

THE CMTA REPORT

Providing information on Charcot-Marie-Tooth disease (a.k.a. Peroneal Muscular Atrophy or Hereditary Motor Sensory Neuropathy), the most common inherited neuropathy. Contents © 1993, CMTA. All rights reserved.

Noted CMT Researcher Receives Service Award

Dr. James R. Lupski of the Institute for Molecular Genetics, Baylor College of Medicine, received the distinguished research award for his work in discovering the CMT gene located on chromosome 17 and for designing the test which is the prototype for the diagnostic DNA blood test now offered by Genica Pharmaceuticals.*

The award was presented to Dr. Lupski by Karol B. Hitt, President of the CMTA, at the October patient/family conference at Methodist Hospital at Methodist Hospital, Baylor College of Medicine. Dr. Lupski gave one of the presentations at that meeting and discussed recent CMT genetic research.

Dr. Lupski received the CMTA's award for his discovery that the majority of CMT patients have a duplication of genetic material which is the causative agent for CMT Type IA. Furthermore, in isolating the CMT gene, PMP-22 located on chromosome 17, he found that point mutations in that gene can also produce CMT.

Dr. Lupski is a member of the CMTA's Medical Advisory Board.



Dr. James Lupski & Karol Hitt

**Genica Pharmaceuticals can be reached by calling: 1-800-394-4493, ext 106. Ask for Sarah Quiry, customer service representative. A physician must order the shipping kit and the cost of the test is \$ 395.00.*

Karol Hitt Chosen National Consultant to UC Davis Study

CMTA President Karol B. Hitt has been selected to be a national consultant to The Rehabilitation Research and Training Center in Neuromuscular Diseases at the University of California, Davis.

UC Davis has received another grant for five years of work with individuals with neuromuscular diseases and their families. The purpose of the center is to develop and evaluate new strategies to address lifelong needs for health care, rehabilitation, psychosocial well-being, education, employment, and independent living.

(cont'd on page 2)

Computer Disasters and Other Problems of the Fall

For three weeks this fall, the IBM computer which is the heart and soul of everything that is done in the office of the CMTA was non-functional as we waited for the part which would bring it back to life. During this "down-time" the articles for the newsletter and all other written work basically could not be done. This is by way of explanation and apology for the lateness of the Fall issue of The CMTA REPORT.

One of the things which became painfully apparent during our "outage" was that we should have had a tape backup of our whole system so that we could have functioned with another computer. We did not. To the rescue came Steve Khosrova, a CMTA member and the author of an article in this issue of the newsletter. Since he works with computers, Steve recognized the need for a tape backup system to be installed and he generously donated both the machinery and the time to help connect it, as well as the expertise to help our staff learn to do the procedure.

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Why Suffer Emotionally More Than Necessary?



by Mark Flapan, Ph. D.

Mark Flapan has scleroderma and is founder of the Scleroderma Federation of the Tri-State Area. He is a psychologist with a special interest in the emotional effects of chronic illness, both on the ill person and on family members. This article is reprinted with the permission of the National Organization for Rare Disorders (NORD).

Anger, with the possible exception of fear, disrupts the human body more than any other emotion and should be labeled, "dangerous to your health." Similarly, anger disrupts human relationships more than any other emotion and should be labeled, "dangerous to your social well-being." Anger takes its toll on both the human body and on human relationships; whether or not you have a chronic illness. But with a chronic illness, more than without one, you need your body working for you and you need supportive relationships with family members and friends as well. So no matter how you look at it, it's to your advantage to live

Hitt as Consultant- cont'd from pg. 1

The center plans to study impairment and disability as it relates to physical performance and nutritional status in neuromuscular diseases, the evaluation and treatment of swallowing disorders, dyspnea, respiratory control, and rehabilitation therapeutics in neuromuscular disease.

Another phase of the five year study will address handicaps which affect employment opportunities and the vocational rehabilitation of individuals who have neuromuscular disorders. The study will also look at mainstreaming of children and young adults with neuromuscular disorders, especially as it involves teacher sensitivity and social skills.

The final focus of the study will involve the research training of current and future researchers and the dissemination of information.

UC Davis has worked with CMT patients in previous studies and the CMTA has published the results of their previous five year study in the newsletter and CMT FACTS II. This is the first time that the center has chosen lay persons to act as consultants and Karol will be working in the portion of the study designed to help train future researchers and increase the dissemination of information.

with the least possible amount of anger. Therefore, in this article, I'll suggest ways of understanding yourself and ways of relating to others that may help you lessen your anger.

If you have a chronic illness, more things anger you than if you were well and healthy. You may get angry when frustrated by tasks you find difficult or impossible to do - tasks which everyone else can do without a second thought. You not only get angry at the object of your frustration, but you get angry at yourself as well. If anyone else got angry at you for the same thing, you wouldn't take it - but you take it from yourself. You take it from yourself because you feel you should be able to do whatever you need to do - disease or no disease.

To put it a little differently, when you can't live up to the "shoulds" in your head, you get angry at yourself. However, if you could set aside your judgmental "shoulds," you would more willingly accept the limitations of your illness. While accepting your limitations won't make your tasks easier physically, it would certainly make them easier emotionally. You would then no longer berate yourself in anger when up against some difficult task, but would be patient with yourself instead.

Even though you feel you've been cheated out of a normal life by your illness, nobody owes you anything because of it.

You not only have "shoulds" for yourself, but you have "shoulds" for other people as well. So when others don't act toward you in ways you think they should, you're as hard on them as you are on yourself. After all - fair is fair.

You may get angry at people who don't treat you in some special way. You feel the world owes you something because of your illness and you should be compensated somehow. As a child, your parents may have given you special consideration when you were sick, and now that you have an illness, you still expect to be treated special. As it happens, you're not a child anymore and others are not your parents, so these expectations are not likely to be met. Even though you feel you've been cheated out of a normal life by your illness, nobody owes you anything because of it.

It may not make sense, but while you would like to be treated special, you still want to be taken as a normal person - no different than anyone else. You don't want to be related to as an invalid or pitied in any way and you get angry at anyone who does. At times it seems as if no matter how others react to you, it's not right somehow.

You may be angry at friends and relatives who aren't as sensitive to your feelings as you think they should be. But why should they be that sensitive to your feelings? How can they understand what it's like to live with a chronic illness? They don't have a disease - you do.

How can they understand what it's like to live with a chronic illness? They don't have a disease - you do.

It may be hard to accept that well and able-bodied people don't know how to relate to someone with a chronic illness. It's difficult for them to understand what they've never experienced. So even with the best of intentions, they're going to say and do things that anger you.

Not only that, if the situation were reversed - you were well and healthy and a friend had some rare disorder - you might not understand her feelings or know how best to relate to her either. So even though your heart was in the right place, you might say and do things that angered her just as your friends say and do things that anger you. I know you would like to think it would be otherwise, - maybe it would be - or maybe it wouldn't.

Friends and relatives are one thing, but your immediate family is something else again. More likely than not, you're

(continued on page 12)

Do you know of anyone famous who is a CMT patient? Do you know anyone famous who would advocate for CMT?



The CMTA is looking for a well-known person to be a spokesperson for CMT. If you know of such a person, contact the CMTA; we will do the rest.

Items of Interest



Item 1. Correction: As a result of a transcriptional error, The Mayo Clinic Dynamine Study article incorrectly referred to Lambert-Eaton Syndrome (also known as Eaton-Lambert Syndrome) as a hereditary disease. It is not hereditary.

Item 2. Please note the additions to the neurotoxic drug list as it now appears in this issue of the newsletter. Until new cards can be printed, you will need to be responsible about adding the drugs to your own medical files.

Item 3. A new book entitled, A Guide to Legal Rights for People with Disabilities by Marc Stolman, Esq. is available to help people with disabilities understand the most often encountered legal topics. This book provides the most comprehensive explanation of the ADA available to date. 150 pages, \$34.95 hardback, \$19.95, softcover. Ask your bookstore to order.

Item 4. Another book entitled Living Beyond Limits by David Spiegel, M.D., covers 20 years' clinical experience in treating patients with chronic illness. Rejecting denial and avoidance as methods of coping with chronic disease, he explores the healing process as a complex of mind-body interactions. Available in libraries or by order from your bookstore.

Item 5. Speak Up for Health, a handbook for parents of teens with chronic illness or disabilities is available for \$ 10, by calling 612-827-2966. The publication includes personal accounts of parents and young adults who have faced the challenges, struggles, and experiences in dealing with the health care system.

Item 6. Also available from PACER Center is a new booklet called Supported Employment: A Step by Step Guide, which helps persons with disabilities and their families obtain or improve supported employment. This publication is a follow-up to PACER'S 1988 booklet, Opportunity Knocking: The Story of Supported Employment, which is a more general look at supported employment. They also publish a manual called Begin the Between: Planning for the Transition from High School to Adult Life. This workbook guides students with disabilities and their families in developing a comprehensive transition plan based on their exploration of the student's skills, interests, and future goals. All these books are under \$ 10 and are available from PACER Center, 612-827-2966.

