First Aid Tool Kit for Autoimmune Encephalitis (AE)

A Patient/Caregiver Resource Guide to Managing Your Care

www.autoimmune-encephalitis.org  (352) 527-2470
Navigating your medical care when you have been diagnosed with an under recognised neuroimmunological disease that presents with psychiatric symptoms, has challenging pitfalls. This pro-active preparedness guide is designed to assist you through that unique journey.

Disclaimer:

IAES, does not advise in medical matters, and is not liable for the medical or behavioral health decisions you make. Medical clinicians who have established expertise in autoimmune encephalitis, (brain swelling by an autoimmune cause), should manage your health care. This often consists of a multi-disciplinarian team. The IAES Doctor’s List contains contact information for physicians who have published research in the field of autoimmune encephalitis. This requirement ensures us that the physician has displayed the expertise required to diagnose, treat and deliver the patient to best outcome/recovery.
Autoimmune Encephalitis is a rare disease

The field of autoimmune neurology is expanding rapidly. However, many neurologists, neuroimmunologists and doctors who encounter these patients first, such as ER physicians and psychiatrists, have never heard of the disease or seen a case. This reality makes the patient vulnerable to misdiagnosis.

Antibody-mediated autoimmune encephalitis are a group of inflammatory brain diseases that are characterized by prominent neuropsychiatric symptoms and are associated with antibodies against neuronal cell-surface proteins, ion channels, or receptors. The discovery of autoantibodies in autoimmune encephalitis has expanded quickly in the past decade since the first antibody, anti-NMDAr, was identified by Dr. Josep Dalmau.

Common clinical features include a change in behavior, psychosis, seizures, memory and cognitive deficits, abnormal movements, dysautonomia, and a decreased level of consciousness. The illness has a relapsing and remitting course (symptoms fluctuate by worsening and then improving) and can be progressive in some types. Brain injury or damage can result in approximately 42% of non-paraneoplastic cases leaving the patient with a permanent deficit.

The speed of recovery, degree of residual deficit, and frequency of relapse vary according to the type of autoimmune encephalitis. Studies regarding the two most common types of AE, anti-NMDAr and LGI1 report an average of 19% residual deficit for anti-NMDAr and 30% for patients with LGI1 at the 2-year mark. Treatment is headed by a Neurologist or Neuroimmunologist (often a team of medical professionals are involved including, Immunologists and Rheumatologists).

In some countries medical insurance covers these diseases. Insurance denials in the United States are averted when the doctor is knowledgeable, codes prior authorizations for treatment correctly, and articulates the treatments for these diseases with accompanied research.

Not everyone can maintain their employment with this disease and many go on Social Security Disability as a result. It all depends on the severity of the illness and treatment outcomes.

Autoimmune encephalitis is under recognised in the medical community even by some of the very best providers in neurology due to its recent identification in 2007. Diagnosing AE is complicated by the fact that it can mimic psychiatric disorders such as bipolar disorder or schizophrenia. Other symptoms such as hallucinations, autoimmune psychosis, (also referred to as autoimmune based depression), loss of memory and stupor/catatonia can result in misdiagnosis as these prominent early features are symptoms of those psychiatric disorders as well.
AE is thought to be more common than originally realized due to those who have gone undiagnosed. Due to the combinations of a newly identified, rare/uncommon illness and the complexity of the diagnostic criteria, misdiagnosis routinely occurs delaying appropriate first line treatments in suppressing the immune system that can bring the disease under control. AE is highly treatable and best outcomes occur with early diagnosis and aggressive immunotherapy.

If your clinician is evaluating for the possible diagnosis of Autoimmune Encephalitis, it can be difficult for the patient to understand the process. Your physician will be running tests to rule out possible causes of your medical condition. This is how an accurate diagnosis is arrived at; to rule out what it is not and gather evidence of what is occurring. Cases of autoimmune encephalitis, brain swelling due to an autoantibody attacking healthy brain cells, can present as mild, moderate or severe and not necessarily in that order.

