Brief Communication

*Clinical Autonomic Research* 2001, 11:265–267

# Postural tachycardia syndrome in syringomyelia: response to fludrocortisone and β-blockers

**Martín Nogués, M.D.,[1] Ricardo Delorme, M.D.,[1] Daniela Saadia, M.D.,[1] Kirsten Heidel,[2] and Eduardo Benarroch, M.D.[2]**

[1]Department of Neurology, Raúl Carrea Institute for Neurological Research (FLENI), Buenos Aires, Argentina; and [2]Department of Neurology, Mayo Clinic, Rochester, Minnesota, USA

Address correspondence and reprint requests to Martín A. Nogués, M.D., Montañeses 2325, (1428) Buenos Aires, Argentina.
Tel: (5411) 4788-3444; Fax: (5411) 4784-7620
E-Mail: mnogues@fleni.org.ar

*Received April 6, 2001; accepted August 16, 2001*

**Orthostatic intolerance is occasionally reported by patients with syringomyelia and is usually attributed to vestibular symptoms or neurogenic orthostatic hypotension. Postural tachycardia syndrome has not been previously described in syringomyelia. A patient with long-standing syringomyelia and a Chiari type I anomaly developed disabling "panic-like" attacks associated to orthostatic intolerance five years after posterior fossa decompression and shunting of the syrinx. A head-up tilt test showed an early phase of postural orthostatic tachycardia followed by progressive arterial hypotension and bradycardia as seen in neurally mediated syncope. A magnetic resonance imaging scan showed a collapsed syrinx from the 3rd cervical to the 12th thoracic vertebra without syringobulbia. Fludrocortisone and β-blockers led to resolution of symptoms. Partial sympathetic denervation of the legs in syringomyelia might explain the occasional occurrence of postural tachycardia syndrome. Postural tachycardia syndrome may be included as a possible cause of orthostatic symptoms in syringomyelia patients.**

*Key words:* postural tachycardia syndrome (POTS), syringomyelia, syncope, autonomic dysfunction.

Autonomic abnormalities in patients with syringomyelia include vasomotor and sudomotor changes in affected limbs, Horner's syndrome, and urinary incontinence [1]. Orthostatic hypotension, sleep-disordered breathing, and sudden death may also occur when syringobulbia is present [2–4]. That syringobulbia is able to cause orthostatic hypotension has been known since the work of Ellis and Haynes [5]. They described a 45-year-old man who, six years after the onset of his illness, started to notice blurring of vision and dizziness when erect. It was thought that the disease was affecting the vasomotor centers within the brain stem. Aminoff and Wilcox [2] reported a case with syringomyelia without bulbar involvement and with widespread autonomic dysfunction, including orthostatic hypotension. They assumed that the Chiari anomaly might have interrupted descending sympathetic fibers or that the intermediolateral columns or autonomic outflow pathways in the spinal cord may have been affected.

We here describe a patient with long-standing syringomyelia and relatively preserved motor function who developed paroxysmal cardiovascular and gastrointestinal symptoms initially interpreted as psychogenic. Our aim is to describe the occurrence of postural orthostatic tachycardia syndrome (POTS) in a patient with syringomyelia, and its complete resolution after treatment with β-blockers and fludrocortisone. This case highlights the value of autonomic studies to define the organic nature of otherwise unexplained episodes and the value of this treatment for very disabling orthostatic symptoms.

## Case report

This 45-year-old right-handed woman was in good health up to age 33, when she noticed numbness of the right leg, headaches triggered by coughing or sneezing, occasional dysphagia, and unsteadiness of gait. In June 1990, a magnetic resonance imaging examination showed a Chiari type I anomaly and a cervicothoracic syrinx extending from the 3rd cervical to the 12th thoracic vertebra. In December 1990, she underwent a syringo-subarachnoid shunt placement, which arrested symptom progression, and, three years later, a foramen magnum decompression and a second syringo-subarachnoid shunt placement, which led to resolution of her headaches.

Over the last four years, she started experiencing episodes of palpitations, dyspnea, nausea, light-headedness, and a tendency to fall, associated with fear and anxiety. These episodes occurred mostly while standing, but sometimes awakened her from sleep. Episodes lasted from minutes to hours and became more severe in the last six months. There were no abnormalities on examination during the events, except for sinus tachycardia, and therefore the episodes were considered psychogenic in origin. Treatment with benzodiazepines or domperidone produced no relief, and a single oral dose of chlorpromazine (25 mg) induced marked arterial hypotension that lasted approximately six hours. On examination, she had a short neck, kyphoscoliosis, and a mild spastic paraparesis, bilateral extensor plantar responses, and reduction in all sensory modalities below level of the

4th thoracic vertebra. Despite these findings, the patient was able to walk, being fully ambulant. Cranial nerves and upper limbs were normal.

