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THE ELIMINATION OF INDIGENOUS WILD POLIOMYELITIS IN CANADA:

A METHODOLOGY FOR DOCUMENTATION

by

ADWOA DESMA BENTSI-ENCHILL

Thesis submitted to  
the School of Graduate Studies and Research  
in partial fulfilment of the requirements for the  
M.Sc. degree in Epidemiology

University of Ottawa

October, 1993



Adwoa Desma Bentsi-Enchill, Ottawa, Canada, 1993



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## ABSTRACT

The Pan American Health Organisation (PAHO) is working towards eradicating indigenous transmission of the wild poliovirus in the Americas. Since 1977, there have been no reported cases of indigenous wild paralytic polio in Canada and the elimination of polio has been generally accepted as a fact. This thesis was conducted to establish the background for a plan to document the elimination of indigenous wild polio in Canada.

Three independent studies were conducted to address the adequacy of polio detection in Canada: (a) a critical review of the existing surveillance system for poliomyelitis in Canada; (b) a retrospective chart review of acute flaccid paralysis (AFP) cases presenting in the Ottawa-Carleton Regional Municipality during a 5-year period (1986-1991); and (c) a cross-sectional survey in Ottawa-Carleton of physicians' awareness about the presentation of poliomyelitis, and their investigative practices for AFP in general.

Polio surveillance in Canada is currently inadequate to detect all possible cases of indigenous poliovirus infection. Surveillance is largely based on passive, physician-initiated reports, a method known to be generally less effective than active surveillance, and less so for rare events. There is no general AFP surveillance and there are also no routine methods of environmental surveillance for poliovirus activity.

For the 5-year chart review period, 35 of 101 AFP cases were found to be polio-compatible. However, stool cultures were ordered for only 4 of the 35 cases; none of these had a second stool culture. Of two polio-compatible cases and two other AFP cases for whom a diagnosis of polio was considered at admission, only one was appropriately investigated with two stool cultures within two weeks of onset of paralysis. There was no definite evidence that any of the AFP cases was a true case of polio but for most of the polio-compatible cases the tests

carried out did not provide conclusive results to support the diagnosis made.

The overall conclusions from the chart review and the physician survey are that among physicians in the study region (a) the index of suspicion for paralytic poliomyelitis is very low, and (b) overall knowledge about the presentation of polio, appropriate diagnostic tests, and requirements for notification are poor, or else not adequately applied.

On the basis of the above findings, the conclusion is that Canada does not currently meet the PAHO requirements for certification of a polio-free status. In addition, there is insufficient evidence from the data obtained for the supposition of wild polio elimination in Canada. Recommendations are presented for improving polio surveillance and for establishing protocols for investigation and control of polio.

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## ACRONYMS COMMONLY USED IN TEXT

ACE	Advisory Committee on Epidemiology
AFP	Acute Flaccid Paralysis
BCDE	Bureau of Communicable Disease Epidemiology
CCDR	Canadian Communicable Disease Reports
CCDSS	Canadian Communicable Disease Surveillance System
CDC	Centre for Disease Control
CID	Childhood Immunization Division
CPS	Canadian Paediatric Society
DSD	Disease Surveillance Division
GBS	Guillain-Barré Syndrome
ICCPE	International Certification Commission on Poliomyelitis Eradication
IMPACT	Immunization Monitoring Program Active (System)
IPV	Inactivated poliovaccine
LCDC	Laboratory Centre for Disease Control
NACI	National Advisory Committee on Immunization
OPV	Oral poliovaccine (Live attenuated poliovaccine)
PAHO	Pan American Health Organisation
PESS	Polio Eradication Surveillance System
VAAESS	Vaccine-Associated Adverse Events Surveillance System
WHO	World Health Organisation

## CHAPTER 1 INTRODUCTION

In May 1985, the Pan American Health Organisation (PAHO) proposed an initiative to eradicate the indigenous transmission of wild-type poliovirus from the Region of the Americas by the end of 1990. Polio eradication was defined as "interruption of the transmission of wild poliovirus".<sup>1 p. 214</sup> By the end of 1989, 6 of the 35 countries in the American Region were still reporting cases of paralytic polio and the goal for eradication was subsequently revised to 1995.<sup>2</sup> Two years have elapsed since the last confirmed case of wild poliomyelitis in the Americas and the achievement of the eradication goal now appears to be a reality; the last case was in a 2-year old boy in Junin, Peru in August 1991 and was reported in September of the same year.<sup>3</sup> For eradication, the emphasis is on the wild or naturally occurring poliovirus as opposed to the vaccine-type virus also capable of causing disease; the different types of virus are discussed in a later section.

Following the initial successes of the PAHO Polio Eradication Program, the World Health Organisation (WHO) adopted, in 1988, a resolution for the global eradication of wild-type poliovirus transmission by the year 2000.<sup>4</sup> This goal when achieved will make poliomyelitis the second human disease eradicated from the world but the first to be eradicated under the WHO Expanded Program of Immunization (EPI) established in 1974. However, it should be noted that the EPI evolved from the activities of the Smallpox Eradication Program which provided valuable lessons for the control of communicable diseases, particularly vaccine-preventable diseases. The eradication of smallpox was achieved almost 11 years after an intensified global effort was initiated by the World Health Assembly in 1966 with the objective of eradication within 10 years. The last case of naturally occurring smallpox was recorded in Merka, Somalia in October, 1977.<sup>5</sup>

As in the case of smallpox, documentation of the eradication of poliomyelitis (regional and global) requires a systematic and active search for cases. Reliable data have to be gathered as evidence of a break in transmission of the virus. An International Certification Commission on Poliomyelitis Eradication (ICCPE) was established by PAHO in 1990 to review documentation and subsequently certify a polio-free status within its member countries.<sup>6</sup>

### **1.1 Guidelines for Documentation**

The criteria set by the ICCPE for certification of polio eradication<sup>7</sup> are paraphrased as follows:

1. Absence of virologically confirmed indigenous wild poliomyelitis cases in the Americas for a period of at least three years under circumstances of adequate surveillance;
2. Absence of detectable wild polioviruses from communities based on stool sampling from normal children, and sewage sampling from high-risk populations where appropriate;
3. On-site evaluation of pre-certification activities by national certification commissions to be set up by each country. National commissions are to determine the appropriate time to submit documents to the ICCPE for final certification;
4. Establishment of appropriate measures to deal with potential importation of cases from polio-endemic areas.

The decision for a criterion of three years as a minimum "polio-free" period was based on the frequent occurrence of inapparent infections of polio which limits its recognition. Smallpox, on the other hand, has characteristic clinical manifestations so for certification purposes two years was used as the acceptable period with no cases; in fact, it is now felt

that one year might have sufficed.<sup>5,8</sup> Currently, two research groups working separately in the United States have developed mathematical models suggesting that the probability of wild poliovirus remaining undetected in a community after 3 to 4 years with effective surveillance of acute flaccid paralysis (AFP) and no culture-confirmed polio cases is less than 5% and that the probability drops further with additional years (Debanne, Robbins et al. unpublished; Rhodes unpublished).

PAHO has outlined a strategy to guide member countries in generating documents for certification of a polio-free status. The four components of the strategy are a) effective AFP surveillance to detect, and respond rapidly to, all suspected cases of polio; b) surveillance of wild poliovirus; c) active AFP case searches in areas of poor surveillance; and d) achievement and maintenance of high immunization levels (including mop-up immunization campaigns in areas of risk such as areas where confirmed or compatible cases of wild polio are detected).<sup>7</sup>

The specific indicators required by the ICCPE<sup>7</sup> for each country's surveillance of AFP and wild poliovirus are:

1. At least 80% of all health units included in the reporting network should be reporting regularly each week.
2. The rate of AFP cases should be approximately 1.0 case reported per 100,000 population < 15 years of age.
3. At least 80% of all AFP cases reported should be investigated within 48 hours of reporting.
4. At least 80% of all AFP cases reported should have two stool specimens taken for virus culture within two weeks of paralysis onset.
5. At least 80% of all AFP cases reported should have stool investigations of at least five contacts.

The current challenge for Canada is to provide evidence of a break in the indigenous transmission of wild poliovirus. This is an achievement deemed by many to have been attained more than five years before the establishment of the PAHO goals; the last confirmed case of indigenous wild poliomyelitis in Canada was reported in 1977.<sup>9</sup> Wild virus cases that have since occurred have been secondary to virus importations. Thus, the three-year requirement for zero detection of indigenous wild polio has been greatly exceeded but it still remains to prove that there has been no possibility of missing polio cases in the existing surveillance system. This thesis examines that possibility in order to determine whether Canada currently meets the certification standards.

## **1.2 Project Objectives and Outline of Activities**

The thesis project comprised three independent studies with the overall objective of establishing the basis for a plan to document a "polio-free" status in Canada. The major issue was the adequacy of polio detection in Canada addressed through an assessment of:

1. The effectiveness of poliomyelitis surveillance as it currently exists in Canada.
2. The potential at the clinical level for missing cases of paralytic poliomyelitis among AFP cases presenting in Canada.

The three stages of the project were:

1. A critical review of the existing programs that together constitute a surveillance system for poliomyelitis in Canada. They are (a) the Canadian Communicable Disease Surveillance System, (b) the Vaccine-Associated Adverse Events Surveillance System, and (c) the Immunization Program Active Surveillance System, a paediatric hospital network for the active surveillance of post-vaccination adverse events and vaccine-preventable diseases.

2. A pilot study aimed at an objective assessment of the management of AFP cases, particularly the investigations carried out to confirm, or to rule out a possible diagnosis of paralytic poliomyelitis. The study consisted of a retrospective review of clinical charts of AFP cases presenting in the Ottawa-Carleton Regional Municipality during the five-year period, 1986-1991.
  
3. A pilot cross-sectional survey of physicians' awareness about the presentation of poliomyelitis, and their investigative practices for acute flaccid paralysis in general. Physicians were selected for this survey from the Ottawa-Carleton Regional Municipality.

The findings from these activities served as the basis for proposing recommendations for (a) an improved system of polio surveillance and (b) a contingency plan for the investigation and control of suspected cases of poliomyelitis in Canada.

### **1.3 Background Literature**

#### **1.3.1 Variants of Poliomyelitis**

The poliovirus is included in the *Enterovirus* genus with the Coxsackieviruses A and B, the ECHO (*enteric cytopathic human orphan*) viruses, and the human enteroviruses 68-72. Man is the only known natural host of the enteroviruses although monkeys and chimpanzees may be infected, most readily, by direct inoculation into the brain or spinal cord.<sup>10,11</sup> Virus transmission is mainly by oro-faecal spread and less commonly by aerosol spread<sup>10-12</sup> and can be prevented by hygienic measures. Prevention of infection is by active immunization; there are two major types of vaccines, the inactivated poliovaccine (IPV) and the live attenuated oral poliovaccine (OPV). As with other enteroviruses, immunity to the poliovirus

is type-specific and long-lasting. Protection against reinfection is mediated through neutralising antibodies detectable shortly after infection or immunization.<sup>11</sup>

There are three antigenic strains or serotypes of the poliovirus; Brunhilde, Lansing, and Leon better known as types 1, 2 and 3 respectively. These serotypes share some common antigens but there are also marked intertypic differences. As well, even within a single serotype, antigenic differences may occur between different isolates due to mutations. Although the pattern of diseases is the same, the type 1 virus is recognised as being more virulent and is reportedly responsible for approximately 85% of all polio illnesses while types 2 and 3 cause only 15% of cases.<sup>13</sup> Type 2 is reported as the least neurovirulent both as wild and attenuated virus.<sup>12</sup>

Infection of a susceptible individual by the poliovirus results in one of four recognised clinical outcomes: (a) an inapparent or subclinical infection; (b) a mild febrile illness or abortive polio, the commonest; (c) aseptic meningitis or nonparalytic polio; and (d) paralytic polio, the most severe form in which the destruction of motor neurons in the spinal cord (and rarely involvement of the brainstem) results in flaccid paralysis.<sup>12,14,15</sup> Throughout this text, poliomyelitis or polio refers to paralytic poliomyelitis unless otherwise stated.

Other enteroviruses are also known to be associated with poliomyelitis-like disease, including paralysis, notably Coxsackievirus A7,<sup>14</sup> enterovirus 71,<sup>10,11,16</sup> and Echovirus 22.<sup>17</sup> As greater control is gained over the transmission of wild polioviruses more emphasis can be placed on investigations into the role of other nonpolio enteroviruses in causing paralytic illnesses.

In addition to naturally occurring polio, disease can also be caused by the virus strain used in the live attenuated oral poliovaccine (OPV) which has the rare capacity to revert to

neurovirulence.<sup>18,19</sup> The risk of OPV-associated disease has been estimated from separate studies by the WHO and the US Centres for Disease Control (CDC), as less than one case per million doses of vaccine distributed.<sup>19,20</sup> It has been recognised that the risk of vaccine-associated polio differs between vaccine recipients and their close contacts to whom the vaccine-type virus may be transmitted. The risk also differs by the order of a particular dose in the series of immunizations.<sup>19</sup> In Canada, risk estimates based on data from 1965 to 1992 are 1 recipient case per 11.8 million doses and 1 contact case per 3.1 million doses (see Table 1). The overall risk estimate is 1 case per 2.4 million doses. These estimates are based on an assumption of maximum risk (i.e., every vaccine-associated case is indeed caused by the vaccine, although that is not always proven).

Table 1

OPV-associated paralytic poliomyelitis in Canada, 1965 - 1992

	Number of cases	Vaccine doses distributed (millions)	Million doses distributed per case
<u>Vaccine recipients</u>			
1965-1969	1	9.0	9.0
1970-1979	2	14.5	7.3
1980-1989	1	16.8	16.8
1990-1992	0	6.7	-
Total	4	47.0	11.8
<u>Contacts of recipients</u>			
1965-1969	1	9.0	9.0
1970-1979	7	14.5	2.1
1980-1989	6	16.8	2.8
1990-1992	1	6.7	6.7
Total	15	47.0	3.1

For the period from 1980 to 1989, a risk of 1 case per 6.8 million doses was reported for OPV recipients in the United States. However, a recent analysis of the PAHO Polio Eradication Surveillance System (PESS) database reports an estimate of 1 case per 1.5 million doses in OPV recipients,<sup>21</sup> a risk that is considerably higher than the estimates for Canada and the US. The methods of risk estimation in the different studies cited were not compared for this thesis and differences in estimates are not accounted for. Risk estimates reported by dose in the United States for 1973 through 1984 range from 1 case in 520,000 doses for the first dose in both recipients and contacts to 1 case in 12.3 million subsequent doses.<sup>22</sup>

The higher risk in contacts may be due in part to the fact that viruses transmitted by an immunized person are often mutants and therefore may not be considered as safety-tested strains. As a result of the relatively higher rate of reversion to neurovirulence associated with such mutants the herd immunity effect of OPV, whereby attenuated poliovirus is spread to household and community contacts of vaccinees, is described as a disadvantage of the vaccine.<sup>18</sup> Some people however argue for herd immunity as one of the benefits of OPV; in areas of very low immunization coverage the latter may indeed be the case. The susceptibility of contacts to infection is influenced by both individual immunization status, as well as the overall immunization coverage and seroconversion levels in the population. It is important to verify the immunization status of parents and close family contacts prior to immunizing children with OPV. Persons with immunodeficient states have a particularly high susceptibility to severe disease following immunization with the live, attenuated poliovaccine. In the US, children less than one year of age with a primary immunodeficiency were reported to have a risk of OPV-associated polio 2000 times higher than children with non-deficient immunity.<sup>23</sup>

### 1.3.2 Monitoring of Poliovirus Activity

It is estimated that only about 0.1% to 1% of naturally occurring or wild poliovirus infections result in paralysis.<sup>14,15</sup> However, because of the non-specific features of the other manifestations of infection, paralysis offers the highest index of suspicion to trigger early investigation. Thus, paralytic polio is a useful clinical marker of wild poliovirus activity in a community.

Another traditional method for monitoring poliovirus activity consists of serosurveys for neutralizing antibodies to the poliovirus. The role of serodiagnosis in the polio eradication program is limited by the inability to differentiate between antibodies to the wild poliovirus and those formed in response to the OPV strain of the virus. Nonetheless, serosurveys are an important method of identifying individuals with a current poliovirus infection, or who have had a recent poliovirus infection, and of evaluating an individual's immunological response to immunization.

Environmental sampling (of sewage or surface water) and population stool surveys may also be used in monitoring poliovirus activity. Environmental and population surveys were commonly employed in Canada in the 1950s and 1960s but not in the recent routine surveillance of poliomyelitis. Nonetheless, a number of virological studies in Canada have reported the recovery of wild and vaccine strains of the poliovirus, from sewage and surface waters.<sup>24-27</sup> The inability of the vaccine strain of poliovirus type 1 to survive in conventionally treated drinking water has been documented.<sup>28</sup>

Ideally, clinical specimens for laboratory detection of the poliovirus should be obtained at the onset of symptoms or shortly thereafter. The best single specimen for viral isolation or detection is stool because of the persistence of virus in the gut for about six weeks.<sup>11</sup> Rectal

and pharyngeal swabs are also a good source of virus detection<sup>11</sup> but cerebrospinal fluid (CSF) is less reliable.<sup>10</sup>

Viral isolation is carried out in cell cultures of a variety of human and monkey tissues including human fetal diploid lung and kidney cells, primary monkey and Buffalo green monkey kidney cells, HeLa cells and rhabdomyosarcoma cells. Identification based on the characteristic cytopathogenic effects of infected cells is usually presumptive only. Definitive identification may be achieved by the standard method of neutralisation with type-specific antiserum, or by other methods such as complement fixation, hemagglutination inhibition, enzyme-linked immunoassay, and immunofluorescence.<sup>11</sup>

The virus may also be detected directly by assays for viral nucleic acids. Nucleic acid technology is based on the fact that unique regions of nucleic acid (NA) can be identified in most viruses. By using nucleic acid probes (enzyme-labelled or radio-labelled NA sequences) that are complementary to the known NA sequence of a virus, the latter can be detected in a clinical specimen through a process of hybridization with the probe. Although nucleic acid hybridization was developed in the 1960s, the sensitivity of the associated detection methods was lower than that of viral culture and the latter remained the method of preference for the next two decades. Since the 1980s, newer techniques in molecular biology known generically as amplification techniques have enhanced direct virus detection.<sup>29</sup>

Amplification techniques are methods by which specific nucleic acid sequences of a virus can be synthesised in vitro. This consists of creating a base-pair between a short piece of deoxyribonucleic acid, the primer, and the sequence of viral NA to be replicated, the template. The primer-template base-pair is then used as a starting point for replication through the action of the enzyme DNA polymerase. Through amplification, the number of NA molecules available in a specimen to generate signals for detection by routine methods

is greatly increased. Thus, although the actual sensitivity of the detection methods is not changed, the overall sensitivity of viral detection is highly improved and there is also an enhanced ability to distinguish between wild virus and vaccine virus strains.<sup>29,30</sup>

The polymerase chain reaction (PCR) introduced in 1985, is described as "the most promising method [and] perhaps the most sensitive method available" for the direct detection of enteroviruses.<sup>11 p. 362</sup> Among the many advantages of NA technology is the fact that both noninfectious and viable (or infectious) viral particles are detectable whereas viral culture is able to detect only infectious particles. Primers and probes can also be designed to be type-specific or to detect multiple enteroviral serotypes in one test.<sup>30</sup> Subtypes or specific viral genetic markers have been detected and reported with important diagnostic and epidemiological implications.

### **1.3.3 Disease Eradication versus Elimination**

According to Stuart-Harris, "Eradication of an infection implies that the infection has disappeared from all countries of the world because transmission of the causative organism has ceased in an irreversible manner."<sup>31 p. 913</sup> This definition should be qualified by the exception of reference stocks of infectious disease agents held in selected countries for research purposes.

Yekutieli, expanding on an earlier definition by Andrews & Langmuir, defines eradication as "The purposeful reduction in the prevalence of a specific disease to the point of continued absence of transmission within a specified area by means of a time-limited campaign."<sup>5 p. 466</sup> The latter definition may be compared with Stuart-Harris's use of the term elimination (or regional eradication) to indicate "the disappearance of transmission from a small or large area, with a country or a continent ultimately becoming free from infection".<sup>31 p. 913</sup> The

WHO definition of elimination adds to this the "prevention of all disease, despite the continued presence of the causative organism and the continued need to apply preventive measures".<sup>32 p. 89</sup>

I agree with the view that eradication means no control measures are required for a particular disease in any geographical area as is currently the case for smallpox. It should be noted that PAHO's Eradication Program refers to regional eradication only. In an individual country, it is even more appropriate to speak about the elimination of polio thus, I use the term elimination when discussing the polio situation in Canada while eradication is retained for the PAHO program.

#### **1.3.4 Factors Influencing Polio Eradication**

Several biologic, socioeconomic, and geographic factors play a role in the control of communicable diseases. Evans describes three biologic levels of communicable disease control: (a) control of the clinical disease and its associated morbidity, disability, and mortality; (b) control of the infection whether clinical or asymptomatic; and (c) control of the causative organism in the environment and its transmission through it.<sup>33</sup> He further states the geographical levels of control as local, regional, country-wide, continent-wide and global.

Biological factors that contribute positively to the interruption of wild poliovirus transmission include a) the brief period (a few weeks) of virus excretion by infected persons, b) the absence of a natural animal reservoir for the virus, and (c) the availability of relatively safe and effective vaccines. Socioeconomic factors such as the political will of a government, and the availability and allocation of financial and technical resources are also important. In discussing the Smallpox Eradication Program, Fenner notes that "a very important lesson for any future global eradication effort is that in addition to their administrative competence, [the

key persons in the program] spent a great deal of time in the field".<sup>8 p. 918</sup> The commitment of health care workers at all levels of an eradication program, and their perception of the importance of the disease are vital to the success of the program.

### **1.3.5 Progress Towards Eradication**

#### **1.3.5.1 The Global and Regional Situation**

In 1990, the WHO estimated that 150,000 cases of paralytic polio were occurring annually in 70 countries where the disease was still endemic.<sup>34</sup> Excluding OPV-associated cases, 12,917 cases of paralytic polio were reported by the six regions of the WHO in 1991<sup>35</sup> (Table 2). These figures are incomplete as several countries, particularly in the African Region, did not provide data to the WHO; in 1992, the WHO estimated that the worldwide reporting efficiency for poliomyelitis was approximately 10% only.<sup>3</sup>

Table 2

Paralytic polio cases reported to the WHO, 1991

Reporting region	Number of cases
African	1562
American	9
Eastern Mediterranean	2032
European	309
South-East Asian	6404
Western Pacific	2601
<b>Total</b>	<b>12917</b>

Currently, poliomyelitis is largely a disease of non-industrialised countries. In the industrialised countries (including the United States, Canada, Australia, New Zealand and 23 European countries) a dramatic reduction in reported cases was noted from 1955 (76,000 cases) to 1967 (1,013 cases).<sup>18</sup> This achievement was attributed mainly to the use of poliovirus vaccines and improved hygiene.

Polio incidence in the American Region in the 35-year period from 1951 to 1985 has been extensively described by Melnick;<sup>18</sup> 27 countries in which polio was considered to be controlled by 1980 were compared with the 14 remaining countries in which polio was incompletely controlled. Countries with control of polio by 1980 had achieved a significant reduction in incidence subsequent to the introduction of the inactivated poliovaccine in 1955, and the live oral vaccine in 1960. The average annual number of cases was 44,391 in 1951-1955 compared to 470 in 1966-1970, and 34 in 1976-1980. However, this group of countries experienced an increase in incidence of more than eight-fold from 1981 (8 cases) to 1982 (79 cases), an increase mostly limited to one country; in 1982, Jamaica which had reported virtually no cases of polio since 1966, and none since 1975, experienced an outbreak that accounted for 73% of the 79 cases reported.

For most of the period under review however, more than 95% of the cases occurred in the second group of countries, with incomplete control of polio, representing approximately half of the region's population. The six-fold increase in incidence experienced by this group of countries from an annual average of 2,202 cases in 1951-1955, to 14,453 case in 1966-1970 was attributed to the epidemic phase of the disease. Another possibility, which is not clearly defined, is that the increase in incidence may be partly attributed to changes in reporting practices. As these are mostly developing countries, it is reasonable to assume that a number of changes in their health care systems would have occurred during the period under review

with the possible result of inconsistent reporting requirements. From 1981 to 1985, the average annual number of cases was 937.

Since the initiation of the PAHO program in 1985, there has been a steady decline in the incidence of polio in the American Region despite an increase in AFP surveillance. In 1989, 130 cases of wild poliomyelitis were reported in the Americas while only 18 and 9 confirmed cases of wild poliomyelitis were reported in 1990 and 1991 respectively.<sup>6</sup> As at mid-1993, no case had been reported in the American Region since the last confirmed case of wild poliomyelitis was reported in September 1991 in Junin, Peru.<sup>3</sup>

#### **1.3.5.2 Trends in the Control of Poliomyelitis in Canada**

Available data<sup>9,36,37</sup> indicate 15,499 reported cases of paralytic polio in Canada from 1949 to 1992; approximately 70% occurred in the pre-vaccine period, 1949-54. Two major epidemics have been reported in this half of the century. The first peaked at 28.3 cases per 100,000 population in 1953 and ended with the licensing of the IPV in Canada in 1955 (see Figure 1). Inadequate vaccine coverage in 1958 and 1959 was a major contributing factor to the second epidemic which peaked at 10.7 cases per 100,000 population in 1959. The introduction of OPV through clinical trials in four provinces in 1960 and 1961, followed by licensure in 1962 led to a major decline in incidence from 1,887 cases in 1959 to 19 cases in 1964.

From 1965 to 1992, the number of paralytic cases ranged from 0 to 9 annually with a mean of 2 cases and a total of 54 cases (see Figure 2 and Table 3); 35 (65%) of these were attributed to wild poliovirus infection on the basis of epidemiological and laboratory data. Fourteen of the wild virus cases are known to have resulted from virus importation but the rest are only assumed to be indigenous or endemic. The last case attributed to a wild

poliovirus occurred in 1988 but was an import-related case; the last confirmed case of indigenous wild poliomyelitis was reported in 1977.<sup>9</sup>

Nine cases of wild polio in 1978 and two in 1979 occurred in well-defined religious communities in Canada that do not accept immunizations; cases were reported from Alberta, British Columbia, and Ontario. These cases followed an outbreak of polio in the Netherlands among similarly non-immunized religious groups with which the Canadian groups had close contact. The most recent case of paralytic poliomyelitis was reported in a 31-year old female in March, 1992 in Quebec. On investigation this case was classified as a vaccine-associated contact case with poliovirus type 2 infection (the patient's seven-month old son received a first dose of OPV 22 days prior to the onset of her symptoms).<sup>38</sup>

Table 3

Classification of paralytic poliomyelitis in Canada by virus type, 1965-1992

	Number of cases (% for subgroup)
<b>Wild virus cases:</b>	
Endemic	21 (60.0)
Imported	0 ( 0.0)
Import-related	14 (40.0)
<b>Total</b>	<b>35</b>
<b>Vaccine-associated cases:</b>	
Recipients	4 (21.1)
Contacts	15 (78.9)
<b>Total</b>	<b>19</b>

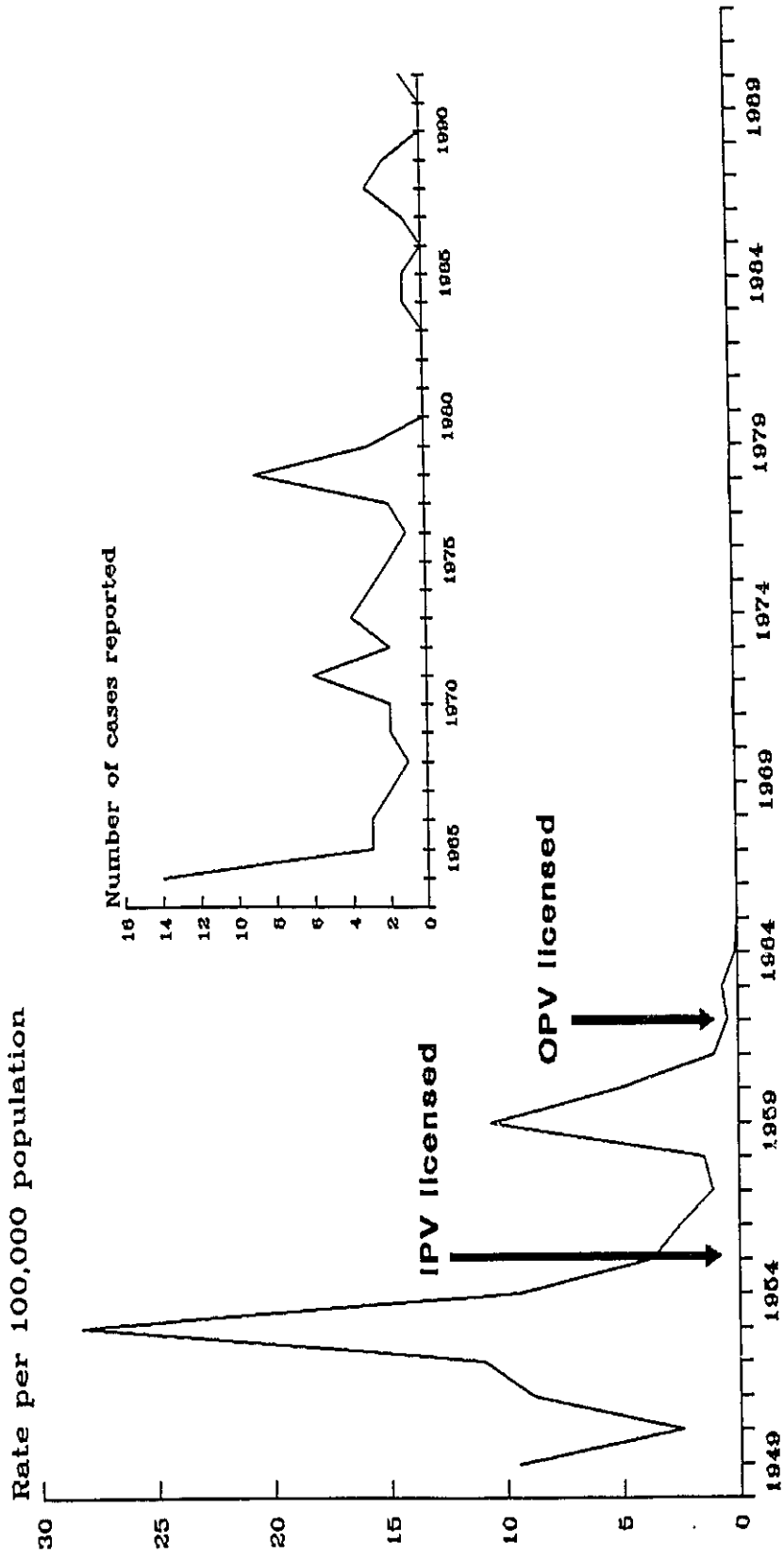


Figure 1 Reporting rate of paralytic polio in Canada, 1949 - 1992. The mini-chart enhances the number of cases reported in 1965 - 1992.

