

A new variant of blepharospasm

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Abstract

Ten patients who were unable to initiate or sustain eye opening in the absence of overt spasm of the orbicularis oculi, were investigated. In five, the problem was isolated. Three had Parkinson's disease and two progressive supra-nuclear palsy for between one to six years before the eye opening difficulty developed. The clinical features and electrophysiological investigation suggested that the disorder is a variant of blepharospasm due to abnormal contraction in the pre-tarsal orbicularis oculi.

Idiopathic blepharospasm consists of repeated involuntary forceful bilateral eye closure in the absence of ocular pathology.¹ The spasms are associated with lowering of the brows below the superior orbital rim (Charcot's sign) indicating contraction of the orbital orbicularis oculi. The cause is unknown, but there is evidence suggesting it is an organic neurological disorder.^{2,3} Blepharospasm may be isolated or associated with muscle spasms in the face, jaw and neck (Meige syndrome); it may occur in generalised dystonia, secondary dystonia and Parkinson's disease, especially the post-encephalitic variety.^{4,5}

Patients with Parkinson's disease may also experience an inability or difficulty in opening the eyes spontaneously or to command. In this condition, which can also occur in progressive supra-nuclear palsy, there is no overt spasm of the orbicularis oculi and the eyes appear to be passively shut with frontalis muscle overaction causing elevation of the brows, in contrast to the lowered brows of typical blepharospasm. In some patients the eyes can be opened by stroking or massaging the upper lids. Periodic blinking when the eyes are open is reduced (in contrast to the increase in blepharospasm) and there is no photophobia. The condition has been described as akinesia or apraxia of eye opening,⁶ levator inhibition or pre-tarsal blepharospasm.⁷ Rarely, it may occur as an isolated abnormality without evidence of a generalised extra-pyramidal disorder. It may also be secondary to right hemisphere infarction. This study was undertaken to provide more information on the clinical features, pathophysiology and treatment of the condition.

Patients and methods

Ten patients were studied. They were unable to open their eyes voluntarily but did not have

obvious blepharospasm. Five had an extra-pyramidal disorder (three Parkinson's disease, two progressive supra-nuclear palsy) and in five the abnormality was isolated. Conventional drug treatment of those with Parkinson's disease or progressive supra-nuclear palsy had not improved eye opening: in three of the remaining five patients benzhexol was ineffective and one tried diazepam without response. Eight of the ten had received botulinum toxin injections; in two this was of benefit when combined with ptosis props, but there was no improvement in the remainder. The eyes themselves were healthy and not uncomfortable and the position of the upper lid skin crease (the site of attachment of the levator palpebrae superioris) and the range of levator function was normal. Diurnal fluctuations, worsening in bright light or whilst walking or travelling and improvement whilst talking (all features of idiopathic blepharospasm) were enquired for and if found, documented as variability (table). On attempted eye opening (which was associated with brow elevation) the orbicularis oculi was closely inspected and auscultated with the bell stethoscope for clinical evidence of contraction.

Electrophysiological investigation

A monopolar electrode was inserted through the upper lid into the levator. This muscle was differentiated from the superior rectus by the antagonistic activity of these muscles during forcible voluntary eye closure, when the superior rectus contracts (Bell's phenomenon) but the levator does not. A concentric EMG needle was also inserted into the pre-tarsal portion of the orbicularis oculi muscle. This study was carried out in seven of the ten patients, five of whom had received botulinum toxin injections to the orbicularis oculi between two to six months previously but an EMG signal was recordable in all. The two EMG signals were displayed simultaneously on a Medelec MS6 with a low filter set at 500 Hz to minimise movement artefact. EMG activity was recorded on electrically stimulated blinks and attempted eye opening.

Blink reflexes were designated abnormal if R1 was bilateral or any of the following abnormalities of R2 were observed: a reduced recovery cycle indicated by lack of reduction in the amplitude when the interval between conditioning and test stimulus was one second: an amplitude of greater than 100 μ V (ipsilateral) or 80 μ V (contralateral): a duration of greater than sixty milliseconds (ipsilateral or contralateral).

