AUTOIMMUNE NEUROLOGICAL DISORDERS
LEADING THE WAY IN AUTOANTIBODY EVALUATIONS
A RAPIDLY EVOLVING SUBSPECIALTY

Muscle fatigue that immobilizes a young mom in the prime of her life. Seizures that strike a child without warning. A 45-year-old man’s puzzling forgetfulness.

Today, patients who present these symptoms can receive appropriate diagnoses — multiple sclerosis (MS), epilepsy and early-onset dementia, respectively — and embark on standard treatment protocols to improve quality of life.

But in cases triggered by the body’s autoimmune response, modern medicine falls short of attacking these diseases at their root causes, offering only temporary relief rather than cures.

Autoimmune neurology is a rapidly evolving subspecialty driven by the discovery that the immune system can target virtually any structure within the central or peripheral nervous system. These conditions manifest as disorders previously thought to be independent and unrelated.

For the spectrum of autoimmune neurological disorders, where an early and accurate diagnosis is paramount, our testing approaches to effectively diagnose and guide treatment for these patients is unparalleled.

A HISTORY OF INNOVATION & DISCOVERY

Recognized as a world leader in the diagnosis and treatment of autoimmune neurologic disorders and demyelinating disease, Mayo Clinic mounts unmatched resources for uncovering novel syndromes, developing new diagnostic biomarkers and unique laboratory tests.

The Mayo Clinic Neuroimmunology Laboratory was the first to introduce comprehensive serological evaluations for the diagnosis of paraneoplastic neurologic autoimmune disorders, a group of disorders in which unusual neurologic signs and symptoms are the initial manifestations of cancer.

The laboratory continues to discover and clinically validate novel autoantibody profiles that inform neurological decision-making and guide the search for cancer.

SYMPTOMS OF AUTOIMMUNE NEUROLOGICAL DISORDERS

- Vision loss
- Spinal cord disorders
- Movement disorders
- Dementia, epilepsy and encephalopathy
- Myelopathy
- Paraneoplastic (cancer-related) neurological disorders
- Nerve, muscle and neurotransmission disorders (neuropathy, myositis and myasthenia gravis)
- Autonomic disorders (e.g., AGID – autoimmune gastrointestinal dysmotility).

Vanda Lennon, M.D., Ph.D., is the founder of the Mayo Clinic Neuroimmunology Laboratory. She is also the Dorothy A. Adair Professor in the Departments of Laboratory Medicine and Pathology, Neurology and Immunology at the College of Medicine at the Mayo Clinic.

COLLEGIAL ACCESS TO MAYO CLINIC NEUROLOGISTS

Neurologists are available for consultation and assistance in the interpretation of autoantibody evaluations. These conversations enable the best diagnosis and treatment approach, as they provide our neurologists the opportunity to ask additional clinical information, as well as address the questions of the ordering neurologist.
# Neural Antibodies Evaluated

