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Dystonia of the Oromandibular, Lingual and Laryngeal Areas

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1. Introduction

Head and neck dystonias, like in other types of dystonias, are defined clinically by the presence of involuntary sustained, forceful muscle contractions leading to characteristic rhythmic movements and abnormal postures.\(^1,2\) Cranio-cervical manifestations of dystonia can have a significant effect on a person’s quality of life by impacting the ability for speech, swallowing and social interaction. Oromandibular, lingual, laryngeal and cervical dystonias can occur as part of a generalized neurodegenerative disorder or can be a focal primary dystonia.\(^3, 4\) Focal primary dystonias of the head and neck can be task specific where the dystonic action is triggered by speaking. In some cases, the dystonic action can be overcome with a “geste antagoniste” or sensory trick. Tactile or sensory stimulation in the region of the affected muscle group causes relaxation of the dystonic muscles.\(^5\) Understanding the particular muscle involved in the dystonia and that muscle’s normal function is important to diagnose and treat cranio-cervical dystonias.

The mainstay of treatment for focal dystonia of the head and neck is Botulinum toxin (BoNT) injection.\(^6\) Since its introduction in 1980 to treat childhood strabismus, BoNT has become the treatment of choice in a number of conditions characterized by focal involuntary muscle over activity. Patients suffering from head and neck dystonias require repeated injections to treat symptoms.\(^7,8\) This present chapter focuses on BoNT treatment protocols for oromandibular, lingual and laryngeal dystonia. BoNT for cervical dystonia is discussed in related chapters.

2. Oromandibular dystonia

Oromandibular dystonia (OMD) is a disorder where repetitive or sustained spasms of the masticatory muscles result in involuntary jaw opening, closing or a combination.\(^1,2\) This disorder can produce painful muscle contractions resulting in abnormal positions of the mouth, jaw or tongue affecting speech and swallow function. OMD can be idiopathic, either focal or as part of generalized dystonia, or secondary to medications, trauma, metabolic disorders or other neurologic movement disorders.\(^12,13\) Focal oromandibular dystonia is rare.\(^3\) In the focal idiopathic form, disease onset typically occurs between 30-70 years of age and occurs twice as frequently in women.\(^13,16\) Initially, dystonic episodes may be triggered by specific tasks such as eating, speaking or swallowing. Later less specific motor tasks

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Induce the symptoms and in advanced stages dystonic movements can occur at rest.17,18 The majority of cases are idiopathic though can occur with phenothiazine use or head injury.19 A 12 year follow up study on a patient with OMD, highlighted the influence of hormonal factors on OMD as more severe symptoms occur during stress or menses.12 Although the exact pathogenesis is unknown, neurotransmitter abnormalities which result in disturbed firing pattern of the basal ganglia are likely involved in the abnormal muscle contraction.20

Oromandibular dystonia can be classified as jaw opening or jaw closing dystonia. Knowing the muscles of mastication, their actions and attachments is important to understanding the symptoms and treatment of OMD. The muscles of mastication are innervated by the trigeminal nerve and include the temporalis, masseter and medial and lateral pterygoids. The temporalis muscle originates in the temporalis fossa of the temporal bone and inserts on the coronoid process and anterior surface the mandible ramus and functions to elevate and retract the mandible. The masseter originates at the zygomatic arch and attaches to the angle and ramus of the mandible and functions to elevate the mandible. The medial pterygoid originates from the medial surface of the lateral pterygoid plate and attaches to the mandible and angle of the mandible. It functions to elevate and protract the mandible. The lateral pterygoid muscle has a superior and inferior head. The superior head originates at the greater wing of sphenoid and attaches to the capsule and articular disk of temporomandibular joint. The inferior head originates at the lateral surface of lateral pterygoid plate superior head and attaches to the neck of mandible. The lateral pterygoid elevates and protrudes the mandible.21

Jaw opening dystonia is caused by sustained contraction of the lateral pterygoid muscle resulting in the inability to close the mouth. Prolonged jaw opening results in difficulty with mastication, swallowing and causes drooling. Patients have difficulty articulating and have unintelligible speech.22 The dystonic movements can impact chewing and swallowing so much the significant weight loss can occur.23 Often idiopathic jaw opening dystonia may be misdiagnosed as dental problems, bruxism or temporo-mandibular joint disorders.12,24

