

THE CMTA REPORT

Fall 1990

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Providing information on Charcot-Marie-Tooth disease (a.k.a. Peroneal Muscular Atrophy or Hereditary Motor Sensory Neuropathy), the most common inherited neurological disease. Contents © 1990, CMTA. All rights reserved.

CMT Foot - Surgical Options

An Overview

Problems with the feet are often the earliest symptoms of Charcot-Marie-Tooth disorder. Beginning as curled toes and high arches, CMT foot problems usually progress. There are three foot deformities typical of CMT, all of which may be present to some degree. These include the cavus foot, the varus heel and claw toes. They usually begin as flexible deformities and progress to fixed deformities. These foot deformities are caused by growth of bone during a state of muscle imbalance. The progression of the deformities can be seen in Figures 1 - 3.

The cavus foot, also called pes cavus or high arches, results from a muscle imbalance between the extrinsic and intrinsic muscles of the foot. The plantar fascia (soft tissue on the sole of the foot) and the plantar ligaments (tissue connecting the bones of the feet) tighten, somewhat like a rubber band, pulling the ends of the foot closer together. This results in the high arch.

Heel varus, also referred to as hindfoot varus, is a condition in which the heel turns in. When viewed from behind it appears that the person is walking on the outside edge of the foot. Heel varus creates instability and balance problems.

Claw toes, or hammer toes, is a condition where the toes cock up. Initially claw toes are seen during walking due to muscle imbalance. Later, this condition becomes rigid and the toes stay cocked. Claw toes cause irritation to the top of the toes as they rub against the shoe. This condition also makes it difficult to find shoes that fit properly.

In addition to these three deformities, painful callouses often develop on the bottoms of the feet and the feet become somewhat rigid. Ankles may become weak and unstable and the "forefoot

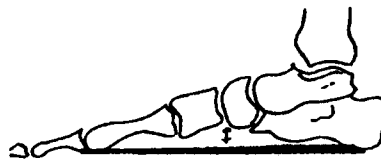


Fig. 1. The normal foot

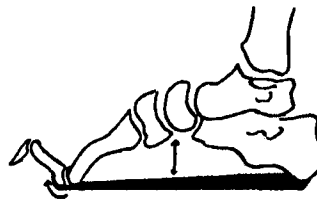


Fig. 2. The CMT foot. Note high arch (pes cavus), claw toes, tightened plantar fascia.

equinus" (foot-drop) may set in. Foot drop results from the wasting of the peroneal muscles (located on the outside of the leg below the knee) and is the cause of high steppage gait.

Surgery's aim is to enable the CMT patient to walk with the entire lower surface of the foot on the ground (plantigrade), to decrease pain, to improve balance and agility, and to halt progression of the deformity.

The Assessment

The progressive nature of CMT makes early assessment and long-term treatment planning prudent. A careful evaluation of the foot is the first step. Distinguishing between fixed and flexible components of each deformity is essential to the proper management of CMT and formulation of an individualized treatment plan.

Options

If the assessment concludes that surgery is indicated, things become more complicated. Several types of surgery may be performed on the CMT foot, depending on the level of involve-

ment. Tendon transfers, lengthenings and sometimes tenotomy (surgical division of a tendon) may be used to eliminate deforming forces (to slow or stop progression of the deformity) and to balance the foot. Osteotomies (surgical division or sectioning of bones) and arthrodesis (fusing joints) are often necessary to correct alignment and to alleviate symptoms, especially in the case of rigid or fixed deformities.

The three foot deformities and respective surgical options will be discussed separately, but the foot is treated as a whole and surgeries, if necessary, are often done at the same time. Specific surgical procedures will not be discussed in detail in this article.

Cavus Foot

Where there is no bony deformity, the goal is to release the tightened tissues and ligaments to relax the bottom of the foot and the cock up tendencies of the toes. Surgical procedures include plantar fasciotomy/fasciotomies, also called plantar release.

When bony deformities are present, some form of osteotomy (removal or cutting of bone) may be performed.

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MEDICAL ALERT



*Certain Drugs
Toxic to the
Peripheral
Nervous System*

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

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

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

VCR Tape Rental

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

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

To order a tape, fill out our "I want to be in touch!" form (see page 11) and send it to us with a check or money order payable to: The CMTA, Crozer Mills Enterprise Center, 600 Upland Ave., Upland, PA 19015.

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CMT Foot (cont'd from pg. 1)

Essentially these remove a section or wedge of bone, then "close" the foot to reposition the foot to plantigrade (walking with the entire bottom of the foot on the floor.) Surgical procedures may include "closing wedge greenstick dorsal proximal metatarsal osteotomies", "closed wedge dorsal osteotomy", "the Japas-V-tarsal osteotomy", the "Cole osteotomy" and the "tarsometatarsal truncated wedge osteotomy". A variety of muscle transfers in various combinations (such as moving the long peroneal muscle to the short; or moving the posterior tibialis to the dorsum of the foot) may also be done to eliminate the deforming force and improve the position of the foot.

Varus Heel

If the heel is flexible, correction of the cavus component should correct the flexible varus heel.

