West Nile/St. Louis Encephalitis Virus Classification Guide

The following algorithms are designed to assist with investigators classifying arboviral disease cases, specifically West Nile and St. Louis encephalitis virus cases. The first algorithm assists with selecting the ‘MEDSIS DSO Type’ for categorization of cases as neuroinvasive or non-neuroinvasive.

The second algorithm assists with case classification. In general, the most common case scenarios are as follows:

- Blood donor with symptoms = Confirmed
- Blood donor without symptoms = Not a Case
- Serologic testing in serum without plaque reduction neutralization (PRNT) testing = Probable
- Serologic testing in CSF for only one virus = Probable
- Serologic testing in CSF for both viruses, when testing was only positive for a single virus = Confirmed
- If PRNT testing was performed and definitive for a single virus = Confirmed
- If PRNT testing cannot distinguish between viruses = Probable Unspecified Flavivirus Group 1

For additional assistance in classifying cases, please contact the Arizona Department of Health Services Vector-borne & Zoonotic Disease Program at vbzd@azdhs.gov or (602)-364-3676.
West Nile/St. Louis Encephalitis Virus Type/Symptom Classification (I)

Based on medical records or interview, select the most compatible symptom profile*

**Fever or chills** (measured or subjective) AND absence of neuroinvasive disease AND absence of a more likely clinical explanation. Other compatible symptoms include: headache, myalgia, rash, arthralgia, vertigo, vomiting.

**Headache, stiff neck AND pleocytosis** (increase in white blood cell count) in CSF with or without fever. *It should NOT present with altered mental status or focal neurologic deficits.*

**Headache AND altered mental status** (from confusion to coma) with or without fever. Many patients can become confused or have altered mental status due to their fever or other conditions or medicines. *Ensure there are some signs or evidence of infection or inflammation of the brain.*

**Rapid-onset** (symptoms present over the course of hours to days from onset) AND Extremity, facial, and/or respiratory muscle weakness AND ‘Flaccid’ or decreased muscle tone in the affected areas (a description of “weakness” alone would NOT qualify as flaccid) (see full definition on slide 4).

**Case with Guillain-Barré syndrome (GBS) diagnosis.** Clinically, it may present with an acute, bilateral, progressive, flaccid weakness of the extremities and/or cranial nerve muscles, and is usually accompanied by reduced or absent reflexes (see full definition on slide 5).

**Neuroinvasive presentations not covered under the categories of AFP, encephalitis, or meningitis, acute flaccid paralysis, GBS (ex. optic neuritis or Bell’s palsy).**

**No fever or chills** AND no symptoms of neuroinvasive disease.

**Unknown symptoms**

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*Note: Symptom profile for Uncomplicated Fever is described in the Arboviral Disease Case Definition: clinical criteria for non-neuroinvasive disease. For additional information, please see the Arizona Department of Health Services, Case Definitions for Communicable Morbidities.*

Last updated: 3/6/2020

Updates only related to neurological classifications.
West Nile/St. Louis Encephalitis Virus Type/Symptom Classification (II) – Helpful Tips

**Meningitis:**

- Meningitis is **infection or inflammation of the tissues that cover the brain** (i.e., the meninges).
- Clinically, this may present with **fever, headache, photophobia or light sensitivity, and/or new nuchal rigidity** (the inability to flex one’s neck forward). Unless there is a concurrent encephalitis (i.e., meningoencephalitis), pure meningitis **should NOT present with prominent altered mental status or focal neurologic deficits.**
- Diagnostically, it may be associated with a cerebrospinal fluid (CSF) pleocytosis, but brain imaging on CT or MRI may be normal
- If meningitis co-exists with an encephalitis (i.e., meningoencephalitis), it should be reported as **encephalitis** instead.

**Encephalitis – including meningoencephalitis:**

- Encephalitis is **infection or inflammation of the brain tissue.**
- Clinically, it may present with **fever, persistent altered mental status, new-onset seizures, and/or focal neurologic deficits** (e.g., aphasia, cranial nerve palsy, focal numbness, focal weakness, etc.).
- Diagnostically, it may be associated with a cerebrospinal fluid (CSF) pleocytosis and/or abnormal brain lesions on magnetic resonance imaging (MRI).
- Sometimes it may co-exist with infection or inflammation of the meninges (i.e., meningitis) resulting in a **meningoencephalitis.**

**Helpful Tip:**

Many patients can become confused or have altered mental status due to their fever or other systemic underlying medical conditions or medicines (e.g., liver or kidney disease, use of tacrolimus) without necessarily having encephalitis. Because of this, some consideration should be taken when classifying a patient with altered mental status as having encephalitis to **ensure there are some signs or evidence of infection or inflammation of the brain.**

**Other neuroinvasive presentation:**

Other neurologic/neuroinvasive presentations **not** covered under the categories of **AFP, GBS, encephalitis/meningoencephalitis, or meningitis.** Some examples of clinical presentations that could fit in the category of other neuroinvasive presentation include optic neuritis or Bell’s palsy.

