Acute Flaccid Myelitis

BASIC EPIDEMIOLOGY

Infectious Agent
There are multiple infectious agents that may be associated with acute flaccid myelitis (AFM). Conditions like AFM may be caused by a variety of factors, including several viruses:

- Enteroviruses
- West Nile Virus (WNV) and viruses in the same family as WNV, specifically Japanese encephalitis virus and South Louis encephalitis viruses, and
- Adenoviruses

Transmission
Mode of transmission is dependent on the infectious agent.

Incubation Period
Incubation period is dependent on the infectious agent.

Communicability
Although the underlying infection may be communicable, the condition of AFM is usually a rare complication.

Clinical Illness
Acute flaccid myelitis is a clinical syndrome characterized by sudden limb weakness (weakness or paralysis in one or more extremities, but not generalized to the entire body) and loss of muscle tone and reflexes. Some patients, in addition to the limb weakness, will experience:

- Facial droop/weakness
- Difficulty moving the eyes
- Drooping eyelids
- Difficulty with swallowing or slurred speech

Numbness or tingling is rare in patients with AFM, though some patients have pain in their arms or legs. Some patients with AFM may be unable to pass urine. The most severe symptoms of AFM are body temperature and blood pressure instability and respiratory failure that can happen when the muscles involved with breathing become weak. This can require urgent ventilator support (breathing machines).

DEFINITIONS

Clinical Case Definition
An illness with onset of acute flaccid limb weakness (low muscle tone, limp, hanging loosely, not spastic or contracted) of one or more limbs

Laboratory Criteria for Diagnosis

- A magnetic resonance image (MRI) showing aspinal cord lesion with predominant gray matter* involvementand spanning one or more vertebral segments
- 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. If still unsure if this criterion is met, consider consulting the neurologist or radiologist directly.

Case Classification

- **Confirmed:**
  - An illness with onset of acute focal limb weakness of one or more limbs **AND**
  - An MRI showing a spinal cord lesion with predominant gray matter*, † and spanning one or more vertebral segments.
- **Probable:**
  - An illness with onset of acute focal limb weakness of one or more limbs **AND**
  - An MRI showing spinal cord lesion where gray matter involvement is present, but predominance cannot be determined
- **Suspect:**
  - An illness with onset of acute focal limb weakness of one or more limbs **AND**
  - An MRI showing spinal cord lesion in at least some gray matter and spanning one or more vertebral segments

Other classification criteria: Autopsy findings that include histopathologic evidence of inflammation largely involving the anterior horn of the spinal cord spanning one or more vertebral segments.

Note: 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. Final case determinations will be emailed to the respective Public Health Region.

SURVEILLANCE AND CASE INVESTIGATION

Case Investigation

Local and regional health departments should investigate all reports of AFM. If an etiology is known and is a reportable condition (e.g., West Nile, varicella, or polio), the case should be investigated according to the etiology.

If the etiology is known and due to a non-reportable condition OR if the etiology is unknown, use this chapter for investigation purposes.

Case Investigation Checklist

- ☐ Confirm the clinical presentation of the patient.
- ☐ Ascertain what testing has been done, including lab testing, lumbar puncture, and MRI.
- ☐ Notify EAIDU of suspect case of AFM at **(800) 252-8239 or (512) 776-7676.**
- ☐ Ask the treating physician, preferably the neurologist, to complete the *Acute Flaccid Myelitis: Patient Summary Form.*
  - EAIDU does NOT recommend that the LHD complete the form themselves.
- ☐ Submit the *Acute Flaccid Myelitis: Patient Summary Form* to EAIDU.
  - CDC also requires a copy of the History & Physical (H&P), MRI report, MRI images (on CD), Neurology consult notes, EMG report (if done), Infectious disease consult notes (if available), vaccination record, and diagnostic laboratory reports for patients reported with suspect AFM.

