

DIAGNOSTIC CRITERIA FOR NMOSD IN ADULTS	
AQP4 <sup>+</sup> NMOSD	
Positive test for AQP4-IgG <sup>a</sup>	
Exclusion of alternate diagnoses	
At least 1 of the following:	
Optic neuritis	
Acute myelitis	
Area postrema syndrome	
Acute brainstem syndrome	
Symptomatic cerebral syndrome w/ NMOSD-typical brain lesion	
Symptomatic narcolepsy OR acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesions.	
AQP4- or AQP4 status unknown NMOSD	
Negative or inconclusive test for AQP4-IgG <sup>a</sup>	
Exclusion of alternative diagnoses	
At least 1 of the following :	
Optic neuritis	
Acute myelitis with LETM	
Area postrema syndrome	
If only 1 of above present, then also 1 of following:	
Acute brainstem syndrome	
Symptomatic cerebral syndrome w/ NMOSD-typical brain lesion	
Symptomatic narcolepsy OR acute diencephalic clinical syndrome WITH NMOSD-typical diencephalic lesions	
PLUS THE FOLLOWING MRI FINDINGS	
Acute optic neuritis	Brain MRI normal or with nonspecific white matter lesions OR optic nerve hyperintense lesion or T1 Gd <sup>+</sup> lesion extending over more than half the optic nerve or involving optic chiasm
Acute myelitis	Associated intramedullary lesion extending over 3 or more contiguous segments (LETM) OR focal spinal cord atrophy over 3 or more contiguous segments and compatible history
Area postrema syndrome	Dorsal medulla/area postrema lesions
Acute brainstem lesion	Periependymal brainstem lesions
Abbreviations: AQP4, aquaporin-4; Gd <sup>+</sup> , gadolinium positive; IgG, immunoglobulin G; LETM, longitudinally extensive transverse myelitis; NMOSD, neuromyelitis optica spectrum disorders. <sup>a</sup> cell-based assay preferred for AQP4 antibody testing	