

Acute Flaccid Myelitis (AFM)/Poliomyelitis

Signs and Symptoms	AFM is characterized by rapid onset of weakness in one or more limbs with distinct abnormalities of the spinal cord gray matter on magnetic resonance imaging. Facial paralysis, oculomotor dysfunction, palpebral ptosis, and dysphagia or dysarthria can be also present. Poliovirus is a cause of AFM. Other viruses that have been associated with AFM include Herpesvirus, Flavivirus, and Adenovirus. Most poliovirus infections are asymptomatic. Up to 25% of infections can cause abortive poliomyelitis (nonspecific mild illnesses including fever, headache, sore throat, or gastrointestinal symptoms, with symptoms disappearing within 2-3 days and no neurological symptoms). Poliovirus infecting the spinal cord or brain stem results in either aseptic meningitis or acute, asymmetric, ascending flaccid paralysis. Paralysis occurs in 1/200 poliovirus infections on average (<1%). It is more common in adults: 1 in 75 adults vs. 1 in 1000 children. In paralytic polio, neurologic symptoms progress within a few days, achieve a plateau for weeks then can resolve partially or fully. Legs are more often affected than arms.
Incubation	AFM: For most common AFM etiologies incubation periods see section 2.F. Polio: 6-20 days (range 3 to 36 days) From contact to onset of paralysis: 11 to 17 days (range 8-36 days)
Case classification	<p>AFM Clinical definition: Onset of acute focal limb weakness AND a magnetic resonance image (MRI) showing spinal cord lesion largely restricted to gray matter and spanning one or more spinal segments, OR Cerebrospinal fluid (CSF) with pleocytosis (white blood cell count >5 cells/mm³) See Section 3.A.</p> <p>Probable case: meets clinical definition, AND CSF findings criteria</p> <p>Confirmed case: meets clinical definition AND MRI findings criteria</p> <p>Polio Probable case: Acute onset of flaccid paralysis of one or more limbs with decreased or absent tendon reflexes, without other apparent cause, and without sensory or cognitive loss.</p> <p>Polio Confirmed case: Meets probable case criteria AND in which the patient has a neurologic deficit 60 days after onset of initial symptoms; or has died; or has unknown follow-up status.</p>
Differential diagnosis	Enteroviruses (polio and non-polio), West Nile Virus, Japanese encephalitis virus, Saint Louis encephalitis, Cytomegalovirus, Epstein-Barr virus, and Adenovirus infections.
Treatment	Supportive, as there is no specific treatment for AFM or Poliomyelitis.
Laboratory	PHL does not perform testing for poliovirus/enteroviruses, but specimens should be sent to PHL to be forwarded to CDC for testing. The following specimens should be collected as early as possible in the course of illness: two stool specimens separated by at least 24 hours, CSF, serum, and a nasopharyngeal (NP). An oropharyngeal swab must be collected in addition to the NP specimen when polio is suspected. In addition, viral respiratory and viral stool cultures should be ordered by the healthcare facility where the patient is hospitalized as soon as possible and results shared with the local health jurisdiction.
Public Health investigation	<ul style="list-style-type: none"> Assess the likelihood of AFM/Polio: confirm compatible clinical symptoms, verify vaccination and travel history, assess exposure risk (e.g. contact with a recent oral polio vaccine recipient), obtain history of recent respiratory or GI illness, and review test findings (i.e. CSF and MRI results). If laboratory testing is indicated, facilitate timely collection/transport of appropriate specimens. If Poliovirus is confirmed as the cause for AFM, DOH and CDC will assist with an extensive contact investigation. See Appendix A. If poliovirus is suspected isolation for 10 days from the onset of illness is indicated and enteric precautions should be followed for six weeks. No specific management is indicated for contacts of AFM cases but they should be educated regarding any specific etiology suspected and advised about when to seek medical care. For a suspected polio case, contacts must be identified and monitored for symptoms. Collection of stool and serum samples from household members and other contacts associated with possible transmission settings may be required. For a confirmed polio case, vaccination should be offered to susceptible contacts with an emphasis on persons who have an ongoing risk of exposure.

Acute Flaccid Myelitis (AFM), Poliovirus infection, and Poliomyelitis reporting overview

OBJECTIVES:

- a) To assure adequately sensitive Poliomyelitis surveillance in Washington State
- b) To describe the clinical characteristics, epidemiology with baseline incidence for all age groups, differential diagnosis, and potential causes of Acute Flaccid Myelitis (AFM), including poliovirus, non-polio enteroviruses, and other viruses.
- c) To integrate AFM and Poliovirus infection and Poliomyelitis surveillance activities to increase the sensitivity and specificity of the epidemiologic surveillance for these conditions.

BACKGROUND

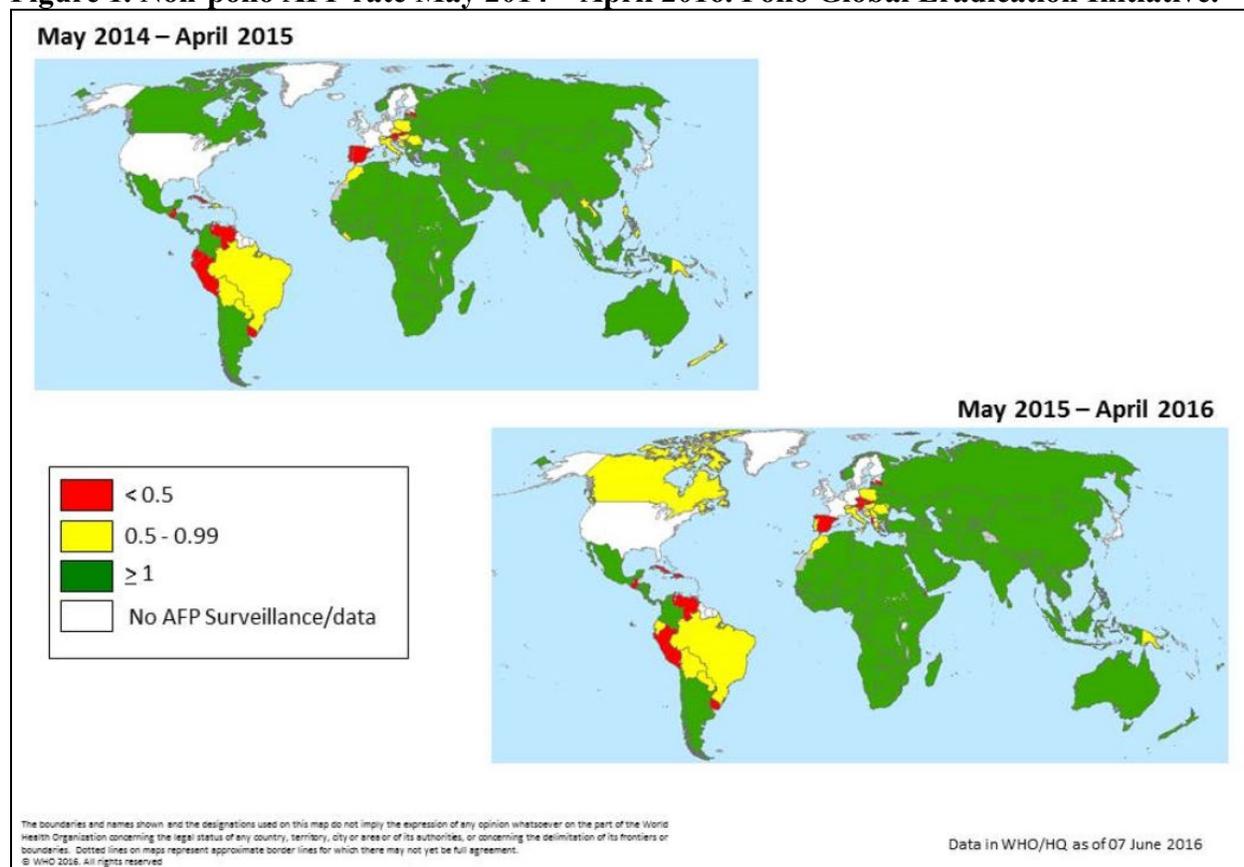
To understand the rationale for a combined approach to reporting and surveillance activities for poliomyelitis and poliovirus infection, AFM, and non-polio enterovirus, it is important to consider the shared concepts that underlie these conditions as well as the ongoing global surveillance activities including polio eradication efforts.

