

Benign Essential Blepharospasm RESEARCH FOUNDATION



anniversary
1981-2011

Serving BEB/Meige and related disorders

MISSION STATEMENT

RESEARCH TO CURE • SUPPORT TO CARE • EDUCATION TO ENLIGHTEN

VOLUME 30 • NUMBER 4 • JULY/AUGUST 2011



HAPPY BIRTHDAY BEBRF!!!

Richard L. Anderson, MD, FACS, Center for Facial Appearances, Salt Lake City, Utah



It is hard to believe that it has been over 30 years since Mattie Lou Koster was told “it couldn’t be done” to start a blepharospasm foundation. She didn’t want others to go through what she had experienced in getting diagnosed and treated for blepharospasm. As always, she was up to the challenge and within months, not years, it was done and the Benign Essential Blepharospasm Research Foundation (BEBRF) was founded. Education and clinical and basic research on blepharospasm was begun and the disease was brought “out of the closet” by the “Texas Tornado” (Mattie Lou Koster). Prior to this time, blepharospasm was so poorly understood that historically some patients had been institutionalized as possessed and still many doctors believed it to be a psychiatric disorder.

1981 was a very good year for blepharospasm as Mattie Lou created the BEBRF, I first published myectomy surgery with 5 year results, and Alan Scott first published the clinical use of oculinum toxin (later to be called Botox) for strabismus and blepharospasm.

Mattie Lou quickly created public awareness regarding BEB and if editors etc. weren’t interested, she would wear them down with her enthusiasm, which became infectious for all of us. She

even got front page coverage in the Wall Street Journal and I woke up one morning with everyone I knew calling me saying my name was on the front page of the Wall Street Journal regarding blepharospasm and myectomy surgery. No one could turn her down, and yearly meetings were created to educate doctors and patients. The “Texas Tornado” even brought a real tornado with her to the second international meeting of the BEBRF that I organized in Iowa in 1982. Alan Scott began encouraging doctors to try neurotoxins for BEB and I began using it in 1982. Fortunately, after we got the dosing figured out, neurotoxins helped most patients or the only operation I would have had time to do would have been myectomy following the Wall Street Journal coverage. While I have published over 400 scientific papers in journals and over 100 book chapters, one paragraph in the Wall Street Journal got more viewers than all of those medical publications.

Mattie Lou surrounded herself with the best doctors in the field and made us be better and work harder. She had us help organize international meetings and do a better job of caring for patients. She organized funding for research on blepharospasm. She never personally complained and always worried about others. She said

Continued on page 11



Richard Anderson, MD and Mattie Lou Koster at The Ophthalmology Academy, 1995

CALL FOR RESEARCH GRANT PROPOSALS

The Benign Essential Blepharospasm Research Foundation (BEBRF) funds research into new treatments, pathophysiology, and the genetics of the following focal dystonias: benign essential blepharospasm (BEB) and Meige syndrome (cranial and oromandibular dystonia). Research into photophobia, dry eye, and apraxia of eyelid opening as they relate to BEB and their treatment will also be considered for funding. Funds up to \$150,000 are available annually. M.D. or Ph.D. required for principal investigator. Non-U.S. citizens working at institutions abroad are also eligible to apply for a research grant. Deadline to apply for this year is August 31, 2011. Grant guidelines and a listing of previous grants may be obtained from: Benign Essential Blepharospasm Research Foundation, Inc., P.O. Box 12468, Beaumont, TX 77726-2468, Phone (409) 832-0788, Fax (409) 832-0890, E-mail bebrf@blepharospasm.org or visit our Web site: www.blepharospasm.org to download forms.



The Benign Essential Blepharospasm Research Foundation (BEBRF) is a non-profit, 501(c)(3) organization founded in 1981 by Mattie Lou Koster, a blepharospasm patient.

Blepharospasm means eyelid spasm. The eyelids unpredictably and involuntarily clamp shut in both eyes, leaving the victim functionally blind until the spasm ceases in a few seconds or a few minutes.

Meige Syndrome is a similar condition in which involuntary muscle spasms in the lower face and jaw cause grimacing and jaw movements.

Hemifacial Spasm generally begins as an involuntary contraction around one eye that gradually progresses down one side of the face to the cheek, mouth and neck. It is not a form of dystonia.

Blepharospasm and Meige are classed as movement disorders and are described as focal dystonias.

BEBRF is a member of the National Organization For Rare Disorders (NORD), WE MOVE, Movement Disorders Society, Dana Brain Alliance, American Brain Coalition, Dystonia Advocacy Network, Dystonia Coalition - ORDR, Harvard Brain Tissue Resource Center, Laurie Ozelius, PhD is the BEBRF Representative to the Harvard Brain Tissue Resource Center

Benign Essential Blepharospasm Research Foundation

637 North 7th Street, Suite 102
P.O. Box 12468
Beaumont, Texas 77726-2468
Phone: 409-832-0788
Fax: 409-832-0890
E-mail: bebrf@blepharospasm.org
Web site: www.blepharospasm.org

Contributions may be sent to:
BEBRF
P.O. Box 12468
Beaumont, Texas 77726-2468

The Editorial Staff reserves the right to edit any and all articles. It is our editorial policy to report on developments regarding blepharospasm, Meige and hemifacial spasm, but we do not endorse any of the drugs or treatments in the Newsletter. We urge you to consult with your own physician about the procedures mentioned.

The Blepharospasm Newsletter is published bi-monthly and mailed to patients, families, doctors, friends of the foundation, and health care providers around the world.

Subscription is \$15.00 U.S. and \$20.00 elsewhere.

The Importance of Brain Research

Mark Hallett, MD, Chairman, BEBRF Medical Advisory Board



Mark Hallett, MD

Despite continuing effort, we still do not understand why patients have blepharospasm. We do know that there is some brain dysfunction, but we do not know where it comes from in the brain itself. Routine imaging of the brain does not show any abnormality, and pathological examination of the brain has not shown any abnormality either. With new neuroimaging techniques using sophisticated magnetic resonance imaging there are some clues to where abnormalities might be, but nothing has yet been identified. To a certain extent, this is good, because this means that there is no severe abnormality. On the other hand, it is frustrating, because there must be some abnormality somewhere that leads to the involuntary muscle spasms.

I am addressing my remarks to the common primary blepharospasm. There are some rare cases of blepharospasm that appear to be due to small strokes in the brainstem or basal ganglia. The blepharospasm comes on suddenly in this situation. In these circumstances, we do know the underlying pathology. There may be some relationship between the pathology in the two types of blepharospasm, but we do not know this one way or the other.

Although it is true that there is no known pathology, the information on which this statement is based is only minimal. There are very few brains available from patients who had blepharospasm. One of the reasons for this is again good; people do not die from blepharospasm, and at the time of death, blepharospasm is only a remote thought. However, unless there are brains available, it will be impossible to identify the relevant pathology. Hence, it would be important for patients with blepharospasm to donate their brains for research.

What will we learn when an abnormality is found? The information should be very valuable in understanding how the spasms come about. And, understanding of the physiology should lead to better treatments. A clear example of this comes from Parkinson's disease, where important pathology was identified in the substantia nigra, a major source of dopamine in the brain. This knowledge was one of the factors leading to the idea of dopamine replacement for therapy, and, of course, this is one of the great successes of modern medicine.

