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Dystonias: rehabilitation

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DESCRIPTION OF THE EVIDENCE COLLECTION METHODOLOGY

Articles in the MedLine (PubMed) database and other research sources were reviewed, with no time limit. The search strategy used was based on structured questions in the PICO format from the initials: Patient, Intervention, Control and Outcome. The descriptors used were: dystonia and (benzodiazepines or baclofen or tizanidine or clodinine): dystonia and (anticholinergics or haloperidol or lisuride); dystonia and (botulinum toxin); dystonia cervical and (speech therapy or speech pathologist or botulinum toxin); focal dystonia and (botulinum toxin); dystonia and (sensory stimulation or sensory rehabilitation); dystonia and (biofeedback or electromyography biofeedback); dystonia and (transcranial magnetic stimulation); (dystonic disorders or dystonia) and (self help devices or assistive technology or assistive technologies or rehabilitation); dystonia and (activity daily living); dystonias and (neurosurgery not intrathecal baclofen)

QUALITY OF EVIDENCE AND STRENGTH OF RECOMMENDATIONS:

A: Experimental or observational studies of highest quality.

B: Experimental or observational studies of lower quality.

C: Case studies (uncontrolled studies).

D: Opinion with no critical evaluation, based on consensus; physiological studies, or animal models.

OBJECTIVE:

To provide information about dystonia and guidance on its treatment and rehabilitation.

PROCEDURES:

Therapies for distonia

CONFLICT OF INTEREST:

The authors have no conflicts of interest to declare.

INTRODUCTION

Movement disorders comprise a group of diseases classified as extrapyramidal and are associated with involuntary movements or abnormalities of skeletal muscle tone, posture, or both. Movement disorders may be divided into two clinical categories: those that show paucity of movement (hypokinesia), and those with excessive abnormal involuntary movements (hyperkinesia).[1]

Dystonias are movement disorders characterized by muscle contractions that cause repetitive torsional movements with varying speed, leading to an abnormal posture. The basal ganglia play an important role in the pathophysiology of dystonia, explaining sensorimotor alterations such as the presence of pain, burning sensations, and paresthesia, which may precede the onset of muscular contractions.[2]

Dystonia can be classified according to age of onset, etiology and anatomical distribution. Based on etiology dystonia may be primary, for instance hereditary or sporadic, or secondary, associated with neurological disorders.[1,2] It can be: focal, when it involves only specific parts of the body, for example blepharospasm and writer's cramp; segmental, when it involves two or more contiguous parts; multifocal, when in involves various unrelated parts of the body; and generalized dystonia involving segmental crural and at least one other part of body.[1,2]

The goal of this guideline is to provide recommendations for the treatment of dystonia.

1. What is the efficacy of muscle relaxants (benzodiazepines, baclofen, tizanidine, clonidine, and anticholinergics) for treating dystonia?

In a study conducted with 11 patients with clinical and electromyographic diagnosis of spasmodic torticollis, the results were compared between treatment with dosages of up to 12 mg/day tizanidine *versus* placebo for 6 weeks with each drug, with an interval of 1 week between each period. There was no significant clinical improvement observed in any of the patients[3] **(B)**.

In a study of 19 patients with tardive dyskinesia due to chronic use of neuroleptics, the effects of clonazepam in dosages of 2 to 4 mg/day were compared with placebo for a period of 12 weeks. A 35% reduction on the dyskinesia scales was observed, being the most obvious benefit for patients in whom dystonic symptoms predominated[4] **(B)**.

In a study of 11 patients with Meige syndrome (blepharospasm/oromandibular dystonia), the use of medications intravenously showed significant improvement in the blepharospasm indices in response to biperiden and clonazepam injection. The evaluation of each patient did not help in predicting the therapeutic potential of these drugs for subsequent oral treatment[5] **(A)**.

In a study of 64 patients diagnosed with idiopathic cervical dystonia, the effectiveness of treatment with botulinum toxin and trihexyphenidyl was compared. Patients were divided into 2 groups of 32 subjects, with one group receiving injections of botulinum toxin combined with placebo pills and the other group saline injections combined with trihexyphenidyl tablets. Clinical evaluations after 12 weeks showed significant improvement in favor of botulinum toxin and significantly less frequent adverse effects with this therapy[6] **(B)**.

