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# CERVICAL DYSTONIA

## Clinical and therapeutic features in 85 patients

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**Abstract** – We studied patients with cervical dystonia (CD) to determine clinical features and response to botulinum toxin A (BoNT/A). Patients were submitted to clinical, laboratory and neuroimaging evaluation. BoNT/A was injected locally in 81 patients using electromyographic guidance. Four patients who had had previous treatment were considered to be in remission. The average ages at onset of focal dystonia and segmental dystonia were greater than for generalized dystonia ($p<0.0003$). The severity of the abnormal head-neck movements were more severe among the patients with generalized dystonia ($p<0.001$). Pain in the cervical area was noted in 59 patients. It was not possible to determine the etiology of the disease in 62.3% of patients. Tardive dystonia was the most common secondary etiology. A major improvement in the motor symptoms of CD and pain was observed in patients following treatment with BoNT/A. The tardive dystonia subgroup did not respond to the treatment. Dysphagia was observed in 2.35% of the patients.

KEY WORDS: dystonia, cervical dystonia, botulinum toxin, dysphagia.

**Distonia cervical: aspectos clínicos e terapêuticos de 85 pacientes**

**Resumo** – Para identificar os aspectos clínicos e a resposta a toxina botulínica A (TxBA), pacientes com distonia cervical (DC) foram submetidos a avaliação clínica, laboratorial e neuroimagem. O tratamento com TxBA foi aplicado a 81 pacientes guiado por eletroneuromiografia. Quatro pacientes, com tratamento prévio, foram considerados em remissão. A média de idade de início dos sintomas de pacientes com distonia focal e segmentar foi maior que a encontrada em pacientes com distonia generalizada ($p<0,0003$). A gravidade das alterações motoras cervicais foi maior entre os pacientes com distonia generalizada que nos pacientes com distonia focal ($p<0,001$). Graus diferentes de dor na região cervical foram relatados por 59 dos pacientes. Não foi possível determinar a etiologia da doença em 62,3% dos pacientes sendo distonia tardia a mais comum. Houve acentuada melhora dos sintomas motores e da dor da DC com a aplicação de TxBA. O subgrupo de pacientes com distonia tardia não respondeu ao tratamento. Disfagia ocorreu em 2,35% dos pacientes.

PALAVRAS-CHAVE: distonia, distonia cervical, toxina botulínica, disfagia.

Dystonia is defined as a syndrome characterized by prolonged muscle contraction causing twisting, repetitive movements or abnormal posture[1]. Most voluntary muscles can be affected and, in the case of the neck muscles, the condition is referred to as cervical dystonia. The term spasmodic torticollis was previously used for this syndrome, but it does not stress the dystonic nature of the disease[2].

A wide range of therapies are available for cervical dystonia, from clinical treatment to brain surgery (pallidotomy and deep brain stimulation) or even peripheral surgery. However, BoNT/A is currently considered the treatment of choice[3].

The objectives of this study were to identify the clinical profile of patients with cervical dystonia who attended the Hospital das Clínicas, UFPR, and to analyze their response to treatment with botulinum toxin A (BoNT/A) in terms of the severity of the motor alterations and pain.

### METHOD

#### Patient selection

Patients with cervical dystonia who attended the Botulinum Toxin and Movement Disorders Outpatient Unit in the Neurology Service, Hospital de Clínicas, Federal University of Paraná, were selected for the study.

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The inclusion criteria were: (1) the presence of cervical or segmental dystonia; (2) the presence of generalized dystonia, hemidystonia or multifocal dystonia, with referral for botulinum-toxin-A treatment for cervical dystonia.

The exclusion criteria were: (1) refusal to submit to diagnostic investigation; (2) failure to agree to the chosen therapy; (3) the presence of hemidystonia, multifocal dystonia or generalized dystonia with referral for surgery or stable with clinical treatment; (4) inability to attend for reassessment; and (5) failure to sign the informed consent form. The exclusion criteria for treatment with BoNT/A were: (1) the presence of myasthenia gravis or other diseases of the neuromuscular junction; (2) the use of aminoglycoside antibiotics; and (3) pregnancy and lactation. All patients signed the informed consent form.

