

The CMTA Report

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

CMTA BOARD

Ann Lee Beyer
Chairman

Jack Walfish
President

William Quinn, DPM
Vice-President

David Grachek
Treasurer

Ardith Fetterolf
Corresponding Secretary

Krista Hall, RN
Recording Secretary

Robert E. Lovelace, MD
Chairman, Medical
Advisory Board

Dan Charny
Diane Freaney
Cynthia Gracey
Steven E. Khosrova
Stephen Scofield
J. Rodman Steele, Jr.

CMTA STAFF

Paul R. Flynn
Executive Director

Patricia L. Dreibelis
Director of Program Services

Georgette Farr
Administrative Assistant

EMAIL CMTA AT:
CMTAssoc@aol.com

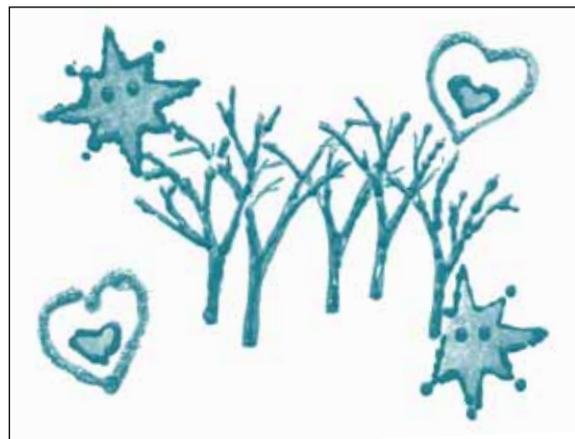
Greeting Card Program Kicks Off New Partnership with Kids In Business

Kids In Business, Inc. of Newark, NJ, will collaborate with the CMTA to raise awareness and funds for their respective programs. The first aspect of this new partnership will be the sale and mass distribution of the Kids In Business (KIB) greeting cards for the holidays. Proceeds from the sale of cards will be split 50/50 between the CMTA and KIB. Commenting on the greeting card program, KIB Executive Director Barbara G. Weiland had this to say: "Just as corporations form alliances to broaden their reach and increase their potential, so too must non-profit organizations join together for their mutual advancement."

Described as festive, colorful and unique, the art work for the greeting cards is truly charming.

The original designs have been chosen to launch the program. Paul R. Flynn, Executive Director of the CMTA has been a volunteer with Kids In Business for more than five years. He and Barbara Weiland realized that by combining efforts, many goals could be achieved. "The CMTA needs a tangible and simple means of raising awareness at the grassroots level," says Flynn. "The sale and use of these cards represents an effective way to begin informing people about CMT."

Kids In Business, which began more than 13 years ago, eventually wants to replicate its successful program elsewhere. KIB provides employment and learning activities to young people and adults to facilitate their learning about business, the workplace, and social responsibility through hands-on experience.



"Forest Fantasy" illustrated above is one of the children's original greeting card designs.

KIB participants give a portion of their profits to Newark charities. To date, they have given more than \$55,000 in profits and matching grants to help combat homelessness, poverty, illiteracy, and AIDS. The KIB school program serves Newark public school children in grades preK-8. The KIB Development Center offers after-school and summertime activities as well as employment for youth and adults.

Holiday greeting cards and note cards are the first aspect of the CMTA-KIB partnership. Plans are underway to market other KIB products in this "proceed sharing" arrangement. For more information, go on-line to either organizations' website: www.charcot-marie-tooth.org or www.kib.net.

Athena Diagnostics will "ring in the holidays" with KIB Greeting Cards!

Begin With Flynn

“Behold! I do not give lectures or a little charity. When I give, I give myself.”

—Walt Whitman, *Leaves of Grass*,
Song of Myself

It has been one year since I took on the role as the first Executive Director of the CMTA. I have learned and experienced many things. Foremost has been the remarkable willingness of members who have “answered the call,” to pitch in, to volunteer. Several have done so without being asked—they saw a need or an opportunity and plunged right in.

The Board of Directors leads this enthusiastic service. Working behind the scenes, committing countless hours, the Board’s often anonymous efforts are the force that propels this organization. I am grateful for their devotion and applaud their tireless dedication. Serving you more directly are our grass roots representatives—the support group leaders. Working in the community, our support group leaders promote awareness about CMT and the Association while serving local member needs. More have joined the ranks of this cadre of leaders, accepting hands-on responsibility for advancing the mission of the CMTA.

Other members have provided assistance working alone on projects or in conjunction with the Board and staff. I want to recognize the independent initiative of several volunteers for

their efforts toward raising money and/or awareness: Kim Colwell, Larry Gomez, Nancy Homyak, Dr. John Hsu, Rex Morgan, Dick Sharpe, Dr. Michael Shy, Buzz Van Almen, and Larry Wechsler.

With all that we are achieving, there still are many opportunities for the CMTA that slip away for lack of “human capital” and requisite stewardship. Given our rapid growth and the increasing needs of our membership, I hope to formalize a team of professional volunteers who offer very specific skills (event management, grant writing, marketing, and public relations) to bolster our office capacity. Of course, managing a team like this will require dedicated support from a volunteer coordinator, an individual I hope to locate in the Greater Philadelphia/Southern New Jersey area. Admittedly, we ask a great deal of our volunteers—that they give of themselves!

Athena Diagnostics Sales Competition: With the help of Vince Bertolino, Neurogenetic Product Manager at Athena Diagnostics, the sales force will be engaged in a marketing program that involves the distribution of CMTA literature to neurologists around the country. The program provides incentives for the sales reps to build awareness about CMT and our Association, and “competition points” for the recruitment of new CMTA members.

CMT Songfest: Staged in Shawnee, OK, by CMTA member Kim Colwell, CMT Songfest involved more than 15 individual performers and groups who sang country and gospel music throughout the day-long event. The talent was truly exceptional, including Kim! The event garnered a full-page story in the area newspaper, which “uncovered” some families with CMT. Kim will be a featured speaker at the kick-off of the Tinker Air Force Base workplace campaign, where she will promote the CMTA as a donor option. Kim’s advocacy role will also involve being interviewed on a cable ministry program, which she hopes will spark interest in the formation of a support group.

Board Officers and New Director: The board recently elected Ann Lee Beyer as Chairman and Jack Walfish as President. In our next newsletter, we will highlight these officers, newly elected board member Cynthia Gracey, and the “organizational vision” for the coming years.

Kim Colwell’s fundraiser in CMT Songfest featured Moccasin Trail (pictured below) and more than 15 other performers.



CMT FACTS IV NOW AVAILABLE!

The latest and largest of the *Facts* series is now available from the CMTA. *CMT Facts IV* is 32 pages in length and contains articles on medical information and research, practical ideas for living with CMT, children and CMT, and legal information, as well as the always popular, "Ask the Doctor" column. The publication will be \$8 for 1998-99 members of the CMTA and \$10 for all others, plus \$1.50 postage and handling.

The *Facts* series was instituted approximately 10 years ago as a condensed version of the factual articles from previous newsletters. This issue features articles from 1995 to 1998. The entire *Facts* series contains all of the important information published since the CMTA's inception.

This edition of the series contains a wealth of information regarding the nature and course of the disorder and would be especially useful for parents who need to present information to schools regarding the education and special needs of a child with CMT. Information from Shriner's hospitals is included along with an assessment of the orthopaedic problems of kids with CMT.

Titles include:

- Treatment of Familial Neuropathies
- Phrenic Nerves and Pulmonary Function in CMT Disease
- The CMT Disability Survey
- An Orthotics Survey of CMT Patients
- Managing Pain in CMT Disorders
- Orthopaedic Considerations for Children with CMT
- SSI Information and The Americans with Disabilities Act



CMTA MEMBERSHIP/ORDER FORM

Name: _____

Address: _____

Phone Number: _____

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

	QTY	COST	TOTAL
Charcot-Marie-Tooth Disorders: A Handbook for Primary Care Physicians		members \$15 non-members \$20	
Membership Dues		\$35	
CMT Facts I <input type="checkbox"/> English <input type="checkbox"/> Spanish		members \$3 non-members \$5	
CMT Facts II <input type="checkbox"/> English <input type="checkbox"/> Spanish		members \$5 non-members \$7	
CMT Facts III		members \$5 non-members \$7	
CMT Facts IV		members \$8 non-members \$10	
VCR Double Tape: Wayne State Conference		members \$25 non-members \$40	
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 Armington Challenge for CMT Research			
Shipping & Handling <i>Orders under \$10 add \$1.50, Orders over \$10 add \$3.00</i>			
TOTAL			

Check payable to the CMTA

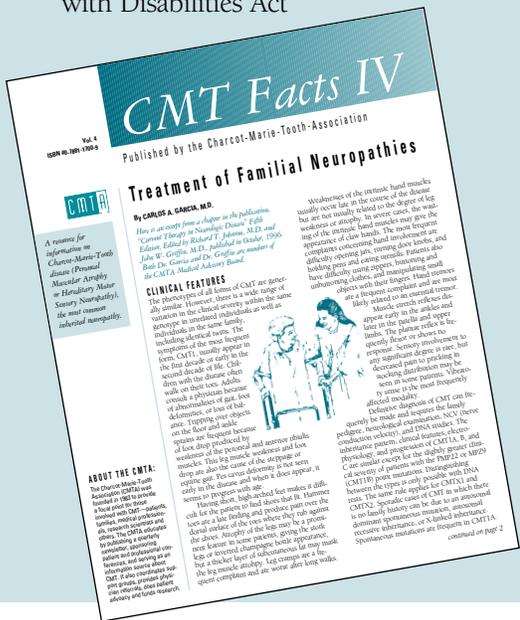
VISA MasterCard

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.



