CHAPTER 02

Neoplasms

This chapter has 326 four-character categories.

Code range starts with 2A00

An abnormal or uncontrolled cellular proliferation which is not coordinated with an organism's requirements for normal tissue growth, replacement or repair.

Coded Elsewhere: Inherited cancer-predisposing syndromes

This chapter contains the following top level blocks:

* Neoplasms of brain or central nervous system
* Neoplasms of haematopoietic or lymphoid tissues
* Malignant neoplasms, except primary neoplasms of lymphoid, haematopoietic, central nervous system or related tissues
* In situ neoplasms, except of lymphoid, haematopoietic, central nervous system or related tissues
* Benign neoplasms, except of lymphoid, haematopoietic, central nervous system or related tissues
* Neoplasms of uncertain behaviour, except of lymphoid, haematopoietic, central nervous system or related tissues
* Neoplasms of unknown behaviour, except of lymphoid, haematopoietic, central nervous system or related tissues
* Inherited cancer-predisposing syndromes

Neoplasms of brain or central nervous system (BlockL1‑2A0)

A benign or malignant neoplasm that affects the brain, meninges, or spinal cord. Representative examples of primary neoplasms include astrocytoma, oligodendroglioma, ependymoma, and meningioma.

2A00 Primary neoplasms of brain

2A00.0 Gliomas of brain

2A00.00 Glioblastoma of brain

Glioblastomas are malignant astrocytic tumours (grade IV according to the WHO classification). They represent the most frequent brain tumours in adults. They may occur at any age, but 70% of cases are seen in patients between 45 and 70 years of age. The tumours are usually located in the brain hemispheres, but can be found anywhere in the central nervous system.

Inclusions: glioblastoma NOS

2A00.0Y Other specified gliomas of brain

2A00.0Z Gliomas of brain, unspecified

2A00.1 Embryonal tumours of brain

2A00.10 Medulloblastoma of brain

A malignant, invasive embryonal neoplasm arising from the cerebellum. It occurs predominantly in children and has the tendency to metastasize via the cerebrospinal fluid pathways. Signs and symptoms include truncal ataxia, disturbed gait, lethargy, headache, and vomiting. There are four histologic variants: anaplastic medulloblastoma, desmoplastic/nodular medulloblastoma, large cell medulloblastoma, and medulloblastoma with extensive nodularity.

2A00.11 Central primitive neuroectodermal tumour

A malignant neoplasm that originates in the neuroectoderm. The neuroectoderm constitutes the portion of the ectoderm of the early embryo that gives rise to the central and peripheral nervous systems and includes some glial cell precursors.

2A00.1Y Other specified embryonal tumours of brain

2A00.1Z Embryonal tumours of brain, unspecified

2A00.2 Tumours of neuroepithelial tissue of brain

2A00.20 Tumours of the pineal gland or pineal region

2A00.21 Mixed neuronal-glial tumours

2A00.22 Choroid plexus tumours

2A00.2Y Other specified tumours of neuroepithelial tissue of brain

2A00.2Z Tumours of neuroepithelial tissue of brain, unspecified

2A00.3 Central neurocytoma of brain

Central neurocytoma is a very rare brain tumour of young adults. It is typically found in the lateral ventricles and occasionally in the third ventricle. Symptoms are those of increased intracranial pressure.

2A00.4 Astroblastoma of the brain

A rare glial neoplasm more commonly found in young adults. It is characterised by tumour cells with characteristics suggestive of an astrocytic origin (positive for GFAP), arranged perivascularly. The cells have broad, non-tapering processes radiating towards a central blood vessel. The biologic behaviour of astroblastomas is variable, so no WHO grade has been established, yet. (Adapted from WHO.)

2A00.5 Primary neoplasm of brain of unknown or unspecified type

2A01 Primary neoplasms of meninges

2A01.0 Meningiomas

2A01.00 Primary malignant meningioma

2A01.0Y Other specified meningiomas

2A01.0Z Meningiomas, unspecified

2A01.1 Mesenchymal tumours of meninges

2A01.2 Primary neoplasm of meninges of unknown or unspecified type

2A02 Primary neoplasm of spinal cord, cranial nerves or remaining parts of central nervous system

2A02.0 Gliomas of spinal cord, cranial nerves or other parts of the central nervous system

2A02.00 Glioblastoma of spinal cord, cranial nerves or other parts of central nervous system

2A02.0Y Other specified gliomas of spinal cord, cranial nerves or other parts of the central nervous system

2A02.0Z Gliomas of spinal cord, cranial nerves or other parts of the central nervous system, unspecified

2A02.1 Tumours of cranial or paraspinal nerves

Coded Elsewhere: Neurofibroma (2F3Y)

2A02.10 Malignant peripheral nerve sheath tumour of cranial or paraspinal nerves

Malignant schwannoma is a tumour of the peripheral nervous system that arises in the nerve sheath.

Exclusions: Malignant nerve sheath tumour of peripheral nerves or autonomic nervous system, primary site (2B5E)

2A02.11 Paraspinal neuroblastoma

2A02.12 Malignant neoplasm of the optic nerve

2A02.1Y Other specified tumours of cranial or paraspinal nerves

2A02.1Z Tumours of cranial or paraspinal nerves, unspecified

2A02.2 Primary neoplasm of spinal cord or cranial nerves of unknown or unspecified type

2A02.3 Benign neoplasm of cranial nerves

This is a tumour of cranial nerves having none of the characteristics of a malignant neoplasm.

2A02.4 Benign neoplasm of spinal cord

2A0Z Other and unspecified neoplasms of brain or central nervous system

Neoplasms of haematopoietic or lymphoid tissues (BlockL1‑2A2)

A neoplasm arising from hematopoietic cells found in the bone marrow, peripheral blood, lymph nodes and spleen (organs of the hematopoietic system). Hematopoietic cell neoplasms can also involve other anatomic sites (e.g. central nervous system, gastrointestinal tract), either by haematogenous spread, direct tumour infiltration, or neoplastic transformation of extranodal lymphoid tissues. The commonest forms are the various types of leukaemia, Hodgkin and non-Hodgkin lymphomas, myeloproliferative neoplasms and myelodysplastic syndromes.

Coded Elsewhere: Symptoms of blood, blood-forming organs, or the immune system (MA00-MA0Y)

Myeloproliferative neoplasms (BlockL2‑2A2)

2A20 Non mast cell myeloproliferative neoplasms

Coded Elsewhere: Acquired thrombocytosis (3B63.1)

2A20.0 Chronic myelogenous leukaemia, BCR-ABL1-positive

Exclusions: Atypical chronic myeloid leukaemia, BCR-ABL1- negative (2A41)

Chronic myelomonocytic leukaemia (2A40)

Other and unspecified myeloproliferative neoplasms (2A22)

Chronic myeloid leukaemia, not elsewhere classified (2B33.2)

2A20.00 Chronic myelogenous leukaemia with blast crisis

2A20.01 Chronic myelogenous leukaemia, Philadelphia chromosome (Ph1) positive

2A20.02 Chronic myelogenous leukaemia, t(9:22)(q34; q11)

2A20.03 Naegeli-type monocytic leukaemia

2A20.0Y Other specified chronic myelogenous leukaemia, BCR-ABL1-positive

2A20.0Z Chronic myelogenous leukaemia, BCR-ABL1-positive, unspecified

2A20.1 Chronic neutrophilic leukaemia

A rare chronic myeloproliferative neoplasm characterised by sustained peripheral blood neutrophilia, bone marrow hypercellularity due to neutrophilic granulocyte proliferation, and hepatosplenomegaly. The neutrophils lack dysplasia and often show toxic granulations. There is no detectable Philadelphia chromosome or BCR/ABL1 fusion gene.

2A20.2 Primary myelofibrosis

Inclusions: chronic idiopathic myelofibrosis

Exclusions: Acute panmyelosis with myelofibrosis (2A60.38)

2A20.3 Chronic eosinophilic leukaemia, not elsewhere classified

A chronic myeloproliferative neoplasm characterised by persistent eosinophilia in the blood, bone marrow and peripheral tissues. Organ damage occurs as a result of leukaemic infiltration or the release of cytokines, enzymes or other proteins by the eosinophils. Chronic eosinophilic leukaemia, not otherwise specified excludes patients with a Ph chromosome, BCR-ABL1 fusion gene or rearrangement of PDGFRA, PDGFRB or FGFR1.

2A20.4 Polycythaemia vera

2A20.5 Non mast cell myeloproliferative neoplasm, unclassifiable

Cases that have definite features of myeloproliferative neoplasms (MPN), but fail to meet the criteria of a specific MPN subtype.

2A20.Y Other specified non mast cell myeloproliferative neoplasms

2A20.Z Non mast cell myeloproliferative neoplasms, unspecified

2A21 Mastocytosis

Mastocytosis is due to a clonal, neoplastic proliferation of mast cells that accumulate in one or more organ systems. Activating mutations of KIT are frequently found. It is characterised by the presence of multifocal compact clusters or cohesive aggregates/infiltrates of abnormal mast cells. The disorder is heterogeneous, ranging from skin lesions that may spontaneously regress to highly aggressive neoplasms associated with multiorgan failure and short survival. Subtypes of mastocytosis are recognised mainly by the distribution of the disease and clinical manifestations. In cutaneous mastocytosis (CM), the mast cell infiltration remains confined to the skin, whereas systemic mastocytosis (SM) is characterised by involvement of at least one extracutaneous organ with or without evidence of skin lesions. Mastocytosis should be strictly separated from mast cell hyperplasia or mast cell activation states without morphological and/or molecular abnormalities that characterize the neoplastic proliferation.

2A21.0 Systemic mastocytosis

Systemic mastocytosis (SM) comprises a heterogeneous group of rare acquired and chronic haematological malignancies that are related to an abnormal proliferation of mast cells in tissue, including bone marrow, with or without skin involvement. SM can be divided into indolent SM (ISM) and aggressive SM (ASM).

2A21.00 Mast cell leukaemia

2A21.0Y Other specified systemic mastocytosis

2A21.0Z Systemic mastocytosis, unspecified

2A21.1 Cutaneous mastocytosis

Cutaneous mastocytosis is characterised by abnormal accumulation and proliferation of cutaneous mast cells. Most types are isolated but cutaneous mastocytosis can occur in association with systemic disease. Clinical forms include cutaneous mastocytoma, urticaria pigmentosa (the most frequent form), pseudoxanthomatous nodular cutaneous mastocytosis, telangiectasia macularis eruptiva perstans and diffuse cutaneous mastocytosis.

2A21.10 Urticaria pigmentosa

Inclusions: Maculopapular cutaneous mastocytosis

2A21.1Y Other specified cutaneous mastocytosis

2A21.2 Mast cell sarcoma

A rare entity characterised by localised but destructive growth of a tumour consisting of highly atypical, immature mast cells.

2A21.3 Extracutaneous mastocytoma

A localised tumour consisting of mature mast cells.

2A21.Y Other specified mastocytosis

2A21.Z Mastocytosis, unspecified

2A22 Other and unspecified myeloproliferative neoplasms

Exclusions: Chronic myelogenous leukaemia, BCR-ABL1-positive (2A20.0)

Atypical chronic myeloid leukaemia, BCR-ABL1- negative (2A41)

Myelodysplastic syndromes (BlockL2‑2A3)

Clonal hematopoietic disorders characterised by dysplasia and ineffective hematopoiesis in one or more of the hematopoietic cell lines. The dysplasia may be accompanied by an increase in myeloblasts, but the number is less than 20% in marrow and blood, which, according to the WHO guidelines, is the requisite threshold for the diagnosis of acute myeloid leukaemia.

Exclusions: Therapy-related myeloid neoplasms (2A60.2)

Drug-induced aplastic anaemia (3A70.10)

2A30 Refractory anaemia

2A31 Refractory neutropaenia

A myelodysplastic syndrome characterised by the presence of at least 10% dysplastic neutrophils in the bone marrow or the peripheral blood.

2A32 Refractory thrombocytopenia

A myelodysplastic syndrome characterised by the presence of at least 10% dysplastic megakaryocytes, found within at least 30 megakaryocytes examined in the bone marrow.

2A33 Refractory anaemia with ring sideroblasts

A myelodysplastic syndrome characterised by an anaemia in which 15% or more of the erythroid precursors are ringed sideroblasts. The ring sideroblast is an erythroid precursor in which one third or more of the nucleus is encircled by granules which are positive for iron stain.

2A34 Refractory cytopenia with multi-lineage dysplasia

A myelodysplastic syndrome characterised by bi-cytopenia or pancytopenia and dysplastic changes in 10% or more of the cells in two or more of the myeloid cell lines.

2A35 Refractory anaemia with excess of blasts

A myelodysplastic syndrome characterised by bi-cytopenia or pancytopenia and dysplastic changes in one or multiple lineages, with 5-19% myeloblasts in the bone marrow, 2-19% blasts in the blood, or <20% blasts with the presence of Auer rods.

2A36 Myelodysplastic syndrome with isolated del(5q)

A myelodysplastic syndrome characterised by anaemia with or without other cytopenias and/or thrombocytosis and in which the sole cytogenetic abnormality is del(5q). Myeloblasts are <5% in the bone marrow and <1% in the blood.

Inclusions: 5 q- syndrome

2A37 Myelodysplastic syndrome, unclassifiable

A subtype of myelodysplastic syndrome which at disease presentation lacks findings appropriate for classification into any other MDS category, or has an MDS-associated cytogenetic abnormality and cytopenia, but lack sufficient dysplastic changes in any lineage and have <15% ring sideroblasts.

2A38 Refractory cytopenia of childhood

The most common subtype of the myelodysplastic syndromes affecting children. It is characterised by persistent cytopenia with less than 5% blasts in the bone marrow and less than 2% blasts in the peripheral blood.

2A3Y Other specified myelodysplastic syndromes

2A3Z Myelodysplastic syndromes, unspecified

Myelodysplastic and myeloproliferative neoplasms (BlockL2‑2A4)

A category of clonal haematopoietic disorders that have both myelodysplastic and myeloproliferative features at the time of initial presentation.

2A40 Chronic myelomonocytic leukaemia

A myelodysplastic/myeloproliferative neoplasm which is characterised by persistent monocytosis, absence of a Philadelphia chromosome and BCR/ABL1 fusion gene, fewer than 20 percent blasts in the bone marrow and blood, often myelodysplasia, and absence of PDGFRA or PDGFRB rearrangement.

Inclusions: Chronic monocytic leukaemia

Exclusions: Myeloid neoplasm associated with PDGFRA rearrangement (2A50)

Myeloid neoplasm associated with PDGFRB rearrangement (2A51)

2A41 Atypical chronic myeloid leukaemia, BCR-ABL1- negative

A myelodysplastic/myeloproliferative neoplasm characterised by the principal involvement of the neutrophil series with leukocytosis with circulating immature myeloid cells, fewer than 20 percent blasts in the bone marrow and blood, and severe dysgranulopoiesis. The neoplastic cells do not have a Philadelphia chromosome or the BCR/ABL1 fusion gene.

2A42 Juvenile myelomonocytic leukaemia

A myelodysplastic/myeloproliferative neoplasm of childhood that is characterised by proliferation principally of the granulocytic and monocytic lineages. Myelomonocytic proliferation is seen in the bone marrow and the blood. The leukemic cells may infiltrate any tissue, however liver, spleen, lymph nodes, skin, and respiratory tract are the most common sites of involvement.

2A42.0 Juvenile myelomonocytic leukaemia in complete remission

2A42.Y Other specified juvenile myelomonocytic leukaemia

2A42.Z Juvenile myelomonocytic leukaemia, unspecified

2A43 Refractory anaemia with ring sideroblasts associated with marked thrombocytosis

A provisional entity that encompasses cases with morphologic and clinical characteristics of refractory anaemia with ring sideroblasts, marked thrombocytosis, and abnormal megakaryocytes.

2A44 Myeloproliferative and myelodysplastic disease, unclassifiable

This entity includes cases that have clinical, laboratory, and morphologic features that support the diagnosis of both a myelodysplastic syndrome and a myeloproliferative neoplasm, but do not meet the criteria for any of the other entities included in the myelodysplastic/myeloproliferative neoplasm category.

2A4Y Other specified myelodysplastic and myeloproliferative neoplasms

2A4Z Myelodysplastic and myeloproliferative neoplasms, unspecified

Myeloid and lymphoid neoplasms with eosinophilia and abnormalities of PDGFRA, PDGFRB or FGFR1 (BlockL2‑2A5)

A group of rare myeloid and lymphoid neoplasms characterised by rearrangement of the PDGFRA, PDGFRB, or FGFR1 genes, resulting in the formation of fusion transcripts and aberrant tyrosine kinase activity. Eosinophilia is a characteristic finding but it is not always present.

2A50 Myeloid neoplasm associated with PDGFRA rearrangement

2A51 Myeloid neoplasm associated with PDGFRB rearrangement

Myeloid neoplasms characterised by the rearrangement of the PDGFRB gene. Patients usually present with a picture resembling chronic myelomonocytic leukaemia and, less often atypical chronic myeloid leukaemia or chronic eosinophilic leukaemia.

2A52 Myeloid or lymphoid neoplasms with FGFR1 abnormalities

Hematologic neoplasms characterised by the rearrangement of the FGFR1 gene, resulting in translocations with an 8p11 breakpoint. Patients may present with a myeloproliferative neoplasm, acute myeloid leukaemia, lymphoblastic lymphoma/leukaemia of T or B-cell lineage, or acute leukaemia of mixed phenotype.

2A5Z Myeloid and lymphoid neoplasms with eosinophilia and abnormalities of PDGFRA, PDGFRB or FGFR1, unspecified

2A60 Acute myeloid leukaemias and related precursor neoplasms

Acute myeloid leukaemia is characterised by clonal expansion of myeloid blasts in the peripheral blood and bone marrow. Clinical manifestations are fever, pallor, anaemia, hemorrhages and recurrent infections.

2A60.0 Acute myeloid leukaemia with recurrent genetic abnormalities

2A60.1 Acute myeloid leukaemia with myelodysplasia-related changes

An acute myeloid leukaemia with at least 20% blasts in the bone marrow or blood, and either a previous history of myelodysplastic syndrome, multilineage dysplasia or typical myelodysplastic syndrome-related cytogenetic abnormalities. There is no history of prior cytotoxic therapy for an unrelated disorder, and there is absence of the genetic abnormalities that are present in acute myeloid leukaemia with recurrent genetic abnormalities.

2A60.2 Therapy-related myeloid neoplasms

Inclusions: therapy-related myelodysplastic syndromes

2A60.20 Therapy related acute myeloid leukaemia or myelodysplastic syndrome

2A60.2Y Other specified therapy-related myeloid neoplasms

2A60.2Z Therapy-related myeloid neoplasms, unspecified

2A60.3 Acute myeloid leukaemia, not elsewhere classified by criteria of other types

Acute myeloid leukaemias specified by morphological criteria should only be classified as such, if recurrent genetic abnormalities, prior history of a myelodysplastic syndrome or myelodysplastic/myeloproliferative neoplasm, or history of cytotoxic chemotherapy and/or radiotherapy are absent.

Exclusions: Acute myeloid leukaemia with recurrent genetic abnormalities (2A60.0)

Therapy-related myeloid neoplasms (2A60.2)

Acute myeloid leukaemia with myelodysplasia-related changes (2A60.1)

2A60.30 Acute myeloid leukaemia with minimal differentiation

An acute myeloid leukaemia (AML) in which the blasts do not show evidence of myeloid differentiation by morphology and conventional cytochemistry.

2A60.31 Acute myeloid leukaemia without maturation

An acute myeloid leukaemia (AML) characterised by blasts without evidence of maturation to more mature neutrophils.

2A60.32 Acute myeloid leukaemia with maturation

An acute myeloid leukaemia (AML) characterised by blasts with evidence of maturation to more mature neutrophils.

2A60.33 Acute myelomonocytic leukaemia

An acute leukaemia characterised by the proliferation of both neutrophil and monocyte precursors.

2A60.34 Acute monoblastic or monocytic leukaemia

Acute monoblastic leukaemia and acute monocytic leukaemia are myeloid leukaemias in which 80% or more of the leukaemic cells are of monocytic lineage including monoblasts, promonocytes and monocytes; a minor neutrophil component, <20%, may be present.

2A60.35 Acute erythroid leukaemia

Inclusions: Erythroleukaemia

2A60.36 Acute megakaryoblastic leukaemia

An acute myeloid leukaemia in which at least 50% of the blasts are of megakaryocytic lineage.

Inclusions: Acute myeloid leukaemia, M7

Acute megakaryocytic leukaemia

2A60.37 Acute basophilic leukaemia

An acute myeloid leukaemia in which the immature cells differentiate towards basophils. This is a rare leukaemia.

