ACUTE FLACCID MYELITIS

Acute Flaccid Myelitis (AFM) is a variant or sub-type of transverse myelitis. AFM is inflammation of the spinal cord and generally presents with unique clinical and MRI features that are not typical of classical transverse myelitis. AFM abnormalities noted on MRI are predominantly found in the gray matter of the spinal cord. In 2013, an outbreak of what is now believed to be this sub-type of transverse myelitis occurred in California and more cases were reported in the summer and fall of 2014 across the United States. The enterovirus (EV-D68) has been suspect in many of these cases however, it has not been definitively proven that it is this particular virus that has caused the paralysis,[1] although several cases of AFM occurred at around the same time as an outbreak of the EV-D68 virus.[2]

[1] Centers for Disease Control and Prevention. Summary of Findings: Investigation of Acute Flaccid Myelitis in U.S. Children, 2014-15. http://www.cdc.gov/ncird/investigation/viral/2014-15/investigation.html. August 21, 2015. Accessed September 9, 2015.

[2] Greninger AL, Naccache SN, Messacar K et al. A novel outbreak enterovirus D68 strain associated with acute flaccid myelitis cases in the USA (2012–14): a retrospective cohort study. Lancet Infec Dis. 2015;15:671-82.

EPIDEMIOLOGY

There are no conclusive studies that identify the actual numbers of individuals specifically affected by this variant of transverse myelitis, but from August 2014 to July 2015, the CDC verified 120 reports of children in 34 states who developed AFM, but the CDC does not include individuals over the age of 22 in their numbers or patients effected prior to August 2014.[1] There have been reports of AFM in both children and adults,[2] but recent cases have primarily affected children under the age of 18.[3]

Until the recent characterization of AFM in 2014, it is likely that many individuals with initial presentation of flaccid weakness and paralysis have been diagnosed as transverse myelitis in previous years.

[1] Centers for Disease Control and Prevention. Summary of Findings: Investigation of Acute Flaccid Myelitis in U.S. Children, 2014-15. http://www.cdc.gov/ncird/investigation/viral/2014-15/investigation.html. August 21, 2015. Accessed September 9, 2015.

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[3] Sejvar JJ, Pastula DM, Cortese MM et al. Acute Flaccid Myelitis: Interim Considerations for Clinical Management. http://www.cdc.gov/ncird/downloads/acute-flaccid-myelitis.pdf. Updated November 4, 2014. Accessed September 9, 2015.

SIGNS & SYMPTOMS

The predominant presentation is weakness that may affect the limbs, face, oral or eye muscle. Weakness varies greatly ranging from subtle to very severe. AFM may result in total paralysis, partial paralysis, or weakness of just one limb. The combination of paralysis and how individuals present are widely variable. The limbs or muscle structures of individuals with AFM appear weak, flaccid, or limp and are not spastic as seen in classic cases of transverse myelitis. Since it is

markedly the gray matter of the spinal cord that is inflamed in individuals with AFM, sensory, bowel and bladder functions can remain intact, however there are individuals that have both upper and lower motor neuron involvement.

DIAGNOSIS

Acute Flaccid Myelitis is diagnosed as a sub-type of transverse myelitis based upon clinical exam, MRI findings, lumbar puncture, and may also include a nerve conduction study (EMG) 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 component is present. While EV-D68 has been a suspected pathogen in many cases, it has not been conclusively proven. Enterovirus is not always related to nor is it always the cause of AFM.[2]

[2] Greninger AL, Naccache SN, Messacar K et al. A novel outbreak enterovirus D68 strain associated with acute flaccid myelitis cases in the USA (2012–14): a retrospective cohort study. Lancet Infec Dis. 2015;15:671-82.

ACUTE TREATMENTS

Specific treatments and intervention for AFM have not yet been identified. The treatments available for transverse myelitis have been used, but have not been proven to benefit AFM.

