

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 © 1993, CMTA. All rights reserved.

Nutrition And Immunology Findings In CMT

*Editor's note: The following article is excerpted from a letter written to Dr. Gareth Parry, the Editor of the **PHYSICIAN'S HANDBOOK** (to be published by the CMTA in 1993), from Dr. Lowell Williams. The information is of such high interest and value to CMT patients and their physicians that we are publishing it now for all to read. Dr. Williams has conducted CMT research at Columbus Children's Hospital for many years. The article that follows is Dr. William's findings and observations from her work in nutrition and immunology in the CMT patient.*



Dr. Lowell Williams

Nutrition Background

The original work showing a fatty acid deficit in CMT blood and nerves was done by Dr. Peter Dyck (Mayo Clinic) and associates. In our study of fatty acid supplementation in Type I CMT, we first confirmed his findings of low essential linoleic acid in serum of CMT I, and then tried to bring this component back to normal by adding linoleic acid to the diet for one year, after a three month blinded placebo period. We also measured nerve conduction velocities and hand and feet muscle strength. Unfortunately, the added linoleic acid did not work biochemically nor measurably by physical tests, although many claimed to "feel" better. Therefore, dietary additions do not change the disease course during one year. However, added fatty acids (we used EFAMOL capsule, but there are cheaper generic forms) do help to improve the common CMT complaint of dry flaky skin, constipation, and may possibly benefit function over a longer period of time than one year.

Sugar metabolism is not completely normal in some CMT patients either. Many complain about fatigue and fast heart rate

after a high sugar meal even though they have a normal glucose tolerance test. Enclosed is my article on pyruvate metabolism (see list at end of article) I know some others were not able to find the defect we described, and the enzyme fault clearly does not appear to be in all CMT I. Since now we know there is a genetic difference in Type I, the PDH defect may segregate with one form. If this is a clinical complaint in the individual CMT patient, I advise a diet low in pure sugar. Of course, diabetes may be present in some CMT patients, so sugar intolerance may be pre-diabetic. If associated with weight loss and excessive drinking, a complete diabetic workup is in order.

Nutrition Advice

A weight loss program is recommended if the patient is overweight to reduce the strain on bearing muscles and joints as well as for better general health. A regular diet of moderate meals eaten every four hours (to include a late afternoon tea break) high in meat and fiber and generally low in sugar works well. I advise a

daily multiple vitamin capsule (any brand) with an option of additional fatty acids after suitable explanations. I counsel CMT patients that that they should not expect to have "the second wind" of others. Since CMT patients have lower available fats in their tissues they can not get that spurt of activity that regular athletes use and call "going beyond the pain". In CMT patients the pain means that they are burning up muscle tissue. Gentle regular exercise is the key. Exercise and when you feel pain stop. (I often write to school gym teachers about this.) Swimming and bike riding are excellent exercise, since there is less weight bearing than in running or walking. An absolute rule is to keep moving and keep active, mentally as well as physically.

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Immunology Background

Enclosed you will find several articles (see list at the end of article) on altered immunity of CMT patients. While the IgA deficiency present in a percentage of CMT has never been explained, the reproducible immunologic event is T-lymphocyte activation. However, it occurs in a cyclic form over time alternating between elevated and normal. The response of the peripheral immune cells appears to be normally generated activation as would follow stimulation from a foreign protein. The foreign protein in CMT has not been identified and is probably not normal peripheral myelin or its components, judging from the work of others. As stated, it may be either genetically altered self-proteins as in autoimmune disease, or possibly the result of a latent viral infection, organism unknown.

I have learned that Dr. Roger Lebo (Univ. of CA SF) has reported interesting data of pregnancy increasing CMT symptoms. I know personally that this is so from my six pregnancies. Also, accidents and surgical procedures tend to increase CMT disability. I too, have heard that story many times from CMT patients. It makes sense when one factors in the information of immune system stimulation in CMT. Since immune cells are already working under an increased load, the added burden of pregnancy or surgical trauma stresses available resources to a breaking point. When this happens, the underlying CMT disease, which appears to be held to a slow progression in most people by their hyperactive immunity, becomes stronger leading to increased symptoms of cramping, spasms, weakness etc. This scenario can occur in other autoimmune disease too. Therefore, our data supports an autoimmune element in CMT. Unfortunately, we do not yet know the identity of the foreign proteins in CMT. (I believe they will be the gene products of the chromosomal genes currently under investigation.)

Advice To CMT Patients

Recognize that CMT is a chronic disease that depletes and stresses immune resources. Patients may need more sleep than others, more regular hours, and less exertion than others their age. It is best to adjust to this and learn to live productively with the disorder. In addition, CMT patients may develop some chronic infection in sinuses, urinary tract, skin (especially the feet), or other areas. When this happens be sure that the infection is treated more vigorously than

it might be in others. Tell your doctor that CMT does alter your immunity. If you take care of yourself wisely, you will live a normal life span. It is important to emphasize the positive.

Journal articles by Dr. Williams:

"Expression of Schwann cell and peripheral T-cell activation epitopes in hereditary motor sensory neuropathy" L.L. Williams et al., *Journal of Immunology*, 36(1992)147-155

"Altered immunoregulation in Charcot-Marie-Tooth Disease, Type I, and its possible role in peripheral demyelination", *Charcot-Marie-Tooth Disorders: Pathophysiology, Molecular Genetics and Therapy*, pp. 231-239, 1990 Alan R. Liss, Inc.

"Activated T cell in type I Charcot-Marie-Tooth disease: evidence for immunologic heterogeneity", L.L. Williams et al., *Journal of Neuroimmunology*, 16(1987) 317-330

"HLA in Charcot-Marie-Tooth Disease", L.L. Williams, *Annals of Neurology*, vol. 8, # 4, Oct. 1980, p. 452

"Isolation of a new strain of Cytomegalovirus from Explanted Normal Skin", *J. Gen. Virol.*, 1980, 47, 519-523 (printed in Great Britain)

"Detection of Cytomegalovirus Genomes in Human Skin Fibroblasts by DNA Hybridization", *J. Gen. Virol.*, 1980, 51, 435-438 (printed in Great Britain)

"Dietary essential fatty acids, vitamin E, and Charcot-Marie-Tooth Disease", L.L. Williams et al., *Neurology*, Vol. 36, # 9, pp. 1200-1205, Sept. 1986

"Essential Fatty Acid Dietary Addition in Hereditary Neuropathy", L.L. Williams, et al., *Prog. Lipid Res.*, Vol. 25, pp. 607-610, 1986 printed in Great Britain.

"Pyruvate oxidation in Charcot-Marie-Tooth disease", L.L. Williams, *Neurology*, Vol. 29, # 11, pp. 1492-1498, Nov. 1979

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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 Hilt and Pat Dreifelber. The layout is by Chesapeake Bay Design, 1418 Fremont Ave., Redwood, CA 93555.

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

The famous green letter

In December, we sent a letter to over 2,000 people on our mailing list. These names were individuals and institutions that we had not had any communication from in at least one year. We did this mailing because we do not want to send the newsletter to households and organizations who have no interest in receiving it. Since the *CMTA Report* is sent non-profit bulk mail, the Post Office discards undeliverable copies. The Post Office will not return or forward bulk mail.

The CMTA must keep its mailing list current. To do this we need to hear from our readers at least once a year. Everyone who received a Fall 1992 "green letter" had previously been sent an annual membership renewal form on the anniversary of his/her last contribution. Some members objected to the "tone" of our letter. If we offended we apologize; however, urgency was required as we began changing our database January 1, 1993. Also, we must clear the database of deceased, disinterested, and unlocatable individuals.

Your money funds our programs and it must be spent prudently. The newsletter should be sent to all who wish to receive it and not thrown out by individuals and the Post Office. No CMT patient is denied the newsletter because of an inability to contribute, but he/she must return his/her renewal form to remain on the mailing list. The CMTA is very aware that some patients are on disability, unemployed, or otherwise unable to contribute. We will always send the newsletter to CMT patients, regardless of the ability to pay. That is one of our missions.

Unfortunately, due to a computer glitch, people whose membership month was in June as well as a few random members were accidentally placed on the list. Our instructions to the computer were to generate all names of persons from whom we have had no contact since June 1991. The computer did that, but also included persons who had contributed in June, 1992. We believe it "read" the month and not the year. Since dates do not appear on your labels, we had no way of knowing an error had been made. As previously stated, we were working with a list of slightly more than 2,000 names.

