CHAPTER 162

Transverse Myelitis

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Synonyms

Transverse myelitis
Acute transverse myelitis
Acute complete transverse myelitis
Acute partial transverse myelitis
Longitudinally extensive transverse myelitis
Idiopathic transverse myelitis
Secondary transverse myelitis

ICD-10-CM Code

G37.3	Acute transverse myelitis in demyelinating disease of central nervous system (Acute transverse myelitis NOS)
TM may also have been coded as:	
G04.0	Acute disseminated encephalitis (Encephalitis and Encephalomyelitis: postimmunization)
G04.8	Other encephalitis, myelitis and encephalomyelitis (Postinfectious encephalitis and encephalomyelitis NOS)
G05.0	Encephalitis, myelitis or encephalomyelitis in bacterial diseases classified elsewhere
G05.1	Encephalitis, myelitis or encephalomyelitis in viral diseases classified elsewhere
G05.2	Encephalitis, myelitis or encephalomyelitis in other infectious and parasitic diseases classified elsewhere
G05.8	Encephalitis, myelitis or encephalomyelitis in other diseases classified elsewhere
G35	Multiple sclerosis
G36.0	Neuromyelitis optica (Devic)
G82.2	Paraplegia, unspecified (Paralysis of both lower limbs NOS, Paraplegia [lower] NOS)
G82.5	Tetraplegia, unspecified (Quadriplegia NOS)

Definition

Transverse myelitis (TM) is a focal inflammation across the spinal cord along one or more levels, in the absence of a compressive lesion. This inflammation can cause damage to the ensheathing nerve cell fiber myelin, with resultant neurological dysfunction including weakness, sensory impairments, and autonomic problems including the bowel and bladder.[1–3] The diagnosis may incorporate the terms *idiopathic*, where no specific bacterial, viral, or other obvious inflammatory cause can be found, or *secondary*, where there is an antecedent or associated disease. Other commonly encountered descriptors include *acute*, *acute partial*, *acute complete*, and *longitudinally extensive*. There are few population-based studies available, and comparative or meta-analysis of the literature is difficult because of the various presentations of TM being reported. It appears, however, that acute TM is rare, with only 1400 new cases annually in the United States, or 1.34 to 4.6 cases per million population per year.[1–5] With availability of improved diagnostic tools, unveiling of the disease over time and longer follow-up periods, the etiology often becomes clearer, including those originally labeled as *idiopathic*. An older study from 1993 in the United States on acute or subacute noncompressive myelopathy reported that 45% of the cases were parainfectious, 21% multiple sclerosis, 12% spinal cord ischemia, and 21% idiopathic.[4] A 2005 French multicenter retrospective study applying the TM Consortium Working Group criteria[1] for acute TM to 288 subjects had a more even spread. Systemic disease (systemic lupus erythematosus, Sjögren syndrome, antiphospholipid syndrome) was implicated in 20.5%, spinal cord infarct in 18.8%, multiple sclerosis 10.8%, infectious or parainfectious 17.3%, neuromyelitis optica 17%, and idiopathic acute TM 15.6%.[6] Another study from France in 2012 on acute partial TM with median follow-up period of 104.8 months reported etiology of the cases as being 62% multiple sclerosis, 1% postinfectious myelitis, 1% neuromyelitis optica, 1% Sjögren syndrome, and 34% undetermined or idiopathic.[7]

There is a female predominance of about 60% to 75%[1,6–11] and a bimodal age distribution in the second and fourth decades. Patients having TM related to multiple sclerosis, post-infectious TM, or idiopathic TM are younger, whereas those with TM related to spinal cord infarcts or delayed radiation effects are older.[2,7,8,10,12] TM may recur, with reported rates ranging from 17.5% to 61%,[10,11] and relapse appears to be more common with acute partial TM.[12]

According to one magnetic resonance imaging (MRI) study, idiopathic acute TM most commonly affects the cervical region (60%), followed by the thoracic region (33%).[6] The onset of TM can be acute (within hours or days) or subacute (between 1 and 4 weeks).[2–4] The period from onset to maximum weakness in idiopathic TM has been reported to range from 10 hours to 28 days, with a mean of 5 days.[13] Subacute presentations, progressing over days to weeks and ascending, are associated with a good to fair prognosis. Acute and catastrophic presentations with back pain have a poorer outcome.[14]

Recovery is often related to the clinical presentation and may or may not be complete. In general, one third of patients with acute TM make a good recovery, another third have fair recovery, and the rest either fail to improve or die.[3,5,13] In idiopathic TM treated with methylprednisolone using the Medical Research Council (MRC) scale for muscle strength, 37.5% were reported to have complete recovery or minimal residual deficit (MRC 5-4), 43% had partial recovery (MRC 3), and 19.4% had severe disability or absent recovery (MRC 0-2). Factors associated with poor outcomes include severe initial symptoms with spinal shock, delayed presentation to the hospital after maximum deficits have already occurred, development of syringomyelia, and extensive MRI lesions.[6,13] If no recovery has occurred by 1 to 3 months, complete recovery is less likely.[4,14]

