

THE CMTA REPORT

Winter 1992

Vol.7/No.1

Providing information on Charcot-Marie-Tooth disease (a.k.a. Peroneal Muscular Atrophy or Hereditary Motor Sensory Neuropathy), the most common inherited neurological disorder. Contents © 1992, CMTA. All rights reserved.



NUTRITION AND CMT

The question of diet and the CMT patient is one that we are frequently asked here at the CMTA. Therefore, we were delighted that this issue was addressed at the Dallas, Texas CMT patient/family conference by Janice Nance, R.D., L.D. Ms Nance is a nutritionist at the Texas Scottish Rite hospital in Dallas. We are grateful to Ms Nance for editing this article.

In her presentation, which related specifically to CMT children, but which is equally applicable to adults, Ms Nance stressed the necessity of avoiding obesity and staying lean. The causes of obesity are threefold, and they are: overeating, inactivity and metabolic abnormalities. The first two are far more common than the third.

There are several reasons why obesity should be prevented and they are:

- Obesity decreases the availability of oxygen to tissues. Oxygen is necessary for the proper functioning of every cell in your body.

- Obesity compromises physical performance. Physical activity becomes more difficult.
- Obesity raises the energy cost of activity. The obese person requires more energy to perform a task than a lean person.
- Obesity increases the demands on the heart and lungs. They must work harder.
- Obesity increases the stress on joints. CMT joints are already stressed from the disorder.
- Obesity causes a decrease in the self-esteem of the person.

Editor's note: During the CMT patient/family conference at Johns Hopkins Medical Center, Baltimore, MD Dr. John Griffin affirmed Ms Nance's position by saying, "Lean and mean should be the motto of the CMT patient."

The primary way of maintaining proper body weight is through eating management. Ms Nance set forth a four part program for weight control. The first goal is to maintain a positive eating/feeding relationship. Meals should be balanced and the atmosphere should be pleasant with no stress. Meals should not be the time for quarreling and the various threats that can arise from parental frustration with childish eating quirks. The second suggestion is to maintain a structure of meals and snacks. Serve meals and snacks on a regular schedule. The third part of the program is to teach the child orderly and positive eating. In other words view proper eating as a necessary and pleasant part of life.

The last part of the program, but certainly not the least, is to moderate the

caloric density (the number of calories/bite of food), and to learn the difference between true hunger versus the need for satisfaction. Caloric density refers to the number of calories in a food. Food is divided into three categories... proteins, carbohydrates, and fat with fat having twice the calories per gram of proteins and carbohydrates. Fats are indeed very calorically dense foods. Many calories can be avoided by serving for snacks low calorie fruits and vegetables instead of high calorie chips, cookies, nuts, ice cream etc. Many calories can also be saved by recognizing that many times we eat when we are not truly hungry, but rather when we are bored, upset, tired, etc.

Ms Nance concluded her presentation by advocating that the most natural and the freshest foods obtainable be eaten. The ratio of fat to protein to carbohydrate she recommended is a diet of 25% fat, 20% protein and 55% carbohydrate. Finally, the question was raised about vitamin supplements, and the answer was that they are unnecessary. A well balanced diet will naturally provide the necessary vitamins and minerals.

Editor's note: CMT patients are advised to avoid mega doses of some vitamins. See the Drug Alert on page 2.



IN THIS ISSUE:

Nutrition and CMT	cover
Medical Alert.....	p. 2
Tape Rental	p. 2
Tidbits.....	p. 2
CMT Patient Profiles.....	p. 3,4 & 7
Support Groups.....	p. 5
Ask the Doctor	p. 6
Ilizarov Procedure.....	p. 8
Physician Referrals	p. 9
Florida Conference.....	p. 11
<i>I want to be in touch!</i>	p. 11
CMT & Discrimination	p. 11

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, Macrodantin)
Nitrous Oxide
(chronic repeated inhalation)
Penicillin
(Large IV doses only)
Pyridoxine (Vitamin B6)
Vincristine

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

VCR Tape Rental

The CMTA has available for rental four lectures which were taped at patient conferences sponsored by the Foundation. The tapes are for play on a VHS VCR. Beta tapes are not available. The speakers are authorities in their fields and lecture topics include: Neurology, Physical Therapy, CMT Genetics, and Orthopedic Surgery.

Single lecture tapes (1 hr., 15 min.) rent for \$10, and the double lecture tapes (2 hr., 30 min.) rent for \$15. The rental fee includes prepaid return postage.

To order a tape, fill out our "I want to be in touch!" form (see page 11) and send it to us with a check or money order payable to: The CMTA, Crozer Mills Enterprise Center, 601 Upland Ave., Upland, PA 19015.

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Interesting Tidbits of Information



ITEM 1 A new information network of people and organizations providing assistance to travelers with disabilities is growing across North America and overseas as well. The three year old network is called "Travelin' Talk" and spans North America from Anchorage to Miami and Ottawa to Honolulu. The network's founder, Rick Crowder, a paralyzed veteran and columnist for two national publications for the disabled, publishes a quarterly newsletter to update members on the resources available to travelers and to share tips and stories of the ways people are helping each other.

For more information, write Travelin' Talk, P.O. Box 3534, Clarksville, Tennessee, 37043-3534 or call 1-615-552-6670.

ITEM 2 Another new organization is the CMT Belgium which was signed into law as a non-profit organization in October, 1991. The group is being led by Etienne Kinds and he can be written to at CMT Belgium, Pastoriestraat 228, 8200 St. Andries, Belgium. We offer our congratulations to this new member of the CMT family!

ITEM 3 A lawyer in a California law office unearthed this article in a legal magazine.

