



**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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EMAIL CMTA AT:
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Fighting for Funding and Awareness in South Florida

BY CHARLES F. HAGINS, EXECUTIVE DIRECTOR

Three years ago, I traveled south to Palm Beach, Florida, to meet with J. Rodman Steele, to discuss the possibility of a fundraiser in his wealthy seaside community.

On February 22, 2005, that discussion became reality when now Board member, J. Rodman Steele, hosted a dinner and presentation to raise money for the CMTA's research fund.

Over one hundred Floridians descended upon The Breakers in Palm Beach, Florida, to hear various presentations which subtly encouraged donations to the ascorbic acid clinical trials. I began the event with a discussion about the difficulty of raising money for a little known disorder. Conditions such as CMT, which are out of the public mainstream and do not provide income for drug companies, have difficulty getting funding for treatments and research. I asked the attendees to try to put themselves in the situation of someone with CMT so they could understand, personally, the need and value of clinical trials.

Following my presentation, Dr. Michael Shy, Wayne State University, and Dr. Stephen Scherer, University of Pennsylvania,



Dr. Stephen Scherer, Archit Khanuja, and Dr. Michael Shy were the guest presenters at the Florida fundraiser.

nia, explained from a scientific point of view, why ascorbic acid is the most exciting option for CMT Type IA patients to date. They emphasized what the possible outcomes would be from such a study.

Nothing, however, prepared me, or the audience, for the insight given to us by sixteen-year-old, Archit Khanuja, who shared how CMT affected her as she grew up in a Florida community. Archit put a human and youthful face on the problems of CMT and emotionally touched the whole audience.

Following the presentations, the doctors circulated throughout the dining room with microphones, repeating and

answering questions from the attendees.

The event was not an easy undertaking and many people and organizations were responsible for its success, primarily the law firm of Ackerman, Senterfitt of West Palm Beach, Florida,

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New Website Coming Soon...

BY DANA SCHWERTFEGER

Not as quickly as we'd like, perhaps, but soon, and we hope you'll bear with us a bit longer. When we started the redesign, we hoped to be ready for launch in April. We still might be, but we'd rather err on the side of caution and test things thoroughly than rush and put up something that doesn't work quite right, so the early part of May now looks like the more probable date.

Will it be worth the wait? Absolutely! We're not just cutting and pasting from the old site. We've updated and expanded the information about CMT and added several new pages. The new site will also be constantly updated with new infor-

mation as it becomes available, but that's just the beginning.

You'll be able to do a lot more online—everything from renewing your membership to reading the latest newsletter (and all the previous issues in the archives). The physician lists will be online, too, and you'll be able to submit the names of physicians we should add and help us keep our lists up-to-date.

We've also compiled a list of over 300 helpful links where you can find doctors, medical information, useful products, resources for parents, state and federal agencies, disability advocates, and much more.

For kids, there will be a page where they can read per-

You can now read
The CMTA Report online
several weeks before the
printed version goes out.

sonal narratives written by other kids and submit their own. One of the forums on the new bulletin board will also be reserved for them. We've reserved another forum for parents, and we hope both forums will become places where members support one another and share information and experiences.

We know how important a resource the Internet can be for CMT patients, doctors, friends

Why Does the CMTA Have Membership Dues?

BY PAT DREIBELBIS

The Charcot-Marie-Tooth Association is a membership-supported nonprofit organization established in 1983 to serve families dealing with CMT. Membership allows the organization to provide publications, books, fact sheets and our newsletter, *The CMTA Report*. These dues also help support the day-to-day operations of the CMTA, including our toll-free phone number, postage, and website design and maintenance.

CMTA membership also helps support the staffing needs of the association. Every time

you call, write, or email the association, CMTA staff members are here to respond to your requests, answer your questions, or refer you to someone who can help you.

Membership dues provide you with discounts at CMTA events and also provide you with an uninterrupted subscription to *The CMTA Report*. CMTA membership gives a voice to CMT families throughout the United States and the world, on issues which concern the disorder and those who deal with it daily.

Most nonprofit organizations have a membership dues structure to help the organization function as a viable entity. Without membership dues, the Association would not be able to provide many of its important programs and services.

Who are CMTA members? Paid members are persons with CMT, family members, members of the medical community, and general friends of the association. CMTA members are people like you and me who share the same concerns and want to help others in dealing with this disorder.

and families, and the CMTA is committed to making it easier for you to access more and better information in one concise location.

We're also committed to using your membership dues and the money you donate in the most effective manner, so when we're up and running, we'll give you the details on how to register your email address. Why? You'll need to be registered to access certain areas of the site, and think of the money we can save on printing and postage if we do membership reminders and annual campaigns via email! You can even opt to read *The CMTA Report* online several weeks before the printed version comes out.

Thanks again for being patient, and we hope you get as much out of the new site as we've put into it. *

If you are not currently a CMTA member or you have never supported the CMTA, please become a member today. In this issue, you will find a membership application and order form. Please complete the form and send it to the CMTA today. To those of you who faithfully send in your membership and even include something extra for those who cannot pay, we thank you.