We apologize for errors and regret if we offended anyone. That was not our intent. We are working with you to advance the cause of CMT among the patient community, the medical community and the general public. §

FIGHTING THE SYSTEM FROM WITHIN

Two CMT attorneys, one a judge and the other a practicing lawyer, know first hand how complicated the legal process can be and how slowly the system can work. These legal professionals undertook the task of making their courthouses handicapped accessible.

Martin Thompson, 34, an associate judge for Municipal Court in Austin, Texas, routinely issues search warrants, orders wire taps, handles traffic cases and reviews documents for higher court cases such as felonies. What he couldn't routinely or easily do was get into his courtroom to handle these magistration cases. Now, because of Martin's efforts and those of the active disabled community in Austin, he has simple lever door-knobs which give him access to the courtroom. As he was quick to point out, however, that victory was not quickly or easily won and it is only the beginning of the many changes that need to be made to accomodate the disabled. As Martin said, "I was hopeful when they appointed



Photo by: Larry Kalkwood, Austin American-Statesman

The Honorable Martin Thompson

me that they had some sense that they needed someone with my perspective." Although the ADA (Americans with Disabilities Act) was put into effect in July 1992, people and institutions are slow to change and many simply do not know what is required of them. Martin hopes to help Austin's court buildings become accessible both to clients and to the professionals who use them.

Martin believes that the legal profession is a dream he can successfully make come true. He decided to become a lawyer after he began to lose feeling in his hands, feet and legs while attending Columbia University in New York. He allowed his mind "to do the walking" while attending law school at the University of Texas in Austin. A law school classmate of Thompson's said of him, "He didn't fault people without disabilities for not feeling his challenges. He could always present his disability in a way that an open-minded individual could understand." It is exactly this ability to convey the challenges of access for the disabled in an understandable manner that should serve Martin as he continues to work from within the system to "change the system."

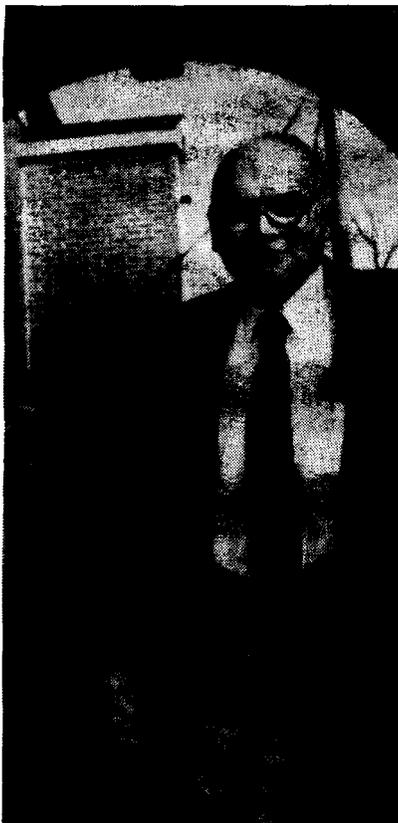
Meanwhile, half the country away in a little county in Virginia, R. Gordon Bradwick, 70, has been fighting his own battle to improve accessibility to a court house built in 1854. As a lawyer with CMT, Gordon has had to reach the court rooms which are located on the second floor of

the court house without the help of an elevator. Because the building was of historic interest, resistance to changing the exterior was fairly strong. Before Gordon's campaign, deputies had to carry wheelchair bound persons up the flights of stairs or conduct court in the clerk's small office downstairs by literally making him/her move out into the hall.

Gordon began a campaign locally and at the state level to add an elevator to the courthouse. However, the problem was not solved until a new circuit court judge took office and simply ordered the county board of supervisors to add the elevator. In addition, some lever door handles have also been added.

The battles are not over yet. Gordon is now trying to get an elevator in the office building next door to the courthouse and is urging the installation of a retriever system to provide access to the huge books which contain the deeds of trust and other records that he routinely needs. As he observed, it is not easy to do these things alone, and he has been encouraged and aided by the Council on Aging of Middlesex County where 30% of the residents are over the age of 60.

Gordon is a graduate of Syracuse University and the Syracuse University School of Law and has been practicing law for 45 years. Since Gordon is now partially retired, he recognizes that the need to bring about change is more important for others than it is for himself. §



R. Gordon Bradwick

RESEARCH UPDATE

(Editor's note: A group of CMT scientists met and conferred at the November 1992 meetings of the Human Genetics Society. The meeting was organized by Dr. Norine Stirpe, MDA, Tucson, AZ. This article was written by Dr. Kenneth Fischbeck, Univ. of PA, Philadelphia, PA. There were 20 participants and they represented five foreign countries as well as the United States. The article is rather technical, but we felt it should be published as written.)

In 1989 the most common form of inherited motor and sensory neuropathy (CMT1A) was mapped to chromosome 17. In 1991 duplication of a segment of the short arm of chromosome 17 was identified in patients with CMT1A. In early 1992 the gene for the protein PMP22, which is highly expressed in peripheral nerve and mutated in Trembler mice, was found to be contained within the CMT1A duplication and thus likely involved in the pathogenesis of the disease. At this workshop new evidence was presented confirming the role of the PMP22 gene in CMT1A, and indication of its involvement in another hereditary disorder of peripheral nerve. In addition, a mechanism was proposed that may explain the relatively high frequency of the CMT1A duplication and occurrence of de novo mutations in sporadic patients.

Chromosome 17 duplications are found in over half of all families with hereditary neuropathy, in about 70% of families with hereditary demyelinating neuropathy, and in more than 90% of families with hereditary demyelinating neuropathy genetically linked to chromosome 17. Although several patients have been found with larger, cytogenetically detectable duplications, and smaller duplications, the large majority of CMT1A patients appear to have the same duplication of an approximately 1.5 Mb chromosomal segment. The common factor in the larger duplication patients with demyelinating neuropathy is involvement of the PMP22 gene duplication, indicating that increased dosage of this gene is the critical factor in the development of the neuropathy.

The common duplicated segment has been cloned and mapped. A direct repeat of approximately 20 kb has been identified at both ends of the duplicated segment, consistent with a mechanism of duplication by unequal recombination.

One family linked to chromosome 17 but showing no duplication has been found to have a point mutation in the PMP22 gene. Interestingly, the mutation in this family produces the same amino acid substitution as has been identified in Trembler mice. The phenotype is somewhat more severe than in other CMT1A families, but fully consistent with this diagnosis. Screening of DNA samples from a large set of patients with demyelinating neuropathy but lacking the duplication led to the identification a new-onset CMT1A family with a de novo point mutation in the PMP22 gene. These findings indicate that CMT1A can be produced by either specific alteration or increased dosage of PMP22.

Hereditary neuropathy with liability to pressure palsies (tomaculous neuropathy, HNPP) is an inherited disorder of peripheral nerve that is both clinically and pathologically distinct from CMT. Surprisingly, HNPP has now been found to be associated with a 1.5 Mb deletion of the CMT1A duplication segment in three families. The HNPP deletion appears to be the reciprocal recombination product of the CMT1A duplication and therefore, may result from the same mechanism of unequal recombination. This finding also indicates that deletion of the PMP22 gene, like duplication and point mutation, can cause damage to peripheral nerve, albeit with different clinical and pathological manifestations.

Various methods are available for detection of the CMT1A duplication, including polymerase chain reaction, routine Southern blot, pulsed-field gel electrophoresis, and fluorescence in situ hybridization of the interphase nuclei. Prenatal diagnosis of CMT1A based on duplication detection was described. Although diagnostic testing is currently available on an investigational basis, several workshop participants expressed concern that such testing be based on more than one technique, that it be accompanied by appropriate neurological evaluation, and that further clinical assessment be done before it is made generally available. §

Editor's note: There is a 22 item bibliography that accompanies this report. If you would like a copy of the bibliography, please send \$1.00 and a stamped self-addressed envelope to the CMTA requesting it.



Dr. Kenneth Fischbeck

Editor's note: This letter was written to Karol Hitt, CMTA President in January 1993 from Thomas B Bird, MD, Chief Neurology Section, VA Medical Center, Professor Neurology and Medical Genetics, Univ. of WA Medical Center.