"A New York appeals court upheld, as part of a divorce settlement, a permanent monthly maintenance award to a wife with Charcot-Marie-Tooth Syndrome, a genetic, chronic and progressive muscular atrophy. The wife stopped working ... and was collecting Social Security disability benefits. The husband also collected annual disability benefits. The appeals court found no error in awarding permanent maintenance since the wife's condition precluded her from working or being retrained and she could not become self-supporting. The appeals court rejected the husband's claim that the award was excessive because it was more than 15% of his gross income, finding there was no set formula for fixing maintenance. Further, the lower court did not err in assessing attorney's fees against the husband..."

ITEM 4 In response to patient request, the CMTA has generated a business sized card for use by CMT patients whose hand function makes it difficult to easily open small packets of sugar, cream, ketchup, etc. The card can be handed to serving persons, and it requests them to serve such condiments in opened containers. To obtain these cards send a \$0.29 stamp and \$1.00 for each 10 cards ordered. Mail to: CMTA/CARDS, 601 Upland Ave., Upland, PA 19086.

ITEM 5 NICHCY is a free information and referral service assisting parents, educators, care-givers, and others to ensure that all children and youth with disabilities have a better opportunity to reach their fullest potential. They are authorized by Congress to provide a national source of information and technical assistance regarding the implementation of the Education of the Handicapped Act and related Federal laws. To obtain NICHCY's publication list write to: NICHCY, P.O. Box 1492, Washington, D.C. 20013-1492 or call 800-999-5599.

ITEM 6 The CMTA has been in contact with **The Myelin Project**, a non-profit group devoted to accelerating research into the remyelination of nerve cells. This project came into existence because of the personal efforts of Augusto Odone, whose son Lorenzo became afflicted with a genetic demyelinating disorder, (ALD), adrenoleukodystrophy. The Myelin Project has created a task force which includes several laboratories throughout the world specializing in myelin repair research. At this point, the research is centered around myelin regeneration in the central nervous system (brain and the spinal cord). Since CMT is a disorder of the peripheral nerves, it is not included in the research; but knowledge gained from this work could be of benefit to all demyelinating disorders. We are also in contact with a pharmaceutical company which is working on myelin regeneration. The CMTA will continue to network with these and other organizations with whom we share mutual interests. Any scientific advances will be reported in *The CMTA Report*. ✠

PATIENT PROFILES:

Rhonda Serafini



Rhonda Serafini

Rhonda Serafini is a radio disc jockey in Pittsburgh, Pennsylvania who has adapted her life to having CMT, but who refuses to allow her disorder to ruin her plans.

Rhonda first had to deal with CMT at the age of two when she was fitted for corrective shoes because she could not walk. She was actually diagnosed with CMT at the age of 13 when she was being tested before surgery for scoliosis. After surgeries, casts, and braces, she found herself with low self-esteem. To help combat this, she entered her first beauty pageant at the age of 15. She didn't even place in that first pageant, but she remembers it as being a great experience which encouraged her to enter 12 more.

She won two photogenic awards which got her interested in modeling. She did several fashion shows and catalogs and also modeled for a photographer's ad-



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.

vertising brochure. At the shoot for that brochure, the male model she was posed with was considerably taller than her 5'3" body so they placed her on three Sears catalogs. Because of her balance problems, she could barely keep herself on them. Now, because of her leg weakness, she doesn't do runway modeling anymore, but she has learned to like print work better.

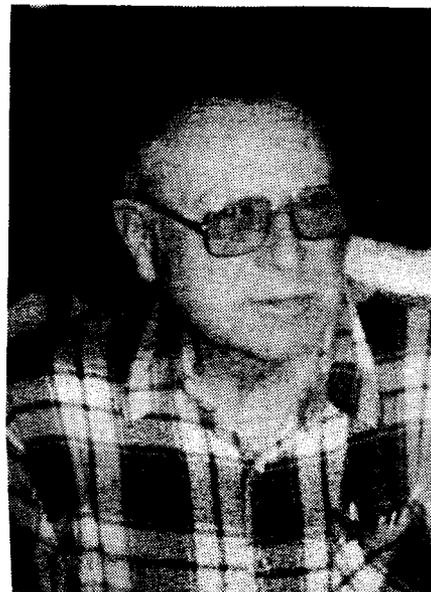
Rhonda currently works for Sunny 99, WDSN, which is an adult contemporary rock station. She only works on weekends because of her weakness, but she tries to do all the public relations work that she can, like broadcasting from local businesses. She enjoys being recognized as "Chelsey Lane."

Away from the job, Rhonda is single, has two cats, and loves to play pool, read, and watch old movies. She also works with the Special Olympics as a swim team coach and helps, when needed, at the track and field events. She also enjoys community theater work and was a ventriloquist in her last role.

When asked about any embarrassing moments in her life, Rhonda recalls one pageant that she was in. She couldn't wear heels with her pageant gowns because of her weak ankles, and because her feet are very wide, her flats were always pretty well stretched out. As she and her mother were leaving for the pageant, they walked out of their room past a group of cute guys who were eyeing her in her tight gown and whistling. She slinked past them without saying a word. They watched her from the balcony as she gracefully got into the car while giving them a sexy smile. She thought she had given a great performance. As she looked down at her feet, she realized she was missing her right shoe. She opened the car door only to find that her shoe had come off her foot as she got in the car and it had been flattened courtesy of the car's tire. Her audience had seen the whole klutzy act and had gotten a big laugh out of it. Her dramatic exit had become more of a comedy routine.

To all people dealing with CMT, Rhonda says, "Never sit back and let it take over. You have control... exercise, eat right, and keep a good sense of humor. And above all, keep faith in God. That's the best medicine." ❧

Ralph Stappenbeck College Student at 81



Ralph Stappenbeck

After receiving an associate's degree in general studies in 1985, Ralph Stappenbeck discovered an interest in international affairs which led to his enrollment at Utica College, at age 75, to pursue his bachelor's degree in international studies. Ralph, a long time resident of New Hartford, New York, credits the college faculty with opening up new thoughts for him and treating him like any other student. Ralph hopes to receive his degree in December, 1991, and said in his letter to the CMTA, "If I do not complete graduation now, I wonder if I will be able to climb school stairways next year with a crutch (or two) and my school books."

