

# Rare Disease of Public Health Significance

## 1. DISEASE REPORTING

### A. Purpose of Reporting and Surveillance

1. To understand the epidemiology of emerging and uncommon diseases in Washington State residents and to inform public health and health care about conditions that have been diagnosed in residents.
2. To assist in the diagnosis and treatment of cases.
3. If applicable, to identify potentially exposed close contacts, health care workers, and laboratory personnel and to provide counseling.
4. To identify sources of transmission and to prevent further transmission.
5. To raise the index of suspicion of a possible bioterrorism event if no natural exposure source is identified.

### B. Legal Reporting Requirements

1. Health care providers: *Burkholderia*, emerging condition with outbreak potential, novel influenza, monkeypox, SARS, smallpox, or viral hemorrhagic fever **immediately notifiable to local health jurisdiction**, influenza-associated death or varicella-associated death notifiable in 3 days, other rare diseases notifiable in 24 hours
2. Hospitals: *Burkholderia*, emerging condition with outbreak potential, novel influenza, monkeypox, SARS, smallpox virus, or viral hemorrhagic fever agents **immediately notifiable to local health jurisdiction**, influenza-associated death or varicella-associated death notifiable in 3 days, other rare diseases notifiable in 24 hours
3. Laboratories: *Burkholderia*, novel influenza virus, SARS-associated coronavirus, or smallpox **immediately notifiable to the local health jurisdiction**, other rare disease agents notifiable in 24 hours
4. Veterinarians: **Suspected human cases notifiable immediately to the local health jurisdiction**; animal cases of some conditions notifiable to Washington State Department of Agriculture (see: <http://apps.leg.wa.gov/WAC/default.aspx?cite=16-70>).
5. Local health jurisdictions: emerging condition with outbreak potential, novel influenza, SARS, smallpox or viral hemorrhagic fever **immediately notifiable to the Washington State Department of Health (DOH) Communicable Disease Epidemiology Section (CDES)**.

### C. Local Health Jurisdiction Investigation Responsibilities

1. Responsibilities are dependent on the disease under investigation.
2. Report all cases to CDES through the Public Health Issues Management System (PHIMS) as a Rare Disease of Public Health Significance.

## 2. THE DISEASES AND THEIR EPIDEMIOLOGY

According to the 2011 revision of WAC 246-101-010 “‘Other rare diseases of public health significance’ means a disease or condition, of general public health concern, which is occasionally or not ordinarily seen in the state of Washington including, but not limited to, viral hemorrhagic fevers, Rocky Mountain Spotted fever, and other tick borne diseases. This also includes a communicable disease that would be of general public concern if detected in Washington.” Included are conditions requested for immediate reporting nationally in accordance with International Health Regulations, which address reporting of any public health emergency of international concern (infectious, chemical, biological, or radiological). Exposures may be endemic or outside the state or country.

Conditions that should be reported through PHIMS to Communicable Disease Epidemiology Section (CDES) as Rare Diseases of Public Health Significance include:

- African tick bite fever ^
- African sleeping sickness ^
- Amoebic meningitis
- Anaplasmosis
- Babesiosis \*
- Burkholderia* infection (Meliodosis or Glanders)
- Chagas disease
- Coccidioidomycosis \*
- Cryptococcus gattii* \*  
see: <http://www.doh.wa.gov/notify/nc/cgattii.htm>
- Domoic acid poisoning \*  
see: <http://www.doh.wa.gov/notify/nc/sfpoison.htm>
- Ehrlichiosis ^
- Histoplasmosis ^
- Monkeypox
- Paragonimiasis ^
- Prion disease \*  
see: <http://www.doh.wa.gov/notify/nc/prion.htm>
- Severe acute respiratory syndrome-associated coronavirus disease (SARS)
- Smallpox
- Tickborne rickettsioses (including Rocky Mountain spotted fever) \*
- Tick paralysis \*
- Typhus
- Vaccinia transmission \*
- Varicella death \*
- Viral hemorrhagic fevers
- Highly antibiotic resistant organisms including vancomycin resistant *S. aureus*
- Any emerging condition with outbreak potential

\* Indicates condition endemic to the state recently identified in a Washington State resident

^ Indicates condition not endemic to the state recently identified in a Washington State resident

Additional conditions can be included in this category for the convenience of local health jurisdictions to document their work load and may be reported to CDES through PHIMS if desired. Other rare diseases entered in PHIMS as separate conditions include anthrax, arboviral encephalitis (e.g., dengue, western equine encephalitis), brucellosis, cholera, hantavirus pulmonary syndrome, influenza-associated deaths, novel influenza infections in humans, plague, polio, Q fever, rabies (human case), West Nile virus, and yellow fever. There is also separate reporting for the category Unexplained Critical Illness or Death. The Washington State Communicable Disease Report has a summary of cases: <http://www.doh.wa.gov/notify/forms/>.

### 3. CASE DEFINITIONS

National cases definitions for some Rare Diseases of Public Health Significance can be found at: [http://www.cdc.gov/ncphi/diss/nndss/casedef/case\\_definitions.htm](http://www.cdc.gov/ncphi/diss/nndss/casedef/case_definitions.htm).

