

Unexplained Critical Illness or Death

1. DISEASE REPORTING

A. Purpose of Reporting and Surveillance

1. To identify emerging pathogens in Washington State.
2. To raise the index of suspicion of a possible bioterrorism event.
3. To recognize critical illnesses or deaths with potential public health impact.

Note that public health resources cannot support the work to diagnose all cases of unexplained critical illness or death.

B. Legal Reporting Requirements

1. Health care providers: notifiable to local health jurisdiction within 24 hours
2. Health care facilities: notifiable to local health jurisdiction within 24 hours
3. Laboratories: no requirements for reporting
4. Local health jurisdictions: notifiable to the Washington State Department of Health (DOH) Communicable Disease Epidemiology (CDE) within 7 days of case investigation completion or summary information required within 21 days

C. Local Health Jurisdiction Investigation Responsibilities

1. For sporadic cases: Review the clinical presentation, laboratory findings, and autopsy report if available. Secure appropriate specimens (if available) for immediate or future testing if warranted.
2. For clusters of patients with a similar illness: Review the clinical presentations, laboratory findings, and autopsy reports (if available) for all of the patients. Consult with CDE to determine if testing at the Washington State Public Health Laboratories (PHL) is appropriate. If so, facilitate the transport of appropriate specimens to PHL for testing.
3. Report all persons who meet the clinical case definition to CDE. Complete the case report form (<http://www.doh.wa.gov/Portals/1/Documents/5100/210-051-ReportForm-Unexplained.pdf>) and enter the data into the Public Health Issues Management System (PHIMS) as Unexplained Critical Illness or Death. If a cause of death is determined to be a notifiable condition, transfer the case in PHIMS to the reportable condition.

2. THE DISEASES AND THEIR EPIDEMIOLOGY

Background

The purpose of this surveillance system is to identify emerging pathogens and detect bioterrorism or other events of public health significance. Whether a diagnosis is established may reflect the intensity of clinical investigation and testing. Particularly with deaths outside a health care facility, basic microbial testing may not have been done.

Public health resources cannot support investigation of all such cases, particularly when few diagnostic tests were done on the patient initially. Clusters of unexplained severe illnesses and single cases of unusual syndromes are of greatest public health priority.

As examples of emerging conditions, in 1993, two individuals from New Mexico with critical illness presented to the same physician. Upon the initiation of an investigation of these and subsequent cases, hantavirus infection was identified and the reservoir was determined within 6 weeks of the first case presentation. In 1999, a cluster of encephalitis cases in New York City was found to be caused by West Nile virus, the first time the agent had been recognized on the continent.

Unexplained Critical Illness and Death in Washington

Requirements for the reporting of unexplained critical illness or death were instituted in December of 2000. To maximize the likelihood of identifying cases with public health importance, reporting is limited to persons aged 1–49 years who were previously healthy and who have evidence of an infectious process. Since the condition was made reportable, DOH has received 0–6 reports annually. The most common clinical syndromes reported are central nervous system (meningitis, encephalitis), respiratory, and sepsis/multiorgan failure.

Etiologies

Potential etiologies of unexplained critical illness and/or death are extensive and will not be addressed in a comprehensive way in this guideline. Below is a list of syndromes followed by infectious disease diagnoses which should be considered of highest public health significance.

Respiratory syndrome: anthrax, plague, tularemia, tuberculosis, pertussis, influenza, hantavirus, cryptococcal disease. (Multiple other potential causes that can be difficult to diagnose include: legionellosis, Mycoplasma infection, psittacosis, coccidioidomycosis.)

Meningoencephalitis: meningococcal disease, *H. influenzae* type B, rabies, arboviral disease, cryptococcal disease. (Multiple other potential causes that can be difficult to diagnose include: ehrlichiosis, leptospirosis, and amoebic meningoencephalitis.)

Fever with rash: meningococcal disease, Rocky Mountain Spotted fever, measles, varicella, hemorrhagic fevers. (Multiple other potential causes that can be difficult to diagnose include: ehrlichiosis, bartonellosis.)

Hepatic syndrome: hepatitis A-E, dengue, yellow fever. (Multiple other potential causes that can be difficult to diagnose include: leptospirosis, Q fever.)

Myocarditis: Q fever, influenza (Multiple other potential causes include: Chagas, ehrlichiosis.)