Autoimmune Encephalitis can be a challenging diagnosis to make. A full neurological evaluation is required for a diagnosis. This consists of a lumbar puncture, laboratory testing (particularly neural autoantibody testing where a negative antibody result does not rule out a diagnosis of Autoimmune Encephalitis), EEG, MRI with and without contrast, Ultrasounds and Cat scans for tumor searches are fundamental in the diagnosis. Experts in the field perform an FDG-PET scan for the deepest look into the brain. This can show active disease, if the disease is responding to treatment and/or a comparative FDG-PET can be performed to confirm that the disease is no longer present. Outside of autoimmune encephalitis clinics, these tests can take many months. Because prompt/accurate diagnosis is key to best outcomes, considering traveling to an AE clinic is advised. *Discounted air travel and accommodations around AE clinics can be located on the IAES website.

Autoimmune Encephalitis is considered a neurological autoimmune disease. To clarify, the immune system creates an antibody that sees healthy brain cells as foreign and attacks them. This occurs throughout the brain. The brain now malfunctions. All areas of brain function can be affected. Because these attacks are relapse/remitting, (remitting not to be confused with remission) and can target different areas of the brain and the brain function that area is responsible for, a fluctuation of symptoms is seen due to the relapse/remitting nature of its presentation. We can change in how our symptoms present from day to day and different times of day. An acquired brain injury due to neuronal brain damage/death can occur. Atrophy and injury or damage can be seen on scans or slowing seen on EEG. The many symptoms associated with AE often confuse the practitioner who is unfamiliar causing them to modify your report of symptoms to fit into a diagnostic criterion they are familiar with. Misdiagnosis is costly in recovery, your time and money.

There is no ‘cookie cutter’ treatment protocol. Guidelines for treatment protocol are included in the following peer-reviewed studies. Some types of AE may respond better with specific treatments in the treatment protocol then other types. Best outcomes for an accurate diagnosis and aggressive
When a person becomes sick, the disease has a rapid onset. Since the first doctors to see these patients may not be aware of this group of disorders, IAES is providing the following articles to help you, your family and your provider regarding how autoimmune encephalitis is diagnosed. These articles should be shared with physicians if autoimmune encephalitis is suspected and the physician is unfamiliar with the disease. Even when a patient has a confirmed diagnosis, since the disease is relapse-remitting, you will likely encounter Physicians who are unfamiliar with AE should an emergency room visit become necessary.

Therefore, to help you advocate for your health we are providing research that will be helpful to you in understanding the diagnostic and treatment process and to assist physicians, so a misdiagnosis can be avoided.
Your AE Resource Kit

IAES recommends you create a notebook/binder that you can take with you to the emergency room should that occasion arise.

A clinical approach to diagnosis of autoimmune encephalitis  Francesc Graus, Josep Dalmau and others 2017.
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5066574/

The Diagnosis and Treatment of Autoimmune Encephalitis  by Eric Lancaster, Department of Neurology University of Pennsylvania.

Antibody-Mediated Encephalitis Josep Dalmau, M.D., Ph.D., and Francesc Graus, M.D., Ph.D. 2018

Diagnostic and Therapeutic Approach to Autoimmune Neurologic Disorders A. Sebastian López-Chiriboga, MD1 Eoin P. Flanagan, M. 2018

Paraneoplastic and Other Autoimmune Disorders of the Central Nervous System  by: Andrews McKeon (make sure to see The treatment algorithm Figure 2, Table 4)
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3726118/

*Create a binder divider for: Psychiatric Symptoms in AE

Include the following articles. This information should be provided to a physician so misdiagnosis can be avoided:

Neuropsychiatric symptoms in autoimmune encephalopathies: a clinician’s guide
Fátima Carvalho, João Massano, and Rui Coelho

Red Flags: Clinical Signs for Identifying Autoimmune Encephalitis in Psychiatric Patients Julia Herken and Harald Prüss 2017
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5311041/

A Transdiagnostic pattern of Psychiatric symptoms in Autoimmune Encephalitis Carsten Finke, 2019
https://www.thelancet.com/journals/lanpsy/article/PIIS2215-0366(19)30038-0/fulltext?fbclid=IwAR0bjju0L0K6AuKpKiOPv0TlfQQbW4ySOKTLzoOtLAb3pBhiW_DX-nk#back-bib6

