CELEBRATING OUR 15th ANNIVERSARY

SPRING 1998

The CMTA Report

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

EMAIL CMTA AT:
CMTA@charcot-marie-tooth.org

Buuck Family Provides Lead Gift for Armington Research Challenge

Bob and Gail Buuck have made a $35,000 gift to the CMTA research fund. This generous commitment will count toward the $150,000 the Association must raise before June 30th to meet the requirements of the Armington Research Challenge (detailed on page two). The Armington Family's intent was for the challenge to be a catalyst for increased research support. It has been that and more. With the initiation of the challenge nearly three years ago, more than $140,000 has been dedicated to CMT research in the form of fellowships and summer intern grants. This increased research activity piqued the interest of Bob Buuck.

In October 1997, Bob and Gail traveled from their home in Minneapolis to the CMTA patient/family conference in Detroit, which was hosted by Dr. Michael Shy of Wayne State University. It was there that the Buucks learned about exciting new developments in CMT research, particularly in Mike Shy's lab. Dr. Agnes Jani, first recipient of the Armington fellowship grant, works closely with Dr. Shy in the use of adenoviral vectors for possible gene therapy in the P_0 mouse, which mimics one form of CMT.

The Detroit meeting was the first the Buucks had attended. Gail enjoyed the opportunity to interact with other CMTA members, exchanging personal experiences and perspectives on CMT "management." Bob was especially impressed with the comprehensive program at Wayne State covering patient diagnosis and care and basic research. With a background in the biotech/medical devices field, Bob has a keen understanding of proper lab set-up and techniques.

Bob is very pleased to see the CMTA taking a leadership role in CMT research. "It's not enough to fund research," says Buuck. "The CMTA must become a leading source for all relevant data pertaining to CMT. It's a huge undertaking, but it must be done to properly guide research grant decision-making." The CMTA is fortunate to have both the financial support and professional advice of Mr. Buuck. Dr. Agnes Jani is the recipient of a research grant made possible by the Buuck family.

> *"The CMTA must become a leading source for all relevant data pertaining to CMT."*

Armington Research Challenge—*Take It Personally!*

Help the CMTA to meet its June 30th goal by sending in your contribution for CMT research today.

Begin With Flynn

> *"If you want to find out who you really are, become a volunteer. You will gain far more than you give. It also happens to be the greatest gift you can give yourself."*
>
> **—Andy Fuller,**
> **Courage Center volunteer**

This issue of *The CMTA Report* is a favorite of mine because it pays special tribute to our volunteers. The above quote, which I read while visiting the Courage Center in Minneapolis, MN (story on page 14) really captures the essence of what it means to volunteer. Charitable service, whether it's our Board of Directors, Medical Advisory Board members, support group leaders or "project" volunteers, is the engine that drives the CMTA. We are experiencing an exciting and rapid period of growth. There is an increased demand for support groups (San Francisco, Dallas, and Atlanta, to name a few), hundreds of people are joining the organization, particularly via the Internet, and many members are coming forward to help with fundraising. The CMTA is building momentum.

Volunteers are contributing their time and talents to advance the organization. Cynthia Gracey, Maureen Horton, and Rex Morgan are writing for the newsletter. For the Westchester Broadway Theater event, Dick Sharpe helped get radio public service announcements, Michele Vogtman designed the program, and Jeff Beyer, Luba Kerod, and Dan Max obtained items and services for auction. *Cycle for CMT,* started by former board member Joe Beernink, is being continued by Mike Reardon (see page 7 for details). Special recognition goes to this year's Volunteer of the Year, Dr. Michael Shy of Wayne State University in Detroit. Mike wrote an excellent grant application to the National Institutes of Health for conference funding, recruited the speakers, and organized the program for our international conference, and hosted a patient/family conference last October at WSU. Mike's department chair is starting to wonder who Mike really works for... To all our volunteers, many thanks for your dedication and enthusiasm!

Paul Flynn, Executive Director, and Nancy Homyak met Prof. Sarah Blacher Cohen (seated) in Albany, NY after seeing her most recent play "Soul Sisters." Dr. Cohen, a CMTA member, has offered the use of her plays to raise funds for the organization.

Armington Research Challenge—*Take It Personally:* The CMTA is in the third year of a four-year "challenge" with the Evenor Armington Foundation. The original terms of the challenge, $25,000 for each of the years to be matched on a 1:1 basis, were far exceeded—more than $45,000 was raised in each of the first 2 years. This overwhelming response enabled us to "renegotiate" the terms of the challenge. The Armington Family accepted our proposal: the Foundation will provide $50,000 in 1998 and 1999, if the CMTA will match that on a 3:1 basis—$150,000 in each year.

Third International Conference on CMT Disorders: Scheduled for October 21–24, 1998, the conference is being chaired by Medical Advisory Board members Dr. Robert Lovelace, Columbia University, NY and Dr. Michael Shy, Wayne State University, MI. The CMTA Board of Directors and MAB members saw this as an opportune time to host this meeting because considerable advances have occurred in the understanding of CMT. The genetic causes of most of the demyelinating forms of CMT have been identified. The development of viral vectors to introduce genes into the peripheral nervous structure, and the identification of trophic factors to promote nerve regeneration and remyelination, are making "gene therapy" for CMT a realistic possibility in the future.

The conference, which is a collaborative effort with the prestigious New York Academy of Sciences, will focus on the latest developments in research and clinical aspects of CMT. It will be of interest to geneticists, molecular biologists, clinicians, morphologists and physiologists. The leading CMT investigators from around the world will be presenting their work, including Dr. Phillip Chance, University of Washington,

continued on page 4

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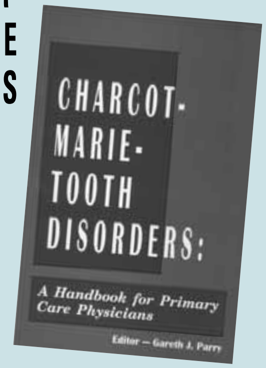

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- *Charcot-Marie-Tooth Disorders in Children,* by Harold Marks, MD, A.I. duPont Institute;
- *The Clinical Features of Charcot-Marie-Tooth Disorders*, by Carlos Garcia, MD, Louisiana State University School of Medicine;
- *Genetics of Charcot-Marie-Tooth Disorders*, by Jeff Vance, PhD, MD, Duke University;
- *Physical Therapy: Conservative Management*, by Carol Oatis, PT, PhD, Beaver College;

and eight more chapters, each written by an expert from the CMTA's Medical Advisory Board.

VIDEO TAPE OF WAYNE STATE CONFERENCE NOW AVAILABLE!

A tape of the Wayne State CMT conference held on October 18, 1997 in Detroit, Michigan is now available. The tape contains over five hours of presentations from the conference. Featured on the tape are Dr. Michael Shy, neurologist and head of the Wayne State CMT clinic, discussing Types I and 2; Dr. William Quinn, podiatrist and member of the CMTA Board of Directors, discussing surgical options; Dr. Steven Hinderer, physiatrist, discussing rehabilitation and CMT; and Dr. John Kamholz, neurologist, presenting an overview of CMT research from an historical perspective. Interviews and excerpts from workshops round out the material. *See order form at right.*

cut here

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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.

BEGIN WITH FLYNN

(continued from page 2)

Dr. James Lupski, Baylor College of Medicine, Dr. Gareth Parry, University of Minnesota, Jeff Vance, Duke University, and Christine Van Broeckhoven, University of Antwerp, Belgium. The program includes more than 50 presenters, several of whom reside in Europe and the Middle East.

Sarah Blacher Cohen: Dr. Cohen, a professor of English and an acclaimed playwright, has been on the faculty at the University at Albany for 25 years. Sarah recently called Pat Dreibelbis, Director of Programs, asking for some advice—where to buy shoes. "I've had the same shoes for longer than I care to admit… it's terribly frustrating." After Pat supplied referrals for shoe stores and podiatrists, the conversation turned to Sarah's plays and the possibility of doing benefits for the CMTA. Cohen's plays include: "Molly Picon's Return Engagement," "Sophie, Totie and Belle" and "Sophie Tucker: Red Hot Yiddishe Mama," both co-written with Chicago playwright Joanne Koch. Her comedy about disability (a personal account of living with CMT), "The Ladies' Locker Room" would provide the perfect context for a CMTA fundraiser. Those interested in helping to host a benefit should call the office at 1-800-606-CMTA.

Dr. Paul Donohue: CMTA members may recall that Dr. Donohue, whose medical Q&A column appears in nearly 300 newspapers, wrote about CMT in early January, generating nearly 900 calls to the office. Another piece appeared on May 18th that will include our web site address. We are grateful for the exposure and education that Dr. Donohue is helping to provide. Most importantly, we thank him for the advance notice.

—Paul R. Flynn, Executive Director

Volunteers in Action

This issue of *The CMTA Report* salutes the many volunteers who make this organization what it is today. With a membership of almost 13,000 and an office staff of 2 1/2, it is clear that much of the organization's work must be done by volunteers. Fortunately, the CMTA has never lacked volunteer help and so we salute many of the people who help us "in the field."

Cynthia Gracey, who has taken the time to write articles for the newsletter and who presented at the Florida conference. **Bill Shaw,** our resident "news hound," who sends us clippings and exciting pieces of information about research studies around the world that might impact on CMT. **Kay Flynn and Nancy Homyak and the Westchester Support Group**, who worked for weeks getting the Westchester Broadway Theater fundraiser up and running and ultimately made it a success. **Maureen Horton,** who has become a one-woman crusader for information about HNPP and who has contributed several excellent articles to our newsletter on the subject. **Dr. Gareth Parry**, who has acted as her medical reference and editor. **Dr. Carlos Garcia**, who is always available to answer difficult medical questions posed by members of

Another contributor to the wonderful fundraiser in Elmsford, NY, was James Ranti, a member of the Westchester County support group. Jim played classical trumpet selections before the performance as attendees gathered in the lobby of the theater.

