

## EVENT DEFINITION FORM

**Event:** Chilblain – like lesions

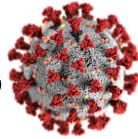
**Outcome/covariate:** **outcome**

**Version:** 1.0

**Status:** final

### Contributing authors

authors	Role	Date
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Miriam Sturkenboom	Codemapper review	August 13, 2020
Leila Belbachir	Medical review	August 22, 2020
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Carlos Durán	Rev. narrow/possible assignment	29-03-2021
Miriam Sturkenboom	Inclusion of codes used in final ACCESS report	23-08-2021



## 1. Event definition

There is not yet an internationally clinical definition and guidelines for chilblains and chilblain-like lesions.

### Chilblains<sup>[1]</sup>

Chilblains, also referred to as perniosis or pernio, is a condition of the skin which manifests as erythematous to violaceous macules, papules, plaques, or nodules in sites of cold exposure and damp environments (idiopathic chilblains). The most common sites for involvement are the fingers and toes and is frequently accompanied by a sensation of itching, burning, or pain. It is postulated that pernio results from an abnormal vascular response to cold exposure. Cold-induced vasoconstriction of vasospasm resulting in hypoxemia that stimulates an inflammatory response is potential mechanism for the formation of skin lesions. There are reports of an causative correlation between systemic diseases and chilblains. The most frequent reported and studied relationship is the relationship between (chronic) chilblains and lupus erythematosus also known as 'chilblain lupus erythematosus (CHLE)'.

### Chilblain-like lesions

During the recent COVID-19 pandemic patients with little or no symptoms presented themselves with chilblain-like lesions located on the toes and fingers. These patients had no underlying autoimmune disease (such as lupus erythematosus), Raynaud's phenomenon or previous episodes of idiopathic chilblains. It mostly affected children and young adults and the lesions took place later in the course of the (suspected) COVID-19 disease. The chilblain-like lesions manifests as multiple red-violaceous edematous lesions with papules and macules located on acral regions such as toes, the feet (heel, sole) and/or the fingers, asymptomatic or associated with pruritis of mild pain. Because of the presentation similar with chilblains it is referred to as pseudo-chilblain of chilblain-like lesions. The lesions disappeared after a few weeks without treatment.<sup>[2][3][4][5][6][7]</sup>

### *Signs and symptoms chilblains*

- Edematous, erythematous to violaceous macules, papules, plaques, or nodules, particularly located on acral site
- Onset associated with cold exposure
- Greatest severity of symptoms during cold months of the year
- Improvement with elimination of cold exposure (however, chronic disease courses do not rule out pernio)
- Acute, idiopathic, cases of pernio typically resolve within one to three weeks
- Chronic pernio (as a result of a systemic disease) may manifest as recurrent acute episodes or episodes that persist for longer than several weeks

## 2. Synonyms / lay terms for the event

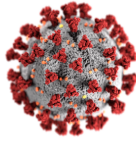
Perniosis

Pernio

Perniones

Covid toes

Acro-ischemic lesions



Acro-ischemia  
Acral chilblain-like lesions  
Pseudo chilblain

### 3. Laboratory tests that are specific for event<sup>[8][9][10][11]</sup>

There are no serologic tests that confirm a diagnosis of pernio. Laboratory investigations are reserved for the evaluation for concomitant medical disorders. Thus, for patients with chronic (lasting more than several weeks) or recurrent pernio and patients with other signs or symptoms that suggest the possibility of an underlying systemic diseases.

laboratory work-up may include:

- Complete blood count
- Serum protein electrophoresis with immunofixation
- Antinuclear antibody
- Inflammatory markers
- D-dimers
- Antinuclear antibodies
- Complement
- Cryoglobulines
- Antiphospholipid antibodies
- Coagulation study results (PT, PTT and trombin time)
- Renal function
- Hepatic function
- Thyroid function
- Serologic testing

### 4. Diagnostic tests that are specific for event<sup>[12]</sup>

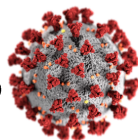
#### 1. Skin biopsy

A biopsy primarily serves to confirm the presence of features consistent with pernio or to detect histologic evidence that suggests a different disorder. There are no pathognomonic histopathologic findings of pernio. A diagnosis of pernio is supported by the detection of the following histopathological features: papillary dermal edema and superficial and deep, moderate to dense lymphocytic infiltrate surrounding blood vessels and eccrine glands. necrotic keratinocytes and lymphocytic vasculitis (edema of vessel walls and lymphocytic infiltration of vessel walls) may be present.

