

# The CMTA Report

AUGUST 2002

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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.*

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## Swimmer Conquers the Chesapeake Bay!

By PAT DREIBELBIS

Steve O'Donnell promised that he would swim the Chesapeake Bay against the current and the choppy waters despite his CMT in order to raise awareness of CMT and to raise money for research. He was as good as his word when he swam the 4.4 mile course on June 16th in just over 2 hours. Steve had practiced for months by swimming in a pool at 5:30 in the morning for almost two hours. He had even trained in the bay near Sandy Point State Park and in the Pacific Ocean while in Hawaii and he swam despite a broken toe he managed to acquire while on vacation.

Steve's efforts to swim the bay to raise money for CMT research were written up in the local Columbia (Maryland) Flier. The article quoted Steve as saying, "I'm not Jerry Lewis and I don't have a television show. That's why I'm swimming across the bay." To date, Steve has raised more money for CMT than any other single person. His efforts have brought in a total of \$56,576 from 189 donors as of July 23, 2002.

In the last issue of the newsletter, we referred to Steve as a "hero" in his superman cape. The Columbia Flier article would seem to endorse that concept. They reported that Steve and two other bystanders helped pull a woman out of a burning car in Savage, MD recently. The car had flipped over on Route 1 and the "heroes" pried open the vehicle's door and res-



*Steve emerges from the bay following his swim.*

cued the driver right before the car became engulfed in flames. Steve modestly said he merely did what any person would have done under the same circumstances.

Steve was diagnosed with CMT a few years ago and has always led an active life. He participated in triathlons until his ankles could no longer support his running. O'Donnell is currently a coach for his 6-year-old son's baseball team and runs his own steel distribution company, but he is still challenged, as are many CMT patients, by small buttons, snapping his fingers, or standing on tiptoe.

When asked by the journalist how he would manage a physical task that would be too much for most people to even consider, Steve pointed to his heart and said, "It's all in here." His heart, his training, his dedication, and his determination all came together on June 16th when he and 600 others took on the Chesapeake Bay. No one was surprised that Steve O'Donnell finished the course or that he finished it in only 2 hours, 4 minutes. That's the kind of man he is—a hero.

*Steve would like to extend a personal thanks to all of you who supported his swim effort: "Your support helped me pull across the bay and push harder for a cure. See you next year!" He can be reached at [steve@stevenodonnellinc.com](mailto:steve@stevenodonnellinc.com)*

# A Visit to the Wayne State CMT Clinic in Detroit, Michigan

## *The CMTA.... Living our Mission*

**Mission Statement:**  
To improve the quality of life for people with CMT and to find a cure.

**Vision Statement:**  
A world without CMT.

*Tyler experiences electrodiagnostic testing during the clinic visit.*

On Thursday, June 27, 2002, CMTA Board member, Patrick Torchia and his cousin, Tom, and Tom's son, Tyler arrived at 8:30 AM for their first visit to the CMT Clinic at Wayne State. They had driven from Johnstown, PA, to be evaluated and to find out which type of CMT they had.

Upon arrival, they were met by Dr. Michael Shy, a neurologist, and Karen Krajewski, a genetic counselor, who explained what tests they would be having and what other physical evaluations would be done that day. The first item on the agenda was a routine history taking and a neurological clinical examination.

The first actual test was a nerve conduction velocity (NCV) test, which is typically used as part of the initial evaluation for diagnosing CMT. The test involves mild shocks on the arms and legs to stimulate the peripheral nerves, which lie under the skin. The intensity of the stimulus is similar to a mild slap with an open-faced hand. Waveforms generated by this stimulus are recorded on an oscilloscope and are used to calculate the speed by which the nerves carry information. This test usually allows patients to be tentatively subgrouped into CMT1 or CMT2.

The second test they received was quantitative muscle testing (QMT). For QMT the patient sits in a chair or lies on a bed and manually contracts certain muscles in the arm or leg by pulling against a strap that is connected to a



*Dr. Shy explains clinic procedure to Tom and Patrick Torchia.*

computer. The computer records the precise force generated by the muscles in units of force called Newtons.

The third test is called quantitative sensory testing (QST). In this test, a device is placed on the foot or finger and either vibrates or turns cold. The patient is asked to identify whether he feels a vibration or a cold sensation and the computer calculates the threshold or minimal level at which the patient can distinguish a vibratory or cold sensation in the hand or foot.

The fourth test is called motor unit number estimation (MUNE). This test is performed exactly like the NCV test, except that it involves stimulation of a single nerve at a time. A computer takes values generated by the stimulation and calculates the number of functioning nerves involved in stimulating the muscle under the recording electrode.

The final test is the 9-hole-peg test. The time it takes the patient to put nine pegs into a pegboard is measured. Two trials are performed for each hand.

The visitors to the clinic are seen not only by neurologists, but also by a physiatrist (MD specializing in rehabilitative and physical medicine), an occupational therapist, and a physical therapist. In addition to the neurological testing, patients can also participate in the gait analysis

*(continued on page 4)*



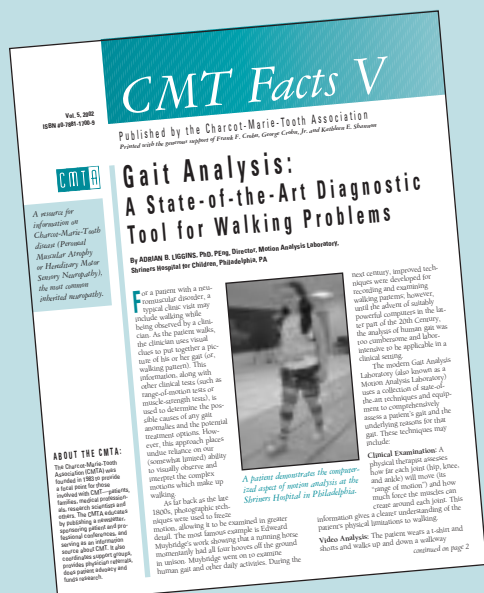
# CMT Facts V Available Now

The fifth publication in the Facts Series is now available. It is the largest edition to date and contains 56 pages of information on various practical topics of interest to CMT patients and their families.

