

THE CMTA Report

MAY/JUNE
2008

Information on Charcot-Marie-Tooth Disorders for patients, families, and the scientific community * www.charcot-marie-tooth.org



Charcot-Marie-Tooth
Association

OUR MISSION:

To generate the resources to find a cure, to create awareness, and to improve the quality of life for those affected by Charcot-Marie-Tooth.

OUR VISION:

A world without CMT.

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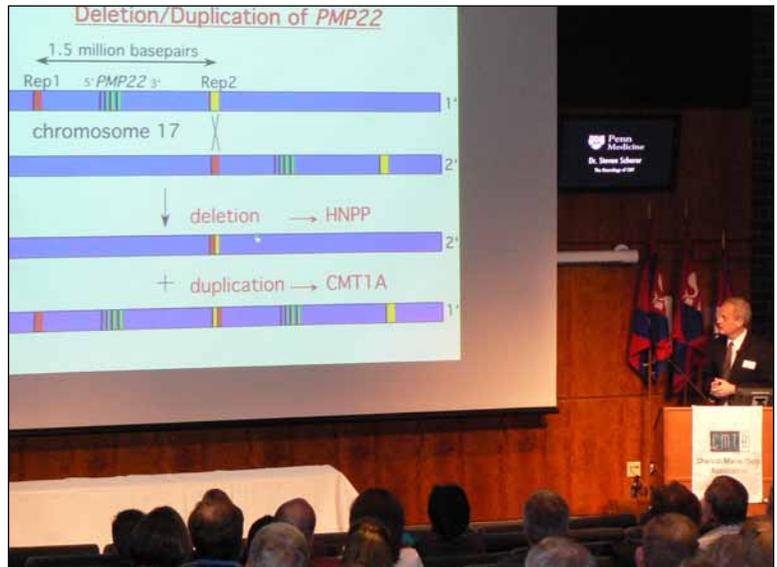
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Two Patient/Family Conferences Highlight April

On April 5, 2008, almost 200 patients, family members, and medical professionals gathered in the BRB auditorium at the University of Pennsylvania to hear presentations on the neurology of CMT, orthopedic corrections for deformities caused by CMT, physical therapy, and bracing options. The conference closed with a presentation by a representative from Canine Partners for Life.

Dr. Steven Scherer began the day with a discussion of the many different types of CMT. There are 33 known types, but there could be hundreds more rarer forms yet undiscovered, he said. Type 1A is the most common throughout the world and Type 1X is the second most common. The axons and nerve cells from the spinal cord to the feet are about 3 feet long, and that's why the axons are so vulnerable and neuropathy is so common. Dr. Scherer concluded his presentation by mentioning the new Strategy to Accelerate Research (STAR)TM initiative, which is looking first for compounds that will decrease the overexpression of PMP22 in



Dr. Steven Scherer discussed various types of CMT. Here, he shows the PMP22 duplication and deletion, both causes of peripheral neuropathy.

Type 1A patients. He said, "It is a sound and logical strategy, and I endorse it."

Many of the presenters answered patient questions, with a few highlighted here:

Q. Are statins safe for CMT patients to take?

A. There are no data to show that statins should not be taken by CMT patients.

Q. Is exercise safe for CMT patients?

A. You should do exercise, but not to the point of exhaustion.

Q. Why is vincristine considered such a dangerous drug?

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COMMEMORATE OUR 25th ANNIVERSARY WITH A COMMITMENT TO THE FUTURE

As the CMTA celebrates the past 25 years of our collective efforts to solve the mystery of CMT, we also look to the future with great anticipation. We believe that through collaborative research efforts such as the Strategy to Accelerate Research (STAR)[™], we are on the very threshold of developing treatments for this complicated disorder. And, even a cure within a decade.

The CMTA wants each of you to be part of our future success. To commemorate our 25th anniversary, we are offering a free year's membership between May 1, 2008 and April 30, 2009, whether it's a

renewal or a new enrollment. It's our way of saying "thank you" to those who have contributed to our past, and "please join us" to new members as we move toward the future.

Now, I'd like to encourage you to make a commitment. While the CMTA is offering a free year's membership, we have reported that the STAR initiative is going to require \$10 million of funding over the next five years to support its initial efforts. That doesn't include the requisite resources to fund the CMTA's continuing educational programs and outreach efforts. I am asking you to consider donating the

\$40 you save on the free membership to the CMTA. While this may seem like a relatively small amount of money, it's an invaluable gesture of your commitment to help us solve the puzzle of CMT. You can, of course, make a larger donation if you are able. Your support is the foundation of all of our efforts.

It's been said that the achievements of an organization are the result of the combined efforts of each individual. Please make your individual commitment. Together we can help end the suffering of millions with CMT.

—Charles F. Hagins
Executive Director, CMTA

TWO CONFERENCES

(continued from page 1)

As 5 to 10 Type 1A patients have had "near death" experiences when taking vincristine.

A second conference was held one week later in Detroit. The format was quite different, and the weekend began with a Friday night reception and tour of the actual working labs at Wayne State University. People were able to see where the CMT research is being done, and they were able to ask questions of the scientists. Then, on April 12, 2008, a group of about 70 patients gathered to hear typical presentations by CMT experts

in the morning and then to attend workshops in the afternoon on such diverse topics as bracing, genetics, and teaching kids about CMT. The afternoon sessions involved small groups and lots of hands-on experiences and time for personal questions.

One of the morning presentations was entitled, "The Psychological Aspects of Living with Chronic Disease" by Sandra Mulhinch. Sandra stated that a diagnosis of CMT is a traumatic experience which assaults one's self image, self esteem, and self worth. It causes concerns about the future because of the progressive loss of function, changes in appearance, and the consequences of hered-

ity and its affect on children. For children who are diagnosed, the concerns are different. They have to deal with CMT and a loss of independence just when they are trying to gain independence from their parents. It's extremely important that a patient, whether child or adult, have support from family and friends, medical staff, and their community (church, Scouts, school, etc.).

Each conference was sponsored by a member of the CMTA Medical Advisory Board: the Philadelphia conference by Dr. Steven Scherer, University of Pennsylvania, and the Detroit conference by Dr. Michael Shy, Wayne State University. ❄

Patricia Dreibelbis, Director of Program Services, Receives Honorary Award

BY ELIZABETH OUELLETTE

After 18 years of dedication and service, Patricia Dreibelbis was officially recognized by the CMTA Board of Directors at a recent meeting in Washington, DC. Pat, as most of us know, carries the distinguished title of Director of Program Services, but her job is in no way connected to a single role. She wears many hats, and her duties include speaking with CMT patients and answering their questions, organizing patient/family conferences, mailing CMT-related literature to the many individuals requesting information, putting together and editing the CMTA newsletter—the list goes on and on.

She initially began her tenure in 1990, as a part-time employee, hired by President Karol Hitt. Hitt wanted a writer to “fluff up” the newsletter, which contained much more scientific writing at the time. Also, working in a small office and listening to Karol answer patients’ questions, Pat gradually learned about CMT both from a patient’s and a medical point of view. For the first 6 years, Pat was the only paid employee of the CMTA. So, naturally, she ended up working on a little bit of everything, from data entry and mailing, to writing articles, and eventually adopting the position of editor of the CMTA newsletter when Karol left the organization.

