



GBS/CIDP
*Foundation
International*

**Guillain-Barré Syndrome and
Chronic Inflammatory
Demyelinating Polyneuropathy**

*Serving patients of GBS, CIDP, MMN and other variants
with support, education, research and advocacy*

Multifocal Motor Neuropathy (MMN)

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Supported by an educational grant from
Talecris Biotherapeutics

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What is Multifocal Motor Neuropathy?

Multifocal Motor Neuropathy (MMN) is an acquired, chronic but treatable condition affecting multiple motor peripheral nerves that connect the spinal cord with the muscles. Damage to these nerves by the patient's immune system results in muscle weakness most often in the arms with minimal or no sensory changes. It develops in a chronically progressive or step-wise manner and over time leads to wasting or atrophy of the muscles controlled by the involved nerve.

Symptoms Males are affected with MMN almost three times as frequently as females and experience an earlier onset with a peak incidence between 50 and 60 years of age. Onset can occur between 20 and 60 years of age. The disease affects one to two people per 100,000 population.

Symptoms progress in a slow or stepwise fashion over 20 or more years. In the beginning, the weakness is most common in the hands as opposed to the legs and asymmetric - or not the same - on each side of the body. The hand weakness can result in significant disability for patients by interfering with their ability to write, button a shirt, handle a fork and knife or turn a key in a lock. Even if symptoms initially occur in the legs, over time the arms become more involved. During the disease course, patients may require the use of a cane or brace but generally do not become wheelchair dependent even late in life. Most patients can continue their jobs, unless the job is physically demanding. Muscles that are used for swallowing, speech and chewing are not involved.

Involved muscles may develop wasting (thinning) or atrophy over time and be associated with twitching of muscle fibers that can be seen under the skin. Patients, particularly at disease onset, have normal sensory function and can feel

pain, cold or hot sensations and touch. In rare cases, some sensory loss may occur over time.

Cause The cause of MMN is not fully known. Evidence, however, supports that damage by the immune system to multiple, focal areas in peripheral nerves underlies this disorder and that immune treatment improves neurological function.

Peripheral nerves carry electrical signals from the spinal cord to muscle. The motor nerve cell or neuron in the spinal cord extends a fiber or axon which, like an electrical wire, is covered by segments of insulation or myelin that abut one to another. In MMN focal areas of myelin and axon damage block the transmission of signals from the brain to the muscle and result in muscle weakness.

At least 30% to 50% of MMN patients have proteins in their blood called antibodies that bind large lipid/fat molecules, GM1, located primarily on the surface of motor axons in focal areas not covered by the myelin segments (nodes of Ranvier). Whether GM1 antibodies participate in nerve damage is not known, although they are useful markers for MMN and help to support the diagnosis. At this time there is no evidence to support that MMN is inherited from parent to child.

Diagnosis The diagnosis of MMN is based on a combination of the patient's clinical findings and electrical testing of peripheral nerve function.

Motor nerves are stimulated with a small amount of electricity at two or more sites (for example the wrist and elbow) and the resulting muscle movement (for example at the base of thumb) is measured. From this study, it can be determined whether the nerve can conduct an electrical impulse and if so, how rapidly. In MMN the conduction of the electrical impulse is blocked in a focal segment of at least two or more nerves while conduction of the impulse in sensory fibers in the

same nerves is normal. Such findings are diagnostic of MMN and help to distinguish this treatable disorder from a form of motor neuron disease known as amyotrophic lateral sclerosis (ALS) or Lou Gehrig's disease in which the immune system has little involvement and for which no effective therapy exists. This test also helps distinguish MMN from other forms of acute and chronic inflammatory neuropathies including Guillain-Barré Syndrome and chronic inflammatory demyelinating polyneuropathy.

Treatment Almost 85% of MMN patients show short and long-term improvement with intravenous immunoglobulin (IVIG), making it the best first line of treatment. Patients typically respond to a course of IVIG within hours to days, with improved strength that may last three to six weeks to months. Repeated doses of IVIG are required in most patients to maintain the improvement although there may be gradual deterioration over years. Other therapies such as corticosteroids are not as effective and in some MMN patients can lead to worsening.

IVIG is the only agent shown to be beneficial in randomized and controlled trials. Immunosuppressant drugs including cyclophosphamide and azathioprine may be effective in individual patients but have not been tested in controlled trials. However, side effects such as cancer of white blood cells associated with long-term use of these drugs limit their benefit in a chronic disorder like MMN.

Physical and occupational therapy are helpful to maintain activities of daily living through exercise as are aids that facilitate specific function such as a hook used to button a shirt. Excessive exercise may result in increased muscle pain and poorer muscle function if not monitored properly.

The GBS/CIDP Foundation International

The organization was founded in 1980 by Estelle and Robert Benson to help patients stricken with Guillain-Barré Syndrome, a paralyzing peripheral nerve disease. Since then it has expanded its outreach to include CIDP, Miller Fisher Syndrome, Multifocal Motor Neuropathy, Lewis Sumner Syndrome and others. With over 180 chapters located in 30 countries, the Foundation's mission is global.

The GBS/CIDP Foundation's medical advisory board is comprised of some of the world's leading experts in the neuromuscular field.

Need Help? If you have MMN or a related demyelinating disorder or know of someone who does and would like assistance or information, please contact the Foundation.

Other services include:

- Local chapters
- Newsletter – The Communicator
- Overview for the Layperson
- Physician referrals
- Educational symposia
- Research funding
- Patient advocacy

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