

# BOTULISM

## Foodborne, Wound, and Infant

### Report Immediately

#### ✓ DISEASE AND EPIDEMIOLOGY

### Clinical Description

In humans, botulism occurs in one of three forms: foodborne botulism, wound botulism, and intestinal (infant and adult) botulism. There is a different site of toxin production for each of the forms, but they all share the flaccid paralysis that results from exposure to botulinum toxin. The spectrum of disease also depends on the dosage of toxin. It ranges from mild to rapidly progressing.

#### Foodborne Botulism

Foodborne botulism is a severe poisoning caused by the ingestion of pre-formed *C. botulinum* toxin. The clinical syndrome is dominated by neurologic signs and symptoms, including blurred or double vision, dysphagia, dry mouth, and muscle weakness. Symmetric, descending flaccid paralysis is classic for botulism. Generally, paralysis first affects the cranial nerves, followed by the upper extremities, the respiratory muscles, and finally, the lower extremities. Patients usually require ventilatory support, which is commonly needed for 2–8 weeks. The clinical symptoms are similar no matter which toxin type is responsible for the illness, but type A has been associated with a higher case-fatality rate than types B or E.

#### Wound Botulism

Wound botulism usually presents with the same clinical picture as foodborne botulism. In wound botulism, the organism multiplies in the wound and produces the toxin, which is then absorbed into the bloodstream.

#### Infant Botulism

Infant (intestinal) botulism has a distinctly different clinical presentation than wound or foodborne botulism. In infant botulism, the *C. botulinum* spores are ingested, and the toxin is formed in the intestines. It is a rare disease, confined exclusively to infants <1 year of age. The earliest clinical sign in infants is constipation, which is followed by poor feeding, decreased sucking, lethargy, listlessness, ptosis (drooping eyelids), difficulty swallowing, a weak cry, and lack of muscle tone, giving rise to the term “floppy baby syndrome.” In some cases, respiratory insufficiency and respiratory arrest may occur. Infant botulism presents with a wide range of severity, from mild illness to sudden death. Some studies suggest that infant botulism may be responsible for up to 5% of cases of Sudden Infant Death Syndrome (SIDS).

### Causative Agent

Botulism is caused by a potent neurotoxin produced by *Clostridium botulinum*, an anaerobic, spore-forming bacterium. While bacterium itself is harmless, *C. botulinum*

toxin is one of the most potent, lethal substances known. There are seven types of botulinum toxins (A–G), but human botulism is primarily caused by types A, B, and E.

### **Differential Diagnosis**

Other conditions and diseases that may have similar symptoms to botulism are drug or chemical poisoning, Guillain-Barré syndrome, myasthenia gravis, paralytic shellfish poisoning, mushroom poisoning, tick paralysis, poliomyelitis, and stroke.

### **Laboratory Identification**

The diagnosis of botulism can be confirmed by culturing the organism itself or by identifying its toxin. In early cases, diagnosis is more likely made by toxin assay, whereas persons in the later stages of disease are more likely to be culture positive.

### **Culture**

Appropriate specimens are:

- Foodborne botulism – gastric aspirate or stool.
- Wound botulism – clinical specimen from the wound.
- Infant botulism – stool, rarely isolated from sputum.

### **Toxin Neutralization**

Botulinum toxin in the patient's serum or stool is demonstrated by a toxin neutralization bioassay in mice. This is performed by injecting serum or buffered supernatant from stool into mice and looking for signs of botulism.

- Foodborne botulism – serum, stool, gastric aspirate, or incriminated food.
- Wound botulism – serum.
- Infant botulism – stool, rarely isolated from sputum.

**UPHL:** Utah Public Health Laboratory is the only laboratory in Utah that offers botulism testing.

### **Testing Protocol**

Botulism testing is time, labor, and resource intensive. Unlike other laboratory tests, the test for botulism is not generally used as a rule-out test. While botulism testing is highly specific, sensitivity is quite low. This means that a positive test can be interpreted as positive in almost all cases, but a negative test is not conclusive. Test sensitivity is decreased when specimen collection is delayed. USL: PH depends on the Utah Department of Health Bureau of Epidemiology (BOE) to screen botulism test requests for adults for clinical compatibility with botulism. The BOE approves tests requests after consultation with the physician and USL: PH. An exception to this is infant botulism: If testing is requested by a physician for a child <1 year of age, USL: PH performs the test and informs the BOE that testing is being conducted.

