

# The CMTA Report

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

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## Profile of a Volunteer

**W**alter Sawyer is the leader of the support group in Port St. Lucie, Florida and has accomplished remarkable things since beginning his tenure about two years ago. At age 53, Walter has lived with the diagnosis for more than fifteen years.

He first got involved with the CMTA about six years ago when his mother needed a ride to a support group meeting in Boston. When he moved to Florida, he joined the West Palm Beach support group prior to its dissolution. Rather than let it fade away, he volunteered to head up a new group.

Walter's expertise was in the field of publicity since he had worked in that area in the military. So, he began by making the rounds of the local radio stations, doing talk shows. Then he branched out, faxing flyers to local doctors and mailing them a copy of his newsletter. The computer is his best tool, enabling him to send faxes with a single keystroke. Each time he pushes a button, a dozen press releases or public service announcements are sent out to local broadcasters. He sends regularly to nine local newspapers and one weekly.

Walter's diligence has paid off with two feature articles in Florida papers. The last one, in *The Stuart News*, has resulted in twelve phone calls about his support group meeting. Currently, he has over thirty participating members.

Only the future will tell how all of Walter's work pays off, but he intends to continue as long as people need his help. His ambitious goal is to someday have a local hospital set up as a CMT medical center.



*Walter Sawyer has found success in getting publicity for CMT and his support group in southern Florida.*

## CONFERENCES COMING THIS FALL

**T**wo regional conferences will be held this fall on the East Coast of the US. The first will be held at A.I.duPont Institute in Wilmington, Delaware on Saturday, September 21, 1996. The second will be held at Yale University in New Haven, Connecticut on Saturday, October 19, 1996. Each conference will offer members of the CMTA the opportunity to meet medical experts in that area who will answer questions and discuss different aspects of dealing with CMT, such as surgical options and therapies that might be beneficial. Make plans now to attend one of the conferences. Tentative plans are to hold spring 1997 conferences in St. Louis, Missouri and Madison, Wisconsin.

## GIFTS WERE MADE TO CMTA IN MEMORY OF:

**Zella Acree**  
Carrolyn Barber

**Floyd Barnhill**  
Flora Jones

**Louise Boswell**  
Anne Russell Owens

**Carmen Brenan**  
Claudia Larrea

**Berdie Cooper**  
Marilyn Prashker

**Irving Fleischer**  
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Mr. & Mrs. P. McGarry  
William Moffett  
Barbara Baker  
Martha Royse  
Marjorie Bailey  
Patty Sykes  
The Adler Family  
Florence Connolly

**Sadie Rech**  
Judith Goldman

**Maurice Sand**  
Judith Goldman

**Rebecca Sand**  
Elizabeth Levy

**Sister of Harriet Schacher**  
Marilyn Prashker

**Jerry W. Smith**  
Bob and Mary Williams

# CMTA Doesn't Slow This Racer Down

**F**red Sigarto of North Royalton, Ohio won the 1995 NE Ohio SCCA and Akron Sports Car Club "B" Street Prepared Autocross Championship. Fred drives a 1972 Datsun 240Z and competes against Corvettes, Porsches, Mustangs and other Datsuns. The amazing aspect of this accomplishment is that Fred does his competing while dealing with Charcot-Marie-Tooth Disorders.

An autocross is a precision driving event in which a driver tries to run a course through

slaloms, offset gates and increasing and decreasing radius turns. The course is generally set up in a parking lot. Orange cones mark the course and a driver tries to drive through it as fast as possible without hitting any cones. Hitting a cone is a 2 second penalty, and in an event in which times are measured in thousandths of a second, a driver doesn't want to knock over any cones.

Cars compete in various classes—stock, street prepared, prepared, and modified,

## Volunteers, Volunteers, Volunteers

**T**his newsletter highlights the accomplishment of some of the CMTA's many volunteers and members. No group so exemplifies the essence of volunteerism as the Board of Directors of the CMTA who direct the operation of the organization and establish its mission and vision. The group met during a retreat in February to set the goals for the CMTA during the year.



*Seated: Jack Walfish, Treasurer; Diane Freaney, President; Diana Eline; Ardith Fetterolf. Standing (l to r): Michael Molinari, Recording Secretary; Dr. William Quinn; Pat Dreibelbis, Office Administrator; Paul Flynn; Ann Beyer, Vice-President; Dr. Robert Lovelace, Chairman, Medical Advisory Board; Steve Khosrova; Ed Butchko, Chairman of the Board; Louise Smith.*



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## CMTA MEMBERSHIP/ORDER FORM

Name: \_\_\_\_\_

Address: \_\_\_\_\_  
\_\_\_\_\_

Phone Number: \_\_\_\_\_

*If outside the US, please see Treasurer's Note on adjacent page.*

*Fred Sigarto in the Firestone parking lot, Akron, Ohio at the 1995 Northeast Ohio (NEOHIO) SCCA Solo II Event.*

depending on the modifications done to the vehicle. Cars and trucks can compete. Anything from a Yugo to a Ferrari or Dodge Viper has run in events. Events run in the rain or sun and women can compete in their own classes or in the same class with men. Since it is a timed event, the driver is either on the gas or the brake at all times...not an easy task for someone with CMT.

