

# Guillain-Barré Syndrome

## **DISEASE REPORTABLE WITHIN 24 HOURS OF DIAGNOSIS**

Per NJAC 8:57, health care providers and administrators shall report by mail or by electronic reporting within 24 hours of diagnosis, confirmed cases of Guillain-Barré syndrome to the health officer of the jurisdiction where the ill or infected person lives, or if unknown, wherein the diagnosis is made. A directory of local health departments in New Jersey is available at <http://www.nj.gov/health/lh/directory/lhdselectcounty.shtml>.

If the health officer is unavailable, the health care provider or administrator shall make the report to the Department by telephone to 609.826.5964, between 8:00 A.M. and 5:00 P.M. on non-holiday weekdays or to 609.392.2020 during all other days and hours.



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## 1 THE DISEASE AND ITS EPIDEMIOLOGY

### A. Etiologic Agent

The cause of Guillain-Barré syndrome (GBS) is unclear; it is thought to be due to an immune response with cross-reactivity to myelin after exposure to an infectious or toxic agent. Infectious agents associated with GBS include a variety of viruses and bacteria; of these, *Campylobacter jejuni* is the most commonly implicated.

### B. Clinical Description

GBS frequently begins with sensory symptoms such as paresthesias (75% of the cases) followed by symmetric flaccid paralysis. Paralysis begins more often in the feet than hands. Facial and oropharyngeal muscle weakness occurs in up to 60% of cases. Overall, 30% of cases have a clinical presentation consistent with respiratory failure. Electrophysiologic studies are abnormal in 95% to 99% of patients. They show features of demyelination and/or axonal damage and no muscle involvement. Complete blood count (CBC) and sedimentation rate (ESR) are normal in most patients. Cerebrospinal fluid (CSF) analysis shows increased protein (>55 mg/dL in 90% of cases), and the majority of patients will have no CSF pleocytosis (<50 white blood count [WBC]/microliter).

### C. Reservoirs

Not applicable.

### D. Modes of Transmission

Modes of transmission are unknown. GBS in and of itself is not known to be communicable from person to person.

### E. Incubation Period

Since the exact cause of GBS is unknown in most cases, the incubation period cannot be determined with great certainty. If a preceding respiratory or gastrointestinal illness is

identified, it usually occurs one to three weeks prior to GBS symptom onset, and GBS may develop up to two months following a symptomatic episode of *C. jejuni* gastrointestinal infection.

#### **F. Period of Communicability or Infectious Period**

GBS in and of itself is not known to be communicable from person to person.

#### **G. Epidemiology**

GBS occurs worldwide. The annual incidence in the United States is close to 1.0 per 100,000 with the attack rate higher in males (0.52 per 100,000 persons) than in females (0.40 per 100,000 persons). GBS among children under 15 years old represents more than half of all acute flaccid paralysis cases. In one study, 41% of the severely affected GBS patients showed serologic evidence for infection with *C. jejuni*, cytomegalovirus, Epstein-Barr virus, or *Mycoplasma pneumoniae*; but only 16% of the mildly affected group showed such evidence. Studies from Sweden are showing that the risk of developing GBS during the two months following a symptomatic episode of *C. jejuni* infection is approximately 100 times higher than the risk in the general population. Several studies have demonstrated seasonal preponderance of GBS in the spring. In New Jersey, an annual average of 12 cases of GBS are confirmed by the New Jersey Department of Health and Senior Services (NJDHSS), with a range of 4 to 20 cases in any given year. Most cases of GBS reported in New Jersey do not have adequate clinical information to identify a risk factor for illness; of the cases with adequate clinical history, approximately half report a history of viral infection preceding the illness.

## **2 NEW JERSEY DEPARTMENT OF HEALTH AND SENIOR SERVICES CASE DEFINITION**

#### **A. Clinical Description**

An illness characterized by symmetric flaccid paralysis.

#### **B. Laboratory Criteria and Case Classification**

##### **CONFIRMED**

A clinically diagnosed case supported by laboratory evidence including

- Electrophysiologic studies showing nerve conduction slowing with features of demyelination and/or axonal damage, AND
- CSF with elevated protein after one week of symptoms, and fewer than 50 WBC/microliter, AND
- Normal CBC and ESR in most patients

**PROBABLE**

A clinically diagnosed case without any laboratory evidence

**POSSIBLE**

Not used

**C. Differences from CDC Case Definition**

There is no formal case definition for GBS provided by the CDC, as GBS is not a nationally notifiable disease.

