D64.0	**Hereditary sideroblastic anaemia** Sex-linked hypochromic sideroblastic anaemia
D64.1	**Secondary sideroblastic anaemia due to disease** Use additional code, if desired, to identify disease.
D64.2	**Secondary sideroblastic anaemia due to drugs and toxins** Use additional external cause code (Chapter XX), if desired, to identify cause.
D64.3	**Other sideroblastic anaemias** Sideroblastic anaemia: · NOS · pyridoxine-responsive NEC
D64.4	**Congenital dyserythropoietic anaemia** Dyshaematopoietic anaemia (congenital) Excludes: Blackfan-Diamond syndrome (D61.0) Di Guglielmo's disease (C94.0)
D64.8	**Other specified anaemias** Infantile pseudoleukaemia Leukoerythroblastic anaemia
D64.9	**Anaemia, unspecified**

D64.0	**Hereditary sideroblastic anaemia** X-linked sideroblastic anaemia, piridoxin responsive sideroblastgic anemia X-linked sideroblastic anaemia with ataxia Autosomal recessive sideroblastic anaemia
D64.1	**Secondary sideroblastic anaemia due to disease** Use additional code, if desired, to identify disease.
D64.2	**Secondary sideroblastic anaemia due to drugs and toxins** Use additional external cause code (Chapter XX), if desired, to identify cause.
D64.3	**Other sideroblastic anaemias** Sideroblastic anaemia NOS
D64.4	**Congenital dyserythropoietic anaemia (CDA)** CDA type I CDA type II Hempas CDA type III CDA, others Excludes: Blackfan-Diamond syndrome (D61.0) Di Guglielmo's disease (C94.0)
D64.5	**Megaloblastic anaemias not elsewhere classified** **Thiamine responsive megaloblastic anaemia** Excludes: Di Gluglielmo-s disease (C90.0)
D64.8	**Other specified anaemias** Leukoerythroblastic anaemia Myelophtisic anaemia
D64.9	**Anaemia, unspecified**

D65 **Disseminated intravascular coagulation [defibrination syndrome]**

Afibrinogenaemia, acquired
Consumption coagulopathy
Diffuse or disseminated intravascular coagulation [DIC]
Fibrinolytic haemorrhage, acquired
Purpura:
· fibrinolytic
· fulminans

Excludes: that (complicating):
· abortion or ectopic or molar pregnancy (O00-O07 , O08.1)
· in newborn (P60)
· pregnancy, childbirth and the puerperium (O45.0 , O46.0 , O67.0 , O72.3)

Coagulation defects, purpura and other haemorrhagic conditions (D65-D69)

D65 **Disseminated intravascular coagulation [defibrination syndrome]**
Afibrinogenaemia, acquired
Consumption coagulopathy
Diffuse or disseminated intravascular coagulation [DIC]
Fibrinolytic haemorrhage, acquired
Purpura:
- fibrinolytic
- fulminans

Excludes: that (complicating):
- abortion or ectopic or molar pregnancy (O00-O07 , O08.1)
- in newborn (P60)
- pregnancy, childbirth and the puerperium (O45.0 , O46.0 , O67.0 , O72.3)

D66 Hereditary factor VIII deficiency Deficiency factor VIII (with functional defect) Haemophilia: · NOS · A · classical ***Excludes:*** factor VIII deficiency with vascular defect (D68.0)	**D66 Hereditary factor VIII deficiency** Deficiency factor VIII (with functional defect) Haemophilia: • NOS • A • classical Excludes: factor VIII deficiency with vascular defect (D68.0)
D67 Hereditary factor IX deficiency Christmas disease Deficiency: · factor IX (with functional defect) · plasma thromboplastin component [PTC] Haemophilia B	**D67 Hereditary factor IX deficiency** Christmas disease Deficiency: • factor IX (with functional defect) • plasma thromboplastin component [PTC] Haemophilia B
D68 Other coagulation defects ***Excludes:*** those complicating: · abortion or ectopic or molar pregnancy (O00-O07 , O08.1) · pregnancy, childbirth and the puerperium (O45.0 , O46.0 , O67.0 , O72.3)	**D68 Other coagulation defects** Excludes: those complicating: • abortion or ectopic or molar pregnancy (O00-O07 , O08.1) • pregnancy, childbirth and the puerperium (O45.0 , O46.0 , O67.0 , O72.3)

D68.0 Von Willebrand's disease Angiohaemophilia Factor VIII deficiency with vascular defect Vascular haemophilia ***Excludes:*** capillary fragility (hereditary) (D69.8) factor VIII deficiency: · NOS (D66) · with functional defect (D66)	**D68.0 Von Willebrand's disease** Angiohaemophilia Factor VIII deficiency with vascular defect Vascular haemophilia Excludes: capillary fragility (hereditary) (D69.8) factor VIII deficiency: • NOS (D66) • with functional defect (D66)
D68.1 Hereditary factor XI deficiency Haemophilia C Plasma thromboplastin antecedent [PTA] deficiency	**D68.1 Hereditary factor XI deficiency** Haemophilia C Plasma thromboplastin antecedent [PTA] deficiency
D68.2 Hereditary deficiency of other clotting factors Congenital afibrinogenaemia Deficiency: · AC globulin · proaccelerin Deficiency of factor: · I [fibrinogen] · II [prothrombin] · V [labile] · VII [stable] · X [Stuart-Prower] · XII [Hageman] · XIII [fibrin-stabilizing] Dysfibrinogenaemia (congenital) Hypoproconvertinaemia Owren's disease	**D68.2 Hereditary deficiency of other clotting factors** Congenital afibrinogenaemia Deficiency: • AC globulin • proaccelerin Deficiency of factor: • I [fibrinogen] • II [prothrombin] • V [labile] • VII [stable] • X [Stuart-Prower] • XII [Hageman] • XIII [fibrin-stabilizing] Dysfibrinogenaemia (congenital) Hypoproconvertinaemia Owren's disease

