

AUTOIMMUNE ENCEPHALITIS **ALLIANCE**



NEWSLETTER

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It has now been two months since I joined the Autoimmune Encephalitis Alliance (AE Alliance) as its Executive Director and CEO. What a journey it has already been! I am humbled by the many people I've had the pleasure to speak with in the patient, caregiver, and clinician communities.

Participating in events such as the World Encephalitis Day Conference in Dallas and the AE Alliance Family Weekend in Durham has provided me a close up look at the struggles and triumphs of this patient community. Collectively, 130 survivors and their caregivers gathered at these two events to share their experiences and learn about the current state of medical treatment for the disease.

I attended a Rare Disease Day conference in Raleigh with 75 speakers and participants from the FDA, the NIH, venture capital firms, medical tech companies, and the pharmaceutical industry. While we have made progress with earlier diagnosis and treatment of autoimmune encephalitis, there is much work to be done on all fronts. I intend that the AE Alliance be a leader in advocacy for patients and physicians dealing with this disease. We need a national awareness campaign, a robust physician training platform, and ultimately significant research investment. It's a huge endeavor, but one I feel excited to undertake.

One of the most frequent questions I've been asked over the last two months is how I came to join the Alliance—what drove me to this particular organization. I'm not an AE survivor, nor do I have family members who have suffered from this disease. I was not a healthcare provider working in the field. So why align with this organization and this disease? The answer is simple—what I share with those of you who suffer (or have loved ones who suffer) from this disease is living with uncertainty about your body's immune system.

In 2014 a breast cancer diagnosis rocked my world. It introduced something new and frightening to my everyday existence—a distrust of my body—an all too frequent nagging concern that my immune system was dysfunctional and compromised and that a recurrence could happen. After all, if no one understands what causes cancer, what's to prevent my body from creating it again? While cancer isn't an autoimmune disorder, it is a dysfunction of the immune system with an equally devastating impact. My specific diagnosis was triple-negative breast cancer, a particularly aggressive form of breast cancer, and one more difficult to treat. Like many suffering with AE, there are currently no targeted therapies to treat it.

The diagnosis is just the beginning of a long road to recovery. I spent a year undergoing surgeries, chemotherapy, and radiation while working full time as an executive overseeing a major non-profit organization. I also spent that year seeing the healthcare industry from a different vantage point—as a patient.

While today I'm cancer-free and credit the care I received at the North Carolina Cancer Hospital (the clinical arm of UNC Lineberger), the experience was a game-changer. I realized that lifestyle changes were necessary. The experience compelled me to re-enter the healthcare field. (Before overseeing the North Carolina Museum of Art Foundation, I had spent over a decade in the pharmaceutical industry.) Since my diagnosis, I've made major changes in my life, leaving my position at the museum and focusing instead on adding to my skill set with certifications in health care coaching and wellness. I'm now a National Board Certified Health and Wellness Coach with a focus on nutrition.

Why the Alliance? Because I see the tremendous opportunity we have to elevate awareness at a national level. Because autoimmune disorders are immune system dysfunctions like cancer. Because I have a driving desire to make a difference with my skills for the good of an important group of patients and their families who need a voice and a platform for excellent care and treatment options. This is why I joined the Alliance and I look forward to working with each of you in this campaign. **You are not alone.**



A handwritten signature in black ink that reads 'Caterri Woodrum'.

Caterri Woodrum

Executive Director and CEO

Dr. Myrna Rosenfeld

**Institute for Biomedical Investigation,
University of Barcelona, Spain**



Myrna wanted to be a doctor for as long as she can remember and, when asked, can't pinpoint any particular reason. No one in her family was in medicine. Medicine was not the expected career path in their community, but her family was supportive of her decision. Her interest in neuroscience developed

in medical school and this led her to choose Neurology as a clinical specialty. However, and without any hesitation, she will tell you that the reason she chose Neuro-oncology for her subspecialty was the strong influence of a mentor, Dr. Nicholas Vick—the Neuro-oncologist in the hospital where she did her internship.

Her interest in treating patients with AE evolved from the research she had carried out since her fellowship training in Neuro-oncology. This work initially focused on autoimmune brain disorders that occurred in patients with cancer, some of which associated with autoantibodies. Over time, she and her colleagues realized that some patients who appeared to have one of these autoimmune disorders did not have cancer nor any of the known antibodies. This realization led them to develop new laboratory techniques that demonstrated, for the first time, the presence of anti-NMDAR antibodies in some patients and subsequently to the discovery of several other autoimmune encephalitis syndromes.