OF INTEREST

The **Hole-in-One Keyholder** from **Ableware™** provides two distinct grasping positions for right or left-handed individuals who have difficulty holding or grasping standard key heads.

It is designed to offer excellent leverage, simplifying the grip and movement required when using most keys. Fingers can be inserted through the hollow center of the five-inch-long handle, or wrapped around the outside. It also creates a brace for keys that are difficult to turn. The Hole-in-One accommodates one or two keys, which fit into slots on the front of the handle and are held in place by a single screw. When a key is needed, it simply slides forward. When not in use, keys can be folded back.

Maddak, Inc.
6 Industrial Rd.
Pequannock, NJ
07440-1993
Tel: 973-628-7600, ext. 9
Website:
www.maddak.com



Why HNPP Is So Hard to Diagnose...

By MAUREEN HORTON, R.N.

Before finally getting diagnosed with HNPP (hereditary neuropathy with liability to pressure palsy), people typically go through multiple tests, doctors, and diagnoses. The process can take months or years. With a genetic test easily available, it seems strange that it should be so complicated. Four factors play a large role in making HNPP hard to diagnose: most physicians still have not heard of it; symptoms may be intermittent; it varies in severity and symptoms even within the same family; and it mimics other diseases.

Typically, as the doctor listens to you describe your symptoms and examines you, she/he is considering what possible diseases you may have. For instance, if you have wrist and hand numbness, is it carpal tunnel syndrome? A strain? A growth? This is part of the “differential diagnosis.” If the physician has never heard of HNPP, it makes it nearly impossible to diagnose it because the doctor is not considering HNPP as part of a differential diagnosis. The symptoms may be diagnosed as something the doctor has heard of or the patient may be sent on to another specialist, for evaluation and testing, to figure out what is going on. (Now, repeat this process over and over until a diagnosis is made.)

Another problem facing people with HNPP is deciding to go to the doctor at all. Most people with HNPP have intermittent pressure palsies (periods of numbness and weakness) lasting minutes to weeks. The symptoms may disappear between the time the doctor’s appointment is made and the date of the appointment. Do you go in? Will the doctor take you seriously if you don’t have symptoms? Should you wait and see if it happens again? If the problems are bothersome enough, that is probably enough to send you to the doctor. If you do go in, does the doctor want to take a “wait and see” approach or pursue the problem?

When you do see a doctor, she/he takes your medical history and checks you over. In taking a history, the doctor asks questions about your chief complaint—why you are there. What is bothering you? How long has it been going on? What makes it better? What makes it worse? Again, as this is going on, the doctor is comparing what she/he sees and hears to symptoms of known diseases. Another part of a history is asking about your family to determine the possibility of a hereditary disease. HNPP

Four factors play a large role in making HNPP hard to diagnose: most physicians still have not heard of it; symptoms may be intermittent; it varies in severity and symptoms even within the same family; and it mimics other diseases.

has many different symptoms. Maybe your father had carpal tunnel problems and you have back and leg problems. It would be hard to link the two seemingly different symptoms as something that runs in the family. Yet, both can be signs of HNPP.

Another stumbling block to diagnosis is that HNPP can look like other diseases that the physician has heard of. It may look exactly like carpal tunnel and the EMG confirms it (90% of people with HNPP are estimated to have carpal tunnel syndrome). Why look any further? Or, maybe the diagnosis is Bell’s palsy or a lumbar disc problem. Again, why look further? Another tidbit: A neurologist may suspect alcohol-related peripheral neuropathy after reviewing the unusual EMG. So don’t be surprised if you are asked more than once about alcohol use.

HNPP was only discovered 49 years ago (the genetic problem, 5 years ago). For most of that time, it was thought to be a very rare disorder, so there was little need to educate all neurologists. Now, HNPP is believed to be much more common. It is time to educate the neurologists and primary care doctors as well. It is time for all of us to learn more about this disease. We all need to work together on this. Educate yourself. Read. Research. Find doctors who can educate you. Then educate all your doctors. As more doctors become aware of this disorder, more people will be diagnosed properly.

Author’s postscript: I was treated for lumbar disk problems and then carpal tunnel problems as two separate disorders. It took 8 years and the development of problems in my relatives to finally be diagnosed. It took another 11 years to finally be educated about HNPP.

Regional Conference Held in Downey, California

Nearly 150 people attended a conference on Charcot-Marie-Tooth disorders held on Saturday, June 27, 1998, at the Rancho Los Amigos Medical Center in Downey, California. One of the highlights of the conference was the opportunity for the attendees to share their personal experiences with CMT and exchange ideas. Another highlight was the personal care and attention provided by conference host Dr. John Hsu and other presenters.

Following Dr. Hsu's morning presentation on orthopaedic procedures for the foot and ankle, he conducted a "mini" clinic: performing gait analysis, assessing foot and ankle deformities, and evaluating orthotics. Most of the surveys that were returned to the CMTA cited the person's appreciation of that personal consultation with Dr. Hsu.

Other well-received presentations included one on social security disability and the application process, an overview of the Athena Diagnostics blood tests, a discussion of the on-going work at Rancho Los Amigos regarding gait analysis, and a review of the CMTA's commitment to funding research.

The workshop concept was so well received that it was difficult at times to get people to leave the workshop and move to the next



scheduled portion of the program. That fact was a compliment to the presenters at the alternative therapy workshop, the orthotics and pedorthics workshop, the physical and occupational therapy workshop, and the pain management workshop.

By the conclusion of the meeting, interest was very high in establishing a support group in the Southern California area and plans are currently underway to get such a group started. If you are interested in helping to form a group in that area, please contact the office.

For anyone who attended the conference or who lives in Southern California and would like to contact the presenters, the following list includes the presenter and his or her phone number.

Dr. John Hsu 562-401-6694
 Lori Rowley, OT 562-401-6271
 Jan Furumasu, PT 562-401-6800
 Sara Mulroy, PhD, PT 562-401-7177
 Darrell Clark, CO 562-401-7655
 Charles Marsden, CPed. 562-693-6261
 Dr. Joseph Sciabarrasi
 (Alternative Therapies) 310-395-2453
 Alice Harmon,
 State Social Security Rep. 213-736-7006
 Craig Kollenda, Athena. 800-394-4493

Ardith Fetterolf, organizer of the Downey Conference, poses with Board member Diane Freany and Dr. John Hsu, orthopaedic surgeon from Rancho Los Amigos.



Dr. John Hsu conducts a "mini" clinic for attendees at the Downey Conference at Rancho Los Amigos.

OF INTEREST

The August 1998 issue of *Seventeen* Magazine contains an article written by Jenn Wilson about her boyfriend, CMTA member, Benjy Feen. The article is entitled “not-so-perfect boyfriend” and focuses on Jenn’s contention that there’s “lot’s more great stuff to my guy than meets the eye.” The article concludes with Jenn’s observation that “Having Benjy in my life is the best. Not only is he affectionate and a great listener; he’s a supreme blast to be with. His sense of humor rules, and, as Benjy told me the first time he asked me out, he gets the *best* parking spaces.”



The Invisible Heroes of CMT

By **DANA SCHWERTFEGER**

In a recent article, Robert Lipsyte, a reporter for *The New York Times*, took up the issue of patients’ rights. Too often, he wrote, patients make the journey alone through the “country of illness,” where they are “scared and sick, vulnerable to bullying billing clerks, snide receptionists, overworked technicians, and doctors and nurses who can rarely take the time to listen.” Mr. Lipsyte went on to describe how each illness has its own group of advocates who compete for money and attention, and he wondered how much better it would be if we all joined forces and formed a patients’ union. Then, much to my surprise, he suddenly referred to “the almost invisible heroes of Charcot-Marie-Tooth.”

