

legs was beneficial, and he was able to walk occasionally without assistance.

Autonomic function tests were performed after weaning from mechanical ventilation.<sup>4</sup> A catheter was inserted into the left radial artery, and vital signs were monitored. Head up tilting to 60° resulted in a blood pressure fall from 134/99 to 51/48 mm Hg. The basal plasma noradrenaline concentration was normal. A rise of plasma vasopressin in response to hypotension was absent, suggesting involvement of the afferent baroreflex pathway. There was no overshoot with Valsalva's manoeuvre, which suggested insufficient baroreflex function. The absence of a blood pressure rise to the cold pressor test suggested involvement of the efferent sympathetic system. A noradrenaline infusion test disclosed sympathetic supersensitivity. No  $\beta$ -1 supersensitivity in response to an isoprenaline infusion test was seen.<sup>4</sup> Increase of his heart rate from 90 to 109 per minute in response to an atropine test showed that the vagal efferents were not completely involved. The results of the remaining autonomic tests were normal.

The control of blood pressure after postural change is mediated by the baroreflex system. Failure of this reflex function can be due to a lesion anywhere in this reflex pathway. Impulses from the baroreceptors, originating from the carotid sinus and aortic arch, travel via the glossopharyngeal and vagal nerves. Most of these baroreceptor afferents terminate in the nucleus tractus solitarius, the major nuclear relay of the baroreflex.<sup>1,2,3</sup> In addition, the nucleus tractus solitarius not only regulates baroreceptor pathways but is also involved in the central regulation of respiration and swallowing.<sup>25</sup> The glossopharyngeal and vagal nerves also convey the sensory impulses from the pharynx, larynx, and the respiratory system to the nucleus tractus solitarius, which contains specific centres for respiration and swallowing.<sup>25</sup> Our patient presented with severe orthostatic hypotension, dysphagia, lingual atrophy, and temporary failure of automatic respiration, but had no pyramidal signs, sensory impairment, cerebellar ataxia, or a neurogenic bladder. Although we cannot discuss the relation of a non-metastatic autonomic neuropathy, neither spinal nor peripheral nerve lesions were found. Investigations of the somatic and autonomic functions suggested a medullary lesion. Severe orthostatic hypotension, the absence of overshoot of Valsalva's manoeuvre, a negative response to the cold water test, and a positive response to the atropine test are consistent with an incomplete failure of the baroreflex arc. Although it is difficult to determine the definite lesion responsible for the failure of the baroreflex arc, a tumour located in the dorsal medulla may cause impairment of the nucleus tractus solitarius, the dorsal vagal nucleus, and the hypoglossal nucleus, which are located in close proximity in the dorsal medulla.<sup>25</sup>

In conclusion, a localised lesion of the dorsal medulla as reported in this case may result in severe orthostatic hypotension, bulbar palsy, and failure of automatic respiration in the absence of pyramidal or sensorimotor signs in the limbs.

AKIKO YAMASHITA  
MASAAKI HIRAYAMA  
YASUO KOIKE  
Department of Neurology,  
Nagoya University School of Medicine,  
Japan  
MEGUMU NUKATA  
MAKOTO HASHIZUME  
Department of Neurology,

Anjo Kosei Hospital, Japan  
AKIRA TAKAHASHI  
Tokai Central Hospital, Japan

Correspondence to: Dr A Yamashita, Department of Neurology, Nagoya University School of Medicine, 65 Tsurumai-cho, Showa-ku, Nagoya, 466 Japan.

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### Tardive dystonia after neuroleptic treatment of Tourette's syndrome

We report a patient with Tourette's syndrome who developed tardive dystonia manifest by a task specific, action induced oromandibular dystonia after 15 years of treatment with haloperidol.

