Oromandibular Dystonia: Demographics and Clinical Data from 240 Patients

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ABSTRACT

Objective To report demographic data from a large cohort of patients with oromandibular dystonia (OMD).

Methods This is a retrospective review of patients with OMD referred to our institution between 1989 and 2015. Demographic (age of onset, gender, and familial history of dystonia) and clinical (type of OMD, associated dystonia, and etiology of dystonia) data were collected from a cohort of 240 individuals.

Results The mean age of onset of OMD was 51.6 years old, with a female predominance (2:1). A family history of dystonia was found in 6 patients (2.5%). One hundred and forty-nine patients (62.1%) had the jaw-opening type of OMD, 48 patients (20.0%) had the jaw-closing type, and 43 patients (17.9%) had a mixed form of OMD. Lingual dystonia was also present in 64 (26.7%) of these patients. Eighty-two patients (34.2%) had a focal dystonia, 131 patients (54.6%) had a segmental dystonia, and 27 patients (11.3%) had a generalized dystonia. One hundred and seventy-one patients (71.3%) had idiopathic OMD.

Conclusion OMD is a chronic and disabling focal dystonia. Our study found a prevalence of female patients, an onset in middle age and a predominantly idiopathic etiology. Unlike other studies, jaw-opening was found to be the most frequent clinical type of OMD.

Key Words Oromandibular dystonia; focal dystonia; movement disorders.
There was a significant female preponderance—165 of 240 (68.8%) patients were women. A total of 149 out of 240 (62.1%) patients had JOOD, 48 (20.0%) patients had JCOD, and 43 (17.9%) had mixed OMD. There was no significant difference between these groups in gender or age of onset. Lingual dystonia was present in 64 (26.7%) patients and was significantly more associated with JOOD and mixed OMD than with JCOD (32%, 26%, and 10%, respectively).

Eighty-two of 240 (34.2%) patients had a focal dystonia; 131 (54.6%) patients had a segmental dystonia; and 27 (11.3%) patients had a generalized form of OMD. Segmental dystonia categories included blepharospasm as part of Meige syndrome (93/240, 38.8%), cervical dystonia (64/240, 26.7%), spasmodic dysphonia (49/240, 20.4%), and limb dystonia (14/240, 5.8%). There was no correlation between OMD groups and focal versus segmental/generalized presentation.

Dystonic movements impaired speech in 153

### Table 1. Demographics and clinical characteristics

| Characteristics                      | OMD (n = 240) |
|--------------------------------------|---------------|
| Females, n (%)                       | 165 (68.8)    |
| Males, n (%)                         | 75 (31.2)     |
| Mean age of onset, y (range, ± SD)   | 51.6 (3.0–85.5, ± 18.6) |
| OMD type, n (%)                      |               |
| JOOD                                 | 149 (62.1)    |
| JCOD                                 | 48 (20.0)     |
| Mixed OMD                            | 43 (17.9)     |
| Lingual dystonia, n (%)              | 64 (26.7)     |
| Etiology, n (%)                      |               |
| Idiopathic                           | 171 (71.3)    |
| Tardive                              | 31 (12.9)     |
| Neurodegenerative                    | 18 (7.5)      |
| Post-anoxic                          | 16 (6.7)      |
| Post-traumatic                       | 4 (1.6)       |
| Associated movement disorders, n (%) |               |
| Blepharospasm                        | 93 (38.8)     |
| Cervical dystonia                    | 64 (26.7)     |
| Spasmodic dysphonia                  | 49 (20.4)     |
| Limb dystonia                        | 14 (5.8)      |
| Generalized dystonia                 | 27 (11.3)     |
| Symptoms, n (%)                      |               |
| Speech impairment                    | 153 (63.8)    |
| Mastication impairment               | 118 (49.2)    |
| Swallowing impairment                | 65 (27.1)     |
| Pain                                 | 78 (32.5)     |
| Dental impairment                    | 40 (16.7)     |
| Family history of dystonia, n (%)    | 6 (2.5)       |

JOOD: jaw-opening oromandibular dystonia, JCOD: jaw-closing oromandibular dystonia, OMD: oromandibular dystonia.

There was a significant female preponderance—165 of 240 (68.8%) patients were women. A total of 149 out of 240 (62.1%) patients had JOOD, 48 (20.0%) patients had JCOD, and 43 (17.9%) had mixed OMD. There was no significant difference between these groups in gender or age of onset. Lingual dystonia was present in 64 (26.7%) patients and was significantly more associated with JOOD and mixed OMD than with JCOD (32%, 26%, and 10%, respectively).

Eighty-two of 240 (34.2%) patients had a focal dystonia; 131 (54.6%) patients had a segmental dystonia; and 27 (11.3%) patients had a generalized form of OMD. Segmental dystonia categories included blepharospasm as part of Meige syndrome (93/240, 38.8%), cervical dystonia (64/240, 26.7%), spasmodic dysphonia (49/240, 20.4%), and limb dystonia (14/240, 5.8%). There was no correlation between OMD groups and focal versus segmental/generalized presentation.