At present, because the majority of cases with acute flaccid myelitis have inflammation of the spinal cord as in classic transverse myelitis, the current acute therapies available for TM (high dose IV steroids, IVIg, and plasma exchange) are being used in AFM. The purpose of the treatments is to attempt to reduce inflammation in the spinal cord and further prevent the individual's immune system from attacking. As is usual with treatment of transverse myelitis, treatment must be individualized. The CDC[3] does not recommend the use of steroids, IVig, or plasma exchange in AFM, but individuals with AFM or caregivers of children with AFM should discuss treatment recommendations with their physician (the CDC recommendation is based on very limited data). Physical therapy is also believed to be significant for recovery in acute flaccid myelitis as it is in transverse myelitis.[3]

[3] Sejvar JJ, Pastula DM, Cortese MM et al. Acute Flaccid Myelitis: Interim Considerations for Clinical Management. http://www.cdc.gov/ncird/downloads/acute-flaccid-myelitis.pdf. Updated November 4, 2014. Accessed September 9, 2015.

PROGNOSIS & MANAGEMENT

Individuals diagnosed with acute flaccid myelitis (AFM) experience different issues (due to largely gray matter involvement) during recovery than traditional transverse myelitis with spastic paralysis. Individuals may not respond to the same therapeutic treatments as TM but may benefit from specific interventions over time. As an example, some individuals may benefit from nerve transplant procedures if there are muscle contractions with electrical stimulation. It is thought that individuals presenting with mild symptoms or weakness are more likely to experience a full recovery. There is limited data to support this and more research is necessary. As with classic transverse myelitis, intensive physical rehabilitation is believed to be essential in the recovery process.

LONG-TERM CARE

The following information is related to transverse myelitis but can applied to acute flaccid myelitis.

After the acute phase, rehabilitative care to improve functional skills and prevent secondary complications of immobility involves both psychological and physical accommodations. There is very little written in the medical literature specifically dealing with rehabilitation after transverse myelitis. However, much has been written regarding recovery from spinal cord injury (SCI), in general, and this literature applies. The physical issues include bowel and bladder management, sexuality, maintenance of skin integrity, spasticity, activities of daily living (i.e., dressing), mobility, and pain.

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.

The long-term management of TM requires attention to a number of issues. These are the residual effects of any spinal cord injury, including TM. 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.

Bladder Function

Bladder function is almost always at least transiently impaired in patients with TM. Immediately after the onset of TM, there is frequently a period of transient loss or depression of neural activity below the involved spinal cord lesion, referred to as “spinal shock,” which lasts about 3 weeks. Following this period, 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), padding for women, intermittent internal self-catheterization, an indwelling catheter or electrical stimulation. Surgical options may be appropriate for some people.

Bowel Function

Another major area of concern is effective management of bowel function. A common problem in spinal cord injury is difficulty with evacuation of stool, although fecal incontinence can also occur. The neurologic pathways for defecation are similar to those of the bladder. Many lacking voluntary control of the bowel may still be able to achieve continence by diet, strategic use of stool softeners and fiber, and the technique of rectal stimulation. Other aids include suppositories and oral medications. A high-fiber diet, adequate and timely fluid intake, and medications to regulate bowel evacuations are the basic components of success. Regular evaluations by medical specialists for adjustment of the bowel program are recommended to prevent potentially serious complications. There are some surgical options, although this is rarely necessary.

Sexual Dysfunction

Sexual dysfunction involves similar innervation and analogous syndromes as those found in bladder dysfunction. Treatment of sexual dysfunction should take into account baseline function before the onset of TM. Of the utmost importance is adequate education and counseling about the known physical and neurologic changes that TM has on sexual functioning. Because of the similarities in innervation between sexual and bladder function, patients with sexual dysfunction should be encouraged to empty their bladders before sexual stimulation to prevent inopportune incontinence. The mainstays of treatment of erectile dysfunction in men are inhibitors of cGMP phosphodiesterase, type 5, which will allow most men with TM to achieve adequate erections for success in intercourse through a combination of reflex and/or psychogenic mechanisms. Although less effective in women, these same types of medications have been shown capable of enhancing a woman's sexual functioning. The most commonly used oral erectile dysfunction drugs are Viagra (sildenafil), Levitra (vardenafil), and Cialis (tadalafil). Although sexual experience is impacted by spinal cord injury, sensual experience and even orgasm are still possible. Lubricants and aids to erection and ejaculation (for fertility) are available. Adjustment to altered sexuality is aided by an attitude of permissive experimentation, as the previous methods and habits may no longer serve.

