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# Bilateral abductor vocal fold paralysis due to myasthenia gravis

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**Abstract**
We report a case of bilateral abductor vocal fold paralysis due to myasthenia gravis in a 61-year-old man who presented with stridor requiring tracheostomy. The stridor had been preceded by several weeks' history of diplopia.

**Key words: Myasthenia gravis; Vocal cord paralysis; Stridor**

## Introduction

Bilateral abductor vocal fold paralysis due to myasthenia gravis is a very uncommon cause of stridor. Delay in presentation and diagnosis results in a poor outcome. Recognition of the condition is essential in order to avoid the complication of respiratory infections and failure. We report a case of a 61-year-old man presenting with stridor and diplopia requiring emergency tracheostomy. The probable diagnosis was myasthenia gravis. Other possible causes are discussed.

## Case history

A 61-year-old man presented to the casualty department in February 1997 with stridor and respiratory distress. Fibre-optic nasendoscopy revealed immobile vocal folds in the paramedian position and an emergency tracheostomy was performed. Bronchoscopy and oesophagoscopy were normal.

A comprehensive history taken later revealed a five-week history of progressive dyspnoea and noisy breathing, and several weeks' history of double vision. There was no recent voice change. There was loss of weight and difficulty in micturition. He had been asthmatic and had Parkinson's disease for three years. He had undergone no previous operations.

Examination revealed an ill man with no neck masses. He had cogwheel rigidity and diplopia. A chest radiograph was normal.

On the second day of admission he had not passed urine. He was found to have a painless distended bladder and a normal-sized prostate. Flexible cystoscopy showed no outflow obstruction and the cause was considered to be neurogenic.

At neurological review, the stridor, diplopia and anuria were considered to be related and a diagnosis of myasthenia was made. This was confirmed by a positive edrophonium test performed by a consultant neurologist. Laboratory investigations revealed normal renal, liver and thyroid functions and the absence of acetylocholinesterase receptor, antinuclear, mitochondrial, smooth muscle, reticulin, liver and kidney microsomal antibodies. A magnetic resonance image (MRI) scan of the mediastinum revealed no thymoma.

He was commenced on methylprednisolone 500 mg on alternate days and pyridostigmine 30 mg t.d.s. and discharged for follow-up in both neurology and ENT clinics.

On review two weeks later, he complained of generalized body weakness and his vocal folds remained fixed in the paramedian position. However, six weeks later the vocal folds were mobile and he could breathe comfortably with the tracheostomy tube occluded. The neurologist's opinion was sought regarding decannulation but, on review four weeks later, there was a relapse with progressive weakness in the neck and worsening diplopia. In the weeks that followed he developed a weak voice, repeated chest infections due to aspiration, progressive diplopia and weakness in the neck and limbs. The vocal folds returned to the fixed paramedian position. He died in October 1997 of respiratory failure and septicaemia.

## Discussion

Bilateral abductor vocal fold paralysis is an uncommon emergency presenting with stridor and severe dyspnoea. It may be a life-threatening condition. The causes are diverse: a review of 389 cases by Holinger *et al.* (1976) identified thyroidectomy as the commonest cause (58 per cent), followed by neurological conditions (22 per cent) such as poliomyelitis, Parkinson's disease, cerebrovascular accident, Guillain-Barré syndrome, multiple sclerosis, CNS infections and neoplasms. The remaining 20 per cent were composed of thyroid neoplasms, congenital malformations, trauma and idiopathic causes. They did not identify any cases due to myasthenia gravis. Sommer and Freeman (1994) reported three cases of the condition associated with diabetes mellitus.

Very few cases of bilateral abductor vocal fold paralysis due to myasthenia gravis have been reported in the literature. Calcaterra *et al.* (1972) reported four cases of stridor out of a series of 147 patients with myasthenia gravis. Six other cases have been reported since that series

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(Colp *et al.*, 1980; Foulkes, 1981; Schmidt-Nowara *et al*, 1984; Fairley and Hughes, 1992; Job *et al.*, 1992; Hanson *et al.*, 1996). Tracheostomy was required in each of these cases except that reported by Fairley where it was avoided by prompt diagnosis and treatment. In one case respiratory arrest was precipitated by anaesthesia with succinylcholine in a patient who had received anaesthesia with the same agent in the past (Colp *et al.*, 1980).

Myasthenia gravis is the commonest primary disorder of neuromuscular transmission with a prevalence of approximately eight per 100 000 in Western Europe and a peak incidence in middle-aged men, although it may present in either sex at almost any age. It is an autoimmune disorder resulting from breakdown in T and B cell tolerance to acetylcholine receptors (Steinman and Mantegazza, 1990). The cardinal features are weakness and fatiguability of muscles with use, characterized by exacerbations and remissions. Ten per cent of patients will have a thymic tumour and a further 70 per cent a hyperplastic thymus.

The most common presenting complaints are ptosis and diplopia and in about 10 per cent of patients only the ocular muscles are ever involved. In the remaining cases there is a progressive involvement of the head and neck muscles causing difficulty in chewing and swallowing. Speech may be affected through involvement of muscles of the tongue, soft palate and larynx; nasal escape is the most marked characteristic and tends to deteriorate with prolonged speech and towards the end of the day. Weakness of neck and respiratory muscles and to a lesser extent the muscles of the arms and upper legs is also common. Characteristically the symptoms fluctuate but deteriorate during the course of the day and in response to stress and systemic infections. The combination of a weak cough and aspiration may lead to repeated chest infections.

The diagnosis of myasthenia gravis is made through a combination of clinical suspicion supported by the presence of acetylcholine receptor antibodies, a positive edrophonium test or repetitive nerve stimulation test. A positive acetylcholine receptor antibodies test provides the strongest evidence of myasthenia gravis but in older patients without a thymoma it is often negative. Intravenous edrophonium may cause a transient improvement in neuromuscular transmission in patients with myasthenia gravis by inhibiting acetylcholinesterase, but the test requires the patient to have sufficient signs to show a clear improvement, so is relatively insensitive. The repetitive nerve stimulation test relies on demonstrating a progressive decline in compound muscle action potentials but it is insensitive and not specific to myasthenia gravis. It is most readily performed on patients with facial or upper limb muscle involvement.

The possible treatments for myasthenia gravis include steroids, anticholinergics (e.g. pyridostigmine, neostigmine), immunosuppressants (e.g. azathioprine), plasmaphoresis and thymectomy. The response to anticholinergics is unpredictable but steroids will usually produce considerable symptomatic relief. Immunosuppressants may take up to six months to achieve a significant effect and, like plasmaphoresis, tend to be reserved for refractory cases. Thymectomy will benefit most patients with myasthenia gravis but appears to be less helpful for patients like ours with a late onset of the disease.

In our patient, diplopia, dyspnoea and dysphagia preceded the onset of stridor by several weeks. We believe his vocal fold paralysis was due to myasthenia gravis rather than Parkinson's disease because of the typical symptoms of diplopia, dysphagia, neck muscle weakness and anuria, together with the positive edrophonium test. Treatment of the myasthenia gravis relieved his vocal fold paralysis and other symptoms initially. However, he relapsed and died of respiratory infections and septicaemia before he could be successfully decannulated.

## Conclusion

This case should remind clinicians to consider myasthenia gravis in the differential diagnosis of vocal fold paralysis. This is only possible after a careful history-taking and thorough neurological examination.

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