As time went on and the organization expanded, Pat took on planning patient/family conferences, scheduling MAB meetings, attending scientific meetings, and writing thank you letters. She also became quite competent at answering patient inquiries about CMT due to her increasing knowledge about this disorder, its effects and causes. Finally, in 1996, a person was hired to do the bookkeeping and data entry, and Pat enjoyed the companionship and assistance of another full-fledged employee.

“Many positive changes have occurred over the years,” Pat reflects. She goes on to explain that, when she first appeared on the scene, “Our organization had about 3000 names in the database, compared to the 28,000 we have today. Grants for research have gone from \$5000 fellowships to \$300,000 grants, and, in the last few years, millions of dollars have been committed to CMT research. The CMTA has grown and made enormous strides in the realms of patient services, research endeavors, and overall recognition as a worthwhile non-profit organization.” Today, with a staff of 5 full-time employees and several volunteers, the Association has spread its wings from Pennsylvania to Chicago and California. As the CMTA expands to different parts of the country and gains

recognition through its new STAR initiative, more employees will surely be hired.

Having served three executive directors, Paul Flynn, Vince Bertolino, and Charles Hagins, Pat has become historian, advisor, and leader in the organization. I see her not only as a tenacious and committed individual who has chosen to devote much of her career to helping others, but also as a grounded and persevering woman who has become the structural backbone of the CMTA.

Pat was recently overheard saying, “I’ve never been as excited about anything the CMTA has done in the past as I am about the new STAR initiative. I really believe that the hope of a cure is more realistic than ever, and I honestly believe that our vision of a world without CMT is a possibility. This is a wonderful time in the history of the CMTA, and I’m proud to be part of it.”

We are very fortunate to have Pat, and she deserves heartfelt thanks from all of us. *

Pat Dreibelbis received a commemorative clock from the CMTA Board of Directors.



Two Recent Articles Address Sleep Apnea and Pulmonary Function in CMT

A recent article entitled, “Disorders of Pulmonary Function, Sleep, and Upper Airway in Charcot-Marie-Tooth Disease” was published in the journal Lung. The authors are Drs. Loufi Aboussouan, Richard Lewis, and Michael Shy. The abstract summarizes the findings in the following manner:

Charcot-Marie-Tooth disease (CMT) encompasses several inherited peripheral motor-sensory neuropathies and is one of the most common inherited neuromuscular diseases. Charcot-Marie-Tooth disease can be associated with several disorders that may be encountered by the pulmonary physician, including restrictive pulmonary impairment, sleep apnea, restless legs, and vocal cord dysfunction. Restrictive pulmonary impairment has been described in association with phrenic nerve dysfunction, diaphragm dysfunction, or thoracic cage abnormalities. Central sleep apnea may be associated with diaphragm dysfunction and hypercapnia (high concentration of carbon dioxide in the blood), whereas obstructive sleep apnea has been reported as possibly due to a pharyngeal neuropathy. Restless legs and periodic limb movement during sleep are found in a large proportion of patients with CMT2, a type of CMT associated with prominent axonal atrophy. Vocal cord dysfunction, possibly due to laryngeal nerve involvement, is found

in association with several CMT types and can often mimic asthma. There may be special therapeutic considerations for the treatment of those conditions in individuals with CMT. For instance, bilevel positive airway pressure may be more appropriate than continuous positive airway pressure (CPAP) for the treatment of sleep apnea in the individual with concomitant restrictive pulmonary impairment. The prominence of peripheral neuropathy as a cause of the restless leg syndrome in CMT may justify treatment with neuropathic medications as opposed to the more commonly

Patients with more severe neuropathy are more likely to develop sleep apnea.

recommended dopaminergic agents. The risk of progression to bilateral vocal cord dysfunction in CMT and the risk of aspiration with laryngeal neuropathy may limit the therapeutic options available for vocal cord paralysis.

The second article was an invited editorial by Michael E. Shy, MD from the Department of Neurology and the Center for Molecular Medicine and Genetics, Wayne State University School of Medicine, Detroit, Michigan. This article was entitled, “Obstructive Sleep Apnea and CMT1A: Answers and More Questions.”

The article suggests that pharyngeal nerves are affected in CMT1A, as the PMP22 duplication is expressed by all Schwann cells myelinating somatic peripheral nerves. However, not all patients develop obstructive sleep apnea-hypopnea syndrome (OSAHS) or have equally severe sleep apnea. The patients with more severe neuropathies are more likely to develop sleep apnea. This can be explained by the positive correlation between sleep apnea and median motor nerve amplitudes. These amplitudes are often used as markers for axonal loss just as nerve conduction studies can be used as a marker of demyelination. Clinical impairment in CMT1A correlates with reduced amplitudes, but not nerve conduction velocity. Thus OSAHS can be considered another example of CMT1A correlating with axonal loss rather than demyelination.

Whether CMT1A patients develop symptoms of daytime sleepiness is important in deciding how aggressively to treat a patient. One of the common complaints in CMT1A is chronic fatigue, out of proportion to obvious pulmonary disease or other obvious causes. Patients frequently complain of an overall loss of energy and sensation of tiredness. We (doctors) may tell patients that their fatigue results from the fact that they require more effort to walk because of ankle weakness or need of increased concentration

WE MADE IT!

Our Board Challenge raised \$365,715 for CMT research. Thanks to everyone who helped make an ambitious goal into an achievable one.



to prevent falling because of proprioceptive deficits. In fact, however, we do not know if these explanations are correct, and we don't really have a good explanation for fatigue in CMT. If fatigue is caused, even in part, by OSAHS and this becomes treatable, we would have important new therapies to offer patients.

Treatment response for sleep apnea is another unanswered question in CMT1A. Positive airway pressure (PAP) is the recommended therapy for most forms of OSAHS. The fixed positive pressure acts as a "physiological splint," forcing the airway to remain open throughout the respiratory cycle. In many patients it requires substantial continuous CPAP pressures of approximately 20 cm H₂O to overcome the obstructed passageway. When pressures rise above 12 to 14 cm H₂O, air leakage and discomfort from CPAP masks or devices increase, which limits the effectiveness of the treatment. Since pharyngeal nerve involvement in CMT1A is likely to cause OSAHS by denervation (axonal loss) rather than by mechanical or anatomical abnormalities, it is conceivable that lower levels of CPAP will be required. If so, CMT1A OSAHS may prove easier to treat than other forms of obstructive sleep apnea. However, there are no data yet on how well CMT1A patients respond to treatment. There are also no data on whether CMT1A patients have a high risk of heart disease or other long-term complications of OSAHS. Finally, there are no data on whether OSAHS is also common in other forms of CMT. *

CMT IN THE NEWS

From PE.com, the Press Enterprise

Abbey Umali of Redlands, CA, was invited to sing the National Anthem at the 6th Annual PossAbilities Triathlon on April 27, 2008. She has a type of Charcot-Marie-Tooth disorder which affects her balance and coordination and causes muscle weakness. She walks with the aid of leg braces and is an example of the PossAbilities message that disabled people can make adjustments and find their place in the community.

