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The CMTA Report

SUMMER 1997

Vol. 12, No. 3 ISBN #1067-0181

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

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Paul Flynn Named Executive Director, Armingtons Increase Challenge Grant

The CMTA Board of Directors voted unanimously to hire Paul Flynn as the first Executive Director of the organization. Paul served on the CMTA Board as chair of the fundraising committee from August 1995 until April 1997, when he left the board to serve as Development Coordinator for the Third International Research Symposium on Charcot-Marie-Tooth disorders. An interview with Paul appears on page 2. Speaking on behalf of the board, chairman Ed Butchko said, "We have set a very ambitious agenda for the next 2 years. In a short time, Paul has been instrumental in advancing the organization and helping to position the CMTA for accelerated growth. Becoming Executive Director is a natural progression for him and a major step for us."

"We are at a turning point. After many years of doing well, the CMTA is poised to do great things." So began the proposal that persuaded the Armington Family to increase their research challenge. The original challenge required that the CMTA raise \$25,000 in each of 3 years to be matched on a one-to-one basis, thereby generating \$150,000. In years one and two, the CMTA raised more than \$45,000, almost doubling the required minimum. The new terms of the challenge signify a bold commitment by the Board to boost research funding. The Evenor Armington Fund will provide \$50,000 in each of the next 2 years provided that the CMTA matches those dollars on a three-to-one basis that would amount to \$400,000.

The parallel announcement of the selection of

"We are at a turning point. After many years of doing well, the CMTA is poised to do great things."

an executive director and the "new" challenge grant is not coincidental. Board President Diane Freaney explains, "It was within the fundraising committee that Paul and I first discussed approaching the Armingtons about increasing the goal. Paul felt that the overwhelming response by members to fund research warranted, actually demanded, that we at least consider this. It will truly be a challenge, but we are extremely confident that this new goal can be met."

In accepting the proposal, the Armingtons have extended the time frame to 4 years and have committed to an additional \$75,000. Meeting the requirements of the research challenge, as well as funding the organization's operating needs, will necessitate significant involvement from our members. "Funding on this scale is serious business," says Paul. "It will take leadership donors stepping forward, not only with their personal support, but also with a willingness to solicit others."

In the coming months, a Development Steering Committee will be formed to work with Paul on all fundraising programs.

EVENOR ARMINGTON CHALLENGE—\$550,000 FOR RESEARCH!

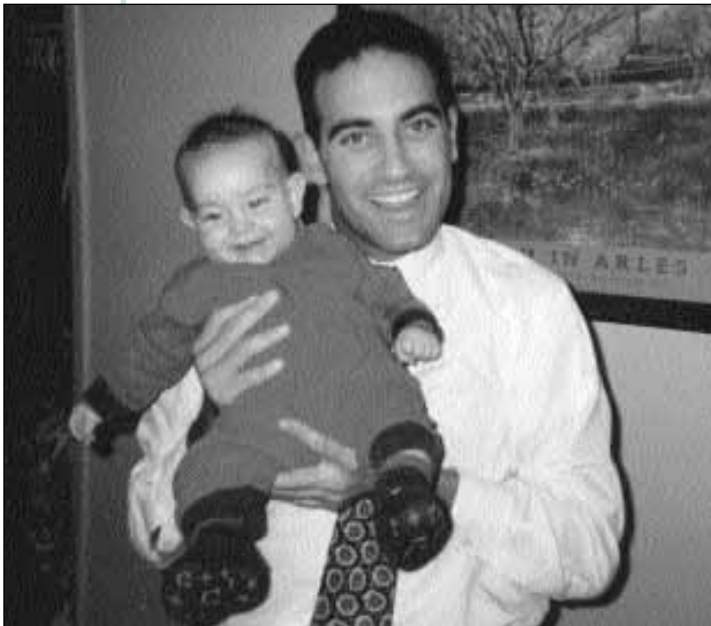
Q & A with Paul Flynn

Are you a member in good standing? If you have not sent in your 1997 \$25 membership dues, please use the attached envelope to do so and your membership card will be mailed back to you.

Paul R. Flynn, recently named Executive Director of the CMTA, embraces his nephew Timothy Raymond Flynn and “the challenge of the next generation.”

Paul R. Flynn will become the first Executive Director of the CMTA, effective September 1, 1997. Naming an Executive Director is the latest move by the Board to formalize the structure and roles of the CMTA staff. In February, Pat Dreibelbis was named Director of Program Services. The next issue of *The CMTA Report* will include, “A Day in the Life...” which will feature Pat.

Paul earned his Bachelor of Arts in Political Science and his Masters in Public Administration with a concentration in Non-Profit Management from Seton Hall University. He worked at Seton Hall Law School for more than five years, having spent the last year as the Acting Director of Development. Including the CMTA, Paul has devoted time to several service organizations. He is a founding board member of Jersey Cares, an organization that matches volunteers with team-oriented service projects, and also is on the board of the Link Community School. Since 1993, Paul has been a volunteer with Kids in Business (KIB) an educational reform initiative within the Newark Public Schools. He is a resident of Newark and is assisting KIB founder, Barbara G. Weiland, with the formation of the Discovery Charter School. Paul's other interests include golf, reading, playing frisbee and almost all types of music. In his immediate family, consisting of younger twin bothers, a baby sister, mom and dad, he and his dad have CMT. Of his extended family, 25 are known to have CMT, ranging from mild to very severe.



How did you become involved with the CMTA?

Ann Beyer [board member] approached me two summers ago when she chaired the nominating committee. Ann has known my grandmother [Kay Flynn] for years since they worked together founding the Westchester Support Group. Ann asked my grandmother if she could recommend a young person, preferably with fundraising experience, for the board... low and behold—here I am!! But seriously, I actually had my reservations at the time about getting involved. Ann and my grandmother's obvious commitment and the realization that I could, should help, led me here.

You've said, this [becoming Executive Director] is the right thing for you. Why is that?

Actually, it's the exact right thing, from the standpoint of timing, personal belief, and professional opportunity. Several months ago, a few mentors were prodding me, not so gently at times, that I should work on making this happen. I'm very pleased with how this evolved; I'm anxious to get started—to prove myself.

How do you see your role?

There's much to do of course, but simply put, it's three things: help set direction and priorities, recruit more volunteer leaders, and raise funds. Correct that, raise LOTS of funds!

What do you see as your most important task?

Awareness. Increasing the level of awareness about the disorder and the organization is the primary task; all else follows naturally. It's frustrating for me personally and to many people I've spoken to that CMT is—you know the mantra Pat—the most commonly inherited peripheral neuropathy... well, let's make it the best known, too!!!



cut here

Okay, that's a glaring problem. What are some other weaknesses and strengths you perceive in the CMTA?

Pat, I know this one bothers you because the CMTA hasn't formally addressed it—utilizing volunteers. We've had members offer their assistance and it became too difficult to coordinate the effort, or it was just plain bad timing... we're going to do a much better job involving members in our mission from now on.

And before I discuss strengths, not to belabor the funds issue, but we really must boost the percentage of dues-paying members.

Now, for strong points. There are many, but two stand-out: commitment and service. I'm not only impressed with, but inspired by, the incredible commitment by the volunteer leaders of this organization. Many years and countless hours barely measure the commitment made by board members, support group leaders and our regular volunteers. It's one of the reasons I am so confident in taking on this responsibility.

Additionally, and you play a major role in this Pat, this organization delivers a VERY HIGH level of service and personal attention. It's evident from the phone calls, letters, and people we meet. It's very gratifying.

So, Paul, I asked you for a photo to include with this interview and you've given me one of you and your nephew. He's cute but...

He sure is and while that's probably reason enough, that's not why I chose the picture. He's the first grandchild on either side of the family. He represents the next generation and, as such, renewed concern about the family's hereditary condition. It may only be anecdotal, and I pray that's the case, but it seems that CMT in our family is worse with each passing generation. If that doesn't bring urgency to our efforts, I don't know what will.

