

# THE CMTA REPORT

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Providing information on Charcot-Marie-Tooth disease (a.k.a. Peroneal Muscular Atrophy or Hereditary Motor Sensory Neuropathy), the most common inherited neuropathy. Contents © 1994, CMTA. All rights reserved.

## Health Insurance... Who Covers What?

A CMTA member recently wrote expressing her concern about the difficulty choosing health insurance which covers the cost of AFO's (durable medical equipment in insurance language). This is a real concern for many of our members, and we decided to do our own investigation of what was currently available across the country. However, we were probably no more successful in finding out answers than many of you have been in trying to make a wise choice with regard to insurance companies.

In the Philadelphia area, we found that the Keystone Health Plan East offers a 60 day short term benefit for physical therapy and is the only plan which covers the full cost of durable medical equipment. Shoe orthotics are not covered and hand bracing and splinting are covered if pre-approved.

Blue-Cross and Blue Shield of Philadelphia cover physical therapy in two different ways. The first is as an outpatient in a member hospital.

After a \$75 deductible, the balance is covered.

The second treatment manner is at a licensed physical therapy office. This coverage is under major medical, and after the individual's deductible, 80% is covered. A molded cast, brace or supportive device is covered and all orthotics would be paid 80% after the initial deductible is met.

Blue Shield of Chicago pays 80% of leg braces after the individual's deductible is met. For physical therapy, they offer \$3,000.00/year after the deductible is met.

Aetna Health Plans, a major health care insurer across the country, gave no specific details as each plan they offer is tailored to the group who purchases it. Aetna establishes plans through employers for groups of 10 or more people.

The Guardian, which offers group insurance for small groups, provided us with the following statement: "The following applies to durable medical equipment. We must have advance written approval for durable medical equipment. We do not cover equipment if it is to alleviate pain or if the occurrence of re-injury is there."

Blue Shield of California responded that coverage depends on what each group

(cont'd on page 2)

## Patient Services... Where are they?

In December of 1993, the Muscular Dystrophy Association put into effect a shift from patient services - specifically durable medical equipment - to research. As a result, CMT patients can no longer depend on the MDA to provide braces, orthopedic shoes when prescribed in association with braces, walkers or other aids to daily living such as commode chairs or bath benches. The MDA continues to offer diagnostic evaluations for suspected CMT patients and upon diagnosis of CMT, the association will provide the following services: an annual or more frequent follow-up visit, up to

a limit of four per year; one consultation to evaluate for the need for physical therapy; one consultation annually to an occupational therapist and one consultation annually to a respiratory therapist. Genetic counseling is also provided to families of inherited neuromuscular diseases. The MDA also provides transportation to appointments at the nearest MDA clinic to your home providing that family or community resources are not available. MDA

continues to assist with the purchase of wheelchairs when they are prescribed by an MDA clinic physician.

With MDA redefining its objectives to emphasize research and not patient services such as orthotic devices and special shoes, where can the CMT patient who has no adequate medical insurance go? After some investigation, we found the following sources of assistance. The Shriner's Hospital network, which is located throughout the country, is the best source for the CMT child. They offer a complete

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#### FALL CONFERENCES

East Coast  
Philadelphia, PA  
September 10, 1994

Southern  
New Orleans, LA  
November 19, 1994

Details in Summer CMTA Facts

## Update on Nutrition Supplements



*This article is reprinted from The Orphan Disease Update of the National Organization for Rare Disorders (NORD) Vol. XI, Ed. 3, Spring 1994.*

On January 1, the new federal law governing food labeling went into effect. Despite heavy lobbying by the nutritional supplement industry to exempt vitamins, minerals and other dietary aids from the new law, nothing disastrous happened to the supplements sold in health food stores. For months the supplement industry had misinformed the public that should the new law go into effect, people would be forced to get their vitamins by prescription. The FDA had argued that the government did not intend to take these products off the market, nor would they require prescriptions. They simply want the supplement manufacturers to tell the truth on their labels, and to stop making false medical claims unless they provide scientific proof to back up their assertions.

Meanwhile, NORD's major concern has not been addressed by changes in law. Because some people with rare disorders, particularly inborn errors of metabolism, need vitamins, minerals and other nutritional supplements, we want to be sure that the products you buy from the health food store contain the ingredients listed on the label, and that they dissolve appropriately in the human body. Since the nutritional supplement industry is not regulated by the government, some companies sell inferior products that do not contain adequate amounts of the labeled ingredients, and they do not assure that the tablets or capsules dissolve in an adequate period of time in the human body.

The generic drug industry is required to abide by very stringent government standards in order to prove that their drugs are absolutely bioequivalent to brand name drugs. But the nutritional supplement industry had no standards until this year when the U.S. Pharmacopoeia (USP) finally issued public standards that address potency, dissolution and disintegration of nutritional supplements.

The USP is a quasi-government agency that sets standards for drugs that are used as the benchmark for quality. Companies that abide by these internationally respected standards will print "USP" on their labels. So the next time you want to

purchase a nutritional supplement, if you don't see USP anywhere on the label, buy another brand. Moreover, if you want to test the dissolution of your supplemental tablets, there is a simple home test. Just put the pills in a glass of vinegar and stir vigorously for 30 minutes. If the tablets don't disintegrate appropriately, take the remainder of the pills back to the store and demand your money back. Capsules, however, have a different standard: Within one hour 75% of their ingredients should dissolve.

The standards for vitamin and mineral supplements were published on November 15, 1993, by the U.S. Pharmacopoeia, and only those companies that choose to meet the standards will use them. This means that nutritional supplement makers are still permitted to sell inferior products that don't meet standards for potency, dissolution and disintegration. Additionally, if they do not actually meet USP standards but they print "USP" on their labels, there is no enforcement mechanism to punish manufacturers for lying. It will therefore be difficult to be a smart consumer and avoid wasting your hard earned money on inferior supplements that don't live up to USP standards. However, if you look for "USP" on supplement labels, you'll stand a better chance of buying quality products. §

### Patient Services - (cont'd from pg. 1)

range of services from diagnostic to surgery to physical therapy to bracing at no cost regardless of the family's ability to pay. Their services are available from birth through age 21. The Easter Seal Society offers physical therapy, occupational therapy and speech therapy to adults and children on an "ability to pay" basis. Other sources are the local service clubs such as Rotary International and the Junior Chamber of Commerce, among others. Each local branch chooses its philanthropic recipients, and you won't know if they could help you if you don't ask. Your own church might even be a source of one time help with braces or a computer to aid in schooling.

The great lesson to be gleaned from any search for assistance is that you will never know if there is help available if you don't ask. §

### Health Insurance - (cont'd from pg. 1)

selects, but that generally there is an 80/20 benefit on physical therapy. Regarding bracing, the representative with whom we spoke said that carriers have an obligation to look at quality of life versus medical necessity. He went on to say that if you don't cover ambulation, it often results in more medical problems. Again, since Blue Shield covers some 1.5 million people in California, benefits will vary from group to group.

Blue Shield of Miami also answered that coverage varies from group to group. Some policies exclude AFO's completely. Medical necessity is the key to all insurance providers. If the brace is deemed medically necessary, it should be covered after the deductible is met.

In Dallas, TX, the spokesperson for Blue Shield said that "functional necessity" is the defining term for coverage. Prosthetics and orthotics are lumped in with durable medical equipment in their coverage. Again, each group defines what it covers, but there is a deductible in all cases.

The final insurance group contacted, Blue Shield of St. Louis, said that each group has an individual contract and all include deductibles and co-payments. Custom-made orthotics that are approved by the medical claims department would be covered.

After researching the health insurance issue about bracing and physical therapy, two items frequently needed by CMT patients, we found that there is no easy answer. There was no one insurance carrier who offered full coverage of everything a patient might need. Each person must ask about coverage at his/her place of employment. Furthermore, he/she should request the employer to include coverage that benefits him/her. Sometimes small additions to a policy cost little or nothing. We urge you to become an informed and active consumer. The benefits could be great. §

#### Just the fax.

Is your office upgrading and replacing your current fax machine? We have a need for an auxiliary fax machine to be used off site this summer. Of course, the donation would be a charitable contribution.

# Items of Interest



**Item 1:** Referrals for a hand surgeon in your state can be obtained from the American Society for Surgery of the Hand. Their new address is 6060 Greenwood Plaza Blvd., Suite 100, Englewood, CO 80111-4801. Their new phone number is 303-771-9236. Please note that this is a new phone number. They will provide you with a list of hand surgeons for your state, which is updated weekly.

