MATTERS ARISING

Ramsay Hunt syndrome: to bury or to praise

"If there is one disease in the neurological literature that is difficult to define and diagnose, it is the cerebellar dysynchrony of Ramsay Hunt." Radermeker, 1974.

Our recent suggestion that the Ramsay Hunt syndrome (dyssynergia cerebellaris myoclonica) is not a useful diagnostic category has provoked considerable controversy. We reached this conclusion following a study of 84 cases of progressive myoclonus epilepsy (PME), of which 13 were previously regarded as Ramsay Hunt syndrome. Review and restudy of this material established the diagnosis of mitochondrial encephalopathy (MERRF) in 11 of the 13 cases; the remaining two cases were not available for reexamination. Those who wish to preserve the Ramsay Hunt syndrome differ widely in their concept of the disorder, which only reinforces our view that the term should be buried.

Tassinari et al recently reported a series of 13 patients with the syndrome of MERRF who had onset of myoclonic or tonic-clonic seizures at ages 6 to 15 years with a mild cerebellar syndrome. Family studies suggested autosomal dominant inheritance and muscle biopsies failed to show evidence of mitochondrial disease. Tassinari's patients are different from the cases that we reclassified as MERRF and we agree that they do not have mitochondrial disease. The clinical, electroencephalographic and genetic features of their patients are, however, identical to those of Unverricht-Lundborg disease (Baltic myoclonus) as described by the original authors and in subsequent definitive studies of Koskineni. There is no doubt that this disorder occurs outside the Baltic region. Although there is as yet no diagnostic laboratory in Unverricht-Lundborg disease, the clinical picture is distinctive and a clinical diagnosis can be made with considerable certainty.

We find the use of the term "Ramsay Hunt syndrome" for such patients is historically inaccurate and diagnostically misleading.

Tassinari et al have emphasised the electroencephalographic features of their patients with normal or mildly slow waking background activity, fast spike-wave discharges, photosensitivity and lack of activation during slow wave sleep. These findings are not different from those of Unverricht-Lundborg disease. Indeed, critical study of the EEG patterns in all the PMEs, including MERRF and Unverricht-Lundborg disease, reveals more similarities than differences.

Tassinari et al have also described vertex spikes during REM sleep. Unfortunately, REM studies of Unverricht-Lundborg disease have not been reported. These spikes are, however, also seen in MERRF and we suspect they may be common to all the PMEs, much like giant external potentials, which they may in fact represent.

Unlike the situation previously, the vast majority of patients with PME can now be accurately diagnosed during life. Whilst occasional undiagnosed patients remain, there is no residual homogeneous group of cases for which the term Ramsay Hunt syndrome is appropriate. Radermeker's frustration at attempting to define the Ramsay Hunt syndrome can at last be put to rest. In retrospect it was a true syndrome, with many causes, although we suspect that most reported cases were probably examples of MERRF. Its widespread use as a diagnostic category has been overtaken by clinical, genetic, biochemical and pathalogical advances in specific diagnosis.

(We) come to bury, not to praise.

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6 Marsden CD, Obeso JA. The Ramsay Hunt Syndrome is a useful clinical entity. Movement Disorders 1989;4:6-12.


Tassinari et al reply: We thank Drs Berkovic and Andermann for the kind comments they made on our recent paper. We are glad to know that the patients previously referred to as Ramsay Hunt syndrome (RHS) by these authors and subsequently found to have a mitochondrial encephalopathy (MERRF) were significantly different from those we have described under the eponym of RHS. This fact supports our statement that RHS and MERRF are two different clinical, EEG and evolutive features. Drs Berkovic and Andermann however criticize the term RHS as applied to our cases. In these patients the main clinical complaint was action myoclonus combined with rare generalised epileptic seizures: indeed this association was described by Bassi Ramsay under the heading "Dyssynergia Cerebellaris Myoclonica." Ramsay Hunt also emphasized the coexistence of "cerebellar ataxia" but, in such cases, the cerebellar component is difficult to define because of the presence of severe intention myoclonus, as recently pointed out by Harding. Thus, in our opinion, the use of the term RHS for our patients is justified.
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