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### Trunkal myoclonus with spontaneous priapism and seminal ejaculation in Wilson's disease

We wish to record an uncommon clinical phenomenon seen in a patient with Wilson's disease which to our knowledge has not been previously reported.

A 25 year old unmarried male was admitted in December 1988 for the treatment of tremor and frequent jerkings of the body. He became symptomatic at the age of 14 years with postural tremor of the right hand. Frequent myoclonic jerks of the trunk appeared at the age of 20 years. During the past two years, he experienced priapism with seminal ejaculation up to three and four times per day, associated with some of the myoclonic episodes of the trunk. There were no other symptoms.

Examination revealed bilateral Kayser-Fleischer rings with postural tremors of the upper limbs, myoclonic jerkings of the limbs and trunk and a dystonic gait. No long tract signs were seen and no abnormality was detected in any other system. Examination of the genitalia did not reveal any abnormality.

The diagnosis of Wilson's disease was established by the bilateral Kayser-Fleischer rings and the low serum ceruloplasmin. The ejaculate during priapism was confirmed to be semen by the detection of spermatozoa. The patient was put on d-penicillamine which caused almost total amelioration of his symptoms over a period of six months.

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### Increased amplitude of F-response in Lambert-Eaton myasthenic syndrome

Increased amplitude of the F response may be due to synchronisation of different motor units activated in this response, as in spasticity, or to reinnervated large amplitude single motor units, as in neurogenic disorders.<sup>1</sup> We describe one case of Lambert-Eaton myasthenic syndrome (LEMS) with F amplitude exceeding M amplitude.

A patient with a recurrence of bronchial neoplasia after surgery was admitted to our unit because of diffuse weakness which he had experienced for some weeks. Muscle weakness without amyotrophy was evident in

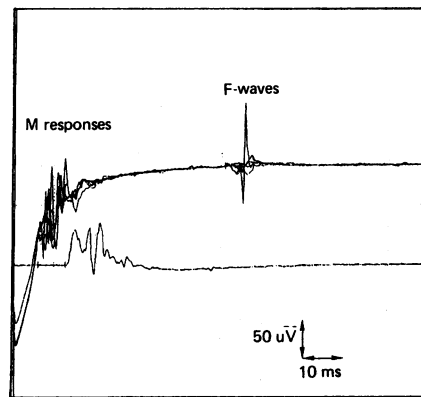


Figure Recording: extensor digitorum brevis. Upper traces: 4 selected traces; stimulation of peroneal nerve at the ankle. Lower trace: stimulation at the peroneal head.

the arms and legs. Facial and ocular motricity were normal and the Babinski sign absent. Sensation was not impaired. Needle electromyography (tibialis anterior, rectus femoris) was performed with concentric needle electrodes. No fibrillations were seen but trigger and delay-line techniques showed fluctuation of amplitude and morphology of individual motor units. Conduction velocities were normal for the sural nerve (47 m/s-15  $\mu$ V) and the peroneal motor conduction velocity was also normal (62 m/s). However, compound muscle action potential amplitude (CMAP) was reduced (100  $\mu$ V) and morphology fluctuated from one stimuli to another in spite of supramaximal stimuli. F waves were recorded at the extensor digitorum brevis by subcutaneous needle electrodes by stimulating the peroneal nerve at the ankle. Stimulation rate was 2/s and more than 20F responses were recorded. Most of them had an amplitude up to 25  $\mu$ V and some were greater than M amplitude (135  $\mu$ V). Minimal F latency was normal for the height (50.9 ms). Repeated stimulation (20 Hz) of the median nerve at the wrist, with recording at the abductor pollicis brevis, was consistent with the diagnosis of LEMS (increment: 532%).

F waves are produced by centrifugal discharges from motoneurons initiated by artificially produced antidromic impulses in the axon by electric stimulation. F-waves studies have proved to be useful in detecting peripheral neuropathies, especially proximal lesions.<sup>1</sup> Some parameters of F waves were studied, including F maximal amplitude, often expressed in per cent of M response (F%). In normal subjects, F is usually lower than 5%.<sup>2,3</sup> Significant increase in the percentage of F response exists in neuropathies of various origin. In chronic spasticity, F amplitude is said to be larger<sup>1</sup> but some studies showed that there was no significant variation but an increased occurrence of F responses.<sup>3</sup>

Nevertheless, in all these cases, F amplitude never exceeded the value of M response, the latter representing the electrical activity of all motor units, the former only part of them. LEMS is a condition in which antibodies directed against calcium channels in the presynaptic nerve terminal membrane are responsive for a decrease in ACh release. At low rates of stimulation, CMAP is reduced in amplitude and shows a decrement in successive responses. At higher rates, usually above 20 Hz, the response becomes strongly in-

cremental.<sup>4</sup> In our patient the interval between the direct stimulation (at the ankle for the peroneal nerve) and the reactivation by the F response was between 45 and 31 ms, estimated by the interval between M and F responses. These intervals correspond to shocks delivered at a frequency between 22 and 32 Hz, which corresponded to the facilitating rate and thus explains the unusually high amplitude of F response.

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### Subarachnoid haemorrhage related to a lumbosacral fusion: a case report

Subarachnoid haemorrhage is a common disorder which is usually caused by the rupture of an aneurysm or arteriovenous malformation. We report an unusual case where the subarachnoid haemorrhage was caused by bleeding into a lumbar pseudomeningocele which developed after lumbosacral fusion.

A 43 year old woman had an L5/S1 discectomy and fusion with a stainless steel (Hartshill) rectangle two years previously. This was complicated by a small dural tear which was apparently repaired at operation with one suture. She made a good post operative recovery and her previous symptoms disappeared. Fourteen months later, while flyfishing, she twisted and immediately developed low back pain and a progressively more severe bilateral sciatica. On examination she had marked neck stiffness, mild pyrexia and bilateral extensor plantars with no other signs. Cerebrospinal fluid (CSF) obtained at lumbar puncture for myelography was blood stained and xanthochromic. Her clotting screen and cranial CT scan were normal. The first myelogram which was done at the referring hospital showed only a hint of a lumbosacral pseudomeningocele. The examination, however, was incomplete. A second myelogram a week later showed a more readily filling lumbosacral pseudomeningocele closely related to the Hartshill rectangle. There was a filling defect on the left side in the pseudomeningocele due to a clot (fig 1b). There were no other abnormalities.

The symptoms persisted for three weeks and the patient had surgical exploration. The cavity of the pseudomeningocele was opened and found to be filled with a blood clot on the left and heavily bloodstained CSF. The anchoring wires of the Hartshill rectangle were engaged into the posterior wall of the