Fred was diagnosed with CMT in 1991, just six months after his sister was diagnosed. Fred is a union carpenter and does all the modification work on his car himself. Fred says that "construction work is getting more difficult to perform. The dexterity, along with the strength, is not there anymore. Working outside in the cold weather or in high places is impossible."

After breaking his leg in three places in a fall at work, Fred began wearing ankle-foot orthoses. "They really help a great deal both at work and in autocrossing. They keep me from putting the power down too early when exiting a turn and spinning the wheels. I'm going to learn to left-foot brake, which should also improve my times."

CMT hasn't slowed Fred down yet and he doesn't plan to let it.

	QTY	COST	TOTAL
<b>Charcot-Marie-Tooth Disorders: A Handbook for Primary Care Physicians</b>		members \$15 non-members \$20	
Membership Dues		\$25	
CMT Facts I <input type="checkbox"/> English <input type="checkbox"/> Spanish		\$3	
CMT Facts II <input type="checkbox"/> English <input type="checkbox"/> Spanish		\$5	
CMT Facts III		\$5	
VCR Tape: CMT Neurology		\$15	
VCR Tape: Physical Therapy & Occupational Therapy		\$15	
VCR Tape: CMT Genetics		\$15	
VCR Tape: Orthopedic Surgery & CMT		\$15	
CMT Informational Brochure <input type="checkbox"/> English <input type="checkbox"/> Spanish		FREE	<i>Send Self- Addressed Stamped Envelope</i>
Physician Referral List: States: _____		FREE	
Letter to Medical Professional with Drug List		FREE	
<b>Contribution to CMT Research</b>			
<b>TOTAL</b>			

Checks payable to the CMTA

VISA  MasterCard

Card Number \_\_\_\_\_ Expiration Date \_\_\_\_\_

Signature \_\_\_\_\_

Mail to the CMTA, 601 Upland Ave., Upland, PA 19015

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

# Ask the Doctor...



## GIFTS WERE MADE TO CMTA IN HONOR OF

**Dr. Rebecca Brezel's  
75th Birthday**  
Bonnie & Mel Hirshowitz

**Eric and Jane Bowden**  
Mr. & Mrs. Keith Petersen

**Shirley Friedman**  
Jerry, Deb, Jeffrey &  
David Bank  
The Meltzers

**Drucilla Lake**  
Betty Lou Smith

**Tom Lynch's Birthday**  
Helen & Charlie Lynch

**Keith & Louise Petersen's  
50th Wedding Anniversary**  
Skippy & Barbara Griffin

**Samantha Lynn Sheriff**  
Mr. & Mrs. Daniel Headrick

**Brian Wiggins**  
Mr. & Mrs. Glenn Wiggins

**Hart Wurzburg's  
75th Birthday**  
Mrs. Leonard Pfaelzer  
Mr. & Mrs. M. Milton Minkin  
Mr. & Mrs. Harry J. Levi  
Ann & Arnie Wolff  
Marjorie & Lewis Lepman  
Lois Lurie  
Mrs. Henry Kahn  
Mr. & Mrs. G. F. Baer  
Mr. & Mrs. George Frank  
Mary & Sam Lawton  
Betty S. Frank  
Mrs. Wilbur A. Grodin  
Irene & Frank Jellinek

**Dear Doctor:**

*I recently talked to a pharmacist who said that there is a drug called Avonex available to help control Multiple Sclerosis. Does this drug have any known or potential benefit for CMT patients? I understand that it helps to control the myelin destruction in MS.*

*He also mentioned that taking large doses of B vitamins might be of benefit. Would this be useful?*

—J.C. Candia, NH

**The doctor replies:**

Avonex, made by Biogen, is a once-a-week treatment for Multiple Sclerosis and will not be available for prescription until July 1996. It is a form of Betaseron or Beta-interferon made in human cells and alters the immunological status in relapsing-remitting diseases such as MS. It is not suitable for the insidiously progressive disorder of CMT, nor would it be expected to control the myelin destruction. It is not approved for this disorder and I would have no good rationale for a treatment trial.

Multivitamins and vitamin B complex are not known to benefit CMT patients in the absence of a deficiency. Vitamin B6 or Pyridoxine in large doses is dangerous and can even cause irreversible neuropathy. It is on our list of contraindicated drugs.



**Dear Doctor:**

*I am 37 years old and was diagnosed with CMT at age 14. I have been leading a very active lifestyle which includes swimming competitively. I am working out five times a week and averaging 3,000 yards per workout. A lot of my swim workouts include rigorous sprint sets which I can handle very well. My question is, can rigorous exercise be harmful to CMT patients? And, could it speed up the muscle and nerve damage. I have read that moderate exercise can be beneficial to CMT patients and I hope that my workouts will be as well. I hope to be swimming all my life and decreasing my time for the 200-yard freestyle, which I currently swim in 2:35.82.*

—D.W. Santa Barbara, CA

**A physical therapist and member of the CMTA Medical Advisory Board replies:**

It is difficult to answer this question without seeing the person's involved muscles and their strength. In general, swimming uses the shoulder muscles of the arms, which are rarely affected by CMT. So there is little risk for problems with arm strength. However, swimming sprints would use a lot of leg muscles below the knee. Really vigorous sprint work could fatigue already weakened muscle to a degree that may be cause for concern. Although studies of the effects of exercise on CMT patients have been inconclusive, there is enough evidence to make us cautious about overworking muscles that have already been weakened.

