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Determinants of disability in cervical dystonia

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ABSTRACT

Background: Cervical dystonia (CD) is characterized by involuntary muscle contractions causing abnormal postures and/or twisting movements of the head and neck. These motor symptoms can have a major impact on disability. Treatment with botulinum toxin injections aims to reduce motor symptoms, and therefore disability. Despite motor improvements, many patients still experience difficulties with performing daily life activities. To optimize treatment, other factors that determine disability should be identified.
Objective: To explore and identify clinical characteristics that relate to disability in CD.
Methods: Data on disability, severity of dystonia, anxiety, depression, pain and quality of life of 96 CD patients was analyzed with a principal component analysis (PCA). Multiple regression analysis was performed to determine which components derived from the PCA explain most of the variance in disability.
Results: PCA revealed five components (disability, psychiatric features, pain, physical function and severity of dystonia), explaining 74.4% of the variance in disability. Multivariate association between disability and the other components was statistically significant (R^2 change 0.433, F change (4 −86) = 22.39, p = .000). Psychiatric features had the largest contribution to disability (standardized beta = 0.555, p = 0.000) followed by pain (standardized beta = 0.232 p = 0.004). Physical functioning (standardized beta = 0.059 p = 0.507) and severity of dystonia (standardized beta = −0.001 p = 0.991) had no significant contribution.
Conclusions: In CD patients, psychiatric features and pain are important determinants of disability. Interventions to reduce psychiatric problems and pain should have a more prominent role in the treatment of CD patients in order to improve disability levels.

1. Introduction

Cervical dystonia is a neurological movement disorder characterized by involuntary muscle contractions causing abnormal postures and/or twisting movements of the head and neck [1]. These motor symptoms are the most distinctive features of CD and can have a major impact on the level of disability. Current treatments are primarily aimed at the reduction of motor symptoms. The preferred treatment is to inject the affected muscles with botulinum toxin (BTX) and is proven to be effective in reducing the involuntary movements and abnormal postures in 70–92% of the patients [2,3]. Improvement of motor symptoms also decrease the level of disability [4] but despite these improvements, many CD patients still experience difficulties with the ability to perform daily life tasks.

Disability according the World Health Organization is an umbrella term covering functions, activities and participation, as well as environmental and personal factors as defined in the international classification of functioning, disability and health (ICF) [5]. Functions are the result of physiological processes and anatomical structures of the body systems. Activities are the execution of a task or action by an individual and participation is involvement in a (social) life situation. Environmental factors are the physical-, social- and attitudinal environment in which people live where personal factors include gender, age, coping styles, social background,

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profession and other factors that influence how disability is experienced by the individual. Thus, disability is a complex phenomenon, reflecting an interaction between these different domains [5].

Over the last ten years there is increasing awareness that in addition to motor symptoms, non-motor symptoms may also play an important role in the management of dystonia [6–9]. A study by Klingelhoefer et al. showed that 95% of the CD patients reported non-motor features like loss of self-confidence, anxiety, depression, insomnia, fatigue and pain [6]. However, only a few studies have investigated the association of non-motor symptoms with disability, so evidence is still limited. Page et al. showed that depression and disfigurement explain 41% of the variance in disability of CD patients [10]. A study by Zetterberg et al. on the self-perceived non-motor aspects of CD indicated that self-efficacy, pain intensity, anxiety and fatigue explain much of the variance in disability in individuals with CD [9]. With evidence pointing towards the contribution of non-motor features, it is not surprising that many CD patients still encounter difficulties with the ability to perform daily life activities despite BTX treatment. Current treatments primarily aim at the reduction of motor symptoms, thus focusing mainly on the functions domain and not the domains of activity, participation, environmental factors or personal factors. Integration of treatment strategies that also focus on other domains of disability, may improve treatment outcomes.

To provide a better understanding of disability in CD patients, more research towards the association of motor and non-motor symptoms with disability is indicated. In this cross-sectional explorative study we aim to identify associations of clinical characteristics which could further unravel disability in CD.