2A60.38 Acute panmyelosis with myelofibrosis

An acute myeloid leukaemia characterised by bone marrow fibrosis without preexisting primary myelofibrosis.

Inclusions: Acute myelofibrosis

Exclusions: Cases that meet criteria for AML with myelodysplasia related changes (2A60.1)

2A60.39 Myeloid sarcoma

Myeloid sarcoma is a rare solid tumour of the myelogenous cells occurring in an extramedullary site.

Inclusions: Chloroma

Granulocytic sarcoma

2A60.3Y Other specified acute myeloid leukaemia, not elsewhere classified by criteria of other types

2A60.3Z Acute myeloid leukaemia, unspecified

2A60.4 Myeloid proliferation associated with Down syndrome

Myeloid neoplasms occurring in individuals with Down syndrome. There is an increased risk of acute leukaemias in both children and adults with Down syndrome. In particular, the incidence of acute myeloid leukaemia in Down syndrome children of less than five years of age is particularly high, it is usually an acute megakaryoblastic leukaemia, and is associated with GATA1 gene mutation. This group of disorders also includes the entity transient abnormal myelopoiesis which occurs in neonates and is associated with GATA1 gene mutation.

2A60.40 Transient abnormal myelopoiesis

A myeloid proliferation occurring in newborns with Down syndrome. It is clinically and morphologically indistinguishable from acute myeloid leukaemia and is associated with GATA1 mutations. The blasts display morphologic and immunophenotypic features of megakaryocytic lineage. In the majority of patients the myeloid proliferation undergoes spontaneous remission.

2A60.41 Myeloid leukaemia associated with Down syndrome

Leukaemia of children with Down syndrome. Encompasses both MDS and AML

2A60.4Y Other specified myeloid proliferation associated with Down syndrome

2A60.4Z Myeloid proliferation associated with Down syndrome, unspecified

2A60.5 Blastic plasmacytoid dendritic cell neoplasm

An aggressive immature hematologic neoplasm formerly known as blastic NK cell lymphoma, composed of cells with a lymphoblast-like morphology. Recent evidence suggests derivation from a plasmacytoid dendritic cell precursor. Patients present with cutaneous tumours and bone marrow involvement.

Inclusions: Blastic NK-cell lymphoma

2A60.Y Other specified acute myeloid leukaemias and related precursor neoplasms

2A60.Z Acute myeloid leukaemias and related precursor neoplasms, unspecified

2A61 Acute leukaemias of ambiguous lineage

An acute leukaemia in which the blasts lack sufficient evidence to classify as myeloid or lymphoid or they have morphologic and/or immunophenotypic characteristics of both myeloid and lymphoid cells.

Precursor lymphoid neoplasms (BlockL2‑2A7)

Neoplasms of immature malignant lymphocytes (lymphoblasts) committed to the B-cell or T-cell lineage. Neoplasms involving the bone marrow and the peripheral blood are called precursor lymphoblastic leukaemias or acute lymphoblastic leukaemias. Neoplasms involving primarily lymph nodes or extranodal sites are called lymphoblastic lymphomas.

2A70 Precursor B-lymphoblastic neoplasms

Neoplasms of lymphoblasts committed to the B-cell lineage.

2A70.0 B Lymphoblastic leukaemia or lymphoma, not elsewhere classified

Precursor B cell neoplasm without defined recurrent genetic abnormality despite appropriate diagnostics

2A70.1 B lymphoblastic leukaemia or lymphoma with t(9:22) (q34;q11.2); BCR-ABL1

A precursor lymphoid neoplasm which is composed of B-lymphoblasts and carries a translocation between the BCR gene on chromosome 22 and the ABL1 gene on chromosome 9. It results in the production of the p190 kd or p210 kd fusion protein. It has an unfavorable clinical outcome.

2A70.Y Other B-lymphoblastic leukaemia/lymphoma with recurrent genetic abnormalities

2A71 Precursor T-lymphoblastic neoplasms

A neoplasm of lymphoblasts committed to the T-cell lineage, typically composed of small to medium-sized blast cells.

2A7Z Precursor lymphoid neoplasms, unspecified

Mature B-cell neoplasms (BlockL2‑2A8)

Non-Hodgkin lymphomas that originate from mature B lymphocytes. May reside in lymph nodes, lymphatic tissue of different organs or bone marrow and blood (then frequently called leukaemia).

2A80 Follicular lymphoma

Follicular lymphoma (FL) is a neoplasm composed of follicle centre (germinal centre) B-cells (typically both centrocytes and centroblasts/large transformed cells), which usually has at least a partially follicular pattern. t(14;18) with BCL2 rearrangement is frequently observed. If diffuse areas of any size comprised predominantly or entirely of blastic cells are present in any case of follicular lymphoma, a diagnosis of diffuse large B-cell lymphoma is also made. Lymphomas composed of centro cytes and centroblasts with an entirely diffuse pattern in the sampled tissue may be included in this category.

Inclusions: follicular lymphoma with or without diffuse areas

Exclusions: Mature T-cell or NK-cell neoplasms (BlockL2‑2A9)

2A80.0 Follicular lymphoma grade 1

2A80.1 Follicular lymphoma grade 2

2A80.2 Follicular lymphoma grade 3

2A80.3 Primary cutaneous follicle centre lymphoma

A primary lymphoma of the skin composed of various numbers of small and large irregular neoplastic follicle center cells. Its morphologic pattern can be nodular, diffuse, or nodular and diffuse. It presents with solitary or grouped plaques and tumours, and it usually involves the scalp, forehead, or trunk. It rarely involves the legs. This type of cutaneous lymphoma tends to remain localised to the skin, and it has a favorable prognosis.

2A80.4 Paediatric type follicular lymphoma

A variant of follicular lymphoma often involving cervical or other peripheral lymph nodes and the Waldeyer ring. It is frequently localised, and often lacks BCL-2 protein expression and never has a BCL2 translocation. It is usually but not exclusively seen in the pediatric population. The prognosis is usually favorable.

2A80.5 Follicular lymphoma in situ

2A80.6 Follicular lymphoma of small intestine

2A80.Y Other specified follicular lymphoma

2A80.Z Follicular lymphoma, unspecified

2A81 Diffuse large B-cell lymphomas

Non-Hodgkin lymphomas are characterised by a proliferation of predominantly large neoplastic B lymphocytes.

Coded Elsewhere: Diffuse large B-cell lymphoma of small intestine (2B80.Y)

2A81.0 Primary mediastinal large B-cell lymphoma

A large B-cell non-Hodgkin lymphoma arising in the mediastinum. Morphologically it is characterised by a massive diffuse lymphocytic proliferation associated with compartmentalizing fibrosis.

2A81.1 Intravascular large B-cell lymphoma

2A81.2 Plasmablastic lymphoma

An aggressive diffuse large B-cell lymphoma frequently arising in the setting of HIV infection and characterised by the presence of large neoplastic cells resembling B-immunoblasts which have the immunophenotypic profile of plasma cells. Sites of involvement include the oral cavity and other extranodal sites

2A81.3 Lymphomatoid granulomatosis

2A81.4 T-cell/histiocyte rich large B-cell lymphoma

A large B-cell lymphoma characterised by the presence of a limited number of scattered neoplastic large B-lymphocytes which are admixed with numerous non-neoplastic T-lymphocytes and frequently histiocytes.

2A81.5 Primary diffuse large B-cell lymphoma of central nervous system

2A81.6 Epstein-Barr Virus-positive diffuse large B cell lymphoma of the elderly

An aggressive diffuse large B-cell lymphoma affecting patients older than 50 years. Epstein-Barr virus is present in all cases. There is no known history of immunodeficiency or prior lymphoma. The majority of patients present with extranodal disease.

2A81.7 Diffuse large B-cell lymphoma associated with chronic inflammation

A diffuse large B-cell lymphoma arising in body cavities or narrow spaces of long standing chronic inflammation. The classic example is the pyothorax-associated lymphoma that arises in the pleural cavity of patients with a history of long standing pyothorax.

2A81.8 ALK-positive large B-cell lymphoma

A usually aggressive large B-cell lymphoma characterised by the presence of monomorphic immunoblast-like neoplastic B-lymphocytes in a sinusoidal growth pattern. The neoplastic B-lymphocytes express the ALK kinase but they lack the 2;5 translocation.

2A81.9 Primary effusion lymphoma

An aggressive non-Hodgkin B-cell lymphoma composed of large cells, presenting as a serous effusion without detectable tumour masses. It is universally associated with human herpes virus 8 (HHV-8)/Kaposi sarcoma herpes virus (KSHV) [HHV-8/KSHV]. It mostly occurs in the setting of immunodeficiency; most cases have been reported in HIV positive patients. The most common sites of involvement are the pleural, pericardial, and peritoneal cavities. The prognosis is extremely unfavorable.

2A81.A Primary cutaneous diffuse large B-cell lymphoma, leg type

An aggressive primary cutaneous B-cell lymphoma, usually involving the lower leg. It is composed of a generally monotonous proliferation of immunoblasts, or less frequently centroblasts, with few admixed reactive cells. This type of lymphoma occurs most often in the elderly who present with rapidly growing tumours, usually on one or both legs. Dissemination to extracutaneous sites is frequent.

2A81.Y Other specified diffuse large B-cell lymphomas

2A81.Z Diffuse large B-cell lymphoma, not otherwise specified

2A82 Mature B-cell neoplasm with leukaemic behaviour

2A82.0 Chronic lymphocytic leukaemia or small lymphocytic lymphoma

An indolent, mature B-cell neoplasm composed of small, round B-lymphocytes. When the bone marrow and peripheral blood are involved, the term chronic lymphocytic leukaemia is used. The term small lymphocytic lymphoma is restricted to cases which do not show leukemic involvement of the bone marrow and peripheral blood.

Inclusions: Small cell B-cell lymphoma

2A82.00 Chronic lymphocytic leukaemia of B-cell type

Chronic lymphocytic leukaemia/small lymphocytic lymphoma (CLL/SLL) is a neoplasm composed of monomorphic small, round to slightly irregular B lymphocytes in the peripheral blood (PB), bone marrow (BM), spleen and lymph nodes, admixed with prolymphocytes and paraimmuno - blasts forming proliferation centres in tissue infiltrates.The CLL/SLL cells usually coexpress CD5 and CD23. In the absence of extramedullary tissue involvement, there must be ?5x109/L monoclonal lymphocytes with a CLL phenotype in the PB. The International Workshop on Chronic Lymphocytic leukaemia (IWCLL) report requires that the lymphocytosis be present for at least 3 months and also allows for the diagnosis of CLL to be made with lower lymphocyte counts in patients with cytopenias or disease-related symptoms {873A}. Whether patients who would have fulfilled the criteria in the past for CLL but who fulfill the criteria only for monoclonal B lymphocytosis (MBL) are better considered to have low stage CLL or MBL remains to be determined. Some may prefer to still consider many of these cases more like CLL. The term SLL is used for non-leukaemic cases with the tissue morphology and immunophenotype of CLL. The IWCLL definition of SLL requires lymphadenopathy, no cytopenias due to BM infiltration by CLL/SLL and <5x109/L PB B-cells {873A}.

Inclusions: Lymphoplasmacytic leukaemia

Exclusions: Lymphoplasmacytic lymphoma (2A85.4)

Coded Elsewhere: Richter syndrome (2A81.Y)

2A82.0Y Other specified chronic lymphocytic leukaemia or small lymphocytic lymphoma

2A82.0Z Chronic lymphocytic leukaemia or small lymphocytic lymphoma, unspecified

2A82.1 B-cell prolymphocytic leukaemia

2A82.10 B-cell prolymphocytic leukaemia in complete remission

2A82.1Y Other specified b-cell prolymphocytic leukaemia

2A82.1Z B-cell prolymphocytic leukaemia, unspecified

2A82.2 Hairy-cell leukaemia

A neoplasm of small B-lymphocytes with hairy projections in bone marrow, spleen, and peripheral blood. Most patients present with splenomegaly and pancytopenia.

Inclusions: Leukaemic reticuloendotheliosis

2A82.3 Splenic B-cell lymphoma or leukaemia, unclassifiable

A small B-cell clonal lymphoproliferative disorder of the spleen that does not fall into any of the other categories of mature B-cell neoplasms.

2A82.Y Other specified mature B-cell neoplasm with leukaemic behaviour

2A82.Z Mature B-cell neoplasm with leukaemic behaviour, unspecified

2A83 Plasma cell neoplasms

Plasma cells, usually secreting monoclonal immunoglobulin (M-protein) and/or immunoglobulin light chains.

2A83.0 Monoclonal gammopathy of undetermined significance

2A83.1 Plasma cell myeloma

A bone marrow-based plasma cell neoplasm usually characterised by a serum monoclonal protein and/or urinary light chains. ”CRAB” criteria (osteolytic lesions, hypercalcaemia, renal failure, and anaemia) separate symptomatic plasma cell myeloma form asymptomatic (smoldering) myeloma.

Inclusions: Kahler disease

Myelomatosis

Medullary plasmacytoma

multiple myeloma

Exclusions: Solitary plasmacytoma (2A83.2)

2A83.2 Solitary plasmacytoma

A single focus of clonal (malignant) plasma cells either in the bone or in another anatomic site without peripheral blood involvement. --2003

Inclusions: Solitary myeloma

2A83.3 Extraosseous plasmacytoma

2A83.4 Plasma cell leukaemia

An aggressive plasma cell neoplasm. It is characterised by the presence of neoplastic plasma cells in the peripheral blood. The peripheral blood plasma cells comprise more than 20% of the peripheral blood white cells or the number of clonal plasma cells in the PB exceeds 2x10?/L.

2A83.5 Monoclonal immunoglobulin deposition disease

2A83.50 Heavy chain deposition disease

A disease of the kidney, caused by proliferation and deposition of pieces of truncated or abnormal alpha, gamma, delta, or mu immunoglobulin heavy chain segments of white blood cells. This disease is characterised by fibrillar or granular tissue deposits and renal dysfunction, which may lead to organ failure. Confirmation is by identifying heavy chain deposition tissue biopsy using immunofluorescence under a microscope.

Exclusions: Heavy chain diseases or malignant immunoproliferative diseases (2A84)

Immunoglobulin heavy chain deficiency (4A01.04)

2A83.51 Light and heavy chain deposition disease

A disease of the kidney, caused by proliferation and deposition of pieces of truncated or abnormal light and heavy chain segments of white blood cells. This disease is characterised by fibrillar or granular tissue deposits and renal dysfunction, which may lead to organ failure. Confirmation is by identification of light and heavy chain deposition tissue biopsy under a microscope.

2A83.52 Light chain deposition disease

A disease of the kidney, caused by the deposition of pieces of truncated or abnormal light chain segments of white blood cells. This disease is characterised by fibrillar or granular tissue deposits and renal dysfunction, which may lead to organ failure. Confirmation is by identification of light chain deposition tissue biopsy under an electron microscope.

Exclusions: Immunodeficiencies with isotype or light chain deficiencies with normal number of B cells (4A01.04)

2A83.Y Other specified multiple myeloma and plasma cell neoplasms

2A83.Z Plasma cell neoplasm, unspecified

2A84 Heavy chain diseases or malignant immunoproliferative diseases

A group of rare disorders of immunoglobulin synthesis associated with B-cell proliferative disorders that produce monoclonal heavy chains and typically no light chains.

2A84.0 Alpha heavy chain disease

The small intestinal morphologic changes are consistent with a mucosa-associated lymphoid tissue lymphoma (MALT lymphoma).

2A84.1 Gamma heavy chain disease

A clonal disorder characterised by the secretion of a truncated gamma chain. In most cases, it is associated with morphologic changes also seen in lymphoplasmacytic lymphomas, but the clinical course is typically more aggressive than in lymphoplasmacytic lymphoma/Waldenstrom's macroglobulinemia.

Inclusions: Franklin disease

2A84.2 Mu heavy chain disease

2A84.Y Other specified malignant immunoproliferative diseases

2A84.Z Heavy chain diseases, unspecified

2A85 Other specified mature B-cell neoplasms or lymphoma

2A85.0 Nodal marginal zone lymphoma

A primary nodal B-cell non-Hodgkin lymphoma which morphologically resembles lymph nodes involved by marginal zone lymphomas of extranodal or splenic types, but without evidence of extranodal or splenic disease. This is a rare entity, and most patients present with localised or generalised lymphadenopathy. The clinical course is indolent.

2A85.1 Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue of stomach

A low grade, indolent B-cell lymphoma, usually associated with Helicobacter pylori infection. Morphologically it is characterised by a dense mucosal atypical lymphocytic (centrocyte-like cell) infiltrate with often prominent lymphoepithelial lesions and plasmacytic differentiation. Some of gastric MALT lymphomas carry the t(11;18)(q21;q21). Such cases are resistant to Helicobacter pylori therapy.

2A85.2 Extranodal marginal zone B-cell lymphoma, primary site skin

A low-grade, extranodal marginal zone B-cell lymphoma of the mucosa-associated lymphoid tissue that arises from the skin. It usually presents with multifocal papular or nodular lesions in the arms or trunk. It rarely disseminates to internal organs or progresses to high grade lymphoma.

2A85.3 Extranodal marginal zone B-cell lymphoma, primary site excluding stomach or skin

2A85.4 Lymphoplasmacytic lymphoma

Neoplasm of small B lymphocytes and plasma cells, mostly residing in the bone marrow. Frequently associated with the production of an IgM serum monoclonal protein, then called Waldenström macroglobulinemia (WM).

Inclusions: primary macroglobulinaemia

Waldenström macroglobulinaemia

Waldenström macroglobulinaemia without mention of remission

Exclusions: small cell B-cell lymphoma (2A82.0)

Chronic lymphocytic leukaemia or small lymphocytic lymphoma (2A82.0)

2A85.5 Mantle cell lymphoma

Mantle cell lymphoma is a rare form of malignant non-Hodgkin lymphoma affecting B lymphocytes in the lymph nodes in a region called the ``mantle zone''. It accounts for 2-10% of lymphomas.

Inclusions: Small cell mantle cell lymphoma

2A85.6 Burkitt lymphoma including Burkitt leukaemia

A highly aggressive lymphoma composed of monomorphic medium-sized B-cells with basophilic cytoplasm and numerous mitotic figures. It is often associated with the presence of Epstein-Barr virus (EBV) and is commonly seen in AIDS patients. Three morphologic variants are recognised: classical Burkitt lymphoma, Burkitt lymphoma with plasmacytoid differentiation, and atypical Burkitt/Burkitt-like lymphoma. All cases express the MYC translocation [t(8;14)].

Inclusions: “Burkitt-like” lymphoma

Coded Elsewhere: HIV - [human immunodeficiency virus] disease associated with Burkitt lymphoma (1C62.3)

2A85.Y Further specified mature B-cell neoplasms or lymphoma

2A86 B-cell lymphoma, mixed features

2A86.0 Malignant lymphoma of B cell type, not elsewhere classified

Coding Note: If B-cell lineage or involvement is mentioned in conjunction with a specific lymphoma, code to the more specific description.

2A86.1 B-cell lymphoma unclassifiable with features intermediate between Burkitt lymphoma and diffuse large B-cell lymphoma

2A86.2 B-cell lymphoma unclassifiable with features intermediate between classical Hodgkin lymphoma and diffuse large B-cell lymphoma

2A86.Y Other specified B-cell lymphoma, mixed features

2A86.Z B-cell lymphoma, mixed features, unspecified

2A8Z Mature B-cell neoplasms, unspecified

Mature T-cell or NK-cell neoplasms (BlockL2‑2A9)

A group of neoplasms composed of T-lymphocytes with a mature (peripheral/post-thymic) immunophenotypic profile and/or NK-cells.

2A90 Mature T-cell lymphoma, specified types, nodal or systemic

2A90.0 T-cell prolymphocytic leukaemia

An aggressive T-cell leukaemia, characterised by the proliferation of small to medium sized prolymphocytes with a mature T-cell phenotype, involving the blood, bone marrow, lymph nodes, liver, spleen, and skin.

2A90.1 T-cell large granular lymphocytic leukaemia

A T-cell peripheral neoplasm characterised by a persistent (>6 months) increase in the number of peripheral blood large granular lymphocytes, without a clearly identified cause.

2A90.2 Chronic lymphoproliferative disorders of NK-cells

Heterogeneous disorders with a chronic clinical course affecting predominantly adults and characterised by the proliferation of large granular lymphocytes with natural killer cell immunophenotype.

