Indexing Metadata/Description

› Title/condition: Neuromyelitis Optica (Devic Syndrome)
› Synonyms: Devic syndrome; Devic’s syndrome; Devic’s disease; neuromyelitis optica spectrum disorder; ophthalmoneuromyelitis
› Anatomical location/body part affected: The optic nerves and the spinal cord are primarily affected, resulting in eye pain and vision loss as well as numbness and paresis of the extremities. The specific location of impairments depends on the location of the spinal cord damage. Other body systems that may be affected by spinal cord damage include bowel and bladder control and respiratory function
› Area(s) of specialty: Acute care, neurological rehabilitation, pediatric rehabilitation
› Description: Neuromyelitis optica (NMO), also known as Devic’s syndrome, is a severe inflammatory demyelinating disorder of the central nervous system (CNS) characterized by optic neuritis and acute transverse myelitis\(^1\) that affects 1 to 2 per 100,000 persons worldwide\(^2\)
  - NMO is an autoimmune disorder with features similar to multiple sclerosis (MS), most notably the occurrence of one or more clinical episodes of optic neuritis in combination with myelitis\(^1\)\(^3\)
  - In NMO, these episodes are usually more acute and severe than in MS
  - At one time NMO was considered a severe form of MS; however, the recent discovery of a disease-specific immunoglobulin G (AQP4-IgG) seems to confirm that it is a separate disease with a distinct etiology\(^1\)\(^4\)
    -- Early diagnosis (differentiation from MS) is considered crucial since early immunosuppressive treatment of NMO is most effective,\(^4\) and some MS treatments have been reported to worsen NMO\(^3\)\(^4\)
  - In 2006, AQP4-IgG serology was introduced into the diagnostic criteria. Diagnosis of NMO required that 2 absolute criteria and at least 2 of 3 supportive criteria be met\(^1\)\(^6\)
    -- Absolute criteria
      - Optic neuritis
      - Acute myelitis
    -- Supportive criteria
      - Negative brain magnetic resonance imaging (MRI) at disease onset
      - Spinal cord MRI with contiguous T2-weighted signal abnormality extending over 3 or more vertebral segments
      - AQP4-IgG seropositive status
  - Further advances have made the 2006 criteria inadequate\(^3\)\(^5\)
    -- An international consensus on diagnostic criteria published in 2015 made use of the high specificity of AQP4-IgG to expand the clinical and neuroimaging spectrum of NMO. By consensus, new nomenclature is now used to refer to the disorder: NMO spectrum disorder (NMOSD)
    -- The new criteria for NMOSD with AQP4-IgG include:
      - At least 1 core clinical characteristic
        - Core clinical characteristics:
          - Optic neuritis
- Acute myelitis
- Area postrema syndrome (i.e., episodes of unexplained hiccups, or nausea and vomiting)
- Acute brainstem syndrome
- Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesions on MRI
- Symptomatic cerebral syndrome with NMOSD-typical brain lesions
- Positive test for AQP4-IgG
- Exclusion of other diagnoses

The new criteria for NMOSD without AQP4-IgG or NMOSD with unknown AQP4-IgG status include:
- At least 2 core clinical characteristics occurring as a result of one or more clinical attacks and meeting the following requirements:
  - At least 1 core clinical characteristic must be optic neuritis, acute myelitis, or area postrema syndrome
  - Dissemination in space (2 or more different core clinical characteristics)
  - Fulfillment of MRI requirements as applicable
    - Acute optic neuritis requires normal brain MRI or optic nerve lesion on MRI
    - Acute myelitis requires intramedullary MRI lesion extending over 3 contiguous segments, or focal spinal cord atrophy
    - Area postrema syndrome requires associated area postrema lesions on MRI
    - Acute brainstem syndrome requires associated brainstem lesions
  - Negative test for AQP4-IgG or testing unavailable
  - Exclusion of alternate diagnoses

These changes allow for an NMOSD diagnosis in patients with involvement of other CNS regions who have not experienced clinical involvement of either optic nerves or spinal cord. The reasons for the changes are:
- In AQP4-IgG seropositive patients, there are no established biological differences between patients with conventional NMO and patients who have not experienced the specified clinical involvement
- NMSOD syndromes affecting CNS regions other than the optic nerve and spinal cord are often precursors to clinical attacks consistent with classical NMO in AQP4-IgG positive patients
- Current immunotherapeutic strategies are the same

• NMO is uncommon in Western populations. Incidence and prevalence are much higher in populations of Asian, Afro-Caribbean, and South American descent(1,5)
• Onset can be at any age, although it most common in childhood and in the fifth decade of life(2)

 › ICD-9 codes: 341.0 Neuromyelitisoptica
 › ICD-10 codes: G36.0 Neuromyelitisoptica (Devic)

(IDC codes are provided for the reader’s reference, not for billing purposes)