Non-infectious cases of severe illnesses are numerous and include toxic exposures, overdoses of pharmaceuticals and of illicit drugs, malignancy, and autoimmune or genetic conditions.

3. CASE DEFINITIONS

A. Clinical Criteria for Diagnosis

Case meets all of the following criteria:

- Critical illness (an illness resulting in admission to the intensive care unit) or death in a person aged 1 to 49,

AND

- Previously healthy with no preexisting known medical condition including:
 - Malignancy;
 - HIV infection;
 - Chronic cardiac, pulmonary, renal, hepatic or rheumatologic disease;
 - Diabetes mellitus;
 - Immunosuppressive therapy or disease;
 - Trauma thought to be related to illness;
 - Evidence of toxic ingestion or exposure;
 - Nosocomial infection prior to the onset of illness;

AND

- Hallmarks of infectious disease including at least one of the following:
 - Fever or history of fever;
 - Leukocytosis (total white blood cell [WBC] count above the range for normal);
 - Histopathologic evidence of an acute infectious process; **or** a physician-diagnosed syndrome consistent with an infectious disease including: encephalitis/meningitis, fulminant hepatitis/hepatic failure, myocarditis, or ARDS/respiratory failure;

AND

- Preliminary testing has not revealed a cause for illness or death.

4. DIAGNOSIS AND LABORATORY SERVICES

If a cluster of cases with a similar clinical presentation is detected, DOH and CDC may assist with additional diagnostic testing. For single cases, resources are limited for additional testing unless an agent of bioterrorism or local acquisition of a non-endemic agent is suspected. Call Communicable Disease Epidemiology (CDE) to discuss the case. CDE may recommend obtaining and storing specimens for possible future testing if additional similar cases occur.

5. ROUTINE CASE INVESTIGATION

A. Evaluate the Diagnosis

Request medical records and review the clinical presentation, physical exam findings, laboratory findings and autopsy report, if available. Clusters of unexplained critical illness or death may require a more extensive investigation including additional laboratory testing. Therefore, it is prudent to request laboratories to hold any remaining clinical specimens from patients who have died. This includes holding respiratory specimens (sputum, bronchioalveolar lavage fluid, pleural fluid), cerebral spinal fluid

(CSF), all serum (early and late collections), other blood specimens, and fixed and fresh tissue collected at autopsy from abnormal organs. Consult with Communicable Disease Epidemiology regarding long term storage of specimens at Washington State Public Health Laboratories for possible future testing.

B. Manage the Case

Infection control measures depend on the suspected agent. Consult with Communicable Disease Epidemiology. In general, follow standard precautions in health care settings. Contact or respiratory precautions may also be appropriate depending on the patient's symptoms. Conditions such as SARS and South American hantavirus infection were transmitted from undiagnosed patients to health care providers.

Autopsies are considered to carry little risk of communicable disease transmission. To avoid risk during autopsy of an unexplained death that may be due to a communicable agent, limit the personnel involved and require appropriate personal equipment including an N95 or higher respirator, full face shield, goggles, gloves, complete body coverage by protective wear, and if appropriate heavy or chain mail gloves to help prevent injury from instruments or bone fragments. Minimize aerosols by using a handsaw rather than an oscillating saw when cutting bone, and by avoiding contact of the saw blade with brain tissue. Use a 10% solution of sodium hypochlorite for disinfection of all exposed surfaces and equipment during and after the autopsy.

C. Identify Potential Sources of Infection

Determine if the case has been in contact with others with similar symptoms. Identify any potential infectious exposures including recent travel, occupational or hobby exposures, or contact with unusual or ill animals. Public health concern would be increased for conditions that appear to be spreading from person to person, that result from unusual exposures, or that occur as a cluster with potential common exposure.

6. MANAGING SPECIAL SITUATIONS

A. Suspected Bioterrorism

If bioterrorism is suspected, Communicable Disease Epidemiology will arrange for specialized laboratory testing, provide guidelines for treatment, prophylaxis and infection control and assist in field investigations.

ACKNOWLEDGEMENTS

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UPDATES

January 2011: Section 1: The Legal Reporting Requirements section was revised to reflect the 2011 Notifiable Conditions Rule revision. Section 2: A section was added listing potential etiologies of highest public health significance. Section 5A: Details were added regarding which specimens to obtain for testing or storage if appropriate.

June 2012: The guideline was reviewed. No significant changes were made.

July 2014: The guideline was reviewed. No significant changes were made.