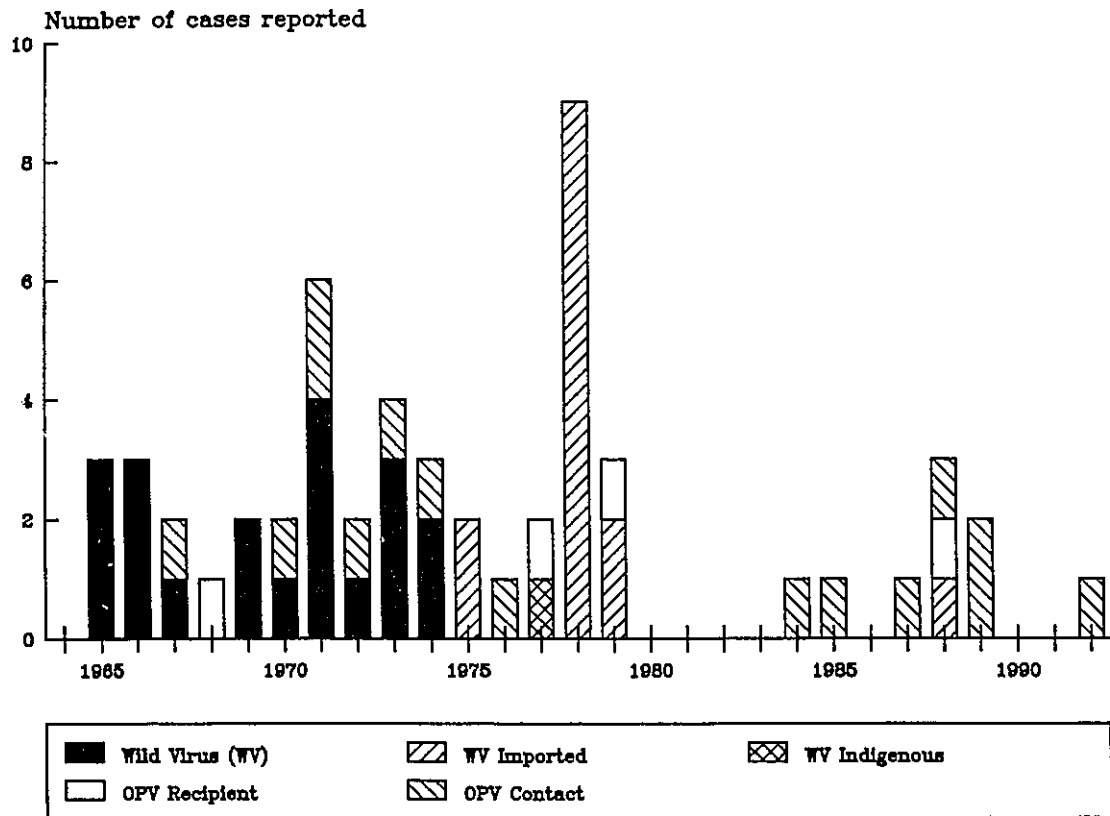


Figure 2 Reported cases of paralytic polio in Canada according to virus type, 1965 - 1992. Prior to 1975, wild virus cases were not differentiated as indigenous or imported.

In spite of the interruption of wild virus transmission in most areas of the Western Hemisphere, there remains a risk of importation of the wild virus by travellers to, and from, other countries where poliomyelitis is still endemic. In Canada, importation of wild poliovirus from Netherlands led to the 1978 outbreak previously mentioned; investigations showed a direct link through travel, and provided laboratory evidence that the virus strain in the two outbreaks was the same.<sup>39,40</sup> From September 1992 to February 1993, the Netherlands experienced another polio outbreak in the same non-immunised religious communities; 68 cases had been reported by the end of the outbreak.<sup>41</sup> Recent investigations conducted in the communities in southern Alberta affected by the 1978 outbreak confirmed a wild virus importation.<sup>42,43</sup> With PCR technology, analysis of the type 3 virus isolated (in 21 of 45 stool samples) showed a close relation to the virus strain identified in the Netherlands outbreak with some intertypic variation.<sup>43</sup> Similar investigations in British Columbia did not detect any wild poliovirus and as at August 1993 no clinical cases had been reported in Canada. The timeliness and accuracy with which this importation was detected is a good example of the implications of PCR technology for the Polio Eradication Program.

Approximately 35% of the cases reported from 1965 to 1992 were below 15 years of age while more than half were 20 years or older (see Figure 3). Both wild and vaccine-associated cases occurred in children as well as adults, however all the recipient cases occurred in children below the age of one year. A comparison of age-specific incidence from 1958 to 1992 shows that in the first decade polio cases occurred almost exclusively in persons below the age of 19 years with a peak in the 1 to 4-year age-group (Figure 4). On the contrary, the latter decades show an upward shift in the age groups more frequently affected by polio; peak age-incidences occurred in early adulthood (20 to 39 years). This shift in age distribution is congruent with what is known about changes in epidemiology as countries progress from a polio-endemic to a non-endemic state.<sup>10</sup>

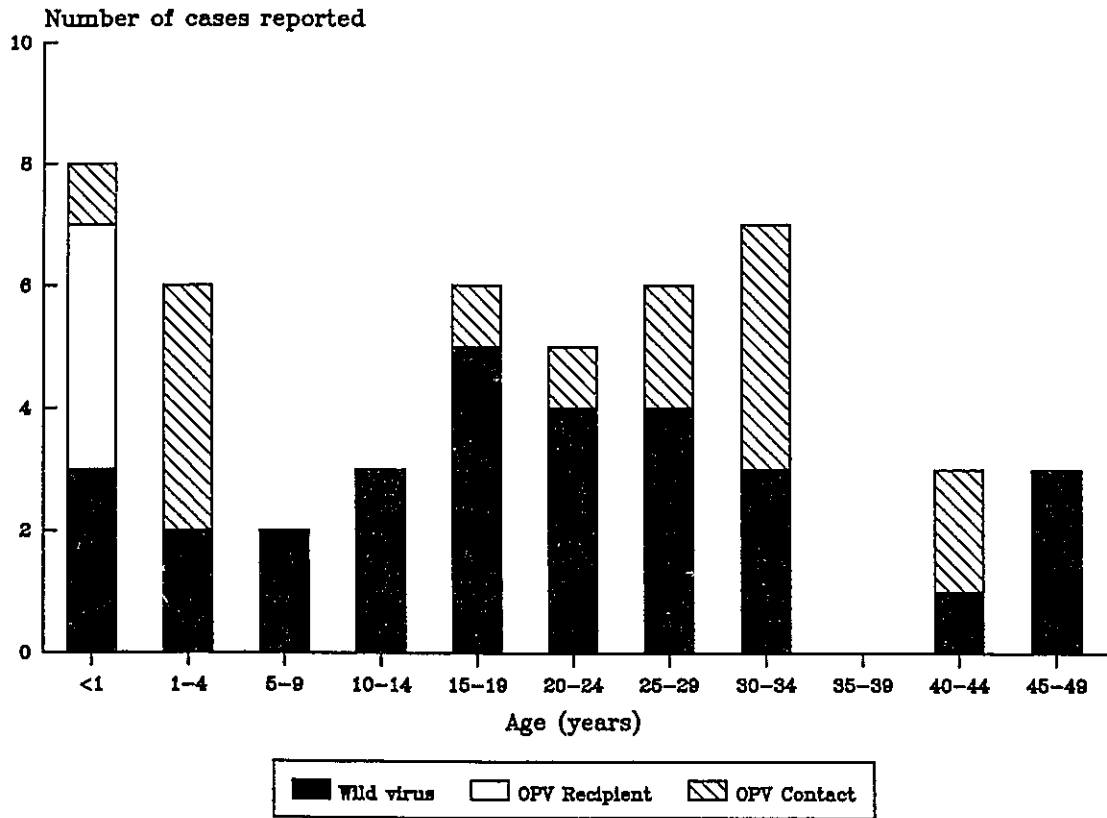


Figure 3 Age distribution of paralytic polio in Canada, 1965 - 1992.  
 Wild virus cases are not differentiated as indigenous or imported cases.

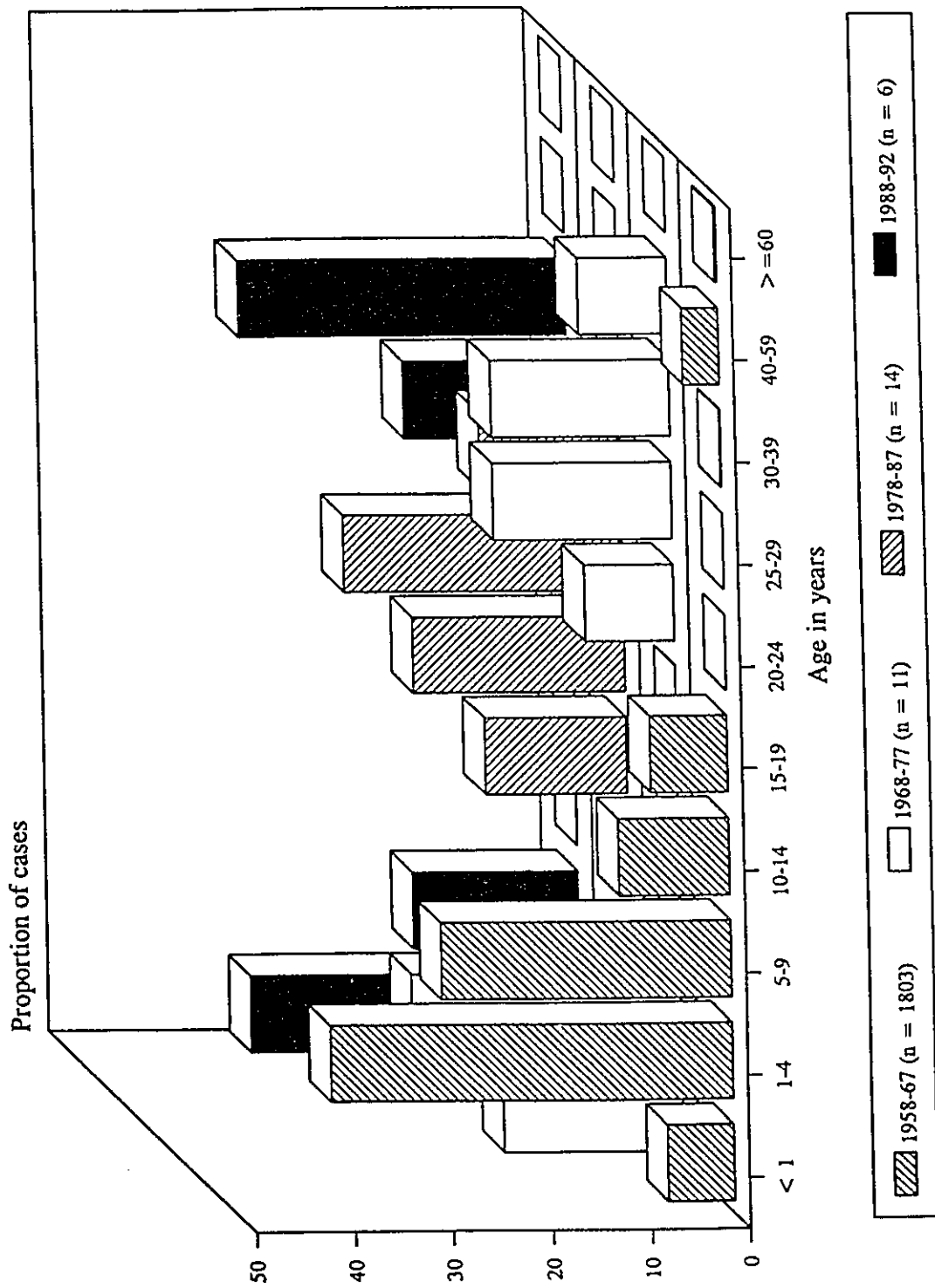


Figure 4 Trends in age distribution of paralytic polio in Canada, 1958 - 1992.

Currently, both IPV and OPV are licensed for use in Canada; recommendations for their use are prepared by the National Advisory Committee on Immunization (NACI)<sup>44</sup> although each provincial or territorial jurisdiction makes an individual decision regarding the particular product to use. Nova Scotia, Newfoundland, and Ontario (representing approximately 42.2% of the Canadian population) use the IPV for routine childhood immunizations at 2, 4, 6 and 18 months with a booster dose at 4-6 years. (Ontario switched from IPV to OPV from the end of 1989 to April 1993 for reasons of unavailability.) The other jurisdictions use OPV at 2, 4, and 18 months followed by a booster at 4-6 years. IPV confers immunity to all three poliovirus serotypes in about 99% of fully immunized persons for at least 10 years while OPV confers long-lasting immunity to all three serotypes in more than 95% of fully immunized persons.<sup>44</sup>

Although there has been no complete evaluation of national immunization coverage, available data indicate that more than 90% of school-aged children (more than 95% in some jurisdictions) are fully immunized against polio (LCDC unpublished data). In addition, more than 80% of children reportedly receive at least three poliovaccine doses by the age of 2 years. In the 1970s, a number of Canadian serosurveys showed variations in antibody status by age and type of virus with mean levels of seroconversion ranging between 82%-97%. The lowest levels of antibody were found for the type 3 virus (79%-86%).<sup>45,46</sup> Analysis by age in a separate study showed the lowest level (65%) in the 4- to 6-year age group while the highest level (97%) was found in the 15- to 17-year age group.<sup>47</sup> In 1991, a survey of 367 military recruits, whose ages ranged from 18 to 21 years, indicated seroconversion levels of 95.4%, 99.7%, and 94.8% to the type 1, 2 and 3 viruses respectively (LCDC unpublished data).

#### 1.4 Relevance of the Study

Mulhern describes three stages of disease eradication based on examples from animal disease eradication. These are (a) the "panic stage" when the attitude of major stakeholders is to do something, (b) the "cooperative stage" when these stakeholders become convinced that something can be done and cooperate willingly, and (c) the "apathetic stage.... the most difficult stage of the eradication program",<sup>8 p. 927</sup> which sets in when infection has been drastically reduced. Following the marked decline in the incidence of paralytic polio, the clinical significance of poliomyelitis in Canada and the United States is very low. Unlike most of the other countries in the American Region which have implemented intensified activities towards polio eradication, these two countries may now be described as being in the apathetic stage.

It is of high public health importance to establish and document the elimination of indigenous poliovirus in Canada. Although Canada has been described (quite reasonably) as polio-free for more than a decade, there has been no detailed documentation of the actual extent of wild poliovirus activity in the country. First of all, continued surveillance for imported cases of poliomyelitis is of importance because of the potential for re-emergence of wild poliovirus transmission. Secondly, the proper investigation of cases and their correct classification will avoid a risk of cases of wild poliomyelitis being misclassified as vaccine-associated. In turn, a perception of a high risk of vaccine-associated polio would have the undesirable effect of reducing public acceptance for the oral poliovaccine with a possible increase in the number of susceptible persons. Thirdly, the certification of polio eradication in the American Region hinges on the efforts of each of its member countries in documenting a polio-free status. Finally, the potential impact of a successful regional eradication program on the WHO global eradication effort cannot be overlooked.

## CHAPTER 2      A REVIEW OF POLIO SURVEILLANCE IN CANADA

### 2.1      Public Health Surveillance

Public health surveillance has been defined as "the continued watchfulness over the distribution and trends of incidence through the systematic collection, consolidation and evaluation of morbidity and mortality reports and other relevant data [and the regular dissemination of data to] all who need to know".<sup>48 pp. 182-3</sup> More recent definitions<sup>49,50</sup> also emphasize its major components as (a) systematic collection of data, (b) analysis of data, (c) the dissemination of data-based information to appropriate persons, and (d) application of these data to the prevention and control of disease.

The applications of surveillance data vary, the relative importance of each depending on the disease under surveillance. For a disease that is essentially under control the most relevant uses of surveillance are (a) to describe the background levels of disease and changes in disease occurrence, (b) to help establish public health interventions for disease control, (c) to evaluate the effectiveness of control programs, (d) to help establish or review priorities for health resources, and (e) to forecast future patterns of disease occurrence or non-occurrence (i.e., elimination or eradication).

Currently, in the Western Hemisphere there are two major goals of polio surveillance: (a) to provide evidence for a break in the indigenous transmission of the wild poliovirus, and (b) to provide the means for the early detection of imported cases and the timely institution of control measures.

### 2.1.1 Surveillance Methods

The methods selected for a surveillance system should be based on the disease under surveillance, and the goal of the program. Generally, the preferred method is one that provides the most accurate information, given the available resources, in a practical and efficient manner. Surveillance methods are broadly classified as passive or active. Passive or spontaneous case reporting is initiated by the reporting source, usually a health care provider or a diagnostic laboratory. An active method, on the other hand, is based on reports sought from these sources at regular intervals by public health authorities.<sup>50,51</sup> Surveillance may also be general or universal when it involves all members of the public health system, or it may be based on a sentinel approach in which a sample of reporting sources are used.

Generally, evaluations of active and passive reporting systems have shown increased reporting with the active approach by figures ranging from 2.0 to 4.6 times the rate of passive reporting.<sup>52</sup> An even higher margin of difference was reported by Halperin et al<sup>53</sup> who found a 14-fold increase in the reported incidence of pertussis during a period of active surveillance in certain areas of Nova Scotia, as compared to other parts of the province where passive surveillance was being used. These authors also reported an 11-fold increase in establishing a diagnosis of pertussis during a period of enhanced laboratory diagnosis.

Active surveillance is generally more expensive and covers smaller populations, therefore the chances of missing rare illnesses are increased. A combined program of active and passive surveillance may be used to achieve greater cost-effectiveness. Furthermore, the higher expense associated with active surveillance can be reduced by limiting surveillance to sentinel sites. It should be recognised that although a sentinel system is relatively inexpensive, it may result in biased information if the target population does not accurately represent the general

community. For a rare disease it is appropriate to define the target population as the high risk population so that sentinel sites need only be chosen to represent that risk category.

### **2.1.2 Attributes of a Surveillance System**

Routine public health surveillance of endemic diseases aims at collecting data on a representative sample of a well-defined target population. Thus, it is not necessary to report every case occurring in the population. As disease incidence decreases however, surveillance tends toward more individual case investigation and the completeness of data becomes essential to the implementation, and evaluation, of prevention and control programs. In an eradication program it is essential that no cases of disease are missed (that is there are no false negative cases) therefore, a high level of sensitivity is desired where the sensitivity of the system is measured as its ability to detect all true cases of the disease under surveillance. Often measures to achieve a high sensitivity mean sacrificing specificity (i.e., more and more cases that are reported and investigated turn out not to have the disease). The loss in terms of resources used to investigate each suspected case or outbreak may, however, be justified by the need to detect all cases.

Other attributes of a surveillance system that need to be considered include the flexibility of the system (meaning its capability to adjust to changes in information needs or operating conditions), the acceptability of the system to participating members, and the timeliness of data collection and transfer at all steps of the reporting chain. These attributes, in addition to the simplicity, representativeness, and usefulness of the system are discussed in subsection 2.7.

### 2.1.3 The Importance of Case Definitions in Surveillance

The importance of disease classification in surveillance is aptly described by the observation that "nomenclature is of as much importance in this department of inquiry as weights and measures in the physical sciences and should be settled without delay", a statement attributed to William Farr, considered the founder of modern surveillance.<sup>54 p. 13</sup>

A uniform case definition in surveillance ensures comparability of data from different sources; surveillance data are usually aggregated for the analyses of such epidemiologic parameters as disease incidence and trends. Therefore, it is important to guard against potential errors from the combination of incomparable data. Case definitions should be well publicized and accessible to all persons participating in a surveillance system. In public health surveillance, uncomplicated case definitions tend to be more acceptable to program participants<sup>50,52</sup> although such definitions may not be appropriate to clinical diagnoses, and may be difficult for physicians to accept because they oversimplify the clinical diagnostic process.<sup>52</sup>

Case definitions vary with the goals of a particular program and the disease incidence, as well as the distinctiveness of the disease from other diseases. A general case definition may be used when disease incidence is relatively high or at the beginning of an epidemic investigation. Such a definition may lead to the inclusion of many non-cases because of a low specificity but it can always be modified to be more precise, and to improve specificity as a greater level of disease control is achieved. Such changes in case definitions should always be borne in mind when examining trend data. In the final stages of eradication too, a specific case definition can be maintained for confirmed cases while a more sensitive and general case definition is re-introduced for the purposes of screening for suspected cases. Such is the case with the current PAHO recommendation for AFP reporting as part of polio surveillance.

With general surveillance, laboratory investigations that form part of the case definition must be readily available, inexpensive, and pose limited demands on the patient. These features may become less important with a rare disease when the cost of investigations is outweighed by the need to confirm each true case in the target population. Clearly, the latter should not preclude the need for proper judgement in the initial investigation of suspected cases, and the need to monitor the indiscriminate, and sometimes unnecessary, utilization of expensive investigative procedures. It is important to determine whether any case of wild polio occurring in Canada now represents an importation or indigenous transmission of the poliovirus as this forms a central part of the elimination effort. Investigations should therefore be aimed at obtaining all relevant information to distinguish between these two possibilities.

## **2.2 The Objectives and Process of the Review**

I conducted a critical review of the current surveillance system for poliomyelitis in Canada with the overall objective of assessing the effectiveness of the system.

The questions raised by the objective were:

1. Is there an effective system to detect all possible outbreaks of poliovirus infection and all cases of paralytic polio?
2. Are there effective control measures for suspected cases?

The sources of information in this review included a) published and unpublished literature, b) a brief survey of Provincial and Territorial Epidemiologists to obtain information on surveillance practices within their respective jurisdictions, and c) interviews with other key persons involved in the planning and implementation of the surveillance programs.

I focused on the surveillance of all forms of poliovirus infection in Canada but also included the surveillance of other specific syndromes of acute flaccid paralysis which have major presenting features similar to those of paralytic poliomyelitis. The surveillance of these other diseases (including Guillain Barré Syndrome (GBS), acute transverse myelitis, other nonpolio enterovirus-related paralyzes, and botulism) was reviewed to assess the potential for detecting missed cases of polio.

I did not include a review of the resources used to operate the surveillance system although that would usually be part of the evaluation of surveillance. A review of resources consists of both direct operating costs for the system (e.g., personnel requirements and administrative costs) as well as indirect costs related to actions based on surveillance data (e.g., immunization or health promotion campaigns). These costs have to be evaluated vis-a-vis the benefits accrued from surveillance and the efficiency of meeting surveillance goals. I considered such analyses to be beyond the scope of this thesis. However, the intention is not to overlook the importance of cost-benefit and cost-efficiency issues in the evaluation of a surveillance system.

### **2.3 The Evolution of Polio Surveillance in Canada**

Poliomyelitis has been a nationally notifiable disease in Canada since 1924.<sup>9,55</sup> Prior to 1949 when tissue culture technology for specific laboratory confirmation was developed, notifications of polio may have included other polio-like illnesses and also, all clinical forms of poliovirus infection were reported as a single entity.<sup>9</sup> In 1949, the Dominion Council of Health recommended separate reporting for paralytic and nonparalytic polio. Since 1958, only paralytic polio has been nationally notifiable; nonparalytic polio is only reportable as viral meningitis or encephalitis with no differentiation from other viral causes.<sup>55</sup>

Surveillance is often justified by the public health importance of a disease. For endemic diseases this is measurable as the burden of morbidity and mortality through indicators like incidence, prevalence, and indices of severity (e.g., case-fatality ratio or hospitalization rate). Although polio typically results in residual paralysis and other neurological problems, these measures of importance are currently not appropriate to polio in Canada because of its extreme rarity. On the other hand, the public health importance of polio may be expressed in terms of a) its preventability, b) the potential for its re-emergence and the associated health risks, and c) the need for documentation leading to certification of a polio-free status.

In 1987, the national Advisory Committee on Epidemiology (ACE) conducted a review of communicable disease priorities for national surveillance.<sup>56</sup> The criteria used were selected for their importance from the national perspective and included among others, WHO interest, incidence, morbidity, potential for outbreaks, socioeconomic impact, and vaccine preventability. Each disease was awarded points on a five-point scale for each of 12 criteria and the total score used as a measure of disease priority, higher scores indicating higher priority. Paralytic polio was ranked fourteenth with 30 points in a range of 8 to 42 points for the 60 diseases reviewed. This indicates the considerable public health importance that paralytic polio held for the committee and there is no evidence of a change in that assessment.

The current surveillance of poliomyelitis in Canada is based on three programs operating independently within broader systems of disease surveillance. They are (a) the Canadian Communicable Disease Surveillance System (CCDSS), (b) the Vaccine-Associated Adverse Events Surveillance System (VAAESS), and (c) the Immunization Monitoring Program Active System (IMPACT), a paediatric hospital-based system for the active surveillance of post-vaccination adverse events and vaccine-preventable diseases. In addition, supplementary data for the laboratory detection of polioviruses (submitted by all Public Health Laboratories)

are available through the Canadian Virus Reporting Program of the Laboratory for Surveillance, Influenza and Viral Exanthemata, Bureau of Microbiology, LCDC.

## **2.4 Overview of the Surveillance Programs**

The three programs are at different stages of development and have varying scopes of operation; poliomyelitis is only one of several diseases/health events reportable under each of these programs. In the following sections the CCDSS is often presented as the basic model because it is the longest established and most comprehensive of the three programs. Also the case definitions and reporting mechanisms used in the other two programs do not differ significantly from those of the CCDSS.

### **2.4.1 The Canadian Communicable Disease Surveillance System**

The CCDSS is administered at the federal level by the Disease Surveillance Division (DSD) of the Bureau of Communicable Disease Epidemiology (BCDE), LCDC. The system is described as a "product of close cooperation between the [LCDC] ... and the communicable disease control epidemiologists in each province and territory".<sup>57, preface</sup> This cooperation is mediated through the ACE which serves as a forum for the provincial and territorial epidemiologists to advise federal authorities on matters related to disease control.

The responsibility for receiving and analyzing notifiable diseases data at the federal level was transferred from the Epidemiology Section of Statistics Canada to the BCDE in 1988; prior to that Statistics Canada only submitted annual reports to LCDC. The transfer of data from Statistics Canada to the computerised surveillance database of the DSD was completed in 1990.

The goals of the CCDSS are:

1. To facilitate the control of the disease under surveillance by identifying:
  - a) prevailing incidence levels, impacts, and trends to assist in the development of feasible objectives for prevention and control of the disease, and the evaluation of control programs;
  - b) epidemiologic patterns and risk factors associated with the disease to assist in the development of intervention strategies;
  - c) outbreaks for the purpose of timely investigation and control.
  
2. To satisfy the needs of government (especially regulatory programs), health care professionals, voluntary agencies, and the public for information on risk patterns and trends in the occurrence of communicable diseases.<sup>57</sup>

These goals are with respect to all notifiable diseases and there are no disease-specific goals although for poliomyelitis it is appropriate to extend the second goal to an external regulatory body such as the ICCPE.

The CCDSS provides ongoing surveillance of paralytic poliomyelitis under provincial and territorial authority. Nonparalytic polio, or aseptic meningitis, is notifiable as viral meningitis although reporting is not separate from other viral causes of meningitis. Indeed, the reporting form for provincial and territorial aggregate data (Report of Notifiable Diseases - Canada Provinces and Territories, LCDC/09-92) and the annual summaries of notifiable diseases indicate that the category of viral meningitis excludes data for poliomyelitis. No basis was found for this conflict in guidelines.

Three provinces (Alberta, British Columbia and Manitoba) also require reporting of positive laboratory findings in cases of asymptomatic poliovirus infection. Ontario has a broader

legislation requiring "all positive laboratory findings with respect to a reportable disease" to be reported.<sup>58 p. 645</sup> The Yukon does not have specific legislation for reporting laboratory isolation of the virus but reportedly, the practice has been that all positive bacterial, viral and serological findings related to notifiable diseases are reported to the territorial authorities.

Nonpolio enterovirus-related paralysis is not notifiable in any of the provinces and territories. At least one province indicated that the general requirement to report "unusual disease circumstances and outbreaks" would cover cases of nonpolio enterovirus-related paralysis. This is not necessarily so as sporadic cases might not be reported under those terms and that is precisely the information needed to quantify AFP. Botulism is nationally notifiable and covered by provincial and territorial disease reporting legislations. GBS is reportable by law only in Ontario, and only when associated with the administration of a vaccine.

#### **2.4.2 The Vaccine-Associated Adverse Events Surveillance System**

The VAAESS is the federal system for post-marketing surveillance of adverse health events following the administration of immunizing agents. The system is administered by the Childhood Immunization Division (CID), a division of the BCDE. The VAAESS was established in 1988 with retrospective classification of data up to January 1, 1987. There are a number of ongoing modifications to the system including changes in the reporting and data handling procedures with effect from 1990.

The broad objective of the VAAESS is to quantify the rates of both well-recognised, and previously undescribed post-immunization adverse events and to identify the risks for these events.

Three reporting categories in this program are relevant to polio and AFP surveillance; GBS, Meningitis/Encephalitis, and the non-specific category of Paralysis. Reporting under this system is dependent on a temporal relation between the adverse event and immunization. Also, the reporting of adverse events is voluntary in all provinces except Ontario where it is mandatory.