Item 7. A directory for disabled travelers is now available. Travelin' Talk Network has published a resource directory of more than 550 pages which lists all the resources a traveler with a disability should ever need to plan their trips and find help should an emergency arise. The new directory is available for \$ 35.00 and comes with a money back satisfaction guarantee. Travelers with disabilities wanting assistance with planning their trip can call 615-552-6670. The book can be ordered from Travelin Talk, P.O. Box 3534, Clarksville, TN, 37043.

Item 8. Dr. Jane Firth, of the Department of Medicine at Sydney University, Australia, has now completed her collection of information on the CMT central nervous system project and will publish the results soon. She has worked with Professor McLeod and Associate Professor Garth Nicholson on this project.

Item 9. The US Equal Employment Opportunity Commission and the US Department of Justice, Civil Rights Division, have published a booklet called, The Americans with Disabilities Act: Questions and Answers. This publication can be ordered for free by calling 1-800-669-3362. Be patient. You will be calling the government.

Item 10. CMT patient, Milda Vizbar, has a Christmas card called " Ring Around the Christmas Tree" available through a catalogue called 1993 Courage Holiday Cards Collection. Courage Cards help thousands of children and adults with disabilities each year through the Courage Center which sponsors rehabilitation and independent living programs. Call 612-588-0811 for further information.

Item 11. From Disability Rights and Resources, Demos Publications, comes some advice for people applying for health insurance. If you have a preexisting condition and are thinking of applying for health insurance, there are at least eight ways in which insurance companies may respond. The specific response you receive will probably be influenced by one or more of the following: the nature of your condition; your current health status and your health history; the company to which you are applying; the specific plan for which you are applying; whether you are applying as an individual, family, or small or large group member; and the state in which you live or are applying for coverage. You may expect any of the following responses:

- 1 The condition will not affect your approval for and receipt of coverage.
- 2 You will be denied coverage temporarily, but with an opportunity to reapply at a specified future time (you will probably have to ask in order to learn this).
3. You will be denied coverage, with no opportunity to reapply (or at least no chance of succeeding if you do).
4. You will be approved for coverage, but with a specified waiting period (for example, six months) during which you will not be covered by the plan for services received in relation to the preexisting condition.
- 5 You will be approved, but coverage will not be available for the preexisting condition until a specified period has elapsed after the effective date of the policy during which you do not receive any treatment or medication for the preexisting condition.
6. You will be approved for coverage, but with a "rider" to your policy permanently excluding coverage for services you receive in relation to the preexisting condition.
7. Special limitations will apply to your coverage; for example, you will be covered for services received in relation to your preexisting condition, but you will have to pay co-insurance, or co-insurance at level. that you would not have to pay for services for other conditions that were not preexisting.
8. A combination of two or more of the foregoing; for example, coverage will be available for services for your condition after a specified time has elapsed in which no treatments for the condition were received or after a specified time whether or not treatment was received. After this point, you will be covered for services for the condition to the extent that the plan normally covers those services.

Item 12. Gerald Weber, DPM, member of the CMTA's Medical Advisory Board, is the Editor-in-Chief of a new publication called The Lower Extremity, a journal of the American College of Foot Orthopedists.

Item 13. A member of a large CMT family is looking for additional relatives,
(continued on page 10)

Research Update

A continuing list of significant CMT genetic research discoveries is indeed good news for the CMT community. It is through these genetic findings that therapies for CMT will be developed. Finding the CMT gene(s) is the first very necessary step which must be accomplished before a therapy can be designed.

The gene responsible for CMT1B on the long arm of chromosome 1 has been identified independently by three groups led by Dr. Roger Lebo in the United States (PNAS), Dr. Hayasaka in Japan (Nature Genetics), and Dr. Baas in Holland (Nature Genetics). This discovery was made possible by the previous sublocalization of the gene in Dr. Lebo's laboratory. An abnormal myelin protein zero (MPZ) gene has been found in 4 of 5 CMT1B families which results in Type IB CMT that is clinically indistinguishable from CMT1A.

X-linked CMT: This form of CMT is sex dependent in its inheritance. Females are carriers (like in hemophilia and Duchenne muscular dystrophy), but over time they may develop mild symptoms of the disease. Weakness usually begins during early childhood or adolescence and may progress to cripple an individual. Clinically, this form of CMT is very similar to CMT I. Dr. Kurt Fischbeck, University of Pennsylvania, in collaboration with researchers at Duke University, Harvard and the University of Iowa, has

localized the gene for CMTX to a very small region of the X chromosome. (Editor's note; At the American Society of Human Genetics meetings in October the genetic defect was reported as a defect in a gap protein junction called Connexin.)"

Also, in another region of chromosome 1, a gene for type 2 CMT was found by Dr. Jeffrey Vance at Duke University. Quoting from the Duke CMT newsletter, "Recently, Dr. Jeff Vance's laboratory localized one gene causing autosomal dominant CMT2 to chromosome 1. They have also demonstrated that like CMT1, more than one gene causes CMT2. The form of CMT2 that is located on chromosome 1 is now designated as CMT2A. These findings were published in the August issue of *Genomics*. We do not yet know the location of the CMT2 gene in the other families. We are actively collecting additional families of the CMT2 type to speed our search for the other CMT2 gene(s). Additional families with CMT2 that would be interested in contributing to this important research effort are needed and we would greatly appreciate their contacting us." (Contact Dr. Vance at Duke University Medical Center, Room 103B, Research Park One, Research Drive, Durham, NC 27710 or call 919-684-5963.)

Continuing from the Duke CMT newsletter, "CMT type IV: Through a collaboration with neurologists in Tunisia, we have been studying autosomal recessive CMT. We are happy to report that we have also recently found the chromosomal location of one of the genes causing CMT IV. This form is not found as frequently in the USA as the other types, but has been described in many families. Work is continuing on this type of CMT to discover where other genes are located that are responsible for recessive CMT. (Editor's note: This gene has been located on chromosome 8.)

None of this work would have been possible if you, the CMT patient, had not cooperated with the research community. The genetic basis for CMT disorders is proving to be highly challenging and interesting to the researchers. So far 5 CMT gene locations have been identified with more to be found. Each one of these locations produces a distinct form of CMT, but the individual patient will have only one form. Similarly, there will only be one genetic form in the extended family. In other words, if the parent has

Call for Participants:

Dr. James Lupski of the Baylor College of Medicine is looking for persons with CMT who have neither parent affected and who are identified as cases of spontaneous mutation. The person should already have been tested by a neurologist and should also have had the diagnostic test for CMT 1A, by Genica Pharmaceuticals. Persons with both positive and negative responses from the Genica test are needed, if they have no family history of CMT. If you qualify to be part of this study, either you or your doctor should write to Dr. James Lupski, Institute for Molecular Genetics, 609 E. Anderson Building, Houston, Tx, 77030.

Dr. Victor Ionasescu is also interested in participants for a study on the X or sex-linked version of CMT. The following criteria are required for a person to be considered. No male to male transmission would be present. A father and his son could not both be affected. Affected males would only pass the disorder to their daughters, not to sons. Affected females can pass the trait to both daughters and sons; however, sons tend to be more affected than daughters. The affected females may have a very mild disease, only detectable on nerve conduction studies. The researchers have 10 families so far with x-linked CMT. They need more in order to find the gene. If you have the requirements listed above, write to: Victor and Rebecca Ionasescu, Division of Medical Genetics, Department of Pediatrics, Iowa City, IA, 52232. The study, including DNA blood test, is free of charge.

CMT 1A, then the children will also have CMT 1A. It is this multiplicity of genes that causes the problem with developing therapies. Each CMT gene causes a different physiologic defect and will therefore need different therapy. That is the challenge; we must meet it. The CMTA has set a goal of a million dollar CMT research fund within three years. We invite you to join us and contribute to CMT research. Simply designate your gift to the CMTA RESEARCH FUND, and it will be used only for research purposes.

CMT in the News

RN Magazine ran a brief article on CMT in the June, 1993, issue of the publication. The organization thanks Board Member Michael Molinari and his wife for their effort in having this article published.

The Newport Daily News, Newport, Rhode Island, ran a two day series on Charcot-Marie-Tooth disease. The second article featured 11 year old CMTA member Elsa Johnson and her family. The Muscular Dystrophy Association reprinted these articles in a summer mailing they did on CMT.

Family Circle published an article on the rescue of George Horner (see patient profile) from his burning apartment in the July 20 issue of their magazine. In the October 12 issue, CMTA President Karol Hitt wrote a letter praising their publishing of the previous article and telling readers how to access our organization for more information.

CMTA Financial Report

Independent Auditor's Report

To the Board of Directors
Charcot-Marie-Tooth Association
Upland, Pennsylvania

We have audited the accompanying balance sheets of Charcot-Marie-Tooth Association (a Pennsylvania nonprofit corporation) as of June 30, 1993 and 1992, and the related statements of support and revenues, expenses and changes in fund balances and cash flows for the years then ended. These financial statements are the responsibility of the Association's management. Our responsibility is to express an opinion on these financial statements based on our audit.

