Janet T. Mills Governor

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AFM Toolkit for Neurologists in Maine August 2022

Acute flaccid myelitis (AFM) is a rare but serious condition. It affects the nervous system causing the muscles and reflexes in the body to become weak. Most AFM cases occur in late summer and early fall between August and October. The US has seen an increase in AFM cases every two years since 2014. In 2022, the United States Centers for Disease Control and Prevention (US CDC) has reported eleven confirmed cases of AFM. Although AFM has been extremely rare in Maine, with four confirmed cases reported since 2014, the condition progresses rapidly and can lead to respiratory failure, so early detection and hospitalization is critical. AFM mostly affects children.

This Toolkit provides some information on what is known about the epidemiology of AFM and additional resources.

Clinicians should report all patients suspected to have AFM, especially after respiratory illness or fever, to Maine Center for Disease Control and Prevention (Maine CDC) as soon as possible. Clinicians who suspect AFM should hospitalize patients immediately, collect lab specimens, diagnose, and begin medical management.

This toolkit includes materials to assist healthcare providers with general information on AFM reporting, testing, and submitting samples. The following materials are included in this toolkit:

- 1. AFM Guide: This document contains information on the background of AFM, signs and symptoms, causes, information for suspected cases, diagnosis instructions, and treatment options.
- 2. Reporting Patients Under Investigation for AFM: This document provides information for providers with Patient(s) Under Investigation (PUI) for AFM. It provides directions for reporting AFM as well as detailed instructions for collecting, storing, and shipping specimens.
- **3.** CDC Vital Signs Report 2020: This document provides an overview of AFM, stresses the importance of prompt recognition, rapid reporting, and conducting age-appropriate neurological exams.
- 4. AFM Frequently Asked Questions: This document contains frequently asked questions for clinicians from US CDC. The questions include various topics from reporting, specimen shipping, infection control practices, vaccines, and others.
- **5. AFM Factsheet for Parents:** This document is a resource for parents or guardians of a child that is diagnosed with or suspected of having AFM.
- 6. AFM Diagnosis and Classification Fact Sheet: This document provides a visual representation of the steps involved in diagnosis, specimen collection, and case classification of AFM.

7. Head, Shoulders, Knees & Toes Fact Sheet: This document provides questions for clinicians to consider while examining children with possible limb weakness.

Other Virtual AFM Resources:

- **CDC MMWR, August 2020:** Clinical Characteristics of Patients with Confirmed Acute Flaccid Myelitis, United States, 2018. Found here: https://www.cdc.gov/mmwr/volumes/69/wr/mm6931e3.htm?s_cid=mm6931e3_w.
- CDC Webinar: Acute Flaccid Myelitis: What Healthcare Providers Need to Know in 2020: https://www.cdc.gov/acute-flaccid-myelitis/hcp/clinicians-health-departments/webinar-2020.html.
- Public Health Grand Rounds: Acute Flaccid Myelitis: <u>https://www.cdc.gov/grand-rounds/pp/2020/20200703-acute-flaccid-myelitis.html</u>.

This toolkit is also available online at <u>www.maine.gov/dhhs/afm</u>.

To report a suspect case of AFM please call Maine CDC at 1-800-821-5821. You can also email <u>disease.reporting@maine.gov</u> with questions or to request a call back. This email is not secure and personal information should not be sent.

Thank you.

Acute Flaccid Myelitis (AFM) Guide



What is AFM?

Acute flaccid myelitis (AFM) is a rare but serious condition. It affects the nervous system, specifically gray matter, which causes the muscles and reflexes in the body to become weak.

Symptoms of AFM

Most people will have sudden onset of:

- Arm or leg weakness
- Loss of muscle tone and reflexes



Some people will also have:

- Facial droop or weakness
- Difficulty moving eyes
- Drooping eyelids
- Difficulty swallowing
- Slurred speech
- Pain in arms, legs, neck, or back



In rare cases, people may also:

- Have numbness or tingling
- Be unable to pass urine

Severe cases:

- Respiratory failure
- Serious neurologic complications

Causes and Prevention of AFM

- US CDC believes viruses, including enteroviruses, likely play a role in AFM. It is currently unknown why a small number of people develop AFM, while most others recover from the virus.
- All stool specimens from AFM patients tested negative for poliovirus.
- There is no specific action to prevent AFM.

