

# Pathology

## LIST OF IMPORTANT ANTIBODIES:

Antibody	Disease	Description
<b>Demylinating CNS</b>		
<b>AQP4 Ab</b>	NMO	Aquaporin-4
<b>MOG Ab</b>	Childhood MS, ADEM, AQP4 negative NMO, AQP4 negative optic neuritis	Myelin oligodendrocyte glycoprotein
<b>Neuromuscular</b>		
<b>AChR Ab</b>	Myasthenia	Acetylcholine Rc Ab Positive in 85% of myasthenia patients
<b>MUSK Ab</b>	Myasthenia	Muscle specific kinase Ab -> inhibits AChR clustering in the motor end plate Positive in 50% of the AChR negative patients More common in women, African Americans, no eye involvement, more neck and bulbar involvement, less responsive to anticholinesterase medications or thymectomy.
<b>LRP4 Ab</b>	Myasthenia	LDL receptor-related protein 4 acts as a receptor for neural agrin, activates MUSK Positive in 9% of double seronegative patients (negative AChR/MUSK)
<b>Striational Ab (RyR Ab - Titin Ab)</b>	Myasthenia	Against striated muscle proteins (titin and rayndaudin) Present only in AChR positive myasthenia, usually in elderly > 60 and patients with thymoma. Sensitive but not specific for thymoma (50% of positives have thymoma, 95% of thymoma patients have titin Ab) Usually associated with more severe course of disease, respond to calcineurin inhibitors (tacrolimus and cyclosporine) Anti RyR can react against both skeletal RyR1 and the cardiac RyR2 receptors
<b>VGCC Ab</b>	Lambert Eaton	Positive in 90% of LEMS Associated with SCLC. Patients with LEMS should be screened with CXR every 6 months for lung cancer.
<b>GAD</b>	Stiff Person Syndrome Stiff Person Syndrome Plus (PERM)	Glutamic acid decarboxylase
<b>Glycine receptor Ab</b>	Stiff Person Syndrome Plus or "PERM" (progressive encephalopathy with rigidity and myoclonus)	
<b>Channels</b>		
<b>VGKC (CASPR2)</b>	Isaacs (neuromyotonia) – Morvan syndrome (Neuromyotonia, encephalopathy, hyperhidrosis) - Limbic encephalitis	Contactin associated protein type 2
<b>VGKC (LGI-1)</b>	Limbic Encephalitis	Leucine-rich, glioma Inactivated protein 1

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		Brief faciobrachial dystonic seizures, memory loss, disorientation, hyponatremia in 60%. CSF with lymphocytosis and OCB in 50% of patients May be associated with ovarian teratoma ( <a href="#">get pelvic MRI</a> )
<b>NMDA</b>	NMDA Encephalitis	Psychiatric features, cognitive dysfunction, seizures
<b>DPPX</b>	DPPX associated encephalitis	dipeptidyl-peptidase-like protein 6, a peptide related to VGKC responsible for attenuation of back-propagation of action potentials Start with diarrhea, weight loss followed by CNS hyperexcitability (hyperekplexia, myoclonus, seizures) over a few months period.
<b>Onconeuroal Ab</b>		
<b>Amphyphysin</b>	Stiff Person Syndrome (paraneoplastic)	SCLC & breast cancer, protein present on cytoplasmic surface of synaptic vesicles.
<b>Hu (ANNA-1)</b>	Encephalomyelitis (limbic, brainstem, cortical, myelitis) – sensory peripheral neuropathy – cerebellar degeneration	SCLC & Neuroblastoma, directed against neuronal nuclear protein (present in all neurons).
<b>Yo</b>	Cerebellar degeneration	Ovarian, endometrial % breast cancer
<b>Ri (ANNA-2)</b>	Cerebellar degeneration – Opsoclonus	Ovarian, endometrial % breast cancer, directed against NOVA protein Most common cause of opsoclonus in adults: Anti Hu, Ri, Yo (SCLC & breast) Most common cause of opsoclonus in children: neuroblastoma with negative anti Hu, Ri, Yo
<b>Ma2</b>	Cerebellar degeneration – Limbic encephalitis – Stiff person syndrome	Testicular tumors
<b>CV2</b>	Cerebellar degeneration – Limbic encephalitis – Peripheral neuropathy	SCLC, thymoma & uterine sarcoma.
<b>Glycoproteins</b>		
<b>GQ1b</b>	Miller Fischer Syndrome – GBS with ophthalmoplegia – Bickerstaff encephalitis – Pharyngo-cervical-brachial GBS	Positive in 90% of patients with MFS
<b>GM1</b>	AMAN – AMSAN – MMN	Associated with axonal variants of GBS
<b>GD1b</b>	Pure sensory variant of GBS	Against gangliosides on sensory neurons in dorsal root ganglia
<b>MAG</b>	Anti MAG neuropathy (Chronic sensory-motor demyelinating neuropathy) – Multiple sclerosis – SLE – MGUS/Waldestrom	Myelin associated glycoprotein (present in peripheral and central myeline) MAG Ab present in 50% of patients with monoclonal gammopathy (MGUS or Waldestrom) with peripheral neuropathy >> <a href="#">test for MAG in patients with MGUS/Waldestrom with neuropathy.</a>
<b>Other</b>		
<b>Endothelial cell Ab</b>	Susac	Triad of encephalopathy, branch retinal artery occlusion, hearing loss
<b>SSA (Ro) – SSB (La)</b>	Sjogren	Axonal neuropathy (pure sensory or sensory-motor), Sensory ganglionopathy, Small fiber neuropathy, Autonomic neuropathy SSA/SSB are only positive in 40% of Sjogren patients presenting with neurological diseases.