Fact Sheet

Facts about rare neuroimmune disorders: The 5 W's

Neuro-immune disorders, such as acute disseminated encephalomyelitis (ADEM), acute flaccid myelitis (AFM), MOG antibody-associated disease (MOG-Ab disease), neuromyelitis optica spectrum disorder (NMOSD), optic neuritis (ON), and transverse myelitis (TM) are conditions in which a person's immune system mistakenly attacks parts of the central nervous system (CNS) – brain, spinal cord, optic nerve.

Who gets these illnesses?

- **ADEM** tends to affect young children, typically ages 4-8, without a significant bias for specific gender or ethnic background.
- **AFM** tends to affect children as well, and increases in cases have occurred every other year since 2012.
- We are still learning about who is more likely to get **MOG-Ab** disease. Some studies have shown that those with MOG Antibody-Associated Disease are on average younger and are likely to be male compared to those with aquaporin-4 (AQP-4) positive NMOSD. Those with MOG-Ab disease may be more likely to have bilateral involvement of the optic nerves.
- **NMOSD** associated with AQP-4 antibodies tends to disproportionately affect non-Caucasian women in their 30-40s.
- **TM** can affect individuals of all ages, ethnicities, and either gender.
- **ON** is more common in women and develops in most patients between the ages of 20 and 45. Additionally, ON typically occurs more frequently in Caucasians than African Americans.

Where in the nervous system do these disorders affect?

- inflammation in optic nerves = optic neuritis (ON)
- inflammation of spinal cord, primarily the white matter of the spinal cord = transverse myelitis (TM)
- inflammation of spinal cord, primarily the grey matter of the spinal cord = acute flaccid myelitis (AFM)
- inflammation of brain = encephalitis
- inflammation of brain and spinal cord (and sometimes optic nerve) = encephalomyelitis (acute disseminated encephalomyelitis (ADEM) is a subtype that may or may not include spinal cord involvement)
- inflammation of brain, optic nerves, and/or spinal cord = neuromyelitis optica spectrum disorder (NMOSD) or MOG antibody-associated disease (MOG-Ab disease)

What does monophasic or relapsing mean?

- Some of these disorders are monophasic, meaning a one-time confused reaction of the immune system, without any further episodes of inflammation (TM, AFM, ADEM, ON).
- Other disorders are known as relapsing, in which a persistently confused immune system can continue to cause inflammatory episodes (NMOSD and MOG-Ab disease, although ADEM, TM, and ON can be initial presentations of these relapsing diseases)
- For the disorders that can be relapsing, people are given long-term therapies to diminish the chance of future episodes or to lessen their impact should they occur.
- Testing for AQP-4 and MOG antibodies can help predict if someone will have a monophasic or relapsing course.
- If antibody testing is negative, the longer one goes without another attack, the more likely it is that the condition is monophasic.

SRNA
PO Box 826962
Philadelphia, PA 19182-6962

phone: +1 (855) 380-3330
email: info@wearesrna.org
website: wearesrna.org

facebook.com/wearesrna
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Which of these conditions tend to be relapsing or recurring?

- Multiple sclerosis and neuromyelitis optica spectrum disorder associated with aquaporin-4 (AQP-4) antibodies are the two most well recognized forms of relapsing CNS autoimmune disorders.
- 60-80% of individuals with optic neuritis and longitudinally extensive transverse myelitis (involvement of greater or equal to the length of 3 vertebrae) have antibodies to aquaporin-4 (AQP-4), a water channel in astrocytes, a type of support cell in the central nervous system.
- A proportion of individuals who test negative for AQP-4 and some of those diagnosed with recurrent ON or ADEM are now known to have antibodies against another target, called myelin oligodendrocyte glycoprotein (MOG). MOG is a protein on myelin and oligodendrocytes, the myelin-producing cells of the central nervous system and are thought to have MOG-Ab disease. Individuals who continue to test positive for MOG antibodies 6-12 months after their initial attack are at risk for recurrent disease and should discuss with their provider if chronic immunosuppression is warranted.

Why do people get these disorders?

This is a central question in neuroimmunology and currently we still don't know for sure. It is hypothesized that these disorders result from a specific set of circumstances, namely 1) a person whose immune system may be primed to overreact or get confused, or a genetic predisposition to auto-immunity and environmental triggers, and 2) a life event, perhaps a bodily stressor, such as an infection, to trigger the attack. We do not yet know the genetics or environmental factors that lead to these conditions.

Author

Dr. Cynthia Wang received her medical degree from University of Texas Southwestern Medical Center in Dallas, Texas and completed a pediatrics and pediatric neurology residency at Mott Children's Hospital, University of Michigan Health System in Ann Arbor, Michigan. Dr. Cynthia Wang completed her James T. Lubin Fellowship under the mentorship of Dr. Benjamin Greenberg at The University of Texas Southwestern and Children's Health. Her research study was a prospective, longitudinal study on acute disseminated encephalomyelitis (ADEM) to identify the clinical characteristics, treatment methods, and follow-up interventions that are associated with better and worse patient-centered outcomes.