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FOCUS ON THE FOUNDATION:
BEBRF AND THE INTERNET

Robert Campbell, PhD, Webmaster



The phrase which comes to mind is, “You’ve come a long way, baby.” In 1981 when Mattie Lou Koster established BEBRF, personal computers were just entering the workplace. The group of ten I worked with managed to get one but we had to prepare a 10-page business case to justify the purchase. In short order, we were all competing for the machine and thus decided we should get another. However, we found that in order to have TWO personal computers for 10 people we had to prepare a 30-page business case. By the time I retired in 2006, it was not unusual for an individual in the workplace, or at home, to have two or more personal computers – and the only constraint was budget.

The release of the first browser with a graphical user interface (GUI), Mosaic, in 1993, paved the way for the use of the Internet and, in particular, the World Wide Web, as a tool for individuals to seek information on virtually any topic. The first information on dystonia appeared on the World Wide Web in 1995 as a single web page. In 1997, BEBRF launched a site dedicated to supplying information about blepharospasm <<http://www.blepharospasm.org/>>. In the same year, an on-line support group in the form of the “Blepharospasm Bulletin Board” was established <<http://www.blepharospasm.org/forums/beb/>>. In 2004, we added the ability to order BEBRF materials and renew subscriptions on-line. In 2005, a photo gallery was added which allowed on-line access to photos of conferences and support groups, and, in particular, photos of patients’ experiences with treatments such as myectomy, brow pins, ptosis crutches and botulinum toxin injection sites <<http://www.blepharospasm.org/gallery3/>>. In 2009, we added the first on-line video, Dr. Soparkar’s lecture on “Dry Eye and Blepharospasm” <<http://www.blepharospasm.org/dry-eye-video/video.html>>.

Where are we now? The web site contains 2.8 gigabytes of information, including 20 videos. Most registrations for regional symposiums are made on-line rather than by mail. The bulletin board had 192 members at the end of 2004 and 1400 at the end of 2010. In 2010, 9,000 messages were posted on topics including: experiences with Xeomin, Myobloc, Dysport and Botox, brow pin surgery, myectomy, sunglasses, coping with ignorance of others, getting disability, driving with blepharospasm, Zytaze, double vision, job interviews, and dependence. As mentioned in the January/February issue, “If the BEBRF has your current e-mail address, you have the means of receiving information that can make a difference to you and other patients. When an important issue needs to be addressed, usually quickly, you will receive an Action Alert explaining it and instructing you on the steps you need to take to contact your senator or congressman.”

What is ahead? For the 30th anniversary of BEBRF, a more user-friendly version of the web site will be released.

BEBRF’s web resources are there for you. View one of the videos <<http://www.blepharospasm.org/video-library.html>>. Check out the videos of presentations at the 2009 conference to see the sort of information you will miss if you do not attend one of the upcoming regional symposiums <<http://www.blepharospasm.org/2009conference/index.html>>. Visit the bulletin board <<http://www.blepharospasm.org/forums/beb/>>. Lurking is quite acceptable – you only have to register if you wish to contribute to the discussion. Find out the latest thinking on apraxia of eyelid opening <<http://www.blepharospasm.org/apraxia-0.html>> or oral medications <<http://www.blepharospasm.org/oral-meds.html>>. Use the site search feature to find information about a topic of interest to you <<http://www.blepharospasm.org/pfsearch-main.html>>. Save the cost of a stamp by renewing your newsletter subscription on-line <<http://www.blepharospasm.org/Merchant2/merchant.mvc?>>>. ☺



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NEWSLETTER DEADLINE DATES

ISSUE	COPY DUE DATE
Sept/Oct	August 3, 2011
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Dry Eyes Associated With Blepharospasm

Seongmu Lee, M.D. and Michael T. Yen, M.D., Cullen Eye Institute, Baylor College of Medicine, Houston, Texas

Dry eyes are not an uncommon finding in patients with blepharospasm. While the precise mechanism for ocular dryness in essential blepharospasm is not known, it is believed to be multifactorial and may be related to and exacerbated by forceful eyelid closure and increased blinking frequency. Additionally, irritation of the ocular surface from a dry eye condition may result in secondary blepharospasm.

The tear film is composed of an external lipid component, an aqueous component, and a mucin component. The aqueous component is produced by the lacrimal gland (tear producing gland) and contains various growth factors, electrolytes, and anti-bacterial substances. The meibomian glands along the eyelid margin produce the lipid component, which plays an important role in slowing the evaporation of tears, lubrication of the surface of the eye, and stabilization of the tear film.

The causes of dry eye can be divided into decreased aqueous tear production and increased evaporative tear loss/dysfunction. Causes of reduced tear production include decreased secretion, tear duct blockage, lacrimal gland disease, and medication side effects. Evaporative forms of dry eye disease include increased exposure of the eye surface resulting from eyelid abnormalities, lipid tear layer deficiency with an unstable tear film, and chronic allergy, toxicity, or inflammation. Changes in tear composition and tear film quality as a result of meibomian gland dysfunction and blepharitis is a common and important cause of evaporative dry eye.

Objective findings of dry eye are usually less than the frequency of complaints in blepharospasm patients with dry eyes. Symptoms of ocular irritation, burning, foreign-body sensation, and blurred vision can be associated with ocular dryness. These symptoms tend to be greatest at the end of the day or after prolonged use of the eyes. Signs of dry eyes can include redness of the eye, decreased tear volume, debris in the tear film, and erosions of the epithelial surface of the cornea. In patients with severe dry eyes, filaments and mucous plaques may be present.

The primary goals of treatment focus on improvement of symptoms, improvement of the stability of the tear film, and reversal of damage to the ocular surface. When meibomian gland dysfunction is present, warm compresses to the eyelids can be very beneficial.

The most common therapy is the application of artificial tear supplements topically to the ocular surface. Numerous preparations are available and include artificial tears, gels, and lubricating ointments. Ointments and higher-viscosity formulas remain in the eye for a longer period of time, providing a longer interval of symptomatic relief. These more viscous formulations, however, may be associated with an increased incidence of blurry vision and lash residue.

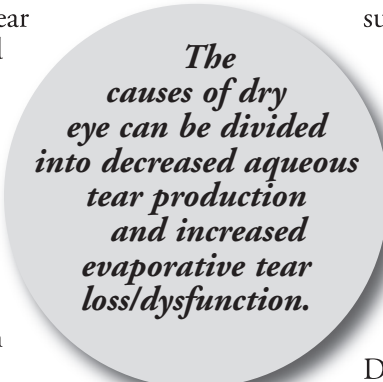
Additionally, artificial tears may contain preservatives that can produce corneal irritation, and any patient that requires application of tear substitutes more than four times a day should use preservative-free formulations.

Occlusion of the puncta (drainage holes for the tears) utilizing silicone plugs, cautery, or laser may be beneficial by helping to retain and preserve tears. Occlusion of the lower punctum may be sufficient, but occlusion of both the upper and lower puncta may be necessary in severe cases. Humidifiers and moisture shields also reduce the evaporation of tears and may be a useful adjunctive treatment. A tarsorrhaphy (suturing part of the eyelids closed) may be useful by reducing the surface area of exposed ocular surface, thereby reducing the evaporation of tears. However, a tarsorrhaphy is usually reserved for patients with severe exposure and decompensation of the ocular surface.

