



**Report Immediately**

# Smallpox

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## Disease Plan

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Last updated: July 20, 2017 by Bree Barbeau

Questions about this disease plan?

Contact the Utah Department of Health Bureau of Epidemiology: 801-538-6191.

## ✓ SMALLPOX CRITICAL CLINICIAN INFORMATION

### Clinical Evidence

#### Signs/Symptoms

- **Ordinary Smallpox**
  - Severe prodromal illness with high fever (102°F-104°F)
  - Enanthem in mouth or on tongue
  - Rash which progresses through successive stages of macules, papules, vesicles, pustules, and then crusted scabs. The center of the vesicles will tend to dimple, a presentation called “umbilication”; rash lasts around 14 days
- **Modified Smallpox**
  - Less severe prodromal illness
  - Rash with fewer, more superficial lesions than those seen in ordinary smallpox; rash lasts around 10 days
- **Flat (malignant) Smallpox**
  - Skin lesions develop slowly, merge together, and remain flat and soft (often described as “velvety” to the touch)
  - Toxemia
- **Hemorrhagic Smallpox**
  - More severe prodromal symptoms with high fever, severe headache, and abdominal pain
  - Development of a dusky erythema after illness onset, followed by petechiae and skin and mucosal hemorrhages
  - Toxemia or death
- **Variola Minor**
  - Less severe symptoms with shorter duration
- **Variola Sine Eruptione**
  - Asymptomatic or brief rise in temperature, headache, and influenza-like symptoms

#### Period of Communicability

- Generally contagious for three weeks, from the development of the earliest lesions in the mouth to the disappearance of all scabs; the first week of rash illness is the most infectious period

#### Incubation Period

- Average 10-14 days; range 7-19 days

#### Mode of Transmission

- Respiratory droplets
- May occur through direct contact with an infected person or contact with an object soiled with infectious particles

### Laboratory Testing

#### Type of Lab Test

- PCR

#### Type of Specimens

- Vesicular or pustular fluid

### Treatment Recommendations

#### Type of Treatment

- None

**Prophylaxis**

- Vaccination

**Isolation Requirements**

**Isolation of Case**

- Standard, contact, and airborne isolation until lesions have dried and crusts have separated

**Case Contact Management**

- Vaccinate and monitor based on protocol outlined in disease plan

## ✓ WHY IS SMALLPOX IMPORTANT TO PUBLIC HEALTH?

Thousands of years ago, variola virus (smallpox virus) emerged and began causing illness and deaths in human populations, with smallpox outbreaks occurring from time to time. Thanks to the success of vaccination, the last natural outbreak of smallpox in the United States occurred in 1949. In 1980, the World Health Assembly declared smallpox eradicated (eliminated), and no cases of naturally occurring smallpox have happened since. Smallpox research in the United States continues and focuses on the development of vaccines, drugs, and diagnostic tests to protect people against smallpox in the event that it is used as an agent of bioterrorism.

The global eradication of smallpox was announced in 1979, marking one of the greatest achievements of modern public health. Although there have been no reported cases since, continued public health interest in this virus remains because of the concern regarding smallpox as a potential agent of bioterrorism. A single confirmed case of smallpox today would likely be the result of an intentional act and would be considered a public health emergency.

## ✓ DISEASE AND EPIDEMIOLOGY

### Clinical Description

Smallpox infection typically begins with a prodromal illness characterized by a high fever (102–104°F), malaise, headache, backache, and abdominal pain. This prodromal period typically lasts 2–5 days and leaves the patient often too sick to carry on with normal activities. A rash begins to develop after the prodromal period. The rash first appears as an enanthem, with tiny red spots developing inside the mouth or on the tongue that are often not noticeable. These spots then change into sores that break open and spread large amounts of the virus into the mouth and throat. Once the sores in the mouth start breaking down, a rash appears on the skin, starting on the face and spreading to the arms and legs, and then to the hands and feet. Usually, it spreads to all parts of the body within 24 hours. The rash follows a characteristic “centrifugal” pattern, with the highest concentration of lesions on the face and distal extremities, including the palms of the hands and soles of the feet. This early rash period lasts about 4 days. Over the next 7-10 days the rash will slowly progress through successive stages of macules, papules, vesicles, pustules, and then crusted scabs. The center of the vesicles will tend to dimple, a presentation called “umbilication”. Between the 7th and 10th day of the rash the pustules reach their maximum size. The pustules are deeply embedded in the skin, giving the feeling of small beads underneath the skin. Three to four weeks after the rash onset the scabs begin to fall off leaving depigmented, often pitted scars.