Symptoms

Patients with TM may present in the ambulatory clinic, urgent care center, or hospital setting with complaints of weakness of the limbs, sensory impairments, pain, and difficulties with the bowel and bladder. Weakness may affect only the lower limbs or all four limbs with varying severity. It may be complete, incomplete, or may present as one of the spinal cord syndromes. The clinical spinal level usually corresponds to the lesion, but lower limb findings do not preclude a lesion at the cervical level. Sensory complaints may include hypersensitivity, numbness, tingling, coldness, burning, or as a circumferential constriction. Pain is a common symptom in one third to one half of patients and may be central or localized, aching or radicular in character. Bowel frequency or constipation may occur, and bladder symptoms include increased frequency, retention, and incontinence.[2,3,13]

The history, including past medical, family, and detailed social, may reveal symptoms of recent infection, immunocompromised or autoimmune condition, space-occupying lesion, demyelinating disease, travel, vaccination, trauma, sexual exposure, animal, insect or tick bites. Whether vaccination triggers TM has been debated. There were only seven cases of TM and eight of acute disseminated encephalomyelitis (ADEM) in the primary vaccination exposure window of 5 to 18 days prior to onset, after 64 million doses within a healthcare network. The incidences were both nonstatistically significant except ADEM with Tdap (tetanus, diphtheria, and pertussis) vaccine at $P = .04$ (translating into 1.16 cases per million doses).[15]

A careful review may yield systemic symptoms, including the upper respiratory tract with cough and difficulty in breathing, chest pain, rashes, joint aches, muscle pain, vision changes, nausea, diarrhea, constipation, and problems with urinary function. Particular attention should be paid to details pointing toward potentially treatable or reversible conditions responsive to antimicrobials or surgical decompression.

There may be a history of invasive spinal intervention for pain management, and TM relating to the infected catheter tip of an intrathecal morphine pump for chronic pain has been reported.[16]

Physical Examination

The physical examination should be broadly systemic as well as focused on neurological findings such as motor weakness, changes in sensation (pinprick, light touch, vibration, position sense, or temperature), tone, muscle stretch reflexes, coordination, and bowel and bladder functioning. Changes affecting the brain, such as cognitive dysfunction and cranial nerve and visual abnormalities, are generally not seen with idiopathic TM.

Fever, tachycardia, and tachypnea may indicate an infectious etiology. Infections, autoimmune, and other conditions that cause acute inflammation of the spinal cord may also manifest in the other body systems. Respiratory, cardiovascular, gastrointestinal, and genitourinary tracts as well as the musculoskeletal and integumentary systems should be assessed accordingly. The findings will assist in determining the level of spinal involvement, guide diagnostic testing, and help rule out other diagnoses.

Functional Limitations

The physiatrist is likely to encounter the patient as a consultation for rehabilitation assessment and management, or referral for a specific problem, such as spasticity or pain intervention. The functional limitations in a patient with TM usually depend on the level of spinal cord involvement and corresponding muscles affected. Debilitation and deconditioning from associated illnesses and prolonged recumbency will also affect function secondarily.

The functional capability review according to spinal level may be influenced by whether the cord injury is unilateral or bilateral and the degree of completeness. High cervical lesions result in tetraplegia with sensory impairment and also affect the phrenic nerve (C3-C5) with diaphragmatic paralysis requiring mechanical ventilation. A patient with C4 innervation preserved may or may not have respiratory difficulties but will be dependent for most self-care activities. Using appropriate technology and devices, whether customized or commercially available, the patient may be able to control the home environment, summon assistance, direct their care, and mobilize in an electric wheelchair with a chin control or a sip-and-puff interface.

A patient with C5 level may be able to self-feed and perform personal grooming with equipment such as a universal cuff for the hand allowing attachment of tools (e.g., fork, spoon, or comb). The patient can independently use a powered wheelchair and propel a lightweight manual wheelchair with hand rim projections (“quad knobs”) for limited distances over level ground. C6 innervation allows independence with upper extremity dressing, bathing with equipment, and functional propulsion of a manual wheelchair indoors. The patient with superior balance and motor control could *theoretically* perform independent or supervised transfers with a sliding board, and self-catheterize with appropriate assistive devices. Driving a specially adapted automatic transmission vehicle with powered steering, hand-controlled accelerator and brake can be achieved with C7-C8 preservation. A C7 level allows independence in all self-care activities with equipment, independent transfers with ability to push off using intact elbow extensor muscles, and the patient may be able to live alone. A patient with C8 and T1 innervation will have improved manual strength and