Membership support helps the Association to maintain and expand the meaningful programs and services we offer to patients and families dealing with CMT. Help us continue to provide quality programs by becoming a CMT member today. *

CMTA MEMBERSHIP/ORDER FORM

Name: _____

Address _____

Phone Number: _____ Email: _____

Members who are current with their dues are considered "active." If you are unsure as to whether you are current with your member dues, please call the office at 1-800-606-CMTA.

	QTY	COST	TOTAL
Charcot-Marie-Tooth Disorders: A Handbook for Primary Care Physicians		active members \$15 inactive members \$20	
Membership Dues		\$40	
CMT Facts I <input type="checkbox"/> English <input type="checkbox"/> Spanish		active members \$3 inactive members \$5	
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Contribution to CMT Research <i>10% will be applied to administrative expenses.</i>			
Shipping & Handling <i>Orders under \$10 add \$1.50, orders \$10 and over add \$4.50</i>			
TOTAL			

Check payable to the CMTA (US residents only).
Foreign residents, please use a credit card or International Money Order.

VISA MasterCard American Express

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Mail to the CMTA, 2700 Chestnut Parkway, Chester, PA 19013 or Fax to 610-499-9267.

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.



Parenting: The Art of Letting Go

BY ELIZABETH OUELLETTE

“He has what? I’ve never even heard of CMT! I don’t believe it. I won’t believe it! There must be some mistake.” These were the first thoughts racing frantically through my mind when the neurologist finally came up with the definitive diagnosis for my then seven-year-old son, Yohan. Neither my husband nor I tested positive for CMT, so why is it that my only child has a genetic mutation causing this life-changing neuromuscular disease? What did I do wrong? How will we deal with this as individuals and as a family?

I would ask myself these and many other questions over and over again, trying to understand, striving to make sense of why an innocent child has to endure such challenges so early in life. I have experienced grief in all its stages—denial, anger, bargaining, depression, and acceptance—and just when I think acceptance will remain strong in my heart, I have setbacks, reverting back to anger and sliding down the scale once more.

My world shattered into millions of pieces that day, and I never thought we’d be able to pick up all the scattered bits to reformulate our dreams, our hopes, and our wishes for our son, while creating a different outlook on all our lives. This reassessment took time, effort,

and a lot of soul searching, but I truly think we are all making headway.

Seeing a child struggling with pain, braces, physical limitations, and obvious differences makes me frantic. My maternal instincts tell me to protect, shelter, cajole, and, especially, do something—anything—to make the world a friendlier, more secure place for him. The more



Encouraged by his personal trainer, Yohan decided to try snowboarding.

his self-esteem plummeted, the more his self-confidence lessened, the more he buried his emotions, the more I would try to make all the bad stuff disappear, pretending that there were no underlying health issues and that everything was peachy keen.

However, kids are too smart and they know when the mask of denial is blotting out reality. As time passed, Yohan became more and more anxious, less

focused, and, simply put, a very unhappy child. What else could I do? Lightening his load did not seem to be the answer, and neither did catering to all his needs. In retrospect, he was becoming more dependent on me for everything, and his teachers in school commented on his lack of autonomy, maturity, and self-motivation. Moreover, I realized that the

secondary gains we both obtained by his being “sick” were actually hindering his development and emotional growth. On the one hand, I was disillusioned by the fact that my inner desire to feel needed and indispensable was actually playing a role in his inability to blossom and to manage the effects of his CMT. On the other hand, his neuromuscular disease quickly became a great way of avoiding uncomfortable situations and often served as an escape from school, from

peer relationships, and from dealing with his CMT in a healthy way.

I thought long and hard about what was playing out before my eyes and decided to get some help from a therapist who counsels families on raising children with medical issues and/or learning differences (both of which pertain to Yohan). Intuitively, I knew what measures needed to be taken, yet I

was searching for someone to help illuminate the way and help me along the path.

I certainly did not want my child to suffer unnecessarily, but making him accountable for his actions, giving him more and more responsibility, and especially “cutting the umbilical cord” so that he may experience the world on his own terms are probably the hardest but most essential jobs I have as a parent of a child with special needs. Both my husband and I were determined to provide Yohan with tools he needed to be independent, self-sufficient, tenacious, and, hopefully, optimistic. After numerous discussions and a lot of trial and error, I now believe that all three of us are on the same wavelength, working together towards common goals. Nevertheless, I still tend to stray at times, fretting over hypothetical possibilities, living much too far in the future, and being obsessed with “what ifs”—what if this happens, what if that happens.

All of this came into play on a recent trip to Vermont. Four years ago, Yohan had tried skiing, but he was very uncomfortable and lacked the flexibility to be successful. He has been content to slide and throw snowballs at his parents since then, but he does have surprisingly good balance, so Steve, his personal trainer at the YMCA, encouraged him to try snowboarding this time around.

I try very hard not to be an overprotective mom, but I privately hoped Steve would not insist. I also silently wished Yohan would dismiss the idea. That was not to happen, however. My husband loves snow sports, and he

CHALLENGE GRANT PROGRESS

The Challenge issued by Board Member, Gary Gasper, has already generated \$11,000 in matching funds. Please join the challenge and help us make, and exceed, our goal. Watch the mail for the Research fundraising letter which encourages each of you to work on making the \$100,000 from Gary into \$200,000 or more!

was excited to have Yohan give snowboarding a try.

