

ICD-11 revision process for Rare Diseases

Diseases of the blood and blood-forming organs

(ICD-10 chapter III, codes D50-D77)

Draft structure n°2

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Introduction and table of contents

You are kindly invited to participate to the World Health Organisation's *International Classification of Diseases* (ICD) revision process. The following document will help you in making your comments. You will find:

1. The rationale and the general methodology of the ICD revision for rare diseases
2. The ICD-11 draft structure for *Haematological diseases*, which represents Orphanet's proposal for a new ICD.
3. Groups of diseases included in the haematological section of ICD10, but that should be moved to another chapter in ICD11.

This second draft document incorporates revisions suggested by experts that reviewed the first version. It is intended to be a **validation step for final corrections** before the draft is sent to the World Health Organization. You are invited to:

1. Check the ICD-11 draft structure indicating whether there are any errors new global structure and the way rare diseases are represented in it. (See the checklist of issues to be addressed on page 5.)
2. Send your feed-back to Orphanet.
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3. Disseminate this invitation to your colleagues who are experts in this field.

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1. Rationale and general methodology

WHO has established various Topic Advisory Groups to serve as planning and coordinating advisory bodies in the update and revision process for specific areas. A Revision Steering Group oversees the overall revision process. An internet-based workspace documents revision proposals that obtain evidence from analysis of available data. WHO collaborates through this platform with all interested parties. Working groups organized by the Topic Advisory Groups (TAG) review the proposals. To learn more about the whole revision process:

<https://extranet.who.int/icdrevision/help/docs/ICDRevision.pdf>

A TAG for rare diseases was established in April 2007 as rare diseases should now be traceable in mortality and morbidity information systems. The production of the basic information to establish a first draft of the classification of rare diseases has been assigned to Orphanet and will contribute to the whole revision process, as rare diseases involve all areas of medicine.

The workplan is as follows:

1. Proposals from TAG for a new ICD structure for this chapter are expected before April 2010.
2. A decision about it, after compilation of all proposals, should be taken in April 2010. This will define the category layers based on consensus hierarchies, called the Alpha version.
3. Work on the Beta version will then start at TAG level to populate the model and finalise the proposals.

In order to prepare the ICD revision process, Orphanet has collected a series of rare diseases classifications mainly based on scientific grounds (aetiology and mechanism). To complement these classifications, Orphanet has developed a strictly clinical in-house classification to meet the needs of the clinicians. All the classifications can be viewed on the Orphanet website. They now serve as a basis to build the ICD-11 proposals. For an overview on the general methodology of Orphanet classification:

<http://www.orpha.net/data/patho/Pro/en/OrphanetClassificationRareDiseases.pdf>

Orphanet is a comprehensive peer-reviewed database of information on rare diseases. Over 5,800 are inventoried, and the database of diseases is updated monthly according to the evolution of knowledge. Each Orphanet entry is indexed with MeSH terms, Orphanet thesaurus of clinical signs and symptoms, ICD-10 codes, and linked to the OMIM database, to an in-house genes database and to PubMed as well as to other websites of interest. For each Orphanet entry there is an identity card with epidemiological data (prevalence rank, mode of inheritance, age of onset) and a set of synonyms. Orphanet produces a peer-reviewed encyclopaedia covering more than 2,600 entries and updated continuously.

2. General principles for ICD revision

The current ICD10 classification is monoaxial, meaning that every entity can figure only at one point in the classification. The rationale for this choice is to avoid double counting, since the ICD is primarily used as a statistic tool. This is a problem however for numerous diseases that can be associated with more than one body system (chapters being broadly organised along them). In such cases, one system must then be given priority, and *exclusion notes* are put in the other relevant chapters to redirect users to the correct code.

In the future ICD11, the classification shall become polyaxial, and every entity shall be assigned a unique identifying number: diseases will be able to figure in all relevant places in the classification (for instance, anaemias associated with organic acidurias will figure among both metabolic and haematological diseases). This system will be fully operational in the electronic version of the future ICD. However, in the paper version, it will still be necessary for space reasons to keep the current monoaxial system; and for statistics, it is still necessary to avoid double counting. Therefore, **the ICD11 will also feature linearisations, i.e. versions allowing for a monoxial approach.** We suggest that the priority specialty should be related to the body system most severely affected by the disease and/or the specialist most likely to be relied on for the management of the disease. In a number of cases however, the choice is questionable and ultimately quite arbitrary. For multisystemic diseases, Orphanet supports the creation of a new dedicated chapter in ICD11.

As a result of the polyaxial nature of the ICD11 coupled with linearisations, many former exclusion notes are no longer necessary and can be converted into inclusions in all relevant places. A few exclusion notes, however, fulfill a different function: they clarify the range of content of particular codes, or remove possible ambiguities. For instance, crossed exclusion notes are used in this proposal between the codes for typical and atypical forms of haemolytic-uraemic syndromes, which are included in different parts of the classification. Such clarifying exclusion notes are to be kept in ICD11.

The Orphanet proposal for a new ICD classification follows a clinical rather than aetiological approach. Groups of diseases are preferentially defined on the basis of shared clinical features. When several possible names are possible for a disease, descriptive names formed in accordance with a clinical approach are to be preferred. The WHO style guide recommends to avoid eponyms in disease nomenclature, except when their use is dominant and well established (e.g. Alzheimer disease, Huntington disease, etc.).

The proposal regularly includes "**default codes**" for conditions without a specific code, or without enough information to allow proper coding. Currently, there are such default codes in every section of the ICD10 for specified conditions (codes ending in .8) and unspecified conditions (codes ending in .9). Those sections will be kept in the future ICD. **We recommend that similar default codes should be created for rare diseases specifically,** to allow them to be identified as such in order to improve statistics about rare diseases.

3. Specific issues for haematological diseases revision

The current ICD10 classification of haematological diseases is covered by the first part of chapter III, made of the block of codes *D50-D77 Diseases of the blood and blood forming organs*.

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As far as possible, we try to harmonise denominations to focus on clinical haematological manifestations. Nevertheless, this is not always possible for several entities that must stand in other chapters, because their name must be the same everywhere; in such cases a looser denomination is often necessary. For instance, we must use *hereditary orotic aciduria* rather than *anaemia due to hereditary orotic aciduria* because the disease must also stand in the chapter for metabolic diseases, where focusing on anaemia rather than other manifestations would be inappropriate.

In the part concerning deficiency anaemias (currently grouped under codes *D50-D55 Nutritional anaemias*), this draft proposal systematically distinguishes constitutional and acquired conditions, contrasting with the current ICD10.

