Varicella Death (Laboratory Confirmed)

1. DISEASE REPORTING

A. Purpose of Reporting and Surveillance

1. To determine mortality rates for laboratory-confirmed varicella-associated deaths.
2. To monitor the epidemiology of severe varicella infections.
3. To facilitate public health action and monitor the impact of the varicella immunization program.

B. Legal Reporting Requirements

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

C. Local Health Jurisdiction Investigation Responsibilities

1. Begin the investigation immediately.
2. Report all *probable* and *confirmed* varicella deaths (see definition below) to CDE. Complete the varicella death report form and enter the data into the Washington Disease Reporting System (WDRS).

2. THE DISEASE AND ITS EPIDEMIOLOGY

A. Etiologic Agent

Varicella-zoster virus (VZV) is the causative agent for varicella (chicken pox). When considering varicella death, it is important to distinguish between varicella-zoster and herpes-zoster. Herpes-zoster (shingles) is caused by the reactivation of VZV and not primary varicella. Herpes-zoster associated deaths do not need to be reported.

B. Description of Illness

Varicella is a febrile rash illness. The varicella rash is characterized as a pruritic, maculopapular, vesicular rash, that over time will become dried crusts. The rash may initially present on the chest, back and face and then spread across the entire body. Typically, around 250 to 500 lesions are seen during the illness. The rash will crust over a three to seven-day period. After crusts form, the individual is no longer infectious. Other common symptoms include malaise, loss of appetite, and headache.

Those who are immunocompromised or are pregnant may be at higher risks of severe disease, complications, or death. Serious complications from varicella disease include secondary bacterial infections, pneumonia, encephalitis, and death.

Varicella can also occur in individuals who have been vaccinated. This is called modified or breakthrough varicella. The rash may present atypically with fewer lesions (<50), mild disease, and a shorter duration of rash. Breakthrough varicella is still contagious, but is much less so compared to an unvaccinated individual.

D. Reservoir

The only reservoir of VZV is humans and disease can only occur in humans. After primary varicella (chicken pox), the virus does remain latent in the sensory-nerve ganglia. Latent VZV could reactivate causing herpes-zoster (shingles). A susceptible person exposed to VZV from shingles may develop chicken pox.

E. Modes of Transmission

Varicella is spread from person to person and is highly infectious. Transmission requires direct contact with someone with either chicken pox or shingles. Viral particulates from the skin lesions and infectious respiratory particulates can be aerosolized and inhaled, causing transmission. In utero infection can also occur.

F. Incubation Period

Ten to 21 days. Most cases occur between 14 to 16 days after contact with the virus.

G. Period of Communicability

It is estimated that varicella communicability begins one to two days prior to the onset of rash and ends after all the lesions have crusted, which is usually three to seven days.

For contact tracing purposes, the deceased person should be considered to have been contagious for the two days prior to rash onset and until all lesions have crusted over or seven days after rash onset, whichever was longer. Please note that for immunocompromised individuals, the duration of communicability may be longer.

H. Treatment

In healthy individuals, there is no recommended antiviral treatment recommended for varicella. Taking aspirin containing medication to relieve fever associated with varicella is not recommended because the use of aspirin or aspirin-containing medication for children with chicken pox has shown to be associated with higher risk of Reye's syndrome. Reye's syndrome severely affects the liver and brain and can cause death. The American Academy of Pediatrics also recommends against treatment with ibuprofen as that as shown to be associated with fatal bacterial skin infections.

For those above the age of 12 and those who are at higher risk of severe disease or complications, oral or intravenous acyclovir can be used.

(For more information, see: https://www.cdc.gov/chickenpox/about/prevention-treatment.html)

I. Immunity

Those who have been vaccinated or those who have been previously infected are considered immune.

The Advisory Committee on Immunization Practices (ACIP) recommends that those who do not have evidence of immunity to be vaccinated. Two doses are recommended.

Here is the ACIP varicella recommendation:

Routine two-dose vaccination:

- First dose at age 12 through 15 months
- Second dose at age four through six years

Second dose catch up vaccination:

- If the second dose is administered after the seventh birthday, the minimum interval between doses is ≥3 months for children age <13 years and four weeks for persons age ≥13 years

Adolescents and Adults (≥ age 13 years) without other evidence of immunity:

- Give two doses four to eight weeks apart
- If it has been more than eight weeks since the first dose, the second dose may be given without restarting the schedule

https://www.cdc.gov/vaccines/vpd/varicella/hcp/recommendations.html

Other evidence of immunity, in addition to documentation of vaccination, is laboratory evidence of immunity or lab-confirmed disease, birth in the US before 1980 (not for those who are pregnant, work in health care settings, or immunocompromised), and diagnosis or verification of history of chickenpox or shingles by a health care provider.

3. CASE DEFINITIONS

A. Clinical Criteria for Diagnosis

An illness with acute onset of diffuse (generalized) maculopapular vesicular rash without other apparent cause. In vaccinated persons varicella that develops more than 42 days after vaccination (breakthrough disease) due to infection with wild-type VZV, is usually mild, with fewer than 50 skin lesions and of shorter duration of illness. The rash may also be atypical in appearance (maculopapular with few or no vesicles).

B. Laboratory Criteria for Diagnosis

Demonstration of VZV DNA by polymerase chain reaction (PCR) tests from a clinical specimen, ideally scabs, vesicular fluid, or cells from the base of a lesion is the preferred method for varicella diagnosis. PC is also useful for confirming breakthrough disease. Other methods, such as DFA and culture, are available for diagnosis but are less sensitive and specific than PCR.

