

2.2 Botulism

Etiology

Toxins produced by *Clostridium botulinum*, a spore-forming obligate anaerobic bacillus, cause botulism. Only a few nanograms of the toxin can cause illness. Human botulism is primarily caused by the strains of *C. botulinum* that produce toxin types A, B and E. Strains of *C. baratii*, which produce type F toxin and *C. butyricum* which produce type E toxin, have also been implicated in human botulism.

Type G has been isolated from soil and autopsy specimens but an etiologic role has not been established. Most cases of infant botulism have been caused by type A or B. Botulinum toxin is considered the most potent lethal substance known to man.

Conditions that promote germination and growth of *C. botulinum* spores include absence of oxygen (anaerobic conditions), low acidity (pH > 4.6), temperatures > 4°C, and high moisture content.

Case Definition

Confirmed Case

A confirmed case requires laboratory definitive evidence with clinical evidence¹ or, in the case of foodborne botulism, clinical evidence and consumption of the same suspect food as an individual that has laboratory confirmed botulism

Foodborne Botulism

Laboratory confirmation of intoxication with clinical evidence¹:

- detection of botulinum toxin in serum, stool, gastric aspirate or food

OR

- isolation of *C. botulinum* from stool or gastric aspirate

AND

- clinical evidence and indication the client ate the same suspect food as an individual with laboratory confirmed botulism

Wound Botulism

Laboratory confirmation of infection:

- lab detection of botulinum toxin in serum

¹Foodborne: Clinical illness is characterized by blurred vision, dry mouth and difficulty swallowing and speaking. Descending and symmetric paralysis may progress rapidly, often requiring respiratory support.

Wound: Clinical illness is characterized by diplopia, blurred vision, and bulbar weakness. Symmetric paralysis may progress rapidly.

Infant: Clinical illness in infants is characterized by constipation, loss of appetite, weakness, altered cry and loss of head control

OR

- isolation of *C. botulinum* from a wound

AND

- presence of a freshly infected wound in the 2 weeks before symptoms and no evidence of consumption of food contaminated with *C. botulinum*

Infant Botulism

Laboratory confirmation with symptoms compatible with botulism in a person less than one year of age:

- detection of botulinum toxin in stool or serum

OR

- isolation of *C. botulinum* from the patient's stool, or at autopsy

Probable Case**Foodborne**

A probable case requires clinical evidence and consumption of a suspect food item in the incubation period (12-48 hours).

Clinical Presentation

The classic presentation is that of a person who develops acute, bilateral cranial neuropathies along with symmetrical descending weakness. The following are cardinal features present in botulism cases:

- Fever is absent (unless a complicating infection occurs).
- The neurologic manifestations are symmetrical.
- The patient remains responsive.
- The heart rate is normal or slow in the absence of hypotension.
- Sensory deficits do not occur (except for blurred vision).

There are four naturally occurring forms of botulism – foodborne (the classic form), wound, infant and adult intestinal toxemia botulism. In addition, two forms of botulism that have also been found and are not naturally occurring are:

Inhalation Botulism

This is a result of inhaling aerosolized botulism neurotoxin.

Iatrogenic Botulism

It is caused by accidental injection of the botulism neurotoxin into the systemic circulation instead of the intended therapeutic location.

Foodborne Botulism

It results from the ingestion of preformed toxin present in contaminated food. Symptoms usually develop between 12 and 36 hours after toxin ingestion. The initial complaints may be gastrointestinal and can include nausea, vomiting, abdominal cramps or diarrhea. Constipation is more likely to occur after the onset of neurologic symptoms.

Dry mouth, blurred vision, and diplopia are the earliest neurologic symptoms. Lower cranial nerve dysfunction manifests as; dysphasia (difficulty speaking), dysarthria (difficulty articulating), and hypoglossal (tongue) weakness. Symmetric weakness then descends to the upper extremities, the trunk, and the lower extremities. Respiratory dysfunction may require ventilation. Autonomic problems may also include; alterations in resting heart rate, loss of responsiveness to hypotension or postural change, hypothermia, and urinary retention. Recovery may not begin for up to 100 days and may take months to conclude. With critical care management, the death rate is about 14%.

