

19-ID-05**Committee:** Infectious Disease**Title:** Revision to the Standardized Case Definition, Case Classification, and Public Health Reporting for Acute Flaccid Myelitis

Check this box if this position statement is an update to an existing standardized surveillance case definition and include the most recent position statement number here: [17-ID-01](#).

Synopsis: This position statement updates the standardized case definition for acute flaccid myelitis.**I. Statement of the Problem**

Acute flaccid myelitis (AFM) is a rare but serious paralytic illness. A clinical case is defined as a person with sudden onset of acute flaccid limb weakness. The AFM case definition, as defined in CSTE position statement 17-ID-01, is based on gray matter lesions in the spinal cord. The causes of AFM remain largely unknown and no laboratory test is available for case confirmation. Furthermore, AFM is not nationally notifiable, which has led to diversity in reporting and interpretation of the legal authority under which AFM is reportable in jurisdictions. Without a standardized system for surveillance and reporting of cases of AFM by clinicians and jurisdictions, understanding baseline incidence and epidemiology of AFM and its public health impact in the United States remains challenging.

This position statement proposes to revise the standardized case definition to improve reporting consistency across jurisdictions and modify case classifications to further characterize the illness:

- The clinical criteria are modified to include patients with acute flaccid weakness in one or more limbs, plus any spinal cord lesion on MRI with at least some gray matter involvement, excluding persons with gray matter lesions in the spinal cord resulting from physician diagnosed malignancy, vascular disease, or anatomic abnormalities.
- The confirmed case classification is revised to include persons with spinal cord lesion predominantly in the gray matter and exclude persons with a clear alternative diagnosis to AFM.
- The probable case classification is revised to remove the requirement for pleocytosis and include persons with spinal cord lesion on MRI where gray matter predominance cannot be distinguished by national experts in AFM surveillance.
- The suspect case classification is created to include persons meeting the clinical criteria but for whom available information is insufficient to classify the case as confirmed or probable.

II. Background and Justification

Acute flaccid myelitis (AFM) is characterized by rapid onset of flaccid weakness in one or more limbs and distinct abnormalities of the spinal cord gray matter on magnetic resonance imaging (MRI). AFM is a subtype of acute flaccid paralysis (AFP), defined as acute onset of flaccid weakness absent features suggesting an upper motor neuron disorder. The term 'AFP' is a generalized 'umbrella' term, and includes multiple clinical entities including paralytic poliomyelitis, AFM, Guillain-Barré syndrome (GBS), acute transverse myelitis, toxic neuropathy, and muscle disorders. The annual rate of AFP among children under 15 years of age is approximately 1 per 100,000 children. Although AFP surveillance is commonly conducted in many countries currently still at risk for ongoing transmission of poliovirus, AFP is not under standardized surveillance or nationally notifiable in the United States. Routine surveillance and assessment for AFP has not been routinely performed since polio was eradicated from the U.S. In the summer and fall of 2014, an apparent increase in reports of AFM occurred in the U.S., and standardized surveillance was established in 2015 to monitor this illness and attempt to estimate baseline incidence (1). Data collected since establishment of standardized surveillance helped with identification of

subsequent increases in reports nationally during 2016 and 2018 and has provided additional valuable information on the clinical presentation to help better characterize clinical features and epidemiology of cases of AFM. Numerous viruses, including polioviruses, flaviviruses, and non-polio enteroviruses are known to be uncommon causes of AFM.

Since the summer/fall of 2014 through December 2018, 549 confirmed cases of AFM from 48 states and the District of Columbia have been reported to CDC, with peaks occurring in 2014, 2016, and 2018. All confirmed patients had distinctive abnormalities of the spinal cord gray matter on MRI, and a majority reported a respiratory or febrile illness in the days before onset of neurologic symptoms (2, 3). One fatality was reported in a confirmed case of AFM during the acute phase of illness in 2017.

Testing of biological specimens, including CSF, respiratory secretions, serum, and stool, has continued through 2018, without identification of a common etiology (4). CDC has expanded the search for potential causes of AFM by broadening laboratory approaches that test for potential infectious, noninfectious, and post-infectious causes, including possibly immune-mediated mechanisms or host responses to AFM. Testing protocols are also being developed to look for AFM biomarkers, and studies are being designed to identify possible mechanisms for AFM.

