

## Chapter 12: Poliomyelitis

Janell A. Routh, MD, MHS; M. Steven Oberste, PhD; Manisha Patel, MD, MS

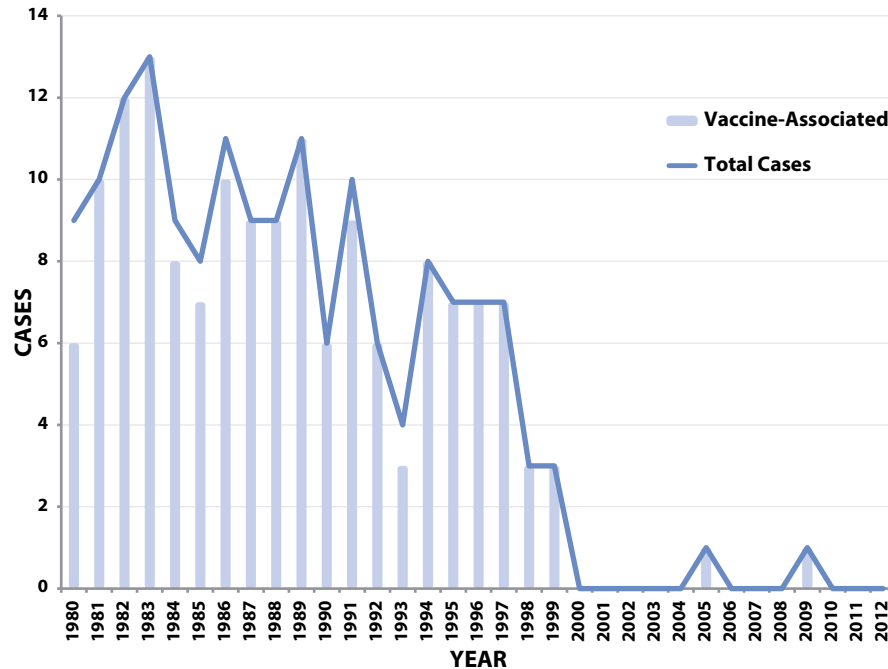
### I. Disease Description

Poliomyelitis is a highly contagious disease caused by 3 serotypes of poliovirus. Infection with poliovirus results in a spectrum of clinical manifestations from inapparent infection to nonspecific febrile illness, aseptic meningitis, paralytic disease, and death. Two phases of acute poliomyelitis can be distinguished: a nonspecific febrile illness (minor illness) followed, in a small proportion of patients, by aseptic meningitis and/or paralytic disease (major illness). The ratio of cases of inapparent infection to paralytic disease among susceptible individuals ranges from 100:1 to 1000:1 or more. Following poliovirus exposure, viral replication occurs in the oropharynx and the intestinal tract. Viremia follows, which may result in infection of central nervous system cells. The virus attaches and enters cells via a specific poliovirus receptor. Replication of poliovirus in motor neurons of the anterior horn and brain stem results in cell destruction and causes the typical clinical manifestations of poliomyelitis. Depending on the site of infection and paralysis, poliomyelitis can be classified as spinal, bulbar, or spino-bulbar disease. Progression to maximum paralysis is rapid (2–4 days); paralysis is usually associated with fever and muscle pain, and rarely progresses after the temperature has returned to normal. Spinal paralysis is typically asymmetric, more severe proximally than distally, and deep tendon reflexes are absent or diminished. Bulbar paralysis may compromise respiration and swallowing. Between 2% and 10% of paralytic poliomyelitis cases are fatal. Infection with poliovirus results in lifelong, type-specific immunity. Following the acute episode, many patients recover muscle functions at least partially, and prognosis for recovery can usually be established within 6 months after onset of paralytic manifestations.