Hold on a second. What’s the connection between CMT and heroism? When I think of heroes, I think of soldiers who risk their lives, and who sometimes die, fighting for their country. I think of firefighters who run into burning buildings to rescue people. I think of people who lay everything on the line for what they believe in. And, I think of outstanding athletes like Babe Ruth and Michael Jordan. But me? A hero? All I’ve ever really done and all I’m ever likely to do is muddle along, day after day, coping with the obstacles CMT puts in my path and trying to live as normal a life as I can.

There are days, I’ll admit, when it seems like

it takes an heroic effort merely to get out of bed. Just once, I’d like to be able to get dressed without fumbling with buttons or having to strap on AFOs. The reality, however, is that my fingers don’t follow instructions very well, and for 20 years, I haven’t been able to take more than a few ungainly steps without my AFOs. When it’s cold, or even cool, my fingers become even more useless, and when it’s hot and muggy, wearing AFOs is a torture of which the Marquis de Sade would approve. Still, recalcitrant buttons are a relatively minor hindrance and putting on AFOs is as natural to me as putting on my pants. So, there must be more to heroism than battling tiny buttons and wearing AFOs.

Maybe it’s the trouble I go through to accomplish what people with an opposable pinch are able to do with much less effort. It’s a fact that fixtures, tools, and you name it—the majority of the things we use every day—were not designed for use by someone with CMT-related hand involvement. Ever notice that keys meant to be turned with thumb and forefinger can be impossible if the lock isn’t well-oiled? Smooth door-knobs can be troublesome, too, as can many bathroom fixtures, especially when your hands are soapy. How about toothbrushes, tweezers, fingernail clippers, and the like? I’ll guarantee they weren’t designed with CMT in mind! I’ve given up trying to use fingernail clippers, but I can manage with toenail clippers if I grip them in

GIFTS WERE MADE TO THE CMTA IN MEMORY OF:

Krystal and Ashley

Ruth Stefonovich

Mary Bastone

Paul R. Flynn
Joseph G. Tucci

Norma Jeanette Bloom

Donald and Jeanette Anstead
Corene and Marilyn Bloom
Merlyn Bloom and Family
Barbara Caveslio
Carrol Knipp and Family
Pauline Lamale
Betty Ling
Evelyn Miller
Robin Paul
Violet Welley

Sophia Lampert Bradwick

Faye Bradwick

Edward Coogan

George Gomez

Les Crawford

Thomas and Nancy Greve

Norma Ewing

Mary D. and Bob Williams

Hy Fetterman

Bob and Bev Warner

Leon Finegold

Mr. and Mrs. Leonard Zobel

Ben Fiscella

George Gomez

Jack Goldman

Kay Flynn
Doris M. Goldman
Celso Luis Lugo

Herbert Grossman

Irma and Jack Jaffe
Julie Leonard

Stanley Hull

Barbara Austin

Stanley Hull (continued)

Mr. and Mrs. Daniel Basta
Richard and Dorothy Deppen
Mrs. K. S. Kidd
Matthew Oehlmann and Family
Ted and JoAnn Parsell
Kenneth and Dorothy Richardson

Donald I. Le Furge

Irene and Henry Buss
Mel and Nina Buss and Family
Mrs. E. Hart
Mrs. Helen Horning
Sam and Pattie Perry and Family
Phyllis and Roger Wildey and Family

Hazel Lester

Mary D. and Bob Williams

Helen Lynch

J. C. Julian

Marie Miller

Jerry Sharp

my fist—not quite how they were intended to be used, but living with CMT often means finding a different way to use something, however awkwardly, to get the job done.

There was also a time when I didn't think twice about using a knife and a fork. Now, it's always a challenge, but I don't want to appear helpless, so I won't ask someone else to cut my food. At home, when I slip and send food skittering across the table, I simply scoop things back onto my plate and try again, but when I lose control of my food in a restaurant, it's a different matter altogether. Unless I tell them, people often don't know I have CMT, and even when I do, they have difficulty fully comprehending what it means. I know people must either think I've had one too many cocktails before dinner or that I'm just a clumsy oaf.

While I'm thick-skinned enough not to be terribly embarrassed by what other people think, I have to agree with Susan B. Adams, whom Mr. Lipsyte quotes in his book, *In the Country of Illness*, as having written, "Sometimes I even think it would be easier to be clearly disabled... At least then, there'd be no pretending, no denial, and no need to put on a cheerful face." Mr. Lipsyte adds that, "Putting on a cheerful face takes an enormous amount of energy... and it is mostly about making other people comfortable." That's certainly true enough.

However, while I don't feel I have to explain my disability to anyone or make other people comfortable, I'm going to keep putting on a cheerful face no matter how much energy it takes. For me, confronting the challenges I face

every day because of my CMT is a matter of self-respect. Okay, pride and stubbornness, too, perhaps, although I do know when to quit and ask for help. But does that make me a hero?

I think maybe it does. If I didn't struggle, if I didn't attempt to do the things that other people can do with far less effort and with far less discomfort, then perhaps life would be easier. Most of us can always ask other people to do things for us that, with a little extra effort, we could do for ourselves. So, why take the easy way out when we can be heroes just by confronting and overcoming the hurdles CMT places in front of us? After all, it's easier (and safer) than running into burning buildings!

It's not nearly as noticeable, however. The trials and tribulations of living with CMT are, as Mr. Lipsyte implies, practically invisible. Even our families and friends don't always know what we go through every day. And what about the people we pass on the street or sit next to in a restaurant? They may notice that we walk a little strangely or that we seem a bit awkward when it comes to using a knife and fork, but they don't have a clue what it's like to have CMT. They don't know what a herculean effort it takes some of us just to walk a few blocks or to cut a piece of steak.

While being clearly disabled would allow others to acknowledge our efforts. I believe it's enough to know in our hearts that we have done something heroic, something of which we can be proud. For, invisible though our heroism may be, when we rise to the challenge of CMT, we ARE heroes.

Josephine O'Grady

John O'Grady

Maki Papavasiliou

Mireille Masson

Marilyn Henry Richardson

Mary D. and B.D.B Williams

Dianne L. Richert

Estelle Waitish

Mrs. Brucie Glenn Rowe

Clay G. Battle

Rebecca Sand

Percy H. Sand

L. June Sim

Marilynn Dodge

Dan Solomons

Robert Warner

Chester M. Sporka

Julia E. Sporka

Edna C. Comstock Summers

Carolyn M. Ahrendt

Mr. and Mrs. H. D. Stevens

Shirley Terry

Karen Rimatzki

Daniel Reese Townsend, Sr.

Ellen T. Burrow

Alan Wechsler

Home Office—A Unit of the
Hearst Corporation

Hugh W. Wilkins

Mr. and Mrs. William H. Anderson

The CMTA lost a dear friend and a loyal supporter this summer when Karen Edelson was killed in an automobile accident. Karen had been a support group leader of the Greater Dallas Area, as well as a practicing podiatrist. The staff and Board of the CMTA send their sympathies to Karen's family and friends.

Dr. Karen Edelson

AAPEX Family Foot Specialists, P.C.

The Altabet Family

Janice Bohannon

Dr. Karen Edelson (continued)

Jeff and Jane Claes

Ethel and Norman Cooper
Dallas County Podiatric Society

Edith and Al Geisinger

Nancy Golar and Jorge Lange

Philip and Betty Heiberger

Terry Heller

Lisa and Timothy Hickey

Frank and Fran Ivker

Dr. and Mrs. Jay Lifshen

Celia and Aaron Lukoff

Medical Staff of Medical Center of
McKinney

Stephanie and Jeffrey Oelke

Bernie and Stan Oswald

Mrs. Gerry Quail

Flo, Harold and Alan Schutzberger

Jack and Jeannine Sellmeyer

Flossie Solomon

Staff at PS 178, Bronx, NY

Texas Podiatric Medical Assoc., Inc.

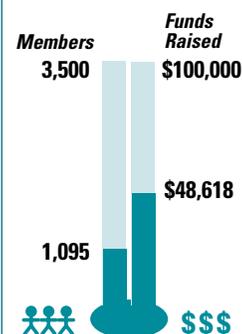
Women's Health Boutique

Leah and Herbert Zarge

Strength in Numbers

MEMBERSHIP GOALS:

OUR 1999 GOALS



IF YOU GIVE AT THE OFFICE:

Federal and some state employees have the opportunity to support the work of the CMTA through payroll deduction in their workplace through the Combined Federal, or state, Campaign. The Charcot-Marie-Tooth Association is listed under Health Charities of America on page 18 of the national directory.

The CMTA can also receive donations in most United Way campaigns. If your area offers donor choice, ask for a donor card and write in The Charcot-Marie-Tooth Association, 601 Upland Avenue, Upland, PA. 19015. We are a federally recognized 501(c)(3) charity and can receive funds through workplace donations.

