



SECTION 13 – NEUROLOGY

- [Guillain-Barré Syndrome](#)
- [Migraine Headache](#)
- [Multiple Sclerosis](#)
- [Traumatic Brain Injury \(TBI\)](#)

GUILLAIN-BARRÉ SYNDROME

Background

This case definition was developed by the Armed Forces Health Surveillance Center (AFHSC) for the purpose of epidemiological surveillance of Guillain-Barré syndrome (GBS). Methodical surveillance of GBS cases facilitates investigations of the potential etiologies of this condition as well as assessment of its impact on military service members. The three case definitions presented below were developed to test their sensitivity, specificity, and positive predictive value (PPV) in identifying “true” cases of GBS. Medical records of possible cases were reviewed by the Military Vaccine Agency (MILVAX), using a standardized medical chart abstraction form based on the Brighton Collaboration case definition, to determine which of the possible cases were confirmed, “true”, GBS cases.^{1, 2}

Clinical Description

Guillain-Barré syndrome (GBS) is clinically characterized by acute, progressive, and generally ascending muscle weakness, loss of deep tendon reflexes, and paralysis. With appropriate medical care, including hospitalization and ventilatory assistance when needed, recovery occurs in most cases although convalescence and rehabilitation may be prolonged. The etiology of GBS is not completely understood. It is hypothesized to be autoimmune in nature because many cases are preceded by acute respiratory or gastrointestinal infections.^{1,3} There has been continuing interest in exploring associations between GBS and antecedent influenza, other respiratory infections, influenza vaccination, and other immunizations.⁴

Case Definition and Incidence Rules - Definition A

For surveillance purposes, a case of Guillain-Barré is defined as:

- *One inpatient medical encounter* with a defining diagnosis of Guillain-Barré (see ICD9 code below) in *any* diagnostic position.

Incidence rule:

For individuals who meet the case definition:

- An individual is considered an incident case only *once per lifetime*.

Exclusions:

- None

¹ Armed Forces Health Surveillance Center. Predictive Value of Surveillance Case Definition of Guillain-Barré Syndrome in Vaccine Safety Assessment. *Medical Surveillance Monthly Report (MSMR)*. 2012 March; 19(3): 8-9.

² Sejvar JJ, Kohl KS, Gidudu J, et al. Guillain-Barré Syndrome and Fisher Syndrome: case definitions and guidelines for collection, analysis, and presentation of immunization safety data. *Vaccine*. 2011; 29(3): 599-612.

³ Hughes RAC, Cornblath DR. Guillain-Barré syndrome. *Lancet*. 2005; 366(9497): 1653-1666.

⁴ Tokars JJ, Lewis P, DeStefano F, et al. The risk of Guillain-Barré Syndrome associated with influenza A (H1N1) 2009 monovalent vaccine and 2009-2010 seasonal influenza vaccines: results from self-controlled analyses. *Pharmacoepidemiol Drug Saf*. 2012. Doi:10.1002/pds.3220.



Case Definition and Incidence Rules – Definition B

For surveillance purposes, a case of Guillain-Barré is defined as:

- *One inpatient medical encounter* with a defining diagnosis of Guillain-Barré (see ICD9 code below) in *any* diagnostic position; AND
- *One outpatient medical encounter* with a defining diagnosis of Guillain-Barré (see ICD9 code below) in *any* diagnostic position.

Incidence rule:

For individuals who meet the case definition:

- An individual is considered an incident case only *once per lifetime*.

Exclusions:

- None

Case Definition and Incidence Rules – Definition C

For surveillance purposes, a case of Guillain-Barré is defined as:

- *One inpatient medical encounter* with a defining diagnosis of Guillain-Barré (see ICD9 code below) in the *primary* diagnostic position; AND
- *One outpatient medical encounter* with a defining diagnosis of Guillain-Barré (see ICD9 code below) in the *primary* diagnostic position.

Incidence rule:

For individuals who meet the case definition:

- An individual is considered an incident case only *once per lifetime*.