D68.3 **Haemorrhagic disorder due to circulating anticoagulants** Haemorrhage during long-term use of anticoagulants Hyperheparinaemia Increase in: · antithrombin · anti-VIIIa · anti-IXa · anti-Xa · anti-XIa Use additional external cause code (Chapter XX), if desired, to identify any administered anticoagulant. ***Excludes:*** long-term use of anticoagulants without haemorrhage (Z92.1)	**D68.3** **Haemorrhagic disorder due to circulating anticoagulants** Haemorrhage during long-term use of anticoagulants Hyperheparinaemia Increase in: • antithrombin • anti-VIIIa • anti-Ixa • anti-Xa • anti-XIa Use additional external cause code (Chapter XX), if desired, to identify any administered anticoagulant. Use additional external cause code (Chapter XX), if desired, to identify any administered anticoagulant. Excludes: long-term use of anticoagulants without haemorrhage (Z92.1)
D68.4 **Acquired coagulation factor deficiency** Deficiency of coagulation factor due to: · liver disease · vitamin K deficiency ***Excludes:*** vitamin K deficiency of newborn (P53)	**D68.4** **Acquired coagulation factor deficiency** Deficiency of coagulation factor due to: • liver disease • vitamin K deficiency Excludes: vitamin K deficiency of newborn (P53) **D68.5** **Primary Thrombophilia** Activated protein C resistance [factor V Leiden mutation] Deficiency: • antithrombin • protein C • protein S Prothrombin gene mutation

D68.8 **Other specified coagulation defects** Presence of systemic lupus erythematosus [SLE] inhibitor **D68.9** **Coagulation defect, unspecified** **D69** **Purpura and other haemorrhagic conditions** ***Excludes:*** benign hypergammaglobulinaemic purpura (D89.0) cryoglobulinaemic purpura (D89.1) essential (haemorrhagic) thrombocythaemia (D47.3) purpura fulminans (D65) thrombotic thrombocytopenic purpura (M31.1)	**D68.6** **Other Thrombophilia** Anticardiolipin syndrome Antiphospholipid syndrome Presence of the lupus anticoagulant Excludes: disseminated intravascular coagulation (D65) hyperhomocysteinemia (E72.1) **D68.8** **Other specified coagulation defects** **Coagulation defect, unspecified** **D68.9** **D69** **Purpura and other haemorrhagic conditions** Excludes: benign hypergammaglobulinaemic purpura (D89.0) cryoglobulinaemic purpura (D89.1) essential (haemorrhagic) thrombocythaemia (D47.3) purpura fulminans (D65) thrombotic thrombocytopenic purpura (M31.1)

D69.0 Allergic purpura Purpura: · anaphylactoid · Henoch(-Schönlein) · nonthrombocytopenic: · haemorrhagic · idiopathic · vascular Vasculitis, allergic	**D69.0 Allergic purpura** Purpura: • anaphylactoid • Henoch(-Schönlein) • nonthrombocytopenic: • haemorrhagic • idiopathic • vascular Vasculitis, allergic
D69.1 Qualitative platelet defects Bernard-Soulier [giant platelet] syndrome Glanzmann's disease Grey platelet syndrome Thromboasthenia (haemorrhagic)(hereditary) Thrombocytopathy ***Excludes:*** von Willebrand's disease (D68.0)	**D69.1 Qualitative platelet defects** Bernard-Soulier [giant platelet] syndrome Glanzmann's disease Grey platelet syndrome Thromboasthenia (haemorrhagic)(hereditary) Thrombocytopathy Excludes: von Willebrand's disease (D68.0)
D69.2 Other nonthrombocytopenic purpura Purpura: · NOS · senile · simplex	**D69.2 Other nonthrombocytopenic purpura** Purpura: • NOS • senile • simplex
D69.3 Idiopathic thrombocytopenic purpura Evans' syndrome	**D69.3 Idiopathic thrombocytopenic purpura** Evans' syndrome
D69.4 Other primary thrombocytopenia	**D69.4 Other primary thrombocytopenia**

Excludes: thrombocytopenia with absent radius (Q87.2) transient neonatal thrombocytopenia (P61.0) Wiskott-Aldrich syndrome (D82.0)	Excludes: thrombocytopenia with absent radius (Q87.2) transient neonatal thrombocytopenia (P61.0) Wiskott-Aldrich syndrome (D82.0)
D69.5 Secondary thrombocytopenia Use additional external cause code (Chapter XX), if desired, to identify cause.	**D69.5 Secondary thrombocytopenia** Use additional external cause code (Chapter XX), if desired, to identify cause.
D69.6 Thrombocytopenia, unspecified	**D69.6 Thrombocytopenia, unspecified**
D69.8 Other specified haemorrhagic conditions Capillary fragility (hereditary) Vascular pseudohaemophilia	**D69.8 Other specified haemorrhagic conditions** Capillary fragility (hereditary) Vascular pseudohaemophilia
D69.9 Haemorrhagic condition, unspecified	**D69.9 Haemorrhagic condition, unspecified**

Other diseases of blood and blood-forming organs (D70-D77)

D70 **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)

D70. Disorders of Neutrophil Number

D70.1 Disorders with Decreased Neutrophil Counts
- D70.1a Acquired Neutropenia
  - Immune-mediated (includes autoimmune, alloimmune, isoimmune)
  - defective production (includes malnutrition, copper deficiency, secondary to infections)
  - drug-related
  - idiopathic
  - NOS (includes defective distribution, hypersplenism)
- D70.1b Congenital Neutropenia
  - Benign ethnic neutropenia (Duffy variants etc.)
  - congenital neutropenia with maturation arrest (includes ELANE2 including cyclic neutropenia, HAX1 including Kostmann syndrome, others)
  - congenital neutropenia with myelokathexis (CXCR4, others)
  - congenital neutropenia associated with syndromal features (G6PT, G6PC3, Barth syndrome, Cohen syndrome, CHH, MAPBPIP, SBDS, etc,)

D70.2. Disorders with Increased Neutrophil Counts
- D70.2a Acquired
  reactive (infection, inflammation, myeloproliferative disorders etc)
- D70.2b Congenital
  mutations in G-CSF receptor
- D70.2c Not otherwise specified (NOS)

D71 **Functional disorders of polymorphonuclear neutrophils**

Cell membrane receptor complex [CR3] defect
Chronic (childhood) granulomatous disease
Congenital dysphagocytosis
Progressive septic granulomatosis