• G-Codes
  • Mobility G-code set
    –G8978, Mobility: walking & moving around functional limitation, current status, at therapy episode outset and at reporting intervals
    –G8979, Mobility: walking & moving around functional limitation; projected goal status, at therapy episode outset, at reporting intervals, and at discharge or to end reporting
    –G8980, Mobility: walking & moving around functional limitation, discharge status, at discharge from therapy or to end reporting
  • Changing & Maintaining Body Position G-code set
    –G8981, Changing & maintaining body position functional limitation, current status, at therapy episode outset and at reporting intervals
    –G8982, Changing & maintaining body position functional limitation, projected goal status, at therapy episode outset , at reporting intervals, and at discharge or to end reporting
    –G8983, Changing & maintaining body position functional limitation, discharge status, at discharge from therapy or to end reporting
  • Carrying, Moving & Handling Objects G-code set
    –G8984, Carrying, moving & handling objects functional limitation, current status, at therapy episode outset and at reporting intervals
–G8985, Carrying, moving & handling objects functional limitation, projected goal status, at therapy episode outset, at reporting intervals, and at discharge or to end reporting
–G8986, Carrying, moving & handling objects functional limitation, discharge status, at discharge from therapy or to end reporting

**Self Care G-code set**
–G8987, Self care functional limitation, current status, at therapy episode outset and at reporting intervals
–G8988, Self care functional limitation, projected goal status, at therapy episode outset, at reporting intervals, and at discharge or to end reporting
–G8989, Self care functional limitation, discharge status, at discharge from therapy or to end reporting

**Other PT/OT Primary G-code set**
–G8990, Other physical or occupational primary functional limitation, current status, at therapy episode outset and at reporting intervals
–G8991, Other physical or occupational primary functional limitation, projected goal status, at therapy episode outset, at reporting intervals, and at discharge or to end reporting
–G8992, Other physical or occupational primary functional limitation, discharge status, at discharge from therapy or to end reporting

**Other PT/OT Subsequent G-code set**
–G8993, Other physical or occupational subsequent functional limitation, current status, at therapy episode outset and at reporting intervals
–G8994, Other physical or occupational subsequent functional limitation, projected goal status, at therapy episode outset, at reporting intervals, and at discharge or to end reporting
–G8995, Other physical or occupational subsequent functional limitation, discharge status, at discharge from therapy or to end reporting

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<tr>
<th>G-code Modifier</th>
<th>Impairment Limitation Restriction</th>
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<tr>
<td>CH</td>
<td>0 percent impaired, limited or restricted</td>
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<tr>
<td>CI</td>
<td>At least 1 percent but less than 20 percent impaired, limited or restricted</td>
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<tr>
<td>CJ</td>
<td>At least 20 percent but less than 40 percent impaired, limited or restricted</td>
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<tr>
<td>CK</td>
<td>At least 40 percent but less than 60 percent impaired, limited or restricted</td>
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<td>CL</td>
<td>At least 60 percent but less than 80 percent impaired, limited or restricted</td>
</tr>
<tr>
<td>CM</td>
<td>At least 80 percent but less than 100 percent impaired, limited or restricted</td>
</tr>
<tr>
<td>CN</td>
<td>100 percent impaired, limited or restricted</td>
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Source: http://www.cms.gov

› **Reimbursement:** No specific issues or information regarding reimbursement have been identified

› **Presentation/signs and symptoms:** Attacks tend to be severe and recurrent, often with incomplete recovery. Symptons depend on the site and extent of the lesion and may include:
  - Loss or blurring of vision in one or both eyes
  - Loss of color vision
  - Paralysis or paresis of one or more extremity and trunk
  - Loss of sensation
  - Loss of bladder or bowel control
  - Urinary retention
  - Intractable nausea and vomiting
  - Intractable hiccoughs
• Fatigue
  › Most individuals with NMO have an unpredictable, relapsing disease course with attacks occurring months or years apart
  › Disability is cumulative, the result of each attack damaging new areas of myelin
  • Some individuals are severely affected by NMO and can lose vision in both eyes and the use of their arms and legs
  • Most individuals experience a moderate degree of permanent limb weakness from myelitis

**Causes, Pathogenesis, & Risk Factors**

**Causes**
- Idiopathic
- Immune-mediated disorder with genetic predisposition

**Pathogenesis**
- Unknown
- Appears to be related to B-cell autoimmunity directed against aquaporin-4 (AQP4), an integral cell membrane protein in the CNS
- However, the presence of serum anti-AQP4 antibodies alone is not sufficient to induce a clinical relapse of NMO, suggesting that other factors including inflammatory mediator(s) are required

**Risk factors**
- Variations in incidence and prevalence in different populations suggest a genetic mechanism; however, no specific gene has been identified
  – Familial cases have linked NMO to human leukocyte antigen (HLA) loci
  • Recurrent disease course occurs in women more than in men with a ratio of > 5-10:1
  • Asian, Afro-Caribbean, or South American descent
- Risk factors for relapses include female sex, older age at onset, short interval between attacks, and systemic autoimmunity
- Younger age and equitable sex distribution are associated with a monophasic (nonrelapsing) course

