Chapter 10: Poliomyelitis

Joanne Cono, MD, ScM; Lorraine N. Alexander, RN, MPH

I. Disease description

Poliomyelitis is a highly contagious disease caused by three serotypes of poliovirus. Infection with poliovirus results in a spectrum of clinical manifestations from inapparent infection to non-specific febrile illness, aseptic meningitis, paralytic disease, and death. Two phases of acute poliomyelitis can be distinguished: a non-specific 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 ranges from 100:1 to 1000:1.

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. A specific receptor is needed for the virus to enter cells. 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 paralysis, poliomyelitis can be classified as spinal, bulbar, or spino-bulbar disease. Progression to maximum paralysis is rapid (2–4 days), usually associated with fever and muscle pain, and rarely continues 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%–10% of cases of paralytic poliomyelitis 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 century. Epidemics of ever-increasing magnitude occurred, with more than 20,000 cases of paralytic poliomyelitis reported in 1952. Following the introduction of effective vaccines, first inactivated poliovirus vaccine (IPV) in 1955, and 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 first recognized. By 1973, for the first time, more cases of vaccine-associated disease were reported than paralytic disease caused by wild poliovirus.[1] 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 OPV, followed by 2 doses of inactivated polio vaccine, IPV.[2] VAPP occurred less frequently under this schedule, and in 2000, this recommendation was updated to a schedule of all IPV.[3,4,5] OPV is no longer manufactured or available in the U.S.

The last U.S. cases of indigenously transmitted wild poliovirus disease were reported in 1979. Since then, apart from six cases of imported poliomyelitis, only one of which has occurred since 1986, all reported cases of paralytic poliomyelitis in the United States have been vaccine-associated (see **Figure 1**).[6, 7] VAPP was a very rare disease, with an average of eight reported cases annually during 1980–1999, or one case reported for every 2.4 million doses of OPV distributed.[6, 7] 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 no cases of VAPP reported in the U.S.

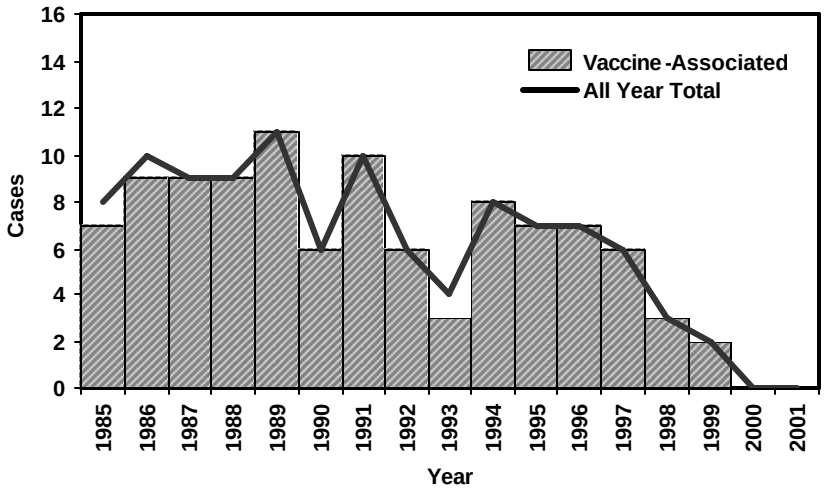

Figure 1: Total number of reported paralytic poliomyelitis cases (including imported cases) and number of reported vaccine-associated cases—United States, 1985–2001

Following the successful implementation of the polio eradication initiative in the Americas beginning 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.[8] In 1988, the World Health Assembly adopted the goal of worldwide eradication of poliomyelitis by the year 2000.[9] By 2001, substantial progress toward eradication has been reported: a more than 99% decrease in the number of reported cases of poliomyelitis was achieved; all polio-endemic and recently endemic countries have conducted National Immunization Days and established sensitive surveillance systems for acute flaccid paralysis. Wild polioviruses continue to be isolated from just seven countries: Afghanistan, India, and Pakistan in Asia, and Egypt, Niger, Nigeria, and Somalia in Africa.[10] Due to the successful implementation of

the global poliomyelitis eradication initiative, the risk of importation of wild polio virus 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.