D72 **Other disorders of white blood cells**

Excludes: basophilia (D75.8)
immunity disorders (D80-D89)
neutropenia (D70)
preleukaemia (syndrome) (D46.9)

D71. Disorders of Neutrophil Function

D71.1 Acquired
Neutrophil dysfunction secondary to systemic infection or organ failure (including uremia, sepsis)

D71.2 Congenital
- Disorders of adhesion (includes leukocyte adhesion disorder (LAD) type 1, LAD type 2, congenital disorder of glycosylation type IIc, LAD type 3
- Disorders of chemotaxis (includes neutrophil actin dysfunction, Rac2 mutation; also see "Hyper IgE syndromes") (excludes congenital deficiencies of complement components, localized juvenile periodontitis)
- Disorders of granule formation or release (includes secondary granule deficiency; also see "Immunodeficiency with hypopigmentation")
- Disorders of oxidative metabolism (includes chronic granulomatous disease, leukocyte G6PD deficiency, myeloperoxidase deficiency, disorders of glutathione metabolism)
- Hereditary, without known mechanism

D71.3 Not otherwise specified (NOS)

D72. Abnormalities of Neutrophil Morphology

D72.1 Acquired
- Neutrophil nuclear anomalies (includes hypersegmentation, hyposegmentation)
- Cytoplasmic anomalies (includes toxic granulation, vacuolization, Döhle bodies)

D72.2 Congenital (Alder-Reilly, May-Heggelin, Pelger-Huet anomalies)

D72.0 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)

D72.1 Eosinophilia

Eosinophilia:
- allergic
- hereditary

D72.8 Other specified disorders of white blood cells

Leukaemoid reaction:
- lymphocytic
- monocytic
- myelocytic

Leukocytosis
Lymphocytosis (symptomatic)
Lymphopenia
Monocytosis (symptomatic)
Plasmacytosis

D72.3 Not otherwise specified (NOS)

D72.9 Disorder of white blood cells, unspecified

D73 Diseases of spleen

D73.0 Hyposplenism

Asplenia, postsurgical
Atrophy of spleen

Excludes: asplenia (congenital) (Q89.0)

D73.1 Hypersplenism

Excludes: splenomegaly:
- NOS (R16.1)
- congenital (Q89.0)

D73.2 Chronic congestive splenomegaly

D73.3 Abscess of spleen

D73.4 Cyst of spleen

D73.5 Infarction of spleen

Splenic rupture, nontraumatic
Torsion of spleen

Excludes: traumatic rupture of spleen (S36.0)

D73. EOSINOPHILS: Disorders of Eosinophil Number

D73.1 Disorders with increased eosinophil counts
- Infections
- Allergic diseases
- Drug reactions
- Neoplasms
- Gastrointestinal disorders
- Musculoskeletal disorders
- Respiratory tract
- Skin diseases
- NOS

D73.2 Disorders with decreased eosinophil counts
- Iatrogenic (corticosteroids, epinephrine)
- Infections
- NOS



D73.8 Other diseases of spleen

Fibrosis of spleen NOS
Perisplenitis
Splenitis NOS

D73.9 Disease of spleen, unspecified

D74 Methaemoglobinaemia

D74.0 Congenital methaemoglobinaemia

Congenital NADH-methaemoglobin reductase deficiency
Haemoglobin-M [Hb-M] disease
Methaemoglobinaemia, hereditary

D74.8 Other methaemoglobinaemias

Acquired methaemoglobinaemia (with sulfhaemoglobinaemia)
Toxic methaemoglobinaemia

Use additional external cause code (Chapter XX), if desired, to identify cause.

D74.9 Methaemoglobinaemia, unspecified

D74. MONOCYTES: Disorders of Monocyte Number

D74.1 Disorders with increased monocyte counts
- Primary hematologic disease
- Inflammatory and immune disorders
- Connective tissue disease
- Infections
- Gastrointestinal disorder
- NOS

D74.2 Disorders with decreased monocyte counts
- Primary hematologic disease (aplastic anemia, hairy cell leukemia)
- NOS



D75 Other diseases of blood and blood-forming organs

Excludes: enlarged lymph nodes (R59.-)
hypergammaglobulinaemia NOS (D89.2)
lymphadenitis:
- NOS (I88.9)
- acute (L04.-)
- chronic (I88.1)
- mesenteric (acute)(chronic) (I88.0)

D75.0 Familial erythrocytosis

Polycythaemia:
- benign
- familial

Excludes: hereditary ovalocytosis (D58.1)

D75.1 Secondary polycythaemia

Erythrocytosis NOS
Polycythaemia:
- acquired
- due to:
  - erythropoietin
  - fall in plasma volume
  - high altitude
  - stress
- emotional
- hypoxaemic
- nephrogenous

D75. Disorders of Lymphocyte Number

D75.1. Disorders with increased lymphocyte counts
- Primary lymphocytosis
- Reactive lymphocytosis
- Mononucleosis syndrome
- Bordetella pertussis
- Stress lymphocytosis
- Hypersensitivity reactions
- Persistent lymphocytosis
- NOS

D75.2 Disorders with decreased lymphocyte counts
- Inherited
- Acquired
  - Aplastic anemia
  - Infection
  - Iatrogenic
  - Systemic disease
  - Nutritional/dietary
- Idiopathic
- NOS

· relative ***Excludes:*** polycythaemia: · neonatorum (P61.1) · vera (D45) **D75.2 Essential thrombocytosis** ***Excludes:*** essential (haemorrhagic) thrombocythaemia (D47.3) **D75.8 Other specified diseases of blood and blood-forming organs** Basophilia **D75.9 Disease of blood and blood-forming organs, unspecified** **D76 Certain diseases involving lymphoreticular tissue and reticulohistiocytic system** ***Excludes:*** Letterer-Siwe disease (C96.0) malignant histiocytosis (C96.1) reticuloendotheliosis or reticulosis: · histiocytic medullary (C96.1) · leukaemic (C91.4) · lipomelanotic (I89.8) · malignant (C85.7) · nonlipid (C96.0)	**D76. Disorders of Lymphocyte Function** D76.1 Combined Immunodeficiency (defined as defects in function of both T and B lymphocytes, but some combined immunodeficiency syndromes may also have reduced number and function of natural killer cells and neutrophils) • T(-) B(+); SCID syndromes with absent or severely decreased T cells; B cells present but with defective function. ▪ gamma c deficiency