- **Variola major** is the more severe form of the disease and can have four very different clinical presentations:
  - **Ordinary smallpox:** Ordinary smallpox was the most common form, accounting for over 85% of all cases during the smallpox era. This form follows the clinical presentation outlined above.

- **Modified smallpox:** Modified-type smallpox occurs in previously vaccinated individuals. In this type, the prodrome stage may still consist of severe headache, backache, and fever, and may last as long as in ordinary type. However, once the skin lesions appear, they generally evolve more quickly, and crusting completes within 10 days as opposed to 14 with ordinary smallpox. There may also be fewer, more superficial lesions than those seen in ordinary smallpox. Patients also do not tend to have a fever during the evolution of the rash.
- **Flat (malignant) smallpox:** Flat-type—or malignant—smallpox is very rare, and is characterized by intense toxemia. It occurs more frequently in children. In contrast to ordinary smallpox, the skin lesions in this type develop slowly, merge together, and remain flat and soft (often described as “velvety” to the touch). They never progress to the pustular stage. The appearance of the lesions suggests a deficient cellular immune response to variola virus, and the majority of flat-type smallpox cases are fatal. If the patient survives, the lesions gradually disappear without forming scabs. Prior vaccination appears to protect against flat-type smallpox.
- **Hemorrhagic smallpox:** Hemorrhagic-type smallpox occurs among all ages and in both sexes, but is more common in adults. Pregnant women appear to be more susceptible. The underlying biological reasons for this type are unclear. Prior vaccination is not protective. This type is differentiated from ordinary smallpox by:
  - Shorter incubation period
  - More severe prodromal symptoms with high fever, severe headache, and abdominal pain
  - Development of a dusky erythema after illness onset, followed by petechiae and skin and mucosal hemorrhages
  - Death usually occurs by the 5th or 6th day of the rash, often before characteristic smallpox lesions develop. Death results from a profound toxemia, leading to multi-organ failure.
- **Variola minor** is a much less severe form of the disease with a shorter duration.
- **Variola sine eruptione** or subclinical infection can occur among vaccinated contacts of a recent smallpox case or in infants with maternal antibodies. Affected persons are asymptomatic or have a brief rise in temperature, headache, and influenza-like symptoms; the transmission of clinical smallpox has not been documented with variola sine eruptione.

## **Causative Agent**

Smallpox is caused by the variola virus, a DNA virus and a member of the Poxviridae family (genus Orthopoxvirus). The two clinical forms of the disease are caused by different strains of the virus, variola major and variola minor.

## **Differential Diagnosis**

The following table describes diseases and syndromes most commonly confused with smallpox.

Condition	Clinical Clues
Varicella (Chickenpox)	Most common in children < 10 years; children usually do not have a viral prodrome
Disseminated herpes zoster	Immunocompromised or elderly persons; rash looks like varicella and usually begins in dermatomal distribution
Impetigo (caused by <i>Streptococcus pyogenes</i> or <i>Staphylococcus aureus</i> )	Honey-colored crusted plaques with bullae are classic but may begin as vesicles; regional not disseminated rash; patients usually not ill
Drug eruptions	Exposure to medications; rash often generalized
Contact dermatitis	Itching; contact with possible allergens; rash often localized in pattern suggesting external contact
Erythema multiforme minor	Target, "bull's eye", or iris lesions; often follows recurrent herpes simplex virus infections; may involve hands and feet
Erythema multiforme major (Stevens-Johnson syndrome)	Major form involves mucous membranes and conjunctivae; may be target lesions or vesicles
Enterovirus infection (particularly hand, foot, and mouth disease)	Summer and fall; fever and mild pharyngitis 1-2 days before rash onset; lesions initially maculopapular but evolve into whitish-grey, tender, flat, often oval vesicles; peripheral distribution
Disseminated herpes simplex	Lesions indistinguishable from varicella; immunocompromised host
Scabies; insect bites	Itching is a major symptom; patient is not febrile and is otherwise well
Molluscum contagiosum	May disseminate in immunosuppressed persons; can occur anywhere on the body; presents as small, raised, and usually white, pink, or flesh-colored lesions with a dimple or pit in the center

