SUMMER 1999

The CMTA Report

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A resource for information on Charcot-Marie-Tooth disease (Peroneal Muscular Atrophy or Hereditary Motor Sensory Neuropathy), the most common inherited neuropathy.

EMAIL CMTA AT:
CMTAssoc@aol.com

The Armington Research Challenge 1999:
Personal Commitment Drives Success

Several hundred members responded to the Armington Research Challenge, taking it personally—some taking it to their friends and family as well, to ensure this final year's success. In 1996, the Evenor Armington Family Foundation presented the CMTA with a $25,000 one-to-one challenge. The terms were increased to a 3:1 arrangement for 1998 and 1999 with the CMTA exceeding the $150,000 goal again to receive the $50,000 matching grant from the Armington Foundation. In the four years of the Challenge, CMTA members gave nearly $550,000 for CMT research: a tremendous accomplishment!

From its inception, the Armington Research Challenge created a powerful context for organizational growth and development. The Challenge was intended to significantly increase funds for CMT research. It did that and more, spurring long-time members and new supporters to not only make generous financial commitments, but also to offer their service and passion to the cause. A wonderful example of this is the initiative demonstrated by California member Lynn Upton. Lynn embraced the idea that everyone can play a role in raising money and generating awareness. Her personal appeal and the newspaper article it inspired appear on page 4.

This final year of the Armington Research Challenge signals a defining moment for the CMTA and potentially for the future pace and direction of CMT research. To effectively promote and "guide" CMT research, the CMTA need not be the biggest funding source. Certainly though, to establish a strong voice and to achieve influence in the research process, funding research grants is necessary. The Armington Challenge began the process whereby the CMTA has developed its leadership capacity in this respect.

This final year of the Armington Challenge signals a defining moment for the CMTA and potentially for the future pace and direction of CMT research.

By virtue of its national scope and the growing use of the Internet worldwide, the CMTA will continue to gain international exposure and influence. This could—in fact should—lead to the CMTA becoming both the world's largest CMT membership organization and quite possibly its largest CMT patient database.

Moving forward, the unique and expanding role of the CMTA is evidenced not only by having successfully met the Challenge, but also by its sponsorship of the Third International Research Conference and the Research Survey and Evaluation Project on CMT Disorders. The survey and evaluation project, with its goal of assembling a "comprehensive inventory" of all current CMT research efforts and an analysis of same, will create a knowledge base for making informed grant decisions. From the project's findings and recommendations and from the expert advice of its Medical Advisory Board, the CMTA will be uniquely positioned to facilitate and promote the power of interdisciplinary research collaboration.

It is difficult to measure the effect that the Armington Challenge has had on the CMTA, particularly since unintended and/or tangential benefits have been manifested along the way. One viewpoint was expressed by John Chernega, reflecting on the perspective held by him and his wife Joan. He said, "I was very impressed by the commitment made by the Armington Family; it motivated us to do something meaningful to help...." Perhaps for all members who gave in response to the Armington Challenge, that sums it up best.

A Salute to Donors!

The CMTA supporters listed below contributed to the successful completion of the fourth and final year of the Armington Research Challenge. We gratefully acknowledge all contributors who helped the CMTA raise $150,000 this year and more than $500,000 overall through the challenge.

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The Chernega Family of State College, PA; committed to the CMTA and CMT research. From left, Joan, Jill Rosenfeld, Jim, Jack, and John. Not pictured: Joy Spiekermann.

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Ashford Blast for Charity Raises $300 for Armington Challenge

In August of 1998, the Knuth family helped organize the first-ever Ashford Parade of vehicles over 20 years old through the streets of the little community of Ashford, Wisconsin. The parade was held in combination with challenge games of golf, horseshoes, and darts. Charitable donations were collected throughout the day and resulted in gifts of $300 being made to the Cancer Foundation and to the Charcot-Marie-Tooth Association.

The Ashford Blast was truly a community event, with donations of money and door prizes being made by area businesses. Friends and family members participated by serving refreshments and staffing the game booths.

The gift to the Armington Challenge was made in honor of Megan Knuth. A second Blast and Parade is scheduled for August 21, 1999.

Right: A parade of old cars and tractors wound its way through the streets of Ashford, Wisconsin, as part of the Ashford Blast, an event designed to raise money for several charities.

Making Strides Before it's Too Late

Local Resident Raising Funds to Fight Degenerative Muscle Disorder

By Brian Simon

For Lynn Upton, every step is just a little more difficult than the last. The 37-year-old El Segundo resident and mother of two boys has a disabling condition known as CMT (formally known as Charcot-Marie-Tooth—named after the three doctors who discovered it in the early part of the century). Though not life-threatening, CMT is a progressively degenerative neuropathy that affects the peripheral nervous system.

While Upton is still able to walk on her own, her muscles are starting to give out sooner. That is the unfortunate nature of CMT. The condition is actually hereditary and afflicts approximately 150,000 Americans.

Lynn Upton

"CMT is a deterioration of the peripheral nerves that control sensory information and muscle functions of the foot and lower leg area and also the hands and forearms," explains Upton. The condition is also known as Perennial Muscular Atrophy. Symptoms include a foot drop walking gait, high arches, hammertoes, balance problems, muscle cramping, fatigue, poor reflexes and scoliosis.

Upton was originally diagnosed with CMT at the age of five. She was always active growing up and has tried to keep fit over the years. Though she has avoided major complications thus far, the disorder is beginning to take its toll. Once an avid walker who took a daily stroll during her lunch hour, Upton is now tired more often and has switched to the slightly less taxing bicycle. "When I walked, it used to feel good, but now it's like I have heavy feet," she says.

It is a bit tricky to chart a course for Upton's future, but her own mother may be a prime indicator. "She has it (CMT) too," says Upton. "She uses a scooter to get around because she can't walk anymore. Her hands are also weakening. Big movements like using the telephone are okay, but writing is almost impossible." It is still unknown at this juncture whether or not either of Upton's two young boys have CMT. However when one parent has it, there is a 50% chance of passing it on to each child.

CMT affects people in varying degrees. Some need wheelchairs while others may not even get diagnosed. Still, the results are often debilitating. Given the progression of her symptoms, the future doesn't bode well for Upton. Unfortunately, not much is available medically. "I've had a few leg braces made, but they're extremely uncomfortable," she admits. Physical therapy and exercise have been the primary courses of action and there is no medication to treat the malady.

Of course, finding a cure entails research and it is difficult to conduct research without money. The sobering fact is that CMT is not well known and pales in the spotlight compared with much more high-profile conditions such as Multiple Sclerosis and Chronic Fatigue Syndrome. Fortunately, there is one organization that is dedicated to finding a cure and has launched fund-raising efforts on behalf of developing research on CMT. The Charcot-Marie-Tooth Association (CMTA) has recently asked its members to step up efforts on the local level to raise dollars.

Upton is reaching out to her El Segundo neighbors and asking for their support with a tax-deductible donation to CMTA. She has gotten the ball rolling by pledging $100 of her own money.

"I want to educate people about CMT and raise awareness," she states. "We are also looking for a celebrity who has CMT that can act as a spokesperson," she states.

Upton's goal is to raise $10,000 by the end of June and she hopes to eventually coordinate an annual event, such as a 10K. Another objective is to locate any local or South Bay-area residents who suffer from the same condition.

To make a donation, please send your check payable to CMTA to: Lynn C. Upton Fundraiser c/o CMTA; 601 Upland Avenue, Upland, PA 19015-2494. Donations may also be made on-line. For more information, visit the website at www.charcot-marie-tooth.org. •

RAISING AWARENESS AND RAISING FUNDS... A RESPONSE TO THE ARMINGTON CHALLENGE

Dear CMTA,

My name is Lynn Upton and I have been reading your newsletter for years. Thank you so much for making the newsletter possible. It's been so helpful in many ways to me personally. I have also been a contributing member for several years and give what I can to the organization. I have been moved recently by the Armington Challenge and particularly by the efforts of the DiCara family. I have been moved enough so as to reach out to my own community. I took a letter to a local newspaper here in El Segundo, CA, and they decided to write an article about me. I hope it will motivate people to send in donations. I will continue to educate and bring forth awareness. I know it's a small first step, but I think I'm headed in the right direction...and who knows, maybe someday I'll be off and running (pun intended!). Please feel free to print this letter and the article. I hope it will help to motivate others to step forward.