Although cases of AFM resemble polio clinically, they would not be considered paralytic poliomyelitis without meeting epidemiologic and laboratory criteria for polio (5). To date, all stool specimens from AFM patients tested at CDC have been negative for poliovirus. Without a biological marker to confirm cases of AFM, classification of cases is challenging. Therefore, as with polio (5), review of AFM case information by experts in national AFM surveillance provides consistency for classification of AFM cases.

III. Statement of the desired action(s) to be taken

CSTE recommends the following actions:

1. Implement a standardized surveillance case definition for Acute Flaccid Myelitis (AFM).
 - A. Utilize standard sources (e.g. reporting*) for case ascertainment for AFM. Surveillance for AFM should use the recommended sources of data to the extent of coverage presented in Section V.
 - B. Utilize standardized criteria for case ascertainment for AFM presented in Section VI and Table VI in Technical Supplement.
 - C. Utilize standardized criteria for case classification for AFM presented in Sections VII and Table VII in Technical Supplement.

*Reporting: process of a healthcare provider or other entity submitting a report (case information) of a condition under public health surveillance TO local, state, or territorial public health. Note: notification is addressed in a Nationally Notifiable Conditions Recommendation Statement and is the process of a local, state, or territorial public health authority submitting a report (case information) of a condition on the *Nationally Notifiable Conditions List* TO CDC.

IV. Goals of Surveillance

To provide a standard case definition to help standardize surveillance for states performing surveillance for AFM. Standardized surveillance for AFM will facilitate interpretation of apparent increases in this condition, better define the etiologic agent(s) and pathogenesis, and improve the tracking of national trends of AFM.

V. Methods for Surveillance: Surveillance for Acute Flaccid Myelitis (AFM) should use the recommended sources of data and the extent of coverage listed in Table V.

The primary source of data for AFM case ascertainment is clinician reporting to public health authorities. Clinicians should report cases meeting the clinical criteria for AFM as described in Section VI. Data from clinicians can be supplemented using data from electronic medical records, hospital discharge or outpatient records, and death certificates.

Table V. Recommended sources of data and extent of coverage for ascertainment of cases of Acute Flaccid Myelitis (AFM).

Source of data for case ascertainment	Coverage	
	Population-wide	Sentinel sites
Clinician reporting	X	
Laboratory reporting		
Reporting by other entities (e.g., hospitals, veterinarians, pharmacies, poison centers), specify:	X	
Death certificates	X	
Hospital discharge or outpatient records	X	
Data from electronic medical records	X	
Telephone survey		
School-based survey		
Vaccine registry		
Other, specify:		

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VI. Criteria for case ascertainment

A. Narrative: A description of suggested criteria for case ascertainment of a specific condition.

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

A1. Clinical Criteria for Reporting

- A person with onset of acute flaccid* limb weakness

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

A2. Laboratory/Imaging Criteria for Reporting

- A magnetic resonance image 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.

A3. Epidemiologic Linkage Criteria for Reporting

Not applicable

A4. 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.

B. 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
- Hospitalized (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.

VII. Case Definition for Case Classification

A. Narrative: Description of criteria to determine how a case should be classified.

A1. Clinical Criteria

- An illness with onset of acute flaccid* weakness of one or more limbs

A2. Laboratory/Imaging Criteria

Confirmatory laboratory/imaging evidence:

- MRI showing spinal cord lesion with predominant gray matter involvement[†] and 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

Presumptive laboratory/imaging evidence:

- MRI showing spinal cord lesion where gray matter involvement is present but predominance cannot be determined
- Excluding persons with gray matter lesions in the spinal cord resulting from physician diagnosed malignancy, vascular disease, or anatomic abnormalities

Supportive laboratory/imaging evidence:

- N/A

A3. Epidemiologic Linkage

N/A

A4. Case Classification

Confirmed:

- Clinically compatible case with confirmatory laboratory/imaging evidence, AND
- Absence of a clear alternative diagnosis attributable to a nationally notifiable condition

Probable:

- Clinically compatible case with presumptive laboratory/imaging evidence, AND
- Absence of a clear alternative diagnosis attributable to a nationally notifiable condition

Suspect:

- Clinically compatible case, AND
- Available information is insufficient to classify 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 (4).

