Botulinum Toxin in Treatment of Focal Hand Dystonia

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Phenomenology and Classification of Dystonia: A Consensus Update

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Mov Disord 2013;28:863-73

ABSTRACT: This report describes the consensus outcome of an international panel consisting of investigators with years of experience in this field that reviewed the definition and classification of dystonia. Agreement was obtained based on a consensus development methodology during 3 in-person meetings and manuscript review by mail. Dystonia is defined as a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both. Dystonic movements are typically patterned and twisting, and may be tremulous. Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation. Dystonia is classified along 2 axes: clinical characteristics, including age at onset, body distribution, temporal pattern and associated features (additional movement disorders or neurological features); and etiology, which includes nervous system pathology and inheritance. The clinical characteristics fall into several specific dystonia syndromes that help to guide diagnosis and treatment. We provide here a new general definition of dystonia and propose a new classification. We encourage clinicians and researchers to use these innovative definition and classification and test them in the clinical setting on a variety of patients with dystonia. © 2013 Movement Disorder Society

Key Words: dystonia; classification; definition
**Phenomenology and classification of dystonia: A consensus update.**

**AXIS I**
Clinical Characteristics

**AXIS II**
Etiology
- Inherited
- Acquired

Jinnah HA, Albanese A. Mov Disord Clin Pract 2014;1:280-4

**Overall prevalence of primary dystonia is estimated at 16.4 per 100,000**

Steeves et al. Mov Disord 2012;27:1789-96

<table>
<thead>
<tr>
<th>Type of dystonia</th>
<th>n</th>
<th>Sex, M/F</th>
<th>Mean age of onset, range (yr)</th>
<th>CPR* per 100000 with 95% CI</th>
<th>Standardized rate†</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blepharospasm</td>
<td>3</td>
<td>0.1</td>
<td>Men: 57.6 (52-63)</td>
<td>5.72 (1.18-16.02)</td>
<td>7.22 (1.49-21.70)</td>
</tr>
<tr>
<td>Cervical dystonia*</td>
<td>2</td>
<td>1:1</td>
<td>Men: 58</td>
<td>3.81 (0.46-13.75)</td>
<td>3.96 (0.48-14.30)</td>
</tr>
<tr>
<td>Writer’s cramp</td>
<td>11</td>
<td>4:5:1</td>
<td>Mean: 41.1 (14-60)</td>
<td>21.00 (10.48-37.57)</td>
<td>21.14 (10.55-37.82)</td>
</tr>
<tr>
<td>Wring tremor</td>
<td>7</td>
<td>2:5:1</td>
<td>Men: 62.6 (48-75)</td>
<td>3.15 (0.55-5.29)</td>
<td>14.85 (5.95-30.59)</td>
</tr>
</tbody>
</table>

*One case in each category.
†Age standardization to world standard population.
‡Crude prevalence rate; CI, confidence interval.

23 of 52,377 screened subjects had isolated dystonia
Das et al. Mov Disord 2007;22:2031-6
Clinical and Demographic Characteristics of Upper Limb Dystonia

Norris et al. Mov Disord 2020 (on line)

- Clinical and demographic characteristics of 367 (mean age at onset: 37.5 ± 18.1) participants in the Dystonia Coalition project with upper limb dystonia were evaluated.
- 67% had task-specific dystonia and 42% noted alleviating maneuver.
- Overall 57% were female, but 77% with musician’s dystonia were men, whereas only 38% of those with writer’s cramp were men.
- Focal onset occurred in 80%; 67% remained focal without spread. Task specificity was most frequent in this subgroup, most often writer’s cramp affecting the dominant limb (83%).
- Focal onset with spread was more frequent in young onset (<21 years).
Risk of spread in adult-onset isolated focal dystonia: a prospective international cohort study.
Berman et al. J Neurol Neurosurg Psychiatry 2020;91:314-20

![Graph showing probability of no dystonia spread over duration (years)]

Number at risk
- Cervical: 286, 270, 231, 176, 122, 87, 41, 20, 8, 4, 1
- Blepharospasm: 116, 87, 65, 43, 25, 17, 5, 3, 2, 1, 0
- Hand: 47, 42, 35, 29, 22, 9, 5, 2, 1, 0
- Laryngeal: 38, 31, 27, 20, 16, 9, 4, 2, 0

Review

Movement Disorders in Musicians

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Abstract: The focus of this article is to review the epidemiology, phenomenology, pathophysiology, genetics, and treatment of movement disorders, particularly task-specific dystonia, in musicians. The goal is to draw attention to this group of neurological disorders among musicians, music teachers, and healthcare professionals and to highlight the importance of early diagnosis, therapeutic options, and preventive measures. To increase professional and public awareness and to facilitate the recognition of music-related neurological problems, we suggest that “medical problems of musicians” be included in curriculum of music schools and medical schools. © 2008 Movement Disorder Society

Keywords: dystonic tremor; Tourette syndrome; musicians, botulinum toxin
Leon Fleisher (1928-2020)

MDS-AOS 4/15/1999
Online Regional Course

MDS-AOS
Online Regional Course
Focal Hand Dystonia, Mirror Dystonia and Motor Overflow

Situburana O, Jankovic J. J Neurol Sci 2008;266:31-3
Dystonic writer’s cramp and alleviating maneuver

Tas et al. Mov Disord 2001;16:1185-9
Using multimodal neuroimaging analyses of resting-state functional connectivity, voxel-based morphometry and tract-based spatial statistics we found that all task-specific focal dystonias were characterized by abnormal recruitment of parietal and premotor cortices. Musician's dystonia was shaped by alterations in primary and secondary sensorimotor cortices together with middle frontal gyrus.
Muscle selection

- History and physical examination
- Muscle afferent block with lidocaine
- Surface and fine wire electromyography (EMG)
- Ultrasound


Botulinum toxin and occupational therapy for writer's cramp.