Sincerely,

Lynn (email:lcupton@west.raytheon.com)

MAKING NEWS:

CMTA Members in the Press

Several CMTA members were featured in news stories in the past few months. In some cases the person actively solicited the coverage and in some cases the columnists simply found a good story irresistible. Each of the persons interviewed made certain that the phone number of the CMTA appeared in print with her article. We thank these featured "stars" for their work in spreading the news about CMT.

Krista Hall, R.N., was featured in *Pulsebeat*, a publication of the Frederick Memorial Healthcare System. Krista reported that she had a history of frequent ankle sprains as a child, but that it was attributed to her active nature. She told the columnist that she wasn't sitting and cutting out paper dolls, but was jumping out of trees and off grapevines. As she got older, she had trouble balancing on her heels and an orthopaedic doctor in New York City referred her to a neurologist, who did nerve conduction studies and diagnosed her condition. As Krista notes in the article, she was lucky that her CMT was diagnosed back then and she eventually found an article in a women's magazine that gave the CMTA's 800 number. She also eventually served on the Board of the CMTA as recording secretary and now serves as liaison between the various support groups in the United States and the Association.

The Journal News of Westchester County, NY, runs a column called "Doing the Right Thing" and they chose to write about **Kay Flynn**, the long-time leader of the Westchester County CMTA Support Group. Kay reported that she fell constantly as a child. Her father took her to the family doctor, who taped her ankles. She continued to fall because she had no balance. It wasn't until after she married that her CMT was diagnosed. One of Kay's grandnephews already uses a wheelchair and respirator, demonstrating the wide variety of symptoms even within the same family. As Kay says, "So many of the young ones are worse at their age than I was. They're using canes. Some can't walk four or five blocks. And they're so young!" Kay has run a monthly CMT support group for the past ten years at Blythedale Children's Hospital in Valhalla, NY. She's had people come to the meeting who are 60 years old and never knew they had the problem. These people say, " I finally can put a name on my problem and know I'm not the only one." That means so much.

Another support group leader, **Libby Bond**, of Arkansas, was featured in the *Northwest Arkansas Times* in an article entitled, "CMT, a common disorder that is difficult to diagnose." Some excerpts from the article follow:

"As a child, Libby Bond knew she was different. She walked awkwardly, couldn't ride a bicycle, wasn't able to hold a pencil the "right" way. She had problems with her bladder control, so she "just learned to watch the clock and knew when to go." She remembers her mother telling her that as an infant, she would shake and she recalls her legs trembling for no apparent reason. She could never keep pace with other children. Compensating for her differences was simply a way of life for Bond. An admitted overachiever, she simply thought that if she worked hard enough, she should be able to keep up."

Libby's search for a diagnosis began in earnest in 1997 when she went for a routine physical and asked for some medication for what she thought was arthritis in her hands. The doctor told her it was definitely not arthritis based on the muscle-wasting. Unfortunately, he didn't uncover the real problem. Finally, a caring friend sent her to a specialist who referred her to the Mayo Clinic in Rochester, MN, and there a name was finally given to her problem.

When Bond heard the diagnosis, she smiled and said, "Thank you. You've just given me my life back...With this, I can cope. Disabling, yes, but life over, no. Not by a long shot."

Unable to work any longer, Bond must conserve her energy as best she can. The energy she does have goes first to her family and second to a support group she has started for people and their families who are affected in some way by CMT. She's especially concerned that the disorder is so little known. It took her years to discover the origin of her symptoms, and she wants to work to educate the public and medical community—particularly since there's a list of drugs and vitamins that can make CMT worse. With CMT often misdiagnosed, this is of real concern to Bond.

OF INTEREST

To better understand the issues facing people with genetic conditions, Patricia Foote, an Alliance of Genetic Support Groups member, is asking for Alliance member participation in a survey. People interested in participating can fill out the survey online at www.hometown.aol.com/ManageMed/gensurvey.html
No identifying information is requested. Survey results will form the core of an article about the psychosocial impact of genetic disorders and will be made available to consumer educators.

Want Comfortable AFOs? Tell Your Orthotist Exactly How You

By DANA SCHWERTFEGER

Each of us is different and has different needs. Your AFOs have to be not just custom-molded, but custom-fitted, and they should be as comfortable as a pair of handmade shoes.

When I recently relocated, one thing I didn't take into account was that I'd be leaving behind an orthotist who made comfortable AFOs. Finding a new orthotist isn't difficult, of course, but getting him or her to make AFOs that are truly comfortable is not always an easy process.

My first experience with an orthotist, for example, was not at all pleasant. She seemed very professional when it came to casting my feet and making the AFOs, but suddenly I found myself standing on hard plastic with my feet crammed into the same ankle-height shoes I'd been wearing without the AFOs inside. I managed to walk out of her office and get halfway home before the agony truly registered. The balls of my feet were on fire and my toes hurt more than ever before. I thought, "This is an improvement? I'll never be able to walk wearing these."

Being something of a "fixer-upper," the first thing I did was remove the insoles from the shoes and put them between my feet and the AFOs. Buying shoes a full size and width larger also helped ease the pressure on my hammertoes, but something still wasn't quite right. Whenever I walked a mile or two or wore the AFOs all day, I noticed that they cut into my ankles and caused painful blisters, and the back of my heels became calloused and sore from rubbing against the plastic. Among other things, the orthotist had not allowed for insoles when casting and molding the AFOs, nor had the trim lines around the tibia and fibula—the leg bones that join on either side of the ankle—been cut to allow for much movement or flexibility.

When I returned to the orthotist several days later, rather than cut the trim lines deeper, she widened them by heating the AFOs and flaring them outward. This took the pressure off my ankles without weakening the AFOs. Another thing I asked her to do was make a shallow depression right under the balls of my feet to take the pressure off the pads right behind my little toes. For some reason, orthotists make the plate of the AFO flat and do not mold it to the contours of the bottom of the foot. So, even with insoles, the pads under the fifth metatarsal become calloused and hurt if I walk or stand for any length of time. As for the pressure on my heels, I cut two strips of PPT about half an inch wide and two inches long and taped them inside the AFOs right behind my heels.

The navicular—the bone located midway in the arch of the foot—is another pressure point that may become irritated by walking or standing. If you have high arches, and most people with CMT do, your AFOs should be molded to provide support through the arches, but you need to keep the navicular bones from pressing against the hard plastic. I ask orthotists to accommodate them by flaring the arches out just a little and then I tape pieces of PPT there, too.

As for insoles, orthotists sometimes glue PPT or a similar material directly to the AFOs. I ask not to have this done. Rarely do insoles made of PPT or any other material retain their elasticity for the life of the AFOs, plus my insoles wear away under my big toes after several months, so when I need to replace them, I can simply throw them away and tape on a new pair. Also, when I received my latest AFOs, the white material that had been glued to them did not provide enough cushioning. I wound up scraping it off and using my own insoles from a pair of sneakers, which I secure to the AFOs with masking tape so they don't slip around.

You'll have to do your own experimentation to find what works best for you. I prefer using insoles made from PPT, but I've also found that the insoles from my New Balance sneakers are very comfortable. Insoles meant to provide extra cushioning generally aren't enough by themselves, and putting them in on top of another

OF INTEREST

Cold Feet?
Wooly Warmers offers fleece-lined socks and knee warmers. Call for a brochure. 1-888-339-6659 or access their website at: www.execulink.com/~ggubbels/wooly.html

Want Them Made

insole makes it too bulky. You still have to get all this inside a shoe or sneaker.

I've also learned to ask the orthotist to pad the straps that go across my shins and to drill holes in the AFOs where they come in contact with my calves. I wear calf-high socks to keep the plastic from touching my skin, and the holes allow some air to circulate. That's really important when the weather is hot; otherwise, my lower legs sweat heavily and become covered with heat rash.

One last thing I sometime do is trim back some excess plastic if the orthotist extends the AFOs too far under my toes. Ideally, I like to have the plastic end just short of my toes. I don't need the support and it's actually more comfortable...but that's me. You have to be the judge of how much support you need. I strongly caution you to think carefully before taking a rasp or hacksaw in hand. It's very easy to ruin a very expensive set of AFOs. Trust me; I know. But, if you need to trim, do so in very small increments.

The better practice, and the easier one, is to get your orthotist to make your AFOs to your personal specifications. I've had AFOs made a number of times since that first fiasco, and now I'm always careful to describe exactly what modifications I'd like made to the AFOs. One orthotist, in fact, became so good at molding and cutting the trim lines that I could wear the AFOs just as he made them. Others seem to have their own concept of what AFOs should be, and that's how they make them.

