

neurologic deficits of botulism were more likely to have a delayed diagnosis of botulism (CDC, unpublished data, 2016).

Key Points for Clinicians

- Be aware of the spectrum of signs and symptoms of botulism, ranging from limited cranial nerve palsies (e.g., ptosis) to respiratory failure and complete extremity paralysis.
- Be aware that the respiratory system might be compromised early in the illness, when respiratory muscles (e.g., diaphragm) are unaffected but the upper airway is compromised from paresis of cranial nerve muscles, resulting in pharyngeal collapse or pooling of secretions.

Recommendations

- Consider botulism when myasthenia gravis or Guillain-Barré syndrome are suspected and in a patient with unexplained symmetric cranial nerve palsies, with or without paresis of other muscles.
- Conduct thorough, serial neurologic examinations to detect the neurologic deficits of botulism and their progression.
- If botulism is suspected, immediately contact the local or state health department's emergency on-call staff to arrange an emergency expert clinical consultation and, when indicated, request botulinum antitoxin from CDC.

Ancillary Testing

Background

Results from routine laboratory tests, including complete blood counts, examination of cerebrospinal fluid (CSF), and radiologic studies, are typically normal in patients with botulism. In Guillain-Barré syndrome, CSF protein concentrations are often elevated, especially by the second week of illness (48). In patients with botulism, mild increases in CSF protein concentrations are not reported frequently (13). Brain imaging might help exclude brainstem strokes that can produce nonlateralizing symptoms. The Tensilon (edrophonium) test, historically used to help diagnose myasthenia gravis, is usually negative in patients with botulism, although minimal responses have been reported (36).

Electrodiagnostic studies such as repetitive nerve stimulation (RNS), electromyography (EMG), and nerve conduction studies (NCSs) can help elucidate the etiology of muscle weakness. RNS involves electrically stimulating a motor nerve at either low (2–3 Hz or possibly 5 Hz) or high frequency (30–50 Hz) and recording the response in the distal muscle. EMG involves inserting a needle electrode into a muscle and recording the electrical activity at rest and with effort, showing motor unit potentials or motor unit action potentials. An

NCS involves providing an electrical stimulus to a nerve and recording the electrical response from a sensory nerve (sensory nerve conduction study) or muscle (motor nerve conduction study) (49). Distinctive classical findings of botulism are an increment in the compound motor nerve action potential amplitude, with RNS rates of 30–50 Hz (50); fibrillation; decreased recruitment of muscle units; decreased duration of muscle unit potentials with EMG; and decreased motor-evoked amplitude on an NCS with otherwise normal findings (49). However, early in the disease course, electrodiagnostic studies might be normal or almost normal and therefore not helpful.

EMG, RNS, and NCSs have several limitations. They are operator-dependent and technically challenging, require specialized training and equipment, are not available at all hospitals, and can take 2 hours to complete; in addition, the results require expert interpretation. Early in the course of botulism, electrodiagnostic testing results are likely to be normal (except for testing by single-fiber EMG) (51,52); late in the course, abnormalities are detected by these tests. EMG requires cooperation from the patient. The entire examination can be painful, especially RNS and particularly at 30–50 Hz (49). Clinicians must remember that patients with botulism who are paralyzed and intubated are still conscious (unless they are sedated); therefore, they should explain to patients who are not sedated why electrodiagnostic testing is being conducted and what they should expect. The sensitivity and specificity of EMG, RNS, and NCSs for diagnosing botulism are unknown. Electrodiagnostic findings in patients with other neuromuscular diseases (e.g., the Miller Fisher variant of Guillain-Barré syndrome) can be similar to those of botulism (50,51). More focused EMG studies, such as single-fiber EMG with measurement jitter, might be more sensitive (although it is nonspecific) than the general EMG but require even more specialized training, more expensive equipment, and more cooperation from the patient (52). The findings of electrodiagnostic studies should always be considered in the context of clinical, epidemiologic, and laboratory data.

Results from electrodiagnostic studies might help with the diagnosis of suspected botulism in settings of conventional, contingency, and crisis standards of care, depending on the situation. During an outbreak, electrodiagnostic studies are rarely needed for a cluster of patients with a clear history of bilateral, symmetric cranial nerve palsies followed by descending paralysis. However, for patients for whom the diagnosis is not clear, electrodiagnostic studies might help distinguish botulism from other neuromuscular diseases (e.g., myasthenia gravis or Guillain-Barré syndrome). This is especially true for sporadic (single) cases, in which increasing the diagnostic certainty of botulism as early as possible in the course of illness helps guide clinicians in making the critical decision to treat with antitoxin for suspected botulism rather than plasmapheresis or immunoglobulin therapy for suspected Guillain-Barré syndrome. Because findings from