Letter to the Editor

Reply to Dr Ravishankar

To the Editor

We appreciate the insightful comments and queries from Dr K Ravishankar regarding our report on 19 patients with the Hypnic Headache Syndrome (1). We also appreciate the opportunity to respond to his questions.

Firstly, each of the patients reported in this study was diagnosed with hypnic headache at the time of the original consultation in the Department of Neurology at the Mayo Clinic. In none of the cases therefore was the diagnosis made in retrospect. The diagnosis was rendered by experienced neurology consultants, with most of the patients in this report having been evaluated by clinicians with a particular interest in headache. The cases were retrieved from the Mayo Clinic database using Hypnic Headache as the key search words. The first author initiated the study after having seen two consecutive patients with this disorder within a 1-year time period. For whatever reason, the cases were not reported previously as case reports or a small series. However, having a larger case series certainly affords one the opportunity to make more meaningful observations.

We certainly agree that these cases and others recently reported in the literature are expanding the clinical spectrum of this disorder, but we feel that this diversity should be embraced as a means to further refine the clinical features of the syndrome rather than be seen as a threat to a simple classification scheme. At the time of this report, only 10 cases had been reported in the literature (5M, 5F), leaving ample space to further expand the clinical spectrum. In fact, since 1997, an additional 31 cases have been reported. Based upon the information available from 25 of these cases, 20/25 were female, 10/25 had unilateral headache, and a number of medications other than lithium have been reported to be effective, including caffeine (1), fluoxetine (2), indomethacin (3), atenolol (1), and cyclobenzaprine (4).

The unsatisfactory response to lithium carbonate in three of four patients in our study does conflict with Professor Raskin's original observation which has been confirmed by others. However, the reasons for this may relate to the retrospective nature of this study and the potential for recall error in patients who had been treated with lithium at some time in the past. It is also possible that side effects precluded an adequate trial duration of the drug or prevented achieving an effective dose. However, the four patients in this study who were exposed to lithium developed intolerable side effects, which promises to be a recurrent problem in the age group commonly affected by this headache syndrome.

We agree with Dr Ravishankar that the time to commencement of the nocturnal headaches can vary between patients. However, there is often considerable consistency within the same individual with headaches recurring near the same time each night. Also, in contrast to cluster headache, hypnic headache occurs exclusively with sleep. For these reasons, we feel that the term “alarm-clock” headache is a more apt description for hypnic rather than cluster headache.

Finally, we agree in principle with Dr Ravishankar's assertion that all rare headache conditions be studied only on a prospective basis. However, careful retrospective analyses (despite the pitfalls) can be valuable in helping to refine the clinical characteristics of a very rare headache disorder such as the hypnic headache syndrome.

Rather than leading to “more dilemmas in confirming the diagnosis”, the 31 cases reported over the last 2 years confirm and highlights Professor Raskin's original observation that the unique feature of this headache disorder is its exclusive occurrence during sleep, hence the term hypnic. Few other headache disorders display such a unique and highly preserved feature. As we have seen with other primary headache disorders, the character of the pain in addition to the intensity, frequency, and duration of painful paroxysms, are more variable both between patients and even within the same patient. Therefore, the intrusion of these headaches only during sleep should be considered as the only absolute criterion when developing classification criteria for this headache disorder.

Rather than hinder our attempts at elucidating the underlying pathophysiology as suggested, we should hopefully be able to exploit the hypnic nature of these headaches to enhance our understanding of the underlying mechanism of hypnic headache specifically, as well as the relationship between sleep and headache in general.

The fact that various treatments have been reported to be effective should be very much
welcomed by both patient and physician. Effective
treatment options expands our therapeutic arsenal,
offers safer and likely better tolerated therapies for
patients, and ultimately may provide further insight
into the pathophysiology of this rare but intriguing
headache disorder.

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DW Dodick, A Mosek, JK Campbell, Mayo Clinic, Mayo Graduate
School of Medicine, Rochester, MN, USA