

# Blepharospasm

## The facts...

Blepharospasm is a type of dystonia resulting in involuntary eye closure that is caused by spasm of the muscles surrounding the eyes. A 'tic', twitching or frequent blinking is typical, sometimes with complete closure of the lids. The severity of blepharospasm varies. It is usually better in the morning, becoming worse later in the day. Bright light, walking, driving, worry or stress can aggravate the condition.

Answering some of the pressing questions posed by our readers are: **John Elston** (JE), consultant ophthalmic surgeon and sister **Alexina Fantato** (AF), from the Oxford Eye Hospital at John Radcliffe Hospital, together with **Dr. Marie-Helene Marion** (MH-M), consultant neurologist and honorary senior lecturer at the Atkinson Morley Neuroscience Centre, St George's Hospital, London. All are leading medical practitioners in the treatment of blepharospasm.



● Alexina Fantato & John Elston



● Marie-Helene Marion

### Q. Can someone function normally after having botulinum toxin treatment?

**JE:** A substantial proportion of patients can function normally or nearly normally after their botulinum toxin (BTX) injections. We can't promise complete normality although they should get an improvement in function, many being able to work and travel.

### Q. Why isn't it satisfactory for everyone?

**MH-M:** Both botulinum toxins (types A or B) are effective for 80% of patients and they are able to lead a normal life, with BTX injections at regular intervals (between 3 to 4 months). There are various reasons why 20% of patients with blepharospasm are not good responders:

- the BTX dose is not optimal (too low or too high)
- the injection sites are not appropriate to the type of blepharospasm
- the blepharospasm is associated with more widespread dystonia around the mouth and tongue (*Meige syndrome*, where the patient may grimace at the same time as his or her eyes are closing).

- the blepharospasm is very severe and associated with mini-spasms of the eyeballs, going up each time that the eyelids attempt to close (*Bell's phenomenon*).
- true biological resistance to BTX is actually extremely rare in cases of blepharospasm.

**JE:** BTX is not a direct treatment for this condition – it treats the symptoms. In blepharospasm, the problem lies with the nerve signal transmission. Basically, what the BTX injections aim to do is to weaken these signals, hence the spasms. For some it is helpful but not necessarily for everyone.

**AF:** It is the single most effective treatment for this condition. Everyone who has blepharospasm should at least try it. Some people come to us having tried oral medication which has not been very successful and these drugs tend to have many side-effects.

### Q. What can be done if the injections aren't satisfactory?

**MH-M:** To optimise the BTX injections, it is best to observe the patient when they are experiencing spasms. Unfortunately most patients are stressed at the time of the consultation and paradoxically don't exhibit their spasms.

# Blepharospasm

## The facts...(continued)

*“Cognitive behavioural therapy is particularly good for developing coping strategies...”*

I ask the patient to sit still, without speaking but focusing on something in front. Within a short while (probably no more than five minutes) the spasms are likely to start. The injection can then be targeted better and located in the exact part of the muscle around the eyes.

**JE:** There are a variety of measures that can be used to augment the injections such as Lundie loops which enables a patient to keep their eyes open and help improve the quality of the response to treatment. The loops are attached to the spectacles to help lift the weakened eyelids following botulinum toxin treatment.

**Q. What immediate aftercare do you recommend?**

**MH-M:** Patients with blepharospasm have a tendency to rub their eyes or to hold their eyelids, in order to keep their eyes open. I would recommend not rubbing the eyes and not washing around the eyes until the day after treatment.

We don't want to push the BTX into the middle of the upper lid to avoid spreading BTX into the levator palpebrae, which is the muscle holding the upper lid up. If it spreads, this increases the risk of ptosis (droopy eyelid).

**JE:** We don't give routine advice to those we are re-treating because they tend to know what is going to happen. However all new patients would be given appropriate advice. Some of our 'old hands' are always happy to chat with a new patient to reassure them.

**AF:** Some patients experience problems with closing their eyes and reduced blinking. They require lubricating drops to manage this condition. I offer a helpline service if anyone has any concerns about their treatment or any possible side effects.

