First Global Multifocal Motor Neuropathy (MMN) Quality of Life (QOL) Patient Survey Identifies Needs in Education and Treatment

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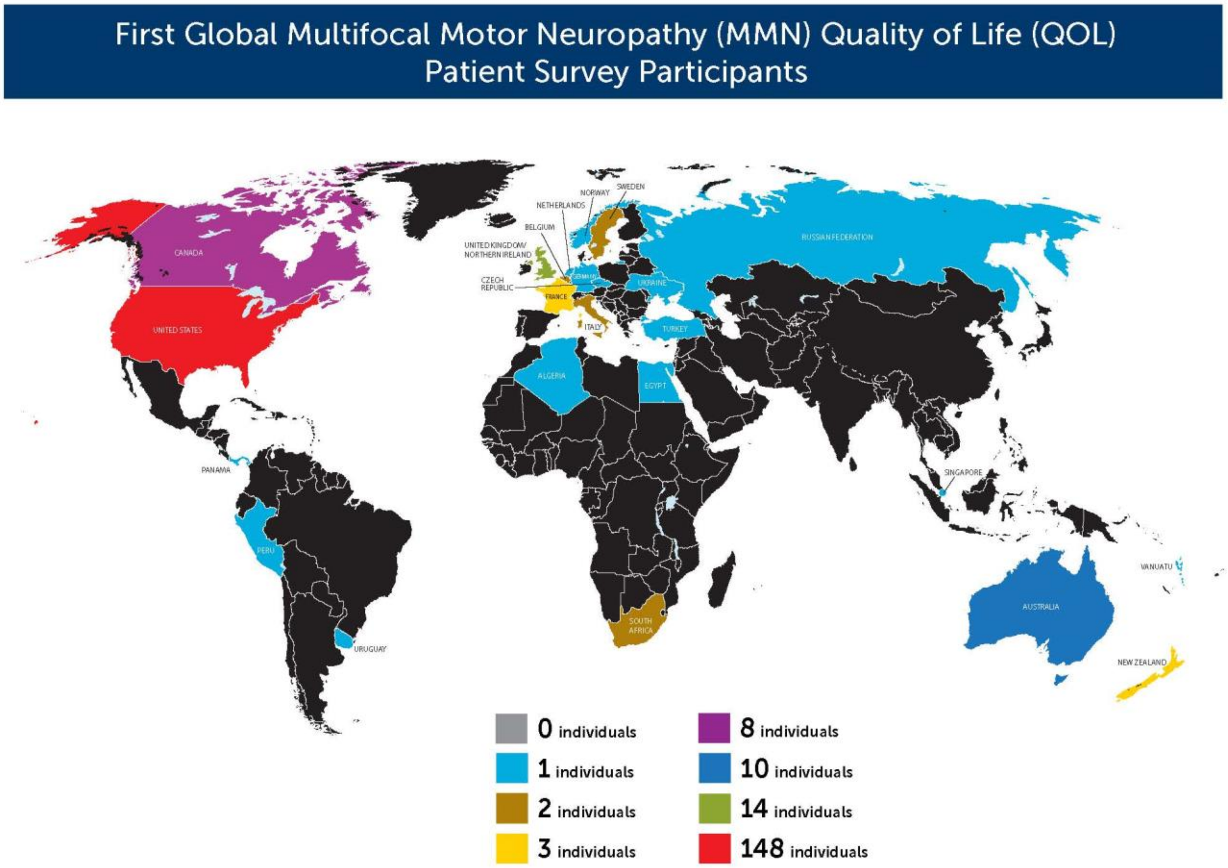
Multifocal Motor Neuropathy (MMN) is a rare condition in which multiple motor nerves are attacked by the immune system resulting in progressive, asymmetric muscle wasting and weakness. MMN affects 0.6 in every 100,000 individuals worldwide and it is twice as likely to affect men than women. While MMN does not usually shorten an individual’s life, it is associated with motor dysfunction and moderate to severe disability.