Recognizing psychiatric presentations of anti-NMDA receptor encephalitis in children and adolescents: A synthesis of published reports 2019, RJ Gurrera
https://www.ncbi.nlm.nih.gov/pubmed/30653785/?fbclid=IwAR2i8lbajmJ594JNH3jawdOse6yxEtC340qVtv0p-EYoN-pzPHiTJQ3p4wM
Many of us who are further along the path on our health journey have found that even in the early stages of Autoimmune Encephalitis, the things on the following pages were helpful for us.
Find a Neurologist who specialises in Autoimmune Encephalitis.

The International Autoimmune Encephalitis Society (IAES) has a Doctor’s list of experts who treat AE on their Web site: www.autoimmuneencephalitis.net. Many of us have had to travel far to find a such a specialist. In some cases, your current provider may need to make the referral. Your insurance may have to approve this which is covered later, in the Insurance part of this kit. Delay in proper identification and aggressive treatment will result in a poorer health outcome and possibly lasting disability that could have been avoided. Be sure to ask your current provider about flying if you opt for this. Air pressure changes associated with flying can impact encephalopathy symptoms in some cases. Ask your neurologist to try and keep your services, labs, testing, etc. at the same facility if possible. Some of us have many specialists spread out all over multiple offices/clinics/hospitals/facilities which uses vital energy and creates a barrier to collaboration of care. Many of us who are ill, are positive for autoimmune disease but continue to show normal labs, MRI’s. This can be confusing. The neurologist will take a full medical history and documentation of the onset of your illness, order tests and labs to gather evidence of what is occurring, rule out possibilities so an accurate diagnosis can be arrived at. If your physician is not an expert in autoimmune encephalitis, unnecessary delay and expense will be spent resulting in poorer health outcomes and unwarranted financial strain.
Give someone a medical power of attorney

If you do not have a **Power of Attorney** (POA), start the process by determining who you want to make medical decisions for you if your cognition becomes impaired. Patients who do not have this in place, have found themselves misdiagnosed as a psychiatric patient. AE patients who are misdiagnosed as psychiatric are prescribed anti-psychotic dopamine antagonist medications which worsen their condition, exacerbate symptoms and delay accurate medical treatment. A full neurological work up must be performed, and all organic medical causes ruled out prior to a psychiatric diagnosis. However, all too often this does not occur. It is not unusual for a patient to be put on a psychiatric hold for their safety.

If your adult child is transferred to a psych ward, you as their parent may not be informed that this has occurred. Any adult placed on a psychiatric hold is no longer able to make their own treatment decisions. Medical information or input from family is not considered and family does not have to be informed of the patient’s treatment plan or anything about their case unless the parents/spouse/family member/or advocate get a POA. If you are in a crisis and do not have a POA in place, you can file for that and obtain emergency medical guardianship. Acquiring guardianship for your adult loved one must be urgently addressed. In a medical emergency when the patient is rendered unable to make their own medical decisions, this can be acquired very quickly. Board members of IAES have witnessed patient’s families not address this need and the patient was legally committed and did not receive appropriate medical/neurological evaluations for their condition.

Free Power of Attorney (POA) forms and guidance on this process can be found at http://powerofattorney.com/medical-power-attorney/. Complete the forms even if you can’t afford the attorney and have your POA keep the forms and keep a copy in your Notebook binder, which will be discussed below. Should you need a POA, the hospital has a process to execute this quickly. If you are misdiagnosed, you will need your advocate to discuss your case with your doctors and make decisions on your behalf to ensure that you receive the medical care you need and deserve.
Do not attempt to manage this on your own

Educating a few family members, friends and/or your Power of Attorney (POA) is essential and a loving and caring thing to do for yourself and others. Patients may undergo treatment for a year or longer depending on the severity of the case, therefore, support from family and friends is a key element to your improved health while on this journey. Cognitive problems associated with AE can occur in a few days or weeks of becoming ill; some types take a relapse-remitting course while other types can be progressive. We know stress exacerbates these disorders. Encephalitis, (brain swelling), is a symptom not the root cause. The root cause is the immune system has created an antibody that is not supposed to be there, and that antibody sees healthy brain cells as foreign. Think of this as a case of ‘friendly fire’ where the immune system is attacking itself. In this case, the immune system is attacking the brain.
Do your best self-care