Ralph first noticed his CMT in 1970 while he was actively employed driving truck and collecting fats and bones from meat markets which had been a family occupation since 1874. He retired at age 68.

In World War II, he was a flight radio operator in military freight/passenger airplanes taking a flight physical every month. He climbed the pyramids, walked the Sahara, went to the Amazon Jungle, met Pope Pius XII, had an extensive tour of the Holy Land, visited the cave of the Holy Family in Egypt and has covered the small half of the world.

(continued on p. 4)

GEOFFREY CLARKSON A Most Unusual Man

If there had been a prize to learn who had come the farthest to the Los Angeles conference this Spring, Geoffrey Clarkson of New Zealand would have had no competition. He came to learn about CMT, and we learned about him, an interesting and delightful individual with a variety of interests and experiences. One of these interests is mountain climbing and not just any mountains, but the most awesome of all, the Himalayas.

Geoffrey sent us his account of last year's climb in the Himalayas and we were impressed that anyone would attempt such a climb. For a CMT patient to undertake such a trek is indeed very impressive. We requested and received an account of this year's climb (November, 1991) and herein are excerpts from his report of the climb up Mt. Naya Kanga (19,180 ft.) When Geff and his party arrived in Nepal they were scheduled to climb another peak, but due to many problems, including a one hundred meter ice wall located at 18,000 feet, this mountain was abandoned for Mt. Naya Kanga.

After learning of the ice wall Geoffrey's reaction was, "So we rushed to our hotel and immediately had to produce our high altitude boots so that they could be fitted with crampons...metal ice-pick type treads that get fixed to the bottoms of the boots. My high altitude boots were super deluxe, specially fitted high altitude snow and ice boots, as the disease that afflicts my legs and feet (CMT) is progressively making it harder and harder for me to operate without special aids. I had broken-in these boots over the previous six months by climbing stairs each day so that with the inner boot holding my foot and leg braces firmly in place and the outer boot with its thermal interlining keeping me warm I felt I might even manage this ice-wall. Though the thought of working my way up 100 meters of ice at 18,000 feet did not fill me with enthusiasm."

The problems of the group continued to multiply. The plane to the mountain flew on a whimsical schedule and at least half of the groups' very necessary equipment (special cold weather tents and clothing) would not fit on the plane. The greatest problem arose when they learned that, while being fitted with the crampons, their boots



Himalayan Mountain Camp

had been stolen. This was a problem for all, but for Geoffrey a far greater problem. They were able to purchase other boots, but Geoffrey's were a very poor and inadequate substitute.

Determining that the problems associated with the original climb were too great, the group then organized the climb of Mt. Naya Kanga.

This climb was eventually accomplished, but not without many physical deprivations. About the high light of the climb Geff writes, "The best was saved for the last. At almost the top of the pass we had to work our way around a massive rock that was too steep to climb up. There was a ledge for one's feet that was probably three to four inches wide. On the downside was a straight drop of several thousand feet on to rocks and ice. So one clung to the rock face with such finger holds as one could find and shuffled sideways until a proper ledge was reached. All anyone else could do was offer vocal encouragement as there was no other way and no room for any physical help to be given. And not being technical climbers we naturally did not have any ropes with us. So no one was tied to anyone else. I had handed my pack over to the guide to carry as I didn't want that weight pulling me backwards. But how our porters made it, each carrying some 100 lbs. on their back, is quite beyond me. I didn't see them do it as there wasn't a large viewing platform from which to watch such suffering. But they all did get across and collapsed at the top of the pass to moan and groan about their lot in life. It was

College Student (cont'd from p. 3)

Ralph, who has been married since 1942 to his wife Margaret, has six children and 10 grandchildren. Ralph has a strong conviction to make the world a better place in which to live. This is evidenced by his work with the Mid-York Library's Literacy Volunteers, his tutoring in the Utica School District, and his work in the soup kitchen of Grace Church.

Ralph says of his rather spectacular achievements given his age and his CMT, "I'm lucky to have inherited native abilities. I have continued to keep my mind active. Perhaps the mind drives the body. If not, what have I lost?"

too cold and windy to sit around admiring the view or our achievement so we set off on what was just as bad a trip down as we had just managed to get up. The route (no trail) was mostly straight down over endless piles of rocks with constant threat of rock slides to keep you on your toes. At least on the way down one went with the rock slides rather than have them come crashing down on top of one."

After completing this climb Geff came to the conclusion that the physical demands for this activity are now too great for him. Very reluctantly he has decided his mountaineering experiences are behind him. This writer must confess that she has never been able to understand the appeal of such an arduous physical experience. However, in concluding his account Geoffrey does articulate his reasons for mountain climbing. He says, "The big appeal of the higher altitudes is the spectacular views of massive mountains and wonders of being virtually alone with them and their power for days on end. But even with the joy that living in these mountains can bring...these high treks are now just too painful and hard for me to manage. So I will have to adjust and instead find empty beaches to roam listening to the crashing waves instead of the awesome, wondrous silence of the snow capped, Himalayan peaks."

We wish Geoffrey well with his new adventures and laud him for his spirit of striving to do all of which he is capable...and gracefully accepting that which he can not do. This is Geoffrey Clarkson, a most unusual man. ❧

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, a growing and vital part of the CMTA program, inform and support anyone who must deal with this often overlooked disorder.

There are already several CMTA support groups. These chapters are spirited and growing stronger, but more groups are needed in other parts of the United States. The CMTA will gladly help you to set up a chapter in your area. For information please contact the CMTA by mail or call 215-499-7486.