From the Daily Advocate, Greenville, OH

An article featured Louise Blickenstaff's collection and use of canes to help her maintain her balance and prevent falls. Louise has a mild form of CMT and has collected canes during her travels. One cane is from Costa Rica, another from Berlin, Ohio and one from Africa (although she hasn't actually been there.) Louise and her husband, Delbert, live in the Brethren Retirement Community and take long walks, sometimes as far as the Shawnee Prairie Nature Preserve. Unfortunately, Louise has taken several falls, which have precipitated the use of the canes. She has visited the Wayne State Clinic, is a member of the Darke County, OH, CMTA support group, and swims at the local YMCA pool several times a week.

From the Bellingham Herald, CA

The newspaper reported on Sherri Backstrom, 46, a single mother of three who grew up around boats and the water, and has now started a company, Waypoint Charter Services, through which she brokers yachts for individuals and groups. A special focus of the company is chartering boats with accessibility for people with disabilities. When she was in her 20s, Sherri was diagnosed with Charcot-Marie-Tooth disorder and her disability makes her enjoy helping other people experience what they might have thought they otherwise couldn't.

Backstrom spent five months on the Sea of Cortez in 2006, with her fiancé, Don Hall, a semi-retired commercial fisherman. They intended to leave and go to Central America, but they never left the sea, exploring the water and the islands. That trip encouraged her to start her business to help others experience the water in the same way she has been able to.

From the Capabilities Newsletter

An article written by Board member Elizabeth Ouellette discussed CMT and its effect on her fifteen-year-old son Yohan. She described the physical symptoms and problems that the disorder can cause for someone trying to keep up with his peers. Yohan wears leg braces and suffers from scoliosis and kyphosis related to his CMT. *

Traveling in London

BY ALAN PAPPALARDO

Recently, I traveled to London to visit a friend. Typically, having CMT and being in a wheelchair make international travel exceedingly difficult. However, London proved to be among the most accessible cities I have ever visited.

Although London was surprisingly accessible, there are a few things disabled tourists should know before they pack their bags. London has an extensive public transportation network, both above and below ground, and it takes some practice to manage the various options.

London's buses are free for those with disabilities, and all of them are equipped with wheel-

chair ramps located outside the center door of the bus. There is a blue button on the outside wall that has the handicap symbol on it. You are supposed to press this button in order to alert the driver to release the ramp; however, in practice it works better to alert the driver

personally. The ramp takes about one minute to cycle, and the doors shut before the ramp extends. Once the ramp extends, only then do the doors open.

Once you are on the bus, it is also prudent to notify the driver personally which stop you want to get off at. Even though there is another blue handicap button on the inside of the bus to press for stops, in my experience the driver never responds to this button, and personal contact is necessary. Most drivers are very friendly and helpful, but I did have some buses pass me by during rush hour because the bus was too full to accommodate a wheelchair.

London's subway system, known as the Underground or "the tube," is slightly more difficult to manage. The Underground was built in the 1860s, resulting in various levels of accessibility. Some of the lines are completely accessible; with their train cars flush with the surrounding dock so that wheelchairs can smoothly enter the train. Before using the Underground, it is wise to consult their website to make sure that your stop and train are accessible.

A wonderful surprise was that London's taxis are all equipped with ramps. For travelers with wheelchairs, the taxi driver has to get out of the car, open the door, and manually pull the ramp out. This process takes about a minute. Every driver I came in contact with was very courteous and willing to help me out.

Of course, one does not visit London simply to experience their transportation options. London boasts many of the world's finest museums, nightclubs, restaurants, and pubs. Many museums are discounted or free for disabled visitors. Most of the exhibits were accessible by elevator or wheelchair lift, but since many of the buildings are very old, some of the exhibits were impossible to get to or require a non-traditional route. For example, Windsor Castle's only elevator was off the normal tourist path (and is the same the Queen has to use).

Many of London's restaurants and pubs are built inside of very antiquated buildings, and so they are simply not accessible. While some pubs may seem accessible, the bathrooms are often located in the basement, so be sure to ask where the bathroom is before committing to a certain nightspot. Also, some older men's bathrooms require users to stand on a step to use the urinals, which can be impossible for CMT patients.

No matter where you go, traveling with CMT is never easy. Having an able-bodied companion is invaluable and greatly increases the quality of the trip. However, London's officials have done a fantastic job of combining modern accessible features with the city's medieval heritage. I fully recommend visiting London, as long as one does the prior research. ✨

A London bus with its wheelchair ramp extended.



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Your gift to the CMTA can honor a living person or the memory of a friend or loved one. Acknowledgment cards will be mailed by the CMTA on your behalf. Donations are listed in the newsletter and are a wonderful way to keep someone's memory alive or to commemorate happy occasions like birthdays and anniversaries. They also make thoughtful thank you gifts. You can participate in the memorial and honorary gift program of the CMTA by completing the form below and faxing it with your credit card number and signature or mailing it with your check to: CMTA, 2700 Chestnut Parkway, Chester, PA 19013.

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In honor of (person you wish to honor)

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Send acknowledgment to:

Name: _____

Address: _____

Amount Enclosed: _____

Check Enclosed VISA MasterCard

Card # _____

Exp. Date _____

Signature _____

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Name: _____

Address: _____

CMTA Support Groups: Still Looking for Leaders!

BY ELIZABETH OUELLETTE

Thank you Susan in Atlanta, Georgia, Debbie in Portland, Oregon, Drusilla in Anaheim, California, and Amanda in central New Jersey! Since my last request for new support groups, these women have all initiated groups in their areas. Thanks for your commitment to CMT patients, their families, and friends.

Despite these new groups, there are still many populous cities and states without resources for those who truly need them. As you know, the CMTA is growing and expanding in the realm of membership, research, public awareness, and fundraising efforts. Our offices receive many calls and e-mail requests each week for information on local support groups in virtually every state in the U.S. The demand is much greater than the supply, so once again, I am personally reaching out to each and every one of you to consider facilitating a CMT support group in your area.

At first, starting a support group may seem to be an overwhelming task. I decided to give it a whirl, assured that the benefits would greatly outnumber the disadvantages, and they do.

What does the CMTA require of those interested in starting a CMT group? You do not have to be an expert in the field of CMT, nor do you have to be trained in leading groups. The most essential requirements

are motivation, a certain time commitment, people skills, and sincere dedication to the group and its members. Remember, you will not be all alone in taking on this endeavor; the CMTA is here to help out. The CMTA is now taking a more active role in reaching out to our groups, assisting the leaders to get off to a good start, and maintaining success over time.

Once you decide to start a group, contact the CMTA, and I will walk you through the organizational steps and answer any questions you have about a potential group. The Association will help publicize your first meeting through e-mail or a written letter and will announce all the details on our web site.

A CMTA support group provides a valuable service by giving support, coping skills, and education as well as socialization and information. When individuals who share a common experience meet face to face, a world of reciprocal understanding and camaraderie is created. A group of this nature allows for the exchange of insightful experiences and life stories not to be found elsewhere. In addition, these meetings pave the way to personal empowerment by providing a structured forum where individuals can help and be helped by others.

Secondly, local professionals, familiar with CMT, are willing to donate their time and knowl-

edge to educate members about every relevant topic of interest to the group's participants. In turn, the participants, the real experts on CMT, are also given the opportunity to enlighten the presenter about the distinctive traits of CMT.

At the time of my first meeting, 2 years ago, I had only met two people with CMT and my son Yohan had never met a single soul with the disorder. Now, in my second year as a support group leader, I remain fiercely committed to the individuals who comprise my group.