### **2.4.3 The Immunization Program Active System**

The Immunization Program Active (IMPACT) System is a hospital-based system for the surveillance of post-vaccination adverse events and vaccine-preventable diseases. The program is contracted by LCDC to the Canadian Paediatric Society (CPS) and unlike the CCDSS and the VAAESS, is targeted at children. The IMPACT system was initiated on a pilot basis (with five paediatric centres) in March 1991, and expanded in July 1992 to include 10 centres across Canada although all centres did not become fully operational until March, 1993 (C. Glover, personal communication, July 1993). The current coverage represents 80% of tertiary care paediatric beds in the country.<sup>59</sup>

The overall program objectives<sup>59</sup> include:

1. To determine the occurrence of serious or unexpected events associated with vaccination of children.
2. To document the course and outcome of such events contributory to the rationale for a national program of assistance to vaccine-damaged individuals.
3. To accumulate epidemiologic information necessary for the interpretation of post-immunization events or pertinent to communicable disease epidemiology and routine immunization programs.

The IMPACT system has several components but the following illustrate its role in polio surveillance in Canada;

1. The surveillance of post-immunization events including the detection and reporting of OPV-associated paralysis. An active search for cases of GBS, aseptic meningitis, and other syndromes of acute paralysis serves the purpose of screening for missed cases of polio.
2. The surveillance of neurologic syndromes including poliomyelitis and all types of nonpolio acute flaccid paralysis. This covers cases not associated with immunization.

The specific objective of the second component is described as directly contributory to Canada's certification by the WHO as free from indigenous wild poliomyelitis.

## 2.5 Case Definitions

### CCDSS

The CCDSS case definitions for paralytic poliomyelitis include two main categories; confirmed and possible cases (See Appendix A). Cases are also classified by the type of virus; wild or vaccine strain. A wild virus case may be further classified as preventable when inadequate immunization or non-immunization is demonstrated in a person eligible for immunization.

Wild virus cases are further subdivided into imported, import-related, and indigenous cases. This subcategorisation is of particular importance to the Polio Eradication Program with its prime goal of eradicating indigenous wild cases. For vaccine-associated cases, five subcategories are provided; recipient, contact, possible contact, no known contact, and immunocompromised cases. The presentation of the subcategory immunocompromised as distinct, and parallel to the other subcategories is potentially misleading. Although an

immunocompromised state is a contraindication to the administration of live vaccines (including OPV), there is still a theoretical possibility of vaccine-associated polio occurring in immunocompromised persons especially if the immune deficiency is unknown prior to immunization. Wild virus infection may also occur in immunocompromised persons in whom seroconversion following IPV is inadequate. Therefore, it may be more appropriate to consider the immune state as an additional description of a case that fits into any of the other categories.

Viral meningitis is defined in very broad terms with no specific reference to poliovirus. Although data on the specific viral cause may be available at the provincial or territorial level, only aggregate data would be tabulated at the federal level. Also, as previously mentioned (subsection 2.4.1) there are conflicting guidelines for reporting of nonparalytic polio in this category. The case definitions for botulism require detection of *Clostridium botulinum* for confirmed cases or "overwhelming clinical and epidemiologic evidence" of botulism.<sup>57 p. 12</sup> This level of specificity limits the potential for detecting missed polio cases among reported cases of botulism.

Polio case definitions in three provinces differ minimally from those recommended by the ACE. Case definitions in Ontario and Manitoba have the basic elements of the federal definition but do not include criteria for further classification of cases as confirmed or possible. This is acceptable as reporting in any jurisdiction is expected to lead to further investigations towards the classification of a case according to federal guidelines. Furthermore, delays in reporting might result from inappropriate attempts to fully classify a case at the initial onset. Ontario and Quebec use case definitions that include either virus isolation, or a four-fold increase between acute and convalescent poliovirus antibody titres as criteria for a confirmed case.

## VAAESS

The VAAESS does not have well formulated case definitions for paralysis and meningitis/encephalitis. GBS is defined only as "progressive weakness of more than one limb and generalized hypo/areflexia" (Health and Welfare Form for Report of a Vaccine-associated Adverse Event, HPB 5127B(6-91). However, diagnosis by a physician is required for each of these events and clinical details are required in addition to the standard information requested for all reports. Six provinces have their own reporting forms which are adapted from the LCDC form with no differences in the case definitions.

## IMPACT

The IMPACT case definitions for paralytic poliomyelitis are essentially the same as the definitions used in the CCDSS (see Appendix B). The case definition for GBS reflects the difficulty with which this diagnosis is often made. Clauses like "weakness typically ascends upwards from feet" and "often involves abdominal and thoracic muscles but cranial nerve involvement is uncommon"<sup>59 p. 43</sup> may be a source of confusion for the nurse monitors who are the primary source of reports. The system is however hospital-based and requires diagnosis of neurological cases by a neurologist so that the responsibility on the nurse monitors to correctly classify a case of GBS is minimal. Unlike the VAAESS, a clear case definition is provided for aseptic meningitis. IMPACT case definitions for other acute paralyzes also depend on pre-assigned clinical diagnoses (see Appendix B).

Other important reporting criteria state that:

1. All suspected cases of polio should be reported without waiting for confirmation or classification of a case (CCDSS, IMPACT).
2. Cases of aseptic meningitis with culture-negative cerebrospinal fluid (CSF), stool isolation of poliovirus and recent OPV vaccination are to be excluded from reports (IMPACT).

## 2.6 Components and Operation of the System

As there is very limited recent experience with polio for reference, most of the information obtained for polio surveillance was based on general communicable disease surveillance. This is reflected in parts of the following discussion but whenever possible the discussion focuses on polio and the acute paralyses already mentioned.

Routine surveillance under the CCDSS and the VAAESS is universal and passive in all jurisdictions. In special instances, a particular province will initiate a short-term active search for cases of infection as was the case in the recent investigation of poliovirus importation in Alberta and British Columbia. IMPACT is the only program of active surveillance for polio and other acute paralyses.

A general surveillance method applies to 46 of the 48 nationally notifiable diseases/disease categories including paralytic poliomyelitis. Federal recommendations for investigation and diagnosis of polio are made for surveillance purposes only. They include (a) collection of multiple stool and other clinical specimens within 2 weeks of onset of symptoms, (b) rapid transport of the specimens to a recognized viral laboratory with facilities for handling of the specimen, (c) adequate neurologic assessment including nerve conduction studies, and (d) follow-up to establish neurologic deficit lasting at least 60 days. Poliomyelitis is cited as the only disease for which an exception is made regarding the absence of data to determine whether a case meets the case definition or not; suspected cases are to be reported "promptly" for later confirmation.<sup>57</sup> All cases of polio are also reviewed by a subcommittee of the National Advisory Committee on Immunization (NACI) for classification. The membership of the reviewing subcommittee includes physicians involved in the care of a particular case.

## 2.6.1 Information Collection and Transfer

### CCDSS

The framework for reporting under the CCDSS is presented in Appendix C-1. The primary reporting sources of poliomyelitis are physicians, or admitting institutions, and laboratories that detect poliovirus in clinical specimens (Level A). Reports are submitted to local health authorities (Level B) who collect all necessary epidemiologic data for forwarding to the provincial or territorial epidemiologist (Level C). The final link in the chain is the submission of reports to the DSD (Level D). Three provinces (Manitoba, Prince Edward Island, and Quebec) and the Yukon report exclusively from Level A to Level C.

Reports received from laboratories are confirmed with attending physicians for further clinical information, and also as a check on duplication of reports. The Level B reporter is responsible for collecting all relevant information on a reported case. This information is recorded on standard reporting forms, the common case report form (Form A, HPB 5130A [12-89]) and the polio case report form (Form E HPB 5130E [12-89]), provided by the federal Health Department.

Beyond this point data are handled differently; both manual and computerised methods are used for data recording and transfer to the next level. Some provinces use a combination of both as shown in Appendix D-1. Provincial and territorial data aggregated by age, gender and month are submitted monthly to the DSD with the exception of Quebec which submits case by case data, on a quarterly basis. Traditionally aggregate data have been reported by fax or mail. As well, several provinces have developed or are at varying stages of developing electronic systems for case-by-case reporting. Most jurisdictions submitting case-by-case data do so monthly except Ontario (weekly) and Alberta (every 2 to 3 weeks). The territories

submit case report forms for direct data entry into the DSD database which is based on the EPIC software system.

### VAAESS

The VAAESS has a similar framework for reporting as the CCDSS except that Level A sources are limited to nurses and physicians (see Appendix C-2). In a few cases, reports of adverse events are received through a vaccine manufacturing company although reports are not accepted from the general public. Reports submitted directly by health care workers to federal authorities are redirected to provincial and territorial health departments to avoid duplication of records, and to ensure that the province or territory from which a case originated is informed. The endpoint of the reporting chain is the CID where data received per standard reporting forms are entered into a database currently based on the SAS software; the division is undergoing a change to the EPIC system. Again there is an exception with Quebec which has made a number of changes in reporting; since September 1991, reports have been submitted by computer diskette, initially monthly but for the past six months on a bimonthly basis.

### IMPACT

The flow of information in the IMPACT system is summarised in Appendix C-3 the endpoint being the IMPACT Coordinating Centre in British Columbia with data entry into a SAS database. Reports are submitted by nurse monitors at individual sites at monthly intervals. Participants in the IMPACT system are expected to have a higher awareness of polio than the general population of health care workers operating under the CCDSS and the VAAESS as the system uses an active approach. If this is so then in time the quality of reporting with regard to completeness and timeliness should be comparatively higher.

## 2.6.2 Data Manipulation

### CCDSS

Generally, the frequency and scope of local distribution of communicable disease summary reports differ by province and territory as summarised in Appendix D-2. Most jurisdictions generate and distribute reports on a monthly basis. For polio, the general indication was that more regular dissemination of information would be initiated at the local level if cases were reported.

At the federal level, notifiable disease data are presented as summary statistics of age and sex incidence by province or territory. Preliminary monthly incidence of polio and other selected diseases are generated for internal distribution within the BCDE. Data are disseminated more broadly through monthly summary reports of all notifiable diseases published in the Canadian Communicable Disease Report (CCDR), and through published annual summaries; summary reports include negative reporting data. The monthly reports are also communicated via an electronic bulletin board system (BBS); provincial and territorial epidemiologists are linked to the BBS and thus have access to the monthly reports, and other communication relayed in special instances. There are no specific reports issued to the provinces and territories the reason being that the analyses are based directly on the data submitted (M.J. Garnett, personal communication, July 1993).

A review of published reports in the CCDR covering an 18-month publication period (January, 1992 to June, 1993) indicated a mean lag time for publication of 5.7 months, and a range of 4 to 8 months. During this period there were only nine monthly reports with no apparent regularity; coincidentally the calendar period of surveillance covered by the reports was also 18 months.

## VAAESS

VAAE reports are followed up for epidemiological and clinical information (when necessary) at the time of reporting, and at 6-month and 1-year follow-up. Data analyses are based on the distribution of adverse events by (a) demographic variables (age, sex and geographic location), (b) immunizing agents (single and combined), (c) dose number of selected immunizing agents, and (d) type of adverse events. Updated quarterly reports are issued to provincial and territorial epidemiologists, vaccine manufacturers, and other special interest groups including WHO and the Bureau of Biologics (the Canadian federal licensing body for vaccines). Reports are also published annually in a variety of scientific journals.

## IMPACT

To date the dissemination of IMPACT data on paralytic syndromes has been limited to monthly summaries distributed to the nurse monitors and site investigators, and liaison persons at the CPS and LCDC, although special reports have been prepared on other reportable diseases.

### **2.7 Attributes of the System**

The attributes of the polio surveillance system in Canada are summarised in this section.

#### Simplicity

I found the current system of polio surveillance to be uncoordinated mainly due to the multiplicity of programs and thus, presenting a significant challenge for evaluation of its effectiveness. Multiple surveillance programs for the same disease have the potential advantage that one can be used as a check on the others. In spite of this, a well-structured combined program would have the additional advantage of simplifying both the operation and the future evaluation of the system for documentation purposes. Data for such a system

could still be based on the existing reporting mechanisms thus retaining the possibility of comparing data from one component of the program with another.

The case definitions and laboratory requirements for classification of polio cases are complicated, nonetheless they are justified by the nature of the disease and the current goals of surveillance. The existing case definitions should be reviewed and standardised for all three programs. As previously pointed out, an eradication program requires a high level of sensitivity in order to detect all cases. To this end, AFP surveillance can be instituted in Canada as a screening phase within the overall surveillance of polio.

#### Flexibility

The flexibility of the system is difficult to assess without the benefit of changing needs in the past. There should be an opportunity to demonstrate the system's capability to adapt to changing needs with the implementation of AFP surveillance and nonparalytic polio (separate from other causes of viral meningitis).

#### Acceptability

This is often judged by the participation of reporters and the accuracy or completeness of reports submitted. In this review accounts of both indicators were mostly anecdotal. All the provinces and territories indicated that the completeness of reporting is very variable at the local level and that laboratories are more consistent than physicians in reporting notifiable diseases. The data on poliomyelitis in the DSD database (1958-1992) is woefully incomplete; both age and sex are unknown for 38.4% of 3,084 recorded cases, and age alone remains unknown for another 2.2%.

The VAAE reports are generally complete but some improvements are needed for assessment of causality, relevant clinical details, immediate outcome and follow-up information.

Particularly for GBS cases and other paralyses, complete information on the clinical presentation and results of investigations (positive and negative) would be very valuable. Lack of this information hinders a retrospective assessment of the accuracy with which polio was ruled out, if it was considered at all as a possible diagnosis. Of particular note is the fact that Quebec submits data by diskette; this limits the amount of clinical detail provided with reports but may be an indication that the general system developed by LCDC is unacceptable to the province.

The level of acceptability of the IMPACT operating system is reported to be high although some difficulties have been encountered in completing the reporting forms (C. Glover, personal communication, July 1993). Ongoing training is a part of this relatively new initiative so improvements are anticipated.

#### Sensitivity

Sensitivity is measured as the proportion of true cases in the target population that are detected by the surveillance system. In order to measure the sensitivity of surveillance, some "gold standard" is needed as a measure of the true incidence of polio. Because polio is currently so rare in Canada, any systematic effort to retrospectively determine all possible cases will require immense resources. One method would be to compare laboratory-based data of virus detection with the number of cases reported to the surveillance programs but available laboratory data on poliovirus isolation are difficult to interpret because of the lack of clinical information.

Generally, the sensitivity of a surveillance system is influenced by three factors: (a) initial contact between a person with the disease and the medical system; (b) the probability that the disease will be diagnosed, which is in turn dependent on a high index of suspicion for the

disease; and (c) the probability that given the right diagnosis, the case will be reported to the system.

With regard to the first factor, there is an extremely high expectation that clinical cases of poliomyelitis would seek medical care and thus come to the attention of a physician. The second factor is not discussed here as a discussion on the sensitivity of polio diagnosis forms a major part of Chapters 3 and 4.

Currently physician reports form a major part of polio surveillance yet underreporting by physicians is a well described phenomenon often related to the perceived importance of the disease and knowledge about reporting mechanisms. In a comprehensive review of public health surveillance in the United States, reporting frequencies for communicable diseases ranged from 6% to 90% among physicians.<sup>51</sup> Konowitz et al,<sup>60</sup> found reporting frequencies as low as 28% among a group of physicians surveyed in the US; most had a general knowledge about disease reporting but the proportion with accurate knowledge of disease-specific requirements ranged from 63%-96%. As well, less than 24% had accurate knowledge of reporting procedures whereas about 30% actually thought they did. With this knowledge, and given the perceived unimportance of polio among many clinicians in Canada, there is the likelihood that the sensitivity of the system is at best moderate. It has been observed in the United States that underreporting by physicians can be improved by ensuring that all laboratories report cases.<sup>61</sup>

### Representativeness

The characteristics of recently reported cases of polio correspond well with what is expected given the current epidemiology of polio in Canada. Specifically, these characteristics are (a) the extremely low numbers reported; (b) the preponderance of vaccine-associated cases over wild cases; (c) the evidence obtained for importation in relation to wild virus cases in 1978,

1979 and 1992-1993; (d) the fact that most wild virus cases have occurred in communities with low immunization coverage; and (e) the relatively widespread age distribution (when compared to polio-endemic countries where polio usually affects children). These facts suggest that polio surveillance in Canada is highly representative of true cases.

### Timeliness

As with the completeness of reports, evaluation of timeliness was based on anecdotal reports because of a lack of empirical data. The timeliness of disease notification is important to the investigation of a case and establishment of an accurate diagnosis, as well as the institution of control measures. For the CCDSS, laboratories were reported as being more timely than physicians in notifying appropriate health authorities of cases. Delays in forwarding aggregate data to LCDC are reported as contributory to the delayed publication of monthly and annual national summaries.

Timeliness of reports to the VAAESS varies by province, the general practice being that reports are accumulated at the local level and forwarded to the LCDC in batches at undetermined intervals. The timeliness of reports is also likely to be influenced by the type of vaccine delivery system (public or private). For the IMPACT system, some limited delay was reported for the submission of reports to the Coordinating Centre. This has been attributed to the incompleteness of information at the investigation sites however the delays reportedly do not affect the preparation of monthly summaries (C. Glover, personal communication, July 1993).

## **2.8 Outcome Measures and Usefulness of the System**

The most obvious outcome in the past has been the knowledge gained on annual incidence and trends, particularly for paralytic polio since 1949 when separate reporting was instituted.

Analysis of surveillance data demonstrated a significant reduction in the incidence of polio with the separate introduction of inactivated and live poliovaccines. Polio surveillance data have led to the recognition of epidemics, and more recently the importation of wild virus thus leading to appropriate control activities. Policy decisions on immunizations against polio have also been guided by surveillance data.

Data from the Canadian Virus Reporting Program enable comparisons between the poliovirus and other enteroviruses, and also between serotypes of the same virus. Trends in virus isolation for 1986-1992 are presented in Figure 5; specific data for various serotypes are not included but are available. From 1989 to 1992, a large number of enteroviruses were reported without typing (more than 65%). Figure 6 shows that whereas coxsackieviruses and echoviruses were more frequently reported from 1986 to 1988, the proportion of poliovirus isolates has increased since 1989; in 1990 the poliovirus constituted approximately 40 percent of all typed viruses reported.

Generally, there are no marked differences in the frequency of detection of the three poliovirus serotypes however there was a slight increase of types 2 and 3 over type 1 in 1989 and 1990 (see Figure 7 which is based on serotyping only). Approximately 75.2% of all poliovirus isolate reports do not include a characterization of the virus as wild or vaccine-type, and are assumed to be vaccine-type (G. Wilson, personal communication, August 1993). The rest are all characterised as vaccine-type. There is an improving trend in poliovirus characterization; from 1986 to 1990, 95.1% to 98.8% of isolates were not characterised but this figure dropped to 59.4% in 1991 while in 1992 all polioviruses reported were characterised as vaccine-type.

A National Centre for Enteroviruses was established in 1991 and provides better opportunity for (a) investigation of outbreaks of enterovirus infection in Canada and (b) typing services

to establish the source of infections where appropriate. Table 4 summarises the virus typing activity of the National Centre in its first two years of operation; the overall number of typing requests received is very small compared to the reported number of virus isolations in the country. It is anticipated that with time the role of the National Centre in enterovirus surveillance will be better defined and publicised, and that more specimens will be submitted for typing when appropriate.

More specifically the National Centre played a major role in the recent investigation of wild poliovirus importation in Alberta resulting in identification of a type 3 virus shown through genomic analysis to be closely identical to the type 3 virus isolated in the 1992 polio outbreak in the Netherlands.<sup>42,43</sup>

Table 4

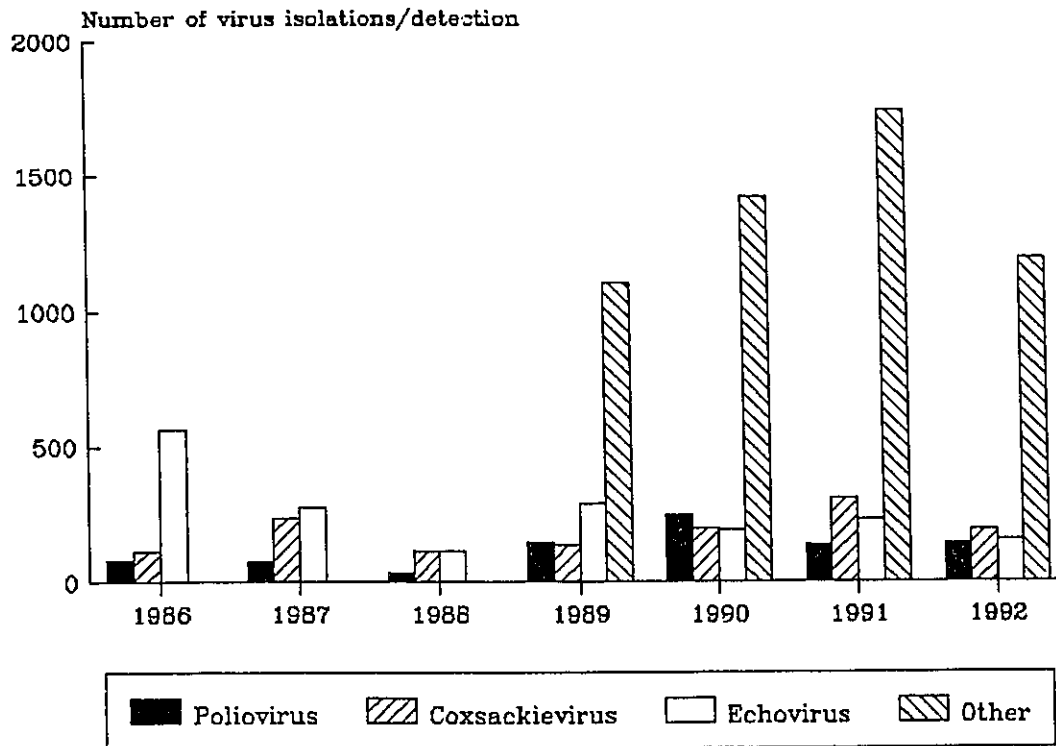
Enterovirus clinical isolate typing at the National Centre for Enteroviruses  
Canada, April 1991 - March 1993

Period	Poliovirus	Coxsackievirus	Echovirus	Total
1991-92	3 <sup>a</sup>	38	158	199
1992-93	8 <sup>b</sup>	42	41	91

Source: Annual Reports of the National Centre for Enteroviruses (1991-1992 and 1992-1993).

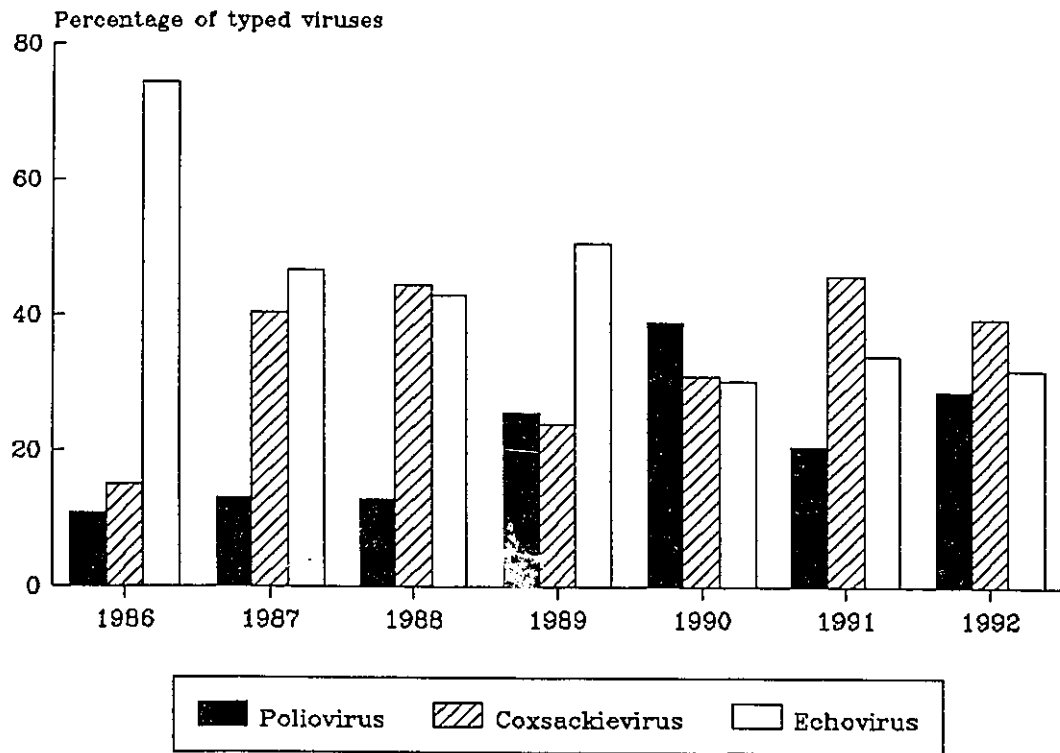
<sup>a</sup> One type 1 and two type 3

<sup>b</sup> One type 1, six type 2, and one type 3



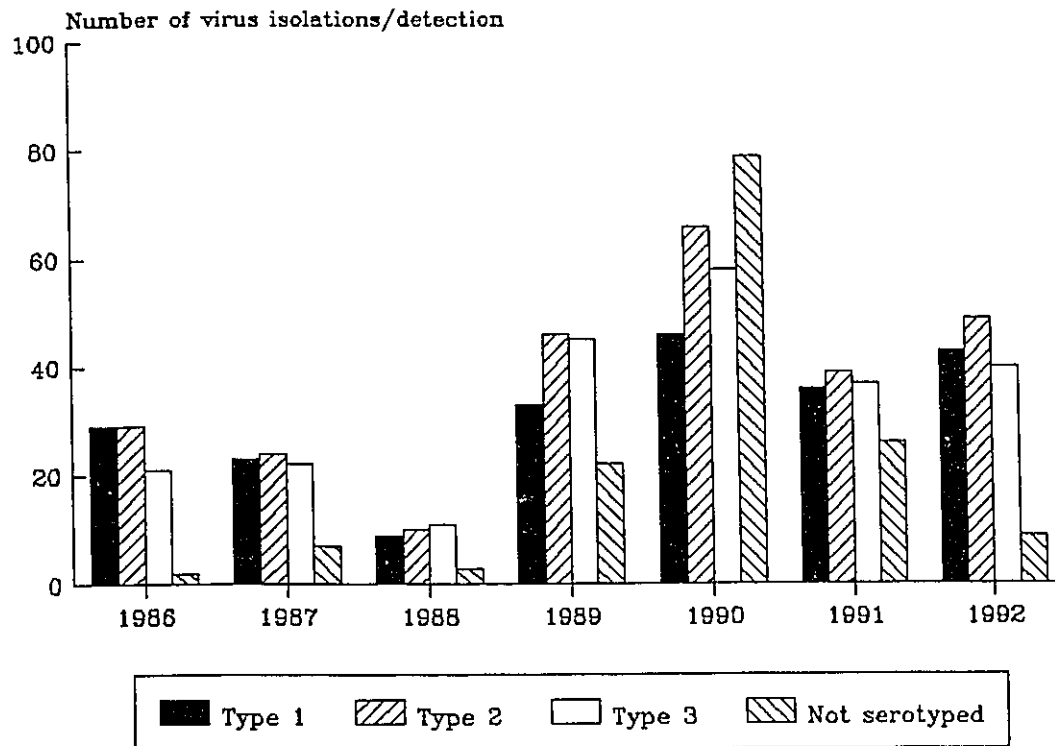
There are no data to differentiate wild and vaccine-type poliovirus

Figure 5 Laboratory evidence of human enteroviral infections in Canada, 1986 - 1992. Source: Canadian Virus Reporting Program, Bureau of Microbiology, LCDC.



There are no data to differentiate wild and vaccine-type poliovirus

Figure 6 Comparison of laboratory evidence for different human enteroviral infections in Canada, 1986 - 1992. Source: Canadian Virus Reporting Program, Bureau of Microbiology, LCDC.



There are no data to differentiate wild and vaccine-type poliovirus

Figure 7 Laboratory evidence of human polioviral infections in Canada, 1986 - 1992. Source: Canadian Virus Reporting Program, Bureau of Microbiology, LCDC.

A detailed review of the seven cases of polio reported in Canada from 1987 to 1992<sup>36-38</sup> shows differences in initiation of polio-specific investigations, namely stool and/or throat viral cultures, and serology. The mean interval between onset of paralysis and submission of stool samples for culture was 16 days (range of 3 to 49 days). Although all cases had at least one stool specimen cultured, only two of the seven (28.6%) had two stool specimens cultured within two weeks of the onset of paralysis. Two more cases had only one specimen cultured within this interval. Thus, a total of 57.1% had at least one stool culture within two weeks.

From 1987 to 1992, the VAAESS received 23 reports of vaccine-associated paralysis, 8 of which were associated with OPV (four contacts and four recipients). Six cases were diagnosed as paralytic polio, all in association with OPV and corresponding to the cases reported to the CCDSS. Two of the 23 cases of paralysis were diagnosed as GBS and there were 10 other cases reported as GBS; none of the GBS cases was in association with OPV.

Cases of AFP reported to IMPACT as at July 1993 are summarised in Table 5. Only 58.3% of these cases had a record of viral culture or serology; approximately 43% of records with viral studies indicated a stool viral culture while for another 43% the only specimen cultured was CSF. One case had serology only with positive results for an enterovirus. For one case the specific specimens cultured were not reported. The high proportion of CSF cultures possibly indicates that the investigations were not polio-specific because CSF offers the least chance for poliovirus detection.

The above findings are very poor when compared with the PAHO requirement for two stool cultures within two weeks of onset of paralysis in at least 80% of AFP cases. Two of the three polio cases with delays in stool culture had CSF investigations and serology within two weeks of onset of paralysis and one case had CSF investigation three weeks after paralysis. The findings suggest that CSF examinations and serology might have been routine and

unrelated to a suspicion of polio. The fact that all CSF cultures were negative (some of the cytological and biochemical findings supported aseptic meningitis) supports the need for rapid investigation of stool and throat specimens which are the best specimens for viral detection. Three of the seven cases were only classified as possible cases of polio partly due to inadequate investigations. There is no indication of stool cultures in contacts although diagnosis may in some cases be based on a positive culture in a contact; over 10% of all confirmed cases reported to PAHO in the last three years were reportedly confirmed on the basis of contact investigations.<sup>62</sup>

Reports on AFP surveillance under the PAHO program also note that inadequate investigation, specifically inadequate collection of stool specimens, led in some instances to the incomplete classification of cases. In 1990 and 1991, 71 and 33 cases respectively were only classified as polio-compatible without enough information for their confirmation.<sup>21</sup> In both the Canadian and PAHO examples the lack of confirmation of cases has stemmed from situations in which a) investigations were not initiated promptly following the onset of illness, or b) appropriate specimens were not collected from cases and/or contacts. Such situations are clearly undesirable in a program of disease eradication.

