National Foundation for Peroneal Muscular Atrophy

THE NFPMA REPORT

Spring/Summer 1988 Vol.2/No.2

Providing information on Charcot-Marie-Tooth disease (or Peroneal Muscular Atrophy), the most common inherited neurological disease

NFPMA Members Examine Psychosocial Aspects of CMT

On January 30,1988 Columbia Presbyterian Medical Center in New York City held a symposium on "Psychosocial Well Being in Muscular Dystrophy and Allied Diseases." Dr. Howard Shapiro, NFPMA Director of Scientic Program, Ann Lee Beyer, and Linda Phillips Goldfarb, both coordinators of NFPMA support groups, addressed the group on behalf of CMT patients. A summary of their remarks concerning the psychosocial effects of CMT from childhood to adulthood follows.

NFPMA speakers on behalf of CMT at the Columbia University Symposium, "Psychosocial Well-Being in Muscular Dystrophy and Allied Diseases III - Perspectives of the Future" were (from left to right): Ann Lee Beyer, coordinator of the NJ support group; Howard K. Shapiro, Ph.D., NFPMA Director of Scientific Program; Linda Phillips Goldfarb, coordinator of the NY support group; and Robert E. Lovelace, M.D., chairman of the NFPMA Medical Advisory Board.

Families and CMT

by Ann Lee Beyer

Unlike some dystrophies, such as Duchennes, Amyotropic Lateral Sclerosis, or some forms of Friedrich's Ataxia, Charcot-Marie-Tooth disease is not fatal and is often referred to by the medical community as a benign disease. Compared with other neuromuscular diseases, it does indeed appear to be benign, but this does not mean that people who have this disease do not suffer. They suffer in many ways.

First, there is the suffering that comes from having a rare disease. Those affected are "... too often misdiagnosed, frequently misunderstood, and repeatedly misdirected." (*NFPMA Report*, Winter 1987)

Then there is the physical and emotional suffering that comes from having a chronic debilitating illness that often requires the need for surgery and braces.

There is also the anguish that takes place when adults finally find out that their chronic foot or hand problems are more than a family trait or the vestiges of an undiagnosed childhood polio and they have passed on this little-known disease to their children and grandchildren, some of whom are now severely impaired. (The average age of the parents in our study when correctly diagnosed was 35.)

Finally, these patients suffer while living in a society that is not geared to coping with chronic illnesses, a society that does not make allowances for the handicapped. As Robert Murphy, a Columbia University anthropologist, asserts in his book *The Body Silent* (New York: Holt, 1987), our society discriminates against those who have disabilities and treats them as though they are less than equal, making it difficult for them to participate fully in everyday life.

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INSIDE:

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Little information is available about CMT disease and even less about how it affects patients' lives. The purpose of this report is to find out the impact of this disease on the lives of 22 CMT families. Interviews were conducted with 22 parents in families where a child has been diagnosed with the disease. Because individuals do not live in isolation, the family and community can help determine how the patient handles the disease, and everyone in turn is affected by their interaction with a CMT patient. Anthropologist Gregory Bateson has argued that the context in which an illness takes place may be as important as the disease itself.

Telephone interviews were conducted with the 22 parents. This was less than ideal because there was an instrument between the interviewer and the person being interviewed. Participants came from an NFPMA support group for patients and family members. Only one parent in each family was interviewed.

This survey was modeled on Levanthal's "Psychosocial Assessment of Children With Chronic Physical Disease" (1984) and modified to fit families with CMT. Several aspects were examined : the severity of the disease and its complications, both in the adult and the child; how the disease affects one within the family; how it affects the adult in the workplace or the child in school; and whether or not it affects either socially. Additionally the family as a whole was considered.

The following preliminary report will soon be published as a study of the psychosocial effects of CMT upon the patient family.

THE PARENT

Of the 22 parents interviewed, 15 have CMT. They range in age from 28 to 72. Of these, four are mildly affected, nine are moderately affected, and two are severely impaired. Seven have not been diagnosed with the disease. For the purposes of this study "mildly" was defined as appearing to be normal, the disease unapparent to others, but accompanied by some problems. That is, the person with hand involvement who has trouble buttoning or zipping clothing or the person with arches who is beginning to have foot and balance problems. "Moderately" affected is the patient who has had surgery or needs braces or orthopedic shoes. These people may have trouble walking, especially on uneven surfaces or getting on and off the bus. "Severely" disabled is the patient confined to a wheelchair or motorized scooter and limited in a major way.

PERFORMANCE

Having CMT can restrict what people can do. Most of the parents who have CMT say it limits them in some way. These limitations range from minor inconveniences to major accessibility problems. Four people report that they also experience fatigue and that this is as limiting as the outward manifestations of the disease.

FEELINGS ABOUT LIMITATIONS

Eight have feelings of anger or depression because of these limitations and four say they get frustrated. Nine are embarrassed at times. Only two report that CMT does not prevent them from doing anything.