We also included in this chapter a classification of haematological malignancies, adapted from the WHO reference classification on this topic:

World Health Organization (WHO), International agency for research on cancer (IARC). *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues. 4th ed.*, edited by S.H. Swerdlow, E. Campo, E., N.L. Harris, E.S. Jaffe, S.A. Pileri, H. Stein, J. Thiele, J.W. Vardiman. Lyon : IARC, 2008. 439 p. ISBN 978-92-832-2431-0

The following classification was also used:

Haemacare: *Proposal for grouping Hematological Malignancies*, Rarecare workshop, Treviso, 2008

This section has significantly more levels than the others, due to the greater need for detail in oncology. It should also be included also in the future ICD chapter for oncology.

4. Checklist for reviewing this revision proposal

- Is the revised structure of the chapter scientifically correct?
- Is it useful or relevant considering the expected uses of the ICD11?
 - Mortality reporting
 - Morbidity reporting
 - Clinical practice
 - Research practice
 - Primary care
 - Public Health Reporting
- Are rare diseases properly represented in the new structure?
- Are there rare diseases lacking in the new structure?
- Are there entities of doubtful status in the new structure?
- Are there entities that could be improperly understood by coders because of their denomination or their place in the hierarchy?
- For diseases that can be included in several medical specialties, do you agree with the choice of the priority specialty for the linearised version of the ICD11?

**ICD-11 revised draft structure
for
Haematological diseases**

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Legend :

Items highlighted in grey: additions to the current ICD10

Items in italics: synonym of the term just above

Items in bold: main divisions of the ICD11 table draft

- ICD10 codes are indicated when there is a corresponding entry in the current ICD10 tabular list.
- Some groups of non-rare disease have been directly transposed from ICD10 and were not revised in-depth.

<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Anaemias			We suggest to harmonise all titles on the following model : Constitutional/acquired anaemia(s) due to [etc.]
Constitutional deficiency anaemias			
Constitutional anaemias due to iron metabolism disorder	D50		
Excludes: Anaemia due to nutritional iron deficiency			
Atransferrinaemia			
Microcytic anaemia with liver iron overload			
Iron-refractory iron deficiency anaemia			
<i>IRIDA syndrome</i>			
Other specified rare constitutional anaemias due to iron metabolism disorder			
Constitutional anaemias due to iron metabolism disorder, unspecified			
Constitutional megaloblastic anaemias due to vitamin B12 metabolism disorder	D51		
Vitamin B12 deficiency anaemia due to congenital intrinsic factor deficiency	D51.0		
<i>Congenital pernicious anaemia</i>			
Vitamin B12 deficiency anaemia due to selective vitamin B12 malabsorption with proteinuria	D51.1		
<i>Imerslund(-Gräsbeck) syndrome</i>			
Congenital megaloblastic anaemia due to transcobalamin II deficiency	D51.2		
Methylcobalamin deficiency type cbl E		metabolic diseases	
Methylcobalamin deficiency type cbl G		metabolic diseases	
Methylmalonic acidaemia - homocystinuria		metabolic diseases	
Methylmalonicacidaemia - homocystinuria, type cbl C		metabolic diseases	
Methylmalonicacidaemia - homocystinuria, type cbl D		metabolic diseases	
Methylmalonicacidaemia - homocystinuria, type cbl F		metabolic diseases	
Other specified rare constitutional megaloblastic anaemia due to vitamin B12 metabolism disorder			

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Constitutional megaloblastic anaemias due to folate metabolism disorder	D52	metabolic diseases	
Folate-related anaemia due to impaired folate absorption/metabolism			
Homocystinuria due to methylenetetrahydrofolate reductase deficiency			
Formiminoglutamic aciduria			
Hereditary folate malabsorption			
Other specified rare constitutional megaloblastic anaemia due to folate metabolism disorder			
Constitutional megaloblastic anaemias, vitamin B12- and folate-independent			
Lesch-Nyhan syndrome	E79.1	metabolic diseases	
Hereditary orotic aciduria	D53.0	metabolic diseases	
Other specified rare constitutional megaloblastic anaemias			
Thiamine-responsive megaloblastic anaemia syndrome			
Constitutional megaloblastic anaemia, unspecified			
Acquired deficiency anaemias			
Acquired anaemias due to iron deficiency	D50		
Acquired anaemia with iron deficiency secondary to blood loss	D50.0		Transposed from ICD10.
Chronic posthaemorrhagic anaemia	D50.0		Transposed from ICD10.
Acute posthaemorrhagic anaemia	D62		Transposed from ICD10.
Congenital anaemia from fetal blood loss	P61.3		Transposed from ICD10.
Plummer-Vinson syndrome	D50.1		Keep as main term the eponym
<i>Kelly-Paterson syndrome</i>			"Plummer-Vinson syndrome" because it is
<i>Sideropenic dysphagia</i>			by far the most common denomination.
Acquired anaemia due to iron deficiency secondary to malnutrition			
<i>Acquired anaemia due to iron deficiency secondary to dietary deficiency</i>			
Acquired anaemia due to iron deficiency secondary to malabsorption			
Other specified acquired anaemias due to iron deficiency	D50.8		Transposed from ICD10.
Other specified rare acquired anaemias due to iron deficiency			
Acquired anaemia due to iron deficiency, unspecified	D50.9		Transposed from ICD10.
<i>Acquired anaemia due to iron deficiency NOS</i>			Transposed from ICD10.
Acquired megaloblastic anaemias due to vitamin B12 deficiency	D51		
Acquired megaloblastic anaemia due to vitamin B12 deficiency secondary to intrinsic factor deficiency	D51.0		
<i>Biermer disease</i>			
<i>Pernicious anaemia</i>			
Excludes: Vitamin B12 deficiency anaemia due to congenital intrinsic factor deficiency	D51.0		See above.