If the varus heel is rigid, an osteotomy may be performed on the heel bones. A wedge of bone is removed from the heel bone to correct it to the straight position. Technically (and with more details) this is a Dwyer osteotomy, closing lateral wedge osteotomy of the calcaneus, or hindfoot osteotomy.

Claw Toes

If the claw toes are still flexible and have not become rigid, cavus correction should fix the claw toes.

Surgical procedures to fix rigid claw toes include transferring the tendons connected to the toes to the bones of the foot, fusing interphalangeal joints (the ones in the middle of the toes) and fusing bones. Procedures include the Jones procedure, modified Jones transfer and the Girdlestone-Taylor Transfer.

Triple Arthrodesis

Triple Arthrodesis is a procedure which stabilizes the hind foot by fusing three joints. There are several types of triple arthrodeses; the condition of the foot dictates which might be appropriate. The basic triple arthrodesis involves fusing three joints in the hind foot (the talonavicular joint, the subtalar joint and the calcaneocuboid joint). This eliminates side-to-side movement of the foot, but the ankle joint is still free so you retain up and down movement of the foot. In the Pantaylor Triple Arthrodesis, the ankle joint is also fused. The Lambrinudi

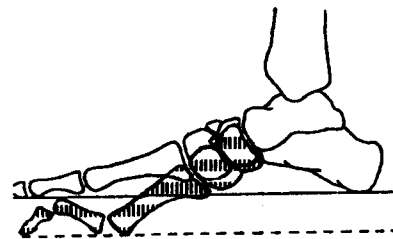


Fig. 3. Advanced CMT foot with rigid deformities, equinus position (foot-drop).

Triple Arthrodesis "tricks" the ankle into thinking that the foot is completely flexed, or pointing down when it has been surgically moved to a position parallel with the ground/floor. The ankle can still move the foot up but not down.

A triple arthrodesis is not done before the foot stops growing— usually age 12 in girls and age 14 in boys. The goal is to prevent progression of the deformity. There is some controversy over whether or not triple arthrodesis offers better long-term stability than combined mid-foot and hind-foot osteotomies.

Summary

The progression of the development of the cavus foot needs to be closely monitored. By identifying progressive pes cavus in the early stages before rigid deformities have occurred, muscle-balancing procedures may suffice. Once deformities have become rigid, surgery becomes even more complex. Most surgeries will require casts for at least six weeks, followed by several months of frequent elevation of the leg and foot to control swelling.

CMT is a progressive disorder. Following foot surgery it is recommended that the patient receive intensive physical therapy under the guidance of a therapist well versed in the CMT foot. This is especially imperative following muscle balancing operations, tissue release, tendon transfers and muscle lengthening operations.

Conditions are dynamic— ever changing. In situations where a second opinion is required, it is not unusual for the opinions to conflict. Surgery may not solve all problems, and there is always the potential for deterioration of the foot correction with time. However, when indicated, foot surgery can drastically improve the quality of life for the CMT patient.

—Jennifer Schoonmaker Hitt

Our thanks to Dr. Bruce Sangeorzan for his review of this article.

CMT Patient Profile

Building Support

The following article appeared in the 7/13/90 issue of *The Sun Sentinel* and is reprinted with its permission.

by Linda Ellis

Every year, as his muscles waste away, George Meyerson's hands grow to look more like claws. His feet, too, grow weaker and more deformed. Meyerson, 70, must find new ways to cope with his disability.

For most of his life, he has struggled alone; until two years ago, he had never met anyone else with Charcot-Marie-Tooth disease.

The disease, named after the doctors who identified it, affects the nerves that control and maintain the muscles of the hands, forearms, feet and lower legs.

The malfunctioning nerves conduct electrical impulses too slowly. As a result, the muscles atrophy.

Although it is the most common inherited neurological disorder, affecting some 125,000 Americans according to the non-profit Pennsylvania-based Charcot-Marie-Tooth Association, CMT has received little publicity.

When at last Meyerson met some fellow sufferers at a support group meeting on Orlando two years ago, he was relieved.

It was like a weight off my shoulders to meet others after all these years," he said. "I used to feel as if I was the only person in the world who had it."

Now Meyerson, who lives in Pembroke Pines with his wife Rose, wants to increase public awareness of the disease and form a South Florida support group.

"I want to help people, especially those who are young enough that treatments can really help," he said. "And I hope that people who don't realize they have it will recognize it."

Meyerson himself was middle-aged before he discovered there was a name for his symptoms.

As a child in Brooklyn, he was just considered clumsy.

"I couldn't catch a baseball like the other kids," he recalled.



George Meyerson

Although his hands were weak, his feet and legs were strong. Meyerson was on his high school track team and loved to dance the Lindy Hop, he said. But during his 20s he began to develop a limp.

"I didn't go to a doctor- I wasn't sick. There was no pain involved," he said. He was unaware that his muscles were gradually wasting away.

Meyerson was diagnosed with Charcot-Marie-Tooth disease when he was 45. It was then he learned he had inherited the disease from his mother, whose only symptom was her hammer toes. Statistically half of a sufferer's children will inherit the gene.

"I watch my three grandsons like a hawk" for signs of the disease, Meyerson said. Their mother, Janice, Meyerson's 41-year-old daughter, recently experienced weakness in her hands.