CDC has developed criteria that can be used to evaluate suspected smallpox cases and to categorize patients into high, moderate or low risk for smallpox. There are three major and five minor smallpox criteria:

**Major criteria**

1. Febrile prodrome occurring 1 to 4 days before rash onset:
  - a. Fever  $\geq 101^{\circ}\text{F}$  ( $38.3^{\circ}\text{C}$ ) **AND** at least one of the following:
    - i. prostration
    - ii. headache
    - iii. backache
    - iv. chills
    - v. vomiting
    - vi. severe abdominal pain
2. Classic smallpox lesions: deep-seated, firm/hard, round, well-circumscribed vesicles or pustules. As they evolve, lesions may become umbilicated or confluent.
3. Lesions in the same stage of development (e.g., all are vesicles or all are pustules) on any ONE part of the body (e.g., the face, arms).

**Minor criteria**

1. Centrifugal distribution of rash: greatest concentration of lesions on face and distal extremities

2. First lesions on the oral mucosa/palate, face, or forearms
3. Severity: Patient appears toxic or moribund
4. Slow rash evolution: lesions evolved from macules to papules to pustules over days (each stage lasts 1 to 2 days)
5. Lesions on the palms and/or soles

A person is considered at **high risk** for smallpox if he or she meets all three major criteria. Immediate action should be taken to make sure that contact precautions and respiratory isolation are implemented. These patients should be reported to local and/or state health authorities immediately. Obtain digital photographs if possible, and consult with dermatology and/or infectious disease experts. Following such consultation, if the patient is still considered to be at high risk, the state health department will immediately report the case to CDC and arrangements will be made for laboratory testing for smallpox virus. Do not proceed with laboratory testing for other diagnoses until smallpox has been ruled out.

A person considered at **moderate risk** for smallpox must have a febrile prodrome and either one other major criterion or four or more minor criteria. These patients should be isolated and be evaluated urgently to determine the cause of the illness. Persons classified as high or moderate risk should be seen in consultation with a specialist in infectious diseases and/or dermatology whenever possible.

Any person who did not have a febrile prodrome is considered at **low risk**, as are persons who had a febrile prodrome and fewer than four minor criteria. These patients should be managed as clinically indicated.

The following chart outlines the risk categories described above:

## **RISK OF SMALLPOX**

### **High Risk of Smallpox → Report Immediately**

1. Febrile prodrome (defined above) **AND**
2. Classic smallpox lesion (defined above) **AND**
3. Lesions in same stage of development (defined above)

### **Moderate Risk of Smallpox → Urgent Evaluation**

1. Febrile prodrome **AND**
  2. One other **MAJOR** smallpox criterion (defined above)
- OR**
1. Febrile prodrome **AND**
  2.  $\geq 4$  **MINOR** smallpox criteria (defined above)

### **Low Risk of Smallpox → Manage as Clinically Indicated**

1. No febrile prodrome
- OR**
1. Febrile prodrome **AND**
  2.  $< 4$  **MINOR** smallpox criteria (defined above)

## Laboratory Identification

Variola virus can be detected in vesicular or pustular fluid by culture or PCR. The diagnosis of an Orthopoxvirus infection can be made rapidly by electron microscopic examination of dried vesicular fluid on a microscope slide, but does not distinguish between vaccinia, variola and other poxvirus infections. PCR and culture testing will confirm the diagnosis.

Laboratory testing for smallpox is warranted only once a case is classified as high risk. For cases that meet the moderate risk classification, the most important laboratory procedure is varicella-zoster virus testing.

NOTE: UPHL is capable of testing low and moderate-risk specimens to rule out smallpox. High-risk specimens will be sent to Colorado public health lab for testing. Testing for smallpox must be coordinated with UDOH, who will work with UPHL and CDC to determine the best course of action.

## Treatment

There is no proven treatment for smallpox disease. Prevention is achieved through vaccination. For purposes of controlling a smallpox outbreak, smallpox vaccine and antivirals can be used.

## Case Fatality

- **Variola major** has a case fatality rate of 30-50% in unvaccinated persons.
- **Hemorrhagic** and **Flat type smallpox** are usually fatal.
- **Variola minor** has an overall case fatality rate of less than 1%.

## Reservoir

Humans are the only known reservoir for the variola virus.