Skin Breakdown

Skin breakdown occurs if the skin is exposed to pressure for a significant amount of time, without sensation or the strength shift position as necessary. Sitting position should be changed at least every 15 minutes. 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. 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.

Spasticity

Spasticity is often a very difficult problem to manage. The goal is to maintain flexibility with a stretching routine using exercises for active stretching and a bracing program with splints for a prolonged stretch. These splints are commonly used at the ankles, wrists, or elbows. Also recommended are appropriate strengthening programs for the weaker of the spastic muscles acting on a joint and an aerobic conditioning regimen. These interventions are supported by adjunctive measures that include antispasticity drugs (e.g., diazepam, baclofen, dantrolene, tizanidine), therapeutic botulinum toxin injections, and serial casting. The therapeutic goal is to improve the function of the individual in performing specific activities of daily living (i.e., feeding, dressing, bathing, hygiene, mobility) by improving the available joint range of motion, teaching effective compensatory strategies, and relieving pain.

Individuals with TM 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 shoe-laces 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.

Pain

Pain is common following transverse myelitis. 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 dysfunction of the spinal cord from the damage caused by

the inflammatory attack. Muscle pain might be treated with analgesics, such as acetaminophen (Tylenol), non-steroidal, anti-inflammatory drugs such as naproxen or ibuprofen (Naprosyn, Alleve, Motrin), or 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).

Nerve pain can be a significant challenge to find effective treatment. Nerve messages traveling through the damaged portion of the spinal cord may become scrambled and misinterpreted by the brain as pain. Besides the treatments listed above, certain antidepressants such as amitriptyline (Elavil), or anticonvulsants, such as carbamazepine, phenytoin, or gabapentin (Tegretol, Dilantin, Neurontin) may be helpful. Stress and depression should also be addressed since these conditions make pain harder to tolerate.

Depression

Individuals with TM should be educated about the effect of TM on mood regulation and routinely screened for the development of symptoms consistent with clinical depression. Warning signs that should prompt a complete evaluation for depression include failure to progress with rehabilitation and self-care, worsening fixed low mood, pervasive decreased interest, and/or social and professional withdrawal. A preoccupation with death or suicidal thoughts constitutes a true psychiatric emergency and should lead to prompt evaluation and treatment. Depression in TM is similar to the other neurologic symptoms patients endure, which are mediated by the effects of the immune system on the brain. Depression is remarkably prevalent in TM, occurring in up to 25% of those diagnosed at any given time, and is largely independent of the patient's degree of physical disability. Depression is not due to personal weakness or the inability to "cope." It can have devastating consequences; not only can depression worsen physical disability (such as fatigue, pain, and decreased concentration) but it can have lethal consequences. Suicide is the leading cause of death in TM. Despite the severity of the clinical presentation of depression in TM, there is a very robust response to combined aggressive psychopharmacologic and psychotherapeutic interventions. With appropriate recognition and treatment of TM depression, complete symptom remission is standard.

MYELITIS HELPLINE

For questions about our organization and rare neuro-immune disorders, such as acute flaccid myelitis, visit the **Myelitis Helpline**, a new online tool developed by the Transverse Myelitis Association.

myelitis.org/mhl

RESOURCE LIBRARY

To access up-to-date resources on acute flaccid myelitis, which include symposium videos, newsletters, podcast recordings, published literature summaries, information sheets and relevant external resources, visit our **Resource Library**.

myelitis.org/living-with-myelitis/resources/resource-library

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