**Clinical assessment**

All the patients were diagnosed with cervical dystonia by one or more neurologists, and this was confirmed by the coordinator for the movement disorders unit (HT). The patients were then assessed by the author by means of a detailed clinical history and a physical and neurological examination to identify clinical characteristics, an association with other movement disorders and neurological diseases, epidemiological data, the time during which the disease had evolved, a history of trauma, the use of medicines, signs and symptoms that might indicate a secondary cause and a family history of dystonia or other movement disorders. Head tremor was classified as dystonic tremor or essential tremor[4].

All the patients were submitted to brain computed tomography and cervical-spine radiography. Additional tests included hemogram, TSH, VDRL, blood glucose test, ESR, electrolyte levels and liver and kidney function in all the patients. Computed tomography of the cervical spine, magnetic resonance imaging of the brain and other laboratory tests were requested according to the clinical assessment of each patient.

The patients were classified according to the clinical presentation of the cervical dystonia (torticollis, laterocollis, retrocollis, anterocollis or combined forms) and whether this was focal dystonia, multifocal dystonia, hemidystonia or generalized dystonia.

For primary dystonia to be diagnosed, the following conditions had to be met: (1) normal perinatal and developmental history; (2) no history of diseases or medication that could have precipitated the appearance of dystonia; (3) no evidence of pyramidal or cerebellar signs, alterations in sensitivity, or cognitive dysfunction on examination; (4) exclusion of secondary causes by specific tests.

The criteria published by Burke et al.[5] were used for tardive dystonia, and those published by Cardoso and Jankovic[6] for posttraumatic cervical dystonia. Secondary causes of cervical dystonia were related to craniocerebral trauma, stroke, encephalitis and brain tumor. Dystonic cerebral palsy was associated with a history of birth trauma and perinatal anoxia.

The presence of dystonia concomitantly with a heredodegenerative disease or neurochemical disorder was considered to be dystonia in a heredodegenerative disease and dystonia-plus[7].

**Treatment**

The clinical treatment patients had been receiving prior to inclusion in the study was continued, and none of the patients were submitted to surgery during the study. Patients received botulinum-toxin-A therapy (Botox®, Allergan, Irvine, CA, USA).

The lyophilized preparation of the toxin (1 flask=100 U=5 ng) was stored at –20ºC and reconstituted with 1 mL of 0.9% saline solution at the time of injection. The 10 U per 0.1 mL solution was administered in a 1cc (1 mL) tuberculin syringe.

Patients who had already used BoNT/A and had reported a subjective improvement continued to receive the same dose. For new patients the initial dose varied from 100 to 280 U. The dose and number of points where the toxin was injected were adjusted for each muscle (Table 1).

The choice of muscle, location and amount of BoNT/A for each muscle were determined based on clinical evaluation with the aid of electromyography (Table 2). To inject BoNT/A, the tuberculin syringe was coupled to a Teflon-coated monopolar electrode needle (37 mm x 27 G) (Oxford Instruments®) connected to a Nihon-Koden multichannel appliance.

**Follow-up**

The patients were assessed on admission and approximately 14, 30 and 120 days after the treatment had started to compare severity, disability and pain using the Toronto Western spasmodic torticollis rating scale (TWSTRS – Severity); Fahn-Marsden dystonia scale (FMS) (for the cervical segment); Jankovic disability scale (JDS); and visual analog pain scale (0=absence of pain, 1–3=mild pain, 4–6=moderate pain, 7–9 = strong pain, 10=disabling pain).

*Table 1. Dose and number of injection points per muscle.*

| Muscle | Dose | Number of points |
|---|---|---|
| Sternocleidomastoid | 15-75 | 2-4 |
| Trapezium | 30-100 | 5-10 |
| Splenius capitis | 15-50 | 2-4 |
| Levator scapulae | 15-50 | 2-4 |
| Paraspinalis | 15-50 | 2-4 |

*Table 2. Muscles chosen for injection of botulinum toxin according to clinical presentation.*

| Subtype | Muscles in which applied |
|---|---|
| Torticollis | Contralateral sternocleidomastoid and ipsilateral trapezium and splenius |
| Laterocollis | Splenius, trapezium and ipsilateral sternocleidomastoid |
| Anterocollis | Bilateral sternocleidomastoid |
| Retrocollis | Trapezium and bilateral paraspinalis |

Adverse effects, latency to start, peak and duration of action of the medication and interference in daily life were measured with a questionnaire.

