CHAPTER 03

Diseases of the blood or blood-forming organs

This chapter has 55 four-character categories.

Code range starts with 3A00

This chapter includes diseases of the blood as well as diseases of blood forming organs.

Exclusions: Complications of pregnancy, childbirth or the puerperium (Chapter 18)

Diseases of the immune system (Chapter 04)

Certain conditions originating in the perinatal period (Chapter 19)

Injury, poisoning or certain other consequences of external causes (Chapter 22)

Human immunodeficiency virus disease (BlockL1‑1C6)

Endocrine, nutritional or metabolic diseases (Chapter 05)

Congenital malformations, deformations or chromosomal abnormalities (Chapter 20)

Other diseases of the blood or blood-forming organs or certain disorders involving the immune mechanism complicating pregnancy, childbirth or the puerperium (JB64.1)

Coded Elsewhere: Neoplasms of haematopoietic or lymphoid tissues (2A20-2B3Z)

Symptoms, signs or clinical findings of blood, blood-forming organs, or the immune system (MA00-MA3Y)

This chapter contains the following top level blocks:

* Anaemias or other erythrocyte disorders
* Coagulation defects, purpura or other haemorrhagic or related conditions
* Diseases of spleen

Anaemias or other erythrocyte disorders (BlockL1‑3A0)

Inclusions: Anaemia, unspecified

Coded Elsewhere: Anaemia complicating pregnancy, childbirth or the puerperium (JB64.0)

Anaemia of prematurity (KA8B)

Nutritional or metabolic anaemias (BlockL2‑3A0)

3A00 Iron deficiency anaemia

A disease caused by chronic or acute bleeding, excessive menstrual bleeding, inadequate intake, substances (in diet or drugs) interfering with iron absorption, malabsorption syndromes, inflammation, infection or blood donation. This disease is characterised by decreased levels of iron present in the body. This disease may present with fatigue, pallor or dizziness. Confirmation is by identification of decreased levels of iron in a blood sample.

3A00.0 Acquired iron deficiency anaemia due to blood loss

Acute or chronic blood loss is a possible cause in every case of iron-deficiency anaemia. Iron deficiency anaemia may be caused by acute bleeding in gastrointestinal tract, uterus or genitourinary system, copious menstrual blood losses (menorrhagia) and multiple blood donations. In many tropical countries, infestations with hookworms lead to intestinal blood losses that in some individuals can be considerable. Iron deficiency may also be caused by several circumstances related to “chronic posthaemorrhagic anaemia”. A diagnosis of iron deficiency should always lead to a search for pathologic causes of blood loss (e.g. tumours in the gastrointestinal tract or uterus, especially if uterine bleedings have increased or changed in regularity).

Exclusions: congenital anaemia from fetal blood loss (KA8C)

3A00.00 Acute posthaemorrhagic anaemia

A disease caused by blood loss such as subsequent to trauma. This disease is characterised by loss of blood from the body leading to low levels of red blood cells/blood in the body. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low levels of red blood cells in a blood sample.

Exclusions: congenital anaemia from fetal blood loss (KA8C)

Anaemia due to acute disease (3A90)

3A00.01 Chronic posthaemorrhagic anaemia

Chronic iron-deficiency anaemia from bleeding may be caused by colon cancer, gastric cancer, peptic ulcer, Meckel diverticulum, hiatal hernia with linear erosions, colonic vascular ectasia, colonic polyps, haemangioma, inflammatory bowel disease, tumours in the gastrointestinal tract or uterus, and chronic menorrhagia. Some infants with severe iron deficiency have chronic intestinal blood loss induced by exposure to cow's milk protein. Repeated phlebotomy for blood tests is a cause of anaemia of prematurity.

3A00.0Z Acquired iron deficiency anaemia due to blood loss, unspecified

3A00.1 Acquired iron deficiency anaemia due to low intake

Iron deficiency is probably the most common nutritional deficiency disorder in the world. Iron deficiency anaemia during pregnancy increases perinatal risks for mothers and neonates; and increases overall infant mortality. Severe anaemia is a major risk factor associated with greatly increased morbidity and mortality for young children and pregnant women. Prompt recognition of the condition, treatment and clinical follow-up of individuals, are crucial in avoiding complications such as high-output heart failure. Maternal iron deficiency during pregnancy increases the risk of iron deficiency in the infant. In less developed countries, the prevalence of iron deficiency during pregnancy is higher than in developed countries, and iron supplementation during pregnancy is beneficial.

3A00.2 Acquired iron deficiency anaemia due to decreased absorption

3A00.3 Acquired iron deficiency anaemia due to increased requirement

3A00.Y Other specified iron deficiency anaemia

3A00.Z Iron deficiency anaemia, unspecified

3A01 Megaloblastic anaemia due to vitamin B12 deficiency

A disease caused by inadequate dietary intake of vitamin B12, impaired absorption of vitamin B12, surgical removal of the small bowel, coeliac disease or inherited mutations affecting absorption of vitamin B12. This disease is characterised by decreased levels of vitamin B12 in the body presenting with or without anaemia. This disease may present with fatigue, pallor, dizziness, seizures, or symptoms of dementia. Confirmation is by identification of decreased levels of vitamin B12 in a blood sample.

3A01.0 Hereditary Vitamin B12 deficiency anaemia

This is a hereditary low blood level of vitamin B12. It can cause permanent damage to nervous tissue if left untreated long enough. Vitamin B12 itself was discovered through investigation of pernicious anaemia, which is an autoimmune disease that destroys parietal cells in the stomach that secrete intrinsic factor.

3A01.1 Neonatal vitamin B12 deficiency anaemia

A disease caused by a lack of vitamin B12 in the mother, which is passed onto the fetus in the antenatal period or to the neonate during breast feeding. This disease is characterised by decreased levels of vitamin B12. This disease may present with increased risk of birth defects or preterm delivery, anaemia, irritability, failure to thrive or apathy. Confirmation is by identification of low levels of vitamin B12 in a blood sample.

Exclusions: Hereditary Vitamin B12 deficiency anaemia (3A01.0)

3A01.2 Vitamin B12 deficiency anaemia due to low intake

A disease caused by insufficient intake of vitamin B12 into the body. This disease is characterised by low levels of vitamin B12 leading to low levels of red blood cells. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low levels of vitamin B12 and red blood cell count in a blood sample.

3A01.3 Vitamin B12 deficiency anaemia due to intrinsic factor deficiency

Exclusions: Vitamin B12 deficiency anaemia due to congenital intrinsic factor deficiency (3A01)

3A01.30 Pernicious anaemia

Biermer's disease, also called acquired pernicious anaemia, is a disorder in vitamin B12 (cobalamin) absorption characterised by megaloblastic anaemia and gastrointestinal symptoms, and that can lead to neurological abnormalities.

3A01.3Y Other specified vitamin B12 deficiency anaemia due to intrinsic factor deficiency

3A01.4 Vitamin B12 deficiency anaemia due to intestinal disease

A number of intestinal disorders can also cause vitamin B12 (cobalamin) deficiency. These include severe pancreatic diseases and small bowel diseases such as malabsorption, ileal disease (including tuberculous ileitis, lymphoma, amyloid, long-term survivors of pelvic irradiation), extensive small bowel resection or bypass, gastric surgery/reconstruction for obesity (bariatric surgery) and Crohn's disease. When jejunal blind loops are present, bacterial overgrowth within the loops competes for cobalamin, leading to cobalamin deficiency. Although not as common currently, infestation with the fish tapeworm, Diphyllobothrium latum, was once a classic cause of cobalamin deficiency.

3A01.5 Drug-induced vitamin B12 deficiency anaemia

3A01.Y Other specified megaloblastic anaemia due to vitamin B12 deficiency

3A01.Z Megaloblastic anaemia due to vitamin B12 deficiency, unspecified

3A02 Folate deficiency anaemia

3A02.0 Hereditary folate deficiency anaemia

3A02.1 Folate deficiency anaemia due to low intake

3A02.2 Folate deficiency anaemia due to increased requirements

3A02.3 Folate deficiency anaemia due to decreased intestinal absorption

A disease caused by determinants affecting intestinal absorption of folate arising after birth. This disease is characterised by low levels of folate in the body leading to incomplete formation of red blood cells resulting in large numbers of immature and incompletely developed red blood cells. This disease may present with fatigue, muscle weakness, loss of appetite, weight loss, diarrhoea, nausea, tachycardia or extremity paraesthesia. Confirmation is by identification of low folate levels in a blood sample.

