

# THE NFPMA REPORT

Summer 1989

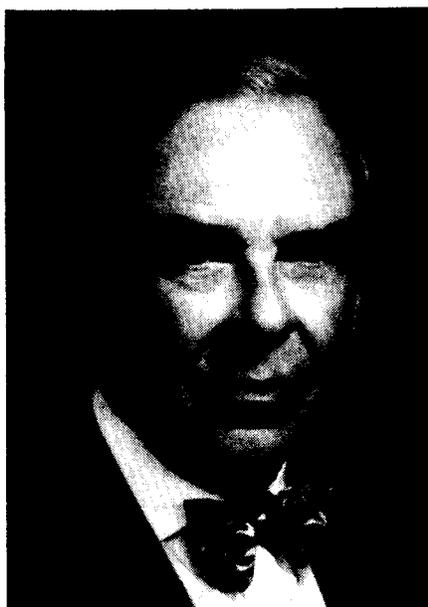
Vol.3/No.3

Providing information on Charcot-Marie-Tooth disease (or Peroneal Muscular Atrophy), the most common inherited neurological disease. Contents © 1989, NFPMA. All rights reserved.

## HAND SURGEON DISCUSSES OPTIONS FOR CMT PATIENTS

*This article was written from a taped discussion between Howard Shapiro, Ph.D. of the NFPMA and James Hunter, M.D.. Dr. Hunter is head of the Hand Clinic in Philadelphia, PA and a hand surgeon of international repute. He is a Professor of Orthopaedic Surgery at Thomas Jefferson University, Philadelphia, Pa.*

During the interview Dr. Hunter discussed hand treatment options for CMT patients. The CMT patient is not frequently referred to hand professionals which is unfortunate as Dr. Hunter feels that much can be done to maintain and improve the quality of life of the CMT patient. Dr. Hunter stressed that early assessment and intervention are vital to preserve and maintain hand function. The cooperative assessment by a hand therapist and a hand surgeon, who are also experienced in treating



Dr James Hunter

peripheral neuropathies, can be the beginning of a program to forestall deformities.

The problems with the CMT hand usually develop gradually with weakness a primary symptom. This is a result of atrophy of the fine intrinsic muscles of the hand resulting in the "claw" hand and flat thumb. There is a loss of coordination and tasks become difficult.

### THE ASSESSMENT

Initially, several procedures are performed which give the physician and therapist a baseline of the hand's functional ability. These include an evaluation of the hand's work

capacity; a sensibility assessment, which is not related to sensation but rather to how the brain is perceiving sensation; complete muscle assessment determining what muscles are at what level of function; and an identification of joint deformities. Dr. Hunter again stressed the importance of early assessment by noting that if the patient is adapting to muscle loss improperly additional harm can be done to other structures not directly involved. Moreover, with early intervention the patient is taught to protect and preserve his/her hand function. Finally, he noted that much knowledge has been gained within the last twenty years about peripheral neuropathies with many resulting new hand therapies.

### TREATMENT

Initial treatment might begin with any number of splinting procedures depending upon the individual's muscle loss and the tasks he/she needs or wants to perform. The splints could be  
(continued, page 2)

IN THIS ISSUE:	
SURGICAL OPTIONS - COVER	
OCCUPATIONAL THERAPY.....	p. 4
RESEARCH NEWS.....	p. 10
MEDICAL ALERT.....	p. 2
GROUP NOTES.....	p. 8
VCR TAPE RENTAL.....	p. 7
GET IN TOUCH.....	p. 7
REMEMBRANCES.....	p. 10

*The NFPMA has moved.  
Our new address is:*

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

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

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

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day and/or night splints, even splints for a specific activity. Splints can include dynamic splints which move with the joint or passive splints which are stationary. The patient is studied for a period of time and is then evaluated. If splinting has helped, the patient might be a candidate for internal splinting which is a surgical procedure. After surgery the patient no longer needs the external splint which is discarded.

### SURGICAL PROCEDURES

The tendon transfer results in an internal splint. (Tendons are tissue that connect muscle to bone.) The surgeon transfers tendons from one location in the hand to another in order to maintain a specific function. Often, tendons are transferred from a strong muscle to a weak muscle and perform a different job than previously. The tendon transfer can be done on the thumb to restore the pinch between the thumb and another finger.