**Overall Contraindications/Precautions**

**Visual loss**
- Low vision increases the risk of falls
- Keep rooms well lit, eliminate clutter, and reposition furniture to make it easier for people with low vision to navigate
- Orthostatic hypotension is a common occurrence in individuals with spinal cord lesions. Monitor for signs and symptoms when performing transfers
- Please see *Clinical Review...Orthostatic Hypotension*. Item number:T908170 for more information on this topic
- Individuals with NMO may experience bladder dysfunction. Catheterization places patients at risk for developing urinary tract infections. Be aware of signs and symptoms such as fever and irritability
- Patients are at an increased risk for developing deep vein thrombosis (DVT) and pulmonary embolism. If the patient has developed DVT, withhold physical therapy for 48 to 72 hours; obtain orders for physician prior to resuming exercise and mobilization
- Pressure ulceration of the skin can occur if pressure relief is not performed at regular intervals. Skin breakdown may occur particularly around the sacral region
- Patients with NMO may require mechanical ventilator assistance depending on the degree of respiratory muscle weakness. These patients are at risk for developing respiratory infections and pneumonia
- Fragile bones
- Long-term steroid therapy can cause osteoporosis
- Autonomic dysreflexia is a potentially life-threatening condition that can occur in patients with spinal cord lesions, especially with lesions above T6. Its occurrence in NMO and transverse myelitis is rare but has been documented
  • The clinician should identify and remove precipitating factors if possible and seek immediate emergency medical care.
  (Please see *Clinical Review...Autonomic Dysreflexia in Adults; Item Number: T708591* for more information on this condition.) Signs and symptoms of autonomic dysreflexia include the following:
  – Elevated blood pressure
  – Pounding headache

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– Bradycardia
– Profuse sweating
– Piloerection (goose bumps), usually above the level of the lesion but sometimes below
– Cardiac arrhythmias, atrial fibrillation, premature ventricular contractions, and arteriovenous conduction abnormalities
– Flushing of the skin, especially in the face, neck, and shoulders; possibly below the level of lesion as well
– Anxiety

› See specific Contraindications/precautions to examination and Contraindications/precautions under Assessment/Plan of Care

Examination

› Contraindications/precautions to examination
  • Stop examination in presence of signs and symptoms of DVT
  • Change positions slowly if orthostatic hypotension is a problem. If symptoms develop, lower the patient to a supine position
  • Be mindful of the potential for significant pain and respect the patient’s complaints
  • Patients are at risk for falls. Arrange the examination environment to accommodate visually impaired individuals and decrease the risk of falls. Keep wheelchair or clinic chair close by when patient is standing
  • Remove precipitating factors and seek emergency medical care if signs or symptoms of autonomic dysreflexia develop during examination

› History
  • History of present illness/injury
    – Mechanism of injury or etiology of illness: What is the reason for the current evaluation/treatment? When did the patient first experience symptoms? Did optic neuritis and transverse myelitis symptoms occur simultaneously? If not, which was first?
      - Optic neuritis and transverse myelitis symptoms can occur at the same time, in rapid succession, or separated by many years(5)
      - What is the level and extent of the patient’s spinal cord lesion?
      - Has the patient had repeated relapses?
      - Most (> 80%) of patients with NMO have repeated relapses(5)
      - A minority have monophasic disease (only experience the initial events)(5)
        - An international consensus panel cautions against making a diagnosis of monophasic disease because criteria that accurately predict a long-term monophasic course cannot be defined. Patients who are AQP4-IgG-seropositive should be assumed to be at risk for relapse indefinitely(35)
      - How has the patient’s disability progressed?
        - Disability is acquired due to relapses. Progressive disability without relapses is rare(5)
        - Deterioration of function has been reported to have a characteristic stepwise progression from motor to sensory to visual to bladder and bowel(6)
        - Most relapses worsen over several days(6)
        - In children, NMO is associated with early recurrence and visual impairment in patients who are AQP4-IgGseropositive, and with physical disability in patients who are seronegative(37)

– Course of treatment
  - Medical management: What treatment has the patient received for this condition? Have there been any complications? Medical management typically consists of medication for treatment and prevention of relapses, symptom control, and rehabilitation
    - Symptom control may include treatment of pain, bowel and bladder symptoms, stiffness, spasms, and spasticity
    - Patients with high cervical spinal cord lesions may need ventilatory support
    - Mobility and visual aids are often needed
    - Patients who do not respond to adequate medication or who relapse rapidly may be treated with plasma exchange
  - Medications for current illness/injury: Determine what medications clinician has prescribed; are they being taken?
    - No drug has been proven to be safe and effective to treat NMO in randomized controlled studies and none has received regulatory approval(38)
Relapses are typically managed with high-dose corticosteroids and supportive care\(^5\). Intravenous methylprednisolone with a gradual taper to oral prednisolone over several months is advocated\(^5\).

Immunosuppression appears to reduce relapse rate\(^5\).