CMTA MEMBERSHIP/ORDER FORM

Name: _____

Address: _____

Phone Number: _____

	QTY	COST	TOTAL
Charcot-Marie-Tooth Disorders: A Handbook for Primary Care Physicians		members \$15 non-members \$20	
Membership Dues		\$25	
CMT Facts I <input type="checkbox"/> English <input type="checkbox"/> Spanish		\$3	
CMT Facts II <input type="checkbox"/> English <input type="checkbox"/> Spanish		\$5	
CMT Facts III		\$5	
VCR Tape: CMT Neurology		\$15	
VCR Tape: Physical Therapy & Occupational Therapy		\$15	
VCR Double Tape: duPont Conference		members \$25 non-members \$40	
CMT Informational Brochure <input type="checkbox"/> English <input type="checkbox"/> Spanish		FREE	Send Self- Addressed Stamped Envelope
Physician Referral List: States: _____		FREE	
Letter to Medical Professional with Drug List		FREE	
Contribution to CMT Research			
TOTAL			

Check payable to the CMTA

VISA MasterCard

Card Number _____ Expiration Date _____

Signature _____

Mail to the CMTA, 601 Upland Ave., Upland, PA 19015

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.

■ CONFERENCE ALERT

Dr. Richard Braver, DPM, will host a seminar entitled, "Let Us Take Care of Foot And Ankle Pain" for CMT patients and family members on September 27, 1997. The seminar is free and will be held on Saturday at 12:00 noon in the offices of the Active Foot and Ankle Care Center, Englewood, NJ. If you have questions or would be interested in attending, please call Thea Capuano at 201-569-7672. They do need an accurate count of participants, so call to register.



I'm Going to Jump, Dammit

by Benjy Feen and Jennifer Wilson

Like most folks with CMT, I get discouraged from time to time. Most recently was when a prominent lay figure in the CMT community, not associated with the CMTA, told me that I should avoid weight training because I would "burn out my nervous system," whatever that means. Now, I've been training since 1992 [see article on Benjy Feen's weight training in the fall 1994 *CMTA Report*, reprinted from *Muscle and Fitness* magazine], when I started while recovering from a car accident, and I put an awful lot of time and energy into my workouts. Through weight-training, I've been able to increase my body weight from 140 to 165, all of it muscle. I've been able to offset the effects of atrophy, regaining some tone in my calves, which were completely paralyzed 4 years ago. I've gained enough strength and energy to carry a 60-pound backpack around Europe for a month, and I feel great!

Even my workouts are fun; for me there's no moment quite as worthwhile as those I spend on the bench under a barbell, wondering whether I'll be able to put it up one... more... time, but knowing I can probably do 5 or 10 pounds more next week. To say the least, it's euphoric.

I'd been aware that some doctors recommend against strenuous activities, but never really worried about their opinions. The benefits of weight training, in my view, far outweighed the potential dangers. Putting on muscle mass increases general metabolism, which keeps body fat low; it stabilizes blood pressure, during rest time as well as during the body's reactions to stress; it improves the effects of circulation; it helps strengthen and maintain connective tissue; and working out even helps personal habits, as I am sure I eat and sleep on a roughly consistent



Why say, "I'll jump 2 feet," and then do it or fail, when you can just say, "I'm gonna JUMP, dammit!" and let yourself find out your genuine limits.

schedule. Then, there's the self-confidence and pride you gain from looking in the mirror and knowing that you have, without a doubt, improved yourself.

But, here was an "expert" in CMT telling me that I was destroying my body, and she warned me that I would call her in a few years, weeping, begging for her help.

The phrase "grain of salt" comes to mind. I guess I was going low-sodium that week because her advice hit me pretty hard, and I just stopped working out. Suddenly, I had more time for school, work, and my girlfriend, and I didn't miss my workouts much at all.

Until, that is, I started noticing a few things. I had less energy; I didn't sleep as well; my diet had gone from pasta and chicken breasts to McDonald's and beer, and, of course, I started getting weaker. It got harder to keep up with my busy schedule. I also began to notice my body fat increasing slightly, which bothered me; I was proud of the physique I'd built, and it was melting away. In a lot of ways, I'd given up.

I'm a system administrator and a webmaster; typing is my life. When you type 13 hours a day, it's evident when your hand strength is failing. The idea of losing my hands completely must have been the shock I needed to get back into the gym; after all, what could I lose by trying? I started working out again, and I wasn't surprised to find that within a week or two, I found I was better rested, eating better, and, of course, my hands seemed to be working better. In fact, I'm back to my old shape again, so I've lost nothing.

I guess I have idealistic standards; then again, why say, "I'll jump 2 feet," and then do it or fail, when you can just say, "I'm gonna JUMP, dammit!" and let yourself find out your genuine limits.

In that vein, my goal is to bench press 300 pounds. Right now I weigh 150 pounds and bench press about 180 to 190. We'll see what happens!

Detroit Conference at Wayne State University

Don't miss this one!!

A conference on Charcot-Marie-Tooth disorders will be held at Wayne State University, Detroit, Michigan on Saturday, October 18, 1997 from 8:00 AM until 4:30 PM in the Elleman Building auditorium. The main lecture hall has a capacity of 125 people and all small group meetings will be in nearby rooms. The entire facility is barrier-free has a parking garage adjacent to it.

This conference is unique in that Wayne State is the site of the CMTA's current research grant and one of the break-out groups will provide attendees with a tour of the clinic and research facilities in which the CMT Type 1A research is being done by Dr. Michael Shy and Dr. Agnes Jani, the Armington Research Fellow.

Registration and coffee starts at 8:00 AM, presentations begin at 9:00 AM. Lunch will be provided at noon. The first session will be a presentation by Dr. Michael Shy, conference host. Vince Bertolini from Athena Diagnostics will be available to discuss the genetic blood tests for CMT and a physiatrist will discuss evaluation and rehabilitation of CMT patients. The workshops in both the morning and afternoon include genetics, pain management, occupational and physical therapy, the research facility tour, and a podiatry talk by CMTA Vice-President, William Quinn, DPM.

After lunch, the same workshops are available, followed by full group presentations on



Dr. Michael Shy and Dr. Agnes Jani will discuss CMT research at Wayne State during the October 18th conference!

orthopaedic surgery and a closing presentation by Dr. John Kamholz, an associate of Dr. Shy.

Question and answer sessions follow each large group presentation.

On Friday night, October 17, 1997, there will be a hospitality suite in the atrium of the Elleman Building. All attendees are invited to meet with Directors of the CMTA and conference presenters.

The Hotel St. Regis, 3071 West Grand Blvd., Detroit, 3/4 of a mile from Wayne State, is offering rooms for \$69.00 plus tax, per night. For registrations, call 1-800-848-4810 and mention that you are attending the conference on CMT at Wayne State.

A map to the conference will be provided with your confirmation letter.

**Return the registration form and payment to:
CMTA, 601 Upland Ave.
Upland, PA 19015**

REGISTRATION FORM: DETROIT CONFERENCE, OCTOBER 18, 1997

Name _____ Phone number _____

Address _____

Names of other attendees _____

Price per person for members of the CMTA is \$25; \$40 for non-members. Number of attendees _____ Total Amount _____

Check enclosed Visa MasterCard Card number _____ Expiration Date _____

Signature _____

OF INTEREST

There is a site on the Internet where you can find out about medications you are taking or might wish to take. The location is: <http://www.cponline.gsm.com>. That is the location for Clinical Pharmacology Online and it has complete information on dosages, contraindications, side effects and drug studies. The information is constantly updated.



Managing Pain in Charcot-Marie-Tooth

by Dr. Becky Parks

From her presentation at the St. Louis Conference

Many patients with Charcot-Marie-Tooth disorders have an understanding why they have weakness, numbness, or other loss of function. However, what is sometimes more difficult for patients to understand is why the diseased nerves give them pain. When peripheral nerves develop disease, they cease to function properly, resulting in loss of strength or sensation. In addition, the abnormal nerves may begin to “short-circuit,” producing sensations not generally experienced by individuals without peripheral neuropathy. Some of these sensations are extremely painful and consist of burning, electrical shock-like, or jabbing sensations; pain in this situation is termed “neuropathic pain.”