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

**California— Los Angeles area
Burbank**
Sherry Brown (818)841-8421

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) 792-1427

California— San Francisco
David Berger (415)491-4801
after 6:00 pm

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

Connecticut
Linda Friedo (203) 374-8478

Florida— Orlando Area
Mary Beeler (407) 295-6215
Carole Wray
Meets 3rd Saturday, every other month

Florida— South (Atlantic Coast)
George Meyerson (305) 431-3979

Georgia— Greater Atlanta Area,
Molly Howard (404) 333-0597
or (404) 564-9433

Georgia— Western
Molly Howard (404) 253-5632

Indiana— Indianapolis
Elaine Donhoffner (317) 841-0241
Robert Birdwell (317) 352-0235

Massachusetts— Boston,
Eunice Cohen (617) 894-9510

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

Michigan— Detroit
Suzanne Tarpinian (313) 883-1123

Missouri— Kansas City
Sandra Toland (816) 926-4213

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 York City— Manhattan
Anne Beyer (201) 391-4624

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 Salzburg (919) 967-3118
(919)286-0411 (x6586) days
Durham Medical Center

Ohio— Cleveland
Norma Markowitz (216) 247-8785

Pennsylvania— Delaware Valley
Rex Morgan, Jr. (215) 672-4169
Meetings: Holy Redeemer Hospital
Meadowbrook, PA

Pennsylvania— Pittsburgh
Garnett McDonald (412) 937-0115

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

Utah— Salt Lake City
Marlene Russell (801) 942-8642

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

support group corner board member profile



Teri & Bob Daino

This issue of the newsletter highlights a support group leader and the newest member of the Board of Directors who happen to know each other quite well. They are group leader, Teri Daino and her husband, Bob.

Teri has been a CMT support group leader in Northern New Jersey for the past two years. She meets every other month on the second Saturday at the Englewood Hospital. She enjoys the opportunity to meet and talk to those new to the CMT family, as well as see old friends. She found it very exciting to represent the NJ chapter in a support group interview segment taped with Sally Jesse Raphael during the 1990 MDA Telethon.

Some of the programs Teri has featured at her meetings have been a healing through yoga talk, a presentation by a physical therapist, a research update, a December holiday party, and therapeutic aquatics.

Some important lessons Teri has learned from her work with the support group are that it is not a good idea to schedule speakers in the month of August (poor attendance) and that fund raising is a topic most people are not anxious to address.

Away from CMT support group concerns, Teri is active in numerous PTO events and works in her school's "Green Circle" child-development program. She also contributes her time and talents to her local church and their groups.

(cont'd on page 6)



Ask the Doctor

Board of Director Profile/Support Group Corner (cont'd from p. 5)

Dear CMTA Report,

I am writing with a question for the "Ask the Doctor" section of the newsletter. I have CMT. It was first diagnosed in 1965 when I was 48 years of age as ALS. The doctors told me at that time that I had three to five years to live. After five years had passed and my condition had not deteriorated greatly, I dismissed my problem and got on with my life. About five years ago, my oldest daughter was diagnosed as having CMT and since it is hereditary, her neurologist suggested that I be tested. The tests confirmed that I had CMT.

In addition to CMT, I have been diagnosed as having Parkinson's disease. Last year I saw a neurologist who said he had treated thousands of patients with Parkinson's disease and over one hundred with CMT, but in all of his experience, he had never encountered one with both of these diseases. So, my question is, am I one of a kind? If so, how do I differentiate between the problems caused by these two diseases?

The Doctor replies:

You are not unique. I care for one other person with both of these disorders. I am not aware of a reported association between these diseases. I think it is coincidence. Parkinsonism causes slowness, tremors, and imbalance. Your neurologist would be the one to help you sort out which disease is causing which problems.

Dear Doctor,

I strongly agree that alcoholism increases the rapid deterioration of CMT. I have personal experience with this. I have been recovering for a little over 2 years, and it seems to me that once I quit using alcohol and drugs, I had rapid deterioration. I chose to go to a wheelchair one year after I became involved with AA. I could no longer stand the severe falls and breaks I was experiencing while trying to remain ambulatory. Is this normal for more rapid deterioration to occur after quitting the drugs and alcohol? Why did I not experience similar problems while I was actively using?

The Doctor replies:

Other recovering alcoholics without CMT have told me similar stories. It is probably a combination of the natural history of CMT and/or alcoholic neuropathy. You probably failed to notice the gradual decline in function as a result of the influence of alcohol and drugs. When you were not using, your ability to notice function, or loss thereof, was sharper.

Dear Doctor,

I had to have a partial hysterectomy performed about 6 months ago due to the ongoing deterioration of my hands. I got to the point where I could not take care of myself during my monthly cycle. I had frequent pain and more atrophy when I was menstruating. Is this common in female CMT patients? I had a number of gynecologists literally laugh me out of their offices when I requested surgery for this problem. I finally found a GYN who understood and would help me with this.

The Doctor answers:

The fluctuations in hormone levels during the menstrual cycle can trigger fluid retention which, in some cases, causes aching pains. I am not aware of any association, however, between the menstrual cycle and changes in nerve function.

Dear Doctor:

Under the heading "Medical Alert," alcohol is listed as one of the drugs that could be harmful to the CMT patient. I recently read that a Canadian study indicates that two drinks a day reduces the risk of heart attack by 26%. Would it be possible to get a clarification on the amount of alcohol that could be harmful to the CMT patient?

Dr. John Griffin addressed this question at the Johns Hopkins' Conference. His reply was:

"Large amounts of alcohol we ought to be careful about in someone who has CMT. An occasional drink is of no concern."



Teri lives in Upper Saddle River, New Jersey, with her two daughters, Laura and Rebecca, and with her husband Bob, who is the subject of the Board of Director's profile.

Robert Daino is the newest addition to the CMTA's Board of Directors, having been elected to that position at the October Board meeting.

Bob is a vice-president and actuary in the reinsurance business. He received his BA in math from Fordham University in 1971 and holds memberships in a number of actuarial societies.

He became interested in the work of the CMTA through his family's involvement and has assumed the Chairmanship of the newly formed Research Fundraising Committee. Bob is currently reading up on the by-laws of the organization and reviewing the past successes and failures in fundraising. He is an enthusiastic and interested Board Member who begins the first year of a three-year commitment to the Board.



