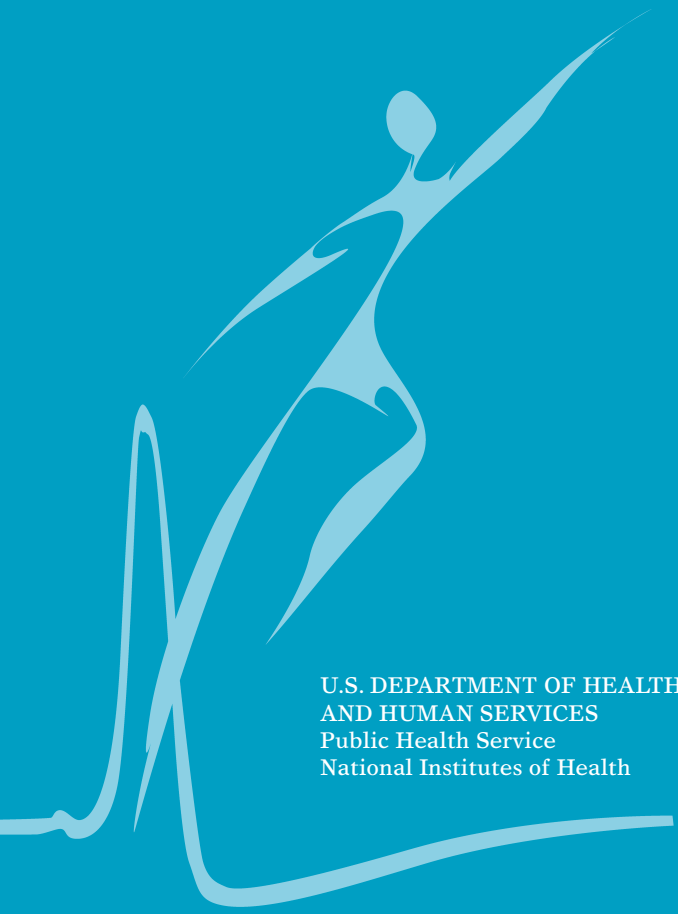
Transverse Myelitis



U.S. DEPARTMENT OF HEALTH
AND HUMAN SERVICES
Public Health Service
National Institutes of Health

Transverse Myelitis

What is transverse myelitis?

Transverse myelitis is a neurological disorder caused by inflammation across both sides of one level, or segment, of the spinal cord. The term *myelitis* refers to inflammation of the spinal cord; *transverse* describes the position of the inflammation—across the width of the spinal cord. Attacks of inflammation can damage or destroy myelin, the fatty insulating substance that covers nerve cell fibers. This damage causes nervous system scars that interrupt communications between the nerves in the spinal cord and the rest of the body.

Symptoms of transverse myelitis include a loss of spinal cord function over several hours to several weeks. What usually begins as a sudden onset of lower back pain, muscle weakness, or abnormal sensations in the toes and feet can rapidly progress to more severe symptoms, including paralysis, urinary retention, and loss of bowel control. Although some individuals recover from transverse myelitis with minor or no residual problems, others suffer permanent impairments that affect their ability to perform ordinary tasks of daily living. Most individuals will have only one episode of transverse myelitis; a small percentage may have a recurrence.

The segment of the spinal cord at which the damage occurs determines which parts of the body are affected. Nerves in the *cervical* (neck) region control signals to the neck, arms, hands, and the muscles that control breathing (the diaphragm). Nerves in the *thoracic* (upper back) region relay signals to the torso and some parts of the arms. Nerves at the *lumbar* (mid-back) level control signals to the hips and legs. Finally, *sacral nerves*, located within the lowest segment of the spinal cord, relay signals to the groin, toes, and some parts of the legs. Damage at one segment will affect function at that segment and segments below it. In individuals with transverse myelitis, demyelination usually occurs at the thoracic level, causing problems with leg movement and bowel and bladder control, which require signals from the lower segments of the spinal cord.

Who gets transverse myelitis?

Transverse myelitis occurs in adults and children, in both genders, and in all races. A peak in incidence rates (the number of new cases per year) appears to occur between ages 10 and 19 years and 30 and 39 years. Although only a few studies have examined incidence rates, it is estimated that about 1,400 new cases of transverse myelitis are diagnosed each year in the United States, and approximately 33,000 Americans have some type of disability resulting from the disorder.

What causes transverse myelitis?

Researchers are uncertain of the exact causes of transverse myelitis. The inflammation that causes such extensive damage to nerve fibers of the spinal cord may result from viral infections or abnormal immune reactions. Transverse myelitis also may occur as a complication of syphilis, measles, Lyme disease, and some vaccinations, including those for chickenpox, pertussis, and rabies. Cases in which a cause cannot be identified are called *idiopathic*.