We conducted our audits in accordance with generally accepted auditing standards. Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the financial statements are free of material misstatement. An audit includes examining, on a test basis, evidence supporting the amounts and disclosures in the financial statements. An audit also includes assessing the accounting principles used and significant estimates made by management, as well as evaluating the overall financial statement presentation. We believe that our audits provide a reasonable basis for our opinion.

In our opinion, the financial statements referred to above present fairly, in all material respects, the financial position of Charcot-Marie-Tooth Association at June 30, 1993 and 1992, and the results of its operations and its cash flows for the years then ended in conformity with generally accepted accounting principles.



Cogen Sklar Levick
Certified Public Accountants
July 12, 1993

Notes

NOTE 1 - SUMMARY OF SIGNIFICANT ACCOUNTING POLICIES

Nature of Organization

The Charcot-Marie-Tooth Association (the Association) was incorporated under the laws of the Commonwealth of Pennsylvania as a nonprofit corporation in October 1983 and is registered with the Commonwealth of Pennsylvania as a charitable organization. The Association was established to sponsor, encourage and undertake scientific investigations of the causes and cures of Charcot-Marie-Tooth Disease. The Association receives the majority of its support from private donations.

Basis of Accounting

The Association uses the accrual basis of accounting. All contributions are considered to be available for unrestricted use unless specifically restricted by the donor.

Volunteers have donated significant amounts of their time in the organization's program and administrative services and its fund raising campaigns. The value of this contributed time is not reflected in these statements since it is not susceptible to objective measurement or valuation.

(continued on page 11)

Balance Sheets - June 30, 1993 and 1992

ASSETS	1993	1992
CURRENT ASSETS		
Cash	\$69,621	\$35,242
Security deposit	<u>165</u>	<u>165</u>
	69,786	35,407
PROPERTY AND EQUIPMENT	<u>4,635</u>	<u>3,886</u>
TOTAL ASSETS	\$74,421	\$39,293
LIABILITIES AND FUND BALANCES		
CURRENT LIABILITIES		
Accounts payable	\$ -	\$1,720
Accrued expenses	<u>250</u>	<u>253</u>
TOTAL LIABILITIES	<u>250</u>	<u>1,973</u>
FUND BALANCES		
Operating balance	59,992	29,764
Board designated	<u>14,179</u>	<u>7,556</u>
	74,171	37,320
TOTAL LIABILITIES AND FUND BALANCES	\$74,421	\$39,293

Statements Of Support And Revenues, Expenses and Changes In Fund Balances Years Ended June 30, 1993 and 1992

	1993	1992
SUPPORT AND REVENUES		
Contributions	\$100,088	\$69,350
Conference fees	3,888	3,744
Interest income	468	274
Miscellaneous	<u>412</u>	<u>-</u>
TOTAL SUPPORT AND REVENUES	<u>104,856</u>	<u>73,368</u>
EXPENSES		
Program services	62,202	51,013
Management and general	2,525	2,143
Fund raising	<u>3,278</u>	<u>2,515</u>
TOTAL EXPENSES	<u>68,005</u>	<u>55,671</u>
EXCESS OF SUPPORT AND REVENUES OVER EXPENSES	<u>36,851</u>	<u>17,697</u>
FUND BALANCES - BEGINNING OF YEAR	<u>37,320</u>	<u>19,623</u>
FUND BALANCES - END OF YEAR	\$ 74,171	\$37,320

Statements Of Cash Flows Years Ended June 30, 1993 and 1992

	1993	1992
CASH FLOWS FROM OPERATING ACTIVITIES		
Excess of support and revenues over expenses	\$36,851	\$17,697
Adjustments to reconcile excess of support and revenues over expenses to net cash provided by operating activities:		
Depreciation	1,101	686
Decrease in:		
Accounts receivable	-	1,092
(Decrease) in:		
Accounts payable	(1,720)	(484)
Other liabilities	<u>(3)</u>	<u>(87)</u>
Net cash provided by operating activities	<u>36,229</u>	<u>18,904</u>
CASH FLOWS FROM INVESTING ACTIVITIES		
Capital expenditures	(1,850)	(4,572)
Net cash used in investing activities	<u>(1,850)</u>	<u>(4,572)</u>
NET INCREASE IN CASH	<u>34,379</u>	<u>14,332</u>
CASH - BEGINNING OF YEAR	<u>35,242</u>	<u>20,910</u>
CASH - END OF YEAR	\$69,621	\$35,242

1993 Year in Review



CMTA Board of Directors

This year, 1993, is the tenth year since the organization of the Charcot-Marie-Tooth Association...and, we have come a long way. Starting in 1983 with a few CMT families, we now have over 5,000 names in our database. This number is continually increasing as more and more patients, families, and medical professionals register with us. With this increased growth, our financial stability is greatly improved. (See accompanying Annual Financial Report.) This financial growth enables us to continue and expand our patient/family outreach and education programs. Our financial position also has allowed us to establish a dedicated fund for research.

In 1993 we organized four Patient/Family Conferences. The sites of these meetings and our hosts were Dr. Harold Marks, A.I. duPont Institute, Wilmington, DE; Dr. Stanley Appel, Baylor Medical Center, Houston, TX; Dr. Ian Alexander, Crystal Clinic, Akron, OH; and Dr. Paul Schultz, Children's Hospital, San Diego, CA. These regional meetings are very enthusiastically attended by patients and family members, as well as medical professionals. We are deeply indebted to our hosts and their institutions. There is a great deal of learning at these meetings, and there is also a great deal of camaraderie and support. It really is beneficial to exchange information and interact with other people who share the same concerns and problems.

The sequel to that "bestseller" CMT FACTS I, CMT FACTS II, was published during 1993. (To order either or both, see Order Form on page 15.) CMT FACTS II contains information not covered in the first volume, and all of the

articles and resources are relevant to the CMT patient/family. Our VCR tape program continues to be a wonderful source of taped presentations about various aspects of CMT and CMT therapies. We would be remiss here if we did not gratefully acknowledge the wonderful work of Walt Kelly. Walt is the basis of our tape program, and we honor him and his work.

The newsletter, *The CMTA Report*, continues to be our primary means of communicating with the lay and professional communities. If you have suggestions for articles, complaints, and/or comments, please write to us. We value your input and welcome reader-submitted articles.

The Medical Advisory Board meets regularly twice a year at the Neurology meetings. These men and women graciously give of their time and knowledge, and we honor and thank them for their

dedication. They truly represent a caring medical community. Dr. Gareth Parry is the editor for *THE PHYSICIAN'S HANDBOOK for CMT DISORDERS*. The book is being written by ten CMT experts and will cover the topics needed by the primary care physician who has a CMT patient in his/her practice. We anticipate publication of the book in early 1994. Its availability will be announced in the *CMTA REPORT*.

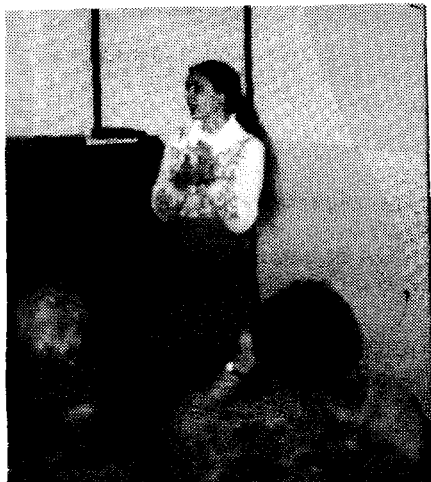
The officers of the CMTA's Board of Directors are Karol B. Hitt, President; J. Rodman Steele, Jr., Vice-President; Diane Freaney, Treasurer; Robert Daino, Recording Secretary; and Donald J. Perrella, Corresponding Secretary. Dr. Robert E. Lovelace is Chairman of the Medical Advisory Board. The other directors of the CMTA are: Rex S. Morgan, Jr., George Crohn, Jr., Ann Lee Beyer, Diana Eline, Gary Griffith, Gerald S. Hartman, and Michael J. Molinari. This year for the first time, the board met for a two day meeting and retreat. Much was discussed and goals were set for one, two and three years. There are vacancies on the Board, and we invite you to apply if you are interested in serving. Historically, we have met in the East as all of our members are located here, but that is not a requirement for membership. Board members are financially responsible for his/her own costs involved in attending a meeting. In your application letter, please submit a brief resume and tell us of any specific interest or experience you feel would be of value to the CMTA. We are particularly in need of someone with public relations experi-



Our youngest conference attendee ever - The one day old Marsh baby of Houston, Texas



Ask the Doctor



Dr. Nina Njus, Akron conference

ence and contacts. We must make CMT and the CMTA better known.

Not only has this been a landmark year for the CMTA, but certainly for the CMT community as a whole. Molecular genetic CMT research is really bursting with discoveries. (See Research Update.) The first direct benefit is the diagnostic testing that is now available for type 1A CMT. As the genes for x-linked CMT and type 2 CMT are found, these discoveries will benefit more and more patients. Ultimately, these discoveries will lead to treatment and possible cures for CMT.

