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1 **The "Drama Mask" Phenomenon in Craniofacial Dystonia**

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18 Dystonia involves sustained or intermittent muscle contractions that cause abnormal movements
19 or postures and are often exacerbated by voluntary actions¹. Blepharospasm, a manifestation of
20 dystonia, spreads in about 50% of cases, with oromandibular involvement as the most common
21 area². In craniofacial dystonia, the involvement of facial musculature can have an outsized
22 impact on social and interpersonal interactions.

23

24 A 52-year-old woman presented to our center with a 7-year history of involuntary eyelid
25 closure. Based on her complaint of “heavy” eyelids that would not stay open, she was initially
26 diagnosed with seronegative myasthenia gravis and treated with medications and thymectomy.
27 The lack of improvement led to subspecialty consultation and a diagnosis of blepharospasm.
28 MRI imaging of the brain was unremarkable. Genetic testing was not pursued. She presented to
our center to continue therapy with botulinum toxin injections.

29 At disease onset, she had isolated involuntary eye closure and her symptoms progressed
30 to involve involuntary contractions of other facial muscles, consistent with craniofacial dystonia.
31 When she chewed food, she had stereotyped eyelid clenching, nose elevation, and lip puckering
32 and elevation (Video 1). In social settings, people would consistently inquire about her reason for
33 disgust, when in fact she was enjoying her food. This caused embarrassment to eat in public.
34 Conversely, when angrily scolding her children, she noticed paradoxical “smiling,” or pulling of
35 the nose upward and angles of the mouth backward, diminishing the conviction and emotion she
36 was trying to convey. These “drama masks” resulted in a frustrating mismatch between her
37 emotional state and outward expression.

38 A trial of clonazepam had minimal impact. Other medications, including trihexyphenidyl,
39 were not tried because of the focal nature of her symptoms and concerns for systemic side
40 effects. Botulinum toxin injections were progressively tailored (Figure 1) and ultimately
41 alleviated her craniofacial dystonia and “drama mask” phenomenon (Video 1).

42 In our case, a patient with chronic blepharospasm experienced new debility as the spread
43 of her dystonia contaminated execution of nonverbal communication. This case highlights the
44 distinct social and emotional impact of craniofacial dystonia and the importance of finely
45 tailoring treatment to aspects of impact to the individual. Muscle selection in this individual
46 required watching several home videos and allowing her to chew food and provoke the dystonia
47 in clinic in order to capture the specific actions.

48 We present an example wherein craniofacial dystonia produced emotionally incongruent
49 facial expressions and impacted emotional expression. Consequently, this “drama mask”
50 phenomenon interfered with effective communication and was socially disabling. This
51 symptomatology is not captured on common scales that assess focal dystonia, is not well
52 described in the literature, and may be underappreciated. This is compounded by the known non-
53 motor effects of dystonia. Depression and anxiety are highly co-morbid with blepharospasm and
54 even more so with segmental dystonia, with one recent study citing social anxiety in 67% of
55 patients with the latter³. Recognizing the emotional and social impacts of craniofacial dystonia is
56 critical to effective and comprehensive treatment.

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- 61 1. Research project: A. Conception, B. Organization, C. Execution;
62 2. Statistical Analysis: A. Design, B. Execution, C. Review and Critique;
63 3. Manuscript Preparation: A. Writing of the first draft, B. Review and Critique;

64 MAR: 1B, 1C, 3A, 3B

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66 AEMN: 1C, 3B

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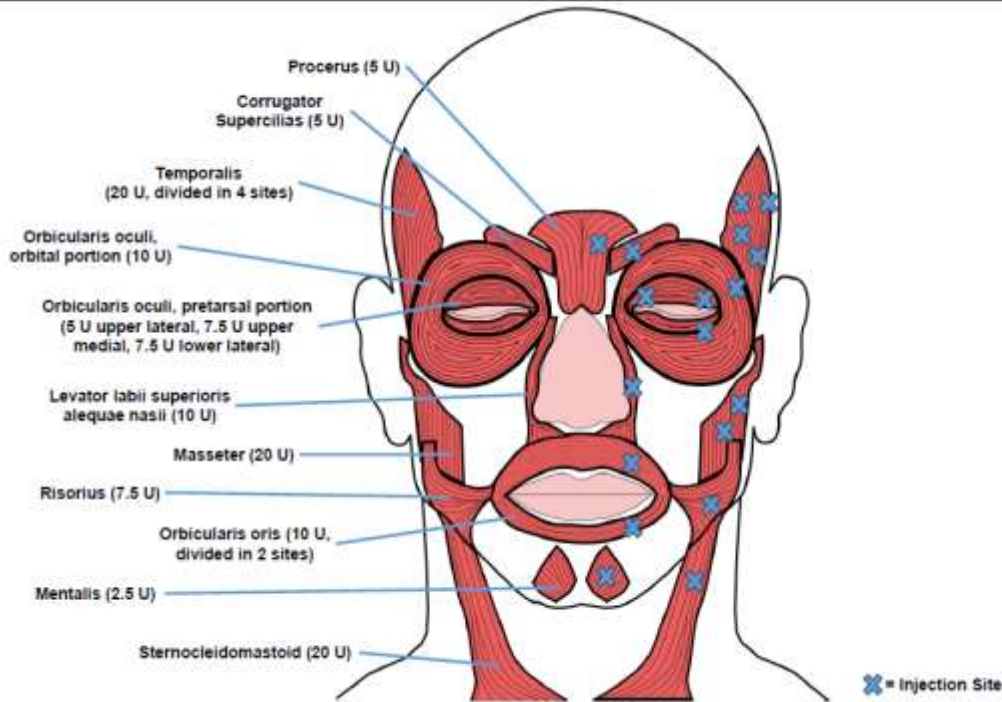
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91 **Figure 1:** Bilateral, symmetric Onabotulinum toxin type A injections into muscles of the face
 92 and neck alleviated the patient’s craniofacial dystonia. Her optimized injection pattern is
 93 displayed on a single side, with each injection site indicated by an X. Units of toxin displayed are
 94 for unilateral injections.

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96 **Legends for Figures and Supplemental Files:**

97 **Video 1:** Assessment before and after botulinum toxin injections. The “drama mask”
 98 phenomenon was induced by eating a cracker and included stereotyped eyelid clenching, nose
 99 elevation, and lip puckering. Peak effect of botulinum injections resulted in a reduction of
 100 dystonia while eating and resolution of the “drama mask” phenomenon, captured by the patient.