## Supplemental Security

**S**upplemental Security Income is a monthly federal cash benefit that may be available if persons are 65 or older or have a disability. SSI may also go to children with disabilities.

Contact your local Social Security Administration Office to schedule an appointment. To find out the closest SSA office, call 1-800-772-1213.

When applying for SSI, it will help if applicants take the following information with them to their initial appointment:

★ Social Security card or a record of your

social security number;

- ★ Birth certificate or other proof of age;
- ★ Information about your home, such as the mortgage, lease, or the landlord's name;
- ★ Payroll slips, bank books, insurance policies, car registration, burial fund records and other information about income and items you own.

If you do not have all the above items, apply anyway. The people in the Social Security office can help obtain needed information.

If you want your SSI monthly checks to go

# R. Bradford Thomas: Profile of a Graduate

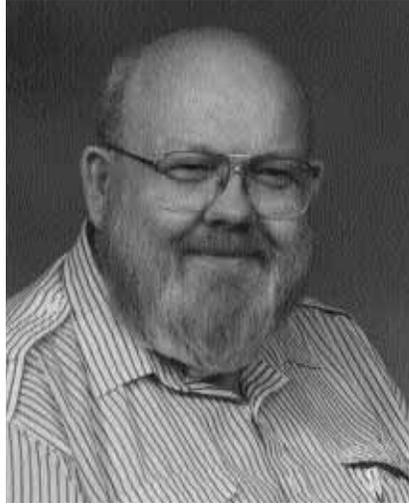
For years, Brad Thomas ignored the symptoms of Charcot-Marie-Tooth Disease, hiding them from his employers and even his doctors. As his muscles grew weaker, his purpose grew stronger. He never let his disability slow him down or keep him from earning a living.

At the age of 64, he didn't let it stop him from earning a doctorate in adult education from Ball State University in Muncie, Indiana.

After careers in computer management and telecommunications, Brad retired in 1988. He had previously earned degrees in chemistry, math and physics. Even though he was considered completely disabled by his doctors, he decided to work on a degree that would enable him to work with adults in accomplishing their educational goals.

Thomas walks slowly and uses a cane. He has AFO's on his legs to help him balance. He can no longer write, but he can use a computer. Because his energy level is low, he takes a nap each day. But Thomas has triumphed in spite of his limitations and he is proud of his degree.

Thomas took 100 hours of courses both full



*At 64, Bradford Thomas claims one more success in life: a doctorate in adult education.*

and part-time for more than five years. He had to rely on others, both classmates who shared their notes with him and his wife, Betty, who spent hours transcribing tape-recorded lectures for him. Professors helped by giving oral exams or allowing his answers to be dictated and then transcribed.

Brad's son and three grandchildren, as well as approximately 15 other members of his family, have all inherited the disease. But Brad has demonstrated to them all that it is important to look at what can be done and not what can't. He hopes to work with elderly people or with the parents of disabled children.

## ■ OF INTEREST

*Genetic Connections: A Guide to Documenting Your Individual and Family Health History* offers a systematic method of recording and maintaining a family health history. The book discusses human genetics and helps the reader identify personal health information such as birth history and life style. A second section contains chapters on anatomy and the physiology of ten body systems. There are explanations of inherited disorders and a set of instructions and questions to answer that help the reader in constructing his or her family health history. *Genetic Connections* is available at bookstores or from Sonters Publishing at 1-800-206-5450 for \$ 34.95.



## Income (SSI)

directly into your bank account, you should bring a checkbook or any other papers that show your name and account number.

To qualify for SSI you must:

- ★ Live in the US or Northern Mariana Islands;
- ★ Be a US citizen or be in the US legally;
- ★ Accept vocational rehabilitation services if you have a disability and services are offered;
- ★ Apply for other benefits you may be eligible to receive.

You must also fall within one of these categories:

- ★ Be 65 or older;
- ★ Have blindness, a severe sight impairment or another physical or mental disability that prevents you from working and is expected to last at least a year or result in death.

The basic monthly SSI check is the same nationwide; however, many states add money. To find out how much the SSI check will be, call 1-800-772-1213.

## RESEARCH NEWS

## Orthopaedic Considerations for Children with CMT

## ■ OF INTEREST

Medi-Span, Inc., a Drug-Disease Monitoring System, evaluates drugs and determines if they are contraindicated in specific diseases. The editor, Jennifer McQuade, Pharm. D., advises patients with CMT to tell their pharmacist as well as their physician that they have CMT. Pharmacists can help prevent medication errors, but will not be aware of your condition unless you tell them.