### II. Background

Poliomyelitis became an epidemic disease in the United States at the turn of the 20th century. Epidemics of ever-increasing magnitude occurred, with more than 20,000 cases of poliomyelitis with permanent paralysis reported in 1952. Following the introduction of effective vaccines—inactivated poliovirus vaccine (IPV) initially in 1955, then oral poliovirus vaccine (OPV) starting in 1961—the reported incidence of poliomyelitis in the United States declined dramatically to <100 cases in 1965 and to <10 cases in 1973. With the introduction and widespread use of OPV (containing live attenuated poliovirus strains), vaccine-associated paralytic poliomyelitis (VAPP) was recognized. By 1973, for the first time in the United States, more cases of vaccine-associated disease were reported than paralytic disease caused by wild poliovirus.<sup>1</sup> This trend continued, and in 1997 the Advisory Committee on Immunization Practices (ACIP) recommended changing to a sequential polio immunization schedule that included 2 doses of IPV, followed by 2 doses of OPV.<sup>2</sup> VAPP occurred less frequently under this schedule, and in 2000, this recommendation was updated to a schedule of all IPV.<sup>3–5</sup> OPV is no longer manufactured or available in the United States. The last US cases of indigenously transmitted wild poliovirus disease were reported in 1979. Since 1986, with the exception of one imported wild-type poliomyelitis case in 1993, all reported cases of paralytic poliomyelitis in the United States have been vaccine-associated (see Figure 1).<sup>6,7</sup> VAPP was a very rare disease, with an average of eight reported cases annually during 1980–1999, or 1 case reported for every 2.4 million doses of OPV distributed.<sup>6,7</sup> The risk of VAPP is highest following the first dose of OPV and among immunodeficient persons. Since changing to an all-IPV immunization schedule in 2000, there have been only 3 cases of VAPP reported in the United States, 1 in an imported case, 1 in a genetically immunocompromised person who was most likely exposed to OPV before its use was discontinued, and 1 fatal case in an immunocompromised child from India who was given OPV as part of routine childhood immunizations in India.<sup>8,9,10</sup>





**Figure 1: Total number of reported paralytic poliomyelitis cases (including imported cases) and number of reported vaccine-associated cases—United States, 1980–2012**

Following the successful implementation of the polio eradication initiative in the Americas that began in 1985, the last case of wild poliovirus-associated disease was detected in Peru in 1991. The hemisphere was certified as free of indigenous wild poliovirus in 1994.<sup>11</sup> In 1988, the World Health Assembly adopted the goal of worldwide eradication of poliomyelitis by the year 2000.<sup>12</sup> By 2001, substantial progress toward eradication had been reported: a >99% decrease in the number of reported cases of poliomyelitis was achieved.<sup>13</sup> In 2020, wild poliovirus remains endemic in just 2 countries: Afghanistan and Pakistan in Asia. Due to the successful implementation of the global poliomyelitis eradication initiative, the risk of importation of wild poliovirus into the United States decreased substantially over the last decade. Nevertheless, the potential for importation of wild poliovirus into the United States remains until worldwide poliomyelitis eradication is achieved. Find more information on the status of poliomyelitis eradication at <http://www.polioeradication.org/>.

Because inapparent infection with OPV or wild virus strains no longer contributes to the establishment or maintenance of poliovirus immunity in the United States, universal vaccination of infants and children is the only means of establishing and maintaining population immunity against poliovirus to prevent poliomyelitis cases and epidemics caused by importation of wild virus into the United States. Population-based surveys conducted during 2009–2010 confirmed that the prevalence of poliovirus antibodies among school-age children, adolescents, and young adults in the United States is high (>90% to poliovirus types 1 and 2, and >83% to type 3).<sup>14</sup> In addition, seroprevalence surveys conducted in two inner-city areas of the United States (areas in which routine coverage was low) during 1990–1991 found that >80% of all children 12–47 months of age had antibodies to all three poliovirus serotypes.<sup>15</sup> Data from 1997–1998 also demonstrated a high seroprevalence of antibody to all poliovirus serotypes among children 19–35 months of age who lived in the inner-city areas of four US cities, with 96.8%, 99.8%, and 94.5% seropositive to poliovirus types 1, 2, and 3, respectively.<sup>16</sup> However, members of certain religious groups that object to vaccination have remained susceptible to poliomyelitis. These groups appear to be at highest risk for epidemic poliomyelitis. The last two outbreaks of poliomyelitis in the United States were reported among religious groups—in 1972 among Christian Scientists<sup>17</sup> and in 1979 among the Amish.<sup>1</sup> Clustering of other subpopulations that object to vaccination can also occur, which could increase the susceptibility to vaccine-preventable diseases, including polio.<sup>18</sup>