The CMTA is actively involved in an assessment of function project of the CMT patient. This project is being conducted by Diana Eline, and 1000 surveys were sent to randomly selected CMT patients. Diana is a trained scientist and will report her findings in a later issue of the CMTA REPORT.

There are many people who make the work of the CMTA possible. We have one employee, Pat Dreibelbis, and we thank her for her loyal service. The rest of the work is done by volunteers, and it is truly a remarkable amount. Some of the people who have assisted us this year are Tom Holmes, Dale Stell, Tom Weston, David Hitt, Laura McCann, Gerry Hitt, George DeBolt, Eli Yaron, David Showers, Dana Schwerfeger and Steve Khosrova. We thank these people for their time and skills. It has been my pleasure to work with all and it is my continuing pleasure to serve the CMT community.

Respectfully submitted,
Karol B. Hitt, President

Dear Doctor,

Would you please comment on the possible neurotoxic effect of Tylenol, aspirin, or Advil as over the counter pain relievers for CMT patients?

The Doctor replies:

There are no reported contraindications in taking these over the counter drugs for persons with peripheral neuropathies.

Dear Doctor:

As a CMT patient, I salute the efforts of the othopedic shoe technician to improve our gait, reference "Ask the Doctor," Summer 1993, THE CMTA REPORT. Fitted for AFO's three years ago, I don't wear them for reasons mentioned below. Unfortunately, I have been unable to find a pair of shoes that meets my needs. The responses to the technician's letter prompted me to consider the requirements for a shoe from the vantage point of a patient who has training as an engineer.

In the normal foot, a torque developed by the weight of the individual acting about the contact point of the shoe heel stretches the leg muscles, partially absorbing the energy of impact and slowing the sole of the shoe enough to avoid slap. The remainder of the energy is absorbed by the shoe and foot when the sole finally hits the pavement. In the CMT leg, the weakened muscles are unable to provide this energy absorbing torque. As the heel strikes, the sole is accelerated downward into the pavement causing the slap. There follows an abrupt impact force which gets transmitted up into the foot and skeletal structure; my brain rattles and vision waivers every time my foot strikes the pavement!

Moving the contact point forward does indeed reduce the torque requirement and causes the sole to strike with less velocity meaning less slapping noise; however, there remains energy to be absorbed. Cushioning and rocker soles are not the ultimate answer because of another problem. I need a heel and sole structure that provides lateral stability; that is, it needs to be wide enough and hard enough to prevent rocking side-to-side. Furthermore, the muscles in the backs of my legs tend to be perpetually taut. When I stand still, I soon have the sensation that I am about to fall over backwards; in compensating, I next find

myself falling forward. So shortening, narrowing, rocking and cushioning the sole/heel assembly work against me.

Which leads me to conclude that what is needed for CMT patients is a shoe design that will provide the necessary combination of energy absorption and stability. I envision a normal hard shoe with an elongated hollowed out area into which is fit a relatively soft rocker. The rocker would protrude above the sole such that it would provide a secondary strike point to the heel and a substitute for the normal leg's torque to slow the sole prior to impact. In other words, the shoe insert would supply the missing energy absorption of the leg muscles. Standing still, the insert would stay compressed such that the outer hard sole could provide the necessary stability. An added feature would be that the insert would contribute to a pushing off effect during walking.

There is yet another big plus to this concept in regards to braces. I have noticed that my foot slapping worsened over a very long period of time, 10-plus years, before I was forced to abandon conventional shoes and go to athletic "walking" shoes. Three years ago, I was advised to get braces. The first set of braces felt quite good, but were a little too stiff. In the process of adjustment, the braces got ruined and a second set was prepared. Whereas the first set easily fit several pairs of my existing shoes, the second set was quite bulky and would not. Steaming and stretching one pair of my shoes didn't help, so a pair of oversize shoes was ordered. After many adjustments of the soles on the new shoes, the much thicker braces still gave too stiff a gait to be comfortable. And they would fit only that one pair of shoes. That and other comfort problems didn't get resolved, so I finally returned to wearing just my athletic shoes. Now, examining those special shoes three years later, I notice that the heels are 3/4" longer than my regular shoes; the braces had to be heavier to offset the greater torque! So, the proposed shoe would lighten up AFO's because they would only have to cope with lateral ankle weakness.

The good news is that there is some existing technology available that could be incorporated into the proposed CMT shoe. I have worn AVIA walking shoes with a rocker insert almost exclusively for over five years. The rocker serves somewhat the function I describe but does not
(continued on page 13)

Patient Profile: George Horner

(Editor's note: Through a series of coincidences, the CMTA has found many new members because an article about the rescue of George Horner, a CMT patient, from his burning apartment appeared in the July 20, 1993 issue of Family Circle. Subsequently, President Karol Hitt had a letter printed in the "Now You're Talking" section of the same magazine and inquiries and responses have been plentiful. We thought an article about the man who quite innocently started all this would be interesting.)

George Ray Horner was born on a small truck farm southeast of Denison, Texas, on January 6, 1916. He was a frail boy with poor balance and very weak ankles, but he was required to do his share of the farm chores and he never knew that there was a serious reason for his weakness. He just accepted it as a fact of his life.

He began his schooling in a rural school at the age of 6 and was never able to excel at physical activities, so he pursued mental activities and became an honor student until the Great Depression forced him to drop out of school and leave home.

From the time of leaving home until he retired in 1981, George followed many paths of employment: bicycle messenger, mail clerk, political campaign worker, political appointee as committee clerk, reception clerk for the Texas House of Representatives, and after a move to Arizona, a fry cook, and door to door salesman until the outbreak of World War II. After several jobs in a company making howitzer gun barrels, he moved to San Francisco and returned to professional cooking, married and became a father.

After his daughter's graduation from college, marriage and a family breakup, he moved back to Texas to work as a security officer, orthopedic shoe salesman, and delinquent Visa accounts collector.

He became ill at the age of 70 with a stomach disorder, subsequently diagnosed as colon cancer. He became so weak from that illness and surgery that he became wheelchair bound, but with funding from MDA he was able to secure an Amigo electric scooter and a carrier for his car so that he could continue to be self-sustaining.

In January of 1989, he watched television reports of the great number of "work-able" handicapped citizens who were unable to find employment. He got an idea to found a non-profit corporation to act as a fundraising foundation to establish work centers for work-able handicapped citizens all over the United States. He founded the Institute of Opportunities for the Handicapped on February 10, 1989 as a Texas Non-Profit Public Charity Foundation. Brochure, board of directors information and plans of operation and goals are available to anyone by calling 817-472-6544.

The profile of George Horner is best summed up by the line, "It's not what we can't do, but what we can that counts." The CMTA salutes this courageous gentleman.

Editor's note: We would have liked a picture of George for this article but everything was burned in the fire.

CMT and Pregnancy



In the October issue of NEUROLOGY, an article entitled, "Pregnancy and delivery in Charcot-Marie-Tooth disease type 1", appeared. This topic is certainly of great interest to any CMT woman of child-bearing age. We will present a summary of the article here, but recommend that, if this is of importance to you, you send for a reprint of the article.

The authors studied the histories of 21 women who had had a total of 45 pregnancies. They found that, "Exacerbation (worsening) of symptoms was reported by 38% of the participants (8 of 21), who noticed a considerable deterioration of muscle weakness in 81% of their pregnancies (17 of 21). The results were examined according to age at onset: 16 patients had early-manifesting CMT with subjective disabilities from childhood or youth, and five had adult onset with a subclinical (mild symptoms) course of CMT up to their deliveries. Fifty percent (8 of 16) of the patients with early-manifesting CMT had a noticeable exacerbation of symptoms in pregnancy. In adult onset CMT, there was no influence of pregnancy on the disease, al-

though two subjects stated that their deliveries initiated the first symptoms."

The authors further stated, "Assuming that in long-standing CMT, weakness is more likely to be exacerbated by gestation (pregnancy), one could expect that those with exacerbation of symptoms in pregnancy were older than patients who remained stable throughout their pregnancies. On the contrary, we found that the average age at delivery of women who reported marked deterioration of symptoms in pregnancy (group 1) was 22.3 years, compared with 34.3 years in group 2 and 25.8 years in group 3. (editor's note: the three groups are: Group 1 = exacerbation of symptoms in at least one pregnancy. Group 2 = exacerbation in the period immediately after delivery of at least one pregnancy. Group 3 = no influence of any pregnancy on the patient.) This means that having children at a younger age cannot be recommended to minimize the negative influence of CMT."

The authors summarized, "In conclusion, there is no evidence of a deleterious

(harmful) effect of CMT 1 on the course and outcome of pregnancy and labor. The risk of a pregnancy-related exacerbation of CMT is 50% in patients with early-onset disease, whereas pregnancy has no influence on adult-onset CMT. Worsening is temporary in about 35% and persistent progression occurs in 65% of gestations (pregnancies) affected by exacerbation. Despite difficulties prior to pregnancy, the majority of women emphasize the positive aspects of a fulfilled family life and would encourage other patients not to avoid pregnancy, given that sufficient support is available."

