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Acute Flaccid Myelitis (AFM) 2022 Case Definition | CDC  
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NOTE:  
A surveillance case definition is a set of uniform criteria used to define a disease for public health surveillance. Surveillance case definitions enable public health officials to classify and count cases consistently across reporting jurisdictions. Surveillance case definitions are not intended to be used by healthcare providers for making a clinical diagnosis or determining how to meet an individual patient’s health needs.  
CSTE Position Statement(s)  
21-ID-02  
Background  
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 of 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. 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. Standardized surveillance was established in 2015 to monitor this illness and attempt to estimate baseline incidence (1). Data collected since standardized surveillance was established have helped to identify subsequent increases in reports nationally during 2016 and 2018 and have provided additional valuable information on the clinical presentation to help better characterize clinical features and epidemiology of cases of AFM.  
From the summer/fall of 2014 through December 2020, 650 confirmed cases of AFM from 49 states and the District of Columbia were reported to the Centers for Disease Control and Prevention (CDC), with peaks occurring in 2014, 2016, and 2018. All confirmed patients had distinctive abnormalities of the spinal cord gray matter on MRI (2), and a majority reported a respiratory or febrile illness in the days before onset of neurologic symptoms (3, 4, CDC unpublished data). One fatality was reported in a confirmed case of AFM during the acute phase of illness in 2017.  
Testing of biological specimens, including cerebrospinal fluid (CSF), respiratory secretions, serum, and stool, has continued through 2020, without identification of a common etiology (3, 4, CDC unpublished data). Numerous viruses, including polioviruses, flaviviruses, and non-polio enteroviruses have been associated with AFM, but viral isolation from cases is not consistent or common. However, data collected since 2014 suggest that enteroviruses, specifically enterovirus D68 (EV-D68), are important factors in the epidemiology of AFM. Although the CDC AFM laboratory has expanded its focus from direct pathogen detection to identification of indirect evidence for infection and possible immune correlates of disease, exploration of the relationship between EV-D68 and AFM continues (5). 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 (6). 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 (6), review of AFM case information by experts in national AFM surveillance provides consistency for classification of AFM cases.  
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  
Suspect  
Meets clinical criteria with supportive laboratory/imaging evidence,  
AND  
Available information is insufficient to classify case as probable or confirmed.  
Probable  
Meets clinical criteria with presumptive laboratory/imaging evidence.  
Confirmed  
Meets clinical criteria with confirmatory laboratory/imaging evidence,  
OR  
Meets other classification criteria.  
Other Criteria  
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,  
AND  
Excluding persons with gray matter lesions in the spinal cord resulting from physician diagnosed malignancy, vascular disease, or anatomic abnormalities,  
AND  
Absence of a clear alternative diagnosis attributable to a nationally notifiable condition.\*\*  
\*\* 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.  
Case Classification Comments  
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 (6).  
Related Case Definition(s)  
Acute Flaccid Myelitis (AFM) | 2020 Interim Case Definition, Approved October 9, 2020  
Acute Flaccid Myelitis (AFM) | 2020 Case Definition  
Acute Flaccid Myelitis (AFM) | 2018 Case Definition  
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NNDSS receives and shares case data from state, local, and territorial health departments to help public health monitor, control, and prevent serious diseases.  
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