IN THE FAMILY

Ten people report that their ability to function within the family is limited, affecting their performance of everyday tasks or participation in family activities. Because of balance difficulty, carrying a child, using a ladder, walking up and down stairs, carrying groceries, or even taking out the garbage can present problems. For those with

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ATTENTION !

A researcher at Harvard Medical School is soliciting reports of people with genetic diseases who have been discriminated against when seeking life or health insurance, employment, and educational resources other than human services (legal, government, or health). This is a study of societal attitudes involving genetics, and strict confidentiality will be maintained.

Contact: Paul R. Billings, M.D., Ph.D.
Director, Clinic for Inherited Diseases
Harvard Medical School
New England Deaconess Hospital
110 Francis Street
Boston, Massachusetts 02215

Phone: (617) 732-9719

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hand involvement, something as simple as tying a child's shoelace, holding objects in one hand without dropping them, or even using a key, can be extremely frustrating. Several people tell of needing help fastening clothing. One woman tells of having trouble diapering her children and buttoning the small buttons on their clothes. The physicians she visited told her her problem was "nerves" and put her on tranquilizers. Another woman reports that she is unable to get her braces on by herself.

Affected parents also report that going to family outings such as plays, sports events, or picnics is often difficult and sometimes impossible. Curbs, stairs, hills, and having to walk long distances present obstacles that have to be thought about and planned for. Theaters and stadiums with their steep steps and lack of hand rails are not designed to accommodate people with handicaps, even mild ones. One father who is moderately impaired mentioned not being able to go to theStautue of Liberty because of the stairs at the top. A woman with a mild case of CMT says that she cannot take part in the sailing activities which her family enjoys.

THE WORKPLACE

Travel, everyday office tasks, taking care of small children, or standing at a lecturn to give a talk can be difficult for the person with CMT. One woman whose job requires traveling finds managing suitcases and luggage conveyor belts at airports very difficult. Another person who talks to groups finds standing awkward. A man who manages the parts department of an auto dealership has a problem with papers and pencils slipping from his hands. He has to concentrate to keep this from happening as he deals with customers and suppliers.

It also seems that pregnancy can exacerbate Charcot-Marie-Tooth Disease. Two women told of their disease getting worse while they were pregnant. One lost her job because she needed extra time to recover after the birth of her baby. Another found that the hospital nursing she had previously done had become too strenuous. In all, nine parents said that their CMT created problems for them at work.

SOCIAL ADJUSTMENT

The same restrictions that limit people from doing things with families also limit people socially. Those who are severely affected find that needing a wheel chair or motorized scooter to get around can prevent them from being able to visit friends, attend a church function, or even go to the post office or bank because the homes and buildings involved are not barrier-free. Several avoid social

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MEDICAL ALERT

Certain Drugs Toxic to the Peripheral Nervous System

On April 9,1988, the NFPMA held a meeting at the Scottish Rite Children's Hospital in Atlanta, Georgia for the purpose of organizing a support group for the Greater Atlanta area. Neurologist Donal A. Costigan, M.D., addressed the group about the etiology of CMT. In his lecture Dr. Costigan mentioned that certain prescribed medications could have a negative effect upon CMT patients and that **patients should always alert their physicians to their specific medical history.** For the benefit of our readers Dr. Costigan has provided a list of drugs that can produce clinical deterioration in CMT patients.

Adriamycin
Amiodarone
Chloramphenicol
Cis-platinum
Dapsone
Diphenylhydantoin (Dilantin)
Disulfiram (Antabuse)
Glutethimide (Doriden)
Gold
Hydralazine (Apresoline)
Isoniazid (INH)
Nitrofurantoin (Furadantin, Macrodantin)
Nitrous Oxide (chronic repeated inhalation)
Penicillin (Large IV doses only)
Pyridoxine (Vitamin B^6)
Vincristine

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functions where dancing is a major part of the activity. Two find dating awkward. Three say that fatique stops them from doing social things. Two find that they just cannot manage the activities that their friends do. One woman has become an introvert because of CMT.

THE CHILD

In the families we interviewed there are a total of 27 children. The ages range from three and a half to 52. Eleven children are mildly impaired (four females; seven males). Eight have moderate involvement (five females; three males). Six offspring are severely impaired (six females; six males). Two who have been diagnosed exhibit no symptoms.

One family has three children with the disease and three families have two children with it. Of the parents who have more than one child with CMT, two are moderately affected and one has it severely. In the other family neither parent has CMT and there is no family history of it.

IN THE FAMILY

Children who have chronic illnesses often have special needs. Children in wheelchairs or braces, especially if they are young, need help with everyday tasks. They also need help with their physical therapy if it has been prescribed. Corrective surgery can immobilize a child for months, and this means someone must be with this child at all times. This often puts a strain on the family, especially the primary caretaker. One mother with a master's degree had to give up her teaching job because the school would not give her enough leave of absence to provide her child with the post-operative care she needed. She lost income, seniority, and tenure. Parents of 12 of the children report that their children require extra attention.

