Acute Flaccid Myelitis (AFM)

Signs and symptoms	Most patients have febrile illness 1–2 weeks before onset of acute flaccid limb weakness, described as: • Rapid onset (hours to a few days) of loss of muscle tone and reflexes in one or more limbs Can be cranial nerve changes: facial or eyelid droop, trouble swallowing or speaking, cry hoarse or weak Often respiratory or gastrointestinal symptoms: fever, rhinorrhea, cough, vomiting or diarrhea. May be stiff neck, headache, or pain in the affected limb(s), uncommonly numbness or tingling. The most severe symptoms of AFM are: • Respiratory failure, requiring mechanical ventilation • Serious neurologic complications such as body temperature changes and blood pressure instability that could be life threatening
Case classification	Illness meeting any of the criteria below should be considered a possible AFM case and reported: • Clinical **AND** laboratory/imaging criteria for reporting, **OR** • Death certificate listing AFM as the cause of death or a contributing cause of death, **OR** • Autopsy findings that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord.
	Clinical Criteria • An illness with onset of acute flaccid* weakness of one or more limbs, AND • Absence of a clear alternative diagnosis attributable to a nationally notifiable condition ** Low muscle tone, limp, hanging loosely, not spastic or contracted.* **Laboratory/imaging evidence:** • MRI showing spinal cord lesion with at least some gray matter involvement 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
Differential diagnosis	Other viruses, synovitis, neuritis, limb injury, Guillain-Barre syndrome (GBS), transverse myelitis, stroke (including spinal stroke), tumor, acute cord compression, conversion disorder. Consider a diagnosis of AFM in late summer or early fall, especially in patients with preceding viral symptoms.
Treatment	Supportive, with no specific treatment for AFM or poliomyelitis. Clinicians should expedite neurology and infectious disease consultations to discuss treatment and management considerations.
Exposure	Person-to-person fecal-oral transmission
Laboratory testing	The provider should order viral respiratory and viral stool cultures locally. CDC requires consultation to test. Submit specimens to PHL for forwarding to CDC for testing and typing enterovirus, rhinovirus, and poliovirus. Collect as soon as possible: • Two stool specimens collected 24 hours apart (minimum 1 gram in sterile container) • CSF (minimum 0.15 mL, spun and processed, in cryovial) • Serum collected at same time as CSF if possible (0.5 mL minimum, spun and processed) • Nasopharyngeal or oropharyngeal swab (synthetic swab with synthetic shaft, in VTM) • If fatal, fresh-frozen tissue or fixed tissue (formalin 3 days then in 100% ethanol, **room temp**.) **Except for fixed tissue,** freeze all specimens, ship frozen on dry ice according to PHL requirements: https://doh.wa.gov/public-health-provider-resources/public-health-laboratories/lab-test-menu
Public health actions	• Assess the likelihood of AFM and polio: confirm compatible clinical symptoms, obtain history of recent respiratory or GI illness, verify vaccination and travel history, and review test results (i.e., CSF and MRI results). For a suspected polio case, please refer to the Polio guideline. • If laboratory testing is indicated, facilitate timely collection/transport of appropriate specimens. • There is no specific management for contacts of AFM cases, but they should be educated regarding any specific etiology suspected and advised about when to seek medical care. See Appendix A for Case Investigation Checklist

Acute Flaccid Myelitis (AFM)

1. DISEASE REPORTING

A. Purpose of Reporting and Surveillance

1. To identify cases of Acute flaccid myelitis (AFM) and establish an incidence baseline and burden of the condition in Washington State.
2. To help identify causes of AFM in the United States.
3. To understand the impact of AFM among all age groups.
4. To establish and maintain a surveillance system sensitive enough to reliably detect cases of paralytic poliomyelitis.

