DYSAUTONOMIA SUPPORT NETWORK

Autoimmune Autonomic Ganglionopathy

What is AAG?

- Autoimmune autonomic ganglionopathy (AAG) is a rare auto-immune disorder that causes pure autonomic neuropathy (damage to the small peripheral nerve fibers), which can lead to selective or generalized autonomic failure
- AAG used to be called pandysautonomia, until they determined the pathology behind it
- In most cases, high levels of ganglionic acetylcholine receptor antibodies are seen (g-AChR), but some patients may not have these antibodies present. There is evidence to suggest that the levels of antibodies correlate with severity of autonomic dysfunction
- About 60% of cases are followed by an illness or infection
- Symptom onset may be sudden or gradual
- Research is still ongoing to determine diagnostic testing and treatment

What are the symptoms?

- Severe orthostatic hypotension
- Fainting (syncope)
- GI dysmotility and constipation
- Urinary retention
- Fixed and dilated pupils (may also be sluggish or may constrict and then redilate while light is still on)
- Dry mouth and eyes
- Some people present with more POTS-like symptoms

For more information visit
http://www.dysautonomiainternational.org/page.php?ID=124

https://www.ncbi.nlm.nih.gov/pubmed/27112689

Additional sources,
https://rarediseases.info.nih.gov/diseases/11917/autoimmune-autonomic-ganglionopathy

How is it diagnosed?

- Tilt table test and other autonomic testing
- g-AChR antibody test

How is it treated?

- No standard treatment has been extensively studied and established
- Treatments used in other autoimmune diseases, such as IVIG, IV corticosteroids, plasmapheresis, immunosuppressive drugs
- Treatment for symptom management of dysautonomia is also used such as **midodrine, florinef, droxidopa, octreotide, and mestinon.**
- **Non-pharmacological treatments like exercise, increasing salt and fluid intake, compression stockings and good sleep habits can also help.**
- **Some patients may occasional resolve on their own (particularly after illness or infection)**