The emergence of circulating vaccine-derived polioviruses (cVDPVs) causing an outbreak of poliomyelitis was first reported in Hispaniola in 2000.<sup>19</sup> One or more cVDPV outbreaks have been reported each year since.<sup>20</sup> These outbreaks have occurred in regions where OPV is being used and overall routine polio vaccination rates are low. The vaccine polioviruses are able to replicate in the intestinal tract of inadequately immunized persons, and may be transmitted to others with inadequate immunity. During these multiple infections, the viruses may regain some of the properties of wild polioviruses, such as transmissibility and neurovirulence. Clinical disease caused by these VDPVs is indistinguishable from that caused by wild polioviruses. Outbreak control measures in these outbreaks have relied upon vaccination with OPV. A circulating VDPV was identified in an undervaccinated Amish community in the United States in 2005.<sup>21</sup> In 2013, the World Health Organization (WHO) set a target of a polio-free world by 2018.<sup>22</sup> Among the three WPV types, type 2 was declared eradicated in September 2015. To remove the risk for infection with cVDPVs type 2, all OPV-using countries simultaneously switched in April 2016 from trivalent OPV (tOPV) to bivalent OPV (bOPV), which contains only types 1 and 3 polioviruses.<sup>23</sup> Please refer to The Pink Book [<https://www.cdc.gov/vaccines/pubs/pinkbook/index.html>] for recommendations for poliovirus vaccination in light of this switch.

### III. Importance of Rapid Identification

Rapid investigation of suspected poliomyelitis cases is critical for identifying possible wild poliovirus transmission. Rapid detection of wild or virus-related cases permits the timely implementation of controls to limit the spread of imported wild poliovirus or cVDPVs and maintain the eradication of wild poliovirus in the United States. Moreover, rapid investigation of suspected cases will allow collection of specimens for poliovirus isolation, which is critical for confirming whether a case of paralytic poliomyelitis is the result of wild or vaccine-related virus infection.

### IV. Importance of Surveillance

The poliomyelitis surveillance system serves to 1) detect importation of wild poliovirus into the United States and 2) detect the presence of vaccine-derived poliovirus in the United States.

### V. Disease Reduction Goals

No cases of paralytic polio due to indigenously acquired wild poliovirus have been reported in the United States since 1979. There have been three reported cases of VAPP in the United States since 2000, when the use of OPV was discontinued. High coverage with poliovirus vaccine is required to maintain elimination of poliomyelitis in the United States until global eradication is achieved.

### VI. Case Definition

#### *Poliomyelitis, paralytic*

The following case definition for paralytic poliomyelitis has been approved by the Council of State and Territorial Epidemiologists (CSTE) and was published in 2010.<sup>24</sup>

#### *Case classification*

**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.

**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 are reviewed by a panel of expert consultants before final classification occurs. Confirmed cases are then further classified based on epidemiologic and laboratory criteria.<sup>25</sup> Only confirmed cases are included in Table 1 in the *Morbidity and Mortality Weekly Report* (MMWR). Suspected cases under investigation are enumerated in a footnote to the MMWR table.

### *Poliovirus infection, non-paralytic*

The following case definition for non-paralytic poliovirus infection has been approved by CSTE and was published in 2010.<sup>24</sup>

#### *Case classification*

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

## VII. Laboratory Testing

Laboratory studies, especially attempted poliovirus isolation, are critical for confirming whether a case of paralytic poliomyelitis is the result of wild or vaccine-related virus infection. Refer to Chapter 22, “Laboratory Support for Surveillance of Vaccine-Preventable Diseases” (<https://www.cdc.gov/vaccines/pubs/surv-manual/chpt22-lab-support.html>) for detailed information on laboratory testing for poliovirus including specific information on specimen collection and shipment.

