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Paroxysmal myoclonic dystonia with vocalisations

Sir: Feinberg *et al*[1] described four patients whom they distinguished from the large majority of patients with Gilles de la Tourette syndrome on the basis of four "atypical" characteristics: (1) paroxysmal bursts of regular, repetitive, rhythmic, stereotypic, coordinated, simultaneous and bilateral myoclonus and vocalisations often with tonic symptoms; (2) presence of hyperactivity and attentional and learning disorders; (3) interference of symptoms with voluntary functioning; and (4) ineffectiveness of haloperidol. I have examined Case 1 and have considered him not to have the lightning-like jerks of myoclonus. Rather, he has virtually all the features of Gilles de la Tourette syndrome. I presented videotapes of his paroxysmal bursts of abnormal movements at the Unusual Movement Disorder Seminar held 29 May 1986, at the meeting of the American Academy of Neurology. The audience of 160 neurologists was in complete agreement that this young man suffered from the tic syndrome, commonly known as Gilles de la Tourette syndrome.

Let me address the four features of the disorder that the authors considered atypical. The first are the "paroxysmal bursts of regular, repetitive, rhythmic, stereotypic, coordinated, simultaneous and bilateral abnormal myoclonus (sic) and vocalisations often with tonic symptoms." Paroxysmal bursts of stereotypic and coordinated movements are the hallmark of tics,[2] and, in fact, are not encountered in other movement disorders. Paroxysmal dyskinesias of dystonia and chorea are well recognised,[3] and paroxysmal tremor has been reported,[4] but none of these are coordinated sequences of complex movements that are so typical of tics. Vocalisations, also, are a classical feature of the Gilles de la Tourette syndrome, and are only encountered elsewhere in Meige syndrome and as a feature of akathisia. The vocalisations of Case 1 included coprolalia, which is almost diagnostic for Gilles de la Tourette syndrome. Bursts of repetitive, rhythmic, bilateral movements are not commonly seen as part of the motor tic spectrum, but I see no reason why this phenomenology cannot be included within the realm of motor tics. Indeed, a minority of patients seen by me with otherwise typical features of tics have this feature. Tonic symptoms have long been recognised as a feature of tics,[2 5 6] and today are commonly referred to as dystonic tics.

It is not clear why Feinberg *et al* listed "presence of hyperactivity and attentional and learning disorders" as atypical for patients with tics. Several investigators report that attention deficit disorder occurs in approximately 50% of patients with Gilles de la Tourette syndrome.[7–9]

Interference of voluntary functioning by symptoms does occur in Case 1. When he has a burst of the repetitive, rhythmical flexion movements of his arms, he stops speaking, other than occasional vocalisations. There is no loss of contact with the environment; rather, it appears as if his mind is actively and compulsively engaged in other activity, which is what he and other patients with tics who have this symptom inform me. Like bursts of repetitive movements, these simultaneously mind-occupying states should be considered within the spectrum of tics.

The final point raised by Feinberg *et al* is the ineffectiveness of haloperidol to suppress these paroxysmal motor bursts. But responsivity to medication is not an acceptable criterion for the diagnosis of tics! As an aside, and for a point of information, based on a telephone conversation I had with the patient on 8 May 1986, he considers himself 80% improved on fluphenazine and clonidine. Thus, the former drug, which blocks dopamine, similar to haloperidol, has benefitted the patient.

I would like to propose that authors who wish to describe new or variants of movement disorders should also submit a videotape showing the abnormal movements. The demonstration of the videotape can most easily be accomplished in the newly founded journal *Movement Disorders* which includes a video format in addition to the classical written format. Those interested in this new journal can contact its publisher, Raven Press. I have obtained written permission from Case 1 to publish his videotape, and I will append it as part of the review of tics by Jankovic and Fahn.[6]

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Feinberg et al reply

We are pleased to respond to Dr Fahn's comments on our paper. The fact that Case No 1 was presented at the Academy of Neurology meetings highlights the atypical nature of the symptoms. We also have presented Case No 1, in addition to the three other patients described in our report, including videotapes, to neurologists interested in movement disorders at the University of Michigan Medical School, Mount Sinai School of Medicine and the Basal Ganglia Club in New York City. None of over 150 physicians at any of these meetings opined that the symptoms represented Gilles