The Neuropathy Action Foundation (NAF) wanted to learn more about the impact of MMN on patients, and whether there might be gaps in education or treatment that, if addressed, could help individuals better control their disease and take charge of their care. To do this, the NAF recently conducted the first global Multifocal Motor Neuropathy (MMN) Quality of Life (QOL) Survey.

Incidence and Demographics

The MMN QoL survey was completed by 211 individuals around the world. Survey respondents were about evenly split between men and women (47% vs.53%) despite the higher reported incidence in men. Survey participants predominantly identified as white or Caucasian (89%); 2% of survey participants were African Americans, 2%

were Asian/Pacific Islander, 2% were Hispanic, and 5% identified as other/multiple ethnicities. Most of the respondents in this survey were from the United States. Amongst the US participants, responses were highest from Texas, Florida, and California.



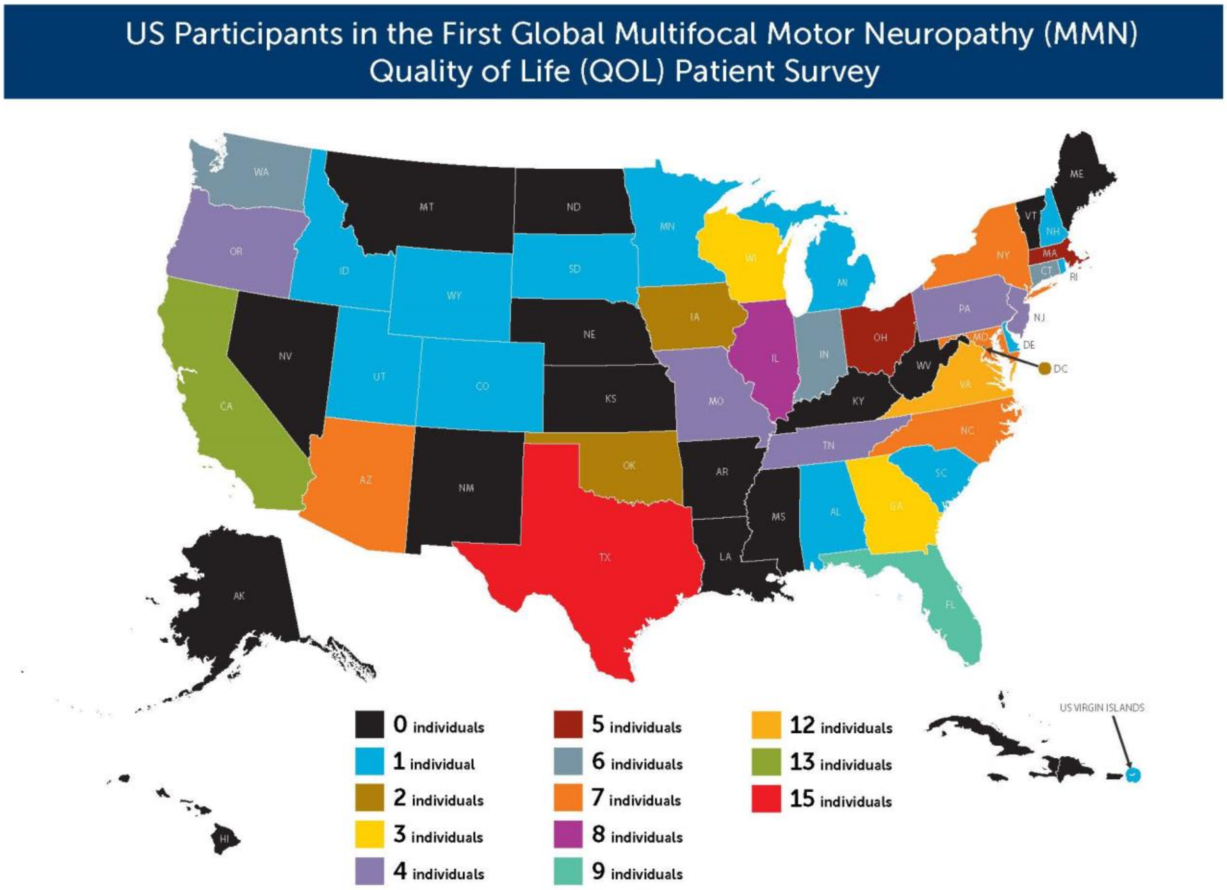
First Global Multifocal Motor Neuropathy (MMN) Quality of Life (QOL) Patient Survey Participants

