NEUROMYELITIS OPTICA SPECTRUM DISORDER

WebMD FOCUS ON

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THE LATEST ON

NEUROMYELITIS OPTICA SPECTRUM DISORDER

UP TO 99%

Percentage of people with neuromyelitis optica spectrum disorder (NMOSD) who have eye pain and spinal cord inflammation.

SOURCE: National Center for Advancing Translational Sciences

8 in 10

Number of people with neuromyelitis optica spectrum disorder who are women.

SOURCE: National MS Society

OUT WITH THE OLD, IN WITH THE NEW

In a small study, stem-cell transplant significantly improved neuromyelitis optica spectrum disorder. People with the condition received chemotherapy to wipe out cells that might contribute to the disease. Next, they received an infusion of their own stem cells. These immature cells have the potential to develop into any type of human cell. The idea behind this research was that the stem cells would mature into disease-free blood cells. Five years after the infusion, 80% of those treated have not relapsed. People reported a higher quality of life and improved physical and neurological function. Most of those treated no longer carry a protein—called AQP4-IgG—linked to the disease.

SOURCE: *Neurology*

up to 4.4

Number of people in every 100,000 who have neuromyelitis optica spectrum disorder. Some research suggests the disease may be more common in people of African and Asian descent.

SOURCE: National Center for Advancing Translational Sciences

3 in 100

Number of people with NMOSD who have a relative who also has the condition.

SOURCE: National Organization for Rare Disorders

nearly 30%

Percentage of people whose neuromyelitis optica spectrum disorder is first misdiagnosed as multiple sclerosis.

SOURCE: *JAMA Neurology*

GETTY IMAGES

POSTPARTUM PROBLEMS

For new mothers with neuromyelitis optica spectrum disorder, the first year after childbirth may bring greater risk of relapse. In a study of 19 women, who had 30 pregnancies, only eight women avoided an attack of neuromyelitis for a year after giving birth. This relapse rate is significantly higher than the rate for women who haven't recently delivered a baby. If you are pregnant or planning to become pregnant, talk to your doctor about preventing postpartum relapse.

SOURCE: *Multiple Sclerosis and Related Disorders*

Talking to your healthcare provider about NMO/NMOSD and potential treatment with ENSPRYNG

If you have received a diagnosis of NMO/NMOSD (neuromyelitis optica or neuromyelitis optica spectrum disorder), the questions below can help you prepare to have a meaningful conversation with your healthcare provider about your symptoms, how to manage them, and potential treatment options going forward.

Your experience with NMO/NMOSD

- What symptoms are you living with today?
- What types of daily activities are impacted by these symptoms?
- What happened during your last relapse? Were you hospitalized? Were you treated with medication afterward (i.e., high-dose steroid, plasma exchange)?
- What kinds of life changes have you made?
- Are you currently on medications to help prevent future relapses or treat residual symptoms after a previous relapse?
- What concerns do you have about how NMO/NMOSD will impact you in the future?

Your potential NMO/NMOSD treatment

- What was ENSPRYNG shown to do in clinical trials?
- How does ENSPRYNG work?
- Can I take ENSPRYNG with my current medications?
- What are the potential side effects of ENSPRYNG?
- How is ENSPRYNG treatment given?

Important Safety Information

What are the most common side effects of ENSPRYNG?

The most common side effects of ENSPRYNG include:

- sore throat, runny nose (nasopharyngitis)
- rash
- fatigue
- extremity pain
- headache
- upper respiratory tract infection
- nausea
- inflammation of the stomach lining (gastritis)
- joint pain (arthralgia)

Please see the brief summary of the ENSPRYNG full Prescribing Information on the following pages.

Turn to ENSPRYNG Access Solutions® for answers and support

Get help understanding your insurance coverage and assistance options from ENSPRYNG Access Solutions®.* You can find answers to your questions, whether they're about financial support, insurance, or ENSPRYNG treatment. Once you've been prescribed ENSPRYNG, you'll be teamed up with a Patient Navigator who will be with you every step of the way.