**Q. Is there any other treatment that helps control the symptoms?**

**MH-M:** Anticholinergics, such as trihexyphenidyl, (although there is a shortage of supply). They can be replaced by procyclidine 5mg which should be taken half to one tablet three times a day.

These drugs should not be taken with alcohol, which enhance any side effects.

**AF:** Cognitive behavioural therapy is particularly good for developing coping strategies. There are some patients who have symptoms that may not suggest true dystonic blepharospasm but currently receive botulinum toxin injections. Although these symptoms are very real, they may have a different condition. It may be possible to reduce the use of botulinum toxin over time if their symptoms are being managed in a different way.

**Q. If medication is proposed, what are the side effects or contra indicators?**

**MH-M:** In order to reduce side effects, I use a combination of drugs, such as anticholinergics and clonazepam, especially for *Meige syndrome*, starting with very low doses and gradually building up in order to reduce any side effects. If there are any, I wait for them to settle before increasing the dosage to one tablet three times a day. The side effects include a dry mouth,

blurred vision, constipation, urinary retention and memory loss. The contra indications are glaucoma and prostatic adenoma. It is best not to take these drugs if pregnant as there is research linking the development of cleft palate in new-born children. Similarly, we avoid using the drug for patients over the age of 70 years. BTX injections are also avoided in pregnancy.

**Q. Does everyone who has blepharospasm also have blepharitis?**

**JE:** Blepharitis is more common in people with blepharospasm and is a chronic disorder of the surface environment of the eye and affects the eyelids. Some research speculates that people with a genetic predisposition to dystonia could develop a condition of the eye, such as blepharitis, that may result in blepharospasm.

*“I offer a helpline service if anyone has any concerns about their treatment...”*

# Blepharospasm

## The facts...(continued)

**Q.** Can patients build up a resistance to botulinum toxin type A if they have had injections for a long time, perhaps years?

**MH-M:** I recommend an interval of 10 weeks between injections. I have known some of my patients for ten or fifteen years and have never known one case of true intolerance or allergy to botulinum toxin.

**JE:** No, not on the doses we use as it is not a true immune resistance response.

### SURGICAL OPTIONS

**JE:** There are relatively few disadvantages for minor surgical procedures, such as control of excess skin or adjusting the height of the eyelid if the muscle becomes stretched. A more interventionist procedure is when a brow suspension is undertaken which requires insertion of material into the eyelid and brow to help keep the eyes open.

- **Orbicularis oculi myectomy** is a very radical procedure involving stripping out the muscles around the eye.

Not many have been performed in the UK and results have been disappointing.

- **Facial nerve avulsion** is effective but very mutilating. The primary aim of this type of surgery is to stop the nerve from stimulating the muscle, thereby stopping the patient from shutting their eyes.

In the vast majority of cases of blepharospasm, surgery is not appropriate and has a relatively small role to play in the management of the condition. The more complex cases, from brow suspension onwards, do carry the risk of permanent damage without a guarantee of success and these are not reversible. The less invasive surgical procedures will allow the botulinum toxin injections to be more effective.

*"In the vast majority of cases of blepharospasm, surgery is not appropriate..."*

# Blepharospasm

## Trial of Oxford device



● Dr Manoj Parulekar

*The 'geste antagonistique' phenomenon is a well known, if not well understood, technique to help relieve the spasms associated with dystonia.*

Many members report relief in using 'tricks' to apply pressure to their facial area in order to reduce the strength of spasms. Some patients use swimming goggles, peaked caps, tight hair bands or simply touching their cheeks.

The Oxford Eye Hospital has been working hard over the past two years to gain the approval needed to trial a simple device that attaches to spectacles to provide a touch-like pressure to the side of the face. It is hoped that the device, which will be trialled on 100 patients later this year, will provide relief for the spasms associated with blepharospasm.

The Society too has supported the trial by raising the £15,000 required to run the project. We have also provided the researchers with a list of eighty members with blepharospasm who are keen to be involved in the trial.

Though the money was raised several years ago, the relocation of the Oxford Eye Unit, the daily workload of the researchers and the lengthy and formal approval processes required to trial any medical device in the UK, have meant that the trial has not yet started.

