

Botulism rev Jan 2016

***Initial calls or reports regarding suspect botulism should be referred to EAIDB for immediate evaluation and for approval of testing and/or release of antitoxin, if appropriate. If botulism is highly suspected (e.g., antitoxin is released or testing is approved) then the case will be forwarded to the appropriate regional or local health department for further investigation. ***

Please contact DSHS EAIDB at 512-776-7676 or 512-221-6852 (after hours).

BASIC EPIDEMIOLOGY

Infectious Agent

Botulism is caused by neurotoxins produced by the bacterium *Clostridium botulinum*. *C. botulinum* bacteria form spores which can survive under a wide range of adverse environmental conditions, including high temperatures, such as boiling for less than ten minutes. Bacterial growth, however, (as opposed to spore survival) occurs only under anaerobic conditions and low acid (generally pH>4) and the toxin itself is produced as the bacteria are multiplying. There are seven types of botulinum toxin, designated A– G. Types A, B, and E are the most common sources of human disease. The toxin is heat-labile, and can be inactivated by boiling for ten minutes.

Transmission

Foodborne botulism: caused by ingestion of pre-formed toxin. Most implicated foods are low acid, home-canned items inadequately processed during canning and not heated before consumption. Rarely, commercial products are implicated, usually after a breakdown in standard canning procedures. Examples of implicated foods include:

- Home-canned asparagus, beans, and other vegetables (including low acid tomatoes and salsa), usually processed inadequately by the water-bath method;
- Fish that has been improperly canned, dried, smoked or stored;
- Sausages or other prepared meats, such as jerky, that are improperly processed (inadequate sodium nitrite) and improperly stored;
- Chopped or whole garlic, herbs, olives or vegetables bottled in oil;
- Among Alaskan Natives, traditionally preserved foods including fermented (putrefied) whale blubber, salmon heads, salmon eggs, and other marine products;
- Possibly home-pickled fish, eggs, vegetables and olives that have been inadequately prepared without the correct concentrations of salt and/or vinegar.
- Rare commercial canned products (e.g., commercially canned chili in 2007); products may be recalled even without cases if improper processing carries a risk of botulism.

Wound botulism: results from a local *C. botulinum* infection in devitalized tissue at a wound site, where semi-anaerobic conditions develop. Wound botulism has been rare, but increasingly reported, especially in injectors of "black-tar" heroin.

Infant botulism: occurs when *C. botulinum* spores are ingested in food or soil and germinate in the preformed gut of infants under the age of 1 that have yet to develop mature intestinal flora. The germination of spores results in an intestinal infection in these infants, where the botulinum toxin is produced within the intestine and then enters the bloodstream causing symptoms. This is also known as intestinal botulism.

Inhalational botulism: does not occur naturally. There have been only three reported cases in humans worldwide. Studies done with monkeys have shown that the toxin can be absorbed through the lung mucus membrane into the bloodstream. It is believed that if botulinum toxin were to be used as a bioweapon, it would be by this route.

Iatrogenic botulism: occurs from an accidental overdose or as an adverse event following the therapeutic or cosmetic injection of botulinum neurotoxin. Examples of therapeutic uses of botulinum toxin include treatment for hemifacial muscle spasms, focal dystonia, focal spasticity, autonomic disorders, Frey's syndrome and oculomotor disorders.

Incubation Period

Foodborne botulism: The incubation period for foodborne botulism can vary from 12 hours to several days, but is usually 12–36 hours. A short incubation is associated with more severe disease and larger toxin dose ingested.

Wound and iatrogenic botulism: The incubation period can be up to two weeks or longer.

Infant botulism: The incubation period is unknown.

Inhalational botulism: Thought to be 12–36 hours after inhalation, but may take several days after exposure to low doses of toxin.

Communicability

No instance of secondary person-to-person transmission has been documented.

Clinical Illness

Early symptoms tend to be nonspecific and providers often do not suspect botulism until the symptoms become more severe. The hallmark symptoms of botulism are bilaterally symmetrical cranial nerve palsies, which result in slurred speech (dysarthria), difficulty swallowing (dysphagia), double vision (diplopia), and/or drooping eyelids (ptosis); the symptoms progress in a descending manner, causing weakness and possibly paralysis, including loss of respiratory function.