Transverse myelitis often develops following viral infections. Infectious agents suspected of causing transverse myelitis include varicella zoster (the virus that causes chickenpox and shingles), herpes simplex, cytomegalovirus, Epstein-Barr, influenza, echovirus, human immunodeficiency virus (HIV), hepatitis A, and rubella. Bacterial skin infections, middle-ear infections (*otitis media*), and bacterial pneumonia also have been associated with the condition.

In post-infectious cases of transverse myelitis, immune system mechanisms, rather than active viral or bacterial infections, appear to play an important role in causing damage to spinal nerves. Although researchers have not yet identified the precise mechanisms of spinal cord injury in these cases, stimulation of the immune system in response to infection indicates that an autoimmune reaction may be responsible. This theory is further reinforced by the observation that some individuals who experience transverse myelitis also

have such autoimmune diseases as systemic lupus erythematosus, Sjogren's syndrome, and sarcoidosis. In autoimmune diseases, the immune system—which normally protects the body from foreign organisms—mistakenly attacks the body's own tissue. This can cause inflammation and, in some cases, damage to myelin within the spinal cord.

In addition, some cancers may trigger an abnormal immune response that may lead to transverse myelitis.

In some people, transverse myelitis represents the first symptom of an underlying demyelinating disease of the central nervous system such as multiple sclerosis (MS) or neuromyelitis optica (NMO). A form of transverse myelitis known as "partial" myelitis, because it affects only a portion of the cross-sectional area of the spinal cord, is more characteristic of MS. Neuromyelitis optica typically causes both transverse myelitis and optic neuritis (inflammation of the optic nerve that results in visual loss), but not necessarily at the same time. All individuals with transverse myelitis should be evaluated for MS or NMO because people with these diagnoses may require different forms of treatment, including therapies to prevent future attacks.

What are the symptoms of transverse myelitis?

Transverse myelitis may be either *acute* (developing over hours to several days) or *subacute* (usually developing over 1 to 4 weeks). Initial symptoms usually include localized lower back pain, sudden *paresthesias* (abnormal sensations such as burning, tickling, pricking, or tingling) in the legs, sensory loss, and *paraparesis* (partial paralysis of the legs). Paraparesis may progress to *paraplegia* (paralysis of the legs and lower part of the trunk). Bladder and bowel dysfunction is common. Many people also report experiencing muscle spasms, a general feeling of discomfort, headache, fever, and loss of appetite. Depending on which segment of the spinal cord is involved, some individuals may experience respiratory problems as well.

From this wide array of symptoms, four classic features of transverse myelitis emerge: 1) weakness of the legs and arms, 2) pain, 3) a change in sensation, and 4) bowel and bladder dysfunction. Most people will experience weakness of varying degrees in their legs; some also experience it in their arms. Initially, people with transverse myelitis may notice that they are stumbling or dragging one foot or that their legs seem heavier than normal. Coordination of hand and arm movements, as well as arm and hand strength, may also be compromised. Progression of the disease may lead to full paralysis of the legs, requiring the individual to use a wheelchair.

Pain is the primary symptom of transverse myelitis in approximately one-third to one-half of all individuals. The pain may be localized in the lower back or may consist of sharp, shooting sensations that radiate down the legs or arms or around the torso.

People with transverse myelitis who experience sensory disturbances often use terms such as *numbness*, *tingling*, *coldness*, or *burning* to describe their symptoms. Up to 80 percent of those with transverse myelitis report areas of heightened sensitivity to touch, such that clothing or a light touch with a finger causes significant discomfort or pain (a condition called *allodynia*). Many also experience heightened sensitivity to changes in temperature or to extreme heat or cold.

Bladder and bowel problems may involve increased frequency of the urge to urinate or have bowel movements, incontinence, difficulty voiding, the sensation of incomplete evacuation, and constipation. Over the course of the disease, the majority of people with transverse myelitis will experience one or several of these symptoms.

How is transverse myelitis diagnosed?

Physicians diagnose transverse myelitis by taking a medical history and performing a thorough neurological examination. Because it is often difficult to distinguish between someone with an idiopathic form of transverse myelitis and one who has an underlying condition, physicians must first eliminate potentially treatable causes of the condition.

When a spinal cord problem is suspected, physicians seek first to rule out structural *lesions* (damaged or abnormally functioning areas) that could cause spinal cord compression or otherwise affect its function. Such potential lesions include tumors, herniated or slipped discs, *stenosis* (narrowing of the canal that holds the spinal cord), abscesses, and abnormal collections of blood vessels.