Because inapparent infection with wild virus strains no longer contributes to establishing or maintaining 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 have 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 > 85% to type 3).[11,12] 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.[13] More recent data also demonstrate a high seroprevalence of antibody to all poliovirus serotypes among children aged 19–35 months who lived in the inner-city areas of four cities in the United States, with 96.8%, 99.8%, and 94.5% seropositive to poliovirus types 1,2, and 3, respectively.[14] However, members of certain religious groups objecting to vaccination have remained susceptible to poliomyelitis. These groups appear to be 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[15] and in 1979 among the Amish.[1]

Since 1999, there have been a few poliomyelitis outbreaks caused by vaccine-derived polioviruses.[16, 17, 18] These outbreaks have occurred in regions where OPV is being used and overall polio vaccination rates are low. The vaccine polioviruses are able to replicate unchecked in the guts of inadequately immunized persons, until regaining some of the infectious features of wild polioviruses. Clinical disease caused by these vaccine-derived viruses is indistinguishable from that caused by wild polioviruses. Outbreak control measures in these outbreaks rely upon vaccination with OPV.

One such outbreak of vaccine-derived type 1 OPV strain poliomyelitis occurred in the Dominican Republic (13 confirmed cases) and Haiti (8 confirmed cases, including 2 fatal cases), during 2000-2001.[16] Twenty of these cases occurred in unvaccinated or incompletely vaccinated cases, and all cases occurred in communities with very low (7%–40%) rates of coverage with OPV. The infecting vaccine-derived poliovirus strain was genetically traced back to OPV administered 1998–1999 and had biological properties indistinguishable from those of wild polioviruses. The World Health Organization assisted the National Ministries of Health in conducting OPV immunization campaigns in both countries; the campaigns successfully contained the outbreak. Other vaccine-derived poliomyelitis outbreaks have recently occurred in the Philippines [17] and Madagascar.[18] In all 3 outbreaks, the presumed risk factors for the outbreak included gaps in OPV coverage and absence of circulating wild poliovirus.

III. Importance of rapid identification

Rapid investigation of suspected poliomyelitis cases is critical to identifying possible wild poliovirus transmission. Rapid detection of wild virus-associated cases permits the timely implementation of controls to limit the spread of imported wild poliovirus 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 ruling out or confirming paralytic poliomyelitis, whether wild virus-associated or vaccine-related.

IV. Importance of surveillance

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

V. Disease reduction goals

No cases of paralytic polio due to indigenously acquired wild poliovirus have been reported in the United States since 1979. The goal of maintaining elimination of paralytic poliomyelitis due to indigenous acquisition of wild poliovirus has been established for each year until the goal of global eradication is met.[16] There have been no reported cases of vaccine-derived paralytic polio in the U.S.

VI. Case definition

The following case definition for paralytic poliomyelitis has been approved by the Council of State and Territorial Epidemiologists (CSTE), and was published in 1997.[20]

Clinical case definition

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.

Case classification

Probable: A case that meets the clinical case definition.

Confirmed: A case that meets the clinical case definition and in which the patient has a neurologic deficit 60 days after onset of initial symptoms, 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. Only confirmed cases are included in **Table 1** in the *Morbidity and Mortality Weekly Report (MMWR)*.[20]

Suspected cases under investigation are enumerated in a footnote to the quarterly immunization table of the *MMWR*.[20]

Confirmed cases are further classified based on epidemiologic and laboratory criteria.[18]

Indigenous case: Any case which cannot be proved to be imported.

Imported case: A case which has its source outside the United States. A person with poliomyelitis (United States resident or other) who has entered the United States and had onset of illness within 30 days before or after entry.[18]

VII. Laboratory testing

To increase the probability of poliovirus isolation, at least two stool specimens should be obtained 24 hours apart from patients with suspected poliomyelitis as early in the course of disease as possible (ideally within 15 days after onset).

Laboratory studies, especially attempted poliovirus isolation, are critical to rule out or confirm the diagnosis of paralytic poliomyelitis.

For additional information on laboratory support for surveillance of vaccine-preventable diseases, see Chapter 19, "Laboratory Support for Surveillance of Vaccine-Preventable Diseases."

Virus isolation

The likelihood of poliovirus isolation is highest from stool specimens, intermediate from pharyngeal swabs, and very low from blood or spinal fluid. The isolation of poliovirus from stool specimens contributes to the diagnostic evaluation but does not constitute proof of a causal association of such viruses with paralytic poliomyelitis.[1] Isolation of virus from the cerebrospinal fluid (CSF) is diagnostic but is rarely accomplished. To increase the probability of poliovirus isolation, at least two stool specimens and two throat swabs should be obtained 24 hours apart from patients with suspected poliomyelitis as early in the course of the disease as possible (i.e., immediately after poliomyelitis is considered as a possible differential diagnosis), but ideally within the first 14 days after onset of paralytic disease. Specimens should be sent to the state or other reference laboratories for primary isolation. Laboratories should forward isolates to CDC for intratypic differentiation to determine whether the poliovirus isolate is wild or vaccine-derived.

