

Rome, May 13th 2017, Blepharospasm

Anna Rita Bentivoglio, MD, PhD

Movement Disorders Unit

Fondazione Policlinico Universitario Agostino Gemelli

Institute of Neurology

Università Cattolica del Sacro Cuore

Rome, Italy

Tel: 00390630155633

email: annarita.bentivoglio@policlinicogemelli.it

Blepharospasm (BSP) is a form of focal dystonia that manifests with spasms of the eyelids, involuntary closure of the eye, and enhanced spontaneous blinking, in any combination. We have advanced in our knowledge of the disorders since the first descriptions of BSP as a form of dystonia. Although the dystonia is focalized to a very small body district, BSP can be very distressing: 1. Interferes with our facial expression and is perceived as disfiguring from many patients, 2. BPS can be so severe that patients may be functionally blind and unable to live a normal social life.

Etiology: Blepharospasm is one of the most frequent focal dystonias (20-133 cases/1.000.000), is more common among women, age at onset range 50-70 years (peak 60), tends to spread to contiguous body parts (generally within 5 years from onset) and often involves lower facial muscles (Meige's syndrome). Head and limb tremor are also frequently concomitant. **The clinical presentation** of BPS consist in stereotyped, bilateral, and synchronous spasms of periocular muscles (orbicular oculi, procerus, frontalis, sometimes also others) causing involuntary eyes closure (intermittent or sustained), or eyelids apraxia (difficulties in opening the eyes) and or enhanced spontaneous eyes blinking. The severity is variable (from excessive blinking to functional blindness) and so the consequences on life quality. Peculiar characteristics of BPS are of the orbicularis oculi muscles. Are sensory tricks and gestes antagonistiques, increased spontaneous blink rate (> 27 b/min) and blink rate at rest > during speaking (in healthy patients the opposite). In the last years it become evident that non motor symptoms are very common and may be really invalidating. Among non motor symptoms must be outlined sensory symptoms (dry and burning eyes, photophobia), psychiatric (anxiety, depression, sleep disorders), mild and selective neuropsychologic impairment. Primary BPS far more common than secondary, it sporadic in most cases but familial BPS: in 25% of BPS patients, at least one family member with dystonia, however none of the genes linked to focal adult onset dystonia correlated to isolated BPS. Blepharospasm can be secondary to focal brain damage (basal ganglia, cortex), Parkinson's disease and other neurodegenerative conditions, drug-induced dyskinesias, neuromuscular disorders (Myasthenia Gravis, etc.)