2A90.3 Aggressive NK cell leukaemia

A rare, highly aggressive, Epstein-Barr virus-associated leukaemia, also known as aggressive NK-cell leukaemia/lymphoma; it may represent the leukemic counterpart of nasal type extranodal NK/T-cell lymphomas. It affects primarily teenagers and young adults. It is characterised by the systemic proliferation of NK cells in the peripheral blood, bone marrow, liver, and spleen.

2A90.4 Systemic Epstein-Barr Virus-positive T-cell lymphoma of childhood

This neoplasm of childhood is characterised by a clonal proliferation of EBV-infected T-cells with an activated cytotoxic phenotype. It can occur shortly after primary acute EBV infection or in the setting of chronic active EBV infection (CAEBV).

2A90.5 Adult T-cell lymphoma or leukaemia, human T-cell lymphotropic virus type 1-associated

A peripheral (mature) T-cell neoplasm linked to the human T-cell leukaemia virus type 1 (HTLV-1). Adult T-cell leukaemia/lymphoma is endemic in several regions of the world, in particular Japan, the Caribbean, and parts of Central Africa.

Coded Elsewhere: Adult T-cell leukaemia or lymphoma, skin (2B0Y)

2A90.6 Extranodal NK/T-cell lymphoma, nasal type

An aggressive, predominantly extranodal, mature T-cell non-Hodgkin lymphoma. It is characterised by an often angiocentric and angiodestructive cellular infiltrate composed of EBV positive NK/T cells. The nasal cavity is the most common site of involvement. Patients often present with midfacial destructive lesions (lethal midline granuloma). The disease may disseminate rapidly to various anatomic sites including the gastrointestinal tract, skin, testis, and cervical lymph nodes. It is also known as angiocentric T-cell lymphoma. The term polymorphic reticulosis has been widely used to describe the morphologic changes seen in this type of lymphoma. However, the latter term may also apply to lymphomatoid granulomatosis, which is an angiocentric and angiodestructive EBV positive B-cell lymphoproliferative disorder.

2A90.7 Enteropathy associated T-cell lymphoma

An uncommon mature T-cell lymphoma of intraepithelial lymphocytes. It usually arises from the small intestine, most commonly the jejunum or ileum. Other less frequent primary anatomic sites include the duodenum, stomach, colon, or outside the gastrointestinal tract. Type II of this lymphoma may occur sporadically outside the context of celiac disease.

Inclusions: Enteropathy type intestinal T-cell lymphoma

Intestinal T-cell lymphoma

2A90.8 Hepatosplenic T-cell lymphoma

An extranodal, mature T-cell non-Hodgkin lymphoma that originates from cytotoxic T-cells, usually of gamma/delta T-cell type. It is characterised by the presence of medium-size neoplastic lymphocytes infiltrating the hepatic sinusoids. A similar infiltrating pattern is also present in the spleen and bone marrow that are usually involved at the time of the diagnosis.

2A90.9 Angioimmunoblastic T-cell lymphoma

A mature T-cell non-Hodgkin lymphoma, characterised by systemic disease and a polymorphous infiltrate involving lymph nodes and extranodal sites. The clinical course is typically aggressive.

Inclusions: AILD - [angioimmunoblastic lymphadenopathy with dysproteinaemia]

2A90.A Anaplastic large cell lymphoma, ALK-positive

A T-cell peripheral lymphoma composed of usually large, pleomorphic, CD30 positive T-lymphocytes with abundant cytoplasm characterised by the presence of a translocation involving the ALK gene and expression of ALK fusion protein. Most patients present with peripheral and/or abdominal lymphadenopathy, and often have advanced disease and extranodal involvement.

Inclusions: Anaplastic large cell lymphoma, CD30-positive

2A90.B Anaplastic large cell lymphoma, ALK-negative

A T-cell peripheral lymphoma morphologically indistinguishable from anaplastic large cell lymphoma, ALK-positive. It is characterised by the absence of the translocation involving the ALK gene and lacks expression of ALK fusion protein.

Exclusions: Primary cutaneous CD-30 positive T-cell lymphoproliferative disorders (2B03)

2A90.C Peripheral T-cell lymphoma, not otherwise specified

A heterogeneous category of nodal and extranodal mature T-cell lymphomas, which do not correspond to any of the specifically defined entities of mature T-cell lymphoma in the current classification.

Inclusions: T-zone variant Peripheral T-cell lymphoma

Lymphoepithelioid lymphoma

Follicular variant Peripheral T-cell lymphoma

Mature T-cell or NK-cell lymphoma, primary cutaneous specified types (BlockL3‑2B0)

Inclusions: Primary cutaneous peripheral T-cell lymphoma

Exclusions: Skin infiltration by nodal or non-cutaneous extranodal lymphoma (2E08)

2B00 Subcutaneous panniculitis-like T-cell lymphoma

Subcutaneous panniculitis-like T-cell lymphoma is a neoplasm of alpha/beta, usually CD8+ T-cells, mainly confined to the subcutis, presenting clinically as subcutaneous nodules which are usually not ulcerated.

2B01 Mycosis fungoides

A peripheral (mature) T-cell lymphoma presenting in the skin with patches/plaques or less commonly with tumours or erythroderma. It is characterised by epidermal and dermal infiltration of small to medium-sized T-cells with cerebriform nuclei.

2B02 Sézary syndrome

A generalised peripheral (mature) T-cell neoplasm characterised by the presence of erythroderma, lymphadenopathy, and neoplastic, cerebriform T-lymphocytes in the blood. Sézary syndrome is an aggressive disease.

2B03 Primary cutaneous CD-30 positive T-cell lymphoproliferative disorders

Primary skin disorders characterised immunohistologically by infiltration by neoplastic CD30+ lymphocytes.

Inclusions: Primary cutaneous CD30+large T-cell lymphoma

2B03.0 Primary cutaneous CD30 positive anaplastic large cell lymphoma

An anaplastic large cell lymphoma limited to the skin at the time of diagnosis. Most patients present with solitary or localised skin lesions, which may be tumours, nodules or papules. The t(2;5) translocation that is present in many cases of systemic anaplastic large cell lymphoma, is not found in this disease.

2B03.1 Lymphomatoid papulosis

Lymphomatoid papulosis is a proliferation of T-cells, often clonal, characterised clinically by the appearance of crops of dome-shaped papules and nodules which tend to ulcerate and then heal with scarring.

2B0Y Other specified primary cutaneous mature T-cell or NK-cell lymphomas and lymphoproliferative disorders

2B0Z Primary cutaneous T-cell lymphoma of undetermined or unspecified type

2B2Y Other specified mature T-cell or NK-cell neoplasms

2B2Z Mature T-cell or NK-cell neoplasms, unspecified

2B30 Hodgkin lymphoma

Malignant lymphomas, previously known as Hodgkin's disease, characterised by the presence of large tumour cells in an abundant admixture of nonneoplastic cells. There are two distinct subtypes: nodular lymphocyte predominant Hodgkin lymphoma and classical Hodgkin lymphoma. Hodgkin lymphoma involves primarily lymph nodes.

2B30.0 Nodular lymphocyte predominant Hodgkin lymphoma

Nodular lymphocyte predominant Hodgkin lymphoma (NLPHL) is characterised by a nodular, or a nodular and diffuse proliferation of scattered large neoplastic cells known as popcorn or lymphocyte predominant cells (LP cells) —formerly called L&H cells for lymphocytic and/or histiocytic Reed-Sternberg cell variants. At present an overlap between NLPHL and T-cell-rich large B-cell lymphoma cannot be excluded.

2B30.1 Classical Hodgkin lymphoma

Classical Hodgkin lymphoma is a B-cell lymphoma characterised histologically by the presence of large mononuclear Hodgkin cells and multinucleated Reed-Sternberg (HRS) cells.

A monoclonal B-cell lymphoproliferation in the vast majority of cases. It is characterised by a bimodal age distribution (15-30 years of life and late life) and is often associated with EBV infection. In less than 5% of cases it is a monoclonal proliferation of T-lymphocytes. Morphologically, it is characterised by the presence of Reed-Sternberg cells and mononuclear Hodgkin cells. The Reed-Sternberg and mononuclear Hodgkin cells are CD30 positive in nearly all cases and CD15 positive in the majority of cases.

Inclusions: Classical Hodgkin lymphoma, type not specified

2B30.10 Nodular sclerosis classical Hodgkin lymphoma

A subtype of classical Hodgkin lymphoma characterised by collagen bands surrounding lymphoid nodules. The lymphoid nodules contain lacunar and Reed-Sternberg cells. Mediastinal involvement occurs in 80% of patients. The prognosis of nodular sclerosis Hodgkin lymphoma is slightly better than that of mixed cellularity or lymphocyte depleted subtype.

2B30.11 Lymphocyte-rich classical Hodgkin lymphoma

2B30.12 Mixed cellularity classical Hodgkin lymphoma

A subtype of classical Hodgkin lymphoma with a mixed inflammatory stroma containing Hodgkin and Reed-Sternberg cells.

2B30.13 Lymphocyte depleted classical Hodgkin lymphoma

2B30.1Z Classical Hodgkin lymphoma, unspecified

2B30.Z Hodgkin lymphoma, unspecified

2B31 Histiocytic or dendritic cell neoplasms

True histiocytic malignancies are vanishing diagnoses due to improved understanding of the provenance of malignant cells.

2B31.0 Juvenile xanthogranuloma

It is characterised by the presence of lipid-laden, foamy histiocytes and Touton-type giant cells in the dermis.

2B31.1 Histiocytic sarcoma

Inclusions: Malignant Histiocytosis

2B31.2 Langerhans cell histiocytosis

A neoplastic proliferation of Langerhans cells which contain Birbeck granules by ultrastructural examination. Three major overlapping syndromes are recognised: eosinophilic granuloma, Letterer-Siwe disease, and Hand-Schuller-Christian disease. The clinical course is generally related to the number of organs affected at presentation.

Inclusions: Histiocytosis X

2B31.20 Langerhans cell histiocytosis involving the skin

2B31.2Y Other specified Langerhans cell histiocytosis

2B31.3 Langerhans cell sarcoma

A neoplastic proliferation of Langerhans cells with overtly malignant cytologic features. It can be considered a higher grade variant of Langerhans cell histiocytosis (LCH) and it can present de novo or progress from antecedent LCH.

2B31.4 Interdigitating dendritic cell sarcoma

A neoplastic proliferation of spindle to ovoid cells which show phenotypic features similar to those of interdigitating dendritic cells. The clinical course is generally aggressive.

2B31.5 Follicular dendritic cell sarcoma

A neoplasm composed of spindle to ovoid cells which have morphologic and immunophenotypic characteristics of follicular dendritic cells. It affects lymph nodes and other sites including the tonsils, gastrointestinal tract, spleen, liver, soft tissues, skin, and oral cavity. It usually behaves as a low grade sarcoma. Recurrences have been reported in up to half of the cases.

2B31.6 Indeterminate dendritic cell tumour

A very rare dendritic cell tumour composed of spindle to ovoid cells with a phenotype that is similar to the Langerhans cells. Patients usually present with cutaneous papules, nodules, and plaques. Systemic symptoms are usually absent. The clinical course is variable.

2B31.7 Fibroblastic reticular cell tumour

A very rare dendritic cell tumour affecting the lymph nodes, spleen, and soft tissues. Morphologically it is similar to the interdigitating dendritic cell sarcoma or follicular dendritic cell sarcoma. The tumour cells are positive for cytokeratin and CD68. Clinical outcome is variable.

2B31.Y Other specified histiocytic or dendritic cell neoplasms

2B31.Z Histiocytic or dendritic cell neoplasms, unspecified

2B32 Immunodeficiency-associated lymphoproliferative disorders

Post-transplant lymphoproliferative disorder (PTLD) is a polyclonal (benign) or clonal (malignant) proliferation of lymphoid cells that develops as a consequence of immunosuppression in a recipient of a solid organ or bone marrow allograft. PTLDs comprise a spectrum ranging from early, Epstein-Barr virus (EBV)-driven polyclonal lymphoid proliferations to EBV-positive or EBV- negative lymphomas of predominantly B-cell or less often T-cell type. In other Immunodeficiency-associated lymphoproliferative disorders, association with EBV is less pronounced.

Inclusions: PTLD - [Post transplant lymphoproliferative disorder]

2B32.0 Post-transplant lymphoproliferative disorder, early lesion

A lymphoproliferative disorder arising as a result of post-transplant immunosuppression therapy. It is characterised by the lack of tissue destruction and the architectural preservation of the involved tissues. It includes two morphologic variants: plasmacytic hyperplasia and infectious mononucleosis-like lymphoproliferative disorders.

2B32.1 Reactive plasmacytic hyperplasia

2B32.2 Post-transplant lymphoproliferative disorder, Infectious mononucleosis-like

2B32.3 Polymorphic post-transplant lymphoproliferative disorder

2B32.Y Other specified immunodeficiency-associated lymphoproliferative disorders

2B32.Z Immunodeficiency-associated lymphoproliferative disorders, unspecified

2B33 Malignant haematopoietic neoplasms without further specification

Coding Note: Only to be designated in cases with incomplete diagnostics.

2B33.0 Acute leukaemia, not elsewhere classified

Coding Note: Only to be designated in cases with incomplete diagnostics.

2B33.1 Myeloid leukaemia

2B33.2 Chronic myeloid leukaemia, not elsewhere classified

Coding Note: Only to be designated in cases. with incomplete diagnostics

2B33.3 Lymphoid leukaemia, not elsewhere classified

Coding Note: Only to be designated in cases with incomplete diagnostics.

2B33.4 Leukaemia, unspecified

Coding Note: Only to be designated in cases with incomplete diagnostics.

2B33.5 Malignant lymphoma, not elsewhere classified

Coding Note: Only to be designated in cases with incomplete diagnostics.

2B33.Y Other malignant haematopoietic neoplasms without further specification

Coding Note: Only to be designated in cases with incomplete diagnostics.

2B3Z Neoplasms of haematopoietic or lymphoid tissues, unspecified

Malignant neoplasms, except primary neoplasms of lymphoid, haematopoietic, central nervous system or related tissues (BlockL1‑2B5)

Coding Note: For use of this category, reference should be made to the mortality coding rules and guidelines in the Reference Guide.

Exclusions: Neoplasms of brain or central nervous system (BlockL1‑2A0)

Neoplasms of haematopoietic or lymphoid tissues (BlockL1‑2A2)

Malignant neoplasms, stated or presumed to be primary, of specified sites, except of lymphoid, haematopoietic, central nervous system or related tissues (BlockL2‑2B5)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

A usually aggressive malignant mesenchymal cell tumour most commonly arising from muscle, fat, fibrous tissue, bone, cartilage, and blood vessels. Sarcomas occur in both children and adults. The prognosis depends largely on the degree of differentiation (grade) of the tumour. Representative subtypes are liposarcoma, leiomyosarcoma, osteosarcoma, and chondrosarcoma.

Exclusions: Mesenchymal tumours of meninges (2A01.1)

Tumours of cranial or paraspinal nerves (2A02.1)

2B50 Chondrosarcoma, primary site

Exclusions: Osteosarcoma, primary site (2B51)

2B50.0 Chondrosarcoma of bone or articular cartilage of limbs

2B50.1 Chondrosarcoma of bone or articular cartilage of pelvis

2B50.2 Chondrosarcoma of bone or articular cartilage of ribs, sternum or clavicle

2B50.Y Chondrosarcoma of bone or articular cartilage of other specified sites

2B50.Z Chondrosarcoma of bone or articular cartilage of unspecified sites

2B51 Osteosarcoma, primary site

A usually aggressive malignant bone-forming mesenchymal tumour, predominantly affecting adolescents and young adults. It usually involves bones and less frequently extraosseous sites. It often involves the long bones (particularly distal femur, proximal tibia, and proximal humerus). Pain with or without a palpable mass is the most frequent clinical symptom. It may spread to other anatomic sites, particularly the lungs.

2B51.0 Osteosarcoma of bone or articular cartilage of jaw

2B51.1 Osteosarcoma of bone or articular cartilage of limbs

2B51.2 Osteosarcoma of bone or articular cartilage of pelvis

2B51.Y Osteosarcoma of bone and articular cartilage of other specified sites

2B51.Z Osteosarcoma of bone and articular cartilage of unspecified sites

2B52 Ewing sarcoma, primary site

A small round cell tumour that lacks morphologic, immunohistochemical, and electron microscopic evidence of neuroectodermal differentiation. It represents one of the two ends of the spectrum called Ewing's sarcoma/peripheral neuroectodermal tumour. It affects mostly males under age 20, and it can occur in soft tissue or bone. Pain and the presence of a mass are the most common clinical symptoms.

2B52.0 Ewing sarcoma of bone or articular cartilage of limbs

2B52.1 Ewing sarcoma of bone or articular cartilage of pelvis

2B52.2 Ewing sarcoma of bone or articular cartilage of ribs

2B52.3 Ewing sarcoma of soft tissue

A rare malignant neoplasm of the soft tissues. It is typically a disease of children and young adults. It is characterised by t(11:22) (q24: q12) resulting in the expression of EWS/FLI-1 chimeric transcript. Most commonly occurs in the paravertebral region, chest wall, pelvis and lower extremities.

2B52.Y Ewing sarcoma of bone and articular cartilage of other specified sites

2B52.Z Ewing sarcoma of bone and articular cartilage of unspecified sites

2B53 Fibroblastic or myofibroblastic tumour, primary site

2B53.0 Myxofibrosarcoma, primary site

2B53.1 Fibroblastic or myofibroblastic tumour of skin

2B53.Y Other specified fibroblastic or myofibroblastic tumour, primary site

2B53.Z Fibroblastic or myofibroblastic tumour, primary site, unspecified

2B54 Unclassified pleomorphic sarcoma, primary site

A pleomorphic sarcoma characterised by the presence of fibrohistiocytic cells and spindle cells arranged in a storiform pattern.

2B54.0 Unclassified pleomorphic sarcoma of skin

A rare malignant neoplasm arising from the skin. It is characterised by the presence of spindle cells in a storiform pattern and histiocytes with abundant cytoplasm.

Inclusions: malignant fibrous histiocytoma of skin

2B54.1 Unclassified pleomorphic sarcoma of retroperitoneum or peritoneum

2B54.Y Unclassified pleomorphic sarcoma, primary site, other specified site

2B54.Z Unclassified pleomorphic sarcoma, primary site, unspecified site

2B55 Rhabdomyosarcoma, primary site

Rhabdomyosarcoma is a malignant soft tissue tumour which develops from cells of striated muscle. It is the most common form of tumour found in children and adolescents.

2B55.0 Rhabdomyosarcoma of the oral cavity or pharynx

2B55.1 Rhabdomyosarcoma of respiratory or intrathoracic organs

2B55.2 Rhabdomyosarcoma of male genital organs

2B55.Y Rhabdomyosarcoma, other specified primary site

2B55.Z Rhabdomyosarcoma, unspecified primary site

2B56 Angiosarcoma, primary site

2B56.0 Angiosarcoma of heart

2B56.1 Angiosarcoma of skin

A malignant tumour arising from the endothelial cells of the blood vessels. Microscopically, it is characterised by frequently open vascular anastomosing and branching channels. The malignant cells that line the vascular channels are spindle or epithelioid and often display hyperchromatic nuclei. Angiosarcomas most frequently occur in the skin and breast. Patients with long-standing lymphoedema are at increased risk of developing angiosarcoma.

2B56.2 Angiosarcoma of breast

A malignant vascular neoplasm arising from the breast.

2B56.3 Angiosarcoma of liver

A malignant vascular neoplasm arising from the liver.

Inclusions: Kupffer cell sarcoma of liver

2B56.Y Angiosarcoma, other specified primary site

2B56.Z Angiosarcoma, unspecified primary site

2B57 Kaposi sarcoma, primary site

A malignant neoplasm characterised by a vascular proliferation which usually contains blunt endothelial cells. Erythrocyte extravasation and hemosiderin deposition are frequently present. The most frequent site of involvement is the skin; however it may also occur internally. It generally develops in people with compromised immune systems including those with acquired immune deficiency syndrome (AIDS).

Coded Elsewhere: Human immunodeficiency virus disease associated with Kaposi sarcoma (1C62.3)

2B57.0 Kaposi sarcoma of lung

2B57.1 Kaposi sarcoma of skin

A Kaposi sarcoma arising from the skin. It presents with patches, plaques, or nodules.

2B57.2 Kaposi sarcoma of gastrointestinal sites

2B57.Y Kaposi sarcoma of other specified primary sites

2B57.Z Kaposi sarcoma of unspecified primary site

2B58 Leiomyosarcoma, primary site

2B58.0 Leiomyosarcoma of retroperitoneum or peritoneum

2B58.1 Leiomyosarcoma of uterus

2B58.2 Leiomyosarcoma of stomach

This is a malignant nonepithelial tumour that arises from cells lining the stomach that develop into smooth-muscle.