Dear Karol,

For your information, I have enclosed a preprint of a paper related to CMT that will appear in *CMT* this month. The senior author is Dr. Philip Chance at the Univ. of Utah along with myself and several colleagues from the Univ. of WA. The important new finding in several families is the reciprocal of the previously known chromosome 17 duplication in CMT. These families have hereditary liability to pressure palsies and have a deletion, instead of a duplication, of the same DNA on chromosome 17 that is involved in CMT. The family members are not typical of classic CMT, in that they do not have distal weakness and atrophy and generally slow nerve conduction velocity. However, they are especially sensitive to modest pressure on their peripheral nerves, and are likely to develop recurrent peroneal palsy with foot-drop, ulnar nerve palsy at the elbow and carpal tunnel syndrome. This finding is additional evidence of the importance of the peripheral myelin protein gene on 17p in the pathogenesis of hereditary nerve disorders.

Best Wishes,

Thomas B. Bird

RESEARCH UPDATE

Pilot Prenatal CMT Test Available

Dr. Roger Lebo, University of California, San Francisco, is doing a pilot prenatal diagnostic study to detect the presence of the CMT1A gene. If the CMT1A duplication on chromosome 17 is found in an affected relative, then the fetus can be tested for CMT. Thus far, this test has diagnosed five of five fetuses using samples obtained routinely by both amniocentesis and chorionic villus sampling. Dr. Lebo is using *in situ* hybridization to provide the most rapid result using cells from the placenta or sloughed off by the fetal skin. Unlike tests that rely on DNA polymorphisms (normal differences like blue or brown eyes), this test detects the DNA change that causes CMT in every fetus arising from a parent with the duplication on chromosome 17. Results are confirmed by a more time consuming polymorphic test when the *in situ* hybridization test is positive. Dr. Lebo's laboratory is also testing CMT family members who are at risk for CMT. In another pilot study, samples of 50 adult CMT patients from 50 unrelated families gave definitive results. For information about these tests contact Dr. Lebo at Room U-253, University of California, 533 Parnassus Avenue, San Francisco, CA 94143-0720 or call him at 415-476-5481.

Consider these excerpts from *Life's Little Instruction Book* by H. J. Brown, Jr.

- Be brave. Even if you're not, pretend to be. No one can tell the difference.
- Pray not for things, but for wisdom and courage.
- Be willing to lose the battle in order to win the war.
- Never take action when angry.
- Never deprive someone of hope; it may be all he or she has.
- Instead of using the word *problem*, try substituting the word *opportunity*.
- When faced with a serious health problem, get at least three medical opinions.
- Choose a charity and support it generously with your time and money.
- Compliment three people each day.
- When facing a difficult task, act as though it is impossible to fail. If you're going after Moby Dick, take along the tartar sauce.

CMT Patients Needed for Drug Study At Mayo Clinic

Dr. Anthony J. Windebank, a neurologist at the Mayo Clinic Peripheral Nerve Center, is supervising a study of a drug therapy for type I CMT. The drug being studied is Dynamine (3,4 di-aminopyridine). Dynamine has been used for a period of years to successfully treat patients with Lambert-Eaton myasthenic syndrome, a rare form of myasthenia gravis.

Myasthenia gravis is a totally different disorder from CMT, but for theoretical reasons, the investigators believe that Dynamine might be a therapy for type I CMT. It is known that Dynamine blocks potassium channels in demyelinated nerve fibers. If the potassium channels are blocked, there is an improvement in the conduction of impulses along the nerve fiber. Hence, the muscle would get a greater stimulus from the nerve and be able to function better.

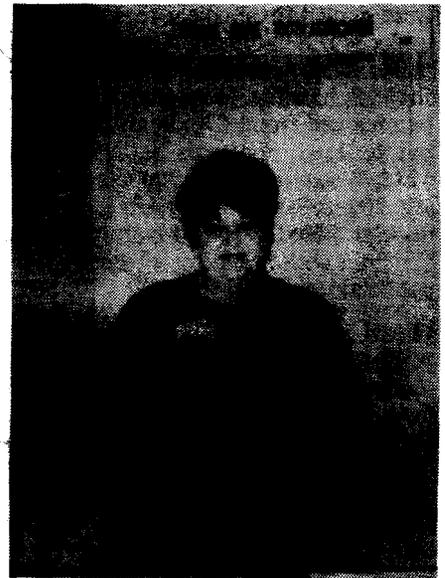
Dr. Windebank is currently conducting clinical trials of Dynamine at the Mayo Clinic, Rochester, MN. To be eligible for the study, you must be at least 18 years of age, with type I CMT. The investigators are particularly keen on recruiting female subjects. The study involves staying at the Mayo Clinic for about two weeks.

Upon arrival at the Clinic, a complete neurological evaluation is done, including an EMG. The patient is then put on either Dynamine or a placebo (a sugar pill with no effect on the body) for five days. Neurological measurements are again done and then the patient receives no medication for five days. After this, the patient is placed on the other medication for five days. The final neurological measurements are done after the second five day drug course. This results in the participant receiving the placebo and Dynamine. If the Dynamine has been effective while taken by the patient, the patient can receive a supply of it to continue the therapy at home. While at the Mayo Clinic, patients are admitted to the Clinical Research Center. All costs for the study are paid for by the research project except for the participant's travel costs. You must be able to get to and from Rochester, MN on your own.

Particularly, if you are a woman, 18 years or older with type I CMT, and are interested in participating in this study, call Dr. Anthony Windebank at 507-284-4349 or write to: Dr. Windebank, Mayo Clinic, Rochester, MN 55905. §

MEETINGS, MEETINGS, MEETINGS

During the Fall, CMTA President Karol Hitt represented the CMTA at three meetings. In Toronto during October the organization had a booth at the annual meetings of the American Neurological Association. Later in October a similar booth was maintained at the Prescription Footwear Association meetings in Florida. Carole Wray, a member of the Orlando support group, also worked in our booth as a CMTA representative. At both of those meetings our literature was distributed to the program participants who requested it. From the PFA meetings we are compiling a list of footwear manufacturers and retail stores who specialize in footwear for the problem foot. This list can be ordered by sending in the order blank located on page 11.



Carole Wray

The third meeting was in December at Cold Springs Harbor, NY. This conference was a seminar for lay people who interact with the scientific community and the public. It was three days of intensive genetic studies including laboratory work. The conference was funded by a government grant and was very intense and very informative. The CMTA felt privileged to be invited to the meetings. The other attendees represented a variety of organizations and occupations, but all had a common interest in genetic disorders. The ethics of genetic testing were discussed at length, and the CMTA will feature articles about this in later issues. §



ITEM 1: More than half of all children with disabilities in families with limited income who should get federal Supplemental Security Income

(SSI) benefits are not receiving them. Children who are eligible get monthly checks and, in most states, free health care through Medicaid. The Children's SSI Campaign is a national outreach effort to enroll more children. Families across the country need to know that new SSI eligibility rules allow more children to qualify than ever before. It is especially important to find 452,000 children who applied between 1980 and 1990 and were denied for medical reasons. Now, under a U.S. Supreme Court decision, called *Zebley*, these children can apply for back benefits. More information can be obtained from: Children's SSI Campaign, 1101 Fifteenth St, NW, Suite 1212, Washington, DC 20005 or by calling: 202-467-5730. They offer a 12 page booklet called "New Opportunities for Children with Disabilities" among their publications.

ITEM 2: A book entitled, HEREDITY AND YOUR FAMILY'S HEALTH by Aubrey Milunsky, M.D. is available from The Johns Hopkins University Press, 701 West 40th St., Suite 275, Baltimore, MD 21211-2190 or call 1-800-537-5487. The cost of the book is \$ 18.95 and postage and handling is \$ 2.00. The book is described as the most comprehensive, compassionate, and informed guide for all concerned about the risks of inherited disease. (Editor's note: We have not seen this book to evaluate its readability for the lay public, but it comes highly recommended by several physicians.)

ITEM 3: JC Penney offers a catalogue called Easy Dressing Fashions with velcro brand fasteners that make dressing a breeze. The catalogue also includes name-brand favorites in no-hassle designs. A very wide range of sizes is offered in most of the clothing and shoes. To receive a catalogue, call toll free 1-800-222-6161.

ITEM 4: From the Progressive Neuromuscular Diseases Newsletter of the University of California, Davis, comes this review of fatigue and weakness in neuromuscular diseases. "Used in

the global sense of the word, weakness is a spectrum ranging from a complaint of fatigue through frank outright paralysis. Like pain, muscle fatigue is a subjective symptom, while weakness is a measurable observation. From a practical standpoint, fatigue can be thought of as short lasting weakness. In successfully treated myasthenia gravis, for example, there is rarely any muscle wasting or longlasting loss of strength. Fatigue occurs following prolonged muscle contractions, with a relatively rapid return of strength following rest. From an experimental standpoint, muscle fatigue is a gradual linear reduction of muscle force (strength) generating capacity. Complete muscle exhaustion occurs when the target force can no longer be maintained at all. At this point, the muscle fibers are depleted of their main fuel, glycogen. The causes of muscle fatigue remain unknown although the primary problem appears to be a breakdown in calcium metabolism....many clinicians now feel that fatigue, as a physical impairment, may be more of a disability than actual weakness in muscle wasting neuromuscular diseases."

