CLINICIAN GUIDANCE: ACUTE FLACCID MYELITIS (AFM):

CURRENT SITUATION

As the winter season approaches, an increase in the circulation of respiratory pathogens, including enteroviruses, is expected. Enterovirus D-68 (EV-D68) is believed to be the main enterovirus responsible for the uncharacteristic increase in acute flaccid myelitis (AFM) cases observed during 2014, 2016, and 2018, which are referred to as peak years. Although there has been an increase in EV-D68 detections in the United States this year, the number of reported cases of AFM has remained relatively low, to date. As of November 1, 2024, the Centers for Disease Control and Prevention (CDC) has received 26 reports of suspected AFM, with 15 confirmed cases in 11 states. In past years, increases in EV-D68 respiratory disease have preceded cases of AFM by about 2 weeks. Therefore, vigilance for possible increases in EV-D68 respiratory disease and AFM is important.[1]

Acute flaccid myelitis (AFM) is a clinical syndrome characterized by flaccid limb weakness and abnormalities of the spinal cord gray matter on an MRI. AFM is a form of acute flaccid paralysis (AFP), which can have numerous varying etiologies. Prior to successful vaccination efforts, the most common cause of AFM worldwide was poliovirus; however, AFM may be caused by other viruses including West Nile virus, herpesviruses, and other viruses including non-polio enteroviruses.

It is imperative for clinicians to obtain whole stool samples from all patients with suspected AFM to rule out poliovirus infection. This is particularly important if the patient is under-vaccinated or unvaccinated against polio.[1] The identification of a paralytic polio case in an unvaccinated person in New York in 2022 reinforced the need to also consider polio in the differential diagnosis of patients with sudden onset of limb weakness.[1]

Reporting

All potential cases of AFM should be reported to the local public health authority based on the location of the healthcare provider. More information on where and how to report can be found here.

Carson City Health and Human Services (CCHHS)	Carson City, Douglas, and Lyon	(775) 887-2190 (24 hours)
Central Nevada Health District (CNHD)	Churchill, Mineral, Eureka, and Pershing	(775) 866-7535 (24 hours)
Northern Nevada Public Health (NNPH, formerly WCHD)	Washoe	(775) 328-2447 (24 hours)
Southern Nevada Health District	Clark	(702) 759-1300 (24 hours)
Nevada Division of Public and Behavioral Health	All other counties	(775) 684-5911 (M-F 8a.m. – 5 p.m.)

1. Centers for Disease Control and Prevention. *Remaining Vigilant for AFM/polio*, (September 18, 2024).
2. Centers for Disease Control and Prevention. *Enhanced AFM Surveillance*, (July 31, 2024).
3. Centers for Disease Control and Prevention. *Case Definitions for AFM,* (June 5, 2024).
4. Centers for Disease Control and Prevention. *Specimen Collection Instructions for Clinicians.* (June 5, 2024).
5. Centers for Disease Control and Prevention. *Clinical Overview of AFM.* (June 3, 2024).

(DPBH) Office of State Epidemiology (OSE)		(775) 400-0333 (after hours)

Clinical Overview of AFM

Introduction:

In 2014, the term acute flaccid myelitis was created to describe patients with sudden onset of acute flaccid limb weakness without a known cause and with lesions in gray matter of the spinal cord. It is important to note that there can also be some white matter involvement, and most cases occur in children.

Viral causes include:

- Non-polio enteroviruses (EV-D68, EV-A71)
- Flaviviruses (West Nile virus, Japanese encephalitis virus)
- Herpesviruses
- Adenoviruses

Please note: the clinical presentation is similar to poliomyelitis, but poliovirus has not been detected in any specimens from patients with AFM.

Potential cases of AFM reported to the local health authority are classified by public health personnel according to the following criteria:

Clinical Criteria:

Most patients had preceding febrile illness 1-2 weeks before onset of acute flaccid limb weakness. Patients meeting the following criteria should be considered a possible AFM case and reported by clinicians to their local health department:

- Respiratory or gastrointestinal illness (GI) with symptoms of fever, rhinorrhea, cough, vomiting or diarrhea. AND
- Rapid onset of weakness occurs within hours to a few days.
- Acute flaccid* weakness in one or more limbs and is more proximal than distal.
- Loss of muscle tone and reflexes in the affected limb(s).

**Low muscle tone, limp, hanging loosely, not spastic or contracted.*

It is important to watch for cranial nerve abnormalities which can present as:

- Facial or eyelid droop
- Difficulty swallowing or speaking
- Hoarse or weak cry

Some patients may complain about stiff neck, headache, or pain in the affected limb(s). In uncommon cases, people can also have numbness or tingling.