Jaw closing dystonias can occur alone or in association with jaw opening dystonias as general OMD dysfunction.14 Involuntary contraction of the masseter muscle results in sustained trismus and jaw clenching. Jaw closing dystonias have been described in musicians who play wind instruments and develop task specific dystonia in response to attempting to play their instrument.25

Assessment of patients with OMD requires exclusion of brain injury or other secondary causes. MRI is performed to evaluate for stroke or mass involving the basal ganglia. Basic laboratory tests are also performed. A blood ceruloplasmin level and slit lamp exam to rule out Wilson’s disease should be performed.26 Also a thorough evaluation of the temporomandibular joint should be performed to evaluate for other disorders. A non-reducing TMJ disk displacement disorder can mimic jaw clenching (closing) dystonia.3

Therapeutic options for OMD include systemic medications, BoNT injections, speech therapy and the use of oral sensory devices. Both jaw opening and jaw closing OMD can be treated with oral anti dystonic therapies such as tetrabenazine, diazepam and carbamazepine. Anticholinergic drugs reduce muscle spasm by centrally inhibiting the parasympathetic system. Benzodiazeppine decreases monosynaptic and polysynaptic reflexes by increasing presynaptic GABA inhibition a similar action to Baclofen. Anticonvulsants
such as carbamazepine reduce severe muscle spasm by decreasing polysynaptic response. Carbidopa/levodopa in low dose may help dopa-responsive dystonia.\textsuperscript{26} Effectiveness of medical therapy for OMD is variable in the literature but only approximately 17\% of patients with OMD responded reported significant benefit from medical therapy.\textsuperscript{14}

For focal dystonias, BoNT has been shown to be superior to medical therapy. For jaw opening dystonias the target muscle is the lateral pterygoid muscle.\textsuperscript{25} The technique is an intraoral injection performed by following the ramus of the mandible to locate the lateral pterygoid and injecting approximately 45 units on each side.\textsuperscript{26} For jaw closing dystonia, BoNT is injected into the masseter muscle. Palpation of the muscle at the angle of the mandible is performed and 20 units of BoNT are injected into each site. The use of BoNT injections have been shown to improve the quality of life and have a significant perceived benefit from patients.\textsuperscript{29} In a study comparing jaw opening and jaw closing dystonias, jaw opening dystonias have been shown to respond better to BoNT therapy.\textsuperscript{14} Jaw deviation dystonia may likewise occur and BoNT injections are done at the temporalis muscle (best approached injecting anterior fibers of muscle) ipsilateral to the jaw deviation and at the lateral pterygoid muscle in the contralateral side.\textsuperscript{27} Side effects of BoNT injection for OMD include jaw weakness, loss of smile, jaw tremor, dysphagia and nasal regurgitation, but side effects are decreased with dose adjustment and accurate injection.\textsuperscript{3,30}

“Geste antagoniste” or oral sensory feedback devices can be used in the treatment of OMD and as an adjunct to BoNT therapy. The use of an oral sensory device has been shown to decrease the frequency and dose of BoNT required to treat OMD.\textsuperscript{24} For jaw opening dystonia, the device is a custom molded retainer that fits the mandibular teeth. Over the molars there is an extra prominence that, when the patient bites down, stimulates the lateral pterygoid muscle to overcome the dystonic action and results in relaxation of the muscle.\textsuperscript{24} For jaw closing dystonia, a prostodontist device that fits over the teeth prevents complete jaw closing. This device helps to inhibit masseter muscle firing to overcome jaw closing dystonia.\textsuperscript{25}

OMD can dramatically affect communication and swallowing function and it is therefore important to address the social, emotional and nutritional impact of the disorder. A multidisciplinary approach with combining medical treatment with speech therapy and a nutritionist are important in address the needs of patients with OMD.