Ruth Levitan, visiting relatives in the Philadelphia area, stopped in to the CMTA office to discuss with Pat Dreibelbis, Director of Program Services, the new support group she is co-sponsoring with Miriam Diaz in the Berkeley, CA, area.

The "tie quilt" is the second such effort by Gloria Charny, the proceeds of which benefit the CMTA. The creative handiwork of Gloria takes many hours to complete. The quilt is displayed by Daniel Leahy and his mom Peg (off camera, and never known by family members, including grand nephew Paul Flynn, to be the least bit shy).

the CMTA. **Dick Sharpe**, who used his connections in communications' to get free public service announcements on several radio stations for the Westchester, NY, fundraiser. **Jim Ranti**, who played many musical selections before the presentations of "Scattered Thoughts" and "Parusia." **The Board of Directors of the CMTA**, who establish policy, attend meetings, represent the organization at medical conferences, set the budget (and try to keep us "on" budget), set up and run patient/family conferences, encourage and attend support group meetings, and do a million other things. **The Medical Advisory Board of the CMTA**, a group of more than 50 doctors who attend meetings, answer questions for the newsletter, review possible drug complications, and provide peer review for the CMTA's research grant proposals. **Nora Burrows, Bob Budde, Rosemary Mills, Angela Piersimoni, Miriam Diaz, and Ruth Levitan**, new support group leaders whose enthusiasm for the task and high degree of organization and commitment energize us all. **Walter Sawyer, Freda Brown, Sue Tarpinian, and Kay Flynn**, long-time support group leaders who have assumed the new task of being regional leaders of the support group network. Playwright and Professor, **Sarah Blacher Cohen**, whose published work, *The Ladies' Locker Room*, will be the basis of a fundraiser in the near future. **Buzz Van Almen**, who videotaped the entire Wayne State Conference and whose work is the basis of the new videotapes now available. **Beth Troutman**, who has designated the CMTA as the recipient of the Women's Civic Group of Hershey's fundraising efforts. **Dan Bachman**, a student at WITC-Rice Lake, who manned a booth on CMT at the Health Fair on campus on February 26, 1998. **Larry Gomez,** who recruited Park-Hill Chemical Corporation and the Westchester County Police PBA as sponsors or the theater event. **Anita Flynn**, who had the plaque for "Above and Beyond" made and who acted as overseer to many of the details of the WBT event. Fine artist **Tom Garrick**, who generously contributed a beautiful piece for the silent auction. **Jack Volk**, who worked his report writing magic for a research project.

If we've omitted you, please forgive us and let us know so we can properly thank you in the next issue of the newsletter. We would like to see many more names in this acknowledgement, which will appear regularly in the newsletter. We could use your help with fundraising, support groups, getting more publicity for the organization, and things we probably haven't even thought of. No job is insignificant; no effort unappreciated. Thanks to all of you who help and to all of you who will soon!

Beth Troutman (left) and Deb Alexander of the Women's Civic Club of Hershey show a $2,000 check that their club presented to the the CMTA in April. The CMTA was this year's President's choice for their fundraising efforts. Troutman, President of WCCH, incidentally, is affected by CMT.

In Tune and On the Go

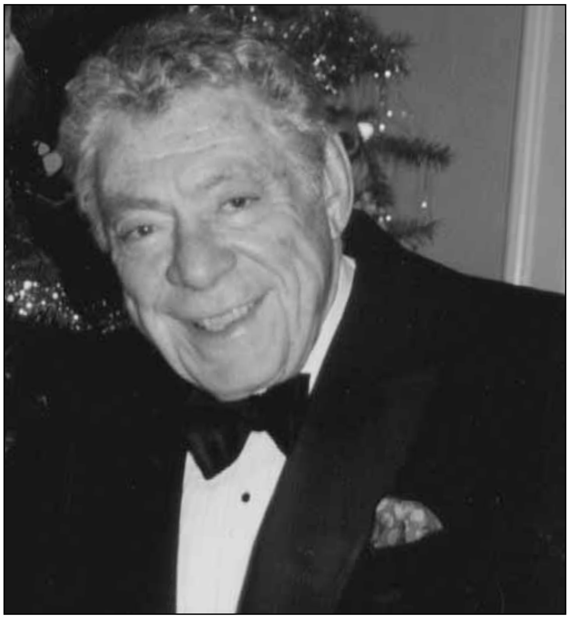
When *The CMTA Report* last profiled Larry Wechsler some 10 years ago, he was leading the ultra-busy life of the successful Manhattan executive. As president of Revlon's U.S. Beauty Division, he had a stylish corner office overlooking Madison Avenue. He also had a schedule that took him around the globe, meeting and greeting the top people in the cosmetics industry like Elizabeth Arden, Helena Rubenstein, and Charles Revson.

These days, however, Larry is playing a decidedly different tune. Quite literally. Since retiring from Revlon 8 years ago, Larry, 73, has taken up the saxophone again—an instrument he played in his youth. "It's been great fun," he says, speaking from his weekend home in Millbrook, NY, about 90 minutes north of New York City. "I'm nowhere near a professional level but I'm getting better. Hopefully, in a short period of time, I'll make my debut somewhere."

What made him decide to toot his horn again? A visit to the New School of Jazz in NYC that reignited his interest in music. "I dug out my old horn, got it reconditioned, and started taking lessons again," he says.

Aside from the joy of playing jazz, Larry has derived some physical benefits, too. "One of the things about my CMT that bothers me the most is my hands. Playing the saxophone has helped me a lot because I'm using my fingers a lot more."

Now don't think for a minute that all this saxophoning has turned Larry into a mellow fellow. This is one retired executive who's still on the go. "I like keeping active," he says. "I don't feel like slowing down. I don't want to and I think it's a serious mistake to 'stop doing.'"



Larry Wechsler, "distinguished alumnus" of the CMTA Board of Directors, looking distinguished in black-tie and ready-to-go… as always!

Keeping active means traveling with his wife Jan (Vietnam, Laos, and the Middle East are a few recent destinations), golfing, consulting (for Christian Dior) and—pant-pant—teaching. For the past 6 years Larry has taught a fully accredited course, "Creative Marketing Design," at the Parsons School of Design in NYC. "It's been a great experience," he says. "I'm fortunate to call upon some friends of mine who are people I worked with over the years. I bring in a series of guest lecturers, people who are very highly placed in different industries—advertising, broadcasting, senior management.

"I've been able to bring to the class a happy merger of subject matter and professionals who are doing it in the field in real life."

Larry is also in his 15th year studying tai chi, the ancient Chinese martial art that's been called "mediation in motion." Unlike the hard martial arts that involve a lot of chopping and loud "hi-ya!"-ing, tai chi is characterized by soft, slow, flowing movements that emphasize force, rather than brute strength.

And Larry thinks that can be a good thing for people with CMT. "Tai chi gives one a sense of balance that is particularly useful for people with CMT," he says. "There is a wonderful sense that one gets, even when learning how to stand. I think it would be great to get people with CMT to be taught the fundamentals of tai chi."

Larry combines tai chi with regular workouts in a gym. "I lift weights and stretch. I work out 2 to 3 times a week. Anyone who says someone with CMT shouldn't work out is crazy, they have no idea of the benefits. It is absolutely vital to keep your sense of balance, to keep your physical strength, and to keep your spirits up."

"The key is knowing when to stop. Not beating yourself into a sense of exhaustion or injuring yourself."

That doesn't figure to happen to Larry anytime soon. "Hey, I dug out my ski boots last year and that's something I've been considering!" he says.

—*Rex Morgan*

Editor's Note: Maybe Larry should take up mountaineering, too! He could probably get some good tips from his son, Brad. Brad Wechsler is the current co-chairman of IMAX Films, producers of the critically acclaimed film, Everest.

Cycle For CMT

By MIKAEL REARDON

Last year, former CMTA board member Joe Beernink demonstrated commitment and courage in his Denver to Aspen 200-mile bicycle race across the mountainous terrain of the Colorado Rockies. Many CMTA members and friends showed tremendous support and generosity by supporting Joe in his efforts to illustrate what a person with CMT can accomplish with determination and will power. A cyclist myself for many years, I personally was heartened to read about another person with CMT pushing himself to the limit despite his condition.

With the support and assistance of the CMTA, I would like to continue to develop the Cycle For CMT idea by forming a team comprising association members, family, and friends who would participate in long-distance cycling events such as the Denver to Aspen Classic, 24-hour races, and cross-state relays, and possibly multi-day events. Participation could be in the form of cycling, road support for the cyclists, or basic organizational and logistical help. The goal of these collective efforts is to inspire those with CMT and parents of children with CMT to explore every avenue of physical fitness and to experience the joy of sport. An organized team effort could also benefit the CMTA by expounding our cause publicly through media coverage and corporate sponsorship.

I invite all CMTA members and their families who would be interested in this endeavor to contact me, Mikael Reardon, by e-mail at reardonm@frb.gov or through the CMTA office. The immediate goal is to participate in an event sometime this summer, with others following in the fall, so we need to hear from you soon for this to become a reality.

GIFTS WERE MADE TO THE CMTA IN MEMORY OF:

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Paul Flynn

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Eileen Giberson

Michael Claflin
Nancy R. Bauard

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Mr. & Mrs. E. A. Jumper's 50th Anniversary
Jessie Dezern

Debbie Militello's Birthday
Elna S. Crook

Miss Hanna Pearlstine's 95th Birthday
Mimi and Donald Vineburg and Family

Susan Solomon's Birthday
Ted and Mary Ann Simon

Mrs. A. Hart Wurzburg's 75th Birthday
(see page 9)

Pressure Palsies of HNPP

By MAUREEN HORTON, R.N.

HNPP causes pressure palsies—periods of numbness and/or weakness that last for a while and go away. Some people have only one or two episodes. Others have frequent episodes. As there is no cure, the number one treatment for HNPP is avoidance of the activities that cause the pressure palsies, or doing those activities in shorter intervals. For those with frequent episodes, life with HNPP can be quite challenging.