Patient Navigators

Your Patient Navigator will work with your healthcare provider's office and/or specialty pharmacy to help you get your medicine, explain insurance coverage, and answer any questions about ENSPRYNG.

Financial Support

Genentech is committed to helping you get the ENSPRYNG you are prescribed. Your Patient Navigator can help you find assistance options so you can pay for ENSPRYNG.

Injection Training Support

ENSPRYNG Clinical Educators are nurses who provide supplemental injection training on how to self-administer ENSPRYNG in person or virtually.

To learn more about ENSPRYNG patient support programs, call

1-844-NSPRYNG (844-677-7964)

or enroll online at www.ENSPRYNG.com/learnmore.

You will be directly connected with someone who can help you get the help you need. Support is available Monday-Friday, 9 AM-8 PM EST.

*You and your healthcare provider are responsible for completing and submitting all required paperwork to your health insurance plan. Genentech (and any partner brand) cannot guarantee your plan will cover any treatments.

These are not all the possible side effects of ENSPRYNG.
Call your doctor for medical advice about side effects.
You may report side effects to FDA at 1-800-FDA-1088.
You may also report side effects to Genentech at 1-888-835-2555.

For more information, go to www.ENSPRYNG.com or call 1-844-NSPRYNG.

ENSPRYNG™
satralizumab-mwge
subcutaneous injection 120 mg/mL

Patient Information

ENSPRYNG™ (satralizumab-mwge) injection, for subcutaneous use

What is the most important information I should know about ENSPRYNG?
ENSPRYNG may cause serious side effects including:

1. **Infections.** ENSPRYNG can increase your risk of serious infections some of which can be life-threatening. Talk to your healthcare provider if you are being treated for an infection or call them right away if you think you have signs of an infection, with or without a fever, such as:
   - chills, feeling tired, muscle aches, cough that will not go away or a sore throat
   - skin redness, swelling, tenderness, pain or sores on your body
   - diarrhea, belly pain, or feeling sick
   - burning when you urinate or urinating more often than usual

   Your healthcare provider will check if you have an infection and treat it if needed before you start or continue to take ENSPRYNG.

- Your healthcare provider should test you for hepatitis and tuberculosis (TB) before you start taking ENSPRYNG.
- All required vaccinations should be completed before starting ENSPRYNG. People using ENSPRYNG should not be given 'live' or 'live-attenuated' vaccines. 'Live' or 'live-attenuated' vaccines should be given at least 4 weeks before you start ENSPRYNG. Your healthcare provider may recommend that you get a 'non-live' (inactivated) vaccine, such as some of the seasonal flu vaccines. If you plan to get a 'non-live' (inactivated) vaccine, it should be given, whenever possible, at least 2 weeks before you start ENSPRYNG.

2. **Increased liver enzymes.**
   Your healthcare provider should order blood tests to check your liver enzymes before and while you are taking ENSPRYNG. Your healthcare provider will tell you how often you will need to have these blood tests. Make sure you get all of your follow-up blood tests as ordered by your healthcare provider. Your healthcare provider will tell you if you need to wait to start ENSPRYNG if your liver enzymes are increased.
3. **Low neutrophil count.**
   ENSPRYNG can cause a decrease in your neutrophil counts in your blood. Neutrophils are white blood cells that help the body fight off bacterial infections. Your healthcare provider should order blood tests to check your neutrophil count while you are taking ENSPRYNG.

See "What are the possible side effects with ENSPRYNG?"

What is ENSPRYNG?

ENSPRYNG is a prescription medicine used to treat neuromyelitis optica spectrum disorder (NMOSD) in adults who are aquaporin-4 (AQP4) antibody positive.

It is not known if ENSPRYNG is safe and effective in children.

Do not take ENSPRYNG if you:

- are allergic to satralizumab-mwge or any of the ingredients in ENSPRYNG. See **"What are the ingredients in ENSPRYNG?"** at the end of this Medication Guide for a complete list of ingredients in ENSPRYNG.
- have an active hepatitis B infection.
- have active or untreated inactive (latent) TB.