D76.0 Langerhans' cell histiocytosis, not elsewhere classified Eosinophilic granuloma Hand-Schüller-Christian disease Histiocytosis X (chronic) **D76.1 Haemophagocytic lymphohistiocytosis** Familial haemophagocytic reticulosis Histiocytoses of mononuclear phagocytes other than Langerhans' cells NOS **D76.2 Haemophagocytic syndrome, infection-associated** Use additional code, if desired, to identify infectious agent or disease. **D76.3 Other histiocytosis syndromes** Reticulohistiocytoma (giant-cell) Sinus histiocytosis with massive lymphadenopathy Xanthogranuloma	▪ JAK3 deficiency ▪ L7Ralpha deficiency ▪ CD45 deficiency ▪ CD3 delta, epsilon or theta deficiency ▪ NOS. • T(+) B(-); SCID syndromes with T cells present but with defective function; B cells absent or severely decreased in number. ▪ RAG1/2 deficiency ▪ DCLRE1c/Artemis deficiency ▪ Adenosine deaminase (ADA) deficiency ▪ Reticular dysgenesis ▪ NOS. • T(+) B (+); Combined immunodeficiency with normal or modestly decreased T cells and B cells but variable functional defects in both T cells and B cells. ▪ DNA ligase IV deficiency ▪ Cernunnos/XLF deficiency ▪ CD40 ligand deficiency ▪ CD40 deficiency ▪ Purine nucleoside phosphorylase (PNP) deficiency ▪ CD3 gamma deficiency ▪ CD8 alpha deficiency ▪ DOCK8 deficiency ▪ ZAP-70 deficiency ▪ Calcium channels deficiencies ▪ MHC class I deficiency ▪ MHC class II deficiency

- Winged helix deficiency
- CD25 deficiency
- STAT5b deficiency
- NOS.

- Combined immune deficiency associated with severe autoimmune syndrome that may include severe eczyma (e.g. Omenn syndrome).

D76.2. Predominantly Antibody Deficiencies

- Severe reduction in all serum immunoglobulin isotypes with profoundly decreased or absent B cells.
  - Btk deficiency
  - Mu chain deficiency
  - Lambda 5 deficiency, Ig alpha deficiency, Ig beta deficiency, BLNK deficiency, Thymoma with immunodeficiency (Good's syndrome), Myelodysplasias, and NOS.
- Common variable immunodeficiency: Severe reduction in serum IgG and IgA with variable B cell number.
  - mutations in TACI, BAFFR or Msh5
  - ICOS deficiency
  - CD19 deficiency
  - X-linked lymphoproliferative syndrome
  - NOS.
- Severe reduction in serum IgG and IgA with normal/elevated IgM and normal numbers of B cells.
  - CD40 ligand deficiency

  - CD40 deficiency
  - Activation induced cytidine deaminase (AICD) deficiency
  - UNG deficiency
  - NOS
- Isotype or light chain deficiencies with normal B cells number
  - Ig heavy chains deletions
  - Kappa chain deficiency,
  - Isolated IgG subclass deficiency
  - IgA deficiency associated with IgG subclass deficiency
  - Selective IgA deficiency
  - NOS.
- Specific antibody deficiency with normal Ig concentrations and normal B cell numbers
- Transient hypogammaglobulinemia of infancy with normal numbers of B cells

D76.3 Diseases of immune dysregulation

- Immunodeficiency with hypopigmentation
  - Chediak-Higashi syndrome (C-H syndrome may also have neutrophil dysfunction)
  - Griscelli syndrome type 2 a
  - Hermansky-Pudlak type 2.
- Hemophagocytic syndromes
  - Familial Hemophagocytic Lymphohistiocytosis (excludes malignant histiocytic disorders, Histiocytosis X and related disorders).
    - Perforin mutation
    - munc 13-4 mutation

	○ syntaxin 11 mutation ○ NOS. • HLH associated with other genetic immunodeficiency syndromes ○ Chediak-Higashi syndrome ○ Griscelli syndrome ○ X-linked lymphoproliferative disease. • Acquired hemophagocytic lymphohistiocytosis syndromes. ○ Infection-Associated HLH ○ Autoimmune disease-associated HLH (Macrophage activation syndrome) ○ Lymphoma-associated HLH ○ Idiopathic HLH. • Lymphoproliferative syndrome without autoimmunity ○ SH2D1A deficiency (XLP1) ○ XIAP deficiency (XLP2) ○ ITK deficiency ○ NOS. • Lymphoproliferative syndrome with autoimmunity: Autoimmune lymphoproliferative syndrome (ALPS). ○ ALPS type 1a (CD95/Fas defects), ○ ALPS type 1b (Fas ligand defects), ALPS type 2a (Caspase 10 defects), ALPS type 2b (Caspase 8 defects), ALPS NRAS

	(defects in NRAS) ○ Autoimmune polyendocrinopathy candidiasis and ectodermal dystrophy (APECED) ○ Immune dysregulation, polyendocrinopathy, enteropathy X-linked (IPEX) D76.4. Autoinflammatory Syndromes including Periodic Fever: • Familial mediterranean fever • TRAPS syndrome • Hyper IgD syndrome • Cryopirin/NALP3 deficiency associated syndromes ○ INCA/NOMID ○ Muckle-Wells ○ familial cold autoinflammatory syndrome), ○ PAPA syndrome ○ Blau syndrome ○ Majeed/CRMO syndrome. D76.5. Complement Deficiencies including deficiencies of complement regulatory proteins • C1q, C1r, C1s deficiency • C4 deficiency • C2, C3, C5, C6, C7 deficiency, • C8a, C8b deficiency, • C9 deficiency • C1 inhibitor deficiency • Factor I deficiency • Factor H deficiency • Factor D deficiency

