Letters

progression to incapacitation within 20 years. At least seven different types have been reported in literature,² with subclassification of this disorder according to onset, inheritance pattern and major clinical features. Our patient was considered to have the autosomal recessive adult onset form of limb girdle type muscular dystrophy. The diagnosis was based on the clinical presentation, muscle biopsy, EMG, laboratory studies and the pedigree. Reported associated conditions of the Wolff-Parkinson-White syndrome such as AV blocks, bundle branch blocks, Ebstein anomaly or mitral valve prolapse were looked for, but were not found in our patient.

An abnormal ECG is very often the earliest evidence of myocardial involvement in muscular dystrophies and therefore an accurate parameter of dystrophic heart disease.⁷ Cardiac lesions in limb girdle type muscular dystrophy are rare. Rhythm and conduction disturbances are the most frequent of them, especially atrial tachycardias, atrial flutter, heart blocks, abnormal QRS configuration and abnormal AV conduction.^{6,7} Seldom found features include persistent atrial standstill, severe cardiomyopathy and hypertrophy.^{2,6} Cardiac involvement in muscular dystrophies is progressive and probably associated with the severity of skeletal muscle disease; it may be accompanied with elevation of some serum enzymes like CK-MB (these changes are absent with pure affection of the conduction system). Mechanisms of the rhythm disturbances are still unknown but various explanations have been proposed. Among them are fibrosis,² fatty infiltration and sino and AV nodal arteriopathies.⁵ Early detection of cardiac abnormalities may help to prevent sudden death in limb girdle type muscular dystrophy and should therefore be searched for, followed up and treated. When the Wolff-Parkinson-White syndrome is associated with haemodynamically significant tachyarrhythmias, treatment with a combination of procainamide and verapamil or quinidine and propranolol might be beneficial. If pharmacological treatment fails, pacemaker implantation or surgical ablation of the accessory pathway have been recommended.

The authors thank Dr JR Azpiri and Dr S Ordoñez for kindly supporting the field studies and for providing the technical equipment. We are in debt with Miss Aracely Herrera for her secretarial assistance.

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References

- 1 Perloff KH, Leon AC, O'Doherty D. The cardiomyopathy of progressive muscular dystrophy. *Circulation* 1966;33:625-48.
- 2 Kariv I, Scherf L, Solomon M. Familial cardiomyopathy: with special consideration of electrocardiographic and vectorcardiographic findings. *Am J Cardiol* 1964;13:734-49.
- 3 Perloff JK. Cardiac rhythm and conduction in Duchenne's muscular dystrophy: a prospective study of 20 patients. *J Am Coll Cardiol* 1984;3:1263-8.
- 4 Perloff JK. Neurological disorders and heart disease. In: Braunwald E, ed. *Heart Disease* 2 ed. Philadelphia: WB Saunders Co, 1984:1704-21.
- 5 Beckman R, Schmit B. Das Herz bei Muskel-erkrankungen. *Med Klin* 1976;71:1135-45.
- 6 Weber D, Gould L, Schaffer A. A family with idiopathic myocardial hypertrophy. *Am J Cardiol* 1966;17:419-25.
- 7 Agarwal RK, Misra DN, Varma RK. Wolff-Parkinson-White syndrome with paroxysmal atrial fibrillation in pseudo-hypertrophic muscular dystrophy (Duchenne type). *Indian Heart J* 1978;25:346-8.
- 8 Bensaid H, Marsaud G, Monassier JP, Blanc G. High degree atrioventricular block associated with Wolff-Parkinson-White syndrome during a Duchenne de Boulogne type myopathy. *Ann Cardiol Angeiol (Paris)* 1975;24:67-74.
- 9 Puddu J, Musafia A. L'electrocardiogramme dans la dystrophie musculaire progressive. *Arch Mal Coeur* 1939;32:958-66.
- 10 Shrader W, Pankley G, Davis R, Thelodiges A. Familial idiopathic cardiomegaly. *Circulation* 1961;24:599-600.