There are a number of medications that can

provide various degrees of relief of neuropathic pain. One class of medications which can be effectively used to cut down on pain is the antidepressants. The antidepressant medication that is most widely used for the purpose of controlling pain is amitriptyline, although nortriptyline, desipramine, and trazodone may also be used. These medications, which generally need to be taken only once daily (usually at night), cut down on the transmission of “pain” signals to the brain. They may also function to greatly improve one’s sleep at night. The most common side effect from these medications is drowsiness. Although this side effect generally wears off after a few weeks, some patients never adjust and the medications have to be discontinued. Other side effects may include dry mouth and weight gain. Your physician should discuss all of the side effects (there are many) with you prior to having you start these medications.

Manage Chronic Pain with Mind and Bo

From the Mayo Clinic Women’s HealthSource

Can you think your migraines away? Can you wish away the aches in your joints or the twinges in your shoulder? Yes... at least to a certain extent, according to the latest findings from pain research.

What you think and feel and even how you were raised can determine how you experience the physical sensation of pain. That’s why pain specialists are now combining traditional physical therapy and pain medication with less mainstream techniques such as meditation, guided imagery, deep breathing, and progressive muscle relaxation. This combination of therapies strikes pain where it resides—in both your body and your mind.

The power of thought

Research begun more than 30 years ago has uncovered new alternatives for managing pain. In 1965, two scientists proposed the central nervous system—which transmits pain signals from your nerve endings to your brain—has “gates” that open and close to allow pain messages to

either reach your brain or be blocked. The theory has gained wide acceptance.

More recent research suggests these psychological factors can determine how your brain receives pain messages.

Mood. Your attitude and mindset may explain why the headache you get the day your boss gives you a raise doesn’t hurt as much as the one you get the day your car breaks down on the expressway.

Personality. Your thoughts and feelings color your perception of pain. Your tense or angry reaction to pain can cause additional stress, leading to headaches, muscle spasm, and jaw pain.

Fears. Believing your pain is beyond your control can make it seem worse. If you’re worried that your joint pain will keep you up all night, it probably will.

Energy level. Your arthritis may act up more after a long work day than when you’re taking a relaxing vacation.

How you were raised. If your parents were stoic about pain, you’re more likely to brush it

Disorders

Another type of medication that is often very effective is anticonvulsants. These medications are most commonly used to treat seizures but are also quite effective for pain control. They also cut down on the transmission of pain signals to the brain. These medications usually need to be taken more frequently than the antidepressants and thus are sometimes not as effective for busy people for the simple reason that they may forget to take the medication on a regular basis. The most commonly prescribed anticonvulsants for the treatment of pain are carbamazepine (brand name Tegretol), clonazepam (brand name, Klonopin), valproic acid (brand names, Depakene, Depakote) and Neurontin. Side effects may include dizziness, nervousness, nausea, or unsteadiness; however, there are many potential side effects and, as always, any medication should be thoroughly discussed with your physician prior to its initiation.



These are just some of the medications available to treat neuropathic pain. Effective pain control also includes the patient's participation, including a willingness to try to accurately report how much pain he/she is experiencing. With patience and a good line of communication with your health care provider, relief from chronic pain is very possible.

Dr. Becky Parks discusses pain medication with a group of CMTA members at the St. Louis Conference.

dy Techniques

off. Your pain may seem worse if your parents treated every cut and scrape as a catastrophe.

Using relaxation techniques to control pain

Relaxation techniques, used in conjunction with pain medications and physical therapy, can diminish, and in some cases, eliminate your pain.

Meditation. Although it's one of the most effective relaxation techniques, meditation requires diligent practice. The most common approach is transcendental meditation, in which you use a repeated word or sound, called a mantra, to keep your mind free of distracting thoughts for 20 minutes at a time. Eventually you achieve a state of calm and control.

In a type of meditation called "mindfulness," you focus on your breathing and pay attention to your intrusive thoughts—even pain—but remain nonjudgmental. You're aware of your pain but are emotionally detached from it. Eventually it becomes less noticeable. Meditation is usually a solitary

pursuit that requires two 20-minute sessions daily.

Guided imagery. This technique involves using your mind to place yourself in a tranquil setting, walking on a beach or resting under a tree. When you're fully relaxed, the next step is to send yourself a positive message about your pain. This is often easier to do with guided imagery tapes available in bookstores.

Deep breathing. Take a deep breath, fill your lungs with air for a count of about 4 seconds, then exhale for 4. Repeat to release muscle tension and lower your blood pressure. You can use deep breathing anytime, anywhere.

Progressive muscle relaxation. This is one of the easiest techniques to learn. All you need is a quiet, comfortable place to lie down. Then, you make a tight fist and hold it for 5 seconds. After noting the tense sensation, release your fist and feel the tension leave. Repeat the tense-release technique on the rest of your muscles to relax your whole self. Because this technique may focus too much on painful joints and muscles, it may not be appropriate if you have a joint or muscle disorder.



**THE CMTA'S
HOMEPAGE IS**
www.charcotmarie-tooth.org

**YOU CAN EMAIL
THE CMTA AT**
CMTA@charcotmarie-tooth.org

**YOU CAN REACH
CMTnet
ON THE INTERNET**

@URL<http://www.ultranet.com/~smith/CMTnet.html>

OF INTEREST

President Clinton, in a major policy statement at the White House on July 14, urged Congress to pass legislation to prohibit health insurance companies from discriminating against persons on the basis of the information contained in a genetic test or even the fact that the person had requested a genetic test. Clinton remarked that the advantages of genomic research should not be lost because people are afraid to be tested due to a concern that their health insurance could be canceled, denied, or made prohibitively expensive.



Charcot-Marie-Tooth Disorders: Clinical Presentation, Disease Course, Treatment

Florian Thomas, MD, PhD
St. Louis University, St. Louis, Missouri

From a presentation that Dr. Thomas made at the first meeting of the St. Louis, MO, support group of the CMTA.

Charcot-Marie-Tooth disorders represent a group of neuromuscular diseases that are inherited in several different patterns. They are characterized by weakness that typically begins in the feet during late childhood and later involves the upper legs and arms.

Owing to its insidious onset, some patients remain unaware of their disorder or are diagnosed only later in life. Often patients contact neurologists because of loss of balance, muscle weakness, and foot deformities. Some children are referred by their teachers because of clumsiness or toe walking.

As the disease progresses, patients complain of tripping over objects because of foot drop. Ankle sprains and fractures are frequent. Because of a foot deformity with hammer toes and high arches, patients may have difficulty finding comfortable shoes; this also causes painful calluses. They may notice increasing difficulty using their hands to manipulate small

objects, such as zippers, forks, or pencils. Some patients complain of cold feet, which is often associated with loss of hair or edema (abnormal accumulation of fluid) in the legs.

This condition does not affect life span or intellectual function, and usually does not involve other organs; it does not usually affect a person's ability to earn a living. The several subtypes differ in severity. Over time, the disorders lead to a variety of other medical problems, such as chronic pain and orthopedic problems. Rarely, patients can become wheelchair dependent.

The foot deformity frequently causes pain and foot ulcers. Pain can also result from pressure or strain of various structures associated with bones, joints, and tendons. Gait abnormalities can cause lower back pain due to strain. Patients frequently suffer from leg cramps. On occasion, weakness reaches the thigh muscles as well. Patients may develop scoliosis and kyphosis of the spinal column and children have been reported to suffer from hip deformities. Frequent sprains may cause further problems by making the ligaments more lax.

Treatment consists of different orthotic devices, which include high-top shoes or boots (for milder cases) and ankle braces. As ankle

Studies Seeking Participants

Dr. John Hsu, a member of the CMTA's Medical Advisory Board, is interested in doing a walking (gait) study of the Charcot-Marie-Tooth patient to understand better which muscles are affected and how the muscles that are working function during standing and walking. In conjunction with the pathokinesiology laboratory at Rancho Los Amigos Medical Center in Downey, California, Dr. Hsu would like to study muscle activity and joint motion during gait.