	• Properdin deficiency • MBP deficiency • MASP2 deficiency • CD46 deficiency • CD59 deficiency • Paroxysmal nocturnal hemoglobinuria. D76.6 Other Well-Defined Immunodeficiency Syndromes • Defects in Innate Immunity associated with recurrent infections (may also include patients with some types of combined immunodeficiency). o Ectodermal dysplasia with immunodeficiency (caused by NEMO deficiency or IkB alpha deficiency) o IRAK4 deficiency o MyD88 deficiency o Warts hypogammaglobulinemia infections myelokathexis (WHIM) syndrome o Epidermodysplasia verruciformis o UNC93b1 deficiency o TLR3 deficiency. • Defects in Acquired Immunity. o Wiskott-Aldrich syndrome o DNA repair defects (including Ataxia-telangiectasia, Ataxia-telangiectasia like disease) o Nijmegen breakage syndrome, Nijmegen breakage like syndrome, o Bloom syndrome. • Defect of Thymus development

D77* **Other disorders of blood and blood-forming organs in diseases classified elsewhere** Fibrosis of spleen in schistosomiasis [bilharziasis] (B65.-+)	(DiGeorge syndrome). • Immune defects with bone abnormalities including Cartilage-hair hypoplasia and Schimke syndrome • Hyper IgE syndromes. o Autosomal dominant Stat3 deficiency (Job's syndrome) o Autosomal recessive Tyk2 deficiency o Dock 8 deficiency (also included in combined immunodeficiency group). • Chronic mucocutaneous candidiasis • Hepatic veno-occlusive disease with immunodeficiency Hoyerall-Hreidarsson syndrome **D77. Diseases of the Spleen** D77.1 Asplenia • post-surgical • congenital D77.2 Atrophy D77.3 Laceration, spleen • Rupture, spleen • Traumatic • Non-traumatic D77.4 Accessory (supernumerary) spleen D77.5 Splenosis D77.6 Ectopic spleen D77.7 Pseudocyst D77.8 Epithelial cyst D77.9 Splenomegaly • Lymphoid hyperplasia • Congestion D77.10 Chronic congestive splenomegaly

D77.11 Hypersplenism
D77.12 Storage diseases
D77.13 Acute septic splenitis (septic spleen)
D77.14 Abscess
D77.15 Splenic infarction
D77.16 Peliosis

D78. Tumor-like Conditions of the Spleen

D78.1 Splenic hamartoma
D78.2 Inflammatory pseudotumor
D78.3 Sclerosing angiomatoid nodular transformation (SANT)

D79. Primary Neoplasms of the Spleen (Exclusive to Spleen)

Littoral cell angioma

Certain disorders involving the immune mechanism (D80-D89)

Includes: defects in the complement system
immunodeficiency disorders, except human immunodeficiency virus [HIV] disease
sarcoidosis

Excludes: autoimmune disease (systemic) NOS (M35.9)
functional disorders of polymorphonuclear neutrophils (D71)
human immunodeficiency virus [HIV] disease (B20-B24)

D80 Immunodeficiency with predominantly antibody defects

D80.0 Hereditary hypogammaglobulinaemia

Autosomal recessive agammaglobulinaemia (Swiss type)
X-linked agammaglobulinaemia [Bruton] (with growth hormone deficiency)

D80.1 Nonfamilial hypogammaglobulinaemia

Agammaglobulinaemia with immunoglobulin-bearing B-lymphocytes
Common variable agammaglobulinaemia [CVAgamma]
Hypogammaglobulinaemia NOS

Certain disorders involving the immune mechanism (D80-D89)

Includes: defects in the complement system
immunodeficiency disorders, except human immunodeficiency virus [HIV] disease
sarcoidosis

Excludes: autoimmune disease (systemic) NOS (M35.9)
functional disorders of polymorphonuclear neutrophils (D71)
human immunodeficiency virus [HIV] disease (B20-B24)
human immunodeficiency virus [HIV] disease complicating
pregnancy childbirth and the puerperium (O98.7)

D80 Immunodeficiency with predominantly antibody defects
D80.0 Hereditary hypogammaglobulinaemia
Autosomal recessive agammaglobulinaemia (Swiss type)
X-linked agammaglobulinaemia [Bruton] (with growth hormone deficiency)
D80.1 Nonfamilial hypogammaglobulinaemia
Agammaglobulinaemia with immunoglobulin-bearing B-lymphocytes
Common variable agammaglobulinaemia [CVAgamma]
Hypogammaglobulinaemia NOS

D80.2 Selective deficiency of immunoglobulin A [IgA]

D80.3 Selective deficiency of immunoglobulin G [IgG]

D80.2 Selective deficiency of immunoglobulin A [IgA]	**subclasses**
D80.3 Selective deficiency of immunoglobulin G [IgG] subclasses	**D80.4 Selective deficiency of immunoglobulin M [IgM]**
D80.4 Selective deficiency of immunoglobulin M [IgM]	**D80.5 Immunodeficiency with increased immunoglobulin M [IgM]**
D80.5 Immunodeficiency with increased immunoglobulin M [IgM]	**D80.6 Antibody deficiency with near-normal immunoglobulins or with hyperimmunoglobulinaemia**
D80.6 Antibody deficiency with near-normal immunoglobulins or with hyperimmunoglobulinaemia	**D80.7 Transient hypogammaglobulinaemia of infancy**
D80.7 Transient hypogammaglobulinaemia of infancy	**D80.8 Other immunodeficiencies with predominantly antibody defects** Kappa light chain deficiency
D80.8 Other immunodeficiencies with predominantly antibody defects Kappa light chain deficiency	**D80.9 Immunodeficiency with predominantly antibody defects, unspecified**
D80.9 Immunodeficiency with predominantly antibody defects, unspecified	
D81 Combined immunodeficiencies ***Excludes:*** autosomal recessive agammaglobulinaemia (Swiss type) (	**D81 Combined immunodeficiencies** Excludes: autosomal recessive agammaglobulinaemia (Swiss type) (