### 4. DIAGNOSIS AND LABORATORY SERVICES

Appropriate diagnostic testing depends on the suspected agent. Commercial laboratory tests may be unreliable for many of these rare diseases so confirmation by a reference laboratory may be appropriate. See Section 7 for brief reviews of diagnostic testing for selected conditions. Consult with Communicable Disease Epidemiology Section (CDES) for assistance with diagnosis and testing.

Note that Washington State Public Health Laboratories (PHL) require all clinical specimens have two patient identifiers, a name **and** a second identifier (e.g., date of birth) both on the specimen label and on the submission form. Due to laboratory accreditation standards, specimens will be rejected for testing if not properly identified. Also include specimen source and collection date.

Consult with CDES regarding appropriate shipping temperature. Use the applicable PHL form.

Microbiology form: <http://www.doh.wa.gov/EHSPHL/PHL/Forms/Microbiology.pdf>

Parasitology form: <http://www.doh.wa.gov/EHSPHL/PHL/Forms/Microbiology.pdf>

Serology/Virology form: <http://www.doh.wa.gov/EHSPHL/PHL/Forms/SerVirHIV.pdf>

Food lab form: <http://www.doh.wa.gov/EHSPHL/PHL/forms/FoodBacteriology.pdf>

### 5. ROUTINE CASE INVESTIGATION

The case investigation depends on the suspected agent. In general, evaluate the diagnosis for a reported case including obtaining copies of laboratory reports. Call Communicable Disease Epidemiology Section (CDES) to arrange confirmatory testing. See Section 7 for brief descriptions of investigations for selected conditions. Consult with CDES for assistance with performing a public health investigation for other agents. The reporting form for Rare Disease of Public Health Significance is available at: <http://www.doh.wa.gov/notify/forms/>

### 6. CONTROLLING FURTHER SPREAD

Infection control measures depend on the suspected agent (see Section 7). Consult with Communicable Disease Epidemiology Section if needed.

## 7. MANAGING SPECIFIC DISEASES

Below are brief descriptions of select conditions endemic to the United States or of particular public health concern. Conditions with a national case definition have year of last revision listed: [http://www.cdc.gov/ncphi/diss/nndss/casedef/case\\_definitions.htm](http://www.cdc.gov/ncphi/diss/nndss/casedef/case_definitions.htm). There is indication whether Department of Health (DOH) can provide or arrange testing at CDC. Although any cases are reported as Rare Diseases electronically through Public Health Issues Management System (PHIMS) there are separate guidelines for *C. gattii*, domoic acid shellfish poisoning, and prion disease (see Section 2).

### A. Babesiosis

#### 1. Disease and its epidemiology:

- Agent is hemoprotzoan (inside red cells) parasite of genus *Babesia*, most commonly *B. microti* but also including *B. duncani* (WA1) which was first described from Washington State and *B. divergens*-like agents identified in the United States.
- Illness can be asymptomatic; symptoms include fever, chills, muscle aches, enlarged spleen and liver, and hemolytic anemia, with more severe illness in the elderly and immunocompromised
- Four locally acquired cases ever identified in Washington; 1 was transfusion associated and 3 had presumed tick exposure
- Reservoir is rodents and hard ticks
- Transmission is through bites of infected ticks, rarely person-to-person only by transfusion (usually from an asymptomatic donor)
- Incubation period is variable, probably 1-8 weeks
- Communicability is only through transfusion
- Treatment is with appropriate antibiotics

#### 2. Case definition (2011: [http://www.cdc.gov/ncphi/diss/nndss/casedef/babesiosis\\_current.htm](http://www.cdc.gov/ncphi/diss/nndss/casedef/babesiosis_current.htm))

*Suspect*: case with laboratory detection or supportive laboratory but insufficient clinical or epidemiologic information for *Probable* or *Confirmed* classification

*Probable*: case with one or more of fever, anemia, or thrombocytopenia AND supportive serology

OR

blood donor or blood recipient: with epi link to confirmed case with confirmatory laboratory detection (below) but not meeting clinical criteria OR with confirming serology but without fever, anemia, and thrombocytopenia

*Confirmed*: a case with one or more of fever, anemia, thrombocytopenia, chills, sweats, headache, myalgia, or arthralgia AND laboratory detection by: parasites in blood smears, PCR, nucleic acid amplification, or isolation in animal inoculation

#### 3. Diagnosis and laboratory services: DOH can confirm the agent or arrange testing.

Submit blood smears, whole blood in EDTA (purple top) tube, and serum in separation or

clot tube with parasitology form (Section 7 Q).