Botulism is frequently misdiagnosed in adults, most often as polyradiculoneuropathy (Guillain-Barré or Miller-Fisher syndrome), myasthenia gravis, or other diseases of the central nervous system.

The signs and symptoms of infant botulism are constipation, poor feeding and/or weak sucking, drooping eyelids (ptosis), weak cry, dilated and/or sluggishly reactive pupils, poor head control, hypotonia ("floppy baby syndrome"), and respiratory difficulty.

DEFINITIONS

Note: There are 4 different categories of botulism used for reporting cases in NEDSS: Botulism, foodborne; Botulism, wound; Botulism, infant; and Botulism, other unspecified.

BOTULISM, FOODBORNE

Clinical Case Definition

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

Laboratory Confirmation

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

Case Classifications

- **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.
- **Probable:** A clinically compatible case with a history of ingestion of a food item known to carry a risk for the botulism toxin.

BOTULISM, WOUND

Clinical Case Definition

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 can progress rapidly.

Laboratory Confirmation

- Detection of botulinum toxin in serum, **OR**
- Isolation of *Clostridium botulinum* from wound.

Case Classifications

- **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 either 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, INFANT

Clinical Case Definition

An illness of infants (<1 year), characterized by constipation, poor feeding, and “failure to thrive” that can be followed by progressive weakness, impaired respiration, and death.

Laboratory Confirmation

- 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, OTHER UNSPECIFIED

Clinical Case Definition

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

Laboratory Confirmation

- Detection of botulinum toxin in clinical specimen, **OR**
- Isolation of *Clostridium botulinum* from 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.

SURVEILLANCE AND CASE INVESTIGATION

Note: EAIDB should be directly involved in the evaluation and testing of suspect foodborne botulism cases because antitoxin (H-BAT) can only be released by the CDC after consultation with the central office of a state health department. This is done to ensure that the state health department is aware that there is a suspect case of foodborne botulism, as there is always the possibility of additional cases and a food source still in commerce.

EAIDB does not have to be directly involved in the evaluation of suspect infant botulism cases because these are almost always isolated cases and do not represent public health threats, but should be contacted for approval and coordination of specimens submitted to the DSHS laboratory. Physicians can contact the California Infant Botulism Treatment and Prevention Program directly, and antitoxin will be released without consultation with EAIDB. EAIDB does receive notification of the antitoxin release in these situations and will notify the appropriate regional or local health department for further investigation.

For suspect botulism consultations, please contact DSHS EAIDB at 512-776-7676 or 512-221-6852 (after hours).

*** If botulism is highly suspected (e.g., antitoxin is released or testing is approved) then the case will be forwarded to the appropriate regional or local health department for further investigation.

Case Investigation

Local and regional health departments should investigate all reports of suspect botulism. Investigations should include an interview of the case or a surrogate to get a detailed exposure history. Please use the Botulism Foodborne Alert Summary or the Infant Botulism Investigation Form available on the DSHS website: <http://www.dshs.state.tx.us/idcu/investigation/>.

Case Investigation Checklist

- Confirm laboratory results meet the case definition (may have been done by EAIDB).
- Verify that the laboratory has sent a specimen to the DSHS laboratory (may have been done by EAIDB).
- Review medical records or speak to an infection preventionist or healthcare provider to describe course of illness and outcome of case.
- Interview the case or surrogate to identify potential sources infections:

Foodborne Botulism

- Interview the case and others who may be able to provide pertinent information about foods eaten.
- A home visit is strongly recommended when home-canned foods are implicated, or if the source is not readily apparent.
- Ask about possible exposures 1–10 days before onset of symptoms, including:
 - Home-canned, vacuum-packed, or traditionally preserved foods. Foods to suspect as a source of illness are those eaten less than two days before onset, those that are low in acid (fish, meat, and vegetables), and those that were not eaten by other persons who remain well.
 - Commercially canned, vacuum packed foods or mishandled commercial products (e.g., refrigerated soup not kept cold after purchase); such products are implicated only rarely. For implicated foods, determine the brand, manufacturer, package size, lot number and location/date of purchase.