**Item 2:** The Alliance of Genetic Support Groups offers several publications for purchase which might be of interest. The most significant offering might be Health Insurance Guide which is available for \$10.00 by calling 1-800-336-GENE. They also offer pamphlets such as Informed Consent: Participation in Genetic Research Studies and other pamphlets related to support groups and genetics' issues. Call for further information.

**Item 3:** Some interesting quotes which came out of the ongoing health care reform issue follow:

*"We're confusing the fact that we have the finest physicians and hospitals in the world with the fact that we have the stupidist financing system for health care in the world."*

Hillary Rodham Clinton, 2/4/94

*"Whether health care reform is on-budget or off does not make a bit of difference to the 70,000 Americans who lost their insurance today, or the millions who are stuck in jobs because they have a pre-existing condition, or those who live in a county without a single doctor. Health care reform being off on on-budget will not stop the tremendous cost-shifting that raises health care expenses; it will not help small businesses give their workers health insurance coverage; and it will not streamline our inefficient administrative system. Only getting on with the real business of health care reform will do that."*

Senator Jay Rockefeller 2/14/94

*"There is no health finance tooth fairy who will give us health care for free. Let's grow up and do our duty; taxes and/or employer mandates. They are the options. Look no further."*

Rep. Pete Stark, 2/7/94

*"They're going up against the best that the trade associations have to offer. They're the most sophisticated. They're the best financed. And they're good at protecting what they have. If you're going to take on the AMA and the hospital association and the insurance companies, just get ready to understand they might not fight fair. And why would you expect them to fight fair if you're taking their livelihood away from them?"*

Rep. Dan Rostenkowski 2/23/94

*"The time has come to be counted, to stand up, to take a stand and to fight with us if you want to get something done. This is a fight, and if you want health reform, you're going to have to fight for it."*

President Clinton 2/16/94

**Item 4:** Hundreds of resources are now available to parents of children with special needs and the professionals providing services to them through LINC'S, a free electronic Bulletin Board System (BBS). Callers may access LINC'S-BBS with a computer and a modem, making available an expansive local and national resource directory, Internet electronic mail and newsgroups, local e-mail and information files. The 24 hour BBS number is 408-294-6933, settings are N-8-1, up to 14,400 baud. There are no on-line or registration fees. For additional information, or if you don't have a computer and need information, call PHP -The Family Resource Center for Children with Special Needs at 408-288-5010.

**Item 5:** More thoughts from Life's Little Instruction Book, Volume II by H. Jackson Brown, Jr.

- If you want to do something and you feel in your bones that it's the right thing to do, do it. Intuition is often as important as the facts.
- Pay as much attention to the things that are working positively in your life as you do to those that are giving you trouble.
- When you find someone doing small things well, put him or her in charge of bigger things.
- Every so often let your sense of adventure triumph over your good sense.



## Problems With Life Insurance?

Recently a patient wrote to us about her problem in obtaining life insurance. In her letter she stated, "Life insurance companies take their insurance rating information from a common pool. My experience is that the information they have in this pool is often faulty and inaccurate. As a result of this information, CMT people are generally charged outrageous premiums or are refused coverage due to their illness."

We feel that this is a question that should be investigated and are asking your assistance. We intend to coordinate with the law firm of Anderson, Kill, Olick and Oshinsky to evaluate the life insurance situation and to determine whether legal action is warranted, including the possibility of a class action suit on behalf of all people with CMT.

If you have been denied life insurance or are paying an unusually high premium, we need that information for our study. Please send your name and address, the name of the life insurance carrier, and brief description of the problem to:

Suzan Charlton, Esq.  
Anderson, Kill, et al.  
Suite 7500  
200 Pennsylvania Ave NW  
Washington, DC 20006.

We urge you to do this—  
it could benefit the whole CMT  
community. §

- Use a favorite picture of a loved one as a bookmark.
- Never lose your nerve, your temper or your car keys.

**Item 6:** There is a catalogue called Comfort House which offers products to make your life easier. The catalogue can be ordered by calling 1-800-359-7701. One of the items which we have already seen and used are kitchen tools with handles designed for comfort and performance. The extra plump handles give more grasping comfort so that you can chop, slice, and peel with less hand pressure and greater control. §

## A Support Group Fundraiser An Unusual Response A Patient Profile

*(Editor's note: We are constantly amazed at how one thing leads to another with regard to information flowing in and out of this office. This winter, a member of the Delaware Valley Support Group mailed out a flyer announcing a no-bake bake sale to raise money for the research fund. It contained a poem suggesting that bakeless cakes are much better to buy from a dietary standpoint and the money still goes to a good cause. In response, we received an improved poetic version from a patient who is a published poet. We wrote to encourage a few more poems from the patient and received such a wonderful letter that she became the focus of a patient profile. Here follows the sequence as it happened.)*

Rebecca Sand of the Delaware Valley Support Group of the CMTA devised and mailed to some 400 people a poem entitled, "The Sweet Smell of Bake-Less." The poem was successful in generating money for the research fund. A response from poet Ann Gasser of Reading, PA, (in addition to her contribution) was an improved version of our poem. It follows:

Stay out of the kitchen,  
Stay off of your feet;  
An oven-less cake is  
The very best treat.

There's nothing to mix.  
No wait while it bakes;  
A soupcon of caring  
Is all that it takes.

It never collapses,  
Tempt's palate or lips,  
And the very best thing of all is  
It stays off your hips.

A simple donation  
(Invisible cakes)  
For CMT Research  
Is all that it takes!

In response to our request to print her improved version of the poem, Ann sent a wonderful letter which forms the basis for the following patient profile.



Ann Gasser

Ann Gasser was gifted with creative energies early in her life and has found them to be a source of tremendous reward to this day. The following words are excerpted from a letter to the editor:

"In 1981, a series of small falls while painting scenery triggered the exacerbation of a relatively mild case of CMT to a debilitating problem of gradually decreasing mobility. By the end of the summer, I began to have excruciating pain in my lower back.

The next three years were spent being referred from one doctor to another, going through one test after another, trying to find the cause of the pain. I had known for 12 years that I had CMT, but it had never given me any pain or discomfort, and although I could never run fast or jump high, the only problems I had ever had were trying to find shoes that fit my high instep. I was getting extremely frustrated and happened to see an MDA ad on TV in which they mentioned CMT. I went to their clinic and after a couple of visits, I was told I had arthritis of the hips. They sent me to an orthopedic surgeon who advised replacing both hips and said there was hardly a week that went by when he did not see a patient with CMT. Being naive, I believed him. I did get a second opinion from another orthopedic surgeon who said the x-rays did not look that bad, but if the pain bothered me, to go ahead. While I was still in the hospital after the right hip was replaced, I began to realize that this surgeon had misled me, and when I asked him point blank how many hips he had replaced on CMT patients, he admitted that I was the first. From that time on, I have had to use a

cane to walk (now two canes) and needless to say, I did not have the second hip replaced.

When my problems first started in 1981, I tried to find other traces of CMT in my relatives. I knew my father could not have had it because he was athletic into his seventies and could perform the Russian dance with great agility - which I doubt anyone with CMT could do. My mother, on the other hand, had always had trouble climbing stairs without using the railing to pull herself up, and when she got into her fifties, had started to have poor balance. In the years following, she fell a number of times, breaking an ankle, a wrist, a leg, etc. - sometimes multiple breaks, and she had to use a cane, then two canes, then a walker, then a wheeled walker, and finally a wheelchair. Her mother had only lived to seventy-two, but I remember her walking through a room "touching" things to keep her balance, and she always wore high shoes, saying she had "weak ankles." When I would mention any of these things to my mother or her sister, wondering if my mother and grandmother had CMT, her sister said, "We never had anything like that in our family!" None of their family would even listen when I asked about family members with disabilities. And my mother said, "It had to start somewhere - how do you know it didn't start with you?"

Their denials made me feel angry, frustrated, and very much alone. At one point, I was very depressed and felt my husband would be better off without me because he likes to go places and do things that were becoming increasingly difficult for me to do. I thought if I were not around he could find someone else to help him lead a fuller life. I started to write out some of my feelings and discovered that I felt very much better when they had been expressed. My thoughts seemed to form themselves into rhyme at times and into free verse at other times. The writing became therapy.

In 1983, I noticed in our local Sunday paper that they were starting a poetry column. I decided to send in a poem I had written about a boy emerging from a coma after an automobile accident. "At a Hospital Bed" was my first published poem, and it was the first poem in the new poetry column. When the poem ap-

peared in the paper, I got a letter inviting me to join a group of local poets who meet each month. After several months of thinking it over, I went to one of their meetings, and have been involved ever since.