4. Routine case investigation: Identify travel exposures, tick bites, and exposure to tick habitats with emphasis on locally acquired infection. Identify blood transfusions and test all blood donors or recipients associated with the case.
5. Controlling further spread: No isolation or restrictions apply. Case should defer donating blood. Educate those sharing a case's exposure about signs and symptoms of babesiosis.
6. Routine prevention: When in risk areas wear long pants and a long-sleeved shirt, use tick repellent when necessary, check for and remove ticks, and monitor for symptoms.
7. Resources: Babesiosis - <http://www.dpd.cdc.gov/dpdx/HTML/Babesiosis.htm>  
Tick removal - [http://www.cdc.gov/ncidod/dvbid/lyme/ld\\_tickremoval.htm](http://www.cdc.gov/ncidod/dvbid/lyme/ld_tickremoval.htm)

### **B. *Burkholderia mallei* (Glanders) and *B. pseudomallei* (melioidosis)**

#### 1. Diseases and their epidemiology:

- Glanders:
  - i. Agent is gram-negative rod *Burkholderia mallei*; potential agent of bioterrorism
  - ii. Illness in humans generally includes malaise, fever, chills, fatigue, multiple skin nodules, regional lymphadenopathy. Inhalational exposure can result in pneumonia, pleuritic chest pain, cervical adenopathy, pulmonary abscesses or pleural effusions. Infections can cause septicemia.
  - iii. Primarily disease of horses, mules, and donkeys. Not found in environment; does not persist in water, soil, or plants. Rare human cases seen in Asia, Africa, the Middle East, and South America; none in U.S. since 1940s.
  - iv. Human cases are generally via occupational exposure. Transmission is by contact of open skin wounds or mucous membranes with infected animals, rarely by ingesting contaminated meat or inhaling respiratory secretions.
  - v. Incubation period ranges 1 to 21 days (usual 10-14), shorter with high inoculum
  - vi. Communicability can occur via respiratory and cutaneous secretions; rare
  - vii. Treatment is with long courses of appropriate antibiotics
- Melioidosis (Whitmore's disease):
  - i. Agent is soil/water saprophyte *Burkholderia pseudomallei*; potential agent of bioterrorism
  - ii. Illness ranges from none to bronchitis, pneumonia, cutaneous or visceral abscesses (e.g. empyema, osteomyelitis, meningoenzephalitis), septicemia
  - iii. Reservoir is soil and water, particularly in tropics and subtropics
  - iv. Transmission is through wound infection, dust inhalation, water aspiration, contact with rodents, direct or sexual contact with a case patient, breast feeding, bloodborne, or intentional distribution
  - v. Incubation period generally 1 to 21 days, shorter with high inoculum

- vi. Communicability of uncertain duration through blood, body fluids
  - vii. Treatment is with long courses of appropriate antibiotics
2. Case definition
    - Confirmed*: case with laboratory detection
  3. Diagnosis and laboratory services: DOH will forward specimens to CDC to confirm the agent. Submit culture with a microbiology form (Section 7 Q).
  4. Routine case investigation: notify Communicable Disease Epidemiology Section immediately for suspected or confirmed case; obtain appropriate specimens as soon as possible for testing including serum, tissues slides, or culture (consult with Communicable Disease Epidemiology first) with virology/serology form (Section 7 Q).
  5. Controlling further spread: contact precautions in medical settings. Educate those sharing a case's exposure about signs and symptoms of glanders and melioidosis
  6. Routine prevention: when in risk areas for melioidosis avoid soil and water contact, particularly for those with open skin wounds or chronic medical condition (diabetes, renal disease)
  7. Resources:
    - a. <http://www.cdc.gov/nczved/divisions/dfbmd/diseases/melioidosis/>
    - b. <http://www.cdc.gov/nczved/divisions/dfbmd/diseases/glanders/>

### C. Coccidioidomycosis

1. Disease and its epidemiology:
  - Agent is *Coccidioides immitis*, a soil fungus infecting humans and other animals
  - Illness is typically asymptomatic; symptoms include fever, cough, chest pain, erythema nodosum or erythema multiforme, and muscle or joint aches with rare complications such as meningitis or bone infection particularly in persons with immunocompromise or in women who are pregnant; rare cases in Washington following travel, possibly even rare endemic cases
  - Reservoir is soil in dry areas of southwestern United States and parts of Mexico
  - Transmission is by inhaling dust or disturbed soil (e.g., with construction, farming, field training, digging, dust storms, or earthquakes) or in laboratory settings where *Coccidioides* cultures are handled
  - Incubation period is 1-3 weeks
  - Communicability: N/A
  - Treatment is with appropriate anti-fungal medications if necessary
2. Case definition (2011: [http://www.cdc.gov/ncphi/diss/mndss/casedef/coccidioid\\_current.htm](http://www.cdc.gov/ncphi/diss/mndss/casedef/coccidioid_current.htm))
  - Confirmed*: case with clinically compatible illness with laboratory confirmation by culture, histopathology, molecular test, serology, or coccidioidal skin-test conversion

3. Diagnosis and laboratory services: DOH can arrange serology, microscopy, and confirmation of culture. Submit serum, tissues slides, or culture (consult with Communicable Disease Epidemiology first) with virology/serology form (Section 7 Q).
4. Routine case investigation: identify others potentially exposed at the same time as the case; identify and evaluate laboratory personnel handling a *Coccidioides* culture
5. Controlling further spread: No isolation or restrictions apply.
6. Routine prevention: Avoid dusty environments in risk areas.
7. Resources: <http://www.vfce.arizona.edu/>

