Infant Botulism

Disease Plan

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Last updated: February 18, 2016, by Cindy Burnett

Questions about this disease plan?

Contact the Utah Department of Health Bureau of Epidemiology: 801-538-6191.
Infant botulism results when an infant (usually between the ages of 6 weeks to 6 months) ingests botulism spores which colonize the gastrointestinal tract and produce botulism toxin. Although the illness is rare (on average only 2–4 cases are reported each year in Utah), it is potentially life-threatening and often results in prolonged hospitalization and lengthy rehabilitation. Timely treatment with Botulism Immune Globulin (Baby BIG®) is critical in reducing duration of hospitalization and improving the outcomes of affected infants. In addition, investigation of cases can identify possible clusters and novel exposures (such as contaminated baby food). Public health investigators also play an important role in educating parents of affected infants about precautions that should be taken when the infant comes home from the hospital to prevent other household members and/or pets from being exposed to botulism toxin.

DISEASE AND EPIDEMIOLOGY

Clinical Description
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, drooling, lethargy, listlessness, ptosis (drooping eyelids), difficulty swallowing, a weak cry, and lack of muscle tone (“floppy baby syndrome”). Paralysis is generally symmetric and descending. 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 the 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 infant botulism is primarily caused by types A and B. Most cases of infant botulism in Utah are type A, which is more prevalent in the Western United States. Other novel *Clostridia* strains, including *C. baratti* and *C. butyricum*, can also produce a botulinum-like toxin and cause infant botulism.

Differential Diagnosis
There are many conditions and diseases that have presentations similar to infant botulism. Infants with botulism are often diagnosed with dehydration, sepsis, or meningoencephalitis after presentation of lethargy and irritability. Other conditions which should be considered in the differential diagnosis are drug or chemical poisoning, metabolic disorders, spinal muscular atrophy (SMA), Reye’s syndrome, congenital myasthenia gravis, poliomyelitis, stroke, and
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Guillain-Barré Syndrome (although this is not usually seen in children younger than one year of age).

**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 include stool or enema.

**Toxin Neutralization**
Appropriate specimens include stool or enema. It may be possible to identify toxin in serum, however, stool is the preferred specimen.

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.

**Note regarding proper specimen collection:** Fecal specimens should be collected in a sterile container with a tight, screw-capped lid, such as a urine specimen container. Do not use containers with fixatives, such as those used for ova and parasite collection. If no stool can be collected spontaneously or through gentle digital stimulation, an enema should be collected. However, glycerin suppositories yield an unsatisfactory specimen and should not be used.

**Utah Public Health Laboratory (UPHL):** UPHL is the only laboratory in Utah that offers botulism testing. Both culture and toxin neutralization tests are available.

**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. The lead enteric epidemiologist or medical officer at the Utah Department of Health (UDOH) Bureau of Epidemiology (BOE) screens all botulism test requests. Test requests are approved after consultation with the requesting physician and UPHL.

**Treatment**
Most infant botulism patients require hospitalization. Botulism Immune Globulin (BabyBIG®) is effective in reducing the length of hospital stay and cost for infant botulism patients. It is available through the California Department of Health, Infant Botulism Treatment and Prevention Program (IBTPP). Prompt treatment with BabyBIG® immediately ends toxemia and enables nerve regeneration to begin. The treatment is most effective when provided as soon as possible after symptom onset. Treatment should not be delayed until laboratory results are available. Physicians requesting BabyBIG® should contact the IBTPP on-call physician directly.
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at (510) 231-7600 (available 24 hours/day). More information is available on the IBTPP website at http://www.infantbotulism.org/. Requesting BabyBIG® does not replace reporting requirements to public health.

Case Fatality
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 and can travel through dust. Although botulism spores can be found in a variety of foods, honey is the only food which has been definitively linked to development of infant botulism.

Transmission
Infant botulism occurs when C. botulinum spores germinate and produce toxin in the anaerobic conditions of the 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. The stools of affected infants contain toxin, and may pose a risk to household members and pets.

Susceptibility
All infants are potentially susceptible to infant botulism. Both formula-fed and breast-fed infants are at risk, however, onset of illness in formula-fed infants occurs at a significantly younger age than breast-fed infants (7.6 weeks vs. 13.7 weeks). Introduction of solid foods may aid colonization with C. botulinum by causing changes in the gut microflora. In addition, infants with slow gut motility (defined as having less than one stool per day) may be at increased risk for developing infant botulism.

Infants treated with BabyBIG® will have a protective level of toxin-neutralizing antibody for at least six months following administration of the medicine, and during this time an infant cannot get botulism again. There have been no reported cases of an infant acquiring botulism more than once. However, in some cases, infants who are prematurely discharged from hospital care may have a relapse of breathing and feeding difficulties.

Incubation Period
The incubation period for infant botulism is 2-4 weeks.