The topics are gathered by theme and the text begins with articles on AFOs and foot issues. The second set of topics concerns emotional issues and is followed by articles on pain and CMT. Other topics include pregnancy, social security issues, vitamins and herbs, physical and occupational therapy considerations and general information such as genetic testing, medical terminology and how to find information on the Internet. A separate section is devoted specifically to HNPP (hereditary neuropathy with liability to pressure palsies).

The issue concludes with seven pages of questions to medical professionals and their answers.

This compilation of articles from previous newsletters is available to current members of the CMTA for \$12 and to non-members for \$15. This is an excellent opportunity to complete your collection if you already own Facts I-IV or to begin your library of informational publications. Please note that because of an increase in postage rates, we have had to raise the amount we charge for postage and handling.



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## VISIT TO WAYNE STATE CLINIC

*(Continued from page 2)*

laboratory, where a videotape of their walking is processed through special software that allows a specialist to see 3D views of their gaits.

The three participants were finished with their comprehensive testing by 5:30 that afternoon. With only half an hour for lunch, it was a busy day.

There is no direct cost to patients to participate in the study, and even those without health insurance can be seen with the financial support that the clinic makes available. The clinic has been operating for 6 years. The doctors involved in the study at the clinic believe that it is important to follow patients over time to see how the disease develops and to be able to evaluate trends that one phenotype might demonstrate compared with another. The clinic is open only on Thursday and the doctors can see two to five patients per day. In an average year, they see approximately 200 patients. Fully one-half of the patients they see come from outside of Michigan and outside of the United States.

As a result of the tests that were administered and a family history that was taken, Patrick Torchia was tentatively diagnosed with CMT type 1. His cousin Thomas was asymptomatic and Tom's son was believed to have inherited his CMT, not from his father's side of the family, but from his mother's.

*The CMTA acknowledges with gratitude a gift of \$10,000 to our research fund from the estate of Jane H. Wood. The gift was sent by her daughter, Suzanne W. Greene.*

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# CMTA Remembrances

Your gift to the CMTA can honor a living person or the memory of a friend or loved one. Acknowledgment cards will be mailed by the CMTA on your behalf. Donations are listed in the newsletter and are a wonderful way to keep someone's memory alive or to commemorate happy occasions like birthdays and anniversaries. They also make thoughtful thank you gifts. You can participate in the memorial and honorary gift program of the CMTA by completing the form below and faxing it with your credit card number and signature or mailing it with your check to: CMTA, 2700 Chestnut Parkway, Chester, PA 19013.

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# Numbness in CMT Disorders

By **GARETH J. PARRY, MD, Neurologist,**  
**University of Minnesota**

Each of the Charcot-Marie-Tooth (CMT) disorders is characterized by varying degrees of degeneration of peripheral nerve axons. In CMT Type I, the primary pathology is degeneration of the myelin sheath, but later there is associated axonal degeneration. In CMT Type II, the disorder directly affects the axon, resulting in primary axonal degeneration. Peripheral nerve axons are relatively simple structures whose function is to transmit electrical impulses to and from the brain; motor nerves transmit impulses from the brain to muscles, while sensory nerves transmit impulses from the skin and other body tissues back to the brain.

When nerves degenerate, those parts farthest from the brain almost always are the first to be affected. Thus, symptoms usually begin in the feet and spread up the legs to about the level of the knees and then begin to involve the hands and spread up the arms in a similar fashion. It is very unusual for the nerves to the body or the face and scalp to be involved to any great extent. Deviation from this pattern is very rare in CMT disorders other than hereditary neuropathy with liability to pressure palsies (HNPP), a condition I will discuss separately.

Involvement of motor nerves predominates in all CMT disorders, resulting in weakness and muscle atrophy (wasting). Involvement of sensory nerves is ubiquitous and yet, sensory symptoms are not as prominent. However, some degree of loss of sensation can always be found on careful examination in CMT patients. It is only rarely disabling, but it can be very annoying. Other symptoms that result from sensory nerve involvement include paresthesias (tingling), pain and loss of balance. The latter particularly occurs in the dark or when the eyes are closed.

It is not clear why sensory symptoms are so minor in CMT patients. It may be that since the sensory loss evolves so slowly, over years or decades, in some patients the brain learns to ignore the symptom. The brain also has the capacity to amplify signals received from the peripheral nerves so that if any signal is

received, no matter how small, a sensation may be felt and any sensory loss will be inapparent to the patient, but can be found on careful examination. In many patients the numbness is worse in the hands than in the feet. This is probably because the hands are normally more sensitive and we feel any sensory loss more acutely. Even in those patients who complain more of their hands, neurological examination will show a greater degree of sensory loss in the feet. Nor is it clear why sensory symptoms are so variable with some patients having very annoying symptoms, while others are completely unaware of sensory involvement.

Sensory loss appears to be more severe when the axons degenerate and, therefore, may occur earlier and be more prominent in CMT Type II. In CMT Type I, the earliest abnormality is slowing in the speed of conduction and, yet, the signal may still eventually reach the brain and be amplified so sensation is relatively normal. Later, as the associated axonal degeneration occurs, the sensory loss may become more obvious. HNPP differs from other CMT disorders in that it is characterized both by acute episodes of demyelination and by a more

slowly progressive degeneration of peripheral nerve axons. HNPP patients frequently complain of episodic numbness and tingling, lasting from minutes to several days. These episodes are usually precipitated by minor trauma to the nerve such as compression or stretching. In more severe episodes, the sensory symptoms are associated with weakness, but sensory symptoms usually predominate. These symptoms may occur in any part of the body, head or limbs. By contrast, the slowly evolving degeneration of the nerves that also occurs in HNPP patients is more like the other CMT disorders in that it begins in the feet and mainly produces symptoms of weakness although sensory loss can always be found on examination. This lends support to the idea that sensory symptoms are, at least in part, dependent on the pace at which they develop.

