Key Points—Acute Flaccid Myelitis in the U.S., 2014-2016

Note: Newly added information is in red.

Key Points

- CDC continues to receive reports of acute flaccid myelitis (AFM) cases. From January 1 to September 30, 2016, a total of 89 people in 33 states across the country were confirmed to have AFM.
  - Most patients are children.
- While the AFM case count for 2016 is less than the 2014 case count, CDC is concerned about the increase in cases in recent months. However, it is currently difficult to interpret trends in AFM data since reporting started in 2014 and is voluntary. Since AFM reporting is relatively new, there initially may be more variability in the data from year to year making it difficult to interpret or compare.
- However, AFM remains a very rare disease (less than one in a million) even with an increase in cases.
- CDC has been actively investigating the AFM cases, testing specimens, and monitoring disease activity since 2014, when we first received an increase of case reports from August to December of that year. We are working closely with health professionals to increase awareness and reporting for AFM, and to better understand the AFM cases, risk factors, and possible causes of this illness.
- The cause of the AFM cases is not known. However, the symptoms are most similar to those caused by some viruses, including poliovirus, other enteroviruses, adenoviruses, and West Nile virus.
- CDC understands that Americans may be concerned about this AFM situation. While AFM is rare, severe illness is always a concern to us. We'll continue sharing information as soon as we have it, and post updates on our AFM website.
- There are no specific ways to prevent this AFM illness. However, CDC recommends being up to date on all recommended vaccinations including polio, maintaining good hygiene through washing your hands often with soap and water, avoiding close contact with sick people, and disinfecting frequently touched surfaces, and protecting yourself from mosquito-borne viruses, such as West Nile virus, by using mosquito repellent, wearing long sleeves and long pants, and staying indoors at dusk and dawn.

Additional Points

Case reports and investigations

2016

- From January 1 to September 30, 2016, a total of 89 people in 33 states across the country were confirmed to have AFM.

2015 cases

- From January 1 to December 31, 2015, 21 people in 16 states from across the country were confirmed to have AFM.

2014 cases

- From August 1 to December 31 2014, CDC verified reports of 120 children in 34 states who developed AFM. Most of these cases occurred between August 1 and October 31, 2014.
  - The median age of the children was about 7 years.
  - Nearly all were hospitalized; some were put on breathing machines.
  - Most patients had fever and/or respiratory illness before onset of neurologic symptoms.
  - About 7 out of 10 of the children had elevated white blood cell counts, often with elevated protein levels, in their spinal fluid.

  - About two thirds of the children who have been observed (median 19 days) after their illness reported some improvement in symptoms, though it is not clear what amount of improvement occurred. About one third showed no improvement. Ten children had full recovery of limb function and 3 children had full recovery of strength.
  - CDC and others have tested many different specimens collected from the children with AFM to check for a wide range of pathogens that can result in AFM. However, the cause of the AFM cases has not yet been identified, and is still being investigated.
  - The increase in reports of AFM cases seen in August-October 2014 occurred at the same time as a national outbreak of enterovirus-D68 (EV-D68); however, the relationship between EV-D68 and AFM is still being investigated. Among the people with AFM, CDC did not consistently detect EV-D68 in the specimens collected from them. In 2015 there were no cases of EV-D68 detected and so far in 2016, only limited sporadic cases of EV-D68 have been detected in the United States.
- In 2016, CDC published results from a retrospective case-control study conducted in 2014 among cases of AFM reported by Colorado from August to October, 2014 to determine any association between EV-D68 infection and AFM.
  - The case-control study showed an epidemiologic association between EV-D68 and AFM. However, an association does not prove that EV-D68 caused AFM in this cluster of cases given the limitations of the study. AFM is rare, and there were only 11 cases available to be enrolled in this study. Although EV-D68 was detected in the nasal cavity of 4 children with AFM, it was not detected in cerebrospinal fluid (CSF). Detection of EV-D68 in the nasal cavity does not prove causation of AFM. Virus detected in the CSF would have been a more definitive link to EV-D68. These data suggest a possible causal link, but there is still a gap in epidemiologic data and data from extensive lab testing.

Symptoms

- AFM is a syndrome characterized by sudden onset of limb weakness, sometimes accompanied by cranial nerve dysfunction (such as facial drooping or difficulty speaking). In most cases, distinctive lesions in the gray matter (nerve cells) of the spinal cord may be seen on neuroimaging.