Acute flaccid paralysis (AFP) is a clinical syndrome characterized by rapid onset of weakness, progressing to maximum severity within several days to weeks. The term flaccid indicates the absence of spasticity or other signs of disordered central nervous system motor tracts such as hyperreflexia, clonus, or extensor plantar responses. Muscle disorders, metabolic disorders, Guillain-Barre Syndrome, neuropathies, acute transverse myelitis, and acute traumatic myelitis, are included in the AFP definition.

Although AFP encompasses all cases of paralytic poliomyelitis, and is of great public health importance because of its use in surveillance for poliomyelitis in the context of the global polio eradication initiative, the current WHO case definition which is highly sensitive in detecting AFP tends to decrease specificity in detecting paralytic poliomyelitis.

AFP surveillance is the WHO gold standard for polio surveillance. As an indicator of surveillance sensitivity, at least one case of non-polio AFP should be detected annually per 100 000 population aged less than 15 years (See figure 1). However, AFP surveillance has not been universally adopted. Some industrialized polio-free countries (e.g. Denmark, Canada) with high polio vaccine coverage have chosen to use alternative enterovirus surveillance systems instead.

Figure 1. Non-polio AFP rate May 2014 – April 2016. Polio Global Eradication Initiative.



Originally available at: <http://www.polioeradication.org/>

Because in the United States AFP is not a reportable condition, we do not conduct AFP surveillance in Washington State. Please note that several conditions that can cause AFP are reportable in most states, including Washington. Examples include botulism, rabies, Japanese encephalitis, Lyme borreliosis, and trichinosis.

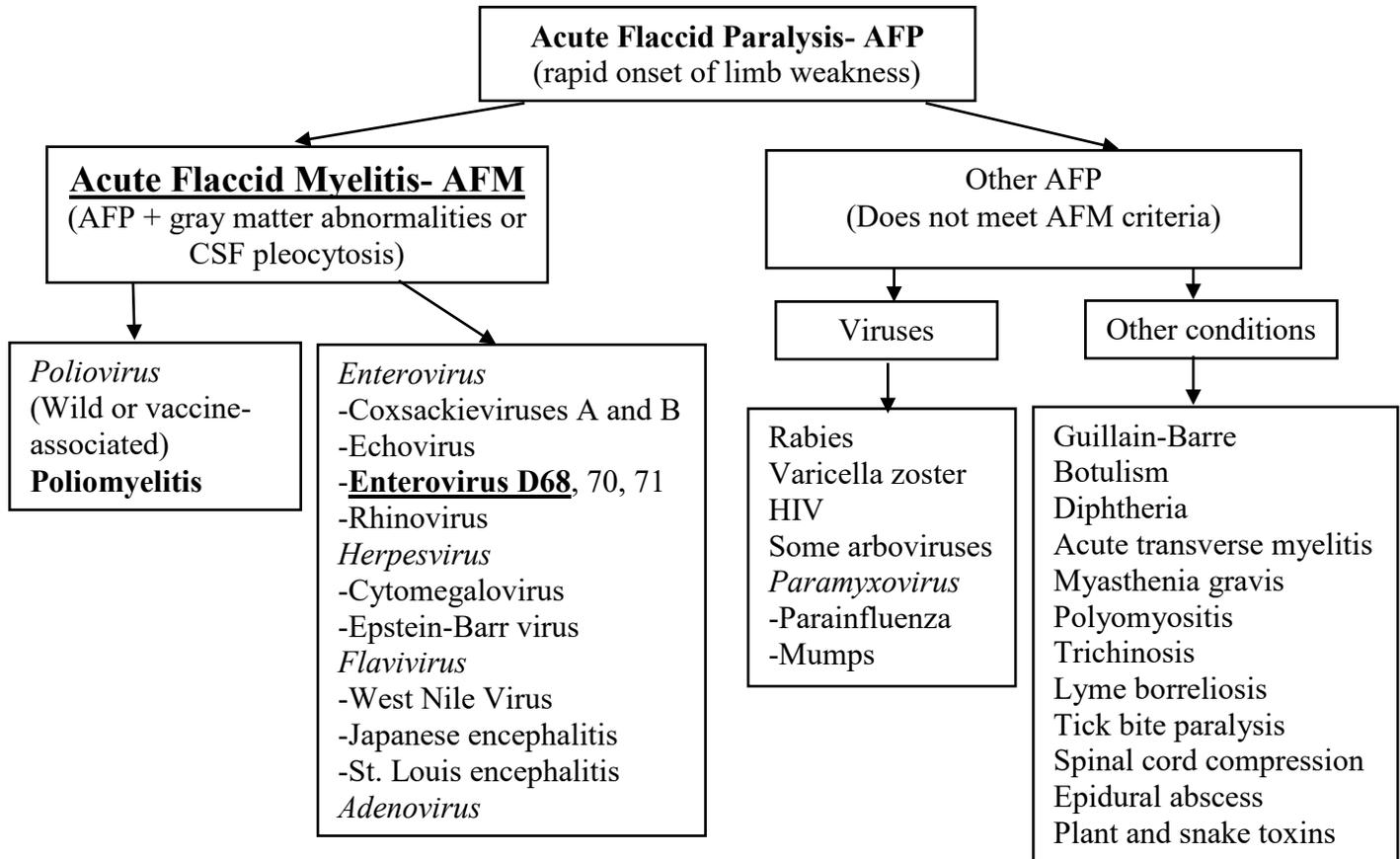
Acute Flaccid Myelitis (AFM) is a subset of AFP that is characterized by rapid onset of weakness in one or more limbs (as occurs in AFP) and distinct abnormalities of the spinal cord gray matter on magnetic resonance imaging (more specific than AFP). An apparent increase in reports of AFM in children in the United States was identified in the summer and fall of 2014, but without a baseline incidence of AFM the interpretation of this increase has been challenging.

