

# The relationship between eye-winking tics, frequent eye-blinking and blepharospasm

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**SUMMARY** A family is reported in which three generations were affected with eye-winking tics and/or blepharospasm. The proband developed eye-winking tics in childhood and then developed excessive blinking progressing to blepharospasm by the age of 21 years. His mother presented with Meige's syndrome and spasmodic torticollis at the age of 59 years; his uncle had blinked excessively from his early forties. His eldest son developed an eye-winking tic with facial grimacing at the age of 8 years, and in another son, a self-limiting period of eye-blinking occurred at the age of 4 years. The recovery cycle of the blink reflex was abnormal in all three generations. Three other children with eye-winking tics have a parent or close relative with frequent eye-blinking or blepharospasm. Five patients with adult-onset blepharospasm or Meige's syndrome are also described who had excessive eye-blinking dating back to childhood. It is suggested that eye-winking tics, frequent blinking and blepharospasm may share common pathophysiological mechanisms; the clinical expression may be age-related.

It is usually possible to distinguish between eye-winking tics, frequent eye-blinking and blepharospasm. Eye-winking tics most commonly present in childhood as a rapid, exaggerated, co-ordinated contraction of the orbicularis oculi. They can be voluntarily controlled to some degree and increase in frequency when the child is bored, tired or anxious. There is little functional visual impairment and they usually disappear spontaneously after months or years. Boys are three times more frequently affected than girls and the tic is usually unilateral.<sup>1</sup> Frequent or excessive eye-blinking is usually secondary to disorders of the ocular surface, tear film, or eyelids; however, it may also occur in otherwise healthy adults as the prodromal sign of essential blepharospasm. It is then usually bilateral, variable in intensity and may be associated with ocular irritation and photophobia.

Blepharospasm consists of repeated, forceful, involuntary sustained contractions of the orbicularis oculi. It most commonly presents in adults and women are more frequently affected than men. Triggers include bright light, reading or watching television and eye movements. It is often associated with oro-mandibular dystonia and is normally intractable, produc-

ing functional blindness. There may be a family history of blepharospasm or other form of focal dystonia.<sup>2</sup> Eye-winking tics, excessive blinking and blepharospasm are regarded as distinct pathophysiological entities.

The blink reflexes are abnormal in blepharospasm; the amplitude of the R1 and R2 components is increased, the duration of R2 is prolonged with an abnormal recovery cycle, suggesting hyper-excitability of brain stem neurons.<sup>3</sup> Blink reflex studies in Gilles de la Tourette's syndrome are also sometimes abnormal suggesting a similar brain stem interneuron hyper-excitability. In this disorder, however, it is possible to correct this by instructing the patient to suppress voluntarily any eye-winking tic before the stimulation takes place<sup>4</sup> (Smith and Lees, unpublished observation). In both blepharospasm and eye-winking tics, evidence suggests that local eye irritation may be the trigger for the development of abnormal movement.

We report here some evidence to suggest that eye-winking tics, excessive blinking and blepharospasm may be more closely related than current opinion would suggest.

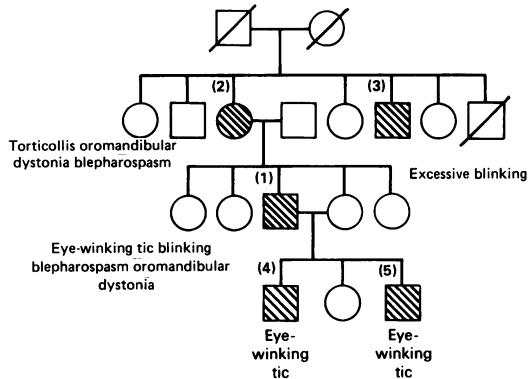
## **Patients**

*Family Cases 1-5: A family with eye-winking tics, excessive blinking and adult-onset focal dystonia (fig)*

All affected members of this family were examined by the authors and video films were taken. The proband, a 32 year

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Pedigree of family with focal dystonia and tics.