There are other operations that can be done, among them the capsulo desis. This involves fascia and tendon fixing across the joint and allows partial joint function to be retained. The procedure is a way of blocking a joint from extreme motions, but allows some motion. This is good for the palsied hand. It can set the stage for tendon transfer, sometimes giving back function the patient has lost. The surgeon's goal is to return prehension (grasping) with power and stability. Particularly, surgery can restore strength so that the thumb and index or middle finger come together with strength to produce a grip with more power than the "claw" grip. A capsulo desis is done on the middle joint of a finger to allow this.

Dr. Hunter pointed out that patients with polio, cerebral palsy, and accidental injuries are also helped with these procedures.

Occasionally joints require fusing (arthrodesis) because another joint will benefit. These decisions are only made after careful study of the patient. Briefly to recount, capsulo desis is done to permit joint motion, arthrodesis to restrict joint motion, and tendon transfers to control the wrist and thumb and to enhance prehension. The recovery period varies with each procedure and is highly individual. Post surgery therapy is vital to recovery and that therapy must be done by a qualified hand therapist. Dr. Hunter will not operate unless the patient agrees to the post-operative therapy.

### THE WRIST

Dr. Hunter discussed the failing wrist and fusion (arthrodesis). If the wrist is failing and needs fusing it comes after splinting and long study. The wrist may be splinted in several different positions to see which position is the most effective and the patient is the judge. The physician can do individual finger splinting to further determine what is most helpful to the patient. Selective splinting gives information about what is effective for the patient and if surgery will help.

### OTHER OPTIONS

If a patient rejects surgery, the physician has the information to selectively splint the hand for optimum function. Dr. Hunter encourages patients to talk with other patients who have experienced hand surgery and therapy. Patients do get into fixed habits and refuse treatment. Sometimes the mind set of the patient precludes any

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therapy. Occasionally, the family is reluctant to allow therapy for a child. They are not looking ahead to the time when that child must perform adult functions and be self-supporting. Dr. Hunter compared CMT with the position of rheumatoid arthritis several years ago. At that time rheumatoid arthritis patients were not sent to hand therapists for treatment. The resulting problems of the arthritic hand stemmed, not from the disease process which was long gone, but from the neglect of the unprotected deformed joints. Over time daily living tasks performed without protective splinting tear the joints apart. The loss of muscle causes the ligaments to stretch and then joints sag and new deformities result. As the joints wear out there is a loss of balance in the hand. Today, the arthritic patient is seen frequently by the hand surgeon for his/her expert care.

The surgeon qualified to treat the hand has been trained in orthopedic techniques, microsurgery and plastic surgical techniques. Some hand surgeons have received their initial training in orthopedic surgery and others in plastic surgery. They then go on to specialize in surgery of the hand and upper extremities. Additionally, and necessary for the CMT patient, some have gone on for further study of peripheral neuropathies.

## CARPAL TUNNEL SYNDROME

The NFPMA has been informed by neurologists that CMT patients should be wary of carpal tunnel syndrome surgery since the CMT disease process can mimic that syndrome. When asked that question Dr. Hunter agreed that carpal tunnel did present a diagnostic problem. A differential diagnosis must be made to determine if the problem stems from carpal tunnel syndrome or CMT. Nerve conduction studies would be an essential part of the very thorough neurological work-up necessary to make an accurate diagnosis.

In conclusion Dr. Hunter stressed that CMT is a progressive disease that frequently reaches a plateau. When the disease is at a plateau, the unchecked and untreated deformity continues to progress. This deformity progression can be forestalled and perhaps halted with early assessment and intervention by the hand therapy team of surgeon and therapist. However, assessment at any time can certainly be of benefit and is an option that every CMT patient should consider. Treatment started now might enable the patient to perform tasks in later years that would be impossible without the care.

— Karol B. Hitt

## REFERRAL SOURCES

To find a qualified hand surgeon in your area contact the American Society for Surgery of the Hand, 3025 South Parker Road, Suite 65, Aurora, CO 80014; (303) 755-4588.