For a copy of this article send \$2.00 and a business size SASE to CMTA/Pregnancy, 601 Upland Ave., Upland, PA 19015. For our medical professional readers the following is the abstract from the original article: "We report the obstetric complications and the influence of pregnancy and delivery in 21 Charcot-Marie-Tooth disease type 1 patients with 45 gestations. Sixteen patients had subjective disabilities from childhood or youth, and 5 with late onset had sub-

(continued on page 11)

A Personal Alternative to Bracing

I would like to share a "FIND" with other people living with CMT, an alternative to braces - sneakers. I know this won't be a suitable answer for everyone, but I want to share the facts so others may consider it. My history will explain the motivation. It has definitely made a difference for me.

Here's my story...

First showing symptoms of CMT at age three, I was pigeon-toed and tripped over my own feet. I wore shoes with corrective inserts until the age of seven, when after being diagnosed with CMT, I underwent a few corrective operations on both legs (bone and tendon work). I then wore below the knee metal leg braces with drop foot supports and Y straps until the age of 16 (1977). I don't remember how often I received new braces, but I do remember the feeling of having those brand new polished shoes and the firm sturdy support they provided before they broke in and loosened up. I clearly remember how noisy they always were. At 16, when I was getting ready for college, the doctors recommended some less visible alternatives. They included new plastic braces, ankle fusion surgery, and high top work boots tied tight around the ankle. The latter was dismissed by many doctors as ridiculous.

The bone fusion method was chosen by my parents. The surgery was first performed on my left ankle, but after several months it became apparent that the main joint would not completely heal. Partial fusion and a staple was holding it together. My surgeon then suggested another try with a bone graft. I didn't agree. I wasn't about to go through that experience again. My feeling at the time was, NOTHING would be worse than having that operation again.

My choice at the time was to walk with crutches, wear the high top workboots and dump the doctors. By the time I started college, walking with the crutches had built up my upper body and I appeared to be someone who broke his leg playing football or skiing. This wasn't so bad. I made the physical vs. psychological trade off. I lived this way for about 9 1/2 years when, after losing a lot of weight, I switched to walking with a cane. This became a very natural order of existence. I felt no desire to see any doctors or change the situation. The physical pain of the unhealed ankle was better than the alternative.



Steven E. Khosrova

A few years ago I dislocated my elbow and was treated by a doctor and physical therapist. It was then that I became more aware of how I had adapted my everyday life style to living with the weaknesses of CMT. I was getting involved in more social activities and felt more pain and restrictions because of the ankle. It was time to learn more about CMT and find out what alternatives were available.

It had been 23 years since I was first diagnosed and 14 since the failed surgery. Modern medicine MUST have made tremendous strides in all that time, or so I thought. While searching for "The Best" doctors in my area, I found that CMT was one of the 40 diseases under the MDA umbrella. In fact I was told it was one of the most common for the MDA Neuromuscular Disease Clinic at New York University Medical Center. NYU is where I had the ankle fusion surgery, so I felt comfortable with the environment. For my first visit with the doctors I was anxious to hear about the wonderful new methods they must be using for diagnosing CMT. Major Bummer - they still used the old electrical tests (NCV/EMG) that I had horrible memories of when I was seven. The rationalization started - "I was just a scared child then, it can't be as bad as I remember" WRONG! I don't think I have to explain further. The rediagnosis confirmed CMT Type 1. Everyone involved with the tests and my visits was wonderful, very understanding and supportive. The Director of the clinic, Dr. Dora Schively, and the Genetic Counselor of the Human Genetics Program at NYU, Elsa Reich, took all the time I needed to answer every question I had about the disease, my condition, and what I should expect in the future. My trust in doctors was renewed.

It was time to seek a surgeon for a second try. Maybe modern medicine would have SOME new methods to fix my ankle.

The first surgeon I visited was recommended as "One of the best". I know we have all heard that one. His plan of action was to perform a bone graft and then "Let the body heal itself" along with the comment "each try reduces the chances of success". Needless to say, that visit ended pretty quickly. My next try was the Staff Orthopedist of the Neuromuscular Disease Clinic - SUCCESS. Dr. Andrew Price was exactly the kind of Doctor I hoped for. Knowledgeable, experienced, open-minded to my suggestions, patient and willing to explain every detail of all my options. Surgery was not the only option he presented, but it was the course I chose because of my increased walking pain. I had the surgery a month later. Because of my obvious concerns over the chances of success, he offered the shotgun approach to the problem. Not only did he use a bone graft, but he used screws for compression and an EBI bone healing system. I'm very pleased with the results. The major problem was taken care of.

When it was time to get back on my feet, it was recommended that I consider supportive braces again. I refused. To me, this would have been regression. I agreed to visit an Orthotics Specialist to hear what he had to say and to see what I had already decided to turn down. I chose a man by the name of Steve Goldstein. We sat in his office for a while and talked over my concerns and physical situation. Steve had an idea to try something he had once seen in a trade show, but had never tried. He offered to bring a pair of shoes that had small vertical pockets in the ankle area. These pockets normally had plastic or graphite "bars" slid in from the top to provide extra support because of weak ankles or while recovering from foot surgery. The shoes are available in several models and don't look like orthopedic shoes at all. After consulting the supplier of the shoes, Steve received a pair of black shoes (that I could wear to work) and manufactured steel inserts to provide the extra support I required. In addition, he made orthotic shoe inserts with a "rocker bar" for additional walking assistance.

I was leaving on a business trip and thought it would be a good opportunity

(Continued on page 10)

to do a test drive. I was very happy and immediately requested another model, the sneakers. This was the first time I had ever worn sneakers. They felt very comfortable. There is a lot of ankle support, but the soles are flexible and the leather covering the forefoot has small ventilation holes to keep cool. Many friends who have known me since grade school have said that they have never seen me walk as well. Strangers have commented how they liked the look and asked where they could buy them. I have been wearing them for over a year now and am still excited by how great they feel. I'm waiting for the manufacturer (Kunzli) to come out with some newer models (fashions). The black shoes are for dress up occasions and the white sneakers are for everyday use. I moved back to the New Jersey office of my company where we are allowed to wear casual attire, so the sneakers have been worn almost every day for the last eleven months.

After sharing this find with other people living with CMT at a support group meeting, I decided to share it with everyone. I know this alternative to braces isn't for everyone. As you can see it was very important for me to find shoes that didn't have a medical appearance. If you are interested, Steve Goldstein welcomes your inquiries. You can contact him at: Med-Tech, 104 E.40th St, Suite 603, New York, NY 10016. Call 1-800-716-9482.

Sincerely,

Steven E. Khosrova

**Due to a shortage of space
Honoraria and Memorials will be
printed in the next issue of
CMTA Report.**

Items of Interest - cont'd from page 3
probably in Pennsylvania. She writes, "After starting on genealogy work with Duke University, I've become extremely motivated to find my lineage. I know my family settled in PA and remained there from 1750 until 1834 when only a few moved to Iowa. Their name is Myers, Minough, or Menough. We are trying to pinpoint CMT to the PA families and to see if there are Myers or Menoughs who gave the CMT gene. Their home was near Allentown, Lancaster County. I know it may aid researchers if they can pool families together to nail these genes." If you are a relative or know someone who might be, contact Mary Weber, 515-499-2042.

Dear CMTA,

I am a 38 year old man who was employed full time as a manager of a shoe store, acting as everything from manager down to janitor and shelf stocker. I participated in recreational sports such as volleyball and basketball. I participated in other normal activities as any other healthy person would. I had very slight manifestations of CMT, although it had never been diagnosed. I had slight hammer toes and a lack of musculature in the webbing between the thumb and index finger and a similar area in the shin area of both legs. None of these problems were disabling.

I then fell off of a step ladder and hit my hip on a chair and my lower back on a stool. I had no surgery or broken bones, but did have serious contusions in those areas for a number of months.

After the fall, there was a prompt acceleration of CMT symptoms such as pain in the back, weakness in the legs, instability and falling which has progressed to the point that within less than two years I have become totally disabled. I have filed a worker's compensation claim and a claim to the effect that the fall from the ladder accelerated my CMT. I would like to hear from other CMT friends that might have court cases along these lines as to how their cases came out and what medical aggravation of CMT was due to a traumatic incident. *(Editor's note: write to him in care of the CMTA)*

I have enjoyed reading the articles in the CMTA Report.

L. W. St. Joseph, MO

Dear CMTA,

I read the spring CMT Report with great interest. It is so exciting to read all that has been accomplished in your research into CMT. I am writing in response to your request that you want to hear from me. So here I am.

I first knew I had a problem at the age of 5 when my father declared I had the "gorramdest looking feet he had ever seen." I was taken to a doctor who prescribed that I sleep on my stomach with my feet hanging over the edge of the mattress to correct my tendency to pronation. Of course, it did not do any good.