old man (Case 1), developed an isolated eye-winking tic at the age of 7, which resolved spontaneously by the age of 11. He was well until the age of 20, when he began to blink excessively and become photophobic. At this stage there were no spasms of eye closure and he was able to continue his work as a London Transport driver. The symptoms recovered spontaneously by the age of 23, but returned again at the age of 28, rapidly worsening to severe blepharospasm. By 29 years of age he was unable to work at all and had become depressed; a number of drug treatments including anticholinergics and benzodiazepines were unhelpful. He has responded well, however, to botulinum toxin A treatment and has been back at work for the last 18 months.

His mother (Case 2) presented at the age of 59 with torsion of the neck, the chin deviating to the left, with no response to anticholinergic drugs. By the age of 61 she had developed frequent eye-blinking, intermittent blepharospasm and oromandibular dystonia. When she attempted to lie flat the cranial dystonia markedly worsened with severe anterocollis. She reported that her younger brother (Case 3) had suffered from excessive eye-blinking in his early forties.

The proband's eldest son (Case 4) developed a winking tic with left-sided facial grimacing at the age of 7 and the younger son (Case 5) has also now developed eye-blinking. The duration and recovery of the R2 components of the blink reflex were abnormal in four family members (table 1). All other investigations were normal including the CT brain scans and wet blood films for acanthocytes.

Table 1 Blink reflex studies in Family 1

Case	Age and sex of patient	Type of movement disorder	Latencies (m/s)								% Recovery Cycle†	
			R1		Ipsilateral R2		Contralateral R2		500 m/s	1 s		
			Rt	Lt	Rt	Lt	Rt	Lt				
4	7M	tic	10	11	36	35	38	37	33		42	
1	32M*	blepharospasm	absent	absent	31	32	32	32	59		51	
2	61F	Meige's syndrome, torticollis	10	10	38	35	41	35	76		65	

\*Botulinum toxin A injection 8 weeks before study.

†% refer to amplitude of R2 component elicited by test stimulus as a percentage of that elicited by conditioning stimulus.

Table 2 Details of families 2, 3 and 4

Family	Case	Age (yr) and sex of onset in proband	Condition	Family history	Case
2	6	4, F	eye tic	Father: (27) excessive blinking	7
3	8	5, F	eye tic	Maternal aunt: (47) blepharospasm (tic as child)	9
4	10	15, M	excessive blinking	Father: (39) blepharospasm oromandibular dystonia	11

Family Cases 6–11: Children with eye-winking tics or excessive blinking and relatives with excessive blinking or blepharospasm (table 2)

A child (Case 6) with mild generalised developmental delay presented with a predominantly right-sided eye-winking tic at the age of 4 years. Her father (Case 7) is 27 and has been aware of excessive blinking for 5 years; on examination he was photophobic with bouts of excessive blinking elicited by fast eye movements.

An otherwise healthy child (Case 8) developed a left eye-winking tic at the age of 5 that resolved spontaneously 2 years later. During this time he was also noticed to widen his palpebral apertures by pulling faces. His maternal aunt (Case 9) developed blepharospasm at 47 years after having a transient eye-winking tic in childhood.

A 15 year old girl (Case 10) developed persistent ocular discomfort and bilateral excessive blinking following the removal of an eyelash from the upper lacrimal punctum of the left eye. Her father (Case 11) developed excessive blinking spontaneously at the age of 39; a year later mild oromandibular dystonia developed.