ITEM 5: In August, 1992, Cephalon announced initiation of its clinical trials in the U.S. for Myotrophin in ALS patients. In September, they presented the results of the UK Phase I clinical trial for Myotrophin, the company's lead compound for ALS and peripheral neuropathy, including CMT.

ITEM 6: As of July 26, 1992, 43 million Americans with disabilities (approximately one out of every six people in the US) have new federal legal protection against employment discrimination under the Americans with Disabilities Act of 1990 (ADA). Considered the most sweeping antidiscrimination law since the Civil Rights Act of 1964, the employment provisions of the ADA mean that people who once were excluded from the job market or faced limited opportunities for job growth because of disabilities are now able to compete in that market on an equal basis with other workers. Careers expert Melanie Astaire Witt has written a new book entitled, Job Strategies for People with Disabilities: Enable Yourself for Today's Job Market. The book contains advice on finding your first job as well as helping current disabled workers to move up their career ladders. The book is published by Peterson's and is \$14.95 in paperback and should be available at any bookstore or by calling 1-800-338-3282 ext 225 for information on a store near you.

ITEM 7: A book entitled, Health Insurance: How to Get It, Keep It, or Improve What You've Got by Robert Enteen, Ph.D. is available for \$12.95 from Demos Publications, (212) 683-0072. The chief health insurance authority of the National MS Society offers valuable information and sound advice to the insured, uninsured and underinsured. He clearly explains how to evaluate coverage, compare costs, select providers, and supplement existing plans to obtain maximum protection at the lowest price. Also available from Demos Publications is a newsletter called Disability Rights Report which will be published bi-monthly and will keep people with disabilities informed about taxes and financial planning, insurance, and legislation. The first several issues promise articles on filing for SSDI, the impact of ADA, a review of health insurance options and alternatives and reports on state and federal legislation that affect people with disabilities. The newsletter costs \$19.95 for a one year subscription. Call Demos for information 212/683-0072.

ITEM 8: A new movie has recently been released entitled, "Lorenzo's Oil." It details the personal efforts of Augusto Odone, whose son Lorenzo is afflicted with the genetic demyelinating disorder adrenoleukodystrophy (ALD). Mr. Odone personally conducted the search for the oil which is being used to treat his son, Lorenzo. Hence, the product is now called Lorenzo's oil and is used by some families to treat ADL. Mr. Odone has also founded a task force called The Myelin Project 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 spinal cord). CMT is not included in this research, but knowledge gained from this work could be of benefit to all demyelinating disorders.

We are in contact with the Project as well as with a pharmaceutical company working on myelin regeneration. Scientific advances will be reported in this newsletter.

ITEM 9: In January 1993, a group of peripheral neurologists met at a "Think Tank" session sponsored by Regeneron Pharmaceuticals in New York. Among the things discussed were drug therapies for hereditary neuropathies including CMT. We are in contact with this company and will keep you informed as progress is made.

§

Letters To The Editor



Dear CMTA Reporters,
Keep up the good work! I have two sources for the CMT foot with regard to shoes.

1. Hitchcock Shoes, INC
225 Beal St
Hingham, MA 02043

These people sell special lasts for extra wide feet and high insteps. The shoes are marginal for hammer toes, but the company is very cooperative and I have found an extra E allows comfort.

2. J.W. Mason (I have no address) sells extra depth and width shoes through orthopedic supply houses. My present shoes come from: Fourroux Shoes and Orthopedics, Inc, 410 Governors Drive, Huntsville, AL 35801. This is a retail outlet. A cooperative outlet might furnish you with the manufacturer's address. The shoes are wide to size and deep enough for a custom insert and hammer toes. Both sources furnish normal looking shoes.

H.N. Huntsville, AL

Dear CMTA,

As many with CMT know, writing can become a difficult and painful task. I found this out when I was 17 years old, three years after I was diagnosed with CMT. I was told by my doctor to cut down on the writing and was given some anti-inflammatory medication. Cutting down on writing when you are a student is tough. I did my best through my last year of high school, but I was still having

problems. Since I was about to start college, something had to be done. My physical therapist suggested getting a computer and possibly getting my doctor to write a prescription for the computer so the insurance would pay for most of it. I talked to my doctor and he said he would write the prescription. It took awhile to convince my dad's insurance that I did need the computer for school, but they finally agreed.

I knew that the computer wouldn't answer all my problems because typing, too, can cause problems. My physical therapist came to my rescue once again. She showed me a new program that her office just got for handicapped people. This program was great. If you type one or two letters of a word, the computer will try to guess what word you are typing. There is a box in the corner that lists six words to choose from. If one of those words is the one you are typing, then all you have to do is hit the number and the word will be completed. After playing with this program a few times, I found that it was a great help. This program is called KEYWHIZ and was developed by Words + out of California. I thought that this program would be easy to get and wouldn't cost much. I was dead wrong. This program costs between \$700 and \$800. My therapist said that she would look into finding some way to get the program. Finally we found the Variety Club, which is a non-profit organization that helps families with handicapped children 18 and under. After filing an application with Variety Club, they

agreed to buy the KEYWHIZ program for me.

I am now 21 years old and a junior in college. I use my computer and my program at least four times a week. I am grateful to both Variety Club and my high school physical therapist.

Words +, INC is located at P.O. Box 1229, Lancaster, CA 93584. Their toll free number is 800-869-8521.

S.T. Kansas City, MO

Dear CMTA,

The information you print in The CMTA Report has been invaluable for me.

I have CMT. My condition has begun to deteriorate this last year and I was desperately seeking knowledgeable medical advice. Then I received the spring 1992 issue of The CMTA Report in which you listed the names of the members of the medical advisory board. Scanning the list, I came across the names of Drs. Lovelace and Myers at Columbia Presbyterian Medical Center in New York City.

I called the MDA clinic at Columbia and was given an appointment with Dr. Lovelace. The examination, evaluation, and therapy I received from Dr. Lovelace, Dr. Myers and their team of physical therapist, occupational therapist and orthotist was unparalleled. Not only is this team professional, knowledgeable, and interested, they are sensitive to the special emotional concerns of the CMT patient.

Today, with the aid of a very inconspicuous AFO, I have more energy to walk, to travel, and to shop. I have truly been given a new lease on life.

It is with heartfelt appreciation and gratitude that I send this gift to the CMTA in honor of Dr. Robert Lovelace, Dr. Stanley Myers and their team at the MDA clinic at Columbia Presbyterian Hospital.

D.S. New York, NY

Dear Editor,

I have a nephew, age 22, who has recently been diagnosed and confirmed as having CMT. His is an unusual case in that we can't find any heredity factor involved. As far back as we can check, there have been no known relatives with CMT. The doctors, after investigation,

(Continued on page 8)



The big problem with your illness, Mr. Hawkins, is that nobody famous has caught it yet.

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MEDICAL ALERT



Certain Drugs Toxic to the Peripheral Nervous System

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

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

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

Referrals Available

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

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

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

Letters... continued

have come up with the decision that he is obviously a "mutant." They were not very diplomatic, but that's another matter.

My purpose in writing is to commend you and your staff for publishing a professional, yet personal magazine. Your list of medicines, which show they are toxic to the peripheral nervous system, is just one example.

I found your entire magazine to be informative and uplifting to a person with CMT. I have epilepsy and receive their magazines and reports, and have found them to be depressing. They have never published a list of medicines that I have seen which are good, bad, or whatever. After I read your current issue, I couldn't help but see the difference.

You can call me an "interested supporter," and I would like to continue receiving your reports. They have helped us find a pedorthist who fit my nephew with supports that have made a tremendous difference in the way he walks.

So, keep up the good work, accept my check and thanks for a job well done. Think positive, that's what I keep telling my nephew, and your Reports confirm this thinking.

R.S. Oak Park, IL

Dear CMTA,

My name is Karen and I met my husband Kevin on September 4, 1991 in Kingman, Arizona. We fell in love immediately. After he and I started our life together, Kevin started playing softball for our church and injured his foot. I made him check it out and while I was at his foot doctor's I asked the doctor why he had scars all over his feet. The doctor then told me about his CMT problem. I had

never heard of CMT before. But, with your literature and your newsletter, I feel a lot better now that I understand CMT. I feel that I can help my husband cope with his body and the changes that he feels.