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Table Atypical blepharospasm: clinical and electrophysiological characteristics

Case	Age	Sex	Duration Yrs	Associations	Variability	Orbicularis oculi contraction		EMG	
						Visible	Audible	Blink reflex	Eye opening
1	50	M	5	torticollis	+	+	+	Abnormal	CC
2	58	M	3	—	+	—	—	Abnormal	CC
3	75	M	14	eye winking tic	+	+	+	N	LNS
4	68	F	4	—	—	+	+	N	LNS
5	56	F	6	depression	—	—	+	—	—
6	64	M	4	PSP (10 years)	—	—	+	Abnormal	CC
7	72	M	5	PD (5 years)	+	+	+	N	LNS
8	77	F	5	PD (9 years)	—	—	+	—	—
9	58	M	1	PD (7 years)	—	—	—	—	—
10	57	M	4	PSP (4 years)	—	—	—	N	CC LNS

PSP = Progressive supra-nuclear palsy

PD = Parkinson's disease

CC = Co-contraction of levator and orbicularis

LNS = Levator contraction not sustained

On attempted eye opening, evidence of an abnormality of the normal reciprocal relationship between the levator and orbicularis oculi (fig 1) was sought, with abnormalities being defined as co-contraction of these muscles or failure to initiate or sustain levator activity.

Results

The results are given in the table and illustrated in figs 2 and 3. In summary, the typical variability of idiopathic blepharospasm was documented in four patients and clinical evidence of abnormal contraction of the orbicularis oculi in seven. Electrophysiological abnormalities were evident in all seven patients tested.

Discussion

Clinically, the patients presented are a recognizable sub-group of those who complain of

visual difficulties due to inability to open the eyes. None of the patients showed any clinical features such as inconsistency of physical signs or response to suggestion or placebo treatment to suggest hysteria as the cause of the problem. Apart from one patient who became depressed there were no psychiatric abnormalities. Of those with isolated eye opening difficulties, one had spasmodic torticollis whilst another had an eye winking tic in childhood. They both showed variability in their disorder and clinical evidence of abnormal orbicularis oculi contraction. These are features consistent with idiopathic blepharospasm⁹ and suggest that these two patients suffer from a variant of blepharospasm. Either variability or abnormal orbicularis oculi contraction was present in the remaining three, five with isolated eye opening difficulties, again suggesting blepharospasm as the cause. Variability or abnormal orbicularis oculi contraction were documented in three of the five with progressive supranuclear palsy or

RECIPROCAL ACTIVITY

Levator



Orbicularis

↑
Close

↑
Open

0.5mv
500 msec

Figure 1 Normal subject; reciprocal activity of the levator and orbicularis oculi muscles demonstrated on closing and opening the eyes.

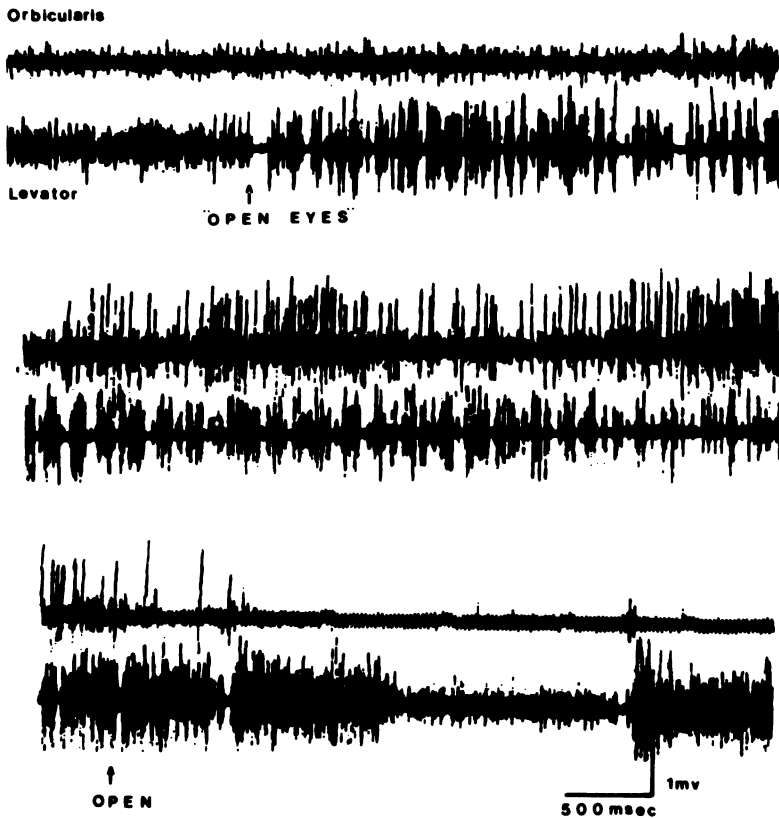


Figure 2 Co-contraction: Following the command "open the eyes" levator activity increases but the orbicularis remains active for over 10 seconds before a further increase in levator activity is seen, orbicularis activity stops and the eyes open (case 1).