Sometimes the child with CMT may not be able to keep up with siblings, both younger and older. Parents often find that their children become annoyed or frustrated because they must depend on others to do things for them. Three parents say that their CMT children tire easily and this can interfere with family activities. Another three parents state that their children with CMT tend to get moody and withdrawn. But as one parent wisely pointed out, "We tend to blame a lot on CMT; whether or not the kids' moods are connected with their having the disease, who knows?"

IN SCHOOL

Ten of the children have experienced some problems in school. The inability to keep up with other children, especially in sports or gym, is frustrating to CMT children. The cruelty of other children who make fun of those with disabilities can be difficult for anyone to handle. Several parents reported that their children had been ridiculed at school. One parent recalled her own experiences as a child being called "Ducky" because of the way she walked. Another parent revealed that children used to walk behind her and say, "Quack! Quack!" The mocking these parents experienced is still happening to the next generation. It still hurts! Even those who have received special attention feel the stigma. One little girl, who was an MDA poster child, alternates between showing off her braces and feeling upset because she is different from the other kids.

Some of the problems are directly related to attitudes of school personnel. Some teachers do not encourage CMT children to catch up on missed schoolwork after an absence. A parent of a child whose IQ is near the genius level tells of teachers who will not challenge her child because they refuse to see her physical disability as just that – a physical problem.

A parent of a severely impaired 25 year-old said that being in a wheelchair prevented her son from attending classes that were above the first floor. The school made no allowances for his condition.

SOCIALLY

There were fewer complaints of problems in this area; however, six parents said that CMT did interfere with their children's social lives. The children who seem to be having the most problems are those with severe CMT. This seems to be an issue for both children and adults. As one mother of a severely impaired child said, "Children are often afraid to approach a child with a handicap." Another parent put it differently," It takes a special person ... to befriend a severely impaired child."

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F. Y. I. ...

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IMPACT ON FAMILY

When parents learn that their children have a hereditary disease, they can experience guilt. Thirteen parents say they do feel guilty although all do not have CMT.They also worry about what will happen to their children. Only two parents said that they were not concerned, but on further questioning admitted that they refuse to think about it. A number of parents with CMT also worry about their own futures. This belief is reflected in the high rating some participants assigned to the impact of the disease on their families.

Parents rated the impact that CMT has on their families using a scale from one to five, with one being little or no stress and five being severe.Two parents rated the impact on their families as a one. Five parents said that they would give their situation a two, and five others scored it a three. Two parents found the impact of CMT on their family a four, and finally, six parents stated that life for them is extremely stressful, thus rating a five. Two people refused to answer.

Of the parents who find CMT extremely stressful, three do not have the disease themselves, and only one of these three has a severely impaired child. Of the remaining three parents, two parents themselves are moderately affected with CMT and one is severely affected. This severely impaired parent has a child who is also severely impaired.

Barbara Sabbeth, Ph.D., in her article, "Understanding the Impact of Chronic Childhood Illness on Families," (*Pediatric Clinics of North America*, Feb. 1984) said, "The impact of an illness from the parent's point of view is often surprisingly different from the impact from the physician's point of view. To add to the complexity, the impact of the disease on the mother from the mother's point of view (that is, according to her self-report) may be different from the impact on the mother from the point of view of an observer." So it is not surprising that three of the parents who did not have CMT rated it a five although only one has a child severely impaired.

Harvard psychiatrist Eric J. Cassell in "The Nature of Suffering and the Goals of Medicine" (*New England Journal of Medicine,* Vol. 306,1982) states, "Suffering occurs when an *impending destruction of the person* is perceived; it continues until the threat of disintegration has passed or until the integrity of the person can be restored in some other manner." In a progressive disease such as CMT, this "reintegration" never takes place as the person is never restored to wellness.

SPORTS

Seventeen parents answered yes to the question, "Knowing that your children might be unable to participate in sports, have you encouraged them to develop abilities other than the physical?" This encouragement takes many forms. Some have fostered intellectual development; others have steered their children towards the arts. One parent has arranged for her daughter to have voice lessons. While making their children aware that they may have some limitations, several parents provide no limits, allowing their children to try whatever they want. A parent of a severely impaired child tells her daughter that every person is gifted ; that she must find and develop her talents, and that she is going to do something important with her life.

FAMILY PLANNING

The next question was, "Knowing that CMT is hereditary, have you thought of not having children or limiting your family?" (Eleven of those interviewed were already parents before they were diagnosed with CMT. One of these even had grandchildren.) Six said that they have thought of limiting or not having children. Sixteen said no.