B. Legal Reporting Requirements

1. **Health care providers and Health care facilities:** notifiable to **local health jurisdictions** within 24 hours
2. **Laboratories:** no requirements for reporting

C. Local Health Jurisdiction Investigation Responsibilities

1. Begin routine case investigation within one working day of notification. If polio is suspected as the cause of AFM begin investigation and notify Office of Communicable Disease Epidemiology (CDE) immediately.
2. Facilitate the transport of specimens to Washington State Public Health Laboratories (PHL) at the direction of CDE and CDC. CDC requires consultation before testing.
3. Implement appropriate infection control measures.
4. Report all *confirmed* and *probable* cases of AFM (see Section 3) to CDE.
    - For AFM, complete the CDC-AFM patient summary form (available at https://www.cdc.gov/acute-flaccid-myelitis/hcp/data-collection.html and send a copy to CDE. This form MUST accompany any specimens submitted to CDC for testing.
    - For polio, complete the polio investigation form (available at http://www.doh.wa.gov/Portals/1/Documents/5100/210-059-ReportForm-Polio.pdf)
5. Enter into the Washington Disease Reporting System (WDRS) as "Acute Flaccid Myelitis (AFM)/Poliomyelitis"

2. THE DISEASE AND ITS EPIDEMIOLOGY

Background

AFP is an umbrella term that includes multiple clinical entities such as poliomyelitis (polio), AFM, Guillain-Barré syndrome, acute transverse myelitis, toxic neuropathy, muscle disorders, and other conditions. Acute flaccid myelitis (AFM) is characterized by

rapid onset of flaccid weakness in one or more limbs and distinct abnormalities of the spinal cord gray matter on magnetic resonance imaging (MRI). Note that 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. AFM is a subtype of a more general condition called acute flaccid paralysis (AFP). AFP is defined as acute onset of flaccid weakness absent of features suggesting an upper motor neuron disorder.

AFM has been tracked since 2014 when the United States recorded the first increase in AFM cases, with two more increases in 2016 and 2018. These increases are thought to be caused by enterovirus D68 (EV-D68). The increase in 2014 coincided with a national outbreak of severe respiratory illness caused by EV-D68, and increased circulation of EV-D68 was observed in 2016 and 2018. There are other etiologies for AFM. Current national AFM case activity is available at: https://www.cdc.gov/acute-flaccid-myelitis/cases-in-us.html

Providers should consider a diagnosis of AFM in late summer or early fall, especially in patients with preceding viral symptoms. AFM is difficult to distinguish clinically from paralytic polio, so stool specimens from all suspected AFP and AFM patients are tested for poliovirus, particularly a case with anterior myelitis, to ensure that any case of poliovirus is quickly identified and investigated. If poliovirus is detected, the case is considered polio, not AFM.

Despite extensive laboratory testing, in many cases a definitive cause for AFM cannot be identified. When a specific pathogen is isolated, it may still be difficult to attribute causality to that pathogen, especially if the virus was not isolated from CSF. Please note: If another notifiable condition is suspected or confirmed to be the cause of AFM, consult the DOH list of notifiable conditions.

A. Etiologic Agent

AFM is most often associated with viruses that can cause inflammation and loss of motor and autonomic neurons located in the anterior horn cells of the spinal cord (i.e., front column of gray matter in the spinal cord). Viruses that have been associated with AFM include: Enteroviruses; West Nile virus (WNV) and other flaviviruses (Japanese encephalitis virus and Saint Louis encephalitis virus); Herpesviruses such as cytomegalovirus and Epstein-Barr virus; and Adenoviruses.

B. Description of Illness

AFM symptoms are similar regardless of the etiologic agent and usually include paralysis, but sensation is spared. Most patients with AFM have sudden onset of limb weakness and loss of muscle tone and reflexes. In addition to the limb weakness, some may also have one or more of the following symptoms: facial droop or weakness, difficulty moving the eyes, drooping eyelids, difficulty with swallowing, or slurred speech. For clinical guidance see: https://www.cdc.gov/acute-flaccid-myelitis/hcp/clinicians-health-departments/clinical-presentation.html.