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Acquired megaloblastic anaemia due to nutritional vitamin B12 deficiency	D51.3		
<i>Acquired megaloblastic anaemia due to dietary vitamin B12 deficiency</i>			
Vegan anaemia	D51.3		
Nutritional vitamin B12 deficiency due to maternal vitamin B12 deficiency	D51.3		
Congenital nutritional vitamin B12 deficiency due to maternal vitamin B12 deficiency	D51.3		
Nutritional vitamin B12 deficiency due to maternal vitamin B12 deficiency in breast-fed infants	D51.3		
Other specified acquired anaemias due to vitamin B12 deficiency	D51.8		
Other specified rare acquired anaemias due to vitamin B 12 deficiency			
Acquired anaemia due to vitamin B12 deficiency, unspecified	D51.9		Transposed from ICD10.
<i>Acquired anaemia due to vitamin B12 deficiency NOS</i>			Transposed from ICD10.
Acquired megaloblastic anaemias due to folate deficiency	D52		
Acquired megaloblastic anaemia due to nutritional folate deficiency	D52.0		
<i>Acquired megaloblastic anaemia due to dietary folate deficiency</i>			
Drug-induced megaloblastic anaemia due to folate deficiency	D52.1		Transposed from ICD10.
Note: Use additional external cause code (Chapter XX), if desired, to identify drug.			
Other specified acquired megaloblastic anaemias due to folate deficiency	D52.8		Transposed from ICD10.
Other specified rare acquired megaloblastic anaemias due to folate deficiency			
Acquired megaloblastic anaemia due to folate deficiency, unspecified	D52.9		Transposed from ICD10.
<i>Acquired megaloblastic anaemia due to folate deficiency NOS</i>			Transposed from ICD10.
Megaloblastic anaemia, unspecified	D53.1		Transposed from ICD10.
Other specified acquired deficiency anaemias	D53		
Acquired megaloblastic anaemia unresponsive to vitamin B12 or folate therapy	D53		Transposed from ICD10
Scorbutic anaemia	D53.2		
Scurvy	E54	nutritional diseases	
Other specified acquired deficiency anaemias	D53.8		
Acquired anaemia due to protein deficiency	D53.0		
<i>Amino-acid deficiency anaemia</i>			
Acquired anaemia associated with deficiency of copper	D53.8		Transposed from ICD10.
Acquired anaemia associated with deficiency of molybdenun	D53.8		Transposed from ICD10.
Acquired anaemia associated with deficiency of zinc	D53.8		Transposed from ICD10.
Acquired deficiency anaemia, unspecified	D53.9		Transposed from ICD10.

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Constitutional haemolytic anaemias			
Constitutional haemolytic anaemias due to enzyme deficiency	D55		
Anaemia due to glucose-6-phosphate dehydrogenase deficiency	D55.0		
<i>Favism</i>			
<i>G6PD deficiency anaemia</i>			
Haemolytic anaemias due to hexose monophosphate shunt and glutathione metabolism anomalies			
Haemolytic anaemia due to glutathione synthetase deficiency	D55.1		
Haemolytic anaemia due to gamma-glutamylcysteine synthetase deficiency			
Haemolytic anaemia due to glutathione reductase deficiency			
Haemolytic anaemia due to 6-phosphogluconate dehydrogenase deficiency			
Haemolytic anaemia due to disorders of glycolytic enzymes	D55.2		
Haemolytic anaemia due to red cell pyruvate kinase deficiency			
Glycogen storage disease type 7		neuromuscular diseases	
<i>Phosphofructokinase (PFK) deficiency</i>			
<i>Tarui disease</i>			
Triose phosphate-isomerase deficiency		metabolic diseases	
Aldolase A deficiency		neuromuscular diseases	The choice of the main code in linearisation is arbitrary.
Phosphoglycerate kinase 1 deficiency		metabolic diseases	
Haemolytic anaemia due to glucosephosphate isomerase deficiency			
Haemolytic anaemia due to glyceraldehyde-3-phosphate dehydrogenase deficiency			
Haemolytic anaemia due to phosphoglycerate mutase deficiency of erythrocyte			
Haemolytic anaemia, nonspherocytic, due to hexokinase deficiency			
Congenital erythropoietic porphyria	E80.0	metabolic diseases	
<i>Günther disease</i>			
<i>Haemolytic anaemia due to uroporphyrin III synthase</i>			
Haemolytic anaemias due to disorders of nucleotide metabolism			
Haemolytic anaemia due to pyrimidine 5' nucleotidase deficiency			
Haemolytic anaemia due to adenylate kinase deficiency			
Haemolytic anaemia due to adenosine deaminase excess			
Other specified rare haemolytic anaemias due to enzyme disorders	D55.8		
Haemolytic anaemia due to enzyme disorder, unspecified	D55.9		
Other constitutional haemolytic anaemias			
Hereditary spherocytosis	D58.0		
<i>Minkowski-Chauffard syndrome</i>			

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Hereditary elliptocytosis	D58.1		
Elliptocytosis, common, hereditary Elliptocytosis, homozygous, hereditary Elliptocytosis, spherocytic Pyropoikilocytosis, hereditary Southeast asian ovalocytosis			
Stomatocytosis	D58.8		
Overhydrated hereditary stomatocytosis Dehydrated hereditary stomatocytosis Rh deficiency syndrome Intermediate stomatocytosis syndrome			
Acanthocytic disorders	E78.6	neurology neurology	
Abetalipoproteinaemia McLeod neuroacanthocytosis syndrome			
Atypical haemolytic-uraemic syndrome	D59.3		This exclusion note is for clarity only.
Excludes: Typical haemolytic-uraemic syndrome			
Other specified rare constitutional haemolytic anaemias			
Haemolytic anaemia, lethal - genital anomalies Diphosphoglycerate mutase deficiency of erythrocyte Alport syndrome - mental retardation - midface hypoplasia - elliptocytosis Familial pseudohyperkalaemia			
Constitutional haemolytic anaemia, unspecified	D58.9		
Acquired haemolytic anaemias			
Autoimmune haemolytic anaemia	D59.1		
Autoimmune haemolytic anaemia, cold-type	D59.1		
Cold agglutinin disease	D59.1		
Paroxysmal cold haemoglobinuria	D59.6		
Autoimmune haemolytic anaemia, mixed-type (cold/warm)			
Autoimmune haemolytic anaemia, warm-type	D59.1		
Evans syndrome	D69.3		Also included among autoimmune thrombocytopenic purpuras. The choice of the main code in linearisations is open to debate.

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Haemolytic disease of fetus and newborn	P55.-	neonatology	
Drug-induced autoimmune haemolytic anaemia	D59.0		Transposed from ICD10.
Note: Use additional external cause code (Chapter XX), if desired, to identify drug.			
Drug-induced nonautoimmune haemolytic anaemia	D59.2		Transposed from ICD10.
<i>Drug-induced enzyme deficiency anaemia</i>			
Note: Use additional external cause code (Chapter XX), if desired, to identify drug.			Transposed from ICD10.
Typical haemolytic-uraemic syndrome	D59.3		
Excludes: Atypical haemolytic-uraemic syndrome			This exclusion note is for clarity only
Other nonautoimmune haemolytic anaemias	D59.4		Transposed from ICD10.
Note: Use additional external cause code (Chapter XX), if desired, to identify drug.			Transposed from ICD10.
Mechanical haemolytic anaemia			Transposed from ICD10.
Microangiopathic haemolytic anaemia			Transposed from ICD10.
Toxic haemolytic anaemia			Transposed from ICD10.
Paroxysmal nocturnal haemoglobinuria	D59.5		
<i>Marchiafava-Micheli disease</i>			
Haemoglobinuria due to haemolysis from other external causes	D59.6		Transposed from ICD10.
Haemoglobinuria from exertion			Transposed from ICD10.
Haemoglobinuria from march			Transposed from ICD10.
Note: Use additional external cause code (Chapter XX), if desired, to identify drug.			Transposed from ICD10.
Other specified acquired haemolytic anaemias	D59.8		Transposed from ICD10.
Other specified rare acquired haemolytic anaemias			
Acquired haemolytic anaemia, unspecified	D59.9		Transposed from ICD10.
Haemoglobinopathies			
Thalassaemias	D56		
Alpha thalassaemia	D56.0		
			Heterozygous (α - / $\alpha\alpha$, α - / α -) and mutational forms of alpha-thalassaemia
			Haemoglobin H disease
			α - / - - and mutational forms of alpha-thalassaemia
			Haemoglobin Constant Spring
			Haemoglobin Paksé
			Alpha-thalassaemia hydrops fetalis
			Alpha thalassaemia - mental retardation, X-linked