2B58.Y Leiomyosarcoma, other specified primary site

2B58.Z Leiomyosarcoma, unspecified primary site

2B59 Liposarcoma, primary site

Liposarcoma, a type of soft tissue sarcoma, describes a group of lipomatous tumours of varying severity ranging from slow-growing to aggressive and metastatic. Liposarcomas are most often located in the lower extremities or retroperitoneum, but they can also occur in the upper extremities, neck, peritoneal cavity, spermatic cord, breast, vulva and axilla.

2B59.0 Liposarcoma of soft tissue of limb

2B59.1 Liposarcoma of retroperitoneum or peritoneum

2B59.2 Liposarcoma of male genital organs

2B59.Y Liposarcoma, other specified primary site

2B59.Z Liposarcoma, unspecified primary site

2B5A Synovial sarcoma, primary site

A malignant neoplasm characterised by the chromosomal translocation t(X;18)(p11;q11). It can occur at any age, but mainly affects young adults, more commonly males. Although any site can be affected, the vast majority of the cases arise in the deep soft tissues of extremities, especially around the knee. Microscopically, synovial sarcoma is classified as monophasic (with a spindle or epithelial cell component) or biphasic (with both spindle and epithelial cell components). Synovial sarcomas can recur or metastasize to the lungs, bones, and lymph nodes.

2B5A.0 Synovial sarcoma of soft tissues of limb

2B5A.1 Synovial sarcoma of respiratory or intra-thoracic organs

2B5A.Y Synovial sarcoma, other specified primary site

2B5A.Z Synovial sarcoma, unspecified primary site

2B5B Gastrointestinal stromal tumour, primary site

This is the most common mesenchymal tumour that arises in the gastrointestinal tract. It is generally immunohistochemically positive for CD117 (KIT), phenotypically paralleling Cajal-cell differentiation, and most examples contain KIT- or PDGFRA- activating mutations. It is most frequent in the stomach and to a lesser degree in the small intestine. The prognosis depends on the tumour size and the mitotic activity.

2B5B.0 Gastrointestinal stromal tumour of stomach

A gastrointestinal stromal tumour that arises from the stomach. It covers a spectrum of benign to malignant mesenchymal neoplasms and includes most gastric smooth muscle tumours, leiomyoblastomas, and tumours formerly called gastrointestinal autonomic nerve tumours.

2B5B.1 Gastrointestinal stromal tumour of small intestine

A gastrointestinal stromal tumour that arises from the small intestine. It usually affects adults over fifty years of age. The majority of cases have spindle cell morphology. The prognosis depends on the tumour size and the mitotic activity.

2B5B.Y Gastrointestinal stromal tumour of other gastrointestinal sites

2B5B.Z Gastrointestinal stromal tumour of unspecified gastrointestinal sites

2B5C Endometrial stromal sarcoma, primary site

A malignant, infiltrating mesenchymal tumour arising from the uterine corpus, cervix, vagina, and the ovary. Based on its morphologic characteristics, it is classified as either a low grade or an undifferentiated (high grade) stromal sarcoma. The low grade endometrioid stromal sarcoma is characterised by the presence of oval to spindle-shape cells that resemble the cells of the endometrial stroma, without evidence of significant atypia and pleomorphism. Numerous small vessels are also present. The undifferentiated stromal sarcoma is characterised by an aggressive clinical course, the presence of significant cellular atypia, pleomorphism, and high mitotic activity.

2B5D Malignant mixed epithelial mesenchymal tumour, primary site

2B5D.0 Malignant mixed epithelial mesenchymal tumour of ovary

Malignant mixed epithelial mesenchymal tumour of the ovary is a rare and very aggressive neoplasm presenting most commonly in postmenopausal women and is composed of adenocarcinomatous and sarcomatous elements and, depending on the types of these elements, can be classified as homologous or heterologous. It often has a poor prognosis.

2B5D.1 Malignant mixed epithelial and mesenchymal tumour of corpus uteri

A primary malignant neoplasm of the uterine corpus characterised by the presence of an epithelial and a mesenchymal component. This category includes carcinosarcoma, carcinofibroma, and adenosarcoma.

2B5D.Y Malignant mixed epithelial mesenchymal tumour, other specified primary site

2B5D.Z Malignant mixed epithelial mesenchymal tumour, unspecified primary site

2B5E Malignant nerve sheath tumour of peripheral nerves or autonomic nervous system, primary site

Exclusions: Malignant peripheral nerve sheath tumour of cranial or paraspinal nerves (2A02.10)

2B5F Sarcoma, not elsewhere classified, primary site

2B5F.0 Sarcoma, not elsewhere classified of uterus

Coded Elsewhere: Endometrial stromal sarcoma, primary site (2B5C)

Leiomyosarcoma of uterus (2B58.1)

Rhabdomyosarcoma of corpus uteri (2B55.Y)

2B5F.1 Sarcoma, not elsewhere classified of retroperitoneum or peritoneum

2B5F.10 Myosarcomas of omentum

2B5F.1Y Other specified sarcoma, not elsewhere classified of retroperitoneum or peritoneum

2B5F.1Z Sarcoma, not elsewhere classified of retroperitoneum or peritoneum, unspecified

2B5F.2 Sarcoma, not elsewhere classified of other specified sites

2B5F.3 Sarcoma, not elsewhere classified, primary site unknown

2B5G Myosarcoma of uterus, part not specified

Coded Elsewhere: Leiomyosarcoma of uterus (2B58.1)

Rhabdomyosarcoma of corpus uteri (2B55.Y)

2B5H Well differentiated lipomatous tumour, primary site

2B5J Malignant miscellaneous tumours of bone or articular cartilage of other or unspecified sites

Exclusions: Neoplasms of haematopoietic or lymphoid tissues (BlockL1‑2A2)

2B5K Unspecified malignant soft tissue tumours or sarcomas of bone or articular cartilage of other or unspecified sites

2B5Y Other specified malignant mesenchymal neoplasms

2B5Z Malignant mesenchymal neoplasm of unspecified type

Malignant neoplasms of lip, oral cavity or pharynx (BlockL3‑2B6)

Squamous cell carcinomas amount to more than 90% of malignant tumours of the oral cavity and oropharynx. As in other parts of the upper aerodigestive tract, there is a strong and synergistic association with tobacco smoking and alcohol abuse. In some regions, particularly the Indian subcontinent, oral cancer is among the most frequent malignancies, largely due to tobacco chewing.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B60 Malignant neoplasms of lip

Malignant neoplasms originating from the transitional epithelium of the lip (excluding oral mucosa and skin of the outer lip) or from the underlying anatomical structures (e.g. orbicularis oris muscle).

Exclusions: Malignant neoplasm of skin of lip (BlockL3‑2C3)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B60.0 Basal cell carcinoma of lip

A basal cell carcinoma arising from the lip.

2B60.1 Squamous cell carcinoma of lip

Squamous cell carcinoma located on or originating in the mucosa or vermilion of the lip, including the vermilion border but excluding the skin of the lip.

Exclusions: Squamous cell carcinoma of skin of lip (2C31)

2B60.Y Other specified malignant neoplasms of lip

2B60.Z Malignant neoplasms of lip, unspecified

2B61 Malignant neoplasms of base of tongue

A primary neoplasm involving the base of the tongue, often associated with chronic alcohol and tobacco use, older age, certain geographic locations, a family history of upper aeordigestive tract cancers and/or certain nutritional deficiencies and infectious agents.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B61.0 Squamous cell carcinoma of the base of the tongue

A carcinoma that arises from the base of the tongue. Representative examples include squamous cell carcinoma, adenoid cystic carcinoma, and mucoepidermoid carcinoma.

2B61.Y Other specified malignant neoplasms of base of tongue

2B61.Z Malignant neoplasms of base of tongue, unspecified

2B62 Malignant neoplasms of other or unspecified parts of tongue

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B62.0 Squamous cell carcinoma of other or unspecified parts of tongue

2B62.1 Malignant neoplasms of lingual tonsil

2B62.10 Squamous cell carcinoma of lingual tonsil

2B62.Y Other specified malignant neoplasms of other and unspecified parts of tongue

2B62.Z Malignant neoplasms of other or unspecified parts of tongue, unspecified

2B63 Malignant neoplasms of gum

Exclusions: Benign osteogenic tumours of bone or articular cartilage of skull or face (2E83.0)

Benign osteogenic tumours of bone or articular cartilage of lower jaw (2E83.1)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B63.0 Squamous cell carcinoma of gum

2B63.Y Other specified malignant neoplasm of gum

2B63.Z Malignant neoplasms of gum, unspecified

2B64 Malignant neoplasms of floor of mouth

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B64.0 Squamous cell carcinoma of floor of mouth

2B64.Y Other specified malignant neoplasm of floor of mouth

2B64.Z Malignant neoplasms of floor of mouth, unspecified

2B65 Malignant neoplasms of palate

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B65.0 Adenocarcinoma of palate

2B65.1 Squamous cell carcinoma of palate

2B65.Y Other specified malignant neoplasm of palate

2B65.Z Malignant neoplasms of palate, unspecified

2B66 Malignant neoplasms of other or unspecified parts of mouth

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B66.0 Squamous cell carcinoma of other or unspecified parts of mouth

2B66.Y Other specified malignant neoplasms of other and unspecified parts of mouth

2B66.Z Malignant neoplasms of other or unspecified parts of mouth, unspecified

2B67 Malignant neoplasms of parotid gland

Salivary gland tumours can show a striking range of morphological diversity between different tumour types and sometimes within an individual tumour mass. In addition, hybrid tumours, dedifferentiation and the propensity for some benign tumours to progress to malignancy can confound histopathological interpretation. These features, together with the relative rarity of a number of tumours, can sometimes make diagnosis difficult, despite the abundance of named tumour entities. The increasing use of pre-operative fine needle aspiration biopsies also needs to be taken into account, as artifactual changes may be superimposed on the tumours. Unfortunately, the morphological variability of these tumours is mirrored by the immunocytochemical profiles, so that special stains are rarely useful in routine diagnosis of salivary gland epithelial neoplasms.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B67.0 Adenocarcinoma of parotid gland

2B67.1 Squamous cell carcinoma of parotid gland

2B67.Y Other specified malignant neoplasms of parotid gland

2B67.Z Malignant neoplasms of parotid gland, unspecified

2B68 Malignant neoplasms of submandibular or sublingual glands

Salivary gland tumours can show a striking range of morphological diversity between different tumour types and sometimes within an individual tumour mass. In addition, hybrid tumours, dedifferentiation and the propensity for some benign tumours to progress to malignancy can confound histopathological interpretation. These features, together with the relative rarity of a number of tumours, can sometimes make diagnosis difficult, despite the abundance of named tumour entities. The increasing use of pre-operative fine needle aspiration biopsies also needs to be taken into account, as artifactual changes may be superimposed on the tumours. Unfortunately, the morphological variability of these tumours is mirrored by the immunocytochemical profiles, so that special stains are rarely useful in routine diagnosis of salivary gland epithelial neoplasms.

Exclusions: Malignant neoplasms of parotid gland (2B67)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B68.0 Adenocarcinoma of submandibular or sublingual glands

2B68.1 Squamous cell carcinoma of submandibular or sublingual glands

2B68.2 Other specified malignant neoplasms of submandibular or sublingual glands

2B68.Z Malignant neoplasms of submandibular or sublingual glands, unspecified

2B69 Malignant neoplasms of tonsil

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

Coded Elsewhere: Malignant neoplasms of pharyngeal tonsil (2B6B.2)

Malignant neoplasms of lingual tonsil (2B62.1)

2B69.0 Squamous cell carcinoma of tonsil

2B69.1 Other specified malignant neoplasms of tonsil

2B69.Z Malignant neoplasms of tonsil, unspecified

2B6A Malignant neoplasms of oropharynx

Malignant neoplasms of the oral cavity and pharynx

Exclusions: Malignant neoplasms of palate (2B65)

Malignant neoplasms of tonsil (2B69)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B6A.0 Squamous cell carcinoma of oropharynx

A squamous cell carcinoma arising from the oropharynx. It predominantly affects adults in their fifth and sixth decades of life and is associated with alcohol and tobacco use. Human papillomavirus is present in approximately half of the cases. It is characterised by a tendency to metastasize early to the lymph nodes. When the tumour is small, patients are often asymptomatic. Physical examination may reveal erythematous or white lesions or plaques. The majority of patients present with locally advanced disease. Signs and symptoms include mucosal ulceration, pain, bleeding, weight loss, neck swelling, and difficulty speaking, chewing, and swallowing. Patients may also present with swollen neck lymph nodes without any symptoms from the oropharyngeal tumour. The most significant prognostic factors are the size of the tumour and the lymph nodes status.

2B6A.Y Other specified malignant neoplasms of oropharynx

2B6A.Z Malignant neoplasms of oropharynx, unspecified

2B6B Malignant neoplasms of nasopharynx

A wide variety of tumours can arise in the nasopharynx, but it is nasopharyngeal carcinoma that has fascinated generations of oncologists, pathologists, scientists and epidemiologists. It shows marked geographic differences, with highest incidence rates in Southern Chinese. In some endemic areas, the incidence has declined by about 30% over the past two decades, suggesting that environmental or lifestyle factors may play a major role and that the disease is, to some extent, preventable. Nasopharyngeal carcinoma shows a very strong association with Epstein-Barr virus (EBV) infection, irrespective of the ethnic origin of the patients. This association has pioneered a new paradigm of utilizing viral serological tests for the diagnosis of cancer and for screening in high-risk populations. Naso -pharyngeal carcinoma is generally responsive to radiation therapy, and the clinical outcome has greatly improved over the years, due to refinements in staging and to improved therapy protocols. The unusual and often deceptive histological features of nasopharyngeal carcinoma have generated controversies over the nature of the tumour and still pose a challenge to surgical pathologists. There have possibly been more names invented for the various histological subtypes of nasopharyngeal carcinoma than any other tumour type.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B6B.0 Squamous cell carcinoma of nasopharynx

2B6B.1 Malignant epithelial neoplasms of nasopharynx, unspecified type

2B6B.2 Malignant neoplasms of pharyngeal tonsil

2B6B.20 Squamous cell carcinoma of pharyngeal tonsil

2B6B.21 Other or unspecified malignant epithelial neoplasm of pharyngeal tonsil

2B6B.2Y Other specified malignant neoplasms of pharyngeal tonsil

2B6B.2Z Malignant neoplasm of pharyngeal tonsil without mention of type

2B6B.Y Other specified malignant neoplasms of nasopharynx

2B6B.Z Malignant neoplasms of nasopharynx, unspecified

2B6C Malignant neoplasms of piriform sinus

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B6C.0 Squamous cell carcinoma of piriform sinus

2B6C.Y Other specified malignant neoplasms of piriform sinus

2B6C.Z Malignant neoplasms of piriform sinus, unspecified

2B6D Malignant neoplasms of hypopharynx

A malignant neoplasm arising in the hypopharynx.

Exclusions: Malignant neoplasms of piriform sinus (2B6C)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B6D.0 Squamous cell carcinoma of hypopharynx and variants

A squamous cell carcinoma arising from the hypopharynx. Signs and symptoms include dysphagia, hemoptysis, and the presence of a neck mass.

2B6D.Y Other specified malignant neoplasms of hypopharynx

2B6D.Z Malignant neoplasms of hypopharynx, unspecified

2B6E Malignant neoplasms of other or ill-defined sites in the lip, oral cavity or pharynx

Exclusions: Malignant neoplasm of oral cavity NOS (2B66)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B6E.0 Squamous cell carcinoma of other or ill-defined sites in the lip, oral cavity or pharynx

2B6E.Y Other specified malignant neoplasms of other or ill-defined sites in the lip, oral cavity or pharynx

2B6E.Z Malignant neoplasms of other or ill-defined sites in the lip, oral cavity or pharynx, unspecified

2B6Y Other specified malignant neoplasms of lip, oral cavity or pharynx

2B6Z Malignant neoplasms of lip, oral cavity or pharynx, unspecified

Malignant neoplasms of digestive organs (BlockL3‑2B7)

A primary malignant neoplasm involving any part of the gastrointestinal system.

Exclusions: Malignant neoplasm metastasis in digestive system (BlockL3‑2D8)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

Malignant neoplasms of lip, oral cavity or pharynx (BlockL3‑2B6)

2B70 Malignant neoplasms of oesophagus

A primary malignant neoplasm involving the oesophagus

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B70.0 Adenocarcinoma of oesophagus

A malignant tumour with glandular differentiation arising predominantly from Barrett mucosa in the lower third of the esophagus. Grossly, esophageal adenocarcinomas are similar to esophageal squamous cell carcinomas. Microscopically, adenocarcinomas arising in the setting of Barrett esophagus are typically papillary and/or tubular. The prognosis is poor.

2B70.00 Barrett adenocarcinoma

Barrett adenocarcinoma is defined as adenocarcinoma of lower oesophagus and gastroesophageal junction associated with Barrett's oesophagus.

2B70.0Y Other specified adenocarcinoma of oesophagus

2B70.0Z Adenocarcinoma of oesophagus, unspecified

2B70.1 Squamous cell carcinoma of oesophagus

A squamous cell carcinoma arising from the esophagus. It can be associated with a long history of tobacco and alcohol abuse and is exceedingly rare before the age of 30. The median age is around 65 in both males and females, but the incidence in males is much higher than in females. It is located mostly in the middle and lower third of the esophagus. Grossly, polypoid, ulcerated, plaque-like and occult lesions have been described. The microscopic features are the same as in other squamous cell carcinomas. Any degree of differentiation may occur, and variation within a single tumour is common. The prognosis is poor.

2B70.Y Other specified malignant neoplasms of oesophagus

2B70.Z Malignant neoplasms of oesophagus, unspecified

2B71 Malignant neoplasms of oesophagogastric junction

Malignant neoplasms that arise from the cells of the oesophagogastric junction (OGJ), which is defined as the point at which the oesophagus ends and the stomach begins. This mainly defines adenocarcinomas that straddle the junction of the oesophagus and stomach. This definition includes many tumours formally called cancers of the gastric cardia.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B71.0 Adenocarcinoma of oesophagogastric junction

An adenocarcinoma that arises from and straddles the junction of the stomach and esophagus. The category of adenocarcinomas of the gastroesophageal junction also includes the majority of adenocarcinomas previously called gastric cardia adenocarcinomas. Squamous cell carcinomas that affect or cross the junction of the stomach and esophagus are classified as carcinomas of the distal esophagus. Adenocarcinoma of the gastroesophageal junction occurs more often in Caucasian middle aged and elderly males. Clinical signs and symptoms include dysphagia, abdominal pain, and weight loss. The prognosis depends on the completeness of the surgical resection, the number of lymph nodes involved by cancer, and the presence or absence of postoperative complications.

2B71.Y Other specified malignant neoplasm of oesophagogastric junction

2B71.Z Malignant neoplasms of oesophagogastric junction, unspecified

2B72 Malignant neoplasms of stomach

A primary or metastatic malignant neoplasm involving the stomach.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

Coded Elsewhere: Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue of stomach (2A85.1)

Leiomyosarcoma of stomach (2B58.2)

Gastrointestinal stromal tumour of stomach (2B5B.0)

2B72.0 Adenocarcinoma of stomach

An adenocarcinoma arising from the stomach glandular epithelium. Gastric adenocarcinoma is primarily a disease of older individuals. It most commonly develops after a long period of atrophic gastritis and is strongly associated with Helicobacter pylori infection. The lack of early symptoms often delays the diagnosis of gastric cancer. The majority of patients present with advanced tumours which have poor rates of curability. Microscopically, two important histologic types of gastric adenocarcinoma are recognised: the intestinal and diffuse type. The overall prognosis of gastric adenocarcinomas is poor, even in patients who receive a curative resection.

2B72.1 Malignant neuroendocrine neoplasm of stomach

A neoplasm with neuroendocrine differentiation that arises from the stomach. It includes well differentiated neuroendocrine tumours (low and intermediate grade) and poorly differentiated neuroendocrine carcinomas (high grade).