3A02.4 Drug-induced folate deficiency anaemia

3A02.Y Other specified folate deficiency anaemia

3A02.Z Folate deficiency anaemia, unspecified

3A03 Other nutritional or metabolic anaemias

A disease caused by nutritional and metabolic determinants leading to anaemia. This disease is characterised by decreased levels of red blood cells within the body. This disease may present with fatigue, pallor or dizziness. Confirmation is by identification of a decreased red blood cell count in a blood sample.

Coded Elsewhere: Disorders of pyrimidine metabolism (5C55.1)

Lesch-Nyhan syndrome (5C55.01)

3A03.0 Hereditary orotic aciduria

Hereditary orotic aciduria is an extremely rare (less than 20 cases identified worldwide) autosomal recessive disorder characterised by retarded growth, anaemia and excessive urinary excretion of orotic acid. It is due to a severe deficiency in the activity of the pyrimidine pathway enzyme uridine 5'-monophosphate (UMP) synthase (bifunctional enzyme containing two activities: orotate phosphoribosyltransferase and orotidine 5'-monophosphate decarboxylase), coded by a single gene (UMPS) localised to chromosome 3q13.

3A03.1 Protein deficiency anaemia

A disease caused by low levels of protein within the body. This disease is characterised by a low red blood cell count in the blood. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of a low red blood cell count in a blood sample.

Exclusions: Lesch-Nyhan syndrome (5C55.01)

3A03.2 Scorbutic anaemia

Scorbutic anaemia, is a common finding in infants and young children with scurvy and is related to impaired iron absorption and coexistent haematopoietic nutrient deficiencies including iron, vitamin B12 and folate.

3A03.3 Copper deficiency anaemia

Anaemia due to copper deficiency arises from impaired utilization of iron and is therefore a conditioned form of iron deficiency anaemia.

3A03.4 Acquired other vitamin B deficiency anaemia

A disease caused by a lack of B vitamins in the body arising after birth. This disease is characterised by low levels of B vitamins leading to low levels of red blood cells in the body. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low red blood cell count and low B vitamin counts in a blood sample.

3A03.40 Acquired pyridoxine deficiency anaemia

A disease caused by determinants arising after birth. This disease is characterised by low levels of pyridoxine (vitamin B6) leading to low levels of red blood cells in the body. This disease may present with fatigue, muscle weakness, loss of appetite, weight loss, diarrhoea, nausea, fast heartbeat or numbness in extremities. Confirmation is by identification of low levels of pyridoxine and low red blood cell count in a blood sample.

3A03.41 Acquired riboflavin deficiency anaemia

A disease caused by determinants arising after birth. This disease is characterised low levels of riboflavin (vitamin B2) leading to low levels of red blood cells in the body. This disease may present with fatigue, muscle weakness, loss of appetite, weight loss, diarrhoea, nausea, fast heartbeat or numbness in extremities. Confirmation is by identification of low levels of riboflavin and low red blood cell count in a blood sample.

3A03.42 Acquired thiamine deficiency anaemia

A disease caused by a lack of thiamine arising after birth. This disease is characterised low levels of thiamine in the body leading to low levels of red blood cells. This disease may present with fatigue, muscle weakness, loss of appetite, weight loss, diarrhoea, nausea, fast heartbeat or numbness in extremities. Confirmation is by identification of low levels of thiamine and low red blood cell count in a blood sample.

3A03.4Y Other specified acquired other vitamin B deficiency anaemia

3A03.5 Acquired vitamin A deficiency anaemia

3A03.6 Acquired vitamin E deficiency anaemia

Inclusions: Haemolytic anaemia due to vitamin E deficiency

3A03.Y Other and unspecified nutritional or metabolic anaemia

Haemolytic anaemias (BlockL2‑3A1)

A disease caused by determinants arising after birth, during the antenatal period or genetically inherited factors leading to premature haemolysis of red blood cells. This disease is characterised by low levels of red blood cells in the body due to abnormal breakdown of the cells. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low red blood cell count in a blood sample.

Congenital haemolytic anaemia (BlockL3‑3A1)

A disease caused by determinants arising in the antenatal period . This disease is characterised by low levels of red blood cells in the body due to abnormal destruction of the red blood cells. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low red blood cell count in a blood sample.

Coded Elsewhere: Haemolytic disease of fetus or newborn (KA84)

3A10 Hereditary haemolytic anaemia

3A10.0 Haemolytic anaemias due to hexose monophosphate shunt or glutathione metabolism anomalies

This is a form of anaemia due to haemolysis, the abnormal breakdown of red blood cells (RBCs), either in the blood vessels (intravascular haemolysis) or elsewhere in the human body (extravascular). This diagnosis is due to is a process that generates NADPH and pentoses (5-carbon sugars) and glutathione metabolism anomalies.

3A10.00 Haemolytic anaemia due to glucose-6-phosphate dehydrogenase deficiency

Glucose-6-phosphate dehydrogenase (G6PD) deficiency is the most common hereditary erythrocyte enzyme deficiency that can manifest with severe neonatal jaundice which can lead to serious neurological consequences, or, most often, with acute haemolytic anaemia following ingestion of certain foods (fava beans), common drugs (some antimalaria drugs, sulphamides, analgesics), or in the course of an infection, in otherwise asymptomatic individuals.

3A10.0Y Other specified haemolytic anaemias due to hexose monophosphate shunt or glutathione metabolism anomalies

3A10.0Z Haemolytic anaemias due to hexose monophosphate shunt or glutathione metabolism anomalies, unspecified

3A10.1 Haemolytic anaemia due to adenosine deaminase excess

3A10.2 Hereditary elliptocytosis

Hereditary elliptocytosis is a group of rare conditions caused by abnormalities in the red cell cytoskeleton and marked by the presence on blood smears of numerous elliptical red blood cells, called elliptocytes. Clinical presentations are highly heterogeneous ranging from asymptomatic forms to more severe forms associated with variable anaemia, from moderate to severe and with pyropoikilocytosis including fragmented red cells, microelliptocytes and microspherocytes.

3A10.3 Familial pseudohyperkalaemia

A disease caused by a genetically inherited mutation. This disease is characterised by a temperature-dependent defect in red cell membrane permeability to potassium that leads to high in vitro potassium levels in samples stored below 37°C leading to elevated potassium levels in the blood that does not reflect the true potassium level. Confirmation is by identification of genetic mutation through genetic testing.

3A10.Y Other specified hereditary haemolytic anaemia

3A10.Z Hereditary haemolytic anaemia, unspecified

3A1Y Other specified congenital haemolytic anaemia

Acquired haemolytic anaemia (BlockL3‑3A2)

A disease characterised by premature destruction of red blood cells arising after birth. This disease is further characterised by low levels of red blood cells in the body due to abnormal destruction of the cells. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low red blood cell count in a blood sample.

3A20 Acquired haemolytic anaemia, immune

A condition characterised by antibodies that are directed against red blood cells in an autoimmune reaction leading to low levels of red blood cells. This condition may present with pallor, fatigue, shortness of breath. Confirmation is by identification of antibodies in a blood sample and positive Coombs test result.

3A20.0 Autoimmune haemolytic anaemia, warm type

Autoimmune haemolytic anaemia (AIHA) is an autoimmune disorder in which various types of auto-antibodies are directed against red blood cells causing their survival to be shortened and resulting in haemolytic anaemia. AIHA can be primary (idiopathic), secondary to infection or associated with diseases such as B-cell lymphomas, other systemic or organ-specific autoimmune diseases, Hodgkin's disease, hepatitis or primary immunodeficiencies, or, in the case of drug-induced AIHA, caused by a reaction to drugs.

Exclusions: Evans syndrome (3A20.5)

Haemolytic disease of fetus or newborn (KA84)

Paroxysmal cold haemoglobinuria (3A20.3)

3A20.1 Autoimmune haemolytic anaemia, cold type

Cold autoimmune haemolytic anaemia comprises two types of autoimmune haemolytic anaemia (AIHA) defined by the presence of cold autoantibodies (autoantibodies which are active at temperatures below 30°C): cold agglutinin disease (CAD), which is the more common, and paroxysmal cold haemoglobinuria (PCH). CAD is more common in people over the age of 55 years, while PCH typically presents in young children. CAD is caused by IgM autoantibodies while PCH is caused by an IgG immunoglobulin.