Numbness or tingling (paresthesia) is rare in patients with AFM, though there can be pain in the arms or legs. Some patients have urinary retention. The most severe symptom of AFM is respiratory failure if the muscles involved with breathing, such as the diaphragm, become weak. This can require urgent ventilator support.

C. AFM in Washington

Washington has had cases reported since 2014, ranging from 0 to 11 cases a year.

D. Reservoirs

Multiple infectious agents associated with AFM are spread person to person. The syndrome can also be caused by trauma, toxins, or other causes (see Appendix B).

E. Modes of Transmission

AFM is a complication of some infections. Mode of transmission depends on the agent.

F. Incubation Period

The incubation period depends on the agent.

G. Period of Communicability

Although the underlying cause may be communicable, AFM is a is a rare outcome of such infections. Recommendations for management of patients with AFM are standard, contact and droplet precautions, consistent with CDC recommendations (see: https://www.cdc.gov/non-polio-enterovirus/hcp/ev-d68-hcp.html). There are no pathogen-specific recommendations currently except for polio.

H. Treatment

There are no FDA-approved drugs or biologics to treat or prevent AFM. Early symptoms of AFM can be subtle and may resemble neurologic diseases with targeted treatments. Based on the available evidence:

- There is no indication that any specific targeted treatment should be either preferred or avoided in the acute medical treatment of AFM.
- There is currently no targeted treatment with enough evidence to endorse or discourage use for the treatment or management of AFM.
- There continues to remain a paucity of prospective clinical trials on treatments for AFM. Current sources are limited to case reports and case-series. Acute treatments that have been used frequently in patients with AFM include intravenous immunoglobulin, corticosteroids, and/or therapeutic plasma exchange.
- Clinicians should expedite neurology and infectious disease consultations to discuss treatment and management considerations.

For details see: Acute Flaccid Myelitis: Interim considerations for clinical management. Centers for Disease Control and Prevention. Available at https://www.cdc.gov/acute-flaccid-myelitis/hcp/clinical-management.html#summary-guidance.

3. CASE DEFINITION

A. Clinical Criteria

- An illness with onset of acute flaccid* weakness of one or more limbs, AND
- Absence of a clear alternative diagnosis attributable to a nationally notifiable condition**

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

** Cases with a clear alternative diagnosis attributable to a nationally notifiable condition should be reported only once as that notifiable condition to avoid duplicate reporting.

B. Laboratory/Imaging Criteria

Confirmatory evidence:

- MRI showing spinal cord lesion with predominant gray matter involvement† 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.

Presumptive evidence:

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

Supportive 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.

† 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.

Note: The categorical labels used here to stratify laboratory/imaging evidence are intended to support the standardization of case classifications for public health surveillance, and should not be used to interpret the utility or validity of any laboratory/imaging test methodology.

C. Other Criteria

- Autopsy findings that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord 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, AND
- Absence of a clear alternative diagnosis attributable to a nationally notifiable condition.**

** Cases with a clear alternative diagnosis attributable to a nationally notifiable condition should be reported only once as that condition to avoid duplicate reporting.

D. Case Classification (2022)

Cases meeting case ascertainment criteria that are reported by the health department to CDC are classified by a CDC panel of experts as:

Suspect:

Meets clinical criteria with supportive laboratory/imaging evidence, **AND**

Available information is insufficient to classify case as probable or confirmed.

Probable:

Meets clinical criteria with presumptive laboratory/imaging evidence.

Confirmed:

Meets clinical criteria with confirmatory laboratory/imaging evidence, **OR**

Meets other classification criteria.

To provide consistency in case classification, experts in national AFM surveillance review case information and assign final case classification for all patients under investigation for AFM. This parallels the review for final classification of paralytic polio cases.