Most of our group leaders across the U.S. have been at this task far longer than I, and along the way, I have gained great respect for these people who forge ahead, personally making these groups happen. Groups make a difference by serving individuals, the community, and society at large.

After reading this article, my wish is that each of you or someone you know will pick up the phone with the intent of putting a CMT group together in your area. The rewards, both personal and social, are immeasurable. Each one of us can make a difference, so let's strengthen our roots and multiply. As Margaret Mead once said, "Never doubt that a small group of thoughtful, committed citizens can change the world. Indeed, it is the only thing that ever has." *

A Wayne State Orthotist and A Physiatrist Answer Questions about Their Specialties

Sean McKale, an orthotist working with people who are seen through the Wayne State Clinic, answered the following questions:

What role does an orthotist play in the treatment of CMT?

An Orthotist is the health care professional responsible for filling the Orthotic(brace) prescription written by the patient's physician. Ultimately, the fit and function of the individuals orthoses are the responsibility of the orthotist. We recommend that patients visit www.abcop.org to find an orthotist local to them. Orthotist found on this website have earned a certificate through a university program, completed a residency, and passed a board exam in or to acquire national certification, while some orthotists may have gained these skills only through practical experience. Skill level and decision making can vary greatly. In general Orthotists are creative problem solvers who work well with their hands to achieve successful outcomes. CMT patients are particularly challenging for orthotists as they often have significant alignment changes and mechanics can be more difficult to control. Many CMT patients also require use of orthotics on both legs, which in itself, can make achieving "normal" mechanics more difficult.

What type of devices do you make for people with CMT?

A variety of lower extremity orthosis are fabricated depending on a patient's individual needs and goals. Improving the patient's bio-mechanics to prevent falling/tripping, increasing stability(balance), and preventing misalignment of other joints (eg. knee, back), while avoiding "over bracing" is consistently the goal. We do find that many CMT patients do better in orthotics fabricated from carbon fiber as it helps to control motion in all planes.

How do you decide what each individual needs?

An orthotic prescription is written based on the patient's deficits; ie what strengths are present. Design must be considered if there are alignment issues or fluctuating swelling, and the patient's goals also need to be addressed. Often patients are concerned with cosmesis (appearance) or they may want to participate in certain activities, for example, golf. It's also important to understand the patient's history and what has and has not worked in the past, and how the patient currently feeling limited.

Will insurance cover the cost of braces?

In many cases insurances will cover the majority of orthotics. Individual co-pays and deductibles may apply. How-

ever in some states it is now legal for insurances to cut Orthotic, Prosthetics and Home Medical Equipment coverage, as a cost savings to the company they are providing insurance coverages to. It is important to pay attention to such "cost savings" when choosing your medical coverage. Medicare in general will cover 80% of the cost. If you have an HMO you most likely have to go through a particular provider.

Dr. Steven Hinderer, a physiatrist from the Wayne State Clinic, answered these questions:

What is a physiatrist (fizz ee a ' trist)?

Physiatrists are licensed physicians (MD or DO) who specialize in the board-certified discipline of physical medicine & rehabilitation (PM&R). PM&R is the branch of medicine emphasizing prevention, diagnosis, and treatment of disorders causing physical and cognitive impairments, including disorders involving the nervous, musculoskeletal, cardiovascular, and pulmonary body systems. The target of treatments prescribed by a physiatrist are to restore normal function when possible, maximize function when full recovery is not possible, and help the people to participate in life roles at home, school, work, and play. We are a "quality of life" specialty.

What types of conditions does a psychiatrist treat?

Impairments and disabilities resulting from:

- Neuromuscular diseases such as neuropathies, multiple sclerosis, muscular dystrophies, ALS, CMT
- Neurotrauma such as spinal cord injury, traumatic brain injury, peripheral nerve injuries
- Stroke
- Cancer
- Amputation
- Burns
- Birth defects (eg, cerebral palsy, spina bifida)
- Connective tissue disorders (eg, rheumatoid arthritis, lupus, scleroderma, dermatomyositis)
- Musculoskeletal problems (eg, neck, back, shoulders, elbows, etc.), including repetitive stress, and work- and sports-related injuries)

Do you perform surgery and/or prescribe medicine?

Psychiatrists do not perform surgery, but we do inject painful joints and trigger points, perform nerve blocks for spasticity with Botox and phenol, and debride wounds and pressure ulcers. Psychiatrists prescribe most medications that other physicians commonly prescribe as well as some specific medications indicated for selective conditions such as traumatic brain and spinal cord injuries.

Is pain normally a part of CMT and how do you treat it?

Pain is a frequent problem for people with CMT. Most often the pain is not directly due to the neuropathy. Instead it results

from other problems caused by the neuropathy, such as the foot and ankle changes affecting joint function and muscle fatigue because of the increased physical effort required to be mobile as the neuropathy from CMT progresses over time. I treat the underlying cause for the pain first. Most commonly I prescribe some type of lower limb orthotic that corrects the alignment of the ankle and foot, and if needed, supports or substitutes for the decreased muscle function and loss of position sensation caused by the neuropathy. Sometimes I prescribe physical therapy to further help restore strength, muscle extensibility, and mobility as well as decrease pain. Occupational therapy can be

helpful when hand function has been affected, using adaptive devices to decrease demand upon weakened hand muscles to prevent cramping and pain.

What difference can you make in the life of a person with CMT?

The evaluation and treatment methods I utilize can decrease or eliminate pain, improve the mechanics and endurance for mobility in multiple settings (home, work, recreation), improve performance of activities of daily living, and prevent joint deformities and degenerative arthritis over time. In summary, I can help to improve and maintain the quality of life for people with CMT. *

VASO-WRAPS

Vaso-Wraps are new noninvasive medical devices that quickly and easily wrap around the leg with Velcro, providing warmth and protection without the use of heating elements.

There are no electrical wires or heating elements that could cause burns. Vaso-Wraps are made of thick soft, custom-quilted nylon. They can be applied or removed easily due to the Velcro closing.

One of their uses is the prevention of night cramps. The Vaso-Wrap keeps the leg warm, which relaxes the calf muscles. This reduces the likelihood of cramping.

For more information about Vaso-Wraps, visit their website at www.vasowraps.com or call 860-416-0590. Consult your insurance provider to see if Vaso-Wraps are covered by your policy or medical spending account.

Editor's note: We were sent a pair, and a CMT patient, a diabetes patient, and one "normal" worker tried them out. We all loved them. It's pretty amazing how they almost immediately produce warmth and we can all imagine using them in colder weather or when our legs are tired and achy. Needless to say, we don't endorse this product, but we do believe it might be useful for some CMT patients.





SUPPORT GROUP NEWS

California – San Francisco

The group met on March 29, 2008, and had a great turnout. A new member, Margot, who is 96, came from Foster City to be part of the meeting. The main topic of discussion was the free membership from the CMTA in honor of the association's 25th anniversary. Another exciting topic was the possibility of a patient/family conference being planned for the San Francisco area in the fall. Finally, Gilles Bouchard talked about STAR, the strategy to accelerate research and the future of the CMTA's research endeavors. The next meeting will be May 17th and Syed Rahman, an orthotist, will discuss bracing options, for CMT patients.

Colorado – Denver Area

The February 23rd meeting was extremely informative. Kathryn Arbour, President of Capabilities demonstrated products that are solutions to many dilemmas that CMT can present. Capabilities' website, www.capabilities.com, sells aids for daily living.