Table 5

AFP cases reported to the IMPACT System, January 1991 - July 1993

Year	GBS	Transverse Myelitis	Other	Cases with viral investigations
1991	5	1	0	2
1992	7	0	1	5
1993	12	3	1	7
Total	24	4	2	14

## 2.9 Discussion

Probably the most important use of polio surveillance in the future is the early recognition of wild virus importation and the associated risk for re-emergence of wild polio. Also important are the early recognition of risk groups and a reassessment of existing prevention and control measures, especially among groups that do not readily submit to those measures. Surveillance data are also required to document polio elimination in Canada for certification purposes. Above all the requirement for reporting, if well publicised and justified will promote awareness about the continuing public health importance of poliomyelitis despite elimination of indigenous transmission.

This review indicates several loopholes in the surveillance system which can conceivably result in polio cases being missed by public health authorities. The system is mainly dependent on passive physician-based reporting, a method which generally has low sensitivity, incompleteness of data, and poor timeliness of notification. Physician-based surveillance is highly influenced by the existing index of suspicion for a disease. As polio is currently very rare, a low index of suspicion is likely to be found across Canada and the appropriate diagnostic work-up may be hard to justify even though it is relatively simple. The frequency of missed polio cases is liable to increase with time, if not in absolute terms at least as a proportion of all polio cases.

The current surveillance of poliomyelitis in Canada while focused on paralytic polio, does not offer a high potential for detecting other forms of poliovirus infection. In addition, the potential for detecting missed cases of polio among other cases of paralysis is not very promising because (a) surveillance for other paralyzes is not universal and is mostly limited to the paediatric population (through the IMPACT system) whereas the age distribution of polio in Canada in the most recent years has been much wider, and (b) reports of other

paralyses have not provided complete information on clinical and investigative details needed to assess the possibility of polio.

There are no routine methods of environmental surveillance through which poliovirus importation can be detected before the rare case of clinically evident polio occurs. Also, laboratory data on poliovirus isolation are difficult to interpret as the associated clinical data are often not available at the central level.

Currently no province or territory has written protocols for the investigation of suspected cases other than the brief recommendations in the federal guidelines. Similarly, there are no written protocols for control measures in the event of a case. The general indication by Provincial and Territorial Epidemiologists was that a suspicion of poliomyelitis would initiate prompt and regular communication with LCDC to develop a plan of action. This remains to be tested; procedures for the recent investigation of suspected poliovirus importation were initiated by LCDC and further developed for implementation by Alberta, British Columbia, and Ontario but investigation was limited to specific communities. There is still the need to define general protocols for case management and control measures.

Although the activities of each of the three surveillance programs are within the scope of their stated objectives, significant improvements are needed in information collection and transfer to completely quantify the occurrence of poliomyelitis. Because of the extremely low proportion of paralytic cases, the inclusion of nonparalytic cases in surveillance should be an important component of risk assessment. With respect to the specific indicators of surveillance outlined by PAHO for certification, the available data indicate that Canada does not currently meet the criteria.

## CHAPTER 3      REVIEW OF HOSPITAL CHARTS

The second part of the project consisted of assessing the potential role of AFP surveillance in documenting a polio-free status in Canada. As outlined in Chapter 1, one of the major components of PAHO's eradication program is the active surveillance of all AFP cases in children under 15 years, and their investigation to rule out polio.

### 3.1      AFP Surveillance and Predictors of Paralytic Poliomyelitis

By definition, surveillance of paralytic polio based on a case definition of acute flaccid paralysis is 100% sensitive. However, data from the surveillance program in the American Region has in the past indicated a very low specificity; this excludes Canada and the United States which have not provided any data to the program. Of 1,930 AFP cases reported to PAHO in 1989, less than 7% were categorised as polio-compatible or confirmed polio. Similarly, in 1990, less than 5% of 2,403 AFP cases reported were either confirmed as polio, or found to be compatible with polio.<sup>2</sup>

Two recent studies have described screening criteria that may be used to improve the application of AFP surveillance in the Americas. Dietz et al studied several clinical variables, individually and in combination, to determine their relative predictive power for poliomyelitis in AFP surveillance.<sup>63</sup> The measure of prediction used was the likelihood ratio which expresses the probability of a specific value of a measure, or test result (positive and negative for dichotomous tests), in the presence of a diagnosis as compared to its probability in those without the diagnosis.<sup>64</sup> The following characteristics were reported as the best predictors of culture-confirmed polio in children below the age of 15 years with AFP; (a) proximal muscle paralysis progressing in less than 4 days together with fever at the onset of paralysis, (b) proximal and unilateral muscle paralysis with fever at onset, and (c) proximal and

unilateral muscle paralysis with progression of paralysis in less than 4 days. Each of these combinations of variables had a positive likelihood ratio (PLR) of 12. When analyzed individually, only one variable (unilateral paralysis) had a PLR higher than the selected cutoff of 5. One variable was also reported with a significant negative likelihood ratio (NLR) of 5.4; paralysis progressing over a period of more than 4 days.

In a separate study by Andrus et al,<sup>65</sup> operational screening criteria for AFP cases reported in the Americas were evaluated with similar findings. The highest specificity achieved for the diagnosis of culture-confirmed paralytic polio was 34% while maintaining a sensitivity of 100%. This was achieved with either (a) fever at the onset of paralysis combined with installation of paralysis (from onset to full extent of paralysis) in less than 4 days, or (b) age less than 6 years with installation of paralysis in less than 4 days. As described in standard epidemiology texts, increases in the specificity of any test (screening procedure, diagnostic test etc.) are associated with a decrease in sensitivity, and vice versa. In this study, while maintaining a high sensitivity (96%), the specificity of AFP surveillance could be improved to 49% by the inclusion of two alternative criteria; (a) age less than 6 years with fever at the onset of paralysis, or (b) age less than 6 years with a rapid progression of paralysis (in less than 4 days). Using age less than 6 years as the sole screening criterion for AFP resulted in a sensitivity of 93% and a specificity of 43%. Further increases in specificity (73% and 82%) were obtained with other combinations of variables although there was a corresponding drop in sensitivity (75% and 64% respectively). Based on these findings, PAHO recommended that AFP case investigation be focused on young children with (a) fever at the onset of paralysis, (b) a rapid installation of the full extent of paralysis, or (c) both of these.<sup>21</sup>

Both studies cited were based on data for children below the age of 15 years thus, the application of these findings to a wider age distribution of AFP cases may be limited. However, variables other than age found to be significant screening criteria are inherent

disease characteristics not known to vary with age and therefore still applicable with a wider age distribution. As these were essentially studies of predictive power, the analyses focused on early features of the disease although Andrus et al reported odds ratio estimates for two late-onset characteristics; neurological sequelae (OR = 14.0) and atrophy (OR = 16.8). These variables are potentially useful for retrospective reviews of AFP cases.

### **3.2 Aims and Objectives of the Chart Review**

In Canada (and other countries like it with a near-zero occurrence of paralytic polio) it is necessary to consider what the potential usefulness of AFP surveillance is. If it is judged to be useful, measures are required to ensure full participation of health care providers in reporting cases. Such measures include providing data on the probability of missing polio cases in the absence of AFP surveillance. Conversely, any data that might be used to rule out the likelihood of such missed cases of polio over a prolonged period in the past may prove to be an important element of the certification process.

To examine these issues, I conducted a retrospective review of medical charts of cases that presented with acute flaccid paralysis in the Ottawa-Carleton region from 1986 to 1991. The chart review was conducted as a pilot study to examine the feasibility of incorporating a) a chart review of AFP cases, and b) ongoing AFP surveillance in a plan for documenting polio elimination in Canada.

The specific objective of the chart review was:

To determine the potential for missed cases of paralytic poliomyelitis among AFP cases presenting in Ottawa-Carleton.

To achieve the stated objective, answers had to be provided to the following questions:

1. What proportion of AFP cases present with polio-compatible features?
2. How often are polio-compatible AFP cases investigated to confirm or rule out poliomyelitis?
3. If no polio-specific investigations are carried out in a case, do either the clinical data or the results of investigative procedures provide convincing evidence for a diagnosis other than paralytic polio?

### **3.3 Methods**

Charts for the review were obtained from six Ottawa area hospitals; the Children's Hospital of Eastern Ontario (CHEO), the Montfort (MFH), Ottawa Civic (OCH), Ottawa General (OGH), Queensway-Carleton (QCH), and Riverside (RH) Hospitals. These hospitals are all referral facilities and were selected on the basis of acute care delivery to paediatric and adult medical cases in the study area. Basic profiles of each hospital, relevant to the review, are provided in Appendix E. Approval to conduct the chart review was obtained from each hospital and guidelines subsequently provided to respective medical records departments for the retrieval of all potentially eligible charts. Charts were reviewed as they were made available with no predetermined sequence.

#### **3.3.1 Sources of Data**

Guidelines for chart retrieval were as follows:

1. The review was limited to in-patient admissions during the five-year period from April 1, 1986 to March 31, 1991; emergency room records were excluded.

2. No limits were placed on the age of AFP cases because of the exploratory nature of the study. This decision is supported by two facts; (a) data for the last 25 years show that more than 50% of paralytic polio cases in Canada have occurred in persons above the age of 20 years and (b) PAHO defines a suspected case of polio as "any case of acute paralysis in a person less than 15 years old for any reason other than trauma, or paralytic illness in a person of any age in whom the diagnosis of poliomyelitis is suspected".<sup>7 p. 10</sup>
  
3. The specific diagnosis for chart identification and retrieval was the main diagnosis assigned at the time of patient discharge. This term is used with respect to the four levels of diagnosis that constitute the standard coding system of the Hospital Medical Records Institute (HMRI) used by all the participating hospitals. The four levels are (a) main, the diagnosis for which the patient was admitted to hospital; (b) primary, meaning other important diagnoses that influenced the patient's length of stay in the hospital; (c) secondary, meaning other important diagnoses that did not affect the length of stay in the hospital; and (d) complications, which refers to any condition or conditions arising from treatment given during the stay in hospital, or the hospital stay itself.
  
4. Diagnoses of interest were those with major presenting features similar to those of paralytic poliomyelitis. In particular, disease categories likely to include trauma-related paralysis, or cerebrovascular conditions were excluded. This was done because of the expectation of a high proportion of false positive polio-compatible cases in those categories.

The categories, and corresponding ICD-9 codes used for chart retrieval are presented in Table 6. For the period under review, diagnoses were coded according to the ninth revision

of the International Classification of Diseases (ICD-9 codes) in which an innovation of dual classification was introduced for certain diagnostic conditions.<sup>66</sup> By this means one code would assign a disease to a classification relating to the underlying disease, and the other code would assign the same disease to a classification relating to the organ system affected. These codes, the primary and secondary codes respectively, enable statistical analyses or chart retrieval by alternative means. Of the selected disease categories, all but polio had a single code. The diagnostic terms and corresponding ICD-9 codes used for paralytic polio are presented below. The code for acute nonparalytic polio (045.2) was excluded.

Primary codes (listed under infectious and parasitic diseases):

045.0; acute paralytic polio specified as bulbar

045.1; acute poliomyelitis with other paralysis

045.9; acute poliomyelitis, unspecified

The secondary code (listed under the nervous system):

323.2; poliomyelitis.

Table 6

Diagnostic categories used in chart retrieval

Diagnostic categories	ICD-9 codes
Botulism	005.1
Paralytic poliomyelitis	045.0, 045.1, 045.9, 323.2
Transverse myelitis	323.9
Monoplegia of lower limb	344.3
Monoplegia of upper limb	344.4
Unspecified monoplegia	344.5
Guillain Barré Syndrome	357.0

Retrieval of charts according to the above guidelines was followed by a screening procedure to eliminate all but the truly eligible charts from the data collection process. This two-tier procedure was necessary because of the possibility of errors in the retrieval system. The anticipated sources of error were (a) incorrect coding of diagnoses (with either the HMRI code or the ICD-9 code), and (b) mistakes in the actual retrieval of charts (e.g., retrieval of a chart with a specified ICD-9 code assigned to a diagnosis other than the main diagnosis, or with an irrelevant date).

The eligibility criteria for chart review were:

1. The record documented an incident admission; readmissions for a given diagnosis were excluded.
2. The patient was discharged from hospital between April 1, 1986 and March 31, 1991 inclusive.
3. The main diagnosis was one of the preselected diagnoses **OR** an acute paralysis of non-traumatic nature was part of the initial presentation. This meant that charts were excluded if the main diagnosis was not one of the pre-selected diagnoses **AND** an acute paresis or paralysis of non-traumatic nature was not documented.

It was considered possible that a diagnosis exclusive of the selection list would be encountered in the event that a diagnosis was miscoded or that a chart was retrieved incorrectly. By using the qualifier or in the last criterion for eligibility, such charts could be included provided the other eligibility criteria were met, and with the following additional provisions:

1. These charts were to be considered a potential source of information on other diagnoses that could be included in the selection list for future reviews.

2. As such diagnoses would not have an equal chance of being retrieved from all participating hospitals, the data pertaining to them would have to be excluded from analyses relating to specific diagnoses.

### **3.3.2 Development of Data Collection Form**

A standardised form was developed for data collection in consultation with a practising neurologist (P. Jacob) who was the study consultant. This process consisted of three steps:

1. Identifying the elements of (a) patient history, (b) clinical examination, and (c) case management (including investigations), that would be relevant to the objectives of the review.
2. Designing a form to include each of the elements above; response options in the form were mostly close-ended.
3. Pretesting of the form with subsequent minor modification.

The data collection form was designed with six sections relating to (a) chart identification and demographics, (b) diagnoses and outcome, (c) pattern of physician referral and public health notifications, (d) presenting symptoms, (e) clinical signs, and (f) laboratory and other investigations (See Appendix F).

The symptoms and signs used in the review were selected on the basis of classical descriptions of polio<sup>10,12,15</sup> and the screening criteria reported in the published studies discussed in subsection 3.2. Symptoms and signs were grouped into two major types of variables; (a) variables that would support a diagnosis of polio, and (b) variables that would support the exclusion of a diagnosis of polio.

Rarely in clinical medicine is one single symptom or clinical sign pathognomonic of a given disease. Thus, the diagnostic weight of each is dependent on its relative frequency of occurrence. To emphasize this, the symptoms and signs presented below are grouped according to their relative frequency of occurrence in paralytic poliomyelitis as compared to other diseases or conditions. The symptoms and signs can however only support or help to rule out a diagnosis of paralytic polio.

A) Factors that generally support a diagnosis of poliomyelitis:

1. A positive history (usually preceding paralysis by one to two weeks) of;
  - a) Prodromal fever
  - b) Respiratory infection
  - c) Gastrointestinal symptoms
  - d) Contact with an OPV vaccinee or travel to a polio-endemic region
  
2. Clinical signs of;
  - a) Fever at the onset of paralysis
  - b) Asymmetrical paralysis
  - c) Rapid evolution of paralysis (from onset to the full extent), within approximately 4 days
  - d) Absent or reduced deep tendon reflexes in the affected limbs only
  - e) Hypotonia
  - f) Residual paralysis after 60 days

B) Symptoms and signs that would generally rule out a diagnosis of polio:

- a) Prolonged evolution of the full extent of paralysis, more than 4 days
- b) Trauma
- c) Rashes

- d) Arthralgia
- e) Sensory loss, especially with a distinct sensory level
- f) Symmetrical paralysis
- g) Global absence or reduction of deep tendon reflexes
- h) Loss of sphincter control of bladder or bowel

Some of these are direct opposing features of the polio-supportive features above while others were included because of the support they might lend to another diagnosis (e.g., rashes or arthralgia might be suggestive of a myositis or polymyositis, especially in a child).

Due to lack of information about the presence of fever at the onset of paralysis, a proxy variable relating to fever at the time of admission was used in this study. Similarly, there was no information on the period of progression of paralysis; the variable used to approximate this value relates to the progression of paralysis after admission. The impact of these proxy variables on the diagnostic conclusions is discussed later (see subsection 3.5).

Data were also collected on paralysis of cranial nerves which is the hallmark of bulbar paralysis, however this form of poliomyelitis is itself relatively uncommon representing about 6 to 25% of paralytic cases.<sup>15</sup> Therefore, the documentation of cranial nerve paralysis requires careful interpretation. In the study by Andrus et al, cranial nerve paralysis was associated with an odds ratio of 0.3 suggesting another diagnosis as more likely than polio.

I also included factors of a less specific nature that are considered by some authorities and many physicians as part of the classical presentation of paralytic polio while others do not consider them to be important clinical features. These are fasciculations, and symptoms and signs of muscle tenderness and they were included because of their potential relevance to the interpretation of the overall clinical picture. Due to poor availability of information however,

interpretation of these data is limited and the data are only presented as part of descriptive analyses.

In the interpretation of any clinical data, reasonable conclusions regarding diagnosis require an assessment of the presence, or absence of a combination of variables rather than any single one. The analysis is always enhanced by specific investigations and often rendered inconclusive without them. The investigations reviewed in this study include both polio-specific investigations (e.g., viral isolation from stool and pharyngeal specimens, and poliovirus-specific serological tests) and other non-specific investigations including protein and leucocyte examinations of CSF, and neurophysiological tests consisting of both nerve conduction studies and electromyographic tests. Data on CSF protein level and leucocyte count are particularly helpful towards the diagnosis of GBS when an acellular rise of total protein is demonstrated, a finding specific to GBS and otherwise known as albuminocytological dissociation.<sup>15</sup> For the neurophysiological tests, data were analyzed collectively after separate evaluation of results with the assistance of the neurology consultant. Other investigations for which data were collected to help evaluate other possible causes of an acute paralysis were radiographic tests and serum creatinine kinase assays.

The forms were pretested through an initial review of five charts from the Riverside Hospital (the first hospital to grant approval for chart review). Subsequent modifications consisted of expanding (a) the list of symptoms and signs, and (b) date variables for investigations, where appropriate, to include both dates of collection, and examination of specimens. Data from the eligible charts used in the pretest were supplemented by a second review. Also a decision was taken to collectively assess investigations during data analysis; during the review of charts no decision was made on the degree to which results supported or excluded polio.

### **3.3.3 Data Collection**

All data collection was performed by the author. Patient charts were almost exclusively written in English with the exceptions listed below; translations were provided by bilingual staff in the local medical records departments.

1. Two charts at Montfort Hospital had discharge notes written in French but with admission notes and some of the progress notes in English.
2. Approximately 10% of charts at the Ottawa General Hospital had referral notes in French.
3. Approximately 10% of charts at the Ottawa General Hospital and the Children's Hospital had some daily progress notes in French.

#### **3.3.3.1 Availability of Information**

Although a positive symptom or sign is generally included in the clinical notes of a patient, this is not always the case when symptoms and signs are absent. A physician's decision to note a symptom or sign as negative is influenced by the knowledge (or presumption) of its relevance to the clinical picture presented. During data collection, symptoms and signs were recorded as absent only when categorically noted as negative. When the information was missing from the chart, the variable was coded as "unknown". For specific variables used in subsequent analyses the assumption was made that a lack of information in the clinical record was equivalent to a negative finding.

#### **3.3.3.2 Validity and Reliability**

Validity was addressed from the point of view of the comprehensiveness of the data collection form because this chart review consisted essentially of transcribing pre-recorded

data. The validity of conclusions arising from the review is influenced largely by the comprehensiveness of the review form which is in turn reflected by the following questions:

1. Do the items on the review form correctly reflect the complete information needed to assess the appropriateness of a stated diagnosis, or an alternative diagnosis?
2. Does the design of the review form permit accurate documentation of signs and symptoms as recorded in the charts?

By consulting with a neurologist in the development of the form and pretesting the form, lapses in comprehensiveness were recognised very early and corrected. Items on the form include not only the direct clinical features of paralytic polio, but also all other important features that would support the other diagnoses under consideration. This ensured that both sides of an argument over diagnostic conclusions are well represented to the extent that the information is available. Although it was necessary to transcribe the relevant data in a well summarised format, steps were taken to avoid loss of information by including data on the specific features, as well as related time periods. Also, subjective data from the patient histories were supplemented by both clinical signs and investigations. It is acknowledged that the validity of the review is limited by the availability and accuracy of information in the clinical charts. However, in view of the overall comprehensiveness of the data, I consider the validity of the review to be high to the extent that is possible with a retrospective approach.

Data collection was repeated for a total of 14 charts in order to measure the reproducibility or reliability of the transcription process. These charts were randomly selected from the three hospitals with the highest numbers of charts (the Civic and General Hospitals and the Children's Hospital) and represent 15-20% of each hospital's eligible charts. The reliability of data collection was measured as the percentage agreement between data collected from each chart on the two separate occasions. Substantially high levels of agreement were

obtained with a mean level of 96.2% (range from 91.3% - 98.8%). The interval between the first and second reviews ranged from 2 to 11 days with a mean of 6.1 days.

### **3.3.4 Data Handling and Analysis**

Variables corresponding to individual items on the review form were entered into a database using the PC version of the Statistical Package for Social Scientists (SPSS/PC+). A second generation of variables, referred to as score variables, were used to compute scores for analysis (see Appendix H for algorithms). Score variables included single variables in the original database as well as composite variables derived from the original ones. Analyses consisted of (a) descriptive analyses including the availability of information in the charts, (b) screening of all cases to identify possible cases of missed polio, and (c) comparative analyses for subsets of the total sample defined by the screening procedure. A possible case of missed polio was defined on the basis of polio-compatible features described in the following screening phases.

#### Screening phase 1

Initial screening was based on individual symptoms and signs listed in subsection 3.3.2. This screening was performed for the total sample of charts.

#### Screening phase 2

The second phase of screening was based on variables or variable combinations reported with a high likelihood ratio for polio in the study by Dietz et al. Because these authors used data for children below 15 years of age only, screening in this phase was performed for the total sample and then for a group consisting of the age group 0-14 years. Although the two procedures result in the identification of the same cases, the proportions identified differ. The five variable combinations used were:

1. Proximal paralysis and fever at onset (PLR = 9.0)
2. Unilateral and proximal paralysis (PLR = 9.6)
3. Unilateral and proximal paralysis, and fever at onset of paralysis (PLR = 12.0)
4. Proximal paralysis, installation of paralysis in less than 4 days, and fever at onset of paralysis (PLR = 12.0)
5. Unilateral and proximal paralysis, and installation of paralysis in less than 4 days (PLR = 12.0)

### Screening phase 3

The variables used in this screening phase were identified by Andrus et al with odds ratios ranging from 1.7 to 14.0 for culture-confirmed polio cases compared with other cases of AFP. This study was also based on data for children below 15 years of age with AFP and, age below 6 years was reported as a significant predictor of polio. For that reason only cases less than 15 years were included in the third phase of screening. The variables are:

1. Installation of paralysis in less than 4 days (OR = 1.7)
2. Prodromal digestive symptoms (OR = 2.2)
3. Prodromal fever (OR = 3.2)
4. Fever at onset (OR = 5.1)
5. Age less than 6 years (OR = 9.3)
6. Neurological sequelae (OR = 14.0)

Cases identified by the screening procedures as possible cases of paralytic polio were compared to the remaining cases on (a) admitting diagnoses, (b) the range of tests ordered, (c) the results of those tests, and (d) final diagnoses. The two subgroups were also compared with respect to the presence of clinical features not supportive of polio.

### **3.4 Results**

It should be noted that the results of comparative analyses presented indicate an inadequate sample for this study as reflected by the wide confidence limits. Issues of statistical power and sample size are discussed under the limitations of the study (subsection 3.5).

#### **3.4.1 Sources of Data**

Of 160 charts retrieved, 59 (37%) were found to be ineligible (see Table 7); they included three cases with a history of polio several years in the past. Not surprisingly, by far the largest number of ineligible charts, approximately 63%, were in the category of ICD-9 code 323.9. This non-specific code used for transverse myelitis also includes many disease categories that do not fit into the more specific subcategories of 323. The most frequent reason for ineligibility was an irrelevant diagnosis (67.8% of ineligible charts). Other reasons were related to chronic cases or readmissions with (a) an irrelevant diagnosis at incident admission (23.7%), or (b) an eligible incident admission but unavailable medical charts (8.5%).

A total of 101 eligible charts (63% of all charts retrieved) were used for actual data collection (Table 8). Approximately 84% of the eligible cases were diagnosed as GBS (ICD-9 code 357.0) while 14% were transverse myelitis (ICD-9 code 323.9). There was one chart with a diagnosis of monoplegia of the upper limb (ICD-9 code 344.4) and one with "viral poliomyelitis syndrome" (ICD-9 code 323.2).

Table 7

Distribution of ineligible charts by hospital and ICD-9 code

ICD-9 code	Diagnostic term (ICD-9)	CHEO	MFH	OCH	OGH	QCH	RH	Total
005.1	Botulism	0	0	0	0	0	2	2
323.2	Poliomyelitis	1	0	0	0	1	0	2
323.9	Unspecified encephalitis, myelitis, and encephalomyelitis	4	1	16	12	1	3	37
344.3	Monoplegia of lower limb	2	0	1	0	2	0	5
344.4	Monoplegia of upper limb	0	0	0	0	0	0	0
344.5	Unspecified monoplegia	1	0	0	0	0	0	1
357.0	Guillain Barré syndrome	0	0	5	2	0	0	7
045.9	Unspecified acute poliomyelitis	0	0	1	0	0	0	1
344.6 <sup>a</sup>	Cauda equina syndrome	2	0	0	0	0	0	2
345.2 <sup>a</sup>	Petit mal status	1	0	0	0	0	0	1
345.9 <sup>a</sup>	Unspecified epilepsy	1	0	0	0	0	0	1
Total		12	1	23	14	4	5	59

<sup>a</sup> These codes were not included in the selection list.

Table 8

Distribution of eligible charts by hospital and ICD-9 code

ICD-9 code	Clinical diagnosis	CHEO	MFH	OCH	OGH	QCH	RH	Total
323.2	Viral polio syndrome	1	0	0	0	0	0	1
323.9	Transverse myelitis	5	0	6	2	0	1	14
344.4	Monoplegia of upper limb	0	0	1	0	0	0	1
357.0	Guillain Barré Syndrome	12	5	33	24	10	1	85

### 3.4.2 Availability of Information

A comparison of recorded information on signs and symptoms showed that on the whole information was more readily available for signs than for symptoms (Tables 9 and 10). No single symptom was recorded in all charts; limb paralysis was the symptom with the highest recording frequency at 95% while the remaining symptoms were recorded at frequencies ranging from 8.9% to 79.2%. Less than 9% of charts yielded information on the risk of poliovirus infection through contact with an OPV vaccinee or travel to an endemic region.

Conversely, of the 14 signs listed, information was recorded in all charts for 3 signs and in at least 95% of charts for 4 other signs. The remaining signs had recording frequencies ranging from 11.9% to 76.2%. Although 87.1% of charts had a record of hyporeflexia or areflexia, further analysis showed that only 17.8% of those cases had this feature in the paralysed limb(s) only. This means that for the rest the sign was not compatible with polio. Information for the variables used to derive scores was available in at least 60% of the charts.

Table 9

## Frequency of symptoms reported by AFP cases

Symptom	Percentage of charts (N = 101) with symptom		
	recorded as:		unrecorded
	Present	Absent	
Prodromal fever	43.6	19.8	36.6
Respiratory infection	50.5	13.9	35.6
Headache	19.8	25.7	54.5
Neck stiffness	1.0	8.9	90.1
Vomiting/Diarrhoea	23.8	43.6	32.7
Constipation	6.9	38.6	54.5
Rashes	3.0	10.9	86.1
Limb paralysis	88.1	6.9	5.0
Muscle pain	31.7	9.9	58.4
Joint pain	7.9	6.9	85.1
Trauma	2.0	14.9	83.2
Bladder involvement	10.9	43.6	45.5
Numbness/paraesthesia	61.4	17.8	20.8
Cranial nerve involvement	17.8	51.5	30.7
Contact or travel risk <sup>a</sup>	1.0	7.9	91.1

<sup>a</sup> Although not a symptom this has been included here because information relevant to it is usually obtained, along with symptoms, as part of the patient history.

Table 10

Frequency of clinical signs recorded at time of admission for AFP cases

Clinical sign	Percentage of charts (N = 101) with sign		
	recorded as:		unrecorded
	Present	Absent	
Fever	4.0	93.1	3.0
Unconsciousness	0.0	100.0	0.0
Generalised rashes	1.0	10.9	88.1
Meningism	0.0	76.2	23.8
Muscle tenderness	4.0	11.9	84.2
Arthralgia	3.0	14.9	82.2
Limb paralysis	88.1	11.9	0.0
Cranial nerve paralysis	29.7	70.3	0.0
Respiratory paralysis	17.8	77.2	5.0
Sensory loss	41.6	55.4	3.0
Fasciculations	1.0	12.9	86.1
Loss of sphincter control	5.9	9.9	84.2
Hypotonia	18.8	40.6	40.6
Areflexia/hyporeflexia	87.1	11.9	1.0

### 3.4.3 General Patient Profile

The breakdown of residence status for cases reviewed was 62% from the Ottawa-Carleton Regional Municipality, 23% from other counties and districts of Southern Ontario, 14% from Northern Ontario, and 1% from Quebec.

The mean age was 43 years with a range of 2 to 88 years; approximately 15% of the cases reviewed were below 15 years while 40% were 50 years or older (see Figure 8). Overall there were 51 males as compared to 50 females, with no significant difference in gender distribution by age group;  $X^2 [8 \text{ df}] = 2.97, p = 0.94$ . GBS was diagnosed in all age groups with a substantial number of cases aged 60 years or more (36.5%). In spite of the small number of transverse myelitis cases the general observation is that the relative proportion of transverse myelitis cases above 50 years is lower than that for GBS cases. Also there were no cases of transverse myelitis below 10 years (Figure 9).

Using limb paralysis and cranial nerve involvement in that order as the reference point, the mean interval from onset of paralysis to admission was estimated as 7.7 days. The duration of admission ranged from less than 24 hours to 172 days with a mean of 28 days. One case died during admission, four of the remaining were transferred to another hospital (i.e., other than the hospital from which records were obtained) for continuing clinical management, and 94 were discharged home or to a rehabilitation unit. For two records, there was no available information about the type of discharge.

