

PURE AUTONOMIC FAILURE

What is Pure Autonomic Failure?

Pure Autonomic Failure (PAF) is a rare form of dysautonomia.¹ It is also known as **Bradbury-Eggleston Syndrome** and is a primary dysautonomia.

PAF causes deterioration of autonomic nervous system cells and therefore causes overall autonomic nervous system dysfunction.²

Recent research has shown a relationship between PAF and later developing other conditions such as MSA, Parkinson's disease, and dementia.¹

The **average age of onset is middle age** and it is more common in males than females.³

Symptoms

In one study, participants reported the following symptoms:⁴

- Orthostatic hypotension
- Supine hypertension (50%)
- Genitourinary dysfunction (50%)
- Constipation (58%)
- Sweating abnormalities (44%)

Incidence/Prevalence

Pure autonomic failure is a rare form of dysautonomia and impacts **fewer than 200,000 people** in the USA.⁵



For references, please scan the QR code. (C) 2022

Diagnosis

Official diagnostic criteria for PAF, per consensus statement, have not been updated since 1996.¹ The diagnostic criteria are fairly vague and include: **neurogenic orthostatic hypotension**, **other autonomic symptoms** are also often present, and that there are **no other neurological symptoms**.¹ Neurogenic orthostatic hypotension is diagnosed through assessing orthostatic vital signs.⁶

PAF is often diagnosed at facilities with access to a full range of complex autonomic function tests in order to help clarify diagnosis. This includes measuring plasma catecholamine levels, Valsalva maneuver, baroreflex testing, QSART, etc.¹

Treatment

Treatment for PAF involves symptom management, specifically controlling blood pressure fluctuations.¹ To address orthostatic hypotension, **non-pharmacological measures** include increasing fluid intake, salt intake, exercise, elevating the head of the bed, and using compression garments.¹

Medications aimed to address orthostatic hypotension include midodrine, droxidopa, fludrocortisone, and pyridostigmine.^{1,2} Supine hypertension may be treated with nitroglycerin, or clonidine.¹

Other symptoms like constipation are managed first with non-pharmacological means like increasing water and fiber intake.^{1,2}



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