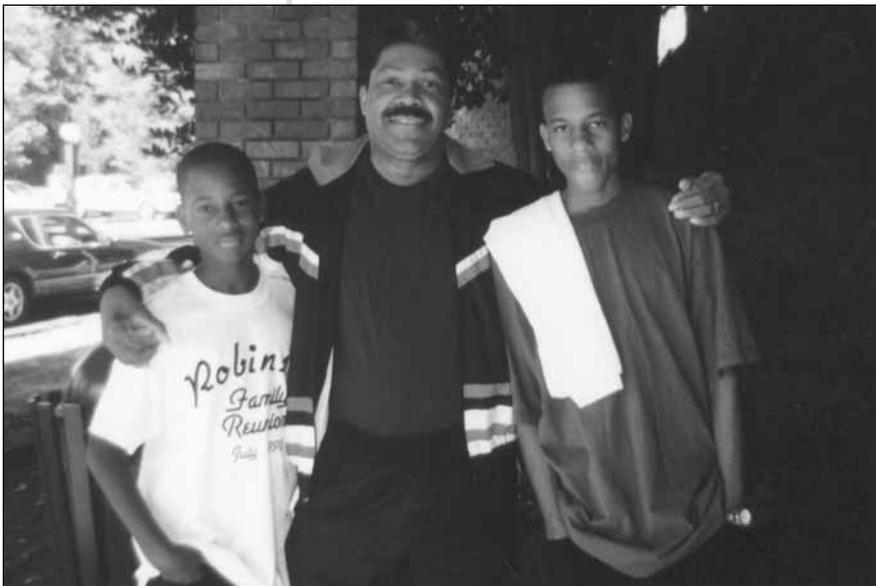
Robinson Family and U of Miami Combine to Advance Research

On Friday evening, July 31, 1998, Dr. Lisa Baumbach spoke to members of the Robinson family at the Best Western King's Quarters in Richmond, VA. The family was gathered there for a reunion and Lisa was there to explain to the family the important role they could play in advancing her research on the unique polymorphism* she believes is expressed in the African-American population.

The Robinson family offered an exciting opportunity for a genetic study of an African-American family because they are a large family with many affected members. First contact with the family came when James Robinson attended the CMTA's patient/family conference in St. Louis, MO, and volunteered to give blood for Dr. Baumbach's study on the different polymorphism found in CMT1A African-American patients. Mr. Robinson is mildly affected. He has six sisters and three brothers and several of his sisters have a long history of foot problems. James also has two nieces and a nephew living in the Philadelphia area who have been diagnosed with CMT.

Between 60 and 70 family members came to the reunion and samples were collected from 32 family members, spanning four generations (22 were blood samples and 10 were cheek swab samples, only). Blood and cheek swabs are processed for DNA; the swab samples are helpful for youngsters and older adults from whom it is difficult to draw blood.

James Robinson, organizer of the Robinson family's "contributions" to research is flanked by his nephews Joshua Rose (left) and Isaiah Rose (right).



Dr. Lisa Baumbach takes a swab sample from Anthony Buffini, while his mother Beverly Buffini holds him.

Dr. Walter Nance and Dr. Jeff Ellison, from the Medical College of Virginia, came to the reunion to help with neurological examinations and medical interviews. A phlebotomist from a Lab Service and Dr. Jeff Best from the Washington Hospital Center Podiatry Surgical Program in Washington, DC, rounded out the professional staff. It was, however, the hard work of James Robinson and his niece, Gloria Glaze, who made all the family arrangements and promoted the necessity of participating in the study, that resulted in this cooperative endeavor. The work of the University of Miami was supported by a postdoctoral grant to Mary Ellen Ahearn from the CMTA.

As of September, all the blood and swab samples have been processed for DNA and tested for the CMT1A duplication and the African-American polymorphism. The final results of this study will be presented at the Third International Conference on CMT in Montreal in October.

* A polymorphism is a genetic variation that differs among people and was once thought to be "benign" or having no effect on the DNA. The polymorphism being investigated in the Robinson family is very specific for African-Americans and seems to indicate that polymorphisms can change the protein structure in DNA.

BOOK REVIEW:

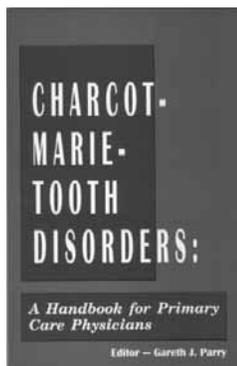
Charcot-Marie-Tooth Disorders: A Handbook for Primary Care Physicians

By David Hilton-Jones, Oxford, United Kingdom

I was very pleased to hear of the existence of this book, for I think that too little has been written about this common disorder in the non-specialist press. It is published by the North American Charcot-Marie-Tooth Association (CMTA) supported by a grant from Athena Diagnostics, who have a discreet one page advert towards the end of the book.

One of my colleagues recently suggested to me that CMT is an “over-medicalised” condition, presumably implying that for many it is a relatively trivial disorder not meriting a great deal of medical input. Having seen many patients, some late on in the course of their disease, who have in the past been given grossly incorrect information about their condition by physicians with no specialist knowledge, and many others who have had inappropriate surgical procedures, not infrequently without the diagnosis of CMT having been established or even thought of, I had to disagree.

There are 10 major chapters which between them cover Clinical Features, Electrodiagnostic Studies, Pathological Changes, Genetics, Features in Children, Orthopaedic Considerations and Management, each by a



different author. Inevitably there is some overlap between chapters and some inconsistency of depth of coverage.

Although their quality is a little variable, the book is well illustrated with photographs, diagrams, and tables, all of which add to its value. **Most doctors will benefit from reading the chapters on physical, non-surgical approaches to therapy, especially the one by a podiatrist which is particularly well illustrated.**

Despite some criticisms above, I think that this is a very welcome addition to the literature concerning CMT. In the UK, I can not see many general practitioners reading it. The main audience will be therapy staff, and I hope, specialists (neurologists, rheumatologists, orthopaedic surgeons) who have dealings with CMT. **I also believe that some patients and families will find it of value and I have started recommending it to them.**

Bolded type reflects editor's emphasis.

(Editor's note: This review was published in Neuro-muscular Disorders, Vol. 8(1998) 59-64. A copy of the complete review is available from the CMTA office. It was edited for space.)

GIFTS WERE MADE TO THE CMTA IN HONOR OF:

R. Gordon Bradwick

Faye Bradwick

Christine and Gerard's Anniversary

Alfred and Gisela Schwab

Stephanie M. Di Cara

Estelle Waitish

Grace Kestler

Ken Dunn
Robert and Phyllis Nussmeier
Robert Nussmeier
Mr. and Mrs. Dick Whitley

Julie Leonard's Birthday

Ellen and Sam Jaffe

Charles Lynch

J. C. Julian

Tom Lynch

J. C. Julian

McDonald Family Members in the US and Canada

Frances J. McDonald Hill

Rev. W. K. Schmidt

Joni and David Schmidt

Dr. and Mrs. A. Schreiber

Mr. Jack Silver

Samantha Sheriff

Mary W. Headrick

Mrs. A. Hart Wurzburg

Mrs. Hugo Sonnenschein

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 occurred.

Armington Research Challenge Ex

Medical professionals are encouraged to support the CMTA both financially and in promoting membership to their patients. To acknowledge the contributions of our professional members, we send out certificates to be posted in their offices.

The Evenor Armington Foundation “challenged” the CMTA membership to elevate its commitment to funding research—and you responded! Having raised more than \$150,000, the CMTA will receive a grant from the Armington Foundation for \$50,000 toward CMT research. In three years, this brings the research funds’ total to nearly \$400,000. The final year of the Armington Challenge Drive commences in 1999...

Sustaining Membership

Achieving organizational goals requires significant support beyond that of general membership. To sustain the mission of the CMTA, leadership commitments are needed. The supporters listed below gave a minimum of \$250 in response to the Membership Appeal. We are pleased to acknowledge the generosity of these members.