As for Yohan, he was completely pumped up to hit the slopes! Unenthusiastically, I rented the snowboarding equipment, grumbling under my breath the whole time, and when father and son left early one morning after Christmas, I stayed home and tried to keep busy, crossing and uncrossing my fingers. I imagined the worst, but several hours later Yohan bounded through the door, still in one piece and with a look of content satisfaction on his face. “I loved it!” he blurted, “I’m going to take this on seriously!” “Phew,” I thought. “What a relief!”

Like many first-time snowboarders, Yohan took a good many tumbles and falls in the snow, but he was confident of his potential (thanks to the motivational and insightful words of his personal trainer), and his determination and resilience took over, making his experience a complete success. I admire Yohan for his bright outlook and positive attitude, and I am also very grateful for my husband’s support and his unwavering confidence in Yohan.

This event also reinforced

what I already know as a parent but still find it difficult to admit. Even though Yohan has CMT, there are many things he can do with the right support and guidance, and he has to be given the freedom to grow and explore a world filled with unknown dangers. New activities may not always turn out as well as hoped, but if we don’t let him try, he’ll never know what he can do.

As I write this, I am reminded of the movie “Ray.” Despite his total and permanent blindness at the age of 7, Ray Charles’ mother treated him the same as any other child. He was made to do his chores, learn to get around without a cane, and fend for himself. “I won’t be here forever,” she repeatedly told him. Harsh though her treatment of the young Ray seemed, their story is filled with wisdom beyond words, and it continues to be a source of hope and inspiration for me. As hard as it may be, I, too, will witness the successes, the failures, and the struggles my son will have to contend with during his journey through life. If there is just one gift with which I wish to leave him, it is the knowledge that he can achieve his heart’s desire. He just has to believe in himself. ❄️

*Editor's note:
This article is not
an endorsement of
this brace, but
merely a presentation
of one option for
CMT patients.*

Helios Brace Benefits

BY MITCHELL S. WARNER, CPO

Many new braces enter the marketplace year after year. CMT patients try to gather information on new bracing to learn what the potential benefits may be. As time goes on, some may try different types of braces, hoping for long-awaited results. Expectations are to achieve a higher level of walking, function, and balance restoration. As I have worked with and designed different types of energy-loading bracing with helical configurations, I learn from follow-up and feedback about long-term benefits, and the results that the brace has produced for them.

PREDOMINANT BRACING GOALS FOR CMT PATIENTS

Most of our patients that are fit with Helios designs have CMT. I would summarize their goals for bracing in this order:

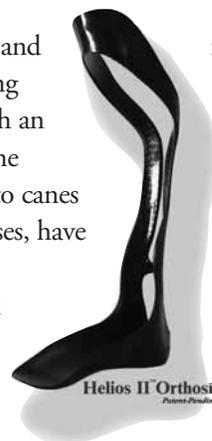
1. Restoration of standing balance. Standing balance becomes critical for the CMT patient because proprioception is affected due to the disease. Proprioception is determined by feedback from sensory nerves in muscles and tendons that give information about limb involvement and positional balance of the body. For many CMT patients impaired proprioception severely

compromises standing and walking balance, causing patients to always touch an object for stability. Some patients have resorted to canes or, in more extreme cases, have gone to a walker.

2. A more functional gait. CMT patients know that they walk with a foot-slap or what is commonly called a high-steppage gait. This happens when the heel cannot strike the ground in the first phase of the human gait cycle. As the foot slaps the ground and makes initial contact, the knee is in a flexed position. When the knee goes down flexed, it comes back up flexed in order to clear the ground. This is what perpetuates a steppage gait cycle. In many cases, foot deformities are present that force weight-bearing on the outside or inside of the foot. This causes further imbalance. Proper alignment of the lower limb and its joints can require a functional device to correct and prevent this. Once heel-strike is obtained, the gait becomes more fluid and more balanced. In addition, balance controls are incorporated into the bracing system through biomechanical alignment which helps restore a center of gravity

for the patient. Foot deformities must also be corrected as much as possible.

3. Comfort. Patients want to be as comfortable as possible in a device. If comfort cannot be obtained, the common reaction will be to discard the device. As the patient continues to wear a brace, comfort and correction are sometimes conflicting; however, with careful diagnostic pre-fitting of braces, and careful evaluation of a pre-diagnostic correction, both comfort and correction can be obtained at the same time.



INQUIRIES

Since I wrote an article for *The CMT Report* in December 2002 about Helios bracing, a common question has been: are the patients you have fit in that time still wearing the devices and how well are they doing?

One such patient is Mr. John Fox, age 79, of Columbus, Indiana. Mr. Fox came to us a little over two years ago and was quite frustrated with bracing. He had tried different types of bracing in the past and still could not obtain the balance and walking function that he desired. When Mr. Fox first



Before bracing

came to us, he presented a mechanical profile of midtarsal collapse, or what we call foot valgus (more commonly known as pronation), severe instability, and an inefficient gait profile.