The easiest way to describe pressure palsies is to say that it is like when an arm or leg "goes to sleep," only it lasts longer. In normal people, the arm or leg becomes numb and changing position takes the pressure off the nerve. After a few seconds of the uncomfortable pins-and-needles sensation, things return to normal.

With pressure palsies, one is never quite sure what will happen. Pressure palsies can come on within seconds of doing a wrong activity or it can take several days of the same activity before the palsy develops. The pressure palsy may start out with a pins-and-needles sensation or it may not. Changing positions may stop the pressure palsy from developing, or it may be too late and the damage is already done. The numbness may be fairly mild or quite severe.... like getting a large dose of novocaine at the dentist (but without the feeling like your lip is sticking out 3 inches). Muscle strength may be affected—partially, significantly, or not at all. It may be difficult to grasp things or get a hand to the mouth to eat or brush teeth. It may take minutes or months to recover from a pressure palsy. Recovery may be complete or partial.

Sitting with legs crossed and leaning on elbows are what doctors tend to mention as causes and activities to avoid. And those are certainly two big culprits. But any pressure (internal or external) or stretch on the nerve or repetitive use can cause the pressure palsies. Here are just a few examples.

In the hands, leaning on the palms is a palsy-inducing position. That can limit gardening and scrubbing a floor or bathtub (OK by me!). But it also can include activities such as sanding a piece of wood, wiping off a kitchen counter, lifting weights, or resting the palm on a hard surface while using a computer mouse.

Chance Finding Spurs Race for Nerve Drugs

Editor's Note: The following article summarizes the report from the April 21, 1998 Wall Street Journal. It has been suggested by members of the CMTA's Medical Advisory Board that the following research has positive implications for the treatment of CMT.

Research teams at Johns Hopkins University in Baltimore and Oregon Health Sciences University in Portland recently made the surprising discovery that medicines originally developed to suppress the immune system and fight organ-transplant rejection can also regrow nerves in the spine or brain damaged by neurological diseases or accidents.

Several companies, including Guilford Pharmaceuticals, Inc., Vertex Pharmaceuticals Inc. of Cambridge, MA and Pfizer, Inc. are racing to develop these experimental nerve drugs, called neuroimmunophilins, that can be used to combat the largely untreatable symptoms of illnesses such as Parkinson's disease, Alzheimer's disease, and stroke.

The new nerve-growth agents, the neuroimmunophilins, have several advantages over the older nerve drugs. They can be given orally, suggesting they will be able to slip easily from the bloodstream into the brain. They also appear to target only damaged nerves while leaving healthy ones intact, suggesting they may have fewer side-effects. Scientists guess they work in a way analogous to hormones, spurring production of growth-boosting proteins and other chemicals normally abundant only during the first years of life when expanding cell growth is needed.

CMTA Medical Advisory Board Member Dr. Gareth J. Parry, Chair, Dept. of Neurology, University of Minnesota, comments on these findings:

"Trophic factors, also known as growth factors, are naturally occurring chemicals secreted by the body which sustain growth and survival of tissues, including nervous tissue such as peripheral nerves. Other chemicals have recently been discovered that also have some of these same prop-

Hand palsies can also involve individual fingers or parts of fingers. Using a knife to cut carrots can cause a pressure palsy. Paper bags are the best to use for carrying. Bags with handles, especially plastic bags with hand holes, can cause episodes just carrying a two-liter bottle of soda. Milk jugs with handles, cans of paint, briefcases, and heavy purses are also problematic. Attempting to open jars with tight lids or doing any gripping can mean trouble. Too much time at a computer can also cause problems.

Arms can be also involved from the shoulder down or just the forearm. Putting an arm across the back of a couch, carrying a purse on the forearm, resting a baby's head on an arm, sleeping on one's back, sleeping on one's side, ironing, stirring, slinging a purse over the shoulder, hammering, and doing any over-head work are all activities that can cause problems.

While crossing legs is a big culprit, any pressure behind the legs can cause foot drop. Sitting on a step or in a car or a chair where the depth is not right can cause such numbness that walking won't be the same as before you sat down. Sometimes just the amount of time spent walking can be enough to cause foot drop.

Sitting itself can be uncomfortable and requires frequent shifting to ease pressure. That can make driving uncomfortable, especially longer trips. Air travel can be miserable as there isn't enough room on airplanes to squirm around.

If getting down on hands and knees to weed the garden won't work, the next idea to compensate might be to squat. Unfortunately, that can cause leg palsies too. To read in bed can mean turning over every 10 minutes. Lying on the couch to watch TV can cause head numbness if the head is resting on the arm of the couch without a pillow. High heels can cause toe numbness.

Self-discipline is needed to avoid the known causes of pressure palsies. And people learn what their own causes are through trial and error, mostly error. Avoiding the episodes can be difficult, even when one knows it will cause problems. Generally, the problem is not the ability to do an activity in the first place. People with HNPP usually have the physical strength and coordination to do it (unless they currently have pressure palsies). The problem is stopping even though the activity is not complete or saying no to doing the activity at all. The desire to do, to help, to be functioning while having a hidden disability can sometimes win out over reason.

For those with frequent pressure palsy episodes, the need to move and change positions and pace activities and say no is a constant challenge. Though the episodes will continue to happen, hang in there!.

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erties. One such group of chemicals are called the neuroimmunophyllins (or neurophyllins). Several of the naturally occurring trophic factors, administered by injection, are currently undergoing clinical trials in diseases such as ALS (Lou Gehrig's disease), Parkinson's disease, and diabetic neuropathy, but none has reached the market yet. The neuroimmunophyllins are much earlier in their development phase but offer the important advantages that they can be given orally and may be less expensive. These molecules also have potential for the treatment of Alzheimer's disease and other degenerative neurological diseases, including CMT. Discussions have already taken place with some drug companies concerning trials of trophic factors in CMT but no decisions have been made. It is important to note that no clinical trials are currently planned and if they do occur they are probably still several years away. Nonetheless, it is exciting to think that there may be a treatment on the horizon for patients with CMT, even if it is a distant horizon."

Reprints of the full article are available. Please send a self-addressed, stamped envelope to: CMTA, 601 Upland Ave. Upland, PA 19015.

A THANK YOU TO LOYAL FRIENDS

For several years, A. Hart and Minna Wurzburg have been asking friends and family to send gifts to the Charcot-Marie-Tooth Association to recognize celebrations such as their birthdays and wedding anniversaries. This year, Minna celebrated her 75th birthday and the following people honored her with gifts to the Association. We are grateful for the Wurzburg's thoughtfulness and the generosity of their many friends and family.

Gifts were made in honor of
Mrs. A. Hart Wurzburg's 75th Birthday

Mr. and Mrs. G. F. Baer
Mary Jean Field
Mrs. Betty S. Frank
Mr. and Mrs. George B. Frank
Jeanne C. Gidwitz
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Mrs. Henriette J. Wolf
Lois Lurie and Bill Zack

A. Hart and Minna Wurzburg

Management and Rehabilitation

By PAOLO VINCI, M.D., ***Specialist in Physical Medicine and Rehabilitation,***
Professor, School for Physiotherapists, University of Rome, Italy

Editor's Note: This information comes from an Internet research source and was sent to the office by Ann Beyer, Chair of the CMTA's Research Committee of the Board of Directors.

1. Improvement of muscle strength, resistance and balance:

- If the muscle to be strengthened is only able to contract with no resistance of any kind, a strengthening exercise program is either ineffective or may cause increased weakness.
- If your muscle is able to contract with weight applied, you should see a physiotherapist to find out the maximum weight the muscle is able to tolerate. Then, it can be exercised three days a week, starting with three sets of four repetitions; this can be increased over a twelve week period to eight repetitions per set. A modest increase in strength may result.
- Strengthening exercises that use heavy weights should always be avoided: increased weakness and muscle cramps may occur. (Please refer to "In deference to Benjy Feen" at end of this article.)
- It is very important to avoid immobilization, if possible, because it causes disuse muscle atrophy as well as loss of the motor patterns you have learned, which compensate for the weakness in some muscle groups: it is advisable to be physically active for 2 or 3 hours every day.
- Exercises designed to improve coordination and balance can help, especially after a period of bed rest (e.g., after illness or a fracture requiring rest in bed).
- It is impossible to reduce the tiring of your muscles: the only therapy is rest.
- Leading the most normal life possible is the best active therapy.

2. Management of deformities and contractures:

Bone and joint deformities are caused by muscle contractures. Deformities are caused by an imbalance of strength in the muscles that act across joints. In CMT, the dorsiflexor muscles (tibialis anterior, peroneus longus, and brevis), which pull your foot up, are affected more than the plantar flexor muscles (tibialis posterior and gastrocnemius), which push your foot down.

As CMT progresses, dorsiflexion of the foot may become impossible, resulting in foot drop. Sometimes the heel will not reach the floor, (pes equinnus) while you are standing. (Your achilles tendon has a contracture.) Usually, this is associated with rotation of your foot (pes equinnus-cavus-varum-supinatum), causing a reduction in size of the area of your foot that contacts the floor. This causes pain along the outside border of your foot.

The same mechanism may occur in the wrists causing a "claw hand" deformity.

Contractures can be prevented and/or postponed by:

- Stretching calf muscles: the best stretching is obtained by standing and walking, with flat shoes and foot orthoses.
- Wear night braces: to keep your feet in the proper position while you sleep.
- Avoid the weight of blankets on your feet: a light duvet or a bar for the blankets is recommended, especially during adolescence.

When stretching is no longer effective and your heel will not reach the ground when you are standing, surgical Achilles lengthening, or other surgical procedures, is necessary. If a contracture forms in one foot only, standing position may be altered and scoliosis may develop.

Contractures may occur in your hands, leading to "claw hand" deformity. A daily self-made stretching exercise of your fingers and wrists is useful to prevent them from becoming fixed in one position.