Residual paralysis lasting 60 days or more from the onset of paralysis was documented for 14 cases whereas 50 cases had no residual paralysis at the time of discharge. For 36 cases this information was lacking because (a) there was no record of the state of paralysis at discharge or just prior to discharge, and there was no follow-up information, or (b) discharge was within the first 60 days and there was no follow-up information.

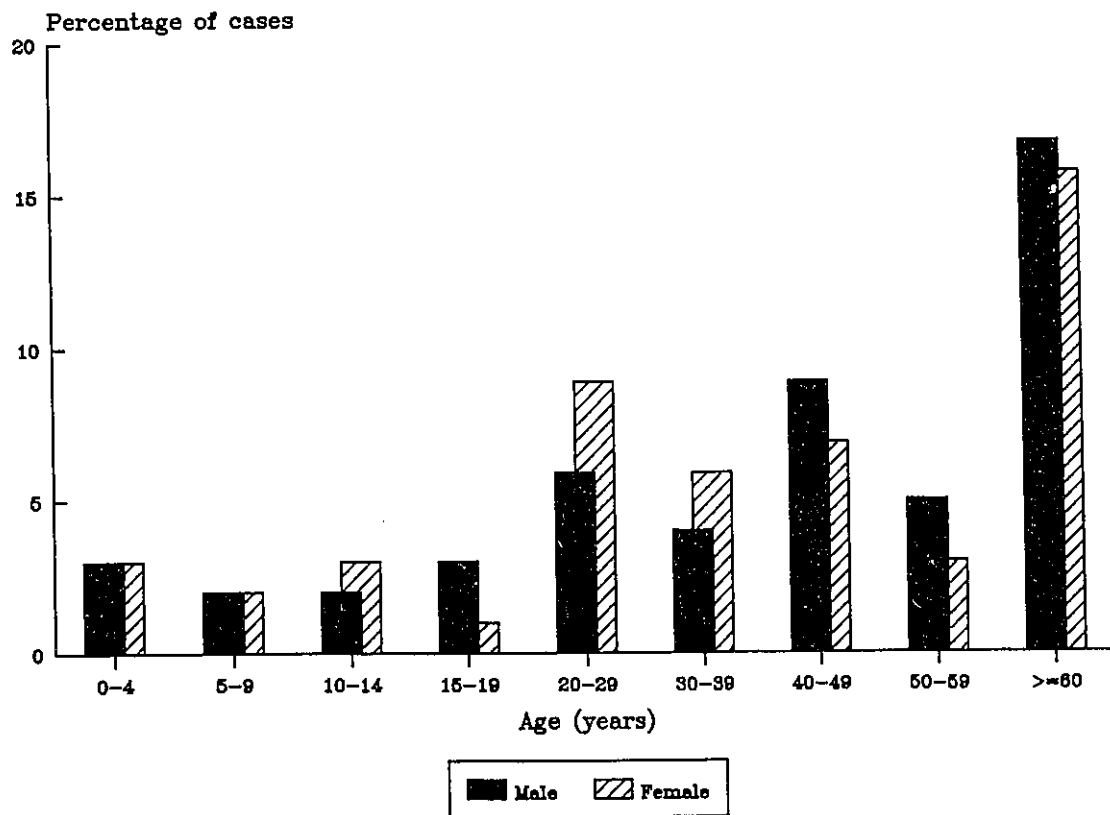


Figure 8 Age and sex distribution of AFP cases in Ottawa-Carleton, 1986 - 1991.

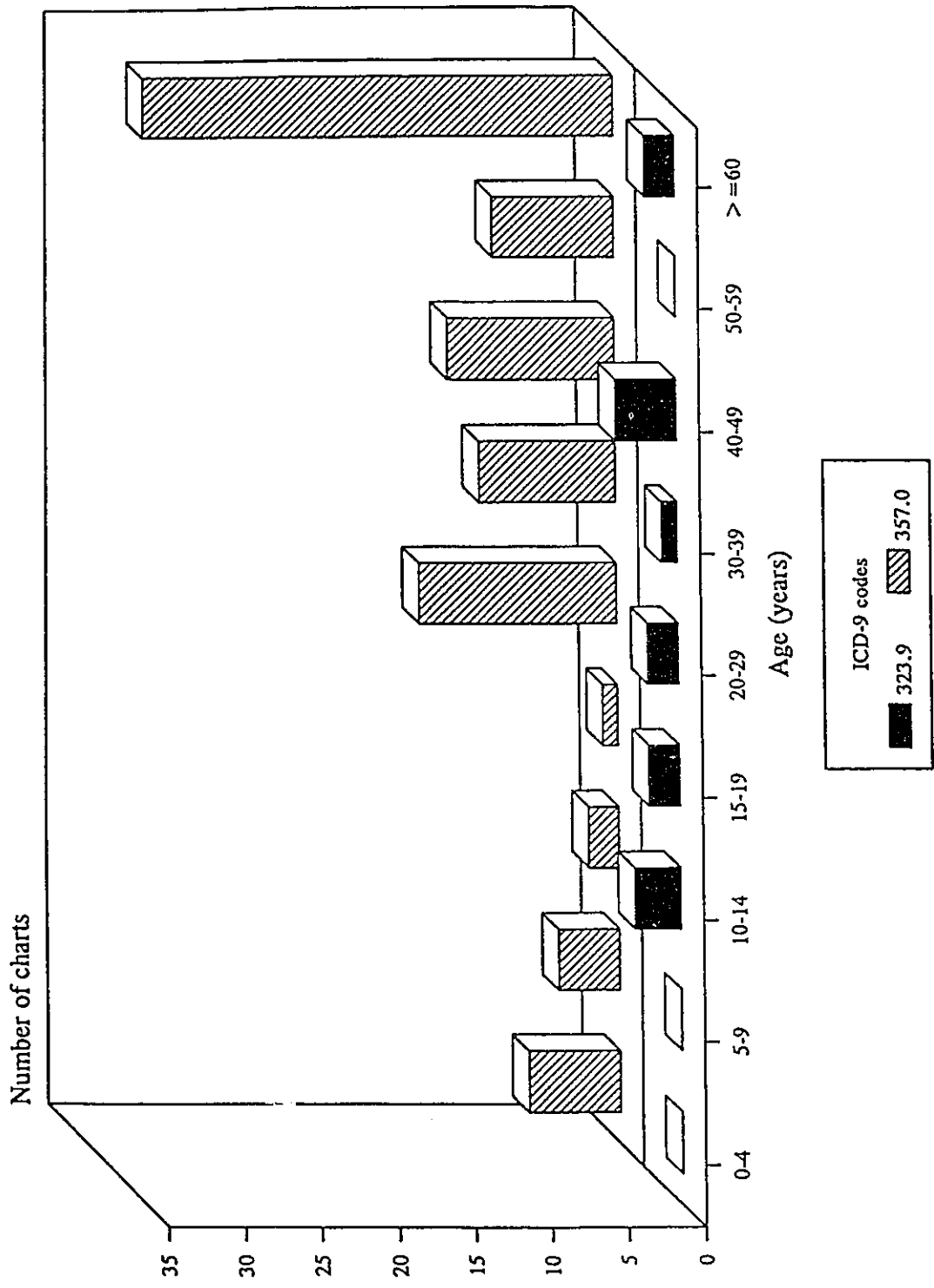


Figure 9 Age distribution of AFP cases in Ottawa-Carleton by ICD-9 code, 1986 - 1991.

### 3.4.4 Physician Care and Public Health Notification

The physician categories involved in patient care varied in frequency according to the role of the physician (i.e., referring, attending or consulting). The data for referring physicians were intended to reflect the types of physician specialty outside of the admitting hospitals who had initial contact with AFP cases. Emergency room physicians were excluded from the referring group as almost all such cases would normally be admitted through the emergency room. The major categories of referring physicians were family and general practitioners, and internists (Table 11). It was not possible to determine from available records what the specialty was for approximately 29% (14) of referring physicians.

Table 11

Categories of physicians involved in care of AFP cases<sup>a</sup>

Physician specialty	Number of cases with physician:		
	Referring <sup>b</sup>	Attending <sup>c</sup>	Consulting <sup>c</sup>
Internal medicine	7	11	31
Paediatrics	1	3	1
Neurology	4	82	29
Family/General practice	17	1	0
Infectious disease	0	1	1
Other	6	4	2
Unknown	14	1	0

<sup>a</sup> Limited to physicians involved directly in clinical care.

<sup>b</sup> Excludes referral by emergency room physicians.

<sup>c</sup> Indicates at least one physician in that category; some cases had none while others had more than one

Eighty-two cases were attended by a neurologist and 11 by a general internist. Twenty-nine cases were seen in consultation by a neurologist; 11 of these were consultation requests from another neurologist who was the attending physician while the remaining 18 were requests from attending physicians in other categories. Only one case was not attended or seen in consultation by a neurologist. The data presented for consulting physicians are only counts of cases seen by a particular specialty and not the actual number of physicians involved in patient care; some cases received consultations with more than one physician in a given specialty. The involvement of infectious disease specialists in patient care was extremely limited for these cases. Few other categories were involved directly in patient care although data were not collected on consultations with certain specialties including radiology, physical and speech therapy, and respirology.

There was only one record of a communicable disease report being issued for a patient with positive serology tests for *Mycoplasma pneumonia*. It was not possible to ascertain where the report was sent, or the indication for sending it. No other records provided information on notification of public health authorities.

#### **3.4.5 Screening for Possible Cases of Missed Polio**

The frequency of occurrence for variables used in each of the screening procedures is presented in Table 12. Also included are a group of six features non-supportive of polio. Overall, the proportion of cases with polio-supportive features was comparable for Phases 1 and 2 in the total sample, and for Phases 2 and 3 for the younger subsample.

##### Phase 1: Based on clinical texts

Approximately 21% of cases had none of the six polio-supportive features used in screening while another 19% had at least three of the features. Conversely, all cases had at least one

of the features that would tend to rule out polio but less than 9% had three or more of the six features. Generally, more cases had polio-supportive features than features that tend to rule out polio.

Phase 2: Based on Dietz et al.

Approximately 27% of the total sample of cases had none of the features with a high PLR for polio while 23% had at least two of the four. The proportions in the total sample are comparable with those obtained in Phase 1 but a much higher proportion of cases below 15 years (67%) had at least half of the polio-supportive features.

Phase 3: Based on Andrus et al.

The proportion of cases with at least half of the polio-supportive features was moderately high with the inclusion of prodromal fever (47%); the reduced frequency of fever at admission partly accounted for the lower proportion (27%) obtained in the second run of this screening phase. Also, the lower proportion of cases with polio-supportive features when compared with Phase 2 is likely to be partly due to the inclusion of the variable, "age below six years"; only eight cases met this criterion.

A total of 35 cases were identified as possible missed cases of polio by one or more of the 12 scores derived for screening. The group of cases identified through screening as possible missed cases of polio (n = 35) was designated as Group A and the remaining cases (n = 66) designated as Group B. Group A cases are presented in a matrix with identifying score variables (see Appendix I). With regard to the consistency of identification, 26 cases were identified in one screening phase only and 9 cases were identified in two phases.

Twelve cases in Group A were only identified on the basis of a history of prodromal fever, respiratory infection, and gastrointestinal symptoms. These symptoms are also frequently

associated with GBS so the accuracy of identification for these cases may be questioned. Nonetheless, the cases were retained in Group A because of the interest in further analyzing subsequent investigative procedures.

#### **3.4.5.1 Demographic Profile of Cases by Groups**

In Group A, 12 cases (34.3%) were below the age of 15 years and 9 cases (31.4%) were 50 years and above. Three cases (4.5%) in Group B were below the age of 15 years whereas 30 cases (45.4%) were 50 years and above. Cases in the younger age groups were found to be significantly more likely to be identified as possible cases of polio ( $p < 0.001$ ). Two of the missed polio cases were eliminated from this last analysis as they were only identified by score variables based on age; all other cases identified by those variables were also identified by one or more other scores. 40% of Group A cases were males compared to 56% of Group B cases.

#### **3.4.5.2 Admitting and Discharge Diagnoses by Groups**

The majority of cases (68%) were assigned a single diagnosis at admission with no differential diagnosis (Table 13). The most frequent admitting diagnosis was GBS; 54% of cases in Group A and 68% in Group B. Polio was considered a possible diagnosis for two cases in each group at admission; a specific diagnosis of polio was recorded as the first differential diagnosis for both of the Group A cases (aged 14 and 60 years) and the second differential for the two cases in Group B (aged 7 and 80 years). For the two Group B cases, an enterovirus-related neuropathy was recorded as the primary diagnosis in one and as the first differential in the other. These four cases were all diagnosed as GBS at discharge and they were all attended by a neurologist. Overall, the relative proportions of admitting diagnoses were similar in Groups A and B.

The discharge diagnoses in Group A consisted of 27 cases (77.1%) of GBS, 7 cases (20.0%) of transverse myelitis and 1 case (2.9%) of viral polio syndrome. In Group B, the discharge diagnoses were 58 cases (87.9%) of GBS, 7 cases (10.6%) of transverse myelitis and 1 case (1.5%) of a monoplegia of the upper limb. There was no significant association between compatibility with polio (a case fitting in Group A or Group B) and the likelihood of being diagnosed as GBS or transverse myelitis ( $X^2$  [1 df] = 1.06,  $p$  = 0.30).

### **3.4.5.3 Analysis of Diagnostic Tests by Groups**

Generally, there were no statistically significant differences between the two groups with respect to either the probability that a specific test was done, or when it was done. The most common test in both groups was CSF analysis of protein content and cells; 77.1% in Group A and 83.3% in Group B (Table 14); also CSF analyses were among the earliest tests done with mean intervals ranging from 1.8 to 4.1 days from the date of admission. Of the 27 GBS cases in Group A, 22 had both CSF protein and CSF leucocytes documented but only 5 cases showed an albuminocytologic dissociation supportive of the diagnosis.

Although on the average serology was done earlier than CSF examinations, it was documented for fewer patients and less than 7% of all cases had two serology tests. More importantly, there was no documentation of polio-specific serology results. For some charts a polio-specific request was documented with a subsequent note from the laboratory indicating a lack of the service in the hospital. None of these charts indicated whether another sample was sent to the Regional Enterovirus Laboratory as recommended by the local laboratory. On the average, the interval between acute and convalescent serology was slightly less than the usual recommendation of at least 14 days.

Approximately 11% of Group A cases, and 9% of Group B cases had a stool culture for poliovirus but a second stool culture was documented in only two Group B cases and none of Group A. The first stool samples were tested at a mean interval of about 7.5 and 5.8 days after admission for Groups A and B respectively. Viral isolation from throat specimens was attempted in 8.6% and 4.5% of Group A and Group B cases respectively. All the results were negative for polio.

A similar percentage of cases in Groups A and B (71.4% and 63.6% respectively) had a neurophysiological examination consisting of a nerve conduction test or an electromyogram or both. None of the results were supportive of polio. Three of the polio-compatible cases had neurophysiological findings compatible with GBS, two in association with albuminocytologic dissociation.

Approximately 55% of Group A cases and 49% of Group B cases had other tests documented. These other tests consisted of one or more of serum creatinine kinase, myelogram, CT scan of spine and/or brain, MRI of spine and/or brain, X-ray of spine, CSF protein and CSF leucocytes prior to admission.

In terms of multiple testing, none of four tests selected as basic for poliomyelitis (serology, stool or throat viral culture, and neurophysiological tests) were carried out for 8 cases in Group A (22.9%) while 14 cases (40%) had more than one of these tests (Table 15). In Group B, 20 cases (31%) had none of these tests while 15 (23%) had more than one. There was no significant difference between the two groups with respect to having none, or at least one of these tests ( $X^2$  [1 df] = 0.63,  $p = 0.426$ ). None of the results for these investigations supported a diagnosis of polio. Of 25 cases in Group A with neurophysiological tests, 13 had results supportive of GBS, 7 had inconclusive results, 2 results were normal and 3 suggested other neurological problems.

Of the four cases for whom a differential diagnosis of polio was recorded on admission, one (in Group A) had none of the four tests mentioned above, and two (one in each group) had only neurophysiological tests, both with GBS-compatible results. The fourth case (in Group B) had a neurophysiological examination, and two stool cultures within 16 days of onset of paralysis but with negative results. The tests recorded for the case diagnosed as viral polio syndrome at discharge were a single stool culture and a throat culture 22 days after the onset of paralysis (but 8 days after admission), and both nerve conduction studies and an electromyogram a day after admission.

Finally, there was no significant difference between the two groups with regard to the presence of any one or more of the clinical features that would rule out polio. Eight cases in group A (22.9%) were found to have two or more of the six features (Table 16); seven were diagnosed as GBS and one as transverse myelitis. All eight cases had at least two of three features; sensory loss, symmetrical paralysis, and progression of paralysis after admission. Although these features directly oppose polio-supportive features and therefore would tend to rule out polio only two of the cases had confirmatory results for GBS based on neurophysiological examination. No other charts documented test results confirming the assigned diagnosis although the diagnosis may have been justified by the total clinical presentation at the time.

Table 12

Frequency of screening variables by total sample of AFP cases and selected subsamples

Combinations of clinical features (Type/number of variables)	Percentage of records with:		
	None	All	> = 50%
<b>BASED ON CLINICAL TEXTS WITH TOTAL SAMPLE (N = 101)</b>			
Prodromal fever, respiratory infection, GIT symptoms, asymmetrical paralysis, progression of paralysis in < 4 days, and residual paralysis (Polio-supportive features, n = 6)	20.8	0.0	18.8
Trauma, prodromal rashes, arthralgia, sensory loss, progression of paralysis in < 4 days, and symmetrical paralysis (Non-supportive features of polio, n = 6)	0.0	0.0	8.4
<b>BASED ON DIETZ ET AL. WITH TOTAL SAMPLE (N = 101)</b>			
Fever at admission, monoplegia, proximal paralysis, and progression of paralysis in < 4 days	26.7	0.0	22.8
<b>BASED ON DIETZ ET AL. WITH SAMPLE &lt; 15 YEARS (n = 15)</b>			
Fever at admission, monoplegia, proximal paralysis, and progression of paralysis in < 4 days	6.7	0.0	66.7
<b>BASED ON ANDRUS ET AL. WITH SAMPLE &lt; 15 YEARS (n = 15)</b>			
Age < 6 years, prodromal fever, GIT symptoms, progression of paralysis in < 4 days, and residual paralysis	0.0	6.7	46.7
Age < 6 years, fever at admission, GIT symptoms, progression of paralysis in < 4 days, and residual paralysis	13.3	6.7	26.7

Table 13

## Distribution of admitting diagnoses for AFP cases in Groups A and B

Diagnosis or diagnostic group	Percentage of charts with admitting diagnosis, in order of importance					
	Group A cases (n = 35)			Group B cases (n = 66)		
	1	2	3	1	2	3
None	0.0	68.6	77.1	0.0	68.2	81.8
Poliomyelitis	0.0	5.7	0.0	0.0	0.0	3.0
Guillain Barré Syndrome	51.4	2.9	0.0	63.6	1.5	3.0
Transverse myelitis	5.7	0.0	0.0	3.0	0.0	1.5
Botulism	0.0	0.0	0.0	1.5	0.0	0.0
Viral neuropathy (unspecified)	2.9	0.0	0.0	1.5	0.0	1.5
Enterovirus-related neuropathy	0.0	0.0	0.0	1.5	1.5	0.0
Post-infectious/infectious neuropathy (unspecified aetiology)	5.7	2.9	0.0	3.0	6.0	1.5
Spinal cord lesions (non-infectious)	8.6	2.9	2.9	4.5	4.5	0.0
Toxic or metabolic neuropathy	0.0	0.0	2.9	0.0	3.0	0.0
Myopathies	5.7	2.9	5.7	0.0	4.5	1.5
Non-specific <sup>a</sup>	14.3	8.6	5.7	12.1	3.0	3.0
Other <sup>b</sup>	5.7	5.7	5.7	10.6	7.6	3.0

<sup>a</sup> Includes demyelinating disease, radiculitis, thromboembolic neuropathy, paraneoplastic neuropathy, intracranial space occupying lesion, and sensory neuropathy

<sup>b</sup> Includes mononeuropathy multiplex, multiple sclerosis, lyme disease, sarcoidosis, leukaemia, motor neurone disease, functional myelopathy, genetically determined neuropathies (e.g., Friedrich's ataxia), and systemic diseases (e.g., porphyria)

Table 14

Frequency of common diagnostic tests for AFP cases in Groups A and B

Investigative procedure	Percentage of charts for which test was done		OR	95% CI	Mean interval from admission to test date (days)		p-value <sup>a</sup>
	Group A (n = 35)	Group B (n = 66)			Group A (n = 35)	Group B (n = 66)	
CSF protein	77.1	83.3	0.68	0.22-2.11	4.1	3.8	n.s.
CSF leucocytes	77.1	81.8	0.75	0.25-2.30	4.1	3.9	n.s.
CSF bacteriology	45.7	54.5	0.70	0.28-1.73	2.8	3.1	n.s.
CSF virology	28.6	18.2	1.80	0.65-5.24	3.1	1.8	n.s.
1st serology	37.1	25.8	1.70	0.65-4.50	1.8	2.4	n.s.
2nd serology	8.6	6.1	1.45	0.20-9.13	13.3	16.0	n.s.
1st stool viral isolation	11.4	9.1	1.29	0.25-5.90	7.5	5.8	n.s.
2nd stool viral isolation	0.0	3.0	0.00	0.00-10.07	n/a	50.5	n/a
Throat viral isolation	8.6	4.5	1.97	0.25-15.44	5.8	5.7	n.s.
Neurophysiology	71.4	63.6	1.43	0.54-3.83	4.9	10.3	n.s.

<sup>a</sup> Based on two-tailed t-test

Table 15

Frequency of selected diagnostic tests for AFP cases in Groups A and B<sup>a</sup>

Number of tests done	Percentage of records	
	Group A (n = 35)	Group B (n = 66)
0	22.9	30.3
1	37.1	47.0
2	28.6	15.2
3	11.4	4.5
4	0.0	3.0

Table 16

Frequency of non-supportive features of polio among AFP cases in Groups A and B

Number of features present	Percentage of records	
	Group A (n = 35)	Group B (n = 66)
0	20.0	6.1
1	57.1	34.8
2	14.3	40.9
3	8.6	15.2
4	0.0	3.0

<sup>a</sup> Four tests were selected as basic tests for poliomyelitis; stool and throat viral cultures, serology, and neurophysiological tests

### 3.5 Limitations

There are a number of important limitations to this retrospective chart review. First is the unavailability of some relevant information in the medical charts which relates to other limiting aspects of the study discussed below. In this regard, it is important to note that very few clinicians found it necessary to record (and possibly to enquire) about a possible risk of poliovirus infection through travel to a polio-endemic region, or through contact with an OPV vaccinee.

A basic assumption of the study was that the absence of information for any variable was equivalent to a true negative finding. I felt that this assumption was justified because of the high probability that all relevant positive findings would be recorded however, this may not be completely accurate. Also two variables, "installation of the full extent of paralysis within four days" and "fever at the onset of paralysis" were substituted by proxy variables selected as the closest approximations; "non-progression of paralysis after admission" and "fever at the time of admission". The progression of paralysis after admission was recorded rather inconsistently in patient charts; I collected data for this variable based on the first mention of cessation in progress which varied considerably. This obviously limits the reliability of the variable. Also, as fever is not necessarily stable over time, a temperature recorded at admission may not reflect the temperature at the onset of paralysis (in this sample the onset was on the average 8 days prior to admission). This is especially so for cases who would have started anti-pyretic treatment prior to admission. The use of these variables is associated with the risk of misclassifying cases; both a risk of wrongly identifying cases as possible missed polio and a risk of not identifying possible missed cases as such.

A second limitation is the fact that data were obtained secondary to and therefore, dependent on the accuracy of clinical judgement and recording of clinical notes at the time of admission. There are a number of factors which may influence these clinical procedures, the two most important being the different practices that exist among individual physicians, and the different levels of physician training (e.g., interns, residents, specialists). It is difficult to assess the overall accuracy of the recorded information although it is expected to be high. The comparison of cases in this review assumes similar levels of clinical accuracy although any variability attributable to the noted differences is not accounted for.

Studies of rare diseases are prone to problems of insufficient statistical power for analyses. Although this study was based on all available charts in the selected area, the sample size obtained is associated with extremely low statistical power. As an example, the power to detect a 5% difference (effect size) with 95% confidence (alpha level) between the given sample sizes of Groups A and B for ordering of a stool culture was only 12.3%. Similar power levels were obtained for other comparisons in the study; calculations were based on a formula in a standard epidemiology text.<sup>67</sup> This level of power is grossly inadequate for concluding that noted group differences were not present by chance alone, or conversely that differences not noted were really not present.

With all other parameters unchanged, the required sample size for a power of 80% is 432. Ideally a smaller effect size and higher power level should be used for calculating the required sample size in order to meet the stringent requirements of disease elimination. With a power of 95%, effect size of 1% and alpha level of 1% a sample of 22,404 AFP cases would be required for the same comparison. It would be difficult to obtain such a sample even across Canada for such a study. For the purposes of discussing the public health importance of the issues raised, I consider the magnitude of differences to be more important

than statistical significance. As this study deals with elimination, any minimum difference noted between recommended or expected practices and actual practices is important.

### 3.6 Discussion

GBS is described as the most important cause of AFP in North America<sup>68</sup> with a reported mean annual incidence of 2.07 per 100,000 population in Ontario.<sup>69</sup> Although there are no true estimates of AFP incidence in Canada, on the basis of clinical experience the proportion of AFP cases diagnosed as GBS in this sample had been estimated as more than 75% prior to the review (P. Jacob, personal communication, August 1992). As expected, GBS was the most frequently occurring final diagnosis among those selected for retrieval of charts.

Overall, more than a third of the AFP cases in Ottawa-Carleton in the five-year period reviewed were identified as meeting clinical criteria for the diagnosis of polio. A retrospective judgement on the possibility of polio was not possible in most cases due to a lack of polio-specific investigations. Although the majority of these cases were diagnosed as GBS, it should be noted that the differentiation between GBS and polio may be difficult in the absence of objective findings of (a) poliovirus in stool or throat samples, (b) albuminocytologic dissociation, and (c) neurophysiological results supportive of either diagnosis.<sup>69</sup> Moreover, the isolation of wild polioviruses from stools of persons meeting the clinical diagnosis of GBS (including symmetrical paralysis) has been reported by Yohannan et al<sup>70</sup> who also comment on the difficulty of differentiating the two diagnoses on the basis of clinical features alone. All four cases reported by the previous authors had received at least three doses of OPV prior to their illness, thus prompting the suggestion that the clinical course of polio may be modified by previous immunization with OPV. Also of note is the fact that all the cases had already been diagnosed as GBS and were being managed as such

when virological confirmation of polio was obtained. The report underscores the possibility of occasional atypical presentations of paralytic polio.

Nine of the cases identified as possible polio had signs of cranial nerve paralysis in association with limb paralysis. As previously noted, bulbar or bulbospinal polio is much rarer than pure spinal polio so the probability of these nine cases being polio may be lower. On the other hand, because bulbar paralysis is a rare form of a disease which is itself rare in Canada, it is likely that there would be an even lower index of suspicion for its diagnosis.

Polio-compatible cases and the other AFP cases did not differ with regard to the pattern of investigations. This may be an indication that for those cases in Group A with polio-specific investigations, the investigations might have been ordered routinely. Another possible reason is that a broad spectrum of tests are ordered for cases presenting with atypical features irrespective of the stated differential diagnoses; these data do not present any conclusive evidence for this. Overall, there were no fully convincing results for a diagnosis of polio in any of the cases reviewed but there was also no convincing evidence for the discharge diagnosis in the majority of cases identified as possible cases of missed polio.

It is important to note that of the four cases for whom polio was considered as a possible diagnosis during admission, only one was adequately investigated for polio. In addition, there was no indication that any of these cases was reported to the public health authorities. These findings indicate a possibility that there was no strong conviction of the diagnosis, or knowledge of appropriate diagnostic tests. Even for the case of viral polio syndrome, this procedure was not adhered to although the retention of the diagnosis (despite the lack of conclusive investigations) suggests some clinical conviction of a poliovirus infection. The lack of notification may be attributable to a low level of awareness among physicians about reporting requirements.

The high involvement of neurologists in patient care tends to support the expectation that AFP cases in the study region would be managed by neurologists. The extremely low involvement of infectious disease (ID) specialists is notable especially for the four cases with polio considered as a differential diagnosis, none of whom received an ID consult. A reason proposed for this finding is that ID consults are often sought only in the acute phase of an infectious disease and only when antibiotic therapy is required. Thus, for suspected non-acute viral infections the average physician would not consider it necessary to seek such a consult (P. Jacob, personal communication, June 1993).

In spite of the acknowledged limitations of the study, these data suggest a low index of suspicion for paralytic poliomyelitis in Ottawa-Carleton together with poor application of both the recommended investigative practices for polio, and the associated reporting requirements. Furthermore, these data do not provide convincing evidence that polio cases have not been missed in the past although none of the polio-compatible cases can be confirmed retrospectively.

The study design does not permit extension of these conclusions to national data. Nonetheless, the findings presented here suggest that the initial investigation of AFP cases may not provide a high probability of identifying a case of polio should one occur. Implementation of an effective AFP surveillance program in Canada is therefore justified, even if difficult to implement. In the short-term, the process of initiating such surveillance could lead to a higher index of suspicion for polio, and improve the frequency of specific testing thereby increasing the quality of data that can be used as evidence to rule out polio. A national retrospective chart review is however not considered to be feasible as the quality of information would most likely be insufficient for definite conclusions and would not justify the resources required for its implementation.

## **CHAPTER 4          PHYSICIAN SURVEY**

The final part of the thesis was a cross-sectional survey of selected categories of physicians practising in the Ottawa-Carleton region. This physician survey was conducted as a pilot study to explore the need for a national survey to study a) the current level of awareness among physicians in Canada about the presentation of paralytic poliomyelitis and b) a need for ongoing education of these physicians about the epidemiology of paralytic poliomyelitis and the process of documenting polio elimination in Canada. Also, the overall results were compared with the more objective findings of the chart review.

### **4.1                  Aims and Objectives of the Physician Survey**

The specific objective of the physician survey was:

To assess the current approach to, and subsequent management of AFP cases in Ottawa-Carleton with particular respect to the diagnosis of paralytic polio.