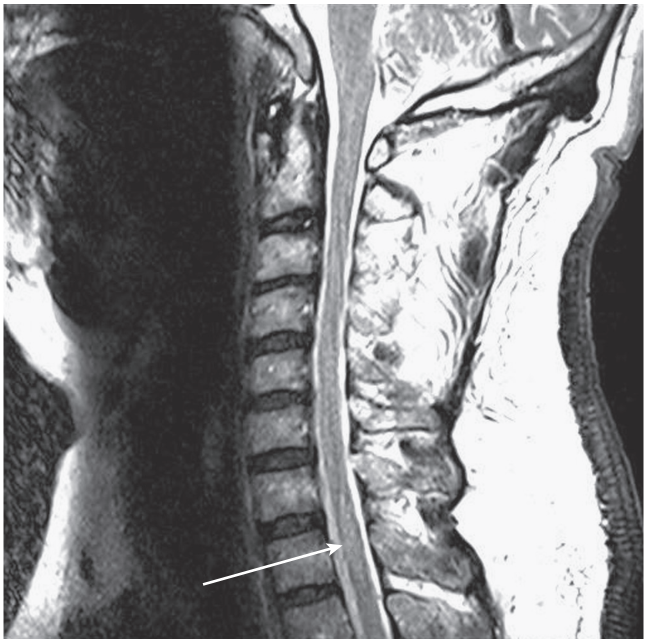

FIG. 162.1 Myelitis: T2-weighted magnetic resonance image of the sagittal cervical spine with fusiform lesion at C7-T1 *(arrow)*.

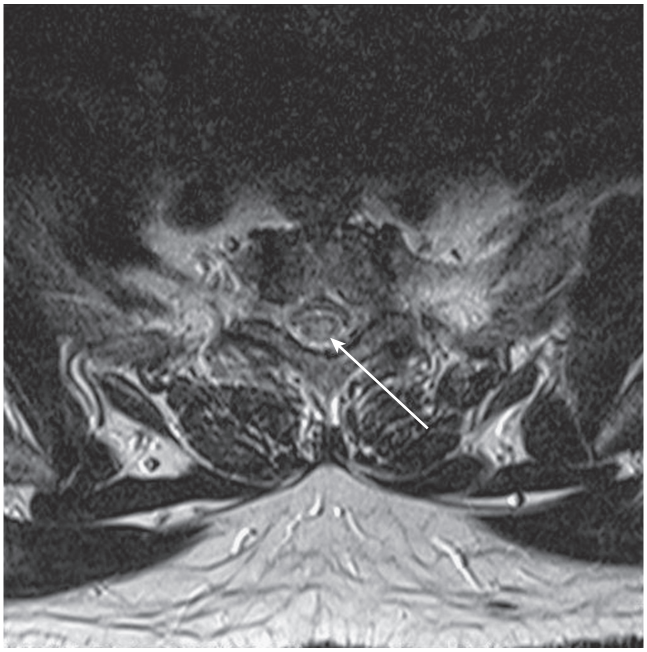

FIG. 162.2 Myelitis: T2-weighted magnetic resonance image of the axial cervical spine showing lesion across most of the spinal cord *(arrow)*.

dexterity for self-care, is independent with a manual wheelchair, and should be able to self-catheterize. Preservation of upper thoracic innervation allows a greater degree of trunk control, increasing stability during use and propulsion of a manual wheelchair. It also adds to ease and independence with bladder and bowel self-management. With bracing of the hips, knees, and ankles (KAFO or knee-ankle-foot orthoses), minimal ambulation can be attempted, although mainly for training and exercise purposes than truly functional. Independent ambulation, even with bracing and bilateral axillary or forearm crutches, is usually not realistic unless the patient has preservation of some upper lumbar innervation. Further preservation of lumbar and sacral innervation will increase ease of ambulation with better trunk and pelvic control. The patient with incomplete spinal injury is less predictable, and functional abilities will largely depend on the degree and nature of neurologic preservation.

Diagnostic Studies

With increasingly greater resolutions and techniques such as T2-weighted fast spin-echo and short-tau inversion recovery (STIR) to enhance or suppress the appearance of fat and tissues of different densities, the best tool when TM is suspected is MRI. MRI not only allows visualization of the lesion but also rules out treatable causes, such as tumor, abscess, and other lesions causing compressive myelopathy. Contrast material can be given to highlight lesions,[17] and myelography may rarely be considered if MRI is not available.

MRI scans show features that help differentiate TM from disorders such as multiple sclerosis (Figs. 162.1 and 162.2). The lesion in TM tends to affect the central region of the cord and involve more than two thirds of the cord diameter, whereas in multiple sclerosis it is usually more peripheral and involves less than half of the cord diameter.[17] TM is more often associated with high signal intensity on T2-weighted images extending longitudinally over more segments.[17,18] The number of segments involved may be from 1 or 2 up to 11, and the entire cord or sometimes only the medulla may be affected.[17,18,20,21] The lesion in TM at times resembles a spinal cord tumor and biopsy may even be attempted during investigation.[17,18,19]