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Beta thalassaemia	D56.1		
Thalassaemia minor <i>Beta-thalassaemia carrier</i> <i>Beta-thalassaemia trait</i> <i>Heterozygous beta-thalassaemia</i>			
Thalassaemia intermedia	D56.1		
Thalassaemia major <i>Cooley's anaemia</i> <i>Mediterranean anaemia</i> <i>Homozygous beta-thalassaemia</i> <i>Compound heterozygotes for $\beta 0$ or $\beta +$ or $\delta\beta$-thal genes</i>	D56.1		
Hereditary persistence of fetal haemoglobin/delta-beta thalassaemia			There is some overlap between HPFH and delta-beta thalassaemia. Reference: Nadkarni A, Wadia M, Gorakshakar A, Kiyama R, Colah RB, Mohanty D. <i>Molecular characterization of delta betathalassaemia and hereditary persistence of fetal hemoglobin in the Indian population.</i> Hemoglobin. 2008;32(5):425-33.
Hereditary persistence of fetal haemoglobin <i>HPFH</i>	D56.4		
Delta-beta thalassaemia	D56.2		
Other specified thalassaemias	D56.8		
Thalassaemia, unspecified	D56.9		
Sickle-cell disorders	D57		
Sickle-cell disease carrier <i>Hb-S trait</i> <i>Hb-S carrier</i> <i>Heterozygous haemoglobin S</i> <i>Heterozygous Hb-S</i>	D57.3		
Sickle-cell disease <i>Sickle-cell anaemia</i> <i>Sickle-cell disorder</i> <i>Homozygous sickle-cell disease</i>	D57.0		

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Double heterozygous sickling disorders	D57.2		
Compound HbS/HbC heterozygotes <i>Hb-SC disease</i>	D57.2		
Compound HbS/HbD heterozygotes <i>Hb-SD disease</i>	D57.2		
Compound HbS/HbE heterozygotes <i>Hb-SE disease</i>	D57.2		
Compound HbS/beta thalassaemia heterozygotes (includes HbS/ $\delta\beta$ -thal compounds) <i>Sickle-cell/beta thalassaemia</i>			
Compound HbS/alpha thalassaemia heterozygotes <i>Sickle-cell/alpha thalassaemia</i>			
Other specified sickle-cell disorders			
Haemoglobin E disease			
Heterozygous HbE carriers			
Compound HbE/ β -thalassemia heterozygotes			
Compound HbE/ α -thalassemia heterozygotes			
Compound HbE /other Hb mutant heterozygotes			
Haemoglobin C disease			
Heterozygous HbC carriers			
Homozygous or compound heterozygotes (HbC/HbC, HbC/ β -thal and other)			
Excludes: Hereditary persistence of fetal haemoglobin			Put among thalassaemias, cf. page above.
Haemoglobin D disease			
Haemoglobin O Arab disease			
Unstable haemoglobins causing haemolytic anaemia			
Haemoglobins causing cyanosis			
Haemoglobins with abnormal oxygen affinity			
Other more complex forms of haemoglobinopathies			
Hereditary congenital methaemoglobinaemia	D74.0		Transposed from ICD10.
<i>Haemoglobin-M disease</i>	D74.0		Transposed from ICD10.
<i>Hb-M disease</i>			Transposed from ICD10.
<i>Congenital NADH-methaemoglobin reductase deficiency</i>	D74.0		Transposed from ICD10.
<i>Recessive hereditary methaemoglobinaemia</i>	D74.0		
Recessive hereditary methemoglobinaemia type 1			
Recessive hereditary methemoglobinaemia type 2			

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Acquired methaemoglobinaemia	D74.8		Transposed from ICD10.
Toxic methaemoglobinaemia	D74.8		Transposed from ICD10.
Note: Use additional external cause code (Chapter XX), if desired, to identify cause.			Transposed from ICD10.
Methaemoglobinaemia, unspecified	D74.9		Transposed from ICD10.
Other constitutional anaemias			
Constitutional aplastic anaemias	D61.0		
Dyskeratosis congenita		to be discussed	
Shwachman-Diamond syndrome		to be discussed	
Blackfan-Diamond disease	D61.0		
<i>Pure aplasia of red cell, congenital</i>			
Fanconi anaemia			
Progressive pancytopenia - immunodeficiency - cerebellar hypoplasia			
<i>Hoyeraal-Hreidarsson syndrome</i>			
WT limb-blood syndrome			
Hereditary sideroblastic anaemias	D64.0		
X-linked sideroblastic anaemia			
X-linked sideroblastic anaemia - ataxia			
Mitochondrial myopathy with sideroblastic anaemia		neuromuscular disorders	
Pearson syndrome		metabolic diseases	
Congenital dyserythropoietic anaemia	D64.4		
Dyserythropoietic anaemia with thrombocytopenia			
Dyserythropoietic anaemia, congenital, type I			
Dyserythropoietic anaemia, congenital, type II			
Dyserythropoietic anaemia, congenital, type III			
Other specified constitutional anaemias			
Other specified rare constitutional anaemias			
Constitutional anaemia, unspecified			
Other acquired anaemias			
Acquired pure red cell aplasia	D60		Transposed from ICD10.
<i>Erythroblastopenia</i>			Transposed from ICD10.
Chronic acquired pure red cell aplasia	D60.0		
<i>Pure red cell aplasia, adult form</i>			

