Utah Department of Health & Human Services

Acute flaccid myelitis (AFM)

Disease plan

Quick links

Last updated: August 18, 2022 by Jared Ripplinger

Questions about this disease plan?

Contact the Utah Department of Health and Human Services Office of Communicable Diseases: 801-538-6191.

Acute flaccid myelitis critical clinician information

Clinical evidence

Signs/symptoms
• Most common:
  ○ Arm or leg weakness
  ○ Loss of muscle tone and reflexes
• Less common:
  ○ Difficulty moving the eyes or drooping eyelids
  ○ Facial droop or weakness
  ○ Difficulty swallowing or slurred speech
  ○ Pain in arms or legs
  ○ Pain in neck or back

Period of communicability • Under investigation; etiologic agent(s) not yet determined
Incubation period • Under investigation; etiologic agent(s) not yet determined
Mode of transmission • Under investigation; possibly spread by the fecal-oral route, respiratory droplets, or nasopharyngeal secretions
Laboratory testing
Type of lab test • Identification/typing of enteroviruses and rhinoviruses • Poliovirus rule-out stool sample testing
Type of specimens • CSF • Serum • Stool • Nasopharyngeal swab
Treatment recommendations
Type of treatment • There is no indication that any specific targeted treatment should be either preferred or avoided in the acute medical treatment of AFM • There are no currently approved drugs or biologics for AFM • Emphasis of clinical treatment is on supportive care • Up to date information on AFM treatments can be found at https://www.cdc.gov/acute-flaccid-myelitis/hcp/clinical-management.html
Time period to treat • Clinicians should immediately admit patients to the hospital because AFM can progress rapidly and require urgent medical intervention, such as assistance with breathing.

Why is acute flaccid myelitis important to public health?

Acute flaccid myelitis (AFM) is a rare but serious neurological syndrome that causes muscle weakness and can sometimes result in permanent paralysis. Cases have been observed since 2014, with peaks in cases in 2014, 2016, and 2018 and are associated with national outbreaks of enterovirus D68 (EV-D68), which typically causes a mild respiratory infection. There was not an AFM outbreak in 2020, as was expected, likely due to social distancing, mask wearing, and handwashing practices adopted due to the COVID-19 pandemic. AFM consists of sudden onset weakness in 1 or more limbs with inflammation of the gray matter in the patient's spinal cord. The Centers for Disease Control and Prevention (CDC) started tracking AFM cases in August of 2014. Since that time, 642 cases have been confirmed in 49 states and the District of Columbia as of October 30, 2020. More than 90% of cases have been in young children.

Disease and epidemiology

Clinical description

Most people with AFM experience rapid onset of weakness in 1 or more limbs and/or a loss of muscle tone and reflexes. Some people with AFM also experience difficulty moving their eyes or drooping eyelids, facial weakness or droop, difficulty swallowing or slurred speech, pain in the arms or legs, and/or pain in the neck or back.

Causative agent

Although causes of related neurologic illnesses with limb weakness have been identified (including viral infections, environmental toxins, genetic disorders, and Guillain-Barre syndrome), specific causes of AFM are still under investigation. AFM cases spiked in 2014, 2016, and 2018 from August to November and coincided with national outbreaks of mild respiratory illnesses among children caused by enterovirus D68 (EV-D68). It is unclear whether the biennial pattern of outbreaks observed between 2014 and 2018 will return. That pattern was not observed in 2020, likely due to public health measures adopted during the COVID-19 pandemic.

EV-D68, coxsackievirus A16, and enterovirus A71 (EV-A71) have been detected in the spinal fluid of a small number of patients with AFM lending evidence they may have a causative role in the development of AFM, but a cause for AFM has not yet been determined. AFM is a syndrome, so it is possible multiple pathogens cause AFM. CDC continues to investigate possible causes of AFM in cooperation with state and local health departments.

Differential diagnosis

AFM symptoms are sometimes similar to illnesses caused by enteroviruses, adenovirus, West Nile virus, and herpes viruses. Poliomyelitis, which can be similar in presentation to AFM, is caused by poliovirus but has been eliminated from most of the world. Poliovirus rule-out testing is conducted before a case can be classified as AFM.