New Support Group Forming in Kansas City, MO

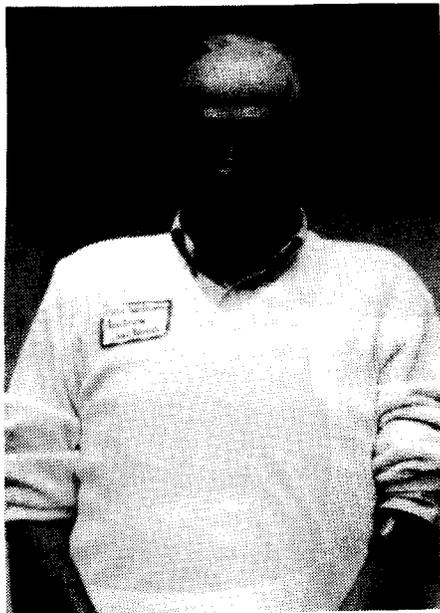
A support group for persons with CMT, or their family members, is forming in Kansas City, Missouri. The group leader will be Sandra Toland. Her phone number is 816-926-4213 and she would love to hear from you if you are interested in attending the meetings.

The first meeting is scheduled for February 1, 1992 in the Research Medical Center of Rockhurst College. The address is 2316 East Meyer Boulevard, Kansas City. The tentative schedule has the meeting lasting from 10am to 1pm on February 1. Please make contact with Sandra if you will be able to be a part of this new group. It is not easy to get a support group going, and it helps to know that the effort is appreciated.



PATIENT PROFILE:

ANDREW JACKSON – Family Historian



Andrew Jackson

Before the Johns Hopkins' conference, the office of the CMTA sent letters to all the members of the association who lived in, or around, the Washington, D.C. area, hoping that one of them would have a story which might intrigue the Washington Post to do an article on CMT and a profile of a CMT patient.

One of the replies we received was from a gentleman who had done, and was doing, some fascinating research into the genealogy of his family, especially as it reflects the incidence of CMT. The man's name is Andrew Jackson and his story follows:

"I inherited this disease from my father. My father's mother, Annie Jackson, was a member of the Mattingly family who trace their roots to the early settlers who came to St. Mary's County, Maryland, in the 1600's.

The origin of the Mattingly family is Anglo-Saxon. Numerous mention of the family's name is in English history as far back as 1086 in the Doomsday Book in London. Other English researchers have traced the name back to the latter part of the 5th Century in Britain. From what I know the family migrated in the 1600's to Maryland from the town of Mattingly, in the County of Hampshire which is 40 miles southwest of London. This town is still in existence today.

Annie's father, George Mattingly, who was born around 1842 had a condition described as "creeping paralysis" a condition that ran in the family from way back. I am certain that this condition was, in fact, Charcot-Marie-Tooth. This is further supported by research on some members of the Mattingly family done by the National Institute of Health in the summer of 1963.

My father and mother had eleven children, nine of whom are still living. Three boys in my parent's family were afflicted with CMT- one died at age 63, one is 54 and myself. None of the girls in the family have CMT, nor do their children. My 63 year old brother was reasonably mobile, with the use of a cane, at the time of his death. My 54 year old brother is still mobile and makes his living as a painter and paper-hanger. However, he has difficulty in climbing ladders and, so, chooses jobs carefully. I am the most severely afflicted. My muscles have atrophied to the point where I have very little use of my extremities and am confined to a battery powered wheelchair for mobility. I cannot effectively write with my hands. It requires special braces to hold utensils for eating. I need specially equipped bathrooms for toileting and I require assistance from an aide in dressing, food preparation, and housekeeping.

All three of the brothers had CMT in varying degrees of severity and the disease manifested itself at different times in our lives. From the standpoint of weakness, I am far worse than my brothers.

I am the only one of my parent's children who has a child with CMT. My oldest son, now 42, began to have noticeable symptoms at the age of 6. He now has difficulty walking and lifting. Around the age of 13, he had surgery to put bone blocks on his ankles to provide support. In addition, he had his Achilles' tendon lengthened in both legs."

Andrew attended the conference at Johns Hopkins and is planning to continue his research into the genealogy of the Mattingly family. As he does this, he will continue to identify and investigate those that had, or have, CMT. □

Call for Participants

Dr. Gregory Stilwell and Dr. John Sherman of Colorado Springs, CO are seeking CMT patients to participate in a Magnetic Resonance Imaging (MRI) study of calf musculature. The investigators hope to use this soft tissue imaging modality to predict two items:

1. The fatty infiltration of muscles affected by CMT to determine rehabilitation potential of those muscles.
2. The success potential of transferring tendons from one side of the leg to the other to help foot function.

To date the investigators have found the muscles affected in the calf by CMT to be quite variable and quite unlike the typical textbook presentation. If the patient has insurance, Dr. Sherman's organization, MRI of Colorado, will submit the insurance claim for the cost of the procedure. For interested participants who have no insurance, there are limited research funds available for the study. If you are interested in participating in this study, contact Dr. Stilwell at 5353 North Union, Suite 101, Colorado Springs, CO, 80918 or call 719-594-9920.

□

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, 49 Henry Ct., Hollywood, MD, 20636.

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.

RUSSIAN SURGICAL TECHNIQUE USED FOR CMT PATIENT

Editor's note: We present the following article written by a CMT patient about an alternative orthopedic surgical procedure which he had. The procedure was developed in Russia by Dr. Gavriel A. Ilizarov in the early 1980's. American doctors have gone to Russia to study Dr. Ilizarov's methods and Dr. Ilizarov has conducted seminars in the United States. The CMTA does not endorse any procedure, but presents this article for informational purposes only.

I have been a reader of your fine newsletter since I was diagnosed with CMT about two years ago at the age of 41. It has been an excellent source of information and encouragement. I am pleased to share with your readers results of an innovative and successful surgical procedure I have had...the Ilizarov procedure.