However over the past four weeks, there has been a huge effort made to cross the final hurdle so that the trial can start as soon as possible. "We are now in the 'final straight' of the race to get the trial started," said Philip Eckstein. "The researchers, Dr Manoj Parulekar and Sister Alexina Fantato and Society staff have done a great job in the last few weeks to try to sort out the final formalities."

"We do understand how frustrating this period has been for our members who are waiting for the trial to finally start", he added. "I am personally hopeful that it will not be much longer."

# My story

## *My eyelids fell shut, heavily*

By Clare Tredgold

*Clare Tredgold recalls when her eyes shut involuntarily with the 'weight and finality of an enormous oak portcullis in a medieval castle' and how this blindness responded to an equally surprising treatment...*

We assume our eyes will open and close when we want them to. The eyelids exist simply to protect the eyes. So, to begin with I didn't pay any real attention to more frequent blinking.

The blinking began to get noticeably worse and lasted several seconds. And, even though I still had no idea what was happening to me, I did realise that I was going to have to stop driving – for the time being. I wasn't safe any more, for me or for other drivers (close your eyes and experience how long several seconds are when you can't see.) But since I worked at home, I reasoned I could manage. I could work, shop (locally), cook, attend to the boys after school, see friends (making light of my eyes) and spend time in the evening with my husband. With plenty of other engaging matters to share together I could still avoid fully facing the truth of what was happening to me – that my eyelids were out of my control and that it was getting progressively worse. For increasing stretches of time my eyes were shut. I could see less and less. My eyelids were making me blind.

Remorselessly time was passing and my lids now were not just closed, they were screwed tight shut most of the time. I tried to force them open. I couldn't. I could only wait until the muscles suddenly relaxed themselves and all was amazingly normal again. For just a few precious seconds. Then the hard, rigid spasms would be right back and I would start the fight all over again. And it was a fight. They were so strong, these minuscule muscles.

Here is an example of how much in denial I was by now. I joined an art class with a friend. Art is my passion. Seeing works of art gives me a rich pleasure and satisfaction and I love doing it. I could not stop

being involved. But as the term wore on I couldn't see to do it. I couldn't see to draw, let alone mix paint and apply it. But I kept on going. I simply sat on my artist's donkey with my drawing board propped up in front of me, scribbled a bit when my eyes flashed open for those precious seconds and returned for more of the same a week later.

My husband and I went to the theatre. I'd have done better to stay at home and listen to a radio play. We went to a brilliantly reviewed film. But I so looked forward to seeing it and was in such denial about my blindness, I'd managed to 'forget' it was Swedish and there would be sub-titles. Not only could I not see it, I couldn't begin to understand what was going on either.

We went to Paris for a romantic escape weekend, me and this so supportive husband of mine. But blind in Paris. Imagine! No gazing at the beauties and delights, no window shopping, no sitting at café tables watching the world go by. My husband's priorities that weekend were negotiating me over the newly washed kerbstones each morning, counting the steps up and down the metro stations and persuading taxi drivers I wasn't a liability (to them at least) if they had seen me trip and stumble as we approached. He took me to art galleries and deposited me in front of particular paintings so I could use all possible fingers to pull up each eyelid for



● *Clare Tredgold after her treatment*

*“Remorselessly time was passing and my lids were not just closed, they were screwed tight shut most of the time!”*

# My story *continued*

## Clare Tregold

that flashing moment of wonderful vision. And one evening he captured the last two seats at a concert so I could listen in perfect peace to a piano recital, with my eyes serenely shut. He also managed to manoeuvre me into a jazz club where we realised the saxophonist was blind too, albeit for a different reason.

Here is another vivid example of my denial, of how frightened I was of facing the truth about what was happening to me. With a friend as Chair, I became Secretary of a local association with several dozen members. A secretary needs to be able to see, to read, to write, to see the keys she's tapping, for goodness sake! A blind secretary is not much good. She is either stupid. Or scared. But I did it. I remember doing that one finger eyelid-lifting trick as I addressed envelopes with the other hand. At meetings I did manage to get down enough of the initials of

*"He managed to manoeuvre me into a jazz club where we realized the saxophonist was blind too..."*

those present in the few moments of sight my eyelids allowed. And afterwards I did manage to make sense of my blindly scribbled minutes, as long as I had enough memory of

the meeting to act as back-up – and as long as my finger could still haul up one eyelid for a vital couple of seconds. It is extraordinary how much strength even a tiny eyelid muscle can exert when crossed. It was a fight to hold that lid up. And it hurt. It is amazing the pain and struggle, physical and emotional, people make themselves endure to avoid the unbearable. I couldn't let myself focus on any of this. I didn't think about the future, I didn't think about blindness. I coped but I didn't think.