Recommendation

The therapeutic arsenal for the treatment of dystonic patients was for years restricted to a limited number of oral medications whose efficacy is highly variable from patient to patient, and have major limitations in their use due to the high frequency of intolerable side effects. Existing studies on this subject are few, and generally old **(D)**. Currently, the treatment of choice for focal dystonias is botulinum toxin[7] (**A**). However, in patients with generalized dystonia, in whom it is not possible to use toxin in all the muscles involved, the combined use of some drugs, notably clonazepam, with careful monitoring of dosages (from 2 to 4 mg/day) and side effects, is recommended and can promote a 35% reduction on dyskinesia scales[5] **(A)**. There is a need for broader studies to better recommend this treatment **(D)**.

2. What is the efficacy of local application of botulinum toxin for cervical dystonia?

Positive results were observed regarding the use of botulinum toxin type A (XEOMIN) in a cervical dystonia study with 233 subjects, 78 who received botulinum toxin 120U, 81 who received botulinum toxin 240U and 74 with placebo treatment. The subjects were evaluated at baseline and follow-up at 4 weeks, 8 weeks, and final visit, using TWSTRS evaluation. One week after treatment, subjects were contacted to investigate changes in symptoms or adverse events. As the primary outcome the difference in the results were analyzed, comparing baseline to followup at week 4. For the secondary outcome the difference in the results between Week 4 and Week 8 follow-up were compared. The final visit showed that both Type A toxin doses were more effective than placebo treatment, with $p < 0.001$ for the 3-item TWSTRS subscale (severity, disability and pain) at all three moments (followup at 4 weeks, 8 weeks, and the final visit). There was no significant difference in the results comparing the applied doses (120U and 240U). The adverse effects, such as neck pain, dysphagia, and muscle weakness, were mostly mild or moderate and did not show a significant result in the study[8] **(A)**.

Positive results were observed regarding the use of botulinum toxin type A (Abobotulinumtoxin A Dysport) in cervical dystonia in a study with 116 subjects, 55 with botulinum toxin application (with 4 cycles of treatment) and 61 with placebo treatment. The subjects were evaluated at baseline and follow-up at 4 weeks, 8 weeks and 12 weeks, using TWSTRS, VAS, SF36, Quality of Life Questionnaire and Subject Evaluation measures. The outcome showed that treatment with botulinum toxin A was more effective than placebo treatment, with $p < 0.001$ in the first followup at week 4, using the TWSTRS scale, $p = 0.019$ at week 12. The adverse effects, such as neck pain, dysphagia, and muscle weakness, were mostly mild or moderate and did not show a significant result in the study[9] **(A)**.

Positive results were found for the use of botulinum toxin (Rimabotulinumtoxin B MIOBLOC) for pain relief in cervical dystonia in three studies. The first (AN072-301), was performed with 109 subjects responsive to treatment with BoNT-A, 36 of whom received BoNT-B 5000U, 37 receiving BoNT-B 10000U, and 36 treated with placebo. The second study (AN072-302), was performed with 77 subjects resistant to treatment with BoNT-A, 39 receiving BoNT-B 10000U and 38 receiving placebo. The third study (AN072-402) was done with 111 subjects, 56 treated with BoNT-B 10000U and 55 with application of BoNT-A 150U. The subjects were evaluated at baseline and follow-up by TWSTRS evaluation. The outcomes showed that treatment with RimabotulinumtoxinB was more effective than placebo or BoNT-A treatment for improving pain, with $p < 0.001$[10] **(A)**.

Positive results were observed regarding the use of Botox (BoNT-A) and Myobloc/Neurobloc (BoNT-B) in a cervical dystonia study with 111 subjects, 55 treated with BoNT-A (150U) and 56 with BoNT-B (10000U). The subjects were evaluated at screening and follow-up at 4 weeks and 8 weeks by TWSTRS and VAS evaluations. The outcome showed that treatment with BoNT-A is as effective as with BoNT-B, with no difference between both series of results, neither in the duration of the effects of the injections, nor in the improvement in pain or adverse effects. However, comparing the results of the evaluations performed at screening and after treatments, both groups showed a significant result, with $p < 0.001$ at followup at 4 weeks and 8 weeks[11] **(A)**.