Medical Advisory Board members Dr. Phillip Chance and Dr. John Hsu have published an article entitled "Inherited Neuropathies" which appeared in *Current Opinion in Orthopedics*, 1995, 6:VI:100-108.

The orthopedic management section of the article contains information regarding the treatment of younger children with CMT:

"Due to the underlying neuromuscular condition and muscle imbalances, fixed contractures and permanent deformities in the child, adolescent or adult with CMT can develop in the spine, hips, hands and feet.

"Foot deformity in the very young child presents as a variation of the normal developmental process and the child stands on the tip toes rather than on the soles of the feet. At this early age, the foot is supple with no fixed contractures. Diagnosis of the condition should be established. Frequently, tip-toe standing can be corrected before it becomes habituated. Casting followed by the use of high-top shoes and braces can bring about resolution at a very early age, generally before 2 years. Surgical treatment like heel cord lengthening is generally not needed. The development of a high arch with prominent metatarsal heads can be seen at 3 to 4 years of age after the child has developed essentially a normal stance and gait. Generally, there are no complaints and the high arch may become a

concern of the family who requests a medical examination. The confirmation of the presence of abnormal neurologic and ancillary diagnostic findings related to CMT is important in starting the treatment process. If pes cavus becomes significant, there will be plantar fascia tightness. The metatarsal heads will also become more prominent with the development of forefoot deformities. The child can continue with normal activities without significant further impairment of the foot at that time.

"Frequent ankle sprains because of dorsiflexor weakness, especially of the peroneals, occur between the ages of 6 to 12 years. Initially, the use of ankle foot orthosis may help; however, the deformity may persist with the development of lateral calluses, forefoot deformities, and pain. Rebalancing the foot by tendon transfers, including the transfer of the posterior tibial tendon, is indicated. If the anterior tibial muscle is active, transfer of its tendon may be useful in further improving the overall studies of the foot.

"Fixed deformities can occur with persistent and long-standing muscle imbalance and are generally seen in older patients. Shortening occurs with the development of fibrous adhesions, leading to the development of fixed contractures. Growth changes in the underlying skeletal structures lead to the development of a fixed bony deformity. When this problem occurs, tendon transfers and soft tissue surgery alone are no longer sufficient. The foot must be reshaped such that it bears weight with good foot contact to the ground and in its most efficient manner. Bony operations including osteotomy alone, osteotomy with wedge resection, or arthrodesis of the joints may become necessary and allow for the foot deformity to be corrected. Unfortunately, the deformity can reoccur if the muscle pull of the foot is not balanced. Therefore, as a secondary procedure, muscle balance must be restored by tendon transfers.

"The results of operative procedures to correct foot deformities in CMT patients are difficult to interpret, not only because of the wide variation in degree of neurologic impairment in the individual patients, but also because of the variety of procedures that have been performed. Nevertheless, it should be emphasized that in the planning of treatment for a patient with CMT, it is important for the orthopaedic surgeon to understand the entire process. With flattening of

## INSECT DNA LINKED TO HUMAN DISEASE



Two disabling genetic diseases that affect nerves in the arms and legs are caused by duplication and deletion, respectively, of a DNA region containing a particular myelin gene. Scientists studying this phenomenon now report that a small segment of DNA, once thought to be present only in fruit flies, is located near a "hot spot" associated with the DNA rearrangement responsible for the two inherited neuropathies. When the DNA region is duplicated, a person develops Charcot-Marie-Tooth disease; when deleted, hereditary neuropathy with liability to pressure palsies (HNPP) results.

Researchers at Baylor College of Medicine in Houston found that the errant DNA near the recombination hot spot belongs to a group of DNA segments known as transposons or jumping genes. Transposons are able to insert and extricate themselves in genomes of many species, sometimes causing breaks in vital genes. It is not known how a piece of reputed insect DNA has found its way into the human genome. The scientists suggest it may have been carried by viruses or germline parasites sometime during evolution.

*Nature Genetics* 12:288-297, 1996.

## VINCRIStINE WARNING!

**D**r. Thomas Bird, Chief, Neurology Section (VA Medical Center), Seattle, Washington, sent the following information about work with vincristine-related neuropathy in CMT. The full article appeared in a publication of the American Cancer Society and is available upon request from the CMTA office.

A general predisposition for vincristine-related neuropathy has been observed in persons with a family history of hereditary neuropathies. We describe three families with autosomal dominant CMT1A, among whom a family member with a neoplastic disease suffered rapid-onset, severe neuropathy after receiving initial doses of vincristine as a part of a routine chemotherapy protocol. All three families have at least one affected member with the 17p11.2-12 duplication.