        - MRI images should, ideally, be sent via the CDC Ambra portal or on a CD are not required to be sent at the time of initial Patient Summary Form and paper 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 DSHS EAIDU.
    - EAIDU will obtain approval from CDC for testing.
- ☐ Collect specimens, if possible, within 24 hours of onset of limb weakness, and to submit to DSHS Austin laboratory (Table 1). CDC has requested LHDs and providers **do not submit directly to the CDC.**
- ☐ DSHS will forward appropriate specimens onto the CDC for testing.
- ☐ Submit MRI images on a CD to appropriate department at CDC as directed by EAIDU.
- ☐ Complete 60 Day Follow Up section of *Acute Flaccid Myelitis: Patient Summary Form* and submit to EAIDU.

Control Measures
Control measures will depend on the causative agent; however, proper hand hygiene will help in controlling spread. Standard precautions in healthcare facilities should be implemented.

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.

MANAGING SPECIAL SITUATIONS

Outbreaks
If an outbreak of AFM is suspected, notify the regional DSHS office or to EAIDU at (**512) 776-7676**.

REPORTING AND DATA ENTRY REQUIREMENTS

Provider, School & Child-Care Facilities, and General Public Reporting Requirements
Acute flaccid myelitis is not currently a reportable condition in and of itself. However, certain illnesses that cause AFM (e.g., polio, varicella, West Nile) may be reportable and should be reported according to Texas Administrative Code requirements for these conditions.

EAIDU requests that patients with suspected AFM be reported within one week to the local or regional health department or the Texas Department of State Health Services (DSHS), Emerging and Acute Infectious Disease Unit (EAIDU) at **(800) 252-8239 or (512) 776-7676.**

Local and Regional Reporting and Follow-up Responsibilities
Local and regional health departments should:

- Fax or email the *Acute Flaccid Myelitis: Patient Summary Form* as soon as possible to EAIDU. The form is needed to facilitate lab testing with CDC.
    - Forms should be faxed or emailed once enough information has been collected to establish that a patient meets the clinical presentation of acute flaccid limb weakness.
        - MRI images upload are not required to be sent at the time of Patient Summary Form and paper medical record information. However, the patient must have an MRI or lumbar puncture done to be submitted for

CDC review. These should be submitted as promptly as possible. The CDC cannot review the patient without these.
    - In the event of a death, copies of the hospital discharge summary, death certificate, and autopsy report should also be sent to DSHS EAIDU.
  - Investigation forms may be faxed to **512-776-7616**, emailed securely to AFMTexas@dshs.texas.gov or mailed to:

    Emerging and Acute Infectious Disease Unit
    Texas Department of State Health Services
    Mail Code: 1960 PO Box 149347
    Austin, TX 78714-9347
- Fax, send secure email, or mail completed *Acute Flaccid Myelitis Patient Summary Form* 60 day follow up, 6 month follow up, and 12 month follow up section once completed.

When an outbreak is investigated, local and regional health departments should:

- Report outbreaks within 24 hours of identification to the regional DSHS office or to EAIDU at **512-776-7676.**

LABORATORY PROCEDURES

Prior to shipping, coordinate with EAIDU Central Office staff regarding specimens shipped.

Clinicians treating patients meeting the AFM case definition should pursue laboratory testing of CSF, blood, serum, respiratory, and stool specimens for enteroviruses, West Nile virus, and other known infectious etiologies at their usual clinical and reference laboratories. Clinicians may contact the local health department and/or DSHS for assistance with any testing that is not available locally. Specimens should not be shipped to DSHS without first consulting with the local health department.

Clinicians should collect specimens from patients suspected of having 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 diagnosis of AFM. The specimens which should be collected include the following:

- Cerebrospinal fluid (CSF) **AND**
- Blood (serum and whole blood), **AND**
- Stool (preferably two stool specimens collected as soon after onset of limb weakness and separated by 24 hours. Please do not send a rectal swab) **AND**
- Nasopharyngeal swab (NP/OP) only if patient tested positive for enterovirus/rhinovirus

CDC advised overnight shipment of available clinical specimens, within 24-48 hours of specimen collection if possible, from patients that meet the clinical case definition. Please ship specimens overnight so they arrive at DSHS Lab in Austin on Tuesday through Friday. Do not ship specimens on Friday or over the weekend.