Before discussing how the Ilizarov has radically improved my situation, the following background may be of interest.

I was born with clubfeet and had numerous surgical procedures during my early teen years. That surgery, together with corrective high-top shoes with custom inserts, was effective in enabling me to enjoy a very active and productive lifestyle. I am married with two children and work as an investment banker in New York City.

In my late thirties, my condition began to steadily deteriorate. During a period of three to four years, I was forced to give up all sports activities (tennis, jogging), leisurely walks at the park or the mall were no longer enjoyable, and eventually simple every day activities like commuting to work or pushing my daughter on her swing became quite a chore. CMT patients are all too familiar with the sedentary way of life which CMT forces one to have.

I knew I had to make a dramatic effort to change things to win back a more acceptable lifestyle. In October 1990, following a sprained ankle, I had the Ilizarov. The procedure is an external fixation system (coded by the AMA as #20692) which promotes bone growth to correct deformities. In my case, the Ilizarov has had the following effects:

1. The heel of my foot has been brought down to the floor (about two inches), and my forefoot has been brought up also about two inches. These corrections have resulted in my having a

straight foot which is essentially flat on the ground vs. my previous classic club-foot position.

2. Raising the forefoot has also had the effect of straightening out my hammer toes to a large extent.

3. My foot was elongated about three inches. This compensated for the otherwise shortening of the foot which normally occurs in correcting a club-foot.

4. My ankle was fused for greater stability.

The combination of these corrections has resulted in my having what I consider to be a "new foot" which looks very normal vs. what I had previously. I can actually wear Nike sneakers and other shoes "off the shelf" instead of very expensive custom made shoes.

Most significantly, I have no pain in my new foot and am starting to enjoy a significant amount of activity again. I've started some treadmill workouts, biking, and fully expect to participate in some racket sports again in the near future. I am enjoying family outings again, and, yes, pushing my daughter on her swing without any problem. These favorable results, I must admit, are far beyond my original expectations.

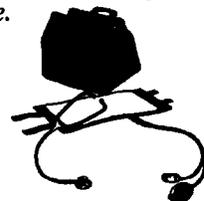
For those CMT patients who may contemplate having the Ilizarov, it is important to recognize that it takes time, and results come slowly...but they do come. In my case, the Ilizarov apparatus remained on for four months and then I was in a cast for four weeks. I have met other Ilizarov patients that required the apparatus to be on for longer and shorter periods...so it all depends on your particular case and how your body responds to the technique. During the time the Ilizarov "pins" are in place, the patient is required to "turn the pins" (the stretching effect) four times a day. No...it doesn't hurt...although intuitively, it is natural to think that one would be in constant agony. In my case, I always viewed whatever discomfort I had from the Ilizarov as being less than the pain I was having trying to walk before committing to the procedure.

I know about the adjustments CMT patients have to make in their daily lives to function as best they can; therefore, I believe in general that CMT patients will do very well with the procedure.

My surgery was performed by an orthopedic surgeon at the Hospital for Joint Diseases, 301 East 17th Street, New York, NY 10003.

– Martin Enowitz, NYC

The CMTA requested an opinion about the Ilizarov procedure from Dr. John Hsu. Dr. Hsu, a member of the CMTA's Medical Advisory Board, is the Chairman of the Department of Surgery and Chief of Orthopaedics at Rancho Los Amigos Medical Center, Downey, CA as well as a Clinical Professor in the Department of Orthopaedics at the University of Southern California School of Medicine.



Dear CMTA:

We are indeed very familiar with the Ilizarov Method of Orthopaedic Reconstruction as Professor Ilizarov has visited our institution and Dr. Stuart Green, one of our orthopaedic surgeons not only has visited Professor Ilizarov's institution on many occasions, but is also in the process of translating some of his works from Russian to English.

In Charcot-Marie-Tooth disease patients, the usual deformity of the foot is secondary to muscle imbalance. This muscle imbalance will need to be corrected by standard and time-proven orthopaedic procedures first, otherwise the abnormal muscle pull will cause the deformity to recur. Attaching the Ilizarov frame, to correct the residual deformity by lengthening the shortened soft tissues is certainly possible but generally not necessary and not without complication and problem. Those that use the Ilizarov treatment for the correction of bony and fixed soft tissue deformities find it to be a very tedious procedure. If a person has a deformity that can be corrected by other methods, for instance traditionally proven surgical methods, then, this is still the preferred way to do things. If there is any doubt or if your patient is enthusiastic about this method, he/she should seek a consultation with one of the clinics around this nation where this method is practiced upon referral of his/her physician.

– John D. Hsu, M.D., F.A.C.S.

Referrals Available From The CMTA

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 envelope indicating the geographic areas needed to: CMTA, Crozer Mills Enterprise Center, 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, phone 303-755-4588.

Honoraria

In Honor Of
Martin Edelheit
Suzanne Tarpinian

Christy Wagner
Joyce Jeronimo
Rebecca Sand
Buzz Van Almen
Rex Morgan, Jr.
Norma Goldblatt
Geoffrey Clarkson
All CMT Victims

Joni Hummel
Patti Geel
Eleene Agnew
Brian Warner

By
Ruth Edelheit
Marlene, Harvey, Lainie & Stacey Goodman
Pamela Gatfield
Helen Spicer
Beatrice Nelson
M/M R. Van Almen
Mrs. Rex Morgan, Sr.
M/M B.J. Eisenstat
Elizabeth Clarkson
Russell & Katherine Rheume
Ann Peterson
Eugene Cyran
Richeleen Snowden
Robert Warner

For your records, our address is:

The CMTA
Crozer Mills Enterprise Center
601 Upland Avenue
Upland, PA 19015
(215) 499-7486

If you are moving, please send us your change of address and enclose a mailing label from a previous CMTA Report. Thank you.