Inclusions: carcinoid and other malignant neuroendocrine neoplasms

Coded Elsewhere: Neuroendocrine neoplasm of duodenum (2B80.01)

Neuroendocrine neoplasms of appendix (2B81.2)

2B72.Y Other specified malignant neoplasms of stomach

2B72.Z Malignant neoplasms of stomach, unspecified

Malignant neoplasms of intestine (BlockL4‑2B8)

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B80 Malignant neoplasms of small intestine

A primary malignant neoplasm involving the small intestine.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

Coded Elsewhere: Enteropathy associated T-cell lymphoma (2A90.7)

Gastrointestinal stromal tumour of small intestine (2B5B.1)

Malignant neoplasm of jejunum (2B80.1Y)

Malignant neoplasm of ileum (2B80.1Y)

Small intestinal leiomyosarcoma (2B58.Y)

2B80.0 Malignant neoplasms of duodenum

A primary malignant neoplasm that affects the duodenum. Representative examples include carcinoma, lymphoma, and sarcoma.

Coded Elsewhere: Lymphoma of duodenum (2B33.5)

Malignant mesenchymal tumour of the duodenum (2B5F.2)

2B80.00 Adenocarcinoma of duodenum

An adenocarcinoma that arises from the duodenum.

2B80.01 Neuroendocrine neoplasm of duodenum

Neoplasms that arise from the cells of neuroendocrine system lining the duodenum.

2B80.0Y Other specified malignant neoplasms of the duodenum

2B80.0Z Malignant neoplasms of duodenum, unspecified

2B80.1 Malignant neoplasms of jejunum or ileum

2B80.10 Adenocarcinoma of jejunum or ileum

2B80.11 Neuroendocrine neoplasms of jejunum or ileum

Neoplasms that arise from the cells of neuroendocrine system lining the jejunum or ileum including well-differentiated (low- to intermediated grade) neuroendocrine tumours. These include carcinoid tumour.

2B80.1Y Other specified malignant neoplasms of jejunum or ileum

2B80.1Z Malignant neoplasms of jejunum or ileum, unspecified

2B80.2 Malignant neoplasms of small intestine, site unspecified

2B80.20 Adenocarcinoma of small intestine, site unspecified

A malignant tumour with glandular differentiation arising from epithelium of small intestine.

Exclusions: Neuroendocrine neoplasms of small intestine, site unspecified (2B80.21)

2B80.21 Neuroendocrine neoplasms of small intestine, site unspecified

Neoplasms that arise from the cells of neuroendocrine system lining the small intestine.

2B80.2Y Other specified malignant neoplasms of small intestine, site unspecified

2B80.Y Other specified malignant neoplasms of small intestine

2B80.Z Malignant neoplasms of small intestine, unspecified

2B81 Malignant neoplasms of appendix

A primary malignant neoplasm that affects the appendix.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

Coded Elsewhere: Lymphoma of the appendix (2B33.5)

2B81.0 Adenocarcinoma of appendix

A malignant neoplasm arising from the glandular epithelium of the appendix. Most are mucinous adenocarcinomas.

2B81.1 Mucinous adenocarcinoma of appendix

An adenocarcinoma, often cystic, with large amounts of extracellular mucin

2B81.2 Neuroendocrine neoplasms of appendix

Malignant neoplasms with neuroendocrine differentiation that arise in the appendix. Most are well differentiated neuroendocrine tumours (low and intermediate grade), i.e. carcinoids. Poorly differentiated neuroendocrine carcinomas (high grade) are exceedingly rare.

2B81.Y Other specified malignant neoplasms of appendix

2B81.Z Malignant neoplasms of appendix, unspecified

Malignant neoplasms of large intestine (BlockL5‑2B9)

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2B90 Malignant neoplasms of colon

Primary malignant neoplasms arising in the colon.

Exclusions: Malignant neoplasms of appendix (2B81)

Coded Elsewhere: Gardner syndrome (LD2D.3)

Malignant Lymphoma of colon (2B33.5)

Gastrointestinal stromal tumour of colon (2B5B.Y)

Kaposi sarcoma of colon (2B57.2)

Leiomyosarcoma of colon (2B58.Y)

2B90.0 Malignant neoplasm of ascending colon and right flexure of colon

2B90.00 Adenocarcinoma of ascending colon or right flexure of colon

A malignant tumour with glandular differentiation arising from epithelium of ascending colon and right flexure.

2B90.0Y Other specified malignant neoplasm of ascending colon and right flexure of colon

2B90.0Z Malignant neoplasm of ascending colon and right flexure of colon, unspecified

2B90.1 Malignant neoplasm of descending colon and splenic flexure of colon

2B90.10 Adenocarcinoma of descending colon or splenic flexure of colon

A malignant tumour with glandular differentiation arising from epithelium of descending colon and splenic flexure.

2B90.1Y Other specified malignant neoplasm of descending colon and splenic flexure of colon

2B90.1Z Malignant neoplasm of descending colon and splenic flexure of colon, unspecified

2B90.2 Malignant neoplasm of transverse colon

2B90.20 Adenocarcinoma of transverse colon

A malignant tumour with glandular differentiation arising from epithelium of transverse colon.

2B90.2Y Other specified malignant neoplasm of transverse colon

2B90.2Z Malignant neoplasm of transverse colon, unspecified

2B90.3 Malignant neoplasm of sigmoid colon

Exclusions: Malignant neoplasms of rectosigmoid junction (2B91)

2B90.30 Adenocarcinoma of sigmoid colon

A malignant tumour with glandular differentiation arising from epithelium of descending colon and splenic flexure.

2B90.3Y Other specified malignant neoplasm of sigmoid colon

2B90.3Z Malignant neoplasm of sigmoid colon, unspecified

2B90.Y Other specified malignant neoplasms of colon

2B90.Z Malignant neoplasms of colon, unspecified

2B91 Malignant neoplasms of rectosigmoid junction

2B91.0 Adenocarcinoma of rectosigmoid junction

A malignant tumour with glandular differentiation arising from epithelium of rectosigmoid junction

2B91.Y Other specified malignant neoplasms of rectosigmoid junction

2B91.Z Malignant neoplasms of rectosigmoid junction, unspecified

2B92 Malignant neoplasms of rectum

Coded Elsewhere: Malignant mesenchymal tumour of rectum (2B5F.2)

2B92.0 Adenocarcinomas of rectum

An adenocarcinoma arising from the rectum. It is more frequently seen in populations with a Western type diet and in patients with a history of chronic inflammatory bowel disease.

2B92.1 Neuroendocrine neoplasms of rectum

Malignant neoplasms with neuroendocrine differentiation that arise in the rectum. Most are well differentiated neuroendocrine tumours (low and intermediate grade), i.e. carcinoids. Poorly differentiated neuroendocrine carcinomas (high grade) are rare.

2B92.Y Other specified malignant neoplasms of rectum

2B92.Z Malignant neoplasms of rectum, unspecified

2B93 Malignant neoplasms of large intestine, site unspecified

2B93.0 Adenocarcinoma of large intestine, site unspecified

2B93.Y Other specified malignant neoplasms of large intestine, site unspecified

2B93.Z Malignant neoplasms of large intestine, site and type unspecified

2B9Y Other specified malignant neoplasms of large intestine

2B9Z Malignant neoplasms of large intestine, unspecified

2C00 Malignant neoplasms of anus or anal canal

A primary malignant neoplasm that arises in the anal canal up to the junction with perianal, hair-bearing skin. Representative examples include carcinomas and melanomas. Tumours of the anal margin are classified with skin tumours.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C00.0 Adenocarcinoma of anus or anal canal

An adenocarcinoma arising in the anal canal epithelium, including the mucosal surface, the anal glands, and the lining of fistulous tracts.

2C00.1 Melanoma of anus or anal canal

A form of cancer that develops in melanocytes, the cells that produce pigment melanin in the skin.

2C00.2 Neuroendocrine neoplasm of anus or anal canal

Neoplasms that arise from the cells of the neuroendocrine system lining the anus and anal canal.

2C00.3 Squamous cell carcinoma of anus or anal canal

A squamous cell carcinoma (SCC) arising from the anal canal up to the junction with the anal margin (perianal, hair-bearing skin). Human papillomavirus is detected in the majority of cases. Homosexual HIV-positive men have an increased risk of developing anal squamous cell carcinoma in comparison to the general male population. The prognosis is generally better for anal margin SCC than for anal canal SCC. The former are classified with skin tumours.

2C00.Y Other specified malignant neoplasms of anus and anal canal

2C00.Z Malignant neoplasms of anus or anal canal, unspecified

2C0Y Other specified malignant neoplasms of intestine

2C0Z Malignant neoplasms of intestine, unspecified

2C10 Malignant neoplasm of pancreas

A primary malignant tumour of the pancreas. Most are adenocarcinomas.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C10.0 Adenocarcinoma of pancreas

An adenocarcinoma which arises from the exocrine pancreas. Ductal adenocarcinoma and its variants are the most common types of pancreatic adenocarcinoma.

2C10.1 Neuroendocrine neoplasms of pancreas

A neoplasm with neuroendocrine differentiation that arises from the pancreas. It includes neuroendocrine tumours (low and intermediate grade) and neuroendocrine carcinomas (high grade).

2C10.Y Other specified malignant neoplasms of pancreas

2C10.Z Malignant neoplasm of pancreas, unspecified

2C11 Malignant neoplasms of other or ill-defined digestive organs

A primary malignant tumour involving a digestive organ or organs not coded elsewhere (including intestinal tract [part unspecified], overlapping lesions of the digestive tract and other ill-defined sites within the digestive system)

Exclusions: Malignant neoplasms of retroperitoneum (2C50)

Malignant neoplasms of peritoneum (2C51)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C11.0 Adenocarcinoma of other or ill-defined digestive organs

2C11.1 Mucinous carcinoma of other or ill-defined digestive organs

2C11.2 Other specified malignant neoplasms of other or ill-defined digestive organs

2C11.Z Malignant neoplasms of other or ill-defined digestive organs, unspecified

2C12 Malignant neoplasms of liver or intrahepatic bile ducts

The most frequent and important hepatic neoplasm is the primary hepatocellular carcinoma (HCC). In many parts of the world, in particular Africa and Asia, it poses a significant disease burden. In these high incidence regions, chronic infection with hepatitis B virus (HBV) is the principal underlying cause, with the exception of Japan which has a high prevalence of hepatitis C infection. HBV vaccination has become a powerful tool in reducing cirrhosis and HCC, but implementation is still suboptimal in several high risk regions. In Western countries, chronic alcohol abuse is a major aetiological factor. Hepatic cholangiocarcinoma has a different geographical distribution, with peak incidences in Northern Thailand. Here, it is caused by chronic infection with the liver fluke, Opisthorchis Viverrini, which is ingested through infected raw fish.

Exclusions: Secondary malignant neoplasm of liver (2D80)

Malignant neoplasm of biliary tract NOS (2C17)

2C12.0 Malignant neoplasm of liver

2C12.00 Combined hepatocellular-cholangiocarcinoma

Combined hepatocellular-cholagiocarcinoma is a tumour containing unequivocal, intimately mixed elements of both hepatocellular carcinoma and cholangiocarcinoma.

Inclusions: Hepatocholangiocarcinoma

2C12.01 Hepatoblastoma

A malignant liver neoplasm that occurs almost exclusively in infants, although isolated cases in older children and adults have been reported. Grossly, hepatoblastoma is solid, well circumscribed, and more often solitary than multiple. Microscopically, most of the tumours are composed exclusively of immature hepatocytic elements. About a fourth of hepatoblastomas contain a stromal component that may be undifferentiated or develop into bone or cartilage.

2C12.02 Hepatocellular carcinoma of liver

A carcinoma that arises from the hepatocytes.

2C12.03 Mesothelial carcinoma of liver

2C12.0Y Other specified malignant neoplasm of liver

2C12.1 Malignant neoplasm of intrahepatic bile ducts

2C12.10 Intrahepatic cholangiocarcinoma

A carcinoma that arises from the intrahepatic bile duct epithelium in any site of the intrahepatic biliary tree. Grossly, the malignant lesions are solid, nodular, and grayish. Morphologically, the vast majority of cases are adenocarcinomas. Early detection is difficult and the prognosis is generally poor.

2C12.1Y Other specified malignant neoplasms of intrahepatic bile ducts

2C12.Z Malignant neoplasms of liver or intrahepatic bile ducts, unspecified

2C13 Malignant neoplasms of gallbladder

A malignant tumour arising from the epithelium of the gallbladder. It is usually associated with the presence of gallstones. Clinical symptoms are not specific and usually present late in the course. Morphologically, adenocarcinoma is the most common type, however squamous cell carcinomas, adenosquamous carcinomas, signet ring carcinomas, and undifferentiated carcinomas can also occur.

2C13.0 Adenocarcinoma of the gallbladder

An adenocarcinoma arising from the gallbladder. It is the most common malignant tumour of the gallbladder, typically in the fundus; it is usually well to moderately differentiated. The incidence is higher in patients with gallstones than in patients without gallstones. Signs and symptoms usually present late in the course of the disease and are reminiscent of those of chronic cholecystitis including right upper quadrant pain. Histologic variants include adenocarcinoma of the intestinal type, clear cell adenocarcinoma, mucinous adenocarcinoma, papillary adenocarcinoma, and signet ring adenocarcinoma.

2C13.Y Other specified malignant neoplasm of gallbladder

2C13.Z Malignant neoplasms of gallbladder, unspecified

2C14 Malignant neoplasms of proximal biliary tract, cystic duct

2C14.0 Adenocarcinoma of proximal biliary tract, cystic duct

2C14.1 Mucinous cystic neoplasm with associated invasive carcinoma of cystic duct

2C14.2 Neuroendocrine neoplasms of cystic duct

2C14.Y Other specified malignant neoplasms of biliary tract, cystic duct

2C14.Z Malignant neoplasms of proximal biliary tract, cystic duct, unspecified

2C15 Malignant neoplasms of biliary tract, distal bile duct

2C15.0 Adenocarcinoma of biliary tract, distal bile duct

An adenocarcinoma that arises from the common bile duct distal to the insertion of the cystic duct.

2C15.1 Mucinous cystic neoplasm with associated invasive carcinoma of distal bile duct

2C15.2 Neuroendocrine neoplasms of distal bile duct

2C15.Y Other specified malignant neoplasms of biliary tract, distal bile duct

2C15.Z Malignant neoplasms of biliary tract, distal bile duct, unspecified

2C16 Malignant neoplasms of ampulla of Vater

2C16.0 Adenocarcinoma of ampulla of Vater

A malignant glandular epithelial tumour arising in the ampulla of Vater

2C16.1 Neuroendocrine neoplasms of ampulla of Vater

2C16.Y Other specified malignant neoplasms of ampulla of Vater

2C16.Z Malignant neoplasms of ampulla of Vater, unspecified

2C17 Malignant neoplasms of other or unspecified parts of biliary tract

Exclusions: Malignant neoplasm of intrahepatic bile duct (2C12)

Coded Elsewhere: Malignant mesenchymal tumours of gallbladder or bile ducts (2B5F.2)

2C17.0 Adenocarcinoma of other or unspecified parts of biliary tract

2C17.1 Mucinous cystic neoplasm with associated invasive carcinoma of other or unspecified parts of biliary tract

2C17.2 Neuroendocrine neoplasms of other or unspecified parts of biliary tract

2C17.Y Other specified malignant neoplasms of overlapping lesion of biliary tract

2C17.Z Malignant neoplasms of other or unspecified parts of biliary tract, unspecified

2C18 Malignant neoplasms of perihilar bile duct

“Includes left, right and common hepatic ducts to the origin of the cystic duct”

2C18.0 Hilar cholangiocarcinoma

Klatskin tumour is an extra-hepatic cholangiocarcinoma arising in the junction of the main right or left hepatic ducts to form the common hepatic duct.

2C18.1 Mucinous cystic neoplasm with associated invasive carcinoma of perihilar bile duct

2C18.2 Neuroendocrine neoplasm of perihilar bile duct

2C18.Y Other specified malignant neoplasms of perihilar bile duct

2C18.Z Malignant neoplasms of perihilar bile duct, unspecified

2C1Z Malignant neoplasms of digestive organs, unspecified

Malignant neoplasms of middle ear, respiratory or intrathoracic organs (BlockL3‑2C2)

Exclusions: Mesotheliomas of peritoneum (2C51.2)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C20 Malignant neoplasms of nasal cavity

From the chapter on Nasal Cavity (H&N BB) Although the nasal cavity and paranasal sinuses occupy a relatively small anatomical space, they are the site of origin of some of the more complex, histologically diverse group of tumours in the entire human body. These include neoplasms derived from mucosal epithelium, seromucinous glands, soft tissues, bone, cartilage, neural/neuroectodermal tissue, haematolymphoid cells and the odontogenic apparatus. Many of the tumours are similar to those found elsewhere in the body but a few, such as the olfactory neuroblastoma, are unique to this site.

Exclusions: Malignant neoplasm of nose NOS (2C29)

Malignant neoplasm of olfactory bulb (2A02)

Malignant neoplasm of posterior margin of nasal septum and choana (2B6B)

Malignant neoplasm of skin of nose (BlockL3‑2C3)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C20.0 Adenocarcinoma of nasal cavity

2C20.1 Malignant neuroepitheliomatous neoplasm of nasal cavity

2C20.2 Melanoma of nasal cavity

2C20.3 Olfactory neuroblastoma

2C20.4 Squamous cell carcinoma of nasal cavity

2C20.Y Other specified malignant neoplasm of nasal cavity

2C20.Z Malignant neoplasms of nasal cavity, unspecified

2C21 Malignant neoplasms of middle ear

Malignant neoplasm originating in the middle ear.

Exclusions: malignant neoplasm of skin of (external) ear (BlockL3‑2C3)

malignant neoplasm of bone of ear (meatus) (BlockL3‑2B5)

malignant neoplasm of cartilage of ear (BlockL3‑2B5)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C21.0 Adenocarcinoma of middle ear

2C21.1 Squamous cell carcinoma of middle ear

2C21.2 Unspecified malignant epithelial neoplasm of middle ear

2C21.Y Other specified malignant neoplasm of middle ear

2C21.Z Malignant neoplasms of middle ear, unspecified

2C22 Malignant neoplasms of accessory sinuses

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C22.0 Adenocarcinoma of accessory sinuses

2C22.1 Squamous cell carcinoma of accessory sinuses

2C22.10 Squamous cell carcinoma of sphenoidal sinus

2C22.1Y Squamous cell carcinoma of other specified accessory sinuses

2C22.2 Malignant epithelial neoplasms of accessory sinuses, unspecified type

2C22.3 Melanomas of accessory sinuses

2C22.Y Other specified malignant neoplasms of accessory sinuses

2C22.Z Malignant neoplasms of accessory sinuses, unspecified

2C23 Malignant neoplasms of larynx

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C23.1 Malignant neoplasms of glottis of larynx

2C23.10 Squamous cell carcinoma of larynx, glottis

A squamous cell carcinoma of the larynx that arises from the glottic area. It may remain localised for a long period then in late disease stage, it may spread to the opposite true vocal cord, supraglottic and subglottic areas, and the soft tissues of the neck. Hoarseness is the presenting symptom.

2C23.1Y Other specified malignant neoplasms of larynx, glottis

2C23.2 Malignant neoplasms of supraglottis of larynx

Exclusions: Malignant neoplasm of anterior surface of epiglottis (2B6A)

2C23.20 Squamous cell carcinoma of larynx, supraglottis

A squamous cell carcinoma of the larynx that arises from the supraglottic area. Signs and symptoms include dysphagia, a sensation of foreign body in the throat, and hemoptysis. It spreads to the space anterior to the epiglottis, pyriform sinus, and base of the tongue.

2C23.2Y Other specified malignant neoplasms of larynx, supraglottis

2C23.3 Malignant neoplasms of subglottis of larynx

A primary or metastatic malignant neoplasm involving the subglottis.

2C23.30 Squamous cell carcinoma of larynx, subglottis

A squamous cell carcinoma of the larynx that arises from the subglottic area. Symptoms include dyspnea and stridor. It spreads to the hypopharynx, trachea, and thyroid gland.

2C23.31 Adenocarcinoma of larynx, subglottis

2C23.3Y Other specified malignant neoplasms of larynx, subglottis

2C23.4 Malignant neoplasm of laryngeal cartilage

2C23.5 Malignant neoplasm of overlapping lesion of larynx

2C23.Z Malignant neoplasms of larynx, unspecified

2C24 Malignant neoplasms of trachea

A primary or metastatic malignant neoplasm involving the trachea

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

trachea carina cancer (2C25)

2C24.0 Adenocarcinoma of trachea

2C24.1 Squamous cell carcinoma of trachea

2C24.2 Malignant epithelial neoplasms of trachea, unspecified type

2C24.Y Other specified malignant neoplasms of trachea

2C24.Z Malignant neoplasms of trachea, unspecified

2C25 Malignant neoplasms of bronchus or lung

A primary or metastatic malignant neoplasm involving the lung.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C25.0 Adenocarcinoma of bronchus or lung

A carcinoma that arises from the lung and is characterised by the presence of malignant glandular epithelial cells. There is a male predilection with a male to female ratio of 2:1. Usually lung adenocarcinoma is asymptomatic and is identified through screening studies or as an incidental radiologic finding. If clinical symptoms are present they include shortness of breath, cough, hemoptysis, chest pain, and fever. Tobacco smoke is a known risk factor.