A structured lifestyle is important. Having a routine and a process for everything can make AE easier to manage. Some AE patients carry a notebook to write everything down in while others rely on cell phones by using notes, memory or calendar apps, to support memory loss issues. (Week Calendar for iPhone to assist with memory and ‘Symple’ app for iPhone to assist with symptoms and medications are good examples). Refer to the ‘Tools for Crisis Management’ page of our website for further patient support ideas.

Many of us have had to re-evaluate what important actions we should implement in our lives to maintain wellness. Rest and minimal stimulation help the brain to heal. Your neurologist will emphasize the importance of getting 8 hours of sleep and napping when tired to help your brain function at its current best. Your body will not have the stamina it once had. Be mindful when scheduling tasks or attending events so you don’t overdo. Your brain has been injured and will signal you that you have done too much by giving you a headache, increased brain fog or other symptoms may increase as a warning to you that you need to rest and give your brain opportunity to ‘re-boot’. Exercise such as walking 45 minutes daily is also highly recommended by neurologists.

Re-evaluate your expectations and help your loved ones and employer understand the new challenges you face. You will require their help and support. Situations will arise that are difficult for you to manage now but were not before you became ill. Turning down a social invitation because the overstimulation causes your symptoms to increase, for example, may appear to friends and family that you are isolating yourself. Help your loved ones understand the disease and how it affects you. That way they can be mad at the disease, not disappointed in you. There are several helpful handouts on the ‘Living with AE’ page of our website that should help friends and family better understand what you are challenged with. You are not alone in your grieving. Be aware that loved ones will also be grieving the loss of ability you used to have.

Many of us have found counseling and spiritual connection helpful. Develop a ‘support circle’ of friends and family. Use your windows of ‘good days’ or time during a good day to batch cook and freeze healthy meals, work on projects important to you and socialize. Some have found that describing Autoimmune encephalitis in simple terms to family and friends to be helpful. An easy explanation is that the immune system is attacking the brain.
Be safe

AE can diminish our cognitive ability. Since symptoms can relapse/remit, (again, when symptoms remit this is not remission but a fluctuation of the disease) pre-empting with safety planning is a way we can stay responsible for our care and make choices for ourselves. Keep an ongoing assessment of safety issues in your journal or cell phone app. Neurological deficits can cause risks for driving, caretaking others, falling, financial issues, safety for appliances, home security and medication administration. An Occupational therapist and Physical therapist can assist in these areas. Plan safety precautions based on your worst days. People cannot see encephalitis outwardly and often providers cannot determine this without thorough neurological testing and neuropsychological evaluations.

A neuropsychological evaluation is an assessment of how your brain is functioning. Neuropsychological tests evaluate functioning in several areas including: intelligence, executive functions (such as planning, abstraction, conceptualization), attention, memory, language, perception, sensorimotor functions, motivation, mood state and emotion, quality of life, and personality styles. Our deficits may be under recognised by the clinician inexperienced in AE or because a neuropsychological evaluation has not yet been performed. You will not likely look different, if you do not have need of a cane, walker or scooter/wheelchair, so some of us have experienced not being believed that our symptoms are impacting the quality of our daily lives to the level we are reporting.

Use structure and a process that mitigates your risks. For example, some of us batch cook and freeze meals on our good days or when we have a friend or family member with us to guide and support us in the kitchen for safety and assistance with our memory deficits/cognitive issues around following a recipe. This way, we can avoid safety hazards around forgetting to turn the stove or oven off or having an uncooked meal because we forgot to turn the oven on. Use of visual reminders and instructions are helpful. Incorporating
the use of crock pots, rice cookers, and microwaves as a cooking alternative to ensure kitchen safety are examples of alternate solutions.

If you have AE and are care giving for young children or an elderly person, if your work requires care giving identify a person in your life who will tell you when you are no longer capable of caring for others or making good decisions. Find a way to be accountable to this. There is no shame in this as this is how responsible people mitigate their risks.