Period of Communicability
While botulism is not spread from person-to-person, patients with infant botulism excrete both C. botulinum toxin and organism in their feces from weeks to months after onset. Scrupulous handwashing should be practiced after diaper changes, and persons with cuts or open wounds
on their hands should wear gloves when changing diapers. Soiled diapers should be disposed of where no person or animal can come into contact with them.

**Epidemiology**

Infant botulism, the most common form of botulism, has only been recognized since 1976. It is a rare disease with an average of 2-4 cases reported each year in Utah. However, Utah has the third highest incidence of infant botulism in the U.S., following Delaware and Hawaii. This may be partly due to increased physician awareness and access to diagnostic testing. Interestingly, eight of 11 states with the highest incidence are located west of the Rocky Mountains.

✓ **PUBLIC HEALTH CONTROL MEASURES**

**Public Health Responsibility**

The public health investigator should ensure that the patient has received BabyBIG®. This is typically obtained by the treating physician directly from the IBTPP. If infant botulism is strongly suspected, but the physician is not sure whether treatment is warranted due to delayed diagnosis, consultation with the IBTPP is strongly encouraged. The phone number for the on-call IBTPP physician is: 510-231-7600.

The investigator should obtain appropriate hospital records, including Admission Notes and Discharge Summary. The patient’s parent or guardian should be interviewed using the Infant Botulism case report form. Education should be provided to all caregivers regarding appropriate handing of diapers.

**Prevention**

The only known prevention measure for infant botulism is to avoid feeding honey to infants 12 months of age or less. Breastfeeding may slow the onset and lessen the severity of illness.

**Chemoprophylaxis**

None.

**Vaccine**

None.

**Isolation and Quarantine Requirements**

**Isolation:** Isolation measures are not required, but meticulous handwashing is. Because the patient may excrete the toxin and organism for weeks to months, close contact with other infants and children should be limited during this time to ensure that other children do not come into contact with fecal material from a leaky diaper. Any contact the patient has with other infants and children during this time should be supervised by an adult.
C. botulinum is not part of the patient’s normal flora and will eventually stop being excreted in the infant’s feces.

Hospital: In the hospital, soiled diapers should be autoclaved and should not be handled by staff with open lesions.

Quarantine: None.

✓ CASE INVESTIGATION

Reporting
Infant botulism cases are not immediately notifiable. Cases should be reported within three working days after identification in Utah.

Reporting Table:
Table of criteria to determine whether a case should be reported to public health authorities.

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Reporting</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Historical Evidence</strong></td>
<td></td>
</tr>
<tr>
<td>History of a fresh, contaminated wound during the two weeks before onset of symptoms</td>
<td>O</td>
</tr>
<tr>
<td>Ingestion of home-canned food within the 48 hours before onset of symptoms</td>
<td>O</td>
</tr>
<tr>
<td><strong>Clinical Evidence</strong></td>
<td></td>
</tr>
<tr>
<td>Diplopia (double vision)</td>
<td>O</td>
</tr>
<tr>
<td>Blurred vision</td>
<td>O</td>
</tr>
<tr>
<td>Bulbar weakness</td>
<td>O</td>
</tr>
<tr>
<td>Impaired respiration±</td>
<td>O±</td>
</tr>
<tr>
<td>Progressive weakness±</td>
<td>O±</td>
</tr>
<tr>
<td>Progressive symmetric paralysis</td>
<td>O</td>
</tr>
<tr>
<td>Healthcare record contains a diagnosis of botulism±</td>
<td>S±</td>
</tr>
<tr>
<td>Death certificate lists botulism as a cause of death or a significant condition contributing to death±</td>
<td>S±</td>
</tr>
<tr>
<td><strong>Laboratory Evidence</strong></td>
<td>S±</td>
</tr>
<tr>
<td>Detection of botulinum toxin in serum</td>
<td>S</td>
</tr>
<tr>
<td>Detection of botulinum toxin in stool±</td>
<td>S±</td>
</tr>
<tr>
<td>Detection of botulinum toxin in patient's food</td>
<td>S</td>
</tr>
<tr>
<td>Isolation of Clostridium botulinum from stool±</td>
<td>S±</td>
</tr>
<tr>
<td>Isolation of Clostridium botulinum from wound</td>
<td>S</td>
</tr>
<tr>
<td><strong>Epidemiologic Evidence</strong></td>
<td>N</td>
</tr>
<tr>
<td>Ingestion of the same food as persons who have laboratory-confirmed botulism</td>
<td></td>
</tr>
<tr>
<td><strong>Special criteria</strong></td>
<td></td>
</tr>
</tbody>
</table>
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<table>
<thead>
<tr>
<th>Request for anti-toxin specific for botulinum toxin</th>
<th>S*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Request for BabyBIG from the Infant Botulism Treatment and Prevention Program</td>
<td>S++</td>
</tr>
</tbody>
</table>