The “before” pictures (*facing page*) show Mr. Fox without braces and the gait sequence presents a severe drop foot and extremely impaired balancing. His head looks down at all times at his feet in order to ambulate and to give him back a sense of proprioception. The gait deviation he employs without braces is lateral trunk bending, and hip hiking, in order for the foot to clear the ground. With these gait deviations in place and with balance impaired, his gait is very slow and awkward. He quite frequently needs to touch objects in order to regain his balance.

In the “after” pictures (*below*) with Helios bracing, Mr. Fox has achieved a normal heel strike, normal balance, and dramatically increased velocity.

We asked Mr. Fox to explain what he felt were the benefits of Helios bracing. John Fox states that the major changes he experienced are:

“My balance is restored. I still had foot drop with previous braces. In the past I needed external objects to grab on to as I walked and stood. I could not walk more than half a city block without fatigue. I had a severe marching gait. I walked with gait deviations which caused me even

CMT and Your Nation’s Capital

The CMTA is making progress in its efforts to increase CMT research at the National Institutes of Health (NIH), located in Bethesda, Maryland, just outside of Washington, DC.

Last year, CMTA’s work with members of the United States Senate and House of Representatives led Congress to direct NIH to report on its efforts on CMT and to look for ways to increase research into CMT.

Recently, NIH released its report and there is good news: NIH plans to conduct what it calls a “Workshop” on peripheral neuropathies. In other words, NIH will bring experts from the National Institute on Neurological Disorders and Stroke (NINDS) and other NIH Institutes together with experts from around the country to map out an agenda for peripheral neuropathies and CMT. Hopefully, from this Workshop an enhanced research strategy will emerge to help find a treatment and cure for CMT.

Also in April 2005, Charles Hagins, Executive Director of the CMTA, submitted testimony to Senate and House Subcommittees about recent scientific developments related to CMT. His testimony encourages Congress to continue its interest in CMT and makes a strong case for the importance of increased research.

The CMTA board will be in Washington, DC, late in April to meet with members of Congress and the Senate. In the next issue of the newsletter, CMT and Your Nation’s Capital will report on the progress achieved in these meetings and other developments. ✨

more fatigue and instability. This was even the case when I had a plastic brace that was hinged with ankle joints. Now, my walking speed has increased dramatically. I can walk four miles per hour for ten minutes on the treadmill. I have a normal gait with no fatigue and can walk two miles. I also use a cross-trainer. I am back to living a normal life. Before I was braced properly I was ready to give up on golf. I couldn’t walk from the golf cart

to the ball. My golf game has even improved. Additionally, as my wife knows, I was depressed. After these braces, I am in a better state of mind. With the Helios, I was able to stand without holding on to anything for the first time in five years.”

While corrective bracing systems yield benefits of restored balance and increased walking function, they are not magical, and the average person does need time to break them in. I would compare it to starting a workout program at a gym or learning to play a new sport. It takes practice. Muscles that are now being used for the first time in years need to be slowly exercised.

(continued on page 8)



After bracing

**FOR MORE
INFORMATION
ABOUT THE
HELIOS BRACE,
CONTACT:**

Ortho Rehab Designs
2578 Belcastro St.,
Suite 101
Las Vegas, NV
89106

HELIOS BRACE

(Continued from page 7)

**UNEXPECTED BENEFITS OF
HELIOS BRACING**

When I started the Helios bracing system, the goal was to restore balance and walking function. As I have now had patients in these braces for over two to four years straight, (with follow-up) many of my patients state that they have had an increase in muscle tone, particularly in the calves and thigh musculature. We have not yet had these results tested. I feel in such cases where there has been increased muscle tone, this has been due to freeing up the toes with a slightly shortened foot-plate in order to let the

metatarsal joints bend. This results in the patient being able to use what musculature is still working in order to push off the ground to help them balance and walk. In my opinion it is very important not to over-brace. Once an orthosis controls the patient too much, it is not ideal, because the potential to use functioning musculature is greatly diminished.

**ONGOING RESEARCH
AND DEVELOPMENT**

As new scientific applications for leg orthoses arise, ongoing research and development is necessary to attain better outcomes. I work with composite materials in bracing, such as carbon fiber, Kevlar, and fiberglass. These

composites are carefully structured for each custom-designed brace; which means that no two braces are exactly alike. The proper engineering of the carbon composites results in a brace being both rigid and flexible where it is structurally desired. Continued research and development looks for improved application and fabrication techniques, and ultimately more advanced results for the brace wearer.

It is my goal to always learn about new advanced materials applications, and to design appropriate bracing systems to meet my patient's needs. With advanced procedures and applicable custom brace design, patients can expect better function and all of the consequential benefits. *

**GIFTS WERE MADE TO THE CMTA****IN HONOR OF****Zohar Birin**

Leon Gelman

Margaret Davis

Pat & Skip Davis

Gail Feeney-Coyle

Mary Feeney

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 KIDS PAGE

Thoughts on CMT

BY YOHAN BOUCHARD, 11 YEARS OLD

Honestly, having CMT really sucks. The pain in my calves and shins hurts and the tingling in my feet and legs is very uncomfortable. I have to wear braces during the day and sometimes at night, but they irritate me so much, I usually have to take them off or I don't sleep very well. I am forced to take a lot of medication for my pain and some nights I just throw it up. Since I am allergic to the material inside my casts, I wear special socks which are bothersome and make my feet sweat.