Myrna feels it takes a village to care for AE patients. It requires the input of multiple disciplines and sub-disciplines, including neurology (sub-specialties in epilepsy, movement disorders, and cognitive disorders), psychiatry, surgery (for tumor removal when present), pediatrics, intensive care medicine, and rehabilitation medicine, among others. She and her colleagues published a position paper* providing guidelines to determine if a patient probably or definitely has an autoimmune encephalitis, which she follows for diagnostic purposes. For treatment, however, she doesn't follow any specific guidelines due in large part to the relative rarity of most autoimmune encephalitis resulting in a lack of sufficient data on which to base treatment guidelines. However, her approach to patients is relatively constant and based on a variety of factors, including the severity of the illness, the presence of co-morbidities, and age, among other factors. Sometimes her treatment approach is circumscribed by socio-economic and insurance issues as well.

Her research has both a clinical and basic neuroscience focus. Clinically, Myrna is identifying and describing the clinical characteristics of new autoimmune encephalitis syndromes. From a basic research perspective, she tries to understand the cellular and molecular mechanisms through which the autoantibodies produce clinical symptoms. This work gives insight into the brain, not just during the active part of the disease but also during the recovery process for syndromes that take months or years. Studies of large numbers of autoimmune encephalitis patients are difficult to do as any one practitioner does not see many of these patients. Therefore, large, multi-center studies would be beneficial in testing treatment approaches, which could result in guidelines that are currently lacking. She sees a need for studies that focus on better understanding what triggers these diseases and how we can identify patients who are predisposed to develop them.

The field of AE has expanded rapidly, and while this is very exciting, she's seen an increasing number of reports and concepts that are not highly rigorous or founded on misinformation and misinterpretation. An example of this is reports of anti-NMDAR antibodies (i.e., IgA and IgM anti-NMDAR antibodies) assumed to be pathogenic in patients with a wide spectrum of disorders, including many that are not even immune disorders. These reports lead to unnecessary testing and often, inappropriate immunotherapy, among other issues. The fact that these IgA and IgM antibodies are completely different from the IgG NMDAR antibodies associated with autoimmune encephalitis, and have not been yet been shown in a scientifically robust manner to have any clinical significance is ignored. Thus, for the field to evolve, we need to have comprehensive definitions of the clinical syndromes and associated antibodies and guidelines for antibody testing. We must demand validation studies before accepting novel concepts, and we need to avoid perpetuating speculative and non-rigorous science.

When Myrna is not working she enjoys gardening and sewing. She finds these activities to be very relaxing because there is always an end product to enjoy. Outside the home, she tries to stay physically active in a variety of ways, and she belongs to a community organization that plans local cultural and political activities.

Thank you, Dr. Myrna Rosenfeld, for giving us the opportunity to share all you do for current and future AE patients.

**Graus F et al. A clinical approach to diagnosis of autoimmune encephalitis. Lancet Neurol 2016; 15:391-404.*

Q:

Our nephew suffers from AE and has been diagnosed a few months ago. He was started on steroids and IVIG. The test came back positive for anti-NMDAR encephalitis and his doctor now wants to start with Rituximab. We are so overwhelmed by all of this and don't know what to expect from the Rituximab. How does it work? When will we see an improvement? And is it safe?

A:

How Rituximab works is a complicated question. Rituximab is an antibody designed to bind to B-cells and cause their destruction. The removal of B cells occurs rapidly in the blood, but a reduction of B cells in tissue may take longer. While Rituximab's primary target is B-cells, it also decreases the numbers of other immune cells over time, including plasma cells and T cells. However, the decrease in these other cell types, as well as antibody levels, tends to take many months (3-6 months). Given the range of effects on the immune system and the different timing for decreases in the various immune cells and antibody levels, you may start to see improvements from Rituximab from as early as a few weeks after dosing in some, and not until many months later in others.

The most common side effects are reactions during the infusion. These are usually mild and can be treated with steroids and antihistamines. Some patients will have a "flu-like" reaction starting several days after Rituximab infusions. Also, given Rituximab is an immunosuppressant and works by decreasing the immune system activity, individuals treated with this

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medication are at increased risk for infections. A rare complication is a drop in the neutrophil count—these are immune cells important for fighting bacteria.

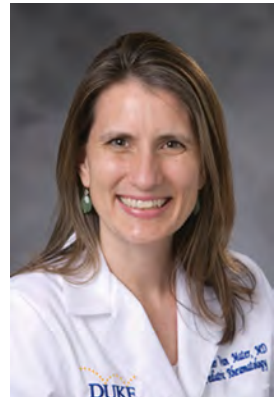
There are two different starting doses commonly used. One gives infusions at a lower dose every week for four weeks, the other gives a higher dose but only has two infusions given two weeks apart. After the “induction” dose, patients may continue on maintenance dosing, which is given approximately every six months. In some people, their immune system recovers from Rituximab faster, and B cells can start coming back earlier than 6 months. Many doctors will follow the B cell count to determine if patients on maintenance dosing need more frequent infusions, with some people needing infusions as often as every three months.

