Acute Flaccid Myelitis

Disease Plan

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Last updated: February 19, 2021 by Jared Ripplinger.

Questions about this disease plan?

Contact the Utah Department of Health Bureau of Epidemiology: 801-538-6191.
## 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 with 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 testing of stool samples

#### 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](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 one 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 one 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 with 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 have spiked every two years from August to November since the fall of 2014 (with the exception of 2020) and have coincided with national outbreaks of mild respiratory illnesses among children caused by enterovirus D68 (EV-D68). 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 that they may have a causative role in the development of AFM. Despite this association in timing, a cause for AFM has not been determined. AFM is a syndrome, so it is possible that multiple pathogens cause AFM. CDC is continuing 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 the polio virus but has been eliminated from most of the world, including the United States. No polio virus has been detected in the stool samples of any AFM patients.
Laboratory Identification
Viral laboratory testing and reporting is performed at the CDC. CDC specimen collection instructions are available at https://www.cdc.gov/acute-flaccid-myelitis/hcp/specimen-collection.html. Specimen submission to CDC must be coordinated by the Utah Department of Health Bureau of Epidemiology 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 particular 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 at the following CDC website: https://www.cdc.gov/acute-flaccid-myelitis/hcp/clinical-management.html. Clinicians can schedule a consult with a neurologist that specializes in AFM through the AFM Physician Consult and Support Portal found at https://wearesrna.org/living-with-myelitis/resources/afm-physician-support-portal/.

Case Fatality Ratio
As of December 2020, two patients in the acute phase of AFM have died, one in 2017 and one in 2020, out of a total of 645 confirmed cases in the United States for a case fatality ratio 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 that 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 2020.

✓ PUBLIC HEALTH CONTROL MEASURES

Public Health Responsibility
Public health’s responsibility in regards to AFM is twofold:

- **Early detection.** Public health should be monitoring respiratory diseases and communicating with clinicians to identify persons with compatible illness to ensure that complete testing, and ascertainment of potential etiologies, can be performed.

- **Rapid assessment and response.** Public health should respond to suspected cases quickly by providing recommendations to providers and coordinating 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
Over 90% of AFM cases have been observed in patients with a mild respiratory symptoms or fever consistent with a recent viral infection. Public health recommends that people practice 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 follow CDC’s recommendations for EV-D68. These precautions include standard, contact, and droplet precautions. For more detail on these interim recommendations visit [https://www.cdc.gov/non-polio-enterovirus/hcp/ev-d68-hcp.html](https://www.cdc.gov/non-polio-enterovirus/hcp/ev-d68-hcp.html).
CASE INVESTIGATION

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

Table of criteria to determine whether a case should be reported to public health authorities

<table>
<thead>
<tr>
<th>Criteria for Reporting</th>
<th>AFM</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Clinical Criteria</strong></td>
<td></td>
</tr>
<tr>
<td>A person with onset of acute flaccid* limb weakness</td>
<td>N</td>
</tr>
<tr>
<td><strong>Laboratory/Imaging Criteria</strong></td>
<td></td>
</tr>
<tr>
<td>A magnetic resonance image (MRI) showing a spinal cord lesion in at least some gray matter** and spanning one or more vertebral segments</td>
<td>N</td>
</tr>
<tr>
<td>Excluding persons with gray matter lesions in the spinal cord resulting from physician diagnosed malignancy, vascular disease, or anatomic abnormalities</td>
<td>N</td>
</tr>
<tr>
<td><strong>Vital Records Criteria</strong></td>
<td></td>
</tr>
<tr>
<td>Any person whose death certificate lists acute flaccid myelitis as a cause of death or a condition contributing to death</td>
<td>S</td>
</tr>
<tr>
<td><strong>Other Criteria</strong></td>
<td></td>
</tr>
<tr>
<td>Autopsy findings that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord spanning one or more vertebral segments</td>
<td>S</td>
</tr>
</tbody>
</table>

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

**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.

Notes:
S = This criterion alone is Sufficient to identify a case for reporting.
N = All “N” criteria in the same column are Necessary to identify a case for reporting.
O = At least one of these “O” (Optional) criteria in each category (e.g., clinical evidence and laboratory evidence) in the same column—in conjunction with all “N” criteria in the same column—is required to identify a case for reporting. (These optional criteria are alternatives, which mean that a single column will have either no O criteria or multiple O criteria; no column should have only one O.)
Case Definition
Acute Flaccid Myelitis (2020)

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.

Laboratory/Imaging Criteria

Confirmatory laboratory/imaging 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 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 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.

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 case as probable or confirmed.

Comments
To provide consistency in case classification, review of case information and assignment of final case classification for all suspected AFM cases will be completed by experts in national AFM surveillance. This is similar to the review required for final classification of paralytic polio cases.
Acute Flaccid Myelitis: Utah Public Health Disease Investigation Plan

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## Criteria for defining a case of Acute Flaccid Myelitis

<table>
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<th>Case Definition</th>
<th>Suspect</th>
<th>Probable</th>
<th>Confirmed</th>
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<tr>
<td><strong>Clinical Evidence</strong></td>
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<tr>
<td>Acute flaccid* weakness of one or more limbs</td>
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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).
A = This criterion must be absent (e.g., NOT present) for the case to meet the classification criteria.
Case Investigation Process

The case investigation process is as follows:

- Upon notification of a potential case, the public health investigator* or AFM coordinator** will contact the hospital infection preventionist and/or clinician and enter 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) (see https://www.cdc.gov/acute-flaccid-myelitis/hcp/specimen-collection.html). The specimen submission form (CDC Form 50.34) must accompany specimens to UPHL (Form 50.34 can be found here: http://www.cdc.gov/laboratory/specimen-submission/form.html).
- The public health investigator, infection preventionist, or patient’s clinician will complete the Patient Summary Form and send to the AFM coordinator (https://www.cdc.gov/acute-flaccid-myelitis/downloads/patient-summary-form.pdf).
- The AFM coordinator will notify CDC of the suspect case, inform them of the expected sample delivery date, and send them the Patient Summary Form.
- The CDC will send test results to UPHL and the AFM coordinator.
- The AFM coordinator will send test results and CDC’s case classification to the public health investigator.
- The public health investigator will update the case in UT-NEDSS (EpiTrax) and send test results and case classification to the clinician.
- The AFM coordinator will communicate 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 AFM Epidemiologist responsible for coordinating reporting, specimen collection, and communication with CDC. If you are unsure who this is, please call 801-538-6191 to verify.

Outbreaks

Not yet determined.

Identifying Case Contacts

Not yet determined.

Case Contact Management

Not yet determined.
REFERENCES


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.
### 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 UDOH)
- Outbreak Associated
- Outbreak Name