Then I met a friend I hadn't seen for a while, a medical friend. And he was horrified by what he could see was happening to me. Suddenly my life started moving again. I saw a new consultant who took one look at me and pronounced: 'This is benign blepharospasm (what a mouthful). I'm going to try botulinum toxin.'

And he did, the very next day – four tiny injections into the muscles around each eye, eight in all, one at each corner. And that's it. In a very few days my eyes began to open. And they began to stay open. Quite soon I was no longer blind. It was simply astonishing. It seemed magical. But it isn't. Not quite.

Botulinum toxin Type A or BNTA was quiet (still is) but I was noisy as I began to welcome back the old abilities, the ones we take for granted until we don't have them any more. I could read a marvellous book I'd first heard of a year ago, I could watch TV with my children, I could get around town by myself, I could socialise, I could go to the cinema and enjoy it all and I could gaze at paintings to my heart's content. And I still can.

Blepharospasm has been known about for a long time, but until BNTA there was no effective treatment. Not until a doctor in California came up with the idea of using the toxin to weaken the connection between the eyelid muscles and the ends of the nerves that are trying to send a faulty movement message to those muscles. When I developed the condition, this new treatment was only a few years old. Now it is many years older and increasing numbers of people are being diagnosed (more quickly than I was) because the treatment works so well and it is getting talked about. The effect of the injection lasts for about three months and the procedure has then to be repeated. I am often asked if it is painful. It does hurt a bit but only for a few seconds and, given these tiny injections are fighting blindness – it is eminently bearable.

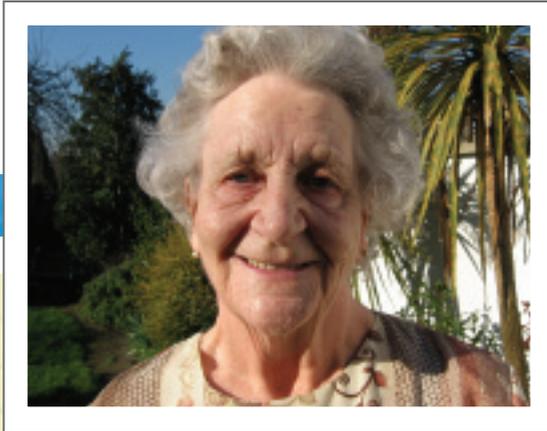
*"...I began to welcome back the old abilities, the ones we take for granted until we don't have them any more."*



*Clare's story has also been published in the magazine Real People, on February 14.*

# Case notes

## Mrs Hill



**Name:** Patricia Hill

**Age:** 68 years

**Address:** Lives in New Malden, Surrey, with husband David. They have four children and ten grandchildren.

**Condition:** Blepharospasm

### Patient Notes:

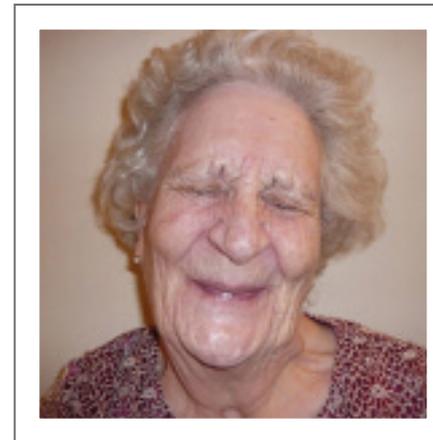
"My eyes really started to bother me about five or six years ago. I was straining to open my eyes even slightly, and eventually I used to sit with my eyes closed entirely all evening. I suffered a stroke affecting the left side of my body two years ago and left me with little peripheral vision in my left eye.