Symptoms usually  
begin in the feet  
and spread up  
the legs to the  
level of the knees.

*(Editor's note: This article first appeared in CMT-news.org and is reprinted by permission of the author.)*

# Charcot-Marie-Tooth Disease in

By **SUSAN IANNACCONE, MD, and NATHANIEL KHO, MD, Texas Scottish Rite Hospital for Children**

## HISTORY

Inherited peripheral neuropathies (or Charcot-Marie-Tooth disorders or Charcot-Marie-Tooth disease [CMT]) are inherited progressive disorders of the peripheral nerves that cause numbness and weakness. Recent advances in genetic research have identified several types of CMT, each of which corresponds with specific genetic mutations. CMT is also known as hereditary motor and sensory neuropathy (HMSN).

### Childhood disease

Since 1871, numerous kinships affected by a heritable neuropathy beginning in childhood have been reported. Although most of these cases were labeled dominant or recessive forms of Charcot-Marie-Tooth disease by Bell (1939), it is likely that several kindreds were examples of hereditary motor and sensory neuropathy of neuronal type commencing in early childhood. In a large series of 228 patients, Harding and Thomas (1980) found that disease onset was within the first decade of life in 62% of CMT/HMSN type 1 cases and in 25% of CMT/HMSN type 2 cases. Similar results were noted in a series of 77 patients by Buchthal and Behse (1997), wherein 70% of those with CMT type 1 had a childhood onset.

### Epidemiology

Charcot-Marie-Tooth disease is among the most common heritable neurologic disorders, but estimates of its frequency vary. An exhaustive study from Norway indicated a prevalence of 3.6 cases per 10,000 people, whereas a worldwide meta-analysis estimated a prevalence of 1 case in 10,000 people. CMT type 2 accounts for about 22% of autosomal dominant neuropathies, CMT type 1A accounts for some 60%, CMT X for about 16%, and CMT type 1b for approximately 1.6%. The other forms are rarer. The total number of affected individuals in the US is unknown.

### Clinical manifestations

Clinical manifestations are quite variable. Motor symptoms always predominate at first. Common complaints include loss of balance, weakness, and foot deformities. Parents or teachers may be concerned about clumsiness or toe walking. Clinical symptoms may begin as early as the first year of life. When symptoms are severe, the neuropathy may be called CMT type 3 or Déjérine-Sottas disease.

Commonly seen are the associated deformities of the feet (high arches or flat feet), hands (clawing), and spine (lordosis and scoliosis). All deformities are caused by loss of power and abnormal tone of the muscles. Hammertoes and high arches often lead to painful calluses and comfortable shoes may be difficult to find. The occurrence of deformities is extremely variable and cannot be predicted on the basis of age. Ankle sprains and fractures are frequent, particularly during childhood, because of joint instability. Children may complain of leg and hand cramps. Activities of daily living, including manipulating small objects such as scissors, spoons, or pens may be difficult. Children without symptoms are often diagnosed during screening of families after one relative has been diagnosed. Children often have a visibly abnormal gait secondary to weakness of the foot dorsiflexors.

Twenty-five percent of patients with CMT type 1 have enlarged and excessively firm nerves, which are found and are often visible in the superficial cervical nerves and are palpable in the arms.

## DIAGNOSIS

Diagnosis of children with CMT is important so that they receive appropriate orthopedic management. The workup may include tests that rule out causes of neuropathies such as endocrinological, infectious, and immunological abnormalities, vitamin and nutritional deficiencies, and nerve compression.

### Pedigree analysis

Establishing inheritance patterns, if available, can narrow the differential diagnosis. This is done by taking a “family history” and constructing a pedigree (family tree). In this way the physician can determine whether the disorder is sporadic (affecting only one person in the family) or whether the disorder may be passed through a recessive or dominant gene. Sporadic cases may not be CMT at all and may require further workup.

### Spinal fluid analysis

A spinal tap may be done in order to obtain cerebrospinal fluid (CSF) for protein analysis. Protein levels are usually normal in patients with Charcot-Marie-Tooth disease, but they may be elevated above 100 mg/dl. This test may be

# Children

recommended if the physician is considering a neuropathy other than CMT, such as chronic inflammatory demyelinating polyneuropathy (CIDP), for example.

## Genetic testing

If the clinical phenotype, family history, and/or electrodiagnostic (EDS) studies (electromyography [EMG]) suggest that the patient might have an inherited neuropathy, the physician may order DNA tests or mutation analysis. This analysis of the blood sample is not “genetic testing,” but rather a diagnostic test to confirm or rule out a diagnosis of CMT. Mutation analysis is now available for as many as five types of CMT. When abnormal, a definitive diagnosis is possible without subjecting the child to multiple, painful procedures. DNA analysis is also called genotyping and it can be very helpful in providing information for relatives who may be at risk of having CMT.

## Electrodiagnostic studies (EMG and nerve conduction study)

Most patients with CMT will have EDS at some point during their workup. However, this can be a traumatic experience, particularly for young children. So in families in which the type of CMT is known, DNA analysis can preclude EDS. This test is often helpful when the type of CMT is unknown. Then, EDS will allow the doctor to differentiate demyelinating versus axonal forms of CMT. When EDS is necessary in young children, sedation may be used, but most neurologists prefer not to use sedation because it is not effective in preventing pain and may prolong the whole procedure.

## Biopsy

Nerve biopsy is a surgical procedure whereby a piece of nerve is removed for examination under the microscope. Usually the sural nerve is sampled because it can be examined with a minimum of aftereffects. Nerve and muscle biopsies are often done at the same time. Biopsy may be recommended when the diagnosis of CMT is in question and, particularly in children, may be done in conjunction with orthopedic surgery.