## Treatment

### Foodborne and Wound Botulism

If diagnosed early, foodborne and wound botulism can be treated with equine-derived botulinum antitoxin that blocks the action of toxin circulating in the blood. This can prevent patients from worsening, but does not reverse the damage and recovery still takes many weeks. The Enteric Disease Epidemiology Branch at the Centers for Disease Control and Prevention (CDC) consults with state health departments on antitoxin release and can be reached through the Emergency Operations Center at 770-488-7100. The decision to administer antitoxin should not wait for laboratory confirmation.

Physicians may try to remove contaminated food still in the gut by inducing vomiting or by using enemas. Wounds should be treated, usually surgically, to remove the source of the toxin-producing bacteria. Good supportive care in a hospital is the mainstay of therapy for all forms of botulism.

### Infant Botulism

Human-derived botulinum antitoxin (BabyBIG) is available through the California Department of Health Services for treating infant botulism.

## Case fatality

The case fatality rate is estimated at 10-46%, depending on when and if antitoxin is administered. In general, the case fatality rate for foodborne botulism is 5–10%. Among hospitalized cases in the U.S., the case fatality rate for infant botulism is <1%.

## Reservoir

*Clostridium botulinum* spores are ubiquitous in the environment. The spores can survive indefinitely in soil under almost any environmental condition. Spores are also found in marine sediment.

## Transmission

Generally, botulism is not communicable from person-to-person. However, minute quantities of the toxin acquired by ingestion, inhalation, or by absorption through the eye or a break in the skin can cause botulism. All materials suspected of containing botulinum toxin must be handled with caution.

### Foodborne botulism

Foodborne botulism is acquired by ingesting toxin produced when *C. botulinum* spores germinate in inadequately processed and prepared food. The most frequent source of foodborne botulism is home canned foods. The toxin is destroyed by boiling.

### Wound botulism

Wound botulism is most frequently reported among chronic injection drug users. Wound botulism also occurs when dirt or gravel containing *C. botulinum* spores germinate and produce toxin within wounds.

### **Infant botulism**

Infant botulism occurs when *C. botulinum* spores germinate and produce toxin in the anaerobic conditions of gastrointestinal tract of infants. This can happen through ingestion of food, soil, or dust contaminated with botulinum spores. This kind of infection is rare in adults because the natural bacterial flora in adult gastrointestinal tracts inhibit the germination of *C. botulinum* spores and thus the production of botulinum toxin. However, *C. botulinum* spores can germinate and produce toxin in intestines of adults whose natural flora have been disrupted by disease or antibiotic use. Honey often contains *C. botulinum* spores. Some cases of intestinal botulism have occurred in infants living in areas of construction and earth disruption.

### **Susceptibility**

Anyone can get botulism. Botulism disease does not result in immunity.

### **Incubation period**

The incubation period is variable depending on the form of botulism. Generally, the shorter the incubation period, the more severe the disease.

**Foodborne botulism** 12–36 hours (ranging from 6 hours to 8 days) after eating contaminated food.

**Wound botulism** usually 7 days, with a range of 4–14 days.

**Infant botulism** anywhere from 3-30 days.

### **Period of communicability**

No instances of person-to-person spread have ever been documented for botulism.

### **Epidemiology**

Botulism occurs worldwide as sporadic cases and as family and general outbreaks. Since 1994, the use of black tar heroin by injection drug users has been associated with an increase in the number of cases of wound botulism. Two cases of foodborne botulism were identified in Utah in October of 2003. Prior to that, the last time foodborne botulism was reported in Utah was in 1993. No cases of wound botulism have been identified in Utah. Infant botulism is slightly more common, with approximately 5 cases reported each year.

## ✓ PUBLIC HEALTH CONTROL MEASURES

### Public health responsibility

#### Prevention

##### Personal Preventive Measures/Education

To avoid future exposures, persons should:

- Be educated about the proper time, pressure, and temperature required to destroy spores, if they are interested in home-canning and other preservation techniques.
- Consider boiling canned food for 10 minutes before eating it to ensure safety.
- Not open bulging containers and not eat or even “taste-test” foods with off odors.
- Not feed honey to children <1 year old.
- Promptly seek medical care for infected wounds.
- Not use injectable street drugs.

Note: Instructions on safe home canning can be obtained from county extension services or from the U.S. Department of Agriculture.

