Key Points: Acute Flaccid Paralysis with Anterior Myelitis – California, June 2012 – June 2014, MMWR

- In August 2012, the California Department of Health (CDPH) was contacted by a San Francisco Bay area clinician who requested poliovirus testing for a man aged 29 years with acute flaccid paralysis (AFP) associated with anterior myelitis (inflammation of the grey matter of the spinal cord). The man had no history of recent travel during the month before the onset of symptoms.
- Within two weeks, CDPH received reports of two additional cases of AFP with anterior myelitis of unknown cause. To identify more cases and determine common causes, CDPH posted alerts in official communications with California local health departments during December 2012, July 2013, and February 2014. Reports of cases of neurologic illness received by CDPH were screened throughout this period, and clinicians were encouraged to submit clinical samples for testing.
- A case was defined as AFP in at least one limb with lesions to the grey matter of the spinal cord and no known cause. During January 2012-June 2014, 23 cases met the criteria.
- After laboratory testing, the cause of AFP in these cases remains undetermined.

General Key Points about AFP

Acute Flaccid Paralysis (AFP)

- AFP is a spectrum of multiple clinical diseases characterized by a sudden onset of limb weakness or paralysis and reduced muscle tone without another obvious cause such as trauma. AFP with anterior myelitis is commonly seen in cases of paralytic poliomyelitis (polio), which is why it's important to rule out polio early during the course of disease.
- AFP can result from a variety of infectious and noninfectious causes. Guillain-Barre syndrome, a disorder caused by an abnormal immune response, is a leading cause of AFP. Viral causes of AFP include enterovirus (polio and non-polio), adenovirus, and West Nile virus. However, these viruses do not commonly cause AFP in people in the United States. It is challenging to identify the cause of AFP, especially with respect to infectious agents. In a significant proportion of cases, the cause is never determined.
- AFP is not a national notifiable disease in the United States, and Centers for Disease Control and Prevention (CDC) does not have a specific surveillance system to collect data on this condition.
- Among children under 15 years of age, in the absence of polio, AFP typically occurs at a rate of at least 1 per 100,000 children annually. Since there are 62 million children under 15 years of age in the US, at least 620 cases of AFP in this age group are likely to occur each year throughout the country.
- Even using advanced clinical diagnostic and laboratory technology, in most cases of suspected infectious AFP, the cause is never determined.

Infectious causes of AFP

- There are more than 100 types of enteroviruses and many types have been routinely detected globally from patients with AFP. Since they are also detected at similar rates in healthy children, they are only rarely thought to be responsible for paralysis. Therefore, a positive identification of an enterovirus in a non-sterile site, such as a throat or stool specimen, is often an incidental finding.

- Enteroviruses most commonly cause aseptic meningitis (inflammation of the tissue covering the brain and spinal cord), less commonly encephalitis (inflammation of the brain), and rarely, acute myelitis and paralysis.

- Enterovirus-D68 was first described in 1962 and infections occur worldwide. Reports over the last decade have routinely associated EV-D68 with respiratory disease, some of which can be severe. CDC is aware of two published reports of children with neurologic illnesses confirmed as EV-D68 infection from cerebrospinal fluid (CSF) testing.

- The emergence of West Nile virus (WNV) in North America in 1999 was unexpected, and WNV quickly became a major public health problem in the United States. Although most persons infected with WNV develop no symptoms at all, or develop a mild illness with fever, a small proportion (roughly 1%) of patients will have infection of the nervous system, including meningitis and encephalitis. While uncommon, WNV anterior myelitis cases are detected nearly every WNV season in North America.

- There are several other viruses that may produce infectious anterior myelitis. Certain strains of adenoviruses have been known to produce AFP with anterior myelitis. Herpesviruses are also a recognized cause of anterior myelitis.

Guidance for the General Public

- Being up to date on all recommended vaccinations is the best way to protect you and your family from serious diseases including polio and acute respiratory illnesses including influenza, measles and whooping cough.