Meyerson himself has been relatively lucky. While some people with the disease are confined to a wheelchair or dependent on leg splints to walk, he gets by with orthopedic shoes.

"I can do everything that any 70 year old can do- maybe more," said Meyerson, who credits his mobility to his athletic youth.

"I always tried to keep up with the boys. I developed my thigh muscles and that

has got me through," he said. The strength of his thigh muscles compensates for the weakness of his stick-thin lower legs and ankles, Meyerson said. He walks with a high-stepping gait to avoid dragging his feet.

Now retired from his job as manager of a sewing factory, Meyerson plays nine holes of golf several mornings a week. He taught himself to play after a professional refused to teach him.

"He said I could never play without the proper grip," said Meyerson, who holds the club in a kind of baseball grip and maintains a respectable handicap of 21.

Meyerson is eager to start a support group so he can urge others to remain active, as he has.

"If I was idle, I think I would be crippled by now," he said. "I want to get everybody off their fannies and get moving."

Members of a support group can share information, he said. For instance, he would suggest that others try the high-top shoes he designed, which have greatly improved his balance- and his golf game.

Karol Hitt, president of the non-profit Charcot-Marie-Tooth Association, said that support groups can provide both practical and emotional benefits.

"Frequently, people who have the disorder feel isolated," she said. "Then they find a group of people who have common problems, and suddenly they are not alone any more."

"Then there's the networking: where to buy shoes, what orthopedic surgeon they saw, what government services someone was able to get for their child in school."

Once Meyerson has established a local support group, he will work with Hitt to bring guest speakers- experts such as Dr. Ram Ayyar, professor of neurology at the University of Miami medical school.

"I'd be delighted to tell them about the latest developments" in medical treatments, Ayyar said. Although there is no cure for the disease, there are therapies.

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1990 in Review



Eunice Cohen & Dr. Addante at Boston conference

The CMTA has changed and grown during 1990. The change came about at the beginning of the year when the Board of Directors, after much consideration, voted to change the name of the organization. The new name more clearly identifies the patient/family population that we serve. This was learned after surveying a random group of neurologists.

In September we began our second year in our new offices. The move has been advantageous and productive for the organization. The office is located in a building which is a "business incubator" and is run by a nonprofit corporation. The result is we have space and services at a reasonable cost. The services we have gained have been excellent and certainly the reception/telephone service is outstanding.

Our growth has come about in many ways. Our philosophy of regional CMT patient/family conferences utilizing area medical experts continued with three meetings. In April we met in Miami at the University of Miami Medical Center. Drs. Ram Ayyar, chief of the neuromuscular clinic, and Michael Gurvey, chief of pediatric orthopedic surgery at Mt. Sinai Medical Center, spent an afternoon teaching and answering questions from the assembled patients and family members. From this meeting George Meyerson, a CMT patient, volunteered to start a south Florida support group. (See CMT Patient Profile).

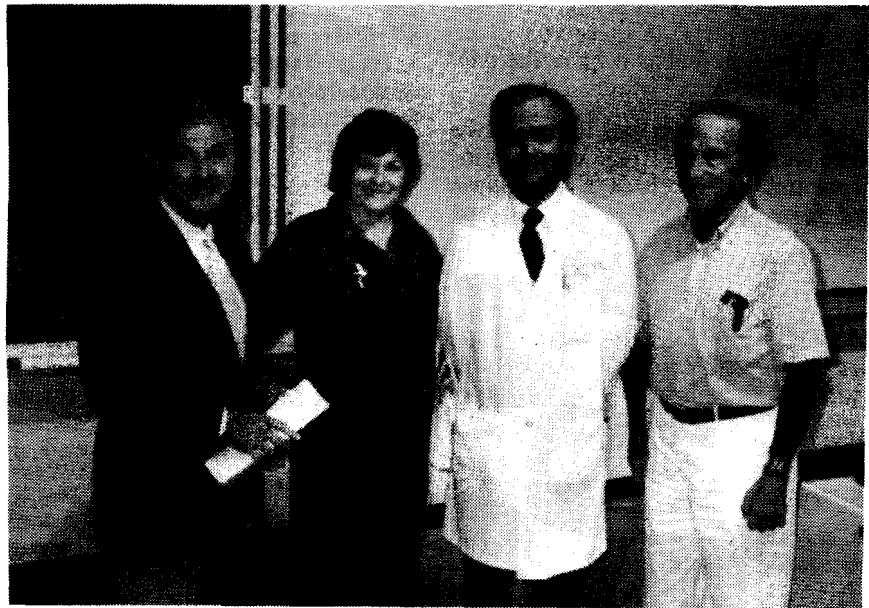
In May with the assistance of Dr. Francis Dyro we sponsored our first Boston area conference. Dr. Dyro, a

neurologist at the Roxboro VA Hospital, Dr. Bruce Korf, a neurogeneticist at Boston Children's Hospital, and Dr. Joseph Addante, a professor of podiatry at the University of Massachusetts, taught us and answered our questions. Eunice Cohen, our Boston support group leader, provided invaluable assistance.