I did not seem to have any trouble walking or running, but when I got into high school, my gym teacher ridiculed me

continuously for being lazy and uncooperative in gym class. I could not execute many of the exercises because of a weakness in my lower legs. Consequently, I got a "D" on my report card in gym for four years. It was frustrating because much as I tried, I couldn't do any better.

When I went to college, I experienced the same problem, but I had a more understanding teacher who gave me passing grades and tried to help me after class. She finally decided that I had a problem and let me go at my own speed. I learned to ski and play tennis, but I wasn't very proficient. About the same time, my friends began to question me as to why I walked funny and why I wasn't able to wear high heels.

My lifestyle changed when my daughter became a physical therapist and noted that when she gave me a feather test on my lower legs, she did not get a response. Finally, when I had to agree with her that I had a problem, she sent me to an orthopedist. He did not identify my problem, so he sent me to a specialist at Boston City Hospital. He told me that when he saw me walking down the hall, he knew my problem. He gave me various tests, and finally told me that I had CMT. He told me that the disease was not life-threatening, but that it was progressive. He prescribed short leg braces, and I was pleased to note that they really helped. This was in 1975.

In 1987, my daughter sent me to another therapist, and he prescribed new braces which included an ankle strap. They were a definite improvement. Also, he suggested my using a cane for added stability. I followed his advice to the letter as I wanted to remain mobile as long as possible. I think I do very well, everything considered. My family and friends are very supportive. It doesn't bother me that people stare at me. I have told some people that I would rather have them ask me why I walk this way than have them whisper about me behind my back.

I am a 76 year old widow with four children. So far there is no sign of CMT in any of my children. The oldest is 48 and the youngest is 37 so it would have probably shown up by now. I have 8 grandchildren.

I am sure my mother had a form of CMT as my father always ridiculed her for being clumsy and stumbling a lot. The last few years of her life she had to resort

(continued on p. 11)

Medical Advisory Board Meets

The Medical Advisory Board of the CMTA met in October during the proceedings of the American Neurology Association's annual meeting. The meeting was chaired by Dr. Robert E. Lovelace. Among those in attendance including Dr. P.K. Thomas from the United Kingdom were the Drs. Walter Bradley, Frances Dyro, Robert Miller, Gerald Weber, Jack Petajan, and Lowell Williams.

Call for Articles

The CMTA Report welcomes your ideas and article suggestions. For example, you may submit a human interest story telling of your experience of living with CMT. Also, medical professionals can forward articles of a clinical or medical nature that would be of general interest to our readership.

Pregnancy & CMT - cont'd from p. 8

clinical CMT when they became pregnant. The rate of obstetric complications in the study group was in accordance with that of the normal population, and there was no deleterious effect on fetal outcome. Of the 21 participants, 38% (8 of 21) reported an exacerbation of CMT in at least one pregnancy. These patients noted increasing weakness in 81% (17 of 21) of their gestations. A temporary worsening occurred in 35% (6 of 17) of these pregnancies, and neurologic disabilities persisted after 65% of the deliveries (11 of 17). Patients who had pregnancy-associated progression in the first gestation (7 of 21) experienced similar deterioration in subsequent pregnancies (10 of 11), ie, there is a high risk for recurrence of exacerbations. Four women (19%) stated that their last deliveries were responsible for either an exacerbation or the onset of the neuropathy. The remaining 9 patients (43%) denied any effect of their gestations on the progression of the neuropathy. Among the patients who had subjective disabilities from childhood or youth, the risk of a noticeable exacerbation in at least one pregnancy was 50% (8 of 16) and affected 81% of their gestations (17 of 21), whereas there was no influence of pregnancy in the 5 patients with adult-onset of CMT, although the first symptoms were noticed postpartum in 2 of them. Neither age at onset, age at delivery, interval between onset and delivery, or order or number of gestations are useful criteria for predicting a pregnancy-related progression of CMT 1."

During the meeting Celeste Chenet of Genica Pharmaceuticals reported on the new diagnostic blood test for type 1A CMT. (For information about the test contact Genica at 800-394-4493.) Dr. Elaine Alexander of Cephalon, Inc. discussed with the attending physicians the feasibility of conducting drug trials on CMT patients using a drug being developed by Cephalon. The drug trial being considered by the MAB neurologists is the use of IGF-1 (or Insulin-like Growth Factor), the potential of this having been presented in September at the Norris ALS Fetschrift (and Memorial) Symposium in San Francisco by Dr. Alexander and her colleagues. This will be shortly published in book form entitled rather appropriately "From Charcot to the Present and into the Future". It is contemplated that we will conduct a multicenter trial, in which Drs. Lovelace (The Neurological Institute, Columbia University) Bradley (Univ. of Miami), Petajan (Univ. of Utah), and possibly Dr. Miller (California Pacific Medical Center) will collaborate. Provisional results could be available by the time of the CMTA international meeting. Other trials being conducted on this chemical are with ALS and Diabetic Neuropathy. (The CMTA will report on this project as it happens.)

Dr. Jack Petejan presented a five part plan for the management of the patient with a chronic illness. Those five components are 1. Nutrition 2. Physical Activity 3. Self-Management (medication, pain management, injury prevention, relaxation, etc.) 4. Patient knowledge of the disease 5. Patient support systems (How the patient interacts with his/her support systems.)

Dr. Williams spoke on her work on the effect of trauma on the immune system of the CMT patient. Dr. Williams presented her hypothesis for the mechanism of this process citing several publications which suggest the immune system is unusually activated in CMT patients. She hypothesized that this inflammatory process may be an important pathologic process in the peripheral nerves. This is secondary to the basic genetic problem. For copies of this work, send \$ 2.00 to the CMTA asking for Dr. Williams' paper.

The meeting concluded with a brief discussion by Dr. Lovelace about the CMTA's proposed Third International CMT Conference in 1995.

Financial Notes - cont'd from p. 5

The Association occasionally receives donations of fixed assets. The value of these assets are not considered material and are therefore not reflected in the Association's funds.

Fund Accounting

To ensure observance of limitations and restrictions placed on the use of resources available to the organization, accounts are maintained in accordance with principles of fund accounting. This is the procedure by which resources for various purposes are classified into funds according to their nature and purposes. The assets, liabilities and fund balances of the Association are reported as follows:

*Operating fund - represents the portion of expendable funds available for support of operations

*Board designated - represents the portion designated by the board of directors to be used for research grants and education

*Restricted - represents the portion of fund restricted by the donors to be used for research grants and education

Property and Equipment and Depreciation

Property and equipment are reported at cost. Depreciation is provided on a straight-line basis over the estimated useful lives of the assets.

NOTE 2 - PROPERTY AND EQUIPMENT

Property and equipment consist of the following:

	1993	1992
Computer equipment	\$6,422	\$4,572
Less:		
Accumulated depreciation	1,787	686
	<u>\$4,635</u>	<u>\$3,886</u>

NOTE 3 - TAX STATUS

The organization is exempt from income tax under Section 501(c)(3) of the Internal Revenue Code and accordingly the financial statements do not reflect a provision for income taxes.

Letters - continued from p. 10

to a walker. I suspect that her sister also had symptoms in a mild form, but there were no signs in either of her two brothers. Members of her family will not discuss the disease with me.

My daughter has prescribed exercises for me - swimming, walking and others to keep my hands and arms nimble. I am now noticing a weakness in my hands, but I am trying to keep this problem at bay with exercises. I swim 3 times a week at a local pool, and one benefit is that I no longer have cramps in my lower legs.

I guess I have written enough about me, but I want you to know what moral support I get from your CMTA letters as well as the financial support I get from MDA who have paid for my braces and 3 pairs of custom-made shoes.

C.M. MA

Write us: Letters / The CMTA
901 Upland Avenue
Upland, PA 19015
215/499-7488

Don't Suffer - cont'd from page 2

mainly angry at those you need and depend upon the most - your husband, your adolescent or adult children or other close family members. You not only get angry at them for what they don't do for you, but more often than not, you get angry at them for what they do do for you and for how they do it.

Even with good intentions, no one is likely to do things for you exactly the way you would do them for yourself, especially when it comes to your personal care. Besides that, no one is likely to do things for you as quickly as you would do them for yourself. They may even forget to do some things altogether.

If, after many times, they still don't do things as you want them done - you get exasperated. And if you have to wait for what you need - you get impatient. So rather than appreciating what family members do for you, you end up being angry at them instead.

...rather than appreciating what family members do for you, you end up being angry at them instead.

If this happens to be the case, it may be you don't realize how much you expect from family members. Since you are your first priority, you expect to be their first priority too. And since you need them to help you with your life, you don't take kindly to their taking time out for their lives. This is most likely to be the case if your illness goes back to early childhood.

Even if your illness came later, you may have expected family members to do your bidding. But now, more than ever, you expect them to do your bidding and you get angry when they don't.

Be that as it may, anger toward those you need and depend upon is a high price to pay for holding on to what may be unreasonable expectations. To live with less anger, you have seen that your expectations take others' needs into consideration. If they don't, you can either modify your expectations or you can continue being angry at family members for not being the way you want them to be.