Country	Number of Participants	Percentage of Total Participants
Algeria	1	0.48%
Australia	10	4.81%

Belgium	2	0.96%
Canada	8	3.85%
Czech Republic	1	0.48%
Egypt	1	0.48%
France	3	1.44%
Germany	1	0.48%
Italy	2	0.96%
Netherlands	1	0.48%
New Zealand	3	1.44%
Norway	1	0.48%
Panama	1	0.48%
Peru	1	0.48%
Russian Federation	1	0.48%
Singapore	1	0.48%
South Africa	2	0.96%

Sweden	2	0.96%
Turkey	1	0.48%
Ukraine	1	0.48%
The United Kingdom of Great Britain and Northern Ireland	14	6.73%
The United States	148	71.15%
Uruguay	1	0.48%
Vanuatu	1	0.48%
Total	208	

Figure: US Participants in the First Global Multifocal Motor Neuropathy (MMN) Quality of Life (QOL) Patient Survey



State	Number of Participants	Percentage of Total Participants
Alabama	1	0.63%
Arizona	7	4.43%
California	13	8.23%
Colorado	1	0.63%

Connecticut	6	3.80%
Delaware	1	0.63%
District of Columbia (DC)	2	1.27%
Florida	9	5.70%
Georgia	3	1.90%
Idaho	1	0.63%
Illinois	8	5.06%
Indiana	6	3.80%
Iowa	2	1.27%
Maryland	7	4.43%
Massachusetts	5	3.16%
Michigan	1	0.63%
Minnesota	1	0.63%
Missouri	4	2.53%
New Hampshire	1	0.63%
New Jersey	4	2.53%

New York	7	4.43%
North Carolina	7	4.43%
Ohio	5	3.16%
Oklahoma	2	1.27%
Oregon	4	2.53%
Pennsylvania	4	2.53%
Rhode Island	1	0.63%
South Carolina	1	0.63%
South Dakota	1	0.63%
Tennessee	4	2.53%
Texas	15	9.49%
Utah	1	0.63%
Virginia	12	7.59%
Virgin Islands	1	0.63%
Washington	6	3.80%
Wisconsin	3	1.90%

Wyoming	1	0.63%
Total	158	

Diagnosing MMN

Diagnosing MMN can be difficult for many reasons. The diagnosis is based on motor symptoms and signs, and because MMN may only affect a single nerve and cause minimal weakness in one limb, it may take a long time to achieve an accurate diagnosis. MMN causes general weakness and typically develops asymmetrically, usually beginning in the hands. The hallmark of the disorder is the diagnosis of "conduction block" on the nerve conduction studies, and missing this sign can contribute to misdiagnosis.

The survey confirmed these ongoing issues with the accurate diagnosis of MMN. The survey data showed that 52% of the respondents had the characteristic symptoms of conduction block. Respondents also reported symptoms of weakness in both the upper body (49.5%) and lower body (47.5%), with 38% of the participants reporting asymmetric upper motor weakness, including weakness, loss of use, and pain in the hands.

While the majority of patients reported that they were diagnosed between the ages of 41 and 65 years (57%), more than 67% of those completing the survey reported that it took more than one year to be diagnosed. Alarmingly, more than 44% reported that it took 2-3 years or longer to be accurately diagnosed. The majority of the misdiagnoses were reportedly done by Neurologists (55%) and Family Care Providers (21%).