These cases show that 17p11.2-12 duplication predisposes patients to severe neurotoxicity from vincristine and that this drug should be avoided in patients with CMT1A. It is therefore essential to obtain a detailed family history for all oncology patients to screen for possible hereditary neuropathies. In patients with unexplained or preexisting familial neuropathy, testing for 17p11.2-12 duplication should be carried out prior to initiating vincristine therapy. Patients with other hereditary neuropathies may also be at risk for severe neurotoxic reactions.

pes cavus, further changes, such as clawing of the toes, may occur. In the child, changes in the foot attitude and shape because of unopposed muscle forces can be rapid. The planning and execution of tendon transfers and possible osteotomies may need to be done over a short period of time. However, for the patient with CMT that presents in adolescence or young adult years, many of the events have developed over a longer period of time. Subtle foot and ankle muscle imbalance can be controlled effectively by the use of braces, exercises, and stretching. Whenever the plantigrade attitude of the foot can be maintained without surgical means, it should be done. Only when there is significant dynamic or fixed deformity should surgical treatment be considered. Conversely, surgical treatment may maintain the integrity of anatomic relationships of the foot and ankle. This treatment will assist in the function of the lower extremity so that activity can be well maintained in this very slowly progressive disorder.”

# For Your Health: Depression



## Symptoms of depression can include:

- Persistent sadness or emptiness
- Loss of interest or pleasure in ordinary activities, including sex
- Decreased energy, fatigue, feeling “slowed down”
- Sleep disturbances (insomnia, waking early or over-sleeping)
- Eating disturbances (loss of appetite and weight or weight gain)
- Difficulty concentrating, remembering or making decisions
- Feelings of hopelessness, pessimism
- Feelings of guilt, worthlessness, helplessness
- Thoughts of death or suicide
- Irritability
- Excessive crying
- Chronic aches and pains that don’t respond to treatment

## Depression can result from the following:

- Biochemical imbalance in the brain
- Psychological distortions or “faulty thinking”
- Unresolved emotion (grief, fear, anger, shame)
- Some triggers that may set off depression include a genetic predisposition, major losses, unresolved feelings, serious illness, relationship or economic difficulties and alcohol or drug abuse.
- The most commonly used treatments for depression are antidepressant medication, psychotherapy or a combination of the two. Which of these is the right treatment for an individual case depends on the nature and severity of the depression and on individual preference. Medication can relieve physical symptoms quickly, while psychotherapy allows the opportunity to learn more effective ways of handling problems.

## To help the depressed person:

- Encourage treatment
- Get expert help
- Listen and validate feelings
- Build a supportive environment
- Structure activity
- Give the person control
- Learn about medications
- Be alert to signs of suicide
- Be watchful at holidays

## ■ OF INTEREST

### Computer Users:

If you are finding it difficult to type on your keyboard, the new Microsoft Natural keyboard may be a solution. It costs about \$90 and can be used on any computer. The keyboard might feel strange at first, but users report enjoying it once they adjust.



# Lori Ley: Portrait of a Female Bodybuilder

**D**ressed in street clothes, Lori Ley looks like any other mother who works at staying physically fit. She defies the stereotype of many of her counterparts who compete as female bodybuilders.

Lori placed second on April 15th in the Western Regional Finals sponsored by the National Physique Committee, even though she has been dealing with CMT for the past two years. It has gotten worse in the last year or so and Lori says she can't button a shirt or put her earrings on. But she continues to run and work out daily and has not changed her routine since before she was diagnosed with CMT.



*Lori Ley poses as part of the bodybuilding competition in the Western Regional Finals.*

Lori and her husband Fred, who is a pilot for Southwest Airlines, moved to Arizona two years ago, where Lori began her fitness regime in earnest between having her babies. Her motive, in addition to slowing the progress of the CMT, was to demonstrate that bodybuilding doesn't have to make someone big and muscle-bound. She uses weights and runs in an effort to tone and sculpt her body.

Although she's frustrated by not being able to open a Kool-Aid packet or a package of Pampers, she plans to continue her working out as long as she can. If nothing else, she knows it helps keep her spirits high and gives her the inner strength to go on.

*Harold Wilson poses with some of his collection of portraits and still lifes.*



# Harold Wilson: Portrait of an Artist

**H**arold Wilson, an Indiana native who moved to Florida eight years ago, is a self-taught artist who has been drawing and painting for as long as he can remember. He was diagnosed with CMT in 1949 and was told he'd be bedfast before he died. Now, he says, "I'm not far from that." Wilson has been wheelchair bound for the past five years and was in braces before that. But until recently, he always had his art.

Wilson can remember as long ago as third grade that a pastel he had done of mountains was taken around and shown off by his teacher. From that point on, without any formal training, he painted portraits, still lifes and landscapes. Unfortunately, now he can no longer paint because, as he says matter-of-factly, "I can't hold a brush."

Because CMT is frequently diagnosed before the age of 30, Wilson is hopeful that his two grown children have escaped the diagnosis. As he says, "You have a hard enough time in this life without being crippled. I would hate to think that I had given them something like this."

Wilson is a member of the Port St. Lucie support group headed by Walter Sawyer. Because Harold can no longer travel to art shows or paint, he is hopeful that area art clubs will be interested in helping him display and sell some of his paintings to generate some much-needed extra income. Harold is 74 years old.

# Letters to the Editor:

## Dear Editor,

This is in response to MWH's letter in the Winter 1996 newsletter.