**Statistical analysis**

The distribution pattern for all the data was tested (normal or non-normal). The statistical differences between the means of the groups were measured using the one-tailed *Student t-test* and ANOVA for normal distributions and the *Mann-Whitney* and *Kruskal-Wallis* tests for non-normal distributions. The correlations were measured using the *Pearson* and *Spearman* correlation coefficients. For the differences between the expected values and the values actually found, the *chi-square* test with *Yates* correction and the *Fisher* exact test were used. The results are given as mean ±SD (standard deviation). The differences were considered significant if $p<0.05$.

*Table 3. Pattern of cervical dystonia in 85 patients.*

| Pattern | Patients |
|---|---|
| **1 – Type** | 33 |
| Torticollis | 21 |
| Laterocollis | 10 |
| Retrocollis | 2 |
| Anterocollis | 0 |
| **2 – Types** | 43 |
| Torticollis + Retrocollis | 19 |
| Torticollis + Laterocollis | 17 |
| Torticollis + Anterocollis | 4 |
| Laterocollis + Retrocollis | 2 |
| Laterocollis + Anterocollis | 1 |
| **3 – Types** | 9 |
| Torticollis + Laterocollis + Retrocollis | 9 |

## RESULTS

### Clinical and epidemiological characteristics

A total of 85 patients with cervical dystonia were included in the study: 51 females and 34 males, giving a ratio of 1.5:1. Forty-five patients (52.9%) only had cervical dystonia while twenty patients (23.5%) also had segmental dystonia, 13 of whom had cranial dystonia, 6 dystonia in the upper limbs, and 1 oromandibular dystonia and dystonia in an upper limb. In the 2 patients (2.4%) with multifocal dystonia, a lower left limb was involved. Hemidystonia was observed in 5 patients (5.9%), and 13 (15.3%) had generalized dystonia.

The age of onset of symptoms varied from 2 to 73 years (mean=34.96±17.4 years), with incidence peaking in the fourth decade of life. The mean age of onset of symptoms in patients with focal dystonia was 40.17±16.42 years, in those with segmental dystonia 37±15.83 years and in those with generalized dystonia 21.53±14.62 years ($p<0.0003$). The incidence of generalized dystonia fell progressively from the first to the fifth decade of life (Fig 1).

With regard to the clinical presentation of cervical dystonia (torticollis, laterocollis, retrocollis and anterocollis), 61.2% of patients had more than one form of cervical dystonia. Of the patients with generalized dystonia, 11 (84.6%) had more than one form of cervical dystonia, with torticollis, which was present in 70 patients (82.4%), being the most prevalent form. The most frequently observed combination was torticollis and retrocollis (Table 3).

The severity of abnormal head and neck movements measured on the TWSTRS scale was greater among patients

