

The Representation of Skin Diseases in the 11th Revision of the International Classification of Diseases (ICD-11)

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Previous ICD Revisions

- ❑ 10th Revision of ICD (ICD-10) drawn up in the 1980s without the benefit of email or modern electronic communication
- ❑ Dermatology input into revision largely conducted by just two dermatologists (Mervyn Elgart and Alan Shrank)
- ❑ Very little opportunity given to make substantive changes from previous revisions
- ❑ The revisions accepted by WHO were largely inconsequential (e.g., lichen striatus, cutis rhomboidalis nuchae)

ICD-11

- ❑ Planning for ICD-11 was started in 2007 in a collaboration between WHO and bioinformatics experts from the US
- ❑ Topic Advisory Groups were assembled in 2009 to oversee revision of specific domains including dermatology
- ❑ RC (Manchester, UK) and Michael Weichenthal (Kiel, Germany) appointed as co-chairs
- ❑ ILDS generously supported RC to spend 8 months working with WHO in Geneva at the outset of the revision process

What is the ICDD*?

- ❑ A detailed dermatologically-focused classification of skin diseases
- ❑ Fully integrated into the 11th Revision of the WHO International Classification of Diseases (ICD-11)
- ❑ One of a family of classifications extracted from the common WHOFIC** Foundation, including the replacement of ICD-10, the ICD-11 for Mortality and Morbidity Statistics (ICD-11 MMS)

* internet search string: [Dermatology ICD-11](#)

** WHO Family of International Classifications

Top level categories in the Diseases of the Skin chapter of WHOFIC Foundation

- ▼ Diseases of the skin
 - ▶ Certain skin disorders attributable to infection or infestation
 - ▶ Inflammatory dermatoses
 - ▶ Metabolic and nutritional disorders affecting the skin
 - ▶ Genetic and developmental disorders affecting the skin
 - ▶ Sensory and psychological disorders affecting the skin
 - ▶ Skin disorders involving specific cutaneous structures
 - ▶ Skin disorders involving certain specific body regions
 - ▶ Skin disorders associated with pregnancy, the neonatal period and infancy
 - ▶ Adverse cutaneous reactions to medication
 - ▶ Skin disorders provoked by external factors
 - ▶ Benign proliferations, neoplasms and cysts of the skin
 - ▶ Disorders of the skin of uncertain or unpredictable malignant potential
 - ▶ Malignant neoplasms involving the skin
 - ▶ Cutaneous markers of internal disorders
 - ▶ Postprocedural disorders of the skin
 - ▶ Adverse cutaneous effects of healthcare related interventions
 - ▶ Impairment of normal functioning resulting from skin disease
 - ▶ Symptoms or signs involving the skin
 - ▶ Absence of skin condition

Top level categories in the Diseases of the Skin chapters

ICD-11 MMS

- ▼ 14 Diseases of the skin
 - ▶ Certain skin disorders attributable to infection or infestation
 - ▶ Inflammatory dermatoses
 - ▶ Metabolic and nutritional disorders affecting the skin
 - ▶ Genetic and developmental disorders affecting the skin
 - ▶ Sensory and psychological disorders affecting the skin
 - ▶ Skin disorders involving specific cutaneous structures
 - ▶ Skin disorders involving certain specific body regions
 - ▶ Skin disorders associated with pregnancy, the neonatal period and infancy
 - ▶ Adverse cutaneous reactions to medication
 - ▶ Skin disorders provoked by external factors
 - ▶ Benign proliferations, neoplasms and cysts of the skin
 - ▶ Disorders of the skin of uncertain or unpredictable malignant potential
 - ▶ Cutaneous markers of internal disorders
 - ▶ Postprocedural disorders of the skin
 - ▶ Malignant neoplasms involving the skin
 - NE81.00 Haematoma of surgical wound of skin
 - NE81.20 Superficial incisional site infection
 - KC50 Neonatal phototherapy burn
 - ▶ Symptoms or signs involving the skin
 - EM0Y Other specified diseases of the skin
 - EM0Z Skin disease of unspecified nature