Richard Brunetti
Robert E. Buuck
Alice T. Cross
Joseph S. Fiore
Paul R. Flynn
Jack A. Graves

Jill Huskisson
Peter T. Jackson
John R. Kathrein
Doris L. Lancaster
Douglas S. Trower
Kathleen Weinstein

Professional Memberships

Dr. Peter C. Conglis
802 Minor Drive
Escondido, CA 92025

Jeffrey M. Leahy, PT
3494 Craig Ave.
Cincinnati, OH 45211

Michael Fritzen, PT
1061 Easton Rd., Apt.11C
Roslyn, PA 19001

Dr. Howard Petusevsky
7769 NW 44th Street
Sunrise, FL 33351

Dr. Joshua Gerbert
16 Fairview Ave.
Corte Madera, CA 94925

Donald E. Robinson, DPM
526 Camden
San Antonio, TX 78215

Amy Gutierrez
LSU School of Medicine/ Neurology
1542 Tulane Ave.
New Orleans, LA 70112-2822

J. Gregory Stilwell, DPM
Kerry E. Berg, DPM
Inter Mountain Foot and Ankle
5353 N. Union, Ste. 101
Colorado Springs, CO 80918

Dr. Steven Hinderer
Rehab. Institute of Michigan
261 Mack Ave. Ste.19
Detroit, MI 48201-2418

Dr. Bonnie Tatar
5750 Centre Ave. Ste. 480
Pittsburgh, PA 15206

Keith Kashuk, DPM
6350 Sunset Drive
South Miami, FL 33143

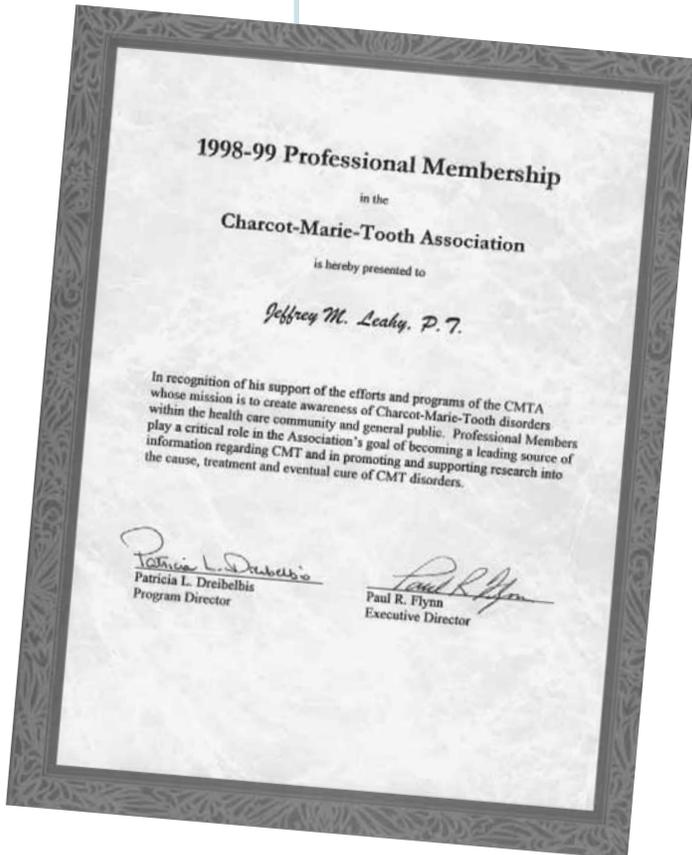
David S. Ungar
Personal Foot Care
34435 Grand River Ave.
Farmington, MI 48335-3309

Dr. Michael R. Kaye
1080 Hwy. 35 South
Forest, MS 39074

James V. Vitale
Maui Family Footcare, Inc.
415 Dairy Rd. Ste. D
Kahului, Maui, HI 96732-2312

Roger W. Kula
LICH Neurocare, PC
339 Hicks St.
Brooklyn, NY 11201-5514

Dr. Lowell L. Williams, retired
Dr. Robert L. Schneider
Box 715
Cashmere, WA 98815



ceeded: \$151,119 Raised

PEOPLE GIVE TO PEOPLE, FOR PEOPLE

CMTA member Mark DiCara, Esq. wants to help his young daughter and advance the goals of the Association. His daughter, Stephanie, who we will learn more about in an upcoming issue, inspired a very generous and thoughtful gesture by one of her dad's clients. Stephanie has an unusual and debilitating form of CMT. Mrs. Estelle Waitish knows of the DiCara Family's struggles because Mark is her attorney. Wanting to help the organization that helps the DiCaras, Mrs. Waitish gave \$500 to the CMTA which will be matched by the foundation of her former employer, The Illinois Tool Works.



The gift is matched 3 to 1 for a total of \$2,000.

This act of generosity is significant for several reasons. It demonstrates the potential for the CMTA to garner support from those "unaffected" by CMT. More importantly, Mrs. Waitish's contribution affirms the axiom of philanthropy: that people give to people, for people. We must continually seek ways to "put a human face" on this unknown disorder. Stephanie is a perfect example of this.

Seven-year-old Stephanie DiCara was the inspiration behind a very thoughtful and generous contribution by Mrs. Estelle Waitish.

September 24, 1998

Dear CMTA.

This donation is being made in honor of Stephanie M. Di Cara. Stephanie's dad is my attorney and that's how I found out about this disease that is afflicting his daughter. Being such a rare disease, it doesn't receive the attention of the media, like AIDS and cancer do.

So, I figured that I would rather do something that is more important for the people you are helping than my getting recognition from a bunch of jaded solicitors.

May God bless and smile on your work.

*Very Sincerely,
Estelle Waitish*

Workplace Campaigns Begin

For several years, the CMTA has been listed as a "donor option" within the workplace fund drives coordinated through the Combined Federal Campaign and the Health Care Charities of America, as well as certain state programs and United Way drives.

CFC Campaign Regions pledged \$27,781 to the CMTA in the Fall of 1997. Of these

pledges, the CMTA can expect to receive between 75% and 80% of the total. This reduction is a factor of campaign administrative costs and "campaign shrinkage" (i.e. unfulfilled pledges). Ten regions contributed more than \$500, while four regions contributed \$1,000 or more. The top four regions were: National Capital Area (\$5,436), Kitsap-Mason

Counties (\$1,579), San Francisco Bay Area (\$1,132), and San Diego County (\$1,128).

If you, a member of your family, or friends of yours participate in workplace campaigns, please consider promoting the CMTA by distributing our literature or by asking colleagues to match your support.

Thank you!!

■ OF INTEREST

Texas Scottish Rite Hospital for Children will hold a CMT children's clinic, November 25, 1998, from noon until 5:00 PM. The clinic is located at Winter Clinic, 2222 Wellborn St., Dallas, TX. Children with CMT who are not patients of Scottish Rite Hospital need a referral from their family physician in order to be seen at the clinic. For further information on the clinic, call 214-559-5000. Ardith Fetterolf, a member of the CMTA's Board of Directors, will be at the clinic to answer questions about the association.



CMTA Support Group News

A new support group is forming in southern Florida in the region of Miami and Ft. Lauderdale. The group will meet at the North Broward Medical Center in Pompano Beach, Florida. The support group leaders will be Al and Marilyn Kent. They would like all interested members to call them at 954-742-5200 (daytime) or 954-472-3313 (evenings). The group held its first meeting on August 29, 1998.

An older support group has been meeting for years, but was inadvertently omitted from the newsletter listings. The group meets in the Willamette Valley region of Oregon at the Brooks Assembly of God Church. They meet the third Saturday of every month except December. Contact Marianne Destefano-Hill at

503-585-3341 or Regina Porter, 503-591-9412.

The first meeting of the Metroplex Charcot-Marie-Tooth Support group was held on September 24, 1998, from 7:00 to 8:30 PM in the Harris Methodist HEB Hospital, 1600 Hospital Parkway, Bedford, Texas. The group serves the Dallas/Ft. Worth area. The leaders are Greta Lindsey (817-281-5190) and Shari Clark (817-543-2068, metro). If you missed this first meeting, please call one of the leaders to inform them of your interest and to have your name added to their mailing list.

A new group is also forming in the Pittsburgh, PA, area. The leaders will be Jay Budde and Don Peebles. Call Jay Budde at 412-323-1400 for more information.

Support Group Highlights

■ **Kay Flynn** in Westchester County, NY, is sending out a questionnaire to members in the New York area, trying to identify how CMT impacts lives, families, and kids.

■ **Angie Piersimoni** in upstate New York had eleven people at her last meeting. Persistence pays off!

■ The **Berkeley, CA**, group met in July and developed goals of starting a teen and youth group, fundraising, and doing some social things once in a while! Miriam, one of the co-leaders, is off to visit family in Florida for a while and Ruth Levitan could use some help with the group. Please consider volunteering.

■ 25 people attended Ellen (Nora) Burrow's July meeting in **Archdale, NC**. Speakers were two people representing orthotics and prosthetics. Their special guest was Miss Ashley Matlock, a finalist in the Miss North Carolina Pageant. Ellen would like to do a CMT/CMTA cookbook. She needs recipe donations. Proceeds would go to the CMTA. Please call the CMTA office at 1-800-606-2682.

■ **Bob Budde** had another good meeting in Kentucky. He brought a family contingent in from Pennsylvania to show his group off.

■ Our **Kansas City, MO**, group had a chiropractor in as a speaker for their last meeting who specializes in a nonsurgical approach to

treating carpal tunnel syndrome. Keep those alternative practices coming. We need to know what's out there!