The most severe symptoms of AFM include:

- Respiratory failure, requiring mechanical ventilation

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- Serious neurologic complications such as body temperature changes and blood pressure instability that could be life threatening[5]

Differential Diagnosis of Flaccid Limb Weakness:

AFM can resemble:

- Synovitis
- Neuritis
- Limb injury
- Guillain-Barre syndrome (GBS)
- Transverse myelitis
- Stroke, including spinal stroke
- Tumor
- Acute cord compression
- Conversion disorder

AFM must be high on the differential diagnosis in late summer or early fall, especially in patients with preceding viral symptoms.

Careful neurological examination, laboratory testing, and MRI of the spine and brain can help guide diagnosis.

Testing and diagnosis:

During the initial evaluation for AFM:

- Collect patient's medical history
- Perform a physical exam along with an age-appropriate neurological assessment
- And ask questions to evaluate limb function impairment

Collect the patient's medical history:

- Collect information on any illness in the past 4 weeks
- Note respiratory and GI symptoms, with or without fever
- Ask about hand, foot, and mouth lesions (possible EV-A71 or similar viral infection)

Physical Exam:

Perform physical exam along with an age-appropriate neurological assessment and make sure to ask questions to evaluate limb function impairment.

Neurological examination should include documentation of:

- Muscle tone (flaccid/loose vs spastic/tight and firm)
- Muscle strength (full strength, move against gravity with some resistance/pressure, move against gravity but with no resistance/pressure, or little limb movement but not against gravity, no muscle movement at all)
- Reflexes in each extremity (hypo-, hyper, or absent)
- Any cranial nerve deficiencies such as for facial, palatal and shoulder asymmetry, hoarseness or hypophonia and dysphagia (if possible)

Note: Sensory exam is often normal in patients with AFM.

Assess the patient's ability to protect their airway:

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- Document respiratory sufficiency
- Negative inspiratory force may be used if the child is old enough and able to cooperate

Check for autonomic manifestations:

- Blood pressure lability
- Body temperature instability

Ask about additional signs and symptoms including:

- Difficulty holding their head up
- Decreased appetite or difficulty swallowing
- Increased sleepiness or inactivity
- Headache or neck. Shoulder, or back pain
    - Patients often complain of this prior or concurrent to weakness
- Pain in extremities
- Bowel or bladder changes, particularly constipation[5]

Laboratory/Imaging Criteria:

Confirmatory laboratory/imaging evidence:

- MRI showing spinal cord lesion with predominant gray matter involvement* and spanning one of more vertebral segments, AND
- Exclude persons with gray matter lesions in the spinal cord resulting from physician diagnosed malignancy, vascular disease, or anatomic abnormalities.

Presumptive laboratory/imaging evidence:

- MRI showing spinal cord lesion where gray matter involvement* is present but predominance cannot be determined, AND
- Exclude persons with gray matter lesions in the spinal cord resulting from physician diagnosed malignancy, vascular disease, or anatomic abnormalities.

Supportive laboratory/imaging evidence:

- MRI showing a spinal cord lesion in at least some gray matter* and spanning one or more vertebral segments, AND
- Excluding persons with gray matter lesions in the spinal cord resulting from physician diagnosed malignancy, vascular disease, or anatomic abnormalities.

**Spinal cord lesions may not be present on initial MRI: a negative or normal MRI performed within the first 72 hours after onset of limb weakness does not rule out AFM. Terms in the spinal cord MRI report such as "affecting mostly gray matter," "affecting the anterior horn or anterior horn cells," "affecting the central cord," "anterior myelitis," or "poliomyelitis" would all be consistent with this terminology.*

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Death Reporting Criteria:

- Any person whose death certificate lists acute flaccid myelitis as a cause of death of a condition contributing to death.
- Autopsy finds that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord spanning one or more vertebral segments.[3]

Laboratory Testing

Clinicians should collect specimens from patients with potential AFM as early as possible in the course of illness, preferably on the day of onset of limb weakness. Requested specimens should include:

- Cerebrospinal Fluid (CSF)
- Respiratory Nasopharyngeal (NP)/Oropharyngeal (OP) swab
- Blood serum
- Stool

For more detailed instructions visit the Specimen Collection Instructions for Clinicians webpage.

All submissions to CDC for diagnostic testing require pre-approval at this time. Do NOT ship specimens directly to the CDC. Contact your local health authority to assist with the coordination of a specimen shipment to CDC through the Nevada State Public Health Laboratory.

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