Case Definition

Heartwater (*Ehrlichia ruminantium;* Cowdriosis)

(Notifiable) **November 2023**

1. **Disease Information**
   - **1.1 General Disease and Pathogen Information:** Heartwater (HW) is an acute, infectious, noncontagious, tick-borne, rickettsial disease of ruminants. The causative agent, *Ehrlichia ruminantium* (formerly *Cowdria ruminantium*), can be transmitted by many species of *Amblyomma* ticks. These three-host ticks also serve as the biological reservoir of HW. This disease primarily affects cattle, sheep, goats, and water buffalo; however, various wild ungulates can also be susceptible. Breeds non-native to endemic countries are more susceptible to infection. HW is endemic in most of Africa south of the Sahara Desert, Madagascar, and in some islands in the Caribbean. This disease is important to the United States due to the proximity of affected islands and the presence of *Amblyomma* ticks in North America that have been shown to be competent vectors for *E. ruminantium*.
   - **1.2 Clinical Signs:** *E. ruminantium* multiply in vascular endothelial cells sufficient to cause severe vascular damage. A hallmark of vascular damage is hydropericardium; hydrothorax, ascites, and pulmonary edema may accompany hydropericardium. Most cases of HW are characterized by fever, respiratory distress, diarrhea, and sometimes neurologic signs. Clinical signs typically manifest within 14 to 28 days after transmission of the disease agent by the tick vector. HW manifests in four forms of clinical disease, determined by host susceptibility and virulence of the *E. ruminantium* strains. Mild or subclinical infections may be seen in young animals, partially immune livestock, some indigenous breeds, and wild ruminants. Transient fever may be the only clinical sign in this form, which is known as “heartwater fever.” The peracute form is characterized by fever followed by respiratory distress, convulsions, occasional severe diarrhea, hyperesthesia, lacrimation, and sudden death. The acute form is the most common form and is characterized by a recurring fever, anorexia, depression, and neurological symptoms including chewing movements, twitching eyelids, protrusion of the tongue, and circling with a high-stepping gait. Acute cases lead to prostration and convulsions followed by death within 1 week of onset of clinical signs. The peracute and acute forms are the most likely to be seen with an introduction to the United States.
2. **Laboratory Criteria**
   - **2.1 Agent Isolation and Identification:** Laboratory diagnosis is based on observation of the organism in conjunction with molecular and culture methods. Blood and plasma can be used for culture. Molecular methods including polymerase chain reaction (PCR) and sequencing can be used for agent identification. The definitive diagnosis of HW is based on the observation of *E. ruminantium* in capillary endothelial cells of the brain.

Antimicrobial therapy may affect observation of the organism, but molecular methods may still be effective for diagnosis.

2.2 Agent Characterization: Further characterization is conducted by whole genome sequencing.

2.3 Serology: Serology is of limited use. Assays can cross-react with other *Ehrlichia* spp. and provide false positives. Serology should not be used to screen animals for importation into HW-free areas or as a diagnostic method for individual animals.

3. Case Classification

3.1 Suspect Case: A susceptible species with

3.1.1 Clinical signs consistent with HW; **OR**

3.1.2 Epidemiology consistent with HW.

3.2 Presumptive Positive Case: A suspect case that has a:

3.2.1 Pathognomonic finding on post-mortem examination; **OR**

3.2.2 Positive PCR result.

3.3 Confirmed Positive Case: An animal from which *E. ruminantium* has been isolated and identified at the National Veterinary Services Laboratories.

4. Reporting Criteria: HW is a U.S. foreign animal disease (FAD) that is reportable immediately under the APHIS National List of Reportable Animal Diseases (NLRAD).

4.1 NLRAD reporting in accordance with the NLRAD Standards for notifiable diseases; and by APHIS to the World Organisation for Animal Health (WOAH); **AND**

4.2 For FAD or Emerging Disease Incidents also follow standard procedures according to the Policy for the Investigation of Potential Foreign Animal Disease/Emerging.