Positive results were observed regarding the use of BoNT-A combined with a specific program of rehabilitation therapy for cervical dystonia in a study with 40 subjects, 20 with botulinum toxin 100-200U (BTX-0 group) and 20 with botulinum toxin 100-200U combined with specific rehabilitation therapy (stretching, massage, biofeedback) for 2 weeks, with daily sessions of 60-90 minutes (BTX-PT group). The subjects were evaluated at baseline and follow-up with TWSTRS, Tsui scale, ADL and Total Pain Score evaluations. After treatment, subjects were contacted to investigate changes in symptoms or adverse events. The outcome showed that both treatments were effective, but the BTX-PT group showed greater efficacy for duration of the effects of the injections, both in improvement of pain and for performance of activities of daily living ($p < 0.001$)[12] **(A)**.

Recommendation

The botulinum toxin (type A or type B) is recommended as the treatment of choice for cervical dystonia, showing greater efficacy when combined with physical rehabilitation therapy (with physiotherapy, occupational therapy, speech-language pathology, etc.) resulting in improvement in pain and disability. Variations in muscle selection and dosage (100U to 1000U) during treatment may generate differing results[8-12] **(A)**.

3. Does intervention with speech-language pathology minimize the effect of collateral dysphagia after application of botulinum toxin in cervical dystonia patients?

Dystonia is a syndrome characterized by sustained muscle contractions causing twisting, repetitive movements, or abnormal postures that can affect most of the voluntary muscles. The most common presentation is cervical dystonia which affects the neck muscles in a localized manner, or combined with other parts of the body. The use of botulinum toxin type A has shown positive results in

studies, the majority of which cite dysphagia as the main side effect after botulinum treatment[9-13] **(A)**,[14] **(B)**.

Recommendation

In clinical practice it is observed that in some cases there is the side effect of dysphagia after application of botulinum toxin in patients with cervical dystonia[9-13] **(A)**,[14] **(B)**. Although transient, dystonia causes serious discomfort for patients, which intervention with speech pathology has shown good results towards minimizing. However, no evidence was found of the action of speech pathology on the neurochemical block in these patients, hence studies in this line of therapeutic intervention are necessary **(D)**.

4. What is the efficacy of local treatment with botulinum toxin in focal dystonia (writer's cramp)?

Positive results were observed with the use of botulinum toxin in focal dystonia/writer's cramp in 40 patients (20 treated with botulinum toxin injection and 20 with placebo), evaluated at baseline and follow-up at 1 year. As the primary outcome measure the patient's opinion was taken into consideration, and a total of 14 patients from a group of 20 who received toxin injections (with an average dose of 178 units in two sessions) opted to continue treatment given the results achieved. The secondary outcome measures considered clinical evaluations that showed improvement results with the follow-up scores of the rating scales as dependent variables and adjusting for baseline covariates, including providing for the baseline values of the assessment scales that showed treatment effects in improvement of pain (during handwriting activity) by visual analogue scale (VAS p = 0.02), improvement by the writer's cramp rating scale (WCRS p = 0.01) and in writing speed (p = 0.04). On the symptom severity scale (SSS) the follow-up scores were close to the limit of significance (p = 0.06), whereas no significant beneficial therapeutic effect was observed in relation to functional status scale (FSS p = 0.14). Side effects such as weakness of the hand have been reported, mostly mild and transient, and pain at the injection site[7] **(A)**.

Recommendation

Considering the improvement both in pain during writing, and severity of cramps (60-90%) observed with injection of botulinum toxin type A (BoNT-A, at a mean dose of 178 units in two sessions), it is recommended as the treatment of choice for most types of focal dystonia (writer's cramp)[7] **(A)**, when available **(D)**.

5. What is the efficacy of therapeutic exercise (sensory/proprioceptive stimulation, muscle strengthening) for dystonia patients?