RE: Her item 1 (bruising from direct contact with hard plastic on AFO): I had two arch inserts made for me 17 years ago. They fit under the arch of each foot, running from my heel to roughly the back of the ball of my foot. They are made of Pelite. They are durable—I have worn them every day for 17 years—weigh virtually nothing, are thin so they do not take up too much space in my shoes (I've had other bulkier supports which I've thrown away because they were too thick, causing my toes to rub against the tops of my shoes), and, most importantly, they support my feet and provide a buffer between my feet and the AFO plastic. They were custom made for me at an orthopedic shoe store.

RE: Item 3 (rashes): To prevent this, I do the following: First, I put on a pair of thin, all-cotton socks. (I call these undersocks.) Second, I put the AFO on. Third, I wear regular dress socks over the AFO (the oversocks). This arrangement has prevented rashes.

I think this is because the cotton undersocks absorb any perspiration. It is also much more comfortable to have a sock, rather than bare skin, against the AFO plastic. I use a thin undersock because it's cooler than wearing two pairs of thick socks and, with a thicker undersock, I found my toes rubbed the tops of my shoes because of the added thickness of the second sock. Thus, I buy the thinnest cotton socks I can find. I find this two-sock arrangement increases my comfort. Also, the outside sock covers the AFO, which I think looks neater than having the AFO show below my pant legs.

Hope this helps.

Sincerely,

—FE, Oakland, CA

## Dear Editor:

Reference: This note is in answer to two letters on braces, from members, in your recent Report (Winter 1996). I have CMT, which was diagnosed in 1957.

A short leg brace can be fabricated out of high-density polypropylene, which works extremely well, does not break, can be bonded for esthetics with naugahyde or vinyl, and is

comfortable. Of course, no design is perfect for every application.

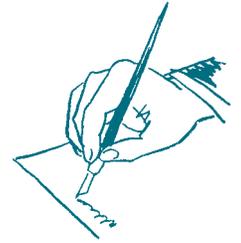
I developed this particular design in 1972, in concert with work being done by others at Los Amigos Hospital in Los Angeles, and I have tested it for 24 years in nearly every conceivable environment. I originally called my design the "Bikini brace" because it was light and flexible. Please note that I am not a doctor, nor was I a member of the Los Amigos team. But at the time I was a member of the Southern California aerospace community, who as a group were doing great developmental work with plastics.

At present there are at least two similar designs being mass produced and sold through medical supply stores and catalogs. Although they look good, they probably will not work satisfactorily without modification; and the modification can probably not be done by the average shop.

For information, the principle of the Bikini brace is a follows:

The CMT leg bone structure and ligaments tend to remain as good as original and the thigh muscles retain most of their strength. The deterioration is (i) in the distal nerves, and subsequently (ii) in the lower (distal) muscles. The brace needs only to compensate for lower leg muscle failure. More simply, the brace must guide the foot to a square landing on the floor during walking; and it must lift the ball of the foot from the floor as the foot is raised. The brace does not, and should not, provide structural strength. The bones and ligaments do that. Once the brace is on, and the foot is in the shoe, the only pressure is on the back of the calf. A strap made out of soft (belt) leather with a dot fastener is more lasting and suitable than velcro. Thickness of the plastic is only 0.090 inches, so normal shoe size is unchanged. Width at the toe is less than or equal to foot width—with no wrap around the ball of the foot. The walking surface of the brace can be covered with orthotic sponge rubber, 1/8- or 1/4- inch thick, for comfort. Again, this will not interfere with normal shoe size. Flexural breaking or fracture of the plastic can be controlled by annealing the brace after it is fitted—but before it is bonded to a vinyl covering.

The Bikini brace will not help the vertical, forward balance problem, which is caused by lack of controlled toe lift. As the body sways



## WRITE TO US!

Pat Dreibelbis, Editor

The CMTA Report

CMTA

601 Upland Ave.

Upland, PA 19015

## The CMTA Report

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**OF INTEREST**

**General Motors** and **Chrysler** offer cash incentives for adaptive expenses incurred by the customer of a newly purchased vehicle. For more information call GM's Motor Assistance Center at 1-800-323-9935 Mon-Fri. 9:00 am - 5:00 pm Eastern time or Chrysler's Physically Challenged Resource Center at 1-800-255-9877, Mon-Fri. 9:00 am - 5:00 pm Eastern time.



## More Letters to the Editor:

backwards, off center, the toe cannot be lifted to compensate. So a quick step backward must be taken, shifting the center of gravity forward. Note that only one type of brace will help improve this—the old-fashioned metal brace with the Klenzak (spring-loaded) ankle joint. With this brace, the toe is always being stressed upward. However, this brace is generally clumsy, unattractive and fatiguing to wear.

As I said in the beginning, nothing is perfect; neither is the Bikini brace. Because the brace is providing precast guidance, walking gait may still look awkward. Some structural strength may be incorporated in the brace to help the gait appearance. I have seen this before, and it does work. But the brace must encase the ankle a little more, and there are negative trade-offs.

However, let me mention that over the past 23 years, I have worn these braces everywhere—on the Sahara desert, in Saudi Arabia, the jungle of Malaysia, sailing, working, swimming, gardening, with tennis shoes, sandals and boots, racing a Porsche and raising two children; and they seem to work. I cannot walk without them. Thanks for printing this letter; I will keep you advised.