S = These criteria alone are 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 alternative, 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**

**Clinical definition**
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 one year.

**CSTE Case Classification Table**

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Case Definition</th>
<th>Confirmed</th>
<th>Probable</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Historical Evidence</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>History of a fresh, contaminated wound during the two weeks before onset of symptoms</td>
<td>N3, A4</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ingestion of home-canned food within the 48 hours before onset of symptoms</td>
<td>A3, A4</td>
<td>N1</td>
<td></td>
</tr>
<tr>
<td>History of injection drug use within the two weeks before onset of symptoms</td>
<td>O3</td>
<td>O3</td>
<td></td>
</tr>
<tr>
<td><strong>Clinical Evidence</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Diplopia (double vision)</td>
<td>O1, 3, O4</td>
<td>O1</td>
<td>O1, O3, O4</td>
</tr>
<tr>
<td>Blurred vision</td>
<td>O1, O3, O4</td>
<td>O1</td>
<td>O1, O3, O4</td>
</tr>
<tr>
<td>Bulbar weakness</td>
<td>O1, O3, O4</td>
<td>O1</td>
<td>O1, O3, O4</td>
</tr>
<tr>
<td>Impaired respiration</td>
<td>O2</td>
<td>O2</td>
<td></td>
</tr>
<tr>
<td>Progressive weakness</td>
<td>O2</td>
<td>O2</td>
<td></td>
</tr>
<tr>
<td>Progressive symmetric paralysis</td>
<td>O1, O3, O4</td>
<td>O1</td>
<td>O1, O3, O4</td>
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<tr>
<td>Healthcare record contains a diagnosis of botulism</td>
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<td></td>
<td></td>
<td>S</td>
</tr>
</tbody>
</table>

02/18/2016
Laboratory Evidence

<table>
<thead>
<tr>
<th></th>
<th>O</th>
<th>O1, O2, O4</th>
<th>O3, O4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Detection of botulinum toxin in serum</td>
<td>O</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Detection of botulinum toxin in stool²</td>
<td>O1, O2, O4</td>
<td>O1</td>
<td></td>
</tr>
<tr>
<td>Detection of botulinum toxin in patient's food</td>
<td>O1, O4</td>
<td>O1</td>
<td></td>
</tr>
<tr>
<td>Isolation of <em>Clostridium botulinum</em> from stool²</td>
<td>O1, O2, O4</td>
<td>O1</td>
<td></td>
</tr>
<tr>
<td>Isolation of <em>Clostridium botulinum</em> from wound</td>
<td>O3, O4</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Epidemiologic Evidence

| Ingestion of the same food as persons who have laboratory-confirmed botulism | A3, A4 | O1 | A? |

Special Criteria

| Age < 1 year² | N2, A4² |

Notes:
- S = These criterion alone are Sufficient to classify a case.
- N = All “N” criteria in the same column are Necessary to classify a case.
- A = These criterion must be absent (e.g., NOT present) for the case to meet the classification criteria.
- O = At least one of these “O” (Optional) criteria in each category (e.g., 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 botulism²
3 = Wound botulism
4 = Other botulism

Case Investigation Process

In most cases, identification of the source of infection is not possible. This is because *C. botulinum* spores are ubiquitous in the soil and dust. Honey has 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. Botulism toxin has been identified in home canned baby foods.

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

1) Laboratory testing
   UPHL is the only laboratory in Utah that provides testing of human and non-human samples for botulinum toxin and *C. botulinum*. The BOE screens all botulism test requests. The BOE approves tests requests after consultation with the physician and UPHL.

2) Treatment
   Healthcare providers work directly with the IBTPP to obtain BabyBIG®. The BOE ensures that this consultation has been initiated when appropriate.

3) Education
   The public health investigator should provide appropriate education to the patient’s family. The IBTPP website (http://www.infantbotulism.org/) is an excellent resource.
Outbreaks
Outbreaks of infant botulism have not been reported. However, geographic clustering of cases has been seen, and would warrant investigation.

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

Case Contact Management
Not applicable.
REFERENCES


VERSION CONTROL

2/18/2016: Created new disease plan specific to infant botulism.
UT-NEDSS Minimum/Required Fields by Tab

MORBIDITY EVENT

Demographic
- Last Name
- First Name
- State
- County
- Date of Birth
- Area Code
- Phone Number
- Birth Gender
- Ethnicity
- Race

Clinical
- Disease
- Onset Date
- Date Diagnosed
- Died
- Date of Death
- Diagnostic Facility

Laboratory
- Test Type
- Test Result
- Accession number

Epidemiological
- Childcare Associated
- Imported From
- Risk Factors
- Risk Factor Notes

Reporting
- Date first reported to public health

Investigation
- If muscle weakness/paralysis present, describe progression
  - if other) Describe progression

Administrative
- State Case Status (completed by UDOH)
- Outbreak Associated
- Outbreak Name