Patients must be between the ages of 7 and 50 years, with a proven diagnosis of CMT, either by nerve conduction studies or muscle

biopsy. The patients should not have other secondary conditions that would interfere with their walking. They should have some form of foot deformity, for instance, a cavus foot, high arch, or drop foot, but not of such severity that they cannot walk independently (use of orthoses during walking is okay.) If they have had surgical procedures done previously, they will need a copy of the operation report.

Preliminary studies have shown that the peroneal longus muscle may be a leading cause of foot deformity.

Patients who are interested in the study need to be able to spend 1 to 2 hours for a

and Genetics

weakness progresses, tendon surgery may be required to stabilize the foot. With longstanding foot deformities, contractures may occur and necessitate bone surgery. Some of these procedures may require an orthopedic surgeon and others can be done by podiatrists. Occasional neuropathic pain, such as tingling or burning, may require the use of specific pain medications with which the neurologist should be familiar. (See article on pain on page 6.)

Patients can also suffer from hand cramps, and hand weakness may become severe. Contractures (a deformity caused by irreversible shortening of muscles and tendons) can often be prevented by splinting but may require tendon-release surgery. Multiple devices are available that allow good functional use of weak hands for general activities. Patients need to access physical and occupational therapy departments for help in these areas. Contractures of the hands, although rare, may require splinting or even surgical release.

While Charcot-Marie-Tooth disorders cause a generalized neuropathy, the ensuing bony deformities can lead to focal entrapment neuropathies causing pain and weakness. These may require nerve-release surgery that is per-

clinical examination, review of records, etc. and be scheduled for a 2- to 3-hour gait study on a separate day. If they wish to review the results, a separate visit will need to be arranged because of the complexity of the test. Ideally, patients should live in the Southern California region. They would need "point of service" insurance or HMO permission to allow for one consultation visit (by Dr. Hsu as orthopaedist and clinician) and payment for the laboratory study. If they want a follow-up visit to discuss the results, they would need to get permission for that, also. The study would be done on a consultation-only basis, so as not to interfere with primary care given by their own physician. If the patients wish, Dr. Hsu will gladly discuss the results of their individual study with them and make recommendations for further care to their primary care physician.

formed by orthopedic surgeons, hand surgeons, or neurosurgeons.

The issue of exercise in CMT remains controversial. While it has been argued that overuse of weak muscles can cause additional weakness, there is convincing evidence from several studies that strengthening exercises have no detrimental effect. However, patients should be instructed what exercises are beneficial and which ones could damage already weak muscles and joints.

Over the past several years, we have started to understand the molecular genetics of Charcot-Marie-Tooth disorders. Although they are highly complex, it has become apparent that the different forms of CMT are all associated with mutations in peripheral nerve genes. These genes all play crucial roles in the normal development and maintenance of the myelin sheath (which provides the crucial layer of insulation around nerve fibers and is required for nerve conduction) or the axon or nerve itself. The scientific process has helped us clarify the different clinical subtypes and understand more clearly how and why these disorders develop. Furthermore, we can now speculate about, and investigate, possible ways of treating these conditions, initially in animal models and, perhaps one day, also in patients.

To speak directly to Dr. Hsu, call 562-401-7166. For referrals, your insurance company should write: Dr. John Hsu, 7626 Golondrinas, Bldg. 304, Downey, CA 90242. Attention: Janet Hirsch, Rancho Faculty Practice Manager. The phone number there is 562-803-1433. Fax: 562-803-3043.

The Center for Research on Women with Disabilities at Baylor College of Medicine in Houston, Texas is looking for women with physical disabilities and chronic physical illness to take part in research studies to improve the quality of life for themselves and others. Participants must be between the ages of 18 and 65 and will be paid for their time. For information about the study, call the center at 1-713-960-0505.

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OF INTEREST

The *CMT Nieuwsbrief*, a publication of CMT International, Belgium, reprinted an article from the Winter 1997 *CMTA Report* by Dr. Gregory Carter. The article was about CMT and breathing difficulties and appeared in their June 1997 issue.



Drs. Phil Chance and John Hsu are two of the volunteer members of the CMTA's Medical Advisory Board who often contribute articles on CMT to medical publications. See "Above and Beyond" on page 12 for more of Dr. Hsu's "contributions."

Inherited Neuropathies

Phillip Chance, MD, and John Hsu, MD

These excerpts are from an article published in Current Opinion in Orthopedics, 1995, 6;VI:100-108. Both Drs. Chance and Hsu are members of the CMTA's Medical Advisory Board. We cite here the sections on some of the less-often discussed variants of CMT. A complete reprint is available from the office of the CMTA for \$2.00 to cover copying and postage costs.

Dejerine-Sottas disease; expansion of the Charcot-Marie-Tooth neuropathy type 1 phenotype spectrum

Dejerine-Sottas disease (DSD), also called hereditary motor and sensory neuropathy type III, is a severe, infantile, and childhood-onset, hypertrophic demyelinating polyneuropathy. The clinical features of DSD overlap with those of severe CMT1. Many cases of DSD appear to represent sporadic cases and are usually thought to result from an autosomal recessive gene. Recent molecular genetic studies have revealed that DSD may be associated with point mutations in either the PO or the PMP22 gene. Interestingly, in patients with the DSD phenotype studied to date, all mutations have been present in the heterozygous state, suggesting that DSD may actually be caused by dominantly acting genetic defects. Molecular studies have not disclosed evidence for recessive inheritance in DSD.

Hereditary neuropathy with liability to pressure palsies

Hereditary neuropathy with liability to pressure palsies (HNPP; also called tomaculous neuropathy, recurrent pressure-sensitive neuropathy, and bulb digger's palsy) is an autosomal dominant disorder that produces an episodic, recurring demyelinating neuropathy. Peroneal palsies, carpal tunnel syndrome, and other entrapment neuropathies are manifestations of this disorder. Motor and sensory nerve conduction velocities may be reduced in clinically affected patients as well as in asymptomatic gene carriers. Pathologic changes observed in peripheral nerves of HNPP patients include segmental demyelination and, in some pedigrees, tomaculous or "sausage-like" formations. Some patients with HNPP may present with signs of chronic neuropathy mimicking CMT1, including pes cavus and mild distal weakness. Other patients may present with episodic, painless brachial plexopathy. (*The brachial plexus is a collection of nerve trunks which control muscles in and receive sensations from the arm and hand.*)

The HNPP locus has been assigned to chromosome 17p11.2-12 and is associated with a deletion in the region of the PMP22 gene which is the same interval that includes the CMT1A duplication. *De novo* deletion of paternal or maternal origin has been detected as a basis for sporadic HNPP.

It has been proposed that the deleted chromosome in HNPP and the duplicated chromosome in CMT1A are the reciprocal products of unequal crossing over. The apparent homogeneity for size of the duplication and deletion in unrelated patients and detection of *de novo* duplication and deletion events suggest that a common mechanism may account for the generation of the duplicated CMT1A chromosome and the deleted HNPP chromosome.



Hints on Conserving Energy

by **Grace R. Young, OTR, Fresno, CA**

Reprinted from Polio Network News, Vol. 13, No.2, with permission of Gazette International Networking Institute, 4207 Lindell Blvd. #110, St. Louis, MO 63108-2915.

As a polio survivor, I have had to modify many areas of my life over the years. I assure, however, that this modifying is not as oppressive as it may sound. By conserving energy in less important areas, I can still do the activities that I enjoy—that's what energy conservation is all about. Here are a few ideas I hope you will find helpful.

Rest before you become aware of fatigue, a difficult task because most of us were taught to ignore pain and fatigue. Before starting an activity, decide how long you will work and when you will take rest breaks. For stressful projects, set a timer and break the activity into segments of half-hour work and 15-minute rests. Lie down during your rest break—listen to music or watch television, meditate, visualize or read, but get off your feet.

Don't become over-ambitious on days when you feel good, or you may then be sidelined for several days. Alternate light and heavy chores throughout the week. Don't do anything heavy if you're planning an evening activity.

