REVISED DIAGNOSTIC CRITERIA FOR NEUROMYELITIS OPTICA

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Background: The authors previously proposed diagnostic criteria for neuromyelitis optica (NMO) that facilitate its distinction from prototypic multiple sclerosis (MS). However, some patients with otherwise typical NMO have additional symptoms not attributable to optic nerve or spinal cord inflammation or have MS-like brain MRI lesions. Furthermore, some patients are misclassified as NMO by the authors' earlier proposed criteria despite having a subsequent course indistinguishable from prototypic MS. A serum autoantibody marker, NMO-IgG, is highly specific for NMO. The authors propose revised NMO diagnostic criteria that incorporate NMO-IgG status. **Methods:** Using final clinical diagnosis (NMO or MS) as the reference standard, the authors calculated sensitivity and specificity for each criterion and various combinations using a sample of 96 patients with NMO and 33 with MS. The authors used likelihood ratios and logistic regression analysis to develop the most practical and informative diagnostic model. **Results:** Fourteen patients with NMO (14.6%) had extra-optic-spinal CNS symptoms. NMO-IgG seropositivity was 76% sensitive and 94% specific for NMO. The best diagnostic combination was 99% sensitive and 90% specific for NMO and consisted of at least two of three elements: longitudinally extensive cord lesion, onset brain MRI nondiagnostic for MS, or NMO-IgG seropositivity. **Conclusions:** The authors propose revised diagnostic criteria for definite neuromyelitis optica (NMO) that require optic neuritis, myelitis, and at least two of three supportive criteria: MRI evidence of a contiguous spinal cord lesion 3 or more segments in length, onset brain MRI nondiagnostic for multiple sclerosis, or NMO-IgG seropositivity. CNS involvement beyond the optic nerves and spinal cord is compatible with NMO.

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Comment from Richard M. Ransohoff, MD, Associate Editor: This report crystallized the recognition that NMO and MS were pathogenetically distinct, by incorporating a new serum biomarker NMO-IgG in the diagnostic criteria.

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Revised diagnostic criteria for neuromyelitis optica

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