On Saturday, April 26th, Elizabeth Ouellette, a CMTA Board Member, was the featured speaker. Her topic was "Pain and Pain Management." Elizabeth also outlined the CMTA's ongoing research project, STAR.

Georgia – Atlanta Area*

The support group for the Metro-Atlanta area met for the first time April 19th. Twenty-nine people gathered to discuss CMT and its effects on the patient. Many questions were

asked in regard to CMT, and the top areas of interest were identified to create the meeting schedule. There are plans to bring local CMT doctors, physical therapists, occupational therapists, counselors, appliance representatives, instructors of yoga and pilates, and nutritionists to the upcoming meetings. Our next meeting is June 21, 2008 at 2:00 pm. at Cliff Valley School on Clairmont Road in Atlanta.

Illinois – Chicago Area

The next meeting will be held on June 21, 2008, at 2:00 pm. The meeting will be held at the Hinsdale Hospital in Hinsdale, IL and will feature a presentation from Ballert Orthopedics. Their topic will be the various types of orthoses useful to CMT patients.

Nevada – Las Vegas Area

The group met on May 17 in the West Charleston Library to hear Dr. A. Scott Martin discuss pain management.

New York – New York City

The group enjoyed a presentation on bracing entitled, "Functional Leg Bracing for CMT Patients" by Mitchell Warner, CPO, on Saturday, May 10, 2008. Mitch is the creator of the Helio orthotic system and demonstrated its usefulness for CMT patients.

New York – Westchester County/Connecticut

The meeting on March 15th featured Barbara Bischoff, Director of Health and Safety, from

the American Red Cross. She demonstrated the use of the Heimlich maneuver for choking victims. The next meeting will be May 17, 2008, and will feature a presentation by Pat Dreibelbis and Dana Schwertfeger from the CMTA office. They will talk about the exciting plans that the organization has for advancing research.

Oregon – Portland Area*

The first meeting of this new support group will be June 12th from 7:00 to 8:30 pm. The location will be 1008 N.E. Division, Suite B, Gresham, OR 97080. The new group leader is Debbie Hagen. She can be reached by email at hagen84@yahoo.com or can be called at 503-886-2538.

Virginia – Harrisonburg

The March 3rd issue of the Shenandoah Journal featured an article about Jeanette Thompson and Ann Long starting a support group for people in the Shenandoah Valley who have CMT. Seventeen people attended their first meeting. The group meets every other month on the second Saturday in the Sunnyside Retirement Community near Penn Laird.

Washington – Seattle

The group met on April 26, 2008 at the University of Washington Medical Center. The speaker was Susan Ewers, MS, CPO, from the Department of Rehabilitation at the University of Washington. *

* New groups

**NEW GROUP
COMING SOON:
Anaheim, CA**

CMT Support Groups

Support Group Liaison: Elizabeth Ouellette, 1-800-606-2682, ext 107

California—Northern Coast Counties (Marin, Mendocino, Solano, Sonoma)

Place: Sutter Medical Center of Santa Rosa
Meeting: Quarterly, Saturday, 1 PM
Contact: Louise Givens, 707-539-2163
Email: ladyblue123@att.net

California—San Francisco Bay Area/Santa Clara County

Place: San Mateo Library
Meeting: Quarterly
Contact: Elizabeth Ouellette, 650-248-3409 (C)
650-559-0123 (H)
Email: elizabetho@pacbell.net

Colorado—Westminster

Place: Capabilities, Westminster, CO
Meeting: 10 AM – noon, Last Saturday of every other month
Contact: Diane Covington, 303-635-0229
Email: dmcovington@msn.com

Florida—Tampa Bay Area

Place: St. Anthony's Hospital, St. Petersburg, FL
Meeting: Second Saturday of Feb, May, Nov, with other special meetings throughout the year
Contact: Lori Rath, 727-784-7455
Vicki Pollyea
Email: rathhouse1@verizon.net
v_pollyea@mindspring.com

Georgia—Atlanta Area

Place: Cliff Valley School Library, 2426 Clairmont Rd, NE
Meeting: Third Saturday of every other month
Contact: Sue Ruediger, 678-595-2817
Email: susruediger@comcast.net

Illinois—Chicago Area

Place: Peace Lutheran Church, Lombard, IL
Meeting: Quarterly
Contact: Alan Pappalardo, 800-606-2682, ext. 106
Email: alan@charcot-marie-tooth.org

Kentucky/Southern Indiana/ Southern Ohio

Place: Lexington Public Library, Beaumont Branch
Meeting: Quarterly
Contact: Martha Hall, 502-695-3338
Email: marteye@mis.net

Michigan—Detroit Area

Place: University Health Center, Wayne State U., School of Medicine
Meeting: Call for schedule
Contact: Lainie Phillips, 248-890-1529
Email: familiaphillips@sbcglobal.net

Minnesota—Benson

Place: St. Mark's Lutheran Church
Meeting: Occasionally
Contact: Rosemary Mills, 320-567-2156
Email: rrmills@fedtel.net

Minnesota—Twin Cities

Place: Call for location
Meeting: Quarterly
Contact: Bill Miller, 763-560-6654
Email: wmill758@msn.com

Mississippi/Louisiana

Place: Baptist Healthplex, 102 Clinton Parkway, Clinton, MS
Meeting: Quarterly
Contact: Flora Jones, 601-825-2258
Email: flojo4@aol.com

Missouri—St. Louis Area

Place: Saint Louis University Hospital
Meeting: Quarterly
Contact: Carole Haislip, 314-644-1664
Email: carole.haislip@sbcglobal.net

Nevada—Las Vegas

Place: West Charleston Library, 6301 West Charleston Blvd.
Meeting: Email for dates 1-3 PM
Contact: Mary Fatzinger
Email: cmt_suppgrp_lvnv@yahoo.com

New York—Greater New York

Place: NYU Medical Center/Rusk Institute, 400 E. 34th St.
Meeting: Second Saturday, 12:30-2:30 PM
Contact: Dr. David Younger, 212-535-4314
Fax 212-535-6392
Website: www.cmtnyc.org
Email: bwine@acm.org

New York—Horseheads

Place: Horseheads Free Library on Main Street, Horseheads, NY
Meeting: Quarterly
Contact: Angela Piersimoni, 607-562-8823

New York (Westchester County)/ Connecticut (Fairfield)

Place: Blythedale Children's Hospital
Meeting: Bimonthly, Jan, March, May, Sept, and Nov; 3rd Saturday
Contacts: Beverly Wurzel, 201-224-5795
Eileen Spell, 732-245-0771
Email: craneomatic@verizon.net
espell@optonline.net

North Carolina—Triangle Area (Raleigh, Durham, Chapel Hill)

Place: Raleigh, NC
Meeting: Quarterly
Contact: Susan Salzberg, 919-967-3118 (afternoons)
Betsy Kimrey
Email: judae@bellsouth.net

Ohio—Greenville

Place: Brethren Retirement Community
Meeting: 4th Thurs. of April, July and October
Contact: Dot Cain, 937-548-3963
Email: Greenville-Ohio-CMT@woh.rr.com

Oregon—Portland Area

Place: 1008 NE Division, Suite B, Gresham, OR
Meeting: Quarterly
Contact: Debbie Hagen
Email: hagen84@yahoo.com