#### D. Ehrlichiosis and Anaplasmosis

1. Diseases and their epidemiology:
  - Agents are obligate intracellular bacteria including in North American *Ehrlichia chaffeensis* causing human ehrlichiosis, *Ehrlichia ewingii* infection (formerly human monocytic ehrlichiosis) and *Anaplasma phagocytophilum* infection (formerly human granulocytic ehrlichiosis or human granulocytic anaplasmosis)
  - Illness typically involves acute onset of fever accompanied by chills, headache, myalgia, malaise, anemia, leucopenia, thrombocytopenia, altered liver function, meningoencephalitis, vomiting, or rash. Travel-acquired cases in Washington residents were reported in 2004 and 2007.
  - Reservoirs of *Ehrlichia* are dogs and white-tailed deer; and for *Anaplasma* are wild rodents, ruminants, and cervids
  - Transmission is by ticks including *Ixodes* species (*Anaplasma*) and *Amblyomma americanum* (*Ehrlichia chaffeensis* and *E. ewingii*)
  - Incubation period is 5-10 days (ehrlichiosis), 5-21 days (anaplasmosis)
  - Communicability: N/A
  - Treatment is with appropriate antibiotics
2. Case definition (2008: [http://www.cdc.gov/ncphi/disss/nndss/casedef/ehrlichiosis\\_2008.htm](http://www.cdc.gov/ncphi/disss/nndss/casedef/ehrlichiosis_2008.htm))
  - Suspect*: case with laboratory evidence of past or present infection with undetermined ehrlichiosis or anaplasmosis but no clinical information available
  - Probable*: clinically compatible case with: evidence for *E. chaffeensis* infection by single serology positive or microscopic identification of morulae; OR evidence for *A. phagocytophilum* infection by single serology positive or microscopic identification of morulae; OR laboratory evidence of past or present infection with undetermined ehrlichiosis or anaplasmosis
  - Confirmed*: clinically compatible case with: evidence for *E. chaffeensis* infection by fourfold rise in serology, culture, PCR, or immunohistochemistry; OR evidence for *E. ewingii* infection by PCR; OR evidence for *A. phagocytophilum* infection by fourfold rise in serology, culture, PCR, or immunohistochemistry
3. Diagnosis and laboratory services: DOH can arrange testing. Submit blood and tissue

samples with appropriate serology and/or microbiology forms (Section 7 Q).

4. Routine case investigation: ask about travel to endemic areas, potential tick habitats, and tick bites.
5. Controlling further spread: Educate those sharing a case's exposure about signs and symptoms of spotted fever rickettsiosis
6. Routine prevention: When in risk areas wear long pants and a long-sleeved shirt, use tick repellent when necessary, check for and remove ticks, and monitor for symptoms
7. Resources: <http://www.cdc.gov/ticks/diseases/ehrlichiosis/> and <http://www.cdc.gov/ticks/diseases/anaplasmosis/index.html>

## E. Monkeypox

1. Disease and its epidemiology:
  - Agent is monkeypox virus in the genus *Orthopoxvirus*
  - Illness is typically fever, headache, muscle aches, backache, and lymphadenopathy followed by vesicular-pustular rash like smallpox; cases in Africa up to 10% fatal
  - Reservoir is presumed primates and squirrels in central and western Africa
  - Transmission is from an infected animal by bites or contact with body or rash fluids
  - Incubation period is about 12 days
  - Communicability: person-to-person spread can occur through respiratory droplets or body fluids
  - Treatment is supportive
2. Case definition (2004: <http://www.cdc.gov/ncidod/monkeypox/casedefinition.htm>)
  - Suspect*: case with exposure (to wild animal or exotic pet) AND fever or unexplained rash AND two or more consistent signs or symptoms within 21 days of last exposure
  - Probable*: case with exposure (to wild animal or exotic pet) AND fever AND either vesicular-pustular rash within 21 days of last exposure or rash not described but IgM elevated
  - Confirmed*: case with viral isolation, PCR, electron microscopy, or immunohistochemistry
3. Laboratory and diagnostic services: DOH can arrange serology, microscopy, and confirmation of culture. Submit serum, tissues slides, or culture (consult with Communicable Disease Epidemiology first) with virology/serology form (Section 7 Q).
4. Routine case investigation: notify Communicable Disease Epidemiology Section immediately for suspected or confirmed case; obtain appropriate specimens as soon as possible for testing including
5. Controlling further spread: contact and droplet precautions, post-exposure smallpox vaccine for those providing direct patient care; notify Environmental Health Program for animal containment issues