- The general public can help protect themselves from infections in general by washing hands often with soap and water, avoiding close contact with sick people, and disinfecting frequently touched surfaces. Mosquito-borne viruses such as WNV can be avoided by protecting oneself from being bitten by mosquitoes (using mosquito repellent, avoiding being outdoors at dusk and dawn, the prime period for mosquitoes to bite).

- If a child appears very sick or seems to demonstrate features of acute limb weakness, caretakers should contact their physicians in order to have the child assessed for possible AFP.

Guidance for Healthcare Professionals

- Although poliovirus is no longer endemic in the United States, it's important to rule out poliovirus infection in cases of AFP that are clinically compatible with polio, particularly those with anterior myelitis, to ensure that any importation of poliovirus is quickly identified and investigated. The World Health Organization and CDC have guidelines for epidemiologic, clinical, and laboratory investigations of AFP to rule out poliovirus as a cause.

- Clinical and epidemiologic investigation should include a careful neurologic exam to characterize specific sensory as well as motor findings; ask patients about recent travel (<30 days before onset) and contact with persons who recently traveled, particularly to regions with polio cases or regions using (oral polio vaccine) OPV. Documented history of vaccination and which polio vaccine was administered should be noted, including dates of administration, number of doses, and manufacturer, if the information is available.

- Specimens should be collected early during the course of disease for laboratory testing. Collection of specimens should follow CDC and WHO guidelines, which include:

    - two stool samples >24 hours apart and <14 days after symptom onset;
    - serum before administration of IVIG;
    - and throat swabs.

- Of patients who had samples tested at CDPH laboratory described here, none met the specifications for ruling out poliovirus infection as recommended by WHO or CDC guidelines. All lacked two stool samples >24 hours apart and <14 days after paralysis onset.

- Paralytic polio cases are immediately reportable to state and local health departments. A confirmed paralytic polio case should be reported to CDC <4 hours after meeting notification criteria. Non-paralytic poliovirus infections are reportable to CDC < 24 hours of detection or isolation.

- Although AFP with anterior myelitis comprises a subset of patients with AFP, these cases can be challenging to distinguish before clinical, imaging, and laboratory results are available. Thus, specimen collection to definitively rule out poliovirus infection from possible alternative diagnoses should be considered among all patients with AFP of unknown cause or a suspected viral cause. This is particularly important for persons, regardless of immunization history, with a history of foreign travel to countries with endemic polio and/or unimmunized persons with a history of travel to countries that use OPV for routine immunization.

    - In 2005, an unimmunized college student acquired vaccine-associated paralytic poliomyelitis (VAPP) in Costa Rica.

- Physicians treating patients presenting with AFP of unknown cause should work with their local and state health departments to rule out polio early during the course of disease. To ensure adequate samples for poliovirus testing, specimens should be collected according to CDC and WHO guidelines.

- CDC can provide assistance for laboratory testing for other potential infectious agents and for unknown cause using pathogen discovery approaches, such as metagenomics.

Links:

- Non-Polio Enteroviruses: http://www.cdc.gov/non-polio-enterovirus/index.html
- 2010 Enterovirus MMWR: http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5948a2.htm.
- 2006 Enterovirus MMWR: http://www.cdc.gov/mmwr/preview/mmwrhtml/ss5508a1.htm.
- 2011 Clusters of Acute Respiratory Illnesses EV68: http://www.cdc.gov/mmwr/preview/mmwrhtml/mm6038a1.htm
- CDC Polio Guidelines: http://www.cdc.gov/vaccines/pubs/surv-manual/chpt12-polio.html.
- WHO Polio Guidelines: https://www.hpsc.ie/hpsc/A-Z/VaccinePreventable/Polio/Guidance/File,2461,en.pdf.
- Guillain-Barre Syndrome: http://www.ninds.nih.gov/disorders/gbs/gbs.htm
- West Nile Virus: http://www.cdc.gov/westnile