Sit whenever possible—for showering, dressing, grooming, working with hobbies, preparing meals, etc. Sitting takes 25% less energy than standing. When you sit, make sure that your work heights are about an inch below elbow level. A drafting chair on casters with a pneumatic seat lift works well, but back it into a corner before hoisting yourself onto the raised seat or the chair may roll out from underneath you. Other ways to obtain a lower work surface: use a beanbag lapboard, a wooden cutting board on your lap, or an adjustable-height, hospital bedside table, which you may find in a thrift store.

Organize your home so that items are easy to reach and you don't have to duplicate motions. In the kitchen, store frequently used items at a height between your hips and your lips. Use vinyl-coated wire racks to store dishes in stacks of their own kind; store pots and skillets individually on wire racks so that you don't

have to lift top items to get to one on the bottom. Use stacking storage bins on wheels and wire shelf units that hook over pantry doors. One- and two-level sliding racks, bins, baskets, and shelf trays can make base cabinets for food staples and cleaning supplies.

Don't put away items you use frequently. After washing pans or skillets, for example, let them dry on top of the stove. Let dishes dry in a rack and then use them in setting the table for the next meal.

Arrange your home to minimize trips. Install floor-to-ceiling pole shelves over the toilet tank in each bathroom for storing towels and washcloths. Store sheets and pillowcases in each bedroom where they are used.

Duplicate supplies used in different areas. For example, store cleanser and sponges under each sink and keep a broom and dustpan in several locations.

Avoid buying deep pile carpets with thick padding. Wheelchair users find plush carpeting difficult to maneuver on, and people with walking problems may have difficulty maintaining their balance on it. Throw away throw rugs that can slip, and remember that ceramic tile floors are slippery when wet.

And, finally, use mechanical help for carrying things. Carrying heavy items changes your center of gravity and stresses your trunk, arm, and leg muscles. A kitchen utility cart on casters, available in most housewares departments, allows you to transport dishes, glasses, silverware, and food from the counter to the table and back again in one trip. Use the cart to carry laundry and cleaning items, and push it along when you straighten up the house. A light-weight luggage cart is useful in many situations. Use one to carry purchases, to transport articles between the car, house, or office, and to move articles from room to room. I keep a luggage cart open and ready to use whenever I have to carry heavy or bulky items around the house. (The definition of "heavy," of course, depends on the strength of the individual.)

You undoubtedly have many other ideas that are appropriate to your particular lifestyle. Look upon energy conservation as just the latest challenge in your life.

GIFTS WERE MADE TO THE CMTA IN HONOR OF:

Madelyn Brown's 40th Birthday
Selma Rothstein

Kay Flynn's Volunteer of the Year Award
Paul R. Flynn

Paul R. Flynn
Thomas Martin
Roughneen

Francine Koehler's 50th Birthday
Mr. & Mrs. Robert
Bernstein

Greg Louer's Birthday
Emily Louer

Samantha Lynn Sheriff
Daniel and Nancy
Headrick

Norma Jean Rodrigues' Birthday
Beverly Sermersheim

Ruth Wendkos' Special Birthday
Mrs. Estelle Bandler



MDA Announces New Regulations for Payment for Durable Medical Equipment

Effective July 1, 1997, the MDA can augment payment for the purchase of wheelchairs or leg braces, up to the medically allowable limit of \$1,700, when prescribed by an MDA clinic physician. For a period of time, MDA did not provide these direct patient services, and their return will be invaluable to CMTA patients.

The maximum allowable amount was determined to be at the level to cover the cost of a manual wheelchair, but can be applied to the cost of a powered wheelchair, scooter, or reclining or other specialized wheelchair, or to the purchase of long or short leg braces (KAFOs and AFOs.)

The criteria for MDA assistance with the purchase of a wheelchair or leg braces are as follows:

1. MDA can assist with the purchase of only one wheelchair or one pair of leg braces when medically essential and prescribed by an MDA clinic physician.

- A. every 3 years for individuals up to 18 years of age
- B. every 5 years for individuals over 18 years of age

2. MDA can assist with payment for only one wheelchair or one pair of leg braces per individual and cannot supplement the cost of an additional wheelchair or pair of leg braces within the time frame defined above.

The above eligibility criteria represent the only conditions that must be met for MDA to consider the provision of a wheelchair or leg braces. Individuals, regardless of age, education, or employment status, will be eligible for MDA assistance. In that regard, no formal application process other than a prescription and statement of diagnosis will be required to obtain a wheelchair or leg braces.

Editor's note: This is a new addition to the CMTA Report and will highlight the instances in which persons associated with the CMTA make that little extra effort to help one another. We encourage contributions to this column from members and support groups who know of someone who has gone out of his or her way to help another person or group. When possible, send both information about the efforts of the person and a picture of the person who is being highlighted. The letter at right was sent to Pat Dreibelbis, Director of Program Services for the CMTA.

ABOVE AND BEYOND

Dear Pat,

Several weeks ago I spoke with you about my concern that a bone fusion had been recommended for my 14-year-old son. You suggested that I contact Dr. John Hsu at Rancho Los Amigos in Downey, CA.

I wanted to report to you that I had an extraordinary experience with Dr. Hsu. After speaking with him by phone, he expressed a desire to see my son along with recent x-rays and original electromyography reports. As luck would have it, he was traveling to Atlanta in February for a conference and said he'd be glad to see my son and examine him at his hotel.

So, on the appointed date, we met him in Atlanta. He did a history, examined my son, and concluded that a tendon transfer was indicated and advised us to do it rather soon while his foot was very flexible. When he asked who we had seen at Emory and we told him Dr. Thomas Moore, he smiled broadly. Dr. Moore was a former associate from Ranchos! Dr. Hsu said that next to himself, he could think of no one better to do the surgery.

What an amazing experience this was, Pat! And how wonderful it was to meet a doctor who cares so much for his young patients that he consulted with us in his hotel room, for free!

Thank you so much, Pat, for being the initiator of all this. You have truly been a blessing to us as we have struggled with what to do about my son's foot deformities. With your help, Dr. Hsu's advice, Dr. Moore's skills, and our persistence, we all may have added years of pain-free walking to our son's uncertain future with CMT.

Thank you.

—B.M. Perry, GA

Footnote: When we called the author of this lovely letter for permission to reprint it in the newsletter, he told us the continuing story of Dr. Hsu's impact. Whenever his son goes to Emory for evaluation, Dr. Moore brings many medical students and interns with him and says, "He's a personal friend of Dr. John Hsu. You've all read many of Dr. Hsu's articles on orthopaedic surgery." Then, Dr. Moore recounts the story of what Dr. Hsu did for the boy. As the father said, "It can't hurt to have "friends" that are well-known and well-respected in the medical community."

BOARD CONNECTIONS

Rex Morgan and Patty Wall: A Multi-Talented Couple

Rex Morgan was a member of the CMTA's Board of Directors for 6 years, but his involvement with the organization reaches far beyond that role. He founded the Philadelphia area support group and was its leader for many years, routinely hosting meetings of 35 or more members. He developed several media campaigns in the early years of the organization and most recently helped to write and design the membership campaign mailing. His ability as a writer comes from his 16 years in magazine, television, radio, and comedy work. He won the 1992 ACE Award as a writer of Nick at Nite's "Global Village News." Currently, he is a technical writer for McNeil Consumer Products (the Tylenol family of products) working on assorted computer projects.

Rex is also a member of the comedy group, "Mixed Nuts." The group was founded at the University of Pennsylvania in 1978 and has appeared on TV and radio and at countless comedy clubs and colleges. Currently, the group is touring the country with "You're Testing My Patients!" an original revue for health-care audiences.

Rex's wife, Patty, has been a graphic artist for 15 years and has willingly joined her hus-



Rex Morgan has a long history of lending his talents to CMTA efforts.

band in volunteering for the CMTA. She designed the cover for Charcot-Marie-Tooth Disorders: A Handbook for Primary Care Physicians, judged the CMTA logo contest several years ago, and in the early years of the organization lent her expertise to the design of many of the brochures, fundraising letters and other publications of the organization.