Rituximab is used alone or with other medications. This partly depends on what the exact disease is and how refractory or hard it is to treat. Because Rituximab does not start working immediately, many patients continue on first-line treatments, such as steroids and IVIG to bridge the first several months until rituximab starts working. For more refractory diseases, oral immunosuppressants such as mycophenolate or azathioprine may also be used in combination with Rituximab.

Rituximab may be used for relapse treatment in individuals who were not treated with it at their initial presentation, and in those who have had a relapse associated with the return of B cells. In people who have been treated recently with Rituximab, it is unclear how helpful it is to redo Rituximab early if peripheral B cells are still depleted from the initial dosing.



Dr. Heather Van Mater, Duke Health



Jackie Stebbins

Lawyers have two great fears in life: missing a deadline and getting sick. Missing a deadline is scary and keeps us awake at night, but getting sick is about the worst. A simple cold or stomach flu can reschedule a deposition, which could change a case's course and other deadlines then it feels like all the dominos start to fall. We're humans, so we're going to get sick, but we're also lawyers, so we plan for everything. Or wait, can we?

I am a lawyer, and I fell deathly ill while in private practice. It was unexpected, there was no accordingly tailored action plan, and it was worse than I ever could have imagined.

In late May 2018, my autoimmune encephalitis (AE) diagnosis forever altered the course of my life. My disease is a sneaky assailant. It took my body and mind, before me or anyone around me could get a serious hold on what was going on.

I suffered from insomnia for quite a while before I decided to take it seriously, but only because I was afraid it would start affecting my work. All signs pointed towards depression and anxiety. Lawyers become depressed from their work-load and stress; that's a fact. Why was I any different, especially with the hours I was working in family law and criminal defense cases? I didn't want to admit to any mental health issues, but the slow and severe deterioration of my health finally made me realize and accept it.

Antidepressants were prescribed for sleep and I continued plowing through work. I told myself I'd take time off in the summer. I just had to make it through my busy spring. By the end of April, I was still an insomniac, with clenched jaws, shaking hands, and ringing ears. In early May, I left work for a week's break. Earlier that day, I had what I now know was a serious anxiety attack at my desk, and I knew I couldn't stay at work any longer. I believed that I needed time for my medication to kick in, which would hopefully allow me to sleep. On the surface, I told myself that I would return to work soon, but a deep down dark thought told me I'd never return to my office as I nearly collapsed out the door.

Six days after I left work, I checked into the psychiatric ward. My decline was evident. I wasn't communicative, and I stared a lot. I couldn't sleep during the day or night, and I believed I was unfit to drive my children. My body stopped working while I tried to swim or bike. I suffered from paranoia and confusion and hallucinated with prescribed sleeping medication. During my 48-hours in the ward, my mind started to slip, and things, like knowing the date and reading a clock, became a challenge. I struggled to read and write. I cried and exhibited serious tremors. Something told me that I didn't belong in the ward, but I was desperate for help to sleep and to feel better.

From the time I left work until my time in the ward, my memory isn't great. Once I left the ward, I nearly ceased to exist as a person, and my memory is bare. My life during that time has been pieced together by me through records and my family's recollections.

For six days after I left the ward, my husband of ten years took care of me like a child and wondered if I had dementia or was possessed because of my cognitive impairment and strange behavior. At a follow-up behavioral appointment, my nurse practitioner immediately believed I had a neurological condition and expedited my referral to a local neurologist. Her astute thinking saved my life. The word "neurological" led my husband and family to think the worst. They believed I had a brain tumor.

My neurologist's diagnosis the next day? He believed I had AE but ordered more testing to rule out other conditions. Over three days, I failed a neurological examination with flying colors, had an MRI image of my brain that was of poor quality because I shook so badly and could not lie still, and I underwent a spinal tap. I have only a few memories of the testing the first day, and I don't remember much after. My family cried for days, and I was oblivious to everything. I repeatedly asked the same questions about what was going on, but was fairly easily reassured and was compliant. My mother repeated over and over, "Thank God she doesn't understand what's going on."



Jackie Stebbins

Just twenty days prior, I had successfully defended a show cause order hearing, but now I could not draw the face of a clock.