This apparent increased in AFM incidence coincided with a national outbreak of severe respiratory illness caused by enterovirus-D68 (EV-D68) among children. Despite the close time association between the increase in AFM cases and the EV-D68 outbreak, a common etiology for the 2014 AFM cases has not been determined. Figure 2 contains a summary of the most common etiologies of AFM and AFP.

As part of important efforts being made nation-wide to improve AFM surveillance in order to determine the burden and impact of this condition among all age groups, we now conduct AFM surveillance in Washington State. In June 2015, the Council of State and Territorial

Epidemiologists (CSTE) adopted a new standardized case definition for acute flaccid myelitis. In order to obtain a more accurate overall incidence of AFM, the new case definition includes all ages. To increase sensitivity in AFM surveillance, CSF findings (i.e. pleocytosis) were also added to the case definition. In June 2017, CSTE updated the standard case definition to clarify that a negative or normal MRI performed within the first 72 hours after onset of limb weakness does not rule out AFM. MRI studies performed 72 hours or more after onset should also be reviewed if available.

Figure 2. Acute Flaccid Paralysis and Acute Flaccid Myelitis most common etiologies



Acute Flaccid Myelitis (AFM), Poliovirus infection, and Poliomyelitis Surveillance Guidelines

1. DISEASE REPORTING

A. Purpose of reporting and surveillance

1. To identify cases of Acute Flaccid Myelitis (AFM) and establish an incidence baseline and burden of the condition in Washington State.
2. To help identify causes of AFM in the United States.
3. To understand the impact of AFM among all ages groups
4. To integrate AFM surveillance activities with those for poliomyelitis and non-paralytic poliovirus infection surveillance activities in order to establish a surveillance system sensitive enough to reliably detect cases of poliomyelitis.
5. To prevent transmission of poliovirus and to distinguish between wild-type polio and vaccine-associated paralytic polio, if a case of poliomyelitis occurs.

B. Legal Reporting Requirements

AFM:

1. Health care providers: notifiable to Local Health Jurisdictions within 24 hours
2. Health care facilities: notifiable to Local Health Jurisdictions within 24 hours
3. Laboratories: no requirements for reporting.
4. Local health jurisdictions: notifiable to the Washington State Department of Health (DOH) Office of Communicable Disease Epidemiology (CDE) within 7 days of case investigation completion or summary information required within 21 days.

Polio:

1. Health care providers: immediately notifiable to local health jurisdiction
2. Health care facilities: immediately notifiable to local health jurisdiction
3. Laboratories: Poliovirus, acute, by IgM positivity or PCR positivity immediately notifiable to local health jurisdiction; specimen submission is required – isolate or clinical specimen associated with positive result (2 business days)

Paralytic polio is designated “immediately notifiable, extremely urgent”, requiring state and local health authorities to notify CDC within 4 hours of their notification.

Non-paralytic polio is designated “immediately notifiable and urgent” requiring state and local health authorities to notify CDC within 24 hours of their notification.

C. Local Health Jurisdiction Investigation Responsibilities

1. Begin routine case investigation within one working day of notification. If polio is suspected as the cause of AFM begin investigation and notify CDE immediately.
2. Facilitate the transport of specimens to Washington State Public Health Laboratories (PHL) at the direction of CDE and CDC for AFM cases including suspected polio.

3. Implement appropriate infection control measures.
4. Report all *confirmed* and *probable* cases of AFM and polio (see definitions below) to CDE.
 - For AFM, complete the CDC-AFM patient summary form (available at <http://www.cdc.gov/acute-flaccid-myelitis/downloads/patient-summary-form.pdf>) and send a copy to CDE. This form MUST accompany any specimens submitted to CDC for testing.
 - For polio, complete the polio investigation form (available at <http://www.doh.wa.gov/Portals/1/Documents/5100/210-059-ReportForm-Polio.pdf>)
5. Enter into the Washington Disease Reporting System (WDRS) as “Polio” for AFM or if poliovirus is the suspected etiology.

2. THE DISEASE AND ITS EPIDEMIOLOGY

Background

The name poliomyelitis: Polios, “gray”; myelos, “marrow” or “spinal cord” is descriptive of the pathologic lesions that involve neurons in the gray matter, especially in the anterior horns of the spinal cord. These polio-like lesions are also seen in AFM due to other etiologies.

Wild polio virus was eliminated from the western hemisphere in 1991 but remains endemic in Afghanistan and Pakistan, with 14 and 8 cases reported in 2017, respectively, and 15 and 4 cases reported in 2018 (data to October 2, 2018). Until poliovirus transmission is interrupted in these countries, all countries remain at risk of importation of polio. Several countries in Africa and the Middle East are still considered vulnerable, given their weak public health and immunization systems combined with travel and trade links with endemic countries.

For up-to-date information regarding worldwide polio transmission see:

<http://polioeradication.org/polio-today/polio-now/this-week/>

In the fall of 2014 (August to October), CDC received an increased number of reports of children across the United States who had developed a sudden onset of weakness in one or more limbs. MRI scans showed inflammation of the gray matter—nerve cells—in the spinal cord, a condition called Acute Flaccid Myelitis or AFM. Sporadic cases have continued to be reported to CDC since then.

The United States has been considered a polio-free country since 1979, and AFP is not a nationally notifiable condition. Therefore, surveillance for AFP has not been systematically conducted in the United States for several decades. In the absence of a baseline incidence for AFP and/or AFM cases, the interpretation of the increase in AFM reports in 2014 was challenging.

A CDC outbreak case definition was developed (onset of acute limb weakness on or after August 1, 2014, and a magnetic resonance image showing a spinal cord lesion largely restricted to gray matter in a patient age <21 years). Using this case definition CDC verified reports of 120 children in 34 states who developed AFM between August 2014 and July 2015. The median age of the children was about 7 years, most patients had experienced fever and/or respiratory illness before the onset of neurologic symptoms. Almost all of these

children were hospitalized, and some required ventilator support.

About 70% of the children had elevated white blood cells count (pleocytosis) in cerebrospinal fluid (CSF), often with elevated protein levels also present. Among a group of these children who were observed for a median of 19 days after their illness, two thirds reported some improvement in symptoms, while about one third showed no improvement. Only two of the children have fully recovered.

Based on the number of cases identified during 2014 using the outbreak definition, the number of cases ≤ 21 years of age in any given state is estimated to range from 1-10 (2 – 3 in the majority of states) in any given year. By including persons of all ages, rather than just those ≤ 21 years of age, a standardized case definition will contribute to a better understanding of the etiology and epidemiology of AFM.

In June 2015, the Council of State and Territorial Epidemiologists (CSTE) adopted a standardized case definition for acute flaccid myelitis. The implementation of an AFM surveillance system will help us to determine an AFM incidence baseline, and to establish the sensitivity of our polio surveillance system. This case definition was revised in 2017 to clarify that a negative or normal MRI performed within the first 72 hours after onset of limb weakness does not rule out AFM, and MRI studies performed 72 hours or more after onset should also be reviewed if available.