** 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.

B. Criteria to distinguish a new case of this disease or condition from reports or notifications which should not be enumerated as a new case for surveillance

Not applicable.

VIII. Period of Surveillance

Surveillance should be ongoing.

IX. Data sharing/release and print criteria

Notification to CDC of all persons meeting the clinical criteria for AFM is recommended.

- States will send core/generic and disease specific data elements to CDC.
 - Data will be used to determine the burden of illness due to AFM and better define the etiologic agent(s) and pathogenesis of AFM.
 - The frequency of reports/feedback to the states and territories will be dependent on the current epidemiologic situation in the United States. Frequency of cases and other factors will influence communications.
1. CSTE recommends the following case statuses* be included in the ‘case’ count released outside of the public health agency:
 - Confirmed
 - Probable
 - Suspect
 - Unknown

* Which case statuses are included in the case counts constitute the “print criteria.”

2. Jurisdictions (e.g., States and Territories) conducting surveillance under this case definition can voluntarily submit de-identified case information to CDC, if requested and in a mutually agreed upon format.

Production of national data summaries and national data re-release for non-NNCs:

- Prior to release of national data summaries CDC should follow the CDC/ATSDR Policy on Releasing & Sharing Data, issued on April 16, 2003 and referenced in 11-SI-01 and custodians of such data should consult the CDC-CSTE Intergovernmental Data Release Guidelines Working Group report (<http://www.cste2.org/webpdfs/drgwgreport.pdf>) which contains data release guidelines and procedures for CDC programs re-releasing state, local, or territorial-provided data.
- CDC programs have a responsibility, in collaboration with states, localities, and territories, to ensure that CDC program-specific data re-release procedures meet the needs of those responsible for protecting data in the states and territories.

X. Revision History

Position Statement ID	Section of Document	Revision Description
17-ID-01	Statement of the Problem	ADDED additional clinical criteria to include any spinal cord lesion on MRI at least partially in the gray matter, and specific diagnoses that should be excluded for reporting. ADDED criteria to probable case classification to include persons with spinal cord lesion on MRI where gray matter predominance cannot be distinguished. REMOVED requirement for CSF pleocytosis for probable case classification. ADDED suspect classification.
17-ID-01	Goals of Surveillance	EDITED “to provide a standard case definition for states electing to perform surveillance for AFM” to “to provide a standard case definition to help standardize surveillance for states performing surveillance for AFM. Standardized surveillance for AFM will facilitate interpretation of apparent increases in this condition, better define the etiologic agent(s) and pathogenesis, and improve the tracking of national trends of AFM.”
17-ID-01	Methods for Surveillance	EDITED “surveillance for acute flaccid myelitis (AFM) should use the recommended sources of data and the extent of coverage listed in Table III” to “the primary source of data for AFM case ascertainment is clinician reporting to public health authorities. Clinicians should report cases meeting the clinical criteria for AFM as described in Section VI. Data from clinicians can be supplemented using data from electronic medical records, hospital discharge or outpatient records, and death certificates.” ADDED “Vaccine registry” as option for source of data ascertainment in Table V.
17-ID-01	Criteria for case ascertainment	ADDED additional clinical criteria to include any spinal cord lesion on MRI at least partially in the gray matter and specific diagnoses that should be excluded for reporting.