E. Case Ascertainment

In addition to the above, illness that meets any of the following criteria should be considered a possible AFM case and should be reported:

- A person meeting clinical AND laboratory/imaging criteria for reporting, OR
- A person whose death certificate lists AFM as the cause of death or a contributing cause of death, **OR**
- A person with autopsy findings that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord

4. DIAGNOSIS AND LABORATORY SERVICES

The clinical presentation (e.g., the location of the weakness, muscle tone, and reflexes) helps differentiate patients with AFM from patients with other forms of acute flaccid paralysis. Magnetic resonance imaging (MRI) is needed for diagnosing cases of AFM.

Clinicians treating patients who meet the AFM case ascertainment criteria should expedite neurology and infectious disease consultations to discuss treatment and management considerations and should pursue laboratory testing for enteroviruses, West Nile virus, and other known infectious etiologies at their usual clinical and reference laboratories. Clinicians may contact the local health jurisdiction for assistance with any testing that is not available locally. Specimens should not be shipped to WA DOH without first consulting with the local health jurisdiction.

A. Laboratory Diagnosis

A CDC neurologist reviews all available clinical, laboratory, and neuroimaging information to confirm AFM on reported cases. For all suspected AFM cases, CSF, serum, a nasopharyngeal or oropharyngeal swab, and stool samples should be collected. PHL does not do enterovirus testing but will forward specimens to CDC to test for enteroviruses and other pathogens. CDC can do typing to confirm EV-D68 if enterovirus is detected.

The laboratory diagnosis of AFM etiologic agents can be made by isolation or detection of an agent from stool, nasopharyngeal/oropharyngeal swab, serum, or CSF; however, enteroviruses are most likely to be detected in **stool cultures**. To rule out poliovirus in a suspect case, a provider should order a viral stool culture locally as well as a viral respiratory culture locally.

B. Tests Available at Washington State Public Health Laboratories (PHL)

PHL does not routinely perform testing for patients with suspected AFM, but send the recommended surveillance specimens for testing at CDC. Specimens intended for testing at CDC must be routed through PHL. More detailed instructions regarding collection and shipping of specimens to be sent to CDC can be found at https://www.cdc.gov/acute-flaccid-myelitis/hcp/specimen-collection.html.

Note that PHL requires all clinical specimens have two patient identifiers, a name **and** a second identifier (e.g., date of birth) both on the specimen label and on the submission form. Due to laboratory accreditation standards, specimens will be rejected for testing if not properly identified. Also include specimen source and collection date. The CDC AFM Patient Summary Form must accompany any specimens submitted for suspected AFM patients, including patients with suspected poliomyelitis: https://www.cdc.gov/acute-flaccid-myelitis/hcp/data-collection.html, and a PHL form: https://doh.wa.gov/public-health-provider-resources/public-health-laboratories/lab-test-menu.

C. Specimen Collection

Testing is done at CDC, and requires consultation to approve testing. If the diagnosis of AFM is being considered, early specimen collection has the best chance to find an etiology for AFM. As soon as possible in the course of illness, preferably on the day of onset of limb weakness, clinicians should collect specimens from patients suspected of having AFM:

1. Two stool specimens collected as soon after onset of limb weakness and separated by 24 hours, minimum 1gram, freeze.
2. Cerebrospinal fluid (CSF), minimum 1mL (collected at the same time, or within 24 hours of serum if feasible), freeze.
3. Serum, minimum 0.4 mL (collected at the same time, or within 24 hours of CSF if feasible), freeze.
4. Nasopharyngeal or oropharyngeal swab, stored in minimum 0.5 ml, 1mL preferred. Store in viral transport media, freeze. Also contact the lab that completed initial viral testing on the first NP/OP swab collected and request that it be frozen and submitted to PHL. This will be sent for additional testing at CDC.