I cannot do as much as other kids my age because CMT makes my feet hurt and my legs ache. CMT causes fatigue, especially in the hot weather, so I have much less energy than my friends. CMT also affects my fine motor skills, so my hands cramp easily, making it hard to write. Sometimes kids make fun of me because they do not understand why I cannot run as fast or walk as far as they can.

Every year my mom gives a fun presentation to my class explaining CMT, so they gain awareness and understand my health problems a little better. So far, I have made more friends who accept me just the way I am thanks to my mom and other



Yohan Bouchard with his parents.

helpful people. I try to be nice to all other kids, especially those who have physical limitations or learning problems because I know just how they feel.

Having physical differences doesn't have to be all that bad. I can relax more at home and participate in special activities other kids don't do, like riding my horse Athos, scuba-diving when on vacation, and zooming around on my very own, very cool electric scooter.

I hope stem cell research will help improve my CMT symptoms and perhaps cure all the pain and suffering people with neuromuscular diseases have to deal with every day. ✨

CMT

BY KELSEY SHINNICK

CMT may affect you in your hands or feet,
 However it affects you, difficulties you meet.
 Some people need canes, braces, or wheelchairs,
 Some may need help opening bottles, or climbing upstairs.
 Trouble with writing or walking,
 Trouble with friends or mocking.
 Just be happy that others know how you feel,
 Don't keep these issues bottled up, these problems are real.
 Don't be depressed or sad,
 These problems should not make you mad.
 Be thankful that your problems aren't worse,
 Think of CMT as a challenge, not a curse.

Kelsey is 12 years old and wrote this poem for people with CMT.



ASK THE DOCTOR

Dear Doctor:

I am a patient who is taking biotin to harden my nails and I'm concerned because it's listed as B-complex. Are there any contraindications that you're aware of?

The Doctor Replies:

There is no known association between biotin and neuropathy. The only B vitamin clearly known to cause neuropathy in excess is pyridoxine (B₆). However, all vitamins should be taken in conventional doses unless otherwise directed.

Dear Doctor:

How great is the neurotoxic risk from a short course of metronidazole? My gastroenterologist has said there is no good alternative other than \$10,000 Vancomycin.

The Doctor Replies:

There are occasions when a concerning medication is the best option. The only drug that should not be taken under any circumstances by CMT patients is the chemotherapy drug vincristine. All others have to have risks and benefits weighed against each other before deciding. In many cases a less risky drug is an equal alternative. Bearing this in mind, metronidazole is at greatest risk with extended use, usually 6 to 8 weeks or more. The risk is even higher with chronic use over many months at high dose. There is little evidence of

high risk with short courses, such as 7 to 14 days, especially if the infection to be treated is potentially serious.

Dear Doctor:

I have advanced CMT, type 1A. I am 29 and not a diabetic. A year ago I was diagnosed with gastric paralysis or gastroparesis after I underwent a gastric emptying study. My neurologist and internist both suspect a connection between CMT and the gastric paralysis. Has this been heard of before? My father, who also has CMT, is also showing symptoms of gastric paralysis. Please let me know if you have heard of a connection between the two.

The Doctor Replies:

This would be very unusual, as most neurologists believe that the autonomic nervous system is largely spared in CMT 1A, though this is beginning to be a subject of research. Gastroparesis caused by CMT 1A would be very unusual, so other potential causes need to be carefully investigated first. I hope that this is helpful.

Dear Doctor:

I have had CMT for 31 years. I am 45 years old and have had night sweats for five months. I started taking soy isoflavone concentrate, 50 mg, once a day. Is it okay to continue taking the soy?

I have read that I should limit my folic acid. If you have any information about either of my concerns, I would appreciate your expertise.

The Doctor Replies:

There is no evidence that soy isoflavones affect CMT neuropathy for better or worse. You have to judge for yourself if they are easing the menopausal symptoms. Pyridoxine (B₆) is the most important vitamin to avoid in high doses, but it is needed in normal dietary and general multivitamin doses. There is no evidence that customary doses of folate or folic acid are hazardous for CMT patients.

Dear Doctor:

Would it be okay for us with CMT to take a stress B-complex vitamin supplement daily? Also, how often should we have our B₁₂ level checked?

The Doctor Replies:

A daily stress B-complex is fine as long as it does not contain excessive doses of pyridoxine (B₆). Most people obtain a sufficient amount of B₁₂ in their diet unless they are strict vegetarians or have a separate disorder that prevents B₁₂ from entering the bloodstream, so there is no need to routinely check B₁₂ unless there is concern that it may be low. *

The GlideAbout

The adult scooter/walking assistant, designed by a man who has CMT

(Editor's note: We tried a sample of the GlideAbout in the CMTA office. The man who works here and has CMT was the most accomplished in using the scooter. He found it simple and smooth to operate. The woman with knee replacements was timid and insecure on the device and I found it tired my thigh muscles to push the scooter on carpet, but was much easier on the sidewalk outside.

This was, by no means, a scientific study and in general, we found the scooter easy to assemble and disassemble to put in the car. We did not get a chance to try it in a WalMart or similar location with level and non-resistant floors.)