Suspect Cases of AFM

- 1. Contact Maine CDC
 - a. If you have a suspect case of AFM, contact Maine CDC as soon as possible to report the suspect case. Clinicians should continue to be vigilant and send information about patients who meet the clinical criteria (sudden onset of flaccid limb weakness) AND laboratory/imaging criteria (MRI showing a spinal cord lesion in at least some gray matter and excluding persons with gray matter lesions in the spinal cord resulting from physician diagnosed malignancy, vascular disease, or anatomic abnormalities) for AFM to Maine CDC regardless of any laboratory results.
 - b. You can contact Maine CDC by phone at 1-800-821-5821 or email <u>disease.reporting@maine.gov</u>. This email is not secure and personal information should not be sent. Urgent questions can also be directed to federal CDC's Emergency Operations Center at 770-488-7100.
- 2. Collect Specimens
 - a. Collect specimens as close to onset of limb weakness as possible. Handle and store as directed.
 - b. Work with Maine CDC to coordinate submission of specimens for testing at federal CDC.
 - i. Specimens should be shipped overnight to arrive at federal CDC Tuesday through Friday.
 - ii. For specimen collection instructions, visit <u>https://www.cdc.gov/acute-flaccid-</u> <u>myelitis/hcp/specimen-collection.html</u>.



- 3. Information to provide to Maine CDC:
 - a. Admission and discharge notes
 - b. Neurology and infectious disease consult notes
 - c. MRI report
 - d. MRI images
 - e. Vaccination history
 - f. Laboratory test results

Diagnosis of AFM

It is important that tests are done as soon as possible after a patient develops symptoms. AFM is diagnosed by examining a patient's nervous system and nerve conduction in combination with:

- Magnetic resonance imaging (MRI)
- Lab tests on the cerebrospinal fluid (CSF)

AFM can be difficult to diagnose because the signs and symptoms are similar to other neurologic diseases, like transverse myelitis and Guillain-Barre syndrome.

Case Definition for AFM

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 (NNC) should be reported only once using the event code for the NNC to avoid duplicate reporting.

Laboratory 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.

+ 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.

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.

Treatment of AFM

There is no specific treatment for AFM. A neurologist may recommend certain interventions on a case-by-case basis.

For more information, see federal CDC's interim considerations for clinical management website: <u>https://www.cdc.gov/acute-flaccid-myelitis/hcp/clinical-management.html</u>.

References and Resources

- Federal CDC Acute Flaccid Myelitis Website: <u>www.cdc.gov/acute-flaccid-myelitis</u>
- Acute Flaccid Myelitis: Interim Considerations for Clinical Management: <u>https://www.cdc.gov/acute-flaccid-myelitis/hcp/clinical-management.html</u>
- Transverse Myelitis Association's AFM Physician Consult and Support Portal: <u>https://myelitis.org/living-with-myelitis/resources/afm-physician-support-portal/</u>
- Maine CDC's AFM website: <u>www.maine.gov/dhhs/afm</u>

Reporting Patients Under Investigation for Acute Flaccid Myelitis

HEALTHCARE PROVIDERS SHOULD

Identify PUI

Identify patient under investigation (PUI) for acute flaccid myelitis (AFM). Patient with:

- o Onset of acute flaccid limb weakness
- \circ $\,$ An MRI showing spinal cord lesions in at least some gray matter $\,$

Contact Maine CDC

Contact Maine CDC to coordinate submission of specimens and information, including copies of:

- MRI images & report
- Neurology consult notes

Collect Specimens

Collect specimens as close to onset of limb weakness as possible and store as directed (freeze as soon as possible after collection).

Specimens to collect for testing AFM PUIs at CDC

ALL submissions to US CDC for diagnostic testing require pre-approval at this time. Please contact Maine CDC at 1-800-821-5821 to coordinate submissions.