In the event of a death, please send the following samples, if possible:

- Fresh frozen tissue, rapidly frozen to -70º F and shipped on dry ice
- Formalin-fixed or formalin-fixed, paraffin embedded tissue
    - Avoid prolonged fixation – fix in formalin for 3 days then transfer to 100% ethanol, store at room temperature in carriers to prevent breakage, do **NOT** freeze

All available tissue specimens may be of interest for testing at CDC. Tissue, particularly spinal tissue, is important for helping determine whether the patient had AFM. Communicable Disease Epidemiology will help facilitate tissue submissions to CDC.

5. ROUTINE CASE INVESTIGATION

Any person noted to have AFM could be a polio case. For every suspect AMF case it is extremely important to immediately obtain information about polio immunizations, recent travel, or exposure to travelers or somebody recently receiving oral polio vaccine (OPV).

A. Evaluate the Diagnosis

Determine the likelihood of the diagnosis:

- Review clinical presentation and physical exam findings (particularly flaccid weakness).

- Review immunization history and risk factors for infection (contact with a case of AFM, travel to an endemic area, possible exposure to somebody who recently received oral polio vaccine).
- Obtain history of any recent viral respiratory and/or gastrointestinal illness.
- Confirm that AFM clinical criteria including CSF findings and/or MRI test results are met.
- If pursuit of laboratory testing is indicated, facilitate timely collection of appropriate specimens and expedite transport of those specimens to DOH Washington State Public Health Laboratories (PHL).
- If a commercial laboratory isolates polio virus in cell culture, request that the laboratory send the cell culture to PHL for confirmatory testing immediately.

B. Identify Source of Infection

Ask about the following exposures in the 3–35 days prior to onset:

- Contact with person with similar symptoms or an AFM diagnosis
- Travel to an area with recent travel to an area with wild or vaccine strain polio cases
- Exposure to a person who recently received oral polio vaccine

C. Identify Potentially Exposed Persons

For non-polio causes of AFM, contact investigation is not usually recommended but may be considered on a case-by-case basis.

If poliovirus is confirmed as the cause for AFM, DOH and the Centers for Disease Control and Prevention will assist with an extensive contact investigation. Refer to Polio guideline if poliovirus is confirmed as the cause for AFM.

6. CONTROLLING FURTHER SPREAD

A. Infection Control Recommendations / Case Management

Standard, contact and droplet precautions are recommended for hospitalized case-patients. This is consistent with CDC's recommendations for EV-D68 (https://www.cdc.gov/non-polio-enterovirus/hcp/ev-d68-hcp.html). There are no pathogen-specific recommendations to add at this time.

B. Contact Management

No specific management for contacts of AFM cases is indicated. Contacts should be educated regarding the specific etiology if an agent is suspected (e.g., vectors for arbovirus, or lack of symptoms or non-specific symptoms associated with uncomplicated enterovirus infection) and advised about when they should seek medical care.

7. MANAGING SPECIAL SITUATIONS

Special situations related to AFM will be handled on a case-by-case basis by CDE.

Prevention Recommendations

Control of polio is accomplished through immunization. Unimmunized persons at risk of exposure, for example during travel to areas with known polio cases or areas where OPV is

used, should maintain strict prevention measures to avoid potential fecal-oral transmission. These include using good hand washing techniques and safe drinking water during travel to areas with endemic wild or vaccine type polio, and maintaining good hygiene practices if in contact with infants who are receiving oral vaccine.

To avoid mosquito-borne viruses that can cause AFM, use mosquito repellents and stay indoors at dusk and dawn, which is the prime period that mosquitoes bite. Remove standing or stagnant water from nearby areas to minimize mosquitoes breeding is also recommended.

Wash hands regularly with soap and water, avoid close contact with sick people, and clean surfaces that a sick person has touched to reduce spread of other known causes of AFM.