## Transmission

Transmission occurs primarily from through the spread of respiratory droplets. However, transmission may occur through direct contact with an infected person or contact with an object soiled with infectious particles.

## Susceptibility

Routine vaccination in the US for smallpox stopped in 1972. Those who have previously been vaccinated are most likely still immune; however, susceptibility among the unvaccinated is universal. Selected military, research, and medical personnel are immune to the disease because of vaccination.

## Incubation Period

The incubation period ranges from 7-19 days; commonly 10-14 days to onset of illness and 2-4 days more to onset of rash.

## Period of Communicability

An infected person is generally contagious for three weeks, from the development of the earliest lesions in the mouth to the disappearance of all scabs. The first week of rash illness is the most infectious period.

## **Epidemiology**

The last case of naturally-acquired smallpox in Utah, the US, and the world occurred in 1945, 1949, and 1977 (in Somalia), respectively. The last two cases of smallpox in the world occurred in 1978 in England and were associated with a breach in laboratory safety. In 1980, global eradication of smallpox was certified by the World Health Organization. Currently, two WHO reference laboratories (the CDC and the Institute of Virus Preparations in Moscow, Russia) hold variola virus stocks under strict security. All laboratory work with the smallpox virus is done under strict biosafety level 4 procedures.

## **PUBLIC HEALTH CONTROL MEASURES**

### **Public Health Responsibility**

- Immediately notify the UDOH epidemiologist on call or the State Epidemiologist to discuss the situation and assess risk of smallpox.
- Rapidly investigate all suspect cases of disease and fill out and submit appropriate disease investigation forms.
- Assure that appropriate testing protocols are followed.
- Rapidly investigate all potentially exposed persons.
- Identify sources of exposure and stop further transmission.
- Identify cases and clusters of human illness that may be associated with a bioterrorism incident.

### **Prevention**

#### **Environmental Measures**

If a patient presents to an emergency department, clinic, or doctor's office with an acute generalized vesicular or pustular rash illness, care should be taken to decrease the risk of disease transmission. Patients should not be left in common waiting areas. The patient should be assessed to determine whether there is a high, medium, or low risk of smallpox.

1. In a doctor's office or clinic, the patient should be placed immediately in a private room with the door kept shut.
2. When admitted or while being held for observation, the facility should institute appropriate airborne isolation and contact precautions and should alert the infection prevention/control department. The patient should be placed in a private room at negative pressure to the rest of the facility (airborne infection isolation). The door should be kept closed at all times, except when staff or the patient must enter or exit.
3. Staff and visitors, regardless of vaccination status, should wear properly fitted respirators (N95 or higher level of protection), gloves, and gowns.

4. The patient should wear a surgical mask whenever they must be outside of the negative pressure isolation room and must be gowned or wrapped in a sheet so that the rash is fully covered.

## **Chemoprophylaxis**

Vaccination after exposure to smallpox has reduced the rate of secondary cases in households by up to 91% when compared to unvaccinated household members. Secondary attack rates were lowest among contacts that were vaccinated within 7 days of their exposure. Contacts that did develop disease usually had much milder symptoms.

## **Vaccine**

The smallpox vaccine is made from a related orthopox virus - the vaccinia virus. The vaccine is highly effective at inducing immunity against smallpox when administered effectively prior to exposure. Smallpox vaccine production ceased in the early 1980s and current supplies of smallpox vaccine are limited. However, recent studies have demonstrated that vaccines stored in the 1960s and 1970s still have excellent potency, even when diluted. Imvamune is another vaccine formulation of a modified vaccinia that is under study as a future generation smallpox vaccine. It is a highly attenuated vaccinia virus that has an excellent safety profile, even in immunocompromised people.

Because the smallpox vaccine is a live-virus vaccine, vaccinia virus is present at the site of vaccination beginning about 4 days after vaccination. Viral shedding from the vaccination site usually occurs 4-14 days after vaccination, but vaccinia can be recovered from the site until the crust separates from the skin. Therefore, appropriate hand hygiene and/or keeping the vaccination site covered with a bandage is necessary to prevent transmission of the virus to contacts of the vaccinee.

## **Isolation and Quarantine Requirements**

**Isolation:** Cases should be placed on standard, contact, and airborne isolation until lesions have dried and crusts have separated.

