Kentucky Public Health
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Acute Flaccid Myelitis

Introduction

Following the increased number of reports of acute flaccid myelitis (AFM) among children that were received by the U.S. Centers for Disease Control and Prevention (CDC) during August–October 2014, CDC has continued to receive sporadic reports of AFM. The apparent increase in AFM cases in 2014 coincided with a national outbreak of severe respiratory illness among children caused by enterovirus-D68 (EV-D68), which resulted in an increased number of children hospitalized. While recent analysis has demonstrated an association between the EV-D68 outbreak and the AFM cases, an etiology for the 2014 AFM cases has not been fully elucidated. As of July 2015, CDC had verified reports of 120 children in 34 states who developed acute flaccid myelitis that met CDC's outbreak case definition. Of note, since January 1, 2016 through July 31, 2016, CDC has confirmed an additional 27 AFM cases out of 49 suspected reports. One of these suspected cases was from Kentucky. Therefore, the CDC and the Kentucky Department for Public Health (KDPH) are re-emphasizing the importance of continued vigilance by clinicians in identifying cases of AFM among all age groups, irrespective of enterovirus status. Reporting of these cases will help public health officials monitor for increases of this illness and better understand potential causes, risk factors, and preventive measures or therapies.

Background

Acute flaccid myelitis (AFM) is a condition that affects the nervous system, specifically the gray matter of the spinal cord. AFM is one of a number of the conditions that can result in neurologic illness with limb weakness. It is one of the clinical entities of acute flaccid paralysis (AFP), http://epirev.oxfordjournals.org/content/22/2/298.full.pdf. AFM is similar in clinical presentation to illness due to poliovirus, but can result from a variety of causes including viral infections, environmental toxins, genetic disorders, and Guillain-Barre syndrome, a neurologic disorder caused by an abnormal immune response that attacks the body's nerves. Oftentimes, however, despite extensive laboratory testing, an etiology for AFM is unable to be identified.

Symptoms

Most patients will have sudden onset of limb weakness 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, or
- Difficulty swallowing or slurred speech.

Numbness or tingling is rare in patients with AFM, though some patients may have pain in their arms or legs. Some patients with AFM may have urinary retention. The most severe symptom of AFM is respiratory failure secondary to involvement of the respiratory musculature. This respiratory failure can require urgent ventilator support.

Diagnosis

A physician can differentiate between AFM and other diseases with a detailed examination of the nervous system, specifically defining the location of weakness, loss of muscle tone, and changes in reflexes. Magnetic resonance imaging (MRI) can be very helpful in diagnosing cases of AFM.

Testing nerve response can also be helpful in supporting a diagnosis of AFM; it is important that the tests are performed at the appropriate time (e.g., 7-10 days after onset of weakness) to be helpful. Finally, by testing the cerebrospinal fluid (and other suggested specimens including stool, blood and respiratory, as defined by the CDC) appropriate information can be obtained to assist in making a diagnosis of AFM.

Specimen Collection and Testing

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.

Specimens should include (*Note. specimens are for investigational purposes and are not intended for timely clinical management*):

- Cerebrospinal fluid (CSF);
- Blood (serum and whole blood);
- A nasopharyngeal aspirate, nasopharyngeal wash, or nasopharyngeal swab with lower respiratory specimen if indicated, and an oropharyngeal swab; and
- Stool for poliovirus testing.

Available clinical specimens should be shipped in insulated containers using cold packs to the Kentucky State Public Health Laboratory, known as the KDPH Division of Laboratory Services (DLS). Specimens will be forwarded to CDC for testing. Clinicians are encouraged to consult with staff at DLS via telephone at 502-564-4446 for further information on handling and processing of samples.

