Subacute Sclerosing Panencephalitis (SSPE)

Public Health Notification Disease Management Guideline Revision Dates

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Case Definition

**Clinical Case**
Diagnosis is usually made clinically, as viral isolation is difficult and there is a paucity of the immune response. All other possible etiologies of encephalitis should be ruled out. May be diagnosed by the demonstration of high measles titres in serum and CSF in the presence of a compatible illness, also role of brain biopsy (characteristic histopathology), EEG (characteristic pattern), and possibly PCR/molecular diagnostic techniques (and prior measles information in these individuals usually remarkable only for early age of occurrence prior to 2 years of age).

**NOTE:** SSPE is chronic encephalitis after measles in normal hosts. It is rare, developing in about 1/100,000 hosts several years after infection, and covers a wide spectrum of severity.
Reporting Requirements

1. Physicians, Health Practitioners and others
   Physicians, health practitioners and others listed in Sections 22(1) or 22(2) of the Public Health Act shall notify the Medical Officer of Health (MOH) (or designate) of all confirmed cases in the prescribed form by mail, fax or electronic transfer within 48 hours (two days).

2. Laboratories
   Although viral isolation to support the diagnosis is rare, when available, all laboratories, including regional laboratories and the Provincial Laboratory for Public Health (PLPH) shall in accordance with Section 23 of the Public Health Act, report all positive laboratory results by mail, fax or electronic transfer within 48 hours (two days) to the:
   - Chief Medical Officer of Health (CMOH) (or designate),
   - MOH (or designate) and
   - Attending/ordering physician.

3. Alberta Health Services
   - The MOH (or designate) of the zone where the case currently resides shall forward the preliminary Notifiable Disease Report (NDR) of all clinical cases to the CMOH (or designate) within two weeks of notification and the final NDR (amendment) within four weeks of notification.
   - For out-of-zone reports, the MOH (or designate) first notified shall notify the MOH (or designate) where the client currently resides by mail, fax or electronic transfer and fax a copy of the positive laboratory report, if available, within 48 hours (two days).
   - For out-of-province and out-of-country reports, the following information should be forwarded to the CMOH (or designate) by phone, fax or electronic transfer within 48 hours (two days) including:
     - name,
     - date of birth,
     - out-of-province health care number,
     - out-of-province address and phone number,
     - attending physician (locally and out-of-province) and
     - positive laboratory report, if available (faxed).
**Etiology (1)**
Subacute Sclerosing Panencephalitis (SSPE) is caused by the accumulation of incomplete measles virus that cannot be cleared by B or T cell mechanisms. The measles genomes are larger and contain multiple mutations. It begins in the cortical grey matter, progresses to subcortical grey and white matter, then to lower structures. SSPE has also been reported in children who did not have a history of natural measles but did receive measles vaccine. Some of these cases may have had unrecognized measles infection under one year of age. (2)

**Clinical Presentation (1,2)**
SSPE is a slow viral infection of the (central nervous system) CNS by a mutant measles virus. Disease is insidious in onset and is characterized by progressive physical and mental deterioration, myoclonia, convulsions, coma, and death.

The first clinical stage involves subtle behavioural changes including aggression and withdrawal. This may be followed by overtly bizarre behaviour and dementia. The second clinical stage involves neurological changes including seizures, movement disorders, and optic changes. Dementia progresses to stupor and coma in either flaccid or spastic postures. A few patients will experience remissions and exacerbations. The disease is almost always fatal.

Onset of SSPE can occur anywhere from 7-12 years after the original measles illness. The average age of onset is nine years and few individuals live longer than three years after diagnosis.(3)

**Diagnosis**
SSPE is only one of a number of degenerative neurological diseases therefore; it requires a high level of diagnostic suspicion. In general, the diagnosis is made by ruling out other degenerative neurological diseases. Laboratory investigation of serum and CSF should be used to determine an elevated antibody titre (IgG) to measles.

EEG may show progressive changes which are typical of SSPE and parallel the slow deterioration of CNS functions. Additional imaging studies (CT and MRI) may be of some value. Brain biopsy material can be examined for measles virus RNA.

With the elimination of indigenous measles disease in Canada due to widespread measles immunization programs, it is important that brain tissue specimens be collected post-humously on all suspect cases of SSPE for virus detection.(4)

Specimens are forwarded by the PLPH to the Viral Exanthemata Laboratory at the National Microbiology Laboratory in Winnipeg.(5) The Viral Exanthemata Lab performs vaccine versus wild-type strain differentiation for measles, rubella, and varicella zoster viruses.(1)

**Epidemiology**
**Reservoir**
Not applicable.

**Transmission**
SSPE is not transmissible.

**Incubation Period (1)**
The average time between exposure and onset of SSPE ranges from 7-12 years.
Period of Communicability
Not communicable.

Host Susceptibility (1)
Previous history of measles disease increases the risk. Over 50% of cases had measles disease before the age of two years. A study done in Israel identified the following as risk factors for SSPE: early measles infection, large family, overcrowding in the home, older age of the mother, higher birth order, fewer years of schooling of the parents, fewer cultural activities, and rural place of birth.

Occurrence
General (1)
Before measles immunization, SSPE was a rare complication of measles infection at 1/100,000 cases. Since the introduction of measles immunization, the rate of SSPE has declined to 0.06/1,000,000 in the United States. Incidence has remained high in the Middle East and India where over 20 cases of SSPE per million people have been reported annually. More males are affected than females (male to female ratio of 4:1). The disease generally occurs in children and adolescents.

Canada (4-6)
SSPE is monitored through the Canadian Paediatric Surveillance Program (CPSP). From 1995 to 1998, no definite cases were reported. In 1999, two cases were reported with very high serum and CSF IgG ratios in the presence of typical clinical manifestations (1). Both cases were males, one Canadian born (immunized) and one foreign born (immunization status unknown). Both had a history of possible early measles infection and both cases were confirmed by serological testing. The Canadian Immunization Guide reports that no cases from whom measles virus was isolated had a vaccine strain.

Alberta (7)
SSPE became reportable in Alberta in 1983. From 1983 to 1994, a total of six cases were reported in the province (three in 1984, and one in 1986, 1990, and 1994). There were no cases reported from 1995 to 2003.

Key Investigation
Single Case/Household Cluster
- Assess measles immunization history.
- Determine measles disease history.

Control
Management of a Case
- Supportive therapy.

Treatment of a Case
- Treatment is generally symptomatic i.e., anticonvulsants may be used.
- Antiviral medication may slow the progression of the disease.

Management of Contacts
- No follow up required.
Preventive Measures (8)

- Prevent wild measles through immunization.
  - Preschool children should receive immunization at 12 months and 4–6 years of age as per the current Alberta Immunization manual. Immunization given between 12 and 15 months of age induces immunity in 94–98% of recipients. The second dose increases the immunity level to about 99%.
  - Two doses of measles-containing vaccine (MMR) given at least one month apart are recommended for children who are one year of age or older and:
    - had vaccine given prior to their first birthday,
    - have no immunization record,
    - do not have reliably recorded measles records (i.e., immigrants),
    - have had immune globulin (IG) administered at the same time as live measles vaccine,
    - have had live measles vaccine administered within five months of being given IG, or
    - were given an inadequate vaccine dosage of measles vaccine.

- Review immunization records of grade nine students and ensure that all individuals have received two doses of a measles-containing vaccine before leaving school as per the current Alberta Immunization Manual.

- A single dose of measles-containing vaccine should be offered to adults born in or after 1970 (Canadian born) or are foreign born and who have not previously received a measles-containing vaccine or had natural measles infection. Individuals at the greatest risk of exposure include those who:
  - know they never received measles vaccine,
  - are healthcare employees who provide direct patient care,
  - are students entering post-secondary institutions, or
  - are travellers who are going to an area where measles is common (i.e., outside of North America, South America or the Caribbean). Vaccines required for travel are not provincially funded.
References