Because MMN only affects motor nerves, the symptoms can mimic those of amyotrophic lateral sclerosis (ALS). Many people ultimately diagnosed with MMN are initially told they have ALS, which can be very alarming, and in this survey, the participants reported that the most common misdiagnosis was indeed ALS (13%) with others being diagnosed as carpal tunnel syndrome (6%), multiple sclerosis (MS) (4%), and ulnar nerve entrapment (3%). Incredibly some patients were told they were suffering from hysteria, too much caffeine, menopause, old age, some were given no diagnosis at all, and still others were told there was nothing wrong with them. The first sign reported by 35% of participants was weakness. Other signs and symptoms included muscle cramps (9%), numbness (10%), pain (10%), and finger or foot drop (11%).

Treatment

When asked about treatment options, 91% of participants reported receiving intravenous immune globulin (IVIg) and 9% reported receiving subcutaneous immune globulin (SCIg) therapy. Other therapies being used to treat MMN were gabapentin (7%), and pregabalin (5%). The participants were knowledgeable about their treatment, as demonstrated by 85% who knew which brand of IVIG they were receiving. Participants reported a wide variance in how frequently they were receiving their treatments and the duration it took to receive treatment, as well as the effects they received from these therapies. A reduction in symptoms was reported within one week of their first Ig treatment by 36% of respondents.

Figure: Frequency of IVIG Treatment

The majority of respondents must go for IVIG infusions every 6 weeks or less.