Laboratory identification

Viral laboratory testing and reporting is performed at the CDC. Instructions for specimen collection are found here. Clinicians should collect specimens from patients under investigation (PUIs) for AFM as early as possible in the course of illness, preferably on the day of onset of limb weakness. Early specimen collection has the best chance to yield a cause of AFM. Specimens include cerebrospinal fluid (CSF), serum, stool (2 specimens collected 24 hours apart), and a nasopharyngeal (NP) swab. Specimen submission to CDC must be coordinated by the Utah Department of Health and Human Services (DHHS) Office of Communicable Diseases, and sent to CDC through the Utah Public Health Laboratory.

Treatment

There are no approved treatments for AFM. Clinical management focuses on supportive care and the patient's clinical presentation. Ninety-eight percent of AFM patients (as of February 2021) required hospitalization because patient health can decline quickly, resulting in paralysis or the need for a ventilator. AFM can also lead to permanent disability. Information on clinical treatment for AFM can be found on the CDC website. Clinicians can schedule a consultation with a neurologist who specializes in AFM through the AFM Physician Consult and Support Portal.

Case fatality

Since AFM surveillance began in 2014, 2 patients in the acute phase of AFM have died; 1 in 2017 and 1 in 2020, out of a total of 645 confirmed cases in the United States, for a case fatality rate of 0.3%.

Reservoir

AFM is a syndrome, and the etiologic agent(s) is/are still under investigation.

Transmission

CDC is actively researching the cause of AFM in close cooperation with state and local health departments because the transmission of AFM has not yet been determined. However, associations between increases in AFM cases and the circulation of enteroviruses such as

enterovirus D68 (EV-D68) and enterovirus A71 (EV-A71) suggest these non-polio enteroviruses may have a role in causing AFM. AFM could potentially spread through the fecal-oral route, respiratory droplets, or nasopharyngeal secretions. The lack of a spike in AFM cases in 2020 suggests interventions such as mask wearing, social distancing, and handwashing could potentially prevent AFM cases as well.

Susceptibility

All humans are thought to be susceptible to AFM and its associated etiologic agent, but most cases have occurred in young children.

Incubation period

An incubation period for AFM has not yet been determined.

Period of communicability

A period of communicability for AFM has not yet been determined.

Epidemiology

There were 16 confirmed cases of AFM in Utah from 2014 through 2021.

Public health control measures

Public health responsibility

Public health's responsibility in regards to AFM is twofold:

- **Early detection**. Public health should monitor respiratory diseases in collaboration with clinical partners and communicate with clinicians to identify persons with compatible illness to ensure complete testing, and ascertainment of potential etiologies, is performed.
- **Rapid assessment and response.** Public health should respond to suspected cases quickly to provide recommendations to providers and coordinate specimen testing at CDC. Public health should continue to report suspected cases promptly to CDC, even when laboratory specimen collection and/or MRI results are still pending, and work with other government agencies to investigate the potential etiology.

Prevention

More than 90% of AFM cases have been observed in patients with mild respiratory symptoms or fever consistent with a recent viral infection. Public health recommends preventive measures for

viral illness in general. These actions include handwashing, not touching the face with unwashed hands, and avoiding contact with those that are sick.

Vaccine

No vaccine is currently available for the prevention or prophylactic treatment of AFM.

Isolation and quarantine

Interim infection control recommendations for healthcare professionals are the same as CDC's infection control recommendations for EV-D68. These precautions include standard, contact, and droplet precautions.

Case investigation

Reporting

AFM is a notifiable condition in Utah. Any potential cases of AFM should be reported to public health within 3 working days of identification.

Report any illness to public health authorities that meets the following criteria:

- Meets clinical criteria for reporting **and** laboratory/imaging criteria for reporting, **or**
- Meets vital records criteria for reporting, **or**
- Meets other criteria for reporting.

Clinical criteria for reporting

- A person with onset of acute flaccid* limb weakness.**

  * Low muscle tone, limp, hanging loosely, not spastic or contracted.
  ** Clinical criteria must be paired with laboratory/imaging criteria to trigger a report to public health.