In 1984, I entered my first poem in a state contest. It won, and I was hooked. Since then my poetry has received recognition about 600 times in the form of prizes, honors, citations, and/or publication, and it has appeared in a number of poetry newsletters, journals, anthologies, and volumes of Prize Poems in several states as well as the annual issues of The National Federation of State Poetry Societies Prize Poems. I have more than a dozen volumes of poetry published and I lead poetry workshops and judge poetry contests.

Many things have been said about "facing your problems" and "not ignoring your problems." When it comes to a chronic disease like CMT, for which there is no cure, I feel that the best way to deal with it is to face it, accept the fact that you have it, but do not beat yourself over the head by focusing constantly on the disease and what cannot be done to help it. In fact, with a disease so little known by most doctors, the more you run from one doctor to another trying to find help, the more dangerous it can be for you. Doctors charge high prices and they feel they must justify those fees by doing something for you, even if it means prescribing treatments or drugs that do you more harm than good (what do they know?).

I feel I learned a valuable lesson a few years ago when I knew a woman who did not have CMT, but another physical problem that bothered her. She went from one doctor to another trying to find a cure for a condition that was not fatal but annoying. Each doctor gave her a new drug or referred her to someone else. After a few years of this, her body chemistry was so altered that her immune system was affected and she died from pneumonia which should have been able to be cured if she had left well enough alone.

### 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.

I have faced my disease and my limitations, and now I am concentrating on things that I can do, and do well. Perhaps if my CMT had not limited other activities, I would never have discovered that I am pretty darn good with a computer and I can write poetry that is appreciated by others.

There are two things that I would like to recommend to people who have CMT.

- Be exceedingly careful about falling. Try your very best not to fall. In at least some of us, our CMT is exacerbated by the trauma connected with falling.
- Find a magnificent obsession to occupy your mind so that you do not think about your physical limitations. Once you have faced your CMT and done what you can to improve your health, forget about it and get on with your life. §

*(Editor's note: If anyone is interested in raising money for the CMT Research Fund through the no-bake sale letter, please write to the office and we will be happy to supply you with the flyer. This is a great project for a support group or an individual. As we go to press, Rebecca Sand's effort has netted over \$600 for CMT research.)*



A Matter of Compassion  
Ann Gasser

Beneath half-open eyelids I can see  
cold reflections in bright floors  
that receive the tender loving care  
we patients would like to know.  
"Just push your button!", I am told  
I often lie in wounded dignity.

It is then I think of ancient Eskimos  
who rowed the infirm to solitary floes  
beneath bright stars.  
Under Northern Lights  
pain numbered to eternal sleep.

The question is, it seems to me,  
"Just who are the savages - they or we?"

## One Person CAN Make a Difference!

Bob Thurston was born in 1935 with a severe and progressive disability called Charcot-Marie-Tooth syndrome. He was the class president of North Yakima High School; he graduated summa cum laude from the University of Washington and he earned an MBA from Harvard University in 1959. His life was a testament to what one man can do, especially when he opens his heart and acts from his convictions. He taught those around him how much can be accomplished when people join together - one helping one.

Among other accomplishments, Bob was a worker with the state Division of Vocational Rehabilitation and a member of the Governor's Committee on Disability Issues and Employment in the State of Washington. The remarkable contributions that Bob Thurston made to the quality of life for disabled people in Washington include access to the voting and political process and the local transit systems, as well as the development of a state government access map. Bob left a legacy which included decreased physical barriers to access, increased opportunities for equal treatment in employment, changes in the attitude of the public toward the disabled, and increased opportunities to live independently with increased accessibility to public transportation.

Bob was a visible affirmation of the capabilities of people, demonstrating that persons with disabilities are indeed able to direct programs and activities that affect themselves and others. He was a role model of commitment, advocacy, philosophical insight and conscience. Among his many recognitions was the Bell Greve Memorial Award for "creative service to persons with disabilities and stimulation of public concern for their rehabilitation."

Bob Thurston died on February 27, 1994, but not without first leaving an indelible mark on his community and his state. §





## Exercise and CMT... One Patient's Experience

by Dana R. Schwertfeger

Though I welcome spring in all its glory, the warmer weather always leads to the unhappy discovery that the bulges around my waist do not all disappear along with winter's bulky sweaters. The fact that there suddenly also seems to be a lot less room in the seat of my pants is no doubt the result of indulging my sweet tooth from Thanksgiving to Easter, but whatever its origin, this unwanted accumulation of poundage around my middle makes my CMT much harder to live with.

I really noticed the extra weight the other day when I went for a walk. When I'm in top condition, I can easily walk three or four miles in an hour, but that day I was huffing and puffing by the second mile. And my feet hurt. I wear AFOs and I'm never particularly light on my feet, but with the added drag of twenty extra pounds, it felt like I had lead weights attached to my shoes. Since body weight figures prominently in calculating the total impact weight of walking or running, it's not hard to figure out that the twenty extra pounds landing with every step

means that walking any distance gives my feet quite a beating.

Of course, when I'm overweight, just standing around isn't any picnic, either. I spend a good part of the day on my feet, and I always feel much better when I'm around my ideal weight, of 170 lbs. Not only do my feet appreciate the lighter load, but my posture is also better. With the AFOs, I tend to bend forward a little at the knee to keep my balance, and the heavier I am the more I've got to rely on the braces to keep me upright. In the process, I also slouch forward more, and after a while, I can feel it in my back and shoulders.

The solution to my annual weight problem has always seemed pretty obvious. I've got to watch what I eat and get more exercise. However, while the need to reduce the amount of fats and sweets in my diet has always been clear, what type and amount of exercise I should get has not. The question of exercise and CMT is seldom addressed, so although I have a good idea what I can't do and shouldn't do, I know very little about what I can and should do. In the seventeen-odd years I've been grappling with the problem, I've found that walking is good all-around exercise even if it's made more difficult by CMT, and I've also learned that it's not only possible, but beneficial, to maintain upper-body strength as well.

As exercise physiologists say, if you can move it, you can strengthen it. I've found that a light to moderate regimen of stretching, calisthenics, and free weights enables me to stay in fairly good shape and even makes it easier for me to walk. I didn't fully understand the relationship between upper-body strength and walking until one day when I went for a walk after doing a back and shoulder routine and discovered that it was unusually difficult for me to walk because those muscles were tired. After a rest period, however, I noticed that walking was much easier. My back and shoulders were stronger and carried more of my weight, and I actually felt lighter on my feet.

While I've enjoyed a great deal of success with my exercise program, I'm also aware of how dangerous exercise can be if it's done improperly. Even for a person in good health, it's unwise to suddenly start an exercise program without first

consulting a doctor who may want to perform stress or other tests to determine what exercise level is appropriate. Those of us with CMT must be aware of some additional precautions. The most important is ditching the "no pain, no gain" philosophy. The worst you should feel is a pleasant soreness, never pain. I work out with weightlifters who regularly push themselves "past the burn" to the point where their muscles are actually being torn, but I never push myself that far. I work only until I get a "pump" and my muscles begin to tire. Otherwise, I could injure myself or work a muscle or a nerve past the point of recovery.

While I always try to pay attention to the feedback from my body - pain is generally an indicator that I'm doing something wrong - I don't always get the message because of CMT, so I think very carefully about each exercise. I have some hand involvement, so I use light weight and I do more reps, and I never put a load on a joint that isn't fully supported by muscle. I don't do squats, for example, because I don't have the lower-leg strength or the balance to work with more than my own body weight. I do a few sets of knee-bends, but I don't go all the way down "in the bucket." And, I always stretch before and after exercise. Flexibility is just as important as strength.

Before heading for the gym, there's one other important thing to remember. You don't go from flab to fitness in a few days. Beginners have a tendency to overdo it the first few times instead of letting their bodies gradually become accustomed to the increased activity, and they often wind up sore and stiff and barely able to move.

Starting out slow is a lesson I still haven't learned, but if I am otherwise faithful to my exercise program, I should have my weight down and be in pretty good shape by sometime around Thanksgiving... §

### CMTA Announces a New Program

Recently we received a call from a CMT family member whose father, a CMT patient, had just died. It was the wish of the father to have his body used for research, and to accomplish this, the family member requested assistance from the CMTA. We called upon a member of our Medical Advisory Board, and as a result, tissue samples from the deceased gentleman were obtained and sent to three research laboratories.

As a consequence of this effort, it is apparent that there is a need for this coordinating service. Therefore, the CMTA has instituted such a program. We have contacted research scientists and alerted them to the availability of this service. If you (a CMT patient) or a CMT family member wishes to be a part of this program, call or write to the CMTA and we will furnish you with the details. This can be done at the time of death or in advance.

Speaking for the research community and the CMT community, both of whom will benefit, we thank this bereaved family and laud their thoughtfulness. We offer our condolences. The patriarch of this family was a true giver.