In meeting the stated objective, the questions raised were:

1.      What is the level of diagnostic accuracy for paralytic poliomyelitis among physicians in Ottawa-Carleton who would see such cases?
2.      Among the same group how adequately would a case of paralytic polio be investigated to confirm the diagnosis?

### **4.2                  Methods**

#### **4.2.1              Survey Population**

In broad terms, all physicians in the Ottawa-Carleton region with the potential to treat, either individually or as part of a team, a patient presenting with an acute, non-traumatic paralysis

were considered potentially eligible for the survey. Although the initial presentation of such patients is directly relevant to their subsequent management, I decided to focus on physician judgements and behaviours that would lead to a definitive diagnosis. Usually, such judgements and behaviours are not limited to the time of first contact with the patient but rather occur over a number of days following initial presentation. Therefore, the survey was limited to categories of physicians considered to have a "primary" role in AFP case-management which was defined with respect to the physician's role in the complete investigation of a case leading to a final diagnosis. The physician categories fitting this role are those most likely to be attending a case, or to be consulted for an opinion by the latter.

An informal survey of a cross-section of physicians in the study region yielded some valuable information on the general patterns of physician practice. The information indicated that generally paediatricians, family physicians and general practitioners would be unlikely to continue with the management of an AFP case beyond the first 48 hours; instead a referral to a neurologist was the expected norm. On the contrary, the observed practice had been that general internists very often maintained their initial role in the management of AFP cases, seeking consultation with neurologists when appropriate. On the basis of the points raised above, the survey was limited to neurologists, general internists and infectious disease specialists.

Eligibility status for inclusion in the survey was defined as a physician with:

1. Full training as a neurologist (paediatric or adult), general internist or infectious disease specialist; physicians undergoing residency training in these selected categories were excluded.
2. An active practice in Ottawa-Carleton during the study period.

3. A clinical practice not limited to (a) a subspecialty for which the survey material would not be directly relevant (e.g., cardiology) or (b) specific diseases of interest (e.g., Parkinson's disease).

A mailing list of "eligible" physicians was obtained on request from the Ontario Medical Association (OMA). This list consisting of 133 general internists, 41 neurologists, and 7 infectious disease specialists was subsequently reviewed for verification. Verification was particularly necessary for physicians designated as general internists as a number of them were proven to be actually sub-specialists. Verification of eligibility was more difficult for physicians whose practice was limited to a specific disease of interest as often this information was not accessible through the general sources used. The verification procedure was based on information obtained from a combination of sources:

1. The current physician listing in the Canadian Medical Directory (1993 edition).
2. Information from the Chief of Medicine (the same individual) of the Ottawa Civic Hospital and the Ottawa General Hospital, the two hospitals to which the majority of general internists are affiliated.
3. The current physician listing in the local telephone directory (January 1993-1994 edition).
4. Telephone calls to physicians' offices. This last step was only employed when the information obtained in the first three steps failed to clarify a physician's specialization.

Through the verification procedure described, approximately 67% of the physicians listed by the OMA were identified as ineligible for this survey; 71% of general internists, 61% of neurologists, and 29% of infectious disease specialists. Seven more physicians were excluded from the survey, despite their eligibility, because of their prior knowledge of the survey through one or more of the following:

1. An involvement in the process of questionnaire development.
2. Participation in a review of the study proposal as part of a particular hospital's approval process for the review of hospital charts (see subsection 3.3).
3. A consultation for verification of the physician list (this was the Chief of Medicine of the Ottawa Civic and General Hospitals).

The final mailing list consisted of 67 physicians; 43 general internists, 19 neurologists, and 5 infectious disease specialists. Only four physicians were identified as being in paediatric practice (four other physicians in paediatric practice were excluded because of prior knowledge) while the rest were all adult physicians.

#### **4.2.2 Data Collection**

I developed a questionnaire for data collection in consultation with the study neurologist and an internist with longstanding experience in the development of case scenarios for medical education and research. The questionnaire was based on hypothetical written case scenarios which were further reviewed by two other practising neurologists after pretesting.

Written case scenarios are often used to examine clinical decision-making and other aspects of physician behaviour and their use in the health sciences for a variety of research questions is extensively reported. Jones et al<sup>71</sup> in reviewing the validity of written case scenarios, identified 74 published articles based on written case scenarios (from 1963 to 1987). The authors however, point out that written case scenarios are known by a variety of names in different formats. Thus, the number of articles identified in their search is probably an underestimate.

There are two major criticisms against case scenarios: (a) they may include selected aspects of reality while neglecting others, and (b) physicians may not respond the same way to the cases as they would in real life.<sup>71-73</sup> The latter criticism relates to the "demand effect" a term used to describe the likelihood of physicians responding on the basis of their concept of social desirability.<sup>71</sup> In addition, written cases are described as lacking in visual cues which may be an important determinant of physician behaviour.<sup>71</sup> On the other hand, written cases have the advantages of (a) the ability to control disease and patient factors by having the study participants review a defined set of cases, and (b) the relative simplicity and low expense of studying large numbers of subjects.<sup>71,73</sup> The intent of this survey was to examine clinical judgements relating to diagnosis and investigations. Therefore, the case scenarios in this study were constructed to exclude details of diagnostic tests. This also ensured elimination of the "cueing effect" which arises when physicians, prompted by response options in a written case, report behaviours at a higher frequency than they would apply in real life.<sup>71</sup>

Two sets of case scenarios were developed for use in the survey instrument (see Appendix J for the complete write-up of the cases). One set consisted of four cases of paralytic polio which served as the test cases. The polio cases were based on actual paralytic polio cases reported in Canada in 1988 and 1989<sup>35,36</sup> with some modifications. Two of the cases were constructed as paediatric cases and the other two as adult cases. In addition, two of the polio cases (one paediatric and one adult) were constructed to create a higher index of suspicion for the diagnosis although caution was taken to avoid excessively leading information. These two cases were designated as "high sensitivity" cases, and the other two as "low sensitivity" cases.

Specifically, the high sensitivity cases both included symptoms of prodromal fever and diarrhoea, with confirmation of fever at examination. The paediatric case also had a history

of recent immigration from a polio-endemic country and a history of receiving immunizations in the previous month. A comparable factor in the adult case was a history of working in a day care nursery. These latter points were included to indicate possible contact with the vaccine-type polio virus. The high sensitivity cases also had a clear presentation of unilateral paralysis which is typical of polio.

The low sensitivity cases had complaints of weakness unilaterally however, the findings on examination included a milder weakness in a second limb. Although bilateral limb involvement is a less common finding in polio, by including the quality of asymmetrical paralysis (based on different power grade levels in the affected limbs) the validity of a diagnosis of polio in these cases is retained. The low sensitivity cases also had a more remote suggestion of contact with the vaccine-type virus in the family information which included the presence of a five-month old baby in both cases. As the recommended age for the second dose of OPV is four months, selecting an age of five months still allows for the maximum interval of 60 days between the time of OPV administration and contact between a suspected case of poliomyelitis and the vaccinee.

The second set of cases consisted of three control cases based on typical presentations of GBS, transverse myelitis, and botulism. All three control cases involved teenagers so the need for separate adult and paediatric cases for each was obviated.

The case scenarios constructed are summarised as follows:

1. Paralytic polio: (a) two high sensitivity (HS) cases, one paediatric (Case 1) and one adult (Case 2); and (b) two low sensitivity (LS) cases, one paediatric (Case 4) and one adult (Case 3).
2. GBS, Transverse myelitis and Botulism: one typical case scenario of each (Cases 5, 6 and 7 respectively).

Each of the seven case scenarios was followed by a standard set of questions relating to (a) diagnosis, (b) investigations, and (c) consultation with other physicians. Each questionnaire consisted of three case scenarios; one each of a high sensitivity and a low sensitivity polio case, and a third case, the control case. The possible combinations of case scenarios in a questionnaire, designated as sets A to F, were:

Set A: Cases 1, 4 and 5

Set D: Cases 2, 3 and 5

Set B: Cases 1, 4 and 6

Set E: Cases 2, 3 and 6

Set C: Cases 1, 4 and 7

Set F: Cases 2, 3 and 7

In addition to the specific questions relating to the cases, respondents were asked to indicate the frequency with which they saw similar cases in their clinical practices and also to indicate their specialties (see Appendix J for example of questionnaire<sup>a</sup>).

The questionnaire was pretested on a group of physicians (located in another province) similar to the target group with respect to their specialization. The pretest survey differed slightly from the actual survey as the questionnaires were distributed by a local physician who was also responsible for obtaining the completed questionnaires. Also, the physicians who were surveyed were selected by convenience. Six physicians agreed to participate in the pretest and five completed questionnaires were received. The responses provided (and additional comments in some cases) formed the basis for further modification of the cases. Questionnaires based on the modified cases were subjected to a final review by two local practising neurologists (different from the neurologist involved in developing the form).

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<sup>a</sup> The example includes two subsequent modifications which are discussed in the context of follow-up in subsection 4.2.3

### 4.2.3 Survey Design

Questionnaires based on sets A, B and C (with paediatric cases) were randomly distributed to the four paediatric physicians and the rest were also randomly distributed to the adult physicians. In order to identify respondents and thereby exclude them from further follow-up, identical code numbers were assigned to each physician's name on the mailing list, and the respective questionnaire mailed to that physician. A cover letter briefly outlining the purpose of the study, the rationale for coding, and the expected return date was mailed with the questionnaire. The cover letter also provided a telephone number at which the author could be reached for information regarding the survey, and was jointly signed by the author and one of the thesis supervisors (I. McDowell). A pre-stamped return envelope addressed to the author was provided with the questionnaire.

The survey design was based on the Total Design Method (TDM) proposed by Dillman<sup>74</sup> who recommends a follow-up sequence comprising (a) a postcard reminder one week after the initial mailout, (b) a letter and replacement questionnaire to non-respondents three weeks after the initial mailout, and (c) a final mailing by certified mail, with another replacement questionnaire, seven weeks after the initial mailout. In this survey, a combination of telephone and mailed reminders was employed with the anticipation that the personal interest generated by the telephone reminders would enhance the response rate. Also, it was anticipated that contact with potential respondents by phone would provide an opportunity to answer questions about the survey. Therefore, the follow up process consisted of five steps as outlined in Table 17.

The first phone contact was used as an alternative to Dillman's third mailing (three weeks after the original mailing) and timed to correspond with the approximate time that a mailed package would have been received by the physicians. During this stage of follow-up,

information was sought about receipt of the original questionnaire (three replacement questionnaires were subsequently mailed out; two to physicians who reported not having received the original, and one to a physician who had discarded the questionnaire based on an incorrect self-assessment of ineligibility). The second mailing, with replacement questionnaires, corresponds to the final mailing in the TDM with the exception that the package was sent by regular mail. In the last stage, a final attempt was made to obtain responses from physicians and the option of providing answers to the survey questions by phone was presented to them.

Table 17

Design method for physician survey

Action implemented	Date <sup>a</sup>	Time since 1st mailing	Time since last contact <sup>b</sup>	Return date requested
1st mailing with return envelope	23.02.93			05.03.93
Mailed reminder	05.03.93	10.0 days		
1st phone follow-up	24.03.93	4.0 weeks	2.5 weeks	
2nd mailing with replacement questionnaire	13.04.93	7.0 weeks	3.0 weeks	22.04.93
2nd phone follow-up	30.04.93	9.5 weeks	2.5 weeks	

<sup>a</sup> For telephone based actions this date refers to the starting date although the process of contacting a physician might have covered a number of days

<sup>b</sup> With reference to the approximate time of receiving mailed material allowing a maximum of one week, or five working days, as processing time for mail

With the replacement questionnaires, a question was added to the cover sheet giving respondents the option of indicating whether the survey would be relevant to their practice or not. If the answer was negative the physician was directed to answer only the question relating to specialty. In addition, an option was given to state other reasons why the questionnaire was not completed. These modifications to the survey were added during the follow-up process in an attempt to further enhance responses (not the completion of questionnaires) so as to avoid unnecessary follow-up of ineligible physicians.

#### **4.2.4 Data Handling and Analysis**

The survey data were entered into two SPSS-based datasets. The first dataset consisted of data from the mailing list as well as the survey itself, for the total sample of physicians surveyed. The variables included gender, year of graduation, eligibility, responses to follow-up indicating intention to complete the questionnaire, and the completion of questionnaires. The actual responses in completed questionnaires constituted the second database. Other variables included were gender, year of graduation, and the date of receipt of responses.

Descriptive analyses were performed for (a) the total sample of physicians surveyed (based on reactions to the survey) and (b) the respondents (based on actual responses to the questionnaire). The analyses included comparisons between subsamples of each of these two groups. Analysis of responses was mainly based on diagnostic accuracy, and investigative and consulting patterns.

I analyzed diagnostic accuracy with respect to the diagnoses that the written cases were intended to simulate although it is acknowledged that the clinical signs and symptoms constituting a typical clinical picture for one disease may simultaneously constitute an atypical presentation of another. Thus, the judgement of accuracy within this study is only with

respect to the agreement between a diagnosis in the completed questionnaire and the diagnosis pre-assigned to a particular case scenario. Physicians were asked to state a primary diagnosis and up to three differential diagnoses so diagnostic accuracy could be assessed at two levels based on (a) the agreement between the pre-assigned diagnosis and the primary diagnosis, and (b) the agreement between the pre-assigned diagnosis and any of the diagnoses offered for a particular case.

In addition, the analysis of diagnostic accuracy was first based on a specific diagnosis of poliovirus infection and then repeated with a broader definition of accuracy for which diagnoses of "enterovirus-related neuropathy", or "anterior horn cell disease" were accepted as accurate for the cases simulating polio. This was done in order to examine the impact of these non-specific diagnoses on the judgements made with respect to the ordering of diagnostic tests. Other diagnoses such as "viral neuropathy" or "post-infectious neuropathy" were not accepted as proxy diagnoses for polio because they were felt to be sufficiently vague as to cover a multitude of clinical conditions. As such, these diagnoses would not yield valid information for a diagnosis of polio.

### **4.3 Results**

As previously mentioned, the survey data were analyzed for the reactions to the survey as well as responses to the questionnaire. The following sections present the results in that order.

#### **4.3.1 Responses to Survey**

Responses were received from 55 physicians (82.1%) and are summarised as follows:

1. Twenty-three physicians (34.3% of the sample initially surveyed) were confirmed as ineligible most often due to a limited practice.
2. Eighteen eligible physicians (40.9% of 44 eligible physicians) returned completed surveys.
3. Five eligible physicians (11.4%) communicated their unwillingness or inability to participate in the survey while nine (20.5%) indicated an intention to complete the survey but did not actually do so.
4. Twelve physicians gave no form of response to the survey but were assumed to be eligible (27.3% of eligible physicians).

The highest level of ineligibility (approximately 44%) was among the general internists (Table 18). The most common reason for ineligibility among this group was a subspecialty or a professional interest in specific diseases only (17 physicians) while one physician was on a leave of absence and another was reported as retired. Two neurologists indicated their specific interest in Parkinson's disease and epilepsy, and Parkinson's disease and Alzheimer's disease respectively. The third was established as a resident in neurosurgery. Only one infectious disease specialist was found to be ineligible based on location at the time of the survey. Twice as many general internists as neurologists completed the survey. However, there was no difference between the proportions that completed the survey (OR = 1.67, 95% CI; 0.38-7.41).

The peak response was in the sixth week following the first telephone follow-up; all the responses received during that week indicated either ineligibility, or inability or unwillingness to complete the questionnaire. Similarly, there was an increase in responses from those not intending to complete the questionnaire following the second phone follow-up. The majority of completed surveys (72.2%) were received in the second to fifth weeks after the initial

mailing; the last two stages of follow-up elicited only 22% (4) of the completed questionnaires.

Among the eligible physicians, the male to female ratio was approximately 14 to 1; 17 of 41 male physicians and only 1 of 3 eligible female physicians completed the survey. About 59% of the eligible physicians had graduated within the last 23 years (Table 19). No significant association was found between completion of questionnaires and the time of graduation from medical school ( $X^2 [1 \text{ df}] = 2.59, p = 0.63$ ).

Table 18  
Responses to survey by physician categories

	General internists (%)	Infectious disease specialists (%)	Neurology (%)
Ineligible	19 (44.2)	1 (20.0)	3 (15.8)
<b>Eligible</b>			
Survey completed	12 (27.9)	0 (0.0)	6 (31.6)
Survey not completed	12 (27.9)	4 (80.0)	10 (52.6)

Table 19  
Completion of questionnaires by physicians' time of graduation from medical school

Decade of graduation from medical school	Number completing questionnaire	Number not completing questionnaire
1940 - 1949	3	1
1950 - 1959	2	3
1960 - 1969	3	6
1970 - 1979	5	6
1980 - 1989	5	10

### 4.3.2 Analysis of Completed Surveys

All six neurologists who completed the survey reported seeing cases similar to the hypothetical ones occasionally. Only five of the general internists reported seeing similar cases occasionally while the rest reported seeing such cases rarely or never.

Based on a specific primary diagnosis of polio, the paediatric high sensitivity polio case was diagnosed with 100% accuracy (Table 20). High levels of accuracy were also obtained for botulism, both when the primary diagnosis alone was considered (83.3%), and when all differential diagnoses were included (100%). The level of accuracy for the adult high sensitivity polio case was 56.3% for primary diagnosis alone, and 62.5% when all differential diagnoses were considered. A similar level of accuracy was obtained for GBS; 57.1% based on all differential diagnoses. However, lower levels of accuracy were obtained for the adult low sensitivity polio case (19% with primary diagnosis only, and 31% with all differential diagnoses). For the paediatric low sensitivity polio case, the primary diagnoses offered were GBS, and a "post-infectious neuropathy" and polio was not recorded as a differential. For the control cases, a diagnosis of polio was made by two separate physicians for the GBS case (primary diagnosis) and the case of botulism (as a differential diagnosis).

When the non-specific diagnoses of "enterovirus-related neuropathy" and "anterior horn cell disease" were accepted as proxy diagnoses for polio, the diagnostic accuracy of polio improved (by an equal margin of 12.5 percentage points) to 75% for the adult high sensitivity polio case, and 43.8% for the adult low sensitivity polio case (see Table 21). These levels of accuracy were obtained with all differential diagnoses included. There was no impact on accuracy for the low sensitivity paediatric case.

The single most frequent diagnosis offered as a primary diagnosis for the polio cases was GBS; overall 10 diagnoses of GBS were made (Table 21), however 8 of these were for the low sensitivity case compared to 2 for the high sensitivity case. A diagnosis of a non-infectious spinal cord lesion (including cord tumours, disc herniation and spinal cord arteriovenous malformations) was also offered quite frequently as a differential diagnosis for the polio cases. Less common diagnoses included multiple sclerosis, radiculitis, thromboembolic neuropathy, mononeuropathy multiplex, lyme disease and an intracranial space-occupying lesion.

A comparison of diagnostic accuracy for the paired polio cases (HS and LS for each respondent) based on specific and non-specific polio diagnoses, and including all differential diagnoses showed that four general internists gave inaccurate diagnoses for both cases (i.e., not in agreement with the preassigned diagnosis of polio). Seven physicians (two neurologists and five general internists) provided accurate diagnoses for the pair of polio cases while seven more (four neurologists and three general internists) gave an accurate diagnosis for one case only. In all instances of a single accurate diagnosis the case involved was the high sensitivity case. Overall, neurologists were slightly more likely than internists to accurately diagnose polio but the difference was not statistically significant (OR = 1.69, 95% CI; 0.33 - 9.74).

Because of the small number of paediatric cases, all paediatric and adult polio cases were combined for the corresponding levels of sensitivity in subsequent analyses. The high sensitivity polio case was accurately diagnosed at least five times more than the low sensitivity case (Table 22); this was the finding both when the specific diagnosis of polio was used (OR = 5.20, 95% CI; 1.03 - 28.52) and when non-specific polio diagnoses were included (OR = 5.50, 95% CI; 1.06 - 31.49). The control cases were also more likely than the low sensitivity polio case to be diagnosed accurately however the difference was

statistically significant only when a specific diagnosis of polio was used (OR = 6.76, 95% CI; 1.29 - 39.33). No significant differences in diagnostic accuracy were found between the control cases and the high sensitivity polio case.

By far the most frequent investigation reported was CSF analysis (80.6% of all cases); even for the 21 accurate diagnoses of polio, the investigations reported were mostly not specific to polio (Table 23). Viral culture was reported for approximately 20% of all polio cases and an unspecified culture for another 14%; a stool specimen was mentioned in all cases and one physician also mentioned a pharyngeal specimen but no specific mention was made of a request for poliovirus culture. For 6 of the 14 cases in which a specific diagnosis of polio was offered as the primary diagnosis, an MRI, myelogram, or CT scan was reported as one of the first two tests in order of importance. CSF analysis, nerve conduction studies and electromyography were also reported ahead of polio-specific tests in a number of cases.

A high percentage of physicians indicated an intention to seek consultation with another physician for the case scenario they had reviewed; 83.3% for the control cases, 88.9% for the low sensitivity polio case, and 99.4% for the high sensitivity polio case. General internists reported an intention to consult another specialist for all the cases they reviewed while neurologists reported such an intention for 12 of the 18 cases reviewed. Overall, the difference in intention to consult was statistically significant ( $X^2 [1 \text{ df}] = 10.34, p = 0.001$ ).

Table 20

Diagnostic accuracy of responses with relation to specific diagnosis of polio<sup>a</sup>

Pre-assigned diagnosis	Number of questionnaires completed	Percentage agreement with pre-assigned diagnosis			Number of questionnaires with polio included in differential diagnoses (control cases only)
		Primary diagnosis only	Primary and differential diagnoses		
Polio child (HS <sup>b</sup> )	2	100.0	100.0	n/a	
Polio adult (HS <sup>b</sup> )	16	56.3	62.5	n/a	
Polio child (LS <sup>b</sup> )	2	0.0	0.0	n/a	
Polio adult (LS <sup>b</sup> )	16	18.8	31.3	n/a	
GBS	7	57.1	71.4	1	
Transverse myelitis	5	0.0	40.0	0	
Botulism	6	83.3	100.0	1	

<sup>a</sup> Based on specific diagnosis of polio (e.g., "enterovirus-related neuropathy" not counted as polio)

<sup>b</sup> High sensitivity (HS) and low sensitivity (LS) cases differed by one or more factors included in the history or clinical information of the former to increase the index of suspicion to the diagnosis

Table 21

Diagnostic accuracy of responses with relation to non-specific diagnosis of polio<sup>a</sup>

Pre-assigned diagnosis	Number of questionnaires completed	Percentage agreement with pre-assigned diagnosis			Number of questionnaires with GBS as primary (differential) diagnosis; test cases only
		Primary diagnosis only	Primary and differential diagnoses		
Polio child (HS <sup>b</sup> )	2	100.0	100.0	0 (0)	
Polio adult (HS <sup>b</sup> )	16	62.5	75.0	2 (6)	
Polio child (LS <sup>b</sup> )	2	0.0	0.0	1 (0)	
Polio adult (LS <sup>b</sup> )	16	25.0	43.8	7 (2)	
GBS	7	57.1	71.4	n/a	
Transverse myelitis	5	0.0	40.0	n/a	
Botulism	6	83.3	100.0	n/a	

<sup>a</sup> Based on non-specific diagnosis of polio; "enterovirus-related neuropathy" and "anterior horn cell disease" counted as polio

<sup>b</sup> High sensitivity (HS) and low sensitivity (LS) cases differed by one or more factors included in the history or clinical information of the former to increase the index of suspicion to the diagnosis

Table 22

Comparison of diagnostic accuracy for different case scenarios

Comparison groups	Specific polio diagnosis only		Non-specific polio diagnoses included	
	OR	95% CI	OR	95% CI
High and low sensitivity polio	5.20	1.03-28.52	5.50	1.06-31.49
Controls and high sensitivity polio	1.30	0.25-6.93	0.74	0.12-4.36
Controls and low sensitivity polio	6.76	1.29-39.33	4.09	0.83-21.56

The specialist category cited most frequently by general internists for consultation was neurology; for 91.7% of cases reviewed (Table 24). In the same group, intended consultations with infectious disease specialists were reported for 36.1% of cases. Neurologists on the other hand, reported consultations with infectious disease specialists most frequently (50% of cases). Of note, neurologists also reported an intention to consult neurosurgeons for 17% of cases; these were cases with differential diagnoses suggesting a space-occupying lesion of the spinal cord. Not surprisingly, general internists were found to be significantly more likely than the neurologists reviewed to seek neurology consultations (OR = 88, 95% CI; 10.90-966.55). However, there was no significant difference between general internists and neurologists with regard to seeking consultations from an infectious disease specialist (OR = 0.57, 95% CI; 0.15-2.06).

Table 23

## Reported frequency of test ordering for polio cases

	Frequency by order of importance				Polio cases with test cited (%)	
	1st	2nd	3rd	4th	All cases (n = 36)	Cases with accurate diagnoses (n = 21)
Culture, unspecified	1	0	1	3	13.9	19.0
Viral\enteroviral culture	3	2	2	0	19.4	28.6
Polioviral culture	0	0	0	0	0.0	0.0
Serology, unspecified	0	1	0	2	8.3	14.3
Viral\enteroviral serology	0	0	1	0	2.8	4.8
Polioviral serology	1	0	1	0	5.6	9.5
CSF analysis	7	12	7	3	80.6	71.4
Nerve conduction study	2	2	4	0	22.2	28.6
Electromyography	0	3	4	2	25.0	28.6
Myelogram	2	3	1	3	25.0	19.0
MRI	4	3	5	0	33.3	28.6
CT scan	4	6	4	2	44.4	23.8
Blood tests	7	1	0	3	30.6	23.8

Table 24

## Reported frequency of intention to consult different specialties

Specialty to be consulted	General internists		Neurologists	
	Number of consultations cited	% of cases reviewed (n = 36)	Number of consultations cited	% of cases reviewed (n = 18)
Neurology	33	91.7	2	11.1
Infectious diseases	13	36.1	9	50.0
Neurosurgery	2	5.6	3	16.7
Intensive care unit	1	2.8	2	11.1
Rehabilitation	0	0.0	3	16.7
Others	6	16.7	0	0.0

#### 4.4 Discussion

As with the chart review, the statistical power obtained with this sample of physicians was extremely low. As an example, comparison of diagnostic accuracy for high and low sensitivity polio cases was based on a power level of approximately 8% for an effect size of 5% at a 95% confidence level. The arguments previously made for public health significance as opposed to statistical significance may be considered applicable to this study as well.

The reactions to the survey were considered an important indication of the interest generated by the survey. In the feedback received most of the surveyed physicians expressed their opinion that the cases simulated are rare in Canada and therefore not applicable to the measurement of patient management. Other aspects of the thesis had also previously elicited an element of perceived unimportance of the disease of interest, poliomyelitis. It is important to note however that these points form the central issue of the thesis. The survey purpose, as stated in the cover letter, was deliberately vague because of the nature of the questions asked. Nonetheless, the rarity of the diseases involved and the poor interest demonstrated by the population of physicians surveyed raise the issue of the validity of responses.

Steps were taken to ensure the content or face validity of the survey through the involvement of experts at all the stages of questionnaire development as described in subsection 4.2.2. One means of assessing the criterion validity (i.e., the agreement between reported judgements and behaviour and those applied in real life situations) would have been to compare each physician's responses with his or her behaviour as documented in patient charts. This is very difficult in any study involving rare diseases because of the difficulty in obtaining an adequate number of records for each physician and was not logistically feasible in the current study. In a broad sense, the findings of the physician survey are in agreement with the more objective findings of the chart review. In spite of the temporal difference and

the inability to compare behaviours for specific physicians, the fact that the results from these independent studies support each other is important for overall conclusions.

In addition, the lack of a cueing effect supports a conclusion that responses to the survey questions are an accurate representation of the judgements and behaviour that these physicians would have taken in real life situations. The low level of diagnostic accuracy for polio, and the low frequency of appropriate investigations are also contrary to the expectation when a demand effect occurs. Again, one possible explanation is that the reported behaviours accurately represent real life physician behaviours. However, the alternative explanation could be that more expensive, more invasive, and non-specific tests for polio (CT scan, MRI, myelogram) were reported at a relatively higher frequency because of the demand effect. In other words, the majority of surveyed physicians might have thought they were expected to order these basic tests in any neurological case. It is possible that in some cases these tests were ordered to rule out the differential diagnoses but the order of importance given to the tests in instances when polio was offered as the primary diagnosis seems inappropriate. This statement is made in light of the fact that the more specific tests of viral culture and serology are under usual circumstances more easily available, less costly and far less invasive to the patient.

These data demonstrate low levels of diagnostic accuracy for polio generally, and especially for atypical cases possibly because of a low level of awareness of polio presentation and the continuing possibility of polio occurring in Canada. The apparent knowledge of appropriate investigations to confirm polio is also low in the population surveyed.

Although the argument can be made that these findings may not be representative of the national situation, there is no obvious reason to expect that physicians in other parts of the country will have a higher index of suspicion or exhibit more appropriate patient management

behaviour for polio. A possible exception to this is the extremely small proportion of physicians who have been most recently involved in the management of polio cases across Canada. Therefore, the overall findings of the physician survey add impetus to the need to increase the level of awareness among clinicians in Canada about polio and the appropriate diagnostic recommendations.

## CHAPTER 5            RECOMMENDATIONS FOR FUTURE ACTION

The independent findings of both the chart review and the physician survey in Ottawa-Carleton demonstrate a low level of awareness for the clinical presentation and diagnostic procedures of paralytic poliomyelitis. Only the review of the polio surveillance system was representative of national data; nonetheless, the overall findings of the thesis provide justification for some definite action to document the true extent of poliovirus activity in Canada. The findings are not only important to the rare individual case of polio but are also important because of the potential for virus transmission and the subsequent health risk to susceptible persons in Canada.

In addition, the implications of these findings for certification of a polio-free status cannot be overlooked. Although an absence of indigenous wild polio cases has been documented in Canada for at least 15 years now, the inadequacies demonstrated in the polio surveillance system, coupled with a lack of effective AFP surveillance should deter reliance on past experience as evidence of polio elimination in Canada. There is currently no system available for the environmental surveillance of polioviruses. Consideration should be given to environmental surveillance as a fundamental component of polio elimination in Canada (and its documentation) because the majority of infected persons have subclinical illnesses but may still excrete the virus for considerable periods of time. The virus in turn may survive in sewage for several weeks acting as a potential source of new infections. The focus of surveillance has to be shifted from a clinical-based approach to environmental surveillance.