The last orthotist I saw was just such a man. To him and others like him, I would say, "Why won't you listen to me when I describe what I need done in order for the AFOs to be comfortable?" I'm the one, after all, who has to wear them and walk on them day after day. If I don't know what I need and what will be comfortable, who does?

You'll probably have varying degrees of success with your orthotists, but you must remember to tell them exactly what you want and need. Each of us is different and has different needs. Your AFOs have to be not just custom-molded, but custom-fitted, and they should be as comfortable as a pair of handmade shoes.

BOOK REVIEW

Numb Toes and Aching Soles: Coping with Peripheral Neuropathy

By JOHN A. SENNEFF

(Editor's Note: I had the good fortune to receive a complimentary copy of this book following a conversation with the author one afternoon several months ago. I believe it was one of those simple pieces of unexplained luck that occasionally befall us, because this is probably one of the most useful books a person working with Charcot-Marie-Tooth disorders, or dealing with it as a patient, might be able to access.)

The book *Numb Toes and Aching Soles: Coping with Peripheral Neuropathy* was not written with the Charcot-Marie-Tooth patient in mind; however, it couldn't have been a more important text had that been its purpose. What distinguishes this text from any other I've ever seen is the sheer magnitude of treatments that it discusses. Not only conventional pain medications, but alternative therapies, nutrients, and experimental drugs are discussed. Not every one of them is applicable to a CMT patient, but so many are that this will be a desk reference for me from now on. Even the newest popular solutions, like magnetic devices, acupuncture, and vitamin therapies, are discussed at length.

One of the interesting devices that the author uses to discuss each drug and therapy is to include quotes from patients who have used that particular treatment. In some cases, the comments are glowing endorsements; in many cases, they are stories of frustration with time and money ill-spent. Above all, what the reader gathers as he/she moves through the book is that Mr. Senneff has done an amazingly fair and studious job of presenting all the possible options for treatment without personally promoting any one of them.

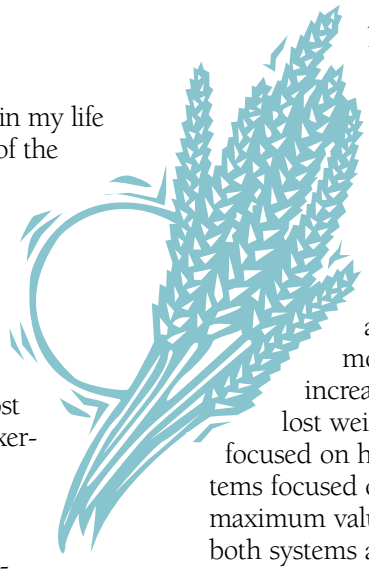
An impressive list of neurologists who are experts in peripheral neuropathy reviewed the material and assisted in reviewing the drafts of the text. They are mentioned in the acknowledgment section of the book. Although Charcot-Marie-Tooth receives only a brief mention in the first section of the book under "Types," the information throughout the text is relevant to anyone with peripheral neuropathy. If you want to know about nerve conduction tests or electromyography or even nerve biopsies, you will find the information there.

Still, the distinction that this book has over others is the amount of space and detail given to a discussion of pain medications and other treatments. Have you been prescribed Elavil, Neurotin, Ultram, Dilantin, Klonopin, or Tegretol? A section of the book is devoted to each of those drugs which are commonly prescribed for CMT patients. Want to compare the effect the drug has on you with the effect that others experience? That is in the book, too. Certainly there are sections of the book specific to diabetes or HIV-induced peripheral neuropathy that will not be very relevant to a CMT patient, but in general the book is a very thorough resource for anyone coping with CMT and the attendant pain.

The book is available from Med Press at 1-888-MED-9898. The cost is $19.95 for the soft bound version and $29.95 for the case bound. There is a $4.00 shipping and handling fee.

A Journey Toward Wellness, Part VI:

By CYNTHIA GRACEY



I have noticed that during the times in my life that I have been overweight many of the symptoms that I attribute to CMT worsen. I notice how much more I stumble, how I tire so much more quickly. I notice how much harder it is to go upstairs, to get out of a chair, to carry groceries or even a purse or a briefcase. With my legs and arms moving like molasses, I most certainly do not have the energy to exercise. When I was pregnant, I really noticed how much harder it was to walk with an extra 10, then 30, then 65 pounds on my five-foot-five small-boned frame. It has been eight years since I lost all the extra weight I gained. Now if an extra five pounds goes on, I notice it immediately.

Recently a friend with CMT told me that each time you take a step you are carrying 20% of your total body weight on the weight-bearing foot. No wonder I was even more tired than usual! No wonder I had no energy by mid-day! No wonder that leaving the house became an effort! I was carrying around an extra bowling ball of weight every second of every day.

Unfortunately, weight issues are often common among the people that I know who have CMT. This should not be surprising, as moving and normal exercise activities can be a challenge. Additionally, being overweight is a problem for Americans in general. According to recent statistics, the average American consumes 540 cans of soft drinks each year and 22 lbs. of pizza, and McDonalds sells 2 million pounds of french fries daily. Americans eat 580 hot dogs per second from Memorial Day to Labor Day. We spend $36 billion each year on potato chips and $50 million on Twinkies. The three greatest calorie sources of the American diet are (1) white bread, rolls, crackers, (2) doughnuts, cookies, cakes, and (3) alcohol (7% of U.S. adult population are active alcoholics). We then try to compensate by spending almost $1.4 million on laxatives and over $3.6 million on vitamins EACH DAY! AT ANY GIVEN TIME THERE ARE 48 MILLION AMERICANS ON A DIET.

Please be assured I am not a great fan of "diets." I never met a diet that I liked and that didn't bring up visions of scarcity and deprivation. Having said that, I am a strong believer that everyone can make some simple changes in lifestyle habits that will ensure the optimal weight for each of our unique bodies.

Over the past 20 years I have experienced many different diets and nutritional systems in an endeavor to increase my vitality and experience of well being. Two very different nutritional and lifestyle systems produced the most profound results. Both systems increased my experience of energy and I lost weight with ease. Both systems focused on how much energy I had. Both systems focused on increasing my ability to get the maximum value from the foods I was eating and both systems added some healthy habits to my daily routine. Neither system had me count calories, or eliminate all the foods I love.

Will these systems work the same way for everyone?

The answer is yes and no. First it is very important to know that we are each biologically unique. What this bio-diversity means is that foods that energize one person may not energize another. There are many factors that must be taken into consideration when creating an energizing food plan for oneself. Body type, blood type, specific sensitivities and allergies, the time of day, and the time of year are each important factors to be analyzed. Obviously the breadth of this conversation is well beyond the scope of this article. The important thing to know is that no one dietary system ENERGIZES EVERYONE OR ANYONE ALL THE TIME. Therefore, it is essential to tune in, to listen to your body, to pay attention to what enhances and what disturbs your energy.

So What Are These Systems?

One is based on principles of Ayurveda, the ancient medicinal system from India. The first step in the Ayurvedic procedure is to answer an extensive questionnaire, and then have a diagnosis and a determination as to your body type. I was told that I had a difficulty absorbing the foods I was eating and some very simple food substitutions were recommended. By eating organic white basmati rice instead of the so-called "healthy" brown rice I had been eating, by switching to refined sugar instead of honey, by taking small amounts of food-based vitamins several times over the day rather than mega nutrients, and doing some of the Indian body care systems, I lost 30 pounds in over three

OF INTEREST

The Exer-Twist

The Exer-Twist equally and simultaneously exercises the left- and right-side muscles of the user's arms, shoulders, and chest.

It is designed to help increase wrist and joint circulation and agility. It conditions and helps warm up wrist muscles that are essential in most racquet sports, and allows people who are otherwise unable to be physically active to exercise upper-body muscles.

The device is operated by a simple twist, like wringing out a wet towel. Resistance can be adjusted for progressive, more difficult exercises, and snap-on grips are available for larger hands or hands that can't easily close around the smaller grips.

Additional product information and an Ableware Homecare Catalog are available from the manufacturers: Maddak Inc., 6 Industrial Rd., Pequannock, NJ 07440-1933. Their number is 973-628-7600 or they can be emailed at: custservice @maddak.com

"Optimal Weight Is a Necessity!"

months and increased my feeling of energy. This system requires finding a qualified practitioner of Ayurvedic medicine.