2C25.1 Small cell carcinoma of bronchus or lung

A highly aggressive subtype of lung carcinoma characterised by the presence of malignant small cells and necrosis. Metastatic disease is usually present at the time of diagnosis.

2C25.2 Squamous cell carcinoma of bronchus or lung

A carcinoma arising from malignant squamous bronchial epithelial cells and characterised by the presence of keratinization and/or intercellular bridges. Cigarette smoking and arsenic exposure are strongly associated with squamous cell lung carcinoma.

2C25.3 Large cell carcinoma of bronchus or lung

2C25.4 Carcinoid or other malignant neuroendocrine neoplasms of bronchus or lung

2C25.5 Unspecified malignant epithelial neoplasm of bronchus or lung

2C25.Y Other specified malignant neoplasms of bronchus or lung

2C25.Z Malignant neoplasms of bronchus or lung, unspecified

2C26 Malignant neoplasms of the pleura

A primary or metastatic malignant neoplasm affecting the pleura. A representative example of primary malignant pleural neoplasm is the malignant pleural mesothelioma. A representative example of metastatic malignant neoplasm to the pleura is when a metastatic carcinoma has spread to the pleura from another anatomic site.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C26.0 Mesothelioma of pleura

Malignant mesothelioma is a fatal asbestos-associated malignancy arising in the lining cells (mesothelium) of the pleural and peritoneal cavities, as well as in the pericardium and the tunica vaginalis.

2C26.Y Other specified malignant neoplasms of the pleura

2C26.Z Malignant neoplasms of the pleura, unspecified

2C27 Malignant neoplasms of thymus

Primary or metastatic malignant neoplasm involving the thymus. This category includes malignant thymomas, thymic lymphomas, primary thymic carcinomas, and metastatic carcinomas from other anatomic sites.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C27.0 Carcinoma of thymus

A diverse group of carcinomas of the thymus gland, previously known as thymoma type C. It includes morphologic variants derived from purely epithelial cells, as well as from cells with neuroendocrine differentiation.

2C27.1 Carcinoid tumour or other neuroendocrine neoplasms of thymus

Thymic neuroendocrine carcinoma is a type of thymic epithelial neoplasm displaying evidence of neuroendocrine differentiation.

2C27.2 Malignant thymoma

A thymoma that has an aggressive clinical course (capsular invasion, infiltration of the surrounding tissues) and can metastasize. Although any morphologic subtype of thymoma may eventually have a malignant clinical course, this term is most often associated with thymoma types B3 and C.

2C27.Y Other specified malignant neoplasms of thymus

2C27.Z Malignant neoplasms of thymus, unspecified

2C28 Malignant neoplasms of heart, mediastinum or non-mesothelioma of pleura

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C28.0 Malignant germ cell neoplasms of heart, mediastinum or non-mesothelioma of pleura

2C28.1 Other specified malignant neoplasms of heart, mediastinum or non-mesothelioma of pleura

2C28.Z Malignant neoplasms of heart, mediastinum or non-mesothelioma of pleura, unspecified

2C29 Malignant neoplasms of other or ill-defined sites in the respiratory system or intrathoracic organs

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C29.0 Squamous cell carcinomas of other and ill-defined sites in the respiratory system and intrathoracic organs

2C29.1 Other specified malignant neoplasms of other or ill-defined sites in the respiratory system or intrathoracic organs

2C29.Z Malignant neoplasms of other or ill-defined sites in the respiratory system or intrathoracic organs, unspecified

2C2Y Other specified malignant neoplasms of middle ear, respiratory or intrathoracic organs

2C2Z Malignant neoplasms of middle ear, respiratory or intrathoracic organs, unspecified

Malignant neoplasms of skin (BlockL3‑2C3)

A primary or metastatic tumour involving the skin. Primary malignant skin tumours most often are carcinomas (either basal cell or squamous cell carcinomas that arise from cells in the epidermis) or melanomas that arise from pigment-containing skin melanocytes. Metastatic tumours to the skin include carcinomas and lymphomas.

Exclusions: Carcinoma in situ of skin (2E64)

Metastatic malignant neoplasm involving skin (2E08)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C30 Melanoma of skin

A primary melanoma arising from atypical melanocytes in the skin. Precursor lesions include acquired and congenital melanocytic nevi, and dysplastic nevi. Several histologic variants have been recognised, including superficial spreading melanoma, acral lentiginous melanoma, nodular melanoma, and lentigo maligna melanoma.

Exclusions: Melanoma of penis (2C81.1)

Melanoma of vulva (2C70.1)

2C30.0 Superficial spreading melanoma, primary

The commonest form of melanoma, superficial spreading malignant melanoma accounts for about 70 per cent of all melanomas. It characteristically presents as an asymptomatic pigmented cutaneous macule which is asymmetrical in shape and displays variability in both hue and pigment intensity. It has a relatively long phase of progressive superficial extension (radial growth) before penetrating deeper into the dermis and entering an invasive vertical growth phase.

2C30.1 Nodular melanoma, primary

Variant of melanoma carrying a poor prognosis due to the fact that there is little or no prodromal radial (superficial) growth phase before deep invasion (vertical growth). The lesion presents as an elevated, reddish, bluish or dark brown coloured, dome- shaped tumour. Ulceration or bleeding from the lesion occurs frequently. It occurs most frequently in the fifth or sixth decade.

2C30.2 Lentigo maligna melanoma, primary

Lentigo maligna malignant melanoma is a form of melanoma which occurs within a lentigo maligna when neoplastic cells no longer remain confined to the epidermis (in situ radial growth) but invade the dermis (vertical growth). The lentigo maligna from which it arises may have been present as an irregularly pigmented macule on sun-exposed skin for many years before dermal invasion supervenes. Clinical differentiation from lentigo maligna may not be possible in the early stages of invasion but as the tumour progresses a focal thickening or nodule within the lentigo maligna will become apparent.

Exclusions: Lentigo maligna (2E63.00)

2C30.3 Acral lentiginous melanoma, primary

Acral lentiginous malignant melanoma is a distinct and uncommon form of melanoma affecting either palmar and plantar skin or the nail apparatus. It is usually preceded by a slowly progressive in situ phase which may be overlooked. It typically presents either as an enlarging area of macular pigmentation on the palms and soles or as a longitudinal pigmented band within the nail plate. More aggressive tumours present as ulcerated nodules which, when involving the nail apparatus, can destroy the nail plate. Acral lentiginous malignant melanoma accounts for a high proportion of melanomas seen in dark-skinned people.

2C30.Y Other specified melanoma of skin

2C30.Z Melanoma of skin, unspecified

2C31 Squamous cell carcinoma of skin

A carcinoma arising from the squamous cells of the epidermis. Skin squamous cell carcinoma is most commonly found on sun-exposed areas. The majority of the tumours are well-differentiated.

Coded Elsewhere: Squamous cell carcinoma of penis (2C81.0)

Squamous cell carcinoma of vulva (2C70.2)

2C31.0 Verrucous squamous cell carcinoma of skin

Verrucous squamous cell carcinoma is a rare variant of well-differentiated squamous cell carcinoma with low malignant potential. It occurs principally on the glans and prepuce of the penis and on the sole of the foot.

2C31.Z Cutaneous squamous cell carcinoma

2C32 Basal cell carcinoma of skin

Basal cell carcinoma or BCC is the most common malignancy in humans. It is believed that BCCs arise from pluripotential cells in the basal layer of the epidermis or, less commonly, hair follicle. BCCs typically occur in areas of chronic sun exposure and presents as slowly enlarging reddish papules, plaques or nodules on the head and neck, although the superficial variant is often located on the trunk. BCCs frequently ulcerate and become crusted. Although they rarely metastasize, they can cause significant local destruction and disfigurement if neglected or inadequately treated, particularly if of the sclerosing or infiltrative subtype.

2C32.0 Nodular basal cell carcinoma of skin

This is the most common form of basal cell carcinoma and is typically located on the head or neck. It starts as a small translucent nodule which will frequently necrose and ulcerate as it enlarges. Telangiectatic blood vessels can often be detected just under the tumour surface. A minority may be pigmented and give rise to difficulty in distinguishing them from melanoma.

2C32.1 Sclerosing basal cell carcinoma of skin

This form of basal cell carcinoma is composed of thin strands, cords and columns of malignant cells which infiltrate between collagen bundles of the dermis. It may infiltrate widely and deeply before it becomes clinically obvious. It typically starts as a pale, poorly-defined indurated plaque which may not come to medical attention until it starts to bleed and crust.

2C32.2 Superficial basal cell carcinoma of skin

Superficial basal cell carcinomas are often multiple and appear as pink or red barely elevated patches varying in size from a few mm to over 10 cm in diameter. A fine pearly border can usually be seen on careful inspection. They occur most frequently on the trunk.

2C32.Y Other specified basal cell carcinoma of skin

2C32.Z Basal cell carcinoma of skin, unspecified

2C33 Adnexal carcinoma of skin

A carcinoma arising from the sebaceous glands, sweat glands, or the hair follicles. Representative examples include sebaceous carcinoma, apocrine carcinoma, eccrine carcinoma, and pilomatrical carcinoma.

Inclusions: Primary cutaneous mucinous carcinoma

Primary cutaneous adenoid cystic carcinoma

Appendageal carcinoma of skin

2C34 Cutaneous neuroendocrine carcinoma

Cutaneous neuroendocrine carcinoma is a primary cutaneous cancer arising from a subset of skin neuroendocrine cells (Merkel cells, giving the name Merkel cell carcinoma (MCC)).

2C35 Cutaneous sarcoma

A group of generally rare malignant neoplasms arising from mesenchymal elements in the dermis including fibroblasts, pilar smooth muscle and vascular endothelium.

Coded Elsewhere: Angiosarcoma of skin (2B56.1)

Kaposi sarcoma of skin (2B57.1)

Cutaneous leiomyosarcoma (2B58.Y)

Dermatofibrosarcoma protuberans (2B53.Y)

2C3Y Other specified malignant neoplasms of skin

2C3Z Malignant neoplasm of skin of unknown or unspecified type

Malignant neoplasms of peripheral nerves or autonomic nervous system (BlockL3‑2C4)

Exclusions: Malignant nerve sheath tumour of peripheral nerves or autonomic nervous system, primary site (2B5E)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C40 Malignant neuroepitheliomatous neoplasms of peripheral nerves or autonomic nervous system

2C41 Malignant perineurioma

2C4Y Other specified malignant neoplasms of peripheral nerves and autonomic nervous system

2C4Z Malignant neoplasms of peripheral nerves or autonomic nervous system, unspecified

Malignant neoplasms of retroperitoneum, peritoneum or omentum (BlockL3‑2C5)

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C50 Malignant neoplasms of retroperitoneum

A primary or metastatic malignant neoplasm involving the retroperitoneum. The vast majority of cases are carcinomas, lymphomas, or sarcomas.

Exclusions: Malignant neoplasms of omentum (2C52)

Malignant neoplasms of peritoneum (2C51)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C50.0 Cystic, mucinous or serous carcinoma of retroperitoneum

2C50.Y Other specified malignant neoplasms of retroperitoneum

2C50.Z Malignant neoplasms of retroperitoneum, unspecified

2C51 Malignant neoplasms of peritoneum

Exclusions: Malignant neoplasms of retroperitoneum (2C50)

Malignant neoplasms of omentum (2C52)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C51.0 Adenocarcinomas of peritoneum

2C51.1 Cystic, mucinous or serous carcinoma of peritoneum

2C51.2 Mesotheliomas of peritoneum

A benign or malignant mesothelial neoplasm that arises from the peritoneum.

2C51.20 Mesothelioma of mesocolon

2C51.21 Mesothelioma of mesentery

2C51.2Y Other specified mesotheliomas of peritoneum

2C51.2Z Mesotheliomas of peritoneum, unspecified

2C51.Y Other specified malignant neoplasms of peritoneum

2C51.Z Malignant neoplasms of peritoneum, unspecified

2C52 Malignant neoplasms of omentum

Exclusions: Malignant neoplasms of retroperitoneum (2C50)

Malignant neoplasms of peritoneum (2C51)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

Coded Elsewhere: Myosarcomas of omentum (2B5F.10)

2C52.0 Cystic, mucinous or serous carcinoma of omentum

2C52.Y Other specified malignant neoplasms of omentum

2C52.Z Malignant neoplasms of omentum, unspecified

2C53 Malignant neoplasm involving overlapping sites of retroperitoneum, peritoneum or omentum

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C53.0 Adenocarcinoma involving overlapping sites of retroperitoneum, peritoneum or omentum

2C53.1 Mesothelioma involving overlapping sites of retroperitoneum, peritoneum or omentum

2C53.Y Other specified malignant neoplasm involving overlapping sites of retroperitoneum, peritoneum or omentum

2C53.Z Malignant neoplasm involving overlapping sites of retroperitoneum, peritoneum or omentum, unspecified

2C5Y Other specified malignant neoplasms of retroperitoneum, peritoneum or omentum

2C5Z Malignant neoplasms of retroperitoneum, peritoneum or omentum, unspecified

Malignant neoplasms of breast (BlockL3‑2C6)

The category refers to primary malignant neoplasms of parenchyma, connective, and soft tissue of the breast, including nipple and areola.

Inclusions: malignant neoplasm of connective tissue of breast

Exclusions: Malignant neoplasm of skin of breast (BlockL3‑2C3)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C60 Carcinoma of breast, specialised type

2C61 Invasive carcinoma of breast

2C61.0 Invasive ductal carcinoma of breast

2C61.1 Invasive lobular carcinoma of breast

An infiltrating lobular adenocarcinoma. The malignant cells lack cohesion and are arranged individually or in a linear manner (Indian files), or as narrow trabeculae within the stroma. The malignant cells are usually smaller than those of ductal carcinoma, are less pleomorphic, and have fewer mitotic figures.

2C61.2 Invasive pleomorphic lobular carcinoma of breast

A grade II invasive lobular carcinoma of the breast, characterised by the presence of neoplastic cells with large and atypical nuclei.

2C61.3 Invasive carcinoma of breast with mixed ductal and lobular features

An invasive ductal breast carcinoma associated with a lobular carcinomatous component. The lobular carcinomatous component may be in situ or invasive.

2C61.4 Invasive carcinoma of breast, unidentifiable type

A carcinoma that infiltrates the breast parenchyma and where the histopathological type could not be identified.

2C62 Inflammatory carcinoma of breast

An advanced, invasive breast adenocarcinoma characterised by the presence of distinct changes in the overlying skin. These changes include diffuse erythema, edema, peau d'orange (skin of an orange) appearance, tenderness, induration, warmth, enlargement, and in some cases a palpable mass. The skin changes are the consequence of lymphatic obstruction from the underlying invasive breast adenocarcinoma. Microscopically, the dermal lymphatics show prominent infiltration by malignant cells. The invasive breast adenocarcinoma is usually of ductal, NOS type. There is not significant inflammatory cell infiltrate present, despite the name of this carcinoma.

2C63 Malignant phyllodes tumour of breast

A phyllodes tumour of the breast characterised by infiltrative margins and a sarcomatous stromal component. The sarcomatous stroma usually displays features of fibrosarcoma. Liposarcomatous, osteosarcomatous, or rhabdomyosarcomatous elements may also be present.

2C64 Solid papillary carcinoma of breast with evidence of invasion

2C65 Hereditary breast and ovarian cancer syndrome

2C6Y Other specified malignant neoplasms of breast

2C6Z Malignant neoplasms of breast, unspecified

Malignant neoplasms of female genital organs (BlockL3‑2C7)

A primary or metastatic malignant neoplasm involving the female reproductive system. Representative examples include endometrial carcinoma, cervical carcinoma, ovarian carcinoma, uterine corpus leiomyosarcoma, adenosarcoma, malignant mixed mesodermal (mullerian) tumour, and gestational choriocarcinoma.

Coding Note: Includes Malignant neoplasm of skin of female genital organs

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C70 Malignant neoplasms of vulva

Squamous cell carcinoma of the vulva occurs predominantly in the older age group. Although the incidence rate of vulvar intraepithelial neoplasia is increasing, that of squamous cell carcinoma of the vulva is declining, reflecting earlier detection and more successful treatment. In addition to human papillomavirus infection, cigarette smoking is a putative risk factor for vulvar squamous cell carcinoma. There are three known precursor lesions: vulvar intraepithelial neoplasia, lichen sclerosis and chronic granulomatous disease. Other important epithelial malignancies of the vulva are Paget disease and Bartholin gland carcinoma. They are much less

common than squamous lesions, and the risk factors are largely unknown. Prominent non-epithelial tumours are malignant melanoma and sarcoma botyoides.

Coding Note: Includes skin of vulva.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C70.0 Basal cell carcinoma of vulva

A slow growing, locally infiltrating carcinoma that arises from the vulva. It is characterised by the presence of malignant cells that resemble the basal cells that are present in the epidermis.

2C70.1 Melanoma of vulva

2C70.2 Squamous cell carcinoma of vulva

An invasive squamous cell carcinoma arising from the vulva. Risk factors include the human papilloma virus and cigarette smoking. Precursor lesions include the vulvar intraepithelial neoplasia, lichen sclerosus with associated squamous cell hyperplasia, and chronic granulomatous vulvar disease such as granuloma inguinale. Symptoms include vulvar pruritus or irritation, discharge, bleeding, and pain. The following morphologic variants have been identified: keratinizing, non-keratinizing, basaloid, warty, verrucous, keratoacanthoma-like, and squamous cell carcinoma with tumour giant cells. Risk factors for recurrence include advanced stage, tumour diameter greater than 2.5 cm, multifocality, capillary-like space involvement, associated vulvar intraepithelial neoplasia grades 2 or 3, and margins of resection involved by tumour.

Coded Elsewhere: Verrucous squamous cell carcinoma of vulva (2C31.0)

2C70.Y Other specified malignant neoplasms of vulva

Coding Note: Includes skin of vulva.

2C70.Z Malignant neoplasms of vulva, unspecified

Coding Note: Includes skin of vulva.

2C71 Malignant neoplasms of vagina

A primary or metastatic malignant tumour involving the vagina. Representative examples include carcinomas and sarcomas.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C71.0 Adenocarcinoma of vagina

An adenocarcinoma arising from the vagina. Morphologic variants include the clear cell, endometrioid, mesonephric, and mucinous adenocarcinoma.

2C71.1 Melanoma of vagina

A primary malignant neoplasm of the vagina composed of malignant melanocytes.

2C71.2 Squamous cell carcinoma of vagina

A squamous cell carcinoma arising from the vagina. Human papillomavirus infection is associated with the development of vaginal intraepithelial neoplasia and invasive squamous cell carcinoma. Signs and symptoms include painless bleeding, postcoital bleeding, and urinary tract symptoms. Morphologically it resembles squamous cell carcinomas in other anatomic sites.

2C71.Y Other specified malignant neoplasms of vagina

2C71.Z Malignant neoplasms of vagina, unspecified

2C72 Malignant neoplasms of uterine ligament, parametrium, or uterine adnexa

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C72.0 Adenocarcinoma of uterine ligament, parametrium, or uterine adnexa

2C72.1 Mucinous or serous carcinoma of uterine ligament, parametrium, or uterine adnexa

2C72.2 Malignant neoplasm involving overlapping sites of female genital organs

Inclusions: Malignant neoplasm of female genital organs whose point of origin cannot be classified to any other existing entity

2C72.3 Carcinosarcomas of uterine ligament, parametrium, or uterine adnexa

2C72.Y Other specified malignant neoplasms of uterine ligament, parametrium, and uterine adnexa

2C72.Z Malignant neoplasms of uterine ligament, parametrium, or uterine adnexa, unspecified

2C73 Malignant neoplasms of ovary

A primary or metastatic malignant neoplasm involving the ovary. Most primary malignant ovarian neoplasms are either carcinomas (serous, mucinous, or endometrioid adenocarcinomas) or malignant germ cell tumours. Metastatic malignant neoplasms to the ovary include carcinomas, lymphomas, and melanomas.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

Coded Elsewhere: Hereditary breast and ovarian cancer syndrome (2C65)

2C73.0 Carcinomas of ovary

2C73.00 Clear cell adenocarcinoma of ovary

A malignant glandular epithelial tumour characterised by the presence of clear and hobnail cells. The tumour is highly associated with ovarian endometriosis, pelvic endometriosis and paraendocrine hypercalcemia.