Rex and Patty have two children, Rachel, 14, and Tyler, 10, and live in the Philadelphia suburb of Horsham.

When asked to work on the membership letter, Rex was enthusiastic. He recognized the value of member-

ship in the CMTA, especially as it relates to receiving the newsletter. "As someone who bangs on a keyboard for a living, I've really enjoyed watching *The CMTA Report* become a useful, attractive newsletter. It used to look like an old science journal, but not anymore! It's a great showpiece for the organization."

Rex and Patty exemplify the best in volunteerism. They have specific gifts and talents, and they share them, unselfishly, with the organization to the benefit of all the members of the CMTA.

Editor's note: This new column highlights past and present CMTA Board members and the members of our Medical Advisory Board.

We Need Your Help!

The Board of Directors of the Charcot-Marie-Tooth Association is actively seeking candidates for the Board. If you are interested and can make the commitment, please call the office of the CMTA (800-606-2682) or email the organization at either CMTAssoc@aol.com or CMTA@charcot-marie-tooth.org. The nominating committee will contact you. They are seeking persons with various qualifications to help in achieving the ambitious goals of the organization.

Cycle for CMT 1: The Follow-up Report

by Joe Beernink

Editor's note: The Spring 1997 CMTA Report detailed Joe Beernink's plans to participate in the Denver-to-Aspen Classic, a 200-mile bike race through the Central Rockies. Here's how his ride went.



Pledges made for Joe Beernink's ride will fund a CMT-oriented summer research internship next year.

The 1997 Denver-to-Aspen Classic is something I will never forget. From a parking lot in South Denver at 3:39 AM, we pedaled west into a gusting headwind in the dark, toward the mountains. As I climbed the 20 miles of Deer Creek Canyon, occasionally joined by other riders, I already knew that I had underestimated the event. 1,700 miles of training, months of sprints, long rides, and mental preparation helped, but in my worst scenarios, I had not imagined such a wind that early in the ride, accompanied by very low temperatures. As I entered Aspen Park, a small town 35 miles from the starting line, I knew I was exerting much more energy than I had planned. I adjusted my pace as I rode the next 30 miles to the top of the 10,000-foot Kenosha Pass, but the winds picked up, making the climb twice as difficult as it should have been.

I knew at the top of Kenosha Pass that I was in trouble. My left knee was causing me severe pain, the result of a problem during training 2 weeks before. The wind not only caused much more air resistance than normal, slowing my pace, but also caused me to use more power at the slower pace than I wanted to.

On the long flat section after the descent down Kenosha Pass, I forced my speed up, trying to make up the lost time. The winds picked up again as I climbed the 10,051-foot Red Hill Pass, gusting to more than 30 mph. One gust nearly knocked me backwards as I neared the summit. Even the descent into Fairplay was slowed by the wind, and I arrived at the checkpoint, a physical and emotional wreck. My worst fears had not included riding into the wind for the first 89 miles. After a long break and a lot of soul searching, I knew that I would not be able to finish. But, I was determined to make the 100-mile mark. I got back on my bike and pushed hard into Hartsel, aided, finally, by a strong tail wind, and the knowledge that Hartsel marked the 105-mile mark. When I arrived there, I retired from the ride, realizing that to push my body any fur-

ther could cause permanent damage to my knees. My time to Hartsel was about 9 hours, with 7 hours and 55 minutes on the bike.

Did CMT make a difference in my ride? No. The injury to my knee had nothing to do with CMT. At least four other people dropped out before I did, so I know I wasn't alone. Where CMT did affect me was the fact that I was doing the ride in the first place. If I didn't have CMT, I might not have tried it. I might have been like so many couch potatoes who let themselves go, even when they are so lucky as not to have to fight the battles those of us with CMT fight all our lives. I have no regrets about attempting the ride, nor do I have any regrets about dropping out. Perhaps with more training and experience, and without the injury, I could have finished. One thing is for sure: the Denver-to-Aspen Classic is not easy and I have the utmost respect for all those who rode or attempted the ride.

People have already asked me if I'm going to try it again next year, and I'm not sure. I would like to do a team event, perhaps a relay across a state, with three to five other people with CMT. I want Cycle for CMT 2 to be bigger than the first one. I don't just mean more pledges; I mean more people involved, more people to be inspirations to those who have just been diagnosed with CMT and to those whose children have CMT. I want the whole world to know CMT is out there but is being beaten back every day through sheer desire and will power.

If you are interested in participating in *Cycle for CMT 2* or know of an event that might be appropriate, please contact me through the CMTA office. The more events we can organize, the better.

For those who supported me in this event, a big thank you. Special recognition goes to my parents, Anthony and Simone Beernink, and my sister Cindy, who drove all the way from Canada to be my support crew. Special mention also goes to the Rocky Mountain Cycling Club who helped me train and supported the event, and the employees of Neodata/EDS who offered their support. Finally, I would like to thank Scott and the gang at Louisville Cyclery in Louisville, Colorado, who got me on the road and kept me on the road all year. Thanks again.

More Contributions to Our 1997 Research Appeal

\$5,000 AND OVER GIFTS

The Evenor Armington
Fund
The Salomon Foundation
INC.

\$2,500-\$4,999 GIFTS

Steve Khosrova

\$300-\$2,499 GIFTS

Anonymous
Robert E. Buuck
Jean R. Iler

\$100-\$299 GIFTS

Anonymous
Edward F. Bachman
Ron & Ann L. Beyer

William S. Buchanan
Leonor Deetjen
John P. Dineen
Mary E. Dodd
E. I. Dreher
Joseph S. Fiore
Paul R. Flynn
Patricia K. Griffith
Daniel D. Headrick
Gay V. Jacobs
Thomas Lynch
Dennis Martin
Shirley A. McKee
Thomas M. Roughneen
Percy A. Sand
Dale Tucker
Richard A. Westsmith
Jane H. Wood

ALL OTHER GIFTS

Anonymous (3 Gifts)
Jane D. Allen
Christine E. Anderson
Sharon G. Arend
Richard J. Barohn
Elda M. Brown
Myrtle H. Browne
Jim B. Burns
William B. Cigliano
John L. Connelly
Margaret P. Crawford

Helen M. Dale
Karen E. Deaton
Morris De Pouw
Terry L. Duncan
Joyce M. Duval
Genevieve M. Eggers
Max D. Ellis
Theresa F. Ember
Paul E. Ertzinger
Brenda M. Gill
Kenneth E. Gilroy
Donald D. Gitell
John A. Gordon
Irene B. Gray
Aaron Greif
Terry M. Henson
Frances J. Hill
Donald J. Janner
Lester M. Johnson
Rainer Kamp
Gracia P. Kather
Howard Kenoyer
Susan C. Klemm
Drucilla Lake
Barbara S. Lanham
Alan B. Latman
Hans A. Lauridsen
Grace M. Loy
Philip E. Mack
Tracey Manuele
Patrick F. Marrs
Michele M. Martinez
James Maxwell
Alice M. Mayhew
Thomas A. McIntyre

Dan Merkel
Ida Miele
Mitchell Moody
Wayne Morrison
Dorothy G. Mulhall
Pauline Nelson
Gary L. Nicholson
Judith M. Nill
Marylynn Orzechowski
Brian R. Ott
John O. Parker
Emma R. Parmee
Daniel E. Patrick
Carol A. Pentecost
Lorraine Pignone-Martin
Harriet G. Redlich
Leonard Reinebach
Elizabeth K. Renninger
Alice C. Rew
Deborah J. Riestler
Carol B. Rosse
E. Gordon Rossiter
Barbara A. Shank
Jerry S. Shulman
Betty R. Smith
Roberta Steckler
J. R. Stillinger
Lee Sullivan
Karen E. Swain
Elizabeth W. Terrell
Joseph A. Unetic
Camille S. Walsh
Kathleen C. Weeks
Earline M. Wisher

*If you have not
already responded to
the 1997 Research
Appeal, we hope
that the increased
Armington Challenge
will encourage you to
do so. Your support is
greatly needed and
appreciated.*

Pen Pals for Kids

The Children's Hopes and Dreams Foundation, Inc. of Dover, NJ, offers a free pen pal program for children with chronic illnesses. The pen pal program helps children find an understanding peer, a shoulder to lean on, or a confidant and new friend. For more information on registering your child, call 1-201-361-7366 or write:

Children's Hopes and Dreams Foundation,
280 Route 46, Dover, NJ 07801 and request their
registration form.