MRI of the brain with contrast enhancement is often performed to help determine whether the MRI findings point toward multiple sclerosis rather than "idiopathic" TM. In idiopathic partial TM, a study that does not show brain lesions translates to the likelihood of evolving multiple sclerosis at 15% to 44%. When brain lesions such as white matter plaques (especially periventricular) are seen, the chance for development of multiple sclerosis increases to 44% to 93%.[22] Asymmetric motor or sensory symptoms and absence of peripheral nervous system involvement at presentation suggest acute myelopathic multiple sclerosis, whereas symmetric symptoms and peripheral nervous system involvement suggest acute TM.[23,24]

Immunoglobulin G antibodies may be useful for determining neuromyelitis optica (Devic's disease) as the etiology in patients with acute complete TM. Longitudinally extensive TM spanning three or more vertebral segments is an important feature and detection of anti-aquaporin 4-specific antibodies (anti-AQP4, AQ4P-Ab, or NMO-IgG) is useful to determine both increased risk for recurrence and conversion to neuromyelitis optica.[13,25]

Other tests include the usual blood counts and chemistry, tests for autoimmune conditions, such as antinuclear antibodies, anti-double-stranded DNA antibodies, anti-Sm antibodies, erythrocyte sedimentation rate, SS-A antibody for Sjögren disease, immunoglobulin levels, and VDRL. Vitamin B_{12} levels may be tested, and *Mycoplasma pneumoniae* or *Mycobacterium tuberculosis* cultures may be performed. Lyme titers and titers for various viruses including human immunodeficiency virus, West Nile virus, poliovirus, hepatitis virus, Epstein-Barr virus, cytomegalovirus, and

Table 162.1 Criteria for the Diagnosis of Idiopathic Acute Transverse Myelitis

Inclusion Criteria	Exclusion Criteria
• Development of sensory, motor, or autonomic dysfunction attributable to the spinal cord • Bilateral signs or symptoms (although not necessarily symmetric) • Clearly defined sensory level • Exclusion of extra-axial compressive etiology by neuroimaging (MRI or myelography; CT of spine not adequate) • Inflammation within the spinal cord demonstrated by CSF pleocytosis or elevated IgG index or gadolinium enhancement. If no inflammatory criterion is met at symptom onset, repeated MRI and lumbar puncture evaluation between 2 and 7 days after symptom onset meet criteria. • Progression to nadir between 4 h and 21 days after the onset of symptoms (if patient awakens with symptoms, symptoms must become more pronounced from point of awakening)	• History of previous radiation to the spine within the last 10 years • Clear arterial distribution clinical deficit consistent with thrombosis of the anterior spinal artery • Abnormal flow voids on the surface of the spinal cord consistent with AVM • Serologic or clinical evidence of connective tissue disease (e.g., sarcoidosis, Behçet disease, Sjögren syndrome, SLE, mixed connective tissue disorder)[a] • CNS manifestations of syphilis, Lyme disease, HIV, HTLV-1, mycoplasma, other viral infection (e.g., HSV-1, HSV-2, VZV, EBV, CMV, HHV-6, enteroviruses)[a] • Brain MRI abnormalities suggestive of multiple sclerosis[a] • History of clinically apparent optic neuritis[a]

[a]Do not exclude disease-associated acute transverse myelitis.
AVM, Arteriovenous malformation; *CMV,* cytomegalovirus; *CNS,* central nervous system; *CSF,* cerebrospinal fluid; *CT,* computed tomography; *EBV,* Epstein-Barr virus; *HHV,* human herpes virus; *HIV,* human immunodeficiency virus; *HSV,* herpes simplex virus; *HTLV-1,* human T-lymphotropic virus 1; *IgG,* immunoglobulin G; *MRI,* magnetic resonance imaging; *SLE,* systemic lupus erythematosus; *VZV,* varicella-zoster virus.
Modified from Transverse Myelitis Consortium Working Group. Proposed diagnostic criteria and nosology of acute transverse myelitis. *Neurology*. 2002;59:499–505.

enteric cytopathic human orphan virus may be elevated. The polymerase chain reaction (PCR) technique is useful for amplifying minute quantities of DNA or RNA. It was used in a recent case report on acute myelitis caused by Zika virus infection, which responded well to high-dose prednisolone.[26]

A lumbar puncture allows assessment of cerebrospinal fluid pressure, and samples for cell count, determination of protein and glucose concentrations, measurement of immunoglobulins, and protein electrophoresis. Oligoclonal bands detected in cerebrospinal fluid are useful in making a diagnosis. In one report, they were present in three of five patients with multiple sclerosis-associated TM, but in none of four patients with parainfectious TM.[4] Nerve conduction studies (NCS), electromyography (EMG), as well as somatosensory and motor evoked potentials may be useful for establishing diagnosis and monitoring progress.[2,27] Urinary system evaluation including cystourethrography, cystoscopy, a baseline renal ultrasound, and urodynamic studies with or without video, have been recommended because of the very high rates of persistent long-term bladder dysfunction.[28,29] Bowel evaluation may require radiography, computed tomography (CT), and MRI scans with or without contrast, or colonoscopy to rule out obstruction. In 2002,

