

## SHORT REPORT

## Orthostatic intolerance and syncope associated with Chiari type I malformation

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*J Neurol Neurosurg Psychiatry* 2005;76:1034–1036. doi: 10.1136/jnnp.2004.048330

The Chiari type I malformation (CM1) is characterized by herniation of cerebellar tonsils to at least 3–5 mm below the plane of foramen magnum and can present with a wide variety of clinical symptoms, frequently including occipital headaches, secondary to bulbar and/or medullary distress. Rarely, syncopal episodes have also been described and attributed to either compression of the midbrain ascending reticular system, or vascular compromise (vertebrobasilar artery compression, hypotension). We report the first case of a CM1 patient with frequently recurring syncope due to postural orthostatic tachycardia syndrome (POTS), a form of orthostatic intolerance, whose symptoms resolved completely after surgical intervention. It is important to stress that it is not clear whether the described association of POTS and CM1 in our patient is a fortuitous finding in an isolated case or a reflection of a more systematic association between the two pathologies.

The Chiari type I malformation is characterized by herniation of cerebellar tonsils to at least 3–mm below the plane of foramen magnum. It usually becomes symptomatic in adulthood and presents with a number of symptoms secondary to medullary distress, such as headaches, often occipital in location, unsteady gait, sensorimotor abnormalities, and lower cranial nerve palsies. Several cases of associated syncope have been reported as the presenting symptom or as one component of the presenting symptom complex.<sup>1–3</sup> Various pathophysiological mechanisms such as compression of the midbrain ascending reticular system, or vascular compromise (vertebrobasilar artery compression, hypotension secondary to cardiorespiratory center compression) have been proposed, although the exact cause of CM1 related syncope is still not well understood.<sup>4</sup> We report the first case of syncope secondary to orthostatic intolerance in a CM1 patient, which resolved after surgical intervention.

## CASE REPORT

A 42 year old female patient presented with gradually worsening occipital headaches associated with nausea and vomiting, chronic fatigue, dizziness, discrete right hemicorporeal hypoaesthesia, and recurrent syncope over a 1 year period. The syncopal episodes always occurred while the patient was in the upright position and were preceded by tachycardia, pallor, sweating, dizziness, oscilloscopy, and/or diplopia and were followed by the appearance of right sensorimotor syndrome, dysarthria, and fatigue, which resolved several hours later. These syncopal episodes occurred several times per week and could be prevented by the patient lying down. The high frequency and related neurological deficits became a serious handicap for the patient, and she was referred for investigation.

The patient had a normal neurological examination, routine blood tests, and cerebral MRI. A prolonged EEG of

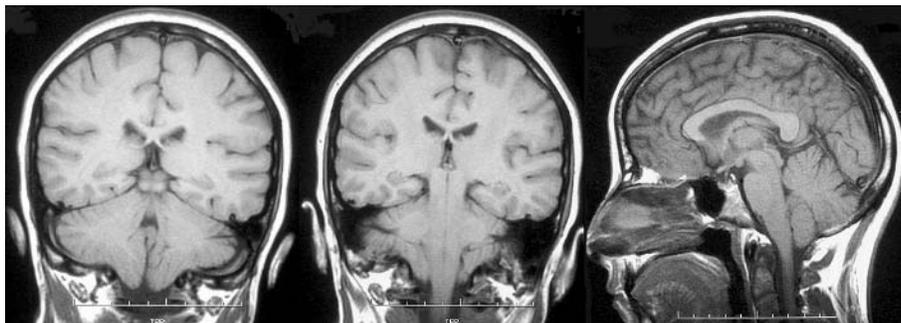
10 hours during which she presented the aforementioned presyncopal symptoms was unrevealing. The cardiac evaluation included echocardiography and a 24 hour Holter recording and were both normal. A Doppler examination of exocranial and endocranial arteries was also normal. A Schellong test (blood pressure (BP) and pulse monitoring every minute for 3 minutes in the resting supine position, followed by standing up immobile and unsupported for up to 20 minutes with BP and pulse monitoring every minute) demonstrated signs of orthostatic intolerance. BP increased from 103/68 mmHg to 154/77 mmHg after movement from the horizontal to vertical position, associated with an increase in heart rate (HR) from 73 to 111 beats/min (38 beats/min increase) and a maximum HR of 130 beats/min during the test (table 1). These BP and HR changes were accompanied by the recurrence of the patient's usual pre-syncopal symptomatology. A peripheral neuropathy was excluded by extensive electroneuromyographic evaluation. The initial MRI was, however, reviewed and reinterpreted as showing a Chiari type I malformation with tonsillar herniation of 7 mm below the plane of foramen magnum (fig 1). A surgical osteodural decompression with posterior fossa craniectomy and C1 laminectomy were subsequently performed and the arachnoid adhesion dissected to establish spontaneous CSF flow from the fourth ventricle followed by a duroplasty. The patient's syncopal episodes, gait unsteadiness, and visual and sensorimotor symptoms all disappeared after surgery and remained absent at the 3 month and 1 year post-surgical follow up visits. The Schellong test at 3 and 12 months (table 1; 12 month data not shown) postoperatively were also normal, although her chronic fatigue symptoms remained unchanged.