## GADGETS

Having trouble opening jars, buttoning buttons, or threading a needle? Where do you go for assistance to enable you to do these things or any other task that is a problem? The answer is a unique free service that is a phone call away. ABLEDATA is an amazing service which will put your specific request or need (e.g. buttoning, zipping a zipper, holding a pencil, etc.) into their data bank. The magic of the computer will then come forth with names of products that will assist you in doing that task which is difficult. There is no charge for the first eight pages of information the computer generates for you.

The telephone number for ABLEDATA is (800) 344-5405. This program is funded by a grant from The National Institute for Disability and Rehabilitation Research.

To request Gadget catalogs, contact:

**Bruce Medical Supply**  
411 Waverly Oaks Rd.  
Waltham, MA 12254  
(800) 225-8446

**Enrichments**  
P.O. Box 579  
Hinsdale, IL 60521  
(800) 323-5547

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**Attention  
CMT Patients!**



Dr. James Lupski, of Baylor Medical Center, requests that CMT patients who have a second inherited condition contact him. Please, when you write give the name of the second condition. Also, CMT patients who have a known chromosomal anomaly are asked to contact Dr. Lupski at the NFPMA, Crozer Mills Enterprise Center, 600 Upland Avenue, Upland, PA 19015. (215) 499-7486.

# OCCUPATIONAL THERAPY AND THE CMT PATIENT

## The CMT Hand (Diagrams)

The intrinsic minus hand (claw hand) has lost the function of both the lumbricals and the interossei muscles. Imbalance of the pull of Extensor digitorum communis causes hyperextension of the MP joints and flexion of the IP joints. When this condition exists, the classic picture of the claw appears. The distal transverse arch is flattened completely which, in turn, renders the thumb useless except for the lateral grasp.

(From The Manual on Management of the Quadriplegic Upper Extremitry, by Maude Malick, OTR, 1978, Harmorville Rehabilitation Center)

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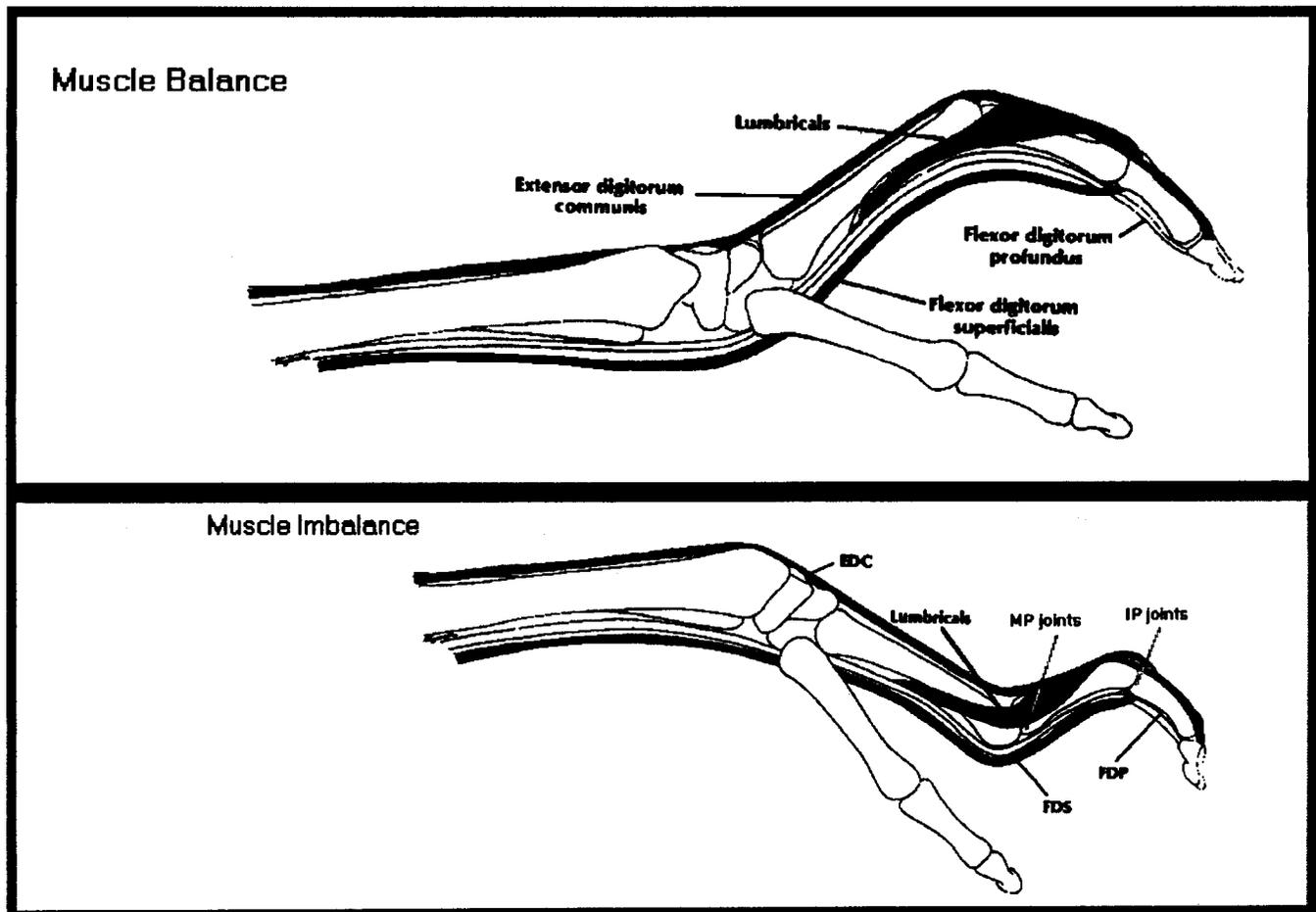
*This article was written from a taped discussion between Howard K. Shapiro, Ph.D., of the NFPMA and Joanne Cassidy, O.T.R.. Ms Cassidy is a Registered Occupational Therapist, Chief of Occupational Therapy at Thomas Jefferson Hospital, Philadelphia, PA, and teaches at Thomas Jefferson University.*