Muscle contractures may cause deformities that can only be corrected with bone surgery, especially if they occur while the bones are still growing.

3. Promotion of mobility:

Walking is the best physiotherapy for contractures; so, even in late stages of severe cases requiring a wheelchair, daily exercises with a walker are recommended.

Avoiding becoming overweight because weak lower limb muscles are unable to support an increased body weight while standing or walking.

Orthoses for walking:

- Custom-made foot orthoses are always recommended, even in mild cases, because

CALL FOR PARTICPANTS

For an ongoing scientific project, Drs. Roger Lebo and Florian Thomas are looking for patients who have both CMT1A or CMT undefined, combined with a chromosomal trisomy, i.e. an anomaly in which patients have three copies of a chromosome instead of two, for example Down syndrome. Patients or their families who have this abnormality are asked to contact Dr. Florian Thomas, Department of Neurology, St. Louis University, 314-577-8026. Patient confidentiality will be protected.

in Charcot-Marie-Tooth Disease

they reduce impact on the bones of the foot, (in CMT, plantar muscles on the soles do not protect the bones); the best ones are polyurethane. The orthotics also help prevent calluses by keeping the foot in the correct position, and give more stability while standing or walking.

- The shoe should be higher and stronger than normal, as your ankle needs support in order to prevent sprains. I recommend a 6" to 8" high sport boot, as its synthetic, larger sole works like a shock-absorber.
- When foot-drop and tripping are a problem, an ankle-foot orthoses (AFO) is necessary. I recommend a plastic, low AFO open at the heel, with a complete sole, and the sides joined by foot orthoses.
- For people who don't want to wear AFOs, a 6" to 8" high sport boot, will do the same thing, if the back of the shoe is strengthened with strong leather or polypropylene. A latex pad inserted into the back of the shoe will allow the ankle some flexion while walking and help prevent pain in your Achilles tendons.
- If the quadriceps muscle (above your knee) becomes weak, i.e., when you are unable to straighten your leg while seated, a knee-ankle-foot orthoses (KAFO) is needed, but two canes are necessary to walk, so an assessment of strength in your upper limbs should precede this prescription.

Prevention of falls:

It is very important to avoid falls, because fractures in people with CMT are common and require more time to heal than in normal individuals, and the inactivity may cause worsening of your CMT.

Falls are prevented by proper shoes with orthoses and, overall, by paying attention:

- Watch the floor, even the lowest step can cause a trip or sprain.
- Take caution on uneven ground; a cane may improve balance.
- Beware of carpets and rugs (certain shoes "stick" to theses surfaces).
- Avoid dark places. Balance worsens when you can't see.
- Use a railing while going up and down stairs.
- Avoid haste.

In rare advanced cases, walking outside the house can become unsafe and tiring. When falls are frequent, it is advisable to use a wheelchair outside of the house and a walker inside.

An electric wheelchair or scooter will protect your upper limbs from overwork, saving you energy that may allow you to continue to walk inside at home or at work, and give you more independence.

4. Improvement of hand function:

Sensation and strength in everyone's hands decrease with exposure to cold temperatures. In CMT, cold may cause complete loss of strength and sensation, in both hands and feet, resulting in increased disability, even in moderately low temperatures.

Soft woolen gloves with a calf-skin palm can protect your hands and prevent your hand from slipping on railings, etc. However, sometimes they can restrict flexibility in your fingers.

Most people with CMT have difficulty with activities that require hand control and sensation during daily living. Special tools or simple changes in tools you use can be very helpful. An occupational therapist can give advice for your specific needs.

5. Management of respiratory failure:

- Deep breathing exercises: take a deep breath in (pushing your stomach out), hold 5 seconds, then blow out all the way. Hold 10 seconds, then repeat.
- Deep sighs should be done regularly to air those parts of the lungs that are not ventilated during quiet breathing.
- Assisted cough exercises are useful if you have the flu or respiratory problems in which you produce phlegm.
- Assisted ventilation is rarely necessary, but may be in severe cases of CMT with respiratory failure due to involvement of the phrenic nerve.

(In deference to Benjy Feen, we felt obliged to include a "second opinion" on this matter of weight training. Readers will recall Benjy as the passionate advocate of weight training (articles in the fall of 1994 and summer 1997). Dr. Vinci presents an excellent overview of CMT management and rehabilitation in the broadest scope. Benjy's success, like that of many others, suggests that people with CMT can benefit from weight training. Therefore, rather than avoiding the gym altogether or attempting to become a body builder, find your own ground. Good luck!)

The CMTA Distributes Three $35,

For the first time, the Charcot-Marie-Tooth Association has awarded three $35,000 research grants to deserving applicants working on various aspects of genetic research. The recipient of the Armington Research Fellowship is Dr. Lawrence Reiter, who is working in the laboratory of Dr. James Lupski at the Baylor College of Medicine. His project is entitled, "An EST Approach to the Identification of CMT Candidate Genes." EST or "expressed sequence tags" is a novel approach for the rapid characterization of expressed genes by partial DNA sequencing. Dr. Reiter hopes to generate an EST library from peripheral nerve myelin and axons in order to identify other genes that may be responsible for peripheral nerve disorders. His hypothesis is that the genes responsible for CMT1C and CMT4A could code for integral membrane proteins similar to PMP (peripheral myelin protein) 22, MPZ (myelin protein zero), and Cx (connexin) 32. Patients with peripheral nerve disorders of unknown etiology can then be screened for point mutations in the genes identified by this method.

Dr. Jim Lupski, Medical Advisory Board Member, (standing) will act as mentor to Dr. Lawrence Reiter at his Baylor College of Medicine laboratory during Dr. Reiter's year as the Armington Research Fellow.

The second recipient of a $35,000 grant from the Association is Dr. Mary Ellen Ahearn, who will be working with Dr. Lisa Baumbach-Reardon at the University of Miami School of Medicine. Dr. Ahearn will be working on "Genetic Anticipation in Charcot-Marie-Tooth Disease: Molecular Investigations of the CMT1A Locus." The goals of the research study are: 1) determine the size of the CMT1A duplication in affected individuals from different generations of

VOLUNTEER OF THE YEAR RECIPIENTS:

1998	Dr. Michael Shy
1997	Kay Flynn
1995	Dr. Gareth Parry
1994	Bob Daino
1993	Sue Tarpinian
1992	Rebecca Sand

Dr. Michael Shy Named 1998 Rebecca Sand "Volunteer of the Year"

Dr. Michael Shy addressed the audience in St. Louis at a patient-family conference on CMT, one of his many "contributions" as Volunteer of the Year.

The CMTA was begun as an entirely volunteer organization in 1983 and for 15 years has grown and improved primarily thanks to the efforts of our volunteers. Once in the history of the Rebecca Sand Volunteer of the Year Award, the recipient has been a member of our Medical Advisory Board—Dr. Gareth Parry, for his excellent work in editing the CMT Handbook. For only the second time, the obvious choice and consensus winner of the award is a member of the MAB: Dr. Michael Shy of Wayne State University.

Dr. Shy deserves this award for both his amazing commitment to the membership of this organization and his efforts in advancing research into the genetic causes of the disorder and potential gene therapy. As part of his efforts on behalf of the members of the CMTA, Dr. Shy established a CMT clinic at Wayne State University with the help of Dr. John Kamholz, Dr. Robert Lisak, Dr. Richard Lewis, and others. He also wrote an article for *The CMTA Report* on diagnosing CMT. A further effort on his part resulted in his flying to St. Louis to present at the regional conference held at the Shriners' Hospital there. He also hosted a conference on CMT in Detroit in October of 1997.

In his efforts to promote research on CMT, Dr. Shy has mentored, for 2 years, Dr. Agnes

000 Grants for 1998-99

Dr. Mary Ellen Ahearn (left) and Dr. Lisa Baumbach-Reardon, University of Miami, are studying genetic anticipation in type I CMT.

the study families; 2) further evaluate the presence of possible DNA rearrangements at chromosome 17p11.2 in affected members of the CMT1A kindreds; 3) evaluate the incidence of a novel polymorphism in the African-American CMT population, and determine possible correlations with disease severity; and 4) determine the statistical significance of clinical data collected using the CMT1A Symptom Score (CSS).

The final recipient of a $35,000 grant, made possible by a generous gift from the Buuck family, is last year's recipient, Dr. Agnes Jani of Wayne State University. She is working in the laboratory of Drs. Michael Shy and John Kamholz. The goal of Dr. Jani's project is to use the techniques of molecular biology to correct the genetic defects in CMT1B Schwann cells and, thus, to lessen the clinical effects of the disease. Successful gene therapy will require developing techniques to introduce genes into Schwann cells in the peripheral nerves. To do this, Dr. Jani is working on using adenoviruses, common respiratory viruses. These viruses can be altered into adenoviral vectors that no longer divide and cause disease, but that can be used to introduce new genes into cells. This second year of Dr. Jani's study continues the development of methodology that may ultimately lead to effective gene therapy in humans with CMT.

The continuing support of the CMTA membership is needed to fund these incredible research studies. Please be generous when you respond to the Armington Research Challenge. Do take it personally.

Editor's Note: Watch for a "reader friendly" version of the grant proposals in the next newsletter.

Jani, who is working on the use of adenoviral vectors in P_0 knockout mice as animal models of CMT. This year, he has spent hundreds of hours working on the Third International Conference in Montreal, Canada, scheduled for October, 1998. This conference will bring together hundreds of researchers from around the world to share their discoveries and their goals for CMT research. Dr. Shy has worked tirelessly with the New York Academy of Science to get all of the researchers lined up for the conference. He has submitted a very strong grant application to the National Institutes of Health in an effort to get funding for the conference.

All of the work for the CMTA and CMT research comes after his attention to his clinical practice, his family (a wife and two children), and his own "paperwork." But Mike is no stranger to giving to others. During his college years, he volunteered with VISTA (Volunteers in Service to America) and was a teacher in a "Get Set" program in the Philadelphia school system. He was on staff at the Thomas Jefferson Medical College before coming to Wayne State. His list of honors and awards covers three pages of a curriculum vitae and his published articles, book chapters, case reports, and presentations cover a daunting five pages. To say that Mike has devoted his life to his work would be to understate the obvious.