SAMPLE	MINIMUM AMOUNT	TUBE TYPE	PROCESSING	STORAGE	SHIPPING
CSF	0.15 mL, 0.5-2 mL preferred (collect at same time or within 24hrs of serum if feasible)	Cryovial	Spun and CSF removed to cryovial	Freeze at ≤-20°C	Frozen on dry ice.
Serum	0.5 mL, 1 mL preferred (collect at same time or within 24hrs of CSF if feasible)	Tiger/red top	Spun and serum removed to tiger/ red top	Freeze at ≤-20°C	Frozen on dry ice.
Stool*	1 gram, 10 – 20 grams preferred (2 samples collected 24hrs apart)	Sterile container	N/A	Freeze at ≤-20°C	Frozen on dry ice. Rectal swabs should not be sent in place of stool.
Respiratory (NP)/ Oropharangeal (OP) swab	0.5 mL, 1 mL preferred (minimum amount)	N/A	Store in vial transport medium	Freeze at ≤-20°C	Frozen on dry ice.

* Please include stool specimens along with CSF, serum, and NP/OP swabs to help with identification of pathogens and to support poliovirus surveillance

Contact Maine CDC at 1-800-821-5821.



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Vitalsigns™

Acute Flaccid Myelitis (AFM) Recognize symptoms. Hospitalize immediately.



Want to learn more? www.cdc.gov/vitalsigns/afm2020



Most patients first sought care at an emergency department.

98%

Most patients with AFM were hospitalized. **54%**

Over half were admitted to the ICU. 1 in 4 hospitalized patients required a ventilator.

Overview

Acute flaccid myelitis (AFM) is an uncommon, but life-threatening neurologic condition that affects mostly children and can lead to permanent paralysis. Enteroviruses, particularly EV-D68, are likely responsible for the increase in cases every two years since 2014. AFM is a medical emergency and patients must be hospitalized and monitored in case they progress to respiratory failure. Prompt recognition and immediate action by pediatricians, and emergency department and urgent care providers are critical to achieving the best possible outcomes.

- AFM typically presents with sudden limb weakness. Most patients had respiratory illness or fever before AFM onset.
- Patient health can decline quickly, resulting in paralysis or the need for a ventilator. AFM can lead to permanent disability.
- Patients who tested positive for EV-D68 typically had more severe AFM illness, requiring hospitalized intensive care and ventilation.
- Most cases occur between August and November.



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PROBLEM

Delays in recognition can put patients at risk

A third of patients were hospitalized two or more days after limb weakness.

When clinicians recognize AFM early, they can quickly

- Hospitalize patients and provide optimal medical management and rehabilitation.
- Collect clinical specimens and order a brain and spinal cord MRI. Done early, these help detect the cause and distinguish AFM from other conditions with limb weakness.



THE WAY FORWARD

HEALTHCARE PROVIDERS CAN:

- Suspect AFM in patients with sudden limb weakness, especially between August and November.
- Collect clinical specimens immediately and report cases to the state health department.
- Request specialty consultations such as infectious disease and neurology.

HEALTH DEPARTMENTS CAN:

- Communicate information about AFM to healthcare providers.
- Work with CDC to collect medical information, specimens, MRI images, and classify cases.

PARENTS CAN:

- Seek medical care immediately if a child develops sudden arm or leg weakness.
- Seek support from other parents and families affected by AFM.
- Visit CDC's AFM parent webpage: www.cdc.gov/acute-flaccidmyelitis/parents/index.html

CONTACT AFM SPECIALISTS through the **AFM Physician Consult and Support** Portal: https://bit.ly/2Y2U3VR

For more information 1-800-CDC-INFO (232-4636) TTY: 1-888-232-6348 | Web: www.cdc.gov

Centers for Disease Control and Prevention 1600 Clifton Road NE, Atlanta, GA 30333 Publication date: August 4, 2020

SOURCE: MMWR, August 2020.



Age-appropriate neurological exam

- □ Muscle strength in all four limbs (What is their strength? Can they move the limb against gravity with resistance?)
- □ Muscle tone (Is it loose/floppy?)
- □ Reflexes (Are they hypo-, hyper-, or absent?)
- Cranial nerve assessment (Are there any cranial nerve deficits?)



Reporting Cases



How do I report (alert the health authorities about) a patient under investigation (PUI) for AFM?



