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Acute Neurologic Illness with Focal Limb Weakness of Unknown Etiology in Children

Summary:

The Centers for Disease Control and Prevention (CDC) is working closely with the Colorado Department of Public Health and Environment (CDPHE) and Children's Hospital Colorado to investigate a cluster of nine pediatric patients hospitalized with acute neurologic illness of undetermined etiology. The illness is characterized by focal limb weakness and abnormalities of the spinal cord gray matter on MRI. These illnesses have occurred since August 1, 2014 coincident with an increase of respiratory illnesses among children in Colorado. The purpose of this HAN Advisory is to provide awareness of this neurologic syndrome under investigation with the aim of determining if children with similar clinical and radiographic findings are being cared for in other geographic areas. Guidance about reporting cases to state and local health departments and CDC is provided. Please disseminate this information to infectious disease specialists, intensive care physicians, pediatricians, neurologists, radiologists/neuroradiologists, infection preventionists, and primary care providers, as well as to emergency departments and microbiology laboratories.

Background

The CDPHE, Children's Hospital Colorado, and CDC are investigating nine cases of acute neurologic illness among pediatric patients. The cases were identified during August 9–September 17, 2014 among children aged 1–18 years (median age 10 years). Most of the children were from the Denver metropolitan area. All were hospitalized. Common features included acute focal limb weakness and specific findings on magnetic resonance imaging (MRI) of the spinal cord consisting of non-enhancing lesions largely restricted to the gray matter. In most cases, these lesions spanned more than one level of the spinal cord. Some also had acute cranial nerve dysfunction with correlating non-enhancing

brainstem lesions on MRI. None of the children experienced altered mental status or seizures. None had any cortical, subcortical, basal ganglia, or thalamic lesions on MRI. Most children reported a febrile respiratory illness in the two weeks preceding development of neurologic symptoms. In most cases, cerebrospinal fluid (CSF) analyses demonstrated mild-moderate pleocytosis (increased cell count in the CSF) consistent with an inflammatory or infectious process. CSF testing to date has been negative for enteroviruses, including poliovirus and West Nile virus. Nasopharyngeal specimens were positive for rhinovirus/enterovirus in six out of eight patients that were tested. Of the six positive specimens, four were typed as EV-D68, and the other two are pending typing results. Testing of other specimens is still in process. Eight out of nine children have been confirmed to be up to date on polio vaccinations. Epidemiologic and laboratory investigations of these cases are ongoing.

The United States is currently experiencing a nationwide outbreak of EV-D68 associated with severe respiratory disease. The possible linkage of this cluster of neurologic disease to this large EV-D68 outbreak is part of the current investigation. CDC is seeking information about other similar neurologic illnesses in all states, especially cases clustered in time and place. CDC has particular interest in characterizing the epidemiology and etiology of such cases.

Recommendations

- ◆ Patients who meet the following case definition should be reported to state and local health departments:
Patients ≤21 years of age with
 - 1) Acute onset of focal limb weakness occurring on or after August 1, 2014; AND
 - 2) An MRI showing a spinal cord lesion largely restricted to gray matter.

- ◆ State and local health departments should report patients meeting the case definition to CDC using a brief patient summary form (www.cdc.gov/non-polio-enterovirus/investigation/). State health departments should send completed summary forms to CDC by email at limbweakness@cdc.gov. *(This will be completed by Washoe County Health District.)*
- ◆ Providers treating patients meeting the above case definition should consult with their local and state health department for laboratory testing of stool, respiratory, and cerebrospinal fluid specimens for enteroviruses, West Nile virus, and other known infectious etiologies.
- ◆ Health departments may contact CDC for further laboratory and epidemiologic support by phone through the CDC Emergency Operations Center (770-488-7100), or by email at limbweakness@cdc.gov. Confirmation of the presence of EV-D68 currently requires typing by molecular sequencing.

For more information:

Please visit the CDC enterovirus website (<http://www.cdc.gov/non-polio-enterovirus/>) for general information about enterovirus infections, including EVD-68, and for up-to-date guidance about infection control measures. For information about poliovirus, please visit the CDC poliovirus website (<http://www.cdc.gov/vaccines/vpd-vac/polio/in-short-both.htm>). For information about West Nile Virus, please visit the CDC West Nile Virus website (<http://www.cdc.gov/westnile/>). State and local health departments with questions should contact the CDC Emergency Operations Center (770-488-7100).

Categories of Health Alert Network messages:

- Health Alert** Requires immediate action or attention; highest level of importance
- Health Advisory** May not require immediate action; provides important information for a specific incident or situation
- Health Update** Unlikely to require immediate action; provides updated information regarding an incident or situation
- HAN Info Service** Does not require immediate action; provides general public health information

Reporting and Testing of Extraordinary Illness

Nevada law ([NAC 441A.525](#)) requires extraordinary occurrence of illness be reported to the local health authority. Recent emerging diseases such as Ebola Virus Disease (EVD), Enterovirus-D68 (EV-D68), and acute neurologic illness with focal limb weakness of unknown etiology in children fall into this category. It is mandatory for healthcare providers to report such diseases to Washoe County Health District (WCHD) at 775-328-2447 (Phone) and 775-328-3764 (Fax) once suspected. The following table describes these emerging infectious diseases, reporting criteria, and reporting timeline. WCHD will coordinate with the Nevada State Public Health Laboratory and healthcare facilities for necessary laboratory testing.

Disease Name	Criteria for Reporting	When to report	Preferred Specimen Types
Ebola Virus Disease	<ul style="list-style-type: none"> ◆ Clinical criteria, which includes fever of greater than 38.6 degrees Celsius or 101.5 degrees Fahrenheit, and additional symptoms such as severe headache, muscle pain, vomiting, diarrhea, abdominal pain, or unexplained hemorrhage; AND ◆ epidemiologic risk factors within the 21 days prior to onset of symptoms, such as contact with blood or other body fluids or human remains of a patient known to have or suspected to have EVD; residence in—or travel to—an area where EVD transmission is active (Countries include Guinea, Liberia, Nigeria, Sierra Leone, and Senegal as of September 29, 2014); or direct handling of bats or non-human primates from disease-endemic areas. 	Immediate	Blood (purple top tube)
EV-D68	In children under the age of 18 years with severe respiratory illness, especially with wheezing, and who are admitted to a PICU; AND positive for Rhinovirus/Enterovirus or Enterovirus screening assay	Within 24 hours	Respiratory-NP/OP swab, NP/OP wash or aspirate, stool, rectal swab, cerebrospinal fluid (CSF), serum, etc.
Acute neurologic illness with focal limb weakness of unknown etiology in children	Patients ≤21 years of age with 1) Acute onset of focal limb weakness occurring on or after August 1, 2014; AND 2) An MRI showing a spinal cord lesion largely restricted to gray matter.	Within 24 hours	Stool, respiratory specimens, and CSF