**Healthcare Settings:** Cases should be placed on standard, contact, and airborne isolation. The patient should be placed in a private, airborne infection isolation room with negative pressure ventilation with high-efficiency particulate air filtration. Cases should remain isolated until lesions have dried and crusts have separated.

**Quarantine:** Febrile contacts shall be placed under fever surveillance (quarantine) for 18 days from the last contact or 14 days from successful vaccination (whichever comes first), with monitoring and recording of temperature occurring twice daily (morning and evening). Febrile contacts with or without rash shall be considered the same as a case and shall be handled in the same fashion (isolation). If no rash develops after five days and the fever is diagnosed as being caused by recent vaccination or some other non-smallpox etiology, the contacts may be released from isolation to home but with continued fever surveillance for 18 days following their last contact with a case or 14 days following successful vaccination (whichever comes first).

## ✓ CASE INVESTIGATION

### Reporting

If smallpox were to reoccur in the US, or elsewhere, the most likely circumstances of reintroduction are generally accepted to be:

- An unintentional infection in a laboratory. Currently there are only two WHO-approved smallpox virus research and repository laboratories. However, there is speculation that stockpiles of variola virus may exist elsewhere.
- A bioterrorist attack involving deliberate infection of a person.
- A bioterrorist attack involving intentional release of smallpox virus into the environment.

Any suspect or confirmed case of smallpox, or of any potential exposure to an agent which could cause smallpox, must be reported to the UDOH Bureau of Epidemiology immediately at 1-888-EPI-UTAH. Adverse events following smallpox vaccination are also reportable.

**Table 1. Criteria for Reporting a Case of Smallpox to Public Health Authorities**

Criterion	Reporting		
<b>Clinical Evidence</b>			
fever $\geq 101^{\circ}\text{F}$ ( $38.3^{\circ}\text{C}$ )	N	N	N
fever occurs 1–4 days before rash onset		N	N
prostration	O	O	O
headache	O	O	O
backache	O	O	O
chills	O	O	O
vomiting	O	O	O
severe abdominal pain	O	O	O
rash with deep-seated, firm or hard, round, well-circumscribed vesicles or pustules; as they evolve, lesions may become umbilicated or confluent			
rash with <i>flat, soft, focal</i> lesions; as they evolve, lesions may become confluent and portions of skin may slough			
lesions on any ONE part of the body (e.g., the face, arms) are all in the same stage of development (i.e., all are vesicles, or all are pustules)			
<i>widespread hemorrhage</i> in skin and mucous membranes			
generalized vesicular rash		N	
generalized pustular rash			N
identifiable cause of clinical findings		A	A
<b>Laboratory Evidence</b>			
polymerase chain reaction (PCR) identification of variola DNA in a clinical specimen	S*		

isolation of smallpox (variola) virus from a clinical specimen	*		
polymerase chain reaction (PCR) identification of variola DNA in an isolate from a clinical specimen	S*		
Detection of orthopox-reactive IgM antibodies, 3-56 days post symptom onset			
<b>Epidemiological Evidence</b>			
contact ( $\leq 2$ meters) for ( $\geq 3$ hours) with a laboratory confirmed case of smallpox	N		
contact ( $\leq 2$ meters) with a confirmed, probable, or suspected case of smallpox			
contact with smallpox infected bodily fluids or contaminated objects such as bedding			
worker in a laboratory that contains smallpox virus			

Notes:

S = This criterion alone is sufficient to report a case

N = This criterion in conjunction with all other “N” and any “O” criteria in the same column is required to report a case. A number following an “N” indicates that this criterion is only required for a specific clinical presentation (see below).

O = At least one of these “O” criteria in each category in the same column (e.g., clinical presentation and laboratory findings)—in conjunction with all other “N” criteria in the same column—is required to report a case. A number following an “O” indicates that this criterion is only required for a specific clinical presentation (see below).

A = This criterion must be absent (i.e., NOT present) for the case to meet the case definition.

\*A requisition for any of the “S” or “N” laboratory tests is sufficient to meet the reporting criteria

1 = flat type smallpox

2 = hemorrhagic smallpox

3 = variola sine eruptione

### Case Definition

**Smallpox (pre-event definition):** **\*This definition is to be used for pre-event surveillance. It is more sensitive and less specific than the definition developed by CSTE, which is to be used only for post-event surveillance.**

### Clinical Case Definition

An illness with acute onset of fever  $\geq 101^\circ\text{F}$  ( $38.3^\circ\text{C}$ ) followed by a rash characterized by firm, deep seated vesicles or pustules in the same stage of development without other apparent cause.