Accepted 26 July 1986

Thymoma without myasthenia gravis. Electrophysiological study after thymectomy

Sir: The onset of myasthenia gravis after total extirpation of a thymoma has been reported in the literature.^{1,2} Symptoms and

signs of myasthenia gravis have been observed 2 weeks to 6 years following total removal of thymoma. All the reported patients showed clinical signs and symptoms of fatigability. There appear to be no reports of neuromuscular transmission studies in thymectomized cases that are not followed by clinical myasthenia gravis.

A 46 year old woman with a 7 years history of weight loss of more than 20 kg required admission because of exertional dyspnoea, mild chest discomfort and subjective sensation of tiredness. Radiological examination showed a mass in the anterior mediastinum. Laboratory studies, neurological examination, and clinical test of fatigability were normal. An encapsulated tumour was totally removed. There were no signs of invasion into adjacent structures, or evidence of metastasis. Microscopically, the tumour was a thymoma of the mixed cell type. After thymectomy the patient was asymptomatic, and there was no clinical evidence of myasthenia gravis. Neurological examination was also normal at 3 weeks, 17 months and 2 years after thymectomy.

Repetitive nerve stimulation of the accessory nerve under resting conditions, and repetitive stimulation of the ulnar nerve before and after five minutes of ischaemia on stimulating at 3 Hz^{3,4} did not show significant changes in the amplitude of the compound muscle action potentials. However,

Fig Increased jitter values in seven out of 12 (Top = Three weeks after thymectomy) and in seven out of 20 end-plates evaluated (Bottom = 17 months after thymectomy). Jitter values were calculated manually from superimposed recordings and expressed in μ s as mean consecutive differences of the interpotential interval.⁵ Normal upper limit for EDC muscle in our laboratory is marked by vertical dotted lines. End-plates without (○) and with (●) impulse blocking.

ever, single fibre electromyography (SFEMG)⁵ in extensor digitorum communis muscle (EDC) showed increased jitter and intermittent impulse blocking, without significant differences between both examinations (fig). Thus, a mild to moderate subclinical impairment of neuromuscular transmission was present.

The onset of myasthenia gravis after thymectomy may occur in 0.9 to 2% of all patients with myasthenia gravis and in 10% of patients with thymoma.² Only one out of 140 cases of myasthenia gravis examined by us in the last 8 years developed myasthenic signs and symptoms, 7 weeks after removal of the thymoma.

The electrophysiological findings in our case show that neuromuscular transmission can also be impaired in patients with thymoma but without clinical signs of myasthenia gravis. Moderately increased jitter and impulse blocking were found at 3 weeks and 17 months after thymectomy, whereas signs of clinical fatigability were still absent 2 years after removal of the thymoma. Unfortunately, the anti-acetylcholine receptor antibody (anti-AcChR) levels were not measured and serum for their determination was not available. Thus, the correlation between antibodies level and SFEMG findings is not possible in this case. High anti-AcChR levels have been found in some patients with thymoma but without myasthenia gravis.⁶⁻⁸ These results are in agreement with our findings, although the level of anti-AcChR in the reported patient is not predictable since abnormalities in SFEMG can be found in cases of myasthenia gravis with normal anti-AcChR levels.⁵

Two possible conclusions arise from the present observation. (1) If the patient should develop myasthenia gravis at some future time, the mild SFEMG abnormalities can be attributed to the initial expression of the disease, since it is known that SFEMG is the most precise and earliest method of detecting impairment of neuromuscular transmission.⁵ (2) Should the patient not in due course develop myasthenia gravis it may conclude that an unknown percentage of patients with thymoma may have subclinical impairment of neuromuscular transmission.

We thank Servicios de Medicina Interna and Neurología of Clínica "Puerta de Hierro" (Madrid) which referred the patient for electrophysiological study.

