Clinical criteria for case classification

Vaccinia disease can present as any number of clinical manifestations ranging from self-limited responses to life-threatening events due to receiving or being inadvertently inoculated with vaccinia as a result of smallpox vaccination.

Clinical complications can include one or more of the following:

- **Eczema vaccinatum**: Characterized by localized or generalized popular, vesicular, or pustular rash, which can occur anywhere on the body, with a predilection for areas of previous atopic dermatitis (e.g., face, forearms, antecubital fossa, popliteal fossa). Rash onset may occur concurrently or shortly after development of the Smallpox vaccine lesion and is often accompanied by fever, malaise, lymphadenopathy and prostration or severe systemic illness.

- **Erythema multiforme major (Stevens-Johnsons Syndrome)**: Characterized by systemic symptoms (fever, malaise, prostration) and involvement of 2 or more mucosal surfaces or 10% of the body surface area.

- **Fetal vaccinia (congenital vaccinia)**: Characterized by skin lesions (e.g., vesicular, pustular, or ulcerative) and/or organ involvement in a newborn. The skin lesions are similar to those of generalized vaccinia or progressive vaccinia and can be confluent and extensive.

- **Post-vaccinial encephalitis or encephalomyelitis**: Characterized by onset of symptoms 6-15 days post-vaccination. Symptoms include any change in mental status (confusion, delirium, drowsiness, restlessness, disorientation, amnesia, seizures, loss of consciousness, coma) or sensorimotor function (altered sensation, weakness, paresis, aphasia, incontinence or urinary retention, obstinate constipation).

- **Progressive vaccinia**: Characterized by a painless progressive and ulcerating lesion at the vaccination site that does not heal, often with central necrosis, and with little or no inflammation.

- **Generalized vaccinia**: Characterized by disseminated maculopapular or vesicular rash, frequently on an erythematous base, usually occurring 6-9 days after first-time vaccination. Lesions may occur on any part of the body, most often on the trunk and abdomen, less commonly on the face and limbs. Though usually benign and self-limiting, can develop into severe systemic illness.

- **Inadvertent inoculation**: Characterized by extensive vesicular and pustular lesion(s) at a distant different location on the vaccinee, or anywhere on a close contact, which is not generalized but may involve a large contiguous area.

- **Ocular vaccinia**: Characterized by inflammation of peri-ocular soft tissue or the eye itself (blepharitis, conjunctivitis, keratitis, iritis).

- **Pyogenic (staphylococcal) infection**: Characterized by vesiculo-pustular lesion at the site of vaccination, often spreading peripherally in circumferential fashion, with clearing behind the advancing border. Bacterial lymphangitis and regional lymphadenitis may occur, but most often the lesions are solely superficial infections.
• **Streptococcal infections:** Characterized by a piled up eschar, heaping at the vaccination site. Lymphangitis occurs commonly as does edematous painful regional lymphadenitis.

• **Enteric and anaerobic infections:** Characterized by purulence with or without extensive necrosis at the vaccination site. Necrotic fasciitis has also been encountered in some cases.

• **Other serious adverse events:** Serious to life-threatening events resulting in hospitalization, permanent disability, life-threatening illness, or death in a Smallpox vaccinee, or a close contact of a vaccinee.

**Laboratory criteria for case classification**
None unless laboratory confirmation is indicated to distinguish from other infections or other pox.

**Epidemiological criteria for case classification**
Not applicable.

**Case classification**

**Probable:**
A person with clinical features compatible with the diagnosis where other causes are excluded and supportive information is available.

**Suspect:**
Either of the following:
- A person with clinical features compatible with the diagnosis where further investigation is required
- Or a person with clinical features compatible with the diagnosis where additional investigation of the case did not provide supporting evidence for the diagnosis and did not identify an alternative diagnosis.

**Criteria to distinguish a new case from previous reports**
Not applicable.