Exclusions:

- None

Codes

The following ICD9 code is included in the case definition:

Condition	ICD-9-CM Codes	CPT Codes
Guillain-Barré Syndrome	357.0 (Acute infective polyneuritis – Guillain-Barré syndrome, postinfectious polyneuritis)	



Development and Revisions

The three case definitions outlined above were developed based on reviews of the ICD9 codes and scientific literature. The case definitions were developed by the AFHSC staff for a March 2012 MSMR article.¹

Case Definition and Incidence Rule Rationale

- The three case definitions were developed to test their sensitivity, specificity, and PPV in identifying “true” cases of GBS.

Reports

None

Review

Apr 2012 Case definitions reviewed and adopted by the AFHSC Surveillance Methods and Standards (SMS) working group.

Mar 2012 Case definitions reviewed and published by the AFHSC MSMR staff.

Comments

Results of analysis:¹

Definitions		Sensitivity	Specificity	Positive Predictive Value (PPV)
A	One inpatient medical encounter in <i>any</i> diagnostic position	100%	81%	78%
B	One inpatient <i>and</i> one outpatient medical encounter in <i>any</i> diagnostic position	100%	88%	86%
C	One inpatient <i>and</i> one outpatient medical encounter, both in the <i>primary</i> diagnostic position	92%	92%	88%



MIGRAINE HEADACHE

Background

This case definition was developed by the Armed Forces Health Surveillance Center (AFHSC) for the purpose of epidemiological surveillance of a condition important to military-associated populations. Migraine headaches are a leading cause of medical encounters and lost duty time among military members. During migraine attacks, the military operational capabilities of those affected may be sharply reduced.¹

Clinical Description

Migraine is a neurovascular syndrome that is generally manifested as recurrent, severe headaches that can be debilitating. Migraine headaches are typically throbbing, localized, and very painful. They are often distinguishable from other types of headaches by characteristic symptoms such as nausea, increased sensitivity to light and sound, and dizziness. Migraines are often preceded by neurologic symptoms (“migraine aura”) which may include blurred or obstructed vision, skin numbness or tingling, exaggerated responses to painful stimuli, and momentary loss of consciousness (syncope).²

Case Definition and Incidence Rules

For surveillance purposes, a case of migraine headache is defined as:

- *One inpatient medical encounter* with any of the defining diagnoses of migraine (see ICD9 code list below) in the *any* diagnostic position; or
- *One outpatient medical encounter* with any of the defining diagnoses of migraine (see ICD9 code list below) in the *primary* diagnostic position.

Incidence rules:

For individuals who meet the case definition:

- The incidence date is considered the date of the first inpatient or outpatient medical encounter that includes a defining diagnosis of migraine.
- Each individual is considered an incident case only once per lifetime.

Exclusions:

- None

¹ Armed Forces Health Surveillance Center. Risk factors for migraine after OEF/OIF deployment, active component, U.S. Armed Forces. *Medical Surveillance Monthly Report (MSMR)*. 2009 December; 16(12):10-13.

² Armed Forces Health Surveillance Center. Migraine and other headaches, active components, U.S. Armed Forces, 2001-2007. *Medical Surveillance Monthly Report (MSMR)*. 2008 May; 15(4):6-10.



Codes

The following ICD9 codes are included in the case definition:

Condition	ICD-9-CM codes	CPT Codes
Migraine Headache	346.0 (migraine with aura) [0-3] 346.1 (migraine without aura) [0-3] 346.2 (variants of migraine, not elsewhere classified) [0-3] 346.3 (hemiplegic migraine) [0-3] 346.4 (menstrual migraine) [0-3] 346.5 (persistent migraine aura without cerebral infarction) [0-3] 346.6 (persistent migraine with cerebral infarction) [0-3] 346.7 (chronic migraine without aura) [0-3] 346.8 (other forms of migraine) [0-3] 346.9 (migraine, unspecified) [0-3] <i>Fifth-digit subclassification :</i> 0 without mention of intractable migraine without mention of status migrainosus 1 with intractable migraine, so stated, without mention of status migrainosus 2 without mention of intractable migraine with status migrainosus 3 with intractable migraine, so stated, with status migrainosus	NA

Development and Revisions

The case definition was originally developed by the AFHSC Medical Surveillance Monthly Report (MSMR) staff for the MSMR article referenced above.² An expert on migraine headaches from the Uniformed Services University for Health Sciences was consulted during the development of this definition.