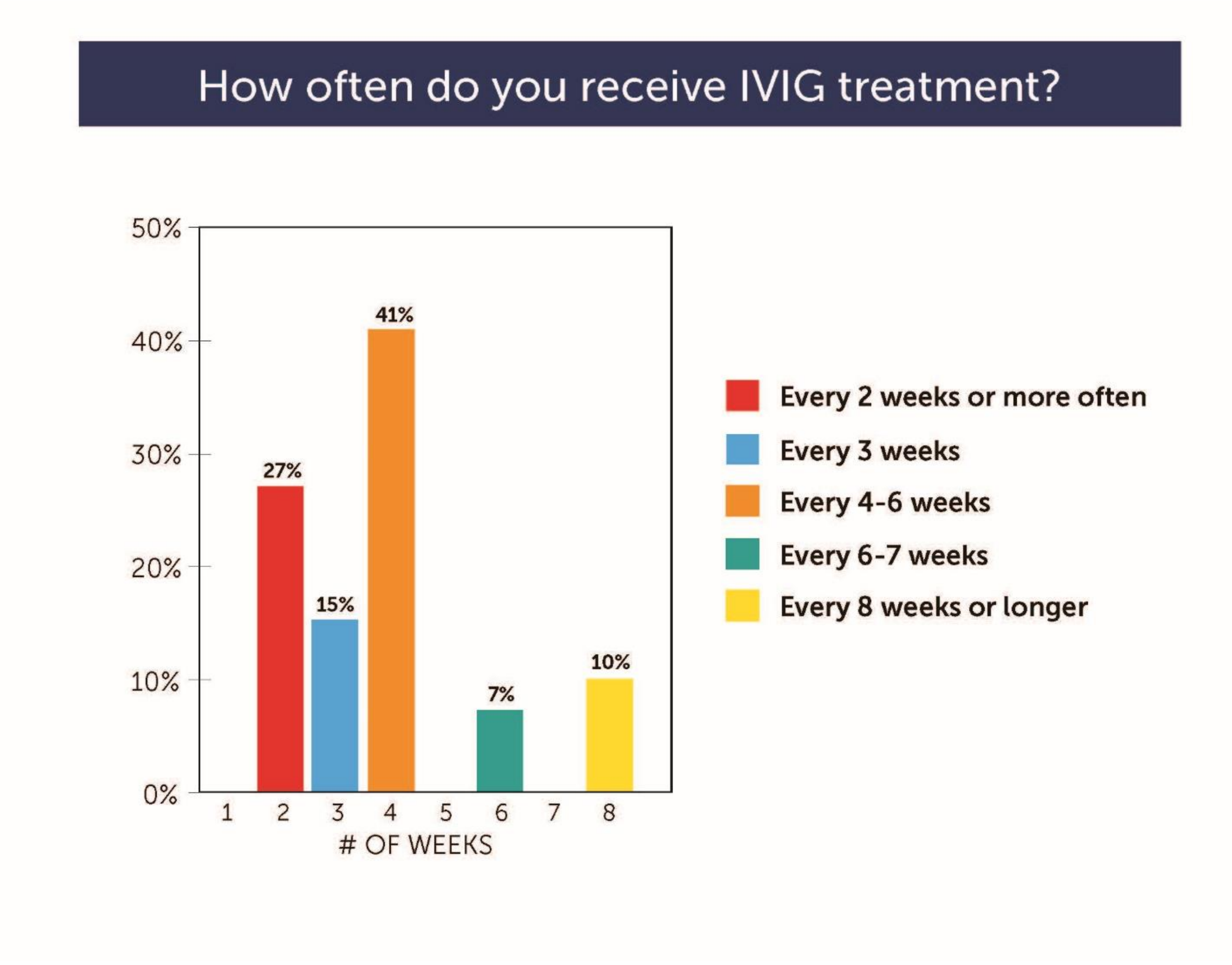
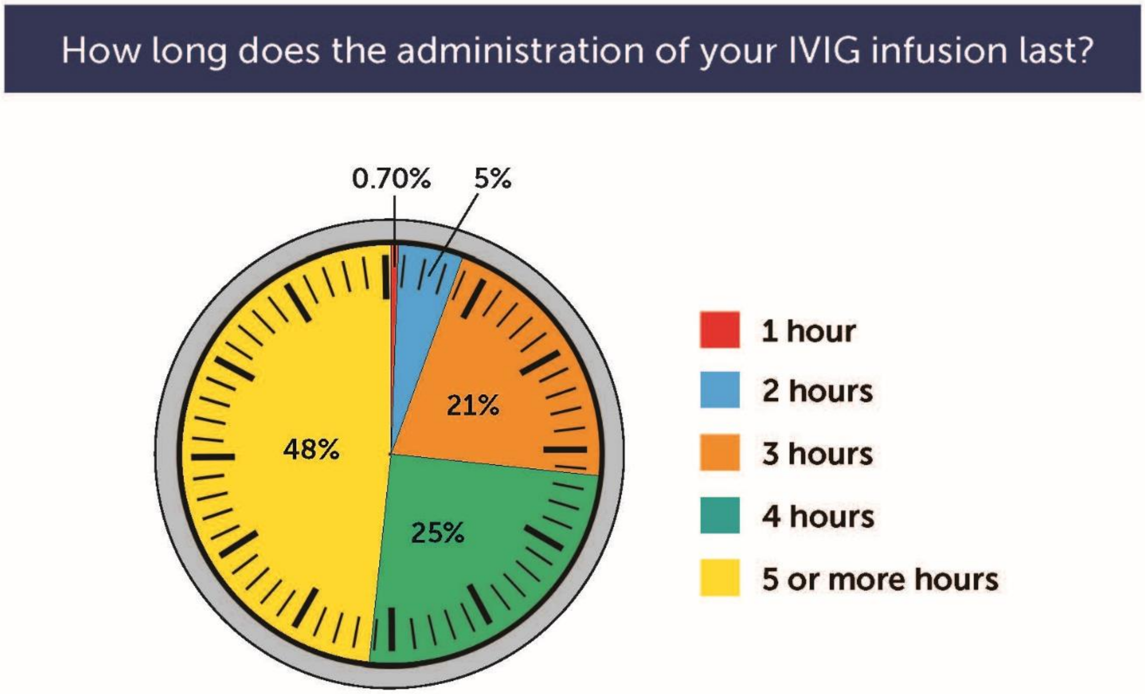


Figure: Length of Treatment

The duration of treatment can add to the burden of MMN for many individuals.



While the majority of patients reported being on IVIG therapy, 81% said they would consider SCIg treatment if it were recommended by their Neurologist. When asked about their insurance coverage 36% reported having access issues to their IVIG therapy and 17% said they had been forced to switch medication brands due to their insurance provider’s coverage requirements. Among the access issues reported were denials by the insurance company because the IVIG therapy was determined to be "not medically necessary". Several survey participants cited problems in the portability of their medical care from one insurance provider to another which resulted in a lapse or delay in therapy. Despite advances in health care access, 18% reported that they had not been able to afford their therapy at some point in time.