#### Chemoprophylaxis

Persons known to have eaten the same contaminated food as the case should be purged with cathartics, given gastric lavage and high enemas and kept under close medical observation. Administration of antitoxin prophylactically is generally not recommended. A decision to release botulinum antitoxin for persons with a plausible epidemiologic link to a botulism case will be made in consultation with CDC.

#### Vaccine

None.

#### Isolation and quarantine requirements

**Isolation:** NA

**Hospital:** NA

**Quarantine:** NA

## ✓ CASE INVESTIGATION

### Reporting

All case of foodborne, intentional or suspected intentional cases of unknown etiology, or clusters of infant botulism are immediately notifiable to public health (within 4 hours). Sporadic cases of infant or wound botulism should be reported by standard notification.

**Table of criteria to determine whether a case should be reported to public health authorities**

| Criterion   | Case Definition |    |          |
|---|-----------------|----|----------|
|   | Confirmed       |    | Probable |
| <i>Historical Evidence</i>  |                 |    |          |
| History of a fresh, contaminated wound during the two weeks before onset of symptoms                  | N3,A4           |    |          |
| Ingestion of home-canned food within the 48 hours before onset of symptoms                            | A3,A4           |    | N1       |
| History of injection drug use within the two weeks before onset of symptoms                           | O3              |    | O3       |
| <i>Clinical Evidence</i>  |                 |    |          |
| Diplopia (double vision)  | O1,O3,O4        | O1 | O1,O3,O4 |
| Blurred vision  | O1,O3,O4        | O1 | O1,O3,O4 |
| Bulbar weakness   | O1,O3,O4        | O1 | O1,O3,O4 |
| Impaired respiration  | O2              |    | O2       |
| Progressive weakness  | O2              |    | O2       |
| Progressive symmetric paralysis   | O1,O3,O4        | O1 | O1,O3,O4 |
| Healthcare record contains a diagnosis of botulism  |                 |    |          |
| Death certificate lists botulism as a cause of death or a significant condition contributing to death |                 |    |          |
| <i>Laboratory Evidence</i>  |                 |    |          |
| Detection of botulinum toxin in serum   | O               | O  |          |
| Detection of botulinum toxin in stool   | O1,O2,O4        | O1 |          |
| Detection of botulinum toxin in patient's food  | O1,O4           | O1 |          |
| Isolation of <i>Clostridium botulinum</i> from stool  | O1,O2,O4        | O1 |          |
| Isolation of <i>Clostridium botulinum</i> from wound  | O3,O4           |    |          |
| <i>Epidemiologic Evidence</i>   |                 |    |          |
| Ingestion of the same food as persons who have laboratory- confirmed botulism                         | A3,A4           | O1 | A?       |
| <i>Special criteria</i>   |                 |    |          |
| Age < 1 year  | N2,A4           |    |          |

S = This criterion alone is Sufficient to identify a case for reporting.

N = All "N" criteria in the same column are Necessary to identify a case for reporting.

O = At least one of these "O" (Optional) criteria in each category (i.e., clinical evidence and laboratory evidence) in the same column—in conjunction with all "N" criteria in the same column—is required to identify a case for reporting. (These optional criteria are

alternatives, which means that a single column will have either no O criteria or multiple O criteria; no column should have only one O.)

\* A requisition or order for any of the “S” laboratory tests is sufficient to meet the reporting criteria.

## **Case definition**

### **Botulism, Foodborne (2010)**

#### **Clinical description**

Ingestion of botulinum toxin results in an illness of variable severity. Common symptoms are diplopia, blurred vision, and bulbar weakness. Symmetric paralysis may progress rapidly.

#### **Laboratory criteria**

- Detection of botulinum toxin in serum, stool, or patient's food, or
- Isolation of *Clostridium botulinum* from stool

#### **Case classification**

*Probable:* A clinically compatible case with an epidemiologic link (e.g., ingestion of a home-canned food within the previous 48 hours)

*Confirmed:* A clinically compatible case that is laboratory confirmed or that occurs among persons who ate the same food as persons who have laboratory-confirmed botulism

### **Botulism, Infant (2010)**

#### **Clinical description**

An illness of infants, characterized by constipation, poor feeding, and “failure to thrive” that may be followed by progressive weakness, impaired respiration, and death

#### **Laboratory criteria**

- Detection of botulinum toxin in stool or serum, or
- Isolation of *Clostridium botulinum* from stool.