6. Routine prevention: animal importation regulations
7. Resources: <http://www.cdc.gov/ncidod/monkeypox/index.htm>

## G. SARS (Severe Acute Respiratory Syndrome)

1. Disease and its epidemiology:
  - Agent is SARS-associated coronavirus responsible for a 2003 outbreak with 8098 cases primarily in Asia but including eight travel-associated cases in the United States
  - Illness is fever > 38.0°C (100.4°F) and flu-like symptoms, sometimes diarrhea, followed by cough and pneumonia, about 10% mortality; no cases in Washington
  - Reservoir is presumed to be wild mammals
  - Transmission is by respiratory droplets or presumably close contact with an infected animal; transmission has occurred in health care settings
  - Incubation period is 2-10 days
  - Communicability is high for certain patients (e.g., transmission among guests at a hotel or to other patients in an emergency department)
  - Treatment is supportive
2. Case definition (2003: <http://www.cdc.gov/ncphi/diss/nndss/casedef/sarscurrent.htm>)
  - Probable:* case with temperature >100.4° F (>38° C) AND pneumonia by x-ray or ARDS or consistent autopsy AND close contact with a confirmed case
  - Confirmed:* case with fever and any consistent respiratory symptom AND antibody detection, isolation, or detection by PCR
3. Diagnosis and laboratory services: DOH can arrange testing. Submit multiple specimens including respiratory (nasopharyngeal wash, nasopharyngeal swab, oropharyngeal swab, bronchoalveolar lavage, tracheal aspirate, pleural fluid, sputum), stool, serum, plasma, and lung tissue with a virology/serology form (Section 7 Q).
4. Routine case investigation: notify Communicable Disease Epidemiology Section immediately for suspected or confirmed case; obtain appropriate specimens as soon as possible including serum as well as lung tissue or respiratory secretions (recommend using appropriate personal protection while obtaining respiratory specimens); obtain travel history and identify risk exposures; identify those sharing exposure with case and those exposed to case; institute isolation and quarantine measures as appropriate
5. Controlling further spread: airborne precautions in health care settings, droplet precautions in home settings; consider quarantine for exposed persons
6. Routine prevention: hand and respiratory hygiene, precautions during travel to risk areas with potential exposure in live animal markets
7. Resources: <http://www.cdc.gov/ncidod/sars/>

## H. Smallpox

1. Disease and its epidemiology:

- Agent is variola virus, considered extinct in nature
  - Illness begins as febrile flu-like illness followed by rash progressing through stages of macules, papules, vesicles, pustules, and scabs; no naturally-occurring cases worldwide since 1977
  - Reservoir was humans, now only laboratory specimens exist
  - Transmission is through respiratory droplets and fomites or through deliberate release of weaponized material; scabs contain virus and are infectious even when dried
  - Incubation period is 7-19 days
  - Communicability is high through respiratory secretions while lesions are present
  - Treatment is supportive; antivirals may be considered
2. Case definition (2004: <http://www.cdc.gov/ncphi/diss/nmdss/casedef/smallpoxcurrent.htm> )
- Suspect:* case with fever followed in 1-4 days by generalized, acute vesicular or pustular rash
- Probable:* case with acute onset of fever  $\geq 101^{\circ}\text{F}$  ( $\geq 38.3^{\circ}\text{C}$ ) followed by a rash characterized by firm, deep seated vesicles or pustules in the same stage of development without other apparent cause OR clinically consistent case with epi link to a confirmed case
- Confirmed:* laboratory confirmed case OR case with acute onset of fever  $\geq 101^{\circ}\text{F}$  ( $\geq 38.3^{\circ}\text{C}$ ) followed by a rash characterized by firm, deep seated vesicles or pustules in the same stage of development without other apparent cause with epi link to a confirmed case
3. Diagnosis and laboratory services: DOH can arrange testing; submit vesicle, scab, skin, and serum specimens with virology/serology form (Section 7 Q).
4. Routine case investigation: notify Communicable Disease Epidemiology Section immediately for suspected or confirmed case; evaluate the diagnosis particularly if lesions are vesicles or pustules that are deep-seated firm well-circumscribed and at the same stage of development. Recommend appropriate personal protective equipment when obtaining specimens. Submit appropriate specimens including sterile collection of skin of vesicle, scraping of base of vesicle touched on glass slides, EM grid touched to based of lesion, scabs, full thickness skin punch biopsies, and 10 ml serum with the virology/serology form (Section 7 Q).
5. Controlling further spread: strict contact and airborne precautions in health care setting; consider quarantine for exposed persons
6. Routine prevention: no routine vaccination
7. Resources: <http://emergency.cdc.gov/agent/smallpox/index.asp>

### I. Spotted fever rickettsioses

1. Disease and its epidemiology:
  - Agent is tick-borne bacterium *Rickettsia rickettsii* for Rocky Mountain spotted fever

(RMSF); various other rickettsial species causes illnesses elsewhere

- Illness for RMSF begins with sudden fever, chills, severe headache, muscle and joint pain, and sometimes gastrointestinal symptoms followed in 80% of cases by a macular rash which may become petechial, first on wrists and ankles and then spreading; there can be delirium, meningoencephalitis, or death. Typically 0-1 cases are reported annually in Washington. An eschar at the site of tick attachment has been reported for some other spotted fever rickettsioses.
- Primary vectors for RMSF are the Rocky Mountain wood tick (*Dermacentor andersoni*) and the American dog tick (*D. variabilis*). Animal hosts are dogs, opossums, wild rabbits, and wild rodents with clinical illness in dogs and some rodents. Most cases occur April through September when ticks are active.
- Transmission for RMSF is by tick saliva, usually requiring 4-6 hours to attach and feed. Exposures in wooded or high grass areas or to dogs may increase the risk.
- Incubation period for RMSF is 2-14 days
- Communicability: N/A
- Treatment for RMSF is with tetracyclines (usually doxycycline). The case-fatality rate is 13-25% if untreated and 4% even with appropriate antibiotic treatment.