One man and two women were aware that they carried a hereditary disease and still decided to have children. In all three cases this decision was made without knowing that they could possibly give birth to a severely impaired child. Also neither of the women realized that CMT can worsen as a result of pregnancy. Both women found that not only had pregnancy exacerbated their disease, but they also now have children with severe involvement and suffer enormous guilt. When one of the women asked her physician about the risks of having children, he brushed off her concerns by telling her that she had no problem because she could still walk.

A number of adults said that if they had known that they had a hereditary disease, they might not have had children. Four parents decided not to have any more children after having children with CMT. Two parents have suggested that their children seriously consider not having children and have encouraged them to seek genetic counseling before making the decision.

EXPENSES

A family that has a member with a severe impairment can have many expenses. Medical care and the need for special equipment such as braces, orthopedic shoes, or a wheelchair can put a financial strain on a family. **However, many patients are not aware that the Muscular Dystrophy Association can provide medical care and may also pick up several expenses. It is worth-**

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while to call the MDA to inquire about the aid they supply.

Severe impairment can also mean costs that are non-medical in nature but still a necessity for the patient with CMT. There may be a need for household help or special adjustments must be made to homes to allow for a decrease in function.Two mothers talked about the difficulties they had after their children's surgery. Because of her own CMT, one woman was unable to care for her recovering child and could not get the help she needed. This same woman moved from a two story house to a more expensive ranch style because she could not manage the stairs.

SPOUSES

Asked if CMT has affected their relationship with their spouse, six people answered yes, for the better. They had become closer. Nine said that it had a negative effect; two husbands left because they could not handle the stress. Seven reported that CMT made no difference in their relationships.

The next topic covered was whether CMT is talked about within the family. Three parents said no and indicated that their spouses were not open to discussing it. However, for most this was not an issue.

LACK of INFO

Do the respondents think there is a lack of information about Charcot-Marie-Tooth disease? Over 70% said yes. Of these, 77.3% found this lack of information stressful. (18.2% - somewhat stressful; 18.2% - moderately stressful; 40.9% - very stressful) Five people mentioned that they have learned more about the disease since they joined a support group. This has reduced the stress.

ADJUSTMENT

When the interviewers asked if the family has adjusted to having CMT, almost 60% indicated that they have adjusted. Initially it was thought that the degree of severity of CMT experienced by the family might have an impact on the ability of the family to adjust. However, this alone in the parent or child does not seem to determine how well the family says they have adjusted.

Two parents in the study are severely impaired. One has adjusted; one has not. The latter has both a severely affected son and a moderately affected daughter, and was abandoned by her husband because he could not handle the situation.

Of the families who have a child with severe CMT, three say they have adjusted. However, one of these was a parent who could not rate its impact on the family.

When all of the data is compared, it seems that there are many factors that go into how well a family adjusts: the attitudes of the parents towards the disease; the ability of the child to cope with his or her limitations; the ability of the parent to cope with a less than perfect self and/or a less than perfect child; and how the parents and the children interact and support each other.

FEELINGS of LOSS

We all go through life with expectations...expectations for what we will do, become, or have. These expectations are not just for ourselves but also include those closest to us. Some of these expectations are realistic; some are not. When a member of a family has an illness such as CMT, many hopes can be shattered and there is a sense of loss or a need to mourn for what he or she no longer has or may never have. Those who can no longer dance and go to events where dancing is a major part of the activities are feeling this loss. Every member of a family with an illness like CMT has to deal with a loss. The husband whose wife can't go boating, the man who can no longer work, the father whose son will never play ball, the mother whose daughter will never become a surgeon, or the child whose brother or sister is unable to take part in children's games— all experience loss.

The percentage of parents who have feelings of loss and a need to mourn, especially when it comes to their children was 68.2%. Almost 32% do not experience it.

Sometimes the feeling of loss gets mired in denial, the coping strategy that serves as a buffer until a person is ready to accept reality. It may be seen in parents who push their children into various (perhaps unrealistic) endeavors, especially athletics. Two parents report that this is an issue in their families. The hope is that the children's involvement in sports will either slow down the disease process or prove that the child does not have it. In both families it is not all right to talk about CMT.

Then there are parents who just cannot cope with having less than perfect children. One woman spoke of her husband who needed the perfect family and let it be known, subtly, of course, that his children and his wife didn't measure up to his standards.

SUMMARY

There are no easy answers. Unfortunately one cannot present the data as a neat package where everything fits into place. One of the weaknesses of social science is

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SHOES

Every CMT patient has his or her own foot story. For some patients buying shoes is only a minor problem, but for others it can be quite a dilemma. CMT patients would do well to visit a pedorthist, a person trained to provide prescription footwear and related devices to patients referred by the medical profession. For information about pedorthists in your area, contact the **NFPMA Report**.

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that it has attempted to take Newtonian thinking, i.e., cause and effect, and apply it to living systems. It doesn't work because of the many variables involved.

First there is the reality of the disease. Depending on its severity, there can be the need for surgery, braces, or a wheelchair. It can be crippling and limit mobility.