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Transient acquired pure red cell aplasia <i>Transient erythroblastopenia of childhood</i>	D60.1		
Other specified acquired pure red cell aplasias	D60.8		Transposed from ICD10.
Acquired pure red cell aplasia, unspecified	D60.9		Transposed from ICD10.
Acquired aplastic anaemias	D61		Transposed from ICD10.
Drug-induced aplastic anaemia	D61.1		Transposed from ICD10.
Note: Use additional external cause code (Chapter XX), if desired, to identify drug.			Transposed from ICD10.
Aplastic anaemia due to other external agents	D61.2		Transposed from ICD10.
Note: Use additional external cause code (Chapter XX), if desired, to identify external agent.			Transposed from ICD10.
Idiopathic aplastic anaemia	D61.3		
Other specified acquired aplastic anaemias	D61.8		Transposed from ICD10.
Acquired aplastic anaemia, unspecified	D61.9		Transposed from ICD10.
Acute posthaemorrhagic anaemia	D62		Transposed from ICD10.
Anaemia in chronic diseases classified elsewhere	D63*		Transposed from ICD10.
Anaemia in neoplastic disease	D63.0* - C00-D48+		Transposed from ICD10.
Acquired sideroblastic anaemias	D64		Transposed from ICD10.
Secondary sideroblastic anaemia due to another disease	D64.1		Transposed from ICD10.
Note: Use additional external cause code (Chapter XX), if desired, to identify disease.			Transposed from ICD10.
Secondary sideroblastic anaemia due to drugs and toxins	D64.2		Transposed from ICD10.
Note: Use additional external cause code (Chapter XX), if desired, to identify drug.			Transposed from ICD10.
Acquired idiopathic sideroblastic anaemia			
Other acquired sideroblastic anaemias	D64.3		Transposed from ICD10.
Sideroblastic anaemia, not otherwise specified	D64.3		Transposed from ICD10.
Pyridoxine-responsive sideroblastic anaemia, not elsewhere classified	D64.3		Transposed from ICD10.
Other specified acquired anaemias	D64.8		
Infantile pseudoleukaemia			
Leukoerythroblastic anaemia			
Other specified rare acquired anaemias			
Acquired anaemia, unspecified			
Anaemia, unspecified	D64.9		

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ICD11 revised table draft				Corresponding ICD10 code	Main code in linearisation, when elsewhere	Comment
Constitutional coagulation defects, purpura and other haemorrhagic conditions						
Coagulation factors defects				D68		
<i>Clotting factors defects</i>						
	Haemophilia A			D66		
	<i>Hereditary factor VIII deficiency</i>					
	Haemophilia B			D67		
	<i>Christmas disease</i>					
	<i>Hereditary factor IX deficiency</i>					
	Von Willebrand disease			D68.0		
	<i>Angiohaemophilia</i>					
	<i>Vascular haemophilia</i>					Obsolete name, to keep only for historical reasons.
	Excludes: acquired von Willebrand disease					The creation of a new code for acquired von Willebrand disease is proposed below.
	Von Willebrand disease, type 1					
	Von Willebrand disease, type 2					
	Von Willebrand disease, type 2A					
	Von Willebrand disease, type 2B					
	Von Willebrand disease, type 2M					
	Von Willebrand disease, type 2N					
	Von Willebrand disease, type 3					
	Fibrinogen deficiency, congenital			D68.2		
	<i>Factor I deficiency</i>					Obsolete name, to keep only for historical reasons.
	Afibrinogenaemia, familial					
	Dysfibrinogenaemia, familial					
	Hypofibrinogenaemia, familial					
	Hypodysfibrinogenaemia, familial					
	Factor II deficiency			D68.2		
	<i>Prothrombin deficiency</i>					
	Dysprothrombinaemia					
	Hypoprothrombinaemia					

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Factor V deficiency <i>Proaccelerin deficiency</i> <i>Owren's disease</i>	D68.2		
Factor VII deficiency <i>Hypoproconvertinaemia</i> <i>Proconvertin deficiency, congenital</i>	D68.2		
Factor X deficiency <i>Stuart-Prower factor deficiency</i>	D68.2		
Factor XI deficiency <i>Rosenthal factor deficiency</i>	D68.1		
Factor XII deficiency <i>Hageman factor deficiency</i>	D68.2		
Factor XIII deficiency <i>Fibrin-stabilizing factor deficiency</i>	D68.2		
Hereditary deficiency of other coagulation factors	D68.2		
Congenital prekallikrein deficiency			
Congenital plasminogen activator inhibitor type 1 deficiency			
Congenital high-molecular-weight kininogen deficiency			
Combined deficiency of factor V and factor VIII			
Combined deficiency of vitamin K-dependent clotting factors			
Antiplasmin deficiency			
Constitutional thrombopathies			
Glanzmann thrombasthenia			
Inherited giant platelet disorder			
Bernard-Soulier syndrome			
Mediterranean macrothrombocytopenia			
<i>Bernard-Soulier carrier syndrome, benign autosomal dominant</i>			
MYH9 macrothrombocytopenia syndromes			
May-Hegglin thrombocytopenia			
Fechtner syndrome			
Epstein syndrome			
Sebastian syndrome			
Amegakaryocytic thrombocytopenia, congenital			

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ICD11 revised table draft	Corresponding ICD10 code	Main code in linearisation, when elsewhere	Comment
Other specified rare constitutional thrombocytopenias	D69.4		
Wiskott-Aldrich syndrome	D82.0		
Chediak-Higashi syndrome	E70.3		Also included in familial haemophagocytic lymphohistiocytosis, see below. Main code in linearisations is open to debate.
Stormorken-Sjaastad-Langslet syndrome			
Thrombocytopenia - absent radius	Q87.2		
Gardner-Morrison-Abbott syndrome			
Thrombocytopenia - chromosome breakage			
Paris-Trousseau thrombocytopenia			
Scott syndrome			
Thrombocytopenia, X-linked			
Hypoplasminogenaemia			
Storage pool platelet disease, unspecific			
Deficiency of P2Y12			
Amegacaryocytosis			
Pseudo-Von Willebrand disease			
Dyserythropoietic anaemia with thrombocytopenia			
Radio-ulnar synostosis - amegacaryocytic thrombocytopenia			bone diseases
Familial platelet syndrome with predisposition to acute myelogenous leukaemia			
Genetic platelet function disease due to a collagen receptor defect			
Bleeding diathesis due to glycoprotein VI deficiency			
Bleeding diathesis due to integrin alpha2-beta1 deficiency			
Hermansky-Pudlak syndrome	E70.3		immunology
Excludes : Hermansky-Pudlak syndrome type 2			
Macrothrombocytopenia with abnormal proplatelet formation, autosomal dominant			

Acquired coagulation defects, purpura and other haemorrhagic conditions

Disseminated intravascular coagulation

D65

Defibrination syndrome

Diffuse intravascular coagulation [DIC]