Table 161.2 Comparison of Findings Based on Etiology

Etiology	Findings	Prognosis
Multiple sclerosis	MRI: lesions small, localized in lateral or posterior cord, more cervical CSF: oligoclonal bands	Clinical outcome good but relapse in 47% at mean of 21 months
Systemic disease (SLE, Sjögren syndrome)	Severe motor and sphincter problems MRI in SLE: large and centromedullary lesions CSF: >30 cells	Clinical outcome poor
Spinal cord infarct	No clear diagnostic criteria acutely; >50 years old; severe motor and sphincter problems MRI: isolated centromedullary lesions CSF: absent or low cells, no oligoclonal bands	Outcome poor or fair in 91% of cases
Parainfectious myelopathy	Severe motor and sphincter problems MRI: large centromedullary lesions, cervicodorsal frequently CSF: >30 cells, no oligoclonal bands Serologic confirmation rarely obtained	Clinical outcome good
Delayed radiation myelopathy	History of irradiation; delay can exceed 10 years MRI: high-intensity cord signals with focal swelling, follow-up cord atrophy CSF: normal	Clinical outcome good in early (10–16 weeks after) radiation myelopathy, poor in delayed
Unknown etiology	Long-term follow-up produces diagnosis in 50% of cases	

CSF, Cerebrospinal fluid; *MRI,* magnetic resonance imaging; *SLE,* systemic lupus erythematosus.
From de Seze J, Lanctin C, Lebrun C, et al. Idiopathic acute transverse myelitis: application of the recent diagnostic criteria. *Neurology*. 2005;65:1950–1953.

the TM Consortium Working Group proposed the criteria in Table 162.1 for the diagnosis of idiopathic acute TM.[1]

A comparison by de Seze[8] of the clinical findings, MRI results, laboratory profiles, and outcomes of patients with acute myelopathy according to etiology is presented in Table 162.2.

Beh et al.[2] have comprehensively listed the different causes of TM as in Table 162.3.

Treatment

Initial

Although the physiatrist may manage stable long-standing TM on an outpatient basis, hospitalization may be necessary during the initial presentation to monitor vital signs, manage respiratory difficulties, bowel or bladder complications, and carry out diagnostic investigations.[13,28] Abnormalities

Table 162.3 Summary of Reported Causes of Transverse Myelitis

1. Acquired demyelinating disorders
   a. Multiple sclerosis
   b. NMO
   c. ADEM
2. Systemic inflammatory autoimmune disorders
   a. SLE
   b. SS
   c. Antiphospholipid syndrome
   d. Behçet disease
   e. Vogt-Koyanagi Harada disease
   f. Ankylosing spondylitis
   g. Mixed connective tissue disease
   h. Others: systemic sclerosis, anti-Jo-1 antibody, urticarial vasculitis, psoriatic arthritis, perinuclear ANCA systemic vasculitis, graft-versus-host disease, common variable immunodeficiency, celiac disease
3. Neurosarcoidosis
4. Parainfectious TM
   a. Viral: hepatitis A, hepatitis B, hepatitis C, hepatitis E, measles, mumps, rubella, varicella zoster, Epstein-Barr, cytomegalovirus, herpes simplex, influenza A/B, lymphocytic choriomeningitis virus, chikungunya, Hanta virus, HIV, human T-cell lymphotropic virus, human herpes virus 6, Japanese encephalitis, Murray Valley encephalitis, St. Louis encephalitis, tick-borne encephalitis, vaccnia. Rocky Mountain spotted fever, dengue virus, enterovirus 71, coxsackievirus A and B, West Nile virus, parvovirus B19, human corona virus, and echovirus
   b. Bacterial: *Mycoplasma pneumoniae, Campylobacter jejuni, Borrelia burgdorferi, Acinetobacter baumanii, Coxiella burnetii, Bartonella henselae, Chlamydia psittaci, Leptospira, Chlamydia pneumoniae. Legionella pneumonia, Orientia tsutsugamushi* (scrub typhus). *Salmonella* paratyphi B, *Mycobacterium tuberculosis, Treponema pallidum, Brucellosis melitensis,* and groups A and B strep to cocci
   c. Fungal: *Actinomyces, Blastomyces, Coccidioides, Aspergillus, Cryptococcus,* and *Cladophialophora bantiana*
   d. Parasitic: *Toxocara* species. *Schistosoma* species, *Gnasthostoma spinigerum, Echinococcus granulosus, Taenia solium, Toxoplasma gondii, Acanthamoeba* species, *Paragonimus westermani,* and *Trypanosoma brucei*
5. Paraneoplastic syndromes
   a. Anti-Ri (ANNA-2) antibody
   b. CRMP-5-IgG antibody
   c. Anti-amphiphysin IgG antibody
   d. Anti-GAD65 antibody
   e. NMDAR antibody
6. Atopic myelitis
7. Drugs and toxins
   a. Tumor necrosis factor-alpha inhibitors

ADEM, Acute disseminated encephalomyelitis; *SLE*, systemic lupus erythematosus; *TM*, transverse myelitis.
From Beh SC, Greenberg BM, Frohman T, Frohman EM. Transverse myelitis. *Neurol Clin*. 2013;31(1):79–138.

of vital signs such as tachypnea or tachycardia may suggest impaired oxygenation or blood flow to be managed urgently. The ability to provide antiviral or antibacterial agents and surgical intervention may also be critical when a specific cause has been identified.