- Preserved or traditionally prepared fish and marine products.
- Items stored in oil (e.g., onions, garlic) or foil (e.g., baked potatoes.).
- Sausages, preserved or traditionally preserved meats, and inadequately refrigerated meats; such products are implicated only rarely.
- Ask if any leftovers of any reported risky food items are still present in the home. Consult with EAIDB regarding possible testing of identified risky food items.
- Identify other potentially exposed persons. Obtain the name, address, and telephone number of every person who may have eaten the suspected food item, shared an environmental exposure or may have the suspect home-processed food in his or her possession.
- Obtain the organization name, contact telephone number, and attendance lists (particularly email or telephone lists) for every suspected gathering, public event, or other shared environmental exposure.
- Use the Botulism Foodborne Alert Summary to record information from the interview.

Wound Botulism

- Interview the case or surrogate to identify potential sources of infections:
 - Ask the patient about illicit injection drug use. Specifically, ask about the types of drugs used and how the drugs are used (e.g., injected into veins, injected into tissues, snorted, etc.).
 - In addition to illicit drug use, interview regarding potential foodborne exposures.

Infant Botulism

- Interview the case or surrogate to identify potential sources of infections:
 - The most common risk factors are exposure to dirt or dust, as with nearby construction, or the ingestion of unpasteurized honey. Although honey was associated with intestinal botulism in the past, it is rarely implicated in cases.
 - Use the Infant Botulism Investigation Form to record information from the interview.
- Fax completed forms to DSHS EAIDB at 512-776-7616 or email securely to an EAIDB epidemiologist.
- Hospitalized cases should be followed until discharge and patient's outcome recorded.
 - Initial reports can be sent to DSHS prior to discharge.
- In the event of a death, copies of the hospital discharge or death summary should also be faxed to DSHS EAIDB.
- All confirmed and probable case investigations must be entered and submitted for notification in the NEDSS Base System (NBS). Please refer to the *NBS Data Entry Guidelines* for disease specific entry rules.

Treatment

Treatment for botulism is based on the clinical picture and should never be delayed pending laboratory confirmation of the diagnosis. All patients require close monitoring of ventilatory status, and severe cases need aggressive supportive therapy. Additional therapies depend on the type of botulism and are outlined below:

- **Foodborne, Wound, Other (non-infant):** The Centers for Disease Control and Prevention (CDC) control the distribution of botulinum antitoxin (H-BAT), which is stocked at United States Public Health Service Quarantine Stations throughout the country. If antitoxin treatment is being considered, EAIDB will immediately consult with the CDC.

- **Infant Botulism:** A human-derived hyper immune globulin (BIG-IV or “Baby BIG”) is approved by FDA for treatment of infants. Baby BIG can be obtained from the California Department of Health Services by calling their 24 hour number at 510- 231-7600. Consultations also available. EAIDB should also be contacted to arrange for testing by the DSHS laboratory. Additional information about infant botulism is available at: <http://www.infantbotulism.org/>

Prevention and Control Measures

Foodborne botulism

- Strict hygienic procedures should be followed when home canning or pickling to properly sterilize products and prevent bacterial growth, thus reducing the contamination of foods.
- Oils infused with garlic, vegetables, fresh herbs or similar moist flavoring should be refrigerated.
- Potatoes which have been baked while wrapped in aluminum foil should be kept hot or thoroughly reheated before being served, or refrigerated immediately.
- Because the botulism toxin is destroyed by high temperatures, persons who eat risky home-canned foods (i.e., low acidic, non-pickled foods) should consider boiling the food for a minimum of ten minutes before eating it to ensure safety. However, if a food is suspected or at risk of containing botulinum toxin, it should be discarded immediately, as uniform heating may not occur throughout the product or be of a sufficient temperature and/or length of time to destroy the toxin. Adequate pickling, the addition of sugar syrup, or sufficient brining should prevent the growth of *C. botulinum*.
- Instructions on safe home canning can be obtained from county extension services or from the United States Department of Agriculture.
 - http://nchfp.uga.edu/publications/publications_usda.html

Wound botulism

- Wound botulism can be prevented by promptly seeking medical care for infected wounds and by not using injectable street drugs.
- Injection drug users and healthcare providers serving them should be educated regarding typical symptoms of botulism and the importance of rapid diagnosis and treatment. Potential routes for education include needle exchange programs, urban hospital emergency departments, or free clinics.