In October Dr. Robert Lovelace, codirector of the neuromuscular clinic at Columbia University Medical Center, and Ann Beyer, a CMTA board member, organized an all day meeting at Columbia Medical Center. Speaking at that meeting were Dr. Lovelace, Dr. Kirk Wilhelmson, a geneticist, Dr. Joanne Borg-Stein, a physiatrist, Dr. Gerald Webber, a podiatrist, Matt Gibble, physical therapist, and Kelley Coch, occupational therapist. These highly qualified professionals gave freely to us their time and talents. These conferences were all brought to the CMT patient/family at no cost to the individual. (At the NY City meeting there was a lunch charge.) The medical professionals who spoke all donated their time. Obviously, we are indebted to all of them and wish to publicly thank them for their outstanding contribution. Our conference philosophy and plans remain the same. Since 1985 we have sponsored regional conferences utilizing area experts accessible to that area's CMT patient. During 1991 we

anticipate continuing this program in cities new to us.

At the annual meeting of the Board of Directors of the CMTA the following officers were elected: Karol Hitt, president; Rodman Steele, Jr., vice president; Diane Freaney, treasurer; Rex Morgan, Jr., recording secretary; and Donald Perrella, corresponding secretary. Patricia Giacomponella was elected to the Board and reelected to the Board were Ann Beyer, Robert Lovelace and Karol Hitt. Other Board members are George Crohn, Jr., Howard Shapiro, Lawrence Wechsler and Lawrence Williams. At this meeting the Board voted to honor an outstanding CMTA volunteer on an annual basis. Hence, the Rebecca Sand Volunteer of the Year award was inaugurated. Of course, the first recipient of that award was Rebecca Sand, a lady who is truly one of the most outstanding people this writer has ever met. (See the Fall '89 CMTA Report) Also honored by the Board and given awards were Drs. Robert Lovelace and Howard Shapiro for coediting the first book devoted solely to Charcot-Marie-Tooth disorders. Dr. Shapiro received an additional award for founding the organization. Dr. Jeffrey Vance of Duke University received an award for his 1989 research locating a CMT gene to chromosome 17, a hitherto unknown site.



Dr. Gurvey, "participant", Dr. Ayyar & George Meyerson at Miami conference



Terry Daino, Dr. Kirk Wilhelmson & Ann Beyer at NYC conference

In October following the annual meeting of the American Neurological Association the Medical Advisory Board of the CMTA met for the first time.

The MAB is chaired by Dr. Lovelace, and that meeting will be reported upon in the next issue of the CMTA Report. Dr. Lovelace has assembled an auspicious group of CMT research and clinical scientists from this country and several foreign countries. We thank them and every other person involved in CMT research and treatment.

Finally, we would be remiss if we did not acknowledge all of our assistance and help from friends and family. We can not begin to name all of the people who have so generously given of themselves, their time, their talents, and their money. This organization could not exist without the help and financial support of its wide circle of friends whether they be an individual, a business, an organization or a corporation. As a result of this support, we are able to serve the CMT community in this country as well as many many foreign countries. We are enriched from our association with all of you.

—Karol B. Hitt, president



Matt Gibble & model patient Gary

Dear Doctor,

In 1959 my mother was diagnosed as having CMT. The doctors also used nerve conduction studies on me (although I had no symptoms) and said I also had the disease. Thirty-one years later my mother (now 81) is confined to a wheelchair with the disorder, but I have shown no symptoms whatever. Is it possible that they were wrong about me or will it perhaps show up when I'm closer to age 50?

My middle sister has had numerous complaints about leg muscles over the years and has had a couple of EMGs but nodefinite answer as to what her problem might be.

My twelve year old son also has the disease. He had problems with his feet as an infant....feet turning in and then his heelcord stopped growing. Problems continued even after surgery to lengthen the heel cord and numerous sets of cast. He has had leg cramps almost nightly for as long as I can remember. Our doctor has finally said my son has the disease.

My mother's maternal grandmother was confined to a wheelchair, but there is no one in the family who knows what her ailment was. We, of course, suspect CMT. No one in the next generation had CMT that anyone was aware of. Finally, out of seven children in my mother's family, only one has CMT, although two died of ALS. What kind of pattern can this possibly suggest?

Also, are tightened heelcords and turned in feet in children in CMT families indications that the children have CMT? My sisters each have a child (adults now) that had feet turning inward in infancy and needed a bar between their shoes to correct the problem. My 17 year old daughter had to wear casts for her feet for the same reason and the neurologist said her Achilles tendons are tight.

C.W., FL

This letter was referenced to Dr. Robert Lovelace, chairman of the CMTA's Medical Advisory Board (MAB). In answer to the questions posed by C.W. this family does have a dominantly inherited neuropathy. The writer probably will develop symptoms of CMT later in life. This is called late age of onset. To
(continued on page 6)

"So, You're Thinking About Starting a Support Group..."



These letters were written by Ann Lee Beyer, the support group coordinator for the CMTA. She can be contacted at 18 Brownstone Way, Upper Saddle River, New Jersey 07450.