## DISCUSSION

We describe a CM1 patient with very frequent recurrent syncope, demonstrated both clinically and neurophysiologically to be due to orthostatic intolerance, who had no further syncopal episodes after surgical decompression of the tonsillar herniation.

Syncope is a relatively rare symptom of CM1 and may occur with or without premonitory symptoms. Various pathophysiological mechanisms have been proposed: (a) vertebrobasilar artery compression secondary to a transient increase of intracranial pressure during a Valsalva manoeuvre, (b) compression of the midbrain ascending reticular system, and (c) compression of cardiorespiratory centers or their efferent/afferent pathways. A certain similarity of symptoms such as dizziness, nausea, and fatigue is seen in patients with CM1, chronic fatigue syndrome, and orthostatic intolerance (OI), leading to a debate on their possible interrelation.<sup>4–6</sup> It has been proposed that hindbrain compression with or without CM1 could be a cause of symptoms in OI

**Abbreviations:** BP, blood pressure; CM1, Chiari type I malformation; HR, heart rate; NTS, nucleus tractus solitarius; OI, orthostatic intolerance; POTS, postural orthostatic tachycardia syndrome



**Figure 1** Pre-operative MR T1 weighted coronal and sagittal acquisitions showing the descent of cerebellar tonsils in the foramen magnum. Surgical decompression resulted in the disappearance of syncopes and POTS.

and that surgical decompression would consequently improve the situation. There is, however, little published literature to support or dispute this hypothesis.<sup>4</sup> In a MRI study on 23 patients with OI, there was no evidence of a higher prevalence of cerebellar herniation in the patient group as compared to controls.<sup>7</sup> However, the patient number may be insufficient to draw final conclusions.

Orthostatic intolerance is defined by some authors as the presence of long standing disabling orthostatic symptoms and >30 beats/min increase in heart rate on standing or during a head up, tilt table test, along with a concomitant exaggerated increase in plasma noradrenaline (norepinephrine). The same clinical entity, in the absence of specification of norepinephrine levels, is also referred to as postural orthostatic tachycardia syndrome (POTS).<sup>6</sup> The usual diagnostic criteria for POTS are: a heart rate increase of at least 30 beats/min within 5 minutes of standing or upright tilt, a heart rate of at least 120 beats/min within 5 minutes of standing or upright tilt, development of orthostatic intolerance symptoms without significant blood pressure changes and absence of a known cause of autonomic neuropathy.<sup>9, 10</sup> Thus our patient with orthostatic intolerance actually manifested the typical clinical features of POTS patients.<sup>8</sup> At the present time, however, there is no definite consensus on diagnostic criteria of OI and POTS, often leading to interchangeable use of both terms by different authors.<sup>6</sup> Both OI and POTS present at a relatively young age (14–45 years) and show a female preponderance. The pathophysiology of

these disorders is not well understood and various aetiologies have been proposed: autoimmune process, reduced blood volume, presence of circulating vasodilator, or a primary abnormality in autonomic regulation at central or peripheral levels. In our patient, POTS leading to syncope seems to have developed in parallel with the other symptoms of CM1 previously described, and disappeared after decompressive surgery, thereby suggesting a central origin, probably by direct hindbrain compression. The appearance of post-syncope right sided sensorimotor deficits is also in favour of a transient CNS insult at the moment of syncope and is recognized as one of the more constant clinical features of CM1 patients.<sup>1</sup> Indeed, a transient malfunction at the brain stem level could affect several structures involved in sympathetic/parasympathetic cardiovascular regulation constituting the baroreflex system. The baroreflex system is one of the major mechanisms assuring the buffering of arterial blood pressure changes.<sup>11, 12</sup> It is mediated by afferent pathways via the ninth and tenth cranial nerves, relaying information from vascular baroreceptors to the nucleus tractus solitarii (NTS), which sends excitatory projections to the caudal ventrolateral medulla, which in turn inhibits the sympathoexcitatory neurones of the rostral ventrolateral medulla. Lesions of the NTS or efferent/afferent baroreflex pathways result in baroreflex failure, leading typically to a marked lability of blood pressure, with hypertension and tachycardia alternating with hypotension and bradycardia. However, depending on the degree and probably the site of injury of the baroreflex mediating system, other clinical presentations could occur, such as orthostatic tachycardia.<sup>11</sup>

To our knowledge, this is the first report of a direct relationship between the CM1 and POTS. Nevertheless, it is important to stress that it is not clear whether the described association of POTS and CM1 in our patient is a fortuitous finding in an isolated case or a reflection of a more systematic association between the two pathologies. Further studies are needed in order to determine the exact relationship between OI/POTS and CM1. We emphasise the fact that our patient's chronic fatigue symptoms have not improved after surgery, thereby suggesting different mechanisms for POTS and chronic fatigue in the context of CM1 in our patient.