Rocker/Balance Board Can Strengthen Ankles in CMT Patients

by Joy Rushfelt

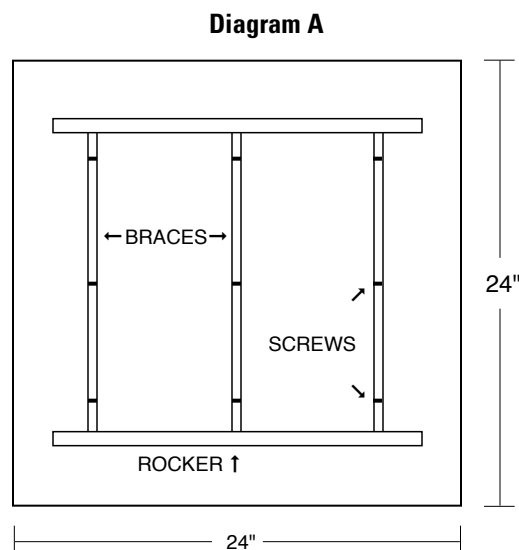
OF INTEREST

Addressing the scarcity of money exclusively earmarked for college students with disabilities, the new edition of HEATH's resource paper, *Financial Aid for Students with Disabilities* (1997), encourages students with disabilities to seek financial aid through the regular (not disability-specific) channels. Using a question-and-answer approach, the 11-page guide provides information about the financial aid process and resources, possible linkages with the vocational rehabilitation and social security systems, and graduate study. A two-page listing of disability-specific scholarships is also included. The publication is available for \$2, prepaid, from: HEATH/American Council on Education, Dept. 36, Washington, DC 20055-0036; tel. 202-939-9320; fax 202-833-4760; email HEATH@ACE.NCHE.EDU. The entire text can be found on HEATH's gopher site at gopher://bobcat-ace.nche.edu.

The following information refers to a home-constructed balance board designed to be used as therapy for strengthening the ankle muscles to improve balance for the CMT patient. [Editor's note: Joy emphasizes that no one who has had an ankle fusion should use this board and no one should ever begin an exercise program without checking with his or her doctor first.] During use of the board, there may also be improved muscle strength in the area of the knees (quads) and the hips. Two sizes are given. The 24" board is recommended for persons under 5'10", children, older adults, or persons with very poor balance. The 28" board is suggested for taller people or more athletic persons with fairly good balance. You may want to start out with the smaller rocker panels and later change to a deeper panel for a more challenging exercise.

Constructing a Rocker Board

Diagram A is a bottom view of the completed board. Dots on the braces indicate an approximate placement for the screws placed from the top of the board down through the braces to attach the rocker panels and the brace assembly to the top. It works best to sand or finish only the edges, and leave the top of the board with some roughness to keep feet from sliding off the board during the exercise. (Always wear shoes.)



MATERIALS NEEDED TO CONSTRUCT A ROCKER BOARD

- 24" or 28" square 5/8" plywood
- 18 deck screws 1 1/4" or 1 1/2" long
- additional wood for the two rocker panels and the three braces

24" version

Rocker panels:

2@3 1/8" wide by 20" long

Braces:

2@1 1/2" wide by 18" long

1@3" wide by 18" long for the center brace

28" version

Rocker panels:

2@4" wide by 24" long

Braces:

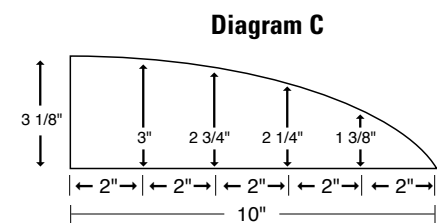
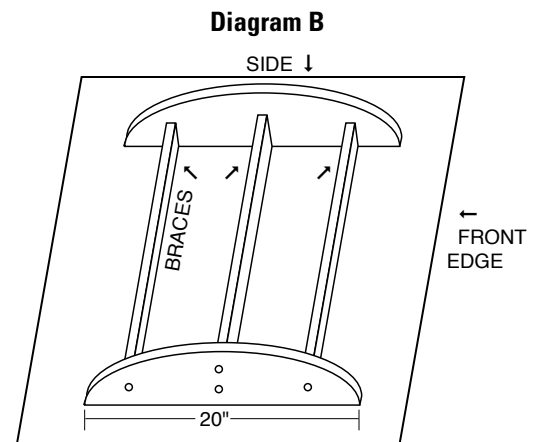
2@1 1/2" wide by 22" long

1@3 3/4" wide by 22" long

28" square of plywood for the top

Diagram B is a perspective view of the bottom. Dots on the rocker panel indicate the screw placement to attach braces to the rocker panels.

Diagram C is a view of 1/2 of the length of the rocker panels for the 24" version. Draw a pattern on paper with the measurements and use a



French curve tool to smooth the arc to a gentle curve. Suggestion: clamp the cutout shaped rocker panels together and sand the edges to assure that the curved edges match. Measurements for a steeper, more challenging rocker panel: try a 3 1/2" center measurement tapering down in a smooth arc as in drawing. For a 28" board, use a 4" center width, again tapered down to the end of the rocker.

Daily Exercises Using the Rocker Board

1. With feet in position as indicated in Figure 1, rock back and forward shifting weight from one foot to the other, with a slight bend in the knees. Do ten repetitions.

2. Change position of feet or turn body around so that left foot is in front. Repeat exercise 1.

3. Center feet as in Figure 2, toes facing the side of the board and rock from side to side. Do ten repetitions.

Suggestion: until your balance improves, use the board next to a high-back chair or chest, balancing yourself with your hands in a "light touch."

(This balance board was used by Joy in an exercise program designed for her by a physical therapist. She presented the concept to the Kansas City support group and another member figured out how to make the rocker board for less than one would cost when ordered from a catalogue.)

Figure 1

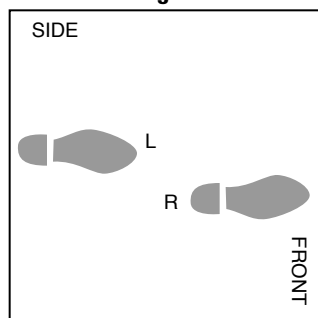
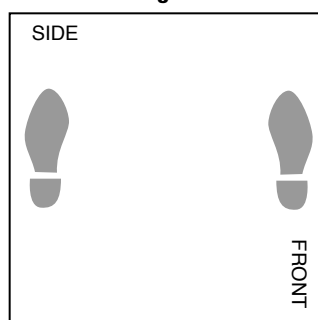


Figure 2



CMTA Support Groups

Alabama/ Greater Tennessee Valley

Place: ECM Hospital, Florence, AL
Meeting: Quarterly
Contact: William Porter, 205-767-4181

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, 707-253-0351

Florida/Boca Raton to Melbourne

Place: Columbia Medical Center,
Port St. Lucie
Meeting: Quarterly
Contact: Walter Sawyer, 561-336-7855

Michigan/Detroit Area

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

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 Center
Meeting: Quarterly
Contact: Carole Haislip, 314-644-1664

New York (Westchester County)/ Connecticut (Fairfield)

Place: Blythedale Hospital
Meeting: Monthly, Saturday
Contact: Kay Flynn, 914-793-4710

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

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

West Virginia/North Central

Place: VFW Conference Room,
Uttica
Meeting: Quarterly
Contact: Joan Plant, 304-636-7152
(evenings)

New Support Group Forms in Boston Area

We are pleased to announce that Dr. Deborah Riester will be the guest speaker at the first meeting of the Burlington, Massachusetts, CMTA Support Group. The first meeting will be held on Tuesday, October 7, 1997, at 7:00 PM in the Lahey-Hitchcock Clinic on Mall Road in Burlington. Dr. Riester's topic will be exercise and the CMT patient. Subsequent meetings will be held every other month on the first Tuesday of the month. For further information about the Burlington Support Group, contact the support group leader, David Prince, at 508-667-9008 (this 508 area code will change to 978 after 9/1/97).