Several anti-inflammatory drugs have been tried for TM without clear success. Although there is insufficient evidence for corticosteroid efficacy, intravenous methylprednisolone is often used to prevent further damage to the spinal cord as a result of swelling.[12,20,21] During the acute phase, it may lead to faster recovery and less disability, and is well tolerated.[22] Cyclophosphamide exerts an immunosuppressive and immunomodulatory effect through suppression of cell-mediated and humoral immunity (on the T cells and B cells).[22] Cyclophosphamide together with methylprednisolone may help in lupus-related TM.[21,30] However, there appears to be an absence of any beneficial effect of immunosuppressive drugs (cyclophosphamide, azathioprine, intravenous immune globulin) in patients with idiopathic acute TM.[6,12] Plasma exchange to remove autoreactive antibodies and other toxic molecules from plasma may be effective with a good clinical response, especially within 20 days of onset and when nonresponsive to high-dose corticosteroids.[12,21] The monoclonal antibody rituximab can be effective in decreasing relapses in TM due to NMO.[12]

Rehabilitation

The management of any spinal cord injury will include rehabilitation, and the more severely affected cases of TM will require a comprehensive multidisciplinary rehabilitation program led by a physiatrist. Physical and occupational therapists on the team can work with strengthening, endurance, balance, coordination, joint range of motion, reconditioning, mobility, and independence with activities of daily living. The goal is one of optimal functioning and independence in the activities of daily living and mobility for the patient. Functional Independence Measure and the Modified Barthel Index are among the more widely used outcome measures during the rehabilitation process.

Assessment for appropriate equipment, such as a well-fitting wheelchair and other assistive and walking devices, is needed. Gait efficiency, stability, and overall mobility can be improved with bracing devices such as an ankle-foot orthosis or KAFO. Education of the patient and family about the disease, resultant impairments, potential complications, rehabilitation plans and prognosis is important. The psychological state of the patient should not be neglected, and there should be monitoring for depression and medications initiated if necessary. Sexual functioning is often affected, and education and counseling, with or without intervention, may be appropriate early. Discharge planning needs and issues potentially affecting the patient's community reintegration, including vocational and recreational, should be assessed.

Recovery to some extent is expected in TM, but it is important to minimize the effects of even temporary impairments and immobility. All muscles and joints should be kept as active as possible, and daily exercises to preserve range of motion of the joints will help prevent contractures and keep joints flexible. Progressive resistive exercises and possibly functional electrical stimulation (FES), also known as neuromuscular electrical stimulation, help maintain strength and decrease muscle atrophy.[31] Exercises for inspiratory muscles should be included and use of an incentive spirometer as needed. Rarely, glossopharyngeal breathing may need to be taught and electrical stimulation of the phrenic nerve diaphragm considered in the high cervical cord patient not showing recovery.[32] Spasticity (see Chapter 154) is a possible complication and regular stretching with use of antispasticity medications such as baclofen, diazepam, gabapentin, and tizanidine, can minimize and decrease development of joint contractures. If pain is present, appropriate medications, thermal (heat, cold), and electrical modalities including transcutaneous electrical stimulation may be helpful. Antiepileptic drugs such as gabapentin, pregabalin, and carbamazepine, may be prescribed as they have good efficacy for neuropathic pain. Amitriptyline may also be useful, although caution is advised with its strong anticholinergic effects. Thorough checks of the skin on a daily basis can help avoid pressure sores and associated infections. Insensate areas, particularly over bony prominences, should be relieved with special cushions and mattresses such as egg-crate foam and alternating pressure overlays, and pressure-relieving ankle-foot orthoses (PRAFO) may be helpful. The many varieties of hydrophilic and antimicrobial wound dressings currently available promote faster healing of skin breakdown.

