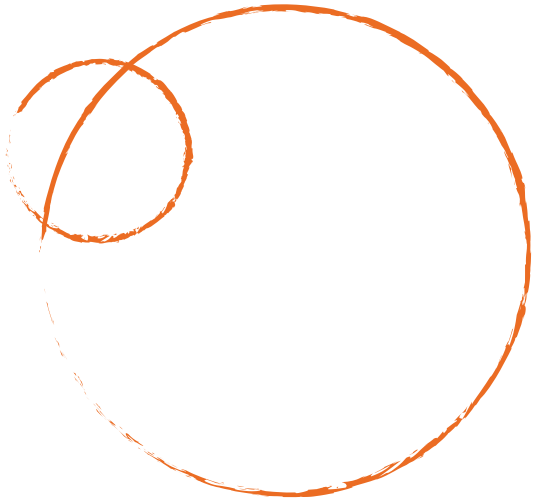
Fact Sheet

Acute Flaccid Myelitis

AFM



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Acute Flaccid Myelitis (AFM) is a type of inflammation in the spinal cord that has specific clinical and MRI features. AFM abnormalities noted on MRI are predominantly found in the grey matter (lower motor neuron) of the spinal cord. In 2012, an outbreak of AFM occurred in California, and more cases were reported in the summer and fall every other year (2014, 2016, and 2018) across the United States. Non-polio enteroviruses have been implicated as potential causal factors in the development of AFM. Enterovirus D68 and enterovirus A71 have been suspected in many of these cases, although other enteroviruses such as coxsackievirus have been implicated as well. Enterovirus D68 most often causes a respiratory illness and circulates in the United States during the summer and fall every other year, which coincides with the increase of cases of AFM seen every other year. The expected biennial increase of enterovirus D68 and subsequent AFM cases for 2020 did not occur and can likely be attributed to personal protection and prevention measures in place due to the COVID-19 pandemic. While some models predicted a spike of enterovirus circulation and therefore AFM, we did not see a spike in cases in 2021. It has not been definitively proven that these particular viruses have directly caused cases of AFM, but the temporal onset of neurological symptoms with infections produced by those viruses implicate them as direct or indirect triggers of the neurological problem.

Epidemiology

There are no conclusive studies that identify the actual numbers of individuals specifically affected by AFM, but to date and since surveillance began in 2014, the Centers for Disease Control and Prevention (CDC) confirmed over 670 reports of those affected by AFM. Not all cases are reported to CDC nor confirmed by CDC, so this number is likely an underestimation. There have been reports of AFM in both children and adults, but AFM cases primarily affect children under the age of 18, with a median age of 6.3 years old. Less than 15% of all AFM cases occur in adults, although this may be an underestimate. Males may be more likely to be diagnosed with AFM. AFM confirmed cases and reports are regularly updated by CDC and can be viewed by state at srna.ngo/cdc/cases.

Until the recent characterization of AFM in 2014, it is likely that many individuals with initial presentation of flaccid limb weakness and/or paralysis with predominantly grey matter lesions of the spinal cord were diagnosed with transverse myelitis or Guillain Barre Syndrome in previous years.

Signs and Symptoms

Most of those diagnosed with AFM report having a respiratory or gastrointestinal illness before the onset of weakness. The predominant presentation is a rapid onset of weakness within hours to a few days that may affect the limbs, face, and the muscles that control breathing and/or swallowing. The weakness may rapidly progress from limb weakness to more severe symptoms, including respiratory failure, so hospitalization for observation is recommended even with mild symptoms. Those with AFM may not be able to breathe, swallow, or move their eyes normally. Weakness varies greatly, ranging from mild to very severe. AFM may result in weakness, partial paralysis, or total paralysis of just one limb or all limbs. The pattern of paralysis and how individuals present are widely variable. Weakness most often occurs in proximal muscles, meaning the muscles closest to the center of the body. Pain in the neck, back, or limb may be an early symptom. Autonomic instability, such as issues with heart rate, blood pressure or temperature regulation, may occur as well. Sensation and bowel and bladder function are generally spared in children with AFM. Some individuals, those more severely affected, may have inflammation in both the white and grey matter of the spinal cord (upper and lower motor neuron) and may experience impaired sensation and/or bowel and bladder dysfunction.

