Meige's Syndrome

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"Meige's Syndrome" is an historical term. In 1910, Dr. Henri Meige described a series of patients with involuntary movements of face and neck, the following year Dr. Oppenheim described patients with involuntary twisting movements of children that involved the entire body, and coined the term "dystonia muscularum deformans," Since that time we have realized that dystonia is not really a muscle disease, and this term has been shortened to "dystonia." Meige's syndrome is a focal dystonia, which manifests itself in involuntary facial movements.

Currently, the dystonic disorders may be classified in many ways. In the context of the Benign Essential Blepharospasm Research Foundation, benign essential blepharospasm (cranial dystonia), and Meige's syndrome (cranial/oromandibular dystonia) are the focus. Patients with oromandibular dystonia have difficulty chewing, eating, and speaking because of an inability to open or close their jaw. In addition, a tongue dystonia may be activated by eating, which causes the tongue to push the food out of the mouth. Because of the eating difficulties, some patients lose weight. Besides speech, swallowing, and respiratory difficulties, patients may also make involuntary vocalizations that include humming, grunting, belching, and gasping. In addition, involuntary jaw closure, jaw opening, or jaw deviation dystonia may occur and may interfere with speaking and chewing.

Benign essential blepharospasm (BEB) is defined as a focal dystonia exhibiting sustained or repetitive involuntary spasm of the muscles of the upper face (corrugator, frontalis, and obicularis oculi muscles). Primary, Essential, or Idiopathic blepharospasm is not associated with any known etiology, while secondary blepharospasm is due to documented pathologic lesion. BEB associated with a strong family history or a gene locus is classified as a primary disorder, and the BEBRF has recently funded a study to hasten the search for a possible genetic predisposition to BEB and other cranial dystonias.

Clinical features of blepharospasm include involuntary eye closing aggravated by bright lights, wind, pollution, smoke, emotional stress, and fatigue. This eye closing may interfere with reading, driving, watching television, and other visual activities, and is rarely associated with retro-orbital pain. Blepharospasm is often associated with other dystonias, such as oromandibular or cervical dystonia, and usually occurs in the 5th to 7th decades of life. There is no clinical difference between the eye findings of primary and secondary blepharospasm with the possible exception that primary blepharospasm is sometimes relieved with a sensory trick (touching face, humming, singing, talking, pinching skin), and this has not been documented in secondary cases.

Cranial Dystonia is usually considered a "Primary or Idiopathic Dystonia," and may represent a genetic or inherited disorder. A recent survey of primary dystonia in Europe found that blepharospasm was present in 28.9% of 957 dystonia patients, second only to cervical (neck) dystonia. Symptoms usually occur between 40-60 years of age, and more than 50% reported spread to include another area of the body. Risk factors for blepharospasm included a history of head or facial trauma or family history of dystonia or tremor disorder.

Defazio and other European researchers have compared patients with primary blepharospasm only to patients with primary blepharospasm and another dystonia location, and found that spread is more likely seen in women, and is associated with head or facial trauma or tooth loss. Patients not exhibiting spread to another location were more likely to report a history of ocular disease or family history of dystonia or tremor (Defazio et al. JNNP 1999; 67:613-9).

Table 1†. Clinical and Molecular Information on the Primary Dystonias

Disease Name/ Gene Symbol	Chromosomal Location	Mode of Inheritance	Phenotype
DYT1	9q34	Autosomal dominant	Childhood and adolescent; limb onset
DYT2	Unknown	Autosomal recessive	In Spanish Gypsies; not confirmed
DYT3	Xq13	X-linked recessive	Parkinsonism-dystonia (Lubag, Philippines)
DYT4	Unknown	Autosomal dominant	Whispering dysphonia in Australian family
DYT5/ Dopa-responsive dystonia	14q22	Autosomal dominant	Dopa-responsive dystonia
DYT6	8p21-p22	Autosomal dominant	Mennonite/Amish dystonia with mixed face/eyes/neck or limb onset; childhood or adult onset
DYT7	18p	Autosomal dominant	German families; adult neck, face or limb onset
DYT8	2q33-q35	Autosomal dominant	Paroxysmal dystonia; paroxysmal dystonic choreoathetosis; may be the same as DYT10
DYT9	1p	Autosomal dominant	Paroxysmal choreoathetosis with episodic ataxia and spasticity
DYT10	Unknown		Paroxysmal kinesigenic choreoathetosis; may be same as DYT8
DYT11	Unknown	Autosomal dominant	Myoclonic dystonia; hereditary alcohol-responsive myoclonus
DYT12	19q13	Autosomal dominant	Early-onset Parkinsonism
LDYT	Mitochondrial DNA		Leber's hereditary optic neuropathy

†Modified from de Leon and Bressman[33]