2. Methods

2.1. Design and participants

This cross-sectional study was performed on the baseline data of patients with idiopathic CD who participated in the Dutch DystoniaNet study that aims to optimize the BTX treatment and investigates the effectiveness of a standardized PT program in 96 patients with cervical dystonia [11]. Patients of 30 years or older and stable on BTX treatment for more than one year were recruited from the neurology departments of 15 Dutch hospitals. Participants were excluded if they had secondary or hereditary forms of dystonia, dystonia in other body parts and if they had deep brain stimulation or selective nerve denervation for the treatment of their dystonia. This project was approved by the local medical ethics committee and written informed consent was obtained from all participants.

2.2. Data collection

Data were collected between November 2012 and June 2015 shortly before the BTX injections because we hypothesized that the effects of PT mainly occur in the period that the effects of BTX wear of [11]. Measurements were performed by one independent assessor (JvdD). In the week prior to the measurements patients filled in self-reported questionnaires. Items that were unclear or missing were discussed with the participant and filled in on the day of the measurements to minimize missing values. Both generic and disease specific instruments were used for the same variable. Generic instruments were used for comparison with other studies. Disease specific instruments were used to investigate how PT affects CD in more detail.

2.3. Outcome measures

2.3.1. Disability

Disability was measured with the disability subscale of the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS). The TWSTRS scale is a widely used disease specific scale in research and is a valid and reliable tool to measure severity, disability and pain in CD (Kendall Tau = 0.85, $p < 0.01$) [12–14]. The disability section is a six point Likert scale consisting of six items like driving a car, reading and performing daily life activities (range 0–5). It has a maximum score of 30 points. Lower scores indicate less disability.

Disability was also measured with the Functional Disability Questionnaire (FDQ). The FDQ is a reliable disease specific scale ($r = 0.93$, $P < 0.001$) with 27 items to measures the impact of CD on daily functioning [15]. Questions are asked about the extent to which CD affects the engagement in and performance of a sample of activities at this moment. Each item is rated on a 5-point scale (range 0–4). It has a maximum of 108 points. Lower scores indicate less disability.

2.3.2. Severity of CD

Severity of CD was measured by the severity subscale of the TWSTRS [12–14]. The TWSTRS scale is a widely used disease specific scale in research and is a valid and reliable tool to measure severity, disability and pain in CD (Kendall Tau = 0.85, $p < 0.01$) [12–14].The TWSTRS severity subscale consists of 11 items scored with a Likert scale (range 0–1 up to 0–5). It has a maximum score of 35 points where lower scores indicate less severity.

Severity of CD was also measured with the Tsui scale [16].It is also a widely used, standardized and reliable scale (ICC = 0.86) to measure the severity of CD. The Tsui scale measures different aspects of abnormal posture and movements. Scores range from range 0–25 where lower scores indicate less severity of dystonia.

2.3.3. Anxiety and depression

Anxiety and depression levels were determined with the Beck's Anxiety Inventory (BAI) and Beck's Depression Inventory (BDI) [17,18]. Both instruments are validated and reliable tools and are rated on a 21 item 4 point Likert scale (BDI: $r = 0.73$ with Hamilton Psychiatric Rating Scale for Depression. BAI: test-retest reliability coefficient of 0.67, $R = 0.54$, $p = 0.05$ with anxiety). The maximum score of both scales is 63 points where lower scores indicate lower levels of depression and anxiety.

2.3.4. Pain

Pain was measured by the TWSTRS pain subscale [12–14]. It scores the usual level of pain, the level of pain at its worse and the level of pain at its least on an 11 point Numeric Rating Scale (NRS) (range 0–10) during the last week. In addition it scores the duration of pain and disability due to pain on a six point Likert scale (range 0–5). Al items are calculated to a total score between 0 and 20 points were lower scores indicate lower levels of pain.

Presence of pain at the present moment was rated on an 11 point NRS. A score of 0 indicates no pain and a score of 10 indicates the worst pain imaginable. The NRS is a validated and reliable tool for the assessment of pain (Spearman r = 0.94 between Visual Analogue Scale and NRS, test-retest reliability ICC = 0.90) [19,20].

2.3.5. Quality of Life

Quality of Life (QoL) was measured with the Craniocervical Dystonia Questionnaire (CDQ-24) and Short Form 36 (SF-36) [23,24]. The CDQ-24 is a validated and disease specific, self-reporting questionnaire to evaluate quality of life of patients with cervical dystonia [21]. It is divided in 5 subscales measuring stigma, emotional problems, ADL functioning, pain and social functioning.

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