Positive serologic test for varicella-zoster immunoglobulin M (IgM) antibody when varicella-like symptoms are present.

Four-fold or greater rise in serum varicella immunoglobulin G (IgG) antibody titer by any standard serologic assay between acute and convalescent sera.

For both unvaccinated and vaccinated persons, PCR is the most reliable method for confirming infection.

C. Case Definition

Varicella death case classification (1998)

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

Varicella case classification (2010)

Probable: A case that meets the clinical case definition, is not laboratory confirmed, and is not epidemiologically linked to another probable or confirmed case.

Confirmed: A case that is laboratory confirmed or that meets the clinical case definition and is epidemiologically linked to a confirmed or a probable case

4. DIAGNOSIS AND LABORATORY SERVICES

A. Diagnosis

Varicella death should be laboratory confirmed. Additionally, it is important to distinguish between varicella-zoster and herpes-zoster. Only varicella-zoster (primary varicella) associated deaths need to be reported. To distinguish between varicella-zoster and herpes-zoster, immunity and history of previous varicella disease should be assessed and if needed, further testing can be pursued.

1. **Detection of VZV nucleic acid by Polymerase Chain Reaction (PCR):** Using PCR to detect VZV in skin lesions (vesicles, scabs/crusts, maculopapular lesions) is the most sensitive method to confirm diagnosis of varicella. Vesicular lesions or scabs are the best samples. It may be difficult to collect specimens from maculopapular lesions. The best samples are ones collected by unroofing a vesicle, especially one that is fresh, fluid filled, then using a polyester swab and rubbing the swab against the base of the lesion. Other sources such as nasopharyngeal secretions, saliva, blood, urine, bronchial washings, and cerebrospinal fluid, however, these are not preferred because of lower sensitivity.
2. **Viral culture of VZV:** VZV can also be isolated from tissue culture, although, compared to the PCR and DFA methodologies, it takes more time and is less sensitive. This is because the viral proteins remain detectable for a longer period even if viral replication has ended. PCR may continue to be positive when there is no growth/replication through viral culture. Viral culture may fail to confirm for as many as 50 percent of varicella infections. VZV can be isolated from blood and CSF, however, often not from crusted lesions.

3. **Serologic test:** Positive IgM can be an indication of recent active VZV infection but it does not distinguish between primary infection, reinfection, or reactivation. This is because IgM only lasts a short period of time after infection and can reemerge after each infection.

B. Services Available at the Washington State Public Health Laboratories (PHL)

In situations where public health action may be indicated, PHL can test for VZV using the RT-PCR methodology.

Refer to: Specimen Collection and Submission Instructions

5. ROUTINE CASE INVESTIGATION

The case investigation for laboratory-confirmed varicella-associated deaths involves reviewing medical records for risk factors, clinical presentations, and laboratory test results and completing the varicella death case report form.

A. Evaluate the Diagnosis

Assess the clinical presentation (e.g., lesion(s), rash, malaise, fever), risk factors (e.g., recent close contact with varicella cases), and immunization history for the patient.

B. Identify Source of Infection

Identify confirmatory laboratory testing and other laboratory test done (serology and PCR/culture). If needed, specimens can be sent to Washington Public Health Laboratories for further testing/confirmation. Collect information about exposures, recent varicella infection, ill close contacts, travel, and medical history of the patient.

C. Identify Potentially Exposed Persons

Since varicella is highly contagious, outbreaks of varicella can be common. However, death in recent years have been rare. It is important to review medical records to determine history of disease and/or vaccination history for all exposed.

D. Environmental Evaluation

Not required. The varicella virus does not persist long enough in the environment to be detected upon investigation.

6. CONTROLLING FURTHER SPREAD

A. Infection Control Recommendations/Case Management

Contact investigations for varicella death can be performed at the discretion of the local health jurisdiction. Consult with Communicable Disease Epidemiology for managing outbreak situations. Exposed persons should discuss their varicella immune status with their health care provider. Symptomatic contacts, especially those who are at high risk of complications, should contact their provider immediately for monitoring of symptoms and potential treatment. All contacts with varicella-like illness should avoid work, school, childcare, and other public settings until after the varicella rash resolves.

7. MANAGING SPECIAL SITUATIONS

Special situations will be handled on a case-by-case basis. Please consult with the Office of Communicable Disease Epidemiology (CDE).

8. ROUTINE PREVENTION

A. Immunization Recommendations

Here is the ACIP varicella recommendation:

Routine two-dose vaccination:

- First dose at age 12 through 15 months
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Second dose catch up vaccination

- If the second dose is administered after the seventh birthday, the minimum interval between doses is ≥3 months for children age <13 years and 4 weeks for persons age ≥13 years

Adolescents and Adults (≥ age 13 years) without other evidence of immunity

- Give two doses four to eight weeks apart
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(For more information: https://www.cdc.gov/vaccines/vpd/varicella/hcp/recommendations.html)

B. Preventing Varicella Transmission and Varicella Death

Vaccination is the best way to prevent varicella disease, transmission, and death. If individuals are concerned, it is recommended that they reach out to their medical provider to ensure that they are up to date with their varicella vaccination and for further medical guidance.

ACKNOWLEDGEMENTS

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UPDATES

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