Before you take ENSPRYNG, tell your healthcare provider about all of your medical conditions, including if you:

- have or think you have an infection. See **"What is the most important information I should know about ENSPRYNG?"**
- have liver problems.
- have ever had hepatitis B or are a carrier of the hepatitis B virus.
- have had or have been in contact with someone with tuberculosis.
- have had a recent vaccination or are scheduled to receive any vaccination.
- are pregnant, think that you might be pregnant, or plan to become pregnant. It is not known if ENSPRYNG will harm your unborn baby.
- are breastfeeding or plan to breastfeed. It is not known if ENSPRYNG passes into your breast milk. Talk to your healthcare provider about the best way to feed your baby if you take ENSPRYNG.

Tell your healthcare provider about all the medicines you are taking, including prescription and over-the-counter medicines, vitamins, and herbal supplements.

How should I take ENSPRYNG?

- ENSPRYNG is provided as a solution in a single-dose, prefilled syringe of 120 mg/mL of satralizumab-mwge.
- See the **Instructions for Use** inside the carton for complete instructions for the right way to prepare and inject ENSPRYNG.
- ENSPRYNG is given by an injection under the skin (subcutaneously). If your healthcare provider decides that you or your caregiver can give your injections of ENSPRYNG, you or your caregiver should receive training on the right way to prepare and inject ENSPRYNG.
- Always inject all of the medicine in the syringe.
- The first 3 injections (loading period) of ENSPRYNG are taken 1 time every 2 weeks.
- After this, injection of ENSPRYNG is taken every 4 weeks (maintenance period). Keep taking ENSPRYNG 1 time every 4 weeks for as long as your healthcare provider tells you to.
- If you miss a dose of ENSPRYNG, talk to your health care provider about restarting dosing.

What are the possible side effects of ENSPRYNG?
ENSPRYNG may cause serious side effects, including:

- See **"What is the most important information I should know about ENSPRYNG?"**
- **Serious allergic reactions.** Serious allergic reactions that may be life-threatening have happened with other medicines like ENSPRYNG. Tell your healthcare provider before taking your next dose if you had hives, rash, or flushing after your injection. Seek medical attention right away if you have any symptoms of a serious allergic reaction, such as:
  - shortness of breath or trouble breathing
  - dizziness or feeling faint
  - swelling of your lips, face, or tongue
  - moderate or severe stomach (abdominal) pain or vomiting
  - chest pain
- The most common side effects of ENSPRYNG include:
  - sore throat, runny nose (nasopharyngitis)
  - rash
  - fatigue
  - extremity pain
  - headache
  - upper respiratory tract infection
  - nausea
  - inflammation of the stomach lining (gastritis)
  - joint pain

These are not all the possible side effects of ENSPRYNG.
Call your doctor for medical advice about side effects. You may report side effects to FDA at 1-800-FDA-1088. You may also report side effects to Genentech at 1-888-835-2555.

How should I store ENSPRYNG?

- Store ENSPRYNG in the refrigerator between 36°F to 46°F (2°C to 8°C) in the original carton.
- Protect from light.
- **Do not** freeze or use the syringe if it has been frozen.
- **Do not** shake.
- ENSPRYNG, if unopened, can be removed from and returned to the refrigerator, if needed. The total combined time out of the refrigerator should not be more than 8 days at a temperature that does not go above 86°F (30°C).

General information about the safe and effective use of ENSPRYNG.

Medicines are sometimes prescribed for purposes other than those listed in a Medication Guide. Do not use ENSPRYNG for a condition for which it was not prescribed. Do not give ENSPRYNG to other people, even if they have the same symptoms that you have. It may harm them. You can ask your pharmacist or healthcare provider for information about ENSPRYNG that is written for health professionals.

What are the ingredients in ENSPRYNG?