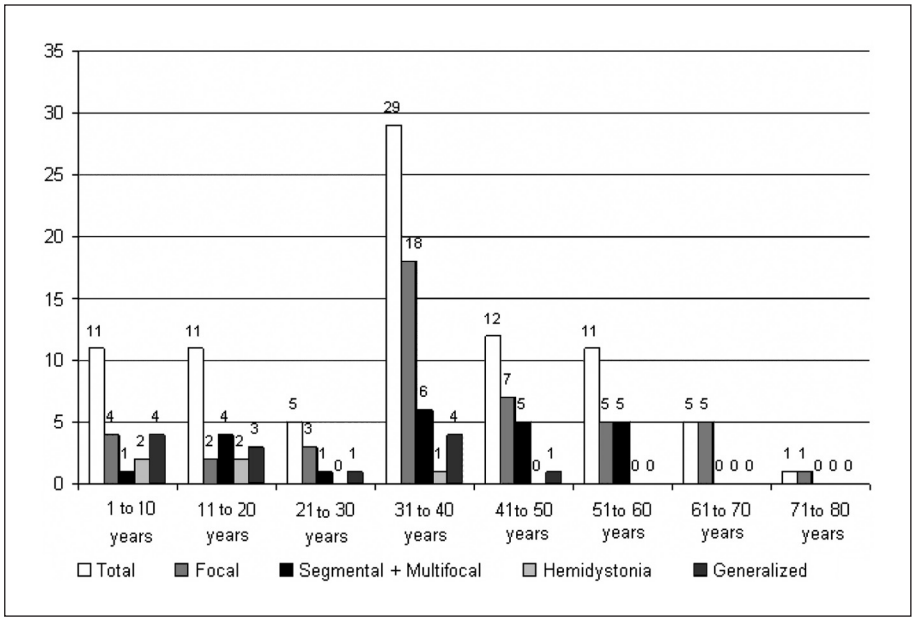


*Fig 1. Incidence of cervical dystonia by age group.*

*Table 4. Etiology by distribution of cervical dystonia.*

| Etiology | Focal | Segmental | Generalized | Hemidystonia | Multifocal | Total |
|---|---|---|---|---|---|---|
| Indeterminate | 31 | 9 | 9 | 2 | 2 | 53 |
| Neuroleptic treatment | 2 | 4 | 1 | | | 7 |
| Perinatal anoxia | 1 | 2 | 2 | 1 | | 6 |
| Craniocerebral trauma | 5 | 1 | | | | 6 |
| Indeterminate with family history | 2 | 3 | | | | 5 |
| Cervical trauma | 3 | | | | | 3 |
| Brain infarct | 1 | 1 | | | | 2 |
| Meningitis | | | | 1 | | 1 |
| Behçet's syndrome | | | | 1 | | 1 |
| Wilson's disease | | | 1 | | | 1 |
| Total | 45 | 20 | 13 | 5 | 2 | 85 |

with generalized dystonia than among those with focal dystonia ($p<0.001$) or segmental dystonia ($p<0.001$). Using the same scale, severity was found to be higher in patients with two or three types of cervical dystonia than in those with only one type ($p<0.001$) and $p<0.0009$, respectively.

Different degrees of pain in the cervical region were reported by 59 (69.4%) of the patients. Of these, 43 (72.9%) reported moderate pain. The presence of spasms and jerks, reported by 60 patients (70.6%), was an aggravating factor ($p<0.0001$).

Comparison of patients with combinations of cervical dystonic movements revealed more complaints of pain in the groups with two and three movements ($p<0.009$ and $p<0.039$, respectively). There was no difference between the number of patients who complained of pain and the level of pain among patients with focal, segmental, multifocal or generalized dystonia or hemidystonia.

"Gestes antagonistes" were reported by only one patient; however, during clinical examination, "gestes antagonistes" such as holding the chin and touching the face were observed in 15 patients (14.6%). These movements rarely lasted for more than a minute.

Some form of tremor was observed in 20 patients (23.5%). Head tremor ('no-no' type) was present in 9 patients (45%) and essential-type tremor of the upper limbs in 3 (15%). A combination of the two types of tremor was observed in 8 patients (40%), and three of these reported a family history of tremor.

Two patients (one with focal dystonia and one with segmental dystonia) had associated Parkinsonism. The latter of these two patients also had a family history of Parkinsonism. Another patient with segmental dystonia had Tourette's syndrome, and his brother and father had a history of tics. The only patient with Behçet's syndrome and hemidystonia also had chorea and hemiballism.

The etiology of the disease could not be determined in 53 patients (62.3%), and the most prevalent etiology found was tardive dystonia (8.2%). In this group of patients, the severity measured on the TWSTRS scale was greater than in patients with other etiologies ($p<0.002$). All the patients with dystonia secondary to neuroleptic treatment had either the isolated form of retrocollis or retrocollis combined with one or two other types of dystonia (five cases and one case, respectively). Five patients (5.9%) without any defined etiology and a mean age at onset of symptoms of 43.5±21.68 years reported cases of cervical dystonia in their families; three of these patients had focal dystonia and two segmental dystonia (Table 4).

**Treatment**

Before treatment with BoNT/A, 48 of the patients (56.5%) had undergone clinical treatment. Anticholinergic agents, particularly trihexyphenidyl (in doses of 6 to 30 mg daily), were the main medication used (42.35%). Eighty-one patients (92.3%) received BoNT/A injections. Four patients (4.7%), who had both been diagnosed and had received BoNT/A previously, with an average disease duration of 3.5±1 years, were considered to be in remission as they did not show any symptoms during the evaluation period (14 months) and therefore did not have any more BoNT/A injections. All the patients who received BoNT/A presented for evaluation at the end of the evaluation period, and 53 (65.4%) presented during the course of the evaluation period.