Exclusions: Immune thrombocytopenic purpura (3B64.10)

Haemolytic disease of fetus or newborn (KA84)

3A20.2 Autoimmune haemolytic anaemia, mixed type, cold and warm

Mixed autoimmune haemolytic anaemia is a type of autoimmune haemolytic anaemia (AIHA) defined by the presence of both warm and cold autoantibodies, which have a deleterious effect on red blood cells at either body temperature or at lower temperatures.

3A20.3 Paroxysmal cold haemoglobinuria

Paroxysmal cold hemoglobinuria is a very rare subtype of autoimmune haemolytic anaemia (AIHA), caused by the presence of cold-reacting autoantibodies in the blood and characterised by the sudden presence of hemoglobinuria, typically after exposure to cold temperatures. PCH is thought to account for at most 2-10% of cases of AIHA.

3A20.4 Alloimmune haemolytic anaemia

A disease caused by determinants such as a blood transfusion that lead to an immune response directed against the person's own red blood cells. This disease is characterised by low levels of red blood cells in the body due to abnormal destruction of the red blood cells. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low red blood cell count in a blood sample.

Exclusions: Haemolytic disease of fetus or newborn (KA84)

3A20.5 Evans syndrome

Evans syndrome is characterised by the association of autoimmune haemolytic anaemia with another haematological anomaly. The thrombocytopaenia may precede, occur concurrently with, or secondary to the autoimmune haemolytic anaemia.

3A20.Y Other specified acquired haemolytic anaemia, immune

3A21 Acquired haemolytic anaemia, non-immune

A disease caused by determinants such as infection, toxic chemicals, drugs and trauma arising after birth. This disease is characterised by haemolysis of red blood cells. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of decreased red blood cell count in a blood sample and negative Coombs test result.

3A21.0 Paroxysmal nocturnal haemoglobinuria

Paroxysmal nocturnal hemoglobinuria (PNH) is an acquired clonal hematopoietic stem cell disorder characterised by corpuscular haemolytic anaemia, bone marrow failure and frequent thrombotic events.

Exclusions: haemoglobinuria NOS (MF94)

Aplastic anaemia with paroxysmal nocturnal haemoglobulinuria (3A70.1)

3A21.1 Microangiopathic haemolytic anaemia

This is a microangiopathic subgroup of haemolytic anaemia (loss of red blood cells through destruction) caused by factors in the small blood vessels. It is identified by the finding of anaemia and schistocytes on microscopy of the blood film.

3A21.2 Haemolytic uraemic syndrome

A disease caused by acquired haematopoietic stem cell mutation defect of the PIGA gene in the X chromosome leading to the premature break down of red blood cells. This disease is characterised by haemolytic anaemia, thrombosis and. This disease may present with haemoglobinuria (blood in the urine will be most noticeable in the morning, and clear as the day progresses), pallor, fatigue, or shortness of breath. Confirmation is by identification of genetic mutation in the PIGA gene through genetic testing.

Exclusions: Hereditary haemolytic uraemic syndrome (3A10)

3A21.Y Other specified acquired haemolytic anaemia, non-immune

3A2Z Acquired haemolytic anaemia, unspecified

3A4Z Haemolytic anaemias, unspecified

3A50 Thalassaemias

A disease caused by genetically inherited autosomal recessive mutations leading to abnormal production of haemoglobin. This disease is characterised by destruction of red blood cells leading to anaemia and abnormal production of haemoglobin. This disease may present with pallor, jaundice, iron overload, fatigue, or shortness of breath. Confirmation is by identification of mutations through genetic testing.

3A50.0 Alpha thalassaemia

Alpha-thalassemia is an inherited haemoglobinopathy characterised by impaired synthesis of alpha-globin chains leading to a variable clinical picture depending on the number of affected alleles, and encompassing the alpha thalassaemia trait, haemoglobin H disease (HbH) and Bart's hydrops fetalis.

Exclusions: Hydrops fetalis due to haemolytic disease (KA85)

3A50.00 Mild alpha thalassaemia diseases

A disease caused by genetically inherited factors affecting the alpha chain of the haemoglobin molecule. This disease is characterised by structural abnormalities of the haemoglobin molecule. This disease may present with mild anaemia: pallor, fatigue, shortness of breath. Confirmation is by identification of changes to the alpha chain by genetic testing.

3A50.01 Thalassaemic alpha-chain variants

3A50.02 Haemoglobin H disease (– α/– – included)

Haemoglobin H (HbH) disease is a moderate to severe form of alpha-thalassemia characterised by pronounced microcytic hypochromic haemolytic anaemia.

3A50.03 Homozygous or compound heterozygous alpha0 thalassaemia

Hb Bart's hydrops fetalis is the most severe form of alpha-thalassemia and is almost always lethal. It is characterised by fetal onset of generalised oedema, pleural and pericardial effusions, and severe hypochromic anaemia.

3A50.0Y Other specified alpha thalassaemia

3A50.0Z Alpha thalassaemia, unspecified

3A50.1 Alpha thalassaemia related syndromes

Alpha-thalassemia-related diseases refers to a group of diseases characterised by alpha-thalassemia and an associated disorder. Three conditions are included in this group: alpha-thalassemia-intellectual deficit syndrome, X-linked (or ATR-X syndrome), alpha-thalassemia-intellectual deficit syndrome and alpha-thalassemia-myelodysplastic disease or ATMDS.

3A50.2 Beta thalassaemia

Beta-thalassemia (BT) is a haemoglobinopathy characterised by deficiency (Beta+) or absence (Beta0) of synthesis of the beta globin chains of haemoglobin (Hb). Three main types of BT have been described: minor, intermedia and major with clinical presentation ranging from asymptomatic forms to microcytic anaemia and splenomegaly due to defective erythropoiesis and haemolysis.

3A50.3 Delta, delta-beta or gamma-delta-beta thalassaemia

Delta-beta-thalassemia is a form of beta-thalassemia characterised by decreased or absent synthesis of the delta- and beta-globin chains with a compensatory increase in expression of fetal gamma-chain synthesis.

3A50.4 Hereditary persistence of fetal haemoglobin

Hereditary persistence of fetal haemoglobin (HPFH) associated with beta-thalassaemia is a haemoglobinopathy characterised by high haemoglobin (Hb)F levels and an increased number of fetal-Hb-containing-cells. The association of HPFH with beta-thalassaemia mitigates the clinical manifestations which vary from a normal state to beta-thalassaemia intermedia.

3A50.Y Other specified thalassaemias

3A50.Z Thalassaemias, unspecified

3A51 Sickle cell disorders or other haemoglobinopathies

Any disorder caused by a HbS mutation in the haemoglobin gene. This disorder is characterised by abnormal rigid sickle-shaped red blood cells decreasing its ability to carry oxygen. This disorder may present with fatigue, shortness of breath, dizziness, headaches, pallor of skin or mucous membranes, and jaundice. This disorder is confirmed by identification of HbS mutation by genetic testing.

Coded Elsewhere: Osteonecrosis due to haemoglobinopathy (FB81.4)

Other sickle-cell disorders with retinopathy (9B71.Y)

3A51.0 Sickle cell trait

A disease caused by genetic inheritance of one abnormal allele of the haemoglobin gene. This disease does not display the severe symptoms of sickle cell disease that occurs in homozygous individuals. Confirmation is by identification of mutation through genetic testing.

3A51.1 Sickle cell disease without crisis

A disorder caused by a HbS mutation in the haemoglobin gene. This disorder is characterised by abnormal rigid sickle-shaped red blood cells decreasing its ability to carry oxygen. This disorder may present with fatigue, shortness of breath, dizziness, headaches, pallor of skin or mucous membranes, and jaundice. This disorder is confirmed by identification of HbS mutation by genetic testing.

3A51.2 Sickle cell disease with crisis

Sickle cell crisis occurs when the sickle cells block blood flow, thus decreasing oxygen delivery to the tissues. This results in intense to severe pain in the extremities, lower back, abdomen, and chest. A crisis can be brought on by illness, stress, dehydration, exposure to temperature changes or high altitudes.

Inclusions: Hb-SS disease with crisis

3A51.3 Compound heterozygous sickling disorders without crisis

A disease caused by genetic inheritance of two heterozygous recessive alleles of the haemoglobin gene leading to abnormal formation of haemoglobin molecule. This disease is characterised by rigid, sickle shaped red blood cells. Confirmation is by identification of mutations through genetic testing.