D80.0)	D80.0)
D81.0 Severe combined immunodeficiency [SCID] with reticular dysgenesis	**D81.0 Severe combined immunodeficiency [SCID] with reticular dysgenesis**
D81.1 Severe combined immunodeficiency [SCID] with low T- and B-cell numbers	**D81.1 Severe combined immunodeficiency [SCID] with low T- and B-cell numbers**
D81.2 Severe combined immunodeficiency [SCID] with low or normal B-cell numbers	**D81.2 Severe combined immunodeficiency [SCID] with low or normal B-cell numbers**
D81.3 Adenosine deaminase [ADA] deficiency	**D81.3 Adenosine deaminase [ADA] deficiency**
D81.4 Nezelof's syndrome	**D81.4 Nezelof's syndrome**
D81.5 Purine nucleoside phosphorylase [PNP] deficiency	**D81.5 Purine nucleoside phosphorylase [PNP] deficiency**
D81.6 Major histocompatibility complex class I deficiency Bare lymphocyte syndrome	**D81.6 Major histocompatibility complex class I deficiency** Bare lymphocyte syndrome
D81.7 Major histocompatibility complex class II deficiency	**D81.7 Major histocompatibility complex class II deficiency**
D81.8 Other combined immunodeficiencies Biotin-dependent carboxylase deficiency	**D81.8 Other combined immunodeficiencies** Biotin-dependent carboxylase deficiency
D81.9 Combined immunodeficiency, unspecified	**D81.9 Combined immunodeficiency, unspecified** Severe combined immunodeficiency disorder [SCID] NOS

Severe combined immunodeficiency disorder [SCID] NOS	
D82 **Immunodeficiency associated with other major defects**	**D82** **Immunodeficiency associated with other major defects**
Excludes: ataxia telangiectasia [Louis-Bar] (G11.3)	Excludes: ataxia telangiectasia [Louis-Bar] (G11.3)
D82.0 Wiskott-Aldrich syndrome	**D82.0 Wiskott-Aldrich syndrome**
Immunodeficiency with thrombocytopenia and eczema	Immunodeficiency with thrombocytopenia and eczema
D82.1 Di George's syndrome	**D82.1 Di George's syndrome**
Pharyngeal pouch syndrome Thymic: · alymphoplasia · aplasia or hypoplasia with immunodeficiency	Pharyngeal pouch syndrome Thymic: • alymphoplasia • aplasia or hypoplasia with immunodeficiency
D82.2 Immunodeficiency with short-limbed stature	**D82.2 Immunodeficiency with short-limbed stature**
D82.3 Immunodeficiency following hereditary defective response to Epstein-Barr virus	**D82.3 Immunodeficiency following hereditary defective response to Epstein-Barr virus**
X-linked lymphoproliferative disease	X-linked lymphoproliferative disease
D82.4 Hyperimmunoglobulin E [IgE] syndrome	**D82.4 Hyperimmunoglobulin E [IgE] syndrome**

D82.8 Immunodeficiency associated with other specified major defects	**D82.8 Immunodeficiency associated with other specified major defects**
D82.9 Immunodeficiency associated with major defect, unspecified	**D82.9 Immunodeficiency associated with major defect, unspecified**
D83 Common variable immunodeficiency	**D83 Common variable immunodeficiency**
D83.0 Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function	**D83.0 Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function**
D83.1 Common variable immunodeficiency with predominant immunoregulatory T-cell disorders	**D83.1 Common variable immunodeficiency with predominant immunoregulatory T-cell disorders**
D83.2 Common variable immunodeficiency with autoantibodies to B- or T-cells	**D83.2 Common variable immunodeficiency with autoantibodies to B- or T-cells**
D83.8 Other common variable immunodeficiencies	**D83.8 Other common variable immunodeficiencies**
D83.9 Common variable immunodeficiency, unspecified	**D83.9 Common variable immunodeficiency, unspecified**
D84 Other immunodeficiencies	**D84 Other immunodeficiencies**
D84.0 Lymphocyte function antigen-1 [LFA-1] defect	**D84.0 Lymphocyte function antigen-1 [LFA-1] defect**
	D84.1 Defects in the complement system

D84.1 **Defects in the complement system** C1 esterase inhibitor [C1-INH] deficiency **D84.8** **Other specified immunodeficiencies** **D84.9** **Immunodeficiency, unspecified** **D86** **Sarcoidosis** **D86.0** **Sarcoidosis of lung** **D86.1** **Sarcoidosis of lymph nodes** **D86.2** **Sarcoidosis of lung with sarcoidosis of lymph nodes** **D86.3** **Sarcoidosis of skin** **D86.8** **Sarcoidosis of other and combined sites** Iridocyclitis in sarcoidosis+ (H22.1*) Multiple cranial nerve palsies in sarcoidosis+ (G53.2*) Sarcoid: · arthropathy+ (M14.8*) · myocarditis+ (I41.8*) · myositis+ (M63.3*) Uveoparotid fever [Heerfordt] **D86.9** **Sarcoidosis, unspecified**	C1 esterase inhibitor [C1-INH] deficiency **D84.8** **Other specified immunodeficiencies** **D84.9** **Immunodeficiency, unspecified** **D86** **Sarcoidosis** **D86.0** **Sarcoidosis of lung** **D86.1** **Sarcoidosis of lymph nodes** **D86.2** **Sarcoidosis of lung with sarcoidosis of lymph nodes** **D86.3** **Sarcoidosis of skin** **D86.8** **Sarcoidosis of other and combined sites** Iridocyclitis in sarcoidosis+ (H22.1*) Multiple cranial nerve palsies in sarcoidosis+ (G53.2*) Sarcoid: • arthropathy+ (M14.8*) • myocarditis+ (I41.8*) • myositis+ (M63.3*) Uveoparotid fever [Heerfordt] **D86.9** **Sarcoidosis, unspecified**