### Laboratory Criteria

- Polymerase chain reaction (PCR) identification of variola DNA in a clinical specimen, OR
- Isolation of smallpox (variola) virus from a clinical specimen *with* variola PCR confirmation

**NOTE:** Laboratory diagnostic testing for variola virus should be conducted in a CDC Laboratory Response Network (LRN) laboratory utilizing LRN-approved PCR tests and protocols for variola virus. Initial confirmation of a smallpox outbreak requires additional testing at CDC.

**NOTE:** The importance of case confirmation using laboratory diagnostic tests differs depending on the epidemiological situation. Because of the low predictive value of a positive lab test result in the absence of a known smallpox outbreak, in the pre-outbreak (pre-event) setting, laboratory testing should be reserved for cases that meet the clinical case definition and are thus classified as being a potential high risk for smallpox according to the rash algorithm poster (<http://emergency.cdc.gov/agent/smallpox/diagnosis/pdf/spox-poster-full.pdf>).

### **Case Classification**

**Suspect:** A case with a febrile rash illness with fever preceding development of rash by 1-4 days.

**Probable:** A case that meets the clinical case definition, or a case that does not meet the clinical case definition but is clinically consistent with smallpox and has an epidemiological link to a confirmed case of smallpox. Examples of clinical presentations of smallpox that would not meet the ordinary type (pre-event) clinical case definition are: a) hemorrhagic type, b) flat type, and c) variola sine eruption.

**Confirmed:** A case of smallpox that is laboratory confirmed, or a case that meets the clinical case definition that is epidemiologically linked to a laboratory confirmed case.

**Smallpox (2010) Post-event definition:** **\*This definition is to be used ONLY during post-event surveillance.**

### **Clinical Case Definition**

An illness with acute onset of fever  $\geq 101^{\circ}\text{F}$  ( $38.3^{\circ}\text{C}$ ) followed by a rash characterized by firm, deep seated vesicles or pustules in the same stage of development without other apparent cause.

Clinically consistent cases are those presentations of smallpox that do not meet this classical clinical case definition: a) hemorrhagic type, b) flat type, and c) *variola sine eruption*. Detailed clinical description is available on the CDC website (<http://emergency.cdc.gov/agent/smallpox/overview/disease-facts.asp>).

### **Laboratory criteria for diagnosis**

- Polymerase chain reaction (PCR) identification of variola DNA in a clinical specimen, OR
- Isolation of smallpox (variola) virus from a clinical specimen (Level D laboratory only; confirmed by variola PCR).

**NOTE:** Indications for laboratory testing of patients with suspected smallpox should be followed as described in detail in the CDC Smallpox Response Plan (<http://emergency.cdc.gov/agent/smallpox/lab-testing/>).

### **Case Classification**

**Suspect:** A case with a generalized, acute vesicular or pustular rash illness with fever preceding development of rash by 1-4 days.

**Probable:** A case that meets the clinical case definition, or a clinically consistent case that does not meet the clinical case definition and has an epidemiological link to a confirmed case of smallpox.

**Confirmed:** A case of smallpox that is laboratory confirmed, or a case that meets the clinical case definition that is epidemiologically linked to a laboratory confirmed case

**Exclusion Criteria**

A case may be excluded as a suspect or probable smallpox case if an alternative diagnosis fully explains the illness or appropriate clinical specimens are negative for laboratory criteria for smallpox.