In 2017, 33 confirmed AFM cases from 16 states were reported to CDC. Current AFM case activity available at: <http://www.cdc.gov/acute-flaccid-myelitis/afm-surveillance.html>

A. Etiologic Agent

AFM is most often associated with viruses that can cause inflammation and loss of motor and autonomic neurons located in the anterior horn cells of the spinal cord (i.e. front column of gray matter in the spinal cord). The symptoms are similar despite the etiologic agent and usually include paralysis but sensation is spared. Viruses that have been associated with AFM include:

- Enteroviruses (both polio and non-polio)
- West Nile virus (WNV) and viruses in the same family as WNV, specifically Japanese encephalitis virus and Saint Louis encephalitis virus
- Herpesviruses, such as cytomegalovirus and Epstein-Barr virus
- Adenoviruses

Poliovirus is an RNA virus that is a member of the family Picornaviridae, genus Enterovirus. AFM caused by poliovirus is known as poliomyelitis which is a notifiable condition in WA State and nationally. There are three serotypes (1, 2, and 3), and all can cause paralysis. Infection with each serotype confers serotype-specific lifelong immunity to disease but there is no cross-protective immunity for the other serotypes. Clinical poliomyelitis can be caused by wild-type viruses (WPV) and, rarely, by attenuated live (oral) vaccine strains (known as vaccine-derived poliovirus or VDPV).

The apparent increase in cases of AFM in 2014, which coincided with a national outbreak of

severe respiratory illness among children caused by enterovirus D68 (EV-D68), highlighted the absence of a known baseline for AFM incidence in the United States. Despite a close association in timing with respiratory illness caused by EV-D68 and extensive testing, a single cause for the apparent cluster of AFM cases could not be established. Although the specific cause of this increase in AFM cases is still under investigation, the illnesses are most similar to others that were confirmed as having a viral etiology.

It is important for public health investigators to recognize that in many cases, despite extensive laboratory testing, a definitive cause for AFM cannot be identified. When a specific pathogen is isolated from one of the patient's samples, it may still be difficult to attribute causality to that pathogen, especially if it was not isolated from CSF.

Please note: If WNV, other arboviral disease, or Influenza is suspected or confirmed to be the cause of AFM, the [WA DOH guidelines available for these conditions](#) should also be consulted.

B. Description of Illness

Most patients with AFM will have sudden onset of limb weakness and loss of muscle tone and reflexes. Some, in addition to the limb weakness, may also experience one or more of the following symptoms: facial droop or weakness (facial paralysis), difficulty moving the eyes (oculomotor dysfunction), drooping eyelids (blepharoptosis or palpebral ptosis), and difficulty with swallowing (dysphagia) or slurred speech (dysarthria).

Numbness or tingling (paresthesia) is rare in patients with AFM, though some patients do experience pain in their arms or legs. Some patients may be unable to pass urine (urinary retention). The most severe symptom of AFM is respiratory failure which can occur when the muscles involved with breathing, such as the diaphragm, become weak. This can require urgent ventilator support.

Clinical presentations of polio infection (including AFM) are much better described in the literature. (See Table 1). Poliovirus enters through the mouth and replicates in the pharynx and gastrointestinal tract. The virus then invades local lymphoid tissue and enters the bloodstream (viremia) and inducing type-specific immunity. For the majority of infections, the virus is contained at this point, but in some individuals the virus may infect cells of the central nervous system.

Some persons have nonspecific mild illnesses including fever, headache, sore throat, or gastrointestinal symptoms (e.g. vomiting, abdominal pain). In 4% to 8% of infections symptoms disappear within a period of 2-3 days, without the occurrence of any abnormal neurological symptoms. This presentation is known as abortive poliomyelitis and it cannot be distinguished from other viral infections and it is usually only detected during outbreaks or epidemics.

Poliovirus can infect the spinal cord or brain stem resulting in aseptic meningitis (1-5%) or acute, asymmetric, ascending flaccid paralysis which occurs in one out of every 200 poliovirus infections on average (<1%). Paralysis occurs more commonly in adults: 1 in 75 adults vs. 1 in 1000 children.

In paralytic polio, neurological symptoms typically progress within a few days, achieve a plateau for weeks, and then can resolve partially or fully. Legs are more often affected than arms. Bulbar paralysis affecting the cranial nerves may accompany extremity involvement

or can occur as the sole paralysis.

Differential diagnoses of poliomyelitis include other viruses. (See list in Section 2.D.)

In most persons with paralytic polio muscle function returns to some degree. Permanent weakness is observed in approximately two thirds of patients with paralytic poliomyelitis. Complete recovery is less likely when acute paralysis is severe and when patients require mechanical ventilation. Paralysis that is still present 60 days after onset will usually be permanent.

Table 1. Distribution and timeline of polio infection clinical presentations*

Clinical/Subclinical Form	Children	All patients	Days after exposure →				
			0	5	10	15	20
Unapparent/Asymptomatic	Up to 72%	Up to 95%	Minor illness		Major illness		
Abortive poliomyelitis	24%	5-8%	Minor illness		Major illness		
Non-paralytic polio (Aseptic meningitis)	1-5%	1-2%	Minor illness		Major illness		
Paralytic polio	<1%	<1%	Minor illness		Major illness		

*Modified from Paul JR. History of Poliomyelitis. New Haven, CT: Yale University Press; 1971

In children, poliovirus infection occur equally in boys and girls, although paralysis is more common in boys. Among adults, women are at greater risk of infection. Both the incidence and severity of poliomyelitis may be increased in pregnant women.

Complications of poliomyelitis include respiratory compromise due to muscle paralysis, myocarditis, gastrointestinal hemorrhage, paralytic ileus, bladder paralysis and urinary retention.

The case fatality rate for paralytic polio is 2-5% among children and 15-30% among adults.

In 20-85% of persons who experienced paralytic poliomyelitis in childhood, a new onset of muscle weakness, pain, atrophy, and fatigue can occur after an interval of 15-40 years, and is known as Post-polio syndrome. The affected muscles are usually the same as those that were affected during the original illness. Post-polio syndrome is not an infectious process. Persons experiencing the syndrome are not contagious.

C. Acute Flaccid Myelitis and polio in Washington

Washington State reported 3 cases of AFM in the fall of 2014. Coxsackie virus A16 was found by CDC laboratory in two stool samples from one patient. The other two patients tested negative for enteroviruses or other causative organism. No cases were reported in 2015 while there were 10 cases in 2016 and 3 cases in 2017.

The last endemic transmission of wild polio virus infection in the United States was in 1979; the last case of wild virus infection identified in Washington occurred in 1977. Vaccine-associated paralytic polio (VAPP) continued to occur sporadically, including in a

Washington resident who contracted the virus in 1993 from a grandchild recently vaccinated with oral polio vaccine (OPV). In 1997, the ACIP recommended routine use of inactivated (IPV) rather than oral polio vaccine to eliminate the risk of VAPP in the United States. In 2000, an all IPV vaccine schedule was implemented which has greatly reduced the occurrence of VAPP. However, an unvaccinated Arizona resident contracted VAPP in 2005 during international travel to a polio-endemic area where oral vaccine was in use.