Sincerely yours,

—DC, Augusta, GA

**Dear Editor:**

I wanted to thank you for publishing the letter from BH in Auburn, CA. His letter really hit home.

I needed to have surgery done to my knee (not CMT related) and in the process of talking with my doctor about a foot question, he proceeded to order me to see a neurologist before he would do the surgery.

The first neurologist insisted that I could not play tennis, button my clothes, tie my shoes or even walk and had better quit my job, buy braces and change my career choice. She really depressed me. What's funny is that before I walked in her office, all I knew was my right ankle turns sometimes on rough hiking trails! I *can* walk 10–15 miles at one time. I was ranked 5th in my city in tennis. I can hold a pencil and dress myself! I DON'T NEED BRACES! On top of that she also charged my insurance company \$3,000 for services. (That cost doesn't include the braces.)

My point in this letter is to let all people with CMT decide on their own how they feel. I am so glad that you printed this letter from BH, as it made me feel more comfortable with my newly discovered diagnosis and my choice not to wear the braces that are somewhere deep in my closet. I am 35 years old and was diagnosed 6 months ago.

Sincerely,

—DR, Columbus, OH

P.S. The second doctor I saw said I wasn't that bad yet and to not worry—but why is it that we always believe the worst news?

**Dear Editor:**

I recently received your newsletter concerning CMT from a patient we have seen in our office. One article in particular I found interesting was the one about AFO's. Since reading the article on AFO's, I thought I would write you about a new material that we have recently started using here in our office. The material is called TPE, short for thermoplastic elastomer. It was developed by Goodyear Tire and Rubber Co. as an alternative for certain tires. This sounds unusual for bracing, but it has worked well for CMT applications. The reason is that TPE is a much softer, malleable product, not like most rigid plastics used in AFO applications.

Most plastics used until this time have been polypropylene or blends of polypropylene and polyethylene. In short, these types of rigid plastic offer little forgiveness for the CMT patient's sometimes flexible foot which is correctable.

The reason I am writing is to inform your readers of alternative plastics for the fabrication of AFO's. We try to combine movement with flexibility by using the TPE. This allows the patient to have motion at the ankle to help with dorsiflexing the foot as well as stabilizing the ankle from side to side. Again, the properties of the TPE make it supportive, but because of its flexibility, it is also more tolerable.

The advantage of TPE plastic is that it provides motion and flexibility. Most practioners should be able to discuss this product because it has been available for almost six years.

—Matt Hall, C.P.  
Progressive Prosthetic and  
Orthopedic Services  
Virginia

# CMTA Contacts

Following is a list of CMTA contact persons and support group leaders. There are many CMTA support groups, but more groups are needed. The CMTA will help you set up a group in your area. For information about forming a group or being a local contact person, please inform the CMTA by mail, fax 1-610-499-7487, or call the office at 1-610-499-7486. This page will appear in the newsletter whenever space permits.

## Alabama/Greater Tennessee Valley

\* Bill Porter 205/386-6579 work  
205/767-4181 home

## Arizona

Lavon Little 602/516-0539

## California

\* Janice Hagadorn 805/985-7332 after 5  
(Oxnard/Thousand Oaks)

\* Denise Miller 805/251-4537  
(Canyon County/Saugus)

\* Freda K. Brown 707/573-0181  
(Santa Rosa)

Gary Oleze 619/944-0550 after 6

Sandra Huntley 310/597-3728

Felice Gail Viggers 805/492-2840

Verna M. Sabo 818/892-6706

Mary Micalizzi 619/441-2432 after 6

Bob Hedge 310/645-2761 9-5

## Colorado

\* Dr. Gregory Stilwell 719/594-9920  
(Denver area)

Roberta Cummings 719/846-5611

## Connecticut

Mary Rehm 203/744-2786

\* Kay Flynn 914/793-4710  
(Fairfield)

## District of Columbia

\* Lorraine Middleton 202/362-4617  
6-9 pm

## Florida

William Brady 904/443-6271

Mary Beeler 407/295-6215 9 am-8 pm

Harold Wilson 407/465-3656

Pat Ports 407/965-3691  
M-W-F 4-9 pm

Joe Ellenbogen 305/921-4660

Edward Carhart 305/567-1066  
9:30-5:30

Beatrice Bannister 407/737-3267

Robyn Cohen 407/622-5829  
M-F 8-9:30 pm anytime weekends

Erika Stilwell 305/232-9066

\* Walter Sawyer 407/335-8624

## Georgia

Nancy Lee McCutchen 770/925-1020

## Kansas

\* Ardith Fetterholf 816/763-2176  
voice mail 816/756-2020

## Louisiana

Bobbie Marberry 504/872-0895

## Maryland

Jean Iler 410/987-5432

Linda Ember Miller 410/882-4019

Robert Kight 410/668-3054

## Massachusetts

Wayne Cardillo 413/298-3156

\* Donald Hay 617/444-1627 9 am-7 pm  
(Boston)