3A51.4 Compound heterozygous sickling disorders with crisis

Compound heterozygous sickling disorders with crisis may present with acute chest syndrome, splenic sequestration, haemolytic crisis, and pain.

3A51.5 Haemoglobin C disease

A disease caused by the bi-parental gene that encodes for haemoglobin C. This disease is characterised by abnormal structure of one of the globin chains of the haemoglobin molecule. This disease may present with mild haemolytic anaemia, increased risk for gallstones, enlarged spleen, episodes of joint pain, and increased risk of infection. This disease is confirmed by identification of the haemoglobin C gene by genetic testing.

Exclusions: Hereditary persistence of fetal haemoglobin (3A50.4)

3A51.6 Haemoglobin D disease

Haemoglobin D (Hb D) disease is characterised by mild haemolytic anaemia and mild to moderate splenomegaly. Prevalence is unknown. Heterozygous forms of Hb D are clinically silent. Molecular testing can be useful to distinguish Hb D homozygosity from cases of heterozygous Hb D in association with beta-(0) thalassaemia.

3A51.7 High affinity haemoglobin

A disease caused by determinants arising after birth, in the antenatal period or by genetically inherited factors leading to high oxygen affinity haemoglobin. This disease is characterised by abnormalities the globin chains that alter the affinity of the haemoglobin molecule for oxygen, affecting the normal loading of oxygen in the lungs and delivery of oxygen to the tissues.

3A51.8 Low affinity haemoglobin

A disease caused by determinants arising after birth, in the antenatal period or by genetically inherited factors leading to low oxygen affinity haemoglobin. This disease is characterised by abnormalities the globin chains that alter the affinity of the haemoglobin molecule for oxygen, affecting the normal loading of oxygen in the lungs and delivery of oxygen to the tissues. This disease may present with fatigue, muscle weakness, loss of appetite, weight loss, diarrhoea, nausea, fast heartbeat or numbness in extremities.

3A51.9 Haemoglobin O disease

A disease caused by the bi-parental inheritance of the gene that encodes for haemoglobin O. This disease is characterised by abnormal structure of one of the globin chains of the haemoglobin molecule. This disease may present with mild haemolytic anaemia, increased risk for gallstones, enlarged spleen, episodes of joint pain, and increased risk of infection. This disease is confirmed by identification of the haemoglobin O gene by genetic testing.

3A51.A Haemoglobin E disease

Haemoglobin E disease is characterised by the synthesis of an abnormal haemoglobin called haemoglobin E (HbE), instead of the normal haemoglobin A (HbA). Subjects heterozygous for HbE (AE) have an asymptomatic condition with no clinical relevance, except for the risk of transmitting E/beta thalassemia if the other parent carries beta thalassemia. The severity of these E/beta thalassemia forms is very variable, the clinical picture ranging from that of beta thalassemia minor through to thalassemia intermedia to thalassemia major. Subjects homozygous for HbE (EE) are asymptomatic.

3A51.B Haemoglobin C/beta thalassaemia compound heterozygosity

Haemoglobin C/beta thalassaemia is a condition resulting from coinheritance of haemoglobin C and beta thalassaemia, both beta globin genes being mutated.

3A51.Y Other specified sickle cell disorders or other haemoglobinopathies

3A51.Z Sickle cell disorders or other haemoglobinopathies, unspecified

Pure red cell aplasia (BlockL2‑3A6)

A condition caused by determinates arising during the antenatal period, after birth or genetically inherited factors, leading to a change in the formation of erythrocytes. This condition is characterised by maturation arrest occurs in the formation of erythrocytes. This condition may present with severe anaemia. Confirmation is by identification of abnormally formed erythrocytes in a blood sample.

3A60 Congenital pure red cell aplasia

A condition caused by determinants arising during the antenatal period, leading to a change in the formation of erythrocytes. This condition is characterised by maturation arrest occurs in the formation of erythrocytes. This condition may present with severe anaemia. Confirmation is by identification of decreased red blood cell count in a blood sample.

3A60.0 Congenital non-inherited pure red cell aplasia

A condition caused by determinates arising during the antenatal period, leading to a change in the formation of erythrocytes. This condition is characterised by maturation arrest occurs in the formation of erythrocytes. This condition may present with severe anaemia. Confirmation is by identification of decreased levels of red blood cells in a blood sample.

3A60.1 Hereditary pure red cell aplasia

A condition caused by determinates arising during the antenatal period, leading to a change in the formation of erythrocytes. This condition is characterised by maturation arrest occurs in the formation of erythrocytes. This condition may present with severe anaemia. Confirmation is by identification of decreased red blood cell count in a blood sample.

3A60.Z Congenital pure red cell aplasia, unspecified

3A61 Acquired pure red cell aplasia

A condition characterised by the near absence of red blood cell precursors in bone marrow, often associated with thymomas and autoimmune disorders

Exclusions: Aplastic anaemia with paroxysmal nocturnal haemoglobulinuria (3A70.1)

3A61.0 Acute acquired pure red cell aplasia

This refers to transient (acute) and acquired type of anaemia affecting the precursors to red blood cells but not to white blood cells. In PRCA, the bone marrow ceases to produce red blood cells.

3A61.1 Chronic acquired pure red cell aplasia

This refers to a chronic and acquired type of anaemia affecting the precursors to red blood cells but not to white blood cells. In PRCA, the bone marrow ceases to produce red blood cells.

3A61.Y Other specified acquired pure red cell aplasia

3A61.Z Acquired pure red cell aplasia, unspecified

3A6Z Pure red cell aplasia, unspecified

3A70 Aplastic anaemia

A disease caused by determinants arising after birth, in the antenatal period or genetically inherited factors leading to the inability of stem cells to generate new mature cells. This disease is characterised by low levels of red blood cells, white blood cells, and platelets. This disease may present with pallor, fatigue, dizziness, increased risk of infection or increased bruising or bleeding.

Inclusions: Medullary hypoplasia

Panmyelophthisis

3A70.0 Congenital aplastic anaemia

A disease caused by determinants in the antenatal period leading to the inability of stem cells to generate new mature cells. This disease is characterised by low levels of red blood cells, white blood cells, platelets. This disease may present with pallor, fatigue, dizziness, increased risk of infection or increased bruising or bleeding.

Inclusions: familial hypoplastic anaemia

Constitutional medullar aplasia

Exclusions: Congenital amegakaryocytic thrombocytopenia (3B64.01)

Coded Elsewhere: Congenital hypoplastic anaemia (KA8C)

Noonan syndrome (LD2F.15)

3A70.1 Acquired aplastic anaemias

A condition occurring secondary to other disorders or via an auto-immune response directed to the bone marrow arising after birth. This disease is characterised by an almost complete absence of hematopoietic stem cells resulting in low levels of red and white blood cells and platelets. This condition may present with fatigue, chronic infections, dizziness, weakness, headaches, and episodes of bleeding, usually in the skin and mucous membranes.

Inclusions: Acquired medullar aplasia

Coded Elsewhere: Paroxysmal nocturnal haemoglobinuria (3A21.0)

Myelofibrosis with myeloid metaplasia (2A20.2)

3A70.10 Drug-induced aplastic anaemia

A disease caused by drug intake. This disease is characterised by inability of stem cells to generate new mature cells leading to low levels of red blood cells, white blood cells, platelets. This disease may present with pallor, fatigue, dizziness, increased risk of infection or increased bruising/bleeding.

3A70.11 Aplastic anaemia due to other external agents

3A70.12 Idiopathic aplastic anaemia

3A70.1Y Other specified acquired aplastic anaemias

3A70.1Z Acquired aplastic anaemias, unspecified

3A70.Z Aplastic anaemia, unspecified

3A71 Anaemia due to chronic disease

A disease caused by chronic diseases such as chronic infection. This disease is characterised by inflammatory responses targeted at red blood cells leading to low levels of red blood cells in the body. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low levels of red blood cells in a blood sample.

Coding Note: Code aslo the casusing condition

3A71.0 Anaemia in neoplastic disease

A disease caused by chronic neoplastic diseases. This disease is characterised by inflammatory responses targeted at red blood cell leading to low levels of red blood cells in the body. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low levels of red blood cells in a blood sample

Coding Note: Code aslo the casusing condition

3A71.1 Anaemia in chronic infectious diseases

A disease caused by chronic infectious diseases leading to decreased levels of red blood cells in the blood. This disease is characterised by a low red blood cell count in the body. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of low red blood cell count in a blood sample.