Bladder (see Chapter 138) and bowel (see Chapter 139) functioning should be assessed, and a bedside ultrasound for post-void residual urine volume is a simple informative procedure, as is a rectal examination. A program may be needed to avert a neglected neurogenic bowel or bladder leading to stool impaction and hydroureter or hydronephrosis. An indwelling catheter can initially be used for bladder drainage but intermittent catheterization, independently or otherwise, should be instituted whenever possible. Long-term follow-up of 2 to 10 years in pediatric patients with TM has shown that residual bladder dysfunction is common even with improvement of paraparesis and lack of urologic symptoms. In one study, 86% had persistent bladder dysfunction and 77% had persistent bowel dysfunction.[28,29]

A bowel program includes adequate fluids, proper diet, activity, and scheduled bowel movements. Upper motor neuron bowels may need a stool softener (e.g., docusate), osmotic laxative (lactulose), or stimulant laxative (senna or bisacodyl) for evacuation. Digital stimulation of the rectum is often effective and needs to be taught. With areflexic lower motor neuron bowels, use of bulk laxatives like psyllium or methylcellulose to produce formed stools may help during digital manual evacuation. Bowel evacuation is often done on a daily basis in the hospital, but frequency can be extended to every 2 or 3 days once an individual returns home. The patient requiring a wheelchair, walker, crutches, or cane will need training, including maneuvering over steps and curbs. If transfers and ambulation require assistance, the training should also include family members or assistants.

For patients with TM at the cervical level especially, various types of equipment and orthoses can be provided to help with self-care activities. Proper bathroom equipment and modifications, such as a tub bench, commode, hand-held shower, raised toilet seat, and grab bars, may make the difference between dependence and independence. Selection of appropriate assistive devices helps maximize function. Some of this equipment can be fairly expensive; hence, timing of purchases must be carefully considered, as they may not be required soon after. Despite a reasonable prognosis for eventual recovery, complacency is to be avoided as it may result in unnecessary secondary complications.

Procedures

Renal ultrasound and urodynamic evaluations are relatively routine procedures to assess and monitor bladder dysfunction. Electrodiagnosis including NCS and EMG are useful for diagnosis and for monitoring recovery. Intramuscular botulinum toxin injections are very effective in the management of spasticity and commonly performed by the physiatrist, as are the alternatives of alcohol or phenol nerve and motor point blocks for spastic limb muscles. An intrathecal baclofen pump may be effective in intractable cases and allows much smaller doses and concomitantly fewer side effects. Many physiatrists are able to manage the settings and refilling of these pumps. Intractable neuropathic pain may respond to an intrathecal morphine pump, which will also require management.[33]

Technology

The field of rehabilitation uses a plethora of devices and technology during the process of restoring or compensating for the impairments and disabilities resulting from conditions such as TM. Some individuals receive FES systems to help maintain fitness and muscle bulk or improve and restore function. FES for the forearm and arm muscles is a routinely employed technique with many devices commercially available. Exercise bicycles for the lower as well as upper limbs (e.g., Ergys3 [Therapeutic Alliances, Inc.], RT300-S [Restorative Therapies, Inc.]) have also long been used, although are not cheap and have a risk for osteoporotic fractures. From simple body weight-support suspension devices, stationary and mobile, allowing for safer ambulation training, to motorized treadmills allowing flexibility in intensity, velocity, and effort, multiple devices from various manufacturers are available.

Robotic assisted gait training devices (e.g., Lokomat [Hocoma Inc.], G-EO System [Reha Technology AG], Gait Trainer GT1 [Reha-Stim Medtec GmbH], Haptic Walker [Fraunhofer-Gesellschaft]) are becoming popular and increasingly promising, although definitive evidence of universal efficacy and applicability is pending.[34,35] They allow severely impaired patients to receive intensive training sessions and reduce therapist sprains, strains, and other injuries. Similar devices are available for upper limb training (e.g., InMotion ARM [Bionik Labs] and Armeo [Hocoma Inc.]).

Neuroprostheses using FES to increase forearm reach, hand grasp, and opening (e.g., H200 [Bioness Inc.]) or compensate for thigh weakness and foot drop (e.g., Para-Step I [Sigmedics, Inc.], L300/L300Plus [Bioness Inc.], WalkAide [Innovative Neurotronics Inc.]) are also available. Less frequently encountered in the United States but more in Europe, the anterior sacral root stimulation Finetech-Brindley device can be very effective for bladder management (Vocare, NeuroControl/NDI Medical). Other devices include implanted electrodes for phrenic nerve stimulation/pacing (Avery Biomedical Devices Inc.) when respiratory muscles have not recovered.

Wheelchairs are ubiquitous equipment, available as either manual, powered, or hybrid, with an almost infinite offering of choices for size, weight, purpose, and even color. Control of the wheelchair can be achieved by hand, chin, or other head part, and by voice activation. Other than locomotion, there are also wheelchairs available for standing purposes, whether for activities at an erect level, or for weight-bearing exercise.

Braces or orthotics have also undergone much development and come with different materials, rigidity or flexibility, weight, and functional goals including for support, pressure relief, positioning, or protection. Powered exoskeleton systems are currently of interest with systems to assist standing and ambulation such as the ReWalk 6.0 (ReWalk Robotics Inc.), Hybrid Assistive Limb or HAL (Cyberdyne Inc.), REX (Rex Bionics), Ekso GT (Ekso Bionics), and Indego (Parker Hannifin Corp). At this time, they are mainly for training and exercise, and limited by the individual's abilities, terrain, device battery, and need for safety supervision including skin breakdown, falls, and equipment failure.[36] The next wave for independent mobility in patients with handicaps could well be that of self-driving or autonomous cars undergoing trials by the major automobile companies and various research laboratories.

