When bilateral optic neuritis, complete spinal cord syndrome, and/or area postrema clinical syndrome are present,1

CONSIDER **NEUROMYELITIS OPTICA SPECTRUM DISORDER (NMOSD)** AS A POSSIBLE DIAGNOSIS

FOR PATIENTS WITH UNTREATED NMOSD, ATTACKS CAN HAVE <u>DEVASTATING</u> AND <u>PERMANENT</u> CONSEQUENCES²

NMOSD IS A RARE ANTIBODY-MEDIATED INFLAMMATORY CNS DISORDER THAT IS **DISTINCT** FROM MULTIPLE SCLEROSIS (MS)¹⁻⁴

	Suggestive of MS	Suggestive of NMOSD
Pathology		
Anti-AQP4 antibody positive	No	Yes
Optic neuritis	Unilateral; localized	Bilateral; extensive
Myelitis	STM	LETM
Area postrema syndrome	No	Yes
Impact		
Relapse recovery	Better recovery (more likely to return to baseline)	Poorer recovery (less likely to return to baseline)
Relapse-dependent disability	Disability progression outside relapses	Relapses directly lead to cumulative disability
Patient demographics		
Median age of onset	30	40
Female to male ratio	2:1	9:1

AQP4, aquaporin-4; CNS, central nervous system; LETM, longitudinally extensive transverse myelitis; MS, multiple sclerosis; STM, short-segment transverse myeliti

NMOSD can be distinguished from MS by testing for the presence of AQP4 antibodies.1

DIAGNOSTIC CRITERIA FOR NMOSD¹

Clinical presentations suggestive of NMOSD¹ Bilateral optic neuritis Optic chiasm involvement Causes an altitudinal visual field defect OR Causes severe residual visual loss (acuity 20/200 or worse) Complete (vs. partial) spinal cord syndrome Especially with paroxysmal tonic spasms Area postrema syndrome Intractable hiccups OR Nausea and vomiting Clinical judgement remains necessary, as **no one characteristic is** representative of NMOSD.1

(Core clinical characteristics (CCC)
Optic n	neuritis
Acute r	myelitis
	ostrema syndrome: unexplained s or nausea and vomiting
Acute I	orainstem syndrome
Sympto	omatic narcolepsy or acute
	phalic clinical syndrome
with NI MRI les	MOSD-typical diencephalic sions
	omatic cerebral syndrome with D-typical brain lesions

With AQP4-IgG	Without/unknown AQP4-IgG
 1. ≥1 CCC 2. AQP4-IgG positive* 3. Exclusion of alternative diagnoses 	 ≥2 CCC occurring as a result of ≥1 clinical attacks and meeting all the following requirements: a. ≥1 CCC must be optic neuritis, acute myelitis with LETM, or area postrema syndrome b. Dissemination in space (≥2 CCC) c. Fulfillment of additional MRI requirements AQP4-IgG negative Exclusion of alternative diagnoses

Additional MRI requirements for NMOSD without/unknown AQP4-IgG status

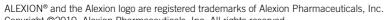
- 1. Acute optic neuritis: requires brain MRI showing:
 - a. Normal findings or only nonspecific white matter lesions, OR
- b. Optic nerve MRI with T2-hyperintense lesion or T1-weighted gadolinium enhancing lesion extending over >1/2 optic nerve length or involving optic chiasm
- 2. Acute myelitis: requires associated intramedullary MRI lesion extending over ≥ 3 contiguous segments (LETM) OR ≥ 3 contiguous segments of focal spinal cord atrophy in patients with history compatible with acute myelitis
- 3. Area postrema syndrome: requires associated dorsal medulla/area postrema lesions
- 4. Acute brainstem syndrome: requires associated periependymal brainstem lesions

IgG, immunoglobulin G; MRI, magnetic resonance imaging

Vigilance and early intervention are key to limit morbidity and mortality from NMOSD.5

References:

1. Wingerchuk DM, et al. Neurology. 2015;85(2):177-189. 2. Jurynczyk M, et al. J Neurol Neurosurg Psychiatry. 2015;86(1):20-25. 3. Kawachi I, et al. J Neurol Neurosurg Psychiatry. 2017;88(2):137-145. 4. Masuda H, et al. J Neurol Sci. 2016;367:375-379. 5. Stavrou M, et al. BMJ Case Rep. 2018.



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