Wishing  
Ann Gasser

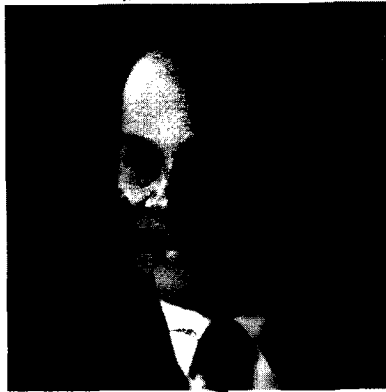


Oh, to fly  
as the soaring hawk,  
rising with the thermal currents  
far above the problems of this world,  
held high in God's hand.

## Patient Profile

*Editor's Note: CMT is found on all the continents and is truly an international problem. We became acquainted with a CMT patient of significant accomplishment in Amman, Jordan, and decided to profile this interesting and accomplished man.*

Elia Atalla was born in Jerusalem in 1932. He was one of six children, two of whom inherited Charcot-Marie-Tooth disorder from their mother. Elia was diagnosed with the disorder in London when he was 27 years old. He graduated with a degree in accounting from the London School of Accountancy and worked with various banking institutions in Jerusalem and Amman. He is best known for his "hands on" management style and in addition to his banking work, he is active in social work and as President of the Amman-Philadelphia International Rotary Club.



Elia Atalla

In his position as president of the Rotary Club, he was honored to preside at a meeting on environmental dangers at which Queen Noor of Jordan spoke on the Rotary's program, Preserve Planet Earth. She addressed the problem of the widespread environmental degradation

resulting from the 1991 Persian Gulf War.

During his research on CMT, Elia found out that the disease was originally in Europe and it traveled to the East, especially his area, via the crusaders and to the West, the USA, via the immigrants. Some areas of the Middle East have no CMT, like Saudi Arabia, because the crusaders did not reach there.

Elia is now 63 years old and after an operation in March of 1993, finds that his handwriting has greatly improved. He is retired and is working for himself on research as well as doing volunteer social work to help others. He has worked to establish a new Rotary club named the Amman Jerash Rotary Club, and it was chartered in December of 1993. As Elia says, "you can see that when there is a will, there is a way to do anything." §

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## How CMT Affects the Body's Function

by Lorraine Middleton  
Support Group Leader, Washington, DC

Dr. Ingrid Gheen addressed the November 15, 1993, meeting of the Washington, DC, area support group. As a physiatrist, Dr. Gheen deals with the way CMT affects the body's functioning.

In a brief overview of CMT symptoms, Dr. Gheen explained that the degeneration of nerves and muscles in the extremities, which proceeds slowly and insidiously for many years, eventually distorts the ability of the feet and legs to produce a proper gait. The muscles which run from the side of the knee and wrap partially around the shin, terminating at the toes, are generally the first to atrophy. The muscles underneath the foot and extending up the back of the leg remain strong and continue to exert a pull downward on the toes. This imbalance between two sets of muscles is what may eventually result in the classic high-arched frozen "CMT foot" with hammer toes and twisted bones. One indication of the weakness of the peroneal muscles that lift the toes is demonstrated when the CMT patient tries to walk on his heels. Most of the people in our support group have some degree of this weakness. In contrast, most can still walk on their toes, demonstrating that the muscles beneath the foot remain strong.

In some patients, in addition to weakness of the peroneal muscles, the intrinsic muscles which hold the foot together may deteriorate. The feet may rotate inward (known as pronation) or outward (known as supination.) Extremely flat feet (pes planar) may be present in some patients, along with the more common high arched foot (pes cavus).

Typically, a CMT patient has many years of gradually worsening symptoms, during which time he or she may adapt the high steppage gait characteristic of CMT. There may be many falls, sprained ankles and even broken bones before the patient investigates special footwear and/or bracing to improve gait. Dr. Gheen emphasized that the primary reasons for utilizing bracing and specialized footwear are to promote safety, improve mobility and prevent further damage to muscles and joints brought on by an inefficient and jarring gait. The human body is designed to produce very efficient locomotion and when anything interferes with that system, excessive fatigue is produced. When joints are forced to work in a less than ideal way, they can become damaged and the normal process of aging is accelerated.

Both bracing and footwear may have to be continuously modified as the foot's condition changes. Since both sensory

and motor neurons are involved in CMT, the brace and footwear must be comfortable and not produce raw or chaffed areas which could become infected without the patient realizing it.

Dr. Gheen's advice is that any CMT patient requiring bracing be seen regularly by a physical therapist in order to correct and maintain tone in weakened muscles in other parts of the foot, ankle and upper leg. Subtle changes in gait can be noted and corrected so as not to further damage joints in the knees and hips. Perhaps most important, since the brace tends to "fix" the foot and ankle in one position for long periods of time, it is important to practice an individually tailored program of exercises to maintain strength and flexibility. A trained physical therapist is the best source for developing such a program.

Dr. Gheen demonstrated the most common types of bracing used by members of our group (there were 25 present). They include: hinged plastic ankle foot orthoses; metal fixed braces; molded in one piece ankle foot orthoses; orthotic inserts; ankle foot orthoses with arch supports molded in; extra wide and deep shoes; tennis shoes (such as New Balance, designed for general athletics); ankle-high boots and Mary Janes with ankle straps. §



## Ask the Doctor



**Dear Doctor:**

How can a doctor differentiate between carpal tunnel syndrome and the problems inherent in the hands and arms of CMT patients? Also, how can I decide whether to have the surgery for carpal tunnel, and if I have the surgery, what is the probability of success given my CMT?

**The Doctor replies:**

*The carpal tunnel syndrome does not appear to occur with increased frequency in patients with Charcot-Marie-Tooth disease. In the cases where I have diagnosed the condition in patients with Charcot-Marie-Tooth disease, I have found that it has been a result of cumulative or repetitive trauma.*

*The diagnosis is a clinical one, based on the occurrence of wrist and hand pain, numbness and tingling in the hand, intermittently, with activities, and at night. These symptoms are identical to those in patients without Charcot-Marie-Tooth disease.*

*Because Charcot-Marie-Tooth disease causes weakness and wasting of all the intrinsic hand muscles, and sensory deficits in all the digits, it is not possible to use those findings to confirm the diagnosis. However, it is possible to look for median nerve irritability at the wrist with provocative maneuvers.*

*Finally, nerve conduction studies may be helpful in confirming the diagnosis by examining for a relative prolongation of the median motor distal latency as compared to that of the ulnar motor distal latency.*

*In the patients with Charcot-Marie-Tooth disease in whom I have diagnosed carpal tunnel syndrome, the surgery has been successful in decreasing or relieving their symptoms.*

*Charles Jablecki, M.D.  
San Diego, CA*

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.

**Dear Doctor,**

Besides cramps in my lower arm, hands, legs and feet, my wrists and ankles hurt (feel sprained), my muscles are tired and sore as if I always have the flu. Besides mocking carpal tunnel in my wrists and hands, I have scoliosis in my lower back. Lately, I wake up in the night with my lower arms so numb that I have to shake them repeatedly to get circulation in them, and, at times, I get sharp pains down my arms.

If the carpal tunnel problems and the scoliosis problems are related to CMT, can they be effectively treated?

My last question is a tough one. I have no functional impairment during a doctor's exam, yet my muscles tire/get stiff and hurt more with repetitive actions. When I overdo, which isn't hard, the pain is so bad that I get severe headaches. If I pace myself slowly, I can have some good days. Please tell me, "how can muscles react so differently at different times?"

**The Doctor replies:**

*Carpal tunnel type problems can be treated if they are indeed carpal tunnel. I would suggest an appropriate neurological exam and a trial of conservative treatment before thinking about surgery.*

*In order to answer the question about scoliosis, I would need to know the age of the patient, the extent of the problem and whether or not there is any pain (I would suspect not) or any problem with breathing.*

*The final question about the difference in muscle reaction is that it is possible with tiredness, as the patient has observed himself.*

**Dear Doctor:**

Can you tell me if Mevacor should be avoided if you have CMT? It seems like I have become much worse very fast since starting mevacor 2 years ago. One of the adverse reactions is myopathy in a few patients. I am 64.