Jim Lawrence 508/460-6928

Jennifer Brelsford 413/538-9579

## Michigan

Robert D. Allard 517/592-5351

Debbie Clements 616/956-1910

\* Suzanne Tarpinian 313/883-1123  
(Detroit)

Laurie Vasquez 517/893-4125

## Mississippi

\* Julia Prevost 601/885-6482

\* Henry/Brenda Herran 601/885-6503  
(Jackson)

Mae Blackledge 601/763-5151

## Minnesota

Rosemary Mills 320/567-2156

Grace Wangaard 612/496-0255

## Missouri

\* Ardith Fetterholf 816/763-2176  
voice mail 816/756-2020

Allan Degenhardt 816/942-1817

Carol Haislip 314/644-1664

Lisa Minzer, RN 314/978-0557

## New Hampshire

Mary Knightly 603/598-5451

## New Jersey

Janet Saleh 908/281-6289

Linda Muhlig 609/327-4392

Gary Orson 609/584-9025  
M-F 6-10 pm & weekends

Russell Weiss 908/536-6700

## New Mexico

Jesse Hostetler 505/536-2890

## New York

Joe Ehman 716/442-4123

Internet: KOLOB@Multicom.org

\* Diana Eline 201/861-0425 before 9 pm  
(New York City)

\* Abby Wakefield 212/722-8052

\* Bernice Roll 716/584-3585  
(Rochester)

\* Kay Flynn 914/793-4710  
(Westchester County)

Amy Gander 518/373-9907

Angela Piersimoni 607/562-8823 after 2

Sharon McAvey 718/380-3792  
afternoon & evening

William Carrington 718/486-6953  
4-11 pm

## North Carolina

Diane Rodden 910/584-3655

\* Susan Salzberg 919/967-3118 5-9 pm  
(Durham)

Raymond Woodie 910/838-3221

## Ohio

Roger Emmons 216/286-6485

Suzanne Lammi 513/339-4312

Norma Markowitz 215/247-8785

## Oklahoma

Leah Holden 405/255-4491

## Pennsylvania

Dennis Devlin 215/269-2600 work  
610/566-1882 home

Patricia Zelenowski 717/457-7067

Camille Walsh 215/747-5321

Janet Fierst 412/487-0757

Mary MacMinn 215/322-1073

Carol Henderson 215/424-1176

Tony Petre 412/647-8324

## Rhode Island

Robert Matteucci 401/647-9154 in pm

## Texas

Karen Edelson, DPM 214/542-0048  
214/542-0122

M-T-Th 8:30-5

Tony Collette 713/699-8432 1-8 pm

## Virginia

\* Mary Jane King 804/591-0516  
(Tidewater)

## West Virginia

\* Joan Plant 304/636-7152 after 6 pm  
(Central)

Barbara Compton 304/636-5456 24 hrs.

## CATALOGS OF INTEREST

### Special Needs Catalogs:

*Enrichments for  
Better Living*  
145 Tower Drive  
PO Box 579  
Hinsdale, IL 60521  
1-800-323-5547

*Independent Living  
Aids, Inc.*  
11 Commercial Court  
Plainview, NY 11803

*OT Ideas, Inc.*  
124 Morris Turnpike  
Randolph, NJ 07869  
1-201-895-3622

Variety of games,  
devices for writing,  
cutting, etc. geared  
for children with small  
motor difficulties.

### Shoe Company Catalogs:

*Masseys*  
601 Twelfth Street  
Lynchburg, VA 24506-  
0088

1-800-462-7739  
Sizes range from 3-14,  
widths AA to EEE.

*Okun Bros. Shoes*  
356 E. South Street  
Kalamazoo, MI  
49007-4883  
1-800-433-6344  
Carries extra-depth  
shoes.



## MEDICAL ALERT:

*These Drugs  
Are Toxic to  
the Peripheral  
Nervous System  
and can be  
harmful to the  
CMT patient.*

Adriamycin  
Alcohol  
Amiodarone  
Chloramphenicol  
Cisplatin  
Dapsone  
Diphenylhydantoin  
(Dilantin)  
Disulfiram (Antabuse)  
Glutethimide (Doriden)  
Gold  
Hydralazine (Apresoline)  
Isoniazid (INH)  
Megadose of vitamin A  
Megadose of vitamin D  
Megadose of vitamin B6  
(Pyridoxine)  
Metronidazole (Flagyl)  
Nitrofurantoin  
(Furadantin, Macrochantin)  
Nitrous oxide (chronic  
repeated inhalation)  
Penicillin (large IV  
doses only)  
Perhexiline (Pexid)  
Taxol  
Vincristine

Lithium, Misomidazole  
and Zoloft can be used  
*with caution.*

Before taking any med-  
ication, please discuss it  
fully with your doctor for  
possible side effects.

# What is CMT?

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

## The CMTA Report

*Information on Charcot-Marie-Tooth Disorders  
from the Charcot-Marie-Tooth Association*

**Crozer Mills Enterprise Center  
601 Upland Avenue  
Upland, PA 19015  
1-800-606-CMTA**

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