Case Definition and Incidence Rule Rationale

- Per incidence rules defined in a 2009 MSMR article, if counting multiple migraine events in one individual, medical encounters must *occur at least one week apart*.³

³ Army Medical Surveillance Activity. Migraines among active duty military personnel, 1998-1999. *Medical Surveillance Monthly Report (MSMR)*. 2000 July; 6(6):14-17.



Reports

None

Review

Mar 2011 Case definition reviewed and adopted by AFHSC Surveillance Methods and Standards (SMS) working group.

May 2008 Case definition developed and reviewed by AFHSC MSMR staff.

Comments

None



MULTIPLE SCLEROSIS

Background

This case definition was developed by the Armed Forces Health Surveillance Center (AFHSC) for the purpose of epidemiological surveillance of a condition important to military-associated populations. Multiple sclerosis (MS) often manifests for the first time in the age range of most military members and MS-related disabilities can degrade the operational capabilities of affected service members.

Clinical Description

Multiple sclerosis is an autoimmune disease of the central nervous system. It is characterized by inflammation, demyelination, and axon degeneration resulting in impaired nerve conduction. The signs and symptoms vary and may include weakness, painful muscle spasms, bladder dysfunction, vision disturbances, impaired speech or swallowing, tremor, poor balance, difficulty with coordination, and cognitive impairment. The clinical course of MS varies from patient to patient; however, the usual course is characterized by recurrent clinical exacerbations. Exacerbations can produce new deficits, worsen persistent deficits, or resolve with complete recovery. In some cases, the disease progresses with worsening disabilities that can be life-threatening.¹

Case Definition and Incidence Rules

For surveillance purposes, a case of multiple sclerosis is defined as:

- *One inpatient medical encounter* with a defining diagnosis of multiple sclerosis (see ICD9 code list below) in *any* diagnostic position; or
- *Two outpatient medical encounters, at least 1 day apart*, with a defining diagnosis of multiple sclerosis (see ICD9 code list below) in *any* diagnostic position; or
- *One inpatient or one outpatient medical encounter* with any of the defining diagnoses of “other demyelinating diseases of the central nervous system” (see ICD9 code list below) in any diagnostic position, followed by *one outpatient medical encounter* with a defining diagnosis of multiple sclerosis (see ICD9 code list below) in *any* diagnostic position.

Incidence rules:

For individuals who meet the case definition:

- The incidence date is considered the date of the first inpatient or outpatient medical encounter that includes a diagnosis of multiple sclerosis or one of the defining diagnoses of “other demyelinating diseases of the central nervous system”.
- An individual can be considered an incident case *once per lifetime*.

(continued on next page)

¹ Armed Forces Health Surveillance Center. Multiple Sclerosis, Active Component, U.S. Armed Forces, 2000-2009. *Medical Surveillance Monthly Report (MSMR)*. 2011 January; 18(1): pp. 12-15.



Case Definition and Incidence Rules (cont.)

Exclusions:

- None

Codes

The following ICD9 codes are included in the case definition:

Condition	ICD-9-CM Codes	CPT Codes
Multiple Sclerosis	340 (multiple sclerosis)	NA
Other demyelinating diseases of the central nervous system	341 (other demyelinating diseases of the central nervous system) 341.0 (neuromyelitis optica) 341.1 (schilder's disease) 341.2 (acute transverse myelitis) - 341.20 (acute transverse myelitis not otherwise specified) - 341.21 (acute transverse myelitis in conditions not classified elsewhere) - 341.22 (idiopathic transverse myelitis) 341.8 (other demyelinating diseases of central nervous system) 341.9 (demyelinating disease of central nervous system, unspecified)	

Development and Revisions

This case definition was developed in January of 2011 by a preventive medicine resident doing a rotation at AFHSC. Neurology experts from the Uniformed Services University of the Health Sciences (USUHS) and the Medical Surveillance Monthly Report (MSMR) staff were consulted during the development of this definition. The definition was developed based on reviews of the ICD9 codes, the scientific literature, and previous AFHSC analyses and is used in the MSMR article on multiple sclerosis referenced above.¹

Case Definition and Incidence Rule Rationale

- This surveillance case definition is designed to identify as many “true cases” of MS as possible while limiting the number of “false positive” cases.