CMTA Receives Bequest

The CMTA is the recipient of a bequest granted to the Association in the will of Sophia Bradwick. This is the first specific bequest ever received by the Association. Sophia died in June, 1991 in St. Petersburg, Florida.

Mrs. Bradwick was the mother of CMTA President Karol Hitt and Association members R. Gordon Bradwick and Marcia L. Bradwick.

Memorials

In Memory Of
David K. Bernard
"Barney"

By
Wayne & Anette Mortimer
Marguerite Kitchen
Warren Jackson
Lorraine Francovich
Hal Plummer
Ralph & Irene Evans
Hope Roberts
Margot Jennings
Sadie & Joseph Jackson
Remo & Lana Fratini
Peter Visloskie Family
Thomas Trelease
Dorothy Smith
Alice Hansen
Sacramento Banjo Band
Richard & Rae Ann Schuster
Reno Emblem Club 372
Hilda Wunner
Mid-America Banjo Enthusiasts
M/M Mac Winslow
Lorraine Holland
Ida Straub
M/M Frank Schweiss
M/M Joseph Marshall
Troop G - Penna. State Police
Frederick Sutton
Memorial Lodge #57
M/M Selkowitz
R. Gordon Bradwick
Faye Bradwick
Karol Hitt
Estelle Bandler
Frederic Royston Trust
Elizabeth Oliver
Arla Van Almen
Sandra Toland
L.J. Linker
Ruth Edelheit
Morris DePouw
The Hamkinsons
Leslie Friedman

Thomas F. Murphy

Robert B. Bradwick
Sophia Bradwick
Robert & Sophia Bradwick
Joseph Goodman
Irving Finkelman
George Phy
Phyllis Keidel
Alvin Toland, Sr.
Jack Goldman
Sophie Jacobson Adler
William Harder
W. Garfield Williams
Milton Friedman

CMTA Remembrances

Your gift to the CMTA can honor a living person or the memory of a friend or loved one. Acknowledgment cards sent in honor of or in memory of will be mailed by the CMTA on your behalf. These donations are a wonderful way to keep someone's memory alive or to commemorate happy occasions like birthdays and anniversaries. They also make thoughtful thank you gifts. You can participate in the memorial and honorary gift program of the CMTA by completing the form below and mailing it with your check to:

CMTA, Crozer Mills Enterprise Center, 601 Upland Ave., Upland, PA 19015.

In honor of: (person(s) you wish to honor) _____

In memory of: (name of deceased) _____

Send acknowledgment to: Name: _____

Address: _____

Occasion:

- | | |
|--------------------------------------|------------------------------------|
| <input type="checkbox"/> Birthday | <input type="checkbox"/> Holiday |
| <input type="checkbox"/> Wedding | <input type="checkbox"/> Thank You |
| <input type="checkbox"/> Anniversary | <input type="checkbox"/> Other |
| <input type="checkbox"/> Memorial | |

Amount Enclosed \$ _____

Gift given by: Name: _____

Address: _____

Check if you would like the amount of your gift to be revealed.

Letters...



to the editor

Dear Friends,

I was delighted to receive the CMTA material. I am 72 years of age and am a CMT patient. My daughter found out about the CMTA through a listing from the United Way Campaign.

I have seen 6 generations of CMT. My grandmother, my mother, myself, my daughter, granddaughter, and a great granddaughter all have CMT. So little was known about this disease. It was heart-warming to find out there is research going on on this disease.

I was left with two children to support when I was 23 years old. All I knew to do in a small town was to work in a department store. Because I was having so much trouble with my feet and legs, I tried from one doctor to another to find some answers. I was always told, "There is nothing I can do for you."

After 60 and 70 hour work weeks, I would hurt so badly that I'd think that every day would be my last. Finally, about 24 years ago, I had surgery on my feet. My biggest problem is being able to balance. I think I do real well since I get around unaided. I do my own housework and keep my lawn mowed. I live alone.

I have seen 21 CMT patients in my family. When I had my foot surgery, the doctor told me to write a history of my family and this disease. I still have so many questions about CMT. I would like to meet Dr. Lupski. I believe the CMT gene affects the females in my family. The only CMT male is a great uncle who is deceased, and all his children were normal.

Again, let me thank you for all the work the CMTA is doing.

Sincerely,
J.P. Mississippi

To Whom It May Concern:

Receiving the most recent report I became aware of the recent research and that CMT is caused by a duplication of DNA on chromosome 17.

I would like to give a bit of a history of what has happened to me since I got married in 1960. During my pregnancy in 1961, I started to lose my balance and walk funny. I related it to my pregnancy

and thought it would disappear after the baby was born. It did not.

I went to several doctors and was diagnosed as having Frederick's Ataxia. I was told not to have any more children and that my condition was fatal. I did not obey. I had another child in 1964 and was rediagnosed as having Spastic Quadra Paresis and was told to pursue physical therapy.

I was periodically checked after that, and after a few more studies, I was told I had Spastic Para Paresis, since the upper part of my body didn't appear to be effected.

My condition is progressive and degenerative. I have the foot drop, the slow walking gait, high arches, hammer toes, along with balance problems and spasticity. I walk with 2 canes.

My father who is now 83 started to have these same problems when he was 75. Today, he is confined to a wheelchair. My mother, who is also 83, fell a lot in her youth and is pigeon-toed, but has no problem walking.

I have a brother who is younger than myself who, about 20 years ago, underwent a spinal fusion and also had orthopedic surgery to straighten his legs. He is confined to a wheelchair.

Of course, reviewing the above, I know I have a genetic disorder. The name- so far the doctors cannot determine that- or the treatment, other than physical therapy. I receive literature from the MS foundation along with your literature which helps make me aware of research.

At present I am 61 years old. It appears that my married daughter, who is 30, is mildly effected. I have been married for 31 years. My husband has been very supportive. However, as time goes on, it gets rougher for both of us. Hopefully, research will do something for me before my time on earth is finished.