- Most commonly, azathioprine is the immunosuppressive therapy used\(^5\).
- More aggressive immunosuppressive therapy may be used for patients who have “breakthrough” disease while on azathioprine. Rituximab\(^5,22\,26\) and mitoxantrone\(^5,10\) have been reported to be effective. Tocilizumab has been reported to be a promising option in pilot studies\(^20\).
- Interferon beta, which is effective for treatment of MS relapses, does not appear to be effective for NMO\(^5\) and in fact can worsen patients with NMO\(^27\).
- Methotrexate is an alternative to azathioprine that has been reported to reduce relapse frequency, stabilize disability, and be well tolerated even in patients who have not responded to one or more other treatments\(^21\).
- Intravenous immunoglobulin (IVIG) may be used to reduce relapse frequency in patients with NMO\(^39\).

- Document medications being taken for symptom control such as pain medications, antispasticity medications, antidepressants, and antibiotics.

**Diagnostic tests completed:** Usual tests for this condition are the following:
- MRI\(^11\)
  - The most characteristic feature seen is a longitudinally extensive cord lesion
  - Occasionally lesions can be seen in the optic nerves
  - Classically NMO has been defined by a lack of brain lesions on initial diagnosis,\(^12\) although recent research findings suggest that many patients with relapsing NMO do have lesions on brain MRI. Any brain lesions seen are atypical for MS.
- Laboratory tests
  - Cerebrospinal fluid may show disease-specific antibodies
  - Blood tests for AQP4-specific IgG
- Pathology
  - Necrosis, demyelination, and cavitation across multiple spinal cord segments may be found
  - Evoked potentials
  - Visual-evoked potentials and somatosensory evoked potentials may be abnormal\(^28\)
- Optic coherence tomography (OCT)
  - Analysis of retinal damage by OCT may be used to help distinguish between NMO and MS. Optic neuritis typically results in more severe retinal nerve fiber layer and ganglion cell layer thinning and more frequent macular edema in NMO than in MS\(^29\).

**Home remedies/alternative therapies:** Document any use of home remedies (e.g., ice or heating pack) or alternative therapies (e.g., acupuncture) and whether or not they help.

**Previous therapy:** Document whether patient has had occupational or physical therapy for this or other conditions and what specific treatments were helpful or not helpful. Document whether previous therapy included education about this condition.

**Aggravating/easing factors** (and length of time each item is performed before the symptoms come on or are eased): Document factors that affect pain, spasticity, or any other symptoms the patient is experiencing.

**Body chart:** Use body chart to document location and nature of symptoms.
- Based on a cohort study, the location of pain in patients with NMO is:\(^30\)
  - Around chest in 35.5%
  - Around waist in 32.3%
  - Entire legs in 29%
  - Back in 29%

**Nature of symptoms:** Document nature of symptoms patient is experiencing. Symptoms may include pain, spasticity, weakness, vision problems, fatigue, bowel and bladder problems, instability, and sensory loss.
- Symptoms occurring in about 33% of patients with recurrent NMO include radicular pain, paroxysmal tonic spasms, and Lhermitte’s sign (i.e., electric shock-like sensation running down spine with forward flexion of neck)\(^22\).
- Symptoms associated with brainstem lesions include nausea, vomiting, hiccupping, vertigo, and diplopia\(^6\)
- Neuropathic pruritus (itching) may occur\(^{(22)}\)

**Rating of symptoms**: Use a visual analog scale (VAS) or 0-10 scale to assess symptoms at their best, at their worst, and at the moment (specifically address if pain is present now and how much)

**Pattern of symptoms**: Document changes in symptoms throughout the day and night, if any (AM, mid-day, PM, night); also document changes in symptoms due to weather or other external variables

**Sleep disturbance**: Document number of wakings/night
- Nocturia is common and can interfere with sleep

**Other symptoms**: Document other symptoms patient may be experiencing that could exacerbate the condition and/or symptoms that could be indicative of a need to refer to physician
- Using self-rating questionnaires for depressive states, daily activity, and fatigue, investigators in Japan found that depression and fatigue are as prevalent in NMO as in MS and were strongly correlated with one another\(^{(40)}\)

**Respiratory status**
- Is there any history of respiratory compromise? Document any use of oxygen and/or mechanical ventilation
- Lesions higher than C4 can cause paralysis of muscles of inspiration and require long-term mechanical ventilation
- Principal muscles used during inspiration
  - Diaphragm – supplied by the phrenic nerve (C3-C5)
  - Intercostals – supplied by the intercostal nerves in the thoracic region
  - Abdominal muscles (T7-T12) assist in cough and forced expiration
  - Patients with cervical lesions may have weak or absent abdominal muscles so cannot produce an effective cough, leading to sputum retention

**Barriers to learning**
- Are there any barriers to learning? Yes__ No__
- If Yes, describe ___________________________

**Medical history**

**Past medical history**
- Previous history of same/similar diagnosis
  - Some patients have repeated episodes of myelitis before optic neuritis occurs and vice versa, and may receive a diagnosis of relapsing myelitis or relapsing optic neuritis before NMO is diagnosed\(^{(5)}\)
  - In the past, patients may have initially received a diagnosis of MS at a time when NMO was considered to be a severe variant of MS
- **Comorbid diagnoses**: Ask patient about other problems, including diabetes, cancer, heart disease, complications of pregnancy, psychiatric disorders, orthopedic disorders, etc.
  - Approximately 25% of patients with NMO have signs and symptoms of another autoimmune disorder such as myasthenia gravis\(^{(2,13)}\), systemic lupus erythematosus (SLE), or Sjögren syndrome\(^{(2)}\)
- **Medications previously prescribed**: Obtain a comprehensive list of medications prescribed and/or being taken (including over-the-counter drugs)
- **Other symptoms**: Ask patient about other symptoms he or she may be experiencing