## MANAGEMENT

### Physical Therapy and Orthotics

Physical therapy (EPT) is often required to prevent and treat joint deformities. The indications



*Grip strength is measured and then improved with exercise.*

for PT and orthotics are essentially the same in children as in adults. Orthotics such as AFOs (ankle-foot orthoses) enable patients to perform activities, while preventing falls that might result in an injury. In addition, orthotics can help prevent contractures such as Achilles tendon shortening.

### Occupational therapy

In the US, occupational therapy (OT) is the specialty that addresses problems with upper extremity function. Occupational therapists are skilled in assessing activities of daily living (ADLs), such as toileting, washing, eating, and dressing. In children, ADLs include skills required to complete schoolwork. Children with CMT often have trouble completing written assignments on time and may experience cramps in their hands while doing homework. Adaptive devices such as thick-handle tools can prevent or reverse such problems. Both physical therapists and occupational therapists are responsible for writing recommendations for school authorities (“school recs”) that are reviewed prior to the opening term. Parents and teachers should review these recommendations together so the child’s best performance in school is assured.

### Nutrition

A well-balanced diet is the goal. Obesity should be avoided, for this may contribute to entrapment of nerves, sometimes involving spinal nerve roots. Obesity also leads to increased load on muscles and increased risks for joint injury, especially of the knees.

On the other hand, malnutrition can be dangerous if it is severe enough to affect the child’s

*(continued on page 8)*

## CMT IN CHILDREN

(Continued from page 7)

ability to fight infection. The child's nutritional state should be assessed by a dietician before any elective surgery, particularly scoliosis repair.

### Pain

Pain may result from joint deformities and overuse of certain muscle groups such as may occur with an abnormal gait or scoliosis. Pain can also emanate directly from the nerves, causing strange sensations called dysesthesias, typically a burning feeling or tingling. Nonsteroidal anti-inflammatory drugs are sometimes used, but dysesthesias tend to respond well to antidepressant or anticonvulsant drugs (such as gabapentin). Cramps involving the legs and hands are common, especially after exertion, and generally go away with gentle massage.

### Surgery

Children with CMT may undergo several surgical procedures including Achilles tendon lengthening, tendon transfers, hammertoe correction, and release of the plantar fascia. Scoliosis is common in these patients after puberty; surgical correction is not usually recommended until skeletal maturity (14 years) and only when the curve is severe.

### Anesthesia

Some anesthetic agents are associated with risk of complications in all patients with weakness. These are agents that can cause "neuromuscular blockade," a direct biochemical effect on the neuromuscular junction. Succinylcholine is one such agent. In a series of 161 surgical procedures on 86 CMT patients, there were no difficulties tolerating anesthetics, even with succinylcholine. The CMTA cites this reference in their handbook for primary care physicians.



*Therapists and doctors routinely check foot flexion and muscle tone.*

They state, however, that in patients who are rapidly becoming weak, it may be inadvisable to use succinylcholine. Other risks, including sensitivity to neuromuscular blocking agents and malignant hyperthermia, are said to be minimal. Children who are being cared for in a pediatric orthopedic center can expect to see an anesthesiologist who is experienced in caring for neuromuscular patients. An experienced pediatric anesthesiologist will usually avoid the use of agents known to cause neuromuscular blockade.

Children with CMT should avoid sedation during dental procedures performed in an office rather than a pediatric facility experienced in neuromuscular disease. Prolonged body and limb positions, as are common during surgical procedures, can result in nerve compression, which could make any underlying neuropathy worse. Regional anesthesia is relatively contraindicated in CMT.

### Medications to avoid

As in adults, certain medications should be avoided, for these cause varying degrees of nerve toxicity. (See list on back page of this newsletter.)

## COMMONLY ASKED QUESTIONS:

### When to perform a genetic test?

When the clinical presentation, family history, and electrodiagnostic studies suggest that the patient might have an inherited neuropathy, genetic testing should be performed for diagnosis.

### When to perform EMG (electromyography and nerve conduction study)?

This is performed routinely in the diagnosis of neuropathy, including CMT. This test is helpful in differentiating peripheral neuropathy (nerve) from a myopathic process (muscle). Patients with negative DNA test on the current available methods for inherited neuropathy will benefit from this procedure by clarifying the underlying neuropathic process (e.g., demyelinating versus axonal forms). In some instances, this is performed as a screening tool among family members for whom DNA analysis is not revealing.

### What is the best physical exercise for patients with CMT?

Strengthening exercises may be beneficial if confined to proximal muscles, such as knee extension and flexion, hip extension and abduction exercises. However, such exercises should not involve use of weights in children. Stabilizing the distal joints via use of proper footwear or an



orthotic device can help prevent injury. Most clinicians recommend aerobic but non-resistive and low-impact exercise. This includes swimming, walking, and bicycling.

### Does early onset disease mean severe disease?

In CMT type 1, progression of the disorder is usually slow even if presenting in early childhood. In some children or adolescents, there may be periods of rapid progression that last for months or years followed by prolonged periods without change (plateau). In some cases, the progression occurs during growth spurts, but not always. The pattern of progression and plateaus is distinctly unpredictable. The possibility of superimposed acquired neuropathy may be considered if a change in clinical progression is noted, especially in adults.

### When to perform surgery?

#### Spine

The decision to undergo scoliosis repair should occur only after full discussion with the pediatric orthopedist, the primary care physician, and the child. Current treatment guidelines for scoliosis in patients affected by CMT disease are the same as in patients afflicted by idiopathic scoliosis despite the presumed neuropathic nature of the disorder. Current guidelines of idiopathic scoliosis as recommended by the Scoliosis Research Society include the following:

- Periodic evaluation in skeletally immature patients (less than 6 months postmenarchal or Risser sign of 0 or 1) with less than 25 degrees of deformity or more mature patients with less than 45 to 50 degrees of deformity.
- Spinal bracing for curvatures between 25 and 45 degrees in skeletally immature patients.
- Spinal fusion for progressive curves in skeletally immature patients refractive to bracing, or curves greater than 50 degrees irrespective of the skeletal maturity.