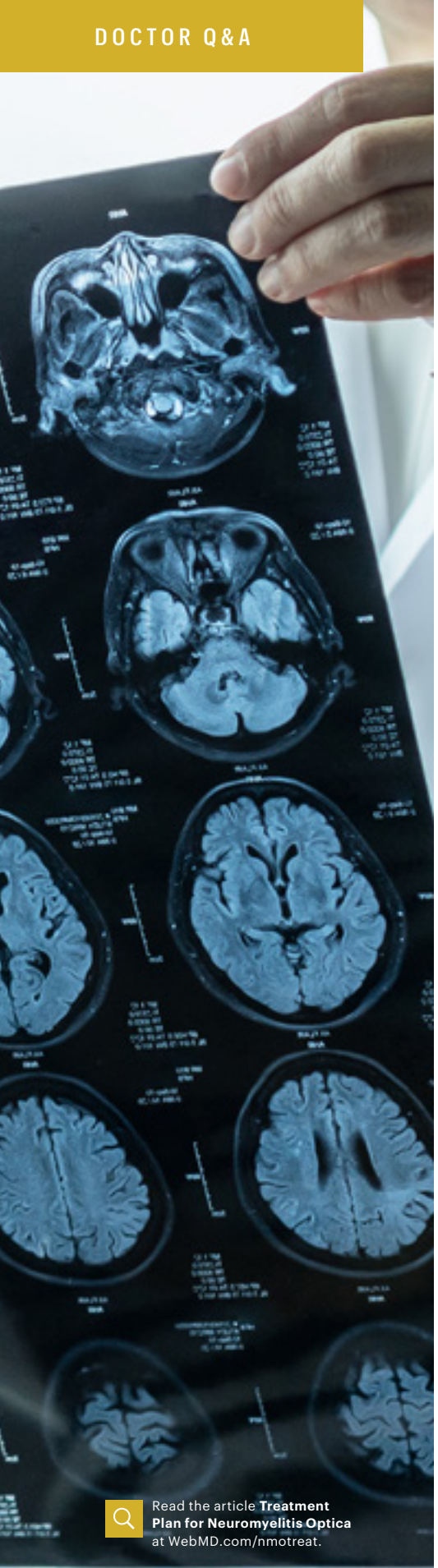
Active ingredient: satralizumab-mwge

Inactive ingredients: L-arginine, L-histidine, poloxamer 188, L-aspartic acid, and Water for Injection.

Manufactured by: **Genentech, Inc.**, A Member of the Roche Group, 1 DNA Way, South San Francisco, CA 94080-4990
ENSPRYNG™ is a trademark of Chugai Pharmaceutical Co., Ltd., Tokyo, Japan
U.S. License No.: 1048
For more information, go to www.ENSPRYNG.com or call 1-844-NSPRYNG.

M-US-00006868(v1.0)

ENSPRYNG™
satralizumab-mwge
subcutaneous injection 120 mg/mL



Read the article **Treatment Plan for Neuromyelitis Optica** at WebMD.com/nmotreat.

THE BASICS

WHAT YOUR DOCTOR WANTS YOU TO KNOW

By Rachel Reiff Ellis
Reviewed by Christopher Melinosky, MD, WebMD Medical Reviewer

Reid Longmuir, MD, assistant professor of ophthalmology and visual sciences at Vanderbilt Eye Institute at Vanderbilt University Medical Center in Nashville, gives a primer on neuromyelitis optica spectrum disorder (NMOSD).

Q. WHAT CAUSES IT?

NMOSD is a condition of the immune system that impacts the nervous system. Antibodies made by the immune system attack the spinal cord, brainstem, and optic nerves—the nerve that goes from your eye to your brain. It is important to understand that NMOSD is not multiple sclerosis (MS). It tends to have less impact on the brain than MS, though it can be difficult at first to distinguish the symptoms from typical MS. It is not genetic, although it has been occasionally recognized in multiple family members. And it is not contagious—you can't catch it from someone else.

Q. WHAT ARE SOME OF THE RISK FACTORS?

If you already have an autoimmune condition, you're at a higher risk of getting NMOSD. It affects women about five to 10 times more than men. You can get it at any age, but typically happens around ages 30 to 40.