Dear Present and Potential Support Group Leaders,

The survey that was sent out was geared to support group leaders. I did not mean to neglect those of you who have expressed interest in starting a Charcot-Marie-Tooth support group but haven't had your first meeting.

The goals of the CMTA are:

- To establish a network of support groups throughout the United States to provide information, advice and encouragement for those who have Charcot-Marie-Tooth and their families.
- To educate the medical community about CMT so that people will be diagnosed early, given accurate information and treatment options.
- To educate the general community so that people will be aware that there is a disorder called CMT and an organization trying to do something about it.

Our hopes are that through support groups and education, those who have CMT will be identified and both funding and research interests will be stimulated. As I have stated before a network of support groups is crucial.

One of the issues that has come up for people wanting to start a support group is that they don't want to know how bad CMT can get. This was one of my fears but the first meeting took away this worry. Most people function very well. One of the main advantages of a support group is sharing information and learning from one another. There are many things people who have CMT know more about than physicians and sharing it makes it available to all.

I hope that you will get the group going. I am here to help. If you haven't seen it, I have written an article on how to start a support group and it is available from the CMTA. I am also available to encourage, support, help over hurdles, and listen. And I would like to hear from you.

Please let me know if you are still interested, how far you have progressed, if you have started but haven't gotten very far, or if you have an idea but haven't gotten beyond the thinking stage, and what I can do to assist you. A short note or a phone call will do. (My number is 201-391-4624.) I can usually be reached early in the morning or in the evening.

Sincerely,

Ann Lee Beyer

Dear Present and Potential Support Group Leaders,

While all of the surveys have not been returned, I am sharing answers to two of the questions because, if you haven't already addressed these issues, they might give you ideas for topics that could be discussed at meetings.

1. The biggest problems of those with CMT:

- Feeling isolated
- Uncertainty of the future
- Lack of information
- No cure
- Acceptance of themselves

2. What people want most from support groups:

- To meet others with CMT
- To both share experience and to learn how to cope
- To exchange and share information
- To talk about problems...including problems with shoes and braces
- To tell their stories and to grieve

All of these issues are important but I think that issues of self acceptance and the need for people to tell their stories and to grieve are extremely important. When we were talking about possible programs in our group, someone said that we really didn't need speakers that often but we needed to talk about things that bothered us, and I would add, accept one another where we are and help one another to work things through.

I have been talking to people who are involved in other support groups. One of the things I am interested in is what works for them. As I find out I will share this information with you. If you think there is something I can help you with, call or write. (201-391-4624)

Sincerely,

Ann Lee Beyer

Ask the Doctor (cont'd from pg. 5)
be more specific we would have to have a copy of the medical records. As to the question of the death of two children from ALS in a known CMT family it is possible that the cause of death was not ALS, but from another form of CMT. Again, medical records would be necessary for an informed opinion. Tightened heel cords and turned in feet in children in CMT families would highly indicate the possibility of CMT in those affected individuals.

Editor's note: See Summer '90 CMTA Report for a discussion about CMT genetics.



Do you know anyone famous who is a CMT patient? Do you know anyone famous who would advocate for CMT? The CMTA is looking for a well-known person to be a spokesperson for CMT. If you know of such a person, contact the CMTA; We will do the rest.

Choosing an Orthopaedic Surgeon

The following letter was written to the CMTA by a CMT mother/patient about her search for an orthopaedic surgeon. The search concluded this spring when her son's surgery was done by Dr. Sigvard T. Hansen, Jr. of Seattle, WA.

Dear CMTA,

I'm glad to know CMTA members are trying to find the best surgeons possible and contacting you for referrals. While I am sorry that the process of finding a surgeon is often frustrating, it is comforting to know that the difficulty of my own search may not be uncommon. As CMT patients we are recognizing more and more that orthopaedic surgery is highly specialized. Many surgeons choose to limit their practice to surgery of the hand, foot, shoulder or hip, etc., and have become highly refined in their particular specialty. Since specialists may also differ according to the procedures and operative techniques they prefer to use when working with a CMT patient, the process of choosing a surgeon can be a confusing one for the lay person. It may be difficult to find the specialists we need and want close to home. I believe that a well-defined communication network is needed to assist CMT patients in their search and to relieve them of the anxiety and monetary cost involved. By sharing personal experiences through the CMTA we can begin to build a national referral system that will connect CMT patients with those orthopaedic surgeons, neurologists and physical therapists who have a strong interest in and high experience level with CMT.

Editor's Note: *The CMTA has been compiling such a list. Additionally, our Medical Advisory Board (MAB) is now in place, and we have access to members of the MAB across the country and several foreign countries.*

Finding a Surgeon

The decision as to who would operate on my son's feet was a tough one that evolved over a long period of time and with a few bitter experiences along the way. I knew I wanted the best surgeon possible but I didn't know what criteria to use in determining who that person would be. Since 1st, 2nd, and even 3rd opinions didn't agree, I began to wonder who was correct. There were

doubts surrounding every opinion. As my experience increased, however, the question gradually changed from "Which surgeon was right, or wrong?" to "Is it possible they all have a valid approach?" I was certainly not prepared to judge. While it is fresh in mind, I'll share some of what I learned during the past three years in the hope the search will be easier for others who wish to make an independent choice.