Dystonia is a movement disorder characterized by sustained muscle contractions causing twisting, repetitive movements or abnormal postures. Although it manifests as a motor disorder, clinical observations suggest the involvement of sensory systems in distonia. Dystonia is commonly associated with sensory symptoms (pain, tension, and burning sensation), which may precede the onset of muscle contractions. The tactile and proprioceptive input to a specific area may improve dystonia: this phenomenon, called "sensory trick" is present in up to 64% of patients with cervical dystonia and has been described in other focal dystonias such as blepharospasm, oro-mandibular dystonia and writer's cramp. Although basic sensation is usually normal in patients with dystonia, a thorough evaluation can demonstrate abnormalities in specific functions such as spatial discrimination, graphesthesia, and stereognosis. It has been suggested that the excessive input of abnormal sensory stimuli due to a lack of good spatial and temporal integration could create a loss in the generation of output of motion, thus causing the abnormal motion characteristics of dystonia. Treatment with BoNT-A, despite showing clinical improvement, produced no change in the somatosensory aspects[3] **(B)**.

Despite the existing evidence on the motor and sensory changes caused by different types of dystonia, no studies were found in which sensory stimulation and muscle strengthening were used as a treatment for dystonia **(D).** In addition, no improvement was found in sensory aspects with neurochemical block by botulinum toxin, despite clinical improvement[3] **(B)**.

In training with specific tasks for focal dystonia in musicians, the extensive use of the muscles involved seems to result in a gain of motor control and reduction of the symptoms of dystonia by cortical organization, which can be modified by repeating the tasks[15] **(B)**.

Recommendation

Training with specific and repetitive tasks for musician's dystonia can promote motor control gains and reduction of symptoms[15] **(B)**. However, greater clinical research is required, as well as a structured study on the possible benefits of clinical practices such as muscle strengthening and sensory/proprioceptive stimulation of regions involved in the dystonia to prove the effectiveness of using these resources in dystonia patients **(D)**.

6. Does the use of electromyographic biofeedback with dystonia patients have efficacy?

Biofeedback applied in 15 patients with torticollis, including 3 evaluation sessions, 15 treatment sessions, 6 home sessions with supervision, and 6 home sessions without supervision, with follow-up 3 months after treatment, showed greater efficacy in reducing muscle activity and relaxing the muscles of the neck, than only relaxation training and Global Postural Recalibration[16] **(B)**.

Auditory biofeedback applied for 7 hours of training in 7 patients diagnosed with writer's cramp-focal dystonia showed improvement in performance and a significant reduction in writing pressure and pain[17] **(B)**.

Recommendation

The use of biofeedback appears to be a therapeutic resource with results in the reduction of muscle activity (reporting 20-30% improvement), relaxation and postural correction, when applied for 15 supervised sessions and 6 unsupervised sessions[16] **(B)**. It also can be used as a resource for reducing pain in patients with writer's cramp, as well as improving performance during writing, with a reduction in pressure from 48.7N to 8.9N[17] **(B)**.

7. What is the efficacy of transcranial magnetic stimulation in dystonia patients?

Dystonia is a disorder of the central nervous system, whose main characteristic is involuntary movements (spasms), which make movement difficult, and can lead to alterations in function and muscle structure in part or all of the body. Studies show that the

motor cortex of patients with dystonic symptoms display cortical hyperexcitability. Therefore, a treatment that could decrease cortical excitability in the motor area could have a positive effect on the clinical picture of these patients, such as transcranial magnetic stimulation (TMS).

TMS is a non-invasive technique based on the principle of electromagnetic induction, i.e., that a powerful and constantly alternating electrical current passing through a coil is capable of generating a magnetic field. This field traverses the bony structures and soft tissues and induces an electric current of focal action within the cerebral cortex. There are two types of TMS: one using a single pulse and one which uses a repetitive pulse-repetitive transcranial magnetic stimulation (rTMS). In single pulse TMS, a single current is delivered to the cortex. With rTMS, the pulses are applied with a constant frequency ranging from 0.1 Hz to 60 Hz; and the higher the frequency and intensity of stimulation, the greater will be the interference in cortical function during the period of stimulation[18] **(B)**.

When applied to the motor cortex, rTMS produces a muscular response in the contralateral limb. Series of rTMS pulses can induce a modulation in cortical excitability, which can vary between inhibition and facilitation of cerebral activity, depending on the frequency and parameters of rTMS used.[19-21] Low frequencies of rTMS (such as 1 Hz or less) applied to the motor cortex, for example, cause a suppression of cortical motor excitability.[22] A 20 Hz stimulation (high frequency), on the other hand, can lead to a temporary increase in cortical excitability[19,23] **(B)**.

