Acute Flaccid Myelitis rev Jan 2018

BASIC EPIDEMIOLOGY

Infectious Agent
There are multiple infectious agents that can cause acute flaccid myelitis (AFM). Conditions like AFM can be caused by a variety of germs, 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 is 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 focal limb weakness. Multiple etiologic agents may cause acute flaccid myelitis.

Laboratory Criteria for Diagnosis

- A magnetic resonance image (MRI) showing a spinal cord lesion largely restricted to gray matter* and spanning one or more spinal segments
- A specific pathogen is not needed to confirm the diagnosis.

* 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. 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 **AND**
  - An MRI showing a spinal cord lesion largely restricted to gray matter*, † and spanning one or more spinal segments.
- **Probable:**
  - An illness with onset of acute focal limb weakness **AND**
  - Cerebrospinal fluid (CSF) with pleocytosis (white blood cell count >5 cells/mm^3, may adjust for presence of red blood cells by subtracting 1 white blood cell for every 500 red blood cells present).

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.

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 EAIDB 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.*
  - EAIDB does NOT recommend that the LHD complete the form themselves.
- ☐ Submit the *Acute Flaccid Myelitis: Patient Summary Form* to EAIDB.
  - 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 on 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 EAIDB.
  - EAIDB 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 EAIDB.
- Complete 60 Day Follow Up section of *Acute Flaccid Myelitis: Patient Summary Form* and submit to EAIDB.

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 EAIDB 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.

EAIDB 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 Branch (EAIDB) 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 EAIDB. 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 probable or confirmed case status.
        - MRI images on CD are not required to be sent at the time of 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 EAIDB.
    - Investigation forms may be faxed to **512-776-7616**, emailed securely to VPDTexas@dshs.texas.gov or mailed to:

      Infectious Disease Control 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 section once completed by the provider at the approximate 60-day patient follow up visit.

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 EAIDB at **512-776-7676.**

LABORATORY PROCEDURES

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.

Along with the specimens listed below, CDC would also require 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 record, and diagnostic laboratory reports for patients reported with suspect AFM.

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)

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
- A CDC specimen submission form 50.34 **FOR EACH SPECIMEN**. Please note that, for the Test Order Name, select "Picornavirus Special Study."

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

*For stool specimens, CDC recommends that healthcare providers rule out poliovirus infection in cases of acute flaccid paralysis (AFP) that are clinically compatible with polio, including those with anterior myelitis. Recommendations for polio testing can be found at: http://www.cdc.gov/polio/us/hcp.html. CDC can do testing for polio if the reporting facility cannot.

TABLES

Table 1: Specimens to Collect from Suspect AFM Cases

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 as whole blood	Freeze at -20°C	Ship on dry ice	
Serum		0.4 mL	Spun and processed; Tiger /red top tube	Freeze at -20°C	Ship on dry ice	
Whole blood		3-5 mL	Unspun; lavender/green top tube (with anticoagulant); collect at same time or within 24 hours as CSF	Refrigerate at 4°C	Ship overnight on cold packs. Ship within **24-48 hours** of collection*	Tubes should be insulated during shipping to ensure they are not in direct contact with cold pack
Stool	Whole stool (preferred)	≥1gram	Collect in sterile container, no special medium required	Freeze at -20°C	Ship on dry ice	Two samples total, collected at least 24 hours apart, both collected as early in illness as possible and ideally within 14 days of illness onset
	Rectal swab	≥1gram	Store in viral transport medium	Freeze at -20°C	Ship on dry ice	Two samples total, collected at least 24 hours apart, both collected as early in illness as possible and ideally within 14 days of illness onset
Optional Specimens						
Respiratory - NP/OP swab		1ml	Store in viral transport medium	Freeze at -20°C	Ship on dry ice	Send only if EV/RV positive for typing
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 specimens cannot be shipped within 24-48 hours of collection, consider recollection, if feasible.**

UPDATES

January 2018

- *Acute Flaccid Myelitis: Patient Summary Form* including updated medical record requirements and 60 day follow up section
- Specimen collection tables were updated to reflect changes to testing procedures at the CDC
- Specimens should be sent through DSHS Austin laboratory and not directly to the CDC