D. Reservoirs for viruses known to be associated with AFM

Agent	Reservoir
Enteroviruses (polio and non-polio)	Humans, usually asymptomatic
West Nile Virus	Bird reservoir: corvids. Vector: Culex mosquitoes
Japanese encephalitis virus	Pigs and wild birds. Vector: Culex mosquitoes
Saint Louis encephalitis	Wild birds and domestic fowl. Vector: Culex mosquitoes
Cytomegalovirus	Humans, CMV strains found in animal species are not infectious for humans
Epstein-Barr virus	Humans. EBV will stay latent in a person for their lifetime
Adenovirus	Humans. Ubiquitous in the environment where contamination by human feces or sewage has occurred.

E. Modes of Transmission for viruses known to be associated with AFM

Agent	Route of transmission
Enteroviruses (polio and non-polio)	Mainly fecal-oral, including contaminated water. Respiratory droplet transmission is also possible. Infants shedding virus in the feces after having received OPV have been the source of exposure for susceptible adults giving child care.
West Nile virus	Mainly bites from infected mosquitoes. Other routes of transmission have been described: Blood transfusions, Organ transplants, Percutaneous injuries in laboratories, Mother to baby via the placenta and breast milk. Not spread through casual contact.
Japanese encephalitis virus	Bites from infected mosquitoes. Not directly transmitted from person-to-person.
Saint Louis encephalitis	Bites from infected mosquitoes. Person-to-person transmission has not been documented.
Cytomegalovirus	Direct contact with body fluids of infected people, such as urine, saliva, or breast milk. Sexual transmission. Transplanted organs and blood transfusions.
Epstein-Barr virus	Oral (contact with saliva of infected persons) Sexual contact. Blood transfusion (rare).
Adenovirus	Respiratory secretions Fecal-oral Waterborne Sexual transmission.

F. Incubation Period for viruses known to be associated with AFM

Agent	Incubation period
Enteroviruses	<u>Polio</u> : 6-20 days (range 3 to 36 days). <ul style="list-style-type: none"> • From contact to onset of paralysis: 11 to 17 days (range 8-36 days) <u>Non-polio</u> : 2-10 days
West Nile Virus	2-14 days
Japanese encephalitis virus	5-15 days
Saint Louis encephalitis	4-21 days
Cytomegalovirus	3 to 12 weeks
Epstein-Barr virus	Unknown for neurological symptoms. (30-50 days for infectious mononucleosis)
Adenovirus	Respiratory tract infection: 2-14 days. Gastroenteritis:3-10 days

G. Period of Communicability for viruses known to be associated with AFM

Agent	Period of communicability
Enteroviruses	<u>Polio</u> : Most infectious from 7-10 days before and after onset of symptoms when virus is present in the throat and excreted in high concentration in feces. After onset of illness, poliovirus might be present in the throat for 1-2 weeks, and in stool from 3-6 weeks. Persons with asymptomatic infections are also communicable. <u>Non-polio</u> : virus can be present 3-4 weeks in the pharynx, and 5-6 weeks in stool.
West Nile Virus	Infected people may develop a short lived (2–3 day) low-level viremia that can be found in donated blood; collection centers screen donated blood to prevent contaminated units from being used in transfusions.
Japanese encephalitis virus	Virus is not usually demonstrable in human blood after onset of disease, but can be isolated from CNS fluid in 1/3 of acute cases; viremia in birds usually lasts 2-5 days; mosquitoes are infective for life; viremia in horses rarely present in high titer for long periods
Saint Louis encephalitis	Virus is not demonstrated in human blood after onset of disease; however, the viremia response in infected birds is typically detected 1-5 days after infection, depending on the viral strain and bird species. Mosquitoes are infected for life.
Cytomegalovirus	CMV virus can persist in body fluids such as urine, saliva, and seminal fluids for many years, or can remain dormant until reactivation of latent infection.
Epstein-Barr virus	The virus is shed in respiratory secretions. Shedding decreases during the year following infection but persists throughout life. Peaks in transmission occur between 1-6 and 14-20 years of age and over 95% of adults are asymptomatic carriers of the virus.
Adenovirus	Children shed non enteric adenovirus in throat and stool samples for 3-6 weeks following lower respiratory infection or generalized febrile illness. Chance of transmission is higher in crowded and closed settings such as day cares, boarding schools and long-term care facilities. Transmission between family members is common. In rare cases, virus shedding may last for 18 months or longer.

H. Treatment

There is no specific treatment for AFM. In general, treatment is supportive and can include respiratory support, ICU (Intensive Care Unit) care, and physical therapy. However, neurologists may recommend certain interventions on a case-by-case basis, and for immunocompromised patients infected with CMV, intravenous ganciclovir or oral valganciclovir are the first line therapy. Treatment for polio is also supportive, and can include respiratory support and physical therapy especially following acute paralytic illness. Splints are used to relieve pain and spasms and prevent the development of deformities.

CDC discourages the use of plasmapheresis, immunosuppressive biologic modifiers, and IVIG (Intravenous Immunoglobulin) to treat patients with AFM due to any cause*. IVIG use

has been studied for post-polio syndrome patients, but is not recommended routinely.**

*Acute Flaccid Myelitis: Interim considerations for clinical management. Centers for Disease Control and Prevention. Available at <http://www.cdc.gov/acute-flaccid-myelitis/downloads/acute-flaccid-myelitis.pdf>

** Huang et al. Intravenous immunoglobulin for post-polio syndrome: a systematic review and meta-analysis. BMC Neurology (2015) 15:39. DOI: 10.1186/s12883-015-0301-9

I. Immunity for viruses known to be associated with AFM

Agent	Immunization
Enteroviruses	Non-polio: No immunizations available. Polio: Whereas Oral Polio Vaccine (OPV) produces intestinal immunity, Inactivated Polio Vaccine (IPV) protects only against paralytic disease and does not prevent intestinal infection or subsequent shedding of the virus. Only IPV is licensed and available in the United States. See Section 7.A. for routine immunization recommendations for prevention of polio.
West Nile Virus	Inactivated vaccine available for horses. Human vaccines in trials.
Japanese encephalitis virus	Inactivated Vero cell culture-derived Japanese encephalitis (JE) vaccine (manufactured as IXIARO) is the only JE vaccine that is licensed and available in the United States. 2 doses should be administered 28 days apart. The last dose should be given at least 1 week before travel.
Saint Louis encephalitis	None available
Cytomegalovirus	None available
Epstein-Barr virus	None available
Adenovirus	A vaccine for adenovirus strains 4 and 7 was developed but is no longer in production for economic reasons.