Q. HOW WILL NMOSD AFFECT ME OVER TIME?

Without treatment, NMOSD typically has numerous relapses. When you compare that to the relapses most MS patients can have, each NMOSD attack leaves significant residual disability. If you don't treat your NMOSD, there is a significant risk of death. But with treatment, that risk is very low. Maintaining follow-up and long-term treatment is critical to achieving the best possible outcome.

Q. HOW COMMON IS IT?

It's a very rare condition in the U.S., but neuromyelitis spectrum disorders represent a much higher percentage of demyelinating disease—diseases that destroy the myelin, or insulation cover, around your nerves—in other parts of the world, particularly in Japan.

Q. HOW DO YOU DIAGNOSE IT?

The testing involves MRI of the brain, orbits (optic nerve), and spine, as well as lumbar puncture (spinal tap), and blood tests to look for the antibodies associated with NMOSD. The classic antibody is aquaporin-4. If you're experiencing vision loss, an eye examination and testing can contribute to the diagnosis. If you've been diagnosed with MS, but treatment does not seem to be working, NMOSD should be considered.

GETTY IMAGES

Read the article **Symptoms of Neuromyelitis Optica** at WebMD.com/nmodiagnosis.

GETTY IMAGES

SIGNS AND SYMPTOMS

KNOW HOW TO SPOT A NEUROMYELITIS OPTICA SPECTRUM DISORDER ATTACK

By Rachel Reiff Ellis

Reviewed by Christopher Melinosky, MD, WebMD Medical Reviewer

When you have neuromyelitis optica spectrum disorder (NMOSD), your body mistakes certain healthy cells as a threat. Specifically, NMOSD pounces on the cells that insulate your optic nerve—the nerve that carries signals from the back of your eye to your brain—and your spinal cord. Rarely, it can also affect your brain.

When NMOSD affects your optic nerve, it swells, a condition called optic neuritis. "Optic neuritis results primarily in sudden, painful vision loss," says Jeffrey Bennett, MD, PhD, professor of neurology and ophthalmology at the University of Colorado Anschutz Medical Campus in Aurora, CO. NMOSD vision loss can range from slightly "fuzzy" vision to legal blindness (20/200 vision or worse). You may just have a blurry spot that doesn't go away when you blink. In 90% of cases, Bennett says, NMOSD sight problems come with a side of eye ache. "It's not necessarily horrible pain, but it causes general discomfort around or behind your eye, and it often gets worse with movement."

Inflammation caused by NMOSD in your spinal cord is called transverse myelitis, and it can affect your movements and sensations. "The spinal cord is an important pathway for the transit of motor-sensory and autonomic nervous system function, which is a fancy way of saying underlying control of bodily function," Bennett says. That means your symptoms can range from weakness, stiffness, and spasms in your arms and legs to bladder and bowel problems. More rarely, you might also feel a tingling feeling wrapping around your torso, or have itchy skin.

If your brain gets involved in your NMOSD, you may have double vision, problems with your facial sensation and movement, uncontrollable nausea, vomiting or hiccups, even narcolepsy.

Many of the symptoms of NMOSD and multiple sclerosis (MS) are the same. In fact, doctors used to consider it a severe type of MS. "There was very little way to distinguish the two based purely on symptoms," Bennett says. MS is more likely to show up on an MRI of the brain, and attacks are often less damaging than NMOSD relapses. "Now that we have a test to differentiate between the two, we know that not only are they different in their cause, but they're different in their response to treatment."

So, which NMOSD symptoms are worrisome enough to warrant a call to your doctor? All of the above, Bennett says. "Any symptoms that suggest a new attack are important for a physician to hear about if you're already diagnosed with NMOSD," he says. NMOSD is an attack-related disorder, so if you're having familiar symptoms, you're likely entering a relapse. The sooner your doctor can start treatment, the better your recovery will be. Bennett says, "In general, any new vision loss, new numbness, new weakness that's lasting for longer than 24 hours—we want to hear about it."