The blepharospasm is a common form of focal dystonia and is characterized by excessive, involuntary closure of the eyelids. Traditionally, dystonia is a pathology caused by the basal ganglia. However, recent studies suggest that blepharospasm is due to pathological changes located in the anterior cingulate cortex (ACC). When rTMS is applied over the ACC for the treatment of blepharospasm, with conventional (C-coil) and H-coil, at 0.2 Hz, 180 stimuli, 15 minutes per session, with a stimulator output equivalent to 100% of active motor threshold, it is effective when compared to placebo using a C-coil in improvement of the clinical aspects: eye blink rate, number of sustained blinks, time of eyelid closure (p = 0.023) both immediately after intervention and 1 hour later (T2 and T3: p = 0.250), indicating that the effects of stimulation remain until 1 hour after the intervention. When compared to placebo stimulation, C-coil and H-coil stimulations are more effective (p = 0.008, p = 0.024, respectively), however there is no significant difference between the two ($p > 0.05$)[24] **(A)**.

Use of rTMS is also effective in improving symptoms reported by patients (p = 0.002), both immediately after the intervention and 1 hour later (T2 and T3: p = 0.659). Comparing placebo stimulation with conventional and H-coil stimulation, there is also evidence of improvement of the symptoms reported by patients (p = 0.007 and p = 0.001, respectively). There is no difference between conventional and H-coil stimuli[25] (**A**). With relation to recovery of blink reflex, rTMS is effective when compared to placebo, but there is no evidence subjects maintain the effects 1 hour after intervention (T2 and T3: p = 0.001). The conventional and H-coil stimulation are more effective than placebo stimulation (p = 0.005 and p = 0.007 respectively), but there is no difference between the two interventions[24] **(A).**

There are no significant adverse events arising from the use of the three types of stimulation in the treatment of blepharospasm[24] **(A)**. The motor threshold for H-coil stimulation is significantly lower when compared to placebo and conventional coil stimulation[24] **(A)**.

Focal dystonia of the hand is a condition characterized by involuntary contraction and co-contraction of the muscles of the hand that causes decreased fine motor control and abnormal posture of the hand. Writer's cramp and musician's dystonia are two prevalent types of focal hand dystonia that involve repetitive and stereotypical movements. Studies show that focal hand dystonia is due to the absence of synaptic inhibition of the central nervous system, which leads to cortical disorganization evidenced by abnormal representation of the hand in the primary motor cortex, as well as compromised sensory integration[25] **(A)**.

The treatment of focal hand dystonia with rTMS (1 Hz) for five consecutive days, with 900 stimuli (at 90% of motor threshold) applied per session to the premotor cortex with a 70 mm, single-phase, figure eight coil improves the cortical silent period, i.e., improves cortical inhibition of patients with symptoms of focal hand dystonia compared to placebo stimulation (p = 0.002), with gains maintained up to 10 days after treatment[25] **(A)**.

In a treatment of 5 consecutive days with biphasic stimulation (1 Hz, 90% of motor threshold) where 9000 total stimuli are given, 1800 stimuli per 30-minute session, with a figure eight coil positioned over the primary somatosensory cortex contralateral to the hand compromised, there is evidence of subjective and objective improvement in writing, especially immediately after the end of the intervention ($p < 0.01$), with up to three weeks duration. Functional MRI results also show greater activation of areas in various regions of both hemispheres, i.e., there is an increase in BOLD (blood-oxygenation-level-dependent) signal bilaterally in the posterior parietal cortex and supplementary motor area ($p < 0.001$)[26] **(B)**.

In the protocol of five consecutive days rTMS also improves writing speed when compared to placebo stimulation (p = 0.05) up to 10 days after intervention (p = 0.007)[25] **(A)**.

There is no evidence of improvement in pressure and flow of writing. There is no evidence of improvement of symptoms reported by patients with focal hand dystonia. And there is no evidence of adverse effects[25] **(A)**.

Recommendation

Low frequency rTMS (less than or equal to 1 Hz) is effective in treating focal dystonias such as blepharospasm and focal hand dystonia. There is evidence of improvement of cortical inhibition in focal hand dystonia, as well as improved writing speed[25] **(A)**. On the other hand, there is greater evidence of improvement in the clinical aspects related to blepharospasm, such as improvement of eye blinks, number of sustained blinks, time of eyelid closure, and also improvement in the symptoms reported by patients[24] **(A)**. And there is no evidence of adverse effects from the use of rTMS in the treatment of focal dystonias[25] **(A)**.