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Addendum

After submission of the paper, a new clinical and electrophysiological examination has been performed 29 months after thymectomy. Clinical test of fatigability was again normal. SFEMG was evaluated in EDC and frontalis muscles. Motor unit fibre density in EDC was normal (1.7). Individual jitter was increased in nine out of 20 endplates in EDC and in four out of 10 endplates in frontalis. Intermittent impulse blocking was found in three (EDC) and two (frontalis) potentials. Thus, mild generalised subclinical neuromuscular transmission impairment was found more than 2 years after removal of the thymoma.

References

- McComas AJ. "Myasthenia gravis preceded by quantal squander miokimia". In: *Neuromuscular Function and Disorders*. London: Butterworths, 1977. Appendix 19-1, pp315-7.
- Namba T, Brunner NG, Grob D. Myasthenia gravis in patients with thymoma with particular reference to onset after thymectomy. *Medicine* 1978;57:411-33.
- Borenstein G, Desmedt JE. "New diagnostic procedures in Myasthenia Gravis". In: Desmedt JE, ed. *New Developments in Electromyography and Clinical Neurophysiology*. Basel Karger, 1973;1:350-74.
- Schumm F, Stöhr M. Accessory nerve stimulation in the assessment of myasthenia gravis. *Muscle Nerve* 1984;7:147-51.
- Stålberg E, Trontelj J. "Single Fibre Electromyography". Old Woking, Surrey: The Miravalle Press, 1979.
- Cuenoud S, Feltkamp TEW, Fulpius BW, Oosterhuis HJGH. "Antibodies to acetylcholine receptor in patients with thymoma but without myasthenia gravis. *Neurology* 1980;30:201-3.
- Limburg PC, The TH, Nummel-Tappel E, Oosterhuis HJGH. "Anti-Acetylcholine receptor antibodies in myasthenia gravis. 1. Relation to clinical parameters in 250 patients". *J Neurol Sci* 1983;58:357-70.
- Gilhus WE, Aarli JA, Janzen RWC, Otto HF, Matre R. "Skeletal muscle antibodies in thymoma patients without myasthenia gravis". *Acta Neurol Scand* 1984;Suppl 98, 69:212-3.

Matters arising

Testing the plantar reflex

Sir: I read with interest the report by Dr Tashiro, in which he described yet another method to produce the upgoing toe sign.¹ Added to those I reviewed some time ago,² this must be the 15th variety, with exclusion of responses that are not even exteroceptive.³ Before others join the game of eponyms⁴ that was so popular at the beginning of this century, I should like to point out there is but a single pathological reflex of the leg. That is when the extensor hallucis longus muscle (inappropriately christened by anatomists, because it acts as a flexor) is released to join the flexion reflex of the leg,^{5,6} as it does in the first year of life.

In some patients almost any stimulation of the skin will evoke the upgoing toe sign, from pricking the sole (as Babinski originally did) to stroking the thigh.⁷ Giving each strip of skin a different name is confusing to neurologists and cruel to students. Babinski himself illustrated the point with the parable of a Spanish nobleman who had eight names and who, on a cold night, was refused accommodation by an inn-keeper who did not realize there was only one person at the other side of the door.⁸ More recently, Denny-Brown was quoted to have replied, on being told that in a particular patient the Oppenheim and Chaddock signs were positive but the Babinski response was negative, "I don't want a Gallup poll; what was the plantar response?"⁹

In addition, there is more to a pathological plantar response than an upgoing toe.¹⁰ First, contraction of the extensor hallucis longus should be synchronous with other components of the flexion synergy of the leg. Secondly, voluntary withdrawal can be confusing but is less constant than the flexion reflex, does not usually involve the tensor fasciae latae muscle, and often precedes or outlasts the stimulus.

I agree with Dr Tashiro that stroking the dorsum rather than the sole of the foot is less likely to produce interference by more or less voluntary reactions. In that case one fails to elicit the normal (downgoing) toe sign, which is a local reflex of the sole,⁵ but the purpose of the examination is to find out whether or not the response is abnormal. For myself I prefer the lateral to the middle part of the dorsum, and a small wooden stick to the handle of a patella hammer. But I