Dr. Agnes Jani and Dr. Michael Shy pause outside the lecture hall at the American Academy of Neurology. Dr. Jani will continue her work in Dr. Shy's lab this year as the result of a grant from the Buuck family.

The Charcot-Marie-Tooth Association is pleased to add to Dr. Shy's list of awards, "The Rebecca Sand Volunteer of the Year Award" for 1998. Congratulations! And thank you for all that you have done and continue to do for the CMTA.

ABC's of the Internet

By STEVE SCOFIELD, CMTA Board of Directors

Okay, this is how it is: Everybody is on the Internet having fun except for you because you just know that the computer is going to hurt you or you will make it blow up. Perhaps you can be convinced otherwise. The message below is from my mother (who is a member of the CMTA) and trust me, if she can conquer the computer and the Internet, so can you.

My mother writes:

One day as I was contentedly reading a mystery, I was informed that I was to receive a personal computer. "No, no, I don't need a computer. . . . I don't want a computer," I exclaimed. Having heard the foreign language "computerese" sometimes spoken by members of my family, I knew I would be lost if I ventured into that alien territory. But, the computer arrived anyway and was installed, and I was told, "There, go to it!"

Later, when no one was around, I put on my little red cape and hood and timidly went out adventuring on the Internet! Oh, my, the wonders to behold. . . . museums, libraries, shopping malls, bookstores, and people! Oh, yes, people!

I found one of my roommates from college who had been a bridesmaid at my wedding 50 years ago! Next came corresponding through e-mail, then finding games to play, newspapers, weather reports, views from outer space, views of Mars. Then learning about an ongoing auction of tens of thousands of items including antiques and curios. I discovered that almost anything could be found with a little exploration of the paths and byways of the Internet.

Here is what you need to do to join me on the Internet.

1) Purchase a computer. You should be able to find a very good used computer with a color monitor and a modem in the newspaper or at the local computer store for less than $500. Tell them you would like at least a computer with a 486 processor running at 66 Mhz (whatever that means), a 15-inch monitor, and an internal modem with a capacity of at least 28.8 kbps (huh?!?).

2) Choose an ISP (an ISP is the way people in the know refer to an Internet Service Provider.) AT&T, MCI, and Sprint are all ISPs. In addition, there are many hundreds of other local companies that provide access to the Internet. Check your yellow pages or ask a neighbor who they use. Call one and set up an account. The standard price that just about everyone charges is $19.95 per month for unlimited access to the Internet. (Trust me, you want unlimited access.)

3) Get help. Get someone—the neighbor, the neighbor's kid, your daughter—to come over and get the thing up and running for you.

That's it! Now, you can join others in the CMT chatroom, go shopping for just about anything, e-mail your friends, and on and on. . . .

Editor's Note: In the next issue of the newsletter, Steve will discuss the CMTA web page and other mysteries of computer use. Steve is a professor in the department of accounting and technology at Texas A&M University in Kingsville, TX.

THE CMTA'S HOMEPAGE IS
www.charcot-marie-tooth.org

EMPHASIS ON ABILITIES

Located in Minneapolis, MN, the Courage Center is a nonprofit organization that works with 18,000 children and adults with disabilities each year. More than 70 services are offered that reach all aspects of a person's life, including a residential independent living program, physical and occupational therapies, drivers' education, job placement, spiritual therapies, family support, sports and recreation, and much more. Courage Center partners with people with disabilities to provide an environment where individuals can learn to live as independently as possible. At left, Jason Gerling participates in one of Courage Center's computer training courses.

CMTA and Rancho Los Amigos Host Conference on CMT

A conference on Charcot-Marie-Tooth Disorders will be held on Saturday, June 27,1998, at the Rancho Los Amigos Medical Center, 7601 E. Imperial Highway, Downey, CA.

The conference will begin with registration and continental breakfast from 8:00–9:00 am. The first presentation will be an overview of ongoing research programs on CMT, followed by a presentation on orthopaedic foot and ankle surgery by Dr. John Hsu, the conference organizer. At 10:15 a representative of Athena Diagnostics will discuss the DNA blood tests that are currently available to diagnose CMT.

At 10:45 am there will be a coffee break followed by the morning workshops on pain management, occupational and physical therapy, and alternative therapies. At noon lunch will be provided in the cafeteria dining room.

At 1:00 PM there will be a presentation on gait analysis and the work of the gait clinic at Rancho. Following that there will be a discussion of the use of orthotics and the role of a pedorthist, a specialist in shoe-fitting and design. Following a soda break the workshops will again be offered. The conference will conclude with a presentation on legal ethics and concerns and closing comments by the staff of the CMTA.

The cost of the conference is $40 for current members of the CMTA and $50 for nonmembers or those who register on the day of the conference (space permitting.) The medical center is barrier-free and there is parking on the ground floor of the Support Service Annex. Segments of the program are in the Annex building. The capacity of the conference is 150 persons.

Commercial shuttles (Super Shuttle and World-Wide Shuttle) are available from the airports. There are many hotels and motels in the area of the medical center, including Days Inn, Embassy Suites, and Firestone Motels. Transportation from the motels would be by taxi or private car.

Return the registration form and payment to:
Downey Conference,
CMTA, 601 Upland Ave.
Upland, PA 19015

REGISTRATION FORM:
DOWNEY CONFERENCE, JUNE 27, 1998

Name ______________________________ Daytime phone number______________

Address ______________________________

____

Names of other attendees ______________________________

____

Price per person for members of the CMTA is $40; $50 for non-members. Number of attendees______________ Total Amount ______________

☐ Check enclosed ☐ Visa ☐ MasterCard

Card number______________________________ Expiration Date ______________

Signature ______________________________

Please mail registration form and payment to: Downey Conference, CMTA, 601 Upland Ave., Upland, PA 19015.
Or return credit card payments by fax: 610-499-7487.

The Little Body That Could

By **LISA M. CURTIS**

I have fond memories of sitting in my mother's lap and listening to the rhythmic quality of her voice as she read stories to me. The one that I will always remember is *The Little Engine That Could*, a simple story really about a train who psychologically convinces himself that he can do anything by chanting, "I think I can*I think I can*I know I can*I can." Following this axiom is how I have lived the past 20 years of my life, since being diagnosed with the progressive disorder, CMT. I have decided to share this story in hopes that my experiences will help others appreciate that life is all about understanding how the perspective you take impacts the path you follow.

I was a smart, loving child, full of life and wonder who was often referred to as "Grace." "Grace" is an endearing term for a child who is clumsy. I am sure my friends and family were not trying to be cruel or unkind, but most people have the perception that females should move with grace and ease. I did not. I can also remember my feet burning constantly from the patent leather shoes that were the rage back then. Once, I remember a look of shocked faces as my father took me off of a pony ride, my leotards torn and my leg bleeding, my face all smiles as I had no idea that my leg was badly cut, I had no sensation at all.

At age 8, I wanted to be a ballerina. All I really remember about that time was the big dance recital where I fell in front of all my family, quickly getting up to avoid further embarrassment. Later, in middle school, there was the constant teasing from fellow students and contempt from gym teachers who thought I was unmotivated because I had the slowest mile in the class.

In high school I was subjected to peer pressures to look, act, and do as everyone else did. Platform shoes were the proverbial straw. I had to have a pair, 3 inches high! Sitting on the ground holding a badly swollen ankle, I should have wondered why shoes were such an important basis for defining one's self, but I didn't. I wondered for years having one calamity after another, all in an attempt to fit in, to march to the beat of everyone else's drum, not realizing that mine was a different beat.

Memories of my freshman year of college are forever embedded in my mind. I had decided to take up a fitness program with my friends that involved jogging, calisthenics, and swimming. After weeks of trying to keep up, saying to myself, "I know I can," I fell and seriously hurt my ankle and knee. A few days later I found myself in the office of a very famous orthopedic surgeon who had just invented the plastic hip joint. After examining me, the doctor asked several of his associates to examine my feet and legs. I remember feeling very strange as a parade of doctors walked in and gazed upon my lower limbs. The minutes seemed like hours as I felt like a fish in a bowl. Finally, they all left except one who politely extended his hand and introduced himself as Dr. Hopmans. I later learned that Dr. Hopmans was a Canadian physician who had treated several cases involving rare muscular disorders. I remember shaking my head as if in a trance as I agreed to undergo a series of painful nerve conduction studies and other medial tests.

> *As a person with CMT you must redirect your course in life as the disorder progresses. This altered perception will allow a new course to be charted and will lead to greater self-fulfillment.*

Three weeks later, I was diagnosed with CMT. I should have felt relieved and validated after years of struggling to live life at the same pace as those around me. Instead, I left the doctor's office determined that I would live my life exactly as I had before, in fact, I tried even harder to prove that I was "The Little Engine That Could."

Later that year, I realized that I wasn't the only one in my family who had this CMT or the attitude of "I can." When I told my friends and family about my diagnosis, I discovered my grandmother had CMT since she was a child. The entire time I was growing up my family never mentioned the "family disease," or who was affected. Why? I'm not exactly sure; my guess is they were ashamed or embarrassed.

Although my grandmother had great pride and inner strength, I believe she never accepted living a life with CMT, and insisted on living her life being the "Little Engine That Could." She

OF INTEREST

Worldview Pictures is a television production company based in Boston and London. They are currently researching a documentary film for the Discovery Channel which will assess the growing concern over the issue of genetic discrimination. They are approaching people whose genetic test results have caused problems for them or their family—in particular, problems with obtaining insurance, or in finding employment.

