

“ELEPHANT MAN’S DISEASE” SUFFERERS LEARN TO COPE

By Joanna Paxinou

Coping with an incurable disease is one of many factors that Neurofibromatosis victims must contend with. Known as Von Recklinghausen’s Disease or the Elephant Man’s Disease, NF sufferers must also deal with ostracism, loss of potential job opportunities, and psychological torment from a society that largely considers the disease offensive.

Because so little is known about the disease and because so much must be done to help NF victims in medical research and in living with NF, a nucleus of three families, spearheaded by NF victims Donna Oettinger, Wayne resident Margery Schaefer and the Huber family, is forming a North Jersey Neurofibromatosis Chapter in cooperation with the New Jersey Self-Help Clearinghouse which assists people in forming their own “self-help” groups.

An inherited neurological disorder that manifests itself in multiple benign tumors just below the skin and on the brain, eyes, internal organs and spinal cord, NF affects 100,000 people in the United States making it among the most common hereditary diseases afflicting people of all nationalities and races. Unable to be detected by prenatal tests and considered incurable, NF can range from slight skin pigmentations (such as the tell-tale sign of six or more “café au lait” spots) to massive disfiguring tumors, which can result in blindness, deafness, paralysis, severe curvature of the spine and bone enlargement and deformation. In about 5-10% of NF patients these tumors become malignant.

With an unpredictable progress, the diseases can also cause learning problems, speech impediments, headaches and seizures. The disease gets progressively worse, although not all patients experience all symptoms. There is no cure but there is treatment to control symptoms, which consists of removal of life-threatening and disfiguring tumors. Despite all these possible consequences, most NF victims manage to lead normal, productive lives.

Disease Known for Over 200 Years

Although there has been an awareness of NF for over 200 years, little research has been done and very few doctors have been trained to even recognize the symptoms. This lack of training and knowledge of the disease on the part of doctors causes many victims to be misdiagnosed and mistreated for many years further adding to the trauma of NF sufferers. There is, therefore, a pressing need for doctors to be educated in detecting the disease.

Generally believed to be genetic, some researchers now feel that spontaneous mutation can also cause it. The National Institute of Neurological and Communicative Disorders and Stroke, a part of the National Institute of Health, is now studying genetics and cell growth in NF patients. Research is also being conducted at Washington University School of Medicine in St. Louis, Mt. Sinai Medical Center in New York City, and at Baylor College of Medicine in Houston, Texas.

The proposed North Jersey Neurofibromatosis Chapter plans to ask doctors to help them set up a two-way referral system by which victims can be referred to knowledgeable local doctors, and social workers and doctors can refer NF victims to their organizations for help. In addition, the group plans to raise funds for research. Through enlightening the public about NF, the group hopes, as Margery stated, “doctors will be more understanding and people more sensitive.”

Traumatic Toll

The toll on the family of NF victims can be traumatic as well as financially and emotionally draining. Besides the painful treatments and possible damage surgically removing tumors can cause, NF victims very often have to contend with people ignorant of the disease who fear “it’s catching.” Very often, especially with children, questions and taunts become crueler, such as calling a six-year-old victim a monster.

Margery, a healthy, vital mother of three, who described years of contending with insensitive people, trying to hide small skin lesions over her body, and not making friends easily because “people are afraid NF is contagious,” commented, “You finally have to live with it—you can’t hide it or yourself.” Finding her family and friends supportive, she has learned to accept herself and her affliction and wants to, along with the other members of the group, help other NF victims.

Asked to sum up the purpose of their proposed chapter, the NF victims stated, “All we want to do is help people. We’re not afraid of it. We want to tell other NF victims and their families ‘Maybe we can help you.’” Ideally, the long-term goals of the chapter are to find a cure or control for this disease and provide a common denominator to help in medical research. As one sufferer commented, “An individual can’t do anything— together maybe we can do something.” The group plans to keep up with new developments and help each other as needed.