The other lifestyle and nutritional system was developed by a group of scientists, doctors and nutritionists for the purpose of increasing vitality, mental clarity, and longevity. A secondary result was weight loss. This system required more focus and was more time- and energy-intensive. I have never regretted any of the time I put into it. I have been more than paid back by how I feel every time I reinstate some of the good habits this system encourages. The philosophy of this system is to feed your body the most easily absorbable and highest grades of foods and nutrients possible. The theory is that our body is much like a Ferrari that operates on high-quality fuels. However, if someone poured sand in the Ferrari's engine it would not run regardless of what kind of gasoline you put in the car. Like the sand-ruined engine, our bodies have been fed foods and exposed to environmental pollutants that "clog our engines." As a result, it is necessary to simultaneously focus on increasing intake of densely nutritious foods and supplements, internal and external cleansing processes, and gentle exercise.

Keep a Journal

A friend of mine, Minx, believes that the single most important action you can take is to get a journal and keep track of what you eat and how you feel as a result of your food choices. She believes that by paying serious and consistent attention to your energy levels and how they correlate with your food choices, you will come a long way toward maximizing the energizing effect that food can have on your body.

I, however, have never been able to keep a food journal no matter how hard I have tried. What I can do is notice how I feel after a meal. Am I energized or ready to take a nap? With this in mind after 40-odd years of experience I know what to eat and when. Regardless of the system that works for you, start paying attention to the immediate energy results of your food choices.

A Word About Water

Our bodies are 65-75% water by weight. Water is involved in nearly every body process—digestion, absorption, circulation (transport of nutrients), and elimination. Without proper hydration muscle cells cannot carry on the energy-producing chemical reactions which keep us going. Therefore, it makes great sense that a lack of water is a major contributor to fatigue, poor physical performance, and even illness. Drink one quart for every 1000 calories consumed OR between 1 and 2 quarts per day. The caveat here is that drinking excessive water beyond this amount can put undue stress on the kidneys. Also, if too much water is consumed at mealtime, hydrochloric acid (necessary for the digestion of protein) and digestive enzymes may become diluted.

An easy way to keep track of how much water you are ingesting is to fill two 36-oz. plastic bottles with water each morning. When they are empty you are within range of having enough, depending on the amount of exercise you have done, the amount of protein you have consumed, the amount of alcohol you have had, etc.

A topic of this magnitude requires serious and lifelong focus. If you are seriously seeking to increase your experience of well being, enhance the energy available to you, and attain your optimal weight, I hope that this article will pique your interest and inspire you to explore further.

WHAT TO EAT

Foods that you digest easily. We are NOT only what we eat but also, more importantly, what we absorb and assimilate.

Foods that are as fresh as possible because their vital life force (or *prana*) is most intact. These include foods that are locally grown and in season because (1) they are part of your specific ecosystem and (2) the sooner they go from harvesting to your plate, the more nutrients they retain.

Foods of varied colors, flavors, and textures to stimulate your palate and to add variety and balance to your meals. Foods that are "close to nature," which means that they are minimally processed and minimally preserved.

"Superfoods," including foods high in anti-oxidants, foods known to contain anti-cancer properties, and foods known to promote longevity based on recent research. Foods that are nutritionally dense (high in micro-nutrients)

"Feel-good foods," including foods from your childhood and ethnic roots. The caveat here is to pay attention and choose only the highest quality, purest, and most nutritionally dense versions available.

Appropriate quantities. Too much of a good thing is not a good thing. This includes water.

Foods at appropriate times. Eating when under stress or just before going to bed is not supportive of one's health. It is important to eat gently when stressed and having CMT adds a certain amount of stress on a constant basis. When we are under stress, adrenaline floods the body, leading to anxiety, agitation, and a sense of emergency. Simultaneously the parasympathetic nervous system (which activates the secretion of saliva and other digestive juices, the movement of the intestines, the functioning of the liver, kidneys, etc.) shuts down so that we cannot properly digest our meal. The equation is simple and basic: more stress to the body = less energy available for digestion. Therefore, when under additional stress, it would be wiser to keep meals simple, choosing readily assimilated fresh fruit and vegetable juices or simple broths and pureed soups.

Three Summer Fellowships Awarded

The Medical Advisory Board of the Charcot-Marie-Tooth Association awarded three $4,000 summer fellowships to: Andrea Robertson, working with Dr. P.K. Thomas at the Royal Free and University College Medical School in London, England; Brooke Tate, working with Dr. Michael Shy at Wayne State University; and Tina Kraljevic, working with Dr. Vern Juel at the University of Virginia.

The study of Andrea Robertson will examine the concept of peripheral nerve regeneration in the Trj mouse (Trembler J—the mouse model for CMT). The abstract of Ms. Robertson's application states: "Type IA Charcot-Marie-Tooth (CMT1A), a demyelinating neuropathy, is due to mutations in the gene for peripheral myelin protein 22 (PMP22), either point mutations, or more commonly to a segmental duplication on chromosome 17p11.2 that leads to the presence of an extra copy of the PMP22 gene. The Trembler and Trembler-J mouse strains also display a demylinating neuropathy resulting from mutations in the PMP22 gene. Although CMT1A is a demyelinating neuropathy, the disability results from axonal loss rather than the demyelination. The explanation for axonal loss is so far unknown. Our previous studies in the Trembler-J mouse suggest that there are abnormalities in Schwann cell/axon and Schwann cell/extracellular matrix adhesion. We now wish to investigate this by stressing peripheral nerve by producing nerve degeneration and examining the changes during regeneration, including the capacity for axonal growth."

The study by Brooke Tate is entitled "An Analysis of Clinical Phenotypes in CMT2 and CMTX." CMTX, a demyelinating form of CMT, is caused by point mutations in the connexin 32 (Cx32) gene on the X chromosome, and accounts for up to 16% of CMT cases. Over 130 different mutations have been identified in the Cx32 gene, which produces a wide spectrum of neuromuscular signs and symptoms. CMT2, an axonal form of CMT, is caused by at

Out and About: The CMTA at Confer-

By ANN LEE BEYER, Chairman of the Board

This has been a busy spring for the CMTA, staffing a booth at the Second World Congress in Neurological Rehabilitation in Toronto, Canada, from April 14-17, then moving across town to the American Academy of Neurology (AAN) meetings the following week. Finally, the spring meetings ended with a patient/family conference at the Texas Scottish Rite Hospital in Dallas, Texas, attended by CMTA members from Texas and Oklahoma, as well as members of the local Dallas/Ft. Worth support group.

Support Group leaders Shari Clark and Greta Lindsey were joined at the registration table in Dallas by conference organizer Dr. Susan Iannaccone (standing).

We were delighted to represent the CMTA with a booth at the Neurological Rehabilitation Congress and were very heartened to see that CMT was on the agenda. One of the many positive results of our Third International Conference on CMT Disorders, held in Canada last October, is an increased awareness of and interest in CMT. This became evident by the number of clinicians and researchers who stopped by our booth to chat, ask questions, take literature, and place their names on our mailing list.

At the AAN meetings, not only was CMT mentioned in the opening address, but four posters on CMT research were presented during the Poster Sessions: three from Wayne State University in Detroit, Michigan, and one from the Universtiy of Utah. The titles of these posters were: "Electrodiagnostic Findings in CMTX: A Disorder of the Schwann Cell and Peripheral Nerve Myelin," "Development of a GDNF (Glial-Cell Derived Growth Factor) Expressing Recombinant Adenovirus to Treat Inherited Demyelinating Peripheral Neuropathies," "CMT1A Phenotype Correlates with Secondary Axonal Degeneration but Not Demyelination," (all from Wayne State), and "Proximal and Distal

least two as yet unidentified genetic mutations and accounts for 10-20% of CMT cases. Although there have been previous attempts to characterize the spectrum of clinical disabilities in patients with CMTX and CMT2, many questions about these disorders remain. Dr. Shy writes, "We have evaluated a large number of patients with CMTX and CMT2 at our CMT clinic at Wayne State University. As part of their evaluation, patients have undergone nerve conduction testing, quantitative motor and sensory testing, and motor unit analysis." Brooke Tate will compile the data that have been obtained to permit publishing the data in manuscript form. Results from this project will enable doctors to perform a thorough clinical comparison of the clinical phenotype in CMT1A, CMT2, and CMTX. Her studies will also provide a baseline for longitudinal studies in CMTX and CMT2 which will provide a baseline for clinical trials in these patients.