Recent studies have suggested a role of inflammation in dry eye, with suppression of secretion and subsequent damaging effects on the lacrimal gland and ocular surface. Unpreserved topical steroids have been shown to be effective in reducing inflammation and restoring a healthy ocular surface in patients with dry eye. However, side effects, including increased risks of glaucoma, cataract, and infection, have limited its widespread use. Topical cyclosporine (Restasis) has been shown to reduce cell-mediated inflammatory responses and thereby effectively improve the ocular signs and symptoms of dry eyes. Doxycycline, azithromycin (Azasite), and omega-3 fatty acids have been reported to improve symptoms and tear film stability via an anti-inflammatory effect.

Autologous serum is an additional treatment option for patients with dry eye, as serum contains essential growth factors that are normally present in tears. Studies have shown improved healing of the ocular surface and enhanced mucin production in dry eye patients treated with autologous serum. The main disadvantage of this therapy is the time-consuming nature of its preparation.

In summary, as our understanding of the mechanisms of dry eye continues to advance, treatment strategies continue to evolve. Dry eyes associated with blepharospasm can be a challenging problem. Numerous treatment options are available, and various trials with careful monitoring and assessment of treatment response may be necessary to find an optimal regimen that provides maximum relief. A meticulous examination to accurately identify the contributing factors and addressing these components is important. In general, mid- to high-viscosity compounds in combination with aggressive lid hygiene and warm compresses are likely to produce a greater reduction in dry eye symptoms. ●



The causes of dry eye can be divided into decreased aqueous tear production and increased evaporative tear loss/dysfunction.


Blepharospasm: New Directions in Research

Joel S. Perlmutter, Washington University, St. Louis, MO

Blepharospasm is a focal dystonia affecting muscles around the eyelids. Although many people that develop these involuntary muscle spasms have antecedent irritation or dryness of the eyes, the primary problem does not reside around the eye but rather in the brain. Much research has focused on identifying where the problem is in the brain, what abnormal function occurs there and what causes it. Only with this type of basic research can we hope to develop a treatment that treats the cause rather than the symptoms of this disorder.

Initially research focused on where in the brain the problem resides. Information for that has come from several sources. The first source has been identifying strokes or brain lesions that have been associated with blepharospasm or other focal dystonias. Multiple sites have been found but the most common anatomical sites has been an area deep in the brain called the striatum – a part of the basal ganglia. Subsequent research has found changes in chemical pathways in this part of the brain, in particular pathways that are, in part, controlled by a chemical messenger called dopamine. Dopamine has different actions on nerve cells depending upon which type of dopamine receptor it hits. A fair amount of research has investigated which of the several types of dopamine receptors may be involved in blepharospasm – the D2-family of dopamine receptor is most suspect. Other drug-induced types of dystonia may also be caused by drugs that interfere with these dopamine-mediated pathways.

Other information supports the notion that dopamine pathways are critical. This other information comes from understanding the actions of brain pathways affected by genetic forms of dystonia. Apparently, the abnormal protein in one of these genetic forms is made in and near dopamine nerve cells. However, these abnormal proteins are also produced in another part of the brain – the cerebellum.

That leads to another area of new research investigating the role of the cerebellum in blepharospasm and dystonia. Neuroimaging studies and anatomy studies now connect cerebellum to the dopamine-mediated pathways in the basal ganglia. Thus these two areas may be highly inter-related. 

**REMEMBER BEBRF
IN YOUR WILL.**

The Preoperative Evaluation of the Patient Considering Microvascular Decompression for Hemifacial Spasm

Raymond F. Sekula, Jr., MD, University of Pittsburgh School of Medicine

INTRODUCTION

Nonoperative treatment options for hemifacial spasm (HFS) include anticonvulsants and serial botulinum toxin injections, and operative treatment options include microvascular decompression (MVD). Unfortunately, anticonvulsants (e.g. carbamazepine, gabapentin, etc.) are ineffective in the treatment of HFS. Serial botulinum toxin injections are tolerated well by some rather than others, but essentially, botulinum toxin injections exchange weakness for cessation of spasms of the treated facial musculature. MVD of the facial nerve addresses the presumed cause of HFS, vascular compression of the facial nerve^{3,6}, and is effective and durable.¹⁰ MVD, however, is associated with risks (e.g., cerebellar hematoma, cranial nerve injury, stroke, and death), albeit infrequent, not associated with serial botulinum toxin injections for HFS. Some practitioners argue that the benefits of MVD for HFS do not outweigh the risks.

In our center, we perform MVD of the facial nerve as the first-line procedure for HFS in patients able to undergo MVD regardless of age or prior history of failed MVD because a well executed MVD of the facial nerve provides the highest likelihood of success (i.e. spasm-free), the best quality of life (i.e. symmetric and normal facial function), and the lowest long-term recurrence rate of hemifacial spasms. Many of our patients have undergone an unsuccessful MVD in the past. Evaluation of the risk/benefit ratio for each patient before operation, however, is essential in optimizing care. With this in mind, this article is dedicated to the preoperative evaluation of the patient with HFS including appropriate supplemental testing in anticipation of MVD.

STEP 1: CONFIRM THE DIAGNOSIS OF HFS

HFS is prevalent in 9.8 per 100,000 persons which means that approximately 30,000 Americans are affected by HFS at the present time.⁷ The diagnosis of HFS is based on the clinical history and neurological examination. HFS, a syndrome of unilateral facial nerve hyperactive dysfunction, is a severe and disabling condition that causes impairments in the quality of life.¹ In most cases of hemifacial spasm, spasms begin insidiously in the orbicularis oculi muscle (i.e. the muscle about the eye) and spread over time to the muscles of the face with variable involvement of the frontalis (i.e. muscle of the forehead) and platysma (i.e. muscle of the neck) muscles. Ultimately, the patient may develop prolonged contractions of all the involved muscles causing severe, disfiguring grimacing with partial closure of the eye and drawing up of the corner of the mouth, the so-called “tonus phenomenon.”⁴ Some patients will report worsening of spasms with fatigue, situations of anxiety, and changes in position of the head (e.g. head to one side or the other on the pillow at night). Patients also frequently complain of new “noises” in the ear and a feeling of “fatigue” of the face of the affected side of the head as the day progresses.

STEP 2. RULE OUT OTHER DISORDERS, WHICH CAN BE CONFUSED FOR HFS WITH AN EMG

Although the diagnosis of HFS is made clinically, electromyography (EMG) (i.e. a n electrical needle test of the face) may help in distinguishing the disorder from other abnormal facial movement disorders such as blepharospasm, tics, partial motor seizures, synkinesis, Meige’s syndrome, and neuromyotonia.^{5,11} The electrophysiologic hallmarks of HFS consist of spontaneous, high frequency (as many as 150 impulses per second), synchronized firing on EMG and an abnormal motor response (AMR) elicited with the trigeminofacial or “blink” reflex. The AMR is the recording of a response in the orbicularis oris muscle to electrical stimuli applied over the supraorbital nerve (and sometimes a stimulus

Continued on next page

The Preoperative Evaluation of the Patient Considering Microvascular Decompression for Hemifacial Spasm

Continued from previous page

to a single facial motor branch) when the trigeminofacial reflex should be limited to the orbicularis oculi.