D89 **Other disorders involving the immune mechanism, not elsewhere classified** ***Excludes:*** hyperglobulinaemia NOS (R77.1) monoclonal gammopathy (D47.2) transplant failure and rejection (T86.-) **D89.0** **Polyclonal hypergammaglobulinaemia** Benign hypergammaglobulinaemic purpura Polyclonal gammopathy NOS **D89.1** **Cryoglobulinaemia** Cryoglobulinaemia: · essential · idiopathic · mixed · primary · secondary Cryoglobulinaemic: · purpura · vasculitis **D89.2** **Hypergammaglobulinaemia, unspecified** **D89.8** **Other specified disorders involving the immune mechanism, not elsewhere classified** **D89.9** **Disorder involving the immune mechanism, unspecified**	**D89** **Other disorders involving the immune mechanism, not elsewhere classified** Excludes: hyperglobulinaemia NOS (R77.1) monoclonal gammopathy (D47.2) transplant failure and rejection (T86.-) **D89.0** **Polyclonal hypergammaglobulinaemia** Benign hypergammaglobulinaemic purpura Polyclonal gammopathy NOS **D89.1** **Cryoglobulinaemia** Cryoglobulinaemia: • essential • idiopathic • mixed • primary • secondary Cryoglobulinaemic: • purpura • vasculitis **D89.2** **Hypergammaglobulinaemia, unspecified** **D89.8** **Other specified disorders involving the immune mechanism, not elsewhere classified** **D89.9** **Disorder involving the immune mechanism, unspecified** Immune disease NOS

Immune disease NOS	

D60	Acquired pure red cell aplasia [erythroblastopenia] Includes: red cell aplasia (acquired)(adult)(with thymoma)	**D60** D60.0 Acquired transient pure red cell aplasia • acute B19 parvovirus infection in hemolytic disease • transient erythroblastopenia of childhood D60.1Acquired chronic pure red cell aplasia, idiopathic D60.2 Acquired chronic pure red cell aplasia, secondary • thymoma • malignancy • viruses including persistent B19 parvovirus infection • drugs • pregnancy • autoimmune disorders • post stem cell transplant • antierythropoietin antibodies D60.3 Acquired pure red cell aplasia NOS D60.4 Inherited pure red cell aplasia • Diamond-Blackfan anaemia D60.5 Fetal red cell aplasia (non-immune hydrops fetalis)
D60.0	Chronic acquired pure red cell aplasia	
D60.1	Transient acquired pure red cell aplasia	
D60.8	Other acquired pure red cell aplasias	
D60.9	Acquired pure red cell aplasia, unspecified	
D61	Other aplastic anaemias Excludes: agranulocytosis (D70)	**D61 *Aplastic anaemia***
D61.0	Constitutional aplastic anaemia Aplasia, (pure) red cell (of): · congenital · infants · primary Blackfan-Diamond syndrome Familial hypoplastic anaemia Fanconi's anaemia Pancytopenia with malformations	D61.0 Hereditary aplastic anaemia • Fanconi anaemia • dyskeratosis congenital • Dchwachma-Diamond syndrome • other rare syndromes D61.1 Hereditary aplastic anaemia NOS D61.2 Acquired aplastic anaemia, idiopathic D61.3 Acquired aplastic anaemia, secondary • drugs (other than cytotoxic drugs) • toxins • viruses • autoimmune disorders • pregnancy • iatrogenic (radiation or cytotoxic drug therapy)
D61.1	Drug-induced aplastic anaemia Use additional external cause code (Chapter XX), if desired, to identify drug.	
D61.2	Aplastic anaemia due to other external agents	

Use additional external cause code (Chapter XX), if desired, to identify cause. D61.3 Idiopathic aplastic anaemia D61.8 Other specified aplastic anaemias D61.9 Aplastic anaemia, unspecified Hypoplastic anaemia NOS Medullary hypoplasia Panmyelophthisis	D61.5 PNH(paroxysmal nocturnal hemoglobinuria) in the setting of bone marrow failure D61.4 Acquired aplastic anaemia NOS • hypoplastic anemia NOS
D62 Acute posthaemorrhagic anaemia Excludes: congenital anaemia from fetal blood loss (P61.3)	**D62 *(D46) Myelodysplastic syndrome (MDS)*** D62.1 Refractory cytopenia with unilineage dysplasia (RCUD) • refractory anaemia (RA) • refractory neutopenia (RN) • refractory thrombocytopenia (RT) D62.2 Refractory anaemia with ring sideroblasts (RARS) D62.3 Refractory cytopenia with mutilleneage dysplasia (RCMD) D62.4 Refractory anaemia with excess blasts (RAEB) • RAEB-1 • RAEB-2 D62.5 Myelodysplastic syndrome associated with isolated del(5q) D62.6 Myelodysplastic syndrome, unclassifiable (MDS-U) D62.7 Childhood myelodysplastic syndrome • refractory cytopenia of childhood (RCC) D62.8 Therapy-related myelodysplastic syndrome • alkylating agents-related • topoisomerase-II inhibitor-related
D63* Anaemia in chronic diseases classified elsewhere D63.0* Anaemia in neoplastic disease (C00-D48+) D63.8* Anaemia in other chronic diseases classified elsewhere	**D63 *Other anaemias*** D 63.1 Anaemia in chronic diseases • inflammation • malignancies • chronic kidney disease

	• endocrine disorders • liver diseases • infections • NOS D63.2 Secondary sideroblastic anaemia • Drugs or toxins • Pyridoxine-responsive NEC • NOS D63.3 Hereditary sideroblastic anaemia D63.4 Congenital dyserythropoietic anaemia D63.5 Anemia NOS
D64 Other anaemias Excludes: refractory anaemia: · NOS (D46.4) · with excess of blasts (D46.2) · with transformation (D46.3) · with sideroblasts (D46.1) · without sideroblasts (D46.0) D64.0 Hereditary sideroblastic anaemia Sex-linked hypochromic sideroblastic anaemia D64.1 Secondary sideroblastic anaemia due to disease Use additional code, if desired, to identify disease. D64.2 Secondary sideroblastic anaemia due to drugs and toxins Use additional external cause code (Chapter XX), if desired, to identify cause. D64.3 Other sideroblastic anaemias Sideroblastic anaemia: · NOS · pyridoxine-responsive NEC D64.4 Congenital dyserythropoietic anaemia	