This chapter outlines measures required in Canada to enhance polio surveillance and to meet PAHO's certification criteria. It should be noted that the recommended measures are not limited to the detection of the wild poliovirus but also have relevance to the vaccine-type virus. Data for the vaccine-type virus will assist in accurately quantifying the risk of OPV-

associated disease. The general recommendations are outlined below and discussed in further detail in subsequent sections.

1. The establishment of AFP surveillance to document the absence of indigenous polio cases for at least three years under circumstances of adequate surveillance. This should include (a) negative reporting of cases and (b) appropriate investigations to rule out the wild poliovirus in all polio-compatible cases.
2. The establishment of environmental surveillance to document the absence of wild poliovirus based on stool and sewage sampling. Two strategies of sampling may be considered: (a) sampling from selected communities at high risk for poliovirus transmission, and (b) random sampling from the general population. The former strategy offers much higher chances for viral recovery however, that alone may be insufficient for conclusions regarding overall poliovirus activity in the country.
3. The development of a written protocol for investigating suspected cases of poliomyelitis. This should be accompanied by a protocol for control measures in the event of positive identification of wild poliovirus.
4. The establishment of a national certification commission to oversee these activities and to collate data for presentation to the international certification commission at an appropriate time. It is hoped that the national commission will develop a protocol for the regular evaluation of the pre-certification activities; such a protocol can also be used in the post-certification period to ensure a minimum risk of re-emergence until global eradication is achieved. The formation of the national commission is not discussed in this thesis.

## 5.1 AFP Surveillance

In cognizance of the time requirements and other difficulties anticipated in establishing a new surveillance program, I recommend that the existing programs be retained with efforts to improve collaboration of activities and allocation of resources. The anticipated advantages of such a collaboration include (a) a greater ease of program evaluation and accumulation of evidence towards documentation of a polio-free status, and (b) a better opportunity for achieving and maintaining awareness of the public health importance of polio until global eradication is achieved. To ensure completeness of accurate data on the polio situation in Canada the establishment of a national database specifically for polio-related information (from all the activities listed above) should be considered.

The current surveillance for paralytic polio under the CCDSS should be retained as the foundation for a collaborative program of AFP surveillance which will also include the IMPACT System and the VAAESS. Cases of both paralytic and nonparalytic poliomyelitis, and other acute paralytic syndromes reported to the three programs should be included in the national polio database. Because of the multiple reporting mechanisms, it will be necessary to have a coordinating centre where data can be processed and monitored for duplication.

Certain modifications may be required in the current surveillance system as a move is made to target AFP as a screening measure for polio. Also, some existing measures need to be emphasised or intensified. Provincial and territorial epidemiologists should be involved in the process of revising the existing system in order to enhance adoption of these changes in their respective jurisdictions. In each jurisdiction, changes to the surveillance system should be well publicised for all local health units, health facilities, and laboratories as well as any ancillary reporting sources that might exist.

The acceptability of AFP as a notifiable disease condition is anticipated to be low initially. During this study, communication with a number of clinicians reflected the opinion that such a diagnostic term is inappropriate in Canada because of the current diagnostic sophistication which enables specific diagnosis of cases presenting with acute paralysis. It is important to acknowledge however that (a) the level of diagnostic sophistication is often useless without prior awareness of, or suspicion for, the possibility of a specific disease and (b) the detection and subsequent reporting of polio-compatible AFP cases is highly dependent on contact between cases and clinicians for medical care. Without prior education of health care workers on Canada's progress in polio elimination the recommended changes in surveillance will be difficult to implement. In addition, measures should be instituted to educate relevant categories of clinicians on the presentation of polio and recommendations for investigation and notification for suspected cases.

Additional recommendations for improving the quality of polio surveillance data are:

1. A case definition for AFP should be developed and well publicised. This definition could include a target age group based on the epidemiology of polio cases reported in Canada since control of the disease was achieved. Helpful data for the decision on the target age group may also be obtained from other non-endemic countries, particularly the United States. Polio-specific investigation of AFP cases may be limited to cases compatible with polio as defined by the best known predictors of culture confirmed polio reported by Andrus et al and Dietz et al.
2. Case definitions for all subcategories of poliomyelitis should be standardised for use in the three programs. This is particularly important if data are to be collated in one database. IMPACT case definitions for other acute paralytic conditions may be adopted for general use. As stated in Chapter 2, the current definition of

immunocompromised cases is potentially misleading and should be reviewed; information on the immune status can be included (when appropriate) as additional description of a case and may be used for other purposes such as the evaluation of vaccine injury compensation claims.

3. Emphasis should be placed on regular negative reporting from all participating health units.
4. The current guidelines for reporting nonparalytic polio under viral meningitis requires clarification to eliminate the conflicting instructions outlined in subsection 2.4.1. Data on polioviral meningitis should be available at both the provincial/territorial and federal level and should be separate from data for other causes of viral meningitis.
5. Particular attention should be given to cases that fit the category of no known contact cases to accurately confirm the virus strain as the vaccine-type; this requires collaboration with the national reference laboratory for viral characterization.
6. To enhance the interpretation of laboratory-based data, all reporting laboratories should be encouraged to obtain basic clinical data (to be decided on) and include such data with poliovirus reports to provincial and territorial health authorities. This is particularly important for symptomatic cases and for asymptomatic cases in whom the wild virus is implicated. If the relevant clinical data are not easily accessible to the laboratories, provincial and territorial health authorities could trace cases with positive viral detection in order to obtain the data. With such clinical data, laboratory-based reports of viral detection can be subjected to more meaningful public health assessment and interpretation.

7. The role of the National Centre for Enteroviruses as a reference laboratory should be well publicised and its virus-typing services made available to all viral laboratories.
8. Available means of providing ongoing education for clinicians should be explored and used to advantage. These may include (a) presentations at appropriate clinical meetings and (b) regular publications in respected clinical journals, an example being a news bulletin on the differential features of paralytic polio and GBS published by the CPS in 1990.<sup>75</sup> Also, collaboration should be sought with recognised groups in the medical profession to enhance their interest and participation in the surveillance and documentation efforts.

## **5.2 Stool and Sewage Sampling for Poliovirus Detection**

1. Environmental surveillance of poliovirus in the general population may be carried out by regular sewage sampling. Virus characterization should include efforts to detect both indigenously circulating wild strains (previously recognised or new) and imported strains.
2. Stool and sewage sampling of populations at high risk for poliovirus transmission should also be carried out on a regular basis, and following suspicion or evidence of virus importation. High-risk populations should be well defined based on immunization coverage levels and demographic factors such as high concentrations of immigrant populations. Due to lack of recent data on seroprevalence of antibodies to poliovirus in Canadian populations, the current definition of high risk groups can only be based on immunization histories. Religious communities known to resist immunization and previously identified as highly susceptible to infection are a particular source of concern with regard to poliovirus transmission. Other

unimmunized individuals with a history of travel to or contact with persons from polio-endemic regions, or regions with outbreaks of wild poliovirus transmission, should be considered at high risk for infection. High risk individuals also include close contacts of confirmed cases who are not fully immunized; when direct stool sampling has not been possible for contacts, sewage sampling at carefully selected sites should be considered.

3. For general surveillance more than one sampling site should be identified for each province or territory; the minimum number and the selection of sites will be determined by the population density. The more appropriate sites include large urban centres and centralised sewage treatment facilities. When sewage sampling is conducted as part of outbreak investigations in a high-risk community, samples may be obtained from sewage facilities at meeting places in the community and timed to maximize the opportunity for obtaining concentrated volumes of virus in sewage.
4. Sampling from sewage treatment plants may be at the intake station or from the activated sludge. It is recommended that a standardised approach be adopted for all sampling sites based on the relative efficiency of viral recovery if such data are available.
5. Methods of sewage sample collection, involving either the "continuous sampling" or the "grab sampling" technique should be standardised across sampling sites based on knowledge of the relative efficiency of viral recovery.
6. Sampling should be carried out throughout the year at regular intervals. During the first year of implementation sampling may be at a monthly interval and subsequently reviewed based on the extent of wild virus recovery. It should be recognised at the

outset that environmental surveillance may have to be sustained for several years until the global eradication of wild poliovirus has been achieved and a decision made to stop immunization against polio.

7. Strict protocols for the identification of viruses (including the use of standardised cell lines and methods of isolation) need to be developed and adopted by all participating laboratories.
  
8. The potential role of environmental surveillance, given the rare occurrence of clinical cases in Canada, has been recognised. At the same time, there are a number of potential setbacks to its implementation such as (a) an imbalance in resources needed versus the efficiency of virus recovery, (b) the difficulty of interpreting negative results of environmental surveillance, and (c) the difficulty of interpreting positive wild virus identification particularly in areas of high transient human traffic flow and with regard to the type of action that could be taken. If a decision is taken to establish environmental surveillance, collaboration will be needed among health care workers in various scientific fields including virologists, epidemiologists, clinicians, statisticians, and engineers. The availability of viral laboratory services in Canada has great advantages for environmental surveillance and as much as possible efforts should be made to establish a network among these laboratories to that end.

### **5.3 Investigation of Suspected Cases and Control Measures**

Investigation and control protocols should be developed for all levels of the public health system including the local, provincial or territorial, and federal levels. Activities at these levels are not to be implemented independently but in a collaborative manner. Protocols

should be widely publicised to the appropriate health care workers at each level. The following points are recommended for the development of protocols.

#### Local level

1. For all polio-compatible AFP cases, two stool specimens should be obtained for viral culture within two weeks of the onset of paralysis. Pharyngeal specimens may be considered an alternative for virology although the emphasis is on stool as the optimum specimen. The importance of including stool specimens from at least five contacts of the case is a debatable point at present. Stool investigation of contacts has the merit of providing data to assist in diagnosis especially when the case is culture-negative or when an opportunity to carry out specific investigations in the case has been missed. Diagnostic serology may also be carried out to augment the efforts at direct viral detection. (LCDC and the National Centre for Enteroviruses are developing guidelines for the proper collection and transfer of specimens; these should be widely distributed in all provinces and territories).
2. All relevant data should be obtained as per the common case and polio report forms. The form may require some limited modification to fit AFP surveillance; emphasis should be placed on including histories for immunization (self and contact) and travel.
3. All cases should be reported to the provincial or territorial public health authorities without waiting for results of investigations.
4. The above procedures should be initiated within 48 hours of reporting to the local health unit.

Provincial/Territorial level

1. Initiation of the above investigations should be confirmed.
2. In the event of **positive poliovirus detection**, proper stool (or pharyngeal) specimens should be submitted to the National Centre for Enteroviruses for confirmation of the result and further virus characterization.
3. In the event of **negative poliovirus detection with positive polio serology**, proper stool (or pharyngeal) specimens should be submitted to the National Centre for Enteroviruses for repeat virology.
4. In the event of **positive wild virus detection**, steps should be undertaken to identify persons or communities at risk for virus transmission based on immunization status or travel histories. Sewage and stool sampling should be carried out in such communities at regular monthly intervals as long as a wild type virus is detected. In the presence of high volumes of vaccine-type virus the detection of a wild-type virus may be particularly difficult with the conventional tissue-culture based methods. Application of molecular methods at the National Centre for Enteroviruses will greatly enhance the sensitivity for detecting wild virus in such instances.
5. In conjunction with the last point, the appropriate categories of physicians, and health institutions in the communities affected should be alerted to actively search for and report AFP cases and to carry out specific investigations for polio-compatible cases.
6. In conjunction with the last two points, viral laboratories in the province or territory affected by an outbreak, or in which an individual case was reported should be alerted to actively search for the poliovirus in stool and pharyngeal specimens as appropriate.

7. Evidence of low immunization coverage against polio should be followed by immunization campaigns wherever possible (the so-called mop-up operation). In Canada, such situations are likely to occur mostly in communities with a well-established resistance to immunization thus, the practicality of such a measure is doubtful, nonetheless the attempt should be made.
  
7. The LCDC should be informed of all suspected cases of poliomyelitis or wild virus transmission promptly without waiting for completion of investigations; an optimum delay of 24 hours is recommended.

#### Federal level

1. In collaboration with the National Centre for Enteroviruses, LCDC should provide materials and resources for specimen collection and transfer, and for other investigation and control measures as needed. An epidemiologist could be assigned to assist provinces and territories in their polio-related activities when necessary. The advantage of having one individual in this capacity is that of sustaining continuity however, this individual's field experience need not be limited to polio especially as the load of work is not anticipated to be large.
  
2. The National Centre for Enteroviruses should continue to act as a reference laboratory for the identification and characterization of polioviruses.
  
3. As part of the ongoing control of polio, LCDC should collate data from all the polio-related activities recommended above to form a national database on the epidemiology of poliomyelitis. This database is to be separate from the general CCDSS database administered by the DSD and should be closely monitored for completeness and accuracy. Also the timeliness of reports and investigations should be documented right

from the local level. Such a database will serve as valuable documentation for the national commission in its task of evaluating pre-certification activities.

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## Appendix A

### Case Definitions for Poliomyelitis, Canadian Communicable Disease Surveillance System

Source: The Advisory Committee on Epidemiology and the Bureau of Communicable Disease Epidemiology. Canadian Communicable Disease Surveillance System; Disease-specific case definitions and surveillance method. CDWR 1991; 17S3 (Suppl) 26-8.

#### Poliomyelitis (paralytic)

##### Confirmed case

Clinically compatible signs and symptoms of paralytic poliomyelitis including all of the following:

1. Acute flaccid paralysis of one or more limbs.
2. Decreased or absent tendon reflexes on the affected limbs.
3. No sensory or cognitive loss.
4. No other apparent cause (including laboratory investigation to rule out other causes of a similar syndrome).
5. Neurologic deficit present 60 days after onset of initial symptoms unless the patient has died.

Associated with the isolation of either vaccine or wild poliovirus from a clinical specimen.

Paralytic poliomyelitis cases are sub-divided into the following categories:

1. **Wild virus:** Laboratory investigation implicates wild-type virus. This group is further sub-divided as follows:
  - (a) **Imported:** travel or residence in a polio-endemic area 30 days or less before onset of symptoms.
  - (b) **Import-related:** epidemiologically linked to someone who has travelled or

resided in a polio-endemic area within 30 days or less before onset of symptoms.

(c) *Indigenous*: no travel or contact as described above.

2. **Vaccine-associated**: Laboratory investigation implicates vaccine-type virus. This group is further sub-divided as follows:

(a) *Recipient*: the illness began 7-30 days after the patient received oral polio vaccine.

(b) *Contact*: the patient was shown to have been in contact with a vaccinee and became ill 7-60 days after the vaccinee received oral polio vaccine.

(c) *Possible contact*: there was no known direct contact with a vaccinee and no history of the patient receiving oral polio vaccine, but the paralysis occurred in an area in which a mass vaccination campaign had been in progress 7-60 days before the onset of paralysis. Within Canada, all provinces routinely using oral polio vaccine meet this criteria [sic] all the time.

(d) *No known contact*: the paralysis occurred in a patient with no known contact with a vaccinated person or recent receipt of polio vaccine in an area where intensive vaccination had not been in progress. In Canada this would include only provinces that do not routinely use oral polio vaccine.

(e) *Immunocompromised*: diagnosis of concurrent medical condition associated with deficient immune function in any vaccine-associated case.

### **Possible case**

Clinically compatible symptoms of paralytic poliomyelitis (as listed above), without isolation of poliovirus from clinical specimens, with serologic evidence of recent poliovirus infection, without evidence for infection with other neurotropic viruses.

**Preventable case**

All wild virus cases in unimmunized or inadequately immunized Canadian residents who are eligible for immunization.

## Appendix B

### Case Definitions for Poliomyelitis and Other Acute Paralyzes, IMPACT System

Source: The Executive Committee of the Immunization Monitoring Program Active System.  
The IMPACT operations manual, July 1992.

#### **Paralytic poliomyelitis**

The IMPACT System uses case definitions compatible with the CCDSS definitions for OPV-associated paralysis and wild poliomyelitis with the following minor exceptions:

1. A case meeting the clinical case definition is classified as probable (equivalent to possible case in the CCDSS).
2. Case confirmation may be supported by a 4-fold rise in serum antibody titre to poliovirus in addition to or instead of virus isolation from a clinical specimen.

"Other noteworthy cases" to be reported are:

#### **Not poliomyelitis:**

Acute paralytic illness in which at least 2 adequate stool specimens were obtained within 2 weeks after onset of symptoms and were negative for poliovirus. Aliquots of the original samples should be held at the laboratory for possible future use. To ensure the accuracy of this categorisation, any patient who dies, is lost to follow-up, or has residual paralysis at 60 days should have aliquots of the original specimens examined by the National Polio Reference Laboratory using all appropriate shipping techniques. If the specimens were adequate and all were negative, these cases should be considered "not polio" and "discarded".

#### **Guillain-Barré Syndrome**

A rapidly progressive, symmetrical motor weakness with loss of tendon reflexes.

Weakness typically ascends upwards from feet and may or may not be associated with some sensory loss. Often involves abdominal and thoracic muscles but cranial nerve involvement is uncommon. CSF changes are limited to elevated protein level.

#### **Aseptic meningitis following live virus vaccination**

Acute febrile illness with headache (irritability in infants) and often with neck stiffness, associated with inflammation of CSF (WBC count in CSF > 7 per ul). Culture of CSF may be positive for mumps or poliovirus of vaccine type.

#### **Other acute paralysis syndromes following vaccination**

Syndromes that can be reported in this category include (a) acute transverse myelitis and (b) peripheral neuropathies

#### **Acute flaccid paralysis, all non- polio types**

All conditions that reasonably mimic poliomyelitis in causing an acute febrile illness with focal weakness or paralysis can be included in this category, such as:

Guillain-Barre syndrome

Transverse myelitis, other acute spinal cord inflammation, including non-polio enterovirus infection

Hemiplegia, monoplegia, paraplegia syndromes (except post-ictal)

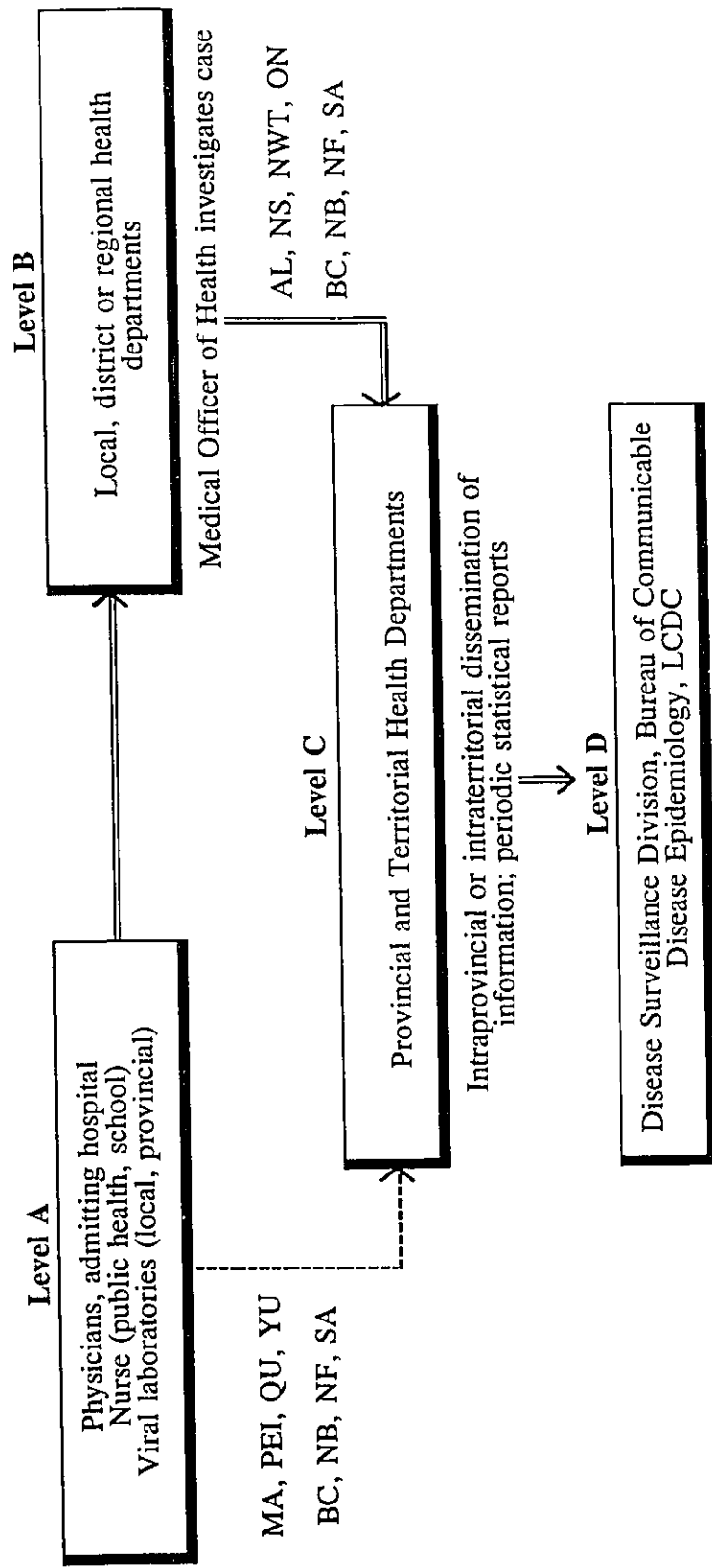
Polyneuropathy syndromes

Appendix C

Channels of Reporting in Surveillance Programs

Figure C-1

Flowchart for the Communicable Disease Surveillance System

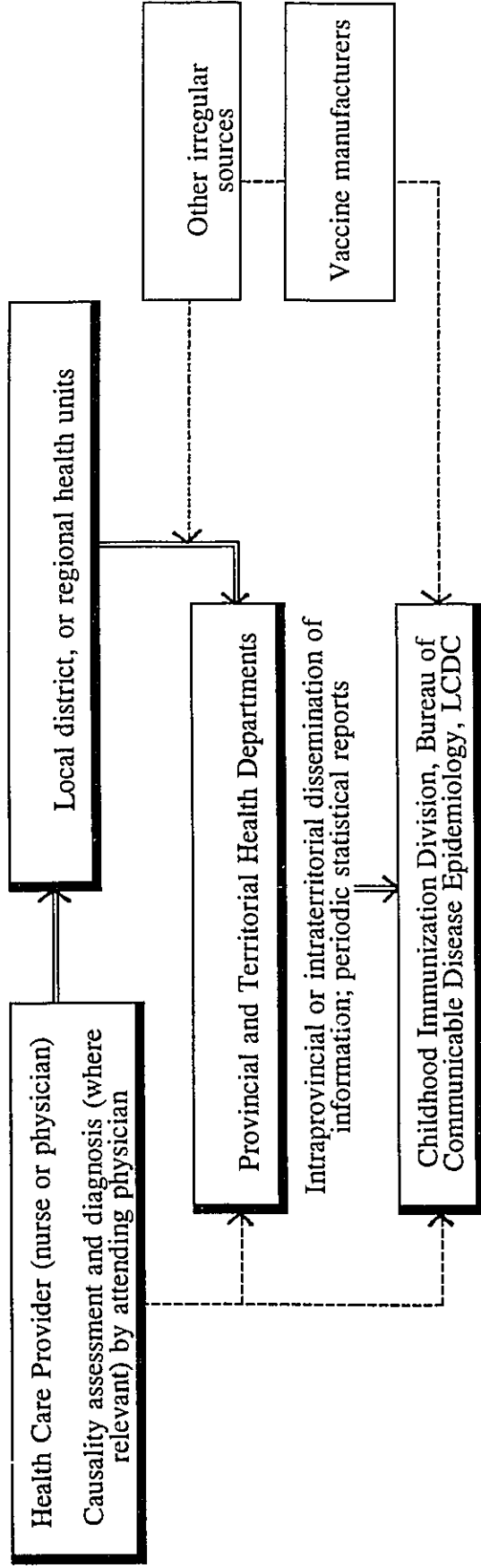


Dissemination of information: Published monthly and annual summaries, Canadian Communicable Disease Report.

Key: Double solid lines indicate main communication channels while single dashed lines indicate supplementary channels.

Figure C-2

Flowchart for the Vaccine-Associated Adverse Events Surveillance System



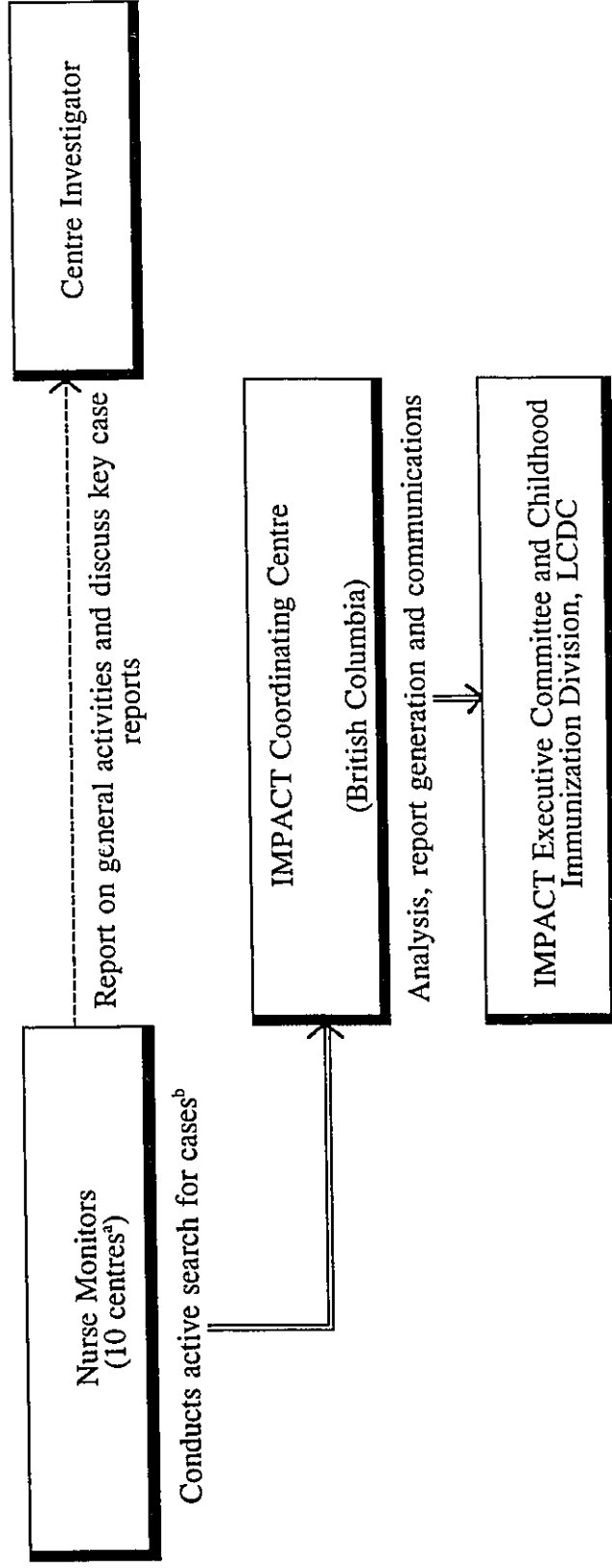
Dissemination of information:

- 1) Quarterly and annual summary reports to Provincial/Territorial Epidemiologists, Manufacturers, Bureau of Biologics, and WHO.
- 2) Published annual summaries in the Canadian Communicable Disease Report.

Key: Double solid lines indicate main communication channels while single dashed lines indicate supplementary channels.

Figure C-3

Flowchart for the IMPACT System



Key: Double solid lines indicate main communication channels while single dashed line indicates a supplementary channel at each site.

<sup>a</sup> The 10 paediatric centres currently participating in IMPACT are located in Quebec(3), Ontario (2), Alberta, British Columbia, Manitoba, Newfoundland and Nova Scotia (1 each).

<sup>b</sup> Assisted by a network of volunteers from the Hospital Admitting Department, infection control nurses, neurology ward staff and physicians, infectious diseases staff, and Medical Records technicians.

Appendix D

Summary of Data Handling in the CCDSS

Table D-1

Availability of communicable disease data handling methods in Canada

Province/Territory	Transfer between levels A, B and C	Transfer of case by case data to LCDC <sup>a</sup>
Newfoundland	Both	Electronic system under development
Prince Edward Island	Manual with electronic system under development	
Nova Scotia		
New Brunswick	Manual	None
Quebec	Electronic	Electronic; by diskettes
Ontario		Electronic
Manitoba		None
Saskatchewan	Electronic	Electronic
Alberta	Electronic	Electronic
British Columbia	Both	Electronic (recently piloted)
Yukon	Manual	Report forms submitted with data entry at DSD
Northwest Territory	Electronic	

<sup>a</sup> All jurisdictions submit aggregate data by mail and fax except Quebec. Quebec data is aggregated by the Disease Surveillance Division, LCDC.

Table D-2

Distribution of communicable disease reports by provinces and territories,<sup>a</sup> Canada

Province/Territory	Frequency	Distribution of reports
Newfoundland	Weekly, monthly	Health units, LCDC, general practitioners, selected physician categories
Prince Edward Island	Monthly	LCDC
Nova Scotia	Monthly	Health regions, LCDC
New Brunswick	Monthly	District Medical Officers of Health, Chief Public Health Officer, physicians
Quebec	Monthly	Health workers in all provincial regions
Ontario	Annually	Medical Officers of Health, LCDC
Manitoba	Monthly, annually	Director, Communicable Disease Control, Regional Health Departments
Saskatchewan	Monthly	Health units, Medical Officers of Health
Alberta	Every 4 weeks, annually	Health units, Medical Services Branch, Provincial Advisory Committee
British Columbia	Monthly	Province wide to health workers
Yukon	Monthly	Medical Services Branch, Territorial Government
Northwest Territory	Monthly, annually	All health workers in territory

<sup>a</sup> The table describes routine practices but most jurisdictions indicate situation-dependent practices with higher frequency of analyses and reporting.