ICDD

- ▼ 14 Diseases of the skin
 - ▶ Certain skin disorders attributable to infection or infestation
 - ▶ Inflammatory dermatoses
 - ▶ Metabolic and nutritional disorders affecting the skin
 - ▶ Genetic and developmental disorders affecting the skin
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 - ▶ Disorders of the skin of uncertain or unpredictable malignant potential
 - ▶ Cutaneous markers of internal disorders
 - ▶ Postprocedural disorders of the skin
 - ▶ Adverse cutaneous effects of healthcare related interventions
 - Impairment of normal functioning resulting from skin disease (EX4H_{DER})
 - ▶ Absence of skin condition
 - ▶ Malignant neoplasms involving the skin
 - ▶ Symptoms or signs involving the skin
 - Other specified diseases of the skin (EM0Y_{DER})
 - Skin disease of unspecified nature (EM0Z)

ICD-11 MMS and ICDD compared

- ❑ There is a considerably larger number of dermatological diagnoses incorporated into the ICD-11 MMS compared with ICD-10
- ❑ ICD-11 MMS may be sufficient for everyday dermatological practice

- ▼ EH90 Pressure ulceration

- EH90.0 Pressure ulceration grade 1

- EH90.1 Pressure ulceration grade 2

- EH90.2 Pressure ulceration grade 3

- EH90.3 Pressure ulceration grade 4

- EH90.4 Suspected deep pressure-induced tissue damage, depth unknown

- EH90.5 Pressure ulceration, ungradable

- EH90.Z Pressure ulcer of unspecified grade**

- ▼ Drug eruptions

- EH60 Exanthematic drug eruption

- ▼ EH61 Drug-induced urticaria, angioedema and anaphylaxis

- EH61.0 Drug-induced urticaria

- EH61.1 Drug-induced angioedema

- 4A84.1 Drug-induced anaphylaxis

- EH62 Lichenoid drug eruption

- ▼ EH63 Stevens-Johnson syndrome and toxic epidermal necrolysis due to drug

- EH63.0 Drug-induced Stevens-Johnson syndrome

- EH63.1 Drug-induced toxic epidermal necrolysis

- EH63.2 Drug-induced Stevens-Johnson and toxic epidermal necrolysis overlap syndrome

- EH64 Drug-induced erythroderma

- EH65 DRESS syndrome

- EH66 Fixed drug eruption

- ▶ EH67 Acne or acneform reactions attributable to drugs

- EH6Y Drug eruption of other specified type**

- EH6Z Drug eruption of unspecified type**

- ❑ The ICDD is directed more at academic centres and specialist services

ICD-11 MMS and ICDD compared

ICD-11 MMS

▼ EB61 Morphoea

EB61.0 Plaque morphoea

EB61.1 Linear morphoea

EB61.Y Other specified forms of morphoea

- ❑ Note extra detail in ICDD
- ❑ Note cross-references to ICD-11 MMS

Foundation URI : <http://id.who.int/icd/entity/1430740369>

Plaque morphoea (EB61.0)

ICDD

▼ Morphoea (EB61)

Plaque morphoea (EB61.0)

Generalised morphoea (EB61_DER)

Linear morphoea (EB61.1)

Progressive hemifacial atrophy (EB61_DER)

Disabling pansclerotic morphoea of childhood (EB61_DER)

Morphoea profunda (EB61_DER)

Extragenital lichen sclerosis with morphoea (EB60_DER)

Atrophoderma of Pasini and Pierini (EW8D_DER)

Other specified forms of morphoea (EB61_DER)

Example of detailed content in ICDD

- ▼ Disorders of skin colour
 - ▼ Acquired hypermelanosis
 - ▼ Physiological hypermelanosis (ED60.0)
 - Suntan (ED60.00)
 - Tanning due to exposure to artificial sources of ultraviolet radiation (ED60.01)
 - Melasma (ED60.1)
 - Hypermelanosis due to endocrine disorder (EW6M_DER)
 - Postinflammatory hypermelanosis (ED60.2)
 - Paraneoplastic hypermelanosis (EW6N_DER)
 - Ashy dermatosis (EW6P_DER)**
 - Post traumatic hypermelanosis (EW6Q_DER)
 - Drug-induced hypermelanosis (EH70_DER)
 - Labial melanin incontinence (DV9S_DER)
 - Oral melanin incontinence (DW05_DER)
 - Occupational melanosis (EX30_DER)
 - Hypermelanosis of other specified aetiology (ED60_DER)
 - Hypermelanosis of unspecified aetiology (ED60.Z)
 - ▶ Acquired melanotic macules or lentigines
 - ▶ Endogenous non-melanin pigmentation
 - ▶ Non-melanin pigmentation due to ingested or injected substance
 - ▶ Non-melanin pigmentation due to exogenous substances
 - ▶ Acquired hypomelanotic disorders
 - ▶ Acquired mixed hyper- and hypomelanotic disorders of skin pigmentation
 - Abnormal skin pigmentation (ED64)
 - ▶ Genetic disorders of skin pigmentation