Then there is the personality of the individual and how he or she handles the disease. One person with severe involvement is very angry and lashes out at everyone; another who is also severely impaired has given up and spends each day in a state of depression. It would be easy to say that it is because of their CMT. However, one woman confined to a wheelchair has a very active social life. She has just made arrangements to go hot air ballooning, wheelchair and all!

The family also plays a role. Barbara Sabbeth states , "Each family has its own mythology, which is passed from one generation to the next, thus linking the family to its past and contributing to its future. Family myths based on fact as well as fiction, include attitiudes toward birth, growth, sickness, and death; they shape and are shaped by the serious illness of a child."

Finally, society's attitude toward those with disabilities has a profound effect on the CMT patient. There is a need for a more aware public so that those with disabilities will be seen as valuable contributors to the society. There is also a practical need for barrier-free buildings so that **all** can have access to the theater, the opera, the bank, the post office, sports events, etc. Schools should be sensitive to the many needs of children, including those who do not fit the "average" profile. A major responsibility lies with the medical community. They need to be better informed in order to make earlier diagnoses. More financial support to CMT research is essential to unravel the mystery of this disease.

This survey of patients and their families is offered as one further step to the realization of these goals. If this insight into the challenges faced by those with CMT enlightens, educates, inspires, or encourages patients and readers, significant progress will have been made.

More psychosocial interviews on next page.

PSYCHOSOCIAL ASPECTS — LOOKING BACK:

Adults Remember CMT in Childhood and Adolescence

by Linda Phillips Goldfarb

For most people childhood and adolescence are carefree times. For a child with a disability this may not always be the case. A child with a disability such as CMT may have a whole set of worries and concerns that would never enter the mind of a healthy child.

This report was based on interviews with 20 adult CMT patients (10 male and 10 female) with regard to their childhood and adolescent experiences. For most of these people, the age of onset was before the age of twenty. It should be noted that many of the women reported age of onset as being in the first decade. This is contrary to statements in the medical literature, which describes typical onset regardless of gender, as being in the second decade. Most reported that the degree of severity at onset ranged from what they described as "mild" to "bad."

Half of the interviewees said that having the disease caused some apprehension towards school. Several people noted that they were afraid to raise their hand in class, as they did not wish to focus attention on themselves and their disability. A few also said they had become introverted. Part of the problem may lie in the fact that teachers frequently misunderstand this disability.

More than half admitted that their relationships with peers were somewhat affected. Fifteen out of 20 said that they felt "different." One woman said, "I felt that the friends who chose me were not reallly the *top people* or they would not have chosen me. If they really knew what I was like, they would have dropped me. In college I was quite popular, and I questioned, 'What was wrong with these people ? Couldn't they see that I was a *subperson* ?' "

Another woman said, " I was ashamed of being unable to go up steps without railings. So I did not participate in certain activities. I could not explain or talk about it and I became more introverted."

Most of the women and some of the men felt that having CMT had affected their self-image. Several people said that they were never able to meet their parents' expectations. One of the women said, " My mother expected me to do everything - learn to ride a bike, roller skate, play tennis, and so on. I failed miserably at all of them. My mother has the disease as well, but she must have felt great guilt for passing it on to me. As a result, I've always felt that it was my fault, that I just didn't try hard enough."

Most of the women and some of the men stated that CMT had definitely affected their high school years. Many said that gym was a major source of anxiety. One man said, "Gym was very embarrassing. The teachers didn't believe that I had a physical problem and I was expected to do what everyone else could do. One gym teacher tried to degrade me in front of the rest of the class."

Few of the people interviewed were able to participate in sports. Among those who could, none did particularly well. "I was always the last to be chosen for any team," was a frequent response. However, several people said that thay were good swimmers. Many described themselves as "clumsy." One man said, "My father and brothers were all good baseball players. I could never measure up to them. I felt that it was my fault, that I was just

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memo:

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Looking Back,

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being lazy."

When asked if they had "overcompensated" in other areas because of their physical limitations, many reported that they had done very well academically. Several of these people went on to list numerous non-physical activities. One man reported that he had meticulously built little model ships, which was an enormous challenge, considering that he has a hand tremor.

For most, CMT was a minor problem with regard to dating, but half of the people said that they would have chosen different careers if they had not had the disease.

Anxiety, frustration, and insecurity seem to be very significant factors. Denial, fear, and anger were somewhat less significant; humiliation and depression were acknowledged as part of the problem, but to a lesser degree. One person stated, "Always having to look for ways of getting around physical barriers puts one in a constant state of anxiety." Another recalled, "Going anywhere was such a chore that the tension and anxiety took away the joy of what I was going to do. I was drained from anticipation, fear, and anxiety from planning which way to go."

One woman who had inherited CMT from her father was unable to speak to him about her fears regarding the progression of the disease. He simply could not deal with it. Two women of the group had entertained thoughts of suicide in adolescence.