3. Lingual dystonia

Lingual dystonia affects the intrinsic muscles of the tongue resulting in repetitive tongue protrusion or tongue contraction.\textsuperscript{31,32} Dystonic movements of the tongue are a well-recognized feature of tardive dystonia which develops secondary to first generation antipsychotic medication.\textsuperscript{33} Primary or idiopathic lingual dystonia is a very rare disabling cranial dystonia that impacts speaking, chewing and swallowing. The movements vary from repetitive to sustained tongue tip protrusion or contraction which can be action induced with speaking, eating and whistling.\textsuperscript{34-36} In severe cases it is associated with tongue biting and has even caused life threatening airway obstruction.\textsuperscript{37} Speech can be unintelligible and lingual dystonia can impact swallowing significantly resulting weight loss.\textsuperscript{38} While idiopathic lingual dystonia does occur, it is very rare and therefore it is important to evaluate for secondary causes. In addition to medications, lingual dystonia has been reported to occur secondary to head injury, electrical injury, varicella infection or part of a
neurodegenerative disease. Action induced lingual dystonia can be a striking and early finding in chorea-acentohcytosis. It is also a characteristic of pantothenate kinase associated neurodegeneration (PKAN), Lesch-Nyhan syndrome and Wilsons disease. In one of the largest series of lingual dystonia cases, Esper et al reported the 41% of cases were secondary to medications, 18% heredodegenerative and post encephalitic, 12% generalized dystonia and 29% focal primary lingual dystonia.

Evaluation of a patient with lingual dystonia requires a complete history especially a detailed drug history. History of trauma, infections and the association of symptoms with a specific action or the improvement of symptoms with a sensory trick are important to elucidate. A full neurologic evaluation is required, as lingual dystonia associated with neurodegenerative diseases often presents with other neurologic symptoms. Laboratory tests including creatinine kinase and ceruloplasmin level and brain imaging should be performed.

While BoNT injections have been the mainstay of treatment for other primary focal dystonias, in lingual dystonia BoNT has been approached with caution. There is limited experience and have been reports of severe dysphagia and dysarthria after injection. Blitzer et al cautioned against using BoNT injections in patients with focal lingual dystonia as results are often disappointing and half of his patients developed significant dysphagia and aspiration pneumonia. Esper et al recently published one of the largest series of patients with lingual dystonia and the use of BoNT. Their results found it to be a safe and effective treatment with a 55% of the patients sustaining a marked improvement and 97.8% of BoNT sessions without any significant adverse effects. They used a submandibular approach to inject the genioglossus muscles. The patient is positioned supine with the head tilted back. The placement of the needle is approximately two fingerbreadths back from the midline body of the mandible and 1-2cm lateral. The needle is placed with EMG guidance through the digastric muscle into the genioglossus muscle approximately 2cm deep. The position is confirmed with EMG when the patient protrudes the tongue. The BoNT is injected into a single location. They recommend starting with 5 units in each genioglossus muscle with an increased by 2.5 units in each successive treatment until the patients achieves a reasonable response.

Oral medications for the treatment of lingual dystonia are similar to the medical treatment of other focal dystonias. Medical management includes tetrabenazine, anticholinergics, benzodiazepines and levodopa with variable success reported in the literature. Sensory tricks such as chewing bubble gum or sucking on a fruit seed may give patients relief from their symptoms or as an adjuvant to pharmacologic therapy. Deep brain stimulation performed in patients with generalized dystonia, has been shown to improve tongue protrusion dystonia. Therefore deep brain stimulation should be considered in severe or medically refractive cases of lingual dystonia.

4. Laryngeal dystonias

Laryngeal dystonia, also referred to as spasmodic dysphonia is a focal, action-induced dystonia that affects the laryngeal muscles. Involuntary muscle contraction of the vocal folds produces vocal strain, breathiness and phonatory breaks. Laryngeal dystonia can be classified as adductor type, abductor type, mixed type or adductor laryngeal breathing
dystonia. The adductor type accounts for 80% of laryngeal dystonia and results from spasms of vocal fold adductor muscles resulting in inappropriate closing of the vocal folds with speech. With the adductor type extreme effort is exerted to achieve fluent speech and the patient’s voice quality is harsh and strained with voice breaks. The abductor type is rarer, with uncontrolled spasms of the vocal fold abductors resulting in speech with sustained breathiness and breathy voice breaks, sometimes to the point of aphonia. Mixed laryngeal dystonia has characteristics of the adductor and abductor types. Adductor laryngeal breathing dystonia is characterized by persistent inspiratory stridor and usually a normal voice with a paroxysmal cough. In all types of laryngeal dystonia, the dystonic muscles contractions are task specific usually affecting speech while sparing other laryngeal tasks such as breathing, singing, swallowing or coughing. Laryngeal dystonia can also be associated with other dystonias such as blepharospasm, cervical dystonia or writer’s cramp.