### *Specimen collection*

Specimen collection and shipping are important steps in obtaining laboratory diagnosis or disease confirmation. Guidelines have been published for specimen collection and handling for viral and microbiologic agents. Information is also available on using CDC laboratories as support for reference and disease surveillance; this information includes

- A central website for requesting lab testing (<http://www.cdc.gov/laboratory/specimen-submission/index.html>)
- The CDC Infectious Diseases Test Directory (<http://www.cdc.gov/laboratory/specimen-submission/list.html>)
- The form required for submitting specimens to CDC (see Appendix 23, Form # CDC 0.5034) and
- Information on general requirements for shipment of etiologic agents (Appendix 24 at <http://www.cdc.gov/vaccines/pubs/surv-manual/appx/appendix24-etiological-agent.pdf>)

State laboratories and CDC provide an online test directory that contains not only a list of orderable tests for that institution, but also detailed information such as appropriate specimen types, collection methods, specimen volume, and points of contact.

Specific instructions for specimen collection and shipping may be obtained from the CDC poliovirus website (<https://www.cdc.gov/olio/us/lab-testing/specimens.html>) or by contacting the CDC Polio/Picornavirus Laboratory at 404-639-5499. Specimens for virus identification should be sent to CDC as directed by the State Health Department.

## VIII. Reporting and Case Notification

### *Case reporting within a jurisdiction*

Each state and territory (jurisdiction) has regulations or laws governing the reporting of diseases and conditions of public health importance.<sup>26</sup> These regulations and laws list the diseases to be reported and describe those persons or groups responsible for reporting, such as healthcare providers, hospitals, laboratories, schools, daycare and childcare facilities, and other institutions. Detailed information on reportable conditions in each jurisdiction is available through CSTE.<sup>27</sup> Contact your jurisdiction/state health department for reporting requirements in your jurisdiction. The *Suspected Polio Case Worksheet* is included as Appendix 14, to serve as a guide for data collection during investigation of reported cases.

### *Case notification to CDC*

Because poliomyelitis has been eliminated from the Americas, each reported case of suspected poliomyelitis should be followed up by local and state health departments in close collaboration with CDC. The jurisdiction in which the patient resides at the time of diagnosis should submit the case notification to CDC. Case notifications should not be delayed because of incomplete information or lack of confirmation; case notifications can be updated electronically as more information becomes available.

Paralytic polio has been classified as “Immediately notifiable, Extremely Urgent,” which requires that local and state health departments contact CDC within 4 hours (Emergency Operations Center, 770-488-7100). Notifications for suspected cases of paralytic polio should be sent to CDC using event code 10410 in the National Notifiable Disease Surveillance System (NNDSS). Reports of nonparalytic polio are designated as “Immediately notifiable, Urgent,” which requires notification to CDC within 24 hours. Notifications for suspected cases of non-paralytic polio should be sent to CDC using event code 10405 in NNDSS.

CDC (Emergency Operations Center, (770-488-7100) will provide consultation regarding the collection of appropriate clinical specimens for virus isolation and serology, the initiation of appropriate consultations and procedures to rule out or confirm poliomyelitis, the compilation of medical records, and most importantly, the evaluation of the likelihood that the disease may be caused by wild poliovirus.

### *Information to collect*

Demographic, clinical, and epidemiologic information are collected to determine:

- whether the suspected case meets the case definition for paralytic poliomyelitis and
- whether the disease may be caused by wild poliovirus

The following data elements are epidemiologically important and should be collected in the course of a case investigation (see Appendix 14 for details on each data category). Additional information may be collected at the direction of the state health department or CDC.

- Demographic information
  - Name
  - Address
  - Date of birth
  - Age
  - Sex
  - Ethnicity
  - Race
  - Country of birth
  - Length of time resident in the United States
- Reporting source
  - County
  - Earliest date reported
- Clinical
  - Hospitalizations: dates and duration of stay
  - Date of onset of symptoms
  - Complications
  - Immunologic status of case-patient
  - Outcome (case survived or died)
  - Date of death
  - Postmortem examination results
  - Death certificate diagnoses
- Laboratory and clinical testing
  - Serologic test
  - Stool test
  - Throat swab test
  - Electromyogram (EMG)
  - Magnetic resonance imaging (MRI)

- Vaccine information
  - Dates and types of polio vaccination
  - Number of doses of polio vaccine received
  - Manufacturer of vaccine
  - Vaccine lot number
  - If not vaccinated, reason
- Epidemiological
  - Recent travel to polio-endemic areas or OPV-using countries
  - Contact with persons recently returning from polio-endemic areas or OPV-using countries
  - Contact with recent OPV recipient
  - Setting (Is case-patient a member of a group objecting to vaccination?)