Dystonic movements impaired speech in 153
(63.8%) patients, mastication in 118 (49.2%) patients, and swallowing in 65 (27.1%) patients. The movements caused pain in 78 (32.5%) patients and dental impairment (wear or early loss of teeth) in 40 (16.7%) patients. Mastication disorders were less present in JCOD than in JOOD and mixed OMD (25%, 56%, and 58%, respectively). Speech impairment was also less frequent in JCOD than in JOOD and mixed OMD (52%, 70%, and 63%, respectively, \( p < 0.05 \)). There was no significant difference in swallowing impairment between these groups. Pain was common in JCOD, less present in mixed OMD and rare in JOOD (71%, 40%, and 18%, respectively, \( p < 0.001 \)). Dental impairment was mostly present in JCOD or mixed OMD and almost absent in JOOD (35%, 37%, and 5%, respectively, \( p < 0.001 \)).

Most patients (171/240, 71.3%) had idiopathic OMD. Thirty-one (12.9%) patients had tardive dystonia, 18 (7.5%) patients suffered from neurodegenerative disease (Parkinson disease \( n = 9 \), pantothenate kinase-associated neurodegeneration \( n = 3 \), GM1 gangliosidosis \( n = 1 \)), 16 (6.7%) patients had a post-anoxic form, and 4 (1.6%) patients presented as post-traumatic OMD. Distribution of dystonia varied depending on etiology (\( p < 0.001 \)). Idiopathic forms were either segmental (63%), focal (29%), or generalized (8%). Tardive dystonia patients were either focal (52%) or segmental (48%) but never generalized. Post-anoxic forms were often generalized (56%). A family history of dystonia was present in 6 patients (2.5%). Five of these patients suffered from an idiopathic focal or segmental dystonia, and one suffered from a generalized dystonia due to a familial mitochondrialopathy.

### DISCUSSION

Two hundred and forty patients diagnosed with OMD were evaluated over a 25-year period at our institution. To our knowledge, this study is the largest reported in the literature. A comparison of our demographic data with other studies is presented in Table 2.

Our results are similar to the data published by others in terms of gender ratio and age of onset.\(^5^6\) The female prevalence has no pathophysiologic explanation.

Focal OMD is rare. OMD is more common as part of a spectrum of cranio-facial segmental or generalized dystonia.\(^5^8\) In our study, 34.2% of patients had focal OMD whereas 54.6% had segmental dystonia and 11.3% presented with generalized dystonia. Sinclair et al.\(^11\) reported similar findings with 12 of 59 (34.3%) patients having a focal OMD and 65.7% of patients presenting with a segmental or generalized form. In our study, the segmental form included blepharospasm (Meige syndrome) in 38.8% of patients, cervical dystonia in 26.7%, spasmodic dysphonia in 20.4%, and limb dystonia in 5.8%. Tan and Jankovic\(^4\) also found blepharospasm (50.0%) and cervical dystonia (57.4%) as more frequently associated movement disorders.

In our study, JOOD was the most frequent form of OMD, 62.1% of patients had JOOD, 20.0% had JCOD, and 17.9% had mixed OMD, as opposed to other studies reported in the literature. Sinclair et al.\(^11\) reported 47.4% of patients with JCOD, 35.6% with JOOD, and 16.9% with isolated lateral JD. Tan and Jankovic\(^4\) reported 52.5% of patients with JCOD, 21.6% with JOOD, 1.9% with isolated lateral JD, and 24.0% with mixed OMD. This difference may

| Table 2. Comparison of demographics with previously published studies |
|-----------------------------|-----------------------------|-----------------------------|
| n                           | Sinclair et al.\(^11\)       | Tan and Jankovic\(^4\)       |
| Female, n (%)               | 59                          | 162                         |
| Age of onset, mean (SD)     | 56.6 (14.0)                 | 57.9 (15.3)                 |
| JOOD, n (%)                 | 21 (35.6)                   | 35 (21.6)                   |
| JCOD, n (%)                 | 28 (47.4)                   | 85 (52.5)                   |
| Mixed OMD, n (%)            | 0                           | 39 (24.0)                   |
| Lingual dystonia, n (%)     | 10 (16.9)                   | -                           |
| Idiopathic OMD, n (%)       | 54 (91.5)                   | 102 (63.0)                  |
| Other movement disorder, n (%) | 23 (65.7)               | -                           |
| Family history, n (%)       | -                           | 10 (6.2)                    |

JOOD: jaw-opening oromandibular dystonia, JCOD: jaw-closing oromandibular dystonia, OMD: oromandibular dystonia, JD: jaw deviation.
Our study shows a prevalence of female patients, an onset of symptoms between 50 and 60 years of age, and an idiopathic etiology in the majority of patients. JOOD was the most frequent type of OMD, as opposed to other reports.

**Conflicts of Interest**

The authors have no financial conflicts of interest.

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