Code Set Determination and Rationale

- The diagnosis of MS often requires two or more clinically distinct episodes of central nervous dysfunction with at least partial resolution between episodes. As such, the first medical encounter for a patient with symptoms of a demyelinating episode suggestive of MS may be characterized and recorded as a nonspecific or “other demyelinating disease of the central nervous system.” For this reason, ICD9 codes 341.xx (other demyelinating diseases of the central nervous system) are used as first encounter criteria for case identification.

Reports

None

Review

Nov 2011	Case definition reviewed and adopted by the AFHSC Surveillance Methods and Standards (SMS) working group.
Jan 2011	Case definition developed by a USUHS preventive medicine resident and reviewed by AFHSC MSMR staff.

Comments

Limitations of case definition and MS surveillance

- Multiple sclerosis is a clinical diagnosis with no confirmatory laboratory or radiologic tests; as such, it can be difficult to diagnose. If many provisional or “rule-out” diagnoses were reported with MS-specific diagnosis codes, the actual incidence rate of MS in military members would be overestimated. Similarly, incidence could be underestimated if active component members sought medical care from sources other than the military health system or purchased care providers; if clinical manifestations of MS were not identified as MS-related or not reported with MS-specific diagnosis codes; or if affected individuals terminated their military service before the case definition criteria was met.¹



TRAUMATIC BRAIN INJURY (TBI)

DoD Standard Surveillance Case Definition for TBI Adapted for AFHSC Use

Background

A case definition for TBI was originally developed in August 2008 by policy makers and medical experts from all Services and from several DoD and Health and Human Services (HHS) agencies. The Defense and Veterans Brain Injury Center (DVBIC) monitors the TBI definition for the military health system (MHS) and, together with the above agencies, has revised the original definition several times since its creation. These revisions are outlined below (see *Development and Revisions* section).

Clinical Description

In the U.S. Military Health System (MHS), traumatic brain injury (TBI) is defined as “traumatically induced structural injury and/or physiological disruption of brain function as a result of an external force that is indicated by new onset or worsening of at least one of the following clinical signs, immediately following the event: any period of loss of or decreased level of consciousness; any loss of memory for events immediately before or after the injury; any alteration in mental state at the time of the injury (confusion, disorientation, slowed thinking, etc.); neurological deficits (weakness, loss of balance, change in vision, praxis, paresis/plegia, sensory loss, aphasia, etc.) that may or may not be transient; intracranial lesion.”¹

Case Definition and Incidence Rules

For surveillance purposes, a case of TBI is defined as:

- *One inpatient medical encounter*, with any of the defining diagnoses of TBI in *any* diagnostic position (see ICD9 code list below); or
- *One outpatient medical encounter*, with any of the defining diagnoses of TBI in *any* diagnostic position (see ICD9 code list below).
- *If analysis requires “deployment-associated” incident case counts*, the initial defining encounter must have occurred while the individual was deployed to, or within 30 days of returning from, a theater of operations of interest and the deployment must have been for 30 days or longer (see *Development and Revisions* section below).¹

Incidence rules:

For individuals who meet the case definition:

- The incidence date is considered the date of the first inpatient or outpatient medical encounter that includes a defining diagnosis of TBI.
- An individual is considered an incident case only once per lifetime.

Exclusions:

- None

¹ Memorandum from the Assistant Secretary of Defense (Health Affairs). Traumatic brain injury: definition and reporting, dated 1 Oct 2007. U.S Department of Defense, Washington, DC.