■ Also in **Missouri**, there is a very interesting lady named Donna Clark who has identified many people with CMT, from her work in a shoe store in Versailles, MO.

If your group is doing something special, contact:

Kay Flynn: Northeast and Mid-Atlantic
Walt Sawyer: North Carolina to Florida,
to Louisiana

Sue Tarpinian: North and Central U.S.
West Coast: Open, but you may contact
Krista Hall

We are planning a Support Group Leaders' national meeting sometime early next year. Your regional leaders will be contacting you. We want to have your thoughts and ideas on when, where, and what! So please respond when they ask.

I had the opportunity to host a meeting last June with our Regional Leaders in Philadelphia. I believe we learned a lot from each other. If you have ideas or concerns about support groups call the CMTA office or email me at lehall@ix.netcom.com

Thanks for all you do out there to help each other. May the force be with you,

—Krista Hall, CMTA Support Group Liaison

CMTA Support Groups

Alabama/Greater Tennessee Valley

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

California—Berkeley Area

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

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

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: Marianne Destefano-Hill (503) 585-3341 or 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)

ARE YOU MOVING???

If you're moving or have moved already and this issue has been forwarded, PLEASE notify the office of your new address. It costs the CMTA a few hundred dollars with each issue to get updated addresses from the post office, along with the additional postage to resend the newsletter. Thank you for your help in "containing costs."



Best Wishes

The good wishes of the Charcot-Marie-Tooth Association go out to our long-time California-North Coast Counties support group leader, Freda Brown, who was injured in a car accident while crossing the street on her scooter several weeks ago. She sustained head injuries and is in rehab. Cards of well-wishes can be sent to the office. They will be forwarded to her family, who will see that she receives them in the rehab center.

■ OF INTEREST

The **Reader's Window** is the first, and only, book holder that actually suspends books above eye level, letting you read comfortably and more efficiently in a recliner or bed. It allows you to rest your neck, back and arms, allowing your mind to focus purely on reading. Turning pages is easy using the device. The book's cover is held against a wooden easel by large Velcro straps. Pages are held by a clear, seamless, optical-quality shield. Moving the shield turns the pages of the book. The shield is guided by a uniquely patented "rack" that's integral with the easel. The frame is crafted of solid ash hardwood, stained as oak or cherry.

Howell Design Inc.
192 Thomas Lane
Stowe, VT 05672
1-800-867-7869



A Journey Toward Wellness, Part III

By CYNTHIA GRACEY

In my last two articles I have tried to provide a context for embarking on an alternative, complementary, or even radically different healing journey. The first article was designed to offer hope that alternative healing modalities existed and that they could be effective. The second article explained that not all of these alternative, complementary, or "radical departures" are going to work for everyone, but one way of ensuring maximum success in your choices is to know your personality type. The more you know how you best energize, process, and receive information (be it physical, emotional, mental, or spiritual), the better equipped you will be for making quality decisions regarding your healing journey. With that as background, I will describe what has been most effective in my pursuit of greater and increasing vitality in my body.

- Breathwork, including yoga breathing, rebirthing, and a synthesis of the works of Stan Grof, MD, and Jacqueline Small, MSW
- Deep body work including Rolfing, cranial sacral therapy, Traiger, and deep neuromuscular therapy
- Visualization
- Diet
- Iyengar yoga

- Aligning the realms of my life with my values and my sense of destiny
- Pursuing that which reminds my body how good it feels to have a body
- Learning new and, sometimes radical, ways of energizing my body
- Learning how to create a sanctuary for my own healing process whether at home or away

Many of the above have a common denominator. Properly practiced, they have the ability to assist me in generating the energies of passion, bliss, and deep peace in my physical body. I have come to believe that these are healing energies, which can create new ways for neurological intelligence to be transmitted through the body.

My first and most profound experience of this was through working with, experiencing, and then studying, the technologies of the "healing breath."

Jacqueline Small, a renowned therapist, author, and group facilitator, has said, "Our greatest healer is right under our nose, moving in and out—our breath."

When coming through a stressful experience, you may notice that we sigh and take a deep breath and our body relaxes. Breathing is

Suggested reading list

THE BREATH

Hendricks, Gay, PhD. *Conscious Breathing: Breathwork for Health, Stress Release, and Personal Mastery*, Bantam, New York, NY, 1997.

Ramacharaka, Yogi. *The Science of Breath*, Yogi Publication Society, Chicago, IL, 1904.

Zi, Nancy. *The Art of Breathing: Thirty Simple Exercises for Improving Your Performance and Well-being*, Bantam Books, New York, NY, 1986.

PERSONALITY TYPING

Kiersey, David and Marilyn Bates. *Please Understand Me: Character and Temperament Types*, Prometheus Nemesis Book Co. Del Mar, CA, 1978.

Kroeger, Otto, and Janet M. Thuesen. *Type Talk: The Sixteen Personality Types That Determine How We Live, Love, and Work*, Dell Publishing, New York, NY, 1988.

Lawrence, Gordon. *People Types & Tiger Stripes*, Center for Applications of Psychological Type, Inc. Gainesville, FL, 1979, 1982, 1993.

Myers, Isabel Briggs with Peter B. Myers. *Gifts Differing: Understanding Personality Type*, Davies-Black Publishing, a division of Consulting Psychologists Press Inc., Palo Alto, CA, 1980.



an anchor, or focus, that is commonly used in many traditional forms of meditation and reflection. Focusing on the breath is an easy and natural way to quiet both mind and body.

“Breath is the bridge which connects life to consciousness, which unites your body to your thoughts.”—*Thich Nhat Hanh*

The Miracle of Mindfulness, p.15

I had learned about many forms of breathing to quiet my mind, balance my energy, and relax. I learned these techniques studying different types of meditative and yoga practices. None of these techniques, however, were doing anything noticeable in slowing down the inexorably steady pace of degeneration that was occurring in my legs and arms.

It was not until I put my extreme skepticism aside and underwent a series of Breathwork sessions called rebirthing that I experienced profound changes in my body, my energy levels, and my experience of being “in” my body. It felt like a “rotor roter” of energy was moving through my body either dynamiting away the energy blockages that were developing in my neurology, or, if that was not possible, creating new neuropathways for the necessary information to move through my body. I actually had moments where I experienced my body as a blissful place to be. I had more energy in my legs and arms. The protein levels in my blood went down. I began to exercise and then slowly jog and finally, after several years, run. I studied breathing techniques with every teacher I could find.

Finally, I began to share what I had learned with others and I call the synthesis of these breathing techniques, Deep Energy Breathwork™. For those who need an intellectual basis for participating in an unfamiliar process, reading about breathwork should provide some momentum to engage in this work. So, I have suggested a reading list on this subject and on personality types, since so many have requested it.

I am planning for a fall healing retreat, jointly ventured with the Upledger Institute, so that those who are interested can experience both Deep Energy Breathwork as well as cranial sacral therapy and the deep and intuitive bodywork that the practitioners at the Upledger Institute are so skilled in. Yes, my next article will be about bodywork and what is available at the Upledger Institute.

As always I will be delighted to address any questions that you might have on any of the above. Send them to Cynthia Gracey, PATH Consulting, 1109 Harbor Drive, Delray Beach, FL 33483 or email me at pathcg@aol.com

The Neurotoxic Drug List

Editor's Note: Callers to the office often request information about the drugs on the toxic list and the conditions for which they might be prescribed. Below, we list each drug on the list with its usual usage. Remember, a mega dose is defined as 10 or more times the recommended daily allowance.