During the early morning hours the day after the spinal tap, I had a grand mal seizure in bed that broke and dislocated my right shoulder. I spent the next five days at CHI-St. Alexius Hospital in Bismarck. My memories from the hospital are either nonexistent or skewed. My health was incredibly fragile, and there was a serious discussion of a medevac to the Mayo Clinic. During this time, my husband, family, and a few close lawyer friends took over my life. This team made all of the decisions for my cases, my role in my law firm, my health care, and whether my husband would sell our home and move us closer to our family. I had no idea any of this was going on. They went into crisis mode—fighting for my health and so no one missed a step at the law firm. The thankfulness I have for my Superman husband, my beloved family, and my friends can never be fully explained.

My health stabilized, so I stayed in Bismarck. Upon my release, I began a week of IV steroids to treat the AE. By the second day of treatment, my mind rallied, and my family saw the signs of me again. I underwent a CT scan that same week. The scan revealed blood clots in my right lung and leg and three broken vertebrae in my back. That week I was able to understand that I was never mentally ill, but that all my health problems were the AE at work.

I went to the Mayo Clinic in June, and my diagnosis and treatment were verified, which was good news; it was the devil we were coming to understand. But the recovery process was slow. I struggled with people distracting me, noise, anxiety, fear, personal interactions, and any public outings. I could not drive, felt broken physically and mentally, had limited movement, and

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was shaky and unsteady. Confined to my home for a year, I had to repair my mind, body, and spirit. The Superwoman efforts required of me to survive and recover were overwhelming. With my husband and family's love and support and my determination to recover for my children's sake, I rose to the challenge.

After the onset of the AE, I never returned to my law firm. I elected to retire from private practice based upon my health conditions. Since fifth grade, I only wanted to be a lawyer. I had sacrificed more than I can explain to get to where I was in my career when AE hit. Losing the career I loved and the law firm I was so proud of, felt like the end of me and everything I knew. Although I have lost in unexplainable ways, I have gained an incredible new perspective on life. I have tremendous hope for the future. I can use the same drive and skills I honed to be a respected lawyer, to accomplish the same goal I had as a lawyer to help others. How I can best do that, only time will tell. I am working on a book to describe my experience with AE and I hope it is out for readers to enjoy in 2020.

Jackie was diagnosed with seronegative AE in 2018.

Seronegative AE patients show similar clinical features to seropositive AE patients, except for autoantibody detection, making diagnosis and treatment more challenging.

More than 25 autoantibodies associated with AE have been identified, and new antibodies are being identified at an astonishing pace; many patients with seronegative AE will probably harbor antibodies that are yet to be isolated.

AEA in the Community



NYC AEA Support Meeting

The NYC Group met on World Encephalitis Day at Lenox Hill Hospital. We were honored to have Dr. Najjar join us, he launched the partnership between Northwell and AEA for this Support Group and he took a lot of time answering all the questions our members had. AEA feels very fortunate to have Joe Calcagno as the NYC Support Group Leader, he is a GAD65 survivor, very compassionate and works tirelessly on supporting others affected by AE.



World Encephalitis Day Conference in Dallas TX

Six nonprofits joined forces and put up a great and informative 3-day conference for (autoimmune) encephalitis survivors and their caregivers in Dallas TX on World Encephalitis Day, the first of many to come.

AEA Family Weekend – March 7 & 8

We started with 1,000 runners participating in the Florence Forth race, AEA’s major fundraising event. The next stop was the Museum of Life and Science. Dr. Stanley Naides, EuroImmun, presented two Service Awards to Dr. Heather Van Mater and Alicia Halbert, for their long and continued service to AE Alliance and AE in general. EuroImmun gave a check for \$10,000 to support AE Alliance in all its endeavors.

Dr. Genalynne Mooneyham and Dr. Suma Shah gave excellent and informative presentations and answered questions. We offered different breakout sessions. Dr. Rebecca Sadun led an Art Therapy class with AE survivors and their siblings, it was truly amazing to see how children through art communicate their emotions. Carol Collins, Community Services and ABI specialist led the Support meeting for survivors and their caregivers, which sparked a great conversation.

Sunday, after a yoga session by Meg Poe NWC-HWC, it was time for giving back and AEA families worked on getting Care Boxes ready. These Care Boxes will be distributed amongst doctors on our Clinicians Network to give to a newly diagnosed AE family. #YouAreNotAlone



Ask Me About AE



'Ask me about AE' t-shirts serve as an invitation for others to learn more about AE and about you.

Buy yours here: customink.com/fundraising/ask-me-about-ae

Post your stories, photos, and videos using the hashtag [#AskMeAboutAE](https://twitter.com/AskMeAboutAE) and let's get loud so more people will know about this disease. We can change and save lives!

Let's do something extraordinary!

- ✓ Want to see physicians better equipped to diagnose and treat AE?
- ✓ Want to ensure patients and caregivers have hope and support?
- ✓ Want to see researchers find a cure?

Invest with us at aealliance.org/donate

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Together, we can shape the course of AE!

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