My son Andrew was seen by five mature and experienced orthopaedic surgeons during the year prior to his surgery in May 1990. However, I learned very little about their training and surgical practice, certainly not enough to make an informed choice. What I did learn is that each of them would approach Andrew in a different way in terms of:

- procedures they would do even though they all had comparable, if not equal, knowledge about the patient;
- technical methods they would use, particularly in working with bone;
- timing of cast changes/size of cast;
- number/size/location of incisions;
- number of operations required;
- post-operative pain management;
- length of hospital stay.

We made an adventure of the out-of-town trips, but they were time-consuming and expensive. Although I learned what each surgeon would recommend for my son, I did not have the courage to ask about the surgeon's experience level with CMT procedures, and went home without the information I truly wanted. Ironically, we chose a surgeon whom we had not met or talked with, but with whom we corresponded, and who provided the name of a patient close in age and degree of CMT deformity to my son. We spoke to this boy and his family, and found that Andrew's surgery would be almost identical to their own experience.

Questions to Ask

When looking for a surgeon, whether it be for the foot or hand, it is helpful to ask a few simple questions about his/her background. You might want to learn the following: Academic background; specialty training on surgery of the foot/ankle (or hand); number of highly deformed feet operated on each year, month or week; special techniques or procedures offered that would

be applicable to CMT, successes with CMT. You may discover that the surgeon has developed a new operation, or learned a new technique and that you are an appropriate candidate! Some CMT patients find this exciting and are more than happy to be one of the "first." They view their participation as a contribution to medical progress and are proud to be involved.

If the CMTA compiled similar information about all the major surgeons (foot/ankle, pediatric orthopaedists, general orthopaedists) that CMT patients are likely to seek help from, I think we could provide a real service for members.

Making the Choice

When choosing a surgeon (CMT or other) the decision can be approached in two ways: (1) by trusting the surgeon implicitly on the recommendation of someone else (CMTA, a satisfied patient, other medical professional); or (2) by studying the subject matter so that a truly informed choice can be made. If the first option is chosen, it is important that the CMT patient be able to accept surgical results without "looking back." He/she needs to move on with life and know the best choice available to him/her was made. If the latter route is chosen, much information is needed to make a genuinely informed decision; information about the disease itself; the patient's unique manifestation of that disease; the surgeon's knowledge of CMT; the surgeon's position on muscle balancing (this is tough in CMT, and there are many variations); the surgeon's attitude toward professional consultations that may suggest other options; the surgeon's ability to individualize for each patient; the surgeon's experience level and confidence with the procedures required; and the surgeon's interest in working with CMT. As information is gathered and options become known the patient begins to define his/her own preferences.

One way to begin an independent selection is to consult the reference section of your library and check a directory of medical specialists for orthopaedic surgeons. There are generally two reference collections, one organized alphabetically according to last name within each medical specialty; the other arranged geographically according to state and city. Directory information is brief, but provides age,

(Continued on page 9)

Support Group Notes



A primary goal of the CMTA is to become a truly successful advocate for those with CMT. Its message must reach the patients, their families, and the medical and research communities. Patient family support groups, a growing and vital part of the CMTA program, inform and support anyone who must deal with this often overlooked disorder.



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

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

California- Burbank

Contact: Sherry Brown (818)841-8421

California- San Diego

Contact: Gary Oleze (619) 792-1427

California- San Francisco

Contact: David Berger (415) 491-4801
After 6:00 pm

Connecticut

Contact: Linda Friedo (203) 374-8478

Florida- Orlando Area

Contact: Mary Beeler (407) 295-6215
Meeting: Third Saturday of every other month

Florida- South (Atlantic Coast)

Contact: George Meyerson (305) 431-3979

Georgia- Greater Atlanta Area,

Contact: Molly Howard (404) 253-5632

Georgia- Western

Contact: Molly Howard (404) 253-5632

Illinois- Chicago,

Contact: Carol Wilcox (312) 445-2263

Indiana- Indianapolis

Contact: Elaine Donhoffner (317) 841-0241
Robert Birdwell (317) 352-0235

Kansas- Parsons

Where: Labette Community College
Parsons, KS
Contact: Tammy Taylor (316) 421-5268

Massachusetts- Boston,

Contact: Eunice Cohen (617) 894-9510

Michigan- Brooklyn

Contact: Robert D. Allard (517) 592-5351

Michigan- Detroit

Contact: Suzanne Tarpinian (313) 883-1123

New Jersey- Central

Where: Princeton Medical Center
Lambert House
Classrooms #1&2

Contact: Janet Selah (201) 281-6289

New Jersey- Northern

Where: Englewood Hospital
Clinic Conference Room
350 Engle Street
Englewood, NJ 07631

Contact: Teresa Daino (201) 934-6241

New York- Long Island

Contact: Lauren Ugell (516) 433-5116

New York- Rochester

Contact: Neale Bachmann (716) 554-6644
Bernice Roll (716) 584-3585

New York- Westchester County

Contact: Kay Flynn (914) 793-4710

Ohio- Cleveland

Contact: Norma Markowitz (216) 247-8785

Pennsylvania- Delaware Valley

Meeting: Holy Redeemer Hospital
Meadowbrook, PA
Contact: Rex Morgan, Jr. (215) 672-4169

Pennsylvania- Pittsburgh

Contact: Garnett McDonald (412) 372-2853

Texas- Greater Dallas Area

Contact: Dr. Karen Edelson, D.P.M.
(214) 542-0048

Utah- Salt Lake City

Contact: Marlene Russell (801) 942-8642

Virginia- Tidewater Area

Contact: Mary Jane King (804) 591-0516
Thelma Terry (804) 838-3279

Letters.....