The protocol of 5 consecutive days of monophasic or biphasic stimulation (1 Hz, 90% of motor threshold), provides evidence that there are gains related to improved subjective and objective writing up to 10 days after intervention[25] **(A)**,[26] **(B)**. There is evidence that rTMS in the primary somatosensory cortex can promote

improvement in symptoms of focal hand dystonia in parallel with increased cortical activity in both hemispheres[26] **(B)**.

Finally, there is a scarcity of controlled studies to better assess the efficacy of rTMS in the treatment of focal dystonias, even more so in relation to the long-term effects of this intervention. It is therefore suggested there be further studies performed on the use of rTMS in treating focal dystonias **(D)**.

8. Do assistive technology resources facilitate the performance of the activities of daily life for the dystonia patient?

The use of assistive technology resources and adaptive equipment can help patients with dystonia perform basic and instrumental activities of daily living, work, and leisure, allowing the patient to achieve the maximum of functional independence possible. Technology is considered "assistive" when it can assist in the functional performance of activities and reduce disability to perform activities of daily living in the different spheres of everyday life, by expanding communication, mobility, control of the environment, and learning and job skills[27,28] **(D)**.

Recommendation

In clinical practice it has been possible to achieve positive results with the use of assistive technology resources, orthoses, and adaptations, including low cost solutions, to achieve functional independence in patients with dystonia, both focal and generalized. However, no evidence was found that supports such a strategy in the practice of rehabilitation of people with dystonia, creating a need for studies in this line of therapeutic intervention **(D)**.

9. Does functional training in the activities of daily life benefit occupational performance in dystonia patients?

Activities of daily living (ADLs) are an important component of the role of personal maintenance. The term "basic activities of daily living" (bADL) corresponds to personal care, relating to the care of the individual for himself, and includes activities such as personal hygiene and self-care, bathing, feeding and eating, dressing, functional mobility (position changes, moving about, and walking), sphincter control, toilet use, care of personal equipment, sexual activity, sleeping, and resting.[29,30] In rehabilitation the progress in the performance of such activities can be measured by functional evaluations and other assessment tools.[30-32] The instrumental activities of daily living (IADL) also make up an important factor in personal maintenance and restoring independence. They are activities related to problem-solving skills, social skills, and interaction with the environment, such as: care for others, care for pets and domestic animals, raising and caring for children, the use of communication equipment, meal preparation and cleanup, home administration, health management (care and maintenance), using environmental control devices, shopping, financial management, and community mobility[29-32] **(D)**.

The training of these activities and of the tasks that the patient hopes to accomplish (independently, as much as possible), as well as the techniques and devices necessary for their implementation, are aimed at providing a basic repertoire of skills for coping with their daily lives during the rehabilitation process. Thus, functional training for patients who have dystonia may benefit their occupational performance considering their social context, personal values and life history, and considering the different occupational and social roles it can play[29-32] **(D)**.

Recommendation

Clinical experience has shown that guidance aimed at functional training of basic and instrumental activities of daily living with patients who have dystonia may benefit their occupational performance (with reduced energy expended during such activities, for example) considering their social context, personal values and life history, as well as considering the different occupational and social roles it can play.[29-32] It is also possible to observe that such functional training combined with the use of assistive technology resources has benefited such intervention in the rehabilitation process. However, no evidence was found that supports such a strategy in the practice of rehabilitation of people with dystonia, creating a need for studies in this line of therapeutic intervention **(D)**. It is observed that application of botulinum toxin can help with performance in activities of daily living by reducing pain during tasks[12] **(A)**.