The final recipient, Tina Kraljevic, will be working on a project entitled, "Pregnancy and Exacerbation of Peripheral Neuropathy in Charcot-Marie-Tooth Disease." Her study follows the printing of a pregnancy survey by Dr. Ted Burns, published in *The CMTA Report*. Significant worsening of peripheral neuropathy in CMT with pregnancy may be relatively common, as was suggested by anecdotal experience at the University of Virginia, in limited medical literature, and in the preliminary findings of the questionnaire sponsored by the CMTA. Ms. Kraljevic's study will evaluate responses of women with CMT to a printed questionnaire to determine how frequently CMT polyneuropathy is worsened by or presents during pregnancy. To address sampling bias and to provide matched controls, a telephone interview will be performed with randomly chosen women with CMT followed in the University of Virginia Neuromuscular Clinics. If the study demonstrates frequent pregnancy-related worsening of CMT, the finding may justify prospective studies, provide important information to the obstetrical community and to pregnant and prospectively pregnant women with CMT, and provide insight into physiological mechanisms for this phenomenon.

Each of these studies will conclude by September 1, 1999, and a report on their findings will appear in the newsletter.

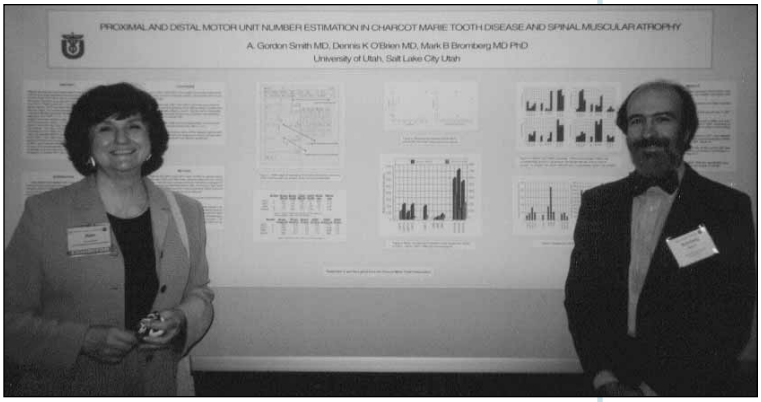
Motor Unit Number Estimation in Patients with Charcot-Marie-Tooth Disease" (University of Utah). There were over 800 poster presentations, and we are very proud that Dr. Agnes Jani, recipient of the Armington and Buuck Fellowships, had her poster chosen as one of the ten best at the conference. Dr. Jani works with Dr. Michael Shy at Wayne State University. Dr. Shy is a member of our Medical Advisory Board and one of the coordinators of the Third International Conference. Another member of our Medical Advisory Board, Dr. Mark Bromberg, presented the poster, "Proximal and Distal Motor Unit Number Estimation in Charcot-Marie-Tooth Disease."

The patient/family conference on CMT at Texas Scottish Rite Children's Hospital in Dallas, Texas, was held on Saturday, May 15, 1999. Dr. Susan Iannaccone hosted the conference and gave the opening presentation on the history of CMT, the CMT children's clinic at Texas Scottish Rite with its 150 children, and the importance of CMT research. Her talk was followed by a creative discussion of genetics by Dr. Gil Wolfe, who actually made inheritance patterns in CMT understandable.

The morning program continued with workshops on nutrition and occupational therapy. One of the most well-received workshops was the one on occupational therapy. Attendees got quite involved and began exchanging coping strategies they use to make living with CMT easier. The discussion was so lively that we plan to introduce a new column in *The CMTA Report* called "Living with CMT," written by our members. We welcome your suggestions.

The afternoon presentations were given by Dr. John Birch on the orthopaedic considerations for CMT and a representative of Athena Diagnostics, who discussed the available DNA tests for CMT. In addition, there was an informative video on how a physical therapist tests muscle strength and weakness, as well as a workshop on the psychological aspects of living and coping with a rare neuromuscular disease.

The program closed with a talk about the significance of the recent Third International Conference on Charcot-Marie-Tooth Disorders, a review of CMTA-supported research, and the importance of the Armington Research Challenge to our basic cause—the treatment and cure of CMT disorders.



Ann Lee Beyer, Chairman of the CMTA Board of Directors, and Mark Bromberg, MD, PhD, pose in front of Dr. Bromberg's poster presentation at the AAN meeting in Toronto, Canada.

CMTA Research Fellowship Report from the University of Miami

By DR. LISA BAUMBACH and MARY ELLEN AHEARN

Mary Ellen Ahearn and Dr. Lisa Baumbach attended the Third International Conference on CMT Disorders last October in Montreal. Both of them have continued their research on CMT Type 1A and the variations in the African-American population.

The extent to which benign (non–disease-causing) genetic variations influence the phenotype (visible physical characteristics) remains an unanswered puzzle. We need to gain a further understanding of the role genetic variation may play in modulating disease presentation, variability and perhaps, thereby, response to therapeutics. A first step toward such a goal is to look for correlation between benign genetic variations and disease genotypes. One disease model to address this question may be the hereditary motor and sensory neuropathies (HMSN, CMT). CMT is clinically heterogeneous and genetically very complex. CMT1A patients may have either a large 1.5-mB duplication in the 17p11.2 region or a point mutation in the PMP-22 gene, but these differences in genotype do not correlate well with differences in disease phenotypes. A novel polymorphism (a genetic variation that differs among people and is thought to be non–disease-causing) in PMP-22 recently described by our lab is a good candidate for an investigation of the effect of a benign mutation on disease phenotype.

This polymorphism is very intriguing, as to date it has only been found in African-Americans. It was first discovered in two individuals who were being tested for the CMT1A duplication. Both patients were youngsters who had shown symptoms of CMT from an early age and both were African-American. One patient had the CMT1A duplication while the other had a novel PMP-22 mutation. In the past year, with the support of the CMTA, we have been fortunate to collect samples from numerous members of a large CMT family (profiled in the Fall 1998 *CMTA Report*).

This family is of special importance to the study as they are a large family with four generations and several affected members. We have collected thirty individuals and are hoping to collect additional family members this summer. We now have samples from a total of seven African-American families and have found the polymorphism in four of four families thus far. These families come from a variety of locations within the United States from the Southeast to the West coast. One of the first patients in which the polymorphism was found is from a Caribbean island, as is one of the newest, untested CMT patients.

To study the frequency of the polymorphism in the African-American population, DNA was obtained from African-American non-CMT subjects from South Florida who represented a mixture of ethnicity and heritage. The polymorphism was found to be present in approximately 35 percent of this control population. It has never been reported in a Caucasian sample. It may be that it was not previously discovered because the vast majority of CMT patients studied are Caucasian. In fact, it has been a very slow process to locate new African-American families for this study.

A number of interesting questions remain to be answered concerning the polymorphism. Does it somehow influence the severity of the disease in those CMT patients who have the polymorphism? Is it a coincidence that the individuals in which the polymorphism was first noted were also the individuals who were significantly affected at an early age? Certainly not all the newer families display this correlation. However, to understand any possible influence of the polymorphism on disease severity, we need a much larger sample size of African-American CMT families. Another future research

continued on page 13

Ask the Doctor

Members of the CMTA's Medical Advisory Board answer questions from readers.

Dear Doctor,

I was in a doctor's office some time ago for an X-ray and evaluation for replacement of my right knee. Since that time, I have had confirmation, via a blood test, that my daughter and granddaughter have CMT 1A. I had EMG testing years ago which revealed some deterioration of the nerves in my legs and CMT was mentioned, although I elected to forego any further testing.

Because of the above-mentioned confirmed diagnosis, I have subscribed to the CMT Association newsletter and am wondering if I should be concerned about the replacement surgery. Are the downsides of replacement greater because of CMT? Does the deterioration of the nerves accelerate? Should I get further testing?

I am really ready to get the knee fixed and just thought I should pursue the information on CMT before I jump out into the frying pan. I would appreciate it if you could advise me if you have knowledge of the hazards involved.

—*B.Y. Texas*

The Doctor Replies:

In the absence of more detailed neurological evaluation, it would be difficult to give meaningful answers. So, the first thing is to get an evaluation by a neurologist who will communicate with your orthopedic surgeon. A general opinion is that if a joint (knee or hip) needs replacement, this should be done early rather than later in a patient with a significant neuropathy (CMT) which will cause ambulatory problems.