Driving is another safety concern. Many of us are no longer able to drive or are highly restricted in our ability to drive. Error on the caution of safety as this is not just your life but the safety of others in the community. Many cities have door to door lift bus services for people with disabilities, look into this option and ask your provider about transportation resources for people with disabilities in your community. If affordable, Uber is another service you can take advantage of regarding alternate transportation.

Having a Plan B and a back up plan for important things is helpful when you cannot attend to it yourself. Wear a medical I.D. bracelet. There are many types available and some allow space for listing multiple diagnoses. Wearing more than one in different colors is also an option. Carry a medical I.D. card. Here is a link to a free medical I.D. card: https://www.myidentitydoctor.com/Free-medical-wallet-card%20.html

List emergency contacts in your cell phone and include an up-to-date list of your current medications in the ‘notes’ section on your cell phone. Discuss emergency care with your neurologist and have him provide an emergency treatment protocol for you to be entered into the computer database and include that in your emergency contacts on your phone. You may also include links to the above articles in your ‘notes’ section under emergency contacts in your cell phone that an ER attendings can access during emergency room visits, although they will need your permission or be given access by a person accompanying you. Have close family and friends keep this information in their phones as well. Post emergency medical telephone numbers and contact information along with emergency medicine to be given or treatment protocol in several visible places in your home. EMTs will usually look on the refrigerator for such medical information.
Behavioral Health

AE can be very debilitating at times. When the brain is under attack and you are in a symptom/disease flare, this can present or mimic a behavioral health problem such as bipolar disorder or schizophrenia. Psychosis, delirium, hallucination, depression, and catatonia can over shadow the neurological symptoms occurring. Your behavior may require hospitalization.

This tool kit, the articles and your health notebook or cell phone should go with you to the ER or hospital to help your providers understand your medical diagnosis. Your POA, legal guardian or family member should be asking for a neurologist to do an evaluation and the attending emergency room doctor to review the articles, patient information in the hospital’s computer system, as well as putting a call into your neurologist. Your medical id bracelet is key in ER visits. Every effort to educate the Emergency Room medical staff of your diagnosis should be made. Advocate that a hospital admission should be to the neurology floor. There are times when patients are referred to the psych ward if their behavior is such that there is a concern for safety for themselves and safety of others. All effort should be made to ensure that the patient’s medical records, which you should have copies of and be contained in your notebook/journal are provided to ensure admittance to neurology.

We try and support that AE patients belong in a neurological bed with first line treatments (that are outlined in the above articles and included in our notebook binder) be ordered to arrest the disease flare. If all aspects of your safety and emergency efforts are in place, there is a higher likelihood of success. However, if you are a danger to yourself or others, you will meet criteria for a psychiatric bed. Family members, friends and your POA will be in place to ensure neurology is consulted in your treatment plan.

Once released from the hospital, you should have a neurological appointment within 7 days to follow up. You can ask the Social Worker at the hospital to make that appointment for you or make sure a member of your ‘support circle’ takes that job and makes the appointment for you.
Autoimmune Psychosis/
Autoimmune Depression

The most URGENT symptom in some of the more common autoimmune encephalitides is suicidal ideation. Suicidal ideation is not uncommon in anti-NMDAr encephalitis and other autoimmune encephalitis types affecting the limbic system. It is the most tragic symptom and requires URGENT emergency immunotherapy treatment. Suicide is especially tragic as it is a preventable death in autoimmune encephalitis. Suicidal ideation often goes unreported by the patient. When patients are no longer in a symptom flare where suicidal ideation played a role and discuss their experience they grapple with processing that thoughts of suicide occurred as this is completely foreign to their nature and feelings. The shock of hearing of this symptom is scary, alien and mysterious to them but above all threatening to their safety and wellbeing. Patients who have no memory of an attempted suicide while in a flare of autoimmune encephalitis are also stunned at what family, physicians or friends later recount about the events that unfolded. Suicidal ideation has no reflection on the person’s love of life but a direct result of the disease.