Clinicians: If you believe your patient has symptoms of AFM, such as acute flaccid weakness, contact Maine CDC at 1-800-821-5821 as soon as possible for instructions on how to report. Urgent questions may also be directed to the US CDC Emergency Operations Center (770-488-7100). Non-urgent questions can be emailed to the AFM team at AFMinfo@cdc.gov. In addition, please collect biological specimens for testing as soon as possible to increase the possibility of finding a cause. These specimens can be tested at a hospital or state public health laboratory (Maine's Health and Environmental Testing Laboratory, HETL) for enteroviruses, West Nile virus and other infectious etiologies known to be associated with AFM. At the same time, additional aliquots of CSF, serum, stool, and respiratory samples should be sent to US CDC for testing for both infectious and noninfectious causes. Additional instructions regarding US CDC-specific specimen collection and shipping can be found on their Specimen Collection Instructions webpage at www.cdc.gov/acute-flaccidmyelitis/hcp/instructions.html. For more information on how to send information about a patient under investigation (PUI) for AFM, see US CDC's document "Reporting Patients Under Investigator for Acute Flaccid Myelitis" https://www.cdc.gov/acute-flaccidmyelitis/downloads/job-aid-for-clinicians-508.pdf.



Should I send information about a patient under investigation (PUI) for AFM even if his/her clinical specimen was negative for enteroviruses?



Yes, US CDC requests that you send biologic specimens and information about all patients under investigation (PUIs) to Maine CDC, regardless of laboratory testing results. US CDC tests specimens to identify the pathogens and biologic mechanisms responsible for AFM. These tests include, but are not limited to, tests for enteroviruses.

Specimen Collecting & Testing



Should I send specimens to US CDC even if the hospital laboratory or state public health laboratory can test for enteroviruses?



Yes, US CDC requests that you send specimens (i.e., cerebrospinal fluid, serum, stool, and respiratory samples) to US CDC for standardized testing and for expanded testing protocols. Contact Maine CDC to coordinate sending of specimens to US CDC for testing. Upon completion, US CDC will share with Maine CDC the results from certain tests, such as enterovirus/rhinovirus testing and typing and stool testing. Maine CDC will then share the results with the clinician. For instructions on how to submit specimens to US CDC, see Specimen Collection

Instructions: <u>https://www.cdc.gov/acute-flaccid-myelitis/hcp/specimen-collection.html.</u>



What happens to the patient specimens that I send to US CDC, and when should I expect to receive the testing results?



All specimens submitted to US CDC help us learn more about AFM, including possible causes and how the immune system responds to this condition. You should not use results from these tests to inform clinical management of your patient because results may not be available in real-time.

- **Respiratory and stool specimens:** As soon as they are completed, US CDC will share with the specimen submitter and Maine CDC the respiratory testing and typing results for enterovirus/rhinovirus and stool testing for poliovirus. Maine CDC will then share the results with the clinician.
- Other specimens: US CDC will use results from other specimens (e.g., CSF and serum) for exploratory testing to learn more about immune responses to AFM. Results will not be immediately available.

Since US CDC testing protocols include several immunoassays that are not approved by the Clinical Laboratory Improvement Amendments (CLIA) nor are intended for clinical diagnosis, US CDC will be unable to provide patient-specific results for certain tests that are performed. However, US CDC will rapidly disseminate results from exploratory testing of samples from multiple cases which may indicate a possible cause of AFM.

Infection Control



What are your interim infection control recommendations for healthcare professionals?

US CDC's interim recommendation for management of patients with acute flaccid myelitis is Standard + Contact + Droplet precautions. This is consistent with US CDC's Recommendations for EV-D68. There are no pathogen-specific recommendations to add at this time.

Case Classification



When should I expect AFM case classification results back from US CDC?

The case classification will be communicated through Maine CDC when the review is complete, generally about 4 weeks after all of the information is received.

The process for case classification requires collection of many different pieces of information, including neurology notes and MRI images, which are then reviewed by several experts. Case classification is used for surveillance purposes and should not interfere with the differential or final clinical diagnosis or treatment of the patient. For more information on the AFM case definitions, visit Case Definitions: <u>https://www.cdc.gov/acuteflaccid-myelitis/hcp/case-definitions.html.</u>

Patient Follow-Up



Will US CDC conduct extended follow-up on cases of AFM after their initial clinical presentation?



Currently, US CDC is working with health departments to collect long-term follow-up information (2 months, 6 months, and 12 months after onset of limb weakness) about confirmed and probable cases of AFM.

Vaccination



Do intramuscular injections play a role in AFM?