As to whether there are downsides to replacement because of CMT, they are slight, but need must be the greatest concern. Will the nerve deteriorate more? With good surgery, this should be no problem. Should the patient get further testing? Absolutely. That should be the first step.

Dear Doctor:

While visiting my gastroenterologist, I was shown a section of a text which listed CMT as causing digestive problems, such as diarrhea and upset stomach. Do you know if intestinal problems are associated with CMT?

The Doctor Replies:

This is a very rare connection, but it is due to involvement of the very tiny nerves called autonomic or vegatative nerves supplying the gastrointestinal tract and viscera (heart, lungs, liver, spleen, kidneys, etc.). Of course, it is more commonly seen as part of diabetic neuropathy and some of the rarer genetic neuropathies such as those related to amyloidosis or leucodystrophy. In my experience, less than 5% (probably 1-3%) have this involvement as part of CMT. However, the patient must see his local neurologist to evaluate any other neurological and especially non-neurological causes.

RESEARCH FELLOWSHIP REPORT

continued from page 12

question is "does the polymorphism exist in other ethnicities who are under-represented in CMT research?"

The other CMT research interest in our lab is genetic anticipation in CMT1A families. Genetic anticipation describes the principle that the age of onset of a disease decreases with successive generations while the degree of disease severity increases with successive generations. This is well documented for several genetic diseases such as Huntington's disease. A pattern of anticipation can be observed in some CMT1A families; our efforts center on how to document the anticipation and then to understand how it is happening. For this, we have been gathering medical records and DNA from a series of families that appear to display anticipation and have the CMT1A duplication. Anticipation studies were the proposed focus of the CMTA Fellowship; however, the possibility of pursuing the African-American polymorphism has resulted in anticipation research taking a back-seat in the past year. We are continuing to pursue both the African-American polymorphism questions and the documentation of anticipation families in the coming months.

I (MEA) thank the CMTA for the opportunity to become involved in this research. I have learned a great deal in the past year. I came to CMT research from a background in the Human Genome Project, which is thrilling in its own way, but does not provide the opportunity to focus research on a single intriguing genetic problem.

CMTA Support Group News

Members of the Kansas City Support Group steering committee gathered at the Kansas City Club for a luncheon hosted by George Owens for visiting Drs. Perelli and Vinci from Italy. Front row: Lester Johnson, Joy Rushfelt and Ardith Fetterolf. Back row: Winifred Johnson, George Owens, Drs. Vinci and Perelli and Lee Ann Bromberg.

California—Los Angeles: Serena Shaffer, Group Leader

This is a brand-new group, which met for the first time on June 26, 1999. They meet at the S.H.A.R.E. Unity Room, 5521 Grosvenor Blvd., Marina Del Rey, CA 90066. Topics for the meeting included: the benefits of exercise, yoga, massage, and Tai Chi, AFO alternatives, and children and CMT Approximately 50 people attended the first meeting. Group leader Serena Shaffer can be reached at 818-841-7763.

California North Coast Counties: Freda Brown, Group Leader

Spring Meeting, May 1, 1999. Gary Fritzsche of Rehab Mobility Services brought a scooter and a power chair (a cross between a scooter and a wheelchair). He discussed customizing equipment for individuals. Also, Deborah Wilde, MS, CPO of Sierra Orthotics, talked about braces for people with CMT. The August 7th meeting will feature an occupational therapist from the MDA Clinic in San Francisco who will demonstrate aids to daily living.

California—San Francisco Bay Area: Ruth Levitan, Group Leader

The San Francisco Bay Area Charcot-Marie-Tooth Support Group celebrated its first anniversary with a party on May 15, 1999. It was a purely "social, sharing, and having fun" time. After the March meeting, which featured a "show and tell" idea-sharing, the decision was made to include that facet of the meeting in all future get-togethers. The group is also working on getting some publicity for the group. Hank Silver and Virginia Drury are placing ads in local papers and contacting the neurology departments of local hospitals.

Florida—Boca Raton to Melbourne: Walter Sawyer, Group Leader

The May meeting of this support group was held in the pool at the Caribbean Shores Resort in Jensen Beach, FL. Sound unusual? The event was a class on aquatic solutions for the person with CMT. The free class demonstrated gentle water exercises with instruction by Deni Gillespie, a COTA/L, who is a member of the support group. The previous meeting in February was conducted by massage therapist Mark Buzzo, who talked about preventing the onset of hammer toes and claw fingers with massage. He also spoke about hot compresses and wax treatments.

Kentucky/Southern Indiana/Southern Ohio: Robert Budde, Group Leader

The Lexington group had another successful meeting in May with 18 people in attendance. The speaker was Mr. James Park, CO/CPed of Central Brace and Prosthetic, Inc. He discussed braces and prosthetics for the person with CMT. In September, the group will meet at the library in Lexington to hear a report from Roger Hopgood, who is taking members' questions with him on a visit to a CMT expert in St. Louis, MO.

Missouri/Eastern Kansas: Ardith Fetterolf, Group Leader

The June 5th meeting of the Kansas City Support Group featured a presentation by Frank DeCastra, PhD, on the topic, "Free to Be Yourself," which was the continuation of a discussion started at the April meeting of the group. Members of the group reported meeting with Dr. Paolo Vinci and Dr. Sandra Perelli, from Rome, Italy, who were in the United States as a follow-up to some work they are doing with orthotic devices for people with CMT. Both doctors attended the Third International Conference on CMT in Montreal, Canada, last October and have continued their interest in working with patients with CMT.

CMTA Support Groups

Alabama/Greater Tennessee Valley
Place: ECM Hospital, Florence, AL
Meeting: Quarterly
Contact: William Porter, 205-767-4181

Arkansas—Northwest Area/ Springdale
Place: Harvey and Bernice Jones Center for Families, Springdale
Meeting: 3rd Saturday of each month
Contact: Libby Bond, 501-795-2318

California—Berkeley Area
Place: West Berkeley Library
Meeting: Quarterly
Contact: Ruth Levitan, 510-524-3506

California—Los Angeles Area
Place: SHARE Unity Room, Marina Del Ray
Contact: Serena Shaffer, 818-841-7763

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

California—Napa Valley
Place: Sierra Vista Convalescent Hospital, Napa
Meeting: Quarterly
Contact: Betty Russell, 707-747-4864

Florida—Boca Raton to Melbourne
Place: Columbia Medical Center, Port St. Lucie
Meeting: Quarterly
Contact: Walter Sawyer, 561-336-8624

Florida—Miami/Ft. Lauderdale
Place: North Broward Medical Center, Pompano Beach, FL
Contact: Al Kent, 954-742-5200 (daytime) or 954-472-3313 (evenings)

Kentucky/Southern Indiana/ Southern Ohio
Place: First United Methodist Church, Lexington, KY
Meeting: Quarterly
Contact: Robert Budde, 606-255-7471

Massachusetts—Boston Area
Place: Lahey-Hitchcock Clinic, Burlington, MA
Meeting: Every other month, the first Tuesday
Contact: David Prince, 978-667-9008

Michigan—Detroit Area
Place: Beaumont Hospital
Meeting: Three times each year
Contact: Suzanne Tarpinian, 313-883-1123

Michigan—Flint
Place: University of Michigan, Health Services
Meeting: Quarterly
Contact: Debbie Newberger/ Brenda Kehoe, 810-762-3456

Minnesota—Benson
Place: St. Mark's Lutheran Church
Meeting: Quarterly
Contact: Rosemary Mills, 320-567-2156

Mississippi/Louisiana
Place: Clinton Library, Clinton, MS
Meeting: Quarterly
Contact: Betty Aultman, 601-825-5626 Julia Provost, 601-825-6482

Missouri/Eastern Kansas
Place: Mid-America Rehab Hospital, Overland Park, KS
Meeting: First Saturday each month except January, July, and September
Contact: Ardith Fetterolf, 816-965-0017, fax: 816-965-9359

Missouri—St. Louis Area
Place: St. Louis University Medical Health Ctr.
Meeting: Quarterly
Contact: Carole Haislip, 314-644-1664

New York (Horseheads)
Place: NYSEG Meeting Room, Rt. 17
Meeting: Quarterly
Contact: Angela Piersimoni, 607-562-8823

New York (Westchester County)/ Connecticut (Fairfield)
Place: Blythedale Hospital
Meeting: Monthly, Saturday
Contact: Kay Flynn, 914-793-4710

North Carolina—Archdale/Triad
Place: Archdale Public Library
Meeting: Quarterly
Contact: Ellen (Nora) Burrows, 336-434-2383

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: Church of the Brethren
Meeting: Fourth Thursday, April-October
Contact: Dot Cain, 937-548-3963

Oregon—Willamette Valley
Place: Brooks Assembly of God Church
Meeting: Monthly
Contact: Regina Porter, 503-591-9412

Texas—Dallas/Ft. Worth
Place: Harris Methodist HEB Hospital
Contact: Greta Lindsey, 817-281-5190 or Shari Clark, 817-543-2068

West Virginia/North Central
Place: VFW Conference Room, Elkins, WV
Meeting: Quarterly
Contact: Joan Plant, 304-636-7152 (evenings)

OF INTEREST

For exercise, some people report good results from walking in water (not "on"). To make this easy, a member wrote to say that she bought an inexpensive above-ground pool and spends some time each summer day walking back and forth in the water. The warm water feels good and the exercise of walking is good for her overall health.