For specimens that should be frozen, please freeze them at -20°C and make arrangements to ship the specimens overnight to DSHS Lab in Austin frozen on dry ice.

For specimens that should be sent refrigerated, please store them at 4°C and make arrangements to ship the specimens overnight to DSHS Lab in Austin on cold packs. Specimens should not have direct contact with the cold packs during shipping.

Specimens from each patient should be shipped with completed hard copies of the following:

- The Acute Flaccid Myelitis Patient Summary Form
- Submitters do not need to complete the CDC specimen submission form 50.34. The DSHS laboratory will complete this.
- A G-2V form must be submitted to the DSHS Austin laboratory for each individual specimen being submitted. For example, if you submit two stool specimens there should be 2 G-2V forms.

If ten or more patient specimens are submitted, please provide an electronic line listing by email. Use the following headers in this order: patient ID number; date of birth; sex; onset date; fatal y/n; specimen ID number; specimen collection date; specimen type; if culture isolate–cell line and passage number.

Prior to shipping, coordinate with Central Office staff regarding specimens shipped.

Additional instructions regarding specimen collection, storage, and shipping can be found at: https://www.cdc.gov/acute-flaccid-myelitis/hcp/instructions.html

TABLES

Table 1: Specimens to collect and send to CDC for testing for Patients Under Investigation (PUIs) for AFM

Specimen Type	Minimum Amount	Collection	Storage	Shipping	Comments
Required Specimens					
Cerebrospinal fluid (CSF)	1 mL	Spun and processed; standard cryovial tube; collect at same time or within 24 hours of serum if possible	Freeze at -70°C	Ship on dry ice	CSF will be used for special studies; EV/RV testing will be batched and results returned as sample amount allows
Serum*	0.4 mL	Spun and processed; Tiger/ red top tube; collect at the same time or within 24 hours of CSF if possible	Freeze at -70°C	Ship on dry ice	Serum will be used for special studies; no individual results will be returned
Stool (Whole stool (preferred)	≥1gram	Collect in sterile container, no special medium required, not a rectal swab†	Freeze at -20°C**	Ship on dry ice	Results for EV/RV and poliovirus testing will be returned as testing completed (within 14 days)
Respiratory - NP/OP swab	1ml	Store in viral transport medium	Freeze at -20°C**	Ship on dry ice	EV/RV testing and typing will be performed and results returned within 10 days of sample receipt
In the event of death, please send the following specimens, if possible					
Fresh-frozen tissue		Place directly on dry ice or liquid nitrogen	Freeze at -70°C	Ship on dry ice	Representative sections from various organs are requested, but particularly from brain/spinal cord (including gray and white matter), heart, lung, liver, kidney, and other organs as available.

Formalin-fixed or formalin-fixed, paraffin-embedded tissue		Avoid prolonged fixation—tissues should have been fixed in formalin for 3 days, then transferred to 100% ethanol	Room temperature	Ship at room temperature with paraffin blocks in carriers to prevent breakage	See comment above regarding frozen tissue

***If any of the serum samples that you are sending to CDC were collected after the patient had received intravenous immune globulin (IVIG), steroid treatments, or plasmapheresis/plasma exchange, please indicate the date of that therapy on the Patient Summary Form.**

† The negative predictive value is very low for rectal swabs since the amount of fecal material collected is much less than for stool.

****All specimens may be stored at -70ºC for ease of shipping.**

NOTE: If specimens cannot be shipped within 24-48 hours of collection, consider recollection, if feasible.

REVISION HISTORY

November 2021
- Minor revisions

January 2021
- Updated case definition
- *Acute Flaccid Myelitis: Patient Summary Form* including updated medical record requirements and 60 day follow up section, 6 month, and 12 month follow up sections.
- Specimens should be sent through DSHS Austin laboratory and not directly to the CDC
- Updated information that CDC will review suspect AFM patients with limb weakness from prior years.
- Updated process for CDC review
- Specimen collection tables were updated to reflect changes to testing procedures at the CDC