Coding Note: Code aslo the casusing condition

3A71.2 Anaemia in chronic kidney disease

A disease caused by chronic kidney disease. This disease is characterised by a low red blood cell count in the blood. This disease may present with pallor, fatigue, or shortness of breath. Confirmation is by identification of a low red blood cell count in a blood sample.

Coding Note: Code aslo the casusing condition

3A71.Y Anaemia due to other specified chronic disease

Coding Note: Code aslo the casusing condition

3A71.Z Anaemia due to chronic disease, unspecified

Coding Note: Code aslo the casusing condition

3A72 Sideroblastic anaemia

Sideroblastic anaemias are a group of disorders in which haemoglobin is insufficiently synthesized, because of defective use of iron (although plasmatic iron levels may be normal or elevated). They are said to be sideroblastic because of the presence of ringed sideroblasts in the blood due to accumulated ferritin in mitochondria. Anaemias may be microcytic hypochrome (in thalassemia and hereditary sideroblastic anaemias), or macrocytic (in idiopathic acquired sideroblastic anaemias).

3A72.0 Congenital sideroblastic anaemias

A disease caused by determinants arising in the antenatal period leading to the production of ringed sideroblasts; abnormal nucleated erythroblasts. This disease is characterised by the inability of to incorporate haemoglobin, which red blood cells need to transport oxygen efficiently. This disease may present with pallor, fatigue, dizziness, and enlarged spleen and liver, heart disease, liver damage, or kidney failure.

3A72.00 Hereditary sideroblastic anaemias

Inclusions: Sex-linked hypochromic sideroblastic anaemia

3A72.01 Hereditary syndromic sideroblastic anaemia

Coded Elsewhere: Thiamine-responsive megaloblastic anaemia syndrome (5C63.Y)

3A72.0Y Other specified congenital sideroblastic anaemias

3A72.0Z Congenital sideroblastic anaemias, unspecified

3A72.1 Acquired sideroblastic anaemias

A disease caused by determinants arising after birth such as myelodysplastic syndromes, antimicrobials, pyridoxine deficiency, lead poisoning, or copper deficiency. Zinc can indirectly cause sideroblastic anaemia by decreasing absorption and increasing excretion of copper. This disease is characterised by the inability of to incorporate haemoglobin, which red blood cells need to transport oxygen efficiently. This disease may present with pallor, fatigue, dizziness, and enlarged spleen and liver, heart disease, liver damage, or kidney failure.

Coded Elsewhere: Refractory anaemia with ring sideroblasts (2A33)

3A72.Z Sideroblastic anaemia, unspecified

3A73 Congenital dyserythropoietic anaemia

Congenital dyserythropoietic anaemias (CDA) result from diverse erythropoietic disorders; they lead to the defective production of red blood cells (RBC) and often mild haemolysis that attests to a qualitative defect of these RBC released into the circulation. Three forms of CDA have been characterised: types I, II and III. The shared symptoms include anaemia of variable severity, intermittent jaundice, splenomegaly and hepatomegaly.

Exclusions: Blackfan-Diamond syndrome (3A60.1)

Di Guglielmo disease (2A60.35)

Polycythaemia (BlockL2‑3A8)

Coded Elsewhere: Polycythaemia vera (2A20.4)

3A80 Congenital polycythaemia

A disease caused by determinants occurring in the antenatal period leading to changes in the concentration of red blood cells. This disease is characterised by having a high concentration of red blood cells in the body leading to slow flow of blood. This disease may present with headaches, blurred vision, red skin, tiredness, high blood pressure, dizziness, periods of confusion, bleeding problems gout or itchy skin. Confirmation is by identification of increased levels of red blood cells in a blood sample.

Coded Elsewhere: Polycythaemia neonatorum (KA8A)

3A80.0 Primary inherited erythrocytosis

A disease caused by genetically inherited factors leading to changes in the concentration of red blood cells. This disease is characterised by having a high concentration of red blood cells in the body leading to slow flow of blood. Confirmation is by identification of mutations by genetic testing.

3A80.Y Other specified congenital polycythaemia

3A80.Z Congenital polycythaemia, unspecified

3A81 Acquired polycythaemia

Secondary polycythaemia is acquired and caused by either natural or artificial increases in the production of erythropoietin, hence an increased production of erythrocytes.

3A81.0 Polycythaemia due to hypoxia, including high altitude

3A81.1 Polycythaemia due to over-transfusion or blood doping

3A81.2 Relative polycythaemia

A disease caused by loss of body fluids leading to apparent increased levels of red blood cells in the blood. This disease may present with headache, vertigo, abnormally enlarged spleen or liver, high blood pressure, or formation of blood clots. Confirmation is by identification of relative blood cell counts in a blood sample.

3A81.Y Other specified acquired polycythaemia

3A81.Z Acquired polycythaemia, unspecified

3A8Z Polycythaemia, unspecified

3A90 Anaemia due to acute disease

Exclusions: Acute posthaemorrhagic anaemia (3A00.00)

3A91 Congenital methaemoglobinaemia

A disease caused by determinants in the antenatal period leading to lack of the enzyme cytochrome b5 reductase. This disease is characterised by elevated levels of methemoglobin within the blood leading to haemoglobin ineffectively releasing oxygen to body tissues. This disease may present with shortness of breath, cyanosis, headache, fatigue, exercise intolerance, dizziness and loss of consciousness. Confirmation is by identification of mutation by genetic testing.

3A92 Hereditary methaemoglobinaemia

Hereditary methemoglobinemia (HM) is a rare red cell disorder classified principally into two clinical phenotypes: autosomal recessive congenital (or hereditary) methemoglobinemia types I and II (RCM/RHM type 1; RCM/RHM type 2). In RCM type 1, well-tolerated cyanosis from birth is the only symptom. RCM type 2, with global loss of Cb5R function, is much more severe; the cyanosis is accompanied by neurological dysfunction (with intellectual deficit, microcephaly, growth retardation, opisthotonus, strabismus and hypertonia), which usually becomes evident during the first four months of life.

3A93 Acquired methaemoglobinaemia

3A9Y Other specified anaemias and erythrocyte disorders

3A9Z Anaemias or other erythrocyte disorders, unspecified

Coagulation defects, purpura or other haemorrhagic or related conditions (BlockL1‑3B1)

A condition caused by determinants arising during the antenatal period, after birth or by genetically inherited factors, leading to coagulation defects. This condition is characterised by increased bruising and bleeding.

Exclusions: Postpartum coagulation defects (JA43.3)

Coagulation defects (BlockL2‑3B1)

Congenital or constitutional haemorrhagic condition (BlockL3‑3B1)

A condition caused by determinants arising during the antenatal period or genetically inherited factors, leading to defects in clotting mechanisms or abnormalities causing structural flaws in the blood vessels. This disease is characterised by spontaneous bleeding or bruising.

Coded Elsewhere: Congenital non-inherited haemorrhagic condition

3B10 Hereditary factor VIII deficiency

A disease caused by a genetically inherited mutation leading to a deficiency in clotting due to lack of factor VIII. This disease is characterised by increasing haemorrhaging and bruising. Confirmation is by identification of mutations by genetic testing.

3B10.0 Haemophilia A

Haemophilia A is the most common form of haemophilia characterised by spontaneous or prolonged haemorrhages due to factor VIII deficiency. Depending on the extent of the factor VIII deficiency, it can be severe (biological activity of factor VIII below 1%), moderately severe (activity of factor VIII between 1% and 5%), or mild (activity of factor VIII between 5 and 40%).

Exclusions: factor VIII deficiency with vascular defect (3B12)

3B10.1 Hereditary factor VIII deficiency with anti-factor VIII inhibitor

A disease caused by a genetically inherited mutation leading to a deficiency in clotting due to lack of factor VIII. This disease also causes anti-factor VIII inhibitor antibodies to be produced when receiving transfusions. Anti-factor VIII inhibitor antibodies develop as the body recognises the factor VIII as foreign, therefore deeming factor VIII infusions ineffective. This disease is characterised by increasing haemorrhaging and bruising. Confirmation is by identification of mutations by genetic testing.