**ACKNOWLEDGEMENTS**

This study was supported by grant nos 32-68105 and 32-104146.

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Competing interests: none declared

**Table 1** Pre-surgical and 3 months post-surgical Schellong test values

	Pre-surgical		Post-surgical (3 months)	
	BP (mmHg)	Pulse (beats/min)	BP (mmHg)	Pulse (beats/min)
Supine				
1 min	113/70	76	114/77	70
2 min	105/69	72	119/74	70
3 min	103/88	73	121/75	69
Standing				
0 min*	154/77	111	113/70	74
1 min	119/77	122	131/74	86)
2 min	121/82	126	120/69	83
3 min	114/80	130	116/77	78
4 min	131/65	129	122/81	84
5 min†	121/74	128	121/80	84
6 min			128/79	82
7 min			119/71	83
8 min			112/69	79
9 min			115/70	85
10 min			115/70	85
15 min			142/85	96
20 min‡			122/67	82

\*On standing up the patient presented her usual symptoms of vertigo, diplopia and sweating. †Test stopped because of imminent syncope. ‡Patient remained asymptomatic throughout the study.

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Received 25 June 2004  
In revised form 17 October 2004  
Accepted 29 October 2004

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## NEUROLOGICAL PICTURE

doi: 10.1136/jnnp.2004.044636

### Pneumorrhachis of the entire spinal canal

**P**neumorrhachis (PR)—the curious phenomenon of intraspinal air—is an exceptional radiographic finding. In this report the first case of PR of the entire spine is presented.

A 19 year old man with a history of diabetes mellitus and bronchial asthma was admitted because of cough, fever, nausea, and vomiting for 3 days. Physical examination and laboratory studies showed cervical subcutaneous emphysema, infection signs, and ketoacidotic decompensated diabetes mellitus with hyperglycaemia.

Subsequent computed tomography (CT) (fig 1) revealed pneumomediastinum, pneumoretroperitoneum, cervical and thoracic subcutaneous emphysema, external pneumocephalus, and PR of the entire spinal canal.

The patient underwent otorhinolaryngological exploration for presumptive diagnosis of nasopharyngeal abscess but no evidence of an infectious process was found.

With intravenous antibiotics for gastrointestinal infection and anti-hectic medication the symptoms resolved and the PR reabsorbed completely. The patient was discharged home asymptotically after 12 days.

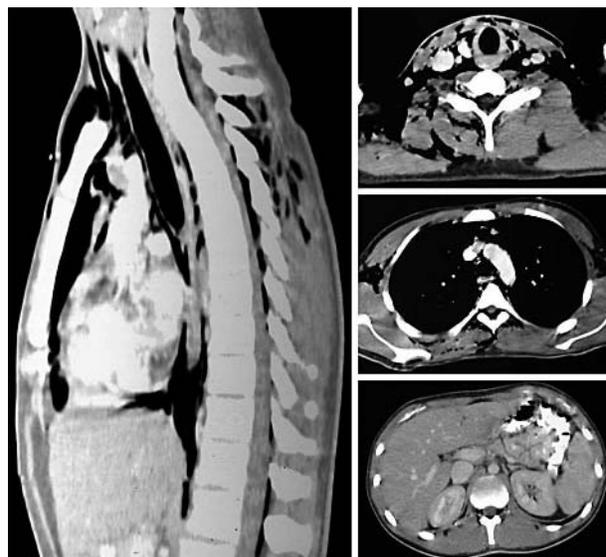
In this case, PR was secondary to tracheobronchial microinjury resulting from violent coughing. The air dissected from the upper respiratory tract into the paraspinal soft tissues of the neck, entered the spinal canal via the neural foramina, and extended epidurally throughout the entire vertebral canal (fig 1).

Only one similar case with air distribution in the cervical and lumbar spine,<sup>1</sup> and in the thoracic and lumbar region,<sup>2</sup> and three cases with both cervical and thoracic PR were previously described.<sup>2–4</sup>

The current case serves to highlight the importance of PR as a rare and usually asymptomatic but eminent epiphenomenon of associated, frequently concealed, severe pathologies, so as to enable adequate patient management.

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**Figure 1** Left panel: sagittal reconstruction of contrast enhanced spinal CT scan demonstrating intraspinal epidural air collection accompanied by subcutaneous emphysema and pneumomediastinum. Transaxial contrast enhanced axial spinal CT images of the cervical (upper right panel), thoracic (middle right panel), and lumbar region (lower right panel) revealing extradural PR of the entire spine associated with distinctive cervical and thoracic subcutaneous emphysema, pneumomediastinum, and pneumoretroperitoneum. Note air dissection via the neural foramina into the vertebral canal.

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