Infant botulism

- Honey can contain spores of *Clostridium botulinum* and may be a source of infection for infants, therefore children less than 12 months old should not be fed honey (raw or otherwise). Honey is safe for persons one year of age and older.

Iatrogenic botulism

- Iatrogenic botulism may be prevented by using commercially manufactured therapeutic botulinum toxin from medically approved sources and by avoiding injections above recommended doses.

Exclusions

No exclusion is required.

MANAGING SPECIAL SITUATIONS

Outbreaks

Botulism outbreaks are rare. Outbreaks of foodborne botulism have potential to be a public health emergency because the contaminated food may be eaten by other people. Rapid investigation of cases and outbreaks is critical for prompt treatment of likely cases, and for the identification of contaminated food vehicles and prevention of additional cases.

If an outbreak is suspected, **immediately** notify DSHS EAIDB at **(800) 252-8239** or **(512) 776-7676** or **512-221-6852 (after hours)**.

- Outbreak investigations should always be done in a collaborative manner involving local health department(s) with suspected or confirmed cases, the appropriate regional health department(s), an EAIDB botulism epidemiologist, DSHS Regulatory Services staff, and any appropriate federal agencies.
- If a food establishment or a commercial product is implicated, EAIDB will notify the DSHS Division of Regulatory Services about the outbreak and the possibility of a common contaminated food source for the case(s).
- Outbreaks of infant botulism are extremely unlikely, but not impossible. A food or formula product containing a high load of spores may be responsible for an outbreak. However, since infant botulism was first recognized in 1976, there has never been an infant botulism outbreak in Texas, and, to our knowledge, there has never been one in the U.S.

The local/regional health department should:

- Review case information collected following the initial notification of any suspect individual case(s) already identified, including laboratory results, Foodborne Botulism Alert Summary forms, clinical histories, food histories, and any other information.
- Contact hospitals and healthcare providers in the appropriate areas of the state, or throughout the state if necessary, to alert them to the possibility of additional cases of foodborne botulism.
- Interview all cases suspected as being part of the outbreak or cluster if not done already.
- Prepare a line list of cases in your jurisdiction. At a minimum, information needed for the line list includes patient name, DSHS specimen identification number, specimen source, date of specimen collection, date of birth, county of residence, date of onset (if known), symptoms, underlying conditions, treatments and outcome of botulism, and risky foods eaten or other risky exposures reported by the case or surrogate.
- Encourage anyone with symptoms be evaluated by a healthcare provider.
- Communicate regularly with all parties involved in outbreak investigation
 - Provide Situation Reports through email.
 - Hold conference calls to discuss the outbreak investigation
- Report findings at conclusion of investigation:
 - Create Outbreak Summary Report.
 - Enter outbreak into NORS at the conclusion of the outbreak investigation. See Reporting and Data Entry Requirements section.

Botulinum Toxin as a Biological Weapon

C. botulinum toxin has been classified as a possible agent of bioterrorism due to its ability to be weaponized and because it is extremely potent and lethal. The toxin is also easy to produce and transport, and affected individuals often need extensive and prolonged intensive care. Dissemination through aerosol or food would be the most likely mode of spread. Aerosol dissemination could result in many cases of illness in a geographic area. Therefore, inhalational botulism produced by an act of bioterrorism should be considered for 2 or more botulism cases linked temporally and geographically but without a likely common foodborne or drug exposure. In such situations immediately call DSHS EAIDB at 512-776-7676 or 512-221-6852 (after hours). The cases should be extensively interviewed to identify possible exposures such as gatherings, public events, specific geographic locations, large buildings, shopping areas, and public transportation.

REPORTING AND DATA ENTRY REQUIREMENTS

Provider, School, Child-Care Facility, and General Public Reporting Requirements

Confirmed, probable and clinically suspected cases are required to be reported **immediately** to the Texas Department of State Health Services (DSHS), Emerging and Acute Infectious Disease Branch (EAIDB) at **(800) 252-8239** or **(512) 776-7676**.