A recent magnetic resonance imaging scan disclosed tonsillar descent, cord atrophy, and a collapsed syrinx from the 3rd cervical to the 12th thoracic vertebra (Fig. 1). A 24-hour electrocardiographic monitoring showed a mean heart rate of 87 beats/min, ranging from 60 to 156 beats/min. The patient's plasma cortisol concentration was 12.5 μg/dl (normal range 7.0–25.0).

The patient underwent autonomic screening for cardiovascular responses to deep breathing, Valsalva maneuver and head-up tilt tests. Expiration:inspiration ratio was 1.5 and the Valsalva ratio 1.6. Head-up tilt (75°) with electrocardiographic and intermittent blood pressure monitoring showed an initial asymptomatic phase, lasting 10 minutes, of excessive sinus tachycardia disproportionate to the decrease in blood pressure. This was followed by progressively more severe tachycardia and hypotension, occurring between 10 and 16 minutes of head-up tilt, and associated with generalized weakness, palpitations, and a tendency to fall (Fig. 2A). At 16 minutes, there was a further decrease in blood pressure with relative bradycardia and syncope. Intravenous infusion of 400 cc saline resulted in attenuation of the cardiovascular responses to tilt (Fig. 2A). She was started on 12.5 mg atenolol twice a day and 0.1 mg fludrocortisone per day. This treatment resulted in a complete resolution of her symptoms and a sensation of well-being during a follow-up period of eight months. Results of a follow-up tilt test were normal (Fig. 2B).

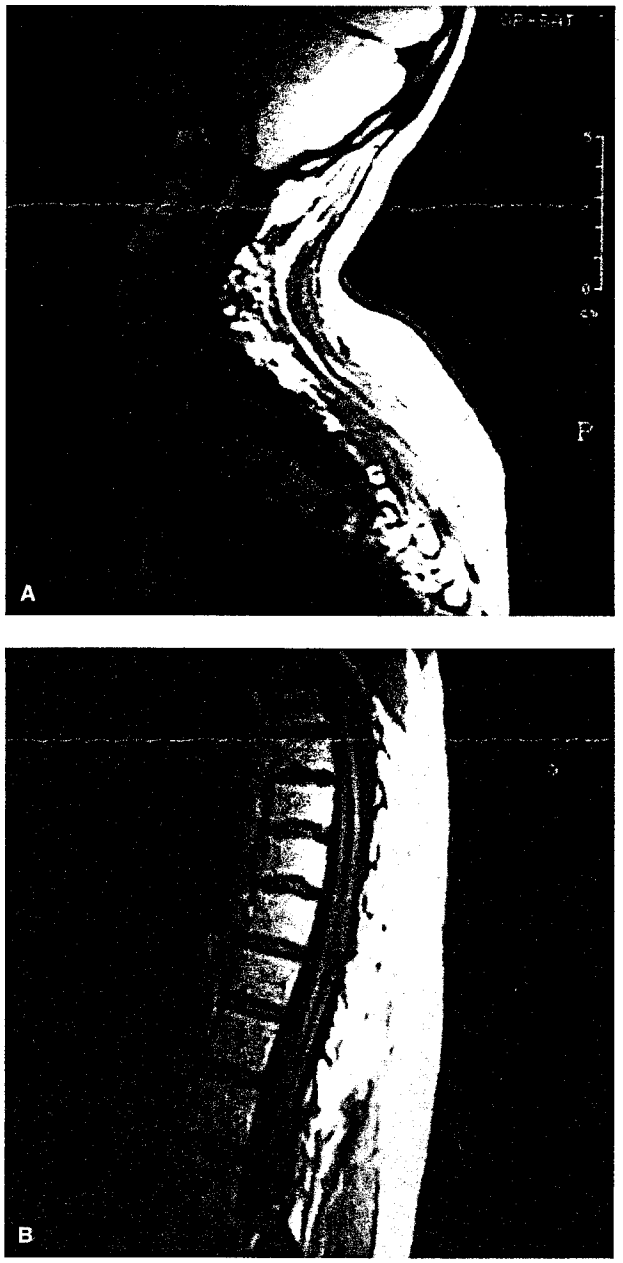


**Figure 1.** (**A**) Cervical spine (by magnetic resonance imaging): Chiari I anomaly, cord atrophy, and a collapsed syrinx; and (**B**) thoracic spine (by magnetic resonance imaging): an extensive syrinx along the thoracic spinal cord.