Codes

The following ICD9 codes are included in the case definition:^{2,3}

Condition	ICD-9-CM codes	CPT Codes
Traumatic Brain Injury (TBI)	310.2 (postconcussion syndrome) 800.0x – 800.9x (fracture of vault of skull) 801.0x – 801.9x (fracture of base of skull) 803.0x – 803.9x (other and unqualified skull fractures) 804.0x – 804.9x (multiple fractures involving skull or face with other bones) 850.x (concussion) 851.0x – 851.9x (cerebral laceration and contusion) 852.0x – 852.5x (subarachnoid, subdural, and extradural hemorrhage, following injury) 853.0x – 853.1x (other and unspecified intracranial hemorrhage following injury) 854.0x – 854.1x (intracranial injury of other and unspecified nature) 907.0 (late effect of intracranial injury <i>without</i> skull or facial fracture) 950.1 - 950.3 (injury to optic chiasm/pathways or visual cortex) 959.01 (head injury, unspecified) <i>Personal history of TBI</i> V15.52 (no extenders); V15.52_0 thru V15.52_9 ; V15.52_A thru V15.52_F (currently only codes in use) V15.5_1 thru V15.5_9; V15.5_A thru V15.5_F* V15.59_1 thru V15.59_9; V15.59_A thru V15.59_F*	NA

*See “Development and Revisions” section below

² ICD9 code 995.55 (shaken infant syndrome) is included in the standard DoD TBI case definition in an effort to be consistent with the CDC. This code is not used by AFHSC as it is not relevant to military surveillance objectives.

³ Case definition and ICD9 codes are based on “TBI: Appendix F-G dated 5/1/10 and Appendix 7 dated 2/26/10: from *Military Health System Coding Guidance: Professional Services and Specialty Coding Guidelines* (Version 3.2) by the Unified Biostatistical Utility working group.



Development and Revisions

Revisions to the DoD's surveillance case definition below are the result of collaborations and consensus of the Defense and Veterans Brain Injury Center (DVBIC), the Traumatic Brain Injury Task Force, the Defense Centers of Excellence for Psychological Health and Traumatic Brain Injury (DCoE), the US Army Public Health Command (USAPHC), formerly the Center for Health Promotion and Preventive Medicine (CHPPM), the Armed Forces Health Surveillance Center (AFHSC), and the Centers for Disease Control and Prevention (CDC).⁴

Jan 2010

- V codes: V15.52_1 thru V15.52_9 and V15.52_A thru V15.52_F were *added* to the surveillance case definition. These codes replaced the V15.5 and the V15.59 series.
- V codes: V15.5_1 thru V15.5_9 and V15.59_A thru V15.59_F are no longer included in the ICD9 coding system. However, the V15.5 and the V15.59 series codes are included in the surveillance case definition for analyses using data from 2010 and prior.

Dec 2008

- ICD9 code 907.0 (late effect of intracranial injury *without* mention of skull fracture) was *added* to surveillance case definition for TBI. The working group felt that adding 907.0 would improve accuracy. The group acknowledged that late effect codes might suggest a consequence of TBI versus a prevalent case and that the purpose of DoD surveillance is to track the prevalence of TBI and not the consequences of TBI.
- The possibility of adding ICD9 code 905.0 (late effect of fracture of skull *and face bones*) was discussed by the working group. The code was not included because the code included facial fractures and the group felt this might add a large number of face and nose fractures that were not accompanied by a TBI.

Oct 2008

In an effort to standardize the codes used by DVBIC, AFHSC, and the CDC, the following changes were made to the TBI case definition. These changes significantly increased the numbers of TBI cases ascertained for routine surveillance.

- ICD9 code 310.2 (postconcussion syndrome) was *added* to the surveillance case definition. The working group added the code because there are some instances where there are no electronic records of earlier encounters associated with other TBI codes. In such instances, code 310.2 would be the only indicator that a TBI had been sustained.

