Neuropathy Action Foundation

Awareness ▪ Education ▪ Empowerment

A Guide to Neuropathy

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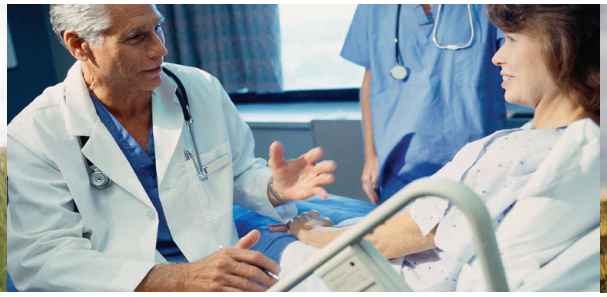
The Neuropathy Action Foundation (NAF), a 501(c)(3) non profit, is dedicated to ensuring neuropathy patients obtain the necessary resources, information and tools to access individualized treatment to improve their quality of life. The NAF increases awareness among physicians, appropriate institutions, the general public and public policy officials that neuropathy can potentially be a serious, widespread and disabling condition, which may be treatable when appropriate medical care is provided.

Our Vision

The Neuropathy Action Foundation (NAF) will be a premiere patient advocacy organization ensuring that neuropathy patients have access to individualized medications, IVIG and other treatments through patient empowerment and advocacy.

Our Goals

Patient Empowerment: The NAF educates and assists neuropathy patients on how to become informed advocates for their healthcare.

Public Awareness and Physician Education: The NAF actively supports programs that create public and physician awareness of neuropathy, the use of IVIG and other remedies to improve patient care through NAF activities and services.

What Is Neuropathy?

Neuropathy means disease of one or more nerves. Neuropathies can be sensory, motor or autonomic. Sensory nerves tell us how things feel. Motor nerves stimulate muscle contraction and initiate movement. Autonomic nerves control functions that our bodies don't consciously regulate, such as breathing and heart rate. Symptoms present depend on the type of nerve and the location.

The symptoms are usually numbness, tingling, pain and/or weakness. Someone with a neuropathy may notice one of these symptoms or he or she may notice that it is harder than normal to do something, such as raising an arm over the head, getting up from a seated position or walking up stairs.

If you experience such symptoms, when you see your doctor he or she will most likely refer you to a neurologist, a doctor who specializes in the diagnosis and treatment of nerve and nervous system disorders. This doctor will take a detailed history, perform an in depth neurological examination and order various tests such as an EMG (electromyography) and nerve conduction studies. This will help him or her to determine the cause of the neuropathy and the best course of treatment for you.

There are several things that cause neuropathies. The information that follows will give you a better understanding of the different types of neuropathies as well as their causes, symptoms, treatment options and prognoses. It is important to recognize that symptoms can be similar in the different types of neuropathies, sometimes making diagnosing the exact type a long and challenging process. This is why a thorough work up is so important, as determining the best treatments is based on a conclusive diagnosis. Please discuss this information with your neurologist.

What Does A Complete Neurological Exam and Workup Consist Of?

Complete health history: It is imperative that the neurologist spend time finding out the history of your symptoms, including type, onset, duration and location. Specific details about what brings on the symptoms, what relieves them and the sensations that occur are imperative to arrive at a definitive diagnosis.

Neurological evaluation: In addition to the history of the symptoms, the neurologist will also do a physical examination of reflexes, strength and the ability to feel various sensations, as well as an evaluation of the autonomic nerves. Again, this aids in an overall diagnosis.

Blood tests: The neurologist will again order some lab tests to find out the cause of the neuropathy. This includes tests to check for vitamin deficiencies, immune responses, blood sugar levels or the presence of any toxins or infections.

EMG: An EMG, or electromyography, electronically measures and records muscle activity. This tells the neurologist the location of any muscle, nerve or neuromuscular junction damage as well as its cause.