## Discussion

We here describe a patient with syringomyelia, whose main disability was a result of episodes of palpitations, dyspnea, lightheadedness, dizziness, and sensation of impending loss of consciousness. The patient's tilt test showed a disproportionate tachycardia during the first 10 minutes after tilting, with complete resolution of symptoms with β-blockers and fludrocortisone. Her symptoms were initially regarded as functional (anxiety or panic attacks). The fact that the patient was fully ambulant and carried on a very active life despite her neurological disease favored the appearance of the aforementioned orthostatic symptoms. Her advanced disease may be confirmed by its long duration, severe neurological disability, and marked spinal cord atrophy dem-

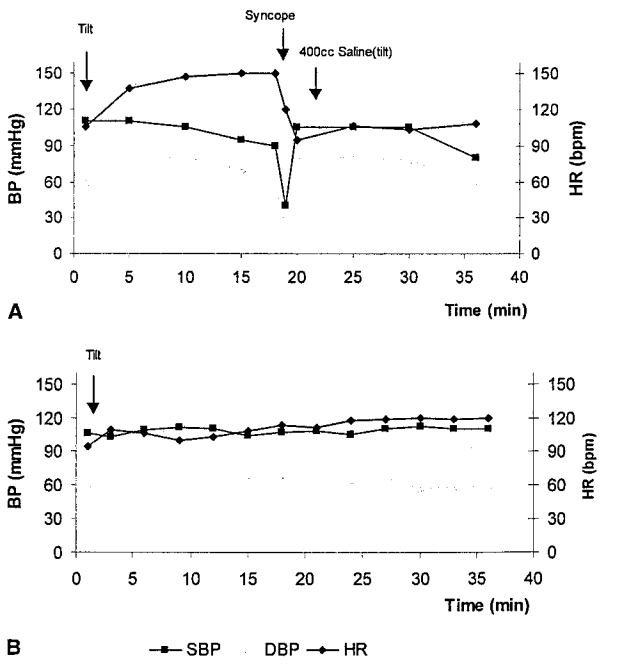


**Figure 2.** (**A**) Baseline head-up tilt test: initial tachycardia without arterial hypotension, followed by hypotension, bradycardia, and syncope. Improved responses after intravenous saline infusion. (**B**) Head-up tilt test after treatment.

onstrated by magnetic resonance imaging. Although it is not appropriate to use the term postural orthostatic tachycardia syndrome (POTS) when a known neurologic condition liable to produce autonomic insufficiency is demonstrated, both symptoms and tilt-test findings are more suggestive of POTS than of neurogenic orthostatic intolerance.

The development in this patient of orthostatic symptoms associated with excessive tachycardia (over 30 beats/min from baseline or over 120 beats/min), within 5 minutes of head-up tilt and in the absence of orthostatic hypotension, is a feature of the postural tachycardia syndrome [5,6]. Whereas the initial response during tilt resembled that of patients with POTS, the late phase resembled that of neurally-mediated syncope. However, the two conditions are not mutually exclusive, and these cases are referred to as having POTS with syncope [7,9]. The fact that the patient was fully ambulant and carried on a normal life favored the appearance of orthostatic intolerance. It is possible that this syndrome may go unrecognized in patients with syringomyelia who are confined to bed or wheelchair.