STEP 3. OBTAIN A BRAIN MRI TO EXCLUDE A STRUCTURAL PROBLEM

Some confusion exists amongst clinicians regarding the utility of MRI in predicting which patients may benefit from proposed microvascular decompression. This confusion exists, in part, because of conflicting reports regarding the ability to detect vascular compression of the facial nerve with MRI and also the significance of vascular compression by MRI. Occasionally, patients are not referred for MVD because the interpreting radiologist, neurologist or neurosurgeon does not note neurovascular compression of the facial nerve. This is a mistake. Lack of visible neurovascular compression can be attributed to technical inadequacies of MRI sequencing or simply the inability of even the most advanced MRI sequencing to detect certain neurovascular conflict (i.e. many compressing vessels are small and venous).⁸ We frequently note apparent compression of a nerve on MRI preoperatively only to discover during the operation a different vessel causing the problem and/or multiple vessels in contact with the nerve. In summary, although we can often see the offending artery compressing the facial nerve by preoperative MRI, we primarily use MRI of the brain with gadolinium to exclude structural lesions including tumors, AVM, Chiari I malformation and other confounding diagnoses.⁹

STEP 4. OBTAIN A COMPREHENSIVE MEDICAL EVALUATION BY YOUR INTERNIST

The ultimate goals of preoperative medical assessment are to reduce the morbidity associated with operation, to reduce the need for prolonged perioperative care, and to return the patient to his or her life without hemifacial spasms. We routinely operate on patients classified as Grades I-IV according to the American Society of Anesthesiologists scale²:

- I. A normal healthy patient
- II. A patient with mild systemic disease
- III. A patient with severe systemic disease
- IV. A patient with severe systemic disease that is a constant threat to life

We work closely, however, with a patient's other physicians to best prepare a patient for the operation.

STEP 5. BEFORE PROCEEDING WITH MVD, CONSIDER THE RISKS OF MVD CAREFULLY

Because MVD for HFS requires the surgeon to dissect about the lower cranial nerves, the risk profile of MVD for HFS (particularly facial, cochlear, glossopharyngeal, and vagus nerves) is different than the risk profile of MVD for other cranial neuralgias (e.g. trigeminal neuralgia). Cranial nerve injuries during MVD may result in facial weakness, hearing impairment, balance troubles, and swallowing difficulty and/or hoarseness, which can affect satisfaction with MVD despite the absence of hemifacial spasms postoperatively. Many clinicians (e.g. neurologists, neurosurgeons, ophthalmologists, plastic surgeons) who routinely treat patients with HFS are appropriately reluctant to refer patients for MVD

because they feel that the risks of MVD outweigh the benefits, and they may have cared for patients who have suffered serious consequences of an MVD in their practice. One patient recently told me that their local neurosurgeon warned them "MVD is a bloody mess of an operation..stick with botulinum toxin injections."

Although MVD can be "a bloody mess of an operation", the operation can be routinely completed with no more than a teaspoon of blood loss in experienced hands. In this author's experience of more than 1000 MVDs for a variety of cranial neuralgias, only one patient has required a blood transfusion. Indeed, in recent years, the risk profile of MVD for HFS has improved with further refinements of the operative technique. In this author's experience of more than 250 MVDs for HFS in the past five years, 92% of patients have become spasm-free following an operation. In that same group of patients, only one patient suffered a stroke, one patient sustained a partial facial nerve injury, and three patients lost their hearing on the affected side of the head. Additionally, no patients sustained infections or cerebrospinal fluid leaks.

STEP 6. ONCE A DECISION HAS BEEN MADE TO PROCEED WITH MVD, A FEW OTHER TESTS ARE REQUIRED TO OPTIMIZE MVD RESULTS

Once a patient has made the decision to proceed with an MVD of the facial nerve, audiometry (i.e. hearing test), acoustic middle ear reflexes, and brainstem auditory evoked potentials (BAEPs) testing should be completed. The audiometric tests are performed preoperatively to obtain a baseline for quantitatively determining deteriorations or improvement in hearing function following MVD. Additionally, preoperative BAEPs provide baseline information for the clinical neurophysiology team so that they may warn the surgeon of any deviations during monitoring of the intraoperative auditory evoked potentials to preserve hearing during the EMG.

CONCLUSION

MVD of the facial nerve for HFS remains the only chance for a cure of HFS. Each patient must carefully consider the risk/benefit profile of such an operation versus continued serial injections with botulinum toxin. Patients should work closely with their physicians to develop a long-term plan for HFS and consult an appropriate neurosurgeon when considering MVD. ●

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Advancing Blepharospasm Research Through Brain Donation

The Benign Essential Blepharospasm Research Foundation, working with other members of the Brain Collective, is proud to partner with the Harvard Brain Bank to help solve the mystery of blepharospasm and other dystonias through brain donation. The Harvard Brain Bank is responsible for collecting, preserving, and distributing human tissues to qualified scientific investigators who are conducting important blepharospasm/dystonia research. Dr. Laurie Ozelius, BEBRF Medical Advisory Board, represents BEBRF on the research committee which reviews applications from researchers requesting brain tissue. Since the majority of scientific research studies can be carried out on a very small amount of tissue, each donated brain provides a very large amount that can be used by many different researchers at institutions throughout the U.S. and the world.

Commonly Asked Questions

1. *Is there a cost involved to participate as a donor?*

No. The BEBRF assumes any and all costs, so there is no expense to the family.

2. *Do I need to live near Massachusetts in order to enroll as a potential brain donor?*

No, but you must live in the United States in order to participate in this program. The Harvard Brain Bank works in conjunction with pathologists and funeral homes throughout the United States and will communicate with the specialists who actually collect the donated brains. They are then transported to the Brain Bank, preserved, stored, and made available for analysis.

3. *Do you only need donated brains from blepharospasm-affected individuals?*

No. In fact, we need brain donations from both persons who have BEB/Meige, as well as from those who do not. The latter are used for comparison purposes and are referred to as “control subjects. So please encourage your family members and friends to also pre-enroll as donors.

4. *If I sign up to be a brain donor, does that preclude me from donating other organs for transplant purposes?*

It may, depending on how long the procedure takes for retrieving the donated organs and how long the donor was on a respirator.

5. *Does the Harvard Brain Bank accept whole body donations?*

No. The facility is not equipped to receive such donations.

6. *Will being a brain donor interfere with funeral arrangements or memorial services in any way?*


Absolutely not. You may have any sort of service or remembrance that you and your family desire, as the brain recovery process does not cause any disruption in those plans.

7. *Will the family of the donor receive any communication from the Harvard Brain Bank after their loved one has passed away?*

Yes. The family will be asked to complete a questionnaire about the donor and will also be asked to grant permission for the donor’s medical records to be sent to the Brain Bank. The medical history and questionnaire are both important so the Brain Bank can reliably give researchers the correct tissue samples needed for their specific studies. When this information is received and after the tissue has been analyzed by the neuropathologist, the family will receive a copy of the final neuropathology report.

8. *If someone has had the Deep Brain Stimulation (DBS) surgery or another type of brain surgery, does that prevent them from participating as a brain donor?*

No, researchers will want to study these brains and the donors medical records will provide needed information for researchers.