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Haemorrhagic disorder due to circulating anticoagulants	D68.3		Transposed from ICD10.
<i>Note:</i> Use additional external cause code (Chapter XX), if desired, to identify any administered anticoagulant.			Transposed from ICD10.
<i>Excludes:</i> Long-term use of anticoagulants without haemorrhage	Z92.1		Transposed from ICD10.
Haemorrhage during long-term use of anticoagulants			Transposed from ICD10.
Hyperheparinaemia			Transposed from ICD10.
Increase in antithrombin			Transposed from ICD10.
Increase in anti-VIIIa			Transposed from ICD10.
Increase in anti-IXa			Transposed from ICD10.
Increase in anti-Xa			Transposed from ICD10.
Increase in anti-XIa			Transposed from ICD10.
Acquired coagulation factor deficiency	D68.4		Transposed from ICD10.
<i>Excludes:</i> vitamin K deficiency of newborn	P53		Transposed from ICD10.
Deficiency of coagulation factor due to liver disease			Transposed from ICD10.
Deficiency of coagulation factor due to vitamin K deficiency			Transposed from ICD10.
Acquired von Willebrand disease			
Other specified coagulation defects	D68.8		Transposed from ICD10.
Familial antiphospholipid syndrome	D68.8		
Coagulation defect, unspecified	D68.9		Transposed from ICD10.
Nonthrombocytopenic purpura	D69.0		
<i>Excludes:</i> Henoch-Schönlein purpura	D69.0	immunology	
Vascular purpura			Transposed from ICD10.
Senile purpura			Transposed from ICD10.
Purpura simplex			Transposed from ICD10.
Autoimmune thrombocytopenic purpura	D69.3		
<i>Thrombocytopenic purpura, idiopathic</i>			

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<i>ICD11 revised table draft</i>					<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
		Evans syndrome			D69.3		Also included among autoimmune haemolytic anaemias. The choice of the main code in linearisations is open to debate.
		Thrombotic thrombocytopenic purpura <i>Moschcowitz disease</i>			M31.1		
		Hereditary or recurrent thrombotic thrombocytopenic purpura (idiopathic or ADAMTS-13 deficient)					The pathophysiology of this disease is incompletely known, so the choice was made to keep the various forms of TTP together, although they include hereditary forms.
		Severe infantile thrombotic thrombocytopenic purpura <i>Schulman-Upshaw syndrome</i>					
		Acquired thrombotic thrombocytopenic purpura due to ADAMTS-13 deficiency					
		Secondary thrombocytopenia <i>Note:</i> Use additional external cause code (Chapter XX), if desired, to identify cause.			D69.5		Transposed from ICD10.
		Acquired thrombocytopenia, unspecified			D69.6		Transposed from ICD10.
		Other specified rare acquired haemorrhagic conditions					
		Other specified haemorrhagic conditions Capillary fragility (hereditary) Vascular pseudothrombophilia			D69.8		Transposed from ICD10.
		Haemorrhagic condition, unspecified			D69.9		
		Constitutional thrombotic disease of haematologic origin					
		Hereditary thrombophilia Protein C deficiency Antithrombin deficiency, congenital Hypercoagulability syndrome, due to glycosylphosphatidylinositol deficiency					

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ICD11 revised table draft	Corresponding ICD10 code	Main code in linearisation, when elsewhere	Comment
Thrombomodulin anomalies, familial Resistance to activated protein C Thrombophilia due to factor V Leiden Thrombocytosis, familial Thrombocytosis, X-linked Thrombocytosis, autosomal dominant			

Acquired thrombotic disease of haematologic origin

Heparin-induced thrombocytopenia			
Purpura fulminans			
Essential thrombocythaemia	D75.2		
Gaisbock syndrome			
Simple cryoglobulinaemia	D89.1		
Protein S acquired deficiency			

Other diseases of blood and blood-forming organs

Diseases of spleen	D73		Transposed from ICD10.
Hyposplenism	D73.0		Transposed from ICD10.
Asplenia, postsurgical	D73.0		Transposed from ICD10.
Atrophy of spleen	D73.0		Transposed from ICD10.
Congenital asplenia	Q89.0	malformations	
Chronic congestive splenomegaly	D73.2		Transposed from ICD10.
Abscess of spleen	D73.3		Transposed from ICD10.
Cyst of spleen	D73.4		Transposed from ICD10.

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Infarction of spleen	D73.5		Transposed from ICD10.
Splenic rupture, nontraumatic	D73.5		Transposed from ICD10.
Torsion of spleen	D73.5		Transposed from ICD10.
Other specified diseases of spleen	D73.8		Transposed from ICD10.
Fibrosis of spleen NOS	D73.8		Transposed from ICD10.
Perisplenitis	D73.8		Transposed from ICD10.
Splentitis NOS	D73.8		Transposed from ICD10.
Disease of spleen, unspecified	D73.9		Transposed from ICD10.
Polycythaemia	D75		
<i>Erythrocytosis</i>			
Familial polycythaemia	D75.0		
Benign polycythaemia	D75.0		
Primary familial polycythaemia, type 1			
Primary familial polycythaemia, type 2			
Secondary polycythaemia	D75.1		Transposed from ICD10.
Erythrocytosis, unspecified	D75.1		Transposed from ICD10.
Acquired polycythaemia	D75.1		Transposed from ICD10.
Polycythaemia due to erythropoietin	D75.1		Transposed from ICD10.
Polycythaemia due to fall in plasma volume	D75.1		Transposed from ICD10.
Polycythaemia due to high altitude	D75.1		Transposed from ICD10.
Polycythaemia due to stress	D75.1		Transposed from ICD10.
Emotional polycythaemia	D75.1		Transposed from ICD10.
Hypoxaemic polycythaemia	D75.1		Transposed from ICD10.
Nephrogenous polycythaemia	D75.1		Transposed from ICD10.
Relative polycythaemia	D75.1		Transposed from ICD10.
Polycythaemia neonatorum	P61.1	neonatology	Transposed from ICD10.
Polycythaemia vera	D45		Also includedn in hronic myeloproliferative diseases. Main code should be here.
Certain diseases involving lymphoreticular tissue and reticulohistiocytic system	D76		
Langerhans cell histiocytosis	D76.0		Open to discussion; histiocytosis could be classified with blood neoplasms as a group. Main code should be here.
Eosinophilic granuloma	D76.0		

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<i>ICD11 revised table draft</i>				<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
		Letterer-Siwe disease				
		Hashimoto-Pritzker syndrome				
		Hand-Schüller-Christian disease				
		Familial haemophagocytic lymphohistiocytosis		D76.1	immunology	
		Familial haemophagocytic reticulosis		D76.1	immunology	
		Chediak-Higashi syndrome		E70.3	immunology	Also included in rare constitutional thrombocytopenias, see above. Main code in linearisations is open to debate.
		Griscelli disease			immunology	
		X-linked lymphoproliferative disease			immunology	
		Secondary haemophagocytic syndrome		D76.2		Transposed from ICD10.
	Note:	Use additional code, if desired, to identify infectious agent or disease.				

