

AUTOIMMUNE AUTONOMIC GANGLIONOPATHY

What is Autoimmune Autonomic Ganglionopathy?

Autoimmune Autonomic Ganglionopathy (AAG), also known as acute pandysautonomia or idiopathic subacute autonomic neuropathy, is a rare form of dysautonomia.¹ Some patients can have a rapid onset, while others have a more progressive presentation of symptoms.²

Incidence/Prevalence

AAG usually affects patients during their middle ages, and affects females more often than males.¹

Symptoms

Symptoms are representative of the autonomic failure of the sympathetic, parasympathetic, and enteric nervous systems.¹

The most prevalent symptom associated with AAG is orthostatic hypotension (OH), a drop in blood pressure that occurs with changes in position.¹

Other symptoms may include: ¹

- Dry eyes and mouth
- Adie's pupil (tonic, sluggishly reactive pupils)
- gastroparesis,
- Neurogenic bladder (bladder dysfunction which may include overflow incontinence, frequency, urgency, urge incontinence, and retention) ^{2,3}
- Paresthesia (a burning or prickling sensation in the extremities)



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Diagnosis

AAG can be diagnosed after a thorough history, medical examination, and autonomic testing.¹ Some patients with AAG have high levels of antibodies for the ganglionic nicotinic acetylcholine receptor (gAChR).² It is believed that these antibodies damage the receptors in the autonomic ganglia, a part of the peripheral autonomic nerve fiber. This interferes with the autonomic transmission, resulting in autonomic failure.^{1,4}

Treatment

Treatment of AAG is targeted at the antibody-mediated pathology, though there is no clear established treatment protocol. Treatment for this form of dysautonomia is usually focused on treating associated symptoms.^{1,4}

The first treatment often used with AAG is antibody depletion through plasma exchange and intravenous immunoglobulin (IVIG).¹ Immunosuppressant medications and corticosteroids can be used in conjunction with antibody depletion.¹

Nonpharmacologic and pharmacologic treatment of OH and artificial tears and saliva for dry mouth and eyes can be used to treat specific symptoms.^{1,4}

Some patients experience a spontaneous recovery without treatment, though for many, recovery is often incomplete and they experience some level of symptoms throughout their life.⁴



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