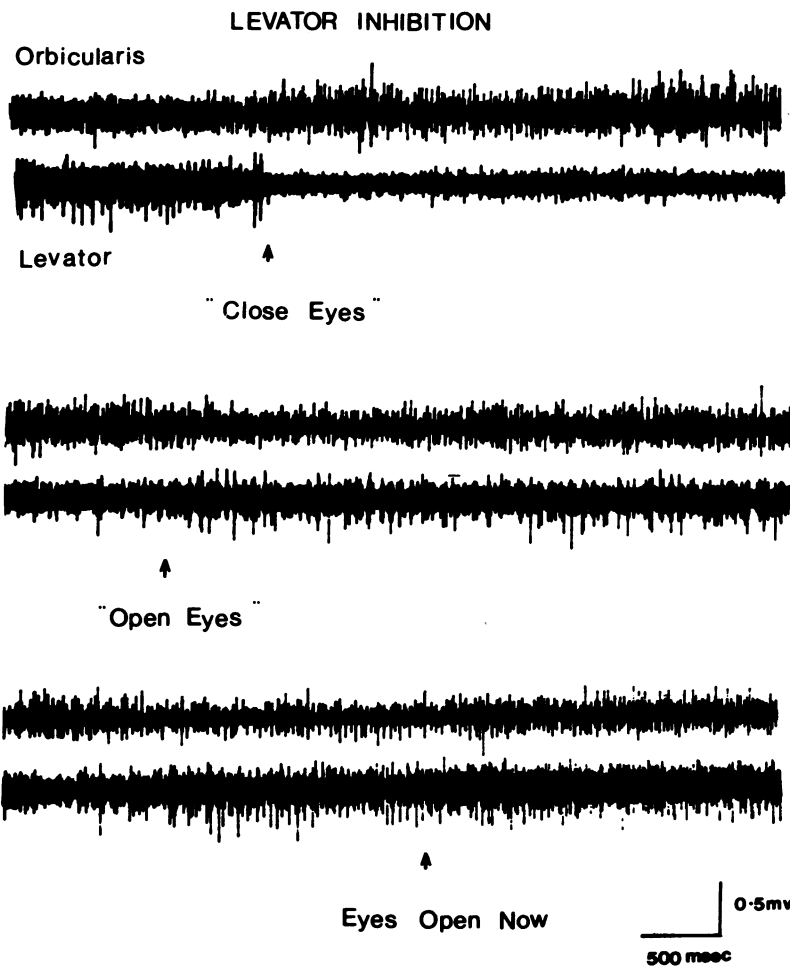


Figure 3 There is co-contraction in the levator and orbicularis oculi at the start of the trace: the levator activity ceases on the order to "close eyes" but on the order "open eyes" relative levator inhibition and co-contraction occur and there is a substantial delay until the eyes are open (case 10).

Parkinson's disease, and since the clinical features as regards eye opening were exactly the same in both groups, it is reasonable to suggest that all the patients have a variant of blepharospasm.

The electrophysiological studies reinforce this view. Although only three had the blink reflex abnormalities regarded as typical of blepharospasm² co-contraction of the levator and orbicularis oculi and/or a delayed or ill sustained levator response was seen in all seven patients tested. All ten patients therefore had either clinical or electrophysiological abnormalities (or both) suggestive of blepharospasm.

It is evident that the term "apraxia of eye lid opening" is inappropriate for these patients since the levator can be voluntarily activated. Likewise the abnormality is not due to an isolated "levator inhibition". Abnormal orbicularis oculi contraction indicated clinically or electrophysiologically is the feature common to all these cases and pre-tarsal blepharospasm is the best description.

If these patients suffer from a variant of blepharospasm botulinum toxin injections would be expected to be beneficial. However, in typical blepharospasm the orbital and pre-septal orbicularis oculi is weakened whereas here the pre-tarsal portion must be treated. The technical difficulty of achieving this may be responsible for the poor results. The combination of global orbicularis oculi weakening and ptosis props appears to be the only useful treatment for these patients but only two out of ten benefited.

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