Dear CMTA,

I want to thank you for sending me the issue of the Interview with Dr. Harold G. Marks. It has helped me to understand our grandson's problem with CMT.

I also want to thank you for sending me the Spring CMTA Report. We never heard of CMT until our grandson was diagnosed as having the disease. We were surprised to read it is genetic.

VT

For the CMTA

This material is presented for educational purposes only and is not meant to either diagnose or prescribe. While there is no substitute for professional medical care for Charcot-Marie-Tooth Disease, these briefs offer current medical opinion that the reader may use to aid and supplement a doctor's treatment.

Memorials

In Memory Of	By
Sophia Jacobson Adler	Ruth Edelheit
Jack Goldman	Ruth Linker
Nora E. Wallace	Nancy Frazier
Fred Bandler	Mrs. Fred Bandler
Harry Brezel	Dr. Karen Edelson
Leo Breeden	Drucilla Lake
Jayne Cahan	Dr. Rebecca Brezel
Lowell Bank	Shirlee Bank
Robert G. Bradwick	M/M Gordon Bradwick
Angela Corio	Joan & Clarence Vater
Jack Friedman	Sondra R. Tracy
Mildred Tolle	John Ronketto
Mildred Tolle	Bonnie L. Lee
Emily P. Beck	Bernie Beck
John Wilson Stuart	Alice Stuart King
Mildred K. Winchell	Phyllis M. Warren
M/M Joseph Galland	Bill Galland

Dear CMTA,

Thanks for the information given to me by the CMTA. I have had Dr. Robert Clark do surgery (5 operations) on both feet. My feet are 100% better. I love the newsletter.

I have enclosed a check. I hope this will help with your work and research.
L.L., MS

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

CMTA, Crozer Mills Enterprise Center, 600 Upland Ave., Upland, PA 19015.

Honorary Gift

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

Send acknowledge to: _____

Name: _____

Address: _____

Occasion:

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

Memorial Gift

In memory of: (name of deceased) _____

Send acknowledge to: _____

Name: _____

Address: _____

Amount Enclosed: \$ _____

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

Gift Given By:

Name: _____

Address: _____

Surgeon Search *cont'd from p. 7*

medical school training, and for some, academic positions and surgical specialties.

Editor's note: *The CMTA has a listing of orthopaedic surgeons who are members of the American Orthopaedic Foot and Ankle Society. For a copy of doctors in your state, send a self-addressed stamped envelope to the CMTA.*

Another step is to look for articles on CMT surgery, note the author(s) and references, and begin reading. This is a little more difficult as a medical library is required to access publications, and the material is often technical. The CMTA is collecting studies on CMT and may be able to provide copies. Many hospitals welcome the lay person to read and photocopy from their library. However, not all studies or articles are considered valid and up-to-date and care must be taken when interpreting what is read. Asking a local orthopaedist or neurologist to act as your advocate in gathering and understanding the information can be very helpful. Physical therapists work closely with patients and surgeons post-operatively and may be a valuable resource in understanding CMT and providing referrals.

When making an independent choice, it is important to seek several opinions to get a feel for how surgeons differ when faced with the same patient. It is extremely important to seek opinions from surgeons who are on a comparable level of expertise with CMT, and travel may be required to accomplish this. Any surgeon who provides a list of patients willing to discuss the actual surgery that has been recommended should be taken seriously.

National Referral System

Wouldn't it be great if the CMTA could recruit some key orthopaedists with a proven record in foot/ankle surgery and convince them to be national consultants on an ongoing basis? Patients and their local orthopaedists would be encouraged to send x-rays and clinical notes to these key people (through the CMTA) for opinions as to what should be done. Or, perhaps, the CMT "experts" could be available for telephone consultations with orthopaedists who already have CMT patients in their practice but would welcome discussion about surgical options. These experts would need to be selected with extreme care, and have an

interest in furthering open communication about CMT with their colleagues.

I hope this letter is beneficial to readers, and encourages them to write to the CMTA about their problems and successes in finding medical specialists.

Sincerely yours,

Rosemary O'Brien

Addendum: Dr. Hansen is a traumatologist, pediatric orthopaedist, and general orthopaedist who sought special training on the foot early in his career and now specializes exclusively in trauma and surgery of the foot/ankle. Young patients are treated at Seattle Children's Hospital, and adults receive care at Harborview Medical Center which is part of the University of Washington. Both hospitals offer auto-injection for post-operative pain management (this is not universally available). Experienced orthopaedists train with Dr. Hansen for foot/ankle surgery. He operates on approximately six markedly deformed feet each week, and about six CMT feet each year. When performing the triple arthrodesis, he frequently uses rigid internal fixation which provides the following benefits: the joints being fused are held so tightly that the surgeon skilled in this method gets a precise result; there is less bleeding; there is less swelling (which makes it possible to transfer tendons in the same operation); there is less pain to the patient. He has a strong bias toward muscle balancing in CMT and balances aggressively (which is very important for some, but not all CMT patients). He readily provides names of patients willing to converse about all of this from the patient standpoint.