**The Doctor replies:**

*Patients with CMT should avoid all drugs that are neurotoxic and other drugs that may be neuro or myotoxic. Mevacor (Lovastatin) is an inhibitor of HMG co-enzyme A reductase and is useful in the treatment of hypercholesterolemia. It does affect the liver*

*and there are a number of reports of it producing muscle weakness with muscle necrosis and myoglobinuria. However, this is rare (<0.5% of patients), and usually occurs in patients who are also on other drugs such as cyclosporin, gemfibrozil, nicotinic acid and erythromycin. Muscle involvement can be detected usually by an elevated serum creatine kinase, (CK). If the patient has weakness and the CK is elevated, the Mevacor should be stopped and the myopathy will improve.*

*If a patient with CMT feels weaker on the drug Mevacor, the serum CK should be obtained, and if it is elevated (compared to a previous study if available), the drug should be discontinued. The serum cholesterol can be controlled usually by other means, such as diet and exercise. Drug effects in patients with CMT vary, depending on the type of neuropathic process and genetic defect. I advise all patients with chronic hereditary neuropathy to avoid all medications except essential therapy. It would be important for the patient to contact the local medical doctor to determine whether Mevacor is essential in the treatment of an elevated cholesterol in that patient.*

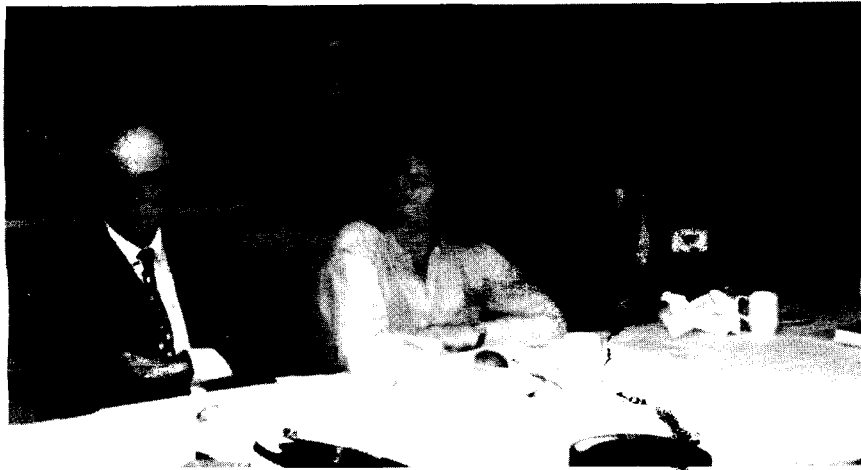
**Dear Doctor:**

I have been having problems lately with pain in my upper back, neck, and shoulders. There appears to be no reason for this pain, so I and my doctor are assuming that my unnatural gait and posture are taking their toll on my back muscles. The pain is not that bad and is really not the problem. The problem is that when this pain gets worse, I sometimes experience terrifying and debilitating episodes (lasting for one to two weeks) of something that my neurologist has diagnosed as vaso-vagal syncope. Could this be connected with CMT somehow and if not, are there exercises out there for the CMT back?

**The Doctor Replies:**

*There are several components to the question. Postural alterations and compensatory mechanisms in patients with Charcot-Marie-Tooth Disease can stress the muscles of the spinal axis (neck, upper and lower back). However, there are a number of other possible contributing factors including age, degree and pattern of weakness, etc. An*





Medical Advisory Board Members meeting in New York City

examination by an appropriate physician is often warranted to rule out other causative factors, e.g. disc disease. At times, patients may respond to a home exercise regimen as well as to judicious use of analgesic - anti-inflammatory medications not contraindicated in patients with CMT Disease. The exercise regimen would emphasize flexibility although some strengthening and endurance training might be of value as long as over stress and fatigue can be avoided. Obviously, given the considerable variation of clinical findings in patients with CMT disease, it is difficult to generalize concerning exercises; however, a gentle progressive program for shoulder mobility, hip rotation, hamstring stretching and heelcord stretching is often of value.

Patients with CMT disease can have autonomic nervous system involvement, but, fortunately, disturbing symptoms related to this are not very common. The pain could trigger vaso-vagal response, but if this persists, then further medical evaluation may be indicated. Diabetes mellitus should be ruled out if not yet done so.

**Dear Doctor,**

I am interested in learning more about the CMT neurotoxic drug list. Why is a drug placed on the list? What is a "mega dose"? Regarding pyridoxine (Vitamin B6), should we avoid multivitamins with B6?

**The Doctor Replies:**

The neurotoxic drug list was compiled by neurologists and is continually monitored by them for additions and corrections. A mega dose is defined as 10 times the RDA (recommended daily allowance). A daily multivitamin capsule

should not be a problem. (Editor's note for further information refer to "CMT FACTS II" pages 7 & 19.)

**Dear Doctor;**

My husband was diagnosed with CMT last year...his doctor suggested that he have a muscle and a nerve biopsy to determine what kind of CMT affected him, and to see if other neuropathies might be present. The doctor mentioned that the nerve biopsy would create a temporary loss in sensation in a small (the size of a quarter) area of his foot.

Unfortunately, the side effects of the biopsy were much more serious than predicted. He was unable to walk at all for three weeks, and required physical therapy for quite a while thereafter to rehabilitate his leg. The nerve biopsy left almost half of his left foot with significantly reduced sensation. This loss seems to be permanent. The progression of his CMT accelerated following these biopsies and I suspect that the injury caused by the surgery is responsible.

I am curious to know whether other CMT patients have had similar experiences with muscle or nerve biopsies and whether routine biopsies may present more risk to CMT patients than to others.

**The Doctor Replies:**

According to your description in the letter, it seems that your husband had a biopsy of the sural nerve, either at the ankle or at the midcalf level. Since you also mentioned a muscle biopsy, the midcalf level that exposes the calf muscle and the sural nerve is the best approach for a combined nerve and muscle biopsy. The sural nerve biopsy at either level produces a permanent loss of sensation in the lateral (external) side of the heel

and foot. This numbness is noticed less frequently in patients with CMT. The reason is that between 30% to 60% of patients with all forms of CMT already have numbness in both feet and hands as part of the disease, according to a survey by the CMT Association. Also, a deficit of sensation to pricking pain, preceding the biopsy, is very frequent in patients with CMT 1. Discomfort, manifested by sensations of pricking, burning, aching, numbness and swelling of the ankle are frequent complaints after a sural nerve biopsy. These symptoms disappear in more than 60% of the patients at three months and in most patients after a year. Why your husband was unable to walk for three weeks is difficult to explain unless some of the symptoms listed above were the cause. Because the sural nerve does not carry fibers that control movements of the foot or leg, weakness in these areas is not seen after sural nerve biopsies.

Acceleration of CMT symptoms, that is, weakness and wasting of the foot and hand muscles, does not occur after sural nerve biopsies. Biopsies of nerve, muscle or any other tissue don't present more risk to CMT patients than to any other patients. In most patients, a sural nerve biopsy is not necessary for making a diagnosis of CMT. The clinical features, the electrodiagnostic studies (nerve conduction velocities), and the DNA analysis for CMT 1A (commercially available at this time) are enough to confirm the diagnosis. In cases where there is no family history of CMT or where the clinical features are not typical of CMT, a sural nerve biopsy may be necessary to make the diagnosis. §

### 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 Surgery of the Hand, 6060 Greenwood Plaza Blvd., Suite 100, Englewood, CO 80111-4801, ph.303/771-9236. §

## Letters to the Editor



Dear CMTA,

As a kid in the 1930's I was a classic klutz: walked funny, lousy athlete, hated Gym classes, etc. I even wore glasses. My mother was concerned about my abnormally high-arched feet so she took me to Chiropodists and Chiropractors who -- depending on their specialty-- either mashed their fists into the arch of my foot or trimmed my toe nails before sending me home. At 17 the army turned me down because of my feet, but not until I was 20 years old did I learn that I had a congenital defect...and I got the diagnosis almost by accident.

My sister thought her infant son might have a problem with this foot and she asked if I would drive them to the NY Orthopedic Hospital for a check-up. I thought, "What the heck, as long as I'm here I might as well get checked, too". Her son was fine but they said I had Freidrich's Ataxia. I didn't know it at the time, but Freidrich's is a very debilitating and life threatening disease.

The doctor suggested an ankle fusion so I could stop tripping over my own feet. I was out of a job and not doing much of anything else with my life, so I agreed and within a month I had the operation. When they took the casts off 3 months later, I had a severe drop -foot.

My first brace was a wire-spring contraption which fastened to the back of my shoes and quickly chewed-up all my shoes. Fortunately my muscles revived before I ran out of shoes, and I only needed the braces for a couple of months.

For the next 10 years I didn't feel particularly handicapped. When I was in college, I bought a motor scooter so I could get from one end of the campus to the other, but, in general, I didn't spend any time thinking about my future. As a matter of fact, I had married one of the nurses I met in the hospital, and she didn't worry about it either...we had 5 children (none of the children ever showed CMT symptoms).

