Reply to Jackson

To the Editor—We appreciate Dr Jackson’s thoughtful comments regarding the diagnostic criteria for encephalitis proposed by the International Encephalitis Consortium (IEC) Working Group [1]. He raises concerns that (1) individual patients may have laboratory or pathologic evidence of encephalitis without meeting the clinical criteria specified in the IEC case definition, and (2) that patients with encephalitis and an additional cause for confusion or obtundation might be inappropriately excluded.

The IEC case definition was developed as a tool for epidemiologic surveillance, research, and outbreak investigation. To identify patients prospectively, we believed that it was important to devise standardized criteria that relied on clinical aspects and initial laboratory testing, exclusive of results of diagnostic testing for specific etiologies. Furthermore, as no etiology is identified in approximately 50% of patients with encephalitis [2], the case definition was specifically formulated to be independent of causality. The case definition was not designed to be comprehensive and, as the article specifies, would not identify patients without altered mentation (eg, those with post-varicella cerebellar ataxia). Moreover, we stand by our emphasis on the importance of clinical manifestations of disease, as a positive test result may well represent a false positive or indicate prior or latent infection, in a patient who does not meet IEC case definition for encephalitis [3–9].

We agree with Dr Jackson that altered mental status is a common presentation for a number of conditions that mimic encephalitis. To maximize specificity, the IEC case definition excludes patients with alternative etiologies (eg, subarachnoid hemorrhage, urosepsis, and bacterial meningitis). This exclusionary criterion is only applicable when there is a clear-cut alternative diagnosis. Patients with depressed mentation and no alternative explanation, including those with metabolic...
derangements who do not normalize with correction of the abnormality, would meet the case definition for encephalitis assuming the requisite number of minor criteria were present.

The IEC encephalitis case definition represents an attempt to standardize recognition of a poorly defined and ambiguous syndrome using evidence-based criteria in the existing literature. The case definition should be viewed as complementary to the diagnostic testing algorithm in the consensus statement and, for individual patients, clinical judgment should be applied. As additional data become available, the proposed case definition may be refined to optimize sensitivity and specificity.

Note

Potential conflicts of interest. All authors: No reported conflicts.

All authors have submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. Conflicts that the editors consider relevant to the content of the manuscript have been disclosed.

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