Initially thought to be psychogenic in origin, the current thinking is laryngeal dystonia is a central neurologic processing disorder. There have been several studies that demonstrate differences on neuroimaging and postmortem tissues in the basal ganglia and brainstem in patients with laryngeal dystonia compared to controls. However these neuroimaging studies can be difficult to interpret and the knowledge regarding the pathologic mechanism are limited.

Understanding the anatomy of the larynx and the function of intrinsic muscles of the larynx can elucidate the symptoms and treatment for different types of dystonia. There are six intrinsic muscles of the larynx and all are innervated by the recurrent laryngeal nerve, except the cricothyroid muscle which is innervated by the external branch of the superior laryngeal nerve. The vocal fold adductors are the lateral cricoarytenoid, thyroarytenoid, cricothyroid and interarytenoid muscles. The thyroarytenoids are broad, thin muscles that lies parallel with and lateral to the vocal fold. It arises in front from the lower half of the angle of the thyroid cartilage, and from the middle cricothyroid ligament. Its fibers pass backward and laterally, to be inserted into the base and anterior surface of the arytenoid cartilage. In addition to adduction, the thyroarytenoid increases vocal fold tension and is the muscle target for BoNT injection in adductor laryngeal dystonia. The only abductor of the vocal folds is the posterior cricoarytenoid muscles. They are paired muscles that extend from the posterior cricoid cartilage to the arytenoid cartilages in the larynx, and abduct the vocal folds by rotating the arytenoid cartilages laterally. This is the muscle targeted for BoNT injection in abductor laryngeal dystonia.

Diagnosis of laryngeal dystonia requires multidisiplinary evaluation by an otolaryngologist, speech therapist and neurologist. A history of progressive symptoms with onset after a stressful life event, association with other dystonias or family history of dystonia supports the diagnosis of laryngeal dystonia. Similar to other focal dystonias, sensory tricks such as humming are effective in reducing the dystonic movement. As mentioned previously, other functions of the vocal folds such as laughing, coughing, or crying are not affected in laryngeal dystonia and unlike other functional voice disorders laryngeal dystonia does not improve with speech therapy alone. Neurological evaluation includes evaluating for other neurodegenerative diseases such as Wilson’s or Parkinson’s disease which can cause secondary laryngeal dystonia. Flexible laryngoscopy and strobe exam are essential to diagnosis, to evaluate for other causes of voice problems and access the severity of the dystonia. During laryngoscopy, glottic function is observed for disruptions, spasms, breathy...
breaks and tremor while the patient speaks sentence segments. Laryngeal EMG is useful to diagnosis laryngeal dystonia though no specific findings are pathognomonic for this form of dystonia. Adductor laryngeal dystonia may show abnormally high activity in the thyroarytenoid and cricothyroid muscles. Posterior cricoarytenoid and thyroarytenoid will have abnormally high activity in abductor laryngeal dystonia. Large polyphasic motor unit potentials with phonation and irregular tremor can also be seen.

The mainstay of treatment for laryngeal dystonia is BoNT injection. Oral medications, speech therapy, surgery and deep brain stimulation are used as adjuvants or in patients non responsive to BoNT injection. Pharmacotherapies for laryngeal dystonia are similar to other types of dystonias and include anticholinergics, benzodiazepines, or baclofen. Long term relief of symptoms with systemic agents is limited in laryngeal dystonia.