If you have personally experienced some kind of genetic discrimination, they would be very interested in talking to you about your experiences. All information will be treated in the strictest confidence. Please contact Dr. Stephen Trombley at 1284 Beacon St. #420, Brookline, MA 02146, telephone: 617-232-1537, email: strombley@worldviewpictures.co.uk never asked or accepted help from others. Instead, she took care of everything and everyone, except herself. To admit that she needed help would in her eyes be a sign of weakness, inadequacy, and failure. She carried this attitude to the limit by lining her home with rails so that she could avoid using a walker, cane, or wheelchair. She would lunge from one side of the room to the other, grasping the rail and steadying herself as she made her way around. Later, as the disease progressed, her pride would not allow her to become mobile with the aid of a wheelchair. Instead she remained home, often alone, throughout the rest of her life.

I loved my grandmother, yet I never understood her stubborn attitude. As a child her behavior seemed so silly to me. However, after living with CMT for the past 20 years I understand how difficult it is to live a life different from others when the disease you have slowly changes you from an able-bodied person to a disabled person over a period of many years. Often it is only after an injury or several embarrassing moments that a person realizes he or she must make the necessary adjustments in order to accommodate the illness.

Suddenly, grandmother's actions made sense to me. I found myself face down in front of a room full of people; their eyes gazing down upon me as I started to rise, only to have my legs buckle and give way again beneath me. I had pushed myself to exhaustion, again, trying to keep pace with those around me, not understanding until that moment that to choose a different route would have not only been acceptable, but would have resulted in far greater personal success for me.

Later, I realized all the repeated accidents, embarrassing moments, and heartache could have been avoided if I had made the necessary adjustments in my life to accommodate my condition. The little engine that was pushing me to succeed at all costs was on the wrong track. As a person with CMT you must redirect your course in life as the disorder progresses. This altered perception will allow a new course to be charted and will lead to greater self-fulfillment.

Now, when I read this story to my children I emphasize that even if the Little Engine can't go as far and as fast as the steam engine, he is actually valuable to those who carry him within their hearts.

ABOVE AND BEYOND: GEORGE PUELLO

The award called "Above and Beyond" was conceived to recognize members of the CMTA community who went out of their way to advance the mission of the association. This time, the recipient of the honor was not even connected to the CMTA prior to his sudden and complete commitment to our Westchester Broadway Theater fundraiser.

George Puello is the artistic director of the Westchester Broadway Theater (WBT) and as such, is no stranger to fundraising for a local homeless shelter, AIDS organizations, and other community-based groups. Before March of this year, he had never heard of CMT or the CMTA. But in a rather roundabout way, that soon changed.

Paul Flynn's cousin, Nancy Homyak (one of the 25 family members who have CMT) has a daughter named Lindsay Budnar, who is 14 years old and a member of a singing group called the Dreamcoats. Nancy had seen a production called "Kids with Heart," which raised money for children with Duchenne's muscular dystrophy and she thought it would be great if the Dreamcoats could do something like that to raise money for CMT.

Enter George Puello. He happens to choreograph the Dreamcoats in addition to his work with the theater. Nancy Homyak arranged for George to meet Paul Flynn, who explained the mission of the CMTA and the need to raise money for the organization. George embraced the concept, agreeing to use one performance of his musicals, "Scattered Thoughts" and "Parusia," to raise money for the CMTA. In addition, he allowed the Westchester County support group, headed by Kay Flynn, to run a silent auction at the performance to raise even more money.

Over 100 people bought tickets for the WBT event and bid on auction items, resulting in more than $5500 in profits for the CMTA. Through all of this, it was the overwhelming generosity of George Puello who opened the theater to the CMTA members, donated his original works for the performance, and emceed the program that made the night so spectacular. With luck and the continuing kindness of George, the fundraiser may become a yearly event.

George Puello and Lindsay Budnar were two of the principals in making the Westchester Broadway Theater fundraiser a financial and artistic success.

CMTA Support Group *News Flash!!!*

We are very excited with everything that's been happening out there with support groups!! Here are some newsworthy items:

■ The Westchester Support Group in the New York area sponsored a benefit at the Westchester-Dinner Theater. It was an evening of great theater, talented performers, and a silent auction. After expenses, the fundraiser netted more than $5000 for the CMTA. Way to go, Kay Flynn and all of the people who made this event possible.

■ Did you know that the Missouri/Eastern Kansas Group has their own library now? Joy Rushfelt of that group has collected many pieces of literature on the subject of CMT. Also, there is a "Balance Board" that can be loaned to members of the group, before they build one themselves. What a great idea!!!

■ Guess what?? Three new groups have had their first meetings, and they are excited!! Bob Budde in Lexington, KY, in the beginning of April. Debbie and Brenda, two R.N.'s in Michigan have started a college campus support group. Nora Burrows in Archdale, NC, had her first meeting and is already planning her speakers for the next eight meetings. I call that commitment!!!

Dot Cain in Greenville, Ohio leads another group that recently had a meeting. Although that group has existed for 3 years, they are trying to get new members to increase their attendance. If you live in that area, try to make a meeting!

■ All you folks out there in the Berkeley, CA Area: There was a first meeting May 23 at the Berkeley Public Library. Ruth Levitan is coordinating events with assistance from Miriam Diaz. Dr. Kirk Wilhelmson was the speaker.

■ Rosemary Mills in Benson, MN, had Dr. Gareth Parry speak at her meeting on May 9th ! Must have been a good one.

■ The big news from the national front is that the CMTA will be having a meeting in June with regional support group leaders. Estelle Benson of the Guillain-Barré Syndrome organization will speak to us. Estelle has had great success with support groups all over the world! Our goal is to develop a more structured support network, so that we can maximize our efforts and serve more people. We are all in this together. . . . families and friends, too! Stay tuned for news of the big support group summit, at a later date.

Thanks to all who lead, or participate, in CMT support groups. Our challenges aren't as great if we have someone to share them with.

In peace and health. —*Krista Hall, R.N.,*
CMTA Support Group Leader Liaison

CMTA Support Groups

Alabama/Greater Tennessee Valley
Place: ECM Hospital, Florence, AL
Meeting: Quarterly
Contact: William Porter, 205-767-4181

California–Berkeley Area
Place: West Berkeley Library
Meeting: Quarterly
Contact: Ruth Levitan, 510-524-3506 or Miriam Diaz, email miriam.diaz@deskprod.com

California–Northern Coast Counties (Marin, Mendocino, Solano, Sonoma)
Place: 300 Sovereign Lane, Santa Rosa
Meeting: Quarterly, Saturday, 1 PM
Contact: Freda Brown, 707-573-0181

California–Napa Valley
Place: Sierra Vista Convalescent Hospital, Napa
Meeting: Quarterly
Contact: Betty Russell, 707-253-0351

Florida/Boca Raton to Melbourne
Place: Columbia Medical Center, Port St. Lucie
Meeting: Quarterly
Contact: Walter Sawyer, 561-336-7855

Kentucky/Southern Indiana/Southern Ohio
Place: First United Methodist Church, Lexington, KY
Meeting: Quarterly
Contact: Robert Budde, 606-255-7471

Massachusetts/Boston Area
Place: Lahey-Hitchcock Clinic, Burlington, MA
Meeting: Every other month, the first Tuesday
Contact: David Prince, 978-667-9008

Michigan/Detroit Area
Place: Beaumont Hospital
Meeting: Three times each year
Contact: Suzanne Tarpinian, 313-883-1123

Michigan/Flint
Place: University of Michigan, Health Services
Meeting: Quarterly
Contact: Debbie Newberger/Brenda Kehoe 810-762-3456

Minnesota-Benson
Place: St. Mark's Lutheran Church
Meeting: Quarterly
Contact: Rosemary Mills, 320-567-2156

Mississippi/Louisiana
Place: Clinton Library, Clinton, MS
Meeting: Quarterly
Contact: Betty Aultman, 601-825-5626
Julia Provost, 601-825-6482

Missouri/Eastern Kansas
Place: Mid-America Rehab Hospital, Overland Park, KS
Meeting: First Saturday each month except January, July, and September
Contact: Ardith Fetterolf, 816-965-0017, fax: 816-965-9359

Missouri/St. Louis Area
Place: St. Louis University Medical Health Ctr.
Meeting: Quarterly
Contact: Carole Haislip, 314-644-1664

New York (Westchester County)/ Connecticut (Fairfield)
Place: Blythedale Hospital
Meeting: Monthly, Saturday
Contact: Kay Flynn, 914-793-4710

North Carolina-Archdale
Place: Archdale Public Library
Meeting: Quarterly
Contact: Ellen (Nora) Burrows, 336-434-2383

North Carolina/Triangle Area (Raleigh, Durham, Chapel Hill)
Place: Church of the Reconciliation, Chapel Hill
Meeting: Quarterly
Contact: Susan Salzberg, 919-967-3118 (evenings)

Ohio (Greenville)
Place: Church of the Brethren
Meeting: Fourth Thursday, April-October
Contact: Dot Cain, 937-548-3963

West Virginia/North Central
Place: VFW Conference Room, Elkins, WV
Meeting: Quarterly
Contact: Joan Plant, 304-636-7152 (evenings)

Ask the Doctor

Members of the CMTA's Medical Advisory Board answer questions from readers.

Dear Doctor:

I have CMT and a lot of respiratory problems (allergic broncho-pulmonary aspergillosis) and pneumonia, off and on. My new doctor gave me ciprofloxacin and it seems under its warnings that maybe I shouldn't take it. Can you respond?

The Doctor replies:

Problems with ciprofloxacin are not clearly established. Only two patients with CMT are known to have had problems with it, but clearly it should be watched. When any new medicines are begun, patients should keep notes on their health and inform their doctors of any changes they notice.

Dear Doctor:

Is there a possible connection between CMT and intestinal problems? I have had a long series of intestinal blockages in spite of very careful eating habits. No other medical conditions have been connected to this condition.

The Doctor replies:

No, not really. The intestines are connected to the autonomic nervous system, which is not part of the system affected by CMT. A gastroenterologist might be able to provide more insight into the problem.