**Social/occupational history**

**Patient's goals**: Document what the patient hopes to accomplish with therapy and in general. Individualized goals should be set from initial evaluation. Some individuals will return to walking, whereas others will not

**Vocation/avocation and associated repetitive behaviors, if any**: What is the patient’s occupation? Is the patient currently employed or attending school? What are the required tasks and responsibilities of his or her various roles? What type of leisure or recreational activities does the patient participate in?
- Patients with NMO participating in a semistructured interview study conducted in the United Kingdom reported frustration due to a change in role including limitations of physical activities, inability to complete activities perceived as their duty, driving restrictions, and resulting financial pressures\(^{(22)}\)
  - Learning to juggle their symptoms, particularly fatigue, with their interests and routine chores and duties was an issue for all participants

**Functional limitations/assistance with ADLs/adaptive equipment**: Document limitations caused by visual impairments as well as those caused by sensorimotor impairments. What adaptive equipment does the patient already have, if any? Identify need for any additional assistive devices
Living environment: stairs, number of floors in home, with whom patient lives (e.g., caregivers, family members).
Identify if there are barriers to independence in the home; any modifications necessary? Has a home safety check been performed by a professional?

Relevant tests and measures:(While tests and measures are listed in alphabetical order, sequencing should be appropriate to patient medical condition, functional status, and setting)

- **Arousal, attention, cognition** (including memory, problem solving): Assess as indicated. Cognition is less commonly affected in NMO than in MS
  - Deficits in sustained attention, concentration, speed of information processing, and verbal memory have been reported in patients with NMO, whereas the areas of spatial reasoning and verbal fluency were relatively spared compared to healthy controls\(^{(14)}\)
  - The Mini-Mental State Examination (MMSE) can be used to screen for cognitive impairment

- **Assistive and adaptive devices**
  - Evaluate the need for assistive or adaptive devices (e.g., neck braces for neck muscle weakness, ankle-foot orthoses for dorsiflexor or plantarflexor weakness, wrist splints for wrist and finger extensor weakness, devices for ADLs such as a shower chair for bathing and a commode for toileting)
  - Ensure that assistive and adaptive devices fit and are being used correctly
  - Assess use of assistive devices for ambulation. Addition of seat to walker may be indicated to accommodate onset of fatigue while walking
  - Assess wheelchair use as indicated
  - Consider pressure-relieving mattresses and cushions for patients with impaired mobility

- **Balance**
  - The Berg Balance Scale or the Functional Reach Scale can be used to assess balance in standing
  - Sitting balance should be assessed in patients with cervical lesions
    - Sitting balance strategies vary depending on the level of the lesion
      - If the lesion is at C5 or above, the patient may have limited ability to maintain balance and will need to use equipment to assist
      - The patient with lower cervical and thoracic lesions may use compensatory strategies to sustain posture against gravity, such as upper extremity assistance. This can impede functional use of the arms

- **Cardiorespiratory function and endurance**
  - Monitor vital signs, ECG, and oxygen saturation as appropriate
  - The Borg Rating of Perceived Exertion (RPE) Scale can be used to assess exertion
  - Use the 6-minute walk for distance test (6MWT) to test endurance if appropriate
  - Document amount of chest and rib movement during inspiration and expiration
  - How much accessory muscle use is present during respiration (observe areas of chest and rib movement during inspiration and expiration)?
    - Auscultate for air entry and breath sounds
    - Assess cough for effectiveness and sputum production. Assess color, consistency, amount, and odor of sputum

- **Circulation:** Assess peripheral pulses

- **Cranial/peripheral nerve integrity:** Deficits of cranial nerve II (optic nerve) are a cardinal feature of NMO. Assess vision or obtain results of vision tests performed by other professionals if available
  - Assess visual fields and extraocular movements
  - Assess for facial weakness, facial numbness, ptosis, and nystagmus\(^{(6)}\)

- **Ergonomics/body mechanics:** Observe body position and body mechanics during general mobility. Where applicable, assess wheelchair biomechanics

- **Functional mobility:** Assess bed mobility and transfers as indicated
  - The Timed Up and Go (TUG) test and the 10-meter walk test (10MWT) have not been validated specifically for use with patients with NMO but are commonly used for patients with similar neurological conditions to assess mobility\(^{(15)}\)
    - FIM may be administered

- **Gait/locomotion:** In ambulatory patients, assess gait, noting synchrony of limb movements, gait speed, posture, and mechanics during walking
  - Safety during walking may be assessed using the Dynamic Gait Index (DGI)
  - With non-ambulatory patients, assess locomotion with wheelchair or scooter

- **Joint integrity and mobility:** Assess for contracture development
• **Motor function** (motor control/tone/learning)
  – Assess muscle tone using Modified Ashworth Scale. Flaccid and spastic paralysis may occur.
  – Spinal lesions spanning many vertebral segments can cause extensive flaccid paralysis of the trunk and limbs (31).
  – Assess coordination of bilateral movements where applicable.