The symptom of suicidal ideation in autoimmune psychosis/autoimmune depression is caused when autoantibodies attack the area of the brain that controls these emotions. It can come on quickly and without warning in its severity and has resulted in successful suicide when sufficient suppression of antibodies was not provided. Most AE patients are seen to emerge from severe depression and/or suicidal ideation 3 days after beginning a course of 60mg daily of prednisone or even one gram of IV solumedrol. Educate your POA and family to watch for symptoms of autoimmune psychosis/depression: sad mood, restlessness, irritability, sleep disturbance, loss of interest and suicidal thoughts. This is a critical part of your safety and self care.
Normally, suppression of antibodies can resolve an autoimmune based depression in a short period of time, therefore, discussing emergency treatment protocol with your neurologist is of vital importance. Many AE patients have a prescription on file at their pharmacy for prednisone including a prednisone taper for emergency use in the event of this type of symptom flare crisis. Other patients have taken the step to have their prescription filled with directions of how much to take for how many days to bring them out of this type of flare. The directions for filling this prescription should be included in your notebook binder and posted in a specific location in your home. Family members, friends and advocates should know where to look for this information.

AE may disrupt your lifestyle, relationships and how you knew yourself to be. Compounding being ill, if you do not have a neurologist who specializes in autoimmune encephalitis, you are likely to be undiagnosed or misdiagnosed or under treated. (Please refer to our Doctor’s list on our website which is constantly being updated). This leaves you in a more vulnerable situation of having to provide evidence to a medical doctor or psychiatrist who is not informed about AE. The symptoms of the disease leave you vulnerable to not being able to advocate for yourself. Facing daily challenges with cognitive disability and other neurological symptoms is a high stress life. Especially if you are fighting a particularly aggressive and/or refractory case of autoimmune encephalitis.
Due to these factors of additional stress the patient is prone to being more vulnerable to symptoms flares as stress is a trigger. We can experience clinical depression that needs to be addressed with your neurologist. Often medications are given to augment immunosuppression and facilitate emotional stability on the short term until treatment for the disease is managed and then these medications are tapered off. There are occasions when medications addressing the symptom of depression are an appropriate treatment in addition to treating the cause which is AE itself. Since some neuroleptic medications and medications used to treat some neuropsychiatric symptoms such as psychosis, can act as antagonists with autoimmune encephalitis it is important that the neurologist prescribe as they know what medications will not exacerbate your symptoms or AE. (per research- Benzodiazepines are successfully used for AE where dopamine antagonist medications should be avoided. Please refer to the handout located on the ‘Living with AE’ page of our website titled: AE is Refractory to anti-psychotic medication’).

Serious depression and thoughts of suicide are mainly rooted and linked to the disease course and how it is manifesting. However, the patient is made more fragile by having to contend with the above described daily life. If you have autoimmune encephalitis and feel depressed, see your neurologist right away, and be sure to tell them you are feeling depressed or suicidal, so your treatment plan can be adjusted.
Health Notebook

Keep a health Notebook that includes:

• Research on Diagnosis and Treatment of Autoimmune Encephalitis
• Research on Psychiatric Symptoms in Autoimmune Encephalitis
• Providers, address, phones and faxes
• Insurance information
• Pharmacy address, phone and fax
• Copies of the articles in your tool kit
• Emergency Treatment protocol from your Neurologist and stated diagnosis
• Medications and supplements with doses and schedule to administer.
• Medications you are allergic to and other allergies
• POA and emergency contacts with a written release of information for the people who may assume your care
• Disc copy of EEG, MRI, or any scans (you can request copies and include the most recent copy in a plastic protective holder)
Create separate sections for labs, specialty, and a daily journal that will read as a time line

The journal should have the date, symptoms, medication changes, provider/hospital visits, daily vitals if you keep these, accidents, job loss etc

Time Line. If you can recreate on a time line your whole medical history this will be helpful. Some keep this electronically as an email file that can be easily accessed. When you attend testing, or have labs, get a diagnosis, etc. be sure to get a copy and keep that in your notebook. The notebook can speak for you when you can’t, can’t remember clearly or ensure that your record is not going to take weeks to transfer from the multiple providers you have seen so will help to expedite your care
Insurance

