

Five things to know about...acute flaccid myelitis

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1) Acute flaccid myelitis is a clinical syndrome characterized by rapid onset of muscle weakness.

In 2018, a number of children with acute flaccid myelitis (AFM) were seen in children's hospitals across North America, with 80 suspected cases in the US and 44 in Canada.^{1,2} AFM results from injury to the anterior horn cells of the spinal cord, and presents clinically with an abrupt onset of limb weakness, pain, paresthesias and areflexia.³ About one-quarter of patients develop cranial nerve and/or bulbar signs.⁴ Clinically, it may resemble Guillain-Barre Syndrome (GBS), but can be distinguished from GBS by its typical MRI appearance (see below) and its motor distribution (asymmetric weakness in AFM versus symmetric and ascending in GBS).³ AFM typically affects children 4-15 years of age, and is usually preceded by a non-specific viral prodromal illness.³⁻⁵ Box 1 outlines the US CDC's case definition of AFM.⁶

2) Enteroviruses are the predominant cause of acute flaccid myelitis in children.

Clusters of AFM were observed in North America in 2014, 2016 and 2018 during the late summer-fall months, coincident with peak enterovirus (EV) season.^{2,4} With the near eradication of polioviruses, EV-D68 and EV-A71 have emerged as the predominant viruses associated with AFM.⁴ EV-D68 was likely responsible for the 2014 and 2016 clusters of AFM in North America⁷ but thus far, a clear culprit has not been implicated for the 2018 cluster.

3) Infectious workup and neuroimaging should be performed urgently.

Infectious workup should include microbiologic testing of cerebrospinal fluid (CSF), respiratory and stool samples, and selectively blood, for EVs. CSF reveals a lymphocytic pleocytosis and normal to mildly elevated protein.⁴ Imaging should include MRI of the brain and spine with contrast. MRI typically shows T2 hyperintensities in the grey matter of the spinal cord and often enhancement of the nerve roots. In GBS, nerve root enhancement is seen but spinal cord grey matter is not involved. Electromyography shows acute motor axonal neuropathy. Cases of suspected AFM should be reported to PHAC.

4) Acute flaccid myelitis can progress rapidly and should be managed as a medical emergency.

Urgent referral to a tertiary care center is warranted as 24-52% of patients require admission to the intensive care unit for ventilatory and/or nutritional support.^{5,8} While their effectiveness is unknown, treatments that may be considered on a case-by-case basis and guided by pediatric neurology and infectious disease specialists, include immunomodulatory therapy, such as high dose intravenous corticosteroids, intravenous immune globulin (IVIG), and/or plasmapheresis.⁴ A vaccine has been developed for EV-A71 but is not licensed in Canada.⁴

5) Acute flaccid myelitis often results in residual impairment.

Canadian and US studies have described persistent deficits in most children after 4-6 months' follow-up, with full recovery in only 8-18%, and 8-14% either requiring assistive devices for ambulation or complete dependence on caregivers.^{5,8}

Box 1. CDC case definition of AFM (no age limitation)⁶

- **Confirmed AFM:** acute focal limb weakness and MRI findings of mainly gray matter lesions involving one or more spinal cord segments.
- **Probable AFM:** acute focal limb weakness and cerebrospinal fluid pleocytosis (>5 cells/mm³)

CDC, Centers for Disease Control and Prevention

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