- Adriamycin** (brand name, Doxorubicin)—leukemia and other cancers
- Alcohol**—no known medical usage
- Amiodarone**—heart arrhythmia
- Chloramphenicol**—an antibiotic used mainly for eye infections
- Cisplatin**—treatment for cancers
- Dapsone**—antibacterial, antiprotozoan, antifungal, antileprosy, dermatitis herpetiformis suppressant
- Diphenylhydantoin** (Dilantin)—anticonvulsant, used to treat epilepsy, heart arrhythmia, and trigeminal neuralgia
- Disulfiram** (Antabuse)—used to help promote sobriety in the treatment of chronic alcoholism
- Glutethimide** (Doriden)—a sedative and hypnotic
- Gold**—used in the treatment of rheumatoid arthritis
- Hydralazine** (Apresoline)—used to treat high blood pressure
- Isoniazid** (INH)—antibacterial, used in the treatment of tuberculosis
- Megadose of vitamin A**—no medical usage
- Megadose of vitamin D**—no medical usage
- Megadose of vitamin B6** (Pyridoxine)—used to treat a very rare vitamin deficiency and as an adjunct to hydralazine or isoniazid. Doses above 200 mg/day can worsen CMT or other neuropathies. Lower doses are probably safe, but there is no evidence that they are helpful to CMT patients
- Metronidazole** (Flagyl)—used to treat very specific bacterial and protozoan infections, amoebic infections, bone infections, endocarditis, genitourinary infections, pneumonia, skin infections, vaginal infections, colitis, and inflammatory bowel disease
- Nitrofurantoin** (Furadantin, Macrofantin)—used to treat urinary tract infections
- Nitrous oxide** (chronic, repeated inhalation)—used as an anesthetic, frequently in dental procedures
- Penicillin** (large IV doses, only)—used as an antibacterial
- Perhexiline** (Pexid)—used to treat angina pectoris
- Taxol**—an injection concentrate for cancer, usually carcinoma of the ovary or breast. Used when other chemotherapies have failed
- **Vincristine**—used to treat cancer and leukemia (*Warning! This drug has caused severe problems, sometimes death, for patients with CMT. Discuss fully with your physician before beginning treatment.*)
- Lithium** (to be used with caution)—used to treat severe manic depression. Lithium can control mood swings characteristic of that disorder
- Misomidazole**—used in the treatment of some cancers
- Zoloft**—an antidepressant for oral administration



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

Ask the Doctor

Dear Doctor,

Someone mentioned to me that the myelin of the vestibular nerve is different from the myelin of other nerves and thus wouldn't account for vertigo in someone with CMT. But the vestibular and auditory nerves are a pair (I think): cranial nerve VIII. My understanding is that cranial nerves are really peripheral nerves and have myelin that comes from Schwann cells.

I know that people with CMT are prone to sensorineural hearing loss (high frequency) due to the ineffective myelin of the cranial nerve VIII.

It isn't acknowledged in the literature that people with CMT are prone to vestibular problems. Is the myelin of the vestibular branch different? If not, might vertigo be a problem that is not yet acknowledged by the medical community?

The Doctor replies:

The patient is right in that the myelin of the cranial nerves is the same as the peripheral nerves except for the first two cranial nerves, the olfactory and optic nerves. The two nerves are extensions of the brain and have oligodendrocytes instead of Schwann cells. The eighth cranial, (acoustic or auditory) nerve is composed of two fiber systems that are blended into a single nerve trunk. The cochlear nerve or the nerve of hearing and the vestibular nerve serves equilibrium, coordination, and orientation in space. It is my experience that most patients with CMT do not complain of hearing loss. However, there is a very rare group of families that have the phenotype of CMT and severe hearing loss that have no duplication of the PMP22 or Po myelin and that probably represent another subtype of CMT. Brain stem auditory evoked responses (BAER), a test that measures nerve conduction of the auditory nerves and the central (brain) connections, have shown decreased conduction velocities in the auditory nerves in patients with CMT. However, decreased nerve conduction velocities do not always correlate with clinical symptoms. Nerve conduction velocities are slow at age 2 and symptoms in the same patients only appear in late childhood and adolescence. Unfortunately, to my knowledge, there has been no systematic study of hearing tests in patients with CMT and no pathologic studies have been done of the auditory system in CMT patients. High-frequency hearing loss is not specific for CMT and frequently occurs in elderly persons. The vestibular part of the acoustic (eighth) nerve has a separate

anatomic connection and separate function. The fibers originate in the inner ear in the vestibular system that senses equilibrium, coordination, and orientation in space. Dizziness and vertigo are frequent complaints in patients with and without CMT. Most causes of vertigo or dizziness are due to lesions in the inner ear rather than in the vestibular nerve or the nerve nuclei. An experienced neurologist with the help of an ENT specialist can find where the problem is. It is my experience that dizziness and vertigo are not more frequent in patients with CMT than in the general population.

Dear Doctor,

CMT runs throughout my whole family—my mom, my sisters, two nieces, my 18 year old daughter, and me. My question is: has anyone seen CMT affect the patella and the surrounding muscles? I am asking because my daughter has trouble with her patellas staying in place. They dislocate so easily and she has to wear a J-brace to help hold them in place. I have two sisters and a niece with this same problem. Is this directly associated with CMT? My daughter had to have knee surgery on one knee last year to try to put the patella in the correct position and to fix the muscle around the kneecap. The surgeon told her the best thing to do is exercise those upper leg muscles by working out in a gym or bike riding. Does that sound correct?

The Doctor replies:

In this case, the problem with the patella may have nothing to do with CMT disease and is a problem that is prevalent in teenage girls and can run in the family. This is not uncommon. When the patella frequently dislocates or dislocates without any significant trauma, then it is time to have reconstructive surgery. Done arthroscopically, it can be quite successful, leaving very few scars. The quadriceps would necessarily be weakened after immobilization and surgery; thus, exercising them, especially when they are normal and not affected by CMT is a reasonable way to go.

Dear Doctor,

I have been wondering if the medical research on CMT has figured out why certain drugs are considered neurotoxic for CMT patients. Is there general knowledge of what happens when these items are ingested by someone with CMT and is there detailed

information on the metabolic activity? How is something designated as neurotoxic?

My reason for asking is that I had one glass of wine with dinner and about a half hour later my feet had a major episode of numbness. This lasted for approximately 2 hours and then my feet were back to normal (for me.) I realize alcohol is on the list of neurotoxic substances, but it seemed to me that there was some sort of metabolic process occurring that caused this to occur and then go away. I formerly was able to drink moderate amounts of alcohol without any noticeable ill effects CMT-wise.

Is anyone able to give an explanation of why the numbness is episodic and what is happening in the nerve cells?

The Doctor replies:

Neurotoxicity is the capacity of chemical or biologic agents to induce functional or structural changes in the nervous system resulting in tissue injury. There are many substances that produce damage to normal peripheral nerves. These substances are heavy metals such as lead, arsenic, mercury and thallium; drugs such as vincristine, thalidomide, pyridoxine, etc. The mechanism in each of these substances is different and in some of them the exact mechanism is known and in others it is not. There are neurotoxicology books that the patient may consult. If these substances damage normal nerves, patients that already have a nerve disease (neuropathy)—either hereditary as in CMT or acquired such as occurs in diabetics—are more susceptible to these substances. The more severe the neuropathy, the more susceptible the patient to the exposure of the neurotoxin.

Chronic and severe alcohol abuse produces a neuropathy with the major effect in the distal part of the axons. Alcoholic neuropathy seems to be the result of B vitamin deficiency and not a direct effect of alcohol. A contribution from the latter is nevertheless not excluded. The axonal damage is usually distal, length dependent, and produces numbness and tingling sensations. It seems like the patient with CMT has, as usual, a progressive course of the disease and that the patient was able to tolerate the alcohol better in the earlier stages of the disease. Episodic numbness occurs early in any sensory neuropathy. CMT is predominantly a motor neuropathy (produces weakness) with a variable sensory (numbness, tingling) component.

Dear Doctor,

My orthopaedic surgeon suggested using ankle weights to exercise the quadriceps and hamstrings to help with degenerative joint disease. Is this safe for someone with CMT to do? I'm worried about harming already weakened ankles.

The Doctor replies:

Knowing the age of a person is important. To have degenerative joint disease, I presume that person is in the 50s-60s. Exercising quadriceps and hamstrings, which are muscles that act around the knee joint, should not affect the weakened ankles as long as the weights are over the ankles and the ankles can support the weights. It is time to stop when the calf muscles become achy, crampy, tired, and painful.

BOOK REVIEW

Genetic Secrets: Protecting Privacy and Confidentiality in the Genetic Era, edited by Mark. A. Rothstein, is a thought-provoking set of articles that address dilemmas of privacy and confidentiality. Topics focus on advances in genetic technology, changes in our healthcare delivery systems, and the increasing pressures to utilize electronic databases for storage of medical information. The views of lawyers, physicians, ethicists, a clinical geneticist, and a genetic counselor are discussed. Recommendations include the development of a legislative framework that includes the following: initiatives at the national and state levels; laws to protect confidentiality of medical records; regulation of access to information; and regulation of authorized disclosures.



Call for Participants

The Charcot-Marie-Tooth Clinic at Wayne State University in Detroit, MI, received a grant to study the correlation between specific mutations in the connexin 32 gene and the severity of disease in patients with CMTX. We are asking patients with CMTX to consider coming through our clinic for evaluation to help us complete this study. The knowledge obtained from the grant will help investigators understand how connexin 32 mutations cause neuropathy so that treatments can ultimately be developed for the disease. There will be no charge for the evaluation, which will include assessment of the patient's CMT and suggestions for improving ambulation skills and evaluation of rehabilitation needs. While travel expenses are not permitted to be covered in the grant, low-cost housing can be made available near the medical center for those who come from outside the Detroit area. Those interested should contact Izabella at (313)-577-1689.

—Michael Shy, M.D.





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.