#### Hip dysplasia

Hip dysplasia is not uncommon in patients with CMT. Symptomatic patients may consider hip reconstructive surgery to relieve pain. However, there is a significant risk of avascular necrosis of the femoral head (destruction due to disruption of the blood supply to the head of the femoral bone) in CMT patients. Therefore, this reconstructive surgery is not recommended for most patients and is rarely necessary in children.

#### Foot deformities

Cavovarus (cavus-high arch, varus-hindfoot inversion) foot deformity is commonly seen with “clawing” of the toes. Surgical treatment is reserved for patients whose pain and discomfort cannot be treated adequately with orthotics. The

principles of surgical management are to correct the deformity itself and to balance the muscle function around the foot and ankle to prevent recurrence. Common procedures include:

- Posterior tibial tendon transfer to the dorsum of the foot.
- Transfer of the peroneus longus to either the peroneus brevis or to the dorsum of the foot.
- Transfer of the long toe extensors posteriorly to the metatarsals to ameliorate clawtoes and forefoot dorsiflexion.

These procedures do not correct the underlying weakness and may perhaps be aggravated by the transfers. The goal of these procedures is to correct the deformity and ameliorate pain. It is therefore important that the patient and his family be counseled prior to embarking on these extensive procedures.

Other procedures such as Achilles tendon lengthening and release of the plantar fascia are less extensive and may be sufficient for some children.

### Is there a cure for CMT?

At this time, there is no cure for hereditary neuropathy (HMSN/CMT).

Experimental therapies are being considered. One approach being considered in an animal model is to introduce recombinant DNA encoding normal PMP22 into nerves. Another idea is to transfer the neurotrophin gene into the spinal cord to prevent secondary axonal changes in animal models of CMT.

## PROMISING NERVE REGENERATION STUDY INITIATED AT OHIO STATE

**D**r. Sarife Sahenk, Professor of Neurology at Ohio State University, is investigating the ability of a new drug, NT-3, to protect and regenerate peripheral nerves.

Dr. Sahenk and her group have shown that neurotrophin-3 (NT-3), which is made by the Schwann cells, or myelin-forming cells in peripheral nerves, can help maintain axons and allow them to regenerate. “We found that treating mice that have defective myelin sheaths with NT-3 resulted in significant regeneration of their nerve fibers,” states Dr. Sahenk. “Our group at Ohio State is now conducting a clinical pilot study to determine whether NT-3 can help patients with a form of hereditary demyelinating neuropathy called CMT.”

Dr. Sahenk believes that in demyelinating neuropathies such as CMT disease, CIDP, MAG neuropathy and Guillain-Barré syndrome, among others, there is a deficiency of NT-3 resulting in breakdown of the axons. They found that normal axons surrounded by sick Schwann cells degenerate and fail to regenerate, whereas they can regenerate if surrounded by normal Schwann cells or given NT-3.

Supplies of NT-3 are being provided by the manufacturer, Regeneron, Inc. “The study should be concluded in the fall,” states Dr. Sahenk, “at which time the results will become available.”

## Aids for Daily Living Can Make Simple

In our continuing effort to provide our readers with practical solutions to tasks which may have become difficult due to loss of dexterity or strength in the hands, we are presenting a collection of adaptive devices which are designed to make tasks requiring thumb and forefinger pinch or hand strength less challenging.

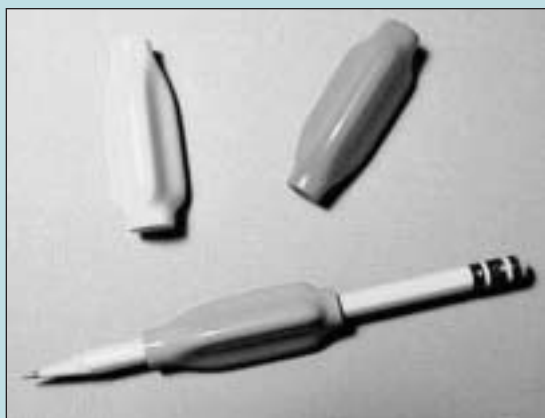
### The Steady Write Pen

This pen is attached to a triangular base which balances and guides the hand. It improves shaky handwriting and can be used by either right or left handers. It is available from Smith & Nephew at 1-800-558-8633 or [www.smith-nephew.com](http://www.smith-nephew.com). The cost is \$8.45.



### Pen/Pencil Holders

A soft holder slides over the end of a pen or pencil and provides a built-up gripping area. It is useful for slight decreases in range of motion in the hands or for limited hand strength. The set of three costs \$4.20 from Smith & Nephew.



### Letter Writing Guide

This plastic 8½ x 11-inch writing guide helps people with limited hand function to write in straight evenly spaced lines on paper. It would be especially helpful in filling out forms with small writing spaces. The cost is \$2.99 from LS&S Group at 1-800-468-4789 or [www.lssgroup.com](http://www.lssgroup.com).



### The Tap Turner

This device provides increased leverage when turning faucet knobs. Designed for people with decreased hand dexterity or strength and for those with limited reach. It costs approximately \$23 from Smith & Nephew.



# Tasks Simpler

## The Bottle Opener

This rubber opener fits over small bottle tops (such as soda bottles) and provides a firm grip on tops and knobs, allowing them to be opened with ease. The opener is useful for people who have difficulties gripping and turning small objects. The price is \$7.00 from Smith & Nephew.