2. Case definition for RMSF (2010):

[http://www.cdc.gov/nceh/diseases/diagnosis/casedef/spottedfever\\_current.htm](http://www.cdc.gov/nceh/diseases/diagnosis/casedef/spottedfever_current.htm)

*Suspect*: case with laboratory evidence of past or present infection but no clinical information available

*Probable*: fever AND one or more of rash, headache, myalgia, anemia, any hepatic transaminase elevation, or thrombocytopenia AND presumptive laboratory of elevated IgG or IgM by IFA, ELISA, dot-ELISA, or latex agglutination

*Confirmed*: fever AND confirmatory laboratory including fourfold risk in IgG tested by IFA, PCR, immunohistochemistry, or culture

3. Diagnosis and laboratory services: DOH can arrange testing. Submit blood and tissue samples with appropriate serology and/or microbiology forms (Section 7 Q).
4. Routine case investigation: ask about travel to endemic areas, potential tick habitats, and tick bites.
5. Controlling further spread: Educate those sharing a case's exposure about signs and symptoms of spotted fever rickettsiosis
6. Routine prevention: When in risk areas wear long pants and a long-sleeved shirt, use tick repellent when necessary, check for and remove ticks, and monitor for symptoms
7. Resources:

[http://www.cdc.gov/ticks/diseases/rocky\\_mountain\\_spotted\\_fever/index.html](http://www.cdc.gov/ticks/diseases/rocky_mountain_spotted_fever/index.html)

## J. Tick paralysis

1. Disease and its epidemiology:

- Agent is a neurotoxin secreted in the saliva of certain ticks.
  - Illness is an acute, ascending, flaccid paralysis. There may be fatigue, muscle aches, numbness in the legs, and in children flu-like symptoms. Paralysis may affect breathing muscles and cause respiratory failure. About 10% of unrecognized tick paralysis cases are fatal. Cases are rare in Washington, most commonly during spring months in girls (with long hair) under 10 years old.
  - Reservoirs in this country are *Dermacentor andersoni* (Rocky Mountain wood tick) in northwestern states, and *D. variabilis* (American dog tick) in southeastern states.
  - Transmission is through an attached tick releasing saliva
  - Incubation period is typically 4-7 days while tick feeds
  - Communicability: N/A
  - Treatment: Prompt removal of the feeding tick usually results in complete recovery within 24 hours. It is important to remove all the mouthparts, which contain the salivary glands. Oxygen therapy or mechanical ventilation may be needed.
2. Case definition:
- Confirmed:* Symptoms consistent with illness and rapid improvement of the patient upon removal of tick.
3. Diagnosis and laboratory services: none
4. Routine case investigation: Inquire about possible exposure to ticks. Carefully check patient for ticks, especially along the hairline.
5. Controlling further spread: N/A
6. Routine prevention: When in risk areas wear long pants and a long-sleeved shirt, use tick repellent when necessary, check for and remove ticks, and monitor for symptoms. Check potentially exposed persons (especially along hair line) and promptly remove any ticks.
7. Resources: <http://www.cdc.gov/mmwr/preview/mmwrhtml/00040975.htm>

## K. Typhus

1. Disease and its epidemiology:
- Agents are *Rickettsia typhi* or *R. felis* (fleaborne – endemic or murine typhus) and *R. prowazekii* (louseborne – epidemic typhus)
  - Illness is febrile rash illness for louseborne with case fatality rate up to 40% if untreated, milder for fleaborne. Washington's last reported case was in 1992 following travel to Asia.
  - Reservoirs are rats for fleaborne (reported from tropics and subtropics), humans for louseborne (Andes region of South America, Burundi, Ethiopia), and rarely flying squirrels in eastern United States
  - Transmission is by bite of a flea or louse feces entering a wound
  - Incubation period is 7 to 14 days

- Communicability for louseborne is through the human lice
  - Treatment is with doxycycline for both and for louseborne also a pediculocide
2. Case definition
    - Probable:* Clinically compatible illness with single antibody titer
    - Confirmed:* Clinically compatible illness with confirmatory laboratory including fourfold antibody rise, PCR positive, or positive immunohistochemical stain
  3. Diagnosis and laboratory services: DOH can arrange testing. Submit serum and tissue samples with a microbiology form (Section 7 Q).
  4. Routine case investigation: notify Communicable Disease Epidemiology Section immediately for suspected or confirmed case; obtain appropriate specimens as soon as possible for testing including
  5. Controlling further spread: delouse a louse-infested patient, educate those sharing a case's exposure about signs and symptoms of typhus
  6. Routine prevention: keep rats away from human habitation