We have had a beautiful little girl and I hope she is not affected. But, thanks to all of you dedicated people, I am prepared in case she is. My only regret is that we are on Welfare and cannot afford to be members. Thank you for all your help and for relieving the worry.

K.F. AZ

Dear Editor,

I am at that stage of CMT where I appear "normal" but absolutely must have the right shoes or I can hardly walk at all. Since I work full time in a professional position, I also need a flat, stable shoe which looks nice but which will stay on my feet. My closet is full of shoes which I cannot wear. I also tried the strap-on-the-pump route but that did not work well for me either.

I am happy to share my find with those who are in the same boat... and that is the Jane shoe from MASSEY'S catalogue which comes in wide width, with strap, with flat broad based heels, etc. They really are perfect at this stage of my life. I still use the old shoe stretcher to accommodate my corn and hammer toes, but they still look very nice.

The very best part is the incredibly low price and the range of colors. If you can publish the picture and the toll free number (1-800-462-7739), I am quite certain Massey will have a hard time keeping them in stock. Thank you for the opportunity to share.

D.C. Baltimore, MD

Jane 1/2 inch Heel \$34.95

The "Mary Jane" is back. Leather uppers feature an instep strap with an adjustable slide buckle for sure fit. Cushioned insoles and trust linings with a poly urea bottom. U.S.A.

52286 Purple
52285 Black
52281 Bone
52251 Blue
52248 Pink
52247 Camel
52236 Khaki
52203 Wine

Order Toll Free
1-800-462-7739



HUNTING AND FISHING? of course!



By Jim Hank

Editor's note: The following article is reprinted with permission of the author and the ALS Society in whose newsletter, Link, it appeared.

Hunting and fishing are possible with disabilities. I know because I'm doing it. I was diagnosed with ALS in August of 1987. The first thing myself and three crazy friends of mine did, upon learning I had the disease, was to go elk hunting in New Mexico. (I've been hunting and fishing ever since.) I couldn't walk on the rough terrain, but I managed the trip to the outhouse like everyone else! My arms weren't strong enough to hold my rifle steady unassisted, but with a little ingenuity, nearly anything is possible. I feel the need to tell you exactly HOW I've been fishing and hunting because I'm afraid that most disabled people have given up on these sports.

With me, the ALS has progressed to the point where there is little I can physically do, other than control the joystick of my electric wheelchair. I have a G-tube and my voice is highly unintelligible. I can't walk and my legs aren't even strong enough to support myself. I don't even have enough strength to raise my arms up. I can usually turn my one hand over, but that's about it. My limbs are nearly worthless to me, and I have a "halo" on my wheelchair that enables me to hold my head up. But the thing to remember is that it is still possible to hunt and fish with this kind of involvement.

My first summer of fishing with ALS, I was able to grasp onto a fishing rod with few problems. My arms were sore at the end of a day, but then that was nothing new! Cranking the handle of a reel was a minor problem, and casting was nearly impossible. To combat the casting problem, we did two things. First, I purchased what is called Van's EZ Cast, a device specially tailored for the handicapped. It straps to the arm of a wheelchair, and allows someone with limited strength to cast a bait. The EZ cast works well for panfish and walleye types of fishing. Secondly, we changed our fishing tactics. We began to troll more and cast less.

As the disease has progressed, we've changed to trolling exclusively, and I've purchased some electric fishing reels. The reels have worked without a hitch. We mounted a standard rod holder to my

chair. This year I'm not able to hold the pole, so we've changed our tactics once again. Now when I get a hit, a friend places the rod between my legs, and places my hands around the rod, but then he puts a hand around mine. That way I can still feel the fish fighting, and I still have the thrill of being in control of the reel.

You can fish from a pontoon boat the easiest. All you need is a piece of plywood to get your wheelchair in the boat, if you are using a wheelchair. Otherwise, all you need are a bunch of strong friends to lift you into nearly any boat.

The way I hunted that first year with ALS was to "still hunt". I'd hide behind a rock or a tree or perhaps just out in the open under a pile of camouflage material. I used what are called "buffalo sticks" to steady the front end of my rifle. Buffalo sticks are simply a couple of sticks tied together about 4 inches from one end. You simply spread them open a bit and place your gun in the "V" formed by the stick ends. You then hold both the "V" and your rifle as you normally would. The sticks simply act as a bipod. Or, you can simply buy a commercially available bipod.

The past three years, however, I've been using an electric rifle rest made by Bob Bowen of Chadron, Nebraska. The SR-77 rifle rest is a wonderful contraption as it comes directly from Bob, but we've customized mine to better accommodate my special needs. The rest comes with a sip n' puff switch to pull the trigger. We changed this to accept any one of the micro-switches from my computer.

The rifle rest also came with a homemade joystick which I found difficult to use, although others might find it adequate. We modified it with a joystick made by KY Enterprises. It's to the point now that it's easier to operate than my wheelchair! Maybe that's why I enjoy shooting so much?!

We've even made a bracket to put the rifle rest in the window of a 4 wheel drive. That way I can hunt from a vehicle if hunting from a makeshift blind is out of the question. Every state I've hunted in has a special handicapped program that makes such a thing legal.

If you or someone you know used to hunt and fish, you need to know that it's still possible. Other patients will undoubt-

edly have special needs in terms of modifications to products, but it's important to know that the "basics" are out there. With a little creative thinking, I believe almost anyone can participate in these sports.

You might wonder how successful I've been. In the five years I've been fishing and hunting with ALS, I've caught over 20 muskies, and countless northern pike and walleye. In the field, 2 mule deer and 2 antelope are mine, as well as 2 turkeys, 2 whitetail deer, and one elk. I've got an elk hunt planned for New Mexico this year. With a little luck, I'll be hanging a trophy over my fireplace when I return!

The following companies make it possible for the disabled to hunt and fish:

Mr. Bob Bowen
SR-77 Enterprises, INC
363 Maple St
Chadron, NE 69337
308-432-2894

Electric Fishing Reel Systems
PO Box 20411
Greensboro, NC 27420
800-654-7168

Ken Yankalevitz
KY Enterprises
3039 E 2nd St
Long Beach, CA 90803
213-433-5244

The following companies offer ready-made outdoor and recreational products to assist disabled people. A couple of them sell Van's EZ Cast.

Access to Recreation, Inc.
2509 E Thousand Oaks Blvd
Suite 430
Thousand Oaks, CA 91362
800-634-4351

IDEA Innovator of Disability
Equipment and Adaptations
1393 Meadowcreek Drive, Suite 2
Pewaukee, WI 53072
800-728-1587

J.L. Pachner, LTD
33012 Lighthouse Ct.
San Juan Capistrano, CA 92675
714-661-2132

I hope my story encourages more disabled to get out and continue doing what they've always done. If anyone needs more information, please contact me through the ALS Association (818-340-7500). I'd be more than happy to assist other disabled people with ideas or advice, enabling them to get out and continue fishing and hunting. §

British CMT Leaders Visit Florida Support Group



While corresponding with British CMT organization leaders about newsletters and conferences, we at the CMTA office became aware of the plans of Don and Margaret Read to visit Florida in the fall of 1992. Currently Don Read chairs the British group and Margaret is the Secretary. We connected Mary Beeler, then leader of the Orlando support group, with the Reads, and they corresponded about the visit. Here is an excerpt from Mary's letter telling us about the actual visit.

"Let me tell you about the Reads. Margaret and Don live in South Wales, UK. We have been corresponding for about a year. Margaret has CMT. We met through CMTA. One hobby of the Reads is bird watching. This was their first trip to the US and one of the things they wanted to do, besides bird watching, was to meet other people with CMT. They are

very involved with CMT - U.K. and had just worked on a convention which was in England in August.

The best group of people with CMT that I could think of was the group of friends I have made in our local support group. So, I decided, why not have a covered dish luncheon. Good food and good fun was enjoyed by all. A total of 25 people attended. We immediately fell in love with the Reads and they are now a part of our CMT extended family. We all shared some of our experiences of living with CMT, or of being in a family of a person with CMT. We found that so many of our life experiences, whether here or in Wales, are very humorous. In my opinion, I believe that people with CMT and their families are some of the greatest people in the world." §

CMTA Report Delivery News

An on-going problem here at the CMTA is the delivery of the newsletter. We learn of this problem from you, our loyal readers, when you do not receive an issue. Sometimes the solution is a simple address correction, but this is not always the case.