Nerve conduction studies: This test finds damage in the peripheral nervous system by measuring the efficiency and speed of electrical signals of the nerves. This tells the neurologist of any abnormality of the nerves. An EMG and nerve conduction studies usually go hand in hand.

MRI: An MRI (magnetic resonance imaging) may be performed to rule out any other causes of the neuropathy, such as trauma or impingement.

Lumbar puncture: A spinal tap may be done to determine the presence of protein or something else in the cerebral spinal fluid, which may indicate the cause of the neuropathy.

Nerve or muscle biopsy: A small piece of nerve or muscle may be taken in order to look for the cause of the damage. These tests are only done if some very specific conditions are suspected or to completely rule out a specific condition.

What Is Idiopathic Neuropathy?

Idiopathic means of no known cause. This type of neuropathy is very common, making up about a third of all neuropathies. This diagnosis simply means that the causative factor is unknown.

Symptoms include numbness, tingling and pain starting in the feet or shins. Balance when standing and/or walking is usually affected. There may also be muscle cramps.

The neurologist will perform various exams and tests to determine if there is a cause so that the best treatment is chosen. Once the neurologist rules out a cause and identifies the neuropathy as idiopathic, the treatment plan is formulated. Again, this

all depends on the location and type of nerves affected.

Treatment consists of over-the-counter pain medications and safety precautions due to balance issues and loss of sensation.

What Is Diabetic Neuropathy?

Diabetic neuropathy can occur when a person with diabetes mellitus has ongoing blood sugar levels that are above normal. The excess sugar in the blood stream can slowly damage the nerve fibers over time. Because virtually any nerve can be affected, symptoms can vary greatly and include numb or painful feet, indigestion, constipation, dizziness, bladder problems and impotence.

Tests include EMG and nerve conduction studies, blood work to check glucose levels as well as other counts and a thorough exam including a neurological assessment. It is important that a thorough workup be done and that assumptions not be made in people with diabetes that high blood sugars are the reason for the neuropathy, as proper treatment depends on the actual cause.

Treatment depends on which nerves are affected and the subsequent symptoms and problems that the person experiences. The first step is to maintain blood glucose levels within normal limits through compliance with diabetic medications and diet. It is vital to prevent further damage and problems from occurring. Further intervention can include proper foot care, treating indigestion and constipation with medications and dietary management, possible antibiotics for any bladder infection and pain relief.

What Is Hereditary Neuropathy?

Hereditary neuropathies are genetic in origin, meaning that they can be passed on or inherited without someone knowing it. Examples include Charcot-Marie-Tooth Disease and Hereditary Neuropathy with liability to Pressure Palsies.

Charcot-Marie-Tooth (CMT) Disease is the more common hereditary disorder and affects both motor and sensory peripheral nerves. Symptoms may include weakness and/or pain in the feet and lower legs and later on in the hands. Deformities can result from loss of muscle bulk, such as foot drop. Symptoms can appear in the teenage years or early to mid-adulthood. Progression is slow.

There are many different types of CMT. It is caused by a mutation in the genes that ultimately affect the structure of the peripheral nerve axon and/or myelin sheath, which in turn affects brain-nerve-muscle communication.

Treatment includes physical and occupational therapy, use of leg braces and use of other assistive devices to facilitate safety and to help offset some of the deformities that can occur.

Hereditary Neuropathy with Liability to Pressure Palsies is a hereditary neuromuscular disorder that makes someone more susceptible to nerve damage from compression or repetitive movements. This pressure can be very minimal and result in numbness or weakness in the limb that was injured. Symptoms last longer and occur more frequently than a limb “falling asleep.”

An example of the resulting injury is carpal tunnel syndrome. Therefore, even people who present with what may appear to be simply carpal tunnel syndrome should have a complete workup to determine that there isn’t a hereditary neuropathy.