For more information on brain donation, contact the
BEBRF Office: 1-409-832-0788, Email:
bebrf@blepharospasm.org 

NEW E-MAILS AND E-MAIL CHANGES

In this information age, the fastest way to communicate is E-mail. If you have a current E-mail address, and you will let us know what it is, we will be able to quickly forward information to you that may be of interest to BEB, Meige or hemifacial spasm patients. To add or change your E-mail information, contact the BEBRF office at: BEBRF, Inc., P O Box 12468, Beaumont, TX 77726-2468. Tel.: (409) 832-0788 Fax: (409) 832-0890E-mail: bebrf@blepharospasm.org

Dystonia Adv

Nilda Rendino, 1st Vice-Pre

The fifth Advocacy Day sponsored by the Dystonia Advocacy Network (DAN) brought together in Washington DC on May 10-11 patients, family members, and friends from its five member organizations (BEBRF, DMRF, NSDA, NSTA, ST/Dystonia) as well as from other dystonia organizations. Because the legislative process is subject to constituent influence, advocates met with their members of Congress or their staff members, most of who were in committees related to the issues important to dystonia patients.

To prepare new advocates for their first visits, Dane Christiansen, DAN's grassroots coordinator, gave a brief refresher course on the Congress and the legislative and budget processes. Dale Dirks, President of Health & Medicine Counsel of Washington, then spoke to them about the importance of advocacy and gave them tips on making an effective congressional visit.

Experienced advocates joined the group to review the issues that they would be presenting the next day on Capitol Hill. Their communications focused on the effect that their specific form of dystonia has on their lives. They were put into teams according to their home states. First time advocates were apprehensive but Maria Sirvent and her husband stated: "I want to thank you again for including us in the Dystonia Advocacy Network Day. Tony and I met some very wonderful people and got quite an education from the whole process. There were many things that we were not aware of about dystonia. You were right about my nerves, after about the second meeting I started to relax. It was, all in all, a wonderful experience and we hope that we helped the effort in some way with our participation."



L-R: Senator Mark Warner (D-VA), Dee Linde, Nilda Rendino, BEBRF Eastern District Director & Advocacy Chair, and Hunter Webster

The Issues:

NATIONAL INSTITUTES OF HEALTH (NIH)

While legislators struggle with the budget and the national debt, we wanted to remind them of the importance of adequately funding the NIH, where the vast majority of federally funded dystonia research is conducted. In addition, Congress established the Cures Acceleration Network (CAN) This Is a landmark initiative to move potential new drugs and therapies through the treatment development pipeline faster. Unfortunately, if Congress doesn't fund it, the NIH director will have to redirect funding from other areas or reduce the effectiveness of the CAN initiative.

DYSTONIA ON DEPARTMENT OF DEFENSE LIST

Dystonia is becoming more prevalent among troops returning from the wars in Iraq and Afghanistan. For this reason the DAN asked Congress to include dystonia as a condition eligible for study under the Department of Defense (DOD) Peer Reviewed Medical Research Program. It was on the list for the first time in FY2010 and again for FY2011. However, it is not a permanent listing so advocates requested support from their legislators to have it included in the FY2012 DOD appropriations bill.

PATIENT ACCESS TO DYSTONIA THERAPIES AND TREATMENT

Follow-on Biologics or Biosimilars: Since generic medications reduce the cost to Medicare, Medicaid and private insurers, Health Care Reform mandated that the Food and Drug Administration (FDA) develop a regulatory pathway for biosimilars, biologic equivalent to generic drugs. However, unlike generic drugs, biologics such as botulinum toxin are extremely complicated and not easily duplicated.

The DAN is urging members of Congress to work with



Representative Ileana Ros-Lehtinen (R-FL) was presented with the 2011 DAN Distinguished Public Service Award. L - R: Sarah Buchanan and Dale Dirks (Health & Medicine Counsel of Washington), Janet Hieshetter (DMRF), Representative Ileana Ros-Lehtinen, Millie Munoz (DMRF), and Nilda Rendino (BEBRF).

Advocacy 2011

President and Advocacy Chair



BEBRF Advocates. Front row, L - R: Annette Dickson, Carol Taberski, Ilana Knopfelmacher, Nilda Rendino, Dee Linde. Back row, L - R: Norb Brouchoud, Sara Jane Brouchoud, Maria Sirvent, Mary Lou Thompson, Barbara Benton, Barbara Johnson, Vicky Nottingham, Larry Phelps. Missing from the photo is Ena Wilmot and Photographer, Tony Sirvent.



L- R: Mary Lou Thompson, BEBRF President, Senator Jim Inhofe (R-OK), Shelly Gray.

the FDA so they develop a biosimilar regulatory pathway that will ensure strong standards of safety such as additional clinical trials and will not require that patients be switched between products without a physician's approval.

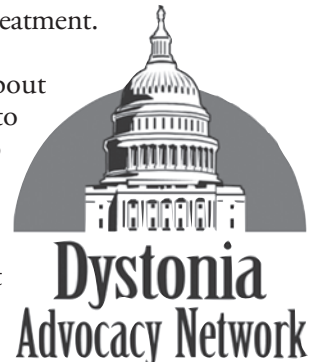
Medicare Physician Reimbursement: Advocates also asked that once and for all Congress fix the way physicians are reimbursed instead of continuing to offer yearly patches that create uncertainty for Medicare beneficiaries and their physicians. The latest short-term fix is scheduled to expire on January 1, 2012. According to the AMA, the scheduled decrease in payment to them could result in approximately 60% of physicians deciding not to treat Medicare patients. This could have serious consequences for dystonia patients, who require

skilled injectors for their botulinum toxin treatments.

Insurance Coverage for Deep Brain Stimulation: Use of these devices is authorized by Federal Law but some private insurers consider the procedure to be experimental or investigational and refuse to cover the service. The procedure is classified by the FDA as a Humanitarian Use Device (HUD) with a Humanitarian Device Exemption and can be used to treat generalized dystonia. This means that the FDA has determined that the device doesn't pose a significant risk of illness or injury to a patient and that probably the health benefit outweighs any risk.

The DAN urged Congress to solve this problem either by having the FDA add language which states that a HUD designation constitutes an explicit FDA approval of the product and that the Center for Medicare and Medicaid Services (CMS) issue a statement to the local insurance carriers telling them that this is an approved treatment.

DAN Website: To learn more about the Dystonia Advocacy Network go to its website. <http://dystonia-advocacy.org/>. To learn more about the legislative process, go there and click on "legislative process". Please provide the BEBRF with your current email address to receive action alerts. YOU can make a difference. 🗳️





BEBRF Birthday Celebration

Georgia's BEBRF 30th Birthday Celebration

Sharon Rakness held a support group meeting in Georgia and celebrated BEBRF's 30th Birthday. The celebration was well attended with 22 people sharing in the festivities.



Birthday Celebration

Cake and balloons for BEBRF's 30th Birthday Celebration in Georgia.



Iowa Support Group Meeting

Celebrating BEBRF's 30th Birthday.

Iowa Support Group Meeting

Guest Speakers: Conley Call, MD, Alycia Marler, Allgeran Rep.



West San Fernando, California Group Celebrates BEBRF 30th Anniversary

Left to right: Volunteers Lenny & Dodie Heller, Coordinators Mark & Carmella Sheeler of the West San Fernando Valley BEB Support Group in Southern California. Every attendee (26) received a cup cake with a candle which were blown out after singing "Happy Birthday!"

Happy Birthday BEBRF!!!