Diagnostic imaging of the brain and spine, using *magnetic resonance imaging* (MRI), can rule out such lesions and check for inflammation. MRI uses computer-generated radio waves and a powerful magnetic field to produce detailed images of body structures, including tissues, organs, bones, and nerves. MRI will almost always confirm the presence of a lesion within the spinal cord whereas the brain MRI may provide clues to other underlying causes, especially MS. If an MRI is not possible (for example, if the individual has a pacemaker), physicians may consider other diagnostic tests such as *computed tomography* (CT, which uses x-rays and a computer scanner to provide cross-sectional images of bones, tissues, and organs) of the spine. *Myelography* involves injecting a dye into the spinal canal to enhance x-ray imaging of the spine. *Myelograms* are used to diagnose spinal nerve injury, herniated discs, and spinal tumors.

Blood tests may be performed to rule out various systemic lupus erythematosus, HIV infection, vitamin B_{12} deficiency, and various other disorders. A blood test for NMO, called NMO-IgG, is also necessary. In some individuals with transverse myelitis, the cerebrospinal fluid that bathes the spinal cord and brain contains more protein than usual and an increased number of leukocytes (white blood cells). A spinal tap may be performed to obtain fluid to study these factors, exclude infections, and to look for markers of such diseases as MS.

If none of these tests suggests a specific cause, the person is presumed to have idiopathic transverse myelitis.

How is transverse myelitis treated?

As with many disorders of the spinal cord, no effective cure currently exists for people with transverse myelitis. Treatments are designed to reduce spinal cord inflammation and alleviate or manage symptoms. Physicians often prescribe anti-inflammatory corticosteroid therapy as soon as the diagnosis is made in order to decrease inflammation and improve the chances and speed of neurological recovery. Although no clinical trials have investigated whether corticosteroids alter the course of transverse myelitis, these drugs often are prescribed to reduce immune system activity because of the suspected autoimmune mechanisms involved in the disorder. Corticosteroid medications that

might be prescribed may include intravenous methylprednisone or dexamethasone (usually for about 5 days); in some cases, oral prednisone is used for a period of time afterwards. In severe cases that do not appear to respond to corticosteroid treatment, other therapies such as plasma exchange or drug therapies may be used to try to salvage neurological function. General painkillers may be prescribed for any pain the individual may have. Other symptoms such as muscle spasms may require additional drug therapies. Bladder dysfunction may require the placement of a urinary catheter to drain the bladder.

Following initial therapy, the most critical part of the treatment for this disorder consists of keeping the individual's body functioning in anticipation of either complete or partial spontaneous recovery of the nervous system. This may require placing the person on a respirator in the uncommon scenario where breathing is significantly affected. People are most often treated in a hospital or in a rehabilitation facility where a specialized medical team can prevent or treat problems that afflict paralyzed patients. Often, even before recovery begins, caregivers may be instructed to move an individual's limbs manually to help keep the muscles flexible and strong, and to reduce the likelihood of pressure sores developing in immobilized areas. Later, if the person begins to recover limb control, physical therapy can help improve muscle strength, coordination, and range of motion.

What therapies are available to help people who have permanent physical disabilities?

Many forms of long-term rehabilitative therapy are available for people who have permanent disabilities resulting from transverse myelitis. Medical social workers, often affiliated with local hospitals or outpatient clinics, are the best sources for information about treatment programs and other resources that exist in a community. Rehabilitative therapy teaches people strategies for carrying out activities in new ways in order to overcome, circumvent, or compensate for permanent disabilities. Rehabilitation cannot reverse the physical damage resulting from transverse myelitis or other forms of spinal cord injury, but it can help people, even those with severe paralysis, become as functionally independent as possible and thereby attain the best possible quality of life.

Common permanent neurological deficits resulting from transverse myelitis include severe weakness, *spasticity* (painful muscle stiffness or contractions), or paralysis; incontinence, and chronic pain. Such deficits can substantially interfere with a person's ability to carry out such everyday activities as bathing, dressing, and performing household tasks.

People living with permanent disability may feel a range of emotions, from fear and sadness to frustration and anger. Such feelings are natural responses, but they can sometimes jeopardize health and potential for functional recovery. Those with permanent disabilities frequently experience clinical depression.

Fortunately, depression is treatable, due to the development of a wide range of medications that can be used with psychotherapeutic treatment.

Today, most rehabilitation programs attempt to address the emotional dimensions along with the physical problems resulting from permanent disability. Individuals typically consult with a range of rehabilitation specialists, who may include physiatrists (physicians specializing in physical medicine and rehabilitation), physical therapists, occupational therapists, vocational therapists, and mental health care professionals.

