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Case Reports

Functional profile of patients with cervical dystonia: a series of cases

Perfil funcional de pacientes com distonia cervical: uma série de casos

Carolina Cerqueira Santos[1], Nildo Manoel da Silva Ribeiro[2]

[1]Corresponding author. Federal University of Bahia. Salvador, Bahia, Brazil. ORCID: 0000-0001-8551-4649. carolinasantos2013.2@gmail.com
[2]Federal University of Bahia. Salvador, Bahia, Brazil. ORCID: 0000-0002-1879-0405. nildoribeiro67@gmail.com

RESUMO | INTRODUÇÃO: A Distonia Cervical (DC) é uma doença dos núcleos da base e tem como consequência movimentos hipercinéticos, sendo a mais comum entre as distonias focais. Além disso, pode ser definida pela presença de contrações involuntárias envolvendo a musculatura cervical. **OBJETIVO:** descrever o perfil funcional de pacientes com DC, oferecendo maior embasamento focado nas habilidades funcionais desses pacientes. **MÉTODOS:** Foi realizado um estudo transversal de pacientes atendidos no Ambulatório de Neurociências localizado no Ambulatório Professor Francisco Magalhães Neto, Salvador, Bahia, no período de novembro a dezembro de 2017. **RESULTADOS:** Foram analisados 6 pacientes: 3 do sexo feminino e 3 do sexo masculino. A maior parte dos pacientes apresentou os sintomas iniciais após algum trauma sofrido, baixo/moderado risco de queda, qualidade de vida (QV) impactada negativamente por limitações físicas, dependência leve e dor moderada. As principais queixas relacionadas à participação social estavam relacionadas a frequentar ambientes públicos, devido à vergonha que sentem por causa dos movimentos involuntários, além da impossibilidade de trabalhar proporcionada pela doença. **CONCLUSÕES:** O perfil funcional dos pacientes com DC ainda é escasso na literatura. Nesse estudo, a população com DC mostrou sofrer não só impactos na QV por fatores físicos, mas, sobretudo, por fatores emocionais e sociais.

PALAVRAS-CHAVE: Distonia. Torcicolo. Comportamento. Funcionalidade.

ABSTRACT | INTRODUCTION: Cervical Dystonia (CD) is a disease of the nuclei of the base and results in hyperkinetic movements, being the most common amongst focal dystonias. In addition, it can be defined by the presence of involuntary contractions involving the cervical musculature. **OBJECTIVE:** make known the functional profile of patients with CD, offering a more focused foundation on the functional abilities of these patients. **METHODS:** A cross-sectional study was carried out of patients attended at the Neuroscience Outpatient Clinic located in the Professor Francisco Magalhães Neto Ambulatory, Salvador, Bahia, from November to December 2017. **RESULTS:** Six patients were analyzed: 3 females and 3 males. Most patients presented initial symptoms after some trauma, low/moderate risk of falls, quality of life (LQ) negatively impacted by physical limitations, mild dependence and moderate pain. The main complaints related to social engagement were related to attending public places, due to the shame that they feel because of the involuntary movements, besides the impossibility of working caused by the pathology. **CONCLUSIONS:** The functional profile of patients with CD is still scarce in literature. In this study, the population with CD showed not only impacts on LQ due to physical factors, but, above all, by emotional and social factors, which makes a biopsychosocial approach indispensable to the assistance of these persons.

KEYWORDS: Dystonia. Torticollis. Behavior. Functionality.

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Introduction

Cervical Dystonia (DC) is a disease that occurs predominantly in female patients aged between 30 and 50 years[1,2]. However, there is no definite etiology for CD: the findings in literature have as the main cause of disability in pain patients, as well as involuntary contractions.

The disabilities generated in people with CD have repercussions on economic and psychosocial factors, which can trigger social stigmas, eventually leading them to isolation and affecting QoL. There is thus a public health problem[3,4,5]. Added to this is the fact that the World Health Organization (WHO) defines health not only as the absence of disease, but as the situation of perfect physical, mental and social well-being[6].

Being aware that there is a gap in literature regarding the functional profile of patients with CD, it is necessary that this framework be investigated and evaluated by health professionals with a biopsychosocial approach, being in agreement with the International Classification of Functioning, Disability and Health (CIF) concerned with health at bodily and social levels[7].

Thus, the objective of this study is to describe the functional profile of patients with DC, in order to obtain a better foundation capable of directing and promoting an effective treatment, that is, focusing on the functional abilities of these patients.

Materials and methods

This is a cross-sectional study of a series of cases of patients with CD submitted to an evaluation that was performed in the Neuroscience department located in the outpatient clinic Professor Francisco Magalhães Neto, at Augusto Viana Street, no number, Canela, Canela Universitary Campus of the Professor Edgar Santos Universitary Hospital Complex, ZIP Code: 40110-060, Salvador, Bahia, from November 2017 to December 2017.