The possibility of chilblain lupus erythematosus is suggested by the detection of vacuolization of the basal layer of the epidermis in addition to the typical findings of pernio

#### 2. Capillaroscopy

Increased nailfold capillary tortuosity and diameter as well as increased apical capillary diameter have been reported in patients with idiopathic pernio.



## 5. Drugs that are used to treat event

None

## 6. Procedures used specific for event treatment

In clinical practice, medium to high potency topical corticosteroids are sometimes prescribed in an attempt to hasten resolution of pernio lesions. Randomized trials to confirm the efficacy of local corticosteroid therapy are lacking.

## 7. Setting (outpatient specialist, in-hospital, GP, emergency room) where condition will be most frequently /reliably diagnosed

Outpatient specialist (dermatologist) or GP

## 8. Diagnosis codes or algorithms used in different papers to extract the events in Europe/USA: seek literature for papers that have studied this event, and see how they extracted/measured the event.

ICD-10-CM: T69.1

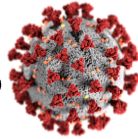
- T69.1XXA (initial encounter)
- T69.1XXD (subsequent encounter)
- T69.1XXS (sequela)

ICD-9-CM

- 991.5

## 9. Proposed codes by Codemapper

Coding system	Code	Code name	Concept	Concept name	Algorithm
ICD10/CM	T69.1	Chilblains	C0008058	Chilblains	Narrow
ICD10/CM	T69.1XXS	Chilblains, sequela	C2886283	Chilblains, sequela	Possible
ICD9CM	991.5	Chilblains	C0008058	Chilblains	Narrow
ICPC2P	A88001	Chilblains	C0008058	Chilblains	Narrow
ICPC2P	A88010	Pernio	C0008058	Chilblains	Narrow
RCD2	SN15.	Chilblains	C0008058	Chilblains	Narrow
SCTSPA	37869000	sabañones	C0008058	Chilblains	Narrow
SCTSPA	238509006	sabañón pustular	C0349505	Pustular chilblain	Narrow
SNOMEDCT_US	37869000	Chilblains	C0008058	Chilblains	Narrow
SNOMEDCT_US	157719006	Chilblains	C0008058	Chilblains	Narrow
SNOMEDCT_US	238509006	Pustular chilblain	C0349505	Pustular chilblain	Narrow
SNOMEDCT_US	269421007	Chilblains	C0008058	Chilblains	Narrow



## 10. Algorithm proposal

### **Broad Algorithm:**

- Concept sets (Chilblains, Chilblains\_sequela)
- Index date = first occurrence of a code from the concept sets (Chilblains, Chilblains\_sequela)

### **Narrow Algorithm:**

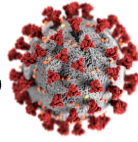
- Concept set Chilblains
- Index date = first occurrence of a code from the concept set Chilblains

## 11. Background rates

none

## 12. References

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8. Kanitakis J, Lesort C, Danset M, Jullien D, CHILBLAIN-LIKE ACRAL LESIONS DURING THE COVID-19 PANDEMIC ("COVID TOES"): HISTOLOGIC, IMMUNOFLUORESCENCE AND IMMUNOHISTOCHEMICAL STUDY OF 17 CASES *Journal of the American Academy of Dermatology* (2020)
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10. Pernio (chilblains) [internet]. [Accessed on 18<sup>th</sup> of june]. Available from: [https://www-uptodate-com.proxy.library.uu.nl/contents/pernio-chilblains?search=pernio&source=search\\_result&selectedTitle=1~30&usage\\_type=default&display\\_rank=1#H692994429](https://www-uptodate-com.proxy.library.uu.nl/contents/pernio-chilblains?search=pernio&source=search_result&selectedTitle=1~30&usage_type=default&display_rank=1#H692994429)
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