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 DEVASTATING AND PERMANENT CONSEQUENCES[2]

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 myelitis.

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

DIAGNOSTIC CRITERIA FOR NMOSD[1]

Clinical presentations suggestive of NMOSD[1]

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 neuritis
- Acute myelitis
- Area postrema syndrome: unexplained hiccups or nausea and vomiting
- Acute brainstem syndrome
- Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions
- Symptomatic cerebral syndrome with NMOSD-typical brain lesions

With AQP4-IgG

1. ≥1 CCC
2. AQP4-IgG positive*
3. Exclusion of alternative diagnoses

Without/unknown AQP4-IgG

1. ≥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
2. AQP4-IgG negative
3. 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

* Cell-based assay strongly recommended.
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.

CA/SOL-N/0002E