10. Is deep brain stimulation (DBS) an effective alternative in the treatment of dystonia?

Deep brain stimulation (DBS) has become an alternative therapy to drug treatment for refractory movement disorders since the 1980s, and since then the number of pathologies in which this technology has been successfully applied is expanding. The locations of the CNS to which DBS is applied vary according to the condition; the stimulation of "pars interna" of the globus pallidus and the subthalamic nucleus was approved by the FDA in April 2003 for treatment of chronic primary dystonia, unresponsive to drug therapy. The advantage of DBS over ablative surgery is the possibility of adjustments and the fact that it is reversible. Thus, in the last decade it has become the surgical treatment of choice for movement disorders. These are criteria for election of dystonic patients for treatment with DBS: symptoms that cause significant disability despite the use of systemic medications at maximally tolerated dosages; good clinical condition; absence of structural brain injury, dementia or severe psychiatric disorders; absence of fixed contractures; age minimum of 7 years. The FDA approval covers primary generalized dystonia, segmental dystonia, hemidistonia, and cervical dystonia, but DBS also shows improvement in patients with Meige syndrome and tardive dystonia[33] **(A)**.

The preferred target for DBS in dystonic patients is the posteroventral lateral portion of the globus pallidus, because, although not completely clear, the pathophysiology of dystonia is related to abnormal activity of the internal globus pallidus. The result obtained is a slow and progressive improvement for days to months, especially when tonic and fixed abnormal postures are present. Because the therapeutic results can last for more than 10 hours after cessation of the stimulus, it is speculated that its effect is not entirely due to the direct action of stimulating nervous tissue, but at least partly due to structural and/or functional changes to the cortical and subcortical circuits caused by the stimulus[33] **(A)**.

Systematic reviews show that DBS is at least partially effective for the treatment of the following: primary segmental dystonia, primary generalized dystonia, cervical dystonia, blepharospasm, Meige

syndrome, tardive dystonia, some forms of secondary dystonia, DYT1-positive and DYT1-negative dystonia, and post-traumatic dystonia. There is no efficacy in dystonia caused by perinatal injury and encephalitis. Unlike what happens in DBS for Parkinson's disease, DBS for dystonia is not associated with cognitive impairment, even showing some improvement in executive function, which can be related both to lower dosages of the medications used as well as the lower average age of these patients[33] **(A)**.

In a review of articles on DBS, in the majority of studies the most commonly used scales were the Burke-Fahn-Marsden dystonia rating scale and the Toronto Western Spasmodic Torticollis Rating Scale. The search yielded 157 studies reporting results with DBS, in a total of 466 patients with different forms of dystonia. A single double-blind study of DBS for primary generalized dystonia showed an average reduction of 41.9% in the severity of dystonia. In contrast, patients with post-traumatic dystonia or dystonic cerebral palsy seem to respond less to DBS, with a few rare exceptions, and the response appears to be unrelated to age, duration of symptoms or initial severity.

As for the evaluation of quality of life after intervention, the studies used objective scales, but do not necessarily reflect the subjective evaluation of the patient. Among 22 patients with primary generalized dystonia, there were significant improvements in general health, physical function, and body pain identified on the SF-36 scale. In a double-blind study of DBS in 40 patients with segmental or generalized dystonia, there was a significant improvement in the SF-36 physical composite score at 3-month follow-up. In relation to the adverse effects of DBS, many studies do not report in sufficient detail, making attempts to quantify such effects difficult. There are reports of lead migration, dysarthria and occasional bleeding.

The most significant and consistent results found are in relation to the cases of generalized dystonia. And although four studies present results of up to 10 years post-DBS showing long-term benefit, data are still lacking related to long-term monitoring of patients who have undergone DBS. Only two of the studies had blinded evaluations, in most studies evaluations were open, which may suggest an observer bias in the results. Thus, DBS has shown benefit for the majority of dystonia patients reported in the literature, even if the improvement rate is still quite variable[34] **(B)**.

Recommendation

There are beneficial, if variable results, for patients with dystonia who undergo deep brain stimulation (average reduction of 41.9% in the severity of generalized dystonia)[33,34] **(B)**. Perhaps such a therapeutic intervention is a possibile alternative in clinical practice with regard to the rehabilitation process of those patients who do not respond to other therapies currently available. However, there are controversies in the results found, mainly with respect to long-term gains, and the need to adapt such treatment to the Brazilian reality **(D)**.

11. Does a psychological approach improve the performance of the activities of daily life in dystonia patients?

It is known that involuntary activation deteriorates according to the patients' emotional state. Thus, it is assumed that the greater the emotional balance these patients attain, the lower the dystonic activation will be, and the better their functional performance. In clinical practice an approach incorporating psychological factors in the rehabilitation has proven crucial in improving the performance of patients with dystonia **(D)**.

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