Read the article **Living With Neuromyelitis Optica** at WebMD.com/nmolife.

LIVING WITH NEUROMYELITIS OPTICA SPECTRUM DISORDER

How I learned to manage this rare condition

By Christine Hà

Reviewed by Michael W. Smith, MD, WebMD Chief Medical Editor

A little more than 20 years ago while I was in college, I noticed a blurry spot in my right eye. At first, I thought it was just a problem with my contacts, but when I went to the optometrist, he said the problem wasn't visual—it was neurological. A visit with a neuro-ophthalmologist revealed that my optic nerve—the nerve that runs from the back of the eye to the brain—was inflamed. So they ran all kinds of tests: MRIs, blood, even a lumbar puncture. They told me an inflamed optic nerve in a young adult can often mean multiple sclerosis (MS). But I had no signs of MS in my brain, so doctors wrote it off as a one-time fluke case of optic neuritis.

Then a few years later, I started to have constant pins and needles in one of my legs. It was the kind of numbness that usually goes away after you get up and walk around, but this time, that brought no relief. Scans showed I still didn't have MS in my brain. But my doctors told me that two different MS-like symptoms at two different points in time on two different parts of my body was enough criteria to diagnose me with MS by default.

Despite starting treatment for MS, I kept getting worse. I had symptom attacks every few months. A bad one at the end of 2002 forced me to leave my job. I had paralysis from my neck down, and couldn't release my bladder, which sent me to the emergency room. It still took nine months of physical and occupational therapy for me to fully recover. And throughout it all, my vision continued to come and go. I lost enough of my sight that I could no longer drive.

I felt a lot of isolation and frustration during those years. I didn't know anyone else with health problems like mine, especially anyone my age. My friends were having fun, going to school, making friends, thinking about career moves—none of them were dealing with a chronic illness like I was. Nothing like this ran in my family, either. It was a lonely experience.

Finally, when the neuromyelitis optica spectrum disorder (NMOSD) antibody test became available, my neurologist suggested I take it, suspecting NMOSD was my true diagnosis. It was such a huge relief when I was positive. Knowing what you have is so much better than not knowing. And for years I thought I had MS that wasn't responding to treatment, when I didn't actually have MS at all. I began a regimen of once-a-year immunotherapy infusions, and I've been stable ever since.

With the use of screen readers, smart home devices, organization, a cane, or a sighted guide, I can navigate my busy schedule as a writer, public speaker, and chef. Having NMOSD has shaped my life in ways I couldn't have predicted, and also helped me find my purpose. In 2012, I won season 3 of *MasterChef* as the show's first blind contestant. My vision loss made my accomplishment unique, which helped elevate my story to a larger audience. Since then, I've been able to use my platform to help advocate and raise awareness for the NMOSD community.

CHRISTINE'S TIPS

SEEK SUPPORT
"It's important to connect with others who can help guide you and give you hope. Ask your doctor about local NMOSD groups, or search out online communities."

ADVOCATE FOR YOURSELF
"Find out as much as you can about NMOSD so you can make informed choices about your care. Choose a doctor you trust and like, who answers your questions in a timely way. Don't stay with a doctor who makes you feel shoved to the side."

SET BOUNDARIES
"As an Asian American woman, I've always had a tendency to want to please everyone and put others first. Learning to prioritize my own needs—and say no—has been critical for my self-care. You can only help others if you're helping yourself first."

CULTIVATE GOOD HEALTH
"Sleep, exercise, relaxation, a healthy diet—they're all important. Taking care of yourself physically will help you feel better mentally, which will help you manage your NMOSD symptoms and flare-ups."

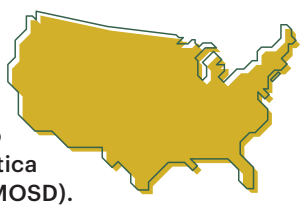
GETTY IMAGES. INSET PHOTO BY JULIE SOEFER PHOTOGRAPHY

STATS & FACTS

By Sonya Collins

Reviewed by Michael W. Smith, MD, WebMD Chief Medical Editor

4,000



Estimated **number of people in the U.S.** who have neuromyelitis optica spectrum disorder (NMOSD).