**Shapiro:** How does occupational therapy (OT) relate to the CMT patient?

**Cassidy:** *The occupational therapist should have a vital role in the care of the CMT patient. The initial visit would be comprised of a thorough hand evaluation. We would look at the hand and see how to increase hand function. This*

*can be done by an exercise program and/or a splinting program designed to support structures, to substitute motion, or regain range of motion. The OT is also concerned with direct functional application in daily living activities. Everyday things like buttons, eating utensils and zippers can be problems for the patient. We also modify athletic equipment such as golf clubs and tennis racquets. The key philosophy of the OT is that the work of daily living is an integral part of the rehabilitation program of the patient. In other words, being able to care for your own daily needs is therapy in itself. When treating trauma patients, early on we have the patient care for him/herself. In*

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chronic illnesses we slowly introduce adaptive measures to allow the patient to remain independent.

**Shapiro:** What do you assess on the patient's first visit?

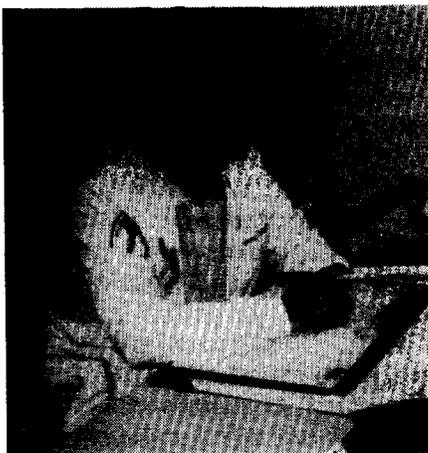
**Cassidy:** A standard basic evaluation includes current hand function, documented grip strength, range of motion, and notation of fixed deformities and weakness. We also discuss current vocational interests, hobbies and daily living tasks. We work very closely with PT's who are more involved with the patients overall physical motion. We too, must consider the patient's whole body range of motion. I read the PT's evaluation as part of my assessment process.

**Shapiro:** Who refers CMT patients to you?

**Cassidy:** Primarily neurologists but sometimes our referrals are from orthopedic surgeons. Here at Jefferson we get referrals from many different sources. Since Jefferson is such a diverse place we are called in from a variety of sources.