The newsletter mailing list is generated from our computer and given to a professional mailing firm. The newsletter is then labeled and mailed to our U.S. readers. If your name and address is correct on your label, there is nothing further we can do to assure its delivery. The next step is to check with your local postmaster to see if the problem is there. This is a great source of frustration to us, and we welcome your suggestions for a solution. It is inevitable that some newsletters will be lost in the mail. We mailed over 4,500 newsletters in Fall, 1992 and we have no way of knowing what percentage was not delivered.

One answer to this problem would be to mail the newsletter first-class mail, but this would more than double the postage costs. There is no easy answer. If you do not receive your *CMTA REPORT* in what you consider to be a timely manner, contact the office and we will certainly mail you another issue.

We apologize for this inconvenience, but please know we are as frustrated as you. §

Honoraria

In Honor Of
 Karol Hitt
 Joyce Ieronimo
 Christopher Lynch
 Her mother
 Ruth Linker
 Debra Czarnecki
 Clyde Goodrich
 Rex Morgan, Jr.
 Pat Dreibelbis
 Joni Hummel
 Karl Henion II
 Geoffrey Clarkson
 Sunny & Stuart Feen
 Betty Lou Smith
 Christopher Lynch
 Gary Borg
 Margaret DeStefano
 Mrs. LW Heinrich
 Peter Heinrich
 Jason Everhart
 M/M Henry Eldredge
 Flora Jones
 Judy & John Grocott
 Craig Haynie
 Relio Thibodeau
 Brenda Covini
 Sandra Pitts
 Linda Baraz
 Brett Nowell

By
 Laura Hitt McCann
 Helen Spicer
 Charles & Helen Lynch
 Ellen Wall
 Doris Goldman
 Carol Fink
 Joyce Rowe
 Leulla Morgan
 Martha Yohe
 Ann Peterson
 Dorothy Ferenbaugh
 Elizabeth Clarkson
 Ruth Feen
 Drucilla Lake
 Charles Lynch
 Monica Borg
 Joseph DeStephano
 William & Linda Melville
 William & Linda Melville
 Mrs. E.A. Jumper
 Douglas Moody
 Mrs. Dwight Waddell
 Robert Hawkes
 Jeanell Haynie
 Emily Nuzzo
 Emily Nuzzo
 Alice Van Huss
 Felice Viggers
 Clyde Goodrich

Memorials

In Memory Of
 Ralph Binford
 James Hitt
 Etta Hitt
 Sara Mills
 Martin Edelheit
 Irving Finkelman
 Betty Gomez
 Morris Reed

By
 Marilyn Dodge
 Laura Hitt McCann
 Laura Hitt McCann
 Mary Walton
 Ruth Edelheit
 Ruth Finkelman
 Kay Flynn
 Cathy Howie
 Thomas Smith
 M/M Jerry Bodenbaugh
 Ross & Glenna Elkins
 Jim & Dana Axmacher
 Denlys of Boston
 Marvin Greenwald
 Joyce Rowe
 Marcia Bradwick
 Elizabeth Oliver
 Dr. Catherine Yack
 Don & Mary Rehm
 Margaret Smith
 R. Gordon Bradwick
 Karol Hitt
 Lawrence Wechsler
 Leslie Friedman
 Mae DiMaio
 Michael Schwartz
 Vincent Farin
 Nancy Finkelman
 Corinne Faschan
 Irene Minter
 Monica Borg
 Jeraldine Taylor
 Fannie Ostrov
 Rebecca Brezel
 Faye Bradwick
 Jean Waldron
 Charles Granieri
 Irving & Beth Bank
 Gary Oetze
 Sarah Blacher Cohen
 Phyllis Warren
 Marilyn Laurence
 Dr. Rebecca Stafford
 Richard Cornforth
 Dr. Karen Edelson
 Ruth & Harry Linker
 David Goldman
 Betty Johnson
 Karen Gabel
 Mrs. Flora Motta
 Lana Wright
 Mildred Oliver
 David Bell
 George Crohn
 Don & Marilyn Conlan
 M/M Michael Schwartz
 Taryn Aldridge
 Drucilla Lake
 Genevieve Minter
 Mary Lane

Millie Squire
 Willie Goodrich
 Sophia Bradwick
 George Phy
 Cynthia Martin
 Betty Gomez
 Roland Smith
 Robert & Sophia
 Bradwick
 Ruth Hochberg
 Milton Friedman
 William Weniger
 Mollye Schwartz
 Lillian Fleming
 Irving Finkelman
 Howard Faschan
 Curtis Ashworth
 Robert Borg
 Billy Wheat
 Lester Ostrov
 Harry Brezel
 Sophia Bradwick
 Herbert Bates
 Monica Granieri
 Mark Albrecht
 Rex Cole, Jr.
 David Blacher
 Mildred Winchell
 Hardin Stults
 Lois Terrell Haines
 Richard Cornforth(Dad)
 Harry Brezel
 Jack Goldman
 Jack Goldman
 Betty Sue Farr
 Andrew Galomb
 Edward Scarsella
 Adelia Wright
 Fred Smith
 Evelyn Bell
 Eleanor Crohn Lowy
 Wayne Hamilton
 Mollye Schwartz
 Doris Adams
 Leo Breeden
 Curtis Ashworth
 Nick & Rose DiRitto

Support Group Notes

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

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

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

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

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

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

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

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

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

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

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

Florida - South
Robyn Cohen (407) 622-5829

Massachusetts - Boston
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) 756-2020

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

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

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

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

Ohio - Cleveland
Norma Markowitz (216) 247-8785

Pennsylvania - Delaware Valley
Rex Morgan, Jr. (215) 672-4169

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

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

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

Virginia - Richmond Area
Dennis Breckenmaker (814) 748-9021
Steve Firestone (804) 745-4123

West Virginia - Central
Joan Plant (304) 636-7153 (after 6pm)

support group corner



HELP WANTED

Wanted: A person with CMT (or a family member), a self-starter, energetic. Someone who likes to meet new people. A person willing to fill a leadership role, to be a facilitator for the CMTA Support Group of Central Florida. Immediate opening. Please call or write today to take advantage of this great opportunity. The rewards are GREAT! You will have an opportunity to meet people outside of your family that have the same neurological disease. You can provide a supportive atmosphere for those with CMT to come and share experiences, concerns and feelings. You can be a comfort to those people newly diagnosed and let them see that patients can live a very productive life. Don't cheat yourself out of the opportunity to be vital part of a very warm and caring Central Florida CMT family. Call today. 1-215-499-7486. §

(Editor's Note: This notice was written by Mary Beeler, the former support group leader of Central Florida. What she doesn't say is that this is an active and well-organized group that should not be allowed to falter for lack of a leader. Come on Florida members!)

Golf Tournament Nets \$900 For CMT Research

On August 24, 1992, Mary Jane King hosted the second Winnie Sorenson Charity Golf Tournament at the Deer Run Golf Course in Newport News, Virginia. The field consisted of 84 ladies from the Tidewater area. The format for the tournament was a two person select shot.

In all, the tournament raised \$1800 for charity with \$900 going to the research fund of the Lupus Foundation of America and \$900 going to the CMTA Research Fund.

Mary Jane King, who is the Tidewater CMT support group leader, was aided in organizing this tournament by her friend, Shirley Redding. §

We want to hear from

YOU!

Write us at:
Editors / The CMTA
201 Highland Avenue
Upland, CA 91786
215/499-7486

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.

Dr. Rhonda Jones' Survey Results Revealed

The preliminary results of the survey on chronic illness and life adjustments by Dr. Rhonda Jones which appeared in the Spring 1992 issue of *The CMTA Report* are provided below for those of you who took part in the survey. A second report of findings will appear in a later issue centering on the responses, both helpful and insensitive, which CMT patients receive from family and friends.

Dr. Rhonda Jones, psychologist, writes: "I am appreciative of the time and effort the participants put into answering this survey. In some instances, people went to considerable effort to elaborate extensively on their experiences. I am grateful to Karol Hitt and the CMT Association for their assistance in creating and administering this survey."

Respondents :

("n" refers to the number of people responding to each question.)