## The Jar Opener

A rubber cone fits over jar tops and lids, gripping them for easier opening. It is useful for people with arthritis or decreased hand strength. It costs approximately \$6 from Smith & Nephew.



## Door Lever Adapter

This device attaches securely to a variety of round door knobs to provide leverage when opening doors. It is easy to grasp with one hand and does not require pinching or twisting of the wrist to operate. It costs approximately \$12.95 from Lindustries, Inc. at 1-781-237-8177.

## Weighted Utensils

These utensils with built-up weighted handles were designed to minimize hand movements caused by tremors. They are also useful for persons with limited grip strength. A set of four is approximately \$28 from Smith & Nephew.

Smith & Nephew, 1-800-558-8633,  
www.smith-nephew.com

LSS&S Group, 1-800-468-4789  
www.lssgroup.com

# The Secret to Happiness

(Excerpted from UPDATE, a publication of Independence Blue Cross.)

What does being happy really mean, and how can we achieve it? If you believe the answers lie in popularity, money, or luxury, new research findings might surprise you. Happiness is “the overall sense of being satisfied with your life as a whole,” says psychologist Ken Sheldon, Ph.D. from the University of Missouri—Columbia. Despite advertising’s claims, wealth and fame don’t necessarily make us happy. According to what Dr. Sheldon and his colleagues wrote in the *Journal of Personality and Society Psychology*, there are three top needs that drive happiness:

- Autonomy (a sense that you’ve chosen to do what you do)
- Competence (you’re good at what you do)
- Relatedness (you feel close to others)

In general, if you’re happy, you feel confident and in control. You’re more optimistic, energetic, decisive and creative. You view the world as a safer place than do those who aren’t happy. You have high self-esteem and a sense of meaning to your life, and you’re more capable of intimacy with others.

You’re also healthier! “Our body’s immune system fights disease more effectively when we are happy rather than depressed,” says social psychologist David Myers, Ph.D., professor at Michigan’s Hope College and author of *The Pursuit of Happiness*.

Your happiness potential is a bit like your cholesterol level. Both are influenced genetically, as well as by diet, sleep and exercise. Your emotional attitude and the quality of your relationships with others also affect happiness. But being healthy doesn’t mean you are happy, and vice versa. “There are plenty of people in poor health who are happy,” says Dr. Sheldon.

“There’s some indication that older people above 60 are happier [than when they were younger]” says Dr. Sheldon. Why? Because in their later years they’re likely to pursue goals based on personal fulfillment rather than obligation.

## Ten Steps to a Happier Life

- 1 Decide to be happy. Think and act happy, so you can create the state of mind for yourself and inspire it in others.
- 2 Take control. Take charge of your time and schedule to make effective change in your life.
- 3 Set goals. Choose ones that urge you forward yet also let you realistically reach them.
- 4 Engage your skills. Seek work and leisure activities that challenge your talents and abilities without overwhelming you.
- 5 Be healthy. Eat well, exercise regularly and get plenty of sleep.
- 6 Feed your soul. Spend time alone, without distractions. Make art, write poetry, meditate, or read inspirational literature.
- 7 Seek out others. Surround yourself with happy people and nurture these relationships.
- 8 Reach out. Be compassionate and focus beyond yourself, to help those in greater need.
- 9 Remain curious. Stay open to new people and experiences so you continue to grow.
- 10 Keep perspective. Take a moment each day to reflect on the positive things in your life that can make you feel grateful.

## Support Group News

### ■ California - Berkeley Area

The July meeting was held at the Albany Library from 2-4 PM on Saturday, July 6. Bruce Dragge shared information on nutrition and its ability to improve the symptoms of CMT. There was the usual opportunity for questions, sharing, and discussion.

### ■ California - Northern Coast Counties

The July meeting featured a presentation by Meganwind Eoyang discussing exercises that can be performed at home to contribute to balance and safety. Future meetings will be held on November 2, 2002, featuring Dr. Jerome Chinn, a neurologist who practices in Santa Rosa. In March 2003, Michelle Mendoza, a physical therapist at the MDA Clinic in San Francisco, will be featured.

### ■ Missouri/Eastern Kansas

A recent meeting of this support group featured a presentation by Dr. Michael Brown on "A Natural Medicine Approach to Charcot-Marie-Tooth Disease." He listed the reasons for lack of health

as stressful, Type A lifestyles, poor food choices, lack of water, lack of exercise, exposure to harmful toxins and pollutants and an inadequate amount of essential nutrients. His primary suggestion was to develop an optimal exercise routine because it will help regulate blood sugar, improve immune function and lymph flow, relieve depression, anxiety and pain, help lower blood pressure and cholesterol, improve bone density and lean muscle mass, and improve stamina and creativity. Proper exercise cannot be replaced by diet, nutrition, or drugs.

### ■ New York - Greater New York

Robert Wine from the support group has informed the office that the group now has a website at [www.cmtnyc.org](http://www.cmtnyc.org). The site has links to other useful sites and articles on various aspects of CMT such as rehabilitation, bracing, and the general medical aspects of CMT. They list their upcoming events and provide a current list of journal articles on CMT. The group meets the second Thursday of each month at the Rusk Institute.

## CMTA Offers Golf Shirts

**B**ecause of the popularity of golf tournaments as fundraising events, the CMTA has ordered golf shirts with the organization's name in our teal color to help raise awareness of our existence. The shirts will be used at the Long Island golf tournament, the Philadelphia area tournament and the Johnstown, PA, one. Additionally, the shirts are being offered to our membership as a way for each person to help spread the word about our organization.

The shirts are white with the Charcot-Marie-Tooth Association in teal on the left breast pocket area. The shirts are available in a wide range of sizes from medium to XX large. They are 100% cotton and are very soft and comfortable. Remember when ordering that the sizes are men's sizes and women should order based on that information.

The price of the shirt is \$15 with the proceeds benefitting the research fund of the CMTA. Ordering a shirt is just one more way you can help support the efforts of the organization to find a cure for CMT.

