

enterovirus-D68, and 13 (34%) for other viruses, primarily from nonsterile sites. cerebrospinal fluid specimen results from 2 patients were positive. One cerebrospinal fluid specimen result was positive for enterovirus-A71; this patient also had a stool specimen result positive for enterovirus-A71. The second patient had a cerebrospinal fluid specimen result positive for enterovirus-D68; this patient also had enterovirus-D68 and parechovirus-A6 identified in a respiratory specimen. Two additional patients had more than one virus detected in a single respiratory specimen, including one with EV-D68 and echovirus 6 and one with rhinovirus-A24 and parechovirus-A6. All stool specimens had negative results for poliovirus. Among the 20 patients who did not meet the acute flaccid myelitis case definition and were classified as noncases, 1 (5%) had a positive cerebrospinal fluid specimen result (echovirus 25), 7 (35%) had positive respiratory specimen results (enterovirus-A71, rhinovirus-A24, rhinovirus-A56, rhinovirus-A90, and enterovirus/rhinovirus not typed), and 6 (30%) had positive stool or rectal swab specimen results (enterovirus-D68, enterovirus-A71, rhinovirus-A90, echovirus 9, echovirus 11, and echovirus 25).

Because some enteroviruses can cause acute flaccid limb weakness, and there was a temporal association with acute flaccid myelitis and a nationwide severe respiratory outbreak of enterovirus-D68 in 2014,² CDC performs enterovirus/rhinovirus testing in an effort to identify causes for acute flaccid myelitis cases. Despite a subsequent peak of acute flaccid myelitis in 2016 (<https://www.cdc.gov/acute-flaccid-myelitis/afm-surveillance.html>), CDC did not receive reports of large outbreaks of severe respiratory illness in 2016. Furthermore, there has been limited detection of pathogens in cerebrospinal fluid in these cases; virus identified in cerebrospinal fluid would be considered causal. Almost all patients with acute flaccid myelitis have reported signs and symptoms consistent with viral illness in the weeks preceding limb weakness. Clinical, laboratory, and epidemiologic evidence to date suggest a viral association. CDC and collaborators continue to investigate risk factors for acute flaccid myelitis and to study the causes and mechanisms of acute flaccid myelitis.

Parents and caregivers are urged to seek immediate medical care for a child who develops sudden weakness of the arms or legs. In the evaluation of a child with acute flaccid limb weakness, clinicians are advised to inquire about recent fever with or without antecedent respiratory or gastrointestinal symptoms and to collect timely specimens for viral testing, including cerebrospinal fluid, serum, respiratory, and stool specimens. Additional information for clinicians is available at <https://www.cdc.gov/acute-flaccid-myelitis/hcp/index.html>. Cases of patients with

acute flaccid limb weakness should be reported to state and local health departments as soon as possible regardless of laboratory or MRI findings.

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REFERENCES

1. Pastula DM, Aliabadi N, Haynes AK, et al. Acute neurologic illness of unknown etiology in children—Colorado, August–September 2014. *MMWR Morb Mortal Wkly Rep.* 2014;63:901-902.
2. Sejvar JJ, Lopez AS, Cortese MM, et al. Acute flaccid myelitis in the United States, August–December 2014: results of nationwide surveillance. *Clin Infect Dis.* 2016;63:737-745.

COMMENTARY

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The above report describes that in the fall of 2018, CDC noted an increase in the number of cases of acute flaccid myelitis, which is a rare polioliike neurologic disease that primarily affects children and young adults and is associated with rapid development of flaccid weakness in one or more limbs, variable cranial nerve involvement, and gray matter lesions involving the spinal cord.¹ Given the increased incidence of this illness and the likelihood that most patients will present to an emergency department, it is critically important for emergency physicians to recognize the characteristics and implications of this disease and the need to report all suspected cases to state and local health departments.

The first case of acute flaccid myelitis was reported in 2012 in California in a child with enterovirus-D68 detected in respiratory tract specimens. In addition to the outbreak described in the above report, similar outbreaks occurred in the fall of 2014 and 2016.^{2,3} Although enterovirus-D68 was the most common virus detected in respiratory samples from patients with acute flaccid myelitis, the cause has not been confirmed in most patients and it remains unclear whether this illness is related to this enterovirus, another virus, or an immunologic response to infection.⁴ Although the cause of acute flaccid myelitis is still unclear, the article highlights that clinical, laboratory, and epidemiologic analyses suggest a likely viral association.

In a recent study by Elrick et al⁴ of North American children receiving a diagnosis of acute flaccid myelitis, it is described as “a poorly understood syndrome” in which an accurate diagnosis can be difficult to make, given the absence of a “biomarker criterion standard for diagnosis.”