The men generally downplayed the emotional impact of the disease on their childhood. When asked to remark about a rather painful medical procedure called an EMG (electromyogram),here too, the men minimized the physical pain commonly associated with this procedure. Perhaps men generally deny pain, whether it is emotional or physical.

The psychosocial implications of CMT for children and adolescents may broadly affect their lives, involving their school activities, relationships with peers, and participation in sports. In addition for the adolescent, CMT may become a factor in their social life and career planning. There was nothing in the data to indicate that CMT limits academic performance. In fact, many patients will "overcompensate" for their disability by actively pursuing other activities where performance is not affected by physical limitations.

In conclusion, it should be noted that subjects were not classified according to their clinical status because their medical records were unavailable to the interviewers. It would be desirable that a more ambitious study be undertaken to better understand the impact of CMT.

GADGET INFO

So you almost strangled yourself yesterday getting the neck button on your shirt closed and last week it took 20 minutes to open a jar of pickles. Well, take heart. We are seeking out sources of household products to help the handicapped . If you have any favorite gadget sources, please let us know. To obtain gadget information try contacting:

Bruce Medical Supply
Dept. CMT
411 Waverly Oaks Rd.
Waltham, MA 02154
(800) 225-8446

Enrichments, Inc.
P.O. Box 579
145 Tower Drive
Hinsdale, IL 60521
(800) 343-9742
In IL, AK, HI (312) 325-1625

Independent Living Aids, Inc.
27 East Mall
Plainview, NY 11803
(800) 262-7827
In KY (800) 537-2118

Psychosocial Aspects for Adults — Next page

ADULT PATIENTS SHARE VIEWS OF CMT

by Linda Phillips Goldfarb

Twenty additional adult CMT patients (10 male and 10 female), who were not previously interviewed in the childhood study, were the subjects of this report. Questions covered a broad spectrum of issues.

Polish artist Zdzislaw Lindner (right) pauses for conversation before one of his paintings at the Tunnel, a New York disco. His art was displayed on May 19 as part of an art show/benefit for the NFPMA. Mr. Lindner, a CMT patient, has lived in the United States for six years and his work can be seen at the On The Wall Gallery in Manhattan.

Photo by Robert Quagliata

As in the childhood study, the women generally reported age of onset in the first or second decade with a smaller percentage of males reporting the same. It is possible that the men were affected earlier, but did not consider the disease to be a problem until more overt symptoms began to manifest themselves, i.e., difficulty in climbing stairs.

Again, as in the first set of interviews, this new group of male CMT patients tended to minimize their problems, both physical and emotional. It was the interviewers' opinion that there was a definite denial factor involved in male responses as evidenced in their reactions to questions about the painful electromyogram.

Both men and women had concerns about self-image and physical attractiveness. Half of those interviewed believed that the disease had no bearing on their feelings of masculinity or femininity. However, a few people made the following comments. "I would feel more feminine if I could put on an attractive pair of shoes. People say, 'You dress so well, but look at your shoes!' " "I wear braces. I think that my boyfriend would really like to see me in high heels and a miniskirt." " I don't see myself as feminine, because being feminine to me means being graceful, and I am clumsy."

Few people had difficulty discussing CMT with their friends, and conversely, very few of their friends had difficulty discussing the disease with them. However, many people said that CMT did limit their activities with their friends.

Most of those interviewed stated that their colleagues knew of their disease. When questioned about disability discrimination at work, most found that this was not a problem. However, one woman had a different story. She worked in a city

(Continued, Page 12)

The NFPMA wishes to extend special thanks to Kathy Jones (second from left), art benefit chairman, for her outstanding efforts in organizing the May 19 event. Seated with her are two other members of the committee, Linda Phillips Goldfarb and Thomas M. Pitegoff. (See back panel for committee.)