**Table 2. Criteria to Determine Whether a Smallpox Case is Classified.**

Criterion	Confirmed			Probable		Suspect	
<b>Clinical Evidence</b>							
fever $\geq 101^{\circ}\text{F}$ ( $38.3^{\circ}\text{C}$ )	N	N	N	N	N	N	N
fever occurs 1–4 days before rash onset	N	N	N	N	N	N	N
prostration	O	O	O	O	O	O	O
headache	O	O	O	O	O	O	O
backache	O	O	O	O	O	O	O
chills	O	O	O	O	O	O	O
vomiting	O	O	O	O	O	O	O
severe abdominal pain	O	O	O	O	O	O	O
rash with deep-seated, firm or hard, round, well-circumscribed vesicles or pustules; as they evolve, lesions may become umbilicated or confluent	N	N	N	N			
rash with <i>flat, soft, focal</i> lesions; as they evolve, lesions may become confluent and portions of skin may slough					N1		
lesions on any ONE part of the body (e.g., the face, arms) are all in the same stage of development (i.e., all are vesicles, or all are pustules)	N	N	N	N			
widespread hemorrhage in skin and mucous membranes					N2		
generalized pustular rash							N
identifiable cause of clinical findings			A	A	A	A	A
<b>Laboratory Evidence</b>							
polymerase chain reaction (PCR) identification of variola DNA in a clinical specimen	N			A		A	A
isolation of smallpox (variola) virus from a clinical specimen		N		A		A	A
polymerase chain reaction (PCR) identification of variola DNA in an isolate from a clinical specimen		N		A		A	A
Detection of orthopox-reactive IgM antibodies, 3-56 days post symptom onset					N3		
<b>Epidemiological Evidence</b>							

**Smallpox: Utah Public Health Disease Investigation Plan**

contact ( $\leq 2$ meters) for ( $\geq 3$ hours) with a laboratory confirmed case of smallpox			N		N	N	
contact ( $\leq 2$ meters) with a confirmed, probable, or suspected							
contact with smallpox infected bodily fluids or contaminated objects such as bedding or clothing							
worker in a laboratory that contains smallpox virus							
generalized vesicular rash						N	

**Notes:**

N = This criterion in conjunction with all other “N” and any “O” criteria in the same column is required to confirm a case. A number following an “N” indicates that this criterion is only required for a specific clinical presentation (see below).

O = At least one of these “O” criteria in each category in the same column (e.g., clinical presentation and laboratory findings)—in conjunction with all other “N” criteria in the same column—is required to confirm a case. A number following an “O” indicates that this criterion is only required for a specific clinical presentation (see below).

A = This criterion must be absent (i.e., NOT present) for the case to meet the case definition.

\*A requisition for any of the “S” or “N” laboratory tests is sufficient to meet the reporting criteria

1 = flat type smallpox

2 = hemorrhagic smallpox

3 = *variola sine eruptione*

**Case Investigation Process**

1. Following immediate notification of the UDOH, the LHD may be asked to assist in investigating any case living within their community, including gathering the following information into UT-NEDSS:
  - a. The case’s name, age, address, phone number, status (e.g., hospitalized, at home, deceased), and parent/guardian information, if applicable
  - b. The name and phone number of the hospital where the case is or was hospitalized
  - c. The name and phone number of the attending physician
  - d. The name and phone number of the infection control official at the hospital
  - e. If the patient was seen by a healthcare provider before hospitalization or seen at more than one hospital, these names and phone numbers.
  - f. Clinical information including symptoms and date of symptom onset
  - g. Available diagnostic laboratory test information that is available
2. Specimen Collection, Transport, Testing, and Submission for Patients with Suspected Smallpox

**Outbreaks**

As smallpox no longer exists as a naturally occurring disease, a single laboratory confirmed case of smallpox would be considered an outbreak and an extensive response involving many different federal, state, and local agencies would be warranted.

**Identifying Case Contacts**

For the purposes of smallpox surveillance and case investigation, contacts are defined as follows:

- **Contact:** A person who has been exposed to the risk of infection
- **Primary contact:** A person with contact with a confirmed, probable, or suspect case of smallpox during the infectious period. Primary contacts include both household and non-household contacts. Risks of smallpox transmission is increased with increased duration of face-to-face contact of less than two meters ( $\leq 6.5$  feet).
- **Secondary contact:** A household member of a primary contact, a non-household contact, and a person who works in the household of a primary contact.

Priority categories for contacts, from highest priority to lowest, are as follows:

1. Case's household family members and others spending three or more hours in the household since the case's onset of fever.
2. Non-household members with contact 2 meters or less ( $\leq 6.5$  feet) with case with rash for 3 or more hours.
3. Non-household members with contact 2 meters or less ( $\leq 6.5$  feet) with case with rash for fewer than 3 hours.
4. Non-household members with any contact with case with rash for 3 or more hours.
5. Non-household members with any contact with case with rash for fewer than 3 hours.