Causes

- While the specific causes of AFM cases reported since August 2014 are still being investigated, the AFM cases are most similar to those caused by viruses, including poliovirus, other enteroviruses, adenoviruses, and West Nile virus. A condition where the body's immune system attacks and destroys body tissue that it mistakes for foreign material may also cause AFM.
  - Enteroviruses can cause neurologic illness, including meningitis, however, more severe disease such as encephalitis, and acute flaccid myelitis are not common. Rather, these viruses most commonly cause mild illness.
  - It is unclear what pathogen (germ) or immune response is causing the disruption of signals sent from the nervous system to the muscles causing weakness in the arms and legs.

Prevention

- There are no specific ways to prevent this AFM illness. However, CDC recommends the following:
  - be up to date on all recommended vaccinations, including polio, which can cause acute flaccid paralysis, and numerous other vaccine-preventable diseases that can cause severe illness and death.
  - protect yourself and others from viral infections in general by
    - washing your hands often with soap and water,
    - avoiding close contact with sick people, and
    - disinfecting frequently touched surfaces.
  - protect yourself from mosquito-borne viruses, such as West Nile virus, by using mosquito repellent, wearing long sleeves and long pants, and staying indoors at dusk and dawn, which is the prime period that mosquitoes bite.
- CDC has not yet determined who is at higher risk for developing AFM, or the reasons why they may be at higher risk.

Treatment

- There is no specific treatment for AFM.

Guidance for Health Professionals

Clinicians should

- be vigilant for and immediately report to their state or local health department any patients who meet the following case definitions, using a patient summary form available on CDC's website (www.cdc.gov/acute-flaccid-myelitis/hcp/data.html):

  Confirmed case
  1. Acute onset of focal limb weakness, AND
  2. An MRI showing a spinal cord lesion largely restricted to gray matter and spanning one or more spinal segments.

  Probable case
  1. Acute onset of focal limb weakness, AND
  2. Cerebrospinal fluid (CSF) with pleocytosis (white blood cell count >5 cells/mm^3).

- report patients meeting the case definition regardless of any laboratory results.
- consult with their local and state health department for laboratory testing of stool, respiratory and cerebrospinal fluid specimens for enteroviruses (including poliovirus), West Nile virus and other known infectious etiologies for patients meeting the above case definitions.
- collect specimens from patients suspected of having AFM as early as possible in the course of illness. Additional instructions regarding specimen collection and shipping can be found at: www.cdc.gov/acute-flaccid-myelitis/hcp/instructions.html.
- refer to CDC's "Interim Considerations for Clinical Management of Patients with Acute Flaccid Myelitis," released November 7, 2014 with consensus from experts in infectious diseases, neurology, pediatrics, critical care medicine, public health epidemiology and virology (www.cdc.gov/acute-flaccid-myelitis/downloads/acute-flaccid-myelitis.pdf).

Health departments should

- report patients meeting the case definition to CDC using a brief patient summary form available on the CDC website (www.cdc.gov/acute-flaccid-myelitis/hcp/data.html). Cases meeting the definition should be reported regardless of any laboratory results.
- ship available clinical specimens to CDC as soon as possible after case identification, so CDC can test and monitor these cases in as real time as possible
- contact CDC by email to arrange further laboratory testing or to discuss any additional questions (limbweakness@cdc.gov)

Guidance for the General Public

- If a child appears to have a sudden onset of weakness in arms or legs, caregivers should contact a healthcare provider to have the child assessed for possible neurologic illness.
- 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 that can cause severe illness and death.
- You can help protect yourself and others from viral infections in general by
  - washing your hands often with soap and water,
  - avoiding close contact with sick people, and
  - disinfecting frequently touched surfaces.
- You can protect yourself from mosquito-borne viruses, such as West Nile virus, by using mosquito repellent, and staying indoors at dusk and dawn, which is the prime period that mosquitoes bite.

What CDC is Doing

CDC has closely monitored the AFM situation since August 2014, when we first received case reports from hospitals and health departments. We are actively working with healthcare professionals and state and local health departments to increase awareness and reporting for AFM, and investigate the AFM cases, risk factors, and possible causes of this illness.