Foundation URI : <http://id.who.int/icd/entity/974973785> [Link to iCat](#)



Ashy dermatosis (EW6P_DER)

Parent

Acquired hypermelanosis

Show all ancestors

Description

Ashy dermatosis is an asymptomatic disorder of skin pigmentation of disputed aetiology characterised by widespread persistent and often symmetrical pale slate-blue macules in healthy individuals. It shares both clinical and histological features with lichen planus but mucous membranes are unaffected. Preceding erythema around the margins of the pigmented macules may be noted in a minority of patients, in which case the disorder may be referred to as erythema dyschromicum perstans. It has been described principally from Central and South America and from Asia.

All Index Terms

- Ashy dermatosis
- Ashy dermatosis of Ramirez
- Dermatosi cenicienta
- Erythema chronicum figuratum melanodermicum
- Idiopathic eruptive macular pigmentation
- Erythema dyschromicum perstans

Hide index terms



Caveats

Taking a closer look at the ICDD

ICDD Chapter 02 Neoplasms

- ▼ 02 Neoplasms
 - ▶ Neoplasms of haematopoietic or lymphoid tissues
 - ▼ Malignant mesenchymal neoplasms
 - Dermatofibrosarcoma protuberans (2V3X_DER)
 - ▼ Angiosarcoma of skin (2B56.1)
 - Epithelioid haemangioendothelioma of skin (2B56_DER)
 - Retiform haemangioendothelioma of skin (2B56_DER)
 - Other specified angiosarcoma of skin (2B56_DER)
 - Angiosarcoma of skin, unspecified (2B56_DER)
 - Kaposi sarcoma of skin (2B57.1)
 - Cutaneous leiomyosarcoma (2V3Y_DER)
 - Other specified malignant mesenchymal neoplasms (2B5Y_DER) *
 - Malignant mesenchymal neoplasm of unspecified type (2B5Z)

* contains 313 entries irrelevant to skin

ICDD Chapter 14 Diseases of the skin

- ▼ Malignant neoplasms of skin
 - ▶ Melanoma of skin
 - ▶ Squamous cell carcinoma of skin
 - ▶ Basal cell carcinoma of skin
 - ▶ Adnexal carcinoma of skin
 - ▶ Cutaneous neuroendocrine carcinoma
 - Radiotherapy-induced skin malignancy (2V48_DER)
- ▼ Cutaneous sarcoma
 - ▶ Angiosarcoma of skin (2B56.1)
 - Cutaneous leiomyosarcoma (2V3Y_DER)
 - Dermatofibrosarcoma protuberans (2V3X_DER)
 - Kaposi sarcoma of skin (2B57.1)
 - Other specified malignant neoplasms of skin (2C3Y_DER)
 - Malignant neoplasm of skin of unknown or unspecified type (2C3Z)
- ▶ Cutaneous markers of internal malignancy
- ▶ Metastatic malignant neoplasm involving skin

Taking a closer look at the ICDD

▼ Dermatology Speciality Linearization

- ▶ 01 Certain infectious or parasitic diseases
- ▼ 02 Neoplasms
 - ▶ Neoplasms of haematopoietic or lymphoid tissues
 - ▼ Malignant mesenchymal neoplasms
 - Dermatofibrosarcoma protuberans (2V3X_{DER})
 - ▶ Angiosarcoma of skin (2B56.1)
 - Kaposi sarcoma of skin (2B57.1)
 - Cutaneous leiomyosarcoma (2V3Y_{DER})
 - Other specified malignant mesenchymal neoplasms (2B5Y_{DER})
 - Malignant mesenchymal neoplasm of unspecified type (2B5Z)
 - ▶ Malignant neoplasms of skin
 - ▶ Malignant neoplasms of female genital organs
 - ▶ Malignant neoplasms of male genital organs
 - ▶ Malignant neoplasms of kidney, except renal pelvis
 - ▶ Metastatic malignant neoplasm involving skin
 - ▶ In situ neoplasms, except of lymphoid, haematopoietic, central nervous system or related tissues
 - ▶ Benign mesenchymal neoplasms