Diagnosis

Acute flaccid myelitis is diagnosed based upon clinical exam, magnetic resonance imaging (MRI) of the spinal cord, and analysis of cerebrospinal fluid (CSF) (usually with increased white blood cells or pleocytosis). On MRI of the spinal cord, AFM lesions are longitudinal throughout the grey matter (the anterior horn cells). Sometimes imaging may appear normal early in the disease, but repeat imaging shows the lesions. In some situations, electrophysiological studies of the nerves and muscle

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(called nerve conduction and electromyogram [NCS/EMG]) may help to determine if there is injury to the lower motor neuron. Testing may also include blood draws, respiratory tract samples, or collection of other bodily fluids to determine if a viral or infectious cause is present.

Acute Treatments

Specific treatments and interventions for AFM have not yet been identified, but some of the treatments available for transverse myelitis have been used, including high dose intravenous (IV) steroids, intravenous immunoglobulin (IVIG), and plasma exchange (PLEX). The purpose of the treatments is to attempt to reduce inflammation in the spinal cord and further prevent the individual's immune system from causing damage. IVIG has antibodies that may limit inflammation or replication of enteroviruses and is widely used in treating AFM. The data on use of steroids or PLEX are mixed. Fluoxetine was used in several centers in the US in 2016 and was well tolerated but was not associated with improved outcomes among treated children. As is usual with treatment of rare neuroimmune disorders in which placebo-controlled trials are difficult to perform, treatment must be individualized. Early initiation of physical and occupational therapy is recommended to decrease the development of secondary consequences of AFM.

Prognosis and Management

Recovery varies among individuals with AFM. Most do not recover fully, but patients do regain strength and motor function over time to varying degrees. The most affected muscle may be the least likely to recover. Again, physical and occupational therapy are also believed to be critical for recovery in AFM.

Long-Term Care

After the acute phase, rehabilitative care to improve functional skills and prevent secondary complications of immobility involves both psychological and physical accommodations. Rehabilitation may begin in the intensive care unit with the goal to transition individuals to an inpatient or outpatient rehabilitation program. Much has been written regarding recovery from spinal cord injury (SCI), in general, and this literature can often apply to those with AFM, although there are some distinct differences that occur in AFM due to the grey matter of the spinal cord being impacted. The issues include respiratory dysfunction, muscle tone, musculoskeletal consequences, skin breakdown, pain, bladder function, bowel function, depression and anxiety, autonomic dysreflexia, and sexual dysfunction, nerve and tendon transfers, and rehabilitation and learning/relearning activities of daily living (e.g., dressing).

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contractures that lead to a decreased range of motion. Assessment and fitting for splints designed to passively maintain an optimal position for limbs that cannot be actively moved is an important part of the management at this stage.

The long-term management of AFM requires attention to a number of issues. These are the residual effects of any spinal cord injury, including AFM. In addition to chronic medical problems, there are the ongoing issues of ordering the appropriate equipment, reentry into school, re-socialization into the community, and coping with the psychological effects of this condition by the patients and their families. During the early recovery period, family education is essential to develop a strategic plan for dealing with the challenges to independence following return to the community.

Respiratory Dysfunction

A subset of patients with AFM may experience marked respiratory and diaphragmatic dysfunction. It may occur when the neurons that control diaphragm movement innervated by the phrenic nerve, or intercostal muscles that partially control the mechanics of breathing, fail due to damage of motor neurons in the spinal cord. Thus, patients can have difficulty breathing and require long-term ventilatory support. Over months or years, patients can be weaned from the ventilator as motor control of the diaphragm comes back, but in some patients it has not yet returned. Strategies such as diaphragm pacing are being used now although there is still need for a validated demonstration of their efficacy. Those with respiratory issues may also experiences issues with heart rate, like tachycardia (high heart rate) and bradycardia (low heart rate) as well as hypertension (high blood pressure) or orthostatic hypotension (low blood pressure), especially with attempts at standing.

Muscle Tone

In those with AFM where only the grey matter is impacted, muscles will be weak and lack muscle tone, so rehabilitation strategies should be adapted to this low level of muscle tone. Serial casting is sometimes used in those with AFM. Due to a lack of muscle movement and tone, those with AFM are at higher risk for contractures, or when limbs become stuck in certain positions. Stretching and maintaining range of motion are important to prevent contractures. Those with AFM with white matter involvement can experience increased muscle tone or spasticity. Spasticity means stiffness or muscle spasms and may be a very difficult problem to manage. Those with spasticity will benefit from strategies used in individuals with other rare neuroimmune disorders or spinal cord injuries.