17-ID-01	Disease specific data elements to be included in the initial report	<p>ADDED clarification of meaning of “flaccid” as footnote.</p> <p>ADDED clarification of meaning of “flaccid” as footnote.</p> <p>REMOVED “Radiographic evidence of spinal cord lesion largely restricted to gray matter** and spanning one or more vertebral segments (if > 1 MRI performed, radiographic details of each MRI)**”</p> <p>REMOVED footnote “*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.”</p>
17-ID-01	Case Definition for Case Classification	<p>ADDED additional clinical criteria to include any spinal cord lesion on MRI at least partially in the gray matter and specific diagnoses that should be excluded for reporting.</p> <p>ADDED clarification of meaning of “flaccid” as footnote.</p> <p>ADDED criteria to confirmed case classification to include “absence of a clear alternative diagnosis”.</p> <p>ADDED criteria to probable case classification to include persons with spinal cord lesion on MRI where gray matter predominance cannot be distinguished.</p> <p>REMOVED requirement for CSF pleocytosis for probable case classification.</p> <p>ADDED suspect classification.</p> <p>REMOVED footnote “*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.”</p>
17-ID-01	Period of Surveillance	<p>EDITED “Surveillance should be ongoing. Reporting should be provided as soon as all necessary data have been ascertained and collected in completed form” to “Surveillance should be ongoing.”</p>
17-ID-01	Data Sharing/release and print criteria	<p>EDITED “Data may be used to measure the burden of AFM” to “Notification to CDC of all persons meeting the clinical criteria for AFM is recommended.</p> <ul style="list-style-type: none"> • States will send core/generic and disease specific data elements to CDC. • Data will be used to determine the burden of illness due to AFM and better define the etiologic agent(s) and pathogenesis of AFM. • The frequency of reports/feedback to the states and territories will be dependent on the current epidemiologic situation in the United States. Frequency of cases and other factors will influence communications.”
17-ID-01	References	<p>ADDED additional reference (McKay, et al)</p>
17-ID-01	Appendix	<p>ADDED appendix to include examples of additional information necessary for case classification as referenced in Sections VI.B and VII.</p>

XI. References

1. CSTE. Revision to the Standardized Surveillance and Case Definition for Acute Flaccid Myelitis. <https://cdn.ymaws.com/www.cste.org/resource/resmgr/2017PS/2017PSFinal/17-ID-01.pdf>.

2. Sejvar JJ, Lopez AS, Cortese MM, et al. Acute Flaccid Myelitis in the United States, August-December 2014: Results of Nationwide Surveillance. Clin Infect Dis. 2016;63:737-45.
3. Lopez A, Lee A, Guo A, Konopka-Anstadt JL, Nisler A, Rogers SL, Emery B, Nix WA, Oberste S, Routh J, Patel M. Vital Signs: Surveillance for Acute Flaccid Myelitis – United States, 2018. MMWR Morb Mortal Wkly Rep. 2019; 68:608-14.
4. McKay, SL, Lee AD, Lopez AS, et al. Increase in Acute Flaccid Myelitis – United States, 2018. MMWR Morb Mortal Wkly Rep. 2018;67:1273-75.
5. CSTE. National Surveillance for Paralytic Poliomyelitis and Nonparalytic Poliovirus Infection. <http://c.ymcdn.com/sites/www.cste.org/resource/resmgr/PS/09-ID-53.pdf>.

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Appendix. Examples of further information necessary for case classification as referenced in Sections VI.B, and VII.

Clinical information necessary for classification of cases meeting the clinical criteria for AFM include:

- 1) Complete neurology consultation notes
- 2) Documentation and results of tone exam
- 3) Documentation and results of reflex exam
- 4) Documentation and results of motor exam
- 5) Images from spinal and brain MRI

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

Criterion	Acute Flaccid Myelitis (AFM)
<i>Clinical Criteria for Reporting</i>	
Acute flaccid* weakness of one or more limbs	S
A magnetic resonance image (MRI) showing spinal cord lesion in at least some of the gray matter† and spanning one 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
<i>Vital Records Criteria for Reporting</i>	
Any person whose death certificate lists acute flaccid myelitis as a cause of death or a condition contributing to death.	S

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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.*

Table VII. Classification Table: Criteria for defining a case of Acute Flaccid Myelitis (AFM).

Criterion	Suspected	Probable	Confirmed
<i>Clinical Evidence</i>			
Acute flaccid* weakness of one or more limbs	N	N	N
Absence of a clear alternative diagnosis		N	N
<i>Laboratory/Imaging Evidence</i>			
MRI showing spinal cord lesion with predominant gray matter involvement† and spanning one or more vertebral segments			N
MRI showing spinal cord lesion where gray matter involvement is present but predominance cannot be determined		N	
Excluding persons with gray matter lesions in the spinal cord resulting from physician diagnosed malignancy, vascular disease, or anatomic abnormalities		N	N
Insufficient information to classify case as probable or confirmed	N		

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Notes:

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*

† *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.*