2C73.01 Endometrioid adenocarcinoma of ovary

An endometrioid adenocarcinoma arising from the ovary. It comprises 10% to 25% of all primary ovarian carcinomas. Grossly, endometrioid carcinoma may present as a cystic or solid mass. Microscopically, the tumour greatly resembles the appearance of the ordinary type of endometrial adenocarcinoma. As a group, endometrioid carcinoma has a prognosis twice as good as that of serous or mucinous carcinoma.

2C73.02 Low grade serous adenocarcinoma of ovary

A slow-growing serous adenocarcinoma that arises from the ovary. It usually originates from borderline neoplastic processes or adenofibromas. It is characterised by the presence of low grade cytologic features and infrequent mitotic figures.

2C73.03 High grade serous adenocarcinoma of ovary

2C73.04 Mucinous adenocarcinoma of ovary

An invasive adenocarcinoma that arises from the ovary and is characterised by the presence of malignant epithelial cells that contain intracytoplasmic mucin. There is cellular atypia, increased layering of cells, complexity of glands, and papillary formations.

2C73.0Y Other specified carcinomas of ovary

2C73.0Z Carcinomas of ovary, unspecified

2C73.1 Dysgerminoma of ovary

A malignant germ cell tumour arising from the ovary. Morphologically, it is identical to seminoma and consists of a monotonous population of germ cells with abundant pale cytoplasm and uniform nuclei. The stroma invariably contains chronic inflammatory cells, mostly T-lymphocytes. It responds to chemotherapy or radiotherapy and the prognosis relates to the tumour stage.

Inclusions: Malignant dysgerminomatous germ cell tumour of ovary

2C73.2 Granulosa cell malignant tumour of ovary

An aggressive granulosa cell tumour that arises from the ovary and metastasizes to other anatomic sites.

2C73.3 Malignant teratoma of ovary

A malignant germ cell tumour arising from the ovary. It usually affects females in their first two decades of life. It contains variable amounts of immature embryonal tissues. Based on the amount of immature neuroepithelial component, immature teratomas are graded from 1 to 3. The stage and grade of the tumour and the grade of the metastatic tumour are the important factors that predict prognosis. The use of cisplatin-based combination chemotherapy has significantly improved the survival rates of the patients.

Coded Elsewhere: Struma ovarii (5A02.Y)

2C73.4 Serous cystadenoma, borderline malignancy of ovary

2C73.5 Endodermal sinus tumour, unspecified site, female

2C73.Y Other specified malignant neoplasms of the ovary

2C73.Z Malignant neoplasms of ovary, unspecified

2C74 Malignant neoplasms of fallopian tube

Tumours of the fallopian tube are much less common than the corresponding ovarian neoplasms; however, histologically the same surface epithelial-stromal tumour subtypes are recognised. Sex cord-stromal and germ cell tumours are rare. Hydatidiform moles and gestational choriocarcinoma are uncommon complications of tubal ectopic pregnancy. The wolffian adnexal tumour is also infrequent and typically occurs in the leaves of the broad ligament. The risk factors appear similar to those of the ovary. Fallopian tube carcinomas are a component of the hereditary breast-ovarian cancer syndrome caused by BRCA1 and BRCA2 germline mutations.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C74.0 Adenocarcinoma of fallopian tube

An adenocarcinoma that arises from the fallopian tube. Histologic subtypes include clear cell, endometrioid, serous, and mucinous adenocarcinoma. It spreads to adjacent organs, regional lymph nodes, and peritoneum.

2C74.Y Other specified malignant neoplasms of fallopian tube

2C74.Z Malignant neoplasms of fallopian tube, unspecified

2C75 Malignant neoplasms of placenta

Exclusions: hydatidiform mole, NOS (JA02)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C75.0 Malignant trophoblastic neoplasms of placenta

A diverse group of pregnancy-related tumours characterised by excessive proliferation of trophoblasts. Representative examples include hydatidiform mole, gestational choriocarcinoma, and placental site trophoblastic tumour.

2C75.Y Other specified malignant neoplasms of placenta

2C75.Z Malignant neoplasms of placenta, unspecified

2C76 Malignant neoplasms of corpus uteri

A malignant neoplasm that affects the uterine corpus. Representative examples include endometrial carcinoma, carcinosarcoma, leiomyosarcoma, and adenosarcoma.

Exclusions: Endometrial stromal sarcoma, primary site (2B5C)

2C76.0 Endometrial endometrioid adenocarcinoma

2C76.1 Endometrial mucinous adenocarcinoma

2C76.2 Endometrial clear cell adenocarcinoma

2C76.3 Endometrial serous adenocarcinoma

2C76.4 Endometrial mixed adenocarcinoma

2C76.40 Endometrial squamous cell carcinoma

2C76.41 Endometrial small cell carcinoma

2C76.42 Endometrial undifferentiated carcinoma

2C76.43 Carcinosarcoma of uterus

2C76.Y Other specified malignant neoplasms of corpus uteri

2C76.Z Malignant neoplasms of corpus uteri, unspecified

2C77 Malignant neoplasms of cervix uteri

Primary or metastatic malignant neoplasm involving the cervix.

2C77.0 Squamous cell carcinoma of cervix uteri

A squamous cell carcinoma arising from the cervical epithelium. It usually evolves from a precancerous cervical lesion. Increased numbers of sexual partners and human papillomavirus (HPV) infection are risk factors for cervical squamous cell carcinoma. The following histologic patterns have been described: Conventional squamous cell carcinoma, papillary squamous cell carcinoma, transitional cell carcinoma, lymphoepithelioma-like carcinoma, verrucous carcinoma, condylomatous carcinoma and spindle cell carcinoma. Survival is most closely related to the stage of disease at the time of diagnosis.

2C77.1 Adenocarcinoma of cervix uteri

An adenocarcinoma arising from the cervical epithelium. It accounts for approximately 15% of invasive cervical carcinomas. Increased numbers of sexual partners and human papillomavirus (HPV) infection are risk factors. Grossly, advanced cervical adenocarcinoma may present as an exophytic mass, an ulcerated lesion, or diffuse cervical enlargement. Microscopically, the majority of cervical adenocarcinomas are of the endocervical (mucinous) type.

2C77.2 Adenosquamous carcinoma of cervix uteri

2C77.3 Neuroendocrine carcinoma of cervix uteri

2C77.Y Other specified malignant neoplasms of cervix uteri

2C77.Z Malignant neoplasms of cervix uteri, unspecified

2C78 Malignant neoplasms of uterus, part not specified

2C7Y Other specified malignant neoplasms of female genital organs

Coding Note: Includes Malignant neoplasm of skin of female genital organs

2C7Z Malignant neoplasms of female genital organs, unspecified

Coding Note: Includes Malignant neoplasm of skin of female genital organs

Malignant neoplasms of male genital organs (BlockL3‑2C8)

A primary or metastatic malignant neoplasm involving the male reproductive system. Representative examples include prostate carcinoma, penile carcinoma, testicular seminoma, and testicular embryonal carcinoma.

Coding Note: Includes malignant neoplasm of skin of male genital organs

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C80 Malignant neoplasms of testis

A primary or metastatic malignant neoplasm that affects the testis. Representative examples include seminoma, embryonal carcinoma, sarcoma, leukaemia, and lymphoma.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C80.0 Choriocarcinoma of testis

A malignant germ cell tumour arising from the testis. It represents the rarest of the testicular germ cell tumours. Histologically it is characterised by the presence of syncytiotrophoblasts and cytotrophoblasts.

Inclusions: Malignant trophoblastic tumour of testis

2C80.1 Embryonal carcinoma of testis

A malignant germ cell neoplasm arising from the testis. It is composed of primitive epithelial cells arranged in solid, papillary, and glandular configurations. Most patients present with a testicular mass, which may be associated with pain. More than half of the patients have metastatic disease at diagnosis.

2C80.2 Germ cell tumour of testis

A germ cell tumour arising from the testis. Representative examples include teratoma, seminoma, embryonal carcinoma, and yolk sac tumour.

2C80.3 Intratubular germ cell neoplasia, unclassified

A non-invasive lesion of the testis, characterised by the presence of malignant large germ cells with abundant cytoplasm in the seminiferous tubules. It may be associated with undescended or atrophic testis and infertility. The vast majority of cases progress to invasive germ cell tumours.

2C80.4 Malignant teratoma of testis

A teratoma that arises from the testis and is composed of immature, fetal type-tissues.

Inclusions: Teratocarcinoma of testis

2C80.5 Mixed seminoma and non-seminomatous germ cell tumour of testis

2C80.6 Non-seminomatous mixed germ cell tumour of testis

2C80.7 Seminoma pure form of testis

A radiosensitive malignant germ cell tumour found in the testis (especially undescended), and extragonadal sites (anterior mediastinum and pineal gland). It is characterised by the presence of uniform cells with clear or dense cytoplasm which contains glycogen, and by a large nucleus which contains one or more nucleoli. The neoplastic germ cells form aggregates separated by fibrous septa. The fibrous septa contain chronic inflammatory cells, mainly lymphocytes.

2C80.Y Other specified malignant neoplasms of testis

2C80.Z Malignant neoplasms of testis, unspecified

2C81 Malignant neoplasms of penis

A primary or metastatic malignant neoplasm that affects the penis. Representative examples include penile carcinoma and penile sarcoma.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

Coded Elsewhere: Verrucous squamous cell carcinoma of penis (2C31.0)

2C81.0 Squamous cell carcinoma of penis

A squamous cell carcinoma arising from the penis. It occurs chiefly in the squamous epithelium of the glans, coronal sulcus, and foreskin. Etiologic factors include phimosis, lichen sclerosus, smoking, ultraviolet irradiation, history of warts or condylomas, and lack of circumcision. Human papilloma virus is present in a subset of penile squamous cell carcinomas. Patients may present with an exophytic or flat ulcerative mass in the glans or a large primary tumour with inguinal nodal and skin metastases. Morphologic variants include the basaloid carcinoma, warty (condylomatous) carcinoma, verrucous carcinoma, and sarcomatoid (spindle cell) carcinoma.

Coded Elsewhere: Verrucous squamous cell carcinoma of penis (2C31.0)

2C81.1 Melanoma of penis

Inclusions: Melanoma of skin of penis

Melanoma of mucocutaneous epithelium of penis

2C81.Y Other specified malignant neoplasm of penis

2C81.Z Malignant neoplasms of penis, unspecified

2C82 Malignant neoplasms of prostate

Prostate cancer contributes significantly to the overall cancer burden, being the most frequent malignant neoplasia in men. The number of cases has continuously increased over the past decades, partly due to the higher life expectancy. An additional factor is the Western lifestyle, characterised by a highly caloric diet and lack of physical exercise. Epidemiological data indicates that black people are most susceptible, followed by white people, while Asian people have the lowest risk. The extent to which prostate cancer mortality can be reduced by PSA screening, is currently being evaluated. Histopathological diagnosis and grading play a major role in the management of prostate cancer

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C82.0 Adenocarcinoma of prostate

An adenocarcinoma arising from the prostate gland. It is one of the most common malignant tumours afflicting men. The majority of adenocarcinomas arise in the peripheral zone and a minority occurs in the central or the transitional zone of the prostate gland. Grading of prostatic adenocarcinoma predicts disease progression and correlates with survival. Several grading systems have been proposed, of which the Gleason system is the most commonly used. Gleason sums of 2 to 4 represent well-differentiated disease, 5 to 7 moderately differentiated disease and 8 to 10 poorly differentiated disease. Prostatic-specific antigen (PSA) serum test is widely used as a screening test for the early detection of prostatic adenocarcinoma.

2C82.Y Other specified malignant neoplasms of prostate

2C82.Z Malignant neoplasms of prostate, unspecified

2C83 Malignant neoplasms of scrotum

Inclusions: malignant neoplasm of skin of scrotum

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C83.0 Squamous cell carcinoma of scrotum

2C83.Y Other specified malignant neoplasms of scrotum

2C83.Z Malignant neoplasms of scrotum, unspecified

2C84 Malignant neoplasms of other specified male genital organs

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C8Z Malignant neoplasms of male genital organs, unspecified

Coding Note: Includes malignant neoplasm of skin of male genital organs

Malignant neoplasms of urinary tract (BlockL3‑2C9)

A primary or metastatic malignant tumour involving the urinary system. Common tumour types include carcinomas, lymphomas, and sarcomas.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C90 Malignant neoplasms of kidney, except renal pelvis

Cancer of the kidney amounts to 2% of the total human cancer burden, with approximately 190,000 new cases diagnosed each year. They occur in all world regions, with a preference for developed countries. Etiological factors include environmental carcinogens (tobacco smoking) and lifestyle factors, in particular obesity. Although renal tumours can be completely removed surgically, haematogeneous metastasis is frequent and may occur already at an early stage of the disease. The pattern of somatic mutations in kidney tumours has been extensively investigated and has become, in addition to histopathology, a major criterion for classification. Kidney tumours also occur in the setting of several inherited cancer syndromes, including von Hippel-Lindau disease.

Exclusions: Malignant neoplasm of renal calyces (2C91)

Malignant neoplasms of renal pelvis (2C91)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C90.0 Renal cell carcinoma of kidney, except renal pelvis

A carcinoma arising from the renal parenchyma. The incidence of renal cell carcinoma has increased by 35% from 1973 to 1991. There is a strong correlation between cigarette smoking and the development of renal cell carcinoma. The clinical presentation includes : haematuria, flank pain and a palpable lumbar mass. A high percentage of renal cell carcinomas are diagnosed when an ultrasound is performed for other purposes. Diagnostic procedures include: ultra sound, intravenous pyelography and computed tomography (CT).

2C90.Y Other specified malignant neoplasms of kidney, except renal pelvis

2C90.Z Malignant neoplasms of kidney, except renal pelvis, unspecified

2C91 Malignant neoplasms of renal pelvis

Abnormal malignant growth of the cells within the renal pelvis.

Inclusions: malignant neoplasm of pelviureteric junction

malignant neoplasm of renal calyces

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C91.0 Urothelial carcinoma of renal pelvis

2C91.Y Other specified malignant neoplasms of renal pelvis

2C91.Z Malignant neoplasms of renal pelvis, unspecified

2C92 Malignant neoplasms of ureter

A primary or metastatic malignant tumour involving the ureter. The majority are carcinomas.

Exclusions: malignant neoplasm of ureteric orifice of bladder (2C94)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C92.0 Urothelial carcinoma of ureter

2C92.Y Other specified malignant neoplasms of ureter

2C92.Z Malignant neoplasms of ureter, unspecified

2C93 Malignant neoplasms of urethra or paraurethral gland

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C93.0 Adenocarcinoma of urethra or paraurethral gland

2C93.1 Squamous cell carcinoma of urethra or paraurethral gland

2C93.2 Urothelial carcinoma of urethra or paraurethral gland

2C93.Y Other specified malignant neoplasms of urethra and paraurethral gland

2C93.Z Malignant neoplasms of urethra or paraurethral gland, unspecified

2C94 Malignant neoplasms of bladder

A primary or metastatic malignant neoplasm involving the bladder.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C94.0 Adenocarcinoma of bladder

A rare adenocarcinoma arising from metaplastic bladder epithelium. It is frequently associated with long-standing local irritation. The majority of cases originate from the trigone and the posterior wall of the bladder.

2C94.1 Squamous cell carcinoma of bladder

A squamous cell carcinoma of the bladder arising from metaplastic epithelium. It represents less than 10% of bladder carcinomas. The exception is the Middle East along the Nile Valley, where it represents the most common form of carcinoma because of the endemic nature of schistosomiasis. Bladder squamous cell carcinoma is often associated with long-standing chronic inflammation of the bladder and usually has a poor prognosis. The diagnosis of squamous cell carcinoma of the bladder should be reserved for those tumours that are predominantly keratin forming.

2C94.2 Urothelial carcinoma of bladder

2C94.Y Other specified malignant neoplasms of bladder

2C94.Z Malignant neoplasms of bladder, unspecified

2C95 Malignant neoplasm involving overlapping sites of urinary organs

Malignant neoplasm of urinary organs whose point of origin cannot be classified to any other existing category

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2C95.0 Adenocarcinoma involving overlapping sites of urinary organs

2C95.1 Squamous cell carcinomas involving overlapping sites of urinary organs

2C95.2 Urothelial carcinoma involving overlapping sites of urinary organs

2C95.Y Other specified malignant neoplasms of overlapping lesion of urinary organs

2C95.Z Malignant neoplasm involving overlapping sites of urinary organs, unspecified

2C9Y Other specified malignant neoplasms of urinary tract

2C9Z Malignant neoplasms of urinary tract, unspecified

Malignant neoplasms of eye or ocular adnexa (BlockL3‑2D0)

A malignant neoplasm affecting the structures of the eye.

Exclusions: Malignant neoplasm of optic nerve (2A02)

Malignant neoplasm of skin of eyelid (BlockL3‑2C3)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2D00 Malignant neoplasm of conjunctiva

A malignant growth of cells within the conjunctiva of the eye.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2D00.0 Melanoma of conjunctiva

A malignant melanoma within the conjunctiva of the eye.

2D00.1 Malignant neoplasm of caruncle

This is a broad group of diseases involving unregulated cell growth of a small, red portion of the corner of the eye that contains modified sebaceous and sweat glands.

2D00.2 Squamous cell carcinoma of conjunctiva

2D00.Y Other specified malignant neoplasm of conjunctiva

2D00.Z Malignant neoplasm of conjunctiva, unspecified

2D01 Malignant neoplasm of cornea

A malignant growth of cells within the cornea of the eye.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2D01.0 Melanoma of cornea

A melanoma within the cornea of the eye.

2D01.1 Squamous cell carcinoma of cornea

2D01.Y Other specified malignant neoplasms of cornea

2D01.Z Malignant neoplasm of cornea, unspecified

2D02 Malignant neoplasm of retina

Abnormal growth of cells comprising the retina with malignant characteristics.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2D02.0 Adenocarcinoma of retinal pigment epithelium

This is a cancer of an epithelium that originates in glandular tissue. Epithelial tissue includes, but is not limited to, the surface layer of skin, glands, and a variety of other tissue that lines the cavities and organs of the body. This diagnosis is with the pigmented cell layer just outside the neurosensory retina that nourishes retinal visual cells, and is firmly attached to the underlying choroid and overlying retinal visual cells.

2D02.1 Malignant neuroepithelial tumours of retina

2D02.2 Retinoblastoma

Retinoblastoma is the most common intraocular malignancy in children. It is a life threatening condition but is potentially curable. It can be hereditary or non-hereditary, unilateral or bilateral (unilateral retinoblastoma, bilateral retinoblastoma, see these terms).

2D02.Y Other specified malignant neoplasm of retina

2D02.Z Malignant neoplasm of retina, unspecified

2D03 Malignant neoplasm of lacrimal apparatus

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2D03.0 Adenocarcinoma of the lacrimal apparatus

2D03.1 Mucoepidermoid carcinoma of lacrimal apparatus

2D03.2 Squamous cell carcinoma of the lacrimal apparatus

2D03.Y Other specified malignant neoplasm of lacrimal apparatus

2D03.Z Malignant neoplasm of lacrimal apparatus, unspecified

2D04 Malignant neoplasm of orbit

A primary or metastatic malignant neoplasm involving the orbit.

Exclusions: Benign neoplasm of orbital bone (2E83.0)

malignant neoplasm of orbital bone (BlockL3‑2B5)

2D05 Malignant neoplasm of choroid

2D06 Malignant neoplasm of ciliary body

Inclusions: Malignant neoplasm of eyeball

2D06.0 Adenocarcinoma of ciliary epithelium

2D06.1 Malignant medulloepithelioma of ciliary body

2D06.2 Other specified malignant neoplasm of ciliary body

2D06.3 Malignant neuroepithelial tumours of ciliary body

2D06.Y Other specified malignant neoplasm of ciliary body

2D06.Z Malignant neoplasm of ciliary body, unspecified

2D07 Malignant neoplasm of iris

2D07.0 Adenocarcinoma of iris epithelium

2D07.1 Malignant neuroepithelial tumours of iris

2D07.Y Other specified malignant neoplasm of iris

2D07.Z Malignant neoplasm of iris, unspecified

2D0Y Other specified malignant neoplasms of eye and ocular adnexa

2D0Z Malignant neoplasms of eye or ocular adnexa, unspecified

Malignant neoplasms of endocrine glands (BlockL3‑2D1)

A malignant neoplasm affecting the endocrine glands. Representative examples include thyroid gland carcinoma, parathyroid gland carcinoma, pituitary gland carcinoma, and adrenal cortex carcinoma.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2D10 Malignant neoplasms of thyroid gland

A primary or metastatic malignant neoplasm affecting the thyroid gland.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

Coded Elsewhere: Thyroid lymphoma (2B33.5)

2D10.0 Follicular carcinoma of thyroid gland

A differentiated adenocarcinoma arising from the follicular cells of the thyroid gland. The nuclear features which characterise the thyroid gland papillary carcinoma are absent. Radiation exposure is a risk factor and it comprises approximately 10% to 15% of thyroid cancers. Clinically, it usually presents as a solitary mass in the thyroid gland. It is generally unifocal and thickly encapsulated and shows invasion of the capsule or the vessels. Diagnostic procedures include: thyroid ultrasound and fine needle biopsy.