Continued from front page

she was retiring after the international BEB meeting I organized in Utah but all of us knew that was just a trick to get more publicity. She continued to attend meetings and organize brainstorming meetings for doctors, researchers, etc. until she could no longer walk. I vividly remember a meeting in Washington DC where we carried her in a chair up the stairs to dinner. She looked like a queen on her throne with doctors as her slaves carrying her. Her daughter, Mary Lou Thompson, became her sidekick in helping blepharospasm sufferers early on and has carried the torch after her mother no longer could. She inherited the same drive and devotion to this cause and is to be commended even more as she doesn't have blepharospasm. I refer to Mary Lou as a blepharospasm wanna be. She even wears the FL41 lenses to be accepted by the group. Mary Smith has done an admirable job of running the BEBRF office for years and helping problem patients and never satisfied bosses.

Mattie Lou, Mary Lou and the BEBRF are responsible for most of the advances in BEB and facial dystonia today. The BEBRF has become a model for other support groups. Without help

from the BEBRF, botulinum toxin would never have been approved in 1989 by the FDA, myectomy surgery would not have been popularized, FL41 lenses wouldn't be utilized and it is unlikely that education, research and many other advances in the field would have occurred or continue to occur. Many patients would still be going undiagnosed and untreated. Just establishing the correct diagnosis and initiating treatment still brings tears to many blepharospasm patients. While a cure has not yet been found, when I first began treating blepharospasm in 1975 patients were brought in wheel chairs because they couldn't open their eyes. Some surgeons were advocating cutting holes in the eyelids so patients would have a peephole. While we are still not satisfied, great progress has been made in this disabling disorder.

Mattie Lou, Mary Lou and the BEBRF have bettered the lives of blepharospasm sufferers and their families worldwide in the last 3 decades. I look forward to seeing the advances by the BEBRF in the next 3 decades and I am very proud to be on the Board of this foundation. Happy 30th birthday BEBRF! 🎈

COMING SOON – BEBRF BIRTHDAY SURPRISE!!!

Please look for your own personal "birthday surprise", coming soon in the mail. A HAPPY BIRTHDAY TO BEBRF from Allergan will arrive shortly. Join together to celebrate our triumphs and progress over the past 30 years and keep the vision alive.



A Patient's Story

Martha Moore, Blakely, Georgia

I am nearly 70 years old, and do not know exactly when my eye problem started. The blinking became so bad that I made my first doctor's appointment in 2007. The doctor said it was rosacea on the eyelids. He worked with me for a year and the problem only got worse. Finally, he sent me to another doctor and he diagnosed it as benign essential blepharospasm. The doctor wanted to give me BOTOX injections, but I thought that was only for removing wrinkles so it took me several weeks to agree to have the BOTOX injections. My first treatment was May 2008 and it worked. I was so happy, even knowing that the treatment would only last for several months.

In October 2008 I had the next set of injections and that was a big mistake. My eyelids were paralyzed and would not open unless I held them up with my fingers. It was devastating. The doctor assured me that the BOTOX would all work out of my system in about 9 months – it took 12. The longer it took, the more depressed I became and my nerves at times were gone. Because everyone was concerned and meant well, they kept trying to tell me what to do. When I used my hands to hold my eyes open, people would ask if I had a headache- do you have a migraine? – do you need help? - do you need to sit down? People were concerned and meant well, but I had had enough. I also got questions and harsh looks and stares!

I could not read, sew, watch TV, or drive for fear of an accident. The wind and light totally shut my eyes down. Crowds and loud noises made me so nervous – I was a nervous wreck. YES! I was very stubborn and determined to beat this blepharospasm/apraxia

of eyelid opening one way or the other. I just wanted to be alone and cry.

August 2010, I had frontalis lid-sling surgery at the Callahan Eye Center and the most devastating thing was that I was blind for 4 days and had unclear eyes for several weeks. The results of the surgery did not make me recover 100%, but the doctor had gotten my eyes open. In January, 2011 the doctor gave me another round of BOTOX injections and discovered that I was developing Meige.

I can read large print, sew a little with a magnifying glass and I can even drive to the grocery store. My life has always been busy and I was always doing something for others, so that part of my life is back. This may have slowed me down, but it has given me time to look back over my life and realize how richly blessed I am. I am even catching up on writing my memoirs. I need to start walking again, inviting friends over for dinner now that I can see where I am going.

I would have never made it this far had it not been for my wonderful, loving and caring husband of 53 years. He is a great nurse and caregiver. My husband reads the Blepharospasm Newsletter to me and we enjoy reading the personal/patients stories. You could change the name in the stories to mine and the story would fit me. This jogs my memory about a father/son story in the Newsletter who thought it might be hereditary. Back in 1970 my daddy's eyes blinked so badly and the wind affected his eyes. My family and I are convinced now that Daddy had blepharospasm, but unlike me, we had no idea what his problem was and had no doctor to treat the problem. 🎈

Ask the Doctor

Disclaimer: Neither the BEBRF nor members of the BEBRF Medical Advisory Board has examined these patients and are not responsible for any treatment.



Q: I have noticed that I have the opposite problem of most blepharospasm patients who seem to do better when they are talking, singing or humming. When I am talking, my eyelids close and stay closed until I stop talking. Do you know the reason for this?

BOTOX® is the cause of tearing, the tearing usually improves as the BOTOX® wears off; i.e., the longer it is since the last BOTOX® injection.

Neil Miller, MD, Johns Hopkins, Baltimore, Maryland

A1: Most patients do note that their eyes stay open while talking. However, I have seen several patients with blepharospasm who have the opposite response as this patient does. It does not change the diagnosis or treatment.

Andrew R. Harrison, MD, University of Minnesota, Minneapolis, Minnesota

A2: There are many different stresses that produce or worsen blepharospasm. Many of my patients tell me that their blepharospasm is worse when they are talking to people. We don't know precisely why this happens.

Neil Miller, MD, Johns Hopkins, Baltimore, Maryland

Q: I would like to know why only one eye overwaters every time I have a BOTOX® treatment. It is very distressing and requires constant dabbing with a tissue to remove the excess water that stays in my eye. I have recently had surgery on my tear duct, since the problem was severe before, and I had Herrick plugs put in years ago when they thought I just had "dry eye", and the surgery was done to remove the plug which in fact was no longer in the duct. It was scar tissue which was clogging the duct and therefore I was over tearing, but more so after BOTOX®. Once I had the surgery, my eye stopped watering, and then I just had my injections, and I had the over tearing in that same eye again.

A: There are many, many causes of tearing, but the most common is dryness of the eyes. We have two types of tear glands. One should always be working, moistening the eyes so one can see well, but when that tear gland stops working or isn't working normally, a second tear gland kicks in. It is like a floodgate with only two positions: open and closed. Thus, if it senses that the eyes are dry, it opens up and the patient gets a flood of tears. Since BOTOX® reduces eyelid closure and blinking, many patients experience dryness of the eyes and secondary tearing due to this mechanism. To find out if this is the case, patients with tearing after BOTOX® injections in the eyelids should consider using artificial tear solution to both eyes 4-5 times a day. If this doesn't help at all, they should see their general ophthalmologist to see if there is another cause for their tearing. It should also be noted that if

Q: I have blepharospasm and mild lower facial spasms. I need extensive dental work done and my dentist wants to do dental implants into my jaw. Can this be safely done without the risk of spread of the dystonia? My other alternative is to have dentures.

A: It is true that dental work is often suspected as being related to the onset of dystonia in the lower face. I do not know whether dental work can worsen the situation once it has started. There may not be any good information on that issue. Either implants or dentures might have similar effect, if there is one, so you should make your decision based on what would be best for you in regard to your teeth.