While a great deal of effort has been made to make these lists as accurate as possible, we apologize for any omissions or misspellings that may have occured.

GIFTS WERE MADE TO THE CMTA IN HONOR OF:

Gaelyn Coyle
Mary Ellen Feeney

Gail Feeney-Coyle
Mary Ellen Feeney

Stephanie Di Cara
Mr. and Mrs. William Atkinson
Tod DiCara
Thomas and Denise Herrmann
Richard J. Kosarek
Mary Martin
Paul, Maria and Jennifer Naye
Mr. and Mrs. William Poling

Stephanie DiCara's First Holy Communion
Reno and Julie Lovison

Marilyn Dodge
Loretta Bennett

Jacqueline Donahue
Mr. and Mrs. Peter Donahue
John and Shirley Reilly

Kay Flynn
Mona G. Coogan

Ruth Gelman
Leon Gelman

Krista Hall's Birthday
K. Laurie Terry

Pam and Robert Kleinman's 25th Wedding Anniversary
Robert and Jennifer Beattie
Alyse Beckman and Family
Mr. and Mrs. Donald D. Capelin
Mrs. Estelle D. Capelin
Niles and Nancy Citrin
Bobby and Betsy Furr
Dr. and Mrs. Richard Giddon
Marsha and Ray Greenberger
James and Maria Jackson
Calvin and Denise Kleinman
Richard and Virginia Kleinman
Mr. and Mrs. Mark Levy
Fred Mack
Diane and Joe Sandler
Mrs. Stephanie Weintraub

Megan Knuth
Ashford Blast for Charity

Diane Higgins Kosik
Irma Higgins

Deirdre O'Donnell
Sherri and Steven O'Donnell

Mr. and Mrs. Percy Sand's 60th Wedding Anniversary
Mrs. Judith Goldman

GIFTS WERE MADE TO THE CMTA IN MEMORY OF:

Mary Abruzzo
Barbara Abruzzo

Mrs. Emily Anderson
Mireille Masson

Cleo Belz
Bob Belz

Jane L. Chase
Steven and Cindy Barrett

James R. Conley, Jr.
Cindy Birk Conley

Betty Essick
Mr. and Mrs. Frank Gunnison

Ralph A. Hall
Arthur H. Benedict
Dr. and Mrs. Karl T. Benedict
Karl T. Benedict, Jr. and Patricia Benedict
Leslie, Bill, Jennifer and Nyles Bragman
Phillips S. Davis
Stacy, Mike, Jason, and Jeffrey Kohn
Sidney Miller
Ellen Raye
John and Barbara Solakian
Erin, Randy, Hannah, and Ilana Springer

Rabbi Norman Kleinman-Jackie and Ann Bitensky
Linda and Phil Bloomberg
Mike and Clarice Davis
Jack Goldman
Dan and Debbie Greenberg and Family
Elliott Greenberg
Roslyn S. Greenberg
Mr. and Mrs. Lee S. Hillman
Barbara and Terry Mitchell and Family
Gert and Abe Nutkis
Sharon, Jeff and Caryn Prosansky and Grandma Shirley
Dr. and Mrs. Jerrold Schwartz
Janet Shlaes
Rochelle and Sheldon Wolfe

My husband and brother
Loretta Bennett

Dr. Jeanette Reilly
Dr. and Mrs. Charles DiGiovanna

Edna Robinson
Diana Pearson

Lou Schwertok
Mr. and Mrs. Frank Gunnison

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, 601 Upland Ave., Upland, PA 19015.

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:_____

_____

The Pain Facts...

(Editor's Note: This article contains excerpts from an article entitled, "Conquering Pain" from the March 1, 1999, edition of Business Week.*)*

Americans alone spend some $3 billion a year on over-the-counter analgesics, and a further $750 million on narcotics prescribed for pain. For most sufferers of chronic pain (defined as persistent pain that lasts more than three months), a life free of agony is elusive. The narcotics most commonly prescribed don't work for the millions who suffer from neuropathic pain arising from damage to the nerves, caused by disease, trauma, or chemotherapy. A breeze across the skin can be excruciating and there is no pharmaceutical recourse.

Most frightening, perhaps, is that for all pain victims, the longer they suffer, the more impossible the problem becomes. Continuing pain eventually rewires the nervous system until it becomes even more sensitive to pain and even harder to treat. The American Pain Society estimates that 45% of the population seeks medical help for persistent pain at some point in their lives. Pain costs the US some $100 billion every year, including 515 million work days lost and 40 million doctor visits.

The sufferers are beginning to be heard. In January, the Veterans Affairs Department announced a new effort to reduce pain for its 3.4 million patients. They instructed VA doctors and nurses to assess and record a patient's pain just as they would blood pressure, pulse, and temperature. Last year, Congress allocated $102 million to the National Institutes of Health for pain research in 1999, a 15% jump from 1998.

A new way of thinking about pain is emerging. Researchers think pain should be treated as a disease in itself, divorced to a large degree from the underlying cause. Instead, pain would be classified by the cellular mechanisms that cause the hurt, which may be the same for different diseases and different for the same disease. Once a pain mechanism is identified, doctors could look through some highly targeted drugs for just the right treatment.

Unfortunately, pain is one of the body's most complex biological functions and it's not uniform at all. The pain response can vary by gender, race, and age, making it hard to predict from one person to the next. Even more problematic, pain is completely subjective—each person's physical and emotional tolerance level is different. That's the great unknown in the study of pain.

Researchers think pain should be treated as a disease in itself.

Pain is identified as one of two types: acute pain and chronic pain. Acute pain is sharp and immediate and comes from an injury to tissue, or it can be triggered by bodily malfunction or severe illness. Acute pain follows this path: 1) Nociceptors in the peripheral nerves sense the injury and in response, release chemical messengers. 2) The messengers travel through superfast nerve conduits to the spinal cord where 3) they are passed directly to the thalamus and into the cerebral cortex. 4) The brain identifies the site of the injury and sends a message back down the spinal column telling the muscles to contract and block the pain. The process can take place in the instant your finger touches a hot stove and pulls away.

Chronic pain is persistent, debilitating pain, such as that from a bad back or diseases of the nerves themselves, which takes a more circuitous route. 1) Pain signals enter the dorsal horn of the spinal cord 2) and transfer back and forth between interconnected nerves that modulate the pain message as it travels up the spinal cord 3) to the cerebral cortex, which assesses the damage and adjusts emotions and other bodily functions, such as breathing. 4) This slower pathway creates duller, more persistent pain. If the pain persists, the entire nervous system may be reprogrammed to create a lower threshold for pain.

VISIT OUR HOMEPAGE
at
www.charcot-marie-tooth.org
The site was provided through the generosity of
MindSpring

Children's *CMTA Report* to Come

The CMTA is planning on publishing a newsletter devoted to children's issues and ideas. For that reason, we are soliciting articles (or ideas for articles) from or about children, as well as letters and "Dear Doctor" questions. We'd be interested in any questions you might have for pediatric neurologists or orthopedic surgeons. If you submit an article, please include a picture for publication. Articles, letters, and pictures should be sent to CMTA, 601 Upland Ave., Upland, PA 19015 or emailed to CMTAssoc@aol.com

WRITE TO US!

Pat Dreibelbis, Editor

The CMTA Report

CMTA

601 Upland Ave.

Upland, PA 19015

The CMTA reserves the right to edit letters for space.

