1. **WHAT FURTHER DIAGNOSTIC TEST WAS CARRIED OUT?**

A 68-year-old man presented with a 2-year history of a postural and action tremor of the upper limbs, and latterly unsteadiness. There were no other neurological symptoms, he was otherwise well, on no medication. A maternal aunt may have had a tremor. On examination, he had a postural and action tremor of the upper limbs which looked similar to an essential tremor, he had mild but definite cerebellar ataxia of gait, and absent deep tendon reflexes in his legs. There were no long tract signs, cognitive or eye movement disturbances, or parkinsonian features.

The following tests were all normal or negative: MR brain scan, nerve conduction studies and electromyography, full blood count, ESR, biochemical screen including thyroid function, autoantibody screen, immunoglobulin screen and electrophoresis, and DNA analysis for the known SCA mutations.

*Richard Davenport, Edinburgh*

2. **WHAT IS YOUR DIAGNOSIS?**

A 40-year-old woman presented 3 months after labour with weakness of her arms only. There were no sensory, cranial nerve or other neurological symptoms, and she was otherwise well on no medication, with no family history. Examination revealed dramatic weakness of both biceps, only able to overcome gravity, and milder weakness of wrist and long finger extensors. Nerve conduction studies revealed motor responses (median and ulnar nerves) of normal amplitude and latency, but with an unusual bifid appearance. Sensory studies were normal. Repetitive stimulation revealed a decremental response. Needle EMG studies were normal. Single fibre EMG showed increased jitter with frequent blocking.

Myasthenia gravis was diagnosed, but antibodies to the acetyl choline receptor and muscle specific kinase (MUSK) were negative, her tensilon test was negative, and there was no response to a therapeutic trial of pyridostigmine.

*Richard Davenport, Edinburgh*
3. PLEASE CONSIDER:
A 3-year-old girl presented with frequent episodes of staring, several times every hour, which had started one year previously. These lasted for a few seconds at a time, and consisted of unresponsiveness and subtle eye flutter sometimes accompanied by head nodding. The EEG showed, repeatedly, the same findings (Fig. 1) (distance between vertical lines = 1 second).
(a) What type of seizures was this girl having?
(b) What, if any, investigations would you perform?
(c) What medications would you start?
(d) If the patient is refractory to the first three consecutive medications that you use what further therapeutic and diagnostic measures would you take?

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Fig. 1

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