<table>
<thead>
<tr>
<th>Antibody</th>
<th>Common Neurologic Findings</th>
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<tbody>
<tr>
<td>ANNA-1 (anti-Hu)</td>
<td>Limbic encephalitis, brainstem encephalitis, automatic neuropathies, sensory neuronopathy</td>
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<tr>
<td>ANNA-2 (anti-Ri)</td>
<td>Limbic encephalitis, brainstem encephalitis, myelopathy, peripheral neuropathy</td>
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<tr>
<td>ANNA-3</td>
<td>Limbic encephalitis, brainstem encephalitis, myelopathy, peripheral neuropathy</td>
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<tr>
<td>AGNA (SOX-1 antibodies)</td>
<td>Limbic encephalitis, neuropathy, Lambert-Eaton syndrome</td>
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<tr>
<td>PCA-1 (anti-Yo)</td>
<td>Cerebellar ataxia, brainstem encephalitides, myelopathies, peripheral neuropathies</td>
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<tr>
<td>PCA-2</td>
<td>Limbic encephalitis, ataxia, brainstem encephalitis, Lambert-Eaton syndrome, peripheral and autonomic neuropathies</td>
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<tr>
<td>PCA-Tr</td>
<td>Cerebellar ataxia, peripheral neuropathies</td>
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<tr>
<td>CRMP-5 IgG (anti-CV2)</td>
<td>Subacute-onset dementia, personality change, aphasia, depression, chorea, ataxia, myelopathy, radiculopathy, neuropathy</td>
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<tr>
<td>AMPH</td>
<td>Limbic encephalitis, aphasia, other subacute-onset dementias, stiff person phenomena, myelopathy, neuropathy</td>
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<td>VGKC complex, including LGI1 and CASPR2</td>
<td>Limbic encephalitis, amnestic syndrome, executive dysfunction, personality change, disinhibition, hypothalamic disorder, brainstem encephalitis, ataxia, extrapyramidal disorders, myoclonus, peripheral and autonomic neuropathy</td>
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<tr>
<td>NMDA receptor</td>
<td>Amnestic syndrome, anxiety, psychosis, seizures, extrapyramidal disorders</td>
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<tr>
<td>AMPA receptor</td>
<td>Limbic encephalitis, nystagmus, seizures</td>
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<tr>
<td>GABA-B receptor</td>
<td>Encephalitis, seizures</td>
</tr>
<tr>
<td>P/Q- and N- type calcium channel antibodies</td>
<td>Lambert-Eaton syndrome, ataxias, encephalomyelopathies</td>
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<tr>
<td>Ganglionic AChR complex</td>
<td>Somatic and autonomic peripheral neuropathies</td>
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<tr>
<td>Striatal antibodies</td>
<td>Typically nonspecific in isolation, but also with myasthenia gravis</td>
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<tr>
<td>Muscle AChR binding and modulating</td>
<td>Myasthenia gravis</td>
</tr>
<tr>
<td>GAD-65</td>
<td>Stiff-man syndrome, ataxia, seizures, brainstem encephalitis, ophthalmoplegia, parkinsonism, myelopathy, limbic encephalitis, other encephalitides; also type 1 diabetes mellitus</td>
</tr>
<tr>
<td>AQP4-IgG</td>
<td>Optic neuritis, longitudinally extensive transverse myelitis; reports of encephalopathies in children</td>
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AUTOIMMUNE NEUROLOGY TESTING

CENTRAL NERVOUS SYSTEM DISORDERS
- Encephalopathy Autoimmune Evaluation, Serum / CSF (Mayo ID: ENS1 / ENC1)
- Dementia Autoimmune Evaluation, Serum / CSF (Mayo ID: DMS1 / DMC1)
- Epilepsy Autoimmune Evaluation, Serum / CSF (Mayo ID: EPS1 / EPC1)

PARANEOPLASTIC, PERIPHERAL NERVE, AND AUTONOMIC DISORDERS
- Paraneoplastic Evaluation, Serum / CSF (Mayo ID: PAVAL / PAC1)
- Autoimmune Dysautonomia Evaluation, Serum (Mayo ID: DYS1)
- Ganglioside Antibody Panel, Serum (Mayo ID: GM1B)

STANDALONE ANTIBODIES
- Glutamic Acid Decarboxylase (GAD65) Antibody Assay, Serum / CSF (Mayo ID: GD65S / GD65C)
- Collapsin Response-Mediator Protein-5-IgG (CRMP-5-IgG) Western Blot Serum / CSF (Mayo ID: CRMWS / CRMWC)

NEUROMUSCULAR JUNCTION DISORDERS
- Myasthenia Gravis Evaluation with MuSK Reflex (Mayo ID: MGRM)
- Myasthenia Gravis (MG) Evaluation, Adult (Mayo ID: MGA1)
- Myasthenia Gravis (MG) Evaluation, Pediatric (Mayo ID: MGP1)
- Myasthenia Gravis (MG)/Lambert-Eaton Syndrome (LES) Evaluation (Mayo ID: MGL1)
- Myasthenia Gravis (MG) Evaluation, Thymoma (Mayo ID: MGT1)

STANDALONE ANTIBODIES
- Acetylcholine Receptor (Muscle AChR) Binding Antibody, Serum (Mayo ID: ARB1)
- Striational (Striated Muscle) Antibodies, Serum (Mayo ID: STR)
- Muscle-Specific Kinase (MuSK) Autoantibody, Serum (Mayo ID: MUSK)

NEUROMYELITIS OPTICA (NMO) SPECTRUM DISORDERS
- Neuromyelitis Optica (NMO)/Aquaporin-4-IgG Fluorescence-Activated Cell Sorting (FACS) Assay, Serum (Mayo ID: NMOFS)

FOLLOW-UP ASSAYS
- Neuroimmunology Antibody Follow-up, Serum / CSF (Mayo ID: PNEFS / PNEFC)

FOR MORE INFORMATION ABOUT AUTOIMMUNE NEUROLOGY TESTING
MayoMedicalLabs.com/neurology