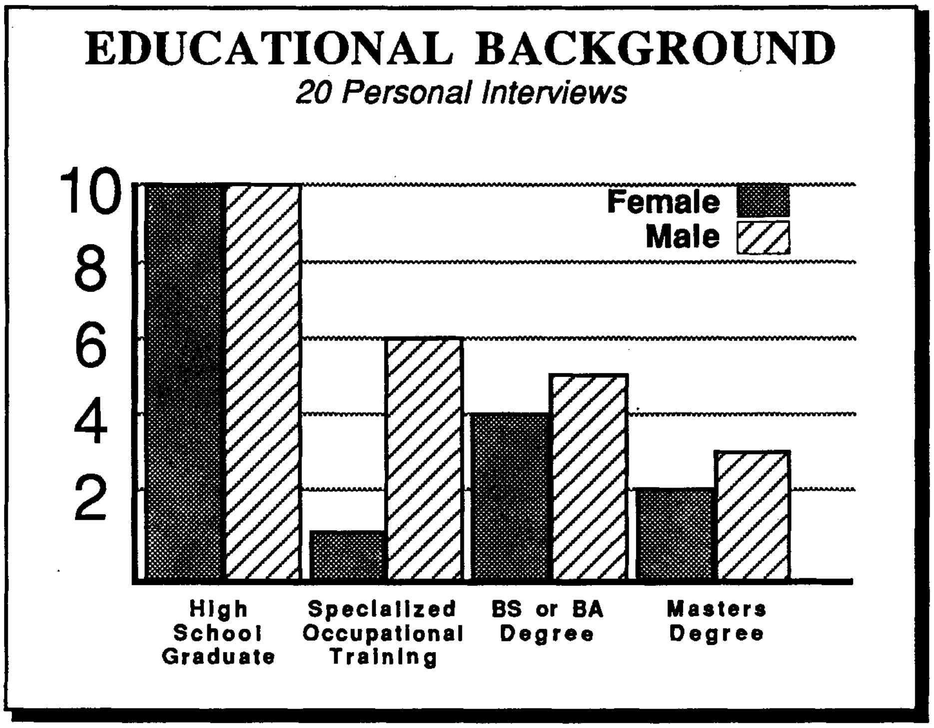
Photo by Robert Quagliata

Adults and CMT,

(Continued from Page 11)

government building with very steep steps and no handrails. She was only able to enter the building by having a co-worker meet her every morning to help her up the stairs. This woman wrote numerous letters to government officials and was eventually able to get the attention of a local congressman who ultimately had railings installed.

The pursuit of higher education was not limited by CMT in these people. Less than one-third had to change careers because of physical limitations. A maintenance supervisor is now limited to performing special non-physical tasks such as training others. One woman who was a biology teacher now works as a lab specialist. Standing all day and having to constantly raise her hand above her head to write on the blackboard had become too difficult. She also found that having to grade papers after school was too much work, as she suffers from great fatigue.



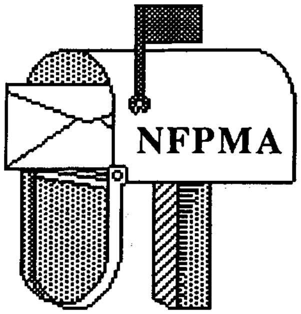
Almost all of the women and many of the men found that CMT had definitely limited their ability to do household chores. Fatigue was frequently cited as a factor. Seven out of ten women stated that CMT had a profound effect on their day to day living, whereas the men tended to minimize this. One of these women said, "I have to plan everything I do in advance. I have to get up much earlier than other people in order to get a parking spot close to my office." Another woman who has difficulty with hand function reported that before she goes shopping, she approximates what she is going to spend in every store and sets up her bills with paper clips to avoid having to fumble around in her wallet. Many others agreed that they had to plan out every day in advance, carefully pacing themselves to avoid faitigue. A woman who lives in a major city said, "I wear leg braces that are rigid. I cannot take buses and subways because of the steps. I am dependent on cabs, and I can't find one before 10:00 AM. I know that I cannot make appointments before 10:30 AM. I have to plan my appointments so that I am not in very crowded areas at peak hours. I don't get around that well and I fear being knocked over in a crowd." Another woman has learned to do much of her shopping through catalogues.

More than half were still able to drive a car although some required special hand controls. Only five were still able to dance without difficulty. One man who loved to dance had to give it up and now can only slow dance holding onto his wife for balance. A young woman who likes to go to discos is glad that free-style dancing is back in style. She used to fear that her partner would swing her out and she would lose her balance.

The issue of family planning was discussed in several ways. Very few people had chosen not to have children even though this is an inherited disease. Some had chosen to limit their families. The issue of a pre-natal screening test (although not presently available) and the decision to abort a fetus bearing the genetic defect were considered very sensitive areas by the participants and something that was difficult to discuss.

The CMT patient experiences many emotions in dealing with this disease. Frustration is a nearly unanimous problem. "People assume that we can do what anyone else can do because we look so normal, and often we can't." Anxiety, fear, and anger are issues for many. Insecurity, denial, and guilt also play a part, although one woman said, "I think that having this disease has given me courage, strength, and determination."

For most of the adult patients interviewed for this study, CMT has a broad and multifaceted affect on their lifestyles, family relationships, career decisions and a variety of other aspects of their personal lives.



NFPMA Mailbox

....The tapes were very interesting and informed me about my condition. Most doctors here do not know what CMT is. Thanks for your trouble and I would be interested in future tapes.

G.B.
Tullahoma, TN

******

....The tape has helped me immensely more than you can possibly imagine—in understanding the surgical options for a child with CMT. For people such as myself with no medical background, the tape allows us to play and replay until we understand and many questions are answered in the process. Then there is the added benefit that time spent in dialogue later with the surgeon can address more complex issues effectively.

R.O.
North Wales, PA

******

I am 28 and was diagnosed with CMT at 17. I do not know much about the disease and would appreciate any information that you could send. The information is for my children's sake since I need to know if there are definite symptoms to look for in them. I can remember from my childhood the difficulty I had keeping up physically with my peers and not understanding why. I want to be able to help my own children cope with this if it happens to them.