WRITE TO US!

Pat Dreibelbis, Editor

The CMTA Report

CMTA

601 Upland Ave.

Upland, PA 19015

The CMTA Report

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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.

Letters to the Editor:

Dear CMTA,

I went to the CMTA's meeting in Connecticut in late October, 1996. I saw Dr. Frank Palermo's presentation on electrical stimulation and was very excited. A couple of weeks later, I drove down to Connecticut and had an appointment with Dr. Palermo. A few days later, I began electrical stimulation at his suggestion. I used the treatment for 11 days and since have been in almost constant pain and have had great difficulty walking any more than very short distances, and now have difficulty climbing stairs. I have also had much muscle atrophy immediately following the treatment. I was an active biker, walker, swimmer, and skier until this treatment.

I would advise anyone who is seeking this treatment to take it slowly, if at all. It seems that other people have had some success with this treatment, but my quality of life has taken a nose-dive since trying it.

I would be very interested in hearing how others who have tried this treatment are doing.

—A.Z., Massachusetts
azirman@harvard.edu

Dear CMTA,

I came across a letter from T.R., Virginia Beach, who was asking about a lighter forearm crutch.

I recently ordered a pair of custom made forearm crutches that are very light and very comfortable. They are made out of a special aluminum by Thomas Fetterman of 1680 Hillside Road, Southampton, PA, 18966. Phone: 215-355-6941, email:tfetter@voicenet.com. Website:www.fetterman-crutches.com. He uses them himself. They also come in neat colors. I'm getting black ones with little purple streaks in them. President Clinton was using them for his bad knee. They come in several styles and prices. I tried them out at an "Ability Expo" in New Jersey. He had the standard forearm crutches there to compare in weight. There was no comparison. He also sells "crutch bags" that can easily attach to crutches so a person can carry essentials like a cordless phone or anything small needed in the house, or a wallet, or glasses, etc. out of the house. And, he sells easy-on and easy-off studded rubber ice grippers for canes and crutches, too.

This spring, I got tendonitis and bursitis of my hips, especially in what was my good leg,

and I cannot walk very well or very far now. I had to switch from one cane to two, but two canes aren't very practical and I kept dropping them especially when I had to use my hands to open a door or pick up or pay for something. I think forearm crutches will be better.

—J.R., New York

Dear CMTA,

I am a 45-year-old female, mother of two, wife and tutor living in Charlotte, North Carolina. I have had CMT since I was 29 and probably before then, unknowingly. I have had other injuries, as well, which puts me in the chronic pain syndrome group.

A neighbor of mine gave me a product called OPC-3. She started taking it for her fibromyalgia and it helped her dramatically. OPC-3 is a natural food supplement that is made up of three bioflavonoids known as oligomeric proanthocyanidins. They are grape seed extract, red wine extract, and pine bark extract. Bioflavonoids are complex plant compounds found mostly in fruits and vegetables. They function as a helper to the body's immune response to inflammation, allergy, and infection.

After 3 weeks, I have to say that I can do much more than before, while suffering less and sometimes not at all. I can still keep up with my swimming! If anyone would like to talk to me about this product, please call me, Emily, at 704-849-2367.

—E.L., North Carolina

Dear CMTA,

I live in England and have been in touch with the CMT organization in the UK, but have not found anyone with CMT III (the recessive form.) I believe the correct name for Type III is Dejerne-Sottas. James is four and was very floppy as a baby. He was a late walker, a late talker, and has never been very mobile. He is now in day splints and his feet have very pronounced arches. Without the splints, he can't stand still and falls over a lot. He is very small for his age and rather skinny. He also has bad eyesight and has to wear glasses. He is already deteriorating quite rapidly.

Daisy has always seemed less affected. She is a fairly steady walker, although not as advanced

as other kids her age. It is not confirmed that she has CMT yet, but her doctors feel it is a distinct possibility. James has had blood tests and has seen a physiotherapist occasionally. It seems difficult to get information from the doctors. They are the only members of the family affected by CMT.

I am looking for information and practical help. I would especially like to hear from families with a child diagnosed with Dejerine-Sottas.

I can be contacted at e-mail address:

—Sacha.tait@ocean.co.uk

■
This was a broadcast e-mail to all CMT public list members. Please respond if you have any information on this topic.

I am a dentist in the Department of Anesthesia at the Faculty of Dentistry, University of Toronto, Canada.

I have heard that those with CMT may have problems with mouth injections (freezing) that are given prior to dental work. These problems may include an inability or difficulty freezing a tooth, pain or swelling afterwards, a prolonged feeling of numbness, or anything that you or your dentist thinks may be out of the ordinary.

If you have any problem associated with local anesthesia in dentistry, please e-mail me at john.suljak@toronto.ca

Also, provide any details regarding your experiences. At present, there is no information about the effects of CMT and dentistry. Your input will provide the groundwork for a possible future study.

Thank you.

—John. P. Suljak, DDS
Department of Anesthesia

■
Dear CMTA,

I was diagnosed with CMT Type I after a serious car accident 4 years ago. Since that time, I have gone downhill quite a bit.

I am particularly interested in hearing from those individuals with CMT I whose condition has gotten worse as a result of an accident or other trauma. Please let me hear from you.

—Beverly Wurzel
914-783-2815 (home)

The CMTA reserves the right to edit letters for space.

Ask the Doctor

Dear Doctor,

What do you know about DHEA prescribed to counteract muscle loss? It is an adrenal gland hormone suggested to have muscle growth capabilities. It might have a negative effect in prostate cancer for men. It's sold over the counter and is written about in the alternative medical press.

The Doctor replies:

DHEA is certainly a substance used by alternative medicine and, currently, we advise caution in its use until more information, preferably scientific, is available. I would have low expectations for significant and sustained muscle preservation. The possibility of producing anxiety, nervousness, or sleeplessness in patients needs to be considered, also.

Dear Doctor,

I have CMTIII and CIDP (chronic inflammatory demyelinating polyneuropathy). My therapists have recommended I use night splints because of lack of dorsiflexion. They wanted me to purchase some from Maramed Orthopaedic Systems, but they look difficult to put on. My fingers are not strong and my dorsiflexion is 7" on one leg. Maramed's assume +10". I believe I will have a time trying to get it on and keeping my foot in it. Is there a particular brand that CMT patients should use?

The Doctor replies:

Night splints can be helpful in preventing contractures, but if difficulty is experienced in putting them on, a physiatrist, podiatrist, or physical therapist should be consulted about finding a more user-friendly brand. No one brand would be recommended for CMT.

Dear Doctor,

I have been diagnosed with CMT and my condition seems to be rapidly deteriorating. I worked as a parts person at a lawnmower shop. I am no longer able to stand. I am also starting to lose use of my hands at times. Does inhaling naphtha, carbon monoxide or working on concrete worsen or cause the onset of CMT?

The Doctor replies:

I would doubt that there is a significant occupational hazard in this case unless other employees are also sick from exposure to neurotoxic substances. Your situation should be discussed in detail with your local neurologist.



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

CORRECTION:

On page 7 of the Spring 1997 CMTA Report, we incorrectly identified Dr. Jack Petajan as Dr. Thomas Bird in the MAB meeting photo. We sincerely apologize for this mistake.

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, Macrochantin)
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.

What is CMT?

- ...is the most common inherited neuropathy, affecting approximately 125,000 Americans.
- ...is also known as peroneal muscular atrophy and hereditary motor sensory neuropathy.
- ...is slowly progressive, causing deterioration of peripheral nerves which 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.
- ...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 the focus of significant genetic research, bringing us closer to answering the CMT enigma.
- ...Type IA and CMTX can now be diagnosed by a blood test.

The CMTA Report

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



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Upland, PA 19015
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