3B10.Y Other specified hereditary factor VIII deficiency

3B10.Z Hereditary factor VIII deficiency, unspecified

3B11 Hereditary factor IX deficiency

A disease caused by a genetically inherited X-linked recessive trait leading to a defective gene located on the X chromosome. This disease is characterised by low levels of the protein factor IX in the body leading to increased haemorrhaging and bruising due to clotting abnormalities. Confirmation is by identification of recessive trait by genetic testing.

3B11.0 Haemophilia B

Haemophilia B is a form of haemophilia characterised by spontaneous or prolonged haemorrhages due to factor IX deficiency. Depending on the extent of the factor IX deficiency, it can be severe (biological activity of factor IX below 1%), moderately severe (activity of factor IX between 1% and 5%), or mild (activity of factor IX between 5 and 40%).

Inclusions: PTC - [plasma thromboplastin component] deficiency

3B11.Y Other specified hereditary factor IX deficiency

3B11.Z Hereditary factor IX deficiency, unspecified

3B12 Von Willebrand disease

A disease caused by inherited genetic mutations. This disease is characterised by quantitative, structural or function abnormalities of Von Willebrand factor leading to abnormalities in coagulation of the blood. This disease may present with prolonged bleeding, easy bruising or, bleeding gums. Confirmation is by identification of mutation through genetic testing.

Inclusions: Factor VIII deficiency with vascular defect

Vascular haemophilia

Angiohaemophilia

Exclusions: factor VIII deficiency with functional defect (3B10)

factor VIII deficiency NOS (3B10)

Acquired von Willebrand disease or syndrome (BlockL3‑3B2)

3B13 Haemophilia C

A disease caused by genetically inherited mutations. This disease is characterised by decreased levels of factor XI leading to abnormalities in coagulation of the blood. This disease may present with prolonged bleeding, easy bruising or, bleeding gums. Confirmation is by identification of mutation through genetic testing.

3B14 Other inherited coagulation factor deficiency with bleeding tendency

Any disease caused by genetically inherited mutations leading to lack of coagulation factors in the blood not elsewhere classified. These diseases are characterised by increased haemorrhaging and bruising as the blood cannot clot properly to control bleeding. Confirmation is identification of mutations by genetic testing.

3B14.0 Hereditary deficiency of factor I

Congenital deficiencies of fibrinogen are coagulation disorders characterised by bleeding symptoms ranging from mild to severe resulting from reduced quantity and/or quality of circulating fibrinogen. Afibrinogenaemia (complete absence of fibrinogen) and hypofibrinogenaemia (reduced plasma fibrinogen concentration) correspond to quantitative anomalies of fibrinogen while dysfibrinogenaemia corresponds to a functional anomaly of fibrinogen. Hypo- and dysfibrinogenaemia may be frequently combined (hypodysfibrinogenaemia).

3B14.1 Hereditary factor X deficiency

Congenital factor X deficiency is an inherited bleeding disorder with a decreased antigen and/or activity of factor X (FX) and characterised by mild to severe bleeding symptoms.

3B14.2 Combined deficiency of vitamin K-dependent clotting factors

Hereditary combined vitamin K-dependent clotting factors deficiency (VKCFD) is a congenital bleeding disorder resulting from variably decreased levels of coagulation factors II, VII, IX and X, as well as natural anticoagulants protein C, protein S and protein Z.

3B14.Z Other inherited coagulation factor deficiency with bleeding tendency, unspecified

3B15 Inherited coagulation factor deficiency without bleeding tendency

A disease caused by a genetically inherited mutation leading to decreased levels of coagulation factor. This disease is characterised by decreased levels of coagulation factor without leading to increased haemorrhaging. Confirmation is by identification of decreased levels of coagulation factor in a blood sample.

3B1Z Congenital or constitutional haemorrhagic condition, unspecified

Haemorrhagic diseases due to acquired coagulation factor defects (BlockL3‑3B2)

Any disease caused by determinants arising after birth. These diseases are characterised by abnormal coagulation of the blood.

Exclusions: vitamin K deficiency of newborn (KA8F.0)

3B20 Disseminated intravascular coagulation

A disorder that is characterised by the systemic intravascular activation of the coagulation system, simultaneously leading to intravascular thrombi, compromising an adequate blood supply to the organs, and to bleeding as the consequence of consumption of platelets and coagulation factors. It may be provoked by a wide range of disorders including infections, inflammatory disorders and malignancy.

Coded Elsewhere: Disseminated intravascular coagulation of fetus or newborn (KA88)

3B21 Haemorrhagic disorder due to circulating anticoagulants and coagulation factors

A disease caused by anticoagulants present in the body that prevent the blood from clotting normally. This disease is characterised by abnormalities in blood clotting. This disease may present with prolonged bleeding, easy bruising or, bleeding gums. Confirmation is by identification of anticoagulants present in a blood sample.

Exclusions: long-term use of anticoagulants without haemorrhage (QC48.0)

3B21.0 Haemorrhage due to thrombin inhibitor other than heparin

A disease caused by any thrombin inhibitors other than heparin that affects normal coagulation of the blood. This disease is characterised by inability of the blood to coagulate leading to bleeding. Confirmation is by identification of thrombin inhibitors in a blood sample.

3B21.1 Haemorrhage due to factor Xa inhibitor

A disease caused by factor Xa inhibitor that affects normal coagulation of the blood. This disease is characterised by inability of the blood to coagulate leading to bleeding. Confirmation is by identification of factor Xa inhibitor in a blood sample.

3B21.Y Haemorrhagic disorder due to other specified circulating anticoagulants

3B21.Z Haemorrhagic disorder due to unspecified circulating anticoagulants

3B22 Acquired haemophilia

Acquired haemophilia is a rare haemorrhagic disease caused by production of anti-factor VIII antibodies and is sometimes associated with other autoimmune disorders, cancers lymphoproliferative syndromes and multiple transfusions during the postpartum period.

3B2Y Other specified haemorrhagic diseases due to acquired coagulation factor defects

3B4Z Coagulation defects, unspecified

Fibrinolytic defects (BlockL2‑3B5)

A disease caused by determinants arising during the antenatal period, after birth or genetically inherited factors, affecting the fibrinolysis system which prevents blood clots from growing and becoming problematic. This disease is characterised by defects in the fibrinolysis system leading to coagulation of the blood. This disease may present with thrombosis.

3B50 Inherited fibrinolytic defects

A disease caused by genetically inherited mutations affecting the fibrinolysis system which prevents blood clots from growing and becoming problematic. This disease is characterised by defects in the fibrinolysis system leading to coagulation of the blood. This disease may present with thrombosis.

Coded Elsewhere: Hypoplasminogenaemia (DA0D.3)

3B50.0 Congenital alpha-2 antiplasmin deficiency

3B50.1 Congenital plasminogen activator inhibitor type 1 deficiency

Congenital plasminogen activator inhibitor type 1(PAI-1) deficiency is a disorder that causes premature lysis of haemostatic clots and a moderate bleeding syndrome. Spontaneous bleeding is rarely observed, whereas moderate haemorrhages of the knees, elbows, nose and gingiva are usually triggered by mild trauma. However, menstrual bleeding may be severe and a prolonged bleeding after surgery is common. The PAI-1 deficiency may be qualitative or quantitative, total or partial.

3B50.Y Other specified inherited fibrinolytic defects

3B50.Z Inherited fibrinolytic defects, unspecified

3B51 Acquired fibrinolytic defects

A disease caused by determinates arising after birth, affecting the fibrinolysis system which prevents blood clots from growing and becoming problematic. This disease is characterised by defects in the Fibrinolysis system leading to coagulation of the blood. This disease may present with thrombosis.

3B60 Non-thrombocytopenic purpura

A descriptive term for purpura caused by determinants other than low platelet count. This should be used for coding only when a more precise diagnosis is not available.

Exclusions: Antineutrophil cytoplasmic antibody-associated vasculitis (4A44.A)

Antiphospholipid syndrome (4A45)

Drug-associated immune complex vasculitis (4A85.03)

Immune complex small vessel vasculitis (4A44.9)

Leukocytoclastic vasculitis (4A44.B)

Purpura or bruising due to vascular fragility (EE40.32)

Thrombotic thrombocytopenic purpura (3B64.14)

Traumatic purpura (EF31)

3B60.0 Hereditary vascular purpura

3B60.1 Acquired vascular purpura

Purpura resulting from vascular factors rather than from abnormalities in the blood such as dysproteinaemias and disorders of platelets and coagulation.