3. CASE DEFINITIONS

ACUTE FLACCID MYELITIS

A. Clinical Criteria

An illness with onset of acute focal limb weakness AND

- A magnetic resonance image (MRI) showing spinal cord lesion largely restricted to gray matter*† and spanning one or more spinal segments, OR
- Cerebrospinal fluid (CSF) with pleocytosis

B. Case Classification (2017)

1. Probable:

- An illness with onset of acute focal limb weakness AND
- CSF showing pleocytosis (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).

2. Confirmed:

An illness with onset of acute focal limb weakness AND

- MRI showing spinal cord lesion largely restricted to gray matter*† and spanning one or more spinal segments

*Terms in the spinal cord MRI report such as “affecting mostly gray matter”, “affecting the anterior horn or anterior horn cells”, “affecting the central cord”, “anterior myelitis”, or “poliomyelitis” would all be consistent with this terminology. If still unsure if this criterion is met, consider consulting the neurologist or radiologist directly.

† Spinal cord lesions may not be present on initial MRI; a negative or normal MRI performed within the first 72 hours after onset of limb weakness does not rule out AFM.

POLIO

A. Poliomyelitis, paralytic (2010)

Case classification

1. Probable:

Acute onset of a flaccid paralysis of one or more limbs with decreased or absent tendon reflexes in the affected limbs, without other apparent cause, and without sensory or cognitive loss.

Please note: All patients with no sensory or cognitive loss that present with a syndrome meeting the clinical criteria for AFM also meet the criteria for consideration as a possible paralytic poliomyelitis case and should therefore be considered immediately notifiable under WAC 246-101 to LHJs.

2. Confirmed:

Acute onset of a flaccid paralysis of one or more limbs with decreased or absent tendon reflexes in the affected limbs, without other apparent cause, and without sensory or cognitive loss; AND in which the patient

- Has a neurologic deficit 60 days after onset of initial symptoms; or
- Has died; or
- Has unknown follow-up status

Comment: All suspected cases of paralytic poliomyelitis will be reviewed by a panel of expert consultants before final classification occurs. Confirmed cases receive further classification based on epidemiologic and laboratory criteria¹. Only confirmed cases are included in Table 1I in the MMWR. Suspected cases are enumerated in a footnote to the MMWR table.

¹Sutter RW, Brink EW, Cochi SL, et al. A new epidemiologic and laboratory classification system for paralytic poliomyelitis cases. Am J Public Health 1989; 79:495-8.

B. Poliovirus infection, nonparalytic (2010)

Case classification

1. Confirmed:

Any person without symptoms of paralytic poliomyelitis in whom a poliovirus isolate identified in an appropriate clinical specimen with confirmatory typing and sequencing performed by the CDC Poliovirus Laboratory, as needed.

4. DIAGNOSIS AND LABORATORY SERVICES

The clinical presentation (e.g. the location of the weakness, muscle tone, and reflexes) helps differentiate patients with AFM from patients with other forms of acute flaccid paralysis. Magnetic resonance imaging (MRI) can be very helpful in diagnosing cases of AFM.

Testing nerve response can also be useful in supporting a diagnosis of AFM but should be performed at the appropriate time (e.g., 7-10 days after onset of weakness) to be helpful. Finally, by testing the CSF, clinicians can look for pleocytosis which is suggestive of AFM. All of these findings together allow a clinician to make the diagnosis of AFM.

A. Laboratory Diagnosis

A CDC neurologist reviews all available clinical, laboratory, and neuroimaging information to confirm AFM on reported cases. For all suspected AFM cases, CSF, serum, and a nasopharyngeal or oropharyngeal swab, and stool samples should be sent to CDC laboratories for enteroviruses and other pathogen testing to look for a specific etiologic pathogen.

The laboratory diagnosis of AFM etiologic agents can be made by isolation of the viruses from stool, nasopharyngeal or oropharyngeal swab, serum, or CSF; however, enteroviruses, including poliovirus, are most likely to be detected in stool cultures. In order to rule out poliovirus in a suspect AFM case, healthcare providers are urged to order a viral stool culture to be performed locally. A viral respiratory culture should also be ordered. Poliovirus is demonstrable in throat secretions as early as 36 hours and in stools as early as 72 hours after exposure in both clinical and inapparent cases. Occasionally, poliovirus may also be isolated from urine, and conjunctival fluids.

Acute and convalescent (early in the course and three weeks later) serologic specimens for polio testing can be obtained. A four-fold rise in the titer between the two specimens suggests poliovirus infection. Poliovirus infection may be ruled out if no antibody is detected in either specimen. However, results can be difficult to interpret because there are some limitations to antibody titers:

- For any patient, neutralizing antibodies appear early and may be at high levels by the time the patient is hospitalized; therefore, a four-fold rise in antibody titer may not be demonstrated.
- Patients who are immunocompromised may have two titers with no antibody detected and still be infected with poliovirus.

Serologic testing cannot distinguish between wild-type and vaccine derived virus.

B. Tests Available at Washington State Public Health Laboratories (PHL)

PHL does not perform testing for poliovirus or other enteroviruses, but the recommended surveillance specimens should be sent to PHL as soon as possible. PHL will forward specimens to CDC for testing. The following table summarizes organisms that have been associated with AFM and testing information:

Table 1. Test information for pathogens known to be associated with AFM

Specimen	Pathogen	Preferred test	Test available at PHL
CSF	Enterovirus	PCR	None
	West Nile Virus	PCR/IgM	IgM by MIA (Microsphere immunoassay)
	Herpes Simplex	PCR/Culture	Culture
	Cytomegalovirus	PCR	None
Respiratory	Enterovirus/Rhinovirus	PCR	None
	Adenovirus	PCR	Culture
	Influenza virus	PCR	PCR
Serum	West Nile Virus	PCR/IgM	IgM by MIA
Stool	Poliovirus	PCR/Culture	None
	Enterovirus (non-polio)	PCR	None

Note that PHL requires all clinical specimens have two patient identifiers, a name **and** a second identifier (e.g., date of birth) both on the specimen label and on the submission form. Due to laboratory accreditation standards, specimens will be rejected for testing if not properly identified. Also include specimen source and collection date.

C. Specimen Collection

Early specimen collection has the best chance to suggest an etiology for AFM. As soon as possible in the course of illness, preferably on the day of onset of limb weakness, clinicians should collect specimens from patients suspected of having AFM due to poliovirus or another enterovirus including:

- a) Two stool specimens collected as soon after onset of limb weakness and separated by 24 hours, minimum 1gram, AND
- b) Cerebrospinal fluid (CSF), minimum 1mL. (Should be collected at the same time, or within 24 hours of serum if feasible)
- c) Serum, minimum 0.4mL. (Should be collected at the same time, or within 24 hours of CSF if feasible)
- d) A nasopharyngeal or oropharyngeal swab, stored in minimum 1mL viral transport media. *Oropharyngeal swab should always be collected in addition to the nasopharyngeal specimen on any patient suspected to have polio.*
- e) For patients suspected to have polio, acute and convalescent serum can be collected. The acute specimen should be collected as soon as possible and forwarded to PHL along with the specimens listed above. The convalescent specimen should be collected 3 weeks after the acute specimen.

