Table e-2. Glossary of Clinical Definitions for NMOSD Diagnosis

Acute myelitis: Acute sensory, motor, or sphincter dysfunction in a pattern consistent with a spinal cord lesion with nadir of deficit reached within 3 weeks from onset

Longitudinally extensive myelitis: Acute myelitis associated with a T2 MRI lesion (Figure 1) extending over 3 or more vertebral segments, optimally detected by MRI of the entire spinal cord with gadolinium obtained within one month of clinical onset

Optic neuritis: Acute unilateral or bilateral optic neuropathy (with or without chiasm involvement) with nadir of deficit (worst measured visual acuity) reached within 3 weeks from onset; may be associated with optic nerve or chiasm imaging abnormality on brain or dedicated optic nerve MRI (Figure 1)

Area postrema syndrome: Episode of 1) hiccups or 2) nausea and vomiting; occurring most of each day for at least 7 days, with or without additional neurological symptoms and not attributable to other cause; 2 days duration suffices if associated with new/acute area postrema/dorsal medulla MRI lesion (Figure 2)

Brain stem syndrome: Acute brain stem symptoms (e.g., ocular motor dysfunction, long tract signs, ataxia) with nadir of deficits reached within 3 weeks and associated with area postrema/dorsal medullary or peri-ependymal lesion(s) in brain stem (Figure 2)

Diencephalon (esp. thalamus, hypothalamus, and peri-3rd ventricular region): Hypersomnolence or narcolepsy-like syndrome or other acute diencephalic clinical syndrome (e.g. anorexia with substantial weight loss; hypothermia) not attributable to other cause and associated with diencephalic "NMO-typical" MRI lesion(s) (Figure 3)

Cerebral syndrome: Acute symptoms and signs referable to the cerebrum (e.g., encephalopathy, hemiparesis, cortical visual loss, etc.) with nadir of deficit reached

within 3 weeks from onset and associated with cerebral “NMO-typical” MRI lesions (Figure 3)

Dissemination in space (DIS): Occurrence of two or more discrete clinical attacks in different CNS regions (optic nerve, spinal cord, brain stem, diencephalon, cerebrum). Recurrent, isolated optic neuritis syndromes (even if both optic nerves are involved) and recurrent transverse myelitis syndromes (even if different cord regions are involved) do not establish DIS. Acute myelitis with extension into the brain stem does not establish DIS.