**Shapiro:** What is splinting?

**Cassidy:** Splinting is a way of supporting a structure. For some splints we use a low temperature, light weight plastic, warming it in water and then molding it to the patient's hand for a specific purpose. This material is very versatile and we use it for many applications. For example, an easy splint to do is a wrist cock-up splint. It is done to stabilize a weak wrist, and gives the fingers more power. Most of the muscles for the fingers run from the arm across the wrist. They lose power going across the dropped wrist. In the claw hand there is a hyperextension of the MP joints and a flexion (cocking-up) of the fingers (see



Joanne Cassidy

diagrams). This corresponds to the hammer toes of the foot. Splints can be made for a specific activity or for general purpose use. Splints are made from a variety of materials. Elastic straps can be used which allow a range of motion. The splints are highly customized and adapted to the patient's needs and usage. Some splints are for resting night time use and some are task specific. A thumb splint could be used to keep the thumb out, to allow pinching with the index finger.

**Shapiro:** What is the age range of patients you have treated?

**Cassidy:** Most of my patients have been in their 40's and 50's. They were adapting to daily living and work related tasks. I have treated one teen-age patient who was a high school golfer. He was a member of the

#### ATTENTION

If you are moving please send your change of address to:

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Upland, PA 19015

It will help us if you enclose your former mailing label from a previous *NFPMA Report*.

school golf team, and we adapted golf clubs for him.

**Shapiro:** What are some adaptive devices that are used in a home setting?

**Cassidy:** A common tool is the button hook to aid in buttoning. Buttoning requires the thumb and index finger, and for some this is very difficult. The button hook goes through the button-hole and grabs the button. It is then pulled through the button-hole by wrist or shoulder action. Another tool is a long handled shoe horn. This aids the patient who wears AFO's and other adaptive footwear. Elastic shoe laces, which don't need tying, and velcro are two great adaptations for shoes. Sock donners assist the patient in putting on socks.

**Shapiro:** Where can the patient purchase these and other gadgets?

**Cassidy:** When a patient is evaluated, most of the gadgets are available here. There are several medical supply catalogs that sell directly to the patient. (see Gadget Info article)

**Shapiro:** What about exercise?

**Cassidy:** If the nerve has some activity, then the muscle can be exercised. The claw hand is a hand out of balance. One group of muscles is stronger and overpowers a weaker group of muscles, and this distorts the appearance and function of the hand. With specific activities we work to maintain and increase strength in the weaker muscle group to maintain balance of the hand. For exercise we use putties and a wide range of rubber bands to exercise the hand using very specific motions. These are very simple tools and very effective. The exercises and activities can be done in idle and spare time.

(Continued, next page)

**Shapiro:** What would be a long term therapy program for a CMT patient?

**Cassidy:** *It is very specific to the individual depending on his/her disease and lifestyle. In an exercise program there should be a regular routine to maintain muscle strength. A couple of short daily sessions are better than one long session. This lessens the chance of over fatiguing an already weakened muscle. If cramping and pain lasts longer than 1/2-1 hour after exercising the muscle has been over exercised. With weak muscles we begin with a light rubber band and only a few repetitions. I tell people to use the rubber band when watching TV, talking on the phone etc. Putty can be used similarly.*

**Shapiro:** What training does an OT receive?

**Cassidy:** *There are two different approaches. The first is a four year program with a six month internship which gives a degree in occupational therapy. Also a student may be a college graduate in a related field and then goes through an 18-24 month certificate program. In addition, there is a six month internship. Students from both courses must pass a national certifying examination. Successful completion of this registers the OT and he/she then is an OTR. Also, most states have a license requirement, and it varies from state to state. Moreover, there is a two year program to be a certified OT assistant. They work under the supervision of an OTR, and carry out prescribed services. Of course, there are also advanced degree programs in OT.*

**Shapiro:** What are the cost ranges for OT services?

**Cassidy:** *They are very variable with the average being \$75.00/hour with a range of*

*\$50.00-100.00/hour depending on location. Major centers are usually higher. Insurance reimbursement for OT services is poor. In-hospital services are generally covered but out patient care is usually not covered. Unfortunately, most insurance plans do not cover chronic conditions. Medicare and some state programs cover OT services. Private coverage varies from state to state and plan to plan. Our national organization has reimbursement for OT's as a priority item. Some states have a mandatory law requiring reimbursement for OT services prescribed by a physician. This is what the national organization is working to establish in all states.*

**Shapiro:** How can a patient find a hand facility?

**Cassidy:** *The patient should call his/her local hospital and ask for the OT department. If there is none ask for the PT department and they will make a referral.*

— Karol Hitt

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#### Donor Choice Information

For the United Way of Southeastern PA the NFPMA's number is 08823. For other areas you may write in the name and the United Way will contact the NFPMA.