Laboratory/imaging criteria for reporting

- A magnetic resonance image (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 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.

** Laboratory/imaging must be paired with clinical criteria to trigger a report to public health.

Epidemiologic linkage criteria for reporting

Not applicable.

Vital records criteria for reporting

Any person whose death certificate lists acute flaccid myelitis as a cause of death or a condition contributing to death.

Other criteria for reporting

- Autopsy findings that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord spanning 1 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

Disease-specific data elements to be included in the initial report

Disease-specific data elements to be included in the initial report are listed below.

- Basic demographics
- Clinical information
    - Date of onset
    - Limb(s) with acute onset of weakness
        - Description of limb weakness: limb(s) affected; weakness symmetric or asymmetric
        - Cranial nerve involvement (e.g., extraocular movement abnormalities, facial weakness)
        - Reflexes and tone (flaccid* or spastic) in affected limbs
    - Hospitalization (include duration)
- Laboratory/imaging data:
    - Date(s) of lumbar puncture(s) (LP)
    - WBC count from CSF (cells / mm3)
    - Protein level in CSF (mg/dL)
    - Date of performance of MRI (if >1 MRI performed, date of each MRI study)†
    - Description of gray matter lesion(s) (may attach MRI report)

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

†Restricted to MRIs performed in the proximate period of the suspected AFM illness; excludes neuroimaging performed for illnesses unrelated (clinically or temporally) to AFM illness.

Criteria to determine if a case should be reported to public health authorities

Criterion	Acute flaccid myelitis (AFM)		
Clinical criteria for reporting			
Acute flaccid* weakness of 1 or more limbs		N	
Laboratory/imaging criteria for reporting			
A magnetic resonance image (MRI) showing spinal cord lesion in at least some gray matter† and spanning 1 or more vertebral segments		N	
Excluding persons with gray matter lesions in the spinal cord resulting from physician-diagnosed malignancy, vascular disease, or anatomic abnormalities		N	
Vital records criteria for reporting			
Any person whose death certificate lists acute flaccid myelitis as a cause of death or a condition contributing to death	S		
Other criteria for reporting			
Autopsy findings that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord spanning 1 or more vertebral segments			N
Excluding persons with gray matter lesions in the spinal cord resulting from physician-diagnosed malignancy, vascular disease, or anatomic abnormalities			N

Notes:

S = This criterion alone is sufficient to report a case.

N = All "N" criteria in the same column are necessary to report a case.

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

† 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.

Case definition (2021)

Acute flaccid myelitis

Clinical criteria

- An illness with onset of acute flaccid* weakness of 1 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 (NNC) should be reported only once using the event code for the NNC to avoid duplicate reporting.

Laboratory/imaging criteria

Confirmatory laboratory/imaging evidence:

- MRI showing spinal cord lesion with predominant gray matter involvement† and spanning 1 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 laboratory/imaging 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 laboratory/imaging evidence:

- MRI showing a spinal cord lesion in at least some gray matter† and spanning 1 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 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. The categorical labels should not be used to interpret the utility or validity of any laboratory/imaging test methodology.

Epidemiologic linkage

Not applicable.

Other classification criteria

- Autopsy findings that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord spanning 1 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 (NNC) should be reported only once using the event code for the NNC to avoid duplicate reporting.

Case classification

Confirmed:

- Meets clinical criteria with confirmatory laboratory/imaging evidence, **or**
- Meets other classification criteria.

Probable:

- Meets clinical criteria with presumptive laboratory/imaging evidence.

Suspect:

- Meets clinical criteria with supportive laboratory/imaging evidence, **and**
- Available information is insufficient to classify a case as probable or confirmed.

Comment: To provide consistency in case classification, review of case information and assignment of final case classification for all suspected AFM cases will be done by experts in national AFM surveillance. This is similar to the review required for final classification of paralytic polio cases.