Isolation of wild poliovirus constitutes a public health emergency and appropriate control efforts must be initiated immediately (in consultation among health-care providers, the state and local health departments, and CDC).

Serologic testing

Serology may be helpful in supporting or ruling out the diagnosis of paralytic poliomyelitis. An acute serum specimen should be obtained as early in the course of disease as possible, and a convalescent specimen should be obtained at least 3 weeks later. A four-fold titer rise between the acute and convalescent specimens suggests poliovirus infection. Non-detectable antibody titers in both specimens may help rule out poliomyelitis but may be falsely negative in immunocompromised persons, who are also at highest risk for paralytic poliomyelitis. In addition, neutralizing antibodies appear early and may be at high levels by the time the patient is hospitalized; thus, a four-fold rise may not be demonstrated. Vaccinated individuals would also be expected to have measurable titers; therefore vaccination history is important for serology interpretation. One of the limitations of serology is the inability to distinguish between antibody induced by vaccine-related poliovirus and antibody induced by wild virus. Serologic assays to detect anti-poliovirus antibodies are available in most commercial and state public health laboratories.

Cerebrospinal fluid (CSF) analysis

The cerebrospinal fluid usually contains an increased number of leukocytes—from 10 to 200 cells/mm^3 (primarily lymphocytes) and a mildly elevated protein, from 40 to 50 mg/100 ml. These findings are non-specific and may result from a variety of infectious and noninfectious conditions.

VIII. Reporting

Each state and territory has regulations or laws governing the reporting of diseases and conditions of public health importance.[22] These regulations and laws list the diseases to be reported and describe those persons or groups responsible for reporting, such as health-care providers, hospitals, laboratories, schools, daycare and childcare facilities, and other institutions. Contact your state health department for reporting requirements in your state.

Reporting to CDC

Because poliomyelitis no longer exists in the U.S. (no cases since 1999), each reported case of suspected poliomyelitis should be followed up by local and state health departments in close collaboration with CDC. At the direction of the state health department, CDC (National Immunization Program, 404-639-8255) 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

- Determine whether the disease may be caused by wild poliovirus or is vaccine-related

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 in U.S.

- 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
  - Serologic test

continued on the next page

Information to collect (con't.)

- 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
  - Contact with persons recently returning form polio-endemic areas
  - 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 non-household 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 [15] and the Amish in 1979,[1] 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. In addition, isolation of wild poliovirus from residents of Canada in 1993 [23] and 1996 [24] 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

All children should receive four doses of IPV given at 2 months, 4 months, 6–18 months, and 4–6 years of age.

All children should complete their primary vaccination for poliomyelitis before entering school. All children who had previously received a primary series with only OPV or only IPV (three doses) should receive a fourth dose of IPV

before entering school (between 4–6 years of age) to complete the recommended schedule.

If the poliovirus vaccines are administered according to their licensed indications for minimum ages and intervals between doses, administration of four doses of IPV or OPV in any combination by 4–6 years of age is considered a complete poliovirus vaccination series.[3]

The primary series of IPV for adults consists of three doses of IPV. Two doses can be given at a 4–8 week interval; the third dose should follow 6–12 months after the second dose.[3]

In circumstances where accelerated protection is needed, the minimum interval between doses of poliovirus vaccine is 4 weeks. Previously vaccinated persons who are considered to be at increased risk of exposure to poliovirus (e.g., travelers to polio-endemic areas, laboratory workers) should receive a single additional dose of IPV.[3]

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 16, "Enhancing Surveillance."

Promoting awareness

Because of the severity of poliomyelitis disease (it is a paralytic 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 the CDC, and the need to obtain stool and serum 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 and serologic testing for poliovirus.

Obtaining laboratory confirmation

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

Active surveillance

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 case finding at area hospitals or any other providers of acute medical care.

XI. Case investigation

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. At the direction of the state health department, CDC should be contacted at 404-639-8255. Timely collection of stool specimens is important in establishing the diagnosis and determining appropriate control measures, in the event of wild poliovirus isolation (see "Virus isolation" in Section VII, "Laboratory testing").

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