Park et al. Toxicon 2019;169:12-17

- 12 WC patients were randomized: 6 received only BoNT therapy and 6 received BoNT & occupational therapy.
- BoNT was injected by movement disorders neurologists in the affected muscles under EMG guidance. The primary outcome was the patient-rated subjective scale at 20 weeks. Secondary exploratory outcomes included the writer's cramp rating scale, writer's cramp impairment scale, the writer's cramp disability scale, handgrip strength and kinetic parameters.
- The patient-rated subjective scale scores at 20 weeks were not significantly different between the two groups. There was a significant decrease (28%) in writer's cramp impairment scale, but the primary endpoint, patient-rated subjective scale, was not achieved. There was worsening as indicated by writer's cramp rating scale and no significant differences in other secondary measures.

An isometric splint customized for a patient fabricated to assist the patient to perform finger movements in the direction opposite to the patient's own dystonic movements.
Self-reported benefit and weakness after botulinum toxin in dystonia.

Kassavetis et al. Parkinsonism Relat Disord 2020;80:10-11

- Questionnaires from 42 patients (26 females; mean age 65 years): 10 patients had blepharospasm, 19 CD and 13 FHD. Mean duration of treatment was 9.5 years.
- The mean benefit for the entire population was 66.0% and the mean weakness in 20.5%. No difference between subtypes of dystonia.
- FHD group had longer maximum benefit (mean duration 87.5 days) compared to blepharospasm (mean duration 46.4 days) or CD (mean duration 52.0 days).
Dystonic Tremor

- A spontaneous oscillatory, rhythmic, although often inconstant, patterned movement produced by contractions of dystonic muscles often exacerbated by an attempt to maintain primary (normal) posture. DT may be relieved by allowing the abnormal dystonic posture to fully develop without resistance ("null point").

Albanese et al. Mov Disord 2013;28:863-73

- The prevalence of any type of tremor was 53.3% (of 2362 individuals with isolated dystonia); the prevalence of DT varied from 36.9-48.4%.

Shaikh et al. Neurology 2020 (on line)

- DT tremor is associated with widespread reductions in functional connectivity compared to ET tremor within cortical, basal ganglia, and cerebellar regions.

DeSimone et al. Brain 2019;142:1644-59

- DT (writer's cramp with writing tremor) exhibits increased tremor variability, instability, and intermuscular coherence, and decreased CTC inhibition compared to tremor associated with dystonia (TAWD; cervical dystonia with hand tremor); CTC functional connection is stronger in TAWD similar to ET.

Panyakaew et al. J Neurosci 2020 (on line)

Botulinum Toxin for the Treatment of Hand Tremor.

Niemann N, Jankovic J. Toxins 2018;10(7):299

- A retrospective review of our patients treated for hand tremor with onaBoNT-A between 2010 and 2018 in ≥ 2 visits.

- 91 patients (ET-53, dystonia-31, PD-6, cerebellar-1), mean age 64.8 years.

- Mean of 7.7 treatments (< 31) over a period of 29.6 months (< 8 years).

- Forearm flexors were injected in 89 (97.8%) patients and 15 (16.5%) were injected in at least one other muscle group.

- 80.2% and 85.7% of patients reported moderate or marked improvement (score 3 or 4 on a 0-4 “peak effect” rating scale ).

- Of 1,095 limb injections, 134 (12.2%) were associated with non-disabling and transient (mean 36 days) weakness.

- Conclusion: OnaBoNT-A injections are safe and effective in the treatment of hand tremor. Avoiding injection into the forearm extensors, prevented extensor wrist and finger weakness without compromising tremor reduction.

BoNT has been also found to be effective in head, voice, PD, and other tremors.

Mittal SO, Lenka A, Jankovic J. Parkinsonism Relat Disord 2019;63:31-41
Safety and long-term efficacy of ventro-oral thalamotomy for focal hand dystonia.

- 171 patients with task-specific focal hand dystonia (TSFD), writer's cramps (92), musician's dystonias (58), and other occupational task-related dystonias (21) underwent unilateral ventro-oral thalamotomy.
- The task-specific focal hand dystonia scale scores improved (p<0.001).
- The mean clinical follow-up period was 25.4 ± 32.1 months (range: 3-165).
- Permanent adverse events developed in 6 patients (3.5%); 18 patients developed recurrent dystonic symptoms postoperatively and 7 of 9 who underwent repeat procedure improved.
- DBS is the preferred procedure for bilateral dystonia, however, TSFD mostly develops unilaterally (25/171, 14.6% bilateral symptoms in this study), so most patients do not require bilateral procedures.

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