The convenience sample consisted of the patients followed in the neuroscience sector with a diagnosis of CD. This study includes part of the project titled "Characterization of patients with cervical dystonia and the impact of the use of botulinum toxin in these patients," approved by the Ethics Committee of the Professor Edgar Santos University Hospital Complex (CAAE: 62719516.7.0000.0049) under the number 1.986.100. However, participants in the current study were not using botulinum toxin within at least three months prior to the evaluations.

Inclusion criteria were considered to be between the ages of 18 years and 65 years, presenting CD, being a patient of the Magalhães Neto Outpatient Clinic and signing the Informed Consent Form (Appendix B). As exclusion criteria, the following points were considered: (i) to have neurodegenerative and osteomioarticular diseases that influenced the results of the tests; (ii) present cognitive impairment according to Mini Mental State Examination (MMSE) lower than: if illiterate, 19 points; 1 to 3 years of schooling: 23 points; 4 to 7 years: 24 points and> 7 years of instruction: 28 points.

The protocol was started with the link for the patients who have the contact number in the CD agenda of the neurosciences outpatient clinic and invitation to participate in the research. With the necessary clarifications, the evaluations were initiated with the application of the MMSE. Then the partner evaluation demographics, manual heart rate measurement using a timer for counting time and blood pressure through a Premium brand sphygmomanometer.

A previously trained researcher applied the scales. The Barthel Index was used to evaluate the level of dependence of the subject to perform basic activities of daily living, being considered the score of 100 for total independence, and the others for light dependence 99 to 90; 89 to 61 moderate; 60 to 21 severe and <20 total. To evaluate the QoL, the SF-36 scale was used in the Brazilian version, with the calculations indicated for each domain (each one of them registering from zero to one hundred), obtaining notes kept separately.

In order to evaluate the balance and possible risks of fall, the Berg scale was used considering the scores of 56 to 54 without risk of falling, 53 to 36 low / moderate risk of fall <36 high risk of fall. Pain was measured using the Visual Analogue Scale

(EVA) in which the score ranges from 0/10 where 0 means no pain, 1-3 mild pain, 4-6 moderate pain, 7-10 severe pain. These results were presented in a descriptive way.

Results

The appointment dialing schedule included the contact number of 112 patients with CD. Connections were made to these numbers and, of these, 38 did not respond in any of the attempts, 42 were wrong numbers, 10 had no interest in participating or were deceased, 1 had no diagnosis of CHD. 15 were interested, however some barriers prevented them from participating, such as distance from the place where they lived in relation to the place of assessment and difficulty in accessing transportation.

Thus, 6 patients were analyzed, being 50% female and 50% male, with mean age of 54.8 ± years, (S=13,8). Two had complete primary level of education, Two complete secondary school, One incomplete primary school and one incomplete secondary school. Three of the participants were divorced and five retirees with an average monthly income less than two minimum wages. In addition, all participants reported having irregular sleep (Table 1).

Table 1. Socio-demographic data. Salvador, BA 2017

Variable	N (±)
Gender	
Male	3
Female	3
Age	54,8 (S= 13,8)
Schooling	
FC	2
FI	1
EMC	2
SI	1
Live	
Alone	1
With Relatives	3
With Children	1
With Family	1
Comorbidites	
HAS	4
Osteomioarticular	3
Other	3
Work	
Yes	1
No	5
Income	1.579
Sleep	
Irregular	6

PC= Patient; M= Male; F= Female; S= Standard deviation; Fk= Full key; KI= Key incomplet; EMC= Complete high school; SI= Accoutat; SAH= Systemic arterial hypertension.

In the results obtained by the Barthel Index, two of the participants obtained total independence, two mild dependence and two moderate dependence. For the Berg scale, the low risk of falls was observed in five of the participants and the high risk of falling in one. In the assessment of pain through VAS, three presented mild pain, one moderate pain and two severe pain. (Table 2).

Table 2. Scoring of scales, questionnaires and evaluation tests. Salvador, BA 2017

PC	BARTHEL	BERG	EVA
1	95	53	3
2	100	45	3
3	85	42	3
4	85	37	5
5	100	38	8
6	95	32	8

PC= Patient.

Regarding aspects that affect QoL, limitations due to physical aspects, followed by functional capacity, pain, social aspects and limitation by emotional aspects are the indices of greatest negative impact for the participants. In the first case, five of the participants scored less than 50. In the other categories, this score represents four of the participants. (Table 3).

Table 3. Score obtained in% of SF-36 by dimensions. Salvador, BA 2017

PC	CF	LAF	D	EGS	V	AS	LAE	SM
1	70	10	52	87	60	50	100	96
2	35	50	74	85	30	25	66,6	44
3	10	25	20	14	25	62.5	33,3	56
4	10	25	42	25	15	25	33,3	44
5	85	25	20	65	60	25	33,3	68
6	35	25	10	65	80	37.5	33,3	32

PC= Patient; CF= Functional capacity; LAF=Limitation for physical aspects; D= Pain; EGS= General health; V= Vitality; AS= Social aspects; LAE= Limitation for emotional aspects; SM= Mental health.

