TO: Neurologists, Pediatricians, Radiologists and Infectious Disease Physicians

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RE: Increase in Guillain-Barre Syndrome and Acute Flaccid Myelitis Cases

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Guillain-Barre Syndrome (GBS)

In October 2015, Nebraska Department of Health and Human Services (NDHHS) was notified of an apparent increase in Guillain-Barre Syndrome (GBS) in Lancaster County. Guillain-Barre is a potentially life-threatening, paralytic illness with an annual incidence of 1-2 per 100,000. The disease results in a loss of myelin on peripheral nerves and is believed to be autoimmune in nature. Past studies have implicated *Campylobacter* gastroenteritis, the influenza vaccine and Zika virus, but in most cases the etiology is unknown. GBS patients frequently require intensive care and ventilator support, with a 5-15% mortality rate, and approximately 20% experience continued disability after one year.

As a result of this concern, NDHHS further investigated cases of Guillain-Barre in the state. Analysis of Nebraska's hospital discharge data from 2007-2013 revealed an average of 39 GBS cases per year, or approximately 2.0 per 100,000. A follow-up chart review by public health medical staff identified 33, 41, and 58 cases in 2013, 2014 and 2015, respectively, for rates of 1.8 per 100,000, 2.3 per 100,000 and 3.2 per 100,000, respectively. These values reflect an overall increase of 75% from 2013 (N=33) to 2015 (N=58). While we have documented an increase in GBS incidence over the past three years, the chart review did not identify an explanation for the increase. We will continue to track this condition in 2016 and beyond to confirm the trend and assess whether this trend is sustained or, rather, a short term anomaly. Clinicians who suspect the diagnosis of GBS should perform a detailed neurologic exam, which should most often include a lumbar puncture for CSF evaluation. This condition was added to our reportable disease list; physicians who diagnose patients with GBS are asked to report the case to a state or local public office for public health chart review.

Acute Flaccid Myelitis (AFM)

The CDC is investigating an increase in AFM in 2016. AFM is a rare neurological illness, most commonly found in children. It is characterized by polio-like symptoms (paralysis, facial droop, slurred speech and difficulty moving the eyes). As of September 2016, 89 people in 33 states were confirmed to have AFM, as compared to 21 cases in 16 states in 2015. Seattle Children's Hospital, for example, has seen eight patients with the neurologic condition since mid-September. Additionally, three recent suspect cases are being investigated in the Washington area. To date, no single pathogen has been consistently detected in CSF, but an array of viral pathogens may be responsible, including poliovirus, non-polio enteroviruses, flaviviruses (i.e. West Nile virus, St. Louis encephalitis, Japanese encephalitis, and possibly Zika), herpes (i.e. CMV, Epstein-Barr virus), and certain strains of adenovirus. All of these should be considered and investigated in the setting of unexplained limb weakness.

Number of confirmed U.S. AFM cases reported to CDC by month of onset, August 2014 - September 2016^** ■ 2014 ■ 2015 ■ 2016 60 55 Number of confirmed AFM cases 50 45 40 35 30 25 20 15 10 5 April February March May lune July January August September October November December

Criteria for Reporting AFM to Public Health

Clinicians should maintain vigilance for AFM among all age groups and report cases of AFM to the Epidemiology Unit at NDHHS (402) 471-2937. Clinicians are also advised to collect specimens from suspected cases as early as possible, ideally on the day of symptom onset. Consult with the Nebraska Public Health Laboratory for proper testing procedures of CSF, blood, stool and nasopharyngeal aspirates.

Month

Report any illness to public health authorities that meets the following criteria:

A person with onset of acute focal limb weakness AND one or both of the following:

- A magnetic resonance image showing a spinal cord lesion largely restricted to gray matter, and spanning one or more spinal segments
- Cerebrospinal fluid (CSF) with pleocytosis (CSF white blood cell count >5 cells/mm3, may adjust for presence of red blood cells by subtracting 1 white blood cell for every 500 red blood cells present); CSF protein may or may not be elevated

At this point, no interventions or treatments have proven effective for AFM. This includes corticosteroids, IVIG, plasmapheresis, interferon and antivirals. Routine clinical management for severe neurologic disease, along with physical and occupational therapy, is the basic standard of care.

Additional Resources

AFM Case Investigation Form:

http://www.cdc.gov/acute-flaccid-myelitis/downloads/patient-summary-form.pdf

CDC Acute Flaccid Myelitis

http://www.cdc.gov/acute-flaccid-myelitis/index.html

CDC AFM Specimen Collection Information

http://www.cdc.gov/acute-flaccid-myelitis/hcp/instructions.html

Nebraska Public Health Laboratory http://nphl.org/