Familial paroxysmal tremor: an essential tremor variant

We read the letter concerning familial paroxysmal tremor by Garcia-Albea et al with considerable interest.1 The authors describe a 24 year old man with a mild 9–10 Hz postural tremor of the upper limbs, in whom intermittent exacerbations of the tremor occurred. The patient’s mother (aged 50) developed essential tremor in her 40s, having previously had a similar paroxysmal arm tremor in late adolescence. The patient had four brothers, two of whom (aged 21 and 30) had mild episodic tremor. The authors knew of only three cases of paroxysmal tremor and considered their patient to have an exceptional presentation of essential tremor.

In fact, the concept of familial paroxysmal tremor is far from new. In 1949, Critchley clearly described episodic tremor in his paper on essential (heredofamilial) tremor and cited Flatau (on page 117) for having suggested the term “intermittent tremor” in such cases.2 Furthermore, Marshall reported in 1962 that in the early stages of essential tremor the amplitude increases “in an episodic fashion, against a background tremor of the same frequency but lower amplitude”.3

The information that we obtained during our recent study of 20 families with hereditary tremor convinced us that this type of tremor typically begins with a feeling of shakiness “inside” which progresses to an intermittent and then persistent tremor.4 Consequently we would consider paroxysmal tremor of the type reported by Garcia-Albea et al to be characteristic of the early natural history of hereditary essential tremor rather than an unusual phenomenon, a view which John Marshall clearly held.5

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Garcia-Albea et al reply: We thank Drs Bain and Findley for their interest in our report on “familial paroxysmal tremor”.1 Their recent study of 20 families with hereditary essential tremor showed that this tremor “typically begins with a feeling of shakiness inside which progresses to an intermittent and then persistent tremor”.2 Many of the 101 patients with essential tremor evaluated in our Movement Disorders Unit reported this type of clinical course (unpublished data). In our opinion, however, the proposed “paroxysmal tremor” has a number of differences from the classic descriptions quoted by Bain et al.3

The term “intermittent tremor” suggested by Flatau4 is defined literally in the report by Critchley5 as follows: “Rarely there may be no trace of tremor while the patient is at rest; alternatively, he may reposit, but as soon as he comes under emotional tension; if he believes he is being watched; or if he concentrates his attention; or it is uncomfortably cold; or if muscular tonus is raised, tremor may appear again.”6 In summary, with the aid of the term “paroxysmal” we tried to define autolimited episodes of tremor lasting from 10 to 60 minutes, that appeared once every three to six weeks, without any apparent precipitating factor, including anxiety.7 Marshall described a subclinical intermittent tremor that consisted of “short bursts of tremor of higher amplitude interspersed among the general background of neurological tremor”.8 The electromyographical record in our patient, showing a synchronous 9–10 Hz tremor during the episodes, seems to be clearly different from the bursts of tremor showed in the figure of a revised chapter by Marshall.9 Out of the episodes, the patient had neither “internal” or “external” tremor or electromyographically registered tremor. In addition, the tremor of the patient’s mother decreased with the passage of time, although she developed, at age 48, a typical essential tremor. This is not the typical evolution of essential tremor suggested by Bain et al.10

Based upon these differences, we think that the proposition expressed in our report is valid.1

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1 Deniliç M, Bajec J. Bilateral tarsal tunnel syndrome. J Neurol Neurosurg Psychiatry 1994;57:239.

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