• **Muscle strength**: Assess strength in the trunk and upper and lower extremities using manual muscle testing (MMT) if abnormal tone or coordination is not present. A dynamometer can be used to assess hand strength.

• **Neuromotor development**: In children, assess for any delay in acquisition of motor skills where indicated. Potential scales to use include:
  – Pediatric Evaluation of Disability Inventory (PEDI)
  – Bruininks-Oseretsky Test of Motor Proficiency, Second Edition (BOT-2)

• **Observation/inspection/palpation** (including skin assessment)
  – Assess skin integrity.
  – Inspect for muscle atrophy.
  – Observe breathing pattern (diaphragmatic, use of accessory muscles).
  – Palpate for muscle contractures.

• **Posture**: Assess body alignment and observe for postural asymmetry in positioning of the head, shoulders, torso, hips, knees, and feet in sitting and standing, with and without assistive devices, if appropriate. Correlate deviations in posture with abnormal findings on motor function, strength, and flexibility. For wheelchair-bound patients, assess posture in their seating systems.

• **Range of motion**: Assess range of motion (ROM) and flexibility of the involved joints. Note whether contracture or spasticity limit ROM.

• **Reflex testing**: Test deep tendon reflexes.

• **Self-care/activities of daily living** (objective testing): Assess independence in self-care tasks as indicated. The Barthel Index can be used. Will vary with level of lesion. Poor vision, reduced mobility, bladder dysfunction and pain can affect patients’ independence (22).

• **Sensory testing**: Sensation is often impaired. Test light touch, temperature, and proprioception.

• **Special tests specific to diagnosis**
  – Expanded Disability Status Scale (EDSS) can be used.
  – Quality of life questionnaires (e.g., SF-36).

**Assessment/Plan of Care**

› **Contraindications/precautions**
  • Patients with this diagnosis may be at risk for falls; follow facility protocols for fall prevention and post fall prevention instructions at bedside, if inpatient. Ensure that patient and family/caregivers are aware of the potential for falls and educated about fall prevention strategies. Discharge criteria should include independence with fall prevention strategies.
  • Clinicians should follow the guidelines of their clinic/hospital and what is ordered by the patient’s physician. The summary below is meant to serve as a guide, not to replace orders from a physician or a clinic’s specific protocols.

• **Aquatic therapy contraindications** (33)
  – Uncontrolled seizure activity.
  – Unstable medical conditions.
  – Severe cardiac precautions.
  – Acute fever.
  – Infectious diseases (e.g., the flu), upper respiratory infections.
  – Severe pulmonary conditions (e.g., vital capacity < 1 L).
  – Behavior that creates safety concerns.
  – Phobia of water.
  – Open wounds (without proper occlusive dressings).
  – Incontinence.
  – Skin infections.
Aquatic therapy precautions\(^{(33)}\)
- The temperature of the pool should not be > 91.4°F (33°C) for individuals with heat intolerance
- Orthostatic hypotension

Electrotherapy contraindications/precautions\(^{(33)}\)
- Do not place electrodes near
  - Carotid bodies, cardiac pacemakers or implantable cardioverter defibrillators, phrenic nerve or urinary bladder stimulators, phrenic nerve, eyes, gonads
- Osteomyelitis
- Hemorrhage
- Impaired sensation, mental status, communication
- Cardiovascular disease
- Malignancy
- Dermatological conditions
- Proximity of electromagnetic radiation
- In pregnant women, near the pelvis, lumbar spine, hips, abdomen
- In patients with stroke or seizures, near the neck
- History of spontaneous abortion in pregnant women

Diagnosis/need for treatment: NMO with impaired mobility due to sensorimotor deficits; limitations in ROM, strength, and endurance; and functional deficits in posture, balance, and gait

Rule out
- MS – It is often difficult to distinguish between cases of NMO and MS in the early phase of the disease
- Other causes of isolated spinal cord syndrome
  - Compression (e.g., tumor, intervertebral disc)
  - Ischemia or infarction
  - Arteriovenous malformation
  - Infection
- Other causes of optic neuritis
  - Retinal disease
  - Ischemic optic neuritis
  - Inflammatory
  - Infection
- Guillain-Barré syndrome
- Myasthenia gravis