My symptoms are not as advanced as one of my sisters who is seven years older than I or my mother's. Can the progression of this disease cease or slow down from one family member to another? The only thing that seems to be getting more intense with me is the muscle and joint pain.

I am glad to know that my family is not alone in dealing with CMT.

B.M.
Largo, FL

Editor's Note: The progression of CMT varies greatly among individuals, even within members of the same family.

******

Very recently your **NFPMA Report** was sent to me. I have had CMT all of my adult life but this was the first time I had ever seen any information concerning it in print. It affects several members of my family and is now beginning to show up in my oldest daughter.

. . . I was amazed to know it affects approximately 125,000 Americans.

My family has traced CMT from our great grandmother, our grandfather, and our father to us. It is very exciting to find out there is information that might be helpful.

N.F.
Danville, IN

******

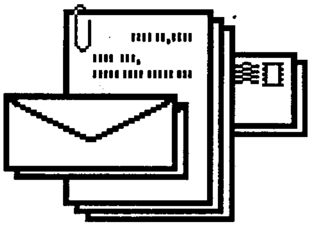
. . . I am very pleased that Dr. Shapiro is one of the leaders in research of CMT, as I remain convinced that every research group should have a member or members who have the disease that is being researched. This, I think, can be very helpful in expediting matters and publicizing the disease. No one knows what it is like until he has been there. Amen. . . .

W.G.
Pound Ridge, NY

******

Thank you for sending me the **NFPMA Report**. In the future I plan to establish an organization similar to the NFPMA here in Japan. Continuous information about your activities is greatly appreciated.

Akio Ohnishi, M.D.
Yahatanishi-Ku, Kitakyushu,
Japan



The **NFPMA Report** is published by the National Foundation for Peroneal Muscular Atrophy, a tax exempt not-for-profit corporation incorporated in the Commonwealth of Pennsylvania (established 1983). Editors of the **NFPMA Report** are Mary Cawley Tracy and Lydia Brennan. Letters and inquiries may be addressed to:

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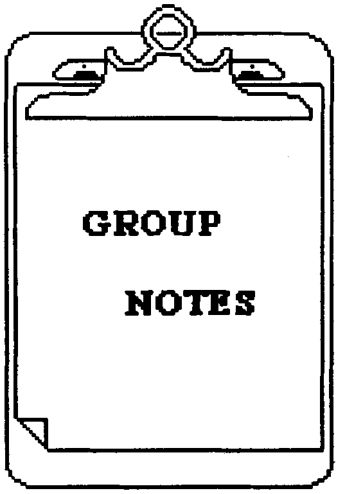

GROUP NOTES

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) 664-6010.

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

New York

Meetings: every other month

Next: October 8

Where: Rusk Institute of Rehabilitation Medicine
Room RR 610 (6th Fl. Research Wing)
400 East 34th Street (at First Avenue)
New York, NY 10016

Time: 1:00 - 4:00 PM

Contact: Linda Phillips Goldfarb (212) 481-3419

New Jersey

Meetings: every other month

Next: October 8, December 10

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

Time: 10:00 AM

Contact: Ann Lee Beyer (201) 391-4624

Delaware Valley

Meeting: This group held its second meeting at the Holy Redeemer Hospital in Meadowbrook, PA on July 30.

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

Tidewater, VA Area

Meeting: early fall, to be announced

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

Contact: Molly Howard (404) 594-1697

Cleveland, Ohio

Contact: Norma Markowitz (216) 247-8785

Orlando, Central Florida Area

Contact: Mary Beeler (407) 295-6215

Fort Pierce Area, Florida (Atlantic Coast)

Contact: Dorothy Stefanovich (407) 461-1016

Greater Dallas, Texas Area

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

San Diego, California

Contact: Gary Oelze (619) 792-1427

Call ☎ *today !*

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Peroneal Muscular Atrophy (CMT) ...

... is the most common inherited neurological disease, affecting approximately 125,000 Americans.

... is also known by its historical name, Charcot-Marie-Tooth disease, for the three doctors who first reported on it in 1886.

... is slowly progressive, causing deterioration of peripheral nerves which control sensory information and muscle function of lower legs and forearm voluntary muscles.

... causes degeneration of peroneal muscles (located on the front of the leg below the knee) subsequent atrophy of additional lower leg and forearm muscle groups.

... 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 in more severe cases may cause scoliosis (curvature of the spine).

...does not affect normal life expectancy.

...has no effective treatment, although physical therapy and moderate physical activity are beneficial.

... is usually inherited in an autosomal dominant pattern, affecting half the children in a family with one PMA parent.

... is present in the world-wide population, with no apparent link to any one ethnic group.

THE NFPMA REPORT

information on Charcot-Marie-Tooth disease from the
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University City Science Center
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Philadelphia, PA 19104

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