### **ATTENTION CMT PATIENTS: STAND UP AND BE COUNTED**

Dr. James Lupski's CMT questionnaire appeared in the Spring '89 issue. If you have not filled this out, we urge you to do so now. If more copies for family members are needed, you may obtain them by writing:

Karol Hitt/NFPMA  
Crozer Mills Enterprise Center  
600 Upland Avenue  
Upland, PA 19015

Please enclose a stamped, self-addressed envelope. Completed forms may be mailed to the same address. Thank you.

#### **For the NFPMA**

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.

### **LETTERS**

**We want to hear from you! Write us at:**

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

## CALL FOR PARTICIPANTS

The Department of Neurology at the University of Vermont is undertaking a study of strength and fatigue in a number of neuromuscular diseases including Charcot-Marie-Tooth disorders. In this study patients will be tested every three months for a period of two years.

The NFPMA Report is published by the National Foundation for Peroneal Muscular Atrophy, a tax exempt not-for-profit corporation.

This issue was designed by:

Chesapeake Bay Designs  
48 Henry Court  
Hollywood, MD 20636

This research study will provide important information on the natural history of the disease and the effect of exercise. Exercise is good for all of us, but patients with CMT are often worried that exercise might be harmful. Research workers at the University of Vermont are interested in proving the beneficial effect of exercise. They have designed an exercise program and a series of tests to assess the effect of the exercise. They are now looking for CMT patients who would be interested in participating in the study. The exercise program is carefully tailored to the individual's needs.

This is a single center study and, unfortunately, there is no re-

search money for travel reimbursement. If you are interested in learning more about the program, please contact:

Mr. Mike Fillyaw,  
Research Physical Therapist,  
Rowell Bldg., Room 307,  
University of Vermont,  
Burlington, VT 05405  
(802) 656-3252

or

Mrs. Patricia Krusinski  
Patient Coordinator  
Department of Neurology  
One S. Prospect St.  
Burlington, VT 05405  
(802) 656-4177.

The study is being funded by a grant from the National Institute of Disabilities Rehabilitation and Research.

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### VCR Tape Rental

The NFPMA will rent 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 and send it to us with a check or money order payable to the NFPMA, Crozer Mills Enterprise Center, 600 Upland Ave., Upland, PA 19015.

### *I want to be in touch!*

Name: \_\_\_\_\_  
Address: \_\_\_\_\_  
Phone Number: \_\_\_\_\_

#### *Tell us about yourself:*

- |   |   |
|---|---|
| <input type="checkbox"/> CMT Patient          | <input type="checkbox"/> Medical Professional |
| <input type="checkbox"/> Interested Supporter | <input type="checkbox"/> CMT Family Member    |
| <input type="checkbox"/> Other                |   |

*Fill out this form and return it today!*

#### *Please check the appropriate boxes:*

- |   |  |
|---|--|
| <input type="checkbox"/> Put me on the mailing list!                              | <input type="checkbox"/> CMT Genetics - \$10       |
| <input type="checkbox"/> CMT Neurology - \$10                                     | <input type="checkbox"/> Orthopedic Surgery - \$10 |
| <input type="checkbox"/> Physical Therapy - \$10                                  |  |
| <input type="checkbox"/> Physical Therapy/Orthopedic Surgery - \$15 (on one tape) |  |
| <input type="checkbox"/> Neurology/Genetics - \$15 (on one tape)                  |  |

## Support Group Notes

A primary goal of the NFPMA 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 NFPMA program, inform and support anyone who must deal with this often overlooked disease.

There are already several NFPMA support groups. These chapters are spirited and growing stronger, but more groups are needed in other parts of the United States. The NFPMA will gladly help you to set up a chapter in your area. For information contact the NFPMA 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.