To order a shirt, see the "CMTA Membership/Order Form" on page 3 of this newsletter.



*Charles Hagins models the new CMTA golf shirt available in white with teal lettering.*



# CMTA Support Groups

## Arkansas—Northwest Area

**Place:** Varies, Call for locations  
**Meeting:** Quarterly  
**Contact:** Libby Bond, 501-795-2240  
**E-mail:** charnicoma57@yahoo.com

## California—Berkeley Area

**Place:** Albany Library, Albany, CA  
**Meeting:** Quarterly  
**Contact:** Ruth Levitan, 510-524-3506  
**E-mail:** rulev@pacbell.net

## California—Los Angeles Area

**Place:** Various locations  
**Meeting:** Quarterly  
**Contact:** Serena Shaffer,  
 818-841-7763  
**E-mail:** CMT\_losangeles@yahoo.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  
**E-mail:** pcmobley@mac.com

## Colorado—Denver Area

**Place:** Glory of God Lutheran Church  
 Wheat Ridge  
**Meeting:** Quarterly  
**Contact:** Marilyn Munn Strand,  
 303-403-8318  
**E-mail:** mmstrand@aol.com

## Kentucky/Southern Indiana/ Southern Ohio

**Place:** Lexington Public Library,  
 Northside Branch  
**Meeting:** Quarterly  
**Contact:** Robert Budde,  
 859-255-7471

## Massachusetts—Boston Area

**Place:** Lahey-Hitchcock Clinic,  
 Burlington, MA  
**Meeting:** Call for schedule  
**Contact:** David Prince, 978-667-9008  
**E-mail:** baseball@ma.ultranet.com

## 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:** Flora Jones,  
 601-825-2258  
**E-mail:** flojo4@aol.com

## Missouri/Eastern Kansas

**Place:** Mid-America Rehab Hospital,  
 Overland Park, KS  
**Meeting:** First Saturday bi-monthly  
**Contact:** Lee Ann Borberg,  
 816-229-2614  
**E-mail:** ardi5@aol.com

## Missouri—St. Louis Area

**Place:** Saint Louis University Hospital  
**Meeting:** Quarterly  
**Contact:** Carole Haislip, 314-644-1664  
**E-mail:** c.haislip@att.net

## New York—Greater New York

**Place:** NYU Medical Center/  
 Rusk Institute, 400 E. 34th St.  
**Meeting:** 2nd Thursday of each month  
**Contact:** Dr. David Younger,  
 212-535-4314,  
 Fax 212-535-6392  
**Website:** www.cmtnyc.org

## New York—Horseheads

**Place:** NYSEG Meeting Room, Rt. 17  
**Meeting:** Quarterly  
**Contact:** Angela Piersimoni,  
 607-562-8823

## New York (Westchester County)/ Connecticut (Fairfield)

**Place:** Blythedale Hospital  
**Meeting:** 3rd Saturday of each month,  
 excluding July & August  
**Contacts:** Diane Kosik, 914-937-2013,  
 Beverly Wurzel,  
 845-783-2815  
**E-mail:** ladydismiles@aol.com or  
 cranomat@frontiernet.net

## North Carolina—Archdale/Triad

**Place:** Archdale Public Library  
**Meeting:** Quarterly  
**Contact:** Ellen (Nora) Burrow,  
 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  
**E-mail:** Greenville-Ohio-CMT@  
 woh.rr.com

## Oregon/Pacific NW

**Place:** Portland, Legacy Good Sam  
 Hospital, odd months  
 Brooks, Assembly of God  
 Church, even months  
**Meeting:** 3rd Saturday of the month  
 (except June and Dec.)  
**Contact:** Jeanie Porter, 503-591-9412  
 Darlene Weston, 503-245-8444  
**E-mail:** jeanie4211@attbi.com or  
 blzerbabe@aol.com

## Pennsylvania—Philadelphia Area

**Place:** University of PA, Founders  
 Building, Plaza Room A  
**Meeting:** Bimonthly  
**Contact:** Amanda Young,  
 215-222-6513  
**E-mail:** stary1@bellatlantic.net



## WRITE TO US!

Pat Dreibelbis, Editor

The CMTA Report

CMTA

2700 Chestnut Pkwy.

Chester, PA 19013

*The CMTA reserves the right to edit letters for space.*

### The CMTA Report

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

# Ask the Doctor

Dear Doctor,

*My brother has CMT and has recently been having seizure-like symptoms. Can seizures be part of having CMT?*

**A Medical Advisory Board Doctor replies:**

CMT and seizures don't usually go together. If someone has CMT and seizures, it is either an unrelated coincidence (seizures are not rare) or some very unusual disease and not typical CMT. It is important to recognize that petit mal seizures are a very specific neurological diagnosis and require EEG confirmation. It does not refer to people who simply feel queasy and light-headed or see flashing lights. Your brother should see his doctor for a complete examination and diagnosis.

Dear Doctor,

*I am 57 years old and was diagnosed with CMT 16 years ago. I am also a diabetic. A year and a half ago, I got a power chair because I am unable to walk or stand without my legs buckling or freezing. I have not read in any journals of my two most severe symptoms. My neurologist seems to be very knowledgeable about CMT and said these symptoms are caused by CMT. They are as follows:*

1. *My calves are very large and swollen and very painful. Often, they spasm and become hard as a rock. They freeze on me. I am told that my calf muscles have atrophied and that tissue, without muscle, has grown where there was once muscle.*

2. *My hamstring muscles are very hard and painful. They feel like inch-wide bands of steel. Stretching exercises tear muscle fiber and that causes swelling.*

*Do you agree that my symptoms are CMT? I would also like to mention that three times a week, for 90 minutes, I have an excellent massage therapist. He is able to soften my spasms and get me out of pain for a short while.*

**A Medical Advisory Board member replies:**

The writer states that his calves are very large, swollen, and painful. This is often associated with "spasms." He is correct that there is replacement of muscle by connective tissue in CMT, but most often there is no calf enlargement. Sometimes loss of innervation of the calf muscle will induce some enlargement, but in most cases in which this process is of long duration, the enlargement disappears.