Exclusions: Antineutrophil cytoplasmic antibody-associated vasculitis (4A44.A)

Antiphospholipid syndrome (4A45)

Capillaritis (EF40.0)

Drug-associated immune complex vasculitis (4A85.03)

IgA vasculitis (4A44.92)

Leukocytoclastic vasculitis (4A44.B)

Purpura or bruising due to vascular fragility (EE40.32)

Thrombotic thrombocytopenic purpura (3B64.14)

Traumatic purpura (EF31)

3B61 Thrombophilia

A disease caused by determinants arising after birth or genetically inherited factors leading to abnormalities in blood. This disease is characterised by abnormality of blood coagulation that increases the risk of thrombosis, clots in blood vessels. This disease may present with deep vein thrombosis or pulmonary embolism. Confirmation is identification of abnormal blood coagulation in a blood sample.

3B61.0 Hereditary thrombophilia

A disease caused by determinants arising after birth or genetically inherited factors leading to abnormalities in blood. This disease is characterised by abnormality of blood coagulation that increases the risk of thrombosis, clots in blood vessels. This disease may present with deep vein thrombosis or pulmonary embolism. Confirmation is identification of abnormal blood coagulation in a blood sample.

Inclusions: antithrombin deficiency

3B61.00 Hyperhomocysteinaemia

A disease caused by deficiencies of vitamin B6, folic acid, or vitamin B12. Genetic defects in 5-MTHF reductase can consequently lead to hyperhomocysteinaemia. This disease is characterised by abnormally high level of homocysteine in the blood. This disease may present with cardiovascular disease, thrombosis, schizophrenia and osteoporosis. Confirmation is by identification of deficiency in a blood sample.

3B61.0Y Other specified hereditary thrombophilia

3B61.1 Acquired thrombophilia

A disease caused by determinants arising after birth. This disease is characterised by abnormality of blood coagulation that increases the risk of thrombosis, clots in blood vessels. This disease may present with deep vein thrombosis or pulmonary embolism. Confirmation is identification of abnormal blood coagulation in a blood sample.

Coded Elsewhere: Antiphospholipid syndrome (4A45)

3B61.Y Other specified thrombophilia

3B61.Z Thrombophilia, unspecified

3B62 Qualitative platelet defects

A disease caused by determinants arising after birth, during the antenatal period or genetically inherited factors. This disease is characterised by abnormalities in coagulation of the blood due to defective platelets. This condition may present with easy bruising, prolonged bleeding or, bleeding gums. Confirmation is by identification of decreased platelets in a blood sample.

Inclusions: Thrombocytopathy

Exclusions: Von Willebrand disease (3B12)

3B62.0 Inherited qualitative platelet defects

A disease caused by genetically inherited mutations leading to abnormalities in platelets. This disease is characterised by abnormal platelet formation or function. Confirmation is by identification of mutations by genetic testing.

Coded Elsewhere: Dense granule disease (3B62.3)

Alpha-delta dense granule deficiency (3B62.4)

3B62.00 Alpha-granule diseases

A condition caused by determinants arising after birth, in the antenatal period. This condition is characterised by defects in the alpha granules in platelets leading to abnormalities in coagulation mechanisms. This condition may present with prolonged bleeding, epistaxis, menorrhagia, easy bruising, anaemia, fatigue or shortness of breath. Confirmation is by identification of platelet defects in a blood sample.

3B62.01 Inherited giant platelet disorder

A disease caused by genetically inherited mutations. This disease is characterised by abnormally large platelets, low platelet count and a bleeding tendency. Confirmation is by identification of mutations through genetic testing.

Coded Elsewhere: MYH9 macrothrombocytopenia syndromes (3B64.01)

3B62.0Y Other specified inherited qualitative platelet defects

3B62.0Z Inherited qualitative platelet defects, unspecified

3B62.1 Bleeding diathesis due to thromboxane synthesis deficiency

A disease caused by thromboxane synthesis deficiency. This disease is characterised by low levels of eicosanoids (lipids), abnormalities in coagulation leading to haemorrhaging. Confirmation is by identification of low levels of eicosanoids in a blood sample.

3B62.2 Isolated thrombocytopenia

3B62.3 Dense granule disease

A condition caused by determinants arising after birth, in the antenatal period. This condition is characterised by defects in the dense granules in platelets leading to abnormalities in coagulation mechanisms. This condition may present with prolonged bleeding, epistaxis, menorrhagia, easy bruising, anaemia, fatigue or shortness of breath. Confirmation is by identification of platelet defects in a blood sample.

Coded Elsewhere: Hermansky-Pudlak syndrome (EC23.20)

Chédiak-Higashi syndrome (EC23.20)

3B62.4 Alpha-delta dense granule deficiency

A condition caused by determinants arising after birth, in the antenatal period. This condition is characterised by defects in the alpha delta dense granules in platelets, leading to abnormalities in coagulation mechanisms. This condition may present with prolonged bleeding, epistaxis, menorrhagia, easy bruising, anaemia, fatigue or shortness of breath. Confirmation is by identification of low levels of alpha delta dense granules in a blood sample.

3B62.5 Haemophagocytic syndrome associated with infection

This is an uncommon hematologic disorder that, typically, clinically manifests as fever, hepatosplenomegaly, lymphadenopathy, jaundice and rash, with laboratory findings of histiocytosis, and the pathologic finding of haemophagocytosis, infection-associated.

3B62.Y Other specified qualitative platelet defects

3B62.Z Qualitative platelet defects, unspecified

3B63 Thrombocytosis

A disease caused by essential thrombocytosis or other myelo-proliferative disorders such as chronic myelogenous leukaemia, polycythaemia, myelofibrosis. This disease can also have secondary causes such as inflammation, surgery, hyposplenism, Splenectomy, asplenia, iron deficiency anaemia or haemorrhage. This disease is characterised by elevated platelet count in the blood. Confirmation is by identification of increased platelet count in a blood sample.

3B63.0 Congenital thrombocytosis

Familial thrombocytosis is a type of thrombocytosis, a sustained elevation of platelet numbers, which affects the platelet/megakaryocyte lineage and may create a tendency for thrombosis and haemorrhage but does not cause myeloproliferation.

Inclusions: Hereditary thrombocytosis

Exclusions: Essential thrombocythemia (3B63.1)

3B63.1 Acquired thrombocytosis

A chronic myeloproliferative neoplasm that involves primarily the megakaryocytic lineage. It is characterised by sustained thrombocytosis in the blood, increased numbers of large, mature megakaryocytes in the bone marrow, and episodes of thrombosis and/or haemorrhage. Progression to a post -essential thrombocythaemia myelofibrosis stage or transformation to acute myeloid leukaemia is rarely observed.

Inclusions: Idiopathic haemorrhagic thrombocythaemia

3B63.10 Secondary thrombocytosis

Coding Note: Code aslo the casusing condition

3B63.1Y Other specified acquired thrombocytosis

3B63.1Z Acquired thrombocytosis, unspecified

3B63.Y Other specified thrombocytosis

3B63.Z Thrombocytosis, unspecified

3B64 Thrombocytopenia

This disease is characterised by decreased levels of platelets within the blood. This disease may present with increased bruising or haemorrhaging. Confirmation is by identification of decreased platelet count in a blood sample.

Coded Elsewhere: Isolated thrombocytopenia (3B62.2)

3B64.0 Congenital thrombocytopenia

A disease caused by determinants arising during the antenatal period leading to low platelet count. This disease is characterised by decreased levels of platelets within the blood. This disease may present with increased bruising or haemorrhaging. Confirmation is by identification of a decreased platelet count in a blood sample.

3B64.00 Congenital non-inherited thrombocytopenia

Coded Elsewhere: Transient neonatal thrombocytopaenia (KA89)

3B64.01 Hereditary thrombocytopenia

A disease caused by a genetically inherited mutation leading to decreased platelet count. This disease is characterised by decreased levels of platelets within the blood. This disease may present with increased bruising or haemorrhaging. Confirmation is by identification of decreased platelet count in a blood sample.

