# Clinical Characteristics of Oromandibular Dystonia: a Multicenter Review of 201 Cases

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## OBJECTIVE

- The overall goal of this study is to better characterize the clinical features of oromandibular dystonia (OMD) in a large multicenter cohort.

## BACKGROUND

- OMD is a rare form of dystonia that affects the masticatory, facial and lingual muscles.
- Diagnosis and treatment are often delayed because OMD is not well recognized, and optimal treatment methods are not known.
- OMD patients tend to be very disabled because of inability to eat and speak.
- The Dystonia Coalition is an NIH-funded multi-center collaboration aimed at advancing clinical research for isolated dystonia syndromes.
- Review of the Dystonia Coalition and Emory data offers a unique opportunity to review a large sample of cases to better describe the clinical characteristics of patients with OMD.

## METHODS

- Study population: We analyzed data collected from 164 OMD patients enrolled across 26 international sites in the Dystonia Coalition and included 37 additional patients evaluated at Emory within the last year for OMD.
- Data Collection: All Dystonia Coalition subjects completed questionnaires for demographics. A movement disorders specialist determined distribution of dystonia, areas affected and severity as determined by the Global Dystonia Rating Scale. A subgroup of the patients also completed the SF 36-Item Health Survey assessing quality of life, Beck Depression II Scale, and Liebowitz social anxiety scale. For Emory patients data on clinical characteristics, treatment type, dosages and response was extracted from chart review.
- Statistical Analysis: Descriptive analysis for sample characteristics was completed. ANOVA for the difference in group mean and etiologic linear regression modeling were performed. All data analysis was using SAS version 9.4.

## RESULTS

**Table 1:** Characteristics of OMD patients

| Characteristics | Dystonia Coalition N=164 | Emory Cohort N=37 |
|---|---|---|
| **Age of Dystonia Onset** | | |
| *Focal* | 54 ± 11 | 60 ± 10 |
| *Segmental* | 49 ± 13 | 50 ± 13 |
| *Generalized* | 19 ± 15 | |
| **Sex** | | |
| *Male* | 34% (n=55) | 32% (n=12) |
| *Female* | 66% (n=109) | 68% (n=25) |
| **Etiology** | | |
| *Idiopathic* | 100% (n=164) | 81% (n=30) |
| *Tardive* | 0 | 19% (n=7) |
| **Distribution** | | |
| *Focal* | 34% (n=56) | 57% (n=21) |
| *Segmental* | 47% (n=77) | 41% (n=15) |
| *Generalized* | 19% (n=31) | 3% (n=1) |
| **Areas Affected** | | |
| *Lower Face* | 45% (n=73) | 40% (n=15) |
| *Jaw* | 80% (n=131) | 89% (n=33) |
| *-opening* | | 51% (n=19) |
| *-closing* | | 16% (n=6) |
| *-deviation* | | 22% (n=8) |
| *Tongue* | 46 (30%) | 41% (n=15) |
| **GDRS Severity** | | |
| *Total* | 17.73 ± 16 | |
| *Lower face* | 2.71 ± 2 | |
| *Jaw and tongue* | 3.56 ± 3 | |
| **Botulinum Toxin** | | |
| *Treated* | | 84% (n=31) |
| *Dose (Onabotulinum toxin A equivalent units)* | | 102 ± 72 |
| *EMG usage* | | 90% (n=28) |
| *Toxin Response* | | |
| *Good (75-100%)* | | 39% (n=12) |
| *Moderate (50-74%)* | | 39% (n=12) |
| *Partial (25-49%)* | | 9.68% (n=3) |
| *Minimal (1-24%)* | | 13% (n=4) |

**Table 2:** Self-reported depression, anxiety and quality of life among patients with OMD in the Dystonia Coalition (N=47)

| Characteristics | Mean | Normal |
|---|---|---|
| **BDI Score*** | 9.25 ± 9 | 0-9 |
| **LSA Score**** | 34.38 ± 27 | 0-30 |
| **SF-36 Quality of Life** | | |
| *Mental Component Score* | 42.4 ± 11 | 50 |
| *Physical Component Score* | 48.4 ± 10 | 50 |

*BDI Score calculated from Beck Depression Inventory II

**LSA Score calculated from Liebowitz Social Anxiety Scale

## RESULTS

**Table 3:** Mean SF-36 sub-scale scores

| Scale | OMD NH Study | Normative data U.S. general population |
|---|---|---|
| Physical function | 74.7 ± 27 | 83.0 ± 24 |
| Role functioning physical | 51.8 ± 44 | 77.9 ± 35 |
| Bodily pain | 67.3 ± 28 | 70.2 ± 23 |
| General Health | 63.6 ± 25 | 70.1 ± 21 |
| Vitality | 56.7 ± 22 | 57.0 ± 21 |
| Social Functioning | 67.3 ± 32 | 83.6 ± 23 |
| Role functioning emotional | 62.7 ± 43 | 83.1 ± 32 |
| Mental health | 59.0 ± 16 | 75.2 ± 18 |

- ANOVA for mean BDI score, LSA score, and SF-36 scores showed no significant difference by distribution of dystonia.
- Linear regression analysis showed that BDI score ($p<0.001$) was a significant predictor of SF-36 mental component score and physical component score controlling for age, sex, total GDRS score and LSA score.
- OMD patients had mean LSA score indicating social phobia, and mean SF-36 scores indicating impaired mental and physical quality of life.
- Treatment with botulinum toxin injections resulted in 50-100% improvement in 78% of OMD patients.

## DISCUSSION

- This cohort of 201 OMD patients is the largest yet described.
- The majority of patients treated with botulinum toxin injection reported improvement in symptoms and 78% reported 50-100% improvement. This contradicts prior reports that botulinum toxin is not an effective treatment for OMD. Prospective studies are needed to confirm this.
- OMD is associated with increased social anxiety and impaired quality of life, with SF36 scores that are worse than those reported in the literature for blepharospasm and CD. We hypothesize that dystonia in the oromandibular region may be a particularly strong predictor of poor quality of life.