

Researcher discusses what parents should know about acute flaccid myelitis

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Recently, cases of a polioliike illness have been back in the news. Acute flaccid myelitis, a rare complication from certain viral infections, causes

paralysis in one or more limbs and strikes mostly children. Keith Van Haren, MD, assistant professor of neurology and neurological sciences at the School of Medicine, has studied the condition and written scientific review articles covering clusters of cases dating back to 2012.

Van Haren talked with science writer Erin Digitale about what parents should know about the disease.

Q: What do we know about the history and causes of this condition?

Van Haren: There are cases of infectious paralysis stretching far back into recorded human history, including, of course, of poliomyelitis. The best interpretation of current and historical evidence suggests these cases are primarily caused by [viral infections](#), and there happen to be several viruses that can do this. Broadly speaking, we could classify poliomyelitis as a form of [acute flaccid myelitis](#); they appear to share similar elements of pathophysiology.

Enteroviruses—including the three human polioviruses, and enterovirus 71—are the most common culprits. Since at least 2012, there is accumulating evidence that enterovirus 68 can also cause this syndrome. And West Nile Virus can cause this acute flaccid myelitis, although it's from a different family of viruses.

As best we can tell, the modern outbreaks go back to at least 2012, when Carol Glaser and her team at the California Department of Public Health began noticing an uptick in polioli-like cases, mostly in kids. It was Carol who first noticed the viral association with enterovirus 68.

The phrase "acute flaccid myelitis" was coined in 2014 by a group of colleagues, including myself, who were trying to come up with an

appropriate descriptive term that would disentangle it from the historical association with poliomyelitis and provide a broader framework for characterizing the [illness](#).

The pattern that we're currently seeing is an every-other-year phenomenon. Different years bring different enteroviruses, just as different years bring different strains of flu. In the years enterovirus 68 has been circulating—in 2012, 2014, 2016 and 2018—we've also seen an increase in cases all clustered in late summer and early fall, which is the time of year that many species of enteroviruses circulate in North America.

Q: What happens in children affected with acute flaccid myelitis?

Van Haren: The syndrome typically begins with what looks like a traditional systemic illness. This is broadly true of many infectious neurological illnesses: What begins as an otherwise ordinary infection takes a different course in a particular patient, and it's not understood why.

Enterovirus infections typically start and end as benign illnesses, with congestion, fever and a sense of malaise all lasting a few days. In a very small number of individuals, this illness is followed by something more ominous. The earliest symptom among patients who actually develop acute flaccid myelitis is a period of significant pain in the limb, or multiple limbs, often described as aching, tingling or electric shocks. Within the next day or so, the limb becomes weak and the weakness can progress very quickly, over the course of an hour or two even, to very weak or complete loss of function. Muscles of the face can also be affected.

The weakness may worsen for the next day or for several days before it reaches its low point and stabilizes. In many cases, the weakened limb does gradually recover, though it may not make a full recovery. Most recovery occurs in the first few months, but recovery may continue for years. We have seen apparent improvement continuing, albeit slowly, even as far out as two years after the injury.

Rehabilitation is sometimes possible once the illness is stabilized. Most rehabilitation efforts are taken on a case-by-case basis, and often include strength training and electrical stimulation devices that deliver tiny electrical pulses, applied directly over the muscle. There are also surgical approaches, in which a nerve that is not working is swapped for a nerve that is working to re-attain some muscle movement, typically in an arm, but it is not appropriate for everyone and is attempted only in very highly specialized centers. It requires a highly skilled team to identify who might benefit and plan the procedure.

Q: How worried should parents be? Is there anything they can or should do to protect their children?

Van Haren: With any infectious illness, the youngest and oldest members of population are most vulnerable. This condition is a bit of an exception, as it is primarily affecting younger children.

To date, the best we can offer is a preventive approach: Try to help keep children healthy and clean with regular hand-washing and limited exposure to very sick people.

If a child is sick, parents should encourage him or her to rest and provide normal, appropriate nourishment and hydration. If the child or parent is noticing acute weakness or significant pain in one limb, they should seek medical care promptly.

It's important to remember how rare this disease is. To put it in context, last year there were about 80,000 deaths across the country from the flu; so far this year, there are around 100 or so total cases of acute flaccid myelitis under investigation. Clinicians and scientists are working hard to understand how to make sure it doesn't become more and, ideally, to eliminate it altogether. Analogous eradication efforts have occurred many times, primarily through vaccination.

Q: What are experts doing to better understand the illness?

Van Haren: The physician community, including child neurologists and infectious disease specialists, is coming together to form working groups to tackle the problem directly. Our general sense now is that this is a serious illness for anyone who is affected, but it remains rare.

Our goal is to understand what's happening well enough to prevent it from becoming more common, and also to develop better modes to treat it. The physician community is seeing a convergence of evidence that suggests enterovirus 68 is responsible for many but not all cases. This community would like more support from [public health](#) agencies and funders to try to understand this disease.

Q: What do we know now about the illness that we didn't know last time there were a significant number of cases?

Van Haren: There has been some really helpful progress in past couple of years in terms of modeling around [enterovirus](#) 68, focusing on the biology of the virus. Scientists have studied the genetic alterations that may have made the virus more prone to attack the human nervous system, and have tested the ability of the virus to do this in a mouse

model. This is a crucial foundation for developing treatments and vaccinations.

It's somewhat disappointing that we don't yet have a good therapeutic or really specific preventive approach. Those are the areas we ought to be making ardent strides toward.

Provided by Stanford University Medical Center

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