Other specified malignant mesenchymal neoplasms (2B5Y_{DER})

Parent

Malignant mesenchymal neoplasms

Show all ancestors ↕

This category is an 'other specified' residual category

All Index Terms

- Other specified malignant mesenchymal neoplasms Hide index terms ⌕
- Chondrosarcoma, primary site ⇒
 - chondrosarcoma of unspecified site
 - cartilage cancer
 - chondrosarcoma NOS
- Extraskelatal myxoid chondrosarcoma of other specified sites ⇒
- Extraskelatal myxoid chondrosarcoma of soft tissues of limb ⇒
- Extraskelatal myxoid chondrosarcoma, site unknown ⇒
 - myxoid chondrosarcoma of unspecified site
 - myxochondrosarcoma of unspecified site
- Chondrosarcoma of bone or articular cartilage of limbs ⇒
- Chondrosarcoma of bone or articular cartilage of pelvis ⇒

Adding detail

- Malignant neoplasms of digestive organs
- Malignant neoplasms of middle ear, respiratory or intrathoracic organs
- ▼ Malignant neoplasms of skin
 - 2C30 Melanoma of skin
 - 2C31 Squamous cell carcinoma of skin
 - ▼ 2C32 Basal cell carcinoma of skin
 - 2C32.0 Nodular basal cell carcinoma of skin
 - 2C32.1 Sclerosing basal cell carcinoma of skin
 - 2C32.2 Superficial basal cell carcinoma of skin
 - 2C32.Y Other specified basal cell carcinoma of skin
 - 2C32.Z Basal cell carcinoma of skin, unspecified
 - 2C33 Adnexal carcinoma of skin
 - 2C34 Cutaneous neuroendocrine carcinoma
 - 2C35 Cutaneous sarcoma
 - 2C3Y Other specified malignant neoplasms of skin
 - 2C3Z Malignant neoplasm of skin of

Postcoordination ?

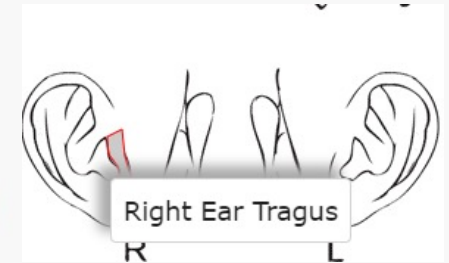
2C32.1 Sclerosing basal cell carcinoma of skin

Code: 2C32.1

Add detail to **Sclerosing basal cell carcinoma of skin**

Laterality (use additional code, if desired)

XK9J	Bilateral
XK8G	Left
XK9K	Right
XK70	Unilateral, unspecified



Specific anatomy (use additional code, if desired)

Search

tragus



Has manifestation

MG30.10

Associated with (use additional code, if desired)

XB17	Occupation as primary factor
XB5G	Occupation as cofactor
XB80	Not occupation-related

Search results Specific anatomy

XA2N71 **Tragus** of pinna

Adding detail

- ❑ The various domains which can be added include body site, occupational relevance and aetiological agents such as drugs, contact allergens, poisons etc.
- ❑ The ampersand (&) links the additional information to the primary code

▼ 2C32 Basal cell carcinoma of skin

2C32.0 Nodular basal cell carcinoma of skin

2C32.1 Sclerosing basal cell carcinoma of skin

2C32.2 Superficial basal cell carcinoma of skin

2C32.Y Other specified basal cell carcinoma of skin

2C32.Z Basal cell carcinoma of skin, unspecified

Postcoordination ?