Discussion

This study evaluated participants with DC, a disease not yet defined pathogenesis. The symptoms of DC triggered after trauma, common in these individuals[4], were found in 4 of the participants. In addition, only one of the six participants presented the initial symptoms early (before the age of 30)1. The others presented symptoms late (between 30 and 50 years)[1,8]. Corroborating with these data, Barbosa et al. (2005) and Camargo et al. (2014) found a greater number of participants who presented initial symptoms late compared to those who presented early.

All participants had some balance deficit with some risk of falling according to the Berg scale: five had low / moderate risk scores and one high risk. Such deficit can be caused due to failures in systems involved with sensory Impults due to the involuntary movements present in the region of head and neck of people with CD. As a consequence, interference may occur in the performance of daily activities.

Crowner[3] portrays the importance of identifying adequate interventions for these deficits to be minimized in subjects with CD, since it is an important factor that can significantly affect the mobility and function of these subjects.

When questioned about sleep quality, all participants reported having irregular sleep due to pain and tension in the neck region. This situation caused discomfort, as the participants reported being tired and irritated because of the lack of sleep. Such information may be decisive for understanding the influence of this disorder on the participants' QoL, deserving greater attention in future studies[9].

The findings in the literature associate the impacts caused on the QOL of the affected subjects to functional disability, as well as to emotional, physical, psychological, pain and limitation factors in social interaction. Although this association was not performed in the present study, it is important to note that individuals with CD and pain may present changes, such as low self-esteem, negative body perception, anxiety and depression[10].

Skogseid et al. (2007) found a positive correlation between the severity of CD and greater pain experienced by the subjects, more physical limitations, worse general health status and social function of the SF-36[11] scale. In the study by Weber W et al. (2014), pain, depression and anxiety had the most significant contributions to the decrease of QoL in patients with CD.

Although not correlated with QOL, the present study found pain as one of the main factors causing functional disability in patients with DC[12], being present to some degree in all participants, as well as in the studies performed by Crowner 2007[1].

Regarding the general state of health, since it includes both the interviewee's perception of their current health condition and their evolution (or not) perspective, the data found shows that only two participants presented negative results. These data demonstrate the positive perception of these participants regarding their own health. However, because it is a subjective perception, this data can not be attributed to the entire population.

The most frequent complaints reported by patients related to social participation deal with the shame of attending public environments, such as restaurants, because of involuntary movements that draw attention to the people around. In addition, these complaints of social isolation become more relevant when participants report that they have stopped working because of the disease.

With these scenarios in view, it is important to stress that work is not only a form of financial maintenance, but also a way of building self-esteem and social value from the moment one feels useful for being able to do so. Only one participant was still working, the others had to retire. Even one participant reported ignorance regarding the laws of retirement, a fact that draws attention to the need for wide dissemination of the rights of the population by health professionals. There have also been complaints about marital relationship, and some participants claim to be inhibited in attempting to re-relate after the onset of symptoms.

Regarding functional independence, evaluated through the Barthel Index, the participants obtained results equivalent to complete independence, mild and moderate dependence. However, when the functional capacity is verified, it is necessary to point out that among the four that presented an inconsistent score with the degree of dependence reported in Table 2, half obtained a significantly negative result[10]. This apparent contradiction falls apart when it is understood that significant part of the patients report that although they present a marked difficulty in functional activities (such as self-care), they need to perform them, since there is no one who can help them, the non-everyday.

With regard to mental health and vitality, half of the participants scored below 50%. Concerning social aspects and limitation by emotional aspects, four of the participants presented unsatisfactory marks, which demonstrates the impact of social and psychological criteria on the QOL of dystonic subjects[13,14,15], since it dialogues with the statement about the central parameters related to the WHO health concept mentioned throughout this work.

Conclusion

In this study, this population presented pain - considered a source of difficulties in the accomplishment of activities -, some deficit of balance and discomfort associated to limitations by physical, social and emotional aspects, that lead to the restriction in social participation, factors that affected the participants' QoL. Irregular sleep quality was also mentioned by the participants as a factor contributing to a change in physical performance during the day. Further studies are necessary, even with a larger number of participants.

Limitations of the study

The sample size was limited in this study, possibly because of the following factors: (i) the participants' lack of mobility, (ii) the in-home confinement due to the embarrassment of going out on the street, (iii) the fear of suffering accidents due to not having company to go to the evaluation site, (iv) limited access to the numbers of patients followed at the outpatient clinic. Another important limitation was that we did not have a specific scale for the evaluation of CD as well as to evaluate depression and quality of sleep, since complaints became frequent as the evaluations were made, although the initial design did not have such scales, being important inclusion in future studies. In addition, the scale chosen for the evaluation of functional balance was not very sensitive for the detection of less severe balance alterations, requiring a more sensitive evaluation instrument.

Contributions of authors

Santos CC participated in the data collection and interpretation and writing of the scientific article. Ribeiro NMS supervised the research and participated in the writing of the scientific article.

Competing interests

No financial, legal or political competing interests with third parties (government, commercial, private foundation, etc.) were disclosed for any aspect of the submitted work (including but not limited to grants, data monitoring board, study design, manuscript preparation, statistical analysis, etc.).

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