Activities that CDC is doing include:

- encouraging healthcare providers to be vigilant for AFM among their patients, and to report suspected cases to their health departments
- verifying reports of suspected AFM cases submitted by health departments using a case definition adopted by the Council of State and Territorial Epidemiologists (CSTE)
- testing specimens, including stool, blood, respiratory and cerebrospinal fluid, from people confirmed to have AFM. Despite extensive testing, CDC does not yet know the cause of the AFM cases.
- working with clinicians and state and local health departments to investigate and better understand the AFM cases, including potential causes and how often the illness occurs
- providing new and updated information to clinicians, health departments, policymakers, the public, and partners in various formats, such as the *Morbidity and Mortality Weekly Report*, the AFM website, and CDC social media
- pursuing an approach that uses multiple research methods to further explore the potential association of AFM with possible causes as well as risk factors for AFM. This includes collaborating with several medical institutions to review MRI scans of children from the past 10 years to determine how many AFM cases occurred before 2014.

AFM surveillance

- AFM is not a nationally notifiable disease, which means that clinicians are not required by law to report known or suspected AFM cases to federal government authorities. Clinicians are voluntarily reporting AFM cases to their health departments using a patient summary form provided by CDC. Health departments submit AFM case reports to CDC and are responsible for these data.

- AFM case reports are submitted by health departments using a standard CSTE case definition. CDC collects and verifies data from these reports in a standardized surveillance system. The main purpose of collecting these data is to better understand the AFM cases, risk factors, and potential causes of the illness.

- CDC continues to analyze AFM data to establish baseline data and assess possible seasonal and annual trends. However, as with new surveillance systems like AFM, there is initially more variability in the data from year to year making it difficult to interpret or compare.
  - For example, variability may be due to changes in awareness among or reporting by states and healthcare providers or changes in case definitions, such as the age limit removal from the AFM case definition in 2015.

- AFM surveillance is unique because there are many known causes of AFM, and none of them have been consistently found in specimens from the 2014, 2015, and 2016 cases. Also, since AFM is a syndrome, surveillance is based on evaluating a collection of symptoms for each case.

- The AFM case counts posted on AFM website represent those cases reported to and confirmed by CDC.
  - The case counts are updated monthly with a one month lag. This allows for the time needed for case review. Confirming an AFM case is an involved process that includes thorough review of patient medical records, MRIs, and consulting with the patient's doctors. For example, the AFM case count for August will be posted on the website in the first week of October.
  - The AFM case counts do not represent real-time reporting. Also, any increases in cases do not reflect changes in real time or mean that the situation is getting worse.
  - CDC is only including the number of states with confirmed cases on our website to protect patient confidentiality, and defers to states to release specific information as they choose.

More information

- Enterovirus D68 Infection in Children with Acute Flaccid Myelitis, Colorado, USA, 2014, EID, August 2016 (wwwnc.cdc.gov/eid/article/22/8/15-1949_article)
- Acute Flaccid Myelitis in the United States—August – December 2014: Results of Nation-Wide Surveillance, CID, June 17, 2016 (http://cid.oxfordjournals.org/content/early/2016/06/17/cid.ciw372.short?rss=1)
- Notice to Clinicians: Continued Vigilance Urged for Cases of Acute Flaccid Myelitis COCA Clinical Reminder, August 27, 2015 (http://emergencylink.cdc.gov/coca/reminders/2015/2015aug27.asp)
- CDC Acute Flaccid Myelitis website: www.cdc.gov/acute-flaccid-myelitis/index.html
- Acute Flaccid Myelitis Among Persons Aged ≤21 Years— United States, August 1–November 13, 2014, MMWR, January 9, 2015 (http://www.cdc.gov/mmwr/preview/mmwrhtml/mm6353a3.htm?s_cid=mm6353a3_w)

- Acute Neurologic Illness of Unknown Etiology in Children — Colorado, August—September, 2014, MMWR, October 3, 2014 (www.cdc.gov/mmwr/preview/mmwrhtml/mm6340a5.htm)
- Acute Neurologic Illness with Focal Limb Weakness of Unknown Etiology in Children, Health Alert Network, September 26, 2014 (http://emergency.cdc.gov/han/han00370.asp)
- Neurologic Illness with Limb Weakness in Children, COCA Call, October 3, 2014 (http://emergency.cdc.gov/coca/calls/2014/callinfo_100314.asp)