Tumor of haematopoietic and lymphoid tissues

Myeloid haemopathy

Leukaemia, myeloid, acute

- Leukaemia, myeloid, acute, with multilineage dysplasia
- Therapy related acute myeloid leukaemia and myelodysplastic syndrome
 - Leukaemia, myeloid, acute, and myelodysplastic syndromes related to alkylating agent
 - Leukaemia, myeloid, acute, and myelodysplastic syndromes related to topoisomerase type II inhibitor
- Leukaemia of ambiguous lineage, acute
 - Leukaemia, undifferentiated, acute
 - Leukaemia, bilineal, acute
 - Leukaemia, biphenotypic, acute
- Acute myeloid leukaemia with recurrent genetic anomaly
 - Leukaemia, myeloid, acute, with abnormal bone marrow eosinophils inv(16)(p13q22) or t(16;16)(p13;q22)
 - Leukaemia, myeloid, acute, with 11q23 abnormalities
 - Leukaemia, myeloid, acute with t(8;21)(q22;q22) translocation
- Unclassified acute myeloid leukaemia
 - Leukaemia, myelomonocytic, acute

C92.0

C95.0

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Leukaemia, monoblastic/monocytic , acute			
Leukaemia, myeloid, acute, with 11q23 abnormalities			
Leukaemia, erythroid, acute			
Leukaemia, megacaryoblastic, acute			
Leukaemia, myeloid, acute, in Down syndrome			
Acute panmyelosis with myelofibrosis			
Leukaemia, basophilic, acute			
Myeloid sarcoma			
Leukaemia, myeloblastic, acute, minimally differentiated			
Leukaemia, myeloblastic, acute, without maturation			
Leukaemia, myeloblastic, acute, with maturation			
Mediastinal (thymic) large B-cell lymphoma			
Lymphoma, intravascular, large B-cell			
Mediastinal diffuse large-cell lymphoma with sclerosis			
Refractory anaemia with excess blasts in transformation			
Myelodysplastic syndromes	D46.7		
Idiopathic acquired sideroblastic anaemia			
Refractory cytopenia with multilineage dysplasia			
Refractory anaemia			
Myelodysplastic syndrome, unclassified			
Aregenerative anaemia			
Refractory anaemia with excess blasts	D46.2		
Refractory anaemia with excess blasts-1			
Refractory anaemia with excess blasts-2			
Myelodysplastic syndrome associated with isolated del(5q) chromosome abnormality			
Refractory anaemia			
Myelodysplastic syndrome, unclassified			
Aregenerative anaemia			
Chronic myeloproliferative disease			
Thrombocythaemia, essential			
Chronic myeloid leukaemia	C92.1		
Myelofibrosis with myeloid metaplasia	D47.1		
Polycythaemia vera	D45	polycythaemia	Also included in polycythaemias.
Leukaemia, neutrophilic, chronic			
Chronic myeloproliferative disease, unclassified	D47.1		

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Chronic eosinophilic leukaemia			
Hypereosinophilic syndromes			
Idiopathic hypereosinophilic syndrome			
Myelodysplastic/myeloproliferative disease			
Leukaemia, myelomonocytic, juvenile form			
Leukaemia, myelomonocytic, chronic			
Leukaemia, atypical, myeloid, chronic			
Myelodysplastic/myeloproliferative disease, unclassified			
Myeloid neoplasms associated with eosinophilia and abnormality of PDGFRA, PDGFRB or FGFR1			
Myeloid neoplasm associated with PDGFRA rearrangement			
Myeloid neoplasm associated with PDGFRB rearrangement			
Myeloid neoplasm associated with FGFR1 rearrangement			
Lymphoid haemopathy			
Cutaneous lymphoma	C84	dermatology	The whole group should also be present in the dermatology chapter. Place of the main code is to be discussed.
Blastic NK cell lymphoma			
Cutaneous T-cell lymphoma			
Indolent cutaneous T-cell lymphoma			
Subcutaneous panniculitis-like T-cell lymphoma			
Primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma			
Mycosis fungoides and variants			
Classical mycosis fungoides			
Granulomatous slack skin			
Folliculotropic mycosis fungoides			
Localised pagetoid reticulosis			
Primary cutaneous CD30+ lymphoproliferative disease			
Primary cutaneous anaplastic large cell lymphoma			
Anaplastic large cell lymphoma			
Lymphomatoid papulosis			
Lymphomatoid papulosis			
Aggressive cutaneous T-cell lymphoma			
Sezary's syndrome			
T-cell leukaemia/lymphoma, adult			

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Extranodal NK/T cell lymphoma, nasal type			
Peripheral T-cell lymphoma, unspecified			
Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma			
Cutaneous gamma/delta-positive T-cell lymphoma			
Cutaneous B-cell lymphoma			
Aggressive cutaneous B-cell lymphoma			
Primary cutaneous diffuse large B-cell lymphoma, leg type			
Indolent cutaneous B-cell lymphoma			
Primary cutaneous marginal zone B-cell lymphoma			
Primary cutaneous follicle center lymphoma			
B-cell lymphoma with common secondary cutaneous involvement			
Burkitt lymphoma			
Myeloma, multiple			
Waldenström macroglobulinaemia			
Mantle cell lymphoma			
Lymphomatoid granulomatosis			
Leukaemia, lymphoblastic, acute	C91.0		
Leukaemia/lymphoma, precursor T lymphoblastic			
Leukaemia, precursor B-cell lymphoblastic, acute			
Leukaemia, precursor T-cell lymphoblastic, acute			
Leukaemia/lymphoblastic lymphoma, precursor B lymphoblastic			
B-cell non-Hodgkin lymphoma			
Burkitt lymphoma			
Diffuse large B-cell lymphoma			
Mediastinal (thymic) large B-cell lymphoma			
Lymphoma, intravascular, large B-cell			
Mediastinal diffuse large-cell lymphoma with sclerosis			
Follicular lymphoma			
Myeloma, multiple			
Waldenström macroglobulinaemia			
Primary effusion lymphoma			
Primary effusion lymphoma associated with the human immune deficiency virus infection			
Primary effusion lymphoma associated with the human immune deficiency virus (HIV) infection			