The mean BoNT/A dose used was 151.05±52.55 U. The greater the severity of the motor symptoms, the more BoNT/A used ($r=0.5$ and $p<0.05$). However, no correlation was found between an increase in BoNT/A dosage and an improvement in symptoms measured on the TWSTRS scale ($r=0.15$ and $p<0.05$). Nevertheless, the group of patients with three types of cervical dystonia benefited from larger dosages of BoNT/A ($r=0.798$ and $p<0.05$).

Thirty-one (36.5%) had not received BoNT/A previously. Of the 54 patients previously treated, 10 (18.5%) had had one treatment session, 25 (46.3%) had had between 2 and 4 sessions and 19 (35.2%) had had more than 5. The previous treatment sessions did not interfere with the assessment of the severity of the disease at the start of the study according to the results using the TWSTRS scale ($p<0.53$), JDS ($p<0.16$) and FMS ($p<0.16$).

There was a similar and highly positive correlation between the results for the response to BoNT/A in relation to the severity of the disease obtained with the three different scales. The correlations observed were as follows: correlation between TWSTRS and JDS: $r=0.82$ and $p<0.05$; between TWSTRS and FMS: $r=0.9$ and $p<0.05$; and between JDS and FMS: $r=0.82$ and $p<0.05$. There was a major improvement in the symptoms of cervical dystonia on the TWSTRS (Fig 2), FMS and JDS scales following treatment sessions with BoNT/A, and these returned to their initial levels at the end of the evaluation. The patients reported a mean time until the medication started to have an effect of 10.07±5.84 days and a mean duration of effect before symptoms returned of 89.15±21.79 days.

BoNT/A also had a beneficial effect in controlling cervical pain. There was a significant improvement in relation to complaints of pain and pain intensity (Fig 3). An improvement in the degree of pain was also observed when the groups with one ($p<0.045$), two ($p<0.0001$) or three ($p<0.026$) types of dystonic movement were analyzed separately. There was no recurrence of previous levels of cervical pain as the analgesic effect of BoNT/A in the group with one dystonic movement was partially maintained at the end of the evaluation ($p<0.094$).

The subgroup of patients with tardive dystonia, whose clinical characteristics and degree of disability differed from those of the other patients in the group, did not respond to treatment with BoNT/A on the TWSTRS scale ($p<0.13$). Patients with posttraumatic cervical dystonia, for whom the duration of trauma before onset of symptoms was 6.66±8.08 years, responded positively to BoNT/A on the TWSTRS scale ($p<0.004$) and reported pain relief ($p<0.002$). In this subgroup there was no change in the analgesic effect of BoNT/A on the 120$^{th}$ day compared with the 30$^{th}$ day ($p<0.3$).

Transitory dysphagia was reported in 2 patients (2.35%). There were no other reports of adverse effects associated with the medication.