Total "n"=178 Males 37%..... Females 63%

Present Age of Respondents: Age at Diagnosis:

0-10..... 0%	0-10..... 6%
11-20... 3%	11-20..... 14%
21-30... 9%	21-30..... 18%
31-40.. 16%	31-40..... 18%
41-50.. 19%	41-50..... 19%
51-60.. 19%	51-60..... 15%
61-70.. 20%	61-70..... 9%
71-80.. 13%	71-80..... 2%
81-90... 2%	81-90..... 0%

1) First physical signs of CMT: (n=177)

A: Foot and ankle weakness

B: Hand weakness

C: Clumsiness

D: Lack of Endurance

A..... 37%	A and D..... 2%
B 5%	B and C 1%
C 20%	C and D..... 2%
D..... 3%	A,B,and C 1%
A and B 5%	A,C,and D 3%
A and C 15%	A,B,C,and D 6%

2) Symptoms were first noticed by: (n=177)

Myself: 56%

Someone else: 38%

Myself and someone else: 6%

3) If noticed by someone else, the someone was*: (n=76)

CMT blood relative 27%
Spouse 5%
Other relative 42%
Peer 3%
Doctor or nurse 19%
Shoe Salesman 1%
Athletic instructor 7%
Dance teacher 1%

*(Some people noted that more than one person noticed the problem.)

4) Those who first noticed the CMT themselves had: (n=109)

Prior knowledge of CMT 13%
No prior knowledge of CMT 87%

5) Those who had prior knowledge of CMT (refer to question 4) had, in almost every case known about CMT from a blood relative with CMT.(n=22)

6) Of those persons diagnosed prior to age 21, they reported excelling in the following areas to compensate for their CMT.

(Note: they could indicate more than one area of excellence.)

Category.....# of people	Category # of people
A. Academics 14	I. Other talent 4
B. Friendships 13	J. Other organizations 2
C. Help at home 10	K. Dating 2
D. Job 8	L. Being class clown 1
E. Sports 8	M. Nintendo 1
F. Clubs 5	N. Computers 1
G. Music 4	O. Drinking 1
H. Art 4	

7) After diagnosis, the feelings over the next few months were reported to be (more than one feeling could be indicated):

Accepting 70	Relieved 13*
Depressed 55	Frightened 2*
Nervous/Worried 53	Embarrassed 1*
Angry 51	Frustrated 1*
Sad 46	Asked "why me?" 1*
Denial 46	Confused 1*
Expected it 25	Challenged 1*
Hopeless 20	

*(Respondents' categories)

8) Those who reported feeling angry were angry at (more than one could be indicated):

Everything, everyone, life in general* 20
Doctors 20
CMT parent 16
Religious figure, God 14
Gym teachers 11
People without CMT 10
Non-CMT parent 6
Siblings without CMT 5
Peers* 2
Non-understanding people* 2
Shoe salesman* 1

*(Respondents' categories)



(cont'd on page 15)



Ask the Doctor

Dear Doctor:

I worked for 8 years in a machine shop that produced water faucets and used vast quantities of zinc and lead. Two years ago, the government closed the factory down because of the excessive lead particles in the air. I have been regularly tested and was found to have high levels of lead in my blood. The amount has been decreasing with each testing.

The question I have is whether or not breathing in the air-borne lead particles would have had any effect on my CMT. I seem to be experiencing increased symptoms, but I'm not sure whether or not that should be attributed to the lead.

The Doctor replies:

Concerning the patient who worked for 8 years in a machine shop that produced excess zinc and lead, it is unlikely that the lead that is now decreasing in her blood should have much effect on her CMT. However, lead toxicity can produce a number of symptoms usually involving the central nervous system and the gastrointestinal tract. Also, chronic intoxications can affect the peripheral nervous system and, therefore, it would be important to know how high the lead levels were and whether there were any associated signs or symptoms such as anemia. Most of the symptoms due to lead toxicity go away with reducing the lead in the body and falling levels would be indicative of improvement. However, this needs to be followed carefully to decide whether a medication that increases removal of lead should be used. If the symptoms of the neuropathy appear to be worse, it would be important to have an examination by a neurologist to see if there are new signs and symptoms that reflect lead toxicity. It would be important to do both electrophysiological studies as well as blood lead levels to document any change in the neuropathy. Personally I think that falling blood levels of lead should not be associated with progressing signs and symptoms of CMT.

Dear Doctor:

I have had pressure headaches for the past 17 years which until recently have responded to the removal of spinal fluid (CSF) by having a spinal tap done at 3 to 3 1/2 month intervals. It has been assumed for the past 17 years that my headaches are related to CMT in "some way." It is assumed that due to the high protein levels, ranging from 101 to over 150 and sometimes near 200 that my body does

not absorb the fluid. Consequently, the CSF builds up which causes increased pressure which in turn causes the headaches. Once the pressure increases, nothing resolves it except a spinal tap.

I am currently trying to decide whether or not I want to have a shunt inserted. Although it sounds like a reasonable solution, it would also bring with it its own set of problems. Finally, I am interested in finding out if CMT researchers might have a need for the spinal fluid which is collected at each procedure.

The Doctor replies:

The description of the headaches suggests that they may reflect the increased cerebrospinal fluid pressure and the likely diagnosis of benign intracranial hypertension. It would be important to know what the CSF pressure is and what the various scanning studies of the brain and spinal cord show. CMT with high CSF protein usually occurs in the demyelinating-hypertrophic types of CMT and may reflect abnormal or large spinal roots or some obstruction of CSF. This needs to be evaluated by a neurologist familiar with this type of problem. There are a number of treatments now available for benign intracranial hypertension and these need to be explored before a shunt should be performed. However, this needs to be determined by the evaluating neurologist and neurosurgeon. If the patient is developing hydrocephalus, a shunt may be indicated. If this is associated with small ventricles and no hydrocephalus, then medical therapy may do as well over time as long as there are no side effects of increased intracranial pressure. Most patients with CMT do not have this type of headache, and it is not likely that they are associated, although there may be an association in the hypertrophic type of hereditary polyneuropathy. In CMT, the CSF protein may be elevated especially in the hypertrophic-demyelinating forms although there are many other factors that may elevate the CSF protein and these need to be ruled out. Spinal fluid is always worth keeping and I would advise your doctor to keep it frozen and let the CMTA know that it is available, should any investigating laboratory wish to study the fluid in a particular way.

Dear Doctor:

Is there a predictable association between CMT and osteoporosis? I have been told by my podiatrist that I am beginning to show signs of osteoporosis

in my feet and that I will develop osteoporosis in my hands and arms as well because I have CMT.

The Doctor replies:

There is no simple answer to this question but, by and large, CMT and osteoporosis are two separate disorders. There is no direct causal relationship, but there might be secondary changes due to disuse that might result in osteoporosis. The patient should be seen by an internist familiar with bone metabolism. Any problem with high level steroid metabolism should be excluded.

Dear Doctor:

The majority of CMT patients seem to have high arched feet. I have the opposite...very flat feet. Please, comment.

The Doctor replies:

About 15% of CMT patients have pes planus (flat feet). There are two reasons for this. The first is that it depends on which nerves in the feet and legs become affected first. The relative rate of progression between the affected nerves must also be considered. Secondly, individuals can have other congenital foot abnormalities independent of CMT which predispose him/her to a flat foot presentation. Also, trauma (injury) can predispose or further direct the abnormality toward a specific structural alignment. Pes cavus (high arched foot) can become pes planus depending on the progression of the neuropathy.

Dear Doctor:

I have a CMT pen-pal living in Israel and her recent letter mentioned that she discovered a person there whose CMT was triggered by an immunization for hepatitis B. My friend works at a hospital which is now requiring that all employees be immunized and she is refusing. Do you know anything about this?

This letter followed an inquiry by the mother of a CMT child who was advised to receive the hepatitis B immunization and whose doctor asked her to check to see if this would be contraindicated.

The Doctor replies:

Some CMT patients also have an autoimmune polyneuropathy as well as CMT. For those patients we must be concerned about vaccinations. Most CMT patients should not have a reaction other than might be expected in anyone. If the CMT patient has a common reaction to an immunization, the problem will be

(continued on next page)

Ask the Doctor - cont'd from p.13

transitory. However, if the CMT patient has chronic inflammatory neuropathy as well as CMT, the patient could be left with a loss of motor and sensory function. There should not be anything neurotoxic in hepatitis B vaccine, however, there is a protein in some flu vaccines that might produce abnormal nerve response. Unless the patient has hypersensitivity or an autoimmune problem, they should be able to tolerate immunizations.

For the pen-pal in Israel who works in a medical setting hepatitis might be a real threat. Hepatitis is a far more serious condition than a reaction to the immunization for hepatitis.

In the case of the child, if the child has benign CMT then there is no reason to not have the vaccination. If the child has Dejerine-Sottas disorder, then the doctor should review how the child reacted to previously received immunizations. Final decisions must rest with the patient's individual physician.

