient presented worsening of clinical evaluation after intervention, the changes of consistencies performed during the therapeutic process impacted positively on QOL, with improvements in the most impacting domains for dysphagia[13]: *fatigue, fear of eating, frequency of symptoms, eating as a burden and desire of eating*. These data highlight the importance of Speech, Language and Hearing intervention in the QOL of these patients, because dysphagia is indicated by some studies as an important part of psychosocial alterations[21]. Other studies demonstrate the importance of eating in the QOL of MJD individuals. Dysphagia affects the QOL of patients and family members[22].

The several diseases which dysphagia is one of the symptoms may present positive correlation with QOL and swallowing disorders, affecting QOL negativelly[13]. It impacts on the social function represented by eating, because of routine restrictions and participation in activities with other people. There is consensus, among several authors, that feelings such as frustration and discouragement may lead dysphagic people to stop eating in public[22,23]. The signs associated with dysphagia, such as cough, suffocation or diet changes, even in no progressive diseases, may result in loss of independency and psychosocial consequences[23]. Likewise, MJD patients, because of great functional incapacity, present several signs which also may reflect on QOL, including limitation and isolation of social coexistence[8].

In the structural clinical evaluation of swallowing, the most prevalent alteration presented by patients was dental failure. It may impair the preparatory and oral phases of swallowing[24]. Dental failure may be associated with socio-economic issues, as the service was performed at a public hospital. Therefore, this is another variable to be considered in the aspects related to QOL and Speech, Language and Hearing therapy, because the patients do not present only the degenerative disease, but also oral structural alterations.

The MJD is a disease, like other neurodegenerative diseases, with no cure. However, as pharmacologic as no pharmacologic treatments should be used with these patients, aiming at improving QOL, minimizing the effects of the symptomatology[25-27].

Even if this is a progressive disease, individuals may understand the disorder and be adapted to the

dysfunction. The Speech, Language and Hearing rehabilitation tries to compensate the functional loss[26], ensuring, as longer as possible, oral eating, total or partial. In a study about the Huntington disease, it was described that a specific rehabilitation program, with bucco-linguo-facial praxias and training of postural strategies and maneuvers, would minimize the swallowing disorder and would contribute to maintain the patients' nutritional status[28].

The study of SWAL-QOL validation to QOL evaluation in patients with MJD identified that there was no correlation between the questionnaire score and the duration of the disease. Although it suggested that dysphagia is slowly progressive[5], the variability of symptoms emergence was supreme when it was prepared the Speech, Language and Hearing therapeutic process to dysphagia in MJD patients. Moreover, as it was found in studies about other chronic or chronic-degenerative diseases, the QOL evaluation in dysphagia and the actions planning impacted on the functionality committed by the disease, causing promotion of general well-being of people[20,25,28,29].

Some studies about the MJD discuss that, although there is impact of dysphagia in the disease, it may not be associated with the period or with the progress of the other symptoms[5,19]. However, as it is a progressive disease, in which dysphagia may appear in it beginning, evaluating QOL is extremely important. This is beyond Speech, Language and Hearing intervention. In this population, it is perceived variability in the emergence of this symptom, which may be discrete, since the beginning of diagnosis or along the disease progression.

CONCLUSION

Speech, Language and Hearing intervention changes positively the perception of all supervised subjects, considering the domains *fear of eating, eating as a burden* and *fatigue*. So, it is possible to infer that there was improvement in QOL and satisfaction in relation to eating process in general. Thus, Speech, Language and Hearing intervention may impact on the QOL of MJD patients. Specifically for the studied cases, it is perceived that the performed treatment was effective in maintaining, mainly, the ingestion and the eating pleasure in the patients' phase of disease.

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