Similarly, if you want to live with less anger toward yourself, you need to ease up on your expectations for yourself as well. This requires making your illness and the limitations that go with it an acceptable rather than unacceptable part

of who you are. In other words, you need to develop an acceptable identity as "a person with an illness."

If you stop to think about it, to do the best you can do and to be the best you can be is more than enough to ask of yourself - with or without a chronic illness. And as an added benefit for accepting your own limitations, you'll be more tolerant of others' "imperfections." In other words, the more you accept yourself for who you are, the more you'll accept others for who they are. That's the way it works.

With a chronic illness, you're not only dependent on family members but on your doctor, too. Although you're dependent on your doctor for your physical well being, he affects you emotionally as well - and one of your emotional reactions may be anger.

You may get angry at your doctor because he doesn't make you better or even keep you from getting worse. Maybe you can't accept that your doctor doesn't know more about your disease and how to treat it than is medically known, that is, more than is written in medical journals and discussed at medical conferences. Maybe you can't accept there is no cure for your disease and you expect him to cure you. In either case, it's your unrealistic expectations, not your doctor's incompetence, that account for your anger. On the other hand, if your doctor is, in fact, not up on your disease, you should, of course, find one who is.

While you may get angry at your doctor for not doing more for you physically, you're more likely to get angry at him for how he affects you emotionally. He may be hard to reach on the phone, he may keep you waiting for hours to see him, he may rush you through appointments. He may say things that upset and frighten you or say so little you imagine all kinds of frightening things. Worst of all, he may attend to your body but ignore your feelings.

...In anger you feel self-righteous, in hurt you feel dependent and vulnerable.

It may be that your doctor is insensitive, inconsiderate and has little interest in you as a person. In this case, you would do well to find a doctor who is better for your emotional well-being - which is as important as your physical well-being for living with a chronic illness.

But maybe your expectations are inappropriate for a relationship between a doctor and patient. Maybe you expect

him to relate to you as if you were his only patient. Maybe you expect him to have the interest, concern and caring for you he would have if you were his child. Maybe you expect him to be God. Discomforting as it may be, you are one of his many patients; you're not his child and he's not God.

You can feel better by changing your expectations of what the other person is supposed to say or do...

But no matter how angry you get at your doctor, whether your expectations are appropriate or not - and you have to decide that - you can only affect his behavior toward you so much. You can't change him into someone he isn't. You can, however, either change your doctor or change your expectations of the doctor you have. Whichever you do, it's better than making anger part of your medical treatment.

You get angry at other people - family members and friends especially - not only because they don't act toward you the way you think they should, but because they hurt your feelings. In a previous article, I suggested ways of lessening hurt feelings by acquiring a fuller understanding of how your illness affects others and by helping others understand how your illness affects you, especially emotionally. What I suggested then would also calm your anger when your anger arises out of hurt feelings.

There are, however, additional ways to relieve anger arising from hurt feelings. This requires being aware of your hurt feelings and not just your anger, which you may not be. For one thing, anger tends to blot out hurt feelings. For another, anger focuses your attention on the other person - what the other person did or didn't do, which hurt feelings focuses your attention on yourself - what you need but didn't get. Besides that, in anger you feel self-righteous, while in hurt you feel dependent and vulnerable. You are, therefore, more likely to acknowledge anger than hurt because you feel better about yourself when you do. This is especially so for men who consider anger a sign of strength and hurt a sign of weakness.

However, by revealing your hurt feelings rather than expressing only your anger, you're more likely to improve the situation with the other person. Blaming someone in anger, more often than not, evokes a defensive or angry response, while revealing hurt feelings tends to

elicit a more sympathetic and caring response. This is so, because no matter why the other person said what he said or did what he did it's unlikely he wanted to hurt your feelings.

There is another advantage to acknowledging hurt feelings. In anger, whether you express it or not, you hold the other person responsible for your feelings. While in acknowledging hurt, even if only to yourself, you're more likely to assume some responsibility for your feelings. This is so, because in acknowledging hurt feelings, you're more apt to recognize your own unfulfilled need for consideration, understanding or sympathy as underlying your hurt and anger.

Maybe you expect others to read your mind rather than asking for what you want.

If you assume responsibility for your feelings, you don't have to depend solely on the other person changing his ways for you to feel better. You can feel better by changing your expectations of what the other person is supposed to say or do to satisfy you. This is important because your own expectations are more in your control than the other person's behavior.

There are still other ways you can improve the situations with those who anger you. It's possible, that in one way or another, you put people off and they then act toward you in ways that make you angry. Maybe you're not doing everything you can to make yourself better, physically or emotionally. Maybe you ask others to do things for you, you could do for yourself. Maybe you expect others to read your mind rather than asking for what you want. Maybe you're so demanding no one can do enough for you. Maybe you don't express appreciation for what others do for you and they resent your ingratitude. And who knows what else?

If you can increase your understanding of how you relate to others and guide or modify your behavior accordingly, you may evoke responses in others that are more to your liking. Isn't taking a good and understanding look at yourself worth living with less anger? I hope so.

Editor's note: "he" and "she" are used interchangeably in this article and are not meant to infer gender preferences.

protrude quite far enough; it would probably need some adjustment in density also. Sears sells "Durashock" work shoes by Wolverine with a patented protruding heel insert to absorb shock. And there is a special Ripplesole available with large transverse ridges that might do the trick if combined with a rigid outer sole.

Hopefully the preceding comments will give the orthopedic shoe technician an idea for a shoe design that will help improve my gait and that of my fellow CMT patients, and to keep us independent of bracing for as long as possible. I would welcome feedback on this; and I would be glad to volunteer as a tester of the new CMT shoe design.

R.H. MI

Editor's note: We gave this letter to Roger Marzano, a certified pedorthist and certified prosthetist, for his expert opinion. Mr. Marzano gave an outstanding presentation on "Footwear and Orthotics for the CMT Patient" at our October, Akron, Ohio conference.

The pedorthist/prosthetist replies:

I must compliment you on your concise description of the problems you have had in finding the correct devices for your ambulation.

I would like to begin by addressing your bracing requirements. The orthotic industry is advancing rapidly in developing components to address your particular needs. I feel that in the hands of the right orthotist, you could be braced quite successfully.

Secondly, the foot structure of most patients with advanced CMT necessitates very specific orthotic modalities. If the orthotist or pedorthist is not knowledgeable about CMT and its management, the chances for successful management are not as great. Your particular type of foot structure requires the understanding of specific forefoot to hindfoot relationships. I feel you could be successfully managed by the right person.

Finally, to address your footwear requirements, I would recommend your searching out a pedorthist to help you in your footwear considerations. They are trained to work on modifications to fine tune them to your requirements. I hope you find the right practitioner because I feel bracing would be in your best interests if done properly.

Roger Marzano, C.P., C.Ped.

Dear Doctor:

I'm a fifty-three year old retired male who's main problems are chronic fatigue and sinus problems. This has become such a problem that I doubt I could handle a normal work week any more. I find that I must take a 20 minute rest most afternoons and all of this is further complicated by minor to moderate insomnia and occasional stress. I've been to four neurologists and not one of them had ever heard of a fatigue and sinus connection to CMT. This really puzzles me, since this complaint is also expressed by other CMT patients and Dr. Williams touched on this briefly in the Winter '93 newsletter. I'm assuming that CMT can cause chronic fatigue, so please advise me if there are any tests to verify this. Also, what actual mechanism triggers the fatigue and what recommendations are given for dealing with the problem..

The Doctor replies:

You wish to know whether chronic fatigue and sinus problems can be related to CMT. The difficulty with your question is that fatigue is a common complaint with a large number of possible causes. Fatigue may result from infection, exercise, muscle weakness, lack of sleep, anemia, poor nutrition, thyroid disease, depression, or many other factors. Not everyone with CMT experiences unusual fatigue. In those that do, it is probably related to decreased strength and the extra effort required for even simple tasks such as walking. Best prevention is plenty of rest, good nutrition, and the avoidance of working or exercising to exhaustion. There is no evidence for an association between CMT and sinus problems.

The CMTA Report is published by the Charcot-Marie-Tooth Association, a registered non-profit 501 (C)(3) health organization. The newsletter is co-edited by Karol Hitt and Pat Dreibelbis. The layout is by Chesapeake Bay Design, 1418 Ticonderoga Ave., Ridgecrest, CA 93555.

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.

Support Group Notes

A primary goal of the CMTA is to become a truly successful advocate for those with CMT. Its message must reach the patients, their families, and the medical and research communities. Patient family support groups help carry out this function.

There are many CMTA support groups, but more groups are needed. The CMTA will help you set up a group in your area. For information about forming a group or being a local contact person please inform the CMTA by mail or call 215-499-7486.

Perhaps there is a group meeting near you. You are cordially invited to join these groups in their upcoming events.