Among many ongoing investigation activities to determine the cause of AFM, US CDC is looking at any possible association between intramuscular injections given in healthcare settings, such as medications and vaccines, and AFM. US CDC collects medical information on all suspected cases of AFM in the United States that are reported to them from health departments. A history of intramuscular injections received in the 30 days prior to limb weakness is recorded for all cases. Of the AFM cases reviewed thus far, approximately 85% have **no** recorded vaccination in the 30 days prior to the onset of limb weakness.



Are there any vaccines or medications to protect against AFM?



At this time, there are no vaccines or medicines, such as antivirals, that have been proven to protect a person from getting AFM or treat a person who has AFM. A summary of experts' current approaches to clinical management of AFM can be found on US CDC's Clinical Management of Patients webpage: <u>https://www.cdc.gov/acuteflaccid-myelitis/hcp/clinical-management.html.</u>

Although there is no vaccine for AFM, being up to date on all recommended vaccinations is essential to prevent a number of severe diseases including polio, which can cause acute flaccid paralysis, and numerous other vaccine-preventable diseases.

Acute Flaccid Myelitis in Children

A PARENT FACT SHEET

Acute flaccid myelitis (AFM) is an uncommon but serious neurologic condition that causes weakness in the arms or legs. If your child develops these symptoms, you should seek medical care right away.

Symptoms of AFM

AFM affects a child's nervous system, specifically the spinal cord. It usually starts with sudden limb weakness and loss of muscle tone and reflexes. Some may also experience:

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

Less common symptoms of AFM include numbness or tingling in the limbs and neck. A child may also have difficulty breathing because of weakness of muscles that support breathing.

AFM Diagnosis

A doctor will review a patient's medical history. They will also carefully examine the nervous system and places of weakness, low muscle tone, and decreased reflexes. Magnetic resonance imaging (MRI), lab testing of the cerebrospinal fluid, and tests to measure how the nerves are working might also be used to diagnose AFM.



Most children with AFM will have sudden onset of arm or leg weakness.



Some children might have arm, leg, or neck pain before the weakness begins.

Causes of AFM

AFM can be caused by viruses. Since 2014, most of the children with AFM (more than 90%) had a respiratory illness or fever before they developed arm or leg weakness.

Increases in AFM cases happened in 2014, 2016, and 2018, with most cases occurring between August and October. At this same time of year, viruses, including enteroviruses, commonly spread. Data collected since 2014 indicate that enteroviruses, and more specifically enterovirus D68 (EV-D68), play a major role in the recent increases in AFM. Respiratory symptoms and fever from viral infections are common, especially in children, and most people recover. It is unclear why a small number of people develop AFM after having a virus.

Acute Flaccid Myelitis in Children

A PARENT FACT SHEET

AFM is sometimes referred to as a "polio-like" illness. This is because the symptoms look similar to those of polio. However, all the stool specimens from AFM patients tested negative for poliovirus. This means that the cases of AFM in the U.S. since 2014 were not caused by poliovirus.

AFM Treatment

There is no specific treatment for AFM, but a doctor who specializes in treating brain and spinal cord illnesses (neurologist) might recommend certain treatments on a caseby-case basis. Doctors will also recommend physical and occupational therapy to help with arm or leg weakness.

Clinicians and public health officials who manage the care of individuals with AFM can review <u>AFM Clinical Guidance | CDC</u>.

Prevention

Even though AFM can be caused by viruses, we do not know what triggers AFM in a person, so there is no specific action that could prevent AFM. Most children who developed AFM had a respiratory illness or fever, likely from a viral infection.

You can lower your child's risk of getting a virus by:

- Washing hands often with soap and water, for at least 20 seconds
- Avoiding touching face with unwashed hands
- Avoiding close contact with people who are sick

You can decrease the risk of spreading viruses by:

- Cleaning and disinfecting frequently touched surfaces, including toys and doorknobs
- Having your child cover coughs and sneezes with a tissue or upper shirt sleeve, not hands
- Keeping sick children at home

How is CDC tracking AFM cases?

Doctors who might have a patient with AFM send medical information and test results to their health departments, who then send this information to CDC's AFM experts to review. The submitted cases are then classified as confirmed, probable, suspect, or not a case of AFM based on criteria for the AFM case definition. CDC sends the classification back to the health department who shares it with the doctor who then shares it with the patient. The process of case classification can take time to complete because of all the information that needs to be gathered and reviewed. However, this process is meant to track the trends of AFM in the United States over time and should not delay treatment or diagnosis.