The first BoNT toxin injection for the treatment of laryngeal dystonia was given by Biltzer in 1984 and since then BoNT injection has become the main treatment modality. For adductor laryngeal dystonia, injection of one or both of the thyroarytenoid muscles is performed. Using EMG, a needle is passed through the cricothyroid membrane in the midline and is angled superior lateral in the direction of the thyroarytenoid muscles. Confirmation is made with EMG by asking the patient to phonate. Injection of 0.625-4 units of botox is performed per muscle depending on the individual. For new patients, an initial dose of 1-2.5 units per thyroarytenoid muscle has been proposed as a starting dose. If the patient does not tolerate bilateral injections, the injections can be staggered several weeks apart. Biltzer et al showed BoNT injections for laryngeal dystonia had an average onset of action to be 2.4 days with peak effect of 9 days. Patients received an average of 15 weeks of benefit and achieved a voice quality 92% of normal. For abductor laryngeal dystonia, BoNT is injected into the posterior cricoarytenoid muscles. Since the posterior cricoarytenoid muscle is the only abductor of the vocal folds, over injection of botulism can result in the inability to abduct the vocal folds during inspiration causing stridor and dyspnea. Therefore, unilateral injection of the posterior cricoarytenoid muscle that appears to have the greatest spasm on laryngoscopy is performed. The technique for posterior cricoarytenoid muscle injection involves rotating the patient’s larynx away from side of injection and passing the needle just posterior to the posterior edge of the thyroid cartilage at the level of the cricoid. The needle is advanced to the posterior plate of the cricoid and correct position is confirmed with EMG by asking the patient to sniff. A dose of 3.75 units is initially used. If injection of one side only is not sufficient, a repeat dose of the same side or a conservative contralateral injection can be performed a later time. Concurrent bilateral injections are avoided because of airway compromise. If stridor or significant narrowing of the glottis occurs no further injections are performed. The dose requirements vary between patients and are not specific to the age or gender of the patient.

Complications of BoNT injections include transient breathy hypophonia, hoarseness, swallowing difficulties and pain. More serious complications include more severe dysphagia or airway compromise. While BoNT is effective in treating the symptoms of laryngeal dystonia, it requires multiple physician visits for repeat injections and the decline in voice quality before the next injection has an impact on the patient’s quality of life.

Various surgical procedures have been developed to provide a long term benefit for patients with adductor laryngeal dystonia. Procedures include a recurrent laryngeal nerve section,
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thyroarytenoid myectomy, expansion laryngoplasty and selective laryngeal denervation-reinnervation. First described by Dedo in the 1970s unilateral recurrent laryngeal nerve section was an initial surgical treatment for laryngeal dystonia. However, with high long term failure rates and poorer voice outcomes this technique has fallen out of favor. Berke et al described a selective denervation of the adductor branches of the recurrent laryngeal nerve bilaterally with immediate reinnevervation using the ansa cervicalis but long term data on this procedure is limited. In general, surgical treatment is reserved for patients who do not respond to BoNT treatment.

Speech and physical therapy are important adjuvants to the treatment of laryngeal dystonia. Where speech therapy has its greatest value is in teaching patients who do not go through injection therapy or are in-between injections to manage the symptoms of the disorder with proper compensatory strategies. These include relaxation techniques, use of diaphragmatic breathing and easy onset tone production, reduced number of words per utterance, with the goals of reducing both excessive adduction and increased subglottal pressure. Sometimes, slight pitch elevation for automatic responses and use of vegetative gestures also help as ‘starters’ of clearer vocalizations. Physical therapy includes myofascial release, integrated manual therapies and laryngeal manipulation.

5. Conclusions
Focal dystonia of the oromandibular, lingual and laryngeal areas are particularly debilitating because they affect the basic human functions of breathing, eating and communicating. While BoNT is effective for the treatment of these forms of dystonia, it is only a temporary treatment and has side effects. Surgery and deep brain stimulation offer the potential for longer lasting symptom relief but there is still much work to be done to perfect techniques. Research is focused on better understanding of the underlying brain pathophysiology causing the focal dystonia and to develop targeted treatments.

6. References
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Dystonia has many facets, and among those, this book commences with the increasingly associated genes identified, including a construct on how biology interacts with the dystonia genesis. The clinical phenomenology of dystonia as approached in the book is interesting because, not only were the cervical, oromandibular/lingual/laryngeal, task-specific and secondary dystonias dealt with individually, but that the associated features such as parkinsonism, tremors and spasticity were also separately presented. Advances in dystonia management followed, and they ranged from dopaminergic therapy, chemodenervation, surgical approaches and rehabilitation, effectively complementing the approach in dystonia at the clinics. A timely critical pathophysiologic review, including the muscle spindle involvement in dystonia, is highlighted at the book’s end.

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