The CMTA Report

is published by the Charcot-Marie-Tooth Association, a registered non-profit 501(C)(3) health organization. Copyright 1998, The CMTA. All rights reserved under International and Pan American Copyright conventions. No part of this newsletter may be reproduced in any form or by any electronic or mechanical means, including information storage and retrieval systems, without permission in writing from the publisher.

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 Editor,

I read with interest a letter from M.M. of Oak Ridge, TN. She was concerned that her son, who has CMT and diabetes, was told that he could not wear braces because he might injure his feet.

With the advent of advanced knowledge in biomechanics and the improvement of shoes, bracing, and accommodative padding, I see no reason that someone should not be able to wear braces to improve ambulation even in cases of severe peripheral neuropathy. While the person might need close and careful monitoring to prevent neuropathic breakdown, there should be no reason that the person should not be able to retain some level of function.

I would encourage these people to seek help from a podiatrist who is board certified in podiatric orthopedics and a local prosthetist/orthotist. This combination of expertise should allow the person to resume fairly normal activities.

—James E. Webb, Jr., D.P.M., Bartlesville, OK

(Editor's note: A second response to the same letter from the spring newsletter follows.)

Dear Editor:

In the spring issue of the newsletter, M.M. of Oak Ridge, TN, wrote that her son has CMT and insulin-dependent diabetes, but has been advised not to wear braces because of the pressure. As an orthopaedic surgeon who specializes in foot and ankle surgery, I treat many patients with diabetes and CMT. I would recommend AFOs for bracing. A good brace fabricator should be able to protect his feet by applying a layer of padding to the AFOs. We often use materials such as plastizole or PPT, which are very conforming materials. They provide excellent protection for the insensate foot, and are readily available and custom moldable. I'd be glad to refer him to an appropriate person in his area, if such a referral is necessary.

—Douglas Wright, M.D., Havre de Grace, MD
dwright@susq-ortho.com

Dear Editor,

Here's an innovative solution to handling the pain associated with CMT. I spent the day at the beach Friday for a work retreat. It was fine. I had a very blissful experience. My feet were hurting very badly. I went wading in the surf. The water is about 50 degrees and cold, but within a few minutes my feet had numbed up and quit hurting. It was really "cool" (no pun intended). I made my feet quit hurting without drugs. I can't

wait to go back! I went in the water looking for bliss and found it in a big way.

—Tony, Internet

Dear Editor,

In the spring issue of the newsletter, the "Doctor" said there could be no connection between an intestinal blockage and CMT disease. Perhaps he was not including anal sphincter paralysis when he rendered that opinion. But to clarify the matter, I wish to inform you that my wife has had rectal incontinence for many years and has consulted with numerous doctors, including neurologists, enterologists, gynecologists, and other experts concerning her condition, and all of them have related her lack of anal sphincter control to her CMT disease.

—P.G., Pacifica, CA

Dear Editor,

I am sure everyone has great stories about the trials and tribulations of living with CMT, but E.C. from Fishkill, NY, got me inspired to write my story in hopes it would help others see things with humor instead of the frustration that is already so much a part of our lives.

I am blessed to be from a large extended family with a significant CMT population. Long before I was diagnosed at age 22, I knew clumsiness to be a sign of an excellent mind. It is a commonly accepted fact in my family. That knowledge got me through many embarrassing moments—falling into raspberry bushes at high school parties, landing face-first in a friend's lap getting into a car, etc. The campus health center at my college was amazed that someone could sprain her ankle during the first day of Tai Chi class and then step off the curb and sprain the other ankle the next day! My friends have lovingly called me Grace since high school.

The highlight of my experience of this nature was a serendipitous string of events one evening in 1990. I was going to a lecture about the playwright Lillian Hellman (*The Children's Hour*) with my housemate. I started the evening by falling Ragedy-Ann-style down all of the stairs in the parking lot and skinned both knees and elbows on my way down. My roommate, a long-time friend used to my problem, cried and laughed with me, picked me up, brushed me off, and we proceeded. During the lecture, I discovered that Lillian Hellman was a kindred soul. After her long-time partner, Dashell Hammett (*The Thin Man*) died, she met a young man at a formal

party. After the party, she walked down Madison Avenue in New York with him. He was amazed by two things. One: she fell down seven times while walking eight blocks, and two: she was completely unfazed because (she told him) her mind was far too busy with more important things than where to step! Her lack of grace, she declared, was a sign of her genius!

Following the lecture, we stopped by my parents' house in hopes of a free dinner. I walked in the house to find my mother laughing with tears pouring down her face as she cooked. My father, a physician, was shedding his white hospital coat and stethoscope. Much to my father's consternation, my mother related to us the cause of her mirth. Earlier in the day, my father (a tall man) had been "holding court" with the younger doctors in the entrance to the Portland VA hospital when, without taking a step, he had fallen down and thrown his full coffee cup into the air, showering anyone within range. He even split his pants "from stem to stern." He noted that those white doctor coats were actually good for something! We later compared skinned knees and declared our genius.

I know that I am fortunate to belong to an extended family who knows that the challenges placed before us are simply pathways to different blessings. I pray that my two-year-old daughter does not have CMT, but I am so happy that she will grow up in a family that can see the humor and potential in the challenges life throws at each of us.

—*"Grace"...*K.M., *Conifer, CO*

Dear CMTA,

I had been using the same AFO for 20 years, but in the last several years, I felt the need for more "assistance." I went to three different orthotists. One offered nothing better than what I already had, the second adjusted and degraded what I already had, the third offered me his great new invention which was carbon fiber spiral braces—at ten thousand dollars a pair!

Fortunately, there was a fourth orthotist. He analyzed what was wrong with my current AFOs and he seemed to know what he was talking about. He offered me rather conventional-looking vacuum-formed AFOS except there were four small L-shaped strips of very thin carbon fiber material embedded in the plastic on either side of the ankle. Apparently, the carbon fiber flexes when you step on the ball of your foot and returns the energy to lift your foot as you take the next step. This is the same action you get with conventional AFOs, but the carbon does it with vigor. The strips are only about 4 inches long, 1/2 inch wide and 1/8 inch thick.

I was only hoping for some modest improvement, but I got a really big boost.

With my old AFOs, I never used a Velcro strap over my shinbone because I didn't like the feel. Instead, I used an elastic band that stretched when I took a step. While this was more comfortable, it also diminished the amount of toe lift. With the new AFOs I do use the Velcro strap, but it's less uncomfortable. The reason, I believe, is that my old AFOs formed an angle of 80 degrees rather than 90. This small difference wasn't terribly relevant 20 years ago, but it had the effect of pushing my knees forward as I walked or tried to stand. As my legs got weaker over the years, it worked against my ability to stand up straight. It also put a strain on the band around my shinbone, which the elastic band relieved.

The good news is that I can now walk pretty well and because the AFOs are held firmly with the Velcro band, I get more support and walk without wondering if my feet are going to go where I want. I can now stand fairly well because the AFOs aren't trying to push my knees forward and the Velcro band provides added support. I also climb stairs better.

I'm 69 and I was ready to use a cane full-time because I couldn't stand without holding on to something and climbing stairs was getting really difficult for me. I want to make two points:

- 1) CMT is insidious and it sometimes takes years to realize that something needs fixing. Perhaps this note will help someone seek a solution to a problem sooner rather than later.
- 2) There apparently can be a significant difference in similar-looking AFOs.

If anyone would like to ask me any additional questions about my AFOs, they can email me at Rkarl619@aol.com

—R.K., *San Diego, CA*

Dear CMTA,

I have been noticing increased difficulty in walking. It's more than just the usual effect of my CMT and I have been told that I need a hip replacement operation. This is, of course, an unwelcome procedure, particularly for someone with CMT. Is there anyone in the membership who has had this procedure and would be willing to tell me about it? Also, does anyone know of any literature on this topic that I could share with my surgeon? He's very skilled in hip and knee replacement (that's all he does), but he told me he has not seen a case of someone with CMT in many years.

—*A Member in South Florida*
(Please send answers to the CMTA office and they will be forwarded.)

■ OF INTEREST

The CMT Association of Australia held a nationwide awareness day on July 26, 1998. Their slogan was "We need your help to stay on our feet." The association had over 100 persons attend the conference at which presentations were given on CMT Genetics, Hand and Foot Surgery, and Physical and Occupational Therapy.



Editor's Note: A phone call to the office produced the following question: Has anyone had to deal with children being cruel to a child whose parent has CMT and, therefore, walks differently? A mother of a young school-age child would like any insight anyone has to offer on this problem. Please write to the CMTA office with your comments.

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 Zolofit 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



601 Upland Avenue
Upland, PA 19015
1-800-606-CMTA

Non-Profit Org.
U.S. Postage Paid
Glen Mills, PA
Permit No. 10

Forwarding and return postage guaranteed.
Address correction requested.