In my early 30's my mild dropfoot was getting worse. I started buying lightweight shoes, and I would slide my feet across the floor instead of lifting my foot for short steps. I decided it was time to get a dropfoot brace. But first I searched for something that I could hide inside my shoes. I found an orthotist in Silver Spring, Md, who crafted a dropfoot brace made of leather which I could wear in-

side my socks. For the next 15 years, I lived (and suffered) with these.

I sweat a lot and when the perspiration hit the leather, it formed a bond with my skin which took the skin off my ankles every summer. I could wear socks under the braces, but if you've ever been in the Washington, DC, area during the hot sticky summers, you know that no sane person would wear two pairs of socks. There was no easy solution, and I just did the best I could. My idea of a relaxing summer weekend was to stay home and go barefoot. But I had a price for going barefoot: I would periodically stub my big toe and take off the top 1/4" of flesh and bleed like a pig (still do that).

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*I sometimes think I could have been more comfortable with a very obvious disability because I could have relaxed with it instead of constantly hiding it.*

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At age 35, I decided to switch occupations and get out of Electrical Engineering and into Technical Sales because it paid better. Since sales would necessitate a fair amount of walking, I made an appointment with a neurologist in Bethesda, MD, to find out what my prognosis might be. His considered opinion was, "If that's what you want to do, do it." So, I did it. Oh yes, he said I probably didn't have Freidrich's because if I did, I probably wouldn't be around anymore...it was probably CMT.

Sometime after that, I had a muscle biopsy which confirmed CMT. I didn't think the biopsy would be much of a procedure, but I found out that climbing steps after a biopsy is very painful for many weeks after the operation.

With my orthotics on, I was able to get around pretty well. My pride coerced me to hide my disability, and I did it so well that not even my family paid much attention to my problems. They might have considered giving me special attention, but probably figured I would be offended if they made concessions. I sometimes think I could have been more comfortable with a very obvious disability because I could have relaxed with it instead of constantly hiding it.

On a couple of occasions over the years, I asked my orthotist if some of the new plastics might be better than leather, but he was anti-plastic, "They break and cut

you". After 15 years of suffering with the leather against my skin, I decided to do my own research. Using the computer terminals at National Library of Medicine, I found a plastic orthotics manufacturer located in Germany, and they referred me to their distributor in New Jersey who sent me a catalog. There it was: a pre-formed brace that came in three different sizes. Now all I had to do was figure out what size I needed and order a pair.

I learned that the distributor would not sell directly to a patient, but I now had my own company so when I placed my order I gave them the distinct impression that I was in the orthotics business. The distributor was very helpful. He explained that the plastic softened at an oven temperature of approximately 350° F and could then be formed over a plaster leg mold (you can't be in the CMT club if you don't have a plaster mold of your leg.)

The cost at that time was only \$40 per brace which was a small fraction of what my leather orthotics cost me (I never knew that MDA would pay for them!). When the braces arrived, they were oversized so I first had to cut them down to size with an electric jig saw. I then laid them on a board in a 350° oven for about 10 minutes and then handformed them to my plaster molds (I wore heavy gloves). Once I started wearing them, I never went back to my \$300 leather braces.

Now remember, I said I was a klutz. What I did with the braces didn't take a lot of skill. In fact, over the years I managed to ruin two braces: one by carelessly overheating (it just melted) and the other by cutting where I shouldn't have. I now have two pairs of orthotics that I've been wearing for over 10 years. Of course, no two braces look alike. But if you own some kind of suitable electric saw and aren't afraid to experiment, you too can do this at home. Five years ago, I got a pair of professionally crafted \$300 vacuum-formed plastic braces which I thought might offer some improvement over what I had fabricated, but I like mine better.

To complete my CMT-life story I should mention that at age 40 it became increasingly difficult to stand without holding on to something for balance. If I'm walking there's no problem, but as soon as I stop I have to grab for something like a

(continued on p.12)

## Some Questions and Answers about Living with Chronic Illness

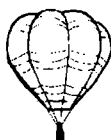


The authors of the book, Sick and Tired of Feeling Sick and Tired, Paul Donoghue and Mary Siegel, are practicing psychologists who have been on a lecture tour with their book and have prepared some questions and answers which they often hear in the course of their presentations. They present them for your information, and they encourage you to send in questions and answers of your own regarding how you deal with an "invisible", or rare, chronic illness.

**Q.** How do I tell my spouse enough about my symptoms and how I'm feeling so that I get empathy about my condition without producing paralyzing over-concern?

**A.** You need your spouse to know you, to be aware of what you are going through, to understand and to comfort. Measuring how much to share to get your needs met without triggering panic in the other can be a sensitive task, challenging your skill in articulating your condition, as well as your understanding of your spouse's limits. Every relationship is unique as is every person, so no formula would apply to all couples. But maybe the focus should be less on how much to share than on how to listen to your spouse's concern. In other words, try to understand your spouse's fear and helplessness without trying to control his reactions. Your spouse needs your understanding as you need his or hers. And your spouse needs to share whatever feelings are stimulated by your illness as much as you do. Illness, like all adversity, challenges people who love one another and who need one another to meet deeply by sharing and listening. Too often this opportunity is missed by overprotectiveness on both sides.

Unanswerable  
by Ann Gasser



Are we reality, or are we dreams?

Without a point of reference,  
who can say?

And does it really matter anyway?

Reality is only what it seems.

**Q.** What do I do when I tell my wife that I feel rotten and she says, "You know nothing can be done, so you just have to get over it,"?

**A.** If you felt "rotten" before telling your wife, you have to feel a whole lot worse after hearing her "pep talk." It is important to find out what she is feeling that makes her say what she does and what her intention is in saying it. She might be impatient with what she hears as complaining. She might feel helpless. She might be fearful that you are going to give up. As to her intention - she might be trying to motivate you, encourage you, or whack you. Whatever she is feeling or intending needs to be discussed and understood.

It is also important that you share how hurtful, frustrating, and ineffective her words are to you. You need to share your pain and your discouragement and you need to be listened to, not prodded; comforted, not made to feel worse. What you want to avoid are more interactions like the one that you quote. Such exchanges breed hurt, resentment, and guilt on both sides. Get beneath the surface of her words and yours and make contact. She has a right to her feelings and needs. You have a right to yours. But, the need to be aired and understood, not acted out in fruitless, repeated parleys.

**Q.** How do I cope when I feel so fatigued and my mind keeps going but my body won't?

**A.** It is very frustrating and discouraging to be unable to do what you want to do - when you have five things that you were going to do today and now you can't get the energy to do one of them. A temptation to ignore fatigue and go ahead anyway might seem brave, but is often a mistake resulting in exhaustion and despair. Better to admit the truth, face the reality that today you are limited by fatigue. Don't panic and don't give in to thoughts that defeat you, "What if I'm always going to be this tired?" "If I don't get these things done, I'm useless." "Why can't I be like Suzy, or Martha, or my sister, or my neighbor?" Simply, honestly, wisely accept that today you are limited. Then think of what you can do, rather than what you can't. If you can catch up on bills, letters, work you can do sitting, do so and forget about cleaning the basement. If you can't go on the hike, maybe you could rent a movie or

read a book. Be gentle with yourself; do what you can do today - leave tomorrow alone until it's today.

**Q.** I had my first symptom 3 years before I was diagnosed. I really thought I was going crazy. Now that I have a diagnosis, I am so angry with the people, including the doctors, who didn't believe that I was sick. I don't know how to let go of the anger.

**A.** At one of our lectures, a woman told us she thought we would appreciate the epitaph that she saw recently on a headstone. It read, "I said I was really sick!" But, being able to say, "I told you so" is not a very satisfying way to release feelings triggered by others' failure to listen to you or trust in you.

The period of time that a person endures seeking a diagnosis is a time-consuming, costly and enervating experience. It threatens self-esteem, shakes self-confidence, and often causes discord in families. It triggers fear, self-doubt, self-dislike and humiliation. For many people, it causes a great deal of anger.

There is nothing wrong with anger. Your anger seems appropriate and even a sign of your self-respect. Your anger only becomes harmful when you turn it against yourself or against others. For example, "giving up in anger" is harmful, as is giving in to self-pity, or overeating or drinking. These behaviors resolve nothing and they don't release anger. They invite guilt and self-dislike. So does kicking the dog or your spouse or friend.

Admit the anger, express it in a forceful letter to a pertinent doctor, for example. Bang your mattress with a tennis racket or smash an old glass against a wall. Then, channel your anger into energetic pursuit of whatever helps you to feel better - exercise, research into your illness, or meaningful conversation with someone who listens.