Cases 12–16: Patients with adult-onset blepharospasm and a previous eye-winking tic or excessive blinking (table 3)

Five patients were seen in this category, three women and two men. The blepharospasm was quite typical in Cases 13–16 and had been preceded by the development of excessive blinking occurring usually around the age of 10 years. This had persisted for between 33 and 57 years (average 43) before it developed into blepharospasm. In one patient there was a family history of oro-mandibular dystonia, in another excessive blinking in a sibling. Case 12 was unusual in that she

Table 3 Details of cases 12–16

Case	Preceding disorder	Age (yr) and sex	Blepharospasm age	Family history
12	excessive blinking	9, F	49	Sister blinks excessively
13	excessive blinking	11, F	63	—
14	excessive blinking	10, M	43	Grandmother has oromandibular dystonia
15	excessive blinking	10, M	67	—
16	tic	7, M	14	Daughter has eye-winking tic

developed a winking tic when aged 7 which has persisted apart from a year of freedom from age 13 to 14. The condition has fluctuated, however, so that in her late 'teens she blinked excessively whereas by her mid-twenties she was housebound by blepharospasm. She is currently only inconvenienced by a largely controllable winking tic. Her eldest daughter developed an eye-winking tic at the age of 5 years.

## Discussion

The fundamental purpose of blinking is the maintenance of the anatomical and optical integrity of the corneal epithelium by the pre-corneal tear film, but normal adults' blink rate (14–17 per minute) is greater than that required for visual purposes, although constant in an individual under fixed conditions. Normal infants and children up to 18 months of age have a low periodic blink rate (2–5 per minute). It is characteristically reduced in patients with Parkinson's disease<sup>5,6</sup> and progressive supranuclear palsy<sup>7</sup> and may be increased in schizophrenia,<sup>8</sup> Gilles de la Tourette's syndrome<sup>9</sup> and to a lesser extent depression.<sup>10</sup> These findings, together with the occurrence of excessive blinking and blepharospasm in disorders which damage the basal ganglia such as Wilson's and Huntington's diseases<sup>11</sup> suggest that normal periodic blinking is a function of the basal ganglia.

The patients described provide clinical evidence that eye-winking tics, frequent blinking and blepharospasm might be more closely related than has previously been considered. Tolosa and colleagues<sup>4</sup> drew attention to the similarities between tics and blepharospasm, claiming an increased incidence of tics in the families of patients with Meige's syndrome. Recently a similar occurrence of childhood tic and adult-onset cranial dystonia occurring within the same kindred has been reported.<sup>12</sup> All the adults (five cases) in the present series showed at different ages more than one of these three abnormalities. In the two cases (1 & 12) who suffered from all three, the tic occurred in childhood, excessive blinking as a teenager and blepharospasm as an adult; the other four developed excessive blinking around the age of ten, and blepharospasm as adults. In seven out of nine cases

another family member had either the same condition (five cases) or a different abnormality of blinking. The age of the affected relative determined the disorder; thus in Case 1, the proband's sons have tics, his mother blepharospasm, the younger uncle excessive blinking or blepharospasm, and Case 7's grandmother had focal dystonia; Case 9's daughter had a tic.

The blink reflex studies carried out in all three affected generations of Family 1 showed an abnormal recovery cycle and one of us (AJL) has found an increased excitability threshold which can be corrected by voluntary suppression in a proportion of patients with Gilles de la Tourette's syndrome. It seems probable that the underlying basal ganglia dysfunction in Case 1 is genetically determined and there is evidence that both tics and blepharospasm may be familial.<sup>12,13</sup> The clinical expression of the underlying movement control varies and may reflect age-related changes in the maturational development of control of blinking. Thus winking tics develop at an age when the normal adult rate of blinking is being established. Excessive blinking seems to develop around the age of ten, which may be another blink control maturational landmark, whereas blepharospasm develops in adults. The precipitant at each age is unclear, but recent evidence points to the possibility that local trauma may be important. Ocular surfaces, tear film or eyelid abnormalities, even when minor, normally increase blinking; in susceptible individuals this may unbalance the control mechanism and produce various degrees of involuntary perpetuation. Many children with eye-winking tics complain of irritation of the affected eye and since minor corneal and lid trauma (such as seen in Case 10) is common, this may be a precipitant in these cases also.<sup>14,15</sup> Although it is impossible to exclude a chance association between two common movement disorders we would like to suggest tentatively that eye-winking tics, excessive blinking and blepharospasm may be age-related manifestations of the same fundamental abnormality.

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