APPENDIX E

Profile<sup>a</sup> of Admitting Hospitals Included in Chart Review

Hospital	Catchment area and/or population	Total in-patient beds <sup>b</sup>	Medical beds <sup>2</sup>	Neurologists <sup>c</sup>
Children's Hospital of Eastern Ontario	Eastern Ontario and western Quebec; 600,000 to 700,000 children	160		8
Montfort Hospital	Eastern part of Ottawa-Carleton, including Vanier and the counties of Prescott and Russell	176	65	5
Ottawa Civic Hospital	Eastern Ontario and western Quebec; approximately 1.2 million	747	246	11
Ottawa General Hospital	Approximately 1 million		149	12
Queensway-Carleton Hospital	Western region of Ottawa, Nepean, Kanata, West Carleton, Goulbourn, and parts of Rideau township; 225,000	211	72	1
Riverside Hospital of Ottawa	The region of Ottawa east of the Rideau River	248	107	2

<sup>a</sup> The information provided below was obtained from the respective administrative, human resources, and/or public relations departments of the hospitals

<sup>b</sup> 1992 estimates for average beds, accounting for seasonal bed closures

<sup>c</sup> Total number of neurologists affiliated with the hospital, including all categories of "active", "associate", "consulting", "honourary", "emeritus", and "courtesy". Excludes the category of "scientific consulting" which provides no patient care.



## PATTERN OF PHYSICIAN REFERRAL

Referred from: \_\_\_\_\_  
(Specialty of physician or department)

Attending physician(s): \_\_\_\_\_  
(Specialty of physician or department)

Consulting physician(s): \_\_\_\_\_  
(Specialty of physician or department)

## NOTIFICATION OF PUBLIC HEALTH AUTHORITIES

(Specify where notice was sent)

\_\_\_\_\_

### A) SYMPTOMS

(Yes, No, Unknown; if yes, note duration between onset and admission)

1. Fever	Y	N	U	___ weeks	___ days
2. Coryza	Y	N	U	___ weeks	___ days
3. Sore throat	Y	N	U	___ weeks	___ days
4. Headache	Y	N	U	___ weeks	___ days
5. Stiff neck	Y	N	U	___ weeks	___ days
6. Nausea/Vomiting	Y	N	U	___ weeks	___ days
7. Diarrhoea	Y	N	U	___ weeks	___ days
8. Constipation	Y	N	U	___ weeks	___ days
9. Rashes	Y	N	U	___ weeks	___ days
10. Weakness/Paralysis	Y	N	U	___ weeks	___ days
11. Muscle Pain	Y	N	U	___ weeks	___ days
12. Arthralgia	Y	N	U	___ weeks	___ days
13. Trauma	Y	N	U	___ weeks	___ days
14. Loss of bladder control	Y	N	U	___ weeks	___ days





CSF viral culture 2    Y    N    Result (Positive Negative) \_\_\_\_\_  
Date taken \_\_\_/\_\_\_/\_\_\_    Date examined \_\_\_/\_\_\_/\_\_\_

4. Viral serology 1    Y    N    Result \_\_\_\_\_  
Date taken \_\_\_/\_\_\_/\_\_\_    Date examined \_\_\_/\_\_\_/\_\_\_

Viral serology 2    Y    N    Result \_\_\_\_\_  
Date taken \_\_\_/\_\_\_/\_\_\_    Date examined \_\_\_/\_\_\_/\_\_\_

5. Stool viral culture 1    Y    N  
Date taken \_\_\_/\_\_\_/\_\_\_    Date examined \_\_\_/\_\_\_/\_\_\_

Stool viral culture 2    Y    N  
Date taken \_\_\_/\_\_\_/\_\_\_    Date examined \_\_\_/\_\_\_/\_\_\_  
Result \_\_\_\_\_

6. Nerve conduction speed/electromyogram

1.    Y    N    Date \_\_\_/\_\_\_/\_\_\_    Result \_\_\_\_\_
2.    Y    N    Date \_\_\_/\_\_\_/\_\_\_    Result \_\_\_\_\_
3.    Y    N    Date \_\_\_/\_\_\_/\_\_\_    Result \_\_\_\_\_

7. Other relevant (specify; X-ray, MRI, Myelogram, CPK studies)

1.    Y    N    Date \_\_\_/\_\_\_/\_\_\_    Result \_\_\_\_\_
2.    Y    N    Date \_\_\_/\_\_\_/\_\_\_    Result \_\_\_\_\_
3.    Y    N    Date \_\_\_/\_\_\_/\_\_\_    Result \_\_\_\_\_
4.    Y    N    Date \_\_\_/\_\_\_/\_\_\_    Result \_\_\_\_\_

## APPENDIX G

### Coding Scheme for Selected Variables

Only those variables used in the algorithms in Appendix H are included in this coding scheme. Variables are listed in alphabetical order to facilitate reference.

Variable name	Variable labels and values
CORYZA	Prodromal respiratory tract infection (0 = No, 1 = Yes, 9 = Unknown)
CRANIALS	Symptoms of cranial nerve involvement (0 = No, 1 = Yes, 9 = Unknown)
CRANONST	Time between onset of cranial nerve symptoms and admission date (days or weeks, 999 = Unknown)
DIARRHEA	Prodromal diarrhea (0 = No, 1 = Yes, 9 = Unknown)
DOADA, DOAMO, DOAYR	Day, month and year of admission
DOBDA, DOBMO, DOBYR	Day, month and year of birth
DRPDA, DRPMO, DRPYR	Day, month and year residual paralysis recorded
FEBRILE	Prodromal febrile illness (0 = No, 1 = Yes, 9 = Unknown)
FEVER	Fever at admission (0 = No, 1 = Yes, 9 = Unknown)
LEVELPAR	Level of limb paralysis (0 = Distal, 1 = Proximal, 3 = Equivalent, 9 = Equivocal)
LPOWERLLL	Lowest grade of power in left lower limb (0 to 5)
LPOWERLUL	Lowest grade of power in left upper limb (0 to 5)

Variable name	Variable labels and values
LPOWRLL	Lowest grade of power in right lower limb (0 to 5)
LPOWRLUL	Lowest grade of power in right upper limb (0 to 5)
PATNPAR	Pattern of limb paralysis (0 = Symmetrical, 1 = Asymmetrical)
PROGPAR	No progression of paralysis after admission (0 = No, 1 = Yes, 9 = Unknown)
RESDLPAR	Residual paralysis (0 = No, 1 = Yes, 9 = Unknown)
SORETHRT	Prodromal pharyngitis (0 = No, 1 = Yes, 9 = Unknown)
VOMITING	Prodromal vomiting or nausea (0 = No, 1 = Yes, 9 = Unknown)
WEAKNESS	Limb paralysis (0 = No, 1 = Yes, 9 = Unknown)
WEAKONST	Time between onset of limb paralysis and admission date (days or weeks, 999 = Unknown)

## APPENDIX H

### Algorithms for Score Variables

These algorithms were programmed in SPSS/PC+ and used to compute scores for identifying possible cases of polio in three screening phases. (Refer Appendix G for coding scheme of relevant variables.)

```
COMPUTE CALCAGE = RND((YRMODA(DOAYR,DOAMO,DOADA) -  
YRMODA(DOBYR,DOBMO,DOBDA)) /365.25).
```

```
COMPUTE AGEX = 0.  
IF (CALCAGE LT 6) AGEX = 1.
```

```
IF (MISSING (WEAKNESS)) WEAKNESS = 0.  
IF (MISSING (CRANIALS)) CRANIALS = 0.  
IF (MISSING (WEAKONST)) WEAKONST = 0.  
IF (MISSING (CRANONST)) CRANONST = 0.  
IF (WEAKNESS = 1 AND WEAKONST GT 0) DAYSONST = WEAKONST.  
IF ((WEAKNESS EQ 0 OR WEAKONST EQ 0) AND (CRANIALS = 1 AND  
CRANONST GT 0)) DAYSONST = CRANONST.
```

```
COMPUTE DAYSRSDL = (YRMODA(DRPYR,DRPMO,DRPDA) -  
YRMODA(DOAYR,DOAMO,DOADA)) + DAYSONST.
```

```
COMPUTE RESIDUAL = 0.  
IF (RESDLPAR EQ 1 AND DAYSRSDL GE 60) RESIDUAL = 1.
```

```
COMPUTE FEVPROG = 0.  
IF (FEVER = 1 OR PROGPARG = 1) FEVPROG = 1.
```

```
COMPUTE HXURTI = 0.
```

```
IF (CORYZA = 1 AND SORETHRT = 1) HXURTI = 1.  
IF (CORYZA = 1 OR SORETHRT = 1) HXURTI = 1.  
IF (CORYZA = 0 AND SORETHRT = 0) HXURTI = 0.  
IF (CORYZA = 0 AND SORETHRT = 9) HXURTI = 0.  
IF (CORYZA = 9 AND SORETHRT = 0) HXURTI = 0.  
IF (CORYZA = 9 AND SORETHRT = 9) HXURTI = 0.
```

COMPUTE HXGIT = 0.

IF (DIARRHEA = 1 AND VOMITING = 1) HXGIT = 1.

IF (DIARRHEA = 1 OR VOMITING = 1) HXGIT = 1.

IF (DIARRHEA = 0 AND VOMITING = 0) HXGIT = 0.

IF (DIARRHEA = 0 AND VOMITING = 9) HXGIT = 0.

IF (DIARRHEA = 9 AND VOMITING = 0) HXGIT = 0.

IF (DIARRHEA = 9 AND VOMITING = 9) HXGIT = 0.

COMPUTE MONOPLEG = 0.

IF (LPOWRRUL = 5 AND LPOWRLUL = 5 AND LPOWRRLL = 5 AND  
LPOWRLLL < 5) MONOPLEG = 1.

IF (LPOWRRUL = 5 AND LPOWRLUL = 5 AND LPOWRRLL < 5 AND  
LPOWRLLL = 5) MONOPLEG = 1.

IF (LPOWRRUL = 5 AND LPOWRLUL < 5 AND LPOWRRLL = 5 AND  
LPOWRLLL = 5) MONOPLEG = 1.

IF (LPOWRRUL < 5 AND LPOWRLUL = 5 AND LPOWRRLL = 5 AND  
LPOWRLLL = 5) MONOPLEG = 1.

### Screening for possible missed polio cases

1. The value of a score variable is the sum of scores for the corresponding composite variables.
2. In Phase 1, a case was identified as a possible case of missed polio if the total score was above a chosen cutoff score; 2, 2, and 4 respectively. For Phases 2 and 3, a case was identified as a possible case of missed polio if the total score was equal to the number of variables used in computing the specific score (i.e., a case had to have positive values for all variables)

#### Screening phase 1 (Based on classical presentation of polio)

```
COMPUTE SCOREA1 = FEBRILE + HXURTI + HXGIT.  
COMPUTE SCOREA2 = PATTNPAR + PROGPAR + RESIDUAL.  
COMPUTE SCOREA3 = 0.  
IF (SCOREA1 GE 2 OR SCOREA2 GE 2) SCOREA3 = SCOREA1 + SCOREA2.
```

#### Screening phase 2 (Based on Dietz et al)

```
COMPUTE SCOREB1 = MONOPLEG + LEVELPAR.  
COMPUTE SCOREB2 = LEVELPAR + FEVER.  
COMPUTE SCOREB3 = MONOPLEG + LEVELPAR + FEVER.  
COMPUTE SCOREB4 = MONOPLEG + LEVELPAR + PROGPAR.  
COMPUTE SCOREB5 = LEVELPAR + PROGPAR + FEVER.
```

#### Screening phase 3 Based on (Andrus et al)

```
COMPUTE SCOREC1 = AGEX + FEVPROG.  
COMPUTE SCOREC2 = AGEX + FEVPROG + RESIDUAL.  
COMPUTE SCOREC3 = AGEX + FEBRILE + HXGIT + PROGPAR + RESIDUAL.  
COMPUTE SCOREC4 = AGEX + FEVER + HXGIT + PROGPAR + RESIDUAL.
```

APPENDIX I

Possible Missed Cases of Poliomyelitis

Cases identified by screening as possible missed cases of polio (n =35)  
 (Y denotes a positive identification)

Case	A1	A2	A3	B1	B2	B4	B5	C1	C2	C3	C4	Case
101								Y		Y		101
102					Y		Y	Y	Y	Y		102
103	Y	Y	Y					Y	Y	Y	Y	103
104		Y										104
106										Y		106
107	Y							Y	Y	Y		107
108		Y		Y		Y						108
109		Y										109
110	Y											110
113		Y						Y				113
114		Y										114
115	Y							Y				115
117	Y	Y	Y					Y	Y			117
118	Y											118
202	Y											202
303		Y										303
314		Y										314
315		Y										315
318		Y										318
319	Y											319
327	Y											327
331	Y	Y	Y									331
333	Y											333
335	Y											335
336	Y											336
337	Y											337
339	Y											339
340		Y										340
401	Y											401
405		Y										405
414		Y										414
417		Y										417
421		Y										421
425		Y										425
427	Y											427

## APPENDIX J

### Written Case Scenarios and Sample Questionnaire for Physician Survey

An example of the complete survey questionnaire follows with copies of the original covering letter, and reminder letters.

#### **CASE 1** (High sensitivity pediatric polio)

A 3-year old male child presents with weakness in the right leg. His aunt gives a 10-day history of a febrile flu-like illness with symptoms lasting approximately one week. At the onset of the febrile illness, the patient had experienced some vomiting and diarrhoea, however, that settled after two days. Yesterday morning, he was noticed to have an unusual gait and "seemed to be favouring the right leg". Today he is unable to walk or stand without support. There is no complaint of pain and no known history of recent trauma. The patient is the last of six siblings and has recently immigrated to Canada with his aunt from Egypt. In the last month the patient has received a full schedule of immunizations for his age as his previous immunization status was not known. The patient's aunt is unaware of a similar history in the patient's immediate family.

On examination, the patient has a mildly elevated temperature but otherwise his general condition is good. The muscle bulk is normal in all limbs and tone is normal except for a markedly reduced muscle tone in the right lower limb. There is a moderate weakness of the right lower limb associated with absent deep tendon reflexes. All other reflexes are normal and plantars are downgoing. There is no apparent loss of sensation and anal tone is normal. There are no other significant findings on examination of other systems.

### **CASE 2 (High sensitivity adult polio)**

A 28-year old woman presents with a five day history of fever, myalgia, headache, and nausea. Her most pressing complaint is weakness in the right lower limb which started the previous day and has grown increasingly worse. She also complains of a numbness and occasional "tremors" in the affected limb. On further questioning, you find she had a diarrhoea at the onset of her symptoms lasting for two days. The patient is unaware of contact with any communicable diseases but states that she works in a day care nursery.

On examination, the patient is mildly febrile but her general condition is otherwise good. Clinical signs are found only on neurological examination. The examination of cranial nerves is normal. No fasciculations are noted in the limbs. Muscle tone is markedly reduced in the right lower limb but normal in all other limbs. Power is normal in the upper limbs. She has a muscle power of grade 2/5 in the right lower limb and 5/5 in the left lower limb. There are absent deep tendon reflexes in the right leg but all other reflexes, including both plantars are normal. There is no sensory deficit and anal sphincter control is normal.

### **CASE 3 (Low sensitivity adult polio)**

Mr. K., a 31-year old man presents with complaints of myalgia, and headache on and off for a week. At the onset of these symptoms he had a single episode of vomiting. Two days prior to presenting at the hospital, he developed some pain and weakness in his left leg. The patient feels that the weakness has grown increasingly worse over the last 24 hours. There are no significant points in Mr. K.'s past medical history, however, he is unable to give any detailed information regarding possible childhood illnesses. Mr. K. lives in the suburbs of Ottawa with his wife, 2-year old child and 5-month old baby.

On examination, his general condition is good. The cranial nerve examination is normal. No muscle tenderness is elicited and there is no sensory deficit. Muscle tone is moderately reduced in the left lower limb and normal in all other limbs. Power is normal in both upper limbs, grade 4/5 to 4<sup>+</sup>/5 in the right lower limb, and grade 3/5 in the left lower limb. The deep tendon reflexes are markedly diminished in the left lower limb but only slightly so in the right lower limb. Plantar response is flexor bilaterally.

#### **CASE 4 (Low sensitivity pediatric polio)**

A 12-year old girl presents with complaints of myalgia, and headache on and off for a week. At the onset of these symptoms she had a single episode of vomiting. Two days prior to presenting at the hospital, she developed some pain and weakness in her left leg. The patient feels that the weakness has grown increasingly worse over the last 24 hours. She denies any impairment of bowel or bladder function. There are no significant childhood illnesses in her past medical history. The patient lives in the suburbs of Ottawa with her parents and three siblings whose ages range from five months to 10 years.

On examination, her general condition is good. The cranial nerve examination is normal. No muscle tenderness is elicited and there is no sensory deficit. Muscle tone is moderately reduced in the left lower limb and normal in all other limbs. Power is normal in both upper limbs, grade 4/5 to 4<sup>+</sup>/5 in the right lower limb, and grade 3/5 in the left lower limb. The deep tendon reflexes are markedly diminished in the left lower limb but only slightly so in the right lower limb. Plantar response is flexor bilaterally.

### CASE 5 (GBS)

A teenage girl presents with a four day history of bilateral weakness in the lower limbs. There are no other complaints associated with the onset of her illness, however she does have a history of an upper respiratory infection three weeks before. She was apparently able to carry on most of her normal activities initially as the weakness was very mild, however, her mother reports a rapid progression in weakness during the last 36 hours or so. She now complains of numbness and a tingling sensation in both legs. The patient has had a "normal childhood" and her immunizations are up to date.

The patient's general condition is good. Cranial nerve examination is normal. The clinical findings include a flaccid weakness with power grade 3/5 to 4/5 in both lower limbs. The muscle weakness is slightly more marked in the proximal than the distal muscles. There is a global absence of deep tendon reflexes in the lower limbs. There appears to be a mild subjective impairment of sensation in the lower limbs, however, there is no clear sensory level in the trunk. There are no sensory or motor deficits in the upper limbs. Tendon reflexes in the upper limbs are all normal and the abdominal reflex is normal in all quadrants. Anal tone is normal. The plantar response is absent bilaterally.

### CASE 6 (Transverse myelitis)

A teenage girl presents with a 36-hour history of gradually increasing weakness in both lower limbs associated with urinary retention for more than 12 hours. Additional symptoms include a mild fever of three days duration and a moderate to severe pain at her mid-back. There is no history of trauma and her past medical history has been otherwise uneventful. She has been an active participant in both school and community activities.

The patient has a mildly elevated temperature but an otherwise satisfactory general condition. A full bladder is palpable on abdominal examination. She has a marked hypotonia in both lower limbs and there is also a complete loss of sensation in the lower limbs and up to the T10 segmental level. Anal tone is absent. There is a symmetrical weakness in both lower limbs with power grade assessed to be 3/5. Tendon reflexes are generally diminished in the lower limbs with extensor plantar responses. There is also a loss of the abdominal reflex below T10. All other findings on neurological examination are normal including tone, power and tendon reflexes in the upper limbs.

#### CASE 7 (Botulism)

A teenage boy presents with complaints of dizziness, blurred vision, a dry mouth and dry, sore throat, and weakness of both arms. On further questioning, you find that his illness started two days ago with an initial complaint of abdominal pain, constipation and nausea. The following day he vomited twice and also began to experience a weakness of his neck muscles which spread to his arms. He has had a urinary retention for more than 24 hours. There has been no fever associated with the illness. The patient received treatment for an infected leg wound about a week ago but otherwise has had no other recent illnesses.

On examination he is afebrile and has bilateral nonreactive dilated pupils, a dry tongue and red, dry oral mucous membranes. He also has a recent wound scar on his left shin with no signs of current infection. There is no apparent reduction in respiratory function. There is, however, a mild abdominal distention with diminished bowel sounds and also a palpable bladder. Cranial nerve examination reveals bilateral diplopia, dysarthria and an absent gag reflex. There is a mild weakness of neck muscles. Muscle power is grade 3/5 to 4/5 in both upper limbs and 5/5 in both lower limbs. There are normal deep tendon reflexes in all limbs and sensation is intact. Plantar responses are normal.

Standard questions for all cases:

Based on the information provided state your initial clinical impression (primary diagnosis) and not more than three differential diagnoses in descending order of likelihood.

Primary Diagnosis:

Differential Diagnoses:

(a)

(b)

(c)

In relation to this case scenario, and assuming you are not immediately referring the patient and have access to appropriate investigative procedures, what investigations would you request? Please list these in order of importance.

(a)

(b)

(c)

(d)

In the context of this patient's care, would you seek an opinion or request a consultation from another physician? If yes, whom i.e., what service(s) would you consult?

(a)

(b)

(c)

February 22nd, 1993

Dr. ... (Address)

Dear Dr. ...

We are conducting a regional survey of physicians' management of acute paralytic syndromes. This pilot survey is an important component of a broader study of the epidemiology and management of acute, non-traumatic, flaccid paralysis in Canada. This study constitutes an MSc. (Epidemiology) Thesis Research and is the result of a collaborative effort between the Laboratory Centre for Disease Control and the University of Ottawa.

The data from the complete study will assist in the accurate interpretation and documentation of national health data. We are currently surveying all practising neurologists, general internists and infectious disease specialists in Ottawa. Although your participation is completely voluntary, it is of utmost importance that each questionnaire be completed and returned.

We enclose three case scenarios describing paralysis of acute onset. Please take a few moments to review the cases and answer the questions on diagnosis and patient management.

Responses will be kept confidential and no personal identification is required from you. Questionnaire identification numbers have been added with the sole purpose of updating our mailing list thus avoiding unnecessary follow-up mail. Please return your completed questionnaire in the enclosed pre-paid, addressed envelope by March 5th, 1993.

We greatly appreciate your time and cooperation in assisting with the study. Thank you very much.

Yours sincerely,

Adwoa Bentsi-Enchill MB. ChB.  
M.Sc. Thesis Student  
Dept. of Epidemiology  
(957-0324)

Ian McDowell PhD.  
Professor  
Dept. of Epidemiology  
(787-6616)

March 4th, 1993

Dr. ... (Address)

Dear Dr. ...

Please accept this as a reminder of my survey on physicians' management of acute paralytic syndromes in Canada. The survey material was mailed to you with an explanatory letter dated February 22nd, 1993. As previously stated, the study constitutes an MSc. (Epidemiology) Thesis Research at the University of Ottawa. As such each completed questionnaire received is of great importance to the completeness of the data I am collecting.

I am requesting that you take a few moments now to complete and return the questionnaire in the pre-paid, addressed envelope provided. If you have already mailed the completed questionnaire please accept my thanks and ignore this reminder.

Yours sincerely,

Adwoa Bentsi-Enchill MB. ChB.  
M.Sc. (Epidemiology) Student  
(957-0324)

April 8th, 1993

Dr. ... (Address)

Dear Dr. ...

You were recently sent a questionnaire on physicians' management of acute paralytic syndromes. **If you have already returned the completed questionnaire, this letter indicates your response has not yet been received. Please accept my thanks, however, and ignore this reminder.**

This pilot survey forms part of a broader study of the epidemiology of selected, acute, non-traumatic paralysis in Canada. As a result of some of the feedback received on the survey, I wish to make the following points regarding your selection to be part of the survey.

**All physicians who may treat (individually or as part of a team) a patient presenting with an acute, non-traumatic, paralysis, possibly with a history of a febrile illness, are potentially eligible for this survey. Of these we have currently selected all practising neurologists, general internists and infectious disease specialists in Ottawa to be surveyed.**

If you have been unable to complete the questionnaire, I would request that you review the first part of the enclosed questionnaire and proceed appropriately.

As previously indicated, the complete study constitutes an MSc. (Epidemiology) Thesis Research and is the result of a collaborative effort between the Laboratory Centre for Disease Control and the University of Ottawa.

Again, I greatly appreciate your time and cooperation in assisting with the study and look forward to receiving your response soon. Thank you.

Yours sincerely,

Adwoa Bentsi-Enchill MB. ChB.  
M.Sc. Thesis Student  
(957-1352/957-0324)

## SURVEY OF THE MANAGEMENT OF ACUTE PARALYTIC SYNDROMES

Thank you for completing this survey. The survey is aimed at obtaining information on certain aspects of the current management of selected acute paralytic syndromes in Canada.

**All physicians in Ottawa who may treat (individually or as part of a team) a patient presenting with an acute, non-traumatic, paralysis, possibly with a history of a febrile illness, are potentially eligible for this survey.**

Enclosed are three case scenarios describing paralysis of acute onset. Please review the cases and answer the questions on diagnosis and patient management. Responses will be kept confidential and no personal identification is required. Identification numbers have been added with the sole purpose of updating our mailing list. Please return your completed questionnaire in the enclosed pre-paid, addressed envelope.



Please start by answering the following question and proceed appropriately. Thank you.



In your medical practice, would you ever treat (individually or as part of a team) a patient presenting with an acute non-traumatic paralysis, with or without a history of a febrile illness?

Yes \_\_\_\_\_

Please review the following hypothetical case scenarios and answer the related questions.

No \_\_\_\_\_

Please turn to page 5, questions 2 and 3.

(Identification Code)

## CASE A

A 3-year old male child presents with weakness in the right leg. His aunt gives a 10-day history of a febrile flu-like illness with symptoms lasting approximately one week. At the onset of the febrile illness, the patient had experienced some vomiting and diarrhoea, however, that settled after two days. Yesterday morning, he was noticed to have an unusual gait and "seemed to be favouring the right leg". Today he is unable to walk or stand without support. There is no complaint of pain and no known history of recent trauma. The patient is the last of six siblings and has recently immigrated to Canada with his aunt from Egypt. In the last month the patient has received a full schedule of immunizations for his age as his previous immunization status was not known. The patient's aunt is unaware of a similar history in the patient's immediate family.

On examination, the patient has a mildly elevated temperature but otherwise his general condition is good. The muscle bulk is normal in all limbs and tone is normal except for a markedly reduced muscle tone in the right lower limb. There is a moderate weakness of the right lower limb associated with absent deep tendon reflexes. All other reflexes are normal and plantars are downgoing. There is no apparent loss of sensation and anal tone is normal. There are no other significant findings on examination of other systems.

Based on the information provided state your initial clinical impression (primary diagnosis) and not more than three differential diagnoses in descending order of likelihood.

Primary Diagnosis:

Differential Diagnoses:

- (a)
- (b)
- (c)

In relation to this case scenario, and assuming you are not immediately referring the patient and have access to appropriate investigative procedures, what investigations would you request? Please list these in order of importance.

- (a)
- (b)

(c)

(d)

In the context of this patient's care, would you seek an opinion or request a consultation from another physician? If yes, whom i.e., what service(s) would you consult?

(a)

(b)

(c)

## CASE B

A 12-year old girl presents with complaints of myalgia, and headache on and off for a week. At the onset of these symptoms she had a single episode of vomiting. Two days prior to presenting at the hospital, she developed some pain and weakness in her left leg. The patient feels that the weakness has grown increasingly worse over the last 24 hours. She denies any impairment of bowel or bladder function. There are no significant childhood illnesses in her past medical history. The patient lives in the suburbs of Ottawa with her parents and three siblings whose ages range from five months to 10 years.

On examination, her general condition is good. The cranial nerve examination is normal. No muscle tenderness is elicited and there is no sensory deficit. Muscle tone is moderately reduced in the left lower limb and normal in all other limbs. Power is normal in both upper limbs, grade 4/5 to 4<sup>+</sup>/5 in the right lower limb, and grade 3/5 in the left lower limb. The deep tendon reflexes are markedly diminished in the left lower limb but only slightly so in the right lower limb. Plantar response is flexor bilaterally.

Based on the information provided state your initial clinical impression (primary diagnosis) and not more than three differential diagnoses in descending order of likelihood.

Primary Diagnosis:

Differential Diagnoses:

(a)

(b)

(c)

In relation to this case scenario, and assuming you are not immediately referring the patient and have access to appropriate investigative procedures, what investigations would you request? Please list these in order of importance.

(a)

(b)

(c)

(d)

In the context of this patient's care, would you seek an opinion or request a consultation from another physician? If yes, whom i.e., what service(s) would you consult?

(a)

(b)

(c)

## CASE C

A teenage girl presents with a four day history of bilateral weakness in the lower limbs. There are no other complaints associated with the onset of her illness, however she does have a history of an upper respiratory infection three weeks before. She was apparently able to carry on most of her normal activities initially as the weakness was very mild, however, her mother reports a rapid progression in weakness during the last 36 hours or so. She now complains of numbness and a tingling sensation in both legs. The patient has had a "normal childhood" and her immunizations are up to date.

The patient's general condition is good. Cranial nerve examination is normal. The clinical findings include a flaccid weakness with power grade 3/5 to 4/5 in both lower limbs. The muscle weakness is slightly more marked in the proximal than the distal muscles. There is a global absence of deep tendon reflexes in the lower limbs. There appears to be a mild subjective impairment of sensation in the lower limbs, however, there is no clear sensory level in the trunk. There are no sensory or motor deficits in the upper limbs. Tendon reflexes in the upper limbs are all normal and the abdominal reflex is normal in all quadrants. Anal tone is normal. The plantar response is absent bilaterally.

Based on the information provided state your initial clinical impression (primary diagnosis) and not more than three differential diagnoses in descending order of likelihood.

Primary Diagnosis:

Differential Diagnoses:

(a)

(b)

(c)

In relation to this case scenario, and assuming you are not immediately referring the patient and have access to appropriate investigative procedures, what investigations would you request? Please list these in order of importance.

(a)

(b)

(c)

(d)

In the context of this patient's care, would you seek an opinion or request a consultation from another physician? If yes, whom i.e., what service(s) would you consult?

(a)

(b)

(c)

**PLEASE ANSWER THE FOLLOWING QUESTIONS AFTER REVIEWING THE  
CASE SCENARIOS AND ANSWERING THE RELATED QUESTIONS.**

1. In your regular practice, how often do you see the kind of patients simulated in the case scenarios you have just reviewed?

Never or rarely \_\_\_\_\_

Occasionally \_\_\_\_\_

Frequently \_\_\_\_\_

Always \_\_\_\_\_

2. Indicate your specialty/specialties.

Neurology \_\_\_\_\_

Internal Medicine \_\_\_\_\_

Infectious diseases \_\_\_\_\_

Other (specify) \_\_\_\_\_

**This last optional question is only for physicians who have not completed the questionnaire.**

3. Do you wish to give any other reasons why you have not completed this questionnaire? If so, kindly state them below.

**THANK YOU FOR RESPONDING TO THIS SURVEY.**

**PLEASE RETURN THE QUESTIONNAIRE IN THE ENCLOSED PRE-PAID,  
ADDRESSED ENVELOPE.**