## **3 LABORATORY TESTING AVAILABLE**

Diagnosis of GBS is made based primarily on clinical grounds. A diagnostic test specific for GBS does not exist.

## **4 PURPOSE OF SURVEILLANCE AND REPORTING AND REPORTING REQUIREMENTS**

**A. Purpose of Surveillance and Reporting**

- To identify cases and better understand the epidemiology of GBS
- To provide information about the GBS, its transmission, and methods of prevention
- To promptly identify clusters or outbreaks of GBS related to a common cause and initiate appropriate prevention measures

**B. Laboratory Reporting Requirements**

Not applicable.

**C. Healthcare Provider Reporting Requirements**

The New Jersey Administrative Code (NJAC 8:57-1.4) stipulates that healthcare providers report (by telephone, confidential fax, or in writing) all cases of GBS to the local health officer having jurisdiction over the locality in which the patient lives or, if unknown, to the health officer in whose jurisdiction the healthcare provider requesting the laboratory examination is located. The report shall contain the name of the disease; date of illness onset; and name, age, date of birth, race, ethnicity, home address, and telephone number of the person being reported. In addition, name, address, institution, and telephone number of reporting official, and other information as may be required by NJDHSS concerning a specific disease.

**D. Health Officer Reporting**

The New Jersey Administrative Code (NJAC 8:57-1.7) stipulates that each local health officer must report the occurrence of any case of GBS within 24 hours of receiving the report. Written or electronic copies of the reports must be made to NJDHSS and may be submitted over the Internet using the confidential and secure Communicable Disease Reporting and Surveillance System (CDRSS).

# 5 CASE INVESTIGATION

**A. Forms**

It is requested that the local health officer complete a Guillain-Barré Syndrome Case Report form, which can be found online at <http://www.state.nj.us/health/forms/cds-9.dot>, by interviewing the clinician, patient, and others who may be able to provide pertinent information. Information requested on this form is needed to determine case status. Be sure to ask about whether the patient has any preceding infection (viral or bacterial, ask specifically about campylobacteriosis), vaccination, or surgery in the past two months. Collect information about clinical symptoms, electrophysiologic studies, laboratory analysis of CSF, treatment with plasmapheresis, and complications associated with the illness.

**B. Entry into CDRSS**

The mandatory fields in CDRSS include disease, last name, county, municipality, gender, race, ethnicity, case status, and report status.

The following table can be used as a quick reference guide to determine which CDRSS fields need to be completed for accurate and complete reporting of GBS. The “Tab” column includes the tabs that appear along the top of the CDRSS screen. The “Required Information” column provides detailed explanations of what data should be entered.

CDRSS Screen	Required Information
<b>Patient Info</b>	Enter the disease name (“GUILLAIN BARRÉ SYNDROME”), patient demographic information, illness onset date, and the date the case was reported to the local health department (LHD).
<b>Addresses</b>	Enter any alternate address (e.g., a daycare address). Use the “COMMENTS” section in this screen to record any pertinent information about the alternate address (e.g., the times per week the case-patient attends daycare). Entering an alternate address will allow other disease investigators access to the case if the alternate address falls within their jurisdiction.

CDRSS Screen	Required Information
<b>Clinical Status</b>	Enter any treatment that the patient received, especially plasmapheresis, and record the names of the medical facilities and physician(s) involved in the patient’s care. If the patient received care from two or more hospitals or healthcare providers, be sure that all are entered so the case can be accessed by other jurisdictions.
<b>Signs/Symptoms</b>	Check appropriate boxes for signs and symptoms and indicate their onset date. Make every effort to get complete information by interviewing the physician, nurse, or office manager abstracting medical information from the patient’s chart, or the patient. Also, information regarding the resolution of signs and symptoms should be entered.
<b>Risk Factors</b>	Enter complete information about risk factors, including previous viral or bacterial infections, specifically campylobacteriosis. Also enter complete information about any surgery or vaccination in the two months preceding the illness.
<b>Contact Tracing</b>	Information regarding contacts is not required for this disease.
<b>Case Comments</b>	Enter general comments (i.e., information that is not discretely captured by a specific topic screen or drop-down menu) in the “COMMENTS” section. <b>NOTE:</b> Select pieces of information entered in the “COMMENTS” section CANNOT be automatically exported when generating reports. Therefore, whenever possible, record information about the case in the fields that have been designated to capture this information; information included in these fields CAN be automatically exported when generating reports.
<b>Epidemiology</b>	Information regarding epidemiology is not required for this disease.