Wound botulism

It lacks the prodromal gastrointestinal symptoms of the foodborne form, but is otherwise similar in presentation. Fever, if present, reflects wound infection rather than botulism. *C. botulinum* infection may also produce skin abscesses.

Botulism has also been reported in individuals with sinusitis as a result of cocaine inhalation. The reported incubation period varies from four to 14 days. The case fatality rate for wound botulism is approximately 15%.

Infant botulism

It is generally the most common form of botulism and affects infants under one year of age, with the majority of cases occurring between six weeks and six months old. Ingested spores germinate in the intestine, where they produce bacteria which then reproduce in the gut and release toxin. Clinical symptoms start with constipation and may include loss of appetite, weakness, altered cry, weak suck, drooling and a significant loss of head control. The illness has a wide spectrum of clinical severity, ranging from mild illness with gradual onset (that never requires hospitalization) to sudden infant death. Progression is more severe in infants that are younger than two months old.

Upper airway obstruction may be the initial sign, and is the major indication for intubation. In severe cases, the condition progresses to include cranial neuropathies and respiratory weakness, with respiratory failure occurring in about 50% of diagnosed cases. The condition progresses for one to two weeks, and then stabilizes for another two to three weeks before recovery starts. Relapses of infant botulism may occur.

Diagnosis

Botulism diagnosis is based primarily on clinical presentation and should be suspected in a person with acute onset of gastrointestinal, autonomic (such as dry mouth or difficulty focusing eyes), and cranial-nerve dysfunction (diplopia, dysarthria, dysphagia). The diagnosis is even more likely if the patient has recently eaten home-canned foods or if family members/companions who have shared the same meals are similarly ill. For confirmation on laboratory specimens go to the public health laboratory web site www.publichealthlab.ca or call 709-777-6583.

Epidemiology

Occurrence

Worldwide outbreaks occur primarily in areas which food products are not processed by methods to prevent toxin formation. In Canada botulism is a rare disease with an average of seven cases per year reported between 2001 and 2004. There has recently

been an upsurge of cases of wound botulism in injecting drug users, especially those that use skin or muscle “popping”; the drug is injected subcutaneously or intramuscularly.

Reservoir

Botulism is caused by a nerve toxin, botulin, produced by the bacterium *Clostridium(C) botulinum*. Botulin is the most lethal substance known with less than one microgram sufficient to cause fatal human disease. *C. botulinum*, commonly found in soil, form spores which allow them to survive in a dormant state until exposed to conditions that can support their growth.

Transmission

There are four mechanisms for botulism toxin to enter the body: foodborne, cutaneous, colonization of the gastrointestinal tract, and inhalational. Foodborne botulism results when food contaminated with spores of *C. botulinum* are ingested. Food contamination can occur if foods are preserved or stored under conditions that allow toxin production. Wound botulism results when *C. botulinum* contaminates the wound and produces toxin. Infant botulism occurs when ingested spores colonize the intestinal tract with subsequent absorption of the toxin. Inhalational cases rarely occur naturally but it is the likely route in a large-scale bioterrorism event.

Incubation Period

In foodborne botulism the symptoms usually occur between 12-36 hours after ingestion of the contaminated food with the range from six (6) hours – ten (10) days. Wound botulism has a longer incubation period, usually four (4) – fourteen (14) days. In infant botulism the incubation period is estimated at three (3) to thirty (30) days.

Communicability

No incidence of person to person transmission has been documented.

Control Measures

Management of Cases

Investigations

- Contact the Microbiologist on call at the public health lab on 709-777-6583 for information on collection and transportation of both food and clinical specimens. Notification of a suspicion of a single case of botulism constitutes a public health emergency and may herald the beginning of a larger outbreak.
- Investigation of a suspect case of botulism includes a search for other possible cases, identification of suspect food exposures, and diagnostic testing of both cases and foods as needed.
- Efforts to locate persons exposed to the same suspect food may lead to early diagnosis and/or instituting an emergency product recall.