#### L. Vaccinia transmission

1. Disease and its epidemiology:
  - Agent is vaccinia (smallpox vaccine) virus
  - Illness involves pustules on the exposed skin or rare complications of smallpox vaccination such as ocular infection or eczema vaccinatum; rare cases in Washington associated with vaccination of military personnel
  - Reservoir is vaccine, in this country currently indicated only for selected military personnel and laboratory workers
  - Transmission is through skin contact or other close contact (sexual contact, breast feeding, sports partner, shared clothing); tertiary transmission has occurred
  - Incubation period is undefined but probably several days
  - Communicability is for duration of lesions; shed scabs may contain viable virus
  - Treatment is supportive; vaccinia immune globulin, cidofovir, and investigational drugs may be needed for severe infections
2. Case definition
  - Confirmed:* laboratory confirmed vaccinia infection in a person not receiving vaccinia immunization
3. Diagnosis and laboratory services: DOH can arrange PCR and culture to confirm vaccinia; serology confirms development of immunity. Submit serum, blood and lesion material with virology/serology form (Section 7 Q).
4. Routine case investigation: immediately notify Communicable Disease Epidemiology Section for suspected or confirmed case; evaluate the diagnosis particularly if lesions are vesicles or pustules that are deep-seated firm well-circumscribed and at the same stage of

development. Recommend appropriate personal protective equipment when obtaining specimens. Submit serum, blood, and lesion material with virology/serology form (Section 7 Q). Identify close contacts likely to have received vaccinia vaccine and persons potentially exposed to the case patient

5. Controlling further spread: strict contact precautions particularly when around unvaccinated persons, educate those sharing a case's exposure about signs and symptoms of vaccinia infection
6. Routine prevention: cover vaccination site, properly dispose of bandages and scabs
7. Resources: <http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5619a4.htm>

#### M. Varicella zoster virus-associated death

1. Disease and its epidemiology:
  - Agent is varicella zoster virus (VZV)
  - Illness with chickenpox is usually mild but can result in fatal complications such as pneumonia, secondary bacterial infection, hemorrhagic complications, or encephalitis; highest risk in neonates (30% mortality for chickenpox infections) and children (0.01% mortality for chickenpox infections); rare cases in Washington
  - Reservoir is humans
  - Transmission is person to person through respiratory droplets and discharge from lesions. Dry scabs may contain viral DNA but not viable virus.
  - Incubation period is 2-3 weeks
  - Communicability is high from 5 days before rash onset until all lesions have crusted
  - Treatment is with acyclovir for severe infection
2. Case definition (1998:  
[http://www.cdc.gov/ncphi/disss/nndss/casedef/varicella\\_deaths\\_current.htm](http://www.cdc.gov/ncphi/disss/nndss/casedef/varicella_deaths_current.htm))
  - Probable:* a probable case of varicella which contributes directly or indirectly to acute medical complications which result in death
  - Confirmed:* a confirmed case of varicella which contributes directly or indirectly to acute medical complications which result in death(For varicella case definition see:  
[http://www.cdc.gov/ncphi/disss/nndss/casedef/varicella\\_current.htm](http://www.cdc.gov/ncphi/disss/nndss/casedef/varicella_current.htm))
3. Diagnosis and laboratory services: DOH can perform viral culture and PCR for VZV on vesicular fluid and scabs from lesions. DOH can arrange testing of serum and tissue specimens from autopsy (if available). For further information regarding laboratory diagnosis see: <http://www.cdc.gov/vaccines/pubs/surv-manual/chpt22-lab-support.htm>
4. Routine case investigation: Review the clinical presentation, physical exam findings, immunization status and exposure history. Recommend that only immune personnel obtain specimens. Submit appropriate specimens (fluid from vesicles obtained using a Dacron swab rubbed on opened lesion, scabs, serum, and tissue from autopsy specimens),

with the virology/serology form (Section 7 Q).

5. Controlling further spread: airborne precautions in health care settings; only personnel with documented immunity should enter the room of the patient or have contact during autopsy. Give post-exposure prophylaxis with VariZIG to high risk exposed contacts (neonates, pregnant women, immunocompromised persons).
6. Routine prevention: universal childhood chickenpox vaccination; shingles vaccine as indicated
7. Resources:  
<http://www.cdc.gov/vaccines/pubs/surv-manual/chpt17-varicella.htm>  
[http://www.cdc.gov/ncidod/diseases/list\\_varicel.htm](http://www.cdc.gov/ncidod/diseases/list_varicel.htm)

## N. Viral hemorrhagic fevers

1. Disease and its epidemiology:
  - Agents are multiple unrelated viruses including arenaviruses (Lassa fever, Argentine hemorrhagic fever, Bolivian hemorrhagic fever, Venezuelan hemorrhagic fever, Brazilian hemorrhagic fever, Lujo virus), filoviruses (Ebola, Marburg), bunyaviruses (Crimean-Congo hemorrhagic fever, Korean hemorrhagic fever), and flaviviruses (Omsk hemorrhagic fever)
  - Illness is severe and affects multiple organ systems with typical symptoms of fever, muscle pain, rash, vomiting, diarrhea, abdominal pain, hemorrhage, thrombocytopenia, organ failure, and shock; with arenaviruses may be pharyngitis, retrosternal chest pain, or proteinuria; no cases reported in Washington
  - Reservoirs when known are various animal (mainly rodent) or insect hosts, each occurring in a limited geographic area
  - Transmission varies with agent; may be through mosquito or tick bites, contact with rodent excretions or ill livestock, contact with body secretions (blood, semen), or bloodborne (e.g., shared syringe); laboratory workers may be at risk from aerosols
  - Incubation period varies with agent, probably a few days to a few weeks
  - Communicability requires close contact, such as with family members or health care providers
  - Treatment is supportive; ribavirin and convalescent-phase plasma have been used to treat some viruses
2. Case definition (2011):  
<http://www.cdc.gov/ncphi/disss/nndss/casedef/virahemorrhagicfever.htm>
  - Suspected:* fever > 40°C AND one or more consistent clinical findings AND epi link by implicated exposure
  - Confirmed:* fever > 40°C AND one or more consistent clinical findings AND detection of VHF virus (ELISA< isolation, PCR, immunohistochemistry)
3. Diagnosis and laboratory services: DOH can arrange testing of serum, fixed tissue (e.g., lung, kidney, spleen, lymph nodes, etc.), and fresh tissue (typically lung, bone marrow).