### San Diego, California

Contact: Gary Oleze (619) 792-1427

### San Francisco, California

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

### Greater Dallas, Texas Area

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

### Parsons, Kansas

Where: Labette Community College  
Parsons, KS

Contact: Tammy Taylor (316) 421-5268

### Indianapolis, Indiana

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

### Detroit, Michigan

Contact: Marrienne Tarpinian (313) 883-1123

### Chicago, Illinois

Contact: Carol Wilcox (312) 445-2263

### Cleveland, Ohio

Contact: Norma Markowitz (216) 247-8785

### Northern New Jersey

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

Contact: Ann Lee Beyer (201) 391-4624

### Central New Jersey

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

Contact: Janet Selah (201) 281-6289

### New York, New York

Where: Rusk Institute of Rehab. Medicine  
Room RR (6th floor Research Wing)  
400 East 34th Street (at First Ave.)

Contact: Linda Goldfarb (212) 496-0001

When: November 18, 1989 (next meeting)

### Rochester, New York

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

### Boston, Massachusetts

Contact: Eunice Cohen (617) 894-9510

### Delaware Valley, Pennsylvania

Meeting: Holy Redeemer Hospital  
Meadowbrook, PA

Contact: Rex Morgan, Jr. (215) 672-4169

### Pittsburgh, Pennsylvania

Contact: Garnett McDonald (412) 372-2853

### Tidewater, Virginia Area

Contact: Mary Jane King (804) 591-0516  
Ellen Morton (804) 851-7046

Where: Riverside Hospital  
School of Professional Nursing  
J. Clyde Morris Blvd.  
Newport News, VA

### Greater Atlanta, Georgia

Contact: Molly Howard (404) 253-5632  
Sue Saye (404) 565-5950

### Orlando, Central Florida Area

Contact: Mary Beeler (407) 295-6215

Meeting: Third Saturday of every other month

### Fort Pierce Area, Florida (Atlantic Coast)

Contact: Dorothy Stefanovich (407) 461-1016

## **LETTERS.... A FORUM FOR NFPMA READERS**

**Dear NFPMA,**

I never thought there would come a day when a sports magazine would ever have an article of interest for me. But the January 1989 issue of Sports Illustrated featured an article by Gene Wojciechowski on what is known as the "Strength Shoe". My husband eagerly read the article, then urged me to read it as well, and am I glad I did.

The shoe, worn by such players as Walter Payton, Vince Coleman, and Ozzie Smith, has an approximately 1 inch platform at the ball of the foot. It is designed to lengthen and strengthen the Achilles tendon, soleus and gastrocnemius muscles thereby "enabling you to run, jump, and move better." For my son, this might mean the possibility of not having to have heel cord lengthening surgery.

Fortunately, our orthopedic surgeon had read the article too. On our next visit to him, I had prescription in hand, then strength shoes on the way when the day was done.

I have now involved the help of our town's new neurologist Dr. Saty Satya-Murti who has done some research in New York on CMT. (He will be the guest speaker for the Midwest Chapter's first meeting.)

If the shoe works for our son Aron, it will be recommended for several other patients in our community who have similar problems.

I will keep the NFPMA, as well as our chapter informed on our progress.

Tammy Taylor, R.N.  
Support Group Leader  
Parsons, KS

Reference:

Wojciechowski, Gene; SHOES THAT GIVE YOUR GAME A LIFT; Sports Illustrated, Dec. 26, 1988-Jan. 2, 1989.

Editor's note: Tammy Taylor recently reported the shoe was not satisfactory for her son. The shoe, like everything else, has to be judged on an individual basis. We would be interested in learning of anyone else's experience with this footwear.

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**To NFPMA,**

Your organization is given as a resource in my latest book entitled CHOICES NOT CHANCES. This is the only up to date book for the lay public addressing all aspects of heredity and health. It contains a wealth of information about genetic disorders which is pertinent and of interest to all those affiliated with your organization. I would very much appreciate a note in your newsletter or other publication informing your readers about the availability of this book. The previous edition written 13 years ago (entitled KNOW YOUR GENES) appeared in nine languages.

Thank you for helping to increase the much needed understanding about genetic disorders.

Aubrey Milunsky  
Boston University School of Medicine  
Center for Human Genetics  
Boston, MA

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**Dear NFPMA,**

The tapes were very good and informative. The tape, although very technical which it has to be, helps to get the message out to CMT people. We can not keep our heads in the sand....we must move forward to whip this disease. Genetic research is super important for the generations to come. Thanks for your continued efforts.