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Last updated: 3/6/2020

*Updates only related to neurological classifications.*
Acute flaccid paralysis (AFP):

Clinical syndrome characterized by

- Rapid-onset (symptoms present over the course of hours to days from onset)
  AND

- Extremity, facial, and/or respiratory muscle weakness

- ‘Flaccid’* or decreased muscle tone in the affected areas (a description of “weakness” alone would NOT qualify as flaccid)

*‘Flaccid’ indicates the absence of spasticity or other signs of disordered central nervous system motor tracts such as hyperreflexia, clonus, or extensor plantar responses:

- In the medical records look for the Neuro exam and for a test for ‘Motor Exam’ and ‘Reflex Exam’. Note documentation of decreased tone muscle strength ($\leq 2^{**}$) and/or hyporeflexia (usually with a score of 1/4 or 1/5***), or areflexia (0/4 ot 0/5***).
- Medical records are required to identify flaccid paralysis. If a neuro exam has not been done then AFP cannot be confirmed. Patient interview, alone, does not confirm AFP.

AFP may result from diverse conditions affecting the lower motor neurons such as anterior (‘polio’) myelitis, neuromuscular junction disorders, or acute neuropathies (such as Guillain-Barré syndrome).

Other supportive documentation includes (not required): Asymmetry of weakness, CSF pleocytosis (WBC $> 5/mm^3$), and Spinal cord magnetic resonance imaging (MRI) documenting abnormal increased signal in the anterior gray matter.

**Muscle Strength**

0 No muscle activation
1 Trace muscle activation, such as a twitch, without achieving full range of motion
2 Muscle activation with gravity eliminated, achieving full range of motion
3 Muscle activation against gravity, full range of motion
4 Muscle activation against some resistance, full range of motion
5 Muscle activation against examiner’s full resistance, full range of motion

***Deep Tendon Reflexes***

0 Absent reflex
1+ Trace, or only seen with reinforcement
2+ Normal
3+ Brisk
4+ Nonsustained clonus
5+ Sustained clonus

Last updated: 3/6/2020

Updates only related to neurological classifications.
Guillain-Barré syndrome (GBS):

GBS is a clinical syndrome characterized by an acute immune-mediated attack on multiple peripheral nerves and/or nerve roots (i.e., an acute immune-mediated polyradiculoneuropathy). Clinically, it may present with an acute, bilateral, progressive, flaccid weakness of the extremities and/or cranial nerve muscles, and is usually accompanied by reduced or absent reflexes. **GBS can be one of many causes of acute flaccid paralysis (AFP).**

*Helpful Tip:* If the case is diagnosed as having GBS, then the clinical syndrome should be reported as GBS, not AFP.
West Nile/St. Louis Encephalitis
Virus Case Classification

Did the individual have clinically compatible symptoms?

- NO → NOT A CASE
- YES →
  
  Was the individual positive by culture, PCR, or NAT testing* OR have at least a four-fold rise in antibody titers between acute and convalescent specimens?

- NO →
  
  Was antibody testing positive or equivocal for IgM?

- YES
  
  Was the individual IgM positive for one virus and negative for the other?

- YES

  Was testing performed on CSF?

- YES →

  CONFIRMED for tested virus.

- NO →

  PROBABLE for positive virus

- NO/UNK →

  PROBABIL for virus with higher titer

- YES →

  PROBABLE CASE†: UNSPECIFIED FLAVIVIRUS GROUP 1

- NO/UNK →

  PROBABLE CASE†: UNSPECIFIED FLAVIVIRUS GROUP 1

*Note: If the case was identified via blood screening, the following DSO questions should be completed:

- “Diagnosis at presentation” should be set to ‘Viremic Blood Donor’

Under Risk Factor Assessment, Within 14 days of onset of symptoms, did the patient? Complete blood donation section as follows:

- “Donate Blood?” should be set to ‘Yes’
- “Identified by blood donor screening?” should be set to ‘Yes’
- “Date” should be filled out based on the Date of Collection reported by the blood bank.

† Due to the availability of the ‘Unspecified Flavivirus Group 1’ morbidity, the suspect case classification is no longer routinely used for arboviral cases.