ACKNOWLEDGEMENTS

This document is a revision of the Washington State Guidelines for Notifiable Condition Reporting and Surveillance published in 2002 which were originally based on the Control of Communicable Diseases Manual (CCDM), 17th Edition; James Chin, Ed. APHA 2000. We would like to acknowledge the Oregon Department of Human Services for developing the format and select content of this document.

UPDATES

January 2010 Section 1: The investigation form link was updated and 2010 case classification information was added.

January 2011: The Legal Reporting Requirements section has been revised to reflect the 2011 Notifiable Conditions Rule revision.

October 2016: AFM guidance and polio guideline were combined. Global epidemiology data was updated. Information regarding trivalent OPV vaccine switch to bivalent OPV vaccine in low and middle income countries was included.

October 2018: The AFM case definition was updated to reflect the June 2017 CSTE revision. Specimen collection instructions for AFM were updated to reflect the latest CDC recommendations. Global epidemiology for polio and national and state-wide epidemiology data for AFM was updated.

December 2022: For 2023 WAC revision combined provider and facility reporting requirement, updated laboratory submission (Section 1B)

April 2023: AFM and Poliomyelitis guidelines separated. AFM Case Definition updated to reflect 2021 CSTE revision, links and references updated.

December 2023: Updated laboratory submission process for 2024 WAC revision.

June 2024: CDC links updated.

APPENDIX A: CASE INVESTIGATION CHECKLIST

- ☐ Confirm the clinical presentation of the patient.
- ☐ Notify WA DOH of suspect case of AFM at 206-418-5500
- ☐ Assess vaccination status for polio and risk of poliovirus exposure (e.g., unvaccinated, or under-vaccinated, travel to polio-endemic area or contact with someone with travel to polio-endemic area, contact with recent OPV vaccinee)
    - Consult polio guideline for suspected polio
- ☐ Ascertain what testing has been done, including lab testing, lumbar puncture, and MRI
- ☐ Ask the treating physician, preferably the neurologist, to complete the https://www.cdc.gov/acute-flaccid-myelitis/downloads/patient-summary-form.pdf
- ☐ Submit the Acute Flaccid Myelitis: Patient Summary Form to WA DOH.
    - CDC also requires a copy of the History & Physical (H&P), MRI report, MRI images, Neurology consult notes, EMG report (if done), Infectious disease consult notes (if available), vaccination records, and diagnostic laboratory reports for patients reported with suspect AFM.
        - MRI images are not required to be sent at the time of initial Patient Summary Form and medical record information.
        - In the event of a death, copies of the hospital discharge summary, death certificate, and autopsy report should also be sent to WA DOH.
- ☐ Collect specimens within 24 hours of onset of limb weakness (or as early in course of illness as possible), and submit to WA State Public Health Laboratories. CDC has requested LHJs and providers do not submit directly to CDC. (See Table 2. below for detailed specimen collection and shipping guidance).
    - WA DOH will forward appropriate specimens to CDC for testing.
- ☐ Submit the requested clinical information (i.e., H&P, MRI report, neurology consult notes) to WA DOH either as an attachment in the WDRS event record or via secure email
- ☐ Submit MRI images via secure electronic imaging platform as directed by WA DOH.
- ☐ Complete 60 Day Follow Up section of Acute Flaccid Myelitis: Patient Summary Form and submit to WA DOH

Control Measures

Standard, contact, and droplet precautions are recommended for hospitalized case-patients. This is consistent with CDC's recommendations for EV-D68. There are no pathogen-specific recommendations to add at this time.

Exclusion

Anyone with a fever should be excluded from work or school until 24 hours have passed fever-free without the use of an anti-fever medication. Anyone with diarrhea should be excluded from work or school until 24 hours have passed diarrhea-free without the use of an anti-diarrheal medication. If the etiology is determined, there may be additional exclusion criteria that apply.

APPENDIX B: MOST COMMON ETIOLOGIES FOR ACUTE FLACCID PARALYSIS AND ACUTE FLACCID MYELITIS