2C32.1 Sclerosing basal cell carcinoma of skin

- Associated with
- Laterality
- Specific anatomy

Code: 2C32.1&XB5G&XK9K&XA2N71

XB5G Occupation as cofactor ✕

XK9K Right ✕

XA2N71 Tragus of pinna ✕

Adding optional detail

▼ EK00 Allergic contact dermatitis

EK00.0 Allergic contact dermatitis due to clothing or footwear

EK00.1 Allergic contact dermatitis due to cosmetics or fragrances

EK00.2 Allergic contact dermatitis due to dental materials

EK00.3 Allergic contact dermatitis due to food flavours or additives

EK00.4 Allergic contact dermatitis due to hairdressing products

EK00.5 Allergic contact dermatitis due to industrial biocides, cutting oils or disinfectants

EK00.6 Allergic contact dermatitis due to metals or metal salts

EK00.7 Allergic contact dermatitis due to allergenic haptens derived from plants or organic matter

EK00.8 Allergic contact dermatitis due to plastics, glues or resin systems

EK00.9 Allergic contact dermatitis due to preservatives or biocides

EK00.A Allergic contact dermatitis due to rubber

- ❑ The various domains which can be added include body site, occupational relevance and aetiological agents such as drugs, contact allergens, poisons etc.
- ❑ The ampersand (&) links the additional information to the primary code
- ❑ Each element can be searched for independently

Code: EK00.4&XB17&XM8RR9

Postcoordination ?

EK00.4 Allergic contact dermatitis due to hairdressing products

Code: EK00.4&XB17&XM8RR9

└ Associated with

XB17 Occupation as primary factor ✖

XM8RR9 Ammonium persulfate ✖

Add detail to **Allergic contact dermatitis due to hairdressing products**

Specific anatomy (use additional code, if desired)

Search

Associated with (use additional code, if desired)

Search

Search results Associated with

XM8RR9 **Ammonium** persulfate

Field-Testing of ICD-11 MMS and ICDD

16 ILDS-affiliated National societies submitted a total of 79 reports reviewing the scope and utility of ICD-11 for coding skin disorders

Number of patients included	3,829
No. of patients codable from MMS	3,612 (94.3%)
No. of patients better coded from Foundation	122 (3.2%)
Total number satisfactorily coded	3,734 (97.5%)
Diagnostic concept present in ICD-11	3,782 (98.8%)
Diagnosis present but not found by field-tester	52 (1.3%)
Index term missing	25 (0.7%)
Concept missing	14 (0.4%)
Not suitable for coding as diagnosis	20 (0.5%)

Specialist review of ICDD

- ❑ Over the past few months ILDS has commissioned reviews of the ICDD from recognized specialist experts but the findings of these have not yet been analysed.
- ❑ We are under no illusions that our task is finished and my successor as dermatology representative of the ICD Medical and Scientific Advisory Committee, Professor Jonathan White, will continue to have plenty of work to do.

Moving forward

- ❑ Further WHO assistance in enhancing functionality of ICDD
 - ❑ Continuing refinement of the classification
 - ❑ Post-coordination to be options added
 - ❑ Providing a better interface with other systems using the ICD API
- ❑ A hope that colleagues in the US and elsewhere would wish to assist in devising the means to incorporate ICDD into their data systems for the eventual benefit of patients with skin disease



Internet Links

ICDD (Dermatology Specialty Linearization): <https://icd.who.int/dev11/l-derma/en>

Internet search : **Dermatology ICD-11**

Email contact: r.chalmers@manchester.ac.uk

Key References

The WHO ICD-11 Classification of Dermatological Diseases: a new comprehensive online skin disease taxonomy designed by and for dermatologists

J.M.L. White, H. Lui, C.G. Chute, R. Jakob, R.J.G. Chalmers

Br Journal Dermatol 2021 (online pre-print version: doi.org/10.1111/bjd.20656)

Developing an international standard for the classification of surface anatomic location for use in clinical practice and epidemiologic research

K.A. Kenneweg, A.C. Halpern, R.J. G. Chalmers, P. Soyer, M. Weichenthal, M.A. Molenda
J Am Acad Dermatol. 2019; **80**:1564-1584. [doi: 10.1016/j.jaad.2018.08.035](https://doi.org/10.1016/j.jaad.2018.08.035)