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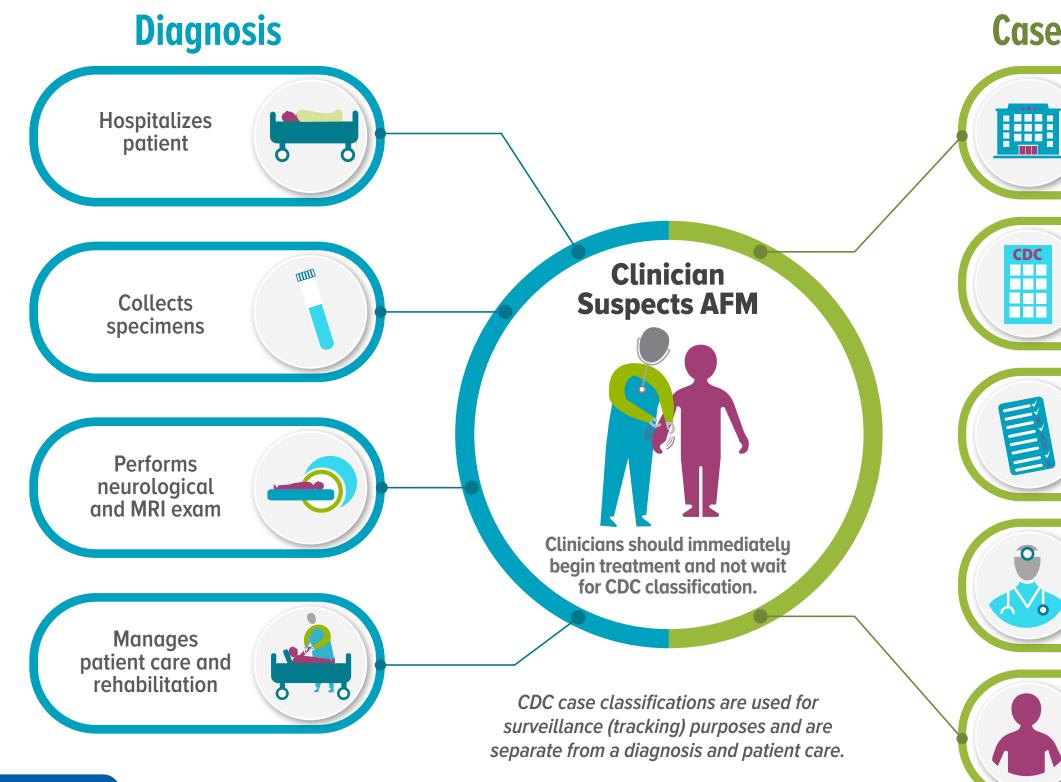


For more information on acute flaccid myelitis, visit **www.cdc.gov/afm**.



AFM is a medical emergency because it can cause problems with breathing. Children who have symptoms of AFM should be seen by a doctor right away.

ACUTE FLACCID MYELITIS: DIAGNOSIS AND CLASSIFICATION





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Case Classification

Clinician reports suspected case to health department

Health department sends report to CDC

CDC reviews, assigns classification. and sends results to health department

Health department sends results to clinician

Clinician shares results with patient

HEAD SHOULDERS KNEES OTOES

Unexplained proximal muscle weakness in children can occur in some neurologic conditions and can be easily missed during exams that only focus on distal strength.

> When examining children with sudden limb, neck, or trunk weakness, remember head, shoulders, knees, and toes.

Lift both arms above the **HEAD**

Muscle Group:



Ask:

- * Are they using one limb less?
- ***** Can they put on a T-shirt?
- Can they give a high-five with each hand?

KNEES

Muscle Group:

Shrug the **SHOULDERS** Muscle Group:

Neck/Shoulder Girdle

Ask:

- Is one shoulder higher than the other?
- Can they throw a ball overhead?
- Can they hold up their head?

Reach down & touch **TOES**

Muscle Group:

Ask:

🚼 Hips

Raise

- Are they limping or dragging a leg?
- * Can they put on pants?
- Can they do a squat and recover?



Ask:

- Are they waddling or falling while walking?
- Can they sit up and stand without support?
- Can they get a toy off the ground while standing?

Don't forget to check both sides and document both proximal and distal muscle strength, tone, and reflexes. See more examples at CDC.gov/AFM/strength

