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Neuromyelitis optica.

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Neuromyelitis optica (NMO, Devic's syndrome) is characterized by concurrence of optic neuritis and transverse myelitis, typically associated with a lesion in the spinal cord extending over three or more vertebral segments. It is an inflammatory, demyelinating central nervous system disorder, and although it is most commonly relapsing, it is distinct from multiple sclerosis in that it is more severe, tends to spare the brain, and is associated with a longitudinally extensive lesion on spinal cord MRI. Furthermore, NMO is associated with a highly specific serum autoantibody marker, NMO-IgG, which targets the water channel aquaporin-4. The disease follows a relapsing course in more than 90% of patients. The relapses account for almost all disability associated with the disease because a secondary progressive course is uncommon, in contrast to multiple sclerosis. We recommend intravenous corticosteroids for acute myelitis and optic neuritis relapses, followed quickly by rescue plasmapheresis for severe, progressive, steroid-refractory events. Overwhelming evidence supports humoral autoimmune mechanisms in the pathogenesis of NMO, and most available data suggest that systemic immunosuppression is required to prevent attacks. We recommend long-term treatment with oral agents such as azathioprine or mycophenolate mofetil for patients with relatively mild disease and rituximab for those with more severe, recent attacks or treatment-refractory disease. We also recommend immunosuppression for at least 5 years in NMO-IgG seropositive patients presenting with a first-ever attack of longitudinally extensive transverse myelitis because they are at high risk for relapse or conversion to NMO.

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