Pennsylvania—Johnstown Area

Place: John P. Murtha Neuroscience Center
Meeting: Bimonthly
Contacts: J. D. Griffith, 814-539-2341
Jeana Sweeney, 814-262-8467
Email: jdgriffith@atlanticbb.net,
cjsweeney@ussco.net

Pennsylvania—Northwestern Area

Place: Blasco Memorial Library
Meeting: Call for information
Contact: Joyce Steinkamp, 814-833-8495
Email: joyceanns@roadrunner.com

Pennsylvania—Philadelphia Area

Place: CMTA Office, 2700 Chestnut St., Chester, PA
Meeting: Bi-monthly
Contact: Pat Dreibelbis or Dana Schwertfeger, 800-606-2682
Email: info@charcot-marie-tooth.org

Virginia—Harrisonburg

Place: Sunnyside Retirement Community, Sunnyside Room
Meeting: Bi-monthly, Second Sat. 1-3
Contact: Anne Long, 540-568-8328

Washington—Seattle

Place: U of Washington Medical Center, Plaza Café—Conference Room C
Meeting: Monthly, Last Saturday, 1-3 PM
Contact: Ruth Oskolkoff, 206-598-6300
Email: rosk@u.washington.edu

A Return Visit to the CMT Clinic at Wayne State University

BY DANA SCHWERTFEGER WITH ELIZABETH OUELLETTE

Last February, Elizabeth, her son Yohan, and I spent two days at the WSU CMT Clinic in Detroit, MI. We wrote about our experiences for the newsletter, including our plans for a return trip this year. What we didn't know then was that our return trip would coincide with the CMT Patient Fair in April, so that turned out to be an added bonus. You might be wondering why we planned a return trip only one year later, especially since so many of you have been told that there is little that can be done to alter the progression of the disorder.

On our first trip we had missed seeing Dr. Steven Hinderer, a physiatrist, and we both wanted to get his opinion regarding what we needed to be doing in terms of maintenance and rehabilitation. While I, at 51, had noticed only a slight change in my condition, Yohan, who is now 15, had grown 7 inches since our last visit, and he was looking for answers to some CMT-related issues like his increased neuropathic pain, sleep-apnea, and poor posture. With Yohan's rapid growth, his scoliosis and kyphosis (rounding of the shoulders) had become worse, and his current treatment plan wasn't working.

In addition to addressing those issues and getting

Yohan new orthotics and AFOs, we both would have a chance to see how if and how our CMT had progressed since our previous visit.

Measuring that progression seemed more important for Yohan than for me. He was growing like a weed, and, except for a slight loss of function in my hands, I hadn't noticed any real change. Unless there was something that could be done about it, determining the extent of my functional loss might seem pointless—except that our test results were also being entered into the CMT North American Database.

When the results were in, my CMT Neuropathy Score (CMTNS) had gone from 20 to 23 on a scale of 1 to 36, with 36 being the worst. As Dr. Shy explained, the reason for the change was a loss of sensation in my lower legs, which he said would affect my proprioception and balance.

It was good to note that my degree of change did not result in additional impairment, but Elizabeth has a different take on the repeated testing and follow-up visit because Yohan is a teen, who continues to develop and mature. Elizabeth writes,

“As a parent of a child with CMT, I was relieved to discover that Yohan's results on the CMTNS had increased by only 2 points, probably due to increased neuropathic pain in his

toes and some loss of sensation. He actually tested better in the strength-related areas, a testimony to all the hard work he and his personal trainer have accomplished over the past 6 months. So, in light of the other physical symptoms he is currently experiencing, having solid evidence that his CMT neuropathy remained in the mild range was comforting news.

“I also learned that Yohan's moderate sleep apnea is a direct result of his CMT. He has been using a CPAP machine at night, but so far, there have been more problems than solutions. We are still working out the kinks, such as finding a face mask that does not cause sores, rashes, and painful redness. Meanwhile, Yohan's daytime fatigue is so severe that until he can tolerate and wear the CPAP machine, we will try the medication Provigil (modafinil), a prescription medication used to improve wakefulness in those who experience excessive sleepiness due to diagnosed sleep disorders.

“During our afternoon visit with Dr. Hinderer, we had a lot of questions about Yohan's feet, legs, and back. Both Dr. Shy and Dr. Hinderer agreed that Yohan needed to be wearing AFOs for stretching and for mild drop foot, and they assured him that with new AFOs his gait would be much better.

“Yohan was thrilled to be upgraded to an AFO made

Sean McKale applies a gel to improve the electrical signals in a new brace for scoliosis.



from a composite material, because the carbon-fiber brace should prove to be much lighter, less cumbersome, and easier to wear than the plastic braces he has today. Sean McKale, an orthotist at Wayne State, made him new orthotics, a yearly ordeal for rapidly growing kids.

“Lastly, in California, Yohan had been fitted for a rigid plastic back brace to arrest the progression of his scoliosis and kyphosis. He wore that rigid brace for all of 2 hours before we called it quits. Not only was it uncomfortable, but when he sat down, it cut into his neck, making him choke and slouch even more!

I took the braces to Dr. Hinderer and Sean McKale for review and examination.

“They both told me it was not made correctly for Yohan’s physique, and then, just when we were feeling a bit discouraged about the possibility of treating his scoliosis, Sean mentioned the Bio-vest, a wearable therapeutic neuromuscular stimulation system (www.wearabletherapy.com). The Bio-vest includes battery-powered electrodes, strategically placed in a form-fitting sleeveless shirt, where they gradually strengthen specific back muscles. The vest, which is also currently used for pain management, will be sent to us in the very near future, and we hope that with continued wear, it will significantly correct Yohan’s posture.”

Yohan always gets the cool stuff, but my visit with Dr. Hinderer was also interesting. He said I appeared to be doing very well, but that I might benefit

from a different style of orthoses. A year ago, Sean had proposed Blue-Rocker Toe-Offs (which have a lateral shank), but my local orthotist said the Toe-Offs wouldn’t correct my ankle instability and made me a pair of AFOs from “pre-preg,” a composite material, and a pair of molded plastic AFOs (which I happened to be wearing). Both were posterior leaf-spring designs with the shank to the rear of the foot.

This year, Dr. Hinderer, Sean, and I discussed the relative merits of bracing designs, and Sean demonstrated a composite design with a medial shank,

Research was transformed from an abstract concept to something happening before our eyes.

meaning the support runs up the inside of the leg. I’m willing to give new designs a try, and Sean used me as a casting model during one of the workshops at the Patient Fair. For the time being, however, those casts are sitting on the shelf because my insurance won’t pay an out-of-network provider. The MDA will pay \$2,000 of the \$2,900 cost, but that leaves me with a \$900 bill. If I had been able to actually try the orthoses, I might be willing to pay out-of-pocket for them, but I’m not prepared to invest that kind of money without assurance that the product will perform better than what I have now.

I may be skeptical about the performance of the braces, but I believe our trip to Detroit was educational, inspirational, and

productive. Elizabeth and Yohan are excited about the potential of this Bio-vest and cannot wait to receive it and report on its effects! Yohan’s AFOs will require another trip to Detroit, but he and Elizabeth feel it is worth the time and energy, so that they are not continually purchasing useless products from well-intentioned physicians and orthotists, who do not know the ins and outs of CMT in its many forms.