The CDC does not include ICD9 code 310.2 in their case definition for TBI because the code likely indicates prevalent cases and they are interested in incident cases only. On the contrary, the military surveillance objectives are to identify those suffering from TBI and TBI-related conditions, i.e., prevalent cases, as well as new occurrences of TBI, i.e., incident cases.
- ICD9 code 802 (fracture of face bones) was *removed* from the surveillance case definition. The working group agreed that, although the 802 series is a possible indicator of a TBI, the inclusion of a diagnosis in the 802 series without any other defining TBI code, would increase the number of false positives.
- ICD9 codes 950.1-950.3 (injury to optic chiasm, optic pathways, visual cortex) and V codes V15.5_1 thru V15.5_9 and V15.5_A thru V15.5_F were *added* to the definition. These changes were made to be consistent with the CDC, to make the data analysis "all inclusive and comparable to civilian numbers."

⁴ Rationale for code decisions obtained from Executive Summaries of the "Surveillance Workgroup meetings for TBI related ICD9 codes" sponsored by the Defense and Veterans Brain Injury Center (DVBIC) and the TBI Component of the Defense Centers of Excellence (DCoE), Oct 8, Aug 27, and Sept 17, 2008.



- ICD9 code 995.55 (shaken infant syndrome) is included in the standard DoD TBI case definition in an effort to be consistent with the CDC. This code is not used by AFHSC as it is not relevant to military surveillance objectives.

Aug 2008 The original case definition for TBI was developed in August 2008 by policy makers and medical experts from all Services and from various agencies including DVBIC, the Traumatic Brain Injury Task Force, the DCoE, the AFHSC, and the CDC.

Deployment-Associated Incident Cases of TBI

If an analysis requires “deployment-associated” incident case counts, AFHSC includes the restriction described above in the case definition. AFHSC includes Department of Defense defined operations associated with deployment in many of its analyses, (e.g., Operations Enduring Freedom (OEF), Iraqi Freedom (OIF), and New Dawn (OND)).

The specification of “within 30-days of returning” represents the best judgment of a reasonable time frame to allow for post-deployment medical evaluations and follow-up to detect incident cases of deployment-associated TBI. When estimating such cases, it should be noted that a causal association for TBI due to an event that occurred during a deployment or direct combat cannot be proven using data available to AFHSC.

Code Set Determination and Rationale

- *TBI DoD Extender Codes.* The TBI V code extenders, (i.e., 1-9 and A-F), are not part of the ICD-9-CM coding system. They are one-character extenders (sixth digits) added to a specific ICD-9-CM code to denote a unique meaning. The codes were developed by TRICARE and adapted for use in the DoD to capture additional detail on the nature of injury, (penetrating vs. non-penetrating), severity of injury, (loss of consciousness, post-traumatic amnesia, Glasgow Coma Scale), and to indicate if the diagnosis was made in the context of the Global War on Terrorism (GWOT). These TBI-specific V-codes became available 28 Jan 08.
- Code lists used by DVBIC, AFHSC, and the CDC were compared and considered in the development of this definition.

Reports

AFHSC reports on TBI regularly in the following reports. Defense Medical Surveillance System and Theater Medical Data Store data are used for all reports.

- Quarterly: “DoD Consolidated TBI Healthcare Encounter Quarterly Report” for the Office of the Assistant Secretary of Defense for Health Affairs (HA)
- Monthly: surveillance case listing for DVBIC
- Monthly: *Deployment- related conditions of special surveillance interest, U.S. Armed Forces, by month and service.* Medical Surveillance Monthly Report (MSMR).

Review

Nov 2010 Case definition reviewed and adopted by AFHSC Surveillance Methods and Standards (SMS) working group.

Nov 2010 Case definition reviewed by DVBIC, Manager of TBI Surveillance Systems



Comments

TBI Severity

In 2008, DVBIC, in collaboration with the CDC, AFHSC, FHP, PHC (formerly CHPPM), and others classified all relevant TBI ICD-9 codes by severity (mild, moderate, severe, penetrating, and unclassified). A pilot test was conducted by AFHSC to determine if all 25 variables outlined in the 1 Oct 07 memo were available in the data sources accessible to AFHSC. Upon completion of this pilot test, it was determined that not all 25 variables could be extracted from the medical records; therefore, AFHSC could only provide DVBIC with a limited dataset in support of the TBI surveillance monthly case listing.