Your insurance company will want to spend as little money as possible on your care. If medical evaluations are currently being done for Autoimmune Encephalitis as a differential diagnosis, call the number on the back of your insurance card and ask to have a nurse case manager assigned to you. Send them the articles in this tool kit and tell them you want your Autoimmune Encephalitis problem addressed correctly and do not want to waste your time or their money on incorrect diagnoses or treatments. Have your neurologist be prepared with showing your insurance company the tests which have been done such as FDG-PET, EEG, Lumbar puncture, MRI with and without contrast, antibody testing for AE, paraneoplastic panel, anti-NMDAr antibody tests in blood and spinal fluid, which will support the diagnosis of Autoimmune Encephalitis. Physician documentation of treatments received, such as a 3-5 day course of IV Steroids or six day trial/short course of 60mg steroid with a finding that suppression of antibodies resulted in improvement, can assist in differentiating an autoimmune basis compared to other disease.

Most insurance companies do not understand autoimmune encephalitis. Due to the diagnosis being uncommon, the diagnostic code is unfamiliar
to the clerical staff responsible for putting prior authorizations through to insurance companies. Additionally, there are a few diagnostic codes for autoimmune encephalitis to choose from, each highly specific, which further increases the possibility of clerical error. These disorders are often coded incorrectly by staff at the hospital, infusion center or doctor’s office which results in an increased chance of clerical error and higher occurrence of treatment denials that are automatically denied because the wrong code was used not because the treatment is not warranted. Sadly, the error goes unidentified as the doctor appeals the denial and moves through the different levels in the appeals process. Eventually a final denial is made. Too often families elect to pay for treatment out of pocket which creates unfathomable financial hardship, never realizing that the treatment would have been covered by their insurance if the correct diagnostic code had been used. Please be aware that a treatment denial stated as ‘investigational’ or ‘experimental’ is often the result of the incorrect diagnostic code having been used on a prior authorization.

The specific diagnostic code for Autoimmune Encephalitis that should be used on a prior authorization is: G04.81 (in the United States). This code includes Limbic encephalitis, ADEM and paraneoplastic autoimmune encephalitis. There is an abundance of research showing these disorders improve with IV Steroids, IVIG, plasmapharesis, Rituxan, and Cytoxan with maintenance medications of Cellcept, Imuran (Azathioprine) over the long term. These treatments are safe and effective in the treatment of autoimmune encephalitis. Due to the abundance of research stating treatments to be safe and effective for G04.81 diagnostic code, your treatments should not be denied while other diagnostic codes may. Be proactive and confirm with the entity that is processing your prior authorization that diagnostic code G04.81 has been used. When you know your physician will be putting in for a prior authorization for one of these treatments or if you are faced with a denial, refer to the ‘How to Appeal an Insurance Denial” page of our website. Lists of peer-reviewed research showing the treatment being requested is safe and effective for autoimmune encephalitis are downloadable there as well as step by step instructions as to how to proceed in overturning a denial. The research should be included with the prior authorization or appeal and will prevent the insurance company from denying your treatment in most cases.

Autoimmune Encephalitis is a life-threatening disease and requires urgent care aggressive treatment for the best documented outcomes. Just as Type 1 diabetes destroys the pancreas, in time, antibodies attacking the brain’s healthy brain cells can and does cause neuronal injury and/or death. If your insurance company denies you access to a specialist, then you must ask them to refer you to a Neuroimmunologist or neurologist who has a documented history in treating autoimmune encephalitis through their published work or mention on their bio. Since experts in the field are few, given that the first antibody in AE was only identified in 2007, the insurance company will be obligated to cover the cost of an expert out of network since they cannot provide you with one that is in network. The insurance company may not want to risk the liability of being wrong and may pay for the consult or out of network provider. If they will not, we encourage
you to find a way to get to a specialist and complete the assessment for Autoimmune Encephalitis.