R.V.A, II, MI

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**Dear NFPMA,**

Being a CMT patient and having a mother who survived to age 91 with it, I want to share some of the problems we faced. The burning sensitive skin should be noted. A dermatologist tells me never to rub those areas..pat them dry. Sitz baths are a relief for the tender mucous membranes. I have good hard plastic braces for my feet to above mid-calf. Plaster casts were taken and the braces were made from the molds. I tried the one size fits all braces that are used after strokes. They should never be used for CMT because the arches aren't right.

J.B., WA

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## Exciting Research News!

by Howard K. Shapiro, Ph.D.

Duke University and affiliated medical centers presented findings which suggest that many CMT patients, perhaps a majority, may share a genetic defect at the same chromosome location. Up to this point molecular geneticists have documented that a small number of CMT kindreds have a genetic neuropathy due to a defective gene on the X chromosome, while a small number of other CMT families have a genetic defect on chromosome one. Dr. Jeffery M. Vance and coworkers looked at DNA samples from six CMT families which did not show evidence of X

chromosome or chromosome one defects. Data from all six families indicated CMT trait linkage to two DNA marker sites known to be on chromosome 17. Patients in these families have the most common clinical type of CMT, autosomal dominantly inherited hypertrophic CMT, or hereditary motor and sensory neuropathy type I. Other laboratory scientists working on CMT genetics have reacted quickly to this exciting news, altering their research plans to include use of chromosome 17 marker site probes to screen CMT kindred DNA samples already at hand. We still do not know how many different genetic varieties of CMT exist. Conceivably, there may be many different CMT genes, each causing some form of peripheral nerve damage. This

would considerably complicate gene mapping studies on CMT, as each genetic variety would require independent investigation. However, the findings reported in the recent chromosome 17 CMT paper suggest that the riddle of CMT genetics may prove to be simpler than expected. This, in turn, will lead to more rapid progress in defining CMT at the genetic and biochemical levels.

No CMT gene has been isolated and no biochemical test for CMT has been developed. Yet, the findings reported by Dr. Vance and coworkers are a significant step towards these goals. Our sincere congratulations to all members of this large research team.

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## NFPMA REMEMBRANCES

Your gift to the NFPMA 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 NFPMA 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 NFPMA by completing the form below and mailing it with your check to:

*NFPMA, 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: \_\_\_\_\_

\_\_\_\_\_

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

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: \_\_\_\_\_

\_\_\_\_\_

## NFPMA Contributions

### Memorials

#### *In memory of*

Jack Goldman  
Dr. Abraham B. Sand  
Arnold Goldman  
Nancy Vigue

#### *Contributor*

Mr. & Mrs. Lawrence Linker  
Mr. & Mrs. Perry Sand  
Mrs. Rebecca Sand  
Mrs. Thelma Brown  
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Ms. Joann Appleton  
Ms. Eva Furman  
Loretta & Morris Woolman  
Ms. Claire Gould  
Mrs. Thelma Brown  
Ms. Joan H. Vater

#### *In memory of*

Thomas J. Henry  
Irving Finkleman  
Bill Cooper  
Dorothy Fellows Bogardus  
Burton Wendkos  
Gack Goldman  
Irving Finkelman  
George Survetnick  
Donni Dodge  
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R. Gordon Bradwick  
Minna & Hart Wurzburg  
Steven E. Firestone  
Mrs. Inez Miller  
Edward Scarsella  
Clarence Vater and family  
Ruth Finkelman  
Mrs. D. Leahy  
Loretta & Morris Woolman

### Honorary Gifts

#### *In Honor Of*

Louise Perry  
Ben Willingham  
Mr. Stephen J. Schaeffer  
Nettie Cooper's 70th Birthday  
Mr. & Mrs. Mitchell Applebaum  
Minna Wurzburg  
Marriage of Miriam & Bruce Medoff  
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Faye Bradwick  
Janice T. Vaughan  
Ruth C. Wendkos  
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Circle #8, Yorkminster Presbyterian Church  
Minna & Hart Wurzburg

## Call for Articles

### ***THE NFPMA 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 NFPMA REPORT** are available at \$2.50 a copy:

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

Write or call the NFPMA  
(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.

# ***THE NFPMA REPORT***

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

**National Foundation for Peroneal Muscular Atrophy**  
Crozer Mills Enterprise Center  
600 Upland Avenue  
Upland, PA 19015

**BULK RATE**  
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