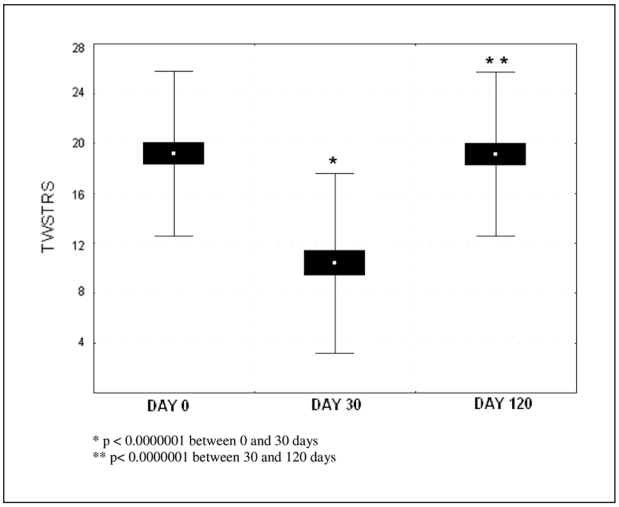


*Fig 2. Response to treatment with botulinum toxin on the TWSTRS scale.*

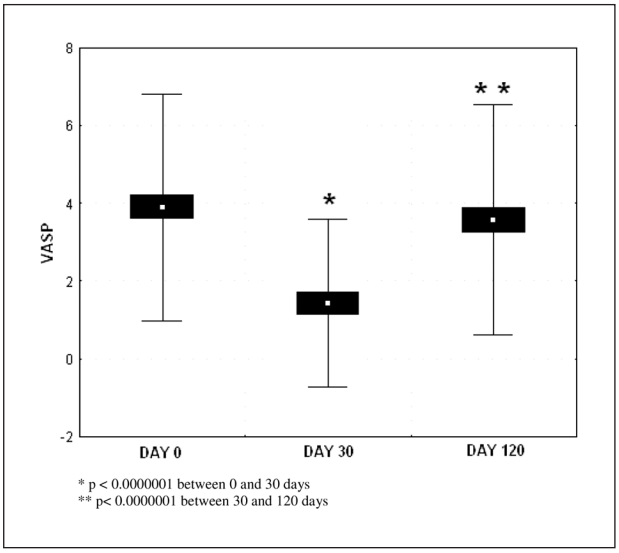


*Fig 3. Response to cervical pain following treatment with botulinum oxin.*

## DISCUSSION

Although cultural and economic issues made it difficult for some patients to present for evaluation during follow-up in this longitudinal study, we were able to assemble a sample that yielded representative data about the clinical characteristics and response to proposed therapy of patients with cervical dystonia. In agreement with previously published studies, we found that cervical dystonia was predominant in females[8,9]. The onset of symptoms occurred mainly between the fourth and sixth decades of life (61.2%)[8,9]. Generalized forms had earlier onset, and focal and segmental forms later onset. Genetic studies have proved helpful in understanding this observation. Dystonias related to the DYT1 and DYT5 genes start as focal dystonias and become predominantly generalized in childhood. Dystonias 7 and 13, which are associated with the DYT7 and DYT13 genes, have later onset and tend to remain focal. When dystonia 13 occurs in childhood, it has a much greater tendency to become generalized[10,11].

Technical and economic limitations prevented us from carrying out genetic tests on the patients in this study.

Some of the patients who were classified as having "indeterminate" etiology may have genetic alterations that were not investigated. Of particular note are the five patients with a family history of focal and segmental dystonia and a mean age of onset of more than 40 years and the two patients with a family history of other movement disorders (parkinsonism and tics).

Tardive dystonia is the main form of secondary dystonia, with the cervical form being the most common[12]. In agreement with published figures, 8.2% of the patients studied were found to have dystonia secondary to the use of neuroleptic agents[9]. The clinical behavior of patients in this group was different from that of the remainder of the sample. All the patients had retrocollis, and 85.71% had more than one type of dystonic movement, two characteristics that could be considered aggravating factors. Our finding that retrocollis was predominant agrees with those of other authors who consider its presence to be highly suggestive of tardive dystonia[13]. Unlike the patients in the study by Molho et al.[13], our patients with tardive dystonia did not respond to BoNT/A treatment, and the size of the sample of patients with tardive dystonia had to be increased to confirm these results.

The natural history of cervical dystonia is not fully understood, and it is important to take great care to separate those patients who are in remission from those who still have the disease. In our study we found that 4.7% of the patients had remission from dystonia during the follow-up period and that all of these had had the disease for less than five years. Other studies have reported that from 12% to 21% of patients with dystonia may have remission[14-16]. The variables that are usually present in sustained remission from cervical dystonia, such as early onset of the disease, predominantly tonic dystonic movements and single-direction head and neck movement were not observed. In addition, there was only one female patient in this group[14,16]. Therefore, longer follow-up of these patients is required to observe the extent to which remission from dystonia is sustained.

Essential tremor is probably the best definition for tremor of the upper members seen in patients with cervical dystonia[4]. The definitions used for tremors varied between studies, making comparisons difficult. We found some type of tremor in 23.5% of the patients, less than the 33% to 71% found in other studies of cervical dystonia patients[9,15]. We found postural tremor in the upper limbs in 3.5% of our patients while Ferraz et al.[17] observed the same type of tremor in 22.2% of their study population. Our finding of dystonic head tremor in 20% of the patients was similar to the 28% found by Chan et al.[8]. The reported incidence of family history of tremor (3.53%) was also different from the 39% observed by Jankovic et al.[9].