**Q.** Sometimes when I go to the doctor, I don't think he is really listening. I think he's a great doctor and I don't want to stop going to him. What do I do to get him to listen?

**A.** We would all like to think that we are good listeners, yet few people really listen and doctors are no exception. But, we desperately need our doctor to slow down, pay close attention and to under-

(continued on p.12)

## Rochester, NY Conference Coming June 4th

A CMT patient/family conference is scheduled for Saturday June 4, 1994, at Strong Memorial Hospital, 601 Elmwood Avenue, Rochester, NY. The conference will begin at 9:15 am with registration. The program will begin at 10:00 am and will conclude at approximately 4:00 pm. The conference will be a full day of information from neurologists, physical therapists, orthotists, and other experts on CMT. Lunch is included in the \$20 registration fee. All speakers will allow time in their presentations for questions from the audience. This conference is being organized by Drs. Ann Moss and Charles Thornton in cooperation with the CMTA.

If you are interested in attending, to register for the conference fill out the form below and return it to the office of the CMTA by June 1, 1994.

| <b>CMT Conference Registration</b>   |  |
|--|--|
| Name(s) of attendees   | _____  |
| Address  | _____  |
| Telephone Number   | _____  |
| Fee for one person   | \$ 20.00   |
| <i>(Fees include morning coffee, lunch, and all postage and mailings.)</i> |  |
| Number of persons attending  | _____  |
| Amount of money enclosed   | \$ _____   |
| Return this form by June 1, 1994 to:                                       | Pat Dreibelbis - CMTA<br>601 Upland Ave.<br>Upland, PA 19015 |

## Dealing with Chronic Illness cont'd

*stand. We don't want to be dismissed, ignored and judged by someone whom we need so much.*

*Two paths of action: First, learn to be a good listener yourself. Practice with your children, or with friends. Give the other person your undivided attention, listen carefully, and then offer back what you have heard to see if what you have understood is really what was meant. Our book and other sources teach this skill. Now you are ready for step two...getting your doctor to listen as you have learned. If you sense that he is not hearing you, say so. "Doctor, I don't think you understood me." or "Doctor, I need you to listen to me rather than dismiss me."*

*Once you know what listening really entails, you are in a position to know when you are not experiencing such attention and can then ask for it. Sometimes even asking, "What did you hear me say?" can give you the satisfaction of confirming whether or not the other is listening.*

*Such expectation of full attention is not impertinence or boldness. You can't expect to be helped if you are not heard. Learn to listen, then expect it and learn to get it from those you need - doctors included. §*

## Letters - cont'd from p.10

wall, a desk, a wife...anything stable. I can't stand in a line or chit-chat at cocktail parties unless I can lean on something. I don't like using a cane, but I have a clever fold-up cane which I use when travelling. It consists of several aluminum tubes with a bungee cord connected through the middle. When it's opened, the contracting bungee cord snaps the tubes into place. When the tubes click-clack into place, airport security guards frequently start reaching for their holsters.

I'm appalled at the number of years it's taken me to figure out some things related to my CMT problems. For instance, I only recently discovered that the lightness of a shoe was less important than the type of last it used. I was a big Hush-Puppy user, but now I'm much more comfortable in Rockport walking shoes (unfortunately the newest models come with a raised back and I can't get them over my orthotic without the raised back folding into the back of the shoe). Finding a comfortable shoe every few years is probably my biggest ongoing problem.

I only recently learned that there was a national CMT organization (is there a turf battle between the various organizations?). I got the information from another handicapped organization whom I contacted via Compuserve. Which brings up another topic... why can't we exchange information by computer? I got a lot out of reading Steven Khosrova's article in the Fall '93 *CMTA Report* wherein he analyzed the dynamics of walking. It's the kind of information that could be permanently available on an electronic bulletin board as opposed to a one-shot newsletter. Is there an existing Electronic Bulletin Board out there somewhere? Since CMT people can't hold pens very well anyway, they should get computer literate in order to communicate!

I'm now 65 and retired in Southern California. I really enjoy swimming and cycling, and I'm so glad that my degree of CMT involvement doesn't interfere with these exercises. It's almost a necessity for me to live in a warm climate because my lower legs turn to ice when the tem-

perature goes below 50 degrees. For 10 years I wondered why my feet got so cold before someone offered me this explanation: there's no muscle stimulation to dilate the blood vessels so the circulation doesn't respond to temperature changes. Again, a bulletin board could permanently include tidbits like that

Perhaps some of you would like to know where I get my orthotics, but I don't want to publish it in the *CMTA Report* because it might blow my cover with the distributor. I've made the information available to the national office. Perhaps they can find a friendly professional (with malpractice insurance) who can order them for you...they now cost \$65 per brace.

Writing this was a real chore, I hope it helps someone.

R.K. Coronado, CA



*Dear Friends,*

I've been taking primrose oil for nine years. While reading an article from my neurologist, I saw that people with CMT have a deficiency of linoleic acid. Checking this out with my doctor who is nutritionally oriented, I found that this was true for me. Perhaps I am the first person to do this. It was not suggested to me by my neurologist. If you have not come across this yet, you might want to tell your readers about it. I have no way to know if it has slowed down the disease, because my symptoms are described as very slowly progressive. I'm 59 and frequently travel around the US putting on seminars. People are amazed at my stamina.

Three years ago I was saved from having to wear braces by finding some special lace up half boots with elastic at the back of the ankle. In case anyone is in this phase, it could keep them walking without further aid for quite some time. Baker's Shoes, 2209 Universtiy Square, Tampa, FL 33612/ University Mall, 1-813-977-9156 or 813-284-8994. The assistant manager, Dawn Massey, ordered shoes, style #97885-06490-3 -Half Boot from Yugoslavia, \$34.00 on sale, \$39 with tax or \$49 at the regular price. She told me that I could give her phone number to anyone who wants to order. With a long skirt or pants, one can look quite fashionable in these boots.

Thank you again for the good work you are doing! Best wishes for much success.

*R.B. Seattle, WA*

*Dear CMTA,*

I would like to respond to your article on pregnancy and CMT with my own experiences. I am 24 and I have mild to moderate CMT. I had been told since early childhood that pregnancy would exacerbate my condition. I did become pregnant inadvertently and was quite alarmed. Relief soon came when I read about pregnancy related complaints of healthy people. I was no different for the first two trimesters. I maintained a full time job with a two hour daily bus com-

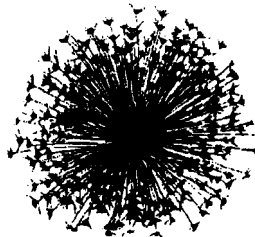
mute then came home to cook and clean. I had never felt better. For the first time in my life, I slept peacefully. In my last trimester, my hips began to hurt at night and I had to prop my knees apart with pillows. That was my only complaint. However, when I hit 7 months, I began feeling weakness in my knees. I fell three times without hurting myself. The fourth time I got banged up pretty badly and quit my job. (I was a temp.) For the next ten weeks. I stayed at home most of the day-leaving only to visit in-laws or to go to the health club. Swimming was wonderful.

My son came almost three weeks early. Labor was unremarkable and manageable. They tried to give me an epidural, but the catheter would not fit into my spinal area. The doctor theorized that my disease may have caused a fat build-up that prevented the epidural from working. I went on without anesthesia and did fine.

One hour after delivering my son, I walked into the shower, dressed myself, and visited the nursery. Now, six months later, I am more active than I have been in years. There is no notice of the weakness I encountered during pregnancy.

My son is beautiful and worth every minute of the pain and fear.

*L.W. Exton, PA*



*Dear CMTA,*

I just received a flyer about the new journal Common Journeys. I haven't yet seen a copy, but you might be interested in it or in passing on the information to newsletter readers. Common Journeys is a journal for those who live with chronic pain or illness and the people who love them. It is a quarterly journal comprised of personal experiences - stories, essays, poems, and thoughts- as they relate to the ways in which chronic pain or illness affects daily life. The subscription cost is \$16 for an individual. A sample copy is \$5. Write to Common Journeys, P.O. Box 17003, Minneapolis, MN 55417.

*J.R. Brooklyn, NY*

*Dear CMTA,*

I have CMT and am beginning to notice the effects in my hands now. When writing, for instance, my hand quickly cramps. It will often be sore for several hours if I write for too long. I have discovered, however, that I can write for much longer when I use a flair pen. I don't need to press down as hard with this type compared to a ball-point pen. Best of all, I use the computer to type as much as possible. This does not cause my hands to hurt because they are in a more relaxed, open position when typing.

Thanks for the newsletters and regional conferences. It is quite helpful to gain knowledge and strategies from others.