I visited opticians and eye specialists at other hospitals for treatment of glaucoma and a wrinkle on the retina but no one seemed to tell me why my eyes kept closing. Even my family and husband David couldn't understand why I kept my eyes tight shut. It wasn't that I didn't want to open my eyes. I just couldn't.

I can't understand why my condition wasn't diagnosed earlier, especially by the Royal Eye Hospital in Kingston. Eventually I saw a neurosurgeon at St. George's Hospital, Tooting, who eventually referred me to Dr. Marion."

### Diagnosis:

Mrs. Hill was referred to Dr. Marion just before Christmas last year and attended privately, but is now back as an NHS patient and was diagnosed as having blepharospasm.



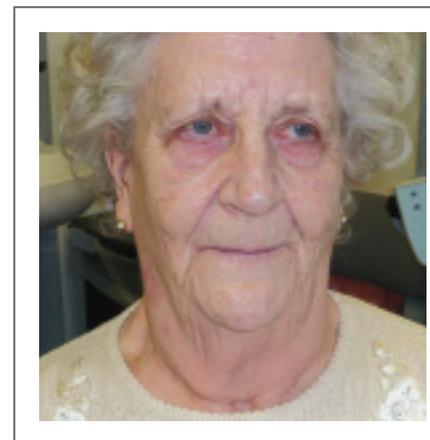
The photograph on the left before Mrs Hill's first botulinum toxin injection, shows how disabled she was, with severe spasms of eye closure and unsuccessful attempts to open her eyes.

The photograph below, one month after the botulinum toxin injections shows how much she has improved; the spasms are less frequent and less severe. Dr Marion says:

"It illustrates that one injection is often not sufficient for severe cases to alleviate the dystonia completely. This requires further injections with optimal dosages."

Dr Marion's webpage can be found on:

[www.londonbtxcentre.co.uk/drmarion.htm](http://www.londonbtxcentre.co.uk/drmarion.htm)



# Notice board

## Coping strategies for blepharospasm

### Useful hints and tips provided by our readers and members of the Blepharospasm support group

“When I was first diagnosed back in 1997, I asked my consultant what advice he could offer for the future control of my condition. He replied: ‘Do what you can do.’ I did not fully appreciate what that meant at the time but I now know the wisdom of his words. One has to face the condition daily as a number of factors can be troublesome, such as wind, bright lights, sun and the change of temperature.” *says William Sutherland B.E.M.*

#### Hot dry gritty eyes? Keep them cool

- *Minims Artificial Tears* (small phials containing one days supply). Can be purchased at chemists and also available on prescription by your GP.
- Try different brands – you may be allergic to the preservative. *Tears Naturelle*, *Viscotears* are preservative free.
- *Clarymist* – spray relief for dry eyes. Spray on closed eyes. May be difficult to find in chemists. Can be purchased direct online [www.quickvit.co.uk](http://www.quickvit.co.uk)
- Blue gel eye mask (Boots, Body Shop). Store in fridge before use.

#### Keep your eyes clean

Wash along eyelashes daily with a solution of (sterile) sodium bicarbonate or Johnsons *No Tear* baby shampoo, using ear buds to apply

#### Can't cope with bright light? Block it out

- Baseball cap, golf visor, sunglasses (a blue tint may be easier on the eye)
- Over-glasses (either tinted or clear) from most opticians for £10 – £20 depending on how stylish. Can also give protection against wind and do not limit visibility in winter

*Averil Newell of Poole says:* “When abroad, I wear a sprung visor to keep the sun out and the added pressure to the side of my head seems to reduce the spasms. Interestingly, the bright sun in Spain causes less irritation to my eyes than here in the UK.”

“I do daily word searches which help me to focus on muscular control around the area of the eyes,” *says Bill Sutherland*

#### Tricks /occupations that can help keep eyes open

- Read aloud (although tricky when sitting on a train!)
- Sing or whistle when walking ● Dancing
- Knitting or sewing
- Cooking and gardening (looking down always helps)
- Playing a musical instrument – but this of course is a gift /talent and not available to all of us

“As soon as I open my mouth, my eyes tend to close,” *says Nikki Parkin* from London Colney “It seems to help when I press the fingers of both hands together in the shape of a diamond with both thumbs placed on my chin and the forefingers on the bridge of my nose.”