Diagnosing a hereditary neuropathy may include a comprehensive history and physical including a neurological exam, EMG, genetic blood testing and nerve biopsy. Treatment consists of over-the-counter pain medication and ergonomic training to avoid pressure-related and repetitive movement injuries from everyday activities.

What Is Immune Neuropathy?

There are several types of immune or inflammatory neuropathies. These are very rare and require a thorough work up. The two most common are Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) and Multifocal Motor Neuropathy (MMN.)

CIDP is believed to be an autoimmune disorder, meaning that the body’s immune system attacks itself. It causes both weakness and sensory abnormalities that develop over several months before stabilizing or improving. Relapses and remissions may occur. The severity of CIDP can vary from mild to severe and it can affect any age group and either gender. CIDP is not painful, although some patients complain of unusual or troubling sensations in the hands or feet.

MMN is a disorder characterized by weakness and muscle atrophy that usually affects the hands and

ankles, but can affect other muscles. The diagnosis can be very confusing since MMN can cause minimal weakness in just a few muscles, or relatively severe weakness in all limbs. Because it only affects strength but not sensation, MMN is sometimes mistaken for ALS (Amyotropic Lateral Sclerosis, or Lou Gehrig's disease). Unlike ALS, MMN is an immune neuropathy and responds to the same treatments.

Both MMN and CIDP occur when the immune system attacks the peripheral nerves. The attack occurs against either the myelin sheath, which is the protective covering over the nerves, or interferes with specialized "ion channels" in nerves that help electrical signals pass up and down the long processes. As a result, electrical impulses that carry the signal to move or feel are blocked and nerves may be injured.

These are both rare neuropathies that are very difficult to diagnose. They usually require the opinion of a neurologist who specializes in neuropathy, called a neuromuscular disease specialist. The diagnosis depends heavily on nerve conduction studies that prove nerve signals are conducting abnormally and on a complete examination of the nervous system by a specialist who can recognize the unusual patterns of muscle weakness and sensory loss. A lumbar puncture is sometimes needed to check for high protein levels, and in rare cases, a nerve biopsy may be required. For MMN, a specialized blood test called anti-GM1 antibodies is useful if it is positive.

The most common treatment for either MMN or CIDP is called IVIG (Intravenous Immunoglobulin). Most, but not all, cases will improve and the symptoms will decrease or even go away in a few weeks up to a few months after IVIG is started. IVIG is given at intervals ranging from every two weeks to every other month. Responses tend to wear off so treatments need to be repeated.

Also, since the diagnosis is often difficult, many treated patients turn out not to have an immune neuropathy. One can even consider the first treatment as part of the diagnosis. Patients who do not respond tend to remain stable, which is the natural course of most neuropathies. Since IVIG is very expensive and the diagnosis can be confusing, it is important that the neurologist check for responses regularly to determine whether ongoing treatments are needed and what doses are necessary.

When IVIG fails or when it does not completely manage the symptoms, a steroid called Prednisone is often added or used to treat CIDP. Prednisone causes more side effects than IVIG but works well to treat the disease. Prednisone is not used for MMN, since it does not work for that particular neuropathy. There are several other medications that can treat immune neuropathy, and these may be used when the condition is severe or difficult to treat.

Treatment of Painful Neuropathies

Pain is an important symptom of many types of neuropathy, especially idiopathic and diabetic neuropathy. Patients usually complain of discomforts like burning or tingling that go along with having numbness. When pain is present, medications that are specialized for managing nerve pain are often used. These include medications like Elavil, Gabapentin, Lyrica and Cymbalta, to name a few. More severe cases may require stronger narcotic medications.

Other Causes of Neuropathy

Neuropathy may also be caused by certain vitamin deficiencies, alcohol abuse, side effects from medications and from certain infections. Once again, diagnosis is based on a thorough neurological examination and specialized testing. Since neuropathy symptoms occur in other neurological disorders, it is not uncommon for people to go through a lengthy process before receiving an accurate diagnosis.

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