#### **Case classification**

*Confirmed:* A clinically compatible case that is laboratory-confirmed, occurring in a child aged less than 1 year

## Botulism, Wound (2011)

### Clinical description

An illness resulting from toxin produced by *Clostridium botulinum* that has infected a wound. Common symptoms are diplopia, blurred vision, and bulbar weakness. Symmetric paralysis may progress rapidly.

### Laboratory criteria for diagnosis

- Detection of botulinum toxin in serum or
- Isolation of *Clostridium botulinum* from wound

### Case classification

*Confirmed:* A clinically compatible case that is laboratory confirmed in a patient who has no suspected exposure to contaminated food and who has a history of a fresh, contaminated wound during the 2 weeks before onset of symptoms, or a history of injection drug use within the 2 weeks before onset of symptoms

*Probable:* A clinically compatible case in a patient who has no suspected exposure to contaminated food and who has a history of a fresh, contaminated wound during the 2 weeks before onset of symptoms, or a history of injection drug use within the 2 weeks before onset of symptoms.

## Botulism, Other (2010)

### Clinical description

Ingestion of botulinum toxin results in an illness of variable severity. Common symptoms are diplopia, blurred vision, and bulbar weakness. Symmetric paralysis may progress rapidly.

### Laboratory criteria

- Detection of botulinum toxin in a clinical specimen or
- Isolation of *Clostridium botulinum* from a clinical specimen

### Case classification

*Confirmed:* A clinically compatible case that is laboratory confirmed in a patient aged greater than or equal to 1 year who has no history of ingestion of suspect food and has no wounds.

### CSTE Case Classification

| Criterion   | Case Definition |    |          |
|---|-----------------|----|----------|
|   | Confirmed       |    | Probable |
| <i>Historical Evidence</i>  |                 |    |          |
| History of a fresh, contaminated wound during the two weeks before onset of symptoms                  | N3,A4           |    |          |
| Ingestion of home-canned food within the 48 hours before onset of symptoms                            | A3,A4           |    | N1       |
| History of injection drug use within the two weeks before onset of symptoms                           | O3              |    | O3       |
| <i>Clinical Evidence</i>  |                 |    |          |
| Diplopia (double vision)  | O1,O3,O4        | O1 | O1,O3,O4 |
| Blurred vision  | O1,O3,O4        | O1 | O1,O3,O4 |
| Bulbar weakness   | O1,O3,O4        | O1 | O1,O3,O4 |
| Impaired respiration  | O2              |    | O2       |
| Progressive weakness  | O2              |    | O2       |
| Progressive symmetric paralysis   | O1,O3,O4        | O1 | O1,O3,O4 |
| Healthcare record contains a diagnosis of botulism  |                 |    |          |
| Death certificate lists botulism as a cause of death or a significant condition contributing to death |                 |    |          |
| <i>Laboratory Evidence</i>  |                 |    |          |
| Detection of botulinum toxin in serum   | O               | O  |          |
| Detection of botulinum toxin in stool   | O1,O2,O4        | O1 |          |
| Detection of botulinum toxin in patient's food  | O1,O4           | O1 |          |
| Isolation of <i>Clostridium botulinum</i> from stool  | O1,O2,O4        | O1 |          |
| Isolation of <i>Clostridium botulinum</i> from wound  | O3,O4           |    |          |
| <i>Epidemiologic Evidence</i>   |                 |    |          |
| Ingestion of the same food as persons who have laboratory- confirmed botulism                         | A3,A4           | O1 | A?       |
| <i>Special criteria</i>   |                 |    |          |
| Age < 1 year  | N2,A4           |    |          |

N = All “N” criteria in the same column are Necessary to classify a case.

A = This criterion must be absent (i.e., NOT present) for the case to meet the classification criteria.

O = At least one of these “O” (Optional) criteria in each category (i.e., clinical evidence and laboratory evidence) in the same column—in conjunction with all “N” criteria in the same column—is required to classify a case.

- 1 = Foodborne botulism
- 2 = Infant botulism
- 3 = Wound botulism
- 4 = Other botulism

## Case Investigation Process

### Foodborne Botulism

Even one case of foodborne botulism constitutes a public health emergency. Public health has five main roles when a case of foodborne botulism is identified or suspected:

#### *Source investigation*

Public health should aggressively investigate for the source of intoxication and immediately remove it from general public consumption, once identified. The investigation should not wait for positive test results. The general recommendations for identifying suspect foods follow:

- Identify all home-canned foods eaten during the week prior to symptoms.
- The most suspect foods are those eaten less than two days before onset and those that were not eaten by other well persons. Keep in mind that additional cases may experience less severe symptoms with a later onset than the first case identified.
- Identify all commercially canned foods eaten during the week prior to the onset of illness. For implicated foods, determine the brand, manufacturer, package size, lot number, and place and date of purchase.
- Identify all sausage and other preserved meats eaten during the week prior to onset of illness.
- Meat products that have not been adequately refrigerated are also suspect.
- Identify all smoked or otherwise preserved fish eaten during the week before onset of symptoms.