Dear Doctor:

My physician requested an opinion about the drug Noroxin (quinolone) for recurring bladder infections. Please comment.

The Doctor replies:

Most antibiotics that are used for chronic bladder and yeast infections have side effects of numbness and paresis. As a result, long term Noroxin therapy is not a good idea, but it should not be a problem for short term therapy.

The following questions and answers were part of a workshop session from the United Kingdom's recent conference. They are reprinted with the permission of the CMT-U.K. publication.

If a patient donates a piece of a nerve for nerve biopsies, does it grow again? It depends on how healthy the nerve is. Nerve biopsies are taken from the side of the nerve.

If small muscles die off, can they be resuscitated? Muscles losing the supply of nerve impulses will atrophy. If they are not rejoined within 3 years, they disappear completely.

How beneficial is physiotherapy? Exercise is good within limitations. It is important to maximise potential, but one can cause damage by over-exercise.

Is physiotherapy only valuable post-operatively? No. It can be very beneficial to keep what muscles you have as active as possible.

What damage does over-exercise cause? It can decrease the patient's potential. A person should find their comfortable level and stay there.

What are the benefits of hydrotherapy? Very good, but it is important to have a designed exercise program.

Is there an "idiot's rule" as to when to stop exercise? When you get tired. When you regret today what you did yesterday.

Why do you feel on top of the world one minute and the next you are finished with no energy? This applies to many people with disabilities. A person who suddenly feels drowsy may have weakness of the diaphragm and breathing difficulties.

For breathing difficulties, would asthma pumps help? They will only help asthma. Breathing difficulties are rare - the dia-

phragm between the ribs is affected. Difficulties occur mainly when lying flat. An oxygen tube can help at night.

Does deterioration cause increase in poor circulation and cold feet? Cold feet are related to lack of movement. Massage can help. A person might also attend a pain clinic. Lumbar sympathectomy can help as it dilates the blood vessels.

Once a person is diagnosed with CMT is there any point in having a regular checkup? With Type I, a person should be seen once a year... Certain deformities can become rigid and they should be caught at the flexible stage. It is nice to keep in touch with the neurologist as adults in case anything comes along, or if the patient suddenly becomes worse. §

East Coast Is Site Of April 1993 CMT Conference

The A.I.DuPont Institute in Wilmington, DE, will be the site of the next CMT patient/family conference on Saturday, April 24, 1993.

The conference will begin with registration at 9:15 am. The presentations will begin at 10:00 am with lunch at 12 noon. The meetings will end at approximately 4:00 pm. Some of the presentations will be a discussion of CMT and children, orthopedic surgery for CMT patients, hand care options, physical therapy, and the psychological impact of CMT. The cost for the full day of presentations including lunch will be \$20.00. Make plans now to attend this very informative and interesting day. Wilmington, DE, is easily accessible and the facility is state-of-the-art in health care. Hotel and travel information will be sent with your reservation confirmation.

Fill out the registration form below and return it to the office of the CMTA by April 19, 1993 in order to insure your place at the conference. §

Wilmington CMT Conference Registration

Name(s) of attendees _____

Address _____

Telephone Number _____

Fee for one person \$ 20.00

(Fees include morning coffee, lunch, and all postage and mailings.)

Number of persons attending _____

Amount of money enclosed \$ _____

Return this form by April 19, 1993 to:

Pat Dreibelbis
CMTA
601 Upland Ave.
Upland, PA 19015

Survey Results - cont'd from p.11

- 9) Those who felt depressed indicated the following (they could indicate more than one):
- Depressed mood:50
 - Low interest in activities:21
 - Poor thoughts of self:43
 - Eating problems:19
 - Difficulty concentrating:32
 - Low sex interest:15
 - Sleep problems:25

- 10) Reactions to the diagnosis included the following (more than one could be indicated):
- Withdrawal:36
 - Sexual acting out: 5
 - Increased drinking: 8
 - Trouble at school: 2
 - Tantrums: 7
 - Drug abuse: 2

- 11) Current severity of symptoms was rated: (n=175)
- Slight:11%
 - Slight to moderate: 3%
 - Moderate:60%
 - Moderate to severe: 8%
 - Severe:17%

- 12) Present physical symptoms are (more than one could be indicated):
- High arch:146
 - Foot flops:111
 - Foot drop:125
 - Thin legs:108
 - Foot rolls to side:113
 - Thin hands: 93
 - Hammer toes:112
 - Bent fingers: 42

- 13,14) Surgeries included:
- 36% had foot surgery (n=177)
 - 5% had hand surgery (n=174)

- 15) Was the surgery helpful?
- Yes: 53%
 - No: 21%
 - Somewhat: 26%

- 16) If the surgery helped, did the surgery help movement? (n=52)
- Yes: 83%
 - No: 13%
 - Somewhat: 4%

- 17) If the surgery was helpful, did it help self-esteem? (n=47)
- Yes: 68%
 - No: 27%
 - Somewhat: 4%

§

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)
- Booklet - CMT FACTS I (\$3)
- Transcript - San Francisco CMT Conference (\$5)
- Letter - to Medical Professionals regarding the drug list (free to members with self addressed stamped business envelope)
- List - Physician Referrals (by state) (free to members with self addressed stamped business envelope) please list states: _____
- Medical Brochure - CMT (gray brochure) (free to members with self addressed stamped business envelope)
- List of Shoe Suppliers (\$1)

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

Total amount enclosed: _____

A copy of the official registration and financial information may be obtained from the Pennsylvania Department of State by calling, toll-free, within Pennsylvania, 1-800-732-0999. Registration does not imply endorsement.

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.

May 30 - June 2, 1993

These are the dates for the 28th Annual Conference, Association for the Care of Children's Health (ACCH) Chicago Marriott Downtown, Chicago, Illinois. The theme of this year's conference is Children's Health Care in Transition: Opportunities for Making a Difference. Health care professionals, family members, advocates, educators, and researchers are invited to register. For further information, contact Elena Widder, 301-654-1205.

Artist's Corner

A Retiree Whose Gift Is Giving

Relio Thibodeau did not want to retire at age 55, but that is just what he was forced to do five years ago when he could no longer work as the supervisor of an electrical shop. He was used to delegating orders and he suddenly found himself at home and wondering what would become of him and his future.

It took him one year to find himself, and in that year he found that life would and could go on.

Relio loves to do woodworking, and he remembered a picture he had kept from the Sacred Heart League. He had promised himself he would make it into a plaque and so he began work on it. When it was finished, it gave it to a friend.

As the cold and damp weather was affecting him, he decided to relocate from Connecticut to Florida. Relio has since converted his garage to his workshop. All his saws and tools are where he can do his work in a sitting down position. When the spirit moves him and he is not hurting too much, he works on making plaques. The gift of that first plaque has grown. In the three years since he moved to Florida, he has given out approximately 1700 plaques, free of charge, working through the local churches in his area. This is his therapy, and as long as his health holds out, he continues to work with local churches.

Relio and his wife, Susan, have four children and three grandchildren. Two of the children have CMT and one granddaughter has also inherited the disorder.

Relio always has a smile on his face, and he often says that he is very thankful for everything he can still do. He appreciates life, and has enjoyed getting back to the simple things which many of us take for granted.

§

CMT...

- is the most common inherited neuropathy, affecting approximately 125,000 Americans.
- is also known as peroneal muscular atrophy and hereditary motor sensory neuropathy.
- is slowly progressive, causing deterioration of peripheral nerves which control sensory information and muscle function of the foot/lower leg and hand/forearm.
- causes degeneration of peroneal muscles (located on the front of the leg below the knee).
- causes foot-drop walking gait, foot bone abnormalities, high arches and hammer toes, problems with balance, problems with hand function, occasional lower leg and forearm muscle cramping, loss of some normal reflexes, occasional partial sight and/or hearing loss problems and scoliosis (curvature of the spine) is sometimes present.
- does not affect life expectancy.
- has no effective treatment, although physical therapy, occupational therapy and moderate physical activity are beneficial.
- is sometimes surgically treated.
- is usually inherited in an autosomal dominant pattern.
- may become worse if certain neurotoxic drugs are taken.
- can vary greatly in severity, even within the same family.
- is the focus of significant genetic research, bringing us closer to answering the CMT enigma.

THE CMTA REPORT

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

Charcot-Marie-Tooth Association

Crozer Mills Enterprise Center
601 Upland Avenue
Upland, PA 19015

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