Prognosis
- There is no cure for NMO
- Researchers in the French Caribbean reported 25% mortality after 10 years\(^{(10)}\)
- Mortality rates are thought to have decreased with implementation of more aggressive treatment
- Mortality is most often due to respiratory complications\(^{(8)}\)
  - Respiratory failure occurs more often in relapsing patients, especially those with high cervical lesions
- The progression of disability is directly related to the severity and frequency of relapses\(^{(10,17)}\)
  - In a study conducted in the French Caribbean, disability progressed, on average, to\(^{(10,17)}\)
    - Blindness after two episodes of optic neuritis
    - Paraplegia or quadriplegia after three episodes of myelitis
- Factors predictive of disability include later age of disease onset, high number of attacks during first year, and short interval between first two attacks\(^{(22)}\)
- NMO has a worse prognosis than MS, making early diagnosis critical for prompt initiation of appropriate (immunosuppressive) treatment\(^{(4)}\)
  - Reaching an EDSS score of 6 is considered a hard clinical endpoint of disease severity, with severe disability and impact on social life. Investigators in Brazil found that patients with NMO have a higher risk and a shorter time to reach EDSS 6 compared to patients with MS\(^{(41)}\)
Young-onset patients in the United Kingdom but not in Japan had a high risk of visual disability while older-onset patients had a higher risk of motor disability. Age at disease onset and genetic factors appear to be prognostic factors in determining clinical outcomes.

Presence of Anti-AQP4 antibody is a prognostic marker and indicates a high risk of further relapses of optic neuritis and myelitis.

Patients with severe damage of the spinal cord, shown on MRI, are more likely to have poor recovery, refractory pain, and a high risk of permanent disability. Patients who have lesions in the upper cervical region may be at risk of respiratory failure.

Referral to other disciplines
- Ophthalmologist or neuro-ophthalmologist
- Neurologist
- Social services for long-term care issues, psychosocial adjustment, and equipment and home needs
- Occupational therapy for ADL training and energy-conservation techniques, as well as to address upper extremity weakness and dysfunction
- Orthotist for bracing needs
- Respiratory therapy as indicated
- Speech therapy for dysphagia management if present
- Wheelchair clinic as indicated
- Multidisciplinary coordination is vital to promote adjustment, coping strategies, and support for people living with NMO.

Treatment summary
- Although NMO is now known to be a unique disorder from MS, rehabilitation protocols for MS are commonly applied to NMO.

Researchers in Israel evaluated the effectiveness of a MS type rehabilitation program for patients with NMO by conducting a retrospective chart review of 15 patients with NMO and 32 patients with MS.

Both groups benefitted from the program.

At discharge, the NMO group showed greater improvement in FIM scores and lower EDSS score.

The authors concluded that inpatient multidisciplinary rehabilitation programs available for MS patients may be successfully implemented for patients with NMO.

Strategies used to treat patients with MS that have been applied to patients with NMO include ROM and strengthening exercises, aquatic therapy, and functional training.

Aquatic therapy has been reported to lead to improvements in gait, balance, posture, strength, and functional abilities in patients with NMO.

In a case report of an aquatic therapeutic intervention in a 37-year-old female, the intervention consisted of 8 aquatic therapy sessions focused on balance and gait training.

- Outcome measures included the Berg Balance Scale, the TUG test, the 10MWT, MMT, amount of assistance required for mobility and transfers, and digital pain scale.
- An aquatic therapy program similar to one used for patients with MS was applied.
- Functional weight-bearing exercises performed in the pool allowed exercise with joint-loading forces and instability reduced by the buoyancy.
- The patient responded well to treatment and was able to continue without exacerbations.

In a case report of an outpatient physical therapy intervention for a 75-year old female with T3 paraplegia secondary to NMO, the intervention consisted of task-specific training with gait, balance, and functional skills in both land and aquatic settings.

- Outcome measures included the 6MWT, 10MWT, and Berg Balance Scale.
- The patient underwent outpatient physical therapy for 3 hours, twice per week, for 3 months.
- Improvements in gait and balance translated clinically into increased mobility and decreased fall risk.

Vibration stimulation can improve walking ability in patients with NMO.

- Based on a case report of treatment of a 36-year-old woman with NMO.
- Vibration stimulation was applied to the lower limb muscles of her more spastic side with an ordinary vibrator.
- The performance of standing up and walking improved with vibration stimulation.
– EMG patterns in the lower legs changed markedly after vibration so that there was reciprocity within antagonistic muscles
– The effect lasted about 30 minutes after the cessation of the vibration
• Compensation strategies addressing visual problems include counting steps to the bathroom, making sure that the patients know their environment, and using tactile cues in the environment
– Assistive devices to help with the depth perception and balance issues that accompany vision loss may be prescribed
• Pelvic floor physiotherapy may improve urinary and fecal incontinence and improve quality of life in patients with NMO\(^{(24)}\)
– Based on a case report of an intervention in Brazil
– A 63-year-old woman with partial denervation of the pelvic floor due to NMO was treated with a physiotherapy program that consisted of electrostimulation, biofeedback, abdominal hypopressive technique (a new form of exercise that targets the core without causing increased pressure to the abdominal wall or pelvic floor), and vaginal cones
– Decreased severity of urinary and fecal incontinence and improved quality of life were reported