Musculoskeletal Complications

Most with AFM will experience musculoskeletal complications associated with muscle weakness and atrophy. Joint subluxation or dislocation, mostly in proximal joints (shoulder and hip) with severe muscle weakness, is common in AFM. Treatment for subluxation includes therapy to strength the muscles surrounding the joint, weight bearing through the joint, and braces or slings to help stabilize the joint. Joint contractures that limit range of motion despite aggressive stretching and bracing may require surgical intervention if they are interfering with function. Those with AFM may have weakness in the neck and/or trunk muscles that can lead to curvature or twisting called scoliosis. Therapy directed at strengthening the muscles in the

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neck and trunk and soft or hard braces may be recommended to slow progression. Proper positioning and support in a custom wheelchair, if used, is also important in decreasing scoliosis. Some with severe scoliosis will ultimately require surgery to correct the scoliosis and stabilize the spine. With growth, differences in limb length in the affected vs unaffected extremity can occur and may require surgical intervention, especially in the lower extremity if walking becomes difficult. It is important to identify an orthopedist to monitor and treat these potential conditions. Decreased bone density, as seen in other spinal cord injuries, can occur, placing those with AFM at increased risk for fractures with only minor trauma. Low bone density is diagnosed with a DEXA scan. Standing and weightbearing as well as adequate calcium and vitamin D intake through diet or supplementation are good first steps to preventing this. Those with significantly low bone density may benefit from medications to prevent ongoing the loss of bone density.

Skin Breakdown

While sensation is frequently intact in AFM, impaired mobility may place individuals at risk for skin breakdown due to a lack of or less movement. Skin breakdown occurs if the skin is exposed to pressure for a significant amount of time, without the strength to shift position as necessary, especially for those who lack sensation. Sitting position should be changed at least every 15 minutes while sitting. This can be accomplished by standing, by lifting the body up while pushing down on armrests, or by just leaning and weight shifting. Wheelchairs can be supplied with either power mechanisms of recline or tilt-in-space to redistribute weight bearing. A variety of wheelchair cushions are available to minimize sitting pressure. During bedrest, repositioning at least every 4 hours is needed to prevent skin breakdown. Redness that does not blanch when finger pressure is applied may signal the beginning of a pressure ulcer. Good nutrition, vitamin C, and avoidance of moisture all contribute to healthy skin. Pressure ulcers are much easier to prevent than to heal.

Pain

Changes in sensation may occur in those with AFM and can manifest as lack of sensation, or numbness, as well as painful sensations called neuropathic pain. For most with AFM, these sensory changes occur acutely and gradually resolve over time. If someone with AFM continues to have nerve pain after this acute phase, a discussion with a health care provider on how to manage it is warranted. There is a long list of medications used to treat these symptoms. The same medication doesn't work for everyone, so trial and error of finding the right medication can be frustrating. Alternative therapies such as acupuncture and meditation have also been utilized, with varying success.

The first step in treating pain effectively is obtaining an accurate diagnosis. Unfortunately, this can be very difficult. Causes of pain include muscle strain from using the body in an unaccustomed manner, nerve compression (i.e., compression of the ulnar nerve at the elbow due to excessive pressure from resting the elbow on an armrest continuously), or pain from muscle contractures and stretching. Muscle pain might be treated with analgesics, such as acetaminophen (Tylenol), non-steroidal, anti-inflammatory drugs such as naproxen or ibuprofen (Naprosyn, Aleve, Motrin), or

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modalities such as heat or cold. Nerve compression might be treated with repositioning and padding (i.e., an elbow pad for an ulnar nerve compression).

Stress and depression should also be addressed since these conditions make any form of pain harder to tolerate.

