



## **Autonomic Nervous System: Chapter 25. Autoimmune autonomic failure (Handbook of Clinical Neurology)**

*Srikanth Muppidi, Steven Vernino*

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Autoimmune autonomic ganglionopathy is a syndrome of panautonomic failure caused by antibodies to ganglionic acetylcholine receptors. The clinical syndrome is characterized by significant postural hypotension, diffuse cholinergic and adrenergic impairment, gastrointestinal dysmotility, urinary retention, and pupillary dysfunction. While acute to subacute onset of disease is commonly seen, chronic, slowly progressive variants have been described. Serological testing for ganglionic acetylcholine receptor antibodies helps confirm the diagnosis. These antibodies cause a similar phenotype of autonomic failure in animal models indicating that an antibody-mediated functional impairment of ganglionic transmission is the underlying etiology. Decrease in antibody levels correlates with clinical improvement. Patients may respond to immunomodulatory therapies such as prednisone, intravenous immunoglobulin, plasma exchange, and oral immunosuppressants. A combination treatment is often required as well as symptomatic therapy.

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