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Mantle cell lymphoma			
MALT lymphoma			
Leukaemia, hairy cell			
Leukaemia, B-cell lymphocytic, chronic			
Leukaemia, B-cell prolymphocytic			
Splenic marginal zone lymphoma			
Plasmacytoma			
Primary plasmacytoma of the bone			
Extramedullary (soft tissue) plasmacytoma			
Heavy-chain disease			
Franklin disease			
Mu heavy-chain disease			
Alpha heavy-chain disease			
Gamma heavy-chain disease			
Nodal marginal zone B-cell lymphoma			
Leukaemia, precursor B-cell lymphoblastic, acute			
T-cell non-Hodgkin lymphoma			
Classical mycosis fungoides			
Sezary's syndrome			
Primary cutaneous anaplastic large cell lymphoma			
Anaplastic large cell lymphoma			
Lymphomatoid papulosis			
Post-transplant lymphoproliferative disease			
Lymphomatoid granulomatosis			
Blastic NK cell lymphoma			
Leukaemia, T-cell prolymphocytic			
Leukaemia, T-cell large granular lymphocyte			
Leukaemia, aggressive NK-cell			
T-cell leukaemia/lymphoma, adult			
Extranodal NK/T cell lymphoma, nasal type			
Enteropathy-type T-cell lymphoma			
Hepatosplenic T-cell lymphoma			
Subcutaneous panniculitis-like T-cell lymphoma			
Peripheral T-cell lymphoma, unspecified			
Angioimmunoblastic T-cell lymphoma			

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Leukaemia/lymphoma, precursor T lymphoblastic			
Anaplastic large cell lymphoma			
Lymphomatoid papulosis			
Plasma cell tumor			
POEMS syndrome			
Myeloma, multiple	C90.0		
Amyloidosis, primary			
Plasmacytoma	C90.2		
Primary plasmacytoma of the bone			
Extramedullary (soft tissue) plasmacytoma			
Monoclonal immunoglobulin depositions disease			
Heavy chain deposition disease			
Light and heavy chain deposition disease			
Light chain deposition disease			
Heavy-chain disease			
Franklin disease			
Mu heavy-chain disease			
Alpha heavy-chain disease			
Gamma heavy-chain disease			
Histiocytic and dendritic cell tumor			
Macrophage or histiocytic tumor			
Histiocytic sarcoma			
Dendritic cell tumor			
Langerhans cell histiocytosis	D76.0		Open to discussion; histiocytosis are also suggested to be included above in the section " Certain diseases involving lymphoreticular tissue and reticulohistiocytic system "
Letterer-Siwe disease	C96.0		
Eosinophilic granuloma	D76.0		
Hashimoto-Pritzker syndrome			
Hand-Schüller-Christian disease	D76.0		
Langerhans cell histiocytosis, pulmonary, adult-form			
Langerhans cell sarcoma	C25.4		
Interdigitating dendritic cell sarcoma			
Follicular dendritic cell sarcoma	C82		

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<i>ICD11 revised table draft</i>	<i>Corresponding ICD10 code</i>	<i>Main code in linearisation, when elsewhere</i>	<i>Comment</i>
Dendritic cell sarcoma, not otherwise specified			
Immunodeficiency-associated lymphoproliferative disease			
Lymphoproliferative disease associated with primary immune disease			
Post-transplant lymphoproliferative disease			
Methotrexate-associated lymphoproliferative disorders			
Primary effusion lymphoma associated with the human immune deficiency virus infection			
Hodgkin lymphoma	C81		
Hodgkin lymphoma, classical			
Nodular sclerosis classical Hodgkin lymphoma			
Mixed cellularity classical Hodgkin lymphoma			
Lymphocyte-rich classical Hodgkin lymphoma			
Lymphocyte-depleted classical Hodgkin lymphoma			
Nodular lymphocyte predominant Hodgkin lymphoma	C81.0		
Composite lymphoma			
Mastocytosis	C96.2		
Excludes: Mastocytosis, cutaneous	Q82.2		To be included in dermatology.
Mastocytosis, systemic			
Mastocytosis, indolent systemic			
Mastocytosis, systemic, with an associated clonal haematologic non-mast cell c lineage disease			
Mastocytosis, aggressive systemic			
Mast cell leukaemia			
Mast cell sarcoma	C96.2		
Extracutaneous mastocytoma	C96.2		

**Groups included in the haematological
diseases section of ICD-10,
to be reclassified in ICD-11**

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Groups included in the haematological diseases section of ICD10, to be reclassified in ICD11

<i>Part of ICD10 tabular list</i>	<i>ICD10 code</i>	<i>Comment</i>
<p>Agranulocytosis Agranulocytic angina Infantile genetic agranulocytosis Kostmann's disease Neutropenia: · NOS · congenital · cyclic · drug-induced · periodic · splenic (primary) · toxic Neutropenic splenomegaly Use additional external cause code (Chapter XX), if desired, to identify drug, if drug-induced. Excludes: transient neonatal neutropenia (P61.5)</p>	D70	Move into immunodeficiencies.
<p>Functional disorders of polymorphonuclear neutrophils Cell membrane receptor complex [CR3] defect Chronic (childhood) granulomatous disease Congenital dysphagocytosis Progressive septic granulomatosis</p>	D71	Move into immunodeficiencies.
<p>Other disorders of white blood cells Excludes: basophilia (D75.8) Excludes: immunity disorders (D80-D89) Excludes: neutropenia (D70) Excludes: preleukaemia (syndrome) (D46.9)</p>	D72	
<p>Genetic anomalies of leukocytes Anomaly (granulation)(granulocyte) or syndrome: · Alder · May-Hegglin · Pelger-Huët Hereditary: · leukocytic: · hypersegmentation · hyposegmentation · leukomelanopathy Excludes: Chediak(-Steinbrinck)-Higashi syndrome (E70.3)</p>	D72.0	<p>Move into immunodeficiencies.</p> <p>Is a morphological anomaly of leukocytes, not a clinical entity (seen i.e. in myeloperoxidase deficiency, an immunodeficiency) Move into thrombocytopenias. Is a morphological anomaly of leukocytes, not a clinical entity. Are morphological anomalies of leukocytes, not clinical entities.</p>

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Groups included in the haematological diseases section of ICD10, to be reclassified in ICD11

<i>Part of ICD10 tabular list</i>	<i>ICD10 code</i>	<i>Comment</i>
Eosinophilia	D72.1	Is a laboratory feature, not a clinical entity.
Eosinophilia: · allergic · hereditary		
Other specified disorders of white blood cells	D72.8	No recent information exists about this disease. Are laboratory features, not entities.
Leukaemoid reaction: · lymphocytic · monocytic · myelocytic Leukocytosis Lymphocytosis (symptomatic) Lymphopenia Monocytosis (symptomatic) Plasmacytosis		
Disorder of white blood cells, unspecified	D72.9	
Other specified diseases of blood and blood-forming organs	D75.8	
Basophilia		Not an entity but a laboratory feature, found mostly in leukaemia.