Shortening of the hamstring muscles occurs in virtually every patient who is confined to a wheelchair, unless there are frequent stretching exercises.

# Letters to the Editor

Dear CMTA,

I would like to comment on your article about canes in the June edition of The CMTA Report. I am 72 and have been diagnosed with CMT1A. At various times in the past I have wanted to use a cane as I have less than normal balance. I have tried the T-shaped handles and a pair of the Fisher handles which are formed for each hand individually.

In each case, I got help with balance, but found my hands became sore and hurt after a short usage of the cane. Also, my hands seemed to "go to sleep" more at night after having used the canes. It would seem to me that would be an obvious problem with most CMT patients as the hands are affected by the disease.

The best help I have with balance outside my home is a rolling walker or a shopping cart.

In the house, the walls and furniture are much handier and less hurtful than canes, for me.

—J. W. Wyoming

Dear CMTA,

If anyone is having trouble with appropriate physical education classes, your local board of education should be your first stop. If your child is attending a public school which receives any federal funding, your child is entitled to an appropriate alternative or modified physical education class as long as PE is part of the regular school curriculum. If your child has trouble with regular PE, contact the school district's administrator in charge of special education. The school is required to provide an appropriate alternative, not just excuse the child from PE.

es applied. Such shortening may be enhanced by the syndrome of “spasticity,” which may result in a preferred flexion posture of hips and knees.

Since a number of heritable nerve and muscle disorders can imitate CMT, it is extremely important to carry out the necessary studies for accurate diagnosis. Spasm of the gastrocnemius muscle is unusual in piriforms of CMT, but CMT is sometimes associated with conditions that may yield spasticity. Medication for spasticity can reduce the incidence of spasms and make life more tolerable.



#### Dear Doctor,

*I am wondering if there is any connection between diabetes and CMT. I seem to know a lot of people who have CMT and are also type 2 diabetics. Is this just a coincidence or is the diagnosis of diabetes a result of our inability to exercise as much as we should?*

#### The Doctor replies:

There is no recognized relationship between CMT and diabetes. Diabetes is a very common problem, affecting an estimated 12 to 15 million Americans. With the 150,000 to 200,000 patients with CMT, the probability of coincidental association is fairly high.

Before you start any campaign, make sure you know what your child would like to do. Some children would rather attempt everything that others do and can do so without harm if the teacher is aware and provides an appropriate environment.

It helps to have a letter from your child's doctor before you begin your campaign, and you have every right to request that the doctor be part of the development of your child's individualized educational plan.

Remember that it's illegal for any child in a school receiving federal funds to suffer through inappropriate physical education. It's also illegal for the child to simply be excused from PE if it is offered to the rest of the class. Kids can't be tortured, or left out.

—D. H. (primary school teacher)

## Project Needs Help with Survey Work

**SEEKING:** Parents with disabilities and their teens

**CONTACT:** Nancy Freed

(510) 848-1112 ext. 174 Voice: (800) 644-2666 TTY: (800) 804-1616

Email: [ParentsandTeens@lookingglass.org](mailto:ParentsandTeens@lookingglass.org)

Website: [www.lookingglass.org](http://www.lookingglass.org)

Berkeley, CA - Through August 2002 -Through the Looking Glass, National Resource Center for Parents with Disabilities, is a community-based nonprofit organization. Through the Looking Glass is conducting a nationwide project to learn more about families in which a parent with a disability is raising a teen (11 to 17 years old). The National Institute on Disability Research and Rehabilitation, part of the Department of Education, funds this project. The staff comprises various specialists who have diverse cultural backgrounds, and nearly 80% of the 40 staff members are disabled, parents of disabled children, or members of families with disabilities.

**WHAT DO WE MEAN BY DISABILITY?** Disability can involve physical, visual, systemic, hearing, cognitive, learning, developmental and mental health issues. Why are we conducting research? Although there are over 10 million families in which one or both parents have a disability, relatively little is known about the experiences of these families.

**WHO CAN PARTICIPATE** Parents with disabilities and Deaf parents who have teenagers (children 11 to 17 years of age). The parent and the teenager are both welcome to participate.

#### WAYS TO GET INVOLVED

- Participate in a national survey of parents with disabilities who are raising teens aged 11 to 17. Your teen can also fill out a survey and receive \$5 in return.
- Surveys are available in a variety of formats: Online and printable at our website, in Spanish, over the phone, and in a version specific to Deaf parents.
- Deaf parents can participate in a face-to-face interview in ASL if you live in or near one of the following cities: San Francisco, Seattle, Santa Fe, New York, Kansas City, or Washington, DC.

#### HOW TO CONTACT US

- Call Nancy Freed at (510) 848-1112 ext. 174, Toll Free (800) 644-2666, TTY (800) 804-1616
- Request a survey by email [ParentsandTeens@lookingglass.org](mailto:ParentsandTeens@lookingglass.org) or contact Nancy Freed at the above telephone number.
- Online and printable survey at our Website: [www.lookingglass.org](http://www.lookingglass.org)

*Through the Looking Glass, The National Resource Center for Parents with Disabilities, is a nonprofit organization in Berkeley, CA that serves families in which a parent or child has a disability. Current projects include The National Parent to Parent Network for parents with disabilities. For more information on these projects please contact Through the Looking Glass.*

## 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, 1B, 1X, HNPP and EGR-2 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*



2700 Chestnut Parkway  
Chester, PA 19013  
1-800-606-CMTA FAX (610) 499-9267  
[www.charcot-marie-tooth.org](http://www.charcot-marie-tooth.org)

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