Mark Hallett, MD, NINDS, NIH, Bethesda, Maryland

Q: I developed Restless Leg Syndrome after the onset of blepharospasm. My mother has a long history of RLS with out blepharospasm as does my daughter - but she has photophobia. My RLS is getting progressively worse. My doctor has to keep modifying my medication. I have read that RLS appears to be in the same basal ganglia area of the brain as blepharospasm. Have there been any studies done on blepharospasm patients with RLS?

A: Although blepharospasm and RLS are considered "movement disorders" and the basal ganglia have been implicated in the mechanism of both, there is no evidence that the two disorders are related. Both, blepharospasm and RLS, tend to occur in the same age group, 5-7th decade of life (about 10% of people over the age of 60 have RLS), and therefore it is not unusual for the two to co-exist in the same individual by chance alone. While RLS is highly genetic, as appears to be the case in this patient, blepharospasm is less frequently inherited.

Q: I was diagnosed with hemifacial spasm in 1997 and have been treated with BOTOX® injections since. The last injections I had around my eyes and my cheek and lip were not helpful at all. My upper lip became paralyzed and it is very difficult to eat or floss my teeth. It is slowly improving as the BOTOX® wears off but now my tongue has started to feel swollen and my neck muscle

Continued on next page

Continued from previous page

twitches along with my eye and facial muscles. I usually notice the feeling of the tongue swelling at the end of the day. My question is: If I get another series of BOTOX® shots around my eye only, could that possibly work as a trigger to stop the lower level facial twitches and the weird tongue feeling?

A: Without examining you it is difficult to make any specific comments or recommendations. Generally, it is a prudent practice to inject botulinum toxin for hemifacial spasm in a way that would control the involuntary twitching but not cause weakness or paralysis of muscles that are required for normal facial function and facial expression. I usually try to stay away from the lips (even if there is twitching around the mouth) in order not to distort the facial expression during smiling or talking. I also found that injecting around the eye not only controls the eyelid twitching but often also improves the involuntary movement in the middle and lower part of the face. So, don't give up on the injections, but do discuss these side effects with your physician so that the appropriate adjustments can be made to avoid or prevent similar adverse effects from subsequent injections.

Joseph Jankovic, MD., Director, Parkinson's Disease Center and Movement Disorders Clinic, Baylor College of Medicine, Houston, Texas

Q: I have been receiving Botox injections since 2002. Recently an article in the BEBRF Newsletter stated that zinc was a mineral that helped with the spasms. I tried zinc supplements without any improvement. Then I noticed I had some leftover PreserVision (Bausch & Lomb) tablets that my mother who has passed away used for macular degeneration. I started taking it but only the two pills in the morning and I've had a noticeable improvement with my spasms. I generally get the injections every 8 - 9 weeks and now it has been more than three months. I have an appointment soon and I know I will need the injections then because my eyes are starting to cause me grief. I think it is fantastic that I've made it this long. Maybe this information will be helpful to others that have tried zinc supplements with no reprieve.

A: If you have found a zinc supplement that works for you relative to your normal diet, you should stick with it. That specific preparation taken at that frequency relative to your usual meals and medications is apparently providing the right dose for you. It may not be right for others, but it is working for you. You recognized the very important point that not all zincs are equal. There are many reasons for this. First, zinc is a positively charged substance, and so it must always be associated with a negatively charged substance in order to be stable with an overall neutral charge. Different

suppliers couple their zinc to different negatively charged substances, and some are handled by us better than others, further facilitating zinc absorption. Second, the amount of zinc in different preparations will differ. So, as a random example, one preparation may offer zinc chloride at 100 mg and another zinc citrate at 25 mg, and the latter may actually be better absorbed, providing more zinc. Third, a preparation, such as a multivitamin, may contain many other positively charged minerals such as Calcium, Iron, Magnesium, and Copper, all of which may compete with the amount of Zinc being absorbed. Finally, if the Zinc preparation doesn't contain Phytase, an enzyme that breaks down Phytates, and if the preparation is taken at the same time as any of the foods we commonly eat containing Phytates, less Zinc will be absorbed. It appears as though you have experienced a 30+ % increase in the duration of your toxin's effect. On average, we are seeing a 29% increase using the zinc-phytase combination available by prescription, although some patients not included in our original study have doubled their duration of toxin effect. We now have one patient who takes half of the amount of zinc we initially prescribed (or she gets too much toxin effect even when we try to decrease her dose), and another who takes twice as much in order to get their maximum benefit. This is clearly an area of therapy still in development, but regularity and reliability of supplement purity will help each patient discover the right supplement dose and preparation for them.

Charles N.S. Soparkar, MD, Baylor College of Medicine, Houston, TX,



JOIN A RESEARCH STUDY

You are invited to participate in a research study on cranial dystonia (including blepharospasm and Meige). For more information, please go to the following web sites: <http://neuro.wustl.edu/patientcare/clinicalservices/movementdisorders/movementdisordersclinical/> and <http://rarediseasesnetwork.epi.usf.edu/dystonia/> Go to "join the study," and you will get all relevant information.

Mark's Ramblings

Mark Sheeler is the Coordinator of the West San Fernando Valley, California Support Group

- 1: Columbus' Mother: "I don't care what you discovered, you still could have written!"
- 2: Abraham Lincoln's Mother: "Again with the stovepipe hat? Can't you just wear a baseball cap like the other kids?"
- 3: Michelangelo's Mother: "Can't you paint on walls like other children? Do you have any idea how hard it is to get that stuff of the ceilings?"
- 4: I went to the local video rental and asked to take out "The Elephant Man". The proprietor said, "I don't think he's your type!" So I asked if I could borrow "Batman Forever?" He said, "No, you'll have to bring it back tomorrow!"
- 5: Marriage is an investment that pays you dividends if you pay interest.
- 6: Marathon runners with bad shoes suffer the agony of de feet!
- 7: I finally got my head together, now my body is falling apart.
- 8: Silence is not only golden, it's often misquoted.
- 9: I'm not saying my wife is a bad cook but she uses a smoke alarm as a timer.
- 10: Real happiness is when you fall in love with a girl and find out later she has money.
- 11: It's not hard to meet expenses, they're everywhere!
- 12: A closed mouth gathers no feet.

I'd better quit while I'm still behind!

Brain Banking

Continued from back cover

Unlike Parkinson's disease, Alzheimer's disease or Huntington's disease, in which specific brain regions degenerate and thus provide clues to where the underlying problem might be, in most forms of dystonia (blepharospasm is a focal dystonia) there is no known neurodegeneration. This is one of the reasons that brain donation is particularly important in dystonia because researchers do not have these obvious clues to guide them in where to look for the defect. Instead careful examination of dystonia brains will be required to identify consistent abnormalities related to the disease and understand how these contribute to their cause. However, several factors work against brain donations for dystonia including: 1. many patients are diagnosed with dystonia when they are young and obviously not thinking about death and brain donation, 2. because genetic forms of dystonia are associated with reduced penetrance, many people who carry dystonia mutations, never show clinical symptoms and so do not think their brains would be useful for donation and 3. unlike the neurodegenerative diseases named above, most dystonia patients die of natural causes rather than from their dystonia thus unless they are enrolled in a brain donation program, the opportunity to donate is often missed. Every brain donated can make a major contribution to our understanding of dystonia and is one of the most important gifts to research, future generations and our hope for better treatments for this disease. ●



More Brain Bank Info...