Coded Elsewhere: Thrombocytopaenia - absent radius (LD2F.1Y)

Familial platelet syndrome with predisposition to acute myelogenous leukaemia (3B62.3)

Congenital thrombotic thrombocytopenic purpura due to ADAMTS-13 deficiency (3B64.14)

Macrothrombocytopenia with mitral valve insufficiency (3B62.01)

3B64.0Z Congenital thrombocytopenia, unspecified

3B64.1 Acquired thrombocytopenia

A disease caused by determinants arising after birth, leading to low platelet count. This disease is characterised by low levels of platelets within the blood. This disease may present with increased bruising or haemorrhaging. Confirmation is by identification of decreased platelet count in a blood sample.

3B64.10 Immune thrombocytopenic purpura

Immune thrombocytopenic purpura (or immune thrombocytopenia; ITP) is an autoimmune coagulation disorder characterised by isolated thrombocytopenia (a platelet count <100,000/microL), in the absence of any underlying disorder that may be associated with thrombocytopenia.

Coded Elsewhere: Evans syndrome (3A20.5)

3B64.11 Secondary thrombocytopenic purpura

This disease is characterised by a relative decrease in levels of platelets within the blood. This disease may present with increased bruising or haemorrhaging. Confirmation is by identification of decreased platelets in a blood sample.

Coding Note: Code aslo the casusing condition

3B64.12 Drug-induced thrombocytopenic purpura

Thrombocytopenic purpura attributable to drug toxicity (e.g. cytotoxic chemotherapeutic or immunosuppressive agents) or to an idiosyncratic drug-associated allergic thrombocytopenia (e.g. quinine, thiazides).

3B64.13 Alloimmune thrombocytopenia

A disease caused by determinants such as a blood transfusion that lead to an immune response to the foreign antigens. This disease is characterised by low levels of platelets in the body due to an immune reactive response to the foreign platelet antigens. This disease may present with increased bruising or haemorrhaging. Confirmation is by identification of decreased platelet count and presence of autoantibodies in a blood sample.

3B64.14 Thrombotic thrombocytopenic purpura

This condition is idiopathic. This condition is characterised by abnormality of blood coagulation causing extensive microscopic clots to form in the small blood vessels throughout the body resulting in low platelet count. This condition may present with seizures, hemiplegia, paresthesias, visual disturbance, and aphasia or anaemia. Confirmation is by identification of thromboses in a blood sample.

3B64.1Y Other specified acquired thrombocytopenia

3B64.Z Thrombocytopenia, unspecified

3B65 Thrombotic microangiopathy, not elsewhere classified

Thrombotic microangiopathies are microvascular occlusive disorders characterised by systemic or intrarenal aggregation of platelets, thrombocytopenia, and mechanical injury to erythrocytes. Thrombotic thrombocytopenic purpura (TTP) and haemolytic–uremic syndrome (HUS) represent a spectrum of thrombotic microangiopathies. In TTP, systemic microvascular aggregation of platelets causes ischemia in the brain and other organs. In HUS, platelet–fibrin thrombi predominantly occlude the renal circulation.

Coded Elsewhere: Thrombotic thrombocytopenic purpura (3B64.14)

Haemolytic uraemic syndrome (3A21.2)

Methylcobalamin deficiency type cbl G (5C50.B)

Hereditary haemolytic uraemic syndrome (3A10.Y)

3B6Y Other specified coagulation defects, purpura or other haemorrhagic or related conditions

3B6Z Coagulation defects, purpura or other haemorrhagic or related conditions, unspecified

Diseases of spleen (BlockL1‑3B8)

3B80 Congenital disorders of spleen

Any condition caused by a failure of the spleen to correctly develop in the antenatal period.

Coded Elsewhere: Structural developmental anomalies of spleen (LB22)

3B80.0 Splenomegaly in storage diseases

A disease caused by storage diseases; genetically inherited metabolic disorders that result from defects in lysosomal, lipid or glycogen function, of the spleen. This disease is characterised by enlargement of the spleen. This disease may present with abdominal pain, chest pain, pallor, shortness of breath fatigue. Confirmation is through medical imaging.

3B81 Acquired disorders of spleen

Any condition caused by determinants acquired after birth, leading to dysfunction of the spleen.

Coded Elsewhere: Injury of spleen (NB91.0)

Primary neoplasms of the spleen (2C11.Z)

3B81.0 Tumour-like conditions of spleen

Any condition caused by determinants acquired after birth, in the antenatal period or genetically inherited factors, leading to tumour-like conditions of the spleen. Confirmation is through medical imaging.

3B81.1 Postsurgical asplenia

A disease caused by underlying diseases, splenectomy or splenic rupture from trauma. This disease is characterised by absence of normal spleen function. This disease may present with increased susceptibility to infection. Confirmation is through medical imaging.

3B81.2 Atrophy of spleen

A disease caused by determinants arising after birth, during the antenatal period or by genetically inherited factors. This disease is characterised by partial or complete degradation of the spleen. This disease may present with increased susceptibility to infection. Confirmation is through medical imaging.

3B81.3 Nontraumatic laceration or rupture of spleen

A disease caused by non-traumatically determinants such as infectious diseases, medical procedures such as colonoscopy, haematological diseases, medications, or pregnancy. This disease is characterised by laceration or rupturing of the spleen leading to lack of function. This disease may present with bleeding and increased susceptibility of infection. Confirmation is through medical imaging.

3B81.4 Splenosis

A diseased caused by determinants arising after birth such as physical trauma or splenectomy . This disease is characterised by autoimplantation one or more focal deposits of splenic tissue in various compartments of the body. Confirmation is through medical imaging.

3B81.5 Splenic cyst or pseudocyst

3B81.50 Pseudocyst of spleen

A disease caused by determinants arising after birth, during the antenatal period or by genetically inherited factors. This disease is characterised by a noncancerous fluid-filled sac, pseudocysts are like cysts, but lack epithelial or endothelial cells. This disease is often asymptomatic but may present with abdominal pain, nausea and vomiting. Confirmation is through medical imaging.

3B81.51 Epithelial cyst of spleen

3B81.5Y Other specified splenic cyst

3B81.5Z Splenic cyst, unspecified

3B81.6 Infarction of spleen

A disease caused by determinants such as trauma, infection, or inherited factors leading to a shortage of oxygen in the spleen. This disease is characterised by death of spleen tissue and loss of function. Confirmation is by medical imaging.

Exclusions: traumatic rupture of spleen (NB91.0)

3B81.7 Infection of spleen

Any condition of the spleen, caused by an infection with a bacterial, viral, fungal, or parasitic source.

3B81.70 Acute septic splenitis

Inclusions: septic spleen

3B81.71 Abscess of spleen

This is a collection of pus (neutrophils) that has accumulated within a tissue because of an inflammatory process in response to either an infectious process (usually caused by bacteria or parasites) or other foreign materials (e.g., splinters, bullet wounds, or injecting needles), in the spleen.

3B81.7Y Other specified infection of spleen

3B81.7Z Infection of spleen, unspecified

3B81.8 Torsion of spleen

A disease caused by abnormal development of splenic suspensory ligaments. This disease is characterised by twisting of the spleen leading to splenic infarction. This disease may present with abdominal pain. Confirmation is through medical imaging.

3B81.9 Fibrosis of spleen

A disease caused by determinants arising after birth, during the antenatal period or by genetically inherited factors. This disease is characterised by formation of excess fibrous connective tissue leading to partial or complete degradation of the spleen. This disease may present with increased susceptibility to infection. Confirmation is through medical imaging.

Coding Note: Code aslo the casusing condition

3B81.A Perisplenitis

A disease caused by bacterial or viral infection, parasite infestation, or cysts. This disease is characterised by inflammation of the peritoneal surface of the spleen. This disease may present with abdominal pain, susceptibility to infection and enlargement of the spleen. Confirmation is by identification of infection in a blood sample.

3B81.B Hypersplenism

A disease caused by determinants such as cirrhosis, malaria, tuberculosis or inflammatory disorders leading overactive spleen function. This disease is characterised by the presence of an enlarged spleen. Confirmation is by identification through medical imaging.

Exclusions: Splenomegaly, not elsewhere classified (ME10.01)

congenital splenomegaly (LB22)

3B81.C Chronic congestive splenomegaly

A form of exaggerated spleen function characterised by splenic enlargement secondary to splenic vein thrombosis and/or portal hypertension

3B81.Y Other specified acquired disorders of spleen

3B8Z Diseases of spleen, unspecified

3C0Y Other specified diseases of the blood or blood-forming organs

3C0Z Diseases of the blood or blood-forming organs, unspecified