Case Definitions

In June 2015, the Council of State and Territorial Epidemiologists (CSTE) adopted a **standardized case definition for acute flaccid myelitis**, http://c.ymcdn.com/sites/www.cste.org/resource/resmgr/2015PS/2015PSFinal/15-ID-01.pdf. As of August 1, 2015, a patient must meet the clinical criteria below to be classified as either a confirmed or a probable case of acute flaccid myelitis:

Confirmed Case

To be classified as a confirmed case, a patient must meet the following criteria:

1. Acute onset of focal limb weakness, AND
2. An MRI showing a spinal cord lesion largely restricted to gray matter (i.e., terms in the spinal cord MRI report such as “affecting mostly gray matter,” or “affecting the anterior horn or anterior horn cells) and spanning one or more spinal segments.

Probable Case

To be classified as a probable case, a patient must meet the following criteria:

1. Acute onset of focal limb weakness, AND
2. Cerebrospinal fluid (CSF) with pleocytosis (i.e., white blood cell count >5 cells/mm^3, adjusting for presence of red blood cells by subtracting 1 white blood cell for every 500 red blood cells present).

Infectious Agents Associated with AFM

Acute flaccid myelitis can be caused by a variety of agents including several viruses:

- Enteroviruses (**poliovirus** and **non-poliovirus**),
- West Nile virus (WNV) and viruses in the same family as WNV, specifically Japanese encephalitis virus and Saint Louis encephalitis virus,
- Herpesviruses, such as **cytomegalovirus** and **Epstein-Barr virus**, and **Adenoviruses.**

Treatment

There is no specific treatment for acute flaccid myelitis but a neurologist may recommend certain interventions on a case-by-case basis. Medical providers are encouraged to use CDC's guidance in the management of patients with AFM, http://www.cdc.gov/acute-flaccid-myelitis/downloads/acute-flaccid-myelitis.pdf.

Prevention

Physicians are requested to provide general preventive measures to their patients such as: 1) encouraging compliance with recommended vaccinations, including **poliovirus**; 2) use of mosquito vector control; 3) observation of personal and/or hand hygiene; and 4) avoiding contact with sick people.

Reporting

Clinicians suspecting AFM in patients meeting the probable or confirmed case definition (irrespective of laboratory testing results) are asked to report these cases to their local health department or to the KDPH Division of Epidemiology and Health Planning at 502-564-3418 or 888-9REPORT (888-973-7678). KDPH also requests that clinicians:

- Consult with the KDPH Division of Epidemiology and Health Planning at 502-564-3418 or 888-9REPORT (888-973-7678) regarding laboratory testing of CSF, blood, serum, respiratory, and stool specimens for enteroviruses, West Nile virus, and other known infectious etiologies, and
- Complete the CDC AFM Patient Summary Form, http://www.cdc.gov/acute-flaccid-myelitis/hcp/data.html, for cases classified as confirmed or probable and submit to KDPH via secure fax at 502-696-3803 to the attention of AFM Surveillance.

For additional information

- About AFM: http://www.cdc.gov/acute-flaccid-myelitis/about-afm.html.
- Acute flaccid myelitis of unknown etiology in California, 2012-2015. Van Haren K. et. al. 2015. ***JAMA*** (http://jama.jamanetwork.com/article.aspx?articleid=2478202).
- MRI findings in children with acute flaccid paralysis and cranial nerve dysfunction occurring during the 2014 enterovirus D68 outbreak. Maloney JA et al 2015. ***Amer. J. of Neuroradiology***. (http://www.ajnr.org/content/36/2/245.full.pdf+html)
- Acute Flaccid Myelitis in the United States, August– December 2014: Results of Nationwide Surveillance http://cid.oxfordjournals.org/content/early/2016/06/25/cid.ciw372.full.pdf+html
- Enterovirus D68 Infection in Children with Acute Flaccid Myelitis, Colorado, USA, 2014 http://wwwnc.cdc.gov/eid/article/22/8/pdfs/15-1949.pdf
- Education and resource materials: http://www.cdc.gov/acute-flaccid-myelitis/references.html
- Specimen collection: http://www.cdc.gov/acute-flaccid-myelitis/hcp/specimens.html