<table>
<thead>
<tr>
<th>Problem</th>
<th>Goal</th>
<th>Intervention</th>
<th>Expected Progression</th>
<th>Home Program</th>
</tr>
</thead>
<tbody>
<tr>
<td>Decreased strength</td>
<td>Increase strength</td>
<td><strong>Therapeutic exercise</strong></td>
<td>Progress each patient as indicated based on degree of weakness and level of lesion</td>
<td>Provide patient with a home program of resistive exercises</td>
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<tr>
<td></td>
<td></td>
<td>Resistive exercise program</td>
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<td></td>
<td></td>
<td>Aquatic therapy</td>
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<tr>
<td>Impaired balance</td>
<td>Improve balance</td>
<td><strong>Therapeutic exercise</strong></td>
<td>Progress each patient as indicated based on disease progression and level of lesion</td>
<td>Provide patient with a home program of balance exercises and instructions in fall prevention strategies, home modifications</td>
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<tr>
<td>Risk of falls</td>
<td>Decrease risk of falls</td>
<td><strong>Therapeutic exercise</strong></td>
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<td></td>
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<td>Balance training</td>
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<td>Functional training</td>
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<td></td>
<td>Compensatory strategies</td>
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<tr>
<td>Decreased endurance</td>
<td>Improve endurance</td>
<td><strong>Therapeutic exercise</strong></td>
<td>Increase aerobic activity as tolerated</td>
<td>Home-based aerobic activities as indicated</td>
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<tr>
<td></td>
<td></td>
<td>Aerobic exercise and recreational activities</td>
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<tr>
<td>Gait deviations, decreased safety with ambulation</td>
<td>Improve walking efficiency and safety</td>
<td><strong>Gait training</strong></td>
<td>Progress each patient as indicated</td>
<td>Provide patient with instructions for home and community use of ambulatory assistive device and exercises to support gait training</td>
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<td></td>
<td></td>
<td>Individualized gait training for improving mobility and function</td>
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<td><strong>Ambulatory assistive device</strong></td>
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<td></td>
<td>Equipment may be recommended to assist walking efficiency</td>
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<tr>
<td>Functional disability</td>
<td>Improve functional mobility</td>
<td><strong>Functional training</strong> Individualized functional activities for improving mobility and function</td>
<td>Progress each patient as indicated</td>
<td>Provide patient with home program of functional exercises</td>
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<td>Improve ADLs</td>
<td>Prescription of assistive devices</td>
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<tr>
<td>Visual impairment</td>
<td>Decrease functional problems caused by vision loss</td>
<td><strong>Therapeutic modalities</strong> Compensation strategies</td>
<td>Progression varies depending on disease progression</td>
<td>Home modifications may include arrangement of furniture, tactile cues, elimination of clutter, brighter lighting, and education of family members and caregivers</td>
</tr>
<tr>
<td>Pain</td>
<td>Decrease pain</td>
<td><strong>Therapeutic modalities</strong> Aquatic therapy</td>
<td>Progress as indicated by patient’s pain</td>
<td>Home use of hot packs or TENS as indicated</td>
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<td>Transcutaneous electric nerve stimulation (TENS)</td>
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<td>Relaxation techniques</td>
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<td>ROM exercises</td>
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<tr>
<td>Complications due to immobility: respiratory compromise, DVT, pressure ulcers</td>
<td>Improve respiration, decrease risk of pneumonia</td>
<td><strong>Therapeutic exercises</strong> Abdominal muscle training, deep breathing and coughing exercises and techniques</td>
<td>Progress each patient based on level of lesion and degree of weakness</td>
<td>Educate patient and caregiver about respiratory risks and need for deep breathing and coughing and position changes</td>
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<td>Lower extremity ROM, positioning, turning schedule</td>
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</tbody>
</table>

**Desired Outcomes/Outcome Measures**

- Maintained or increased ROM
  - Goniometric measures of ROM
- Increased strength
  - MMT or dynamometry
- Improved balance
  - Berg Balance Scale
Improved endurance
- 6MWT, cycle ergometry testing for endurance
- Decreased risk of falls
- DGI

Improved functional mobility and gait
- FIM, TUG test, Functional Reach Test
- Decreased pain
- VAS

Improved respiration, decreased risk of pneumonia
- Auscultation
- Oxygen saturation
- Spirometry, Pulmonary Function Test results

Reduced disability, improved quality of life
- EDSS
- SF-36

Maintenance or Prevention
- Prevention of relapses through use of immunosuppressive medications is considered crucial for preventing disability
- A home program of ROM and strengthening exercises may help prevent atrophy and contractures
- Education about fall prevention
  - Home adaptations to decrease fall risk

Patient Education

Coding Matrix
References are rated using the following codes, listed in order of strength:

| M | Published meta-analysis |
|SR| Published systematic or integrative literature review |
|RCT| Published research (randomized controlled trial) |
|R| Published research (not randomized controlled trial) |
|C| Case histories, case studies |
|G| Published guidelines |
|RV| Published review of the literature |
|RU| Published research utilization report |
|QI| Published quality improvement report |
|L| Legislation |
|PGR| Published government report |
|PFR| Published funded report |
|PP| Policies, procedures, protocols |
|X| Practice exemplars, stories, opinions |
|GI| General or background information/texts/reports |
|U| Unpublished research, reviews, poster presentations or other such materials |
|CP| Conference proceedings, abstracts, presentation |

References