Sincerely,
P.M. Pennsylvania

Dear Editor,

I saw the article in the Detroit Free Press a few weeks ago and the symptoms of CMT very much parallel the ones I am experiencing. The only symptom missing is that my hands have

not assumed the claw-like position. All the rest are present.

I have stork legs along with the numbness, burning and tiring. Chronic fatigue and weakness are also present. I have a hard time pushing doors open for myself.

I was diagnosed by one doctor as having multiple sclerosis. He said I would be in a wheelchair in 6 months. That was 3 years ago. I'm still walking. After reading the MS information, I said, no way, this was not me.

Then I was told that I do not have MS, but a pinched nerve somewhere in my spine, but it is so slight that it does not show up on the x-rays. I've now had 5 MRI's, a spinal tap, and several other tests that I cannot remember now.

I have a handicap sticker for my car, use a cane mainly because I walk like I'm drunk and I want people to know that drunkenness is not the problem. I am 52 years of age and this problem began to come on about 10 years ago.

Please send your available educational material and your newsletter.

Thank you.
J.D. Michigan

Dear Newsletter Staff,

Thank you so much for the expanded information regarding neurotoxic drugs for CMT patients. I have seen the list before, but until you listed the drugs and the conditions for which they are prescribed, I did not realize my internist had prescribed a listed drug on several occasions. How can I weigh the benefits versus the risks of the drug? I am especially interested in Flagyl, prescribed for vaginal infections.

I have made a copy of the list and will discuss it with my doctor on my next visit, requesting that he add it to my medical chart.

Editor's note: Only your doctor, knowing of the possible neurotoxic nature of these drugs, can watch your reaction to the drug and determine the benefits versus the dangers. The patient's plan to add this to her medical chart is a wise one.

V.W California

(continued on pg. 11)

Letters to the Ed. (cont'd from p. 10)

Dear Sir:

I am writing this letter for several reasons; however, first and foremost, is to thank your association for an extremely informative conference held at Johns Hopkins on November 9th. The speakers were excellent and they all presented the required information in a manner that was easily understandable and informative.

I want to request membership in the CMT Association. I believe your Association works very hard to bring its members the most current and reliable information concerning CMT. Again, I thank you, and I look forward to receiving your information.

B.C. Maryland

Dear Editor,

Your Fall 1991 issue of CMTA Report was in my mail this morning and was intriguing enough to make me play hookey from the work I should have been doing.

The report of continuing research by Dr. Lupski was particularly interesting and I would like to see the required criteria for being in his study.

James McFellan's communication on your Letters to the Editor page fascinated me because he brought up a matter which I think is of vital importance to those with CMT--- the relationship between falls and the acceleration of the disease. I know that for years I felt I was indestructible, and even when I found out I had CMT, I was sure that I did not have to take any particular precautions against falling, because falls were not important. I would just pick myself up, dust myself off, and be as good as new... just as I had always done. Right? Wrong! I found out just how wrong I was the hard way.

I am enclosing a letter for James. I hope he gets other mail on this subject. It is one I think should be impressed upon all CMT people.

Sincerely,
A.G. Pennsylvania

LETTERS
We want to hear from YOU!
Write us at:
Letters / The CMTA
601 Upland Avenue
Upland, PA 19015

I want to be in touch!

Name: _____
Address: _____
Phone Number: _____

Tell us about yourself:

- CMT Patient
- Interested Supporter
- Other
- Medical Professional
- CMT Family Member

Please check the appropriate boxes:

- Put me on the mailing list!
- CMT Neurology - \$10 rental
- Physical Therapy - \$10 rental
- Physical Therapy/Orthopedic Surgery - \$15 rental (on one tape)
- Neurology/Genetics - \$15 rental (on one tape)
- Send me a copy of the informative booklet "CMT Facts" - \$3
- CMT Genetics - \$10 rental
- Orthopedic Surgery - \$10 "

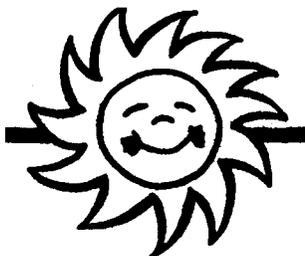
Join the fight against CMT!

Enclosed is my check for:

- \$25 Member
- \$ _____ other

Contributions are tax deductible.

Please make checks payable to the CMTA.



South Florida

CMT Patient/Family Conference

The CMTA is delighted to announce that the next regional CMT patient/family conference will be on Saturday April 25th at the University of Miami Jackson Medical Center, Miami, FL. Registration will begin at 9:15 AM. Dr. Walter Bradley, Chairman of the Department of Neurology, will begin the day with a discussion of "The Neurology of CMT." Following Dr. Bradley will be presentations by a geneticist, rehabilitation specialist, and an orthopaedic surgeon. Lunch will be included in the \$20.00 per person conference fee. Florida state residents will receive a registration form in the mail. Out of state readers who are interested in attending the conference should immediately send a stamped self-addressed envelope to the CMTA. By return mail we will send a conference registration form. Come join us and escape to the sun and sand of Florida. ☀

Attention CMT Patients

We are interested in hearing from anyone who thinks they may have been discriminated against because of their CMT in employment, insurance or housing. If you believe you've been the victim of discrimination in any of these areas, please send a brief explanation along with your name and address to:

CMTA
601 Upland Ave
Upland, PA 19015

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.

The following back issues of *The CMTA Report* are available at \$2.50 a copy:

Fall '91
Summer '91
Spring '91
Fall '90
Summer '90
Winter '90
Fall '89
Summer '89
Spring '89
Winter '89
Fall '88
Spring/Summer '88
Winter '88
Spring '87

Write the CMTA (address below)

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

CMT...

- is the most common inherited neurological disease, 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, affecting half the children in a family with one CMT parent.
- may become worse if certain neurotoxic drugs are taken.
- can vary greatly in severity, even within the same family.
- is the focus of significant genetic research, bringing us closer to answering the CMT enigma.

TO:

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