Bladder Function

Bladder dysfunction may not occur in all individuals with AFM. Those with AFM may have the sensation to urinate but may have difficulty with contraction of the bladder to release urine. If the bladder cannot release urine, this can lead to urinary retention, urinary tract infections, and bladder or kidney stones. Two general problems can affect the bladder. The bladder can become overly sensitive and empty after only a small amount of urine has collected, or relatively insensitive, causing the bladder to become over extended and overflow. An overly distended bladder increases the likelihood of urinary tract infections and, in time, may threaten the health of the kidneys. Depending on the dysfunction, treatment options include timed voiding, medicines, external catheters for males (a catheter connected to a condom), pads, intermittent internal self-catheterization, an indwelling catheter, or electrical stimulation. Surgical options may be appropriate for some people. Common bladder problems include incontinence, frequency, nocturia (frequent urination at night), hesitancy, and retention. Treating incontinence, frequency, and nocturia is often easier than treating hesitancy and retention, where clean intermittent urinary catheterizations are the basic component to success. Urodynamic testing is necessary to determine urine retention to check risk for urinary tract infections, particularly if there is a history of urinary tract infections (UTIs), to guide the urologist in terms of the best management. Also, if a young child with AFM fails to potty train or if any child develops a urinary tract infection or urinary incontinence, they should be evaluated by a urologist. Working with a good urologist is imperative to prevent potential serious complications, particularly one who understands spinal cord disease.

Bowel Function

Another major area of concern is effective management of bowel function. Most of those with AFM have constipation due to decreased gastrointestinal motility and decreased physical mobility. They may have the sensation of needing to have a bowel movement, but may experience difficulty eliminating the stool. Incontinence can occur with constipation when liquid stool seeps around hard stool in the colon. A high-fiber diet, adequate and timely fluid intake, and medications to regulate bowel evacuations are the basic components of success. Stool softeners and oral laxatives can be used, and it is recommended to sit on the toilet at the same time every day after a meal to aid in producing a bowel movement. The use of a cone enema or anal irrigation may aid in the management of constipation or incontinence. Those with ongoing difficulty with bowel function should speak with their health care professional about potential management strategies.

Depression and Anxiety

Individuals and caregivers of children with AFM should be aware and educated about the possible effect of AFM on mood regulation. Those with AFM should be routinely screened for the development of symptoms consistent with clinical depression and

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anxiety. Based on current, limited research, symptoms associated with depression and anxiety may be present as a child makes the normal adjustment following a diagnosis of AFM. If symptoms of depression and anxiety persist, it may warrant longer-term monitoring and possible intervention to assess for clinical anxiety or depressive disorder. Signs of anxiety and depression present differently in children than adults. Signs in children may include changes in appetite or sleeping habits, they may be more withdrawn than before, more irritable, tired, or uninterested in their usual favorite activities. They may also experience problems at school or complain of headaches and stomachaches. Younger children may experience more behavioral issues such as hyperactivity or aggression.

A child exhibiting a preoccupation with death or suicidal thoughts constitutes a true psychiatric emergency and should lead to prompt evaluation and treatment. While the prevalence of depression and anxiety in children with AFM is not known, parent-reported depression symptoms in children with a diagnosis of TM (which likely included children now known as AFM) occurred at a higher rate than the average population. Depression is not due to a child's weakness or the inability to "cope." It can have devastating consequences and could worsen physical symptoms (such as fatigue, pain, and decreased concentration). With appropriate recognition and treatment of depression, complete depressive symptom remission is possible with proper psychotherapy and pharmacotherapy.

During the early recovery period, family education is essential to develop a strategic plan for dealing with the challenges to a child's independence level, appropriate for their age and developmental levels, following a return to home. Ongoing problems typically include ordering the appropriate equipment, dealing with re-entry into school, work, and community, and coping with the psychological effects of an AFM diagnosis for both the child and their family. Being saddened or demoralized by the diagnosis of AFM is possible. A child will go through an adjustment period adapting to the diagnosis and possible disability caused by AFM. The inability to move through grief in a reasonable amount of time such that it interferes with relationships and functional living needs to be addressed and treated.

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Autonomic Dysreflexia

Autonomic dysreflexia can occur when a spinal cord is damaged above the T6 level. Symptoms can include nausea, sweating, fast heart rate, and/or profound blood pressure changes (up or down). Episodes can be triggered by urinary tract infections, catheterizations, constipation, or painful events in the lower extremities. Care should be given to minimize triggers and manage any blood pressure variations during an event. Those with severe paralysis in both the arms and legs will be most susceptible to experiencing autonomic dysreflexia.