*K.V. Walworth, NY*

*Dear CMTA,*

I am wondering if you might have an address at which I can contact the noted Dr. James Lupski. I would appreciate it if you could help me. I would very much like to write to him and express my thanks for his spending all his time and hard work for the research for CMT. The test which he developed is how my CMT was diagnosed and that put an end to my wondering what was wrong. I would also like to thank you for the CMT Association. Your seasonal paper has helped me very much to understand CMT since my diagnosis almost one year ago.

*Thanks*

*M.A. New Vienna, OH*

*(Editor's note: M.A. is a teenager... we were struck by her thoughtfulness and her sincerity.)*

Write us:

Letters - CMTA

601 Upland Ave.  
Upland, PA 19015  
ph. 610/499-7486



### Blood Test Available

The blood test for diagnosing CMT Type 1A found on chromosome 17 is available from Genica Pharmaceuticals. They 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. The cost of the test is \$395.00. §

# CMTA Contacts



Following is a list of CMTA contact persons and support group leaders. 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 610-499-7486.

\*denotes support group leader

## Alabama / Greater Tennessee Valley

\*Bill Porter 205/386-6579W  
205/767-4181

## California

\*Janice Hagadorn 805/985-7332 after 5  
\*Sheila Levitch 805/254-5322  
\*Denise Miller 805/251-44537  
\*Gary Oleze 619/944-0550  
10am-5:30 pm 619/436-2116  
\*David Berger 415/491-4801  
\*Freda K. Brown 707/573-0181  
Eda Adams, will return calls  
916/677-6460  
Felice Gail Viggers, 805/492-2840  
Verna M. Sabo, 818/892-6706  
Mary Micalizzi, after 6pm  
619/441-2432  
Bob Hedge, 9am-5pm 310/645-2761

## Colorado - Denver Area

\*Dr. Gregory Stilwell 719/594-9920

## Colorado:

Roberta Cummings, 719/846-5611

## District Of Columbia:

\*Lorraine Middleton, 6pm-9pm  
202/362-4617  
Robert Kight 410/668-3054

## Florida:

William Brady 904/443-6271  
Mary Beeler, 9am-8pm 407/295-6215  
Harold Wilson 407/465-3656  
Pat Ports, M, W, F, 4pm-9pm,  
407/965-3691  
Joe Ellenbogen 305/921-4660  
Edward Carhart, 9:30am-5:30pm  
305/567-1066  
Beatrice Bannister 407/737-3267  
\*Robyn Cohen 407/622-5829  
8pm-9:30pm M-F  
weekends anytime

## Georgia:

Nancy Lee McCutchen 404/925-1020

## Louisiana

Bobbie Marberry 504/872-0895

## Maryland

Jean Iler 410/987-5432  
Linda Ember Miller 410/882-4019

## Massachusetts

Wayne Cardillo 413/298-3156  
\*Donald Hay 9am-7pm, 617/444-1627  
Jim Lawrence 508/660-6928

## Michigan

\*Robert D. Allard 313/592-5351  
\*Suzanne Tarpinian 313/883-1123  
Laurie Vasquez 517/893-4125

## Mississippi

\*Julia Prevost 601/885-6482  
\*Henry & Brenda Herren 601/885-6503

## Minnesota

Grace Wangaard 612/496-0255

## Missouri

Allan Degenhardt 816/942-1817  
\*Sandra Toland 816/756-2020

## New Hampshire

Mary Nightly 603/598-5451

## New Jersey

\*Janet Saleh 908/281-6289  
\*Teresa Daino 201/934-6241  
Linda Muhlig 609/327-4392  
Gary Orson, Mon-Fri 6pm-10pm  
& weekends 609/584-9025  
Russell Weiss 908/536-6700

## New Mexico

Jesse Hostetler 505/536-2890

## New York

Alan Latman 800/227-1343  
\*Diana Eline 201/861-0425 before 9pm  
\*Abby Wakefield 212/722-8052  
\*Lauren Ugell 516/433-5116  
\*Neale Bachmann /716 554-6644  
\*Bernice Roll 716/584-3585  
\*Kay Flynn 914/793-4710  
Angela Piersimoni, after 2pm  
607/562-8823  
Sharon McAvey, afternoon & evening  
718/380-3792  
William Carrington, 4pm-11pm  
718/486-6953

## North Carolina

\*Susan Salzberg, 7am-3:30pm  
919/286-0411 x 6586  
5pm-9pm 919/967-3118

## Ohio

Roger Emmons 216/286-6485  
Suzanne Lammi 513/339-4312  
\*Norma Markowitz 216/247-8785

## Oklahoma

Leah Holden 405/255-4491

## Oregon

\*Mary Elizabeth York 503/246-4939

## Pennsylvania

\*Dennis Devlin 215/269-2600 work  
610/566-1882 home  
Patricia Zelenowski 717/457-7067  
Camille Walsh 215-747-5321  
Janet Fierst 412/487-0757  
Dennis Devlin, evenings  
610/566-1882  
Mary MacMinn 215/322-1073  
Carol Henderson 215/424-1176

## Rhode Island:

Robert Matteucci  
evenings, 401/647-9154

## Texas

\*Dr. Karen Edelson, D.P.M.  
214/542-0048  
M,T,Th, 8:30am-5pm, 214/542-0122  
Tony Collette, 1pm-8pm, 713/699-8432  
Ken Kerby 817/282-9329

## Virginia

\*Mary Jane King 804/591-0516  
\*Thelma Terry 804/838-3279

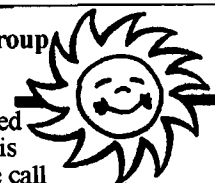
## West Virginia

\*Joan Plant 304/636-7152 after 6pm  
L.Ben Simmers 304/693-7731  
Beverly Simmers 304/364-5309  
Ronald & Rebecca Sampson,  
304/636-7449 24 hours  
Barbara Compton 24 hours  
304/636-5456

## Washington - Naselle

Marlene Russell 206/484-3116

A new support group  
is forming in the  
Los Angeles area.  
If you are interested  
in being part of this  
new group, please call  
Bob Hedge at 310-645-2761.



| Honoraria                 |                            |
|---------------------------|----------------------------|
| In honor of               | By                         |
| M/M Edward Sachs          | Mary Laber                 |
| Christine Schwab          | Gisela Schwab              |
| Eileen Meltzer            | Shirlee Bank<br>and family |
| M/M Edward Sachs          | Mike Wurzburg              |
| Christine Schwab          | Mary Ruotolo               |
| Maggie & James<br>Leonard | Susan Steingarten          |
| Jay & Kim Lamar           | Judith Lamar               |
| Robert Switzer            | Marjorie Cowan             |
| M/M Hart Wurzburg         | M/M George Frank           |
| M/M Hart Wurzburg         | M/M William Luria          |
| Rebecca Sand              | Ada Cipler Hyams           |
| Rebecca Sand              | Pearl & Percy Sand         |
| Rebecca Sand              | Jean Foremann              |
| Percy Sand                | Lionel Ferris              |

| Memorials          |   |
|--------------------|---|
| In Memory Of:      | By  |
| Howard S. Foley    | The Gorrill Family<br>Diane & Bill Armstrong<br>The Aramian Family<br>Ralph Richle<br>Carolyn Wollen<br>Charles Winchester<br>Howard Sheppard       |
| Adelaide Aldridge  | Emily Sanson<br>Rosemary Davis<br>Paul & Maggie von Kempf<br>Frederick & Ruth Etheridge<br>Barbara Winther<br>Marian & George Hopiak<br>JaNell Cook |
| Estelle McDonald   | Roy & Marie Brunson   |
| Jules Schwartz     | Reeva Sterling  |
| Jules Schwartz     | Tillie Wiener   |
| Bertha Koloian     | Richard Koloian   |
| Dr. Gerald Gilbert | Marilyn Prashker  |
| Charles Graham     | Marilyn Prashker  |
| Sophie Nerenberg   | Estelle Bandler   |
| Harold Lauman      | Marilyn & Don Conlan  |
| Leona Haley        | Kay Flynn   |
| Rose Bucchieri     | Kay Flynn   |
| Ralph Rust         | Mr. & Mrs. Donald Rehm  |

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

## 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)
- 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) (please send SASE) please list states: \_\_\_\_\_
- Medical Brochure - CMT (gray brochure) (one copy free with self addressed stamped business envelope)

Total amount enclosed: \_\_\_\_\_ Contributions are tax deductible. Please make checks payable to the CMTA.

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 (Apressoline)  
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, Misomidazole and Zolof  
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, 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 disorders  
from the

### Charcot-Marie-Tooth Association

Crozer Mills Enterprise Center  
601 Upland Avenue  
Upland, PA 19015

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