The authors argue that the CDC definition is appropriately sensitive for epidemiologic assessment but also results in the inclusion of other neurologic diseases that cause acute weakness. They suggest a more restrictive definition of acute flaccid myelitis to establish “a more homogenous population that likely reflects a shared pathophysiology” and describe a 4-item checklist for the restricted type: (1) prodromal fever or viral syndrome; (2) certain pattern of weakness or abnormal weakness; (3) supporting evidence from MRI scans, electromyography or nerve conduction studies, or cerebrospinal fluid analysis; and (4) absence of objective sensory deficits, MRI lesions greater than 1 cm in the cortex and supratentorial white matter, encephalopathy not attributable to fever or certain other conditions, and cerebrospinal fluid protein elevations twice the upper limit of normal without cerebrospinal fluid pleocytosis. In the study, alternative diagnoses were found for approximately 25% of patients who met the CDC acute flaccid myelitis case definition. These diagnoses included transverse myelitis, spinal cord ischemia, Guillain-Barré syndrome, Chiari I myelopathy, meningitis, and other demyelinating syndromes.⁴

Any child or young adult who develops symptoms concerning for acute flaccid myelitis should seek immediate medical attention. A possible diagnosis of acute flaccid myelitis should be considered for any patient who presents with sudden-onset limb weakness in one or more extremities. Acute flaccid paralysis may be accompanied by bulbar findings, such as ophthalmoplegia, dysarthria, dysphagia, and facial paralysis. Most patients will report an antecedent viral syndrome, including fever, respiratory symptoms, and gastrointestinal complaints.⁵ Patients meeting clinical criteria for acute flaccid myelitis should be evaluated for a range of infectious and noninfectious causes. Specimens should be collected in a timely manner, including for cerebrospinal fluid, serum, respiratory, and stool analysis. The local health department may be able to provide guidance on which specimens to obtain and help with coordinating specialized testing. MRI should be considered to evaluate for gray matter disease of the spinal cord. Radiographic or electrophysiologic evidence of anterior horn cell involvement in the setting of flaccid paralysis is the most specific finding in acute flaccid myelitis.⁶

Treatment is mainly supportive because no interventions have been shown to be effective in the management of

acute flaccid myelitis, including antivirals, glucocorticoids, intravenous immunoglobulin, plasma exchange, or other immunomodulatory agents.^{3,7,8} Hospital admission or observation may be indicated to expedite testing, including MRI and cerebrospinal fluid analysis, and to observe for progression of symptoms. Approximately one third of patients will develop acute respiratory failure requiring intubation and mechanical ventilation because of respiratory muscle or bulbar muscle weakness, with some requiring ventilatory support for a prolonged period.⁶ Prognosis and neurologic recovery in acute flaccid myelitis are variable and often incomplete, with many children having persistent motor deficits and muscle atrophy long after onset of disease. All suspected cases of acute flaccid myelitis should be reported to state and local health departments, using the patient summary form available online at <http://www.cdc.gov/acute-flaccid-myelitis/hcp/data.html>.⁹

REFERENCES

1. Vogel G. Outbreaks of polioliike disease pose a puzzle. *Science*. 2018;362:277.
2. McKay SL, Lee AD, Lopez AS, et al. Increase in acute flaccid myelitis—United States, 2018. *MMWR Morb Mortal Wkly Rep*. 2018;67:1273-1275.
3. Van Haren K, Ayscue P, Waubant E, et al. Acute flaccid myelitis of unknown etiology in California, 2012-2015. *JAMA*. 2015;314:2663-2671.
4. Elrick MJ, Gordon-Lipkin E, Crawford TO, et al. Clinical subpopulations in a sample of North American children diagnosed with acute flaccid myelitis, 2012-2016. *JAMA Pediatr*. 2018; <https://doi.org/10.1001/jamapediatrics.2018.4890>.
5. Centers for Disease Control and Prevention. Acute flaccid myelitis: AFM investigation. Available at: <https://www.cdc.gov/acute-flaccid-myelitis/afm-surveillance.html>. Accessed December 24, 2018.
6. Messacar K, Schreiner TL, Van Haren K, et al. Acute flaccid myelitis: a clinical review of US cases 2012-2015. *Ann Neurol*. 2016;80:326-338.
7. Centers for Disease Control and Prevention. Acute flaccid myelitis: interim considerations for clinical management. Available at: <https://www.cdc.gov/acute-flaccid-myelitis/hcp/clinical-management.html>. Accessed December 24, 2018.
8. Messacar K, Schreiner TL, Maloney JA, et al. A cluster of acute flaccid paralysis and cranial nerve dysfunction temporally associated with an outbreak of enterovirus D68 in children in Colorado, USA. *Lancet*. 2015;385:1662-1671.
9. Council of State and Territorial Epidemiologists. Standardized case definition for acute flaccid myelitis. Available at: <http://c.ymcdn.com/sites/www.cste.org/resource/resmgr/2015PS/2015PSFinal/15-ID-01.pdf>. Accessed December 24, 2018.

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