More detailed instructions regarding collection and shipping of specimens to be sent to CDC can be found at <http://www.cdc.gov/acute-flaccid-myelitis/hcp/instructions.html>

Office of Communicable Disease Epidemiology will assist with the determination of which specimens should be collected:

- Any specimen intended for testing at PHL should arrive with a completed Virus Examinations form available at: <http://www.doh.wa.gov/Portals/1/Documents/5230/302-017-SerVirHIV.pdf>
- All specimens intended for testing at CDC, should arrive at PHL with the CDC Patient Summary Form available at: <http://www.cdc.gov/acute-flaccid-myelitis/downloads/patient-summary-form.pdf>

5. ROUTINE CASE INVESTIGATION

Any person noted to have AFM has the potential to be a polio case. Immediately obtaining information about prior immunizations and recent travel or exposure to a recent OPV vaccinee is extremely important for every suspect AFM case.

A. Evaluate the Diagnosis

Determine the likelihood of the diagnosis:

- Review the clinical presentation, physical exam findings (particularly flaccid weakness).
- Review immunization history and risk factors for infection (e.g., recent travel to a polio endemic area or possible exposure to a person that recently received oral polio vaccine).
- Obtain history of any recent viral respiratory and/or gastrointestinal illness.
- Confirm that clinical criteria including CSF findings and/or MRI test results are met for AFM cases.
- If pursuit of laboratory testing is indicated, facilitate timely collection of appropriate specimens and expedite transport of those specimens to PHL.
- If a commercial laboratory isolates polio virus in cell culture, request that the laboratory send the cell culture to PHL for confirmatory testing immediately.

B. Identify Source of Infection

Ask about the following exposures in the 3–35 days prior to onset:

- Contact with person with similar symptoms or an AFM diagnosis
- Travel to a polio endemic area
- Travel to or contact with persons from an area where OPV is used
- Contact with a traveler arriving from an endemic area
- Contact with a person who recently received OPV

C. Identify Potentially Exposed Persons

For non-polio causes of AFM, contact investigation is not usually recommended but may be considered in a case by case basis.

If Poliovirus is confirmed as the cause for AFM, DOH and the Centers for Disease Control and Prevention will assist with an extensive contact investigation. See Appendix A for Contact Identification.

D. Environmental Evaluation

Routine environmental sampling for polio virus is not currently done in the United States.

If Poliovirus infection is confirmed as the cause for AFM, an environmental evaluation might be indicated depending on the specific circumstances of the case.

6. CONTROLLING FURTHER SPREAD

A. Infection Control Recommendations / Case Management

If a person is confirmed to have AFM due to Poliovirus, DOH in consultation with CDC will assist in making other infection control recommendations for the management of the case.

- Each infected person shall remain in isolation for 10 days from the onset of illness. Enteric precautions shall be followed for six weeks.
- Standard precautions for hospitalized case-patients, with contact precautions indicated for hospitalized infants and young children.

B. Contact Management

No specific management for contacts of AFM cases is indicated. Contacts should be educated regarding the specific etiology if one is suspected (e.g. vectors for arbovirus or lack of symptoms or non-specific symptoms associated with uncomplicated enterovirus infection) and advised about when they should seek medical care.

If Polio is suspected or confirmed, DOH in consultation with CDC will assist LHJs with identification and management of contacts. Contacts of a suspected polio case must be identified and monitored for symptoms. Collection of stool and serum samples from household members and other contacts associated with possible transmission settings are likely to be required. Vaccination with IPV should be offered to susceptible contacts of a confirmed polio case with an emphasis on persons who have an ongoing risk of exposure.

Note: If polio is confirmed and no source has been identified, a retrospective survey of hospitals that serve the community at risk should be conducted to review the illnesses of patients with diagnoses that might be consistent with poliovirus infection.

For additional information see Appendix A for Contact Identification and Appendix B for Contact Management recommendations.

7. MANAGING SPECIAL SITUATIONS

Special situations related to AFM will be handled on a case by case basis. Please consult with Office of Communicable Disease Epidemiology.

A. Immunization Recommendations

Table 2: Routine Schedule for Childhood Polio Vaccination

Dose	Age	Minimal Interval
IPV 1	2 months	N/A
IPV 2	4 months	4 weeks
IPV 3	6-18 months	4 weeks
IPV 4*	4-6 years	6 months
IPV 5	4–6 years	Only if 4 or more doses were administered before age 4

*A fourth dose is not necessary if the third dose was administered at age 4 years or older and at least 6 months after the previous dose.

Inactivated polio vaccine (IPV) contains three polio serotypes and is recommended for all children in the United States to be given in a four-dose series with doses at 2 months, 4 months, 6–18 months and 4–6 years. The fourth dose should be administered on or after the fourth birthday and at least 6 months after the previous dose. If 4 doses are administered prior to age 4 years, a fifth dose should be administered at age 4 to 6 years. In the event the third dose was given on or after the fourth birthday a fourth dose is not required for school entry.

IPV is not routinely recommended for U.S. residents ages 18 or older. Adults who have never been vaccinated against polio should receive three doses of IPV if they are:

- Traveling to polio-endemic or high-risk areas of the world.
- Working in a laboratory and handling specimens that might contain polioviruses.
- A health care worker in close contact with a person who could be infected with poliovirus.

Adults at high risk of coming in contact with polio virus who have received the 3 dose primary series should receive a booster dose of IPV.

Although no longer recommended in the United States, OPV is used elsewhere and can cause paralytic disease in unimmunized travelers if exposed (such as through contaminated food or water.) It is important to remember that IPV protects against paralytic polio but not intestinal infection. It is possible that a person vaccinated with inactivated polio vaccine could acquire an intestinal polio infection (wild type or VDPV) during travel to a polio-endemic area or to an area where OPV is used and subsequently transmit the virus to others through the fecal-oral route without becoming ill.

If both OPV and IPV were administered as part of a child’s vaccination series, a total of 4 doses should be administered, regardless of the child’s current age. If only OPV was administered, and all doses were given prior to age 4 years, 1 dose of IPV should be given at age 4 years or older, and at least 4 weeks after the last dose of OPV.

All OPV recipients should avoid close contact with immunodeficient persons for approximately 4-6 weeks after vaccination. If this is not feasible, rigorous hygiene and hand washing after contact with feces (e.g., after diaper changing) and avoidance of contact with saliva (e.g., sharing food or utensils) can be used but may be less effective. Maximum

excretion of vaccine virus occurs within 4 weeks after oral vaccination.

The last case of type 2 wild poliovirus disease occurred in 1999. Following the confirmation of eradication of type 2 wild poliovirus in September 2015, a globally synchronized phase withdrawal of OPV has started. The final goal is the removal of all OPVs in the long term.

From April to May 2016, 155 countries and territories participated in the first phase which was to substitute bivalent OPV (bOPV= type 1 and 3 poliovirus) for trivalent OPV (tOPV) in order to further reduce the risk of disease from circulating vaccine derived polioviruses (cVDPV). Over 90% of cVDPV cases, and approximately 40% of VAPP cases, are due to the type 2 component of tOPV. The type 2 component of tOPV also interferes with the immune response to poliovirus types 1 and 3.