Differences were seen in how often the participants were receiving their treatments and the impact they had from these therapies. Currently, 39% reported receiving their therapy in a hospital, 36% receive their infusion in their home or office, 11% in an infusion center, 10% in a doctor's office, and 4% reported that they received their therapy in another setting. Difficulty finding an infusion center in their local area was also a concern for many patients.

A reduction in symptoms was reported within one week of their first Ig treatment by 36% of respondents. When asked at what point following their treatment they started to feel their symptoms return, 28% reported within 1 -2 weeks, 24% said within 3 weeks, 17% said within 4 weeks, while 12% said their symptoms did not return for more than 4 weeks, and 18% said their symptoms did not return at all in-between treatments. Many of those completing the survey reported improvements following their IVIG therapy, including feeling stronger, having improved balance, and less cramping and pain.

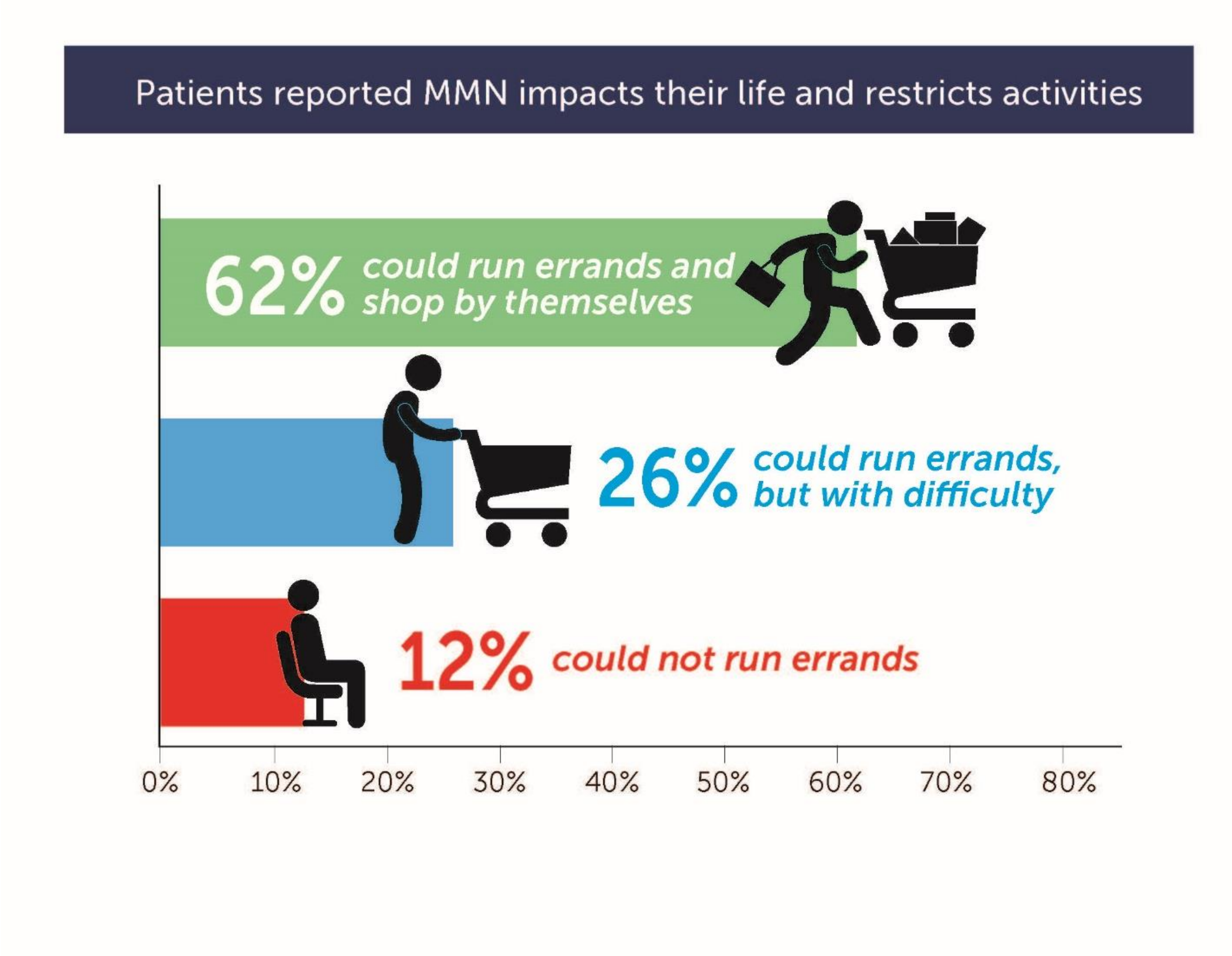
Quality of Life

Participants also reported that MMN impacts their life and restricts their activities. The most commonly reported symptoms included fatigue (21%), muscle cramping (20%), and weakness (53%). Many participants reported numbness, pain, and difficulty walking. Overwhelmingly, 75% said they felt exhausted, 75% said they felt they had no energy, 78% said that they experienced daytime sleepiness, while 58% reported difficulty falling asleep. The impact of sleep loss was evident; 45% of respondents noted that they had avoided or canceled activities with my friends or family because they were tired. Overall. 65% said that they frustrated by being too tired to do the things they wanted to do and 55% admitted that they had limited their social activities because they felt too tired. These feelings directly impacted their ability to do regular activities of daily living. For example, 62% said they were too tired to do their household chores.

More than half reported that MMN often or always interferes with their employment. The data showed that 46% of participants had difficulty typing on a computer or using a telephone, and 46% had trouble concentrating. When asked whether they had to work really hard to pay attention to avoid making a mistake, 38% responded that this was the case.