The following information is requested when an individual contacts the BEBRF about brain donation: Donor information: Name, address, telephone number, date of birth and neurologic diagnosis. Next-of-Kin information: Name, address, telephone number, relationship to donor. Email addresses are also requested if available.

The BEBRF will record an individual's decision to be listed as a brain donor with the Harvard Brain Tissue Resource Center and submit that registration information to them. In addition, the BEBRF will send each registered donor a wallet card along with contact information for the Harvard Brain Bank.

At the time of death, an individual's body becomes part of their estate and its' disposition is decided by the legal next-of-kin or other authorized representative. Although an individual can make a personal request to donate his/her brain, it is the surviving family member(s) or other authorized representative who has the responsibility of deciding whether this donation will be made. The legal next-of-kin or other authorized representative (such as the Executor of the donor's estate), are asked to verify the donor's intention-to-donate. A consent form which will be faxed or read by telephone will authorize the HBTRC to acquire, store, and share the brain and appropriate medical records of the donor with qualified scientists. ●

SUPPORT GROUP MEETINGS

TO GET YOUR SUPPORT GROUP MEETING IN THE NEXT ISSUE OF THE NEWSLETTER, PLEASE NOTIFY THE FOUNDATION OFFICE, BEFORE AUGUST 3, 2011 THE NEXT NEWSLETTER DEADLINE.

CO-AREA REPRESENTATIVES

Chicago, Illinois
Nancy Cummings
4623 N Chester Ave, Chicago, IL 60656
Tel. (773) 589-2797
Email: ngcummings@att.net

Tika Strehle
3441 W Bryn Mawr Ave
Chicago, IL 60659
Tel. (773) 588-6343

Shelly Goddard
996 Castlegate Ct.
Lake Forest, IL 60045
Tel. (847) 284-8634
Email: gowithgoddard@yahoo.com

NEW PHONE NUMBER

Iowa State Coordinator
Sam Lee
Tel. (712) 267-9068

SUPPORT GROUP MEETINGS

North

Milwaukee, Wisconsin
Saturday, September 17, 2011; 1:30 - 3:30 p.m.
Immanuel Lutheran Church — Meeting Room, 135th
& Hampton Road, Brookfield, WI
Contact: Marie Zehnder ... (262) 781-8364, Email:
zehmar@hotmail.com

South

Huntsville, Alabama
Blepharo-Buddies Awareness Support Group
Sunday, July 24, 2011; 1 - 4 p.m.
Dowdle Center, 109 Governors Dr., Huntsville, AL
Speaker: To be Announced.
Contact: Linda Webb... (256) 723-2661 Phone and Fax

Austin, Texas
Saturday, July 23, 2011; 10 a.m. - Noon
To be announced.

Speaker: Michelle Gray with OcuSoft.
Contact: Zoe Fallgren... (512) 268-7426, Email:
zls823@gmail.com

East

Silver Spring, Maryland Symposium
Saturday, July 16, 2011; 8:30 a.m. - 3:15 p.m.
Holy Cross Hospital, 1500 Forest Glen Rd,
Silver Spring, MD
Mark Hallett, M.D., Program Director
Register on-line at www.blepharospasm.org

West

San Jose, California
Saturday, November 5, 2011; 1-3 p.m.
Lincoln Glen Church, Fireside Room, 2700 Booksin
Ave., San Jose, CA
Contact: Kathy Berg... (408) 270-9787, Email:
kberg@pacbell.net

West San Fernando Valley, California
Sunday, October 23, 2011; 1:30 - 4:00 p.m.
Northridge Hospital and Medical Center, 18300
Roscoe, Blvd, Northridge, CA
(lower level auditorium)
Contact: Mark Sheeler... (818) 348-6127, Fax: (818)
348-7990, Email: smarkam@att.net

Albuquerque, New Mexico
Saturday, September 17, 2011; 1 p.m.
El Camino Medical Center, Pinon Rm, 1st Floor, 8100
Constitution PL NE, Albuquerque, NM
Contact: Al Deguio... (505) 298-6129, Email:
deguio@comcast.net

Seattle, Washington
Sunday, July 24, 2011; 2 p.m.
Swedish Medical Center, Cherry Hill Campus 500 17th
Ave, Seattle, WA. Contact: Peter Bakalor... (206) 219-
9053, Email: pdbbebhila@gmail.com

**IF YOU WOULD LIKE TO START A SUPPORT GROUP IN YOUR AREA -
CONTACT YOUR DISTRICT DIRECTOR OR THE BEBRF OFFICE FOR ASSISTANCE.**

Harvard Brain Tissue Resource Center

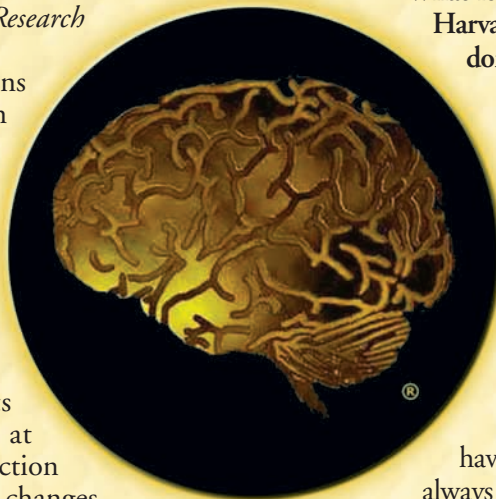
Brain Banking

Laurie Ozelius, PhD, Mt. Sinai School of Medicine, New York, NY

Editor's Note: Dr. Laurie Ozelius is the BEBRF representative to the Harvard Brain Tissue Research Center.

A brain bank collects and stores brains from deceased persons who suffered from neurological diseases and from individuals with no known neurological disease. The donated brains are treated in a standardized fashion whereby one half of the brain is used for neuropathological characterization and confirmation of diagnosis of the donor while the other half is frozen and stored for later distribution to scientists performing approved research aimed at understanding human-specific brain function and development as well as the unique changes underlying the cause of the disease. There are specific ethical guidelines for the collection of brains including consent by the donor before death or by authorization of relatives after death but only in accordance with the deceased's wishes.

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Why I Made the Decision to Send My Brain to Harvard!!!

An Interview with Peggy Raleigh, Rice Lake, Wisconsin

What led you to make the decision to register with the Harvard Brain Tissue Resource Center as a brain donor?

My blepharospasm has been very bad -an extremely severe problem. I've had this since 1984, and I have had every kind of treatment including BOTOX®, drugs and myectomy surgery, and still have problems. I thought that by signing up to donate my brain for research that I can help somebody else so they don't have to go through this.

Why is this important to you?

I want to find a cure so that other people don't have to go through the same thing I have. I am always hoping for something new that would help me.

What does your family think of your decision to be a brain donor?

I've discussed it with them and they think it's wonderful. They will follow through with my decision. ☺

Dated Material Enclosed

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RESEARCH FOUNDATION, INC.
P.O. BOX 12468
BEAUMONT, TEXAS 77726-2468
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It is our editorial policy to report on developments regarding BEB/Meige and related disorders but do not endorse any of the drugs or treatments in the Newsletter. We urge you to consult with your own physician about the procedures mentioned.