Pathophysiological types of POTS include mild orthostatic intolerance, idiopathic POTS, POTS associated with idiopathic autonomic neuropathy, POTS associated with mitral valve prolapse, and hypertensive POTS. In the case we present, the presence of syringomyelia suggests a "secondary" POTS. The mechanisms of POTS are complex and include peripheral denervation resulting in poor vasomotor tone and venous pooling, β-adrenergic receptor supersensitivity, hypovolemia, sympathetic-parasympathetic imbalance, and "brain stem" mechanisms [6,7]. Some patients with POTS display a mild distal sympathetic venomotor neuropathy that results in venous pooling [6–8]. Our patient had no history of previous viral illness or cyclic variations of her symptoms that are usual in idiopathic POTS cases. Hypovolemia and deconditioning with venous pooling are common mechanisms of POTS, and could have contributed to the development of symptoms in our patient. The presence of an extensive syrinx from the 3rd cervical to the 12th thoracic vertebra and cord atrophy, together with the favorable response to volume expansion, suggest that venous pooling and reduced venous return as a result of involvement of sympathetic structures in the spinal cord may have contributed to the appearance of POTS in our patient. Neuronal loss in the intermediolateral columns by a "degenerative" neuronal process might explain the late appearance of symptomatic POTS in our patient. This mechanism has been advocated to explain why patients with syringomyelia may show slowly progressive deterioration despite adequate drainage of the syrinx [12].

It is unlikely that the associated Chiari type I anomaly was responsible for the appearance of POTS in this patient because it had been previously resolved by foramen magnum decompression, and there was no evidence of brain stem or upper cervical spinal cord compression.

The patient's normal expiration:inspiration and Valsalva ratios suggest that cardiac sympathetic and parasympathetic functions were relatively preserved, inasmuch as the tachycardia during phase II of the Valsalva maneuver is affected by both inhibition of cardiovagal and activation of cardiac sympathetic outflows [10]. The occurrence of neurally-mediated syncope [11] during the late phase of head-up tilt testing in the setting of cardiovagal failure has two main implications: (1) it suggests the major contribution of a vasodepressor mechanism, with abrupt interruption of a relatively preserved sympathetic vasomotor tone; and (2) together with the preserved Valsalva ratio, it implies that medullary cardiovascular reflexes were relatively preserved in our patient.

In summary, our case suggests that POTS may be an early manifestation of autonomic dysfunction in syringomyelia. Its detection by means of a head-up tilt test and its appropriate treatment with volume expansion and β-blockers may lead to significant relief of disabling postural symptoms, otherwise misdiagnosed as psychogenic. Further studies measuring the norepinephrine spillover in arms and legs in response to diverse stimuli may help to confirm our hypothesis and detect further cases with this disabling syndrome.

## References


1. Nogués MA. Syringomyelia and syringobulbia. In: *Malformations. Handbook of clinical neurology.* Vinken PJ, Bruyn GW, Klawans HL, eds. Amsterdam: Elsevier; 1987. pp. 443–464.
2. Aminoff MJ, Wilcow CS. Autonomic dysfunction in syringomyelia. *Postgrad Med J* 1972; 48:113–115.
3. Nogués MA, Newman PK, Male VJ, *et al.* Cardiovascular reflexes in syringomyelia. *Brain* 1982; 105:835–849.
4. Nogués MA, Gené R, Benarroch E, *et al.* Respiratory disturbances during sleep in syringomyelia and syringobulbia. *Neurology* 1999; 52:1777–1784.
5. Ellis LB, Haynes FW. Postural hypotension. With particular reference to its occurrence in disease of the central nervous system. *Arch Int Med* 1939; 58:773–798.
6. Low PA, Opfer-Gehrking TL, Textor SC, *et al.* Postural tachycardia syndrome (POTS). *Neurology* 1995; 45(Suppl 5):S19–S25.
7. Low PA, Schondorf R, Novak V, *et al.* Postural tachycardia syndrome. In: *Clinical autonomic disorders.* 2nd ed. Low P, ed. Philadelphia: Lippincott-Raven; 1997. pp. 681–697.
8. Jacob G, Costa F, Shannon JR, *et al.* The neuropathic postural tachycardia syndrome. *New Engl J Med* 2000; 343:1008–1014.
9. Sandroni P, Opfer-Gehrking TL, Benarroch E, *et al.* Certain cardiovascular indices predict syncope in the postural tachycardia syndrome. *Clin Auton Res* 1996; 6:225–231.
10. Novak VA, Novak P, Low PA. Time-frequency analysis of cardiovascular function and its clinical implications. In: *Clinical autonomic disorders.* 2nd ed. Low P, ed. Philadelphia: Lippincott-Raven; 1997. pp. 323–348.
11. Kaufmann H. Neurally-mediated syncope: pathogenesis, diagnosis and treatment. *Neurology* 1995; 45(Suppl 5):S12–S18.
12. Logue V, Rice-Edwards M. Syringomyelia and its surgical treatment—an analysis of 75 patients. *J Neurol Neurosurg Psychiatry* 1981; 44:273–284.