I am always impressed by the knowledge and professionalism of the team at Wayne State, but the high point of the trip for me occurred not at the clinic

but at the Patient Fair reception on Friday evening, when I had a chance to tour the research labs in the Elliman building.

We were escorted in small groups, and while our group was waiting, I spoke with a couple who had traveled from Vancouver in search of answers. They didn’t know a great deal about CMT, but I was able to give them a 10-minute synopsis before we entered the labs.

I always have a million questions of my own, so viewing the presentations and being able to ask the researchers questions was an added bonus, but what really impressed me was the reaction of the Vancouver couple and the other guests. I could literally see them nodding their heads in understanding as they listened, and suddenly “research” was transformed from an abstract concept to something happening before their eyes. I wish everyone could have that experience! *

The Joys of Travel?

BY PAT DREIBELBIS

Getting away is fun...right? That was my original thought process when I began making arrangements for Dana Schwertfeger and myself to fly to Detroit, Michigan, to visit the Wayne State Clinic, tour the research facilities, and attend a patient/family conference.

So, with optimism in hand, I called Northwest Airlines, the major carrier in Detroit, to get round-trip tickets. The problems began when I discovered that there's no way to request special seating on the Internet site. Dana has CMT, wears leg braces, and is tall—he needs the extra space of bulkhead seating. When I called the 800 number for Northwest I was connected to a “customer service representative” (I'll leave it to you readers to decide if that's an appropriate job title in this instance). I explained my situation and the

need to book special seating.

Her first question to me set the mood for the entire exchange. She said: “Can he bend his knees?” I replied that he could and she then said “Northwest doesn't consider a person disabled unless they cannot bend their knees.” I was so surprised/upset/amazed/irritated (choose

Our tickets were printed out, and we moved through security with only the vague hassle of Dana having to be hand inspected because of his braces. When we got to our gate, I took the time to look at our boarding passes and noticed that we were now seated in row 13 right next to each other. Oh, no! He didn't

“Can you bend your knees?
Then you aren't disabled!”

whatever word seems appropriate) that I hardly knew how to respond. What actually came out of my mouth was something sort of sarcastic, which was, “I'll bet the ADA representatives will be interested to hear that!” The magic words “Americans with Disabilities” seemed to trigger some fear in her, because she immediately changed her approach. She still seemed to think that having braces on his legs did not constitute a disability, but she agreed to look for bulkhead seating. She found one seat for him. I would have to sit in the row behind him. Okay. That seemed to work.

(I have to interject an aside here and say that several people in wheelchairs in Detroit were surprised and somewhat delighted to learn that, according to Northwest, they could bend their knees and were, therefore, no longer disabled!)

When we checked in to the airport in Philadelphia, things seemed to proceed fairly easily.

have his hard-won bulkhead seat.

Not a single Northwest agent could be found the entire length of terminal E. So, I had to go back down to ticketing and through security again to address the issue of having been given the wrong seats. This story just gets better and better. When I explained that I hadn't been given the seats I had booked, they told me that “maybe the plane was reconfigured after I booked my seats.” I'm not really sure what that means, but when I asked them if the plane configuration put row 13 as the bulkhead, the answer was “no.” The irony of all this was that when they looked in the computer, it actually read that Dana wore leg braces and required bulkhead seating. Right. But they changed his seat to be back eight rows in the middle of “no leg room” seating. Since I was getting nowhere with the ticketing agents, I asked to speak to a manager. That was no easy task, apparently, but

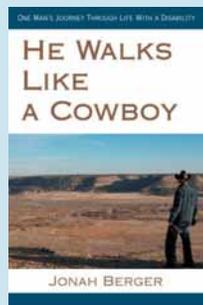
OF INTEREST

A book written by a CMT patient, Jonah Berger, entitled, *He Walks Like A Cowboy*, details his experiences in dealing with the disorder from his early days of childhood to his current age of 35.

In his introduction, Jonah writes:

“In the pages that follow, I humbly share my experiences of walking through this life with a disability, including all of the highs and lows that come along with that. I aim to share my perspective and that of my loved ones. Every person with a disability has a unique story to tell, and it is my hope that to read this story is to grant yourself some insight about life lived with a challenge. The story is mine; the theme is universal.”

The book was published by iUniverse and is \$12.95.



finally they said I could go back up to the gate and speak to someone they were sending there to speak to me.

He was, indeed, a manager, but he also told me that there were already people in the seats that had been originally booked for me. I asked exactly how that could happen...what, after all was the point of reserving seats if they were going to be given to other people? He apologized. I told him the "Can you bend your knees" definition of disability. He apologized again. I was getting really frustrated and was feeling righteously indignant. I finally asked if there were disabled people in our seats. The upshot was that they were, in fact, just random people who hadn't even requested the special seating. He couldn't, or wouldn't, fix the problem, but another agent seemed to be willing to take on the issue. Within about fifteen minutes, we were both in bulkhead seating and no one seemed at all disturbed about lost seats.

How could this happen? How can you make reservations and book special seats and then have them taken away by some mysterious "computer" problem. The real question, I think, isn't how "computers" can make these mistakes. The real question is why people won't accept responsibility for making mistakes and then correct them without so much hassle.

Being in Detroit *was* fun, but getting there really wasn't. (And I didn't even tell you that our flight coming back to Philly had to make an emergency landing because the wing flaps wouldn't open.) *

Facts VI and New Neurotoxic Drugs Cards Now Available

The newest edition of the Facts Series has just been published and is available for purchase. The sixth edition is 64 pages in length and is divided into sections on general information, genetics, diagnosis and treatment, therapies and therapists, bracing, CMT and children, and "Ask the Doctor."

The compilation of articles goes back as far as 2002 and captures the most significant articles from *The CMTA Report* from then to the present. Some of the articles involve numbness in CMT, HNPP phenotypes, current therapies for CMT by Dr. Michael Shy, exercise options, various types of bracing, IEPs for children, and the interesting and diverse questions posed to members of our Medical Advisory Board and answered by them in a ten-page section of the publication.

In keeping with our philosophy of providing information to the members of our association, we are providing this newest issue of the Facts series at the same price as *Facts V*. The cost for members (aren't we all members this year?) is \$12 and for non-members, \$15.

Not only is *Facts VI* now available, but the long-awaited neurotoxic drug list cards are now available as well. Because of the length of the new drug list, we struggled to get them produced in an attractive and wallet-friendly format and are now happy to announce their availability. They are free to anyone who requests one (or two or three), but if you are requesting them when you are not ordering something or making a donation, please send a self-addressed stamped envelope to help us with the processing. *

Facts VI:
64 pages of
important articles
on CMT and the
always popular
Dear Doctor
questions and
answers.



WRITE TO US!

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ASK THE DOCTOR

Dear Doctor,

My endocrinologist did a blood test which revealed a vitamin D deficiency, so he has prescribed a vitamin D regimen of 1.25 mg (about 50,000 units) per week for six weeks. I recall that there is a concern with megadoses of vitamin D being contraindicated for people with CMT. Also, is it common for patients with CMT to have elevated creatine kinase readings (normal for me is 300-400)? While exercising, it was as high as 580. Would statins be contraindicated for me with CMT?