## Case Contact Management

Vaccinating and monitoring contacts of cases and family contacts of contacts will help to protect those at greatest risk for contracting the disease as well as form a buffer of immune individuals to prevent the spread of disease. Large-scale vaccination in potentially exposed communities may become necessary, although it is crucial that cases be identified and isolated. Large-scale vaccination might also be applied to unaffected communities to protect against further spread of smallpox and additional releases as well as to build the public's confidence in its protection and ability to return to normal activities.

## Contract Tracing

Contact tracing should include the following steps:

1. Trace each contact whose name, address, and/or telephone number is known.
2. Use work and school contact numbers, telephone directories, voting lists, neighborhood interviews, site visits, "hangouts", etc., to trace contacts when contact information is unknown or incomplete. If contacts cannot be found through these mechanisms, other sources for notification of potential contacts (such as media announcements) may have to be considered.
3. Locate and interview each primary contact to confirm contact with the suspect, probable, or confirmed smallpox case, the presence or absence of symptoms in the contact (fever and/or rash), and to identify additional contacts that may not have been listed by the case.
4. Identify household contacts of each primary contact of the smallpox case (secondary contacts).
5. Arrange for immediate vaccination of each primary contact and their household contacts (secondary contacts). Either vaccinate contacts in the household (if this is feasible with the vaccine supply, security issues, and resources), or provide a vaccination ticket with identifying information and designate a vaccination facility for the contact(s) to attend as soon as possible. It is extremely important for smallpox outbreak control to prioritize the

vaccination of contacts. In the past, when vaccination was done in the household, the task was given priority over transportation of a case to an isolation facility.

6. If the primary contact is symptomatic with fever or rash, arrangements should be made for prompt vaccination and transportation of the contact to a Type C facility (capable of isolating contagious individuals) or other designated evaluation site for medical evaluation to rule out smallpox. Contacts with symptoms should be counseled, interviewed, and reported as suspect cases using the appropriate smallpox surveillance (case reporting) form, and their contacts should be identified, interviewed, and vaccinated as soon as possible.
7. If the primary contact does not have a fever or rash, vaccinate or arrange for prompt vaccination, and place the contact under surveillance (quarantine) so that if the contact develops a fever or rash, he/she is immediately isolated and evaluated and does not expose other persons to smallpox (see #8 below).
8. If a household member cannot be vaccinated because of contraindications, the household member should be instructed to avoid physical contact with the primary contact until the incubation period of the disease has passed (19 days) or all vaccinated persons in the household are non-infectious for vaccinia virus (after the scab at the vaccine site has separated, 14-21 days after vaccination).
9. Each household contact should be provided with a vaccination ticket and instructed to attend a designated vaccination clinic site as soon as possible.
10. If any contacts have left the state, the contact tracers should notify the supervisor responsible for out-of-state contacts. The supervisor will then notify the appropriate authorities.

#### **Surveillance (Monitoring) of Health Status and Vaccine “Take” of Contacts**

Surveillance of contacts of cases of smallpox will be conducted for early signs of smallpox disease (fever on two consecutive days and/or rash) and for vaccine “take.” Contacts will be provided with a health department phone number to call if they develop any of the severe vaccine adverse reactions shown on the Vaccine Information Statement.

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✓ **VERSION CONTROL**

Update July 2017. Updated Format. Added Critical Clinician Information section. Updates to Sections: Clinical Description, Differential Diagnosis, Public Health Responsibility.

## ✓ UT-NEDSS MINIMUM/REQUIRED FIELDS BY TAB

### Demographic

- First Name
- Last Name
- Date of Birth
- County
- Birth Gender
- Race
- City
- Street Name
- Zip Code
- Ethnicity
- Area Code
- Phone Number

### Clinical

- Date Diagnosed
- Date of Death
- Died
- Disease
- Onset Date
- Clinician Name
- Clinician Phone

### Laboratory

- Organism
- Result Value
- Test Result
- Test Type
- Lab Test Date

- Specimen sent to State Lab
- Specimen source

### Epidemiological

- Imported From
- Occupation

### Investigation

- Contact ( $\leq 2$  meters) for ( $\geq 3$  hours) with a laboratory confirmed case of smallpox
- Contact ( $\leq 2$  meters) with a confirmed, probable, or suspected case of smallpox
- Contact with smallpox infected bodily fluids or contaminated objects such as bedding or clothing
- Worker in a laboratory that contains smallpox virus

### Reporting

- Date first reported to public health

### Administrative

- State case Status
- Outbreak Associated