Sexual Dysfunction

Sexual dysfunction involves similar innervation and analogous syndromes as those found in bladder dysfunction. Treatment of sexual dysfunction in adults diagnosed with AFM should take into account baseline function before the onset of AFM. Most of those who are diagnosed with mild AFM will have intact sexual function. Until we learn more about this issue in AFM, adults with AFM experiencing sexual dysfunction may want to refer to the strategies used in individuals with other rare neuroimmune disorders or spinal cord injuries.

Nerve and Tendon Transfers

Some individuals may benefit from nerve or tendon transfer procedures to improve function, which is when nerves or tendons are taken from one area of the body and are transferred to a denervated nerve or a different tendon. Previous experience derived from obstetric brachial plexus injury has guided some of the approaches in patients with AFM. The value in recovery of selected upper extremity muscle groups in AFM patients appears promising, although there is still need for a well-documented and validated approach to prove their beneficial outcomes. There are some cases reported in the literature of successful nerve and tendon transfers. Additional studies are needed to learn the correct timing for when nerve transfers should occur after onset.

Rehabilitation and Activities of Daily Living

It is important to begin occupational and physical therapies early during the course of recovery to prevent the inactivity-related problems of skin breakdown and soft tissue contractures that lead to a decreased range of motion. Assessment and fitting for splints designed to passively maintain an optimal position for limbs that cannot be actively moved is an important part of the management at this stage.

Activity-based rehabilitation includes weight-bearing exercise, functional electrical stimulation (FES), locomotor training, task-specific practice, and massed practice. Individuals with AFM may not respond to FES, but therapists can adjust FES parameters to try to get a better muscle contraction. FES even without obvious muscle contraction may have benefits. Furthermore, since those with AFM have intact sensation, this can limit their tolerance to the stimulation intensity needed to get motor contraction. Using stimulation with a low frequency and long pulse width can allow for time for the slower moving motor units to respond with greater refractory periods.

Weight-bearing exercise has been shown to improve bone mineral density, range of motion, muscle tone, and bowel function. Vibration during weight bearing may also activate denervated muscles. Weight-bearing exercises can progress to locomotor

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training on the treadmill with appropriate orthotics. Aquatic therapy can be helpful for stretching tight muscles. Task-specific practice involves relearning functions that were lost due to AFM such as bed mobility and coming to a seated position, feeding, dressing, and personal hygiene. Some children who were younger when AFM onset occurred will have to learn these for the first time. Massed practice involves repetition and increased level of activity. It has been shown that children can tolerate up to 5 hours of therapy a day. It is important to keep in mind that AFM can result in risks of skeletal subluxations and decreased bone mineral density which can result in fractures.

Other rehabilitation factors to consider are pulmonary management for those with ventilator dependence, and speech and language pathologists for those children with difficulty swallowing (dysphagia) and talking (dysphonia).

Individuals with AFM may find ordinary tasks such as dressing, bathing, grooming, and eating very difficult. Many of these obstacles can be mastered with training and specialized equipment. For example, long handled sponges can make bathing easier as can grab bars, portable bath seats, and hand-held shower heads. For dressing, elastic shoelaces can eliminate the need to tie shoes while other devices can aid in donning socks. Occupational therapists are specialists in assessing equipment needs and helping people with limited function perform activities of daily living. A home assessment by an experienced professional is often helpful.

Physical therapists assist with mobility. Besides teaching people to walk and transfer more easily, they can recommend mobility aids. This includes everything from canes (single point vs. small quad cane vs. large quad cane) to walkers (static vs. rolling vs. rollator) and braces. For a custom-fabricated orthotic (brace), an orthotist is necessary. Careful thought should go into deciding whether the brace should be an ankle-foot orthosis, whether it should be flexible or stiff, and what angle the foot portion should be in relationship to the calf portion. Some will benefit by a knee-ankle foot orthosis. Each person should be evaluated individually. The best results occur when a physician coordinates the team so that the therapists and orthotists are united on what is to be achieved. The physician best trained to take this role is the physiatrist.

Additional Resources

Myelitis Helpline
srna.ngo/helpline

For questions about our organization and rare neuroimmune disorders, visit the Myelitis Helpline, an online tool developed by SRNA.

Resource Library
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