Local and Regional Reporting and Follow-up Responsibilities

Local and regional health departments should:

- Call DSHS EAIDB immediately when a botulism investigation is being conducted.
- Enter the case into NBS and submit an NBS notification on all **confirmed** and **probable** cases.
 - Please refer to the *NBS Data Entry Guidelines* for disease-specific entry rules.
 - A notification can be sent as soon as the case criteria have been met. Additional information from the investigation may be entered upon completing the investigation.
- Fax completed forms to DSHS EAIDB at **512-776-7616** or email securely to an EAIDB foodborne epidemiologist.

When an outbreak is investigated, local and regional health departments should:

- Report outbreaks **immediately** to the regional DSHS office or to EAIDB at **512-776-7676**.
- Enter outbreak information into the **National Outbreak Reporting System (NORS)** at the conclusion of the outbreak investigation.
 - For NORS reporting, the definition of an outbreak is two or more cases of similar illness associated with a common exposure.
 - The following should be reported to NORS:
 - Foodborne disease, waterborne disease, and enteric illness outbreaks with person-to-person, animal contact, environmental contact, or an indeterminate route of transmission.
 - Outbreaks as indicated above with patients in the same household.
 - Enter outbreaks into NORS online reporting system at <https://wwwn.cdc.gov/nors/login.aspx>
 - Forms, training materials, and other resources are available at <http://www.cdc.gov/nors/>
- To request a NORS account, please email FoodborneTexas@dshs.state.tx.us
 - Please put in Subject Line: NORS User Account Request
 - Information needed from requestor: name, email address, and agency name
 - After an account has been created a reply email will be sent with a username, password, and instructions for logging in.

LABORATORY PROCEDURES

The DSHS laboratory in Austin is the only lab in Texas that can perform confirmatory testing for botulism. Specimens will be accepted by the DSHS lab only with prior approval by EAIDB. Please contact DSHS EAIDB at 512-776-7676 or 512-221-6852 (after hours).

A preliminary laboratory result may be available in 3-5 days after the specimen arrives and a definitive result may take as long as 3 weeks. The decision to treat is based on the clinical picture and should not wait for laboratory confirmation. Generally, if the physician is not considering treatment with antitoxin, there is no need for laboratory testing.

The laboratory must be notified at 512-689-5537 prior to shipping any specimens.

Specimen Collection

- Stool
 - 10-50 grams recommended for an adult
 - ≥ 5 grams recommended for an infant
 - Keep at 2° - 8° C. Do not freeze
 - A sterile water enema can be used to obtain a specimen from a non-stooling patient
- Vomitus or Gastric Aspirate
 - > 10 mL in sterile, leak-proof container
- Serum
 - 10ml minimum for an adult is recommended
 - Not recommended for infant testing
- Wound
 - Tissue from a biopsy or swab from deep in the wound
- Food
 - Only tested if associated with a confirmed botulism case

Submission Form

- DSHS Laboratory G-27A form for specimen submission.
- Make sure the patient's name, date of birth and/or other identifier match exactly what is written on the transport tubes and on the G-27A form.
- Fill in the date of collection and select the appropriate test.
- Payor source:
 - Check “IDEAS” to avoid bill for submitter

Specimen Shipping

- Transport temperature:
 - Stools, Vomitus or Gastric Aspirate, and Serum:
 - Keep at 2° - 8° C
 - Should be shipped cold (on cold packs, not dry ice) by overnight courier
 - Wound:
 - Ship tissue in anaerobic atmosphere
 - Ship swab in anaerobic transport for swabs
 - Ship without refrigeration
 - Food:
 - Should be shipped in original container under current storage conditions (e.g., cold storage submitted cold; frozen storage submitted frozen; etc.)
- All specimens must be triple contained in accordance with federal shipping regulations. All clinical specimens must be accompanied by a specimen submission form (G-27A).
- Ship specimens via overnight delivery.
- DO NOT mail on a Friday unless special arrangements have been pre-arranged with DSHS Laboratory.
- Ship specimens to:

Laboratory Services Section, MC-1947
Texas Department of State Health Services
Attn. BioThreat Team (512) 689-5537
1100 West 49th Street
Austin, TX 78756-3199

Causes for Rejection:

- Incorrect source of specimen.
- Insufficient amount of specimen.
- Missing or discrepant information on form/specimen

UPDATES

January 2016

- Expanded the NORS sub-section in the Reporting and Data Entry Requirements section to include the NORS outbreak definition and the types of outbreaks that should be reported in NORS.