2D10.1 Papillary carcinoma of thyroid gland

A differentiated adenocarcinoma arising from the follicular cells of the thyroid gland. Radiation exposure is a risk factor and it is the most common malignant thyroid lesion, comprising 75% to 80% of all thyroid cancers in iodine sufficient countries. Diagnostic procedures include: thyroid ultrasound, and fine needle biopsy. Microscopically, the diagnosis is based on the distinct characteristics of the malignant cells, which include enlargement, oval shape, elongation, and overlapping of the nuclei. The nuclei also display clearing or have a ground glass appearance.

2D10.2 Poorly differentiated carcinoma of thyroid gland

2D10.3 Undifferentiated carcinoma of thyroid gland

A primary carcinoma of the thyroid gland composed of undifferentiated cells. The malignant cells demonstrate evidence of epithelial differentiation, either by immunohistochemistry or electron microscopic studies. Microscopically, in the majority of cases there is a mixture of spindle, epithelioid, and giant cells. The vast majority of the patients present with a rapidly enlarging neck mass. The clinical course is invariably aggressive.

Inclusions: anaplastic carcinoma of thyroid gland

2D10.4 Medullary carcinoma of thyroid gland

A neuroendocrine carcinoma arising from the C-cells of the thyroid gland. It is closely associated with multiple endocrine neoplasia syndromes. Approximately 10% to 20% of medullary thyroid carcinomas are familial. Patients usually present with a thyroid nodule that is painless and firm. In the majority of cases nodal involvement is present at diagnosis.

2D10.Y Other specified malignant neoplasms of thyroid gland

2D10.Z Malignant neoplasms of thyroid gland, unspecified

2D11 Malignant neoplasms of adrenal gland

Tumours arising from the adrenal cortex include adenomas and carcinomas. These are rare neoplasms but may cause a variety of hormonal symptoms, including hyperaldosteronism, Cushing syndrome, and virilisation. A small fraction of adrenocortical tumours are associated with an inherited tumour syndrome, including Li-Fraumeni syndrome and Carney complex.

Benign and malignant phaeochromocytomas arise in the adrenal medulla and are derived from chromaffin cells of neural crest origin. Phaeochromocytomas may occur in the setting of several hereditary conditions, including multiple endocrine neoplasia types 2a and 2b, von Hippel Lindau disease and neurofibromatosis.

Extra adrenal paragangliomas arise from chromaffin cells in sympathoadrenal and parasympathetic paraganglia. They occur in many parts of the body and can pose a significant challenge to surgeons and oncologists. Some function as chemoreceptors, others are endocrinologically active. Familial paragangliomas are associated with mutations of the mitochondrial complex II genes.

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2D11.0 Adenocarcinoma of adrenal gland

2D11.1 Malignant phaeochromocytoma of adrenal gland

2D11.2 Neuroblastoma of adrenal gland

Neuroblastomas are malignant tumours that form in certain types of the nerve tissue. It most often begins in the adrenal gland. About 1 out of 3 neuroblastomas start in the adrenal glands and about 1 out of 4 begin in sympathetic nerve ganglia in the abdomen. Most of the rest start in sympathetic ganglia near the spine in the chest or neck, or in the pelvis.

2D11.Y Other specified malignant neoplasms of adrenal gland

2D11.Z Malignant neoplasms of adrenal gland, unspecified

2D12 Malignant neoplasms of other endocrine glands or related structures

Exclusions: Malignant neoplasms of adrenal gland (2D11)

Malignant neoplasms of testis (2C80)

Malignant neoplasms of ovary (2C73)

Malignant neoplasm of pancreas (2C10)

Malignant neoplasms of thyroid gland (2D10)

Malignant neoplasms of thymus (2C27)

Malignant mesenchymal neoplasms (BlockL3‑2B5)

2D12.0 Malignant epithelial neoplasms of other endocrine glands or related structures, unspecified type

2D12.1 Adenocarcinoma of other endocrine glands or related structures

2D12.Y Other specified malignant neoplasms of other endocrine glands or related structures

2D12.Z Malignant neoplasms of other endocrine glands or related structures, unspecified

2D1Y Other specified malignant neoplasms of endocrine glands

2D1Z Malignant neoplasms of endocrine glands, unspecified

2D3Y Other specified malignant neoplasms, stated or presumed to be primary, of specified sites, except of lymphoid, haematopoietic, central nervous system or related tissues

2D3Z Malignant neoplasms, stated or presumed to be primary, of specified sites, except of lymphoid, haematopoietic, central nervous system or related tissues, unspecified

Malignant neoplasms of ill-defined or unspecified primary sites (BlockL2‑2D4)

Exclusions: Malignant mesenchymal neoplasms (BlockL3‑2B5)

2D40 Adenocarcinoma of unspecified site

A common cancer characterised by the presence of malignant glandular cells. Morphologically, adenocarcinomas are classified according to the growth pattern (e.g., papillary, alveolar) or according to the secreting product (e.g., mucinous, serous). Representative examples of adenocarcinoma are ductal and lobular breast carcinoma, lung adenocarcinoma, renal cell carcinoma, hepatocellular carcinoma (hepatoma), colon adenocarcinoma, and prostate adenocarcinoma.

2D41 Unspecified carcinoma of unspecified site

2D42 Malignant neoplasms of ill-defined sites

Malignant neoplasms of ill defined sites is used for cases where the documentation refers to a site that includes multiple organ systems and tissue types that should be coded separately.

2D43 Malignant neoplasms of independent, multiple primary sites

Coding Note: Use additional codes to identify individual neoplasms.

2D4Y Other specified malignant neoplasms of ill-defined or unspecified primary sites

2D4Z Unspecified malignant neoplasms of ill-defined or unspecified sites

Malignant neoplasm metastases (BlockL2‑2D5)

Spread of a malignant neoplasm into secondary sites.

2D50 Malignant neoplasm metastasis in brain

A malignant neoplasm that has spread to the brain from another anatomic site or system. The majority are carcinomas (usually lung or breast carcinomas).

Coding Note: Code aslo the casusing condition

2D51 Malignant neoplasm metastasis in meninges

Coding Note: Code aslo the casusing condition

2D52 Malignant neoplasm metastasis in spinal cord, cranial nerves or remaining parts of central nervous system

Coding Note: Code aslo the casusing condition

Malignant neoplasm metastasis in lymph nodes (BlockL3‑2D6)

Exclusions: Neoplasms of haematopoietic or lymphoid tissues (BlockL1‑2A2)

2D60 Malignant neoplasm metastasis in lymph node of a single region

Coding Note: Code aslo the casusing condition

2D60.0 Malignant neoplasm metastasis in lymph nodes of head, face or neck

Coding Note: Code aslo the casusing condition

2D60.1 Malignant neoplasm metastasis in intrathoracic lymph nodes

Coding Note: Code aslo the casusing condition

2D60.2 Malignant neoplasm metastasis in intra-abdominal lymph nodes

Coding Note: Code aslo the casusing condition

2D60.3 Malignant neoplasm metastasis in axillary lymph nodes

Coding Note: Code aslo the casusing condition

2D60.4 Malignant neoplasm metastasis in inguinal lymph nodes

Coding Note: Code aslo the casusing condition

2D60.5 Malignant neoplasm metastasis in intrapelvic lymph nodes

Coding Note: Code aslo the casusing condition

2D60.Y Other specified malignant neoplasm metastasis in lymph node of a single region

Coding Note: Code aslo the casusing condition

2D60.Z Malignant neoplasm metastasis in lymph node of a single region, unspecified

Coding Note: Code aslo the casusing condition

2D61 Malignant neoplasm metastases in lymph nodes of multiple regions

Coding Note: Code aslo the casusing condition

2D6Z Metastatic malignant neoplasm to unspecified lymph node

Coding Note: Code aslo the casusing condition

Malignant neoplasm metastasis in thoracic or respiratory organs (BlockL3‑2D7)

2D70 Malignant neoplasm metastasis in lung

Exclusions: Malignant neoplasms of bronchus or lung (2C25)

2D71 Malignant neoplasm metastasis in mediastinum

The spread of cancer to the mediastinum from an adjacent or distant anatomic site.

Coding Note: Code aslo the casusing condition

2D72 Malignant neoplasm metastasis in pleura

The spread of cancer to the pleura from an adjacent or distant anatomic site.

Coding Note: Code aslo the casusing condition

2D73 Malignant neoplasm metastasis in upper respiratory tract organs

Coding Note: Code aslo the casusing condition

2D7Y Malignant neoplasm metastasis in other specified thoracic organs

Coding Note: Code aslo the casusing condition

2D7Z Malignant neoplasm metastasis in thoracic or respiratory organs, unspecified

Coding Note: Code aslo the casusing condition

Malignant neoplasm metastasis in digestive system (BlockL3‑2D8)

2D80 Malignant neoplasm metastasis in liver or intrahepatic bile duct

Malignant neoplasms that have metastasized to the liver from extrahepatic primary tumours.

Coding Note: Code aslo the casusing condition

2D80.0 Malignant neoplasm metastasis in liver

Coding Note: Code aslo the casusing condition

2D80.1 Malignant neoplasm metastasis in intrahepatic bile duct

Coding Note: Code aslo the casusing condition

2D80.Y Other specified malignant neoplasm metastasis in liver or intrahepatic bile duct

Coding Note: Code aslo the casusing condition

2D80.Z Malignant neoplasm metastasis in liver or intrahepatic bile duct, unspecified

Coding Note: Code aslo the casusing condition

2D81 Malignant neoplasm metastasis in pancreas

A malignant neoplasm that has spread to the pancreas from another anatomic site. Representative examples include metastatic carcinomas from the gastrointestinal tract, metastatic melanomas, and renal cell carcinomas.

Coding Note: Code aslo the casusing condition

2D82 Malignant neoplasm metastasis in extrahepatic bile ducts

Coding Note: Code aslo the casusing condition

2D83 Malignant neoplasm metastasis in ampulla of Vater

Coding Note: Code aslo the casusing condition

2D84 Malignant neoplasm metastasis in the small intestine

The spread of cancer to the small intestine. This may be from a primary intestinal cancer, or from a cancer at a distant site.

Coding Note: Code aslo the casusing condition

2D85 Malignant neoplasm metastasis in large intestine

The spread of cancer to the large intestine. This may be from a primary colon or rectal cancer, or from a cancer at a distant site.

Coding Note: Code aslo the casusing condition

2D86 Malignant neoplasm metastasis in anus

Malignant tumour that metastasized in the anus and anal canal.

Coding Note: Code aslo the casusing condition

2D8Y Malignant neoplasm metastasis in other specified digestive system organ

Coding Note: Code aslo the casusing condition

2D8Z Malignant neoplasm metastasis in unspecified digestive system organ

Coding Note: Code aslo the casusing condition

Malignant neoplasm metastasis in retroperitoneum or peritoneum (BlockL3‑2D9)

2D90 Malignant neoplasm metastasis in retroperitoneum

Coding Note: Code aslo the casusing condition

2D91 Malignant neoplasm metastasis in peritoneum

Coding Note: Code aslo the casusing condition

Malignant neoplasm metastasis in other sites (BlockL3‑2E0)

2E00 Malignant neoplasm metastasis in kidney or renal pelvis

The spread of the cancer to the kidney. This may be from a primary kidney cancer involving the opposite kidney, or from a cancer at a distant site.

Coding Note: Code aslo the casusing condition

2E01 Malignant neoplasm metastasis in bladder

Tumours of the urinary bladder that originate from an extravesical, non-urothelial tract neoplasm

Coding Note: Code aslo the casusing condition

2E02 Malignant neoplasm metastasis in other or unspecified urinary system organs

Coding Note: Code aslo the casusing condition

2E03 Malignant neoplasm metastasis in bone or bone marrow

The spread of a malignant neoplasm from a primary site to the skeletal system. The majority of metastatic neoplasms to the bone are carcinomas.

Coding Note: Code aslo the casusing condition

2E04 Malignant neoplasm metastasis in soft tissue

Coding Note: Code aslo the casusing condition

2E05 Malignant neoplasm metastasis in female reproductive system

Coding Note: Code aslo the casusing condition

2E05.0 Malignant neoplasm metastasis in ovary

The spread of the cancer to the ovary. This may be from a primary ovarian cancer involving the opposite ovary, or from a cancer at a distant site.

2E05.Y Malignant neoplasm metastasis in other female reproductive system organs

Coding Note: Code aslo the casusing condition

2E05.Z Malignant neoplasm metastasis in female reproductive system, unspecified

Coding Note: Code aslo the casusing condition

2E06 Malignant neoplasm metastasis in male genital organs

Coding Note: Code aslo the casusing condition

2E07 Malignant neoplasm metastasis in adrenal gland

A malignant tumour that has spread to the adrenal gland from an adjacent or distant anatomic site. The majority of cases are metastatic carcinomas, and less frequently lymphomas. (NCI05)

Coding Note: Code aslo the casusing condition

2E08 Metastatic malignant neoplasm involving skin

Involvement of the skin by metastatic spread from a known or unknown primary malignant neoplasm. The secondary deposit may result from local migration of malignant cells, or from regional lymphatic or haematogenous spread from more distant sites.

Coding Note: Code aslo the casusing condition

2E09 Malignant neoplasm metastasis in peripheral nervous system

Coding Note: Code aslo the casusing condition

2E0Y Malignant neoplasm metastasis in other specified sites

Coding Note: Code aslo the casusing condition

2E2Z Malignant neoplasm metastasis, unspecified

Coding Note: Code aslo the casusing condition

In situ neoplasms, except of lymphoid, haematopoietic, central nervous system or related tissues (BlockL1‑2E6)

2E60 Carcinoma in situ of oral cavity, oesophagus or stomach

Exclusions: Melanoma in situ neoplasms (2E63)

2E60.0 Carcinoma in situ of lip, oral cavity or pharynx

Exclusions: Carcinoma in situ of aryepiglottic fold, laryngeal aspect (2E62.0)

Carcinoma in situ of epiglottis nos (2E62.0)

Carcinoma in situ of epiglottis, suprahyoid portion (2E62.0)

Carcinoma in situ of skin of lip (2E64)

2E60.1 Carcinoma in situ of oesophagus

Stage 0 includes: For squamous cell carcinoma: Tis (HGD), N0, M0, G1, GX, tumour location: Any. For adenocarcinoma: Tis (HGD), N0, M0, G1, GX. Tis: High-grade dysplasia. N0: No regional lymph node metastasis. M0: No distant metastasis. G1: Well differentiated. GX: Grade cannot be assessed-stage grouping as G1. tumour location: Location of the primary cancer site is defined by the position of the upper (proximal) edge of the tumour in the esophagus.

2E60.2 Carcinoma in situ of stomach

Stage 0 includes: Tis, N0, M0. Tis: Carcinoma in situ: intraepithelial tumour without invasion of the lamina propria. N0: No regional lymph node metastasis. M0: No distant metastasis.

2E61 Carcinoma in situ of other or unspecified digestive organs

Exclusions: Melanoma in situ neoplasms (2E63)

2E61.0 Carcinoma in situ of colon

Stage 0 includes: Tis, N0, M0. Tis: Carcinoma in situ: intraepithelial or invasion of lamina propria. N0: No regional lymph node metastasis. M0: No distant metastasis.

2E61.1 Carcinoma in situ of rectum

Malignant epithelial tumour of rectum that has not invaded adjacent tissue of the large intestine.

2E61.2 Carcinoma in situ of anal canal

Malignant epithelial tumour that has not invaded beyond the epithelium of the anal canal.

Exclusions: Carcinoma in situ of anal margin (2E64.2)

Carcinoma in situ of anal skin (2E64)

Carcinoma in situ of perianal skin (2E64.2)

2E61.3 Carcinoma in situ of gallbladder, biliary tract or ampulla of Vater

an early form of cancer without invasion of tumour cells into the surrounding tissue, usually before penetration through the basement membrane.

2E61.Y Carcinoma in situ of other specified digestive organs

2E61.Z Carcinoma in situ of unspecified digestive organs

2E62 Carcinoma in situ of middle ear or respiratory system

Exclusions: Melanoma in situ neoplasms (2E63)

2E62.0 Carcinoma in situ of larynx

Exclusions: Carcinoma in situ of aryepiglottic fold, hypopharyngeal aspect (2E60.0)

Carcinoma in situ of aryepiglottic fold, marginal zone (2E60.0)

Carcinoma in situ of aryepiglottic fold, NOS (2E60.0)

2E62.1 Carcinoma in situ of trachea

2E62.2 Carcinoma in situ of bronchus or lung

2E62.Y Carcinoma in situ of other specified sites of middle ear and respiratory system

2E62.Z Carcinoma in situ of unspecified sites of middle ear and respiratory system

2E63 Melanoma in situ neoplasms

Stage 0 includes: Tis, N0, M0. Tis: Melanoma in situ. N0: No regional lymph node metastases. M0: No detectable evidence of distant metastases.

2E63.0 Melanoma in situ of skin

Malignant melanoma confined to the epidermis and described as being in radial growth phase.

2E63.00 Lentigo maligna

An atypical proliferation of atypical melanocytes in the dermal-epidermal junction, without infiltration of the papillary or reticular dermis. The melanocytic proliferation is associated with actinic damage and epidermal atrophy. It usually occurs in the sun-exposed skin of elderly people. It is a form of melanoma in situ and in approximately 5% of cases it progresses to lentigo maligna melanoma.

2E63.0Z Melanoma in situ of skin, unspecified

2E63.1 Melanoma in situ of conjunctiva

2E63.Y Other specified melanoma in situ neoplasms

2E63.Z Melanoma in situ neoplasms, unspecified

2E64 Carcinoma in situ of skin

Stage 0 includes: Tis, N0, M0. Tis: Carcinoma in situ. N0: No regional lymph node metastasis. M0: No clinical or radiographic evidence of distant metastasis.

Exclusions: Melanoma in situ neoplasms (2E63)

Coded Elsewhere: Carcinoma in situ of vulva (2E67.1)

Carcinoma in situ of penis (2E67.4)

2E64.0 Intraepidermal squamous cell carcinoma

Malignant squamous neoplasia confined to the epidermis of extragenital skin and known commonly as Bowen disease. It arises most frequently on chronically sun-exposed glabrous skin of the head and neck or lower legs. It typically presents as single or multiple well-demarcated scaly erythematous patches, nodules or plaques which histologically show extensive keratinocytic atypia. It may develop from preexisting actinic keratosis (Actinic intraepidermal squamous cell carcinoma). Although it is most commonly associated with exposure to ultraviolet radiation, other carcinogens such as arsenic and tar may be implicated. Human papilloma virus may represent an additional risk factor in immunosuppressed patients.

2E64.00 Bowen disease of skin

Intraepidermal squamous cell carcinoma due to predisposing factors including chronic human papilloma virus infection, arsenic ingestion, ionising radtiation and chronic immunosuppression.

2E64.01 Actinic intraepidermal squamous cell carcinoma

Intraepidermal squamous cell carcinoma attributable to chronic exposure to ultraviolet radiation and typically developing from a pre-existing actinic keratosis

2E64.0Y Other specified intraepidermal squamous cell carcinoma

2E64.0Z Intraepidermal squamous cell carcinoma, unspecified

2E64.1 Extramammary Paget disease of skin

An intraepithelial adenocarcinoma of apocrine gland-bearing skin and mucous membrane. Clinically it presents as sharply demarcated erythematous plaques most commonly affecting the vulva in women and perianal skin in men.

Coded Elsewhere: Vulvar Paget disease (2E67.11)

2E64.2 Carcinoma in situ of anal margin or perianal skin

Carcinoma in situ of anal margin or perianal skin is most commonly squamous and related to oncogenic HPV strains, HIV infection or both. It may present as warty pigmented patches (Bowenoid papulosis).

2E64.Y Other specified carcinoma in situ of skin

2E64.Z Carcinoma in situ of skin, unspecified

2E65 Carcinoma in situ of breast

Exclusions: carcinoma in situ of skin of breast (2E64)

melanoma in situ of breast (skin