### *Travel history*

Because the last cases of paralytic poliomyelitis due to indigenously acquired wild poliovirus infection in the United States were reported in 1979, it is likely that wild poliovirus in a suspected case-patient is imported, either by the suspected patient directly or by a contact of the case-patient. Results of virus isolation and differentiation may not be available at the time of the case investigation. Therefore, to rule out the possibility of imported wild poliovirus, a detailed travel history of suspected cases and of other household and nonhousehold contacts should be obtained. Any history of contacts with visitors, especially those from polio-endemic areas, might be particularly revealing.

### *Setting*

Because the last two outbreaks of poliomyelitis in the United States were reported among Christian Scientists in 1972<sup>17</sup> and the Amish in 1979,<sup>1</sup> a suspected case of poliomyelitis reported from a group objecting to vaccination should be assigned the highest priority for follow-up and collection of specimens. VDPVs also pose a risk of poliomyelitis in communities with low vaccination coverage. In addition, isolation of wild poliovirus from residents of Canada in 1993<sup>28</sup> and 1996<sup>29</sup> demonstrates the potential for wild poliovirus importation into this continent. The strain isolated in Canada in 1993 was linked epidemiologically and by genomic sequencing to the 1992 poliomyelitis outbreak in the Netherlands, and the 1996 isolate was from a child who had recently visited India.

## **IX. Vaccination**

For specific information about poliovirus vaccination, refer to The Pink Book [<https://www.cdc.gov/vaccines/pubs/pinkbook/index.html>], which provides general recommendations, including vaccine use and scheduling, immunization strategies for providers, vaccine content, adverse events and reactions, vaccine storage and handling, and contraindications and precautions.

## **X. Enhancing Surveillance**

A number of activities can improve the detection and reporting of cases and improve the comprehensiveness and quality of reporting. Additional surveillance activities are listed in Chapter 19, “Enhancing Surveillance.”

### *Promoting awareness*

Because of the severity of poliomyelitis disease, clinicians are often the first to suspect the diagnosis of poliomyelitis and they are the key to timely reporting of suspected cases. However, disease reporting by clinicians is often delayed because it is only after other differential diagnoses are ruled out that the diagnosis of poliomyelitis is considered. Efforts should be made to promote physicians’ awareness of the importance of prompt reporting of suspected cases to the state and local health department and CDC and of the need to obtain stool and throat specimens early in the disease course.

### *Ensuring laboratory capabilities*

Make sure that the state laboratory or other easily accessible laboratory facility is capable of performing, at a minimum, primary virus isolation on appropriate cell lines. The CDC polio laboratory is always available for consultation and/or testing.



### Obtaining laboratory confirmation

Appropriate stool and throat specimens (2 specimens taken at least 24 hours apart during the first 14 days after onset of paralytic disease) should be collected.

### Active surveillance

Active surveillance should be conducted for every confirmed case of poliomyelitis to ensure timely reporting. The diagnosis of a case of poliomyelitis, particularly in a member of a group that refuses vaccination (such as the Amish or Christian Scientists), should prompt immediate control measures as well as active surveillance activities. These activities should include active contact tracing among populations at risk.

### Streamlining reporting using electronic methods

Although many surveillance systems still rely on paper and pencil for data collection, use of data from sources such as electronic medical records, electronic case reporting,<sup>30–36</sup> and clinical laboratory information systems (LIMS) can significantly improve reporting speed, enhance data quality, and reduce workload.

## XI. Case Investigation

Timely collection of stool specimens is important in establishing the diagnosis and determining appropriate control measures, in the event of wild poliovirus isolation refer to the poliovirus website [<https://www.cdc.gov/polio/us/lab-testing/specimens.html>] for specific information on specimen collection and testing.

Guidelines and a worksheet for the investigation of suspected cases of poliomyelitis are included as Appendix 14. Suspected cases of poliomyelitis should be reported immediately to the state health department. CDC's Emergency Operations Center should also be contacted at 770-488-7100.

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