Foodborne Botulism

- Involve environmental/public health inspectors and CFIA.
- Collect food samples and forward to the laboratory for toxin analysis.
- Take a detailed food history of those who are ill, especially foods consumed within the last two or three days. Include consumption of home-preserved foods and traditionally prepared foods. Even theoretically unlikely foods should be considered. *C. botulinum* may or may not cause container lids to bulge and the contents to have “off-odours.” Other contaminants can also cause cans or bottle lids to bulge.
- Collect clinical samples (sera, gastric aspirates and stool) from patients and, when indicated, from others exposed but not ill and forward immediately, with relevant clinical history, to the Public Health Lab **before** administration of antitoxin.
- Identify individuals who may have been exposed to the same source.

Infant Botulism

- Investigate source, in particular, history of honey consumption.
- Identify individuals who may have been exposed to the same source.

Wound Botulism

- Contact the physician to determine the possible source of infection.
- Determine if history of trauma, or Intravenous Drug Use (IDU) and if possible forward sample of drug for testing.
- Identify individuals who may have been exposed to the same source.

Treatment

- Persons with botulism require immediate emergency medical treatment. Treatment must not await laboratory confirmation.
- Botulism antitoxin and immune globulin are not approved for sale in Canada. The antitoxin is available from the provincial vaccine depot through the Medical Officer of Health (MOH) or on call MOH (1-866-270-7437). Antitoxin requires completion of the Health Canada Special Access Program (SAP) form which is available at phone number: 613-941-2108 or fax 613-941-3194 or email address: http://www.hc-sc.gc.ca/dhp-mps/acces/drugs-droques/index_e.html
- In children less than one year of age (< 1 year) human-derived botulism immune globulin (BabyBIG®) is indicated. BabyBIG® is not a licensed product in Canada. To obtain BabyBIG® the following steps are required:
- The physician must first contact the California Department of Health Services (DHS) Infant Botulism Treatment and Prevention Program on call physician at 510-231-7600 to review the indications for such treatment
- Product is obtained from the California DHS at 510-540-2646
- The use of an unlicensed product in Canada requires approval through Health Canada’s SAP (numbers listed above).
- Treatment focus for wound and foodborne botulism is early administration of the botulism antitoxin/immune globulin with immediate access to an intensive care

setting if ventilatory support is required. Giving antitoxin within 24 hours has been shown to decrease need for and duration of mechanical ventilation.

- Antibiotics do not improve the course of the disease. Aminoglycosides and tetracyclines (which can impair neuron calcium entry), have shown to worsen infant botulism. Thus, it is recommended that antibiotics only be used to treat secondary infections.

Wound Botulism

- Administration of antitoxin.
- The wound should be debrided and/or drainage established.
- Appropriate antibiotics (benzyl penicillin or metronidazole) should be administered.
- The best results are obtained when very large doses of antitoxin are given early in the disease process to provide the body with excess circulating antitoxin.
- Other treatment considerations include enemas, laxatives and other cathartics. If ingestion was recent, may induce vomiting and/or gastric lavage.

Management of Contacts

- Botulism is not passed person to person, therefore, direct contacts of the index case do not require follow-up.
- Those who are known to have consumed the suspected food should be purged with a cathartic, given gastric lavage and high enemas, and kept under close medical observation.
- Providing immunoprophylaxis for asymptomatic individuals strongly suspected of foodborne exposure is recommended. This decision should be weighed carefully due to the risk of adverse effects and sensitization to horse serum.
- If antitoxin is required, it should be given within one to two days of ingestion of the suspect food.

Management of Outbreaks

An outbreak management team should be established to direct and coordinate the investigation as well as address infection prevention and control measures. If the outbreak is limited to one region the region is responsible to manage the outbreak; if more than one region is involved the outbreak will be managed by the province or in consultation with the province.