Alabama - Greater Tennessee Valley
Bill Porter 205-386-6579W, 205-767-4181
Meets at ECM Hospital, Florence, AL

California-- Los Angeles Area
Oxnard-Thousand Oaks
Janice Hagadorn (805) 985-7332

Adelanta (High Desert)
Mary'L Michels (619) 246-7807

Canyon Country - Saugus
Sheila Levitch (805) 254-5322
Denise Miller (805) 251-44537

California-- San Diego
Gary Oleze (619) 944-0550

California-- San Francisco
David Berger (415) 491-4801

California-Santa Rosa
Freda K. Brown (707) 573-0181

Colorado-- Denver Area
Dr. Gregory Stilwell (719) 594-9920

Florida- South
Robyn Cohen (407) 622-5829

Massachusetts-- Boston
Donald Hay (617) 444-1627

Massachusetts- Southboro
Jim Lawrence (contact person)
(508) 460-6928

Michigan-- Brooklyn
Robert D. Allard (517) 592-5351

Michigan-- Detroit
Suzanne Tarpinian (313) 883-1123

Mississippi- Jackson
Julia Prevost (601) 885-6482
Henry & Brenda Herren (601) 885-6503

Missouri-- Kansas City
Sandra Toland (816) 756-2020

New Hampshire - Southern
Mary Nightly (contact person)
(603) 598-5451

New Jersey-- Central
Janet Saleh (908) 281-6289
Somerset Medical Center
Sommerville, NJ 08876

New Jersey-- Northern
Teresa Daino (201) 934-6241
Meetings: Englewood Hospital
Clinic Conference Room
350 Engle Street, Englewood, NJ

New Jersey-Millville Area
Linda Muhlig (contact person)
(609) 327-4392

New Mexico
Jesse Hostetler (contact person)
(505) 536-2890

New York - Brooklyn
Alan Latman (contact person)
(800) 227-1343

New York City
Diana Eline (201) 861-0425
Abby Wakefield (212) 722-8052

New York-- Long Island
Lauren Ugell (516) 433-5116

New York-- Rochester
Neale Bachmann (716) 554-6644
Bernice Roll (716) 584-3585

New York-- Westchester County
Kay Flynn (914) 793-4710

North Carolina--Eastern
Susan Salzberg (919) 967-3118
(919) 286-0411 (x6586) days
Durham VA Medical Center

Ohio-- Cleveland
Norma Markowitz (216) 247-8785

Oregon- Portland
Mary Elizabeth York (503) 246-4939

Pennsylvania-- Delaware Valley
Dennis Devlin (215) 269-2600 work
(215) 566-1882 home

Pennsylvania-Duryea
Patricia Zelenowski (contact person)
(717) 457-7067

Texas-- Greater Dallas Area
Dr. Karen Edelson, D.P.M. (214) 542-0048

Utah-- Salt Lake City
Marlene Russell (801) 966-7563 home
(801) 565-1212 work

Virginia-- Tidewater Area
Mary Jane King (804) 591-0516
Thelma Terry (804) 838-3279

West Virginia- Central
Joan Plant (304) 636-7152 (after 6pm)

Washington, DC - Baltimore, MD
Lorraine Middleton (202) 362-4617
Robert Kight (contact person)
(410) 668-3054

Board Member profile



Diane Freaney

CMTA Board member Diane Freaney was diagnosed with CMT in a not uncommon manner. That is, Diane's diagnosis came after her 15 month old daughter Allison was diagnosed. At that time Diane, her two sisters, her mother and grandmother were all diagnosed. Diane relates that they called themselves a family of "funny feet", but no one knew they had a hereditary neuropathy. With Allison's diagnosis many questions were answered, but many were then raised.

In her search for answers, Diane found the CMTA, volunteered her services, and has been the Treasurer since 1989. Accounting is Diane's field and after attending Syracuse University and fulfilling subsequent requirements, she became a certified public accountant (CPA). Currently, Diane is Chief Financial Officer for the Philadelphia Foundation, which manages endowment funds, makes grants to community organizations, and raises additional funds to meet changing community needs.

Diane's past volunteer service includes the Ambler YMCA, Miquon School, and Woman's Way. Education is once again a part of her busy life as she is enrolled in a Master's program at the University of Pennsylvania. The CMTA is fortunate to have Diane's dedication and efforts.

Alert - Alert - Alert

As of January 8, 1994
we will have a
new telephone area code
Our new number:
(610) 499-7486



Support Group Notes

Call Dave Christensen, support group leader for Augusta, Georgia, if you missed the October 23rd meeting but would like to be informed of future meetings. He can be reached at 706-738-5209. Dr. Tom Smith, Chairman of the board of directors of the Podiatry Institute in Tucker, Georgia, spoke at the October meeting and is willing to act as a resource person for the support group. Dave is currently hoping to get a winter meeting scheduled, but he needs to know that CMT patients and families in his area will come out to support the effort.

Don Hay, support group leader in the Boston area would like people in the following zip code areas to contact him about the possibility of setting up a meeting. If you live in 02192, 02194, 02026, 02030, 02090, 02157, 02181, 02161, call Don at 563-6417 and let him know of your interest in a meeting.

Referrals Available

The CMTA has compiled a list of neurologists, orthopedists, physiatrists (a physiatrist is a physician trained in physical medicine and rehabilitation) and podiatrists who have a special interest in CMT. We can also access respiratory specialists. Additionally, we have listings for pedorthists. A pedorthist is a practitioner who provides care to the patient by fitting orthopedic shoes and devices, at the direction of and in consultation with physicians.

To receive any of these referrals send a stamped self-addressed business-sized envelope indicating the geographic areas needed to: CMTA, 601 Upland Avenue, Upland, PA 19015.

For referrals for a hand surgeon contact the American Society for Hand Surgery, 3025 South Parker Road, Suite 3025, Aurora, CO 80014, 303/755-4588.



Drug List Updated

One drug has been added to the neurotoxic drug list. The drug is Taxol - a cancer drug. Note that at the bottom of the list two drugs have been added with the statement that they can be used, but with caution. Those two drugs are Lithium which is prescribed for manic depression or severe headaches, and Misomidazole which is a cancer drug.

CMTA Membership/Order Form

Name: _____

Address: _____

Phone Number: _____

Tell us about yourself:

- CMT Patient Medical Professional
 Interested Supporter CMT Family Member

Enclosed is:

- \$25 \$50
 \$100 other

for my membership in the CMTA (newsletter included in membership)

- At this time I cannot contribute to the CMTA but would like to receive mailings.

Publications and Tapes available from the CMTA

(Check to order)

- VCR Tape - CMT Neurology (\$15)
 VCR Tape - Physical Therapy & Occupational Therapy (\$15)
 VCR Tape - CMT Genetics (\$15)
 VCR Tape - Orthopedic Surgery & CMT (\$15)
 VCR Tapes (2) - Wilmington Del. Conference (\$25)

 Handbook (16 pp.) - CMT FACTS I (\$3)
 Handbook (24 pp.) - CMT FACTS II (\$5)

 Transcript - San Francisco CMT Conference (\$5)
 Letter - to Medical Professionals regarding the drug list (free to members with self addressed stamped business envelope)
 List - Physician Referrals (by state) (free to members with self addressed stamped business envelope) please list states: _____
 Medical Brochure - CMT (gray brochure) (one copy free with self addressed stamped business envelope)

Contributions are tax deductible.
Please make checks payable to the CMTA.

Total amount enclosed: _____

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.

MEDICAL ALERT



Certain Drugs Toxic to the Peripheral Nervous System

This is a list of neurotoxic drugs which could be harmful to the CMT patient.

Adriamycin
Alcohol
Amiodarone
Chloramphenicol
Cis-platinum
Dapsone
Diphenylhydantoin
(Dilantin)
Disulfiram (Antabuse)
Glutethimide (Doriden)
Gold
Hydralazine (Apresoline)
Isoniazid (INH)
Mega Dose of Vitamin A
Mega Dose of Vitamin D
Metronidazole (Flagyl)
Nitrofurantoin
(Furadantin, Macrochantin)
Nitrous Oxide
(chronic repeated inhalation)
Penicillin
(Large IV doses only)
Perhexiline (Pexid)
Pyridoxine (Vitamin B6)
Taxol
Vincristine

Lithium and Misomidazole can be used with caution

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

CMT FACTS II NOW AVAILABLE

The sequel to that "best seller" **CMT FACTS I** is now available and appropriately named **CMT FACTS II**. It covers topics pertinent to the CMT patient and family which were not covered in the first booklet. Some of the articles in this new publication include the American with Disabilities Act, Orthotics, Rehabilitative Medicine, Neurotoxic Drugs and Anesthesia, and Living with a Rare Disorder. The cost of this 24 page booklet is \$5.00 and may be ordered by completing the form on page 15 of this newsletter. **CMT FACTS I** may also be ordered and the cost for this 16 page booklet is \$3.00.

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, occasional partial sight and/or hearing loss problems and scoliosis (curvature of the spine) is sometimes present.
- 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 can now be diagnosed by a blood test.

THE CMTA REPORT

*information on Charcot-Marie-Tooth disease
from the*

Charcot-Marie-Tooth Association

Crozer Mills Enterprise Center
601 Upland Avenue
Upland, PA 19015

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