If you have difficulty walking or standing on cement, marble, brick, or other hard surfaces—if your feet and lower legs are injured or just get sore—then the GlideAbout could be for you!

The GlideAbout is an adult scooter/walking assistant.

According to the company, this device:

- Folds easily and fits in a corner of the trunk of your car



The GlideAbout™

- Is lightweight and can be lifted with one hand
- Glides like ice along hard surfaces with just a gentle push of your leg, so it requires no motor, which makes it convenient: no charging, no gasoline, and no hassle taking it into your favorite stores, such as Costco, Target, Home Depot, grocery stores, or shopping malls

GlideAbout is designed for:

- People with permanent foot problems, such as flat feet, arthritis, hammer toes, or foot deformities

- People with temporary foot problems, such as bunions, corns, or ulcers
- People with tendon or ligament injuries
- People who have undergone recent foot or knee surgery
- People with weak, broken, or sprained ankles
- People with walking casts
- People who just like to ride rather than walk!

GlideAbout

- Is a superior alternative to crutches or a wheelchair, as long as you can momentarily support your own weight
- Allows you to ride in an almost standing position, so you are eye-level with those around you, unlike a wheelchair
- Is easy on your hands and arms, unlike crutches
- Has a soft seat!
- Has a brake (not that you really need it)
- Has a shopping basket, although you can easily pull a grocery shopping
- Is just plain fun, unlike crutches or a wheelchair! ✨

FOR MORE INFORMATION ABOUT THE GlideAbout:

Visit the website www.glideabout.com or email: contact@glideabout.com or call 1-877-GLIDEAB (454-3322).

SOUTH FLORIDA

(Continued from page 1)

which underwrote the cost of the dinner.

Months of preparation were required to make this fabulous evening possible and simply saying “thank you” to the organizers does not seem adequate.

J. Rodman Steele and his committee did an extraordinary job.

The money raised at this event will significantly contribute toward the \$30,000 committed by the CMTA to the first clinical trials of ascorbic acid in North America. It is estimated that the clinical trials will cost approximately \$5,000,000

over four years. Approximately 500 patients will be tested at four different centers, to be named at a future date.

The CMTA hopes that J. Rodman Steele was encouraged by the turnout and the generosity enough to make this South Florida fundraiser an annual event! ✨



SUPPORT GROUP NEWS

New York – Greater New York

The Greater New York support group has a website located at www.cmtnyc.org. The site is an informational and interactive website for individuals with Charcot-Marie-Tooth disease. There are articles on CMT as well as a page to get responses to “Ask the Experts” questions. The group is under the guidance of Dr. David Younger, neurologist.

Pennsylvania – Johnstown

The Johnstown, PA, CMTA support group held its bi-monthly meeting on Saturday, February 26th, at the John P. Murtha Neuroscience and Pain Institute, a member of the Conemaugh Health System. Cathy Crist from PAID (The Pennsylvania Association for Individuals with Disabilities), addressed issues concerning disabilities and the workplace. PAID, <http://www.paid-online.org>, is a nonprofit organization dedicated to addressing the employ-

ment needs of disabled individuals through unique programming, education, and partnership with business and industry.

Some of the issues Cathy covered included the facts that 70% of individuals with disabilities are not employed and that tax incentives exist for employers to hire the disabled. She also discussed SSD eligibility and working, who is hiring locally, and the workforce of local defense contractor, Kuchera Industries, which is over 50% disabled. Cathy’s talk was interesting to all and invaluable to our nonworking members.

Jan Goodard, from John P. Murtha Neuroscience and Pain Institute, spoke of the exciting CMT research projects under development at the Institute. She also discussed the benefits of Conemaugh’s new Art for Healing, a program employing artistic creativity to benefit chronic conditions in general and CMT in particular.



Cathy Crist discussed employment for disabled persons.

J.D. Griffith gave a brief PowerPoint presentation on a potential new treatment for CMT1A, interference RNA (RNAi). The science behind RNAi is baffling to most and remained so following the presentation and video.

We again want to thank The John P. Murtha Neuroscience and Pain Institute, The Conemaugh Health Systems, and Jan Goodard and Barb Duryea, in particular, for their help with our support group, dedication to helping CMT patients, and, now, CMT research. *

CMTA REMEMBRANCES

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.

Honorary Gift:

In honor of (person you wish to honor)

Send acknowledgment to:

Name: _____

Address: _____

Occasion (if desired):

- Birthday Holiday Wedding
 Thank You Anniversary Other

Memorial Gift:

In memory of (name of deceased)

Send acknowledgment to:

Name: _____

Address: _____

Amount Enclosed: _____

Check Enclosed VISA MasterCard

Card # _____

Exp. Date _____

Signature _____

Gift Given By:

Name: _____

Address: _____



CMT Support Groups

Bob Budde, Support Group Liaison, 859-255-7471

Arkansas—Northwest Area

Place: Varies, Call for locations
Meeting: Quarterly. Meetings are not regularly scheduled so call ahead.
Contact: Libby Bond, 479-787-6115
Email: charnicoma57@yahoo.com

California—Berkeley Area

Place: Albany Library, Albany, CA
Meeting: Quarterly
Contact: Gail Whitehouse
Email: gwhite@earthlink.net