Letters to the Editor:

Dear CMTA,

I was diagnosed with CMT at the age of 28. I am now 30 years old and have diminished sensitivity and poor circulation. Just these past few years, I've had worsening infections, affecting the bone and tissue in my left foot. I have required multiple tissue debridement and bone resections. I'm looking for anyone that has CMT, that I could correspond with, between the ages of 20 and 35.

If you are interested in corresponding, please write to Tammy Sawyer, PO Box 1483, Fort Gibson, Oklahoma 74434.

Dear CMTA,

For women who need wide or extra-wide shoes and don't mind shopping out of a catalog, call 1-800-362-8400, Arizona Mail Order Company. Ask for the catalogs of Old Pueblo Traders and Coward. Old Pueblo Traders sells both clothes and shoes, while Coward sells only shoes. Both catalogs handle lots of brand names. I hope this helps someone else.

—E.K.

Dear CMTA,

It has been my misfortune to be born with a genetic, progressive neuro-muscular disease that has left me completely disabled at the age of 46. In the past 25 years, I have enjoyed a very successful career in my chosen profession. I have been able to cope with the physical changes as well as the greatly reduced physical activity. But, something has happened to the easy-going, very happy and positive attitude I have enjoyed over the years. I have become angry and bitter.

I have noticed over the past 1 1/2 years on public assistance that people in my situation seem to be angry and display a bad attitude. I thought they had a problem they needed to correct. I thought they should have control over their emotions. I have since learned differently.

There are people and organizations that the disabled have to deal with every day. These people have an extreme amount of control over our lives. Some of the people who have the power over us have contempt and disrespect for our physical and emotional needs and sensitivities. We can hear the disdain in their voices.

I would like to ask those people in control of our lives to stop and think for a minute. Think what it might feel like to be told "We can't find your check' or "public assistance doesn't cover clinical pain management."

I feel a huge loss of control over my life now. I feel helpless and angry. It is hard enough dealing with a terrible disease and then experiencing the mental abuse by those who probably don't even realize how damaging their words can be.

—C.L. Spokane, WA

Dear CMTA,

I want to thank you for all your wonderful Reports! I look forward to receiving them. I am not able to send the full $35 for dues, but I am sending $20 and hope to remain on your mailing list.

I have been diagnosed with CMT although the genetic test came back negative. My neurologist is certain I have it, especially since he did the nerve conduction velocity studies.

It is interesting that CMT causes deterioration of the nerves and yet causes so much pain and discomfort. I rarely have a complete night's sleep due to the pain and tingling in my feet. It seems like the doctor has tried all kinds of medication and now I'm taking Tegretol, 2 to 3 times a day along with Elavil at bedtime and Klonopin at bedtime. This is after they tried Vicodin and Robaxin! To say the least, I feel like a zombie all day and am tired of feeling this way.

To my amazement, I find it interesting how limited the knowledge of some doctors is. I started taking the CMTA Report to my doctor and he was delighted with the new insight. It just makes me wonder how many doctors are treating a disease they don't know about or don't know how to treat!

I am a 50-year-old woman, mother of three. As of today's date, only my youngest (age 21) has been diagnosed with CMT and underwent very successful surgery about 8 years ago. I'm hoping to gain more information that I can share with my neurologist so I can begin to feel human again.

—C.M. Whittier, CA

Dear CMTA,

Recently, I purchased "Step Stretch," a new product designed by Prism Technology. It comes in a box the size of a shoe box, costs under $30, and is extremely easy to use. "Step Stretch" has a foot rocker shape and provides me with a great lower extremity stretch. I have noted an improvement in my balance and flexibility. I'm 45 and think the product could probably benefit any age group. I purchased the product locally,

but it should be available from the manufacturer at Prism Technologies, San Antonio, TX, 210-520-8051.

—B.B. Emmaus, PA

Dear CMTA,

I was reading a report on human growth hormones and thought you might be interested in a small section related to CMT. My husband, 82 years old, has had CMT since age 35. I thought this information was important, so I'm sending it on to you.

"The implications of this work for helping people is nothing short of mind-boggling. If IGF-1 can regenerate spinal cord motor neurons, it maybe be useful in treating amyotrophic lateral sclerosis (ALS), a devastating disease in which the loss of spinal cord and cortical motor neurons results in complete paralysis and death. It may also be useful for peripheral neuropathies, such as Charcot-Marie-Tooth syndrome."

This information comes from a book entitled, "Grow Young with HGH," by Dr. Ronald Klatz.

—V. A. Wurtsboro, NY

Dear CMTA,

I am a 45-year-old married woman with a 16-year-old daughter. I was diagnosed with CMT in my early 20's although symptoms were present from the age of 3. It is possible I have CMT Type 2, but even this is uncertain. I have severe wasting of the hands and feet, and I also have optic neuropathy attributed to CMT.

Another symptom I have is vocal cord paralysis which results in difficulty speaking. The throat muscles are also weakened. This has also been attributed to CMT. However, I was recently told by a professor of neurology that throat involvement has never been a symptom of CMT. My daughter also has the same condition.

What I am looking for, is to hear from anyone else who has CMT and also throat and voice problems.

Thank you.

Patricia, email:kpnuscin@iaccess.com.au

Dear CMTA,

I am a 72-year-old woman who has had CMT Type 2 for over 15 years and am just now finding that I cannot do all of the things I used to enjoy. It has progressed to my upper legs and I have been told that because I seem to be in perfect health in all other ways, I will probably be using "wheels" to get around in later life. Believe it or not, the things I used to enjoy were being independent and doing house and yard work. If I fall now or get down on the ground, I need a yard bench to help me up by using my arms to push me up from my knees. I feel very tired after working like this, so I don't enjoy it anymore.

These are the water shoes with AFOs inserted that show what I use when taking exercise classes in the water and now when showering.

I have been to see Dr. Michael Shy at Wayne State University and I would like to stay with him for my CMT. He is interested and very compassionate. He suggested that I have the blood test to make sure that my type was 2. It showed that I did not have any of the other types of CMT, so Type 2 was confirmed.

My letter and the attached picture is for B. D. from California who wanted to know how people handled showering and bathing. I have a hand-held shower head and use a bath bench. So, I sit down to bathe. I did discover when I wanted to take a swim and exercise class that the beach and water shoes worked well to get me to the pool edge. I found out that my AFOs fit in very easily. The shoes have rubber soles and a stretchy top that comes up high on the instep. I have started to use these in the shower and now enjoy standing up in the shower. I hope this will help B.D. I now wear the shoes right into the pool for the exercise, but AFOs are not good for swimming.

—J.J. Grand Blanc, MI

The CMTA Report is published by the Charcot-Marie-Tooth Association, a registered non-profit 501(C)(3) health organization.

The opinions expressed in the newsletter are not necessarily those of the Charcot-Marie-Tooth Association. The material is presented for educational purposes only and is not meant to diagnose or prescribe. While there is no substitute for professional medical care for CMT disorders, these briefs offer current medical opinion that the reader may use to aid and supplement a doctor's treatment.

MEDICAL ALERT:

These drugs are toxic to the peripheral nervous system and can be harmful to the CMT patient.

Adriamycin
Alcohol
Amiodarone
Chloramphenicol
Cisplatin
Dapsone
Diphenylhydantoin (Dilantin)
Disulfiram (Antabuse)
Glutethimide (Doriden)
Gold
Hydralazine (Apresoline)
Isoniazid (INH)
Megadose of vitamin A*
Megadose of vitamin D*
Megadose of vitamin B6* (Pyridoxine)
Metronidazole (Flagyl)
Nitrofurantoin (Furadantin, Macrodantin)
Nitrous oxide (chronic repeated inhalation)
Penicillin (large IV doses only)
Perhexiline (Pexid)
Taxol
Vincristine

Lithium, Misomidazole, and Zoloft can be used *with caution.*

Before taking any medication, please discuss it fully with your doctor for possible side effects.

*A megadose is defined as ten or more times the recommended daily allowance.

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).
- … 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).
- … does not affect life expectancy.
- … 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, 1X, and HNPP can now be diagnosed by a blood test.
- … is the focus of significant genetic research, bringing us closer to answering the CMT enigma.

The CMTA *Report*

Information on Charcot-Marie-Tooth Disorders from the Charcot-Marie-Tooth Association

CMTA

601 Upland Avenue
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