SPHINCTER ELECTROMYOGRAPHY IN PATIENTS WITH SUSPECTED MULTIPLE SYSTEM ATROPHY

Hypothesis
Multiple system atrophy (MSA) is a progressive neurological disorder characterised by parkinsonism, cerebellar features and autonomic dysfunction. Histologically, the diagnosis is from glial and neuronal inclusions. Clinically, the diagnosis is made using standard criteria but many of these are not met in the early stages of the disease. The neurological symptoms may mimic Idiopathic Parkinson’s disease (IPD) and it is now thought that about 10% of patients with a diagnosis of IPD actually have MSA. It is known that many patients with MSA first present to urologists with lower urinary tract symptoms and/or erectile dysfunction, before the onset of other neurological symptoms (1). The diagnosis of MSA is important since the results of urological surgery in these patients are poor. Urinary symptoms in MSA are best managed by conservative means with anticholinergics, desmopressin and intermittent catheterisation (2).

In MSA degeneration occurs in Onuf’s nucleus and this leads to denervation of the external urethral and anal striated sphincters. Previous investigators have demonstrated by electromyography (EMG) denervation and reinervation in the urethral and anal sphincters of patients with probable MSA. However, the role of sphincter EMG in the diagnosis of MSA remains controversial (3).

A detailed sphincter EMG service was established in our urology department 8 years ago. Patients with suspected MSA are referred by local neurologists for sphincter EMG and urological assessment.

The aim of this study was to assess the usefulness of sphincter EMG in establishing the neurological diagnosis in patients with suspected MSA.

Materials and methods
A retrospective study was performed on 61 patients (24 female, 37 male, mean age 62 years) who had urethral or anal sphincter EMG performed in our department. Concentric needle electromyography of the striated sphincter muscle was performed using a Dantec Counterpoint machine. The duration of at least 10 individual motor units was measured in each patient. The mean motor unit duration and the percentage of motor units greater than 10ms were calculated for each patient. The normal values were taken from published control data and our own control results.

The most recent neurological diagnosis was recorded from the patient’s records together with genitourinary symptoms.

The value of the sphincter EMG test was measured using sensitivity, specificity and predictive values.

Results
Thirty patients had presented initially with neurological symptoms alone, 12 with primarily urological symptoms and 13 with mixed symptoms (limited data on 7). Twenty-five patients complained of urinary incontinence and 30-reported urinary frequency. At the time of the study 25 patients had been diagnosed as having probable MSA and in 36 the diagnosis remained uncertain or was thought not to be MSA. Five of the patients had undergone 6 transurethral prostatectomy (TURP) procedures and in 4 the result was unsatisfactory. The poor results all occurred in patients who had an abnormal sphincter EMG and were later considered to have MSA. One patient with parkinsonism had a normal EMG and subsequently underwent a TURP with a good result.

From control data, the upper limit of duration for a motor unit was taken to be 10ms. In the patients with an eventual diagnosis of probable MSA; the average motor unit duration was 12.5ms average and 59% of the motor units were >10ms. In those patients who were eventually considered not to have MSA; the mean motor unit duration was 6.7ms (Figure 1). We classified patients by the mean motor unit duration <=10ms normal and >10ms abnormal.
The sensitivity of sphincter EMG for predicting the development MSA was 80% and the specificity 83%. The positive predictive value was 77%.

**Figure 1** Sphincter electromyographic findings in patients with parkinsonism and suspected MSA. The majority of patients with a highly abnormal EMG have a final diagnosis of probable MSA. None of the patients with a completely normal EMG were given a diagnosis of MSA.

**Conclusions**
Sphincter EMG was well tolerated and in no patient was the procedure abandoned. Sphincter EMG provides useful information regarding the likelihood of the neurological diagnosis being MSA. Patients with highly abnormal sphincter EMG were all eventually diagnosed as having MSA. Outflow tract surgery should not be undertaken in men with Parkinsonism and a highly abnormal EMG.

Little is known about the value of repeating the sphincter EMG in patients with parkinsonism and an uncertain neurological diagnosis. Evidence of progressing denervation and reinnervation in the external sphincters would be likely to provide strong evidence for a diagnosis of MSA.

Many of the studies of patients with Parkinson’s disease and urinary symptoms are likely to have contained a significant number of patients who actually had MSA. Sphincter EMG may help reduce this error in the future.

**References**
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