

2015 IPND Neuromyelitis Optica Spectrum Disorder (NMOSD) Diagnostic Criteria

NMOSD with AQP4-IgG

1. At least 1 core clinical characteristic (see reverse)
2. Positive test for AQP4-IgG*
3. Exclusion of alternative diagnoses**

* Using best available detection method (cell-based assay strongly recommended)

** Evaluation for alternative diagnoses guided by "red flags"

NMOSD without AQP4-IgG or Unknown AQP4-IgG Status

1. At least 2 core clinical characteristics (see reverse) resulting from 1 or more clinical attacks and satisfying all of the following requirements:
 - a) At least one of: ON, acute myelitis with LETM, or APS
 - b) Dissemination in space (≥ 2 different core characteristics)
 - c) MRI requirements, if applicable (see below)
2. Negative test(s) for AQP4-IgG* or testing unavailable
3. Exclusion of alternative diagnoses**



Core Clinical Characteristics of NMOSD

Most common:

1. Optic neuritis (ON)
2. Acute myelitis
3. Area postrema syndrome (APS): episode of otherwise unexplained hiccups or nausea and vomiting

Less common:

4. Acute brain stem syndrome
5. Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions
6. Symptomatic cerebral syndrome with NMOSD-typical brain lesions

Supporting MRI Requirements for NMOSD without AQP4-IgG

- 1. Acute optic neuritis:** brain MRI normal or demonstrating only nonspecific white matter lesions; OR 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:** spinal cord MRI showing attack-associated lesion extending ≥ 3 contiguous segments (LETM); OR ≥ 3 contiguous segments of focal cord atrophy in patients with prior history of acute myelitis
- 3. Area postrema syndrome:** dorsal medulla/area postrema MRI lesion
- 4. Acute brain stem syndrome:** peri-ependymal brain stem lesions