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

Place: 300 Sovereign Lane, Santa Rosa
Meeting: Quarterly, Saturday, 1 PM
Contact: Freda Brown, 707-573-0181
Email: pcmobley@mac.com

Colorado—Denver Area

Place: Glory of God Lutheran Church Wheat Ridge
Meeting: Quarterly
Contact: Marilyn Munn Strand, 303-403-8318
Email: mmstrand@aol.com

Florida—Tampa Bay Area

Place: St. Anthony's Hospital, St. Petersburg, FL
Meeting: Quarterly, 2nd Saturday, 10:30 AM
Contact: Lori Rath, 727-784-7455
Email: rathouse1@msn.com

Kentucky/Southern Indiana/Southern Ohio

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

Minnesota—Benson

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

Minnesota—Twin Cities

Place: Call for location
Meeting: Quarterly
Contact: Maureen Horton, 651-690-2709
 Bill Miller, 763-560-6654
Email: mphorton@qwest.net, wmillier7@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: c.haislip@att.net

New York—Greater New York

Place: NYU Medical Center/Rusk Institute, 400 E. 34th St.
Meeting: Third Saturday of every other month, 1-3 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 Hospital
Meeting: Bimonthly, Jan, March, May, Sept, and Nov; 3rd Saturday
Contacts: Beverly Wurzel, 845-783-2815
 Eileen Spell, 201-447-2183
Email: cranomat@frontiernet.net
 espell@optonline.net

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

Place: Church of the Reconciliation, Chapel Hill
Meeting: Quarterly
Contact: Susan Salzberg, 919-967-3118 (evenings)

Ohio—Greenville

Place: Wills Restaurant, 405 Wagner Ave, Greenville
Meeting: Fourth Thursday, April–October
Contact: Dot Cain, 937-548-3963
Email: Greenville-Ohio-CMT@woh.rr.com

Ohio—NW Ohio

Place: Medical College of Ohio
Meeting: Quarterly
Contact: Jay Budde, 419-445-2123 (evenings)
Email: jbudde@fm-bank.com

Oregon/Pacific NW

Place: Portland, Legacy Good Sam Hospital, odd months
 Brooks, Assembly of God Church, even months
Meeting: 3rd Saturday of the month (except June and Dec.)
Contact: Darlene Weston, 503-245-8444
Email: blzerbabe@aol.com

Pennsylvania—Johnstown Area

Place: Crichton Center for Advanced Rehabilitation
Meeting: Bimonthly
Contact: 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@adelphia.net

Pennsylvania—Philadelphia Area

Place: Penn Towers Hotel Conference Room
Meeting: Bimonthly
Contact: Amanda Young, 732-977-9983
Email: astarryoung@yahoo.com

Pennsylvania—State College

Place: Centre County Senior Center
Meeting: Monthly
Contact: Rosalie Bryant
Email: rab296@psu.edu

Planning for Elective Surgery

BY LOUIE BOITANO, MS, RRT, PULMONARY CLINIC, UNIVERSITY OF WASHINGTON, SEATTLE

Almost all patients who are scheduled for elective surgery need a preoperative evaluation to determine their health status and to plan for pre- or postoperative care that will best support them through the surgery. A preoperative evaluation for a person with a neuromuscular condition who is scheduled for surgery must include a thorough respiratory assessment and detailed medical history.

While physical limitations may be apparent to the evaluating physician, mild-to-moderate respiratory limitation can be overlooked if the person does not express any symptoms. An individual with slowly developing respiratory muscle weakness may not even be aware of respiratory limitations.

RESPIRATORY EVALUATION

Pulmonary function testing should include a forced vital capacity test in both sitting and supine positions, if possible, maximum inhalation and exhalation pressure tests, a peak cough flow test, and an arterial blood gas test. These test results are used to measure respiratory muscle strength and to screen for lung or diaphragmatic problems that can affect breathing.

A history of sleep-related problems including snoring, frequent awakening during the night, frequent need to urinate more than once per night, nightmares, awakening with headaches in the morning, or

excessive daytime sleepiness can indicate sleep apnea and/or under-ventilation related to respiratory muscle weakness. The inability to sleep flat on one's back because of feeling short of breath (orthopnea) can indicate diaphragm muscle weakness.

Night-time pulse oximetry to monitor blood oxygen saturation (optimal is >95%) and a diagnostic sleep study may be necessary to screen for sleep-disordered breathing. If a sleep study is found to be positive, an additional positive airway pressure (PAP) study should be performed to determine the level of noninvasive positive pressure ventilation (NPPV) needed to provide optimal night-time breathing support. Ideally, if the surgery is elective, there will be time to become comfortable using NPPV support prior to surgery.

Swallowing function must be evaluated before surgery to determine an individual's ability to prevent anything but air from entering the lungs. Weak swallowing muscles can limit one's ability to protect the airway following surgery. While poor swallowing function will not affect the ability to undergo some surgical procedures, it may determine the level of support in monitoring the individual's airway both during and following the surgery.