International Autoimmune Encephalitis Society can refer you to organizations who provide free air transportation and low-cost accommodations. You can also ask that the insurance company staff your case in their grand rounds. Specifically request insurance consult a neurologist in their rounds to provide your neurologist guidance on why a specialist neurologist/Neuroimmunologist in autoimmune encephalitis is not needed. Have your neurologist request this in writing and ask insurance to respond in writing. Continue weekly calls for support with your Nurse Case Manager as they have to document your progress or lack of progress in each call. They can also be a good resource for records, services, and ongoing support.
Finances/Paying for Care

If you are working and have AE, you are likely having some challenges working or are not able to work. Be sure to document this along with your symptoms in your journal or app. Getting an AE diagnosis is difficult and time consuming due to long appointment waiting times and the amount of testing involved. Often, we are deemed “sick” but do not yet meet a diagnostic category that confirms a disability. Having the medical evaluations done to rule out a diagnosis of AE (a diagnosis is arrived at by ruling out what it is not to confirm what it is) may or may not be enough to qualify you for the Family Medical Leave Act (FMLA), as criteria for the FMLA are different state to state.

Your county should have a Department of Vocational Rehabilitation, which is a federal program to help disabled persons with employment. Google Department of Vocational Rehabilitation and add your state and county to the search. They offer support, testing, job training for persons with disabilities. Be prepared to show proof of your financial circumstance. Be prepared for more testing and the process to take several months. Be prepared to request help or ask if there is a social worker or patient advocate available to assist you in filling out the required forms as these tasks are daunting if you’re contending with a cognitive problem.

Generally, Insurance companies, Medicaid, and Medicare cover encephalitis of all types. It is necessary to have proper coding and well described written articulation of your diagnosis by your experienced treating neurologist. When it comes to hospital bills one no longer needs to sign a repayment plan. Based on your income under the 2015 Law named financial assistance or charity care, any family with a household income under $70,000 a year qualify under the new law. This new law only applies to non-profit hospitals and not providers. This law can also be used to decrease copays and deductibles depending on your income and after insurance has paid.

It States:

26 USC 501 (3) (5) Limitation on charges.

An organization meets the requirements of this paragraph if the organization—

(A) limits amounts charged for emergency or other medically necessary care provided to individuals eligible for assistance under the financial assistance policy described in paragraph (4)(A) to not more than the amounts generally billed to individuals who have insurance covering such care, and

(B) prohibits the use of gross charges.

It normally requires the Hospital bill be written off in total if the family yearly income is under 200% of the poverty level depending on that hospitals Financial Assistance Policy published on the Internet. Contact the hospital’s Chief Financial Officer.
By law they are mandated to do this if you ask for it. Know they will have to use your last year’s income record and this will work against you if your finances changed drastically from when you were well to when you were ill. Be prepared to show your records of this. Often the price the Hospital would charge insurance is greatly reduced when you’re paying out of pocket, no matter what your earnings. The provider does not have to abide by the minimum services rendered rule because insurance is not paying so service quality and treatment may be better and more efficacious.

Department of Assistive and Rehabilitative Services (DARS) is where the process starts to apply for disability. This is a long process even for partial disability and a decision to apply should be discussed with your doctor. You will need their recommendation and an attorney.
International Autoimmune Encephalitis Society

The International Autoimmune Encephalitis Society (IAES) conducts an educational support group for patients with a diagnosis and their loved ones on Facebook. The link to that location is: https://www.facebook.com/groups/251477975360/. The group was nominated in 2016, 2017 and 2018 for “Best in Show” and “Best Health Activist” in the WEGO HEALTH AWARDS.

Our Moto is:

“Empowering victims of Autoimmune Encephalitis and their caregivers through Support and Education”.

You are not alone. IAES can assist you in overcoming many of the challenges in living with AE and guide you to receiving an accurate diagnosis and treatment plan which will lead to the best outcome. The educational support group provides one-on-one assistance and helps you understand the disease, so you can become a strong advocate/self-advocate. Insurance assistance, overturning treatment denials and many other hurdles a patient faces are addressed by volunteer advocates in the group as well as emotional and personal support.

International Autoimmune Encephalitis Society

Website: https://www.autoimmune-encephalitis.org

Facebook page: https://www.facebook.com/IAESociety/

Facebook group: https://www.facebook.com/groups/251477975360/