

## Feature

## Acute flaccid myelitis in the USA

This autumn, the USA has seen another spike in cases of acute flaccid myelitis (AFM), a rare and poorly understood condition that causes polio-like paralysis, mostly in children and adolescents, and rarely in adults. The condition is typified by damage to peripheral nerves in the spinal column, resulting a dense, floppy paralysis and a loss of muscle tone in the limbs, head, and neck. The severity of the paralysis varies from case to case, with some patients exhibiting minor weaknesses in their limbs, while others have difficulty breathing, in some cases leading to respiratory failure.

There have been more than 450 confirmed cases of AFM in the USA since 2014, with the vast majority occurring between August and October on even-numbered years. There have been 165 cases in 36 states so far in 2018. Prognosis also varies greatly, with some children recovering quickly with seemingly few long-term consequences, while others are affected more deeply with little evident recovery.

In 90% of cases, patients have a mild respiratory infection prior to the development of AFM symptoms. Several viruses—including enterovirus A71, the West Nile virus, and multiple strains of the coxsackievirus—have been detected in patients with AFM, but to date no patient with AFM has tested positive for the poliovirus. However, in most cases, the cerebrospinal fluid has not shown a result for viral infection. According to Manisha Patel, a paediatrician and the AFM Epidemiology team lead at the US Centers for Disease Control and Prevention (CDC), there could be multiple reasons for that. "Is it direct infection by the virus of the motor neuron", said Dr Patel, "or indirect infection where it's more this immune-mediated pathogenesis?" Similar to multiple sclerosis, immune-mediated

pathogenesis causes the body's immune system to attack the central nervous system in response to the virus' presence. Another possibility is an autoimmune response (as in Crohn's Disease or inflammatory bowel disease) where the body's own antibodies identify the anterior horn cells—part of the spinal column that contains motor neurons—as threats and begins attacking them.

For patients with AFM, rehabilitation is the main path towards recovery. However, while with most spinal cord injuries patients maintain a certain density of muscle tone in their limbs, that muscle tone is often lost in patients with AFM, according to Rebecca Martin (Manager of Clinical Education and Training at the International Center for Spinal Cord Injury at the Kennedy Krieger Institute, Baltimore, MD, USA). "The kids...can't hold their head up, they can't hold their trunk up, they can't move their arms or legs, and they're very floppy", said Martin. She manages rehabilitation treatments for children with AFM, and while the underlying causes of AFM are still not understood, she works with children to treat the symptoms that they can observe. Such treatments include electric stimulation of muscles to help maintain and rebuild muscle tone, walking exercises, and other developmentally appropriate strengthening and motor control exercises. Patients participate daily in 3 hours of physical therapy, 2 hours of occupational therapy, and therapeutic recreation services. "Some of those may overlap", Martin said, "because if you do the math there, [there is] not a lot of time, especially if you have a little kid who still needs a nap."

According to Martin, children with AFM can make progress, but so far it has proven to be slow and incremental, and the condition has not been recognised long enough to know whether that progress is towards a partial or complete

recovery, or how long it might take. But Martin says that for kids she worked with who were diagnosed with AFM back in 2014, "every year when they come back they look a little bit better and a little bit different".

For Patel, the main message she wants to get across to clinicians who might see children with potential symptoms of AFM is "recognise early, collect specimens early, and let the health department know early". While the understanding of the causes and effects of AFM is still in flux, according to Patel the CDC needs full access to all available evidence—including cerebrospinal fluid, respiratory, blood, and stool samples early on in the disease course, and a thorough description of each patient—to understand the full spectrum of this condition. Due to its rarity, the vast majority of children who come through a paediatrician's office are not going to develop AFM. But for those who do, collecting early samples and sending them to the CDC could be a vital link in unraveling the mystery of the condition.

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A patient undergoing rehabilitation treatment at the International Center for Spinal Cord Injury, Kennedy Krieger Institute (Baltimore, MD, USA)



*Lancet Child Adolesc Health* 2018

Published Online  
December 20, 2018  
[http://dx.doi.org/10.1016/S2352-4642\(18\)30406-1](http://dx.doi.org/10.1016/S2352-4642(18)30406-1)

For up-to-date information on the CDC's investigation into AFM see <https://www.cdc.gov/acute-flaccid-myelitis/afm-surveillance.html>