PLANNING

Once the preoperative evaluation is completed, a plan of care can be developed that best sup-

ports the individual's needs during and after the procedure. The planning team should include the surgeon or physician performing the procedure, the anesthesiologist, if one is needed, and the individual's pulmonary physician. The care plan depends upon the type of surgery, the need to use either general anesthesia or conscious sedation during the procedure, and the individual's degree of respiratory muscle weakness.

Individuals who already use continuous noninvasive respiratory support are also viable candidates for most surgical procedures, if they do not have other complicating conditions such as heart disease. Additional preoperative planning will be needed in determining the feasibility of using the individual's personal home ventilator and the coordination of respiratory support from noninvasive to invasive, if needed, for the surgical procedure, and then from invasive to noninvasive back again at the appropriate time.

The type of procedure and the time it takes are important factors. Procedures such as a gastric tube placement or colonoscopy requiring conscious sedation are relatively minor and short in duration. They can be well supported using NPPV along with airway monitoring by someone experienced in noninvasive respiratory support. In almost all cases requiring general anesthesia, the person must have a breathing tube placed

during the procedure to provide breathing support and to protect the airway.

POSTOPERATIVE CARE

Noninvasive breathing support can be immediately started once the patient awakens from anesthesia and the breathing tube is removed. If the individual is already using NPPV, he or she should have consulted with the team about bringing his or her own ventilator to the hospital to use at this time. Depending upon the type and length of the surgery, resumption of NPPV can occur in the operating room, the post anesthesia recovery unit or the ICU.

Postoperative pain can limit an individual's ability to breathe and cough adequately. This is especially true for individuals with neuromuscular conditions.

However, postoperative pain control may often require pain medications that can also affect an individual's ability to breathe independently. For this reason, a person should continue to use NPPV following surgery with a backup respiratory rate that ensures enough support if breathing becomes depressed by the amount of medication necessary to control the pain.

The placement of a breathing tube in the throat during general anesthesia often causes discomfort after the tube is removed. Because the breathing tube is a foreign object in the airway, the body often reacts to the tube by producing more secretions for a while after the tube is removed.

An individual with a weak cough, evidenced by a peak cough flow less than 200

L/minute, who is also taking pain medication, will have difficulty clearing secretions after surgery. A mechanical cough assist device such as the CoughAssist (J. H. Emerson Co., www.coughassist.com) should be available to aid in secretion clearance, as needed, during postoperative recovery.

An individual undergoing a significant surgical procedure requiring general anesthesia will be closely monitored in post-anesthesia recovery and in the ICU to ensure optimal respiratory support and cough augmentation during the first 24 hours following surgery. *

Reprinted from Ventilator-Assisted Living (formerly called IVUN News) with permission of Post-Polio Health International (www.post-polio.org).

LETTER

Dear CMTA,

Enclosed is a check in memory of my son, Jeffrey Dennis Gerry, who died on November 2, 2004, from colon cancer. Unfortunately, by the time the cancer was diagnosed, it was stage 4 and had spread to his liver.

He was able to undergo only one chemo treatment due to the devastating side effects. Within hours of the treatment, his muscles had been so severely affected that he could no longer walk, even with the aid of his walker. The doctor had intended to use vincristine for treatment and was not aware of its devastation on CMT patients. Unfortunately, the drug used instead caused

basically the same side effects as vincristine. I am in the process of having the doctor report to the CMTA what drug was used and its affects on this CMT patient.

Jeffrey was diagnosed with CMT as a young boy, and suffered its effects his entire life. He struggled to stay on his feet, but was given a tremendous challenge when he broke his leg from a fall when he was 37 years old. He used that challenge to bond with his infant daughter and took care of her for two years until he was able to return to work. Once the cast came off, he immediately went outside and tore down the ramp he had used for his wheelchair. He

refused to use his disability as a crutch. He never asked for special favors and never, ever complained about his limitations. He even went parasailing and had a 4-wheeler that was his vehicle of freedom, allowing him to explore the woods and valleys that he could never have walked.

Please continue the good work and use every avenue available to cure this cruel and devastating disease. My two daughters also have it and two of my four grandchildren. My hope also is that the medical community will become better educated about the side effects of cancer drugs and vincristine in particular.

—B.G.

WRITE TO US!

Pat Dreibelbis, Editor
The CMTA Report
CMTA
2700 Chestnut Pkwy.
Chester, PA 19013
or
CMTAssoc@aol.com

The CMTA reserves the right to edit letters for space.



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

Leflunomide

Metronidazole/Misonidazole

(extended use)

Nitrofurantoin (Macrochant,

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)

Fluoroquinolones

Gemcitabine

Griseofulvin

Hexamethylmelamine

Hydralazine

Ifosfamide

Infliximab

Isoniazid (INH)

Mefloquine

Penicillamine

Phenytoin (Dilantin)

Podophyllin resin

Sertraline (Zoloft)

Statins

Tacrolimus (FK506, Prograf)

Zimeldine (not in U.S.)

a-Interferon

Negligible or doubtful risk:

Allopurinol

Amitriptyline

Chloramphenicol

Chlorprothixene

Cimetidine

Clioquinol

Clofibrate

Cyclosporin A

Enalapril

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.
- ◆ 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 sometimes surgically treated.
- ◆ 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), 1X, HNPP, 2A, 2E, 4E, and 4F 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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