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Philadelphia Department of Public Health

Division of Disease Control

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Health Advisory

Acute Flaccid Myelitis: Identification, Testing, and Reporting Guidelines September 3, 2015

SUMMARY POINTS

- To better understand the epidemiology of unexplained acute flaccid myelitis (AFM), healthcare providers should report any suspected cases of AFM to the Philadelphia Department of Public Health (PDPH)
- Cerebrospinal fluid (CSF), blood, stool, and respiratory specimens should be collected as close to illness onset as possible for laboratory testing. PDPH is available to coordinate testing of clinical specimens.

Last fall, a nationwide increase in acute flaccid myelitis (AFM) was observed. Clinical findings suggested a viral process affecting the spinal cord and producing a polio-like illness. To better understand the epidemiology of AFM, the Philadelphia Department of Public Health (PDPH) is requesting that providers report cases of AFM and collect specimens for testing at public health reference laboratories.

The number of individuals who develop AFM annually in the United States is not well understood. An apparent increase in cases of AFM occurred in August–October 2014, coinciding with a nationwide outbreak of Enterovirus D-68. As of July 2015, 120 pediatric cases of AFM in 34 states have been confirmed by CDC, of which 5 were treated at Philadelphia hospitals. The median age of all cases nationwide is approximately 7 years (range 5 months to 20 years). Almost all patients have been hospitalized with some requiring ventilation and only two completely recovering. Because the link between AFM and a specific pathogen is not yet proven, continued surveillance and investigation is necessary.

Clinical Syndrome & Management

Patients with AFM typically present with acute flaccid paralysis (AFP) of one or more limbs (often asymmetric) and cranial nerve weakness following a febrile illness. Limb weakness tends to progress rapidly with maximal weakness occurring between a few hours and days from symptom onset. Cranial nerve involvement can manifest as facial weakness, paralysis of the eye muscles, and difficulty speaking or swallowing. Spinal lesions identified by MRI are largely restricted to the grey matter of the cord and do not appear to affect the white matter, although lesions can span anterior and posterior segments of the spinal cord. These aspects distinguish AFM from other central nervous system disorders such as Guillain-Barre syndrome and transverse myelitis. Laboratory analyses of cerebrospinal fluid (CSF) demonstrate moderate pleocytosis, normal to mildly elevated protein levels, and glucose levels within normal range in the majority of cases. These findings have led medical experts to suggest that AFM is likely due to a neuroinvasive infectious disease.

Pathogens including enteroviruses (polio virus, enteroviruses 70 and 71, echoviruses, and coxsackieviruses A and B), flaviviruses (West Nile, Japanese encephalitis, and tick-borne encephalitis), herpesviruses, and adenoviruses can infect the central nervous system (CNS) and cause AFP and fever, although illness presentation varies. Patients with AFM should be managed using basic standards of care for severe neurologic disease. At present there are no targeted interventions in the treatment of AFM. Plasmapheresis and immunosuppressive biologic modifiers should be discouraged in AFM management.

Diagnostic Testing

If AFM is suspected, specimen collection should occur as early as possible, ideally on the day of limb weakness onset. In order of priority, specimens include CSF, blood, serum, stool, and nasopharyngeal specimens. Contact PDPH to arrange for testing of specimens at public health reference laboratories.

Reporting Guidelines

Patients who have <u>acute onset of focal limb weakness AND 1</u>) an MRI showing a spinal cord lesion largely restricted to gray matter and spanning one or more spinal segments OR 2) CSF with pleocytosis with no other apparent cause should be reported to PDPH's Division of Disease Control at 215-685-6748. Providers are reminded that encephalitis or meningitis regardless of etiology, Guillain-Barrè syndrome, West Nile virus and other arboviral infections, Lyme disease, and Varicella and herpes zoster are also reportable to PDPH. For more information visit: https://hip.phila.gov/DiseaseControlGuidance/DiseaseSconditions/AFM