Submit specimens with appropriate serology and/or microbiology forms (Section 7 Q).

4. Routine case investigation: notify Communicable Disease Epidemiology Section immediately for suspected or confirmed case; obtain travel history and identify risk exposures, those sharing exposure with case, and those exposed to case
5. Controlling further spread: strict isolation with disinfection of blood and body secretions, patient education because secretions and urine may be viremic for weeks (e.g., semen up to 3 months for Lassa fever), safe handling of bodies, strict laboratory safety, consider quarantine for exposed contacts
6. Routine prevention: varies, may include rodent and mosquito control as well as infection control in health care settings
7. Resources: <http://www.cdc.gov/ncidod/diseases/virlfvr/virlfvr.htm>

#### **O. Highly antibiotic resistant organisms including *Staphylococcus aureus*, vancomycin resistant (VRSA)**

1. Disease and its epidemiology:
  - Agent is organism with atypical resistance pattern e.g., VRSA is *S. aureus* with resistance to vancomycin ( $MIC \geq 16 \mu\text{g/ml}$ )
  - Illness is typical for the bacterial species; one vancomycin intermediate case reported in Washington in 2008
  - Reservoirs vary; for VRSA is humans
  - Transmission varies; for VRSA person to person
  - Incubation period varies; for VRSA is indeterminate
  - Communicability varies; for VRSA is for duration of respiratory and skin infections
  - Treatment varies; for VRSA is with appropriate antiviral drugs and antibiotics
2. Case definition for VRSA (2007:  
<http://www.cdc.gov/ncphi/diss/nndss/casedef/vancomycincurrent.htm>)

*Confirmed:* laboratory-confirmed resistant *S. aureus* ( $MIC \geq 16 \mu\text{g/ml}$  for VRSA).
3. Diagnosis and laboratory services for VRSA: DOH can confirm vancomycin resistance. Submit isolate with microbiology form (Section 7 Q).
4. Routine case investigation varies: in general, obtain isolate for future strain comparison, identify likely nosocomial transmission, identify persons likely to have organism transmitted to or acquired from case patient and obtain cultures
5. Controlling further spread: contact or respiratory precautions as indicated
6. Routine prevention: routine hand or respiratory hygiene as indicated
7. Resources:  
VRSA: [http://www.cdc.gov/HAI/organisms/visa\\_vrsa/visa\\_vrsa.html](http://www.cdc.gov/HAI/organisms/visa_vrsa/visa_vrsa.html)

### P. Emerging condition with outbreak potential

An emerging condition is one whose incidence in humans has recently increased or threatens to increase in the near future. Of particular concern are conditions of high severity with potential person-to-person spread. Public health seeks to rapidly detect such conditions, control their spread, and identify risk factors for acquisition.

Newly emerging conditions have included SARS and 2009 H1N1. Until the condition is identified it cannot be specified for notifiable conditions reporting. Similarly, the transmission, laboratory testing, case definition, and control and prevention measures will have to be determined after the condition is identified. Initial healthcare provider judgment is necessary to recognize and report an unusual condition.

*Suspect:* a newly identified condition with potential for person-to-person transmission

### Q. Specimen Submission Forms for Washington State Public Health Laboratories

Food lab form: <http://www.doh.wa.gov/EHSPHL/PHL/forms/FoodBacteriology.pdf>

Microbiology form: <http://www.doh.wa.gov/EHSPHL/PHL/Forms/Microbiology.pdf>

Parasitology form: <http://www.doh.wa.gov/EHSPHL/PHL/Forms/Microbiology.pdf>; check “Parasitology” box

Serology/Virology form: <http://www.doh.wa.gov/EHSPHL/PHL/Forms/SerVirHIV.pdf>

Note that Washington State Public Health Laboratories (PHL) require all clinical specimens have two patient identifiers, a name **and** a second identifier (e.g., date of birth) both on the specimen label and on the submission form. Due to laboratory accreditation standards, specimens will be rejected for testing if not properly identified. Also include specimen source and collection date.

## 8. ROUTINE PREVENTION

Routine prevention measures depend on the suspected agent. See Section 7 for comments about selected conditions. Consult with Communicable Disease Epidemiology Section for any other conditions.

## ACKNOWLEDGEMENTS

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## UPDATES

September 2008: The definition of “rare diseases of public health significance” was made consistent with the definition provided in WAC 246-101-010.

January 2011: Section 7 includes expanded descriptions of certain rare diseases are included. Reporting requirements were revised to reflect the 2011 Notifiable Conditions Rule revision.