80

Number of **known autoimmune diseases,** including NMOSD.

1 year

The average time from when a person develops their first symptom **to diagnosis of neuromyelitis optica spectrum disorder (NMOSD).**

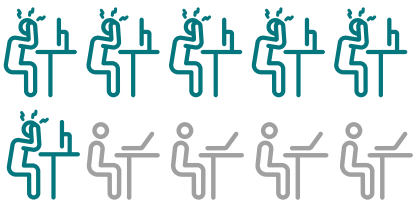
250,000

Estimated number of people worldwide who have neuromyelitis optica spectrum disorder.

35%

Percentage of people with neuromyelitis optica spectrum disorder whose **vision loss is moderate to entirely disabling.**

The chance of a **recurrence after a first attack** of NMOSD happens.



6 IN 10

Number of people with NMOSD who say their **health sometimes limits their work or activities.**

45%

Percentage of people with neuromyelitis optica spectrum disorder **whose pain is moderate to completely disabling.**

40 to 50

Most common ages at which neuromyelitis optica spectrum disorder appears, though it can develop at any age.

1 IN 5

Number of people with NMOSD **who have moderate "brain fog,"** which can affect your ability to think.

GETTY IMAGES

SOURCES: National MS Society, *Multiple Sclerosis and Related Disorders*, National Institutes of Health—National Institute of Allergy and Infectious Diseases

A GROUP EFFORT

Given that neuromyelitis optica spectrum disorder is a rare disease, patients and caregivers can benefit from finding others who understand life with this illness

By Erin O'Donnell

Reviewed by Michael W. Smith, MD, WebMD Chief Medical Editor

Learning that you have neuromyelitis optica spectrum disorder (NMOSD) is life-changing. Connecting with a group of people living with the same challenging chronic disease can help you weather this new reality.

"Rare diseases can be incredibly isolating, and for a lot of folks, going to the first support group meeting is the first time they meet anybody else with that diagnosis," says Benjamin Greenberg, MD, director of the neuroimmunology program at the University of Texas Southwestern Medical Center. Talking with others who "get" the diagnosis can be a relief.

Support group members often share details about their symptoms and how they manage them, offer their takes on doctors and other health care providers, and suggest their favorite resources for learning more about NMOSD. Support group members keep each other updated on opportunities to participate in research studies. And the emotional support can be especially valuable. "There's also a large psychological impact to being diagnosed with a rare disease," Greenberg says, especially given that people with NMOSD can be healthy for long periods and then suddenly experience an attack that could impair their vision, weaken their arms, or alter their ability to urinate. "With this condition, people are just waiting for the other shoe to drop," he adds.

Unless you live in a large metropolitan area, you may have trouble finding an in-person support group for people with NMOSD, Greenberg says. But the COVID-19 pandemic has led many support groups to start gathering virtually using tools such as Zoom or Skype, creating new opportunities for people to connect with others with NMOSD around the country.

To find a group, Greenberg recommends checking with two organizations: the Siegel Rare Neuroimmune Association (wearesrna.org), which has an international network of support groups, and the Guthy-Jackson Charitable Foundation (guthyjackson-foundation.org), which offers regional support groups. The foundation also offers several NMOSD groups designed for specific groups, including men only, women who are pregnant or planning to be, and teens. Guthy-Jackson can also connect you with Facebook groups for people with NMOSD.

Greenberg, who periodically participates in NMOSD groups through the Siegel Rare Neuroimmune Association, notes that support groups vary depending on their leaders and members, and a group may not feel like the best fit for you at first. While this could mean that you need to find another group, Greenberg usually urges his patients to stick with one group for a while. "I remind folks that the topic changes monthly," he says. You might also consider getting involved in planning, and propose topics that you would find personally useful, he adds. "It's a very democratic system and group leaders really push to make sure that everybody's engaged and there's something for everyone."

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