□

Note: Andrew is recuperating well and has been working with a Physical Therapist post-operatively in an ongoing therapy program. This has been vital to the post-operative process.

Rosemary feels it is imperative that the patient be under the care of a competent Physical Therapist at all times, before, during, and after surgery.

Rebecca Sand Receives Volunteer of the Year Award

At their May 12, 1990 meeting, the Board of Directors of the CMTA voted to award the organization's first Volunteer of the Year Award to Ms. Rebecca Sand. Acknowledging of her tremendous service to CMTA over the years, the Board also voted to honor the first recipient by naming it the "Rebecca Sand Volunteer of the Year Award."

Before a gathering of relatives and friends, Ms. Sand accepted the award at the July 14 meeting of the Delaware Valley CMT support group. "Rebecca really is an inspiration to us all," said support group chairman and Board member Rex Morgan. "You can't begin to measure the benefits we've gained as a result of her hard work and selfless dedication. Put simply, she has been a wonderful asset to the CMTA."

In Honor Of Rebecca Sand

Contributions were made by:

Mr/Mrs Michael Woolman
Mr/Mrs Richard Woolman
Mr/Mrs Morris Woolman
Rhoda, Steven, Abby & Shira Sand
Patty Woolman
Mr/Mrs. John Schumacher
Mr./Mrs. I. Miller
Mr/Mrs H. Bruce
Ruth Horwitz
Dr. & Mrs. Simon Levin
Sybil & Irv Whitman
Pauline Goldfarb
Rose Freed
Judith Goldman

LETTERS

We want to hear from YOU!

Write us at:

Letters / The CMTA
Crozer Mills Enterprise Center
600 Upland Avenue
Upland, PA 19015

Referrals Available From The CMTA

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

To receive any of these referrals send a stamped self-addressed envelope indicating the geographic areas needed to: CMTA, Crozer Mills Enterprise Center, 600 Upland Avenue, Upland, PA 19015.

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

☐

I want to be in touch!

Name: _____
 Address: _____
 Phone Number: _____

Tell us about yourself:

- CMT Patient Medical Professional
 Interested Supporter CMT Family Member
 Other

Please check the appropriate boxes:

- Put me on the mailing list! CMT Genetics - \$10
 CMT Neurology - \$10 Orthopedic Surgery - \$10
 Physical Therapy - \$10
 Physical Therapy/Orthopedic Surgery - \$15 (on one tape)
 Neurology/Genetics - \$15 (on one tape)

Join the fight against CMT!

Enclosed is my check for:

- \$25 Member
 \$ _____ other

Contributions are tax deductible.

Please make checks payable to the CMTA.

HELP PERPETUATE THE CMTA'S WORK

REMEMBER THE CMTA IN YOUR WILL

You can give hope to thousands of CMT patients by extending your support of the CMTA's programs beyond your lifetime. Whether your legacy is small or large, you can support our programs of education, service and research by remembering the CMTA in your Will.

To make a bequest of cash or other property to the CMTA, your Will (or supplemental codicil if you do not wish to write a new Will), should state:

"I give and bequeath to the Charcot-Marie-Tooth Association, a not-for-profit corporation, organized under the laws of the Commonwealth of Pennsylvania, and having its principal office at Crozer Mills Enterprise Center, 600 Upland Avenue, Upland, PA 19015, the sum of \$(_____) or (_____) percent of the rest, residue, and remainder of my estate to be used for general purposes of the Organization."

A bequest to the CMTA is fully deductible for estate tax purposes. Additionally, you will be providing hope to CMT patients and families now and in the future. You may wish to learn about other gift giving opportunities by consulting your attorney, accountant, and/or tax or estate planning specialist.

☐

Honoraria

In Honor Of

Martin Edelheit
 by Ruth Edelheit
 M/M R. E. VanAlmen
 by Richard E. VanAlmen, II
 Joan Decker
 by Anita Decker
 Norman Cool
 by Robert Cool
 Sophia L. Bradwick
 by Faye Bradwick
 R. Gordon Bradwick
 by Faye Bradwick
 Dr. Robert E. Lovelace
 by Marilyn Prashker
 Mark, Leah & Ben Willingham
 by a friend

For your records, our address is:

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

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

Call for Articles

The CMTA Report welcomes your ideas and article suggestions. For example, you may submit a human interest story telling of your experience of living with CMT. Also, medical professionals can forward articles of a clinical or medical nature that would be of general interest to our readership.

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

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

Write or call the CMTA
215-499-7486

CMT...

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

THE CMTA REPORT

information on Charcot-Marie-Tooth disease from the

Charcot-Marie-Tooth Association

Crozer Mills Enterprise Center
600 Upland Avenue
Upland, PA 19015

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