Dyshaematopoietic anaemia (congenital) Excludes: Blackfan-Diamond syndrome (D61.0) Di Guglielmo's disease (C94.0) D64.8 Other specified anaemias Infantile pseudoleukaemia Leukoerythroblastic anaemia D64.9 Anaemia, unspecified	**Issues to be discussed with other WGs** ✓ Location and the title of category where MDS was listed. MDS listed under the category D46 in ICD-10 that has been tentatively relocated here under Category D. ✓ Anaemia due to acute blood loss has been deleted as it is certainly not a blood disease. ✓ Inclusion of cytopenia due to hypoproduction such as Inherited or acquired amegakaryocytic thrombocytopenia, Kostmann syndrome, severe congenital neutropenia thrombocytopenia absent radil, refractory neutropenia, refractory thrombocytopenia etc.) in this section needs to be discussed. ✓ PNH in the setting of another specified bone marrow disorder needs to be incorporated in somewhere. ✓ Vitamin B6 (pyridoxine)- responsive sideroblastic anaemia can be transferred to the section of nutritional anemia. ***Code 75 Other diseases of blood and blood-forming organs*** Exclude; Polycythemia vera (D47) Primary myelofibrosis (D47) Essential thrombocythemia (D47) D75.0 Primary familial and congenital polycythemia • Chuvash polycythemia D75.1 Secondary erythrocytosis • physiological overproduction of erythropoietin

	• pathological overproduction of erythropoietin • drug D75.3 Relative erythrocytosis • Gaisbeck syndrome D75.4 Erythrocytosis NOS D75.5 Familial thrombocytosis D75.6 Reactive (secondary) thrombocytosis • transient reactive process • sustained process • thrombocytosis NOS D75.4 Secondary myelofibrosis • neoplasms • infection • myelofibrosis NOS D75.8 Other specified diseases of blood and blood forming organs • Basophilia (relocate to D72) D75.9 Disease of blood and blood-forming organs, unspecified D47 Myeloproliferative neoplasms and others Exclude; Chronic myelogemous leukemia, BCR-ABL 1 positive (C92.1) Monoclonal gammopathy (C90?) D47.1 Polycythemia vera D47.2 Primary myelofibrosis D47.3 Essential thrombocythemia D47.4 Chronic eosinophilic leukaemia, NOS D47.5 Chronic neutrophilic leukaemia D47.6 Mastocytosis • cutaneous mastocytosis • mastocytosis • mast cell sarcoma • extracutaneous mastocytoma D47.7 Myeloproliferative neoplasm, unclassifiable D47.8 Myeloid and lymphoid neoplasms with eosinophilia and

	abnormalities of PDGFRA, PDGFRB or FGFR1 **Issues to be discussed with other WGs** ✓ The section of Myeloproliferative disorders (ICD10) should be relocated into C code (C96.7 seems appropriate). ✓ Chronic eosinophilic leukemia (D47.4) and Chronic neutrophilic leukemia (D 47.5) should be relocated to C95.9 ✓ The section of monoclonal gammopathy should be combined with MDS as pre-neoplastic disorder? The section in which quantitative disorders of monocot should be created and combined with the same disorders of eoshinophila

D65 Disseminated intravascular coagulation [defibrination syndrome] Afibrinogenaemia, acquired Consumption coagulopathy Diffuse or disseminated intravascular coagulation [DIC] Fibrinolytic haemorrhage, acquired Purpura: · fibrinolytic · fulminans Excludes: that (complicating): · abortion or ectopic or molar pregnancy (O00-O07 , O08.1) · in newborn (P60) · pregnancy, childbirth and the puerperium (O45.0 , O46.0 , O67.0 , O72.3)	**D65** Purpura and other haemorrhagic conditions D65.0 Hereditary vascular purpura Hereditary Hemorrhagic Telangiectasia (HHT) Ehlers-Danlos disease Marfan Syndrome Other hereditary vasculopathies D65.1 Acquired vascular purpura Henoch-Schönlein Purpura cryoglobulinaemic purpura benign hypergammaglobulinaemic purpura Other vascular purpura D65.2 Other specified vascular purpura D65.3 Unspecified vascular purpura
D66 Hereditary factor VIII deficiency Deficiency factor VIII (with functional defect) Haemophilia: · NOS · A · classical Excludes: factor VIII deficiency with vascular defect (D68.0)	**D66** Platelet disorders D66.1 Hereditary thrombocytopenia Congenital amegalyocytic thrombocytopenia Thrombocytopenia with absent radius (TAR) syndrome Mutations in MYH9 gene May-Hegglin anomaly Sebastian Syndrome Fectner Syndrome Epstein Syndrome Wiskott-Aldrich syndrome Hereditary thrombotic thrombocytopenic purpura (Upshaw-Schulman syndrome) Other hereditary thrombocytopenia

	D66.2 Acquired thromboocyytopenia primary immune thrombocytopenia secondary immune thrombocytopenia. Drugs SLE Neonatal (alloimune) thrombocytopenia Post-transfusion purpura Evans Syndrome Heparin-induced thrombocytopenia Thrombotic thrombocytpenic purpura Hemolytic uremic syndrome Acquired amegalyocytic thrombocytopenia D66.3 Other specified thrombocytopenia D66.4 Unspecified thrombocytopenia D66.5 Hereditary thrombocytosis Congenital or familial thrombocytosis D66.6 Acquired thrombocytosis Essential thrombocytosis Reactive thrombocytosis D66.7 Other specified thrombocytosis D66.8 Unspecified thrombocytosis D66.9 Inherited qualitative platelet disorders Bernard-Soulier syndrome Glanzmann's thrombasthenia Gray platelet syndrome Isolated δ-storage pool disease Hermansky-Pudalak Syndrome Chediak-Higashi Syndrome

D67 Hereditary factor IX deficiency Christmas disease Deficiency: · factor IX (with functional defect) · plasma thromboplastin component [PTC] Haemophilia B	Other inherited qualitative platelet disorders D66.10 Acquired qualitative platelet disorders Myelodysplastic syndromes Drug-induced functional platelet abnormality D66.11 Other specified qualitative platelet disorders D66.12 Unspecified qualitative platelet disorders **D67** Coagulation defects D67.0 Hereditary von Willebrand disease Type 1Fanconi anaemia Type 2A Type 2B Type 2M Type 2N TYPE 3 With VWF inhibitor Platelet type D67.1 Congenital coagulation defects Hemophilia A With FVIII inhibitor Hemophilia B With FIX inhibitor Other congenital defects of intrinsic pathway Factor XI deficiency Factor XII deficiency Prekallikrein deficiency High-molecular-weight kininogen deficiency Congenital defects of extrinsic pathway Factor VII deficiency Congenital defects of common pathway