#### *Contact investigation*

All case contacts that may have eaten contaminated food should be identified and appropriately managed (explained in detail below).

#### *Laboratory testing*

USL: PH is the only laboratory in Utah that provides testing of human and non-human samples for botulinum toxin and *C. botulinum*. USL: PH also provides this service to other states in its area (Wyoming, Colorado, Arizona, and Montana). Because of the resources required, USL: PH depends on clearance from the BOE (or an epidemiologist from the state requesting the test) before the test is performed. The BOE coordinates with the patient's physician and the local health department to determine cases for which testing will be provided.

#### *Treatment*

The Centers for Disease Control and Prevention has the responsibility to consult with the patient's physician and BOE to ensure that equine-derived botulism antitoxin is released when appropriate. The BOE ensures that this consultation has been initiated when appropriate. Botulism antitoxin is released by the CDC only after consultation with the patient's physician and the BOE. The Enteric Disease Epidemiology Branch at CDC can be reached through CDC's Emergency Operations Center at **770-488-7100**.

*Education*

The public health investigator should educate the patients and their family on the prevention of botulism intoxication.

**Wound Botulism**

Public health has three main roles when a case of wound botulism is identified or suspected:

*Laboratory testing*

USL: PH is the only laboratory in Utah that provides testing of human and non-human samples for botulinum toxin and *C. botulinum*. USL: PH also provides this service to other states in its area (Wyoming, Colorado, Arizona, and Montana). Because of the resources required, USL: PH depends on clearance from the BOE (or an epidemiologist from the state requesting the test) before the test is performed. The BOE coordinates with the patient's physician and the local health department to determine cases for which testing will be provided.

*Treatment*

The Centers for Disease Control and Prevention has the responsibility to consult with the patient's physician and BOE to ensure that equine-derived botulism antitoxin is released when appropriate. The BOE ensures that this consultation has been initiated when appropriate. Botulism antitoxin is released by the CDC only after consultation with the patient's physician and the BOE.

*Education*

The public health investigator should educate the patients and their family on the prevention of botulism intoxication.

**Infant Botulism**

One case of infant botulism is not necessarily a public health emergency. In most cases, investigation of the source of infection is not necessary. This is because *C. botulinum* spores are ubiquitous in the soil and dust. Honey has also been identified as a vehicle of *C. botulinum* spores. However, investigators should keep in mind less common or less frequently identified sources of infant botulism. Investigators should also be vigilant of foodborne botulism cases in an infant that may present as infant botulism. Public health has three main roles when a case of infant botulism is identified or suspected:

### *Laboratory testing*

USL: PH is the only laboratory in Utah that provides testing of human and non-human samples for botulinum toxin and *C. botulinum*. USL: PH also provides this service to other states in its area (Wyoming, Colorado, Arizona, and Montana). USL: PH automatically performs botulism testing that is requested on a child <1 year of age and notifies the BOE that the testing is underway.

### *Treatment*

The California Department of Health Services has the responsibility to consult with the patient's physician and the BOE to ensure that human-derived botulism antitoxin (BabyBIG) is released when appropriate. The BOE ensures that this consultation has been initiated when appropriate.

### *Education*

The public health investigator should educate the patient's family on the prevention of botulism intoxication.

## **Outbreaks**

Botulism usually occurs as sporadic cases; however, outbreaks of foodborne botulism can occur if multiple persons consume contaminated food.

## **Identification of case contacts and management**

### **Foodborne Botulism**

Persons who consumed food items suspected to be the source of foodborne botulism should be immediately contacted, advised to seek health care, and questioned about symptoms. Depending on the time of ingestion, other exposed persons might be candidates for treatment with purgatives, and at the very least, should be under close medical supervision.

### **Wound Botulism**

When additional cases are suspected to be related to a case of wound botulism (e.g. injection drug user), contacts may be identified and investigated.

### **Infant Botulism**

Adult contacts of a case of infant botulism are not at risk of developing disease.

## ✓ REFERENCES

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