Another finding that differed from the literature was the small percentage of patients with "gestes antagonistes" (14.6%), which is as high as 88.9%[18] in the literature. The limited educational background of the patients in our study population may explain this distortion, as these patients are not normally able to relate the onset of dystonia to "gestes antagonistes", and 92% of them were unable to describe when they first realized they were making these gestures[19].

The total number of patients with a family history of movement disorders (11.77%) was also less than the 44% found in the literature[9], probably because of the difficulty most of the patients have in contacting their relatives, their poor socioeconomic background and the absence in the study population (which consists of patients from public health care institutions in Paraná) of any ethnic groups, such as Jews, associated with genetic transmission of dystonias.

The most frequently observed presentation of cervical dystonia was torticollis, followed by laterocollis, retrocollis and anterocollis, as observed in previous studies[8,9]. In addition, as previously described, cases of anterocollis are rare, and a combination of abnormal head and neck postures was more prevalent than only one abnormal posture[9].

Patients with two or three types of dystonic movements had higher severity and pain indexes than those with a single presentation. Patients with generalized dystonia with more than one type of abnormal head movements had higher degrees of severity. Therefore, a larger number of dystonic movements of the neck and larger number of extracervical dystonia sites appear to be aggravating factors in cervical dystonia. In spite of this, patients with generalized dystonia showed a good response to BoNT/A without the need for higher doses.

BoNT/A was very effective at controlling the severity or intensity of pain irrespective of the clinical presentation of the dystonia in the group of patients assessed. The 10-day latency period for the BoNT/A to start having an effect was greater than that reported by Barbosa et al.[20]. The 12-week maximum duration of improvement was similar to the 11 weeks observed by various authors[20,21].

The success of BoNT/A therapy depends on correctly selecting the muscles where it is to be applied and the dose of BoNT/A injected[22]. Although some studies have yielded good results without the use of electromyography, we believe that our use of this technique as a guide to help us choose the muscles in which to inject BoNT/A was important in achieving the excellent clinical response observed in the patients in this study[20]. When used as a guide, electromyography helps locate and choose the muscle, optimizes the BoNT/A dose and reduces the incidence of adverse effects[23].

The BoNT/A dose, which averaged 151 U, could be optimized by choosing the correct muscle group, with larger doses only being of benefit for patients with three combined forms of dystonic movements. Jankovic et al.[24], using an average dosage of 209 U and without the aid of electromyography, obtained very good results, which we managed to reproduce with lower doses of BoNT/A. In our study, a larger proportion of patients had an improvement in their dystonia and fewer complications than in studies by other authors who used electromyography[21].

The choice of the correct muscle groups with the aid of electromyography and the use of low dosages of BoNT/A may account for the few adverse effects reported, as only two patients reported dysphagia. In a study by Jankovic et al.[24], in which the authors used higher average doses of BoNT/A and did not use electromyography guidance, 24% of the patients had adverse effects, and 23% of these had dysphagia. In a study by Barbosa et al.[20], in which the average dose was 191 U and electromyography was not used, 47% of the patients developed dysphagia. Other factors may also have contributed to the low dysphagia indexes in our study population. Firstly, a larger number of application points in each muscle and simultaneous application in both sternocleidomastoids in only 16% of patients (thus reducing diffusion of the medicine to the pharynx) can reduce the incidence of dysphagia[22]. Secondly, it is not easy for patients with cervical dystonia to notice dysphagia, so that it is very often underdiagnosed[25].

The high incidence of pain distinguishes cervical dystonia from other focal dystonias and contributes significantly to patient disability[8]. Different degrees of pain in the cervical region were reported by 69.4% of the patients, a similar figure to those reported previously in the literature[15,26] . BoNT/A was highly effective in controlling pain, and its analgesic effect was sustained for a long time in patients with fewer disability factors.

We conclude that BoNT/A is an effective and safe treatment for cervical dystonia despite the heterogeneous and complex presentation of this disease. Our findings agree with reports in the literature that BoNT/A improves the quality of life of patients with dystonia[27,28].

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