The doctor replies:

There are neuromuscular problems with both too much and too little vitamin D. There is uncertainty about how much vitamin D is excessive, but it is quite a bit and more than the amount in the usual supplements. There is no evidence that moderate or lesser doses are

problematic, but insufficient vitamin D is associated with a variety of problems. CMT patients can have mildly elevated creatine kinase in the range you mention. Statins can increase creatine kinase and cause various levels of muscle problems. It is, however, very controversial how often or how severely they affect nerves.

Dear Doctor,

I would like to know if Botox is bad for patients with CMT. I was diagnosed with CMT in the last year and my neurologist has used Botox to try to help me with the pain in my feet. On the forum there were several people who said it wasn't a good thing for patients with CMT. Can you help me?

The doctor replies:

Pain is not a specific indication for botulinum toxin use, unless the pain is caused by excessive

muscle spasms or activity, most commonly dystonia or spasticity. Botulinum toxin (Botox and Myobloc) works in many cases by intentionally weakening certain muscles to alleviate the symptoms. Because foot and lower leg muscle weakness is already a problem in many CMT patients, there is concern that local injections may weaken them further, though only for 3 to 4 months. I would ensure that the treatment has an intended benefit and discuss the risk of weakening muscles near the injection site.

Dear Doctor,

I have CMT and have been prescribed lisinopril for high blood pressure. Is this a safe medication for me to take?

The doctor replies:

I know of no link between CMT, neuropathy and lisinopril.



LETTERS

Dear CMTA,

There was a query in the last issue of the newsletter about alpha-lipoic acid. I would like to share that I have CMT Type 2 and have been taking 150 mg of alpha-lipoic acid 4 times a day for about 4 years now. I have found that the terrible cramping in my legs and feet has almost completely disappeared, and, when I do get a cramp, it disappears in a matter of seconds when I change position.

After taking it for a few

years, I had nerve testing, and it showed that in some respects I had improved and in others I remained the same. I feel this has helped me and has possibly slowed the progress of the condition, but more than anything, it has been wonderful to be relieved of the cramping which used to keep me up at night.

—A.S. (by email)

(Editor's note: These are the opinions of a patient and do not reflect medical advice. Consult your doctor before beginning any drug usage.)

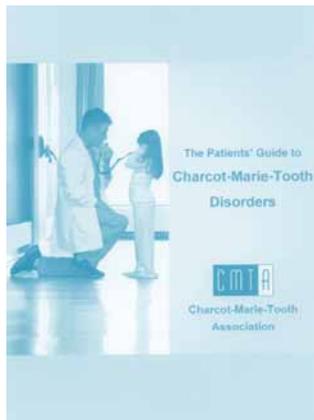
Dear CMTA,

As a long-time member of and frequent contributor to the CMTA, I was pleased to read in the March/April issue of *The CMTA Report* about free memberships for one year. I appreciate your doing that for members and for any other folks interested in joining. I want to sign my brother up, although he does not have CMT. He supports me in many ways, as I do have it.

—J.G., California

Patients' Guide to CMT...
Is Now Available

in print...



and CD...



The long-awaited update to our *Handbook for Primary Care Physicians* is now available in both print and CD format. Each chapter in the book has been written by a leading CMT expert and is directed at a more lay audience than the previous text.

Topics include the clinical features of CMT, electrodiagnostic evaluations, genetics, CMT in children, orthopaedic considerations, new treatments, physical exercise, genetic counseling, toxic medications, and resources for CMT patients and their families. The new book is 178 pages and features full color photos of many conditions and treatments.

The CD, which contains the entire text of the *Patients' Guide*, also contains a slide show by Dr. Steven Scherer, neurologist, University of Pennsylvania, which is intended for doctors who would like a quick overview of CMT and another slide show by Dana Schwertfeger, Director of Member Services, which is intended for patients and families who might need a fuller understanding of CMT.

We pride ourselves on being the best source for up-to-date information on CMT, and these two publications are our latest accomplishments in that area.

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A copy of the official registration and financial information may be obtained from the Pennsylvania Department of State by calling, toll-free within Pennsylvania, 1-800-732-0999. Registration does not imply endorsement.



CMT PATIENT MEDICATION ALERT:

Definite high risk (including asymptomatic CMT):

Vinca alkaloids (Vincristine)

Moderate to significant risk:

Amiodarone (Cordarone)
Bortezomib (Velcade)
Cisplatin and Oxaliplatin
Colchicine (extended use)
Dapsone
Didanosine (ddl, Videx)
Dichloroacetate
Disulfiram (Antabuse)
Gold salts
Lefluonamide (Arava)
Metronidazole/Misonidazole (extended use)
Nitrofurantoin (Macrochantin, Furadantin, Macrobid)
Nitrous oxide (inhalation abuse or vitamin B12 deficiency)
Perhexiline (not used in US)
Pyridoxine (mega dose of Vitamin B6)
Stavudine (d4T, Zerit)
Suramin
Taxols (paclitaxel, docetaxel)
Thalidomide
Zalcitabine (ddC, Hivid)

Uncertain or minor risk:

5-Fluorouracil
Adriamycin
Almitrine (not in US)
Chloroquine
Cytarabine (high dose)
Ethambutol
Etoposide (VP-16)
Gemcitabine
Griseofulvin
Hexamethylmelamine
Hydralazine
Ifosfamide
Infliximab
Isoniazid (INH)
Lansoprazole (Prevacid)
Mefloquine
Omeprazole (Prilosec)
Penicillamine
Phenytoin (Dilantin)
Podophyllin resin
Sertraline (Zoloft)
Statins
Tacrolimus (FK506, Prograf)
Zimeldine (not in US)
a-Interferon

Negligible or doubtful risk:

Allopurinol
Amitriptyline
Chloramphenicol
Chlorprothixene
Cimetidine
Clioquinol
Clofibrate
Cyclosporin A
Enalapril
Fluoroquinolones
Glutethimide
Lithium
Phenelzine
Propafenone
Sulfonamides
Sulfasalazine

What is CMT?

- ◆ is the most common inherited neuropathy, affecting approximately 150,000 Americans.
- ◆ may become worse if certain neurotoxic drugs are taken.
- ◆ can vary greatly in severity, even within the same family.
- ◆ can, in rare instances, cause severe disability.
- ◆ is also known as peroneal muscular atrophy and hereditary motor sensory neuropathy.
- ◆ is slowly progressive, causing deterioration of peripheral nerves that control sensory information and muscle function of the foot/lower leg and hand/forearm.
- ◆ causes degeneration of peroneal muscles (located on the front of the leg below the knee).
- ◆ does not affect life expectancy.
- ◆ is sometimes surgically treated.
- ◆ causes foot-drop walking gait, foot bone abnormalities, high arches and hammer toes, problems with balance, problems with hand function, occasional lower leg and forearm muscle cramping, loss of some normal reflexes, and scoliosis (curvature of the spine).
- ◆ has no effective treatment, although physical therapy, occupational therapy, and moderate physical activity are beneficial.
- ◆ is usually inherited in an autosomal dominant pattern, which means if one parent has CMT, there is a 50% chance of passing it on to each child.
- ◆ Types 1A, 1B, 1C, 1D (EGR2), 1E, 1F, 1X, 2A, 2E, 2I, 2J, 2K, 4A, 4E, 4F, HNPP, CHN, and DSN can now be diagnosed by a blood test.
- ◆ is the focus of significant genetic research, bringing us closer to solving the CMT enigma.

THE CMTA Report



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