Temporary relief may be obtained by taking travel sickness medication (such as *Kwells*), *Tesco Health Juice* (in a green tin) or *Benadryl* anti-histamine tablets.

“Overall, I find deep concentration on a task takes my mind off blepharospasm for a while,” *says Carroll Ashton*, “and any stress makes it worse, as do car journeys, wind or strong light.

#### Droopy eyelids? Prop them up

- Narrow porous tape (Boots own make), attach one end to outer eyelid and stretch gently to a comfortable position near eyebrow
- *Ptosis Props* are bars attached to the inside of the spectacles, which apply gentle pressure to the upper eyelids. A guide that may indicate that this will work for you is to apply pressure to the eyelid area to see if it helps alleviate the spasm. **Contact Premiere Optical on 01255 424100**
- *Lundie Ptosis Loops* work in a similar way but differ in that a person can decide on the flexibility of the lift given. Some opticians treat it as a rigid prop and cement the pivots into the frame which reduce flexibility. **Contact Mr and Mrs Lundie on 01639 750196**

#### Driving? What the DVLA says...

The DVLA has recognised that people with well-controlled blepharospasm should be able to keep their driving licence. **Ask for your DVLA fact sheet by contacting the UK Office on 0845 458 6211**

- **If you would like to know more about the Blepharospasm support group, contact Cathy Palmer on 01903 725448**





# Blepharospasm

## Should you register?

By Beverley Ricketts

*Beverley is a member of the Society and a qualified optician and gives advice on how to register as sight impaired.*

If you have poor eyesight due to blepharospasm, it's a good idea to think about registering with your local authority as severely sight impaired (SSI) or sight impaired (SI). Some people worry that registration is a backward step. They fear that it will lead to them losing their independence, or that family, friends or officials will become interfering or over-protective.

No-one can make you register if you don't want to but it has been shown to be helpful in getting extra support. Obviously if your local authority knows about your needs, then they are more able to help you. A bit of support can make all the difference in keeping your independence.

### What is the legal definition of blindness?

According to the National Assistance Act 1948, a person can be registered as SSI if they are 'so blind that they cannot do any work for which eyesight is essential'. A person may be registered as SI if they are 'substantially and permanently handicapped by defective vision, caused by congenital defect or illness or injury'.

If you are registered as blind this does not necessarily mean that you are, or will be, totally without sight.

For some people with blepharospasm, obtaining registered status can prove difficult. Meeting the criteria for registration is easy when your eyes are closed but when they are open the legal definition is not met. Registration seems to be at the discretion of your ophthalmologist, so discuss this carefully with your consultant.

If the consultant agrees that you may be registered, a certification form should be completed, giving details of the assessment of your vision and the verification that you are SSI or SI.

*A table of agencies that can help is available from the UK Office by calling 0845 458 6211*

### Sight loss and registration

Each local authority keeps a register of blind and partially sighted people living in its area. The register is held by the social services department, or in some areas, a local voluntary society for people with sight problems acting as agents for the local authority. The register is confidential and covered by data protection legislation.

Registration is voluntary but it is helpful in getting extra support. However, anyone having difficulties because of poor eyesight is entitled to ask their local social services department to assess their needs, even if not yet registered. If you are having difficulties because of your poor sight, you should not hesitate to get in touch with your local social services department or low vision service. In some places there are professionals who can teach a range of skills to people with low vision. These include indoor and outdoor mobility, communication and other skills for daily living. The factsheet will help you understand what you may be entitled to. For further information and to obtain a factsheet, please contact the Dystonia Society Helpline on 0845 458 6322 or email [support@dystonia.org.uk](mailto:support@dystonia.org.uk)

The Royal National Institute of Blind People (RNIB) can also be of service. They can be contacted via their helpline on 0845 766 9999. Or you can visit their website via this link: [www.rnib.org.uk](http://www.rnib.org.uk)

*PLEASE NOTE: Not everything mentioned in the fact sheet is available for people registered as SSI or SI. For example, some of the benefits which may be available have their own special rules. If you wish to receive to receive a fact sheet, it will be for guidance only and is not an authoritative statement of law.*