A 28 year old man with Tourette's syndrome since the age of seven had been treated with haloperidol (2-4 mg daily) with good effect since the age of 13. Motor and vocal tics had included eye blinking, facial grimacing, shoulder shrugging, head and neck movements, humming and squeaking noises, and occasional snapping-shut jaw movements. Obsessive-compulsive features included repetitive touching and counting behaviours. At the age of 28, he developed severe dystonic movements of the jaw, mouth, and tongue for the first time, which were activated by speaking. Immediately on beginning to speak he displayed forceful dystonic jaw opening and buccolingual movements associated with a pronounced dysarthria. He could prevent these by speaking with a finger or pencil clenched between his teeth. The mouth movements were absent at rest and were not activated by eating or non-verbal jaw movements. There were also mild choreic movements of the hands and fingers. After stopping haloperidol there was transient exacerbation of jaw movements which lasted for several weeks followed by persistent speech activated jaw opening oromandibular dystonia, which remained unchanged for the next 10 months. Eye blinking and cervical tics exacerbated when haloperidol treatment was stopped, and were treated with clonazepam with partial benefit. Benzotropine and tetra- benazine were unhelpful for the oromandibular dystonia but beginning 10 months after onset there was slow improvement followed by complete resolution of the jaw dystonia, which has now been absent for 18 months.

Tardive dyskinesia and tardive dystonia have only rarely been reported in patients with Tourette's syndrome after chronic neuroleptic treatment.<sup>1,2</sup> Our patient developed an action induced oromandibular dystonia after 15 years of treatment with haloperidol. Diagnosis of tardive dystonia occurring on a

background of Tourette's syndrome may be difficult. In this case, the patient's dystonia differed from dystonic tics sometimes associated with Tourette's syndrome in being highly localised, action induced, repetitive, patterned, not suppressible, and unaccompanied by an urge to move.<sup>3</sup> His severe dysarthria was also more consistent with oromandibular dystonia than a dystonic jaw tic. Because cranial and cervical dystonia have been reported in a few patients with tic disorders not exposed to neuroleptic drugs,<sup>4,5</sup> the possibility of a coincident primary dystonia unrelated to neuroleptic drugs also deserves consideration. The history of prolonged neuroleptic exposure, exacerbation of dystonia after stopping haloperidol, and disappearance of dystonia over 12 months after stopping haloperidol are clinical features much more indicative of tardive dystonia induced by neuroleptic drugs than spontaneous dystonia.

Although tardive dyskinesia has been uncommon in patients with Tourette's syndrome, vigilance for the appearance of new involuntary movements, especially when they appear dyskinetic or dystonic rather than tic-like, is appropriate in the management of Tourette's syndrome with neuroleptic drugs.

DANIEL TARSY  
EDISON MIYAWAKI  
Division of Neurology, Deaconess Hospital,  
and Department of Neurology,  
Harvard Medical School,  
Boston, MA 02215, USA

Correspondence to: Dr D Tarsy, Division of Neurology, Deaconess Hospital, 110 Francis Street, Boston, MA 02215, USA.

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### Should patients with central core disease be screened for malignant hyperthermia?

Malignant hyperthermia is a potentially fatal condition of hypermetabolism in skeletal muscle triggered by certain anaesthetic agents. Although neuromuscular disorders may predispose to anaesthetic complications in their own right, only central core disease and malignant hyperthermia myopathy are associated with the characteristic in vitro muscle responses diagnostic for susceptibility to malignant hyperthermia.<sup>1</sup> Central core disease is characterised by weakness, wasting, and orthopaedic complications. Muscle fibres show rounded areas of reduced oxidative staining, known as cores. Malignant hyperthermia myopathy is a non-specific histological myopathy, sometimes with cores, occurring in clinically asymptomatic patients, susceptible to malignant hyperthermia. Most susceptible patients show normal muscle histology. A few show central core disease or malignant hyperthermia myopathy. Conversely, many patients with central core disease are susceptible to malignant hyperthermia. Indeed an assumption has grown that all patients with central core disease are susceptible to malignant hyperther-



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D Tarsy and E Miyawaki

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