Environmental control units or multiple devices within a s*mart home* controlled using simple touch-pad, infrared or motion-sensitive, and voice-activated mechanisms including automatic doors, curtains, and various electronics such as the television and personal computer, are now commercially available, easy to control, and importantly becoming increasingly affordable.

Apps (applications) that allow easy communication, videophone interactions, and ready access to the Internet are already built-in for many smart phones. Intelligent voice-controlled personal assistants include the Apple Siri, Google Assistant, Amazon Alexa, Microsoft Cortana, and Samsung Bixby.

Surgery

There is no specific curative surgical procedure for TM. However, lesions such as abscesses, herniated disks, spinal stenosis, and tumors may need surgery as soon as possible to relieve pressure and prevent further damage to the cord. Timely management of compressive lesions may reverse or at least halt further neurologic injury to the cord. Pressure sores may require sharp débridement on the unit to remove dead or infected tissue and other debris to accelerate healing.

Tendon transfers may be considered at a later stage to increase an individual's functioning. Nerve transfer in patients with permanent upper limb deficits may be considered to restore or to improve ability to voluntarily activate a muscle. In one case report, a child with TM who underwent multiple fascicle transfers from median and ulnar nerves to the musculocutaneous nerve, spinal accessory to suprascapular nerve, and medial cord to axillary nerve, had excellent recovery of elbow flexion.[37]

Potential Disease Complications

Potential complications from the spinal cord dysfunction of TM are numerous and may require medical or surgical intervention. They include orthostatic hypotension, impaired thermoregulation, autonomic dysreflexia, lung and urinary tract infections, ileus and constipation, electrolyte imbalances, skin breakdown, spasticity and contractures, musculoskeletal and neuropathic pain, injury (including fractures) to bones, muscles and joints due to sensory impairments, heterotopic ossification, osteoporosis, kidney stones, depression, and anxiety. There may be respiratory muscle weakness depending on the level of spinal cord involved, and when severe, mechanical ventilation assistance may be required. The risk of bronchopneumonia and sleep apnea is compounded by any sedating medications or respiration-depressing medications.

Spasticity and joint contractures are common complications with spinal cord injury and management may be straightforward or extremely difficult, requiring several interventions simultaneously. Heterotopic ossification (see Chapter 131) may develop around a joint, especially the elbow, knee, and hip. Gastrointestinal complications include gaseous distension, regurgitation, indigestion, and chronic constipation. Urinary tract infections and urosepsis are also common with a neurogenic bladder, as both retained urine and bladder instrumentation increase infection risk. Autonomic dysreflexia/hyperreflexia may occur, especially for lesions above T6. Pain is a very frequent complaint and may arise from musculoskeletal sources or be neuropathic in nature. Pain management may include medications such as analgesics, nonsteroidal anti-inflammatory drugs, short courses of cyclooxygenase-2 inhibitors, various anticonvulsants, and tricyclic antidepressants.

Overuse syndromes often occur because muscles and joints are overstressed during functional compensation for weakness or even during the process of rehabilitation training. Shoulder pain is a common phenomenon with causes including tendinitis, rotator cuff injury, impingement syndromes, contractures, and inflammatory or degenerative arthritis. Steroid and local anesthetic injections in the joint may sometimes be needed, but topical anti-inflammatory drugs, heat, cold, and other modalities, with proper transfer techniques or specific adaptive equipment such as sliding board, are often helpful. A common complication is ischemic breakdown of the skin if pressure relief is not regularly performed. Awareness and monitoring for deep venous thrombosis and pulmonary embolism should be routine. Prolonged pressure on a peripheral nerve can also cause dysesthesias, pain, or weakness. There may be sexuality, reproduction, and fertility concerns, particularly in younger as well as sexually active patients. The concerns and possible solutions should be discussed, addressed, or referred to a specialist as appropriate.

Depression and anxiety are not uncommon and usually respond to supportive counseling, but may need antidepressants such as the selective serotonin reuptake inhibitor or the serotonin-norepinephrine reuptake inhibitor drugs.

Potential Treatment Complications

Treatment complications may occur because of the medications and equipment required to manage the disease and its complications. Strictures or tracheal irritation can result from tracheostomy tubes, lung infections are common in this population, and the ventilators may break down, resulting in an emergency situation. High-dose corticosteroids frequently used for treatment of inflammation in the spinal cord may result in peptic ulcer disease or gastrointestinal bleeding. Thromboembolism prophylaxis and anticoagulant treatment in the event of this happening may result in serious bleeding complications. Skin breakdown may result at contact and pressure areas with devices or dressings used.

Frequent catheterization results in increased risk for urinary tract infections and accidental creation of false passages in the urethra with development of strictures. If bowel programs are not well managed or carried out gently, there may be discomfort, pain, and anorectal injuries.

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