Physical Therapy: Physiatrists and physical therapists treat disabilities that result from motor and sensory impairments. Their aim is to help individuals increase their strength and endurance, improve coordination, reduce spasticity and muscle wasting in paralyzed limbs, and regain greater control over bladder and bowel function through various exercises. Physiatrists and physical therapists teach paralyzed individuals techniques for using assistive devices such as wheelchairs, canes, or braces as effectively as possible. Paralyzed patients also learn ways to avoid developing painful pressure sores on immobilized parts of the body, which may lead to increased pain or systemic infection. In addition, physiatrists and physical therapists are involved in pain management. A wide variety of medications can alleviate the pain that results from spinal cord injuries such as those caused by transverse myelitis. Some examples include nonsteroidal anti-inflammatory drugs such as ibuprofen or

naproxen; antidepressant drugs such as amitryptyline (tricyclic) and sertraline (a selective serotonin reuptake inhibitor); muscle relaxants such as baclofen or tizanidine; and anticonvulsant drugs such as gabapentin, pregabalin, and carbamazepine.

Occupational Therapy: Occupational therapists help people learn new ways of performing meaningful, self-directed, goal-oriented, everyday tasks such as bathing, dressing, preparing a meal, house cleaning, engaging in arts and crafts, or gardening. They teach people how to develop compensatory strategies, how to make changes in their homes to improve safety (such as installing grab bars in bathrooms), how to change obstacles in their environment that interfere with normal activity, and how to use assistive devices.

Vocational Therapy: In addition to acquainting people with their rights as defined under the Americans with Disabilities Act of 1990 and helping people develop and promote work skills, vocational therapists identify potential employers, assist in job searches, and act as mediators between employees and employers to secure reasonable workplace accommodations.

What is the prognosis?

Recovery from transverse myelitis usually begins within 2 to 12 weeks of the onset of symptoms and may continue for up to 2 years. However, if there is no improvement within the first 3 to 6 months, significant recovery is unlikely. About one-third of people affected

with transverse myelitis experience good or full recovery from their symptoms, regaining the ability to walk normally and experience minimal urinary or bowel effects and paresthesias. Another one-third show only fair recovery and are left with significant deficits such as spastic gait, sensory dysfunction, and prominent urinary urgency or incontinence. The remaining one-third show no recovery at all, remaining wheelchair-bound or bedridden with marked dependence on others for basic functions of daily living. Unfortunately, making predictions about individual cases is difficult. However, research has shown that a rapid onset of symptoms generally results in poorer recovery.

The majority of people with this disorder experience only one episode although in rare cases recurrent or relapsing transverse myelitis does occur. Some individuals recover completely, and then experience a relapse. Others begin to recover, and then suffer worsening of symptoms before recovery continues. In cases of relapse, physicians will re-evaluate possible underlying causes such as MS, NMO, or systemic lupus erythematosus since most people who experience relapse have an identifiable underlying disorder. People with a recurrent/relapsing disorder will usually require some type of ongoing therapy that modulates or suppresses the immune system. The purpose of such therapies is to reduce the chance of future relapses.

What research is being done?

Within the Federal government, the National Institute of Neurological Disorders and Stroke (NINDS), a component of the National Institutes of Health (NIH), has primary responsibility for conducting and supporting research on spinal cord disorders and demyelinating diseases such as transverse myelitis. The NINDS conducts research in its laboratories at the NIH and also supports studies through grants to major medical research institutions across the country.

Some NINDS researchers are seeking to clarify the role of the immune system in the development of demyelination in auto-immune diseases or disorders. Other work focuses on strategies to repair demyelinated spinal cords including approaches using cell transplantation. The knowledge gained from such research should lead to a greater knowledge of the mechanisms responsible for demyelination in transverse myelitis and may ultimately provide a means to prevent and treat this disorder.

NINDS also funds researchers who are using animal models of spinal cord injury to study strategies for replacement or regeneration of spinal cord nerve cells. The ultimate goals of these studies are to encourage the same regeneration in humans and to restore function to paralyzed individuals. Scientists are also developing neural prostheses to help people with spinal cord damage compensate for lost function. These sophisticated electrical and mechanical devices connect with the nervous system to supplement or replace lost motor and sensory function.

Where can I find more information?

For more information on neurological disorders or research programs funded by the NINDS, contact the Institute's Brain Resources and Information Network (BRAIN) at:

BRAIN
P.O. Box 5801
Bethesda, MD 20824
301-496-5751
800-352-9424
www.ninds.nih.gov

Information also is available from the following organizations:

Cody Unser First Step Foundation
P.O. Box 56696
Albuquerque, NM 87187
505-792-9551
www.cufsf.org

Christopher and Dana Reeve Foundation and Resource Center
636 Morris Turnpike
Suite 3A
Short Hills, NJ 07078
800-225-0292
www.christopherreeve.org

Miami Project to Cure Paralysis
1095 NW 14th Terrace
Lois Pope LIFE Center
Miami, Florida 33136
305-243-6001
800-782-6387
www.themiamiproject.org

National Multiple Sclerosis Society
733 Third Avenue
3rd Floor
New York, NY 10017-3288
800-344-4867
www.nationalmssociety.org

Transverse Myelitis Association
1787 Sutter Parkway
Powell, OH 43065-8806
614-766-1806
www.myelitis.org

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