Preparation for the removal of OPVs will also include the introduction of at least one dose of IPV into routine immunization schedules in all countries. The use of IPV will:

- Help to reduce risks associated with the withdrawal of OPV type 2,
- Hasten eradication by boosting immunity to poliovirus types 1 and 3, and
- Facilitate interruption of transmission if monovalent OPV type 2 was ever needed to control an outbreak.

For additional information regarding polio vaccination see the CDC Pink Book:

Centers for Disease Control and Prevention. *Epidemiology and Prevention of Vaccine-Preventable Diseases*. Hamborsky J, Kroger A, Wolfe S, eds. 13th ed. Washington D.C. Public Health Foundation, 2015.

B. Prevention Recommendations

Control of polio is accomplished through immunization. Unimmunized persons at risk of exposure, for example during travel to areas with known polio cases or areas where OPV is used, should maintain strict prevention measures to avoid potential fecal-oral transmission. These include using good hand washing techniques and safe drinking water during travel to areas with endemic polio and maintaining good hygiene practices if in contact with infants that are receiving oral vaccine.

Protection against mosquito-borne viruses that can cause AFM can be obtained using mosquito repellents and staying indoors at dusk and dawn, which is the prime period that mosquitoes bite. Removal of standing or stagnant water from nearby property to minimize the number of mosquitoes is also recommended.

Regular hand washing with soap and water, avoid close contact with sick people, and clean surfaces that a sick person has touched, can help prevent transmission of other known causes of AFM.

UPDATES (To previous poliomyelitis guideline)

January 2010 Section 1: The investigation form link was updated and 2010 case classification information was added.

January 2011: The Legal Reporting Requirements section has been revised to reflect the 2011 Notifiable Conditions Rule revision.

October 2016: AFM guidance and polio guideline were combined. Global epidemiology data was updated. Information regarding trivalent OPV vaccine switch to bivalent OPV vaccine in low and middle income countries was included.

October 2018: The AFM case definition was updated to reflect the June 2017 CSTE revision. Specimen collection instructions for AFM were updated to reflect the latest CDC recommendations. Global epidemiology for polio and national and state-wide epidemiology data for AFM was updated.

ACKNOWLEDGEMENTS

This document is a revision of the Washington State Guidelines for Notifiable Condition Reporting and Surveillance published in 2002 which were originally based on the Control of Communicable Diseases Manual (CCDM), 17th Edition; James Chin, Ed. APHA 2000. We would like to acknowledge the Oregon Department of Human Services for developing the format and select content of this document.

APPENDIX A. POLIOMYELITIS CONTACT IDENTIFICATION

Contact identification should be initiated as soon as poliovirus etiology is highly suspected (unimmunized patient with AFM and a high risk exposure).

Define Contacts:

Exposure is defined as contact with the stool or oral secretions (e.g. saliva) of an infectious person. Some examples of persons who should be considered contacts:

- Persons having contact (or potential for contact) with stool or fecal matter of the case within 30 days before the case's onset of illness, without using infection control precautions
- Persons living in the same household or having close contact with the case (e.g., sharing sleeping arrangements, shared utensils or towels) within 30 days prior to the case's onset
- Children attending the same daycare as the case or frequently playing together

First Steps:

- Inquire about case's activities and occupations during the communicable period 10 days prior to and after onset of symptoms.
- Record case occupation(s) and any other high risk activities with dates, descriptions, and locations:
 - Food handling
 - Provision of childcare in any setting
 - Daycare attendance, employment, or household contact of an attendee or employee.
 - Direct patient care
 - Other activities with high risk for fecal-oral or droplet transmission
- Obtain a contact name and number for a person in each high-risk setting that can help you identify contacts in the setting

Evaluate contact susceptibility:

- Contacts should be surveyed regarding polio vaccination status, immune status, and recent compatible illness.
- Contacts with no written record of a complete polio immunization series must be considered susceptible.
- A complete polio immunization series includes three primary doses and a single booster dose of IPV, when doses are received after 6 weeks of age and at intervals > 4 weeks apart.
- Other countries' vaccination schedules may include OPV. To assess completeness for a person vaccinated outside the United States, please see the WHO list of current vaccination schedules available at:
http://apps.who.int/immunization_monitoring/globalsummary/schedules

The risk for transmission in communities with low vaccination coverage is high. The estimated rate of transmission for wild poliovirus among unvaccinated household contacts is 73%-96%¹

¹Zimmerman K, Middleton D, Burns I, Clover R. Routine vaccines across the life span, 2003 clinical review. J Fam Pract 2003;52 (suppl 1):s1-s21.

APPENDIX B. POLIOMYELITIS CONTACT MANAGEMENT

The suspicion of poliomyelitis or poliovirus infection, particularly in a member of a group that refuses vaccination, should prompt an immediate response. The following active surveillance activities should be initiated in order to assure timely ascertainment of secondary cases and prevention of further transmission:

1. Create a list of all identified contacts.
2. Obtain name, address, and telephone number of every contact.
3. Determine occupation for each contact.
4. Note any school or daycare attendance. (Facility name and location)
5. Document each contact's immunization status to identify all susceptible contacts.
6. Define the community at risk and possible transmission settings based on epidemiologic data.
7. Maintain a line list of all contacts until at least 36 days after the exposure of each person that contains the following information (at a minimum):
 - Immunization history
 - Susceptibility status
 - Dates of screening and follow-up interviews
 - Were recommendations provided?
 - Whether any symptoms developed and description
 - Tests performed and results
 - Disposition of the contact (e.g. hospitalized, follow-up completed)
8. Consult with WA DOH CDE and Immunization Program regarding appropriate strategies for the effective use of OPV and/or IPV.
 - If evidence indicates wild-type poliovirus, an outbreak control program with vaccination planning is required.
 - Communities at risk should be assessed for current vaccination status and, at a minimum, one dose of IPV should be provided for any contact without documentation of a complete polio immunization series
 - All susceptible contacts 6 weeks of age and older with an incomplete or undocumented vaccination series or booster should be vaccinated on an accelerated schedule (4-week intervals)
 - A booster dose of vaccine is recommended for all adults (>18 years of age) in susceptible members of the community at risk and health-care workers at high risk for exposure who have completed a primary series but have not received an adult booster dose
 - OPV should be used in certain situations and will be determined by epidemiologic data
 - OPV should never be administered to immunodeficient patients or their household contacts; IPV should be used in such situations
 - If evidence indicates vaccine-associated disease, no outbreak control program is needed.
9. Track the number of susceptible contacts receiving recommended vaccination(s).
10. Maintain active community-wide surveillance for 2 incubation periods (i.e., 72 days) beyond the onset of the last case in the area.
11. Refer any identified contact that become symptomatic for immediate medical assessment.
12. Manage any susceptible contact that becomes symptomatic as a suspect polio case.
13. Contacts of a confirmed polio case can be quarantined by order of the Local Health Officer. However, this has not been shown to be effective in preventing transmission once a case has occurred and is not usually recommended.