Dear Doctor:

Without giving you a lot of medical and or personal history, I do have a question. I have CMT1A. I have areas on my body, primarily my feet, thigh, and hands that are numb on the surface. Is this just a "skin thing" similar to the feeling in your cheek after dental work? I feel the pressure, but not the actual sensation on my skin. I haven't seen this mentioned in the newsletter before and wonder if it's connected to my CMT.

The Doctor replies:

Yes, decreased sensation, more often noticed in the feet than in the hands, can create a feeling of numbness. The sensory nerves are located on the surface of the skin and their loss of function can create the lack of feeling you describe.

Editor's Note: A letter from a family member of a young man who died earlier this year and had been taking Zoloft was posted on our bulletin board and caused a great deal of concern over the use of the drug. Two members of our Medical Advisory Board have responded to the sister's letter which follows.

The sister writes:

My brother, Mark, died on March 1, 1998, suddenly and unexpectedly. He was only 38 years old when he died. The story begins on February 4th when he saw a doctor about his anger. His disease has always been a problem for him, with a lot of pain in his feet and legs. Nevertheless, he worked on our family's Christmas tree farm. The doctor who saw him prescribed Zoloft to help him with his anger and when CMT was mentioned, the doctor said that Zoloft was one of the safest antidepressants available.

Two weeks into his treatment, Mark began having dizzy spells and shortness of breath. He went back to the doctor to discuss the problems and was asked how his anger was doing. He said there had not been a noticeable change, so the doctor doubled his dose of Zoloft. Six days after the increase, Mark became increasingly ill and fainted while working in the woods.

He was taken to the emergency room complaining of chest pain and difficulty breathing. They did an EKG and found nothing wrong. He got a nosebleed while there and when they did a blood test, his oxygen level was down. They gave him oxygen and he felt well after that. Since Mark felt well after the oxygen, they concluded that he had acute bronchitis and they sent him home with antibiotics and an inhaler. He seemed better that night and felt well the next morning, so his parents left for the weekend.

By that night, however, Mark was complaining of some chest pains and his legs hurting. The next morning, he was very ill and was throwing up most of the day. When his family called the ER, doctors said the antibiotics were making him ill and to give him soda for his upset stomach. My sister wanted his medicine changed, but they said his regular doctor should make that decision. Unfortunately, his family never got through to his doctor.

By Sunday morning, Mark was moaning and breathing with great difficulty. He was on the floor on all fours trying to catch his breath. Suddenly, he stopped breathing and they began CPR on him while 911 was called. Emergency technicians administered an IV and continued the CPR until an ambulance could arrive. They used shock paddles and got a short pulse, but decided to transport him to the hospital. He did not make it and was pronounced dead, never having recovered from the collapse on the living room floor.

We know now that Mark's condition was caused by several bloodclots that were working their way up from his legs and adhering to his lungs, causing the shortness of breath.

DEAR DOCTOR

continued from page 19

The Doctor's Reply:

All of us connected with the CMTA send our heartfelt condolences to the members of Mark's family. His illness and death was recently reported on the Internet and sent to us for our review. We thank the family for the report, but we are concerned that this tragic event may not have been due to the medication, but to some other cause. We don't think it should stop the use of the newer antidepressants, and for that reason, we think this note of caution is warranted.

The reason is that Zoloft and the other new antidepressants have given enormous relief to those who suffer from depression. This includes many with CMT along with the other disorders that cause muscle weakness.

Major depression is associated with loss of pleasure in life and a sense of hopelessness and withdrawal from friends and family. When untreated, these negative feelings can lead to self-destructive behavior, suicide being the most extreme example. CMT causes frustration enough, and right now has no specific treatment, but depression can almost always be effectively treated, and should be.

Our patients with CMT also report that they become discouraged as they try to obtain the health benefits they deserve or attempt to interact in any way with our difficult and inadequate health care delivery system. These frustrations alone may lead to unhappiness and even depression that may actually produce more suffering than the CMT itself. For these reasons, we want to do what we can to see that effective treatment of serious depression continues.

We do know of several patients with CMT who have lost weight and become weaker while taking Zoloft, but we have not heard of any deaths in CMT patients after starting the medication. However, because of the chance of weight loss, we recommend close attention to weight and caloric intake particularly when starting a new medication that might affect appetite.

We also recommend that open and public discussion continue on how medications affect patients with CMT. Support groups and the Internet are ways to share this information with each other. But it is important to remember that while shared information is helpful, there is no substitute for a private conversation between an individual patient and his/her doctor. This is because each individual is unique and his/her own doctor is the only one who knows his/her blood pressure, heart status, and the level of blood counts and liver function. Patients and doctors should know each other better and understand they must unite against CMT.

We need to communicate better with our doctors. The ability to communicate with a doctor or nurse who can help is the best "health insurance."

—*Members, The CMTA Medical Advisory Board*

Pain Clinics: *What you should know before you go*

Many patients with CMT complain that their primary care physician does not understand the chronic pain associated with the disorder, and sometimes tries to tell them they do not have any pain. Pain clinics are relative newcomers to the world of medical care. Their sole task is to treat various types of chronic pain, making them logical destinations for CMT patients. Clinics vary, offering inpatient or outpatient services, and single or multidisciplinary therapies.

Looking for a pain clinic? Consider the following:

CARF accreditation. The Rehabilitation Accreditation Commission (known as CARF) encourages high standards through a process including site visits, record audits, and staff review. For a free list of CARF accredited pain clinics, send a self-addressed, stamped envelope to CARF, 4891 East Grant Rd., Tucson, AZ 85712.

A multidisciplinary approach. Good pain clinics treat the psychological issues that pain raises as well as the physical aspects. At least one physician, one physical therapist, one psychologist, and one nurse should be on staff.

Individualized exercise programs. Exercise is an important part of a pain treatment plan, but not all exercises are appropriate for everyone. Look for a clinic that individualizes exercise programs based on your abilities.

Health insurance coverage. Unless you can pay your expenses outright, make sure the pain center is covered by your health insurance. Some policies cover only CARF-accredited clinics.

Convenient location. For a list of pain clinics near you, send a self-addressed, stamped envelope requesting the list to the American Pain Society, 5700 Old Orchard Rd., Skokie, IL 60077.

Pain clinics do not seek to provide a quick fix. Sometimes they can eliminate pain, but not always. The goal is to minimize suffering so that pain is manageable and not the focal point of one's life.

Letters to the Editor:

Dear CMTA,

In 1977, I was examined at the Mayo Clinic in Rochester, MN, and after a thorough examination, was diagnosed with CMT. Roughly, the disease had shown up when I was 9. From 9 until 16, I wore AFOs. At age 16, my ankles were fused, and at age 21, my AFOs were extended to go over my knees. At 23, I started using a wheelchair only for outdoor/sporting events. For the last 5 years, I have been wheelchair bound. I was born legally blind with optic nerve damage. This condition has not changed and never will. Last year, the Mayo Clinic labeled my disease as CMT6.

Dr. Griffin at Johns Hopkins examined both of my parents. Both have recessive traits for CMT but no other family history of the disease. Dr. Griffin believes that it is the combination of my parents' recessive traits that has resulted in my current condition.

The reason I am sharing this information with you is to ask for your help. Are there other people who have CMT6 or similar physical problems? Do you know of any doctor in the world who is conducting research on this?

Please send your responses to the office and they will be forwarded to me.

—S.S., Scottsdale, Arizona

Dear CMTA,

I enjoy your newsletters and find them very informative. I am particularly interested in information on making life with CMT a little easier. I am 71 years old and was diagnosed with CMT in 1993 following my son's diagnosis. I had long suspected that I had a problem, but didn't know what it was. I could never squat, nor run very well. In 1980, I had surgery on my left foot to lower the arch and correct hammer toes. The podiatrist that did the surgery never mentioned CMT. I was in a leg cast for 6 weeks after the surgery, but it was well worth it, as it relieved the pain in my foot and enabled me to find shoes that fit.

One of the things I have done to make life easier is to have wider, lower steps installed at the front and side entrances of our home, with a handrail added to the side entrance, as well as to the stairs leading to our basement. When I began having trouble getting out of my car, I added a wedge-shaped foam cushion, which makes it much easier. I can't peel vegetables very well and have found that boiling potatoes before peeling them helps a lot. These things may not seem like much, but every little bit helps.

Our 42-year-old daughter was just recently diagnosed with CMT as well. She is very upset about it, as her left foot is completely numb and she has almost constant discomfort in her left leg below the knee. She is afraid that she will eventually have to give up some of the things that she most enjoys, such as travel and her beautiful home (all of the bedrooms are on the second floor), although I have tried to convince her otherwise. Our son who has CMT also has insulin-dependent diabetes. His gait is rather severely affected by the CMT, but his doctor has told him that he cannot wear braces, as the pressure of the braces might injure his feet. I would like to hear the opinions of others who have both diabetes and CMT regarding the use of braces.

—M.M, Oak Ridge, TN

Dear CMTA,

I'd like to let you know about a great pair of scissors that I found in a store called Brookstone. They cost $12.00 plus tax. These scissors have a spring that opens so all you do is squeeze your hand and the paper cuts. Great for people like us who have trouble using scissors. They are called Oxo Good Grips kitchen scissors and can be used by either right- or left-handers. They are made by: OXO International, 1-800-545-4411.

Keep up the great work with the newsletter. I read it cover to cover as soon as it comes in the mail. I also went to Detroit for Dr. Shy's CMT study. What a great group of doctors!

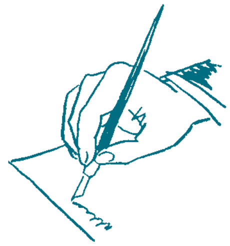
—I.Z., Saylorsburg, PA

Editor's Note: We did an article on Good Grips back in the Winter, 1995, newsletter. They are manufacturers of many kitchen utensils and all are designed with comfort and function in mind. One of the inventors has arthritis and all of the handles are cushioned and easy to hold.