Criteria for defining a case of acute flaccid myelitis

Criterion	Suspect	Probable	Confirmed	
Clinical evidence				
Acute flaccid* weakness of 1 or more limbs	N	N	N	
Absence of a clear alternative diagnosis attributable to a nationally notifiable condition**	N	N	N	
Laboratory/imaging evidence				
MRI showing spinal cord lesion with predominant gray matter involvement† and spanning 1 or more vertebral segments			N	
MRI showing spinal cord lesion where gray matter involvement† is present but predominance cannot be determined		N		
MRI showing spinal cord lesion in at least some of the gray matter† and spanning 1 or more vertebral segments	N			
Excluding persons with gray matter lesions in the spinal cord resulting from physician-diagnosed malignancy, vascular disease, or anatomic abnormalities	N	N	N	
Other evidence				
Autopsy findings that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord spanning 1 or more vertebral segments				N
Excluding persons with gray matter lesions in the spinal cord resulting				N

from physician-diagnosed malignancy, vascular disease, or anatomic abnormalities				
Absence of a clear alternative diagnosis attributable to a nationally notifiable condition**				N
Insufficient information to classify case as probable or confirmed	N			

Notes:
S = This criterion alone is sufficient to classify a case.
N = All "N" criteria in the same column are necessary to classify a case. A number following an "N" indicates that this criterion is only required for a specific disease/condition subtype (see below). If the absence of a criterion (i.e., criterion NOT present) is required for the case to meet the classification criteria, list the absence of criterion as a necessary component.

* Low muscle tone, limp, hanging loosely, not spastic or contracted.
** Cases with a clear alternative diagnosis attributable to a nationally notifiable condition (NNC) should be reported only once using the event code for the NNC to avoid duplicate reporting.
[†]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.

Case investigation process

The case investigation process is as follows:

- Upon notification of a potential case, the public health investigator* or AFM coordinator** contacts the hospital infection preventionist and/or clinician and enters the potential case into UT-NEDSS (EpiTrax).
- The AFM coordinator and/or local health department (LHD) will coordinate specimen collection and shipment to the Utah Public Health Laboratory (UPHL). The specimen submission form (CDC Form 50.34) must accompany specimens to UPHL.
- The public health investigator, infection preventionist, or patient's clinician will complete the Patient Summary Form, and send it to the AFM coordinator.
- The AFM coordinator notifies CDC of the suspect case, informs them of the expected sample delivery date, and sends them the Patient Summary Form.
- The CDC sends test results to UPHL and the AFM coordinator.
- The AFM coordinator sends test results and CDC's case classification to the public health investigator.

- The public health investigator updates the case in UT-NEDSS (EpiTrax) and sends test results and case classification to the clinician.
- The AFM coordinator communicates with the CDC for additional instructions or changes in the case status.

*Public health investigator: Local health department staff responsible for the case investigation
**AFM coordinator: Utah Department of Health and Human Services AFM epidemiologist responsible to coordinate reporting, specimen collection, and communication with CDC. If you are unsure who this is, call 801-538-6191 to verify.

Outbreaks

Not yet determined.

Identification of case contacts

Not yet determined.

Case contact management

Not yet determined.

References

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Version control

V. 11.15—Created disease plan.

V. 02.21—Added Critical Clinician Information section, updated reporting requirements and case definition with new definition released by CSTE in 2020. Revised sections: Why Is AFM Important to Public Health, Disease and Epidemiology, Public Health Control Measures, and Case Investigation with current understanding about AFM. Clarified Case Investigation process with more specific instructions for the public health investigator (LHD-level investigator or epidemiologist) and the AFM coordinator (State-level epidemiologist). Updated links throughout.

V. 02.22—Updated disease and epidemiology to reflect current patterns. Updated reporting criteria and case definition to match 2021 CSTE case definition.

UT-NEDSS (EpiTrax) minimum/required fields by tab

Morbidity event

Demographic

- Last name
- Street
- City
- State
- County
- Zip code
- Date of birth
- Area code
- Phone number
- Birth gender
- Ethnicity
- Race

Clinical

- Disease
- Onset date
- Date diagnosed
- Hospitalized
- Admission date
- Died
- Date of death

Laboratory

- Test type
- Organism
- Test result
- Collection date
- Lab test date

Epidemiological

- Imported from
- Risk factors

Reporting

- Date first reported to public health

Administrative

- State case status (completed by DHHS)
- Outbreak associated
- Outbreak name