CDRSS Screen	Required Information
<p><b>Case Classification Report Status</b></p>	<p>Case Classification options are “REPORT UNDER INVESTIGATION (RUI),” “CONFIRMED,” “PROBABLE,” “POSSIBLE,” and “NOT A CASE.”</p> <ul style="list-style-type: none"> <li>• All cases entered by NJDHSS will be assigned a case status of “REPORT UNDER INVESTIGATION (RUI).”</li> <li>• Cases still under investigation by the LHD should be assigned a case status of “REPORT UNDER INVESTIGATION (RUI).”</li> <li>• Upon completion of the investigation, the LHD should assign a case status on the basis of the GBS case definition (options include “CONFIRMED,” “PROBABLE,” and “NOT A CASE”).</li> </ul> <p>Report status options are “PENDING,” “LHD OPEN,” “LHD REVIEW,” “LHD CLOSED,” “DELETE,” “REOPENED,” “DHSS OPEN,” “DHSS REVIEW,” and “DHSS APPROVED.”</p> <ul style="list-style-type: none"> <li>• Cases entered by NJDHSS will be assigned a report status of “PENDING.”</li> <li>• Once the LHD begins investigating a case, the report status should be changed to “LHD OPEN.”</li> <li>• The “LHD REVIEW” option can be used if the LHD has a person who reviews the case before it is closed (e.g., health officer or director of nursing).</li> <li>• Once the LHD investigation is complete and all the data are entered into CDRSS, the LHD should change the report status to “LHD CLOSED.”</li> <li>• “LHD CLOSED” cases will be reviewed by DHSS and be assigned one of the DHSS-specific report status categories. If additional information is needed on a particular case, the report status will be changed to “REOPENED” and the LHD will be notified by e-mail. Cases that are “DHSS APPROVED” cannot be edited by LHD staff (see section 5C below).</li> </ul> <p>If a case is inappropriately entered (e.g., a different reportable disease or condition was erroneously entered as a case of GBS), the case should be assigned a report status of “DELETE.” A report status of “DELETE” should NOT be used if a reported case of GBS simply does not meet case definition.</p>

### **C. Other Reporting/Investigation Issues**

1. It is not always possible to obtain all the information necessary to determine the case status of a patient. A minimum of three attempts should be made to obtain necessary information. If at this time information is not acquired, the case should be entered into CDRSS with as much information is known, attempts (dates and results of the attempts) should be documented in the “Comments” section, and the case status changed to “NOT A CASE” and report status to “LHD CLOSED.”
2. Case report forms (Guillain-Barré Syndrome Case Report form and medical records) DO NOT need to be mailed to NJDHSS as long as mandatory fields indicated in section 5B are completed.
3. Once an LHD completes its investigation and assigns a report status of “LHD CLOSED,” NJDHSS will review the case. NJDHSS will approve the case by changing the report status to “DHSS APPROVED.” At this time, the case will be submitted to CDC and the case will be locked for editing. If additional information is received after a case has been placed in “DHSS APPROVED,” an LHD will need to contact NJDHSS to reopen the case. This should be done only if the additional information changes the case status of the report.
4. Every effort should be made to complete the investigation within three months of opening a case. Cases that remain open for three months or more and have no investigation or update notes will be closed by NJDHSS and marked as “NOT A CASE.”

## **6 CONTROLLING FURTHER SPREAD**

### **A. Isolation and Quarantine Requirements (NJAC 8:57-1.10)**

Because GBS is not transmitted from person to person, there are no restrictions for case-patients or contacts of case-patients.

### **B. Protection of Contacts of a Case**

There are no restrictions of contacts.

### **C. Managing Special Situations/Outbreak Suspected**

If the number of reported cases in a setting or town/community is higher than usual, or if an outbreak is suspected, please contact IZDP at 609.588.7500 immediately. This situation may warrant an investigation of the clustered cases to determine a course of action to prevent further cases. IZDP staff can perform surveillance for clusters of illness that may cross several jurisdictions and therefore be difficult to identify at a local level.

# 7 PREVENTIVE MEASURES

There are no environmental or personal preventive measures for GBS.

## Additional Information

A GBS FAQ sheet can be obtained at the NJDHSS Web site at <http://www.state.nj.us/health/cd/>.

## References

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