Dear CMTA,

I read the letter from S.L. in the winter report. I am a 39-year-old man with Type I CMT. My feet used to look similar to the ones pictured in the winter report on page 22. I, too, had many reservations about having my feet "done." I had 14 tendon transfers done on each foot (so many because each toe was worked on), the wedge taken out of my arch, the great toe fused, the heel bone cut off and shifted, and the inner/outer tendon transfer. Yikes.

I have no regrets. I started out with bad balance, then I found myself falling over for no



WRITE TO US!

Pat Dreibelbis, Editor

The CMTA Report

CMTA

601 Upland Ave.

Upland, PA 19015

The CMTA reserves the right to edit letters for space.

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LETTERS TO THE EDITOR

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apparent reason. I also sprained my ankle a lot. What really bothered me the most was the pain I used to get in the arch under the foot. That was from the flexor tendon that runs from the heel to the arch, creating the arch. That used to be bad! My doctor detached it and simply moved it back towards my heel a little. That alone was worth it. I had one foot done at a time (highly recommended!) I took 1 month off of work each time, though I could have come back in 3 weeks. I was on crutches for 6 weeks and a cast a total of 8. I then wore one of those boots for another 6 weeks.

I waited 1 year between operations. It really took about 6 months to feel "normal" again. It was tough and sometimes frustrating, but now I kick myself for waiting so long to do it. It was the thing to do for me. Only you can decide if it is for you. Now, I walk with no pain at all. My ankles are a little less flexible, but not so I'd notice. I did not fall down again. I think the best part was when I could finally look at my own feet and see two normal-looking things down there! Except for a few scars....

If you want to ask me anything about the operations, please e-mail me at cougard@prodigy.net.

Editor's Note: The e-mail address for S.L. who wrote the original letter about foot surgeries was incorrect in the winter issue. Her correct email address is leasure1@ix.netcom.com

Dear CMTA,

My son, who is 10 years old and in the fifth grade, was only diagnosed in January. We have struggled all year long and he will still have to go to summer school so that he might have a chance of passing into sixth grade. We recently moved into a new school district and they have never heard of CMT. I can't really complain because I never had either, until it hit home. My son also has ADD. His right hand has lost its muscle and he cannot lift his wrist or fingers. It is his dominant hand. I wish the school district would understand CMT and that this is going to be the way of life for him and would do something constructive. They have said they will start doing oral testing, but that was 3 months ago and they haven't yet. I did get them to purchase a keyboarding program to teach him some skills.

I wish the CMTA would put together some information on CMT and children. It might help with school districts. I did go to the web and print out the information for the principals, teachers and OT. One thing that should be mentioned in the pamphlet is that kids with CMT shouldn't be picked on or called names. My son has come home crying more than a few times because of this. Sometimes the problem is almost more emotional than physical. We need to help these bright and wonderful kids all we can.

—*T.L*

Dear CMTA,

In response to the request from R.K. in the summer 1997 issue (I'm a little late with my reading), I would like to contribute some stories that can make him/her feel good and find comfort, and humor, in some misfortunes that befall all people with CMT.

I was always falling as a child, but I learned to do it with grace and style. It was a necessary defense when, the first day of my freshman year in high school, I looked to the library table where my friends were seated, and proceeded to fall down eight steps. I was escorted through the main hallway in a wheelchair to the hoots and hollers of upper classmen. I very stylishly held my right hand up in a royal manner, bowing my head and waving as if I were a princess in a horse-drawn carriage. I also held the distinct advantage of failing ninth grade physical education with a numerical grade of 10. Even before I was diagnosed at the age of 17, I knew enough about my body to refuse to get up on the balance beam, despite the numerous "spotters" the teacher offered. Fortunately, the principal did not average the mark into my grades, as it really would have dragged down the 95 I got in English!

I nearly drowned after my high school prom when we went to the beach the next day and I could not get past the breaking waves. All of my friends watched and laughed from the shore as I was repeatedly dragged by each pull of the undertow. "That Elissa! She is such a clown! Look at her pretending to drown in 2 feet of water!" I wish I could say that a cute lifeguard saved me, but finally one of my girlfriends saw I was in a panic and pulled me onto the sand.

I was taken out as a "shipoopy girl" in the high school production of "Music Man" when I twisted my ankle and landed in the orchestra pit. I was standing still at the time. I always dreamed of a career in acting and had auditioned for the American Academy of Dramatic Arts. However, I was diagnosed with CMT in April of my senior year and the triple arthrodesis surgery was performed on my right foot the next fall. There went my stage career. Suppose I got the part of Auntie Mame, or something! Imagine opening and closing each act going up those stairs. . .

■ SURVEY

Dorothy Gosling, RN, is working on a survey on CMT and would appreciate more responses than the 200 she has already received through her web page. If you would be interested in completing the survey and helping with the gathering of information about CMT patients, please contact Dorothy through e-mail at dgosling@netcom.ca or write her at CMT Survey, Centenary Post Office, Box 72042, 2898 Ellesmere Road, Scarborough, Ontario, Canada M1E 5G4. The survey is 8 pages long, but don't let that scare you!

The next 25 years I spent working as a dental assistant. How could I get in trouble just sitting in a chair? I fell in a lot of patient's laps, tripped over hundreds of wires, walked into a great deal of sharp instruments. I pulled out a portion of a wall when I got stuck on the wall-mounted blood pressure gauge that hooked under my arm in a fall, knocked over a free-standing cabinet by standing next to it, and hung by my arms from a storage cabinet door when the step-stool collapsed on my legs. One dentist fired me because every morning I tripped over the humidifer in the hallway. So, this one day, I was carrying his hot coffee…

I told him as I left that my last job had been with the Russian Ballet!

I have been married twice and did not fall during the ceremonies or the receptions, although it was everyone's fear. I did, however, have to wear children's shoes at both weddings. The band played the "Buster Brown" theme song instead of the "Wedding March."

The only time I was really concerned with falling was when I was pregnant with my two children. I nearly lost both sons because of falls I took in the first trimester. I had to stay in bed for 2 weeks when I fell on the ice 4 weeks into my second pregnancy. Now that my children are out of my body, they are gratefully on their own.

My 9-year old son has been diagnosed with CMT. He was 5 when we discovered my fears were real. I cried, at first. Then, I thought about my life. I only had to quit my job and go into braces 2 years ago. That's 14 years longer than the doctors had predicted. They said I'd be in a wheelchair and I do use one now. I use it for long distances or to shop. But, sometimes, I just get in it and do an impersonation of Joan Crawford in "Whatever Happened to Baby Jane?" after she was fed the rat on a plate by Bette Davis. My kids choke from laughing.

What sent me to a local support group, though, was the day I walked into my liquor store to get a bottle of wine. The rug buckled ever so slightly and I tripped and knocked over an entire wine display. Then, as I knelt to help clean up, I got glass shards in my knees. My blood began to mix with the flowing, spilled wine. The owner started screaming at me to "just get out of here."

Now, I go to the support group when I can, as difficult as it is to get there some days. There are so many different-looking canes and appliances, but when we start telling stories of all the falls we have taken or tickets we have gotten for getting our foot stuck between the brake and the gas pedal, then we are all the same.

And, believe me, that's not so bad!!!

—E.C., Fishkill, NY

Publications Available from the CMTA

The *CMT FACTS* Series:

CMT FACTS I is the first in a series of publications that contain important articles from previous newsletters, 1985–1990. This first booklet is 16 pages in length and contains articles on *The Genetics of CMT, CMT and Children, Physical Therapy and Rehabilitation, CMT Foot and Surgical Options,* and *the CMT Hand and Occupational Therapy Options.* The booklet is $3 for paying members of the CMTA and $5 for non-members.

CMT FACTS II is a 24-page publication that contains articles on *The Americans with Disabilities Act, Adaptive Gadgets, Rehabilitative Medicine, Vocational Rehabilitation, Orthotics, Tremor and CMT, a Disability Profile of HMSN (hereditary motor sensory neuropathy: another term for CMT), an explanation of the Neurotoxic Drug list, and pages of Ask the Doctor questions and answers.* The booklet is $5 for members and $7 for non-members of the organization.

CMT FACTS III is a 32-page publication containing articles on *CMT and Pregnancy, Prenatal Testing, several articles on research, CMTX-linked, CMT and Hand Surgery Options, Exercise and Sports for Children, Neurotrophic Drugs, and an overview of neuromuscular diseases.* Gadget information, special education information, and Ask the Doctor questions round out the contents. The price is $5 for members; $7 for non-members.

Back issues of *The CMTA Report* are available: $3 for members; $5 for non-members. Some of the topics covered:

Winter 1997:
Report on Type 2 Research Grant
Treatment of Familial Neuropathies
Phrenic Nerves and Pulmonary Function in CMT
CMT: A Primer for Patients

Spring 1997:
Volunteer of the Year Award
Ankle Bracing
Shriners Services for Children with CMT
The CMT DNA Test

Summer 1997:
New Executive Director Named
Managing Pain (2 articles)
CMT Disorders: Clinical Presentation, Treatment and Genetics
Dejerine Sottas and HNPP
MDA Services (bracing and wheelchairs)
Rocker Balance Board

Fall 1997:
Third International Conference
What is HNPP?
The Americans with Disabilities Act
A Journey Toward Wellness
Financial Report
Thoughts on Being a Patient with CMT

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, Macrodantin)
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 medication, please discuss it fully with your doctor for possible side effects.

*A megadose is defined as ten or more times the recommended daily allowance.

What is CMT?

… is the most common inherited neuropathy, affecting approximately 150,000 Americans.

… 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 also known as peroneal muscular atrophy and hereditary motor sensory neuropathy.

… is slowly progressive, causing deterioration of peripheral nerves that 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, which means if one parent has CMT, there is a 50% chance of passing it on to each child.

… Types 1A, 1X, and HNPP can now be diagnosed by a blood test.

… is the focus of significant genetic research, bringing us closer to answering the CMT enigma.

The CMTA Report

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

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