In addition, almost half (49%) of the participants said that MMN often impacts their overall schedule. In response to some basic questions, 5% of participants said they could not turn a key in a lock, almost half (48%) admitted they had difficulty doing so. Most were able to complete daily tasks such as bathing (97%) and brushing their teeth (99%), while complex skills such as buttoning a shirt were challenging. Overall, 85% stated that they could button a shirt, however, 46% have difficulty. The survey also showed that 58% either couldn’t open a medicine bottle or had difficulty doing so.

Figure: MMN Impacts Basic Activities



Simply standing and getting around was reported as a challenge for more than half of those participating. While 56% said they were able to stand for a period of time without a problem, 36% reported have difficulty doing so, and 8% could not stand at all. Almost half (49%) said they were able to go for a walk of at least 15 minutes, whereas 27% had difficulty and 24% could not walk for this length of time. Similar challenges were also noted in climbing stairs; 47% of respondents were able to walk up and down a flight of stairs, while 42% had difficulty, and 11% could not. When asked, 56% clearly stated that MMN impacts their overall quality of life.

MMN has a direct impact the participant's emotional wellbeing. Some 46% reported feeling depressed and 28% of those completing the survey said they had time when they felt hopeless. Half of all respondents said that because of their MMN, they felt left out of things. Feeling embarrassed by their physical limitations was a concern for 59% of respondents and 65% felt that their MMN limited their participation in regular family activities. Additionally, 64% are dissatisfied with their ability to participate in fun activities outside their home.

This survey highlighted some very critical issues relating to the diagnosis, management, and impact on the quality of life individuals with MMN. It also clearly demonstrates that some participants are empowered and knowledgeable about their condition, but that more work need to be done in educating physicians, and health insurers in the proper diagnoses and management of the condition. Perhaps one participant said it best when they told us: "*We are so happy that MMN is being researched and studied. It is very difficult to find information about this condition. Also, had it not been for the IVIG treatments, all of the symptoms/limitations listed were ME! I was unable to do anything for myself. We are very grateful that IVIG is working and that our insurance company covers the cost*."