Education and Preventive Measures

- Wash hands prior to and after preparing food
- Keep all work surfaces, food, utensils, equipment clean during all stages of food preparation especially for canning processes
- Date and label preserves and canned goods and follow proper canning requirements strictly
- Refrigerate all foods labeled “keep refrigerated”

- Methods to control botulism should focus on the inhibition of bacterial growth and toxin production. Manufacturers of commercially canned low acid foods use strict thermal processes which are designed to destroy spores of *C. botulinum*.
- Search for any remaining food from the same source that may be similarly contaminated and submit for laboratory examination.
- The implicated food(s) should be detoxified by boiling before discarding or the containers broken and buried deeply in soil to prevent ingestion by animals.
- Contaminated utensils should be sterilized by boiling or by chlorine disinfection to deactivate any remaining toxins.
- Usual sanitary disposal of feces/diaper from infant cases.
- Educate the public about safe handling of food. For example:
 - Do not use food from damaged or bulging containers. These containers should be returned unopened to the vendor.
 - Foods with off-odours and unusual tastes should not be eaten or 'taste-tested'.
 - Proper storage is one of the keys to food safety. Refrigeration slows down most bacterial growth. Encourage people to check the temperature of their fridge on a regular basis with a refrigerator thermometer. Set the refrigerator at or below 4°C (40°F). Don't overload the fridge - cool air must circulate freely to keep food properly chilled. After grocery shopping, immediately refrigerate or freeze foods as indicated on the label.
 - Storing food in non-airtight containers and at 4°C or lower will prevent growth of the bacterium.
- Boil foods (for at least 10 minutes) and stir home-canned foods to destroy botulinum toxins.
- Take precautions with home-prepared foods stored in oil (e.g., vegetables, herbs and spices). If these products are prepared using fresh ingredients, they must be kept refrigerated (below 4°C) and for no more than 10 days.
- If the above products are purchased from fairs, farmer's markets, roadside stands or have received them as a gift, and prepared more than a week ago, discard them.
- Avoid feeding honey to infants (even pasteurized).
- Provide information to Aboriginal groups regarding food preparation traditions that pose a risk of botulism.
- Promote research to evaluate the safety of traditionally prepared (high-risk) foods, and to identify the precise conditions under which botulinum toxin will be present or absent. Areas to emphasize might include:
 - The importance of refrigeration with home-canning methods,
 - Heating food to temperatures high enough to kill the botulism toxin, and
 - Keep aging meats such as whale, seal or walrus in a cool place (below 4°C), in containers that allow air in and, if aged in oil, keep in a cool place and stir frequently to allow the meat to be in contact with air.
- Where wound botulism occurs in IDUs, educate them regarding safe injection practices.
- Do **NOT** inject into muscle or under the skin.

- Decrease the amount of citric acid used to dissolve the drug. Too much citric acid damages the tissues under the skin leaving them susceptible to bacterial growth.
- Studies have shown that when cocaine is mixed with heroin and when injected at the same site it gives bacteria a better chance to grow so, inject different drugs at different sites on the body.
- Teach IDUs signs and symptoms of infection(s) and to seek physician help especially if infection seems different than ones had in the past.
- Provide fact sheet available at <http://www.inspection.gc.ca/english/fssa/concen/cause/botulisme.shtml>

Reporting Requirements and Procedures

- The laboratory (hospital or public health laboratories) report case/s to the attending physician, the Chief Medical Officer of Health and the Medical Officers of Health (MOH)
- The MOH office will notify, as required, local physicians, nurse practitioners, environmental health officers, community health nurses, communicable disease control nurses (CDCNs) and infection control practitioners (ICP), in the particular region as required for follow-up and case investigation.
- EHO will conduct an investigation of the case under the direction of the MOH and provide case details as per the food history.
- CDCN enters the case details into the electronic reporting system and uses the CNPHI tool, if indicated, for alerts or outbreak summaries

Provincial Disease Control

- Reports the aggregate case data to Public Health Agency of Canada
- Provides an analysis of the case/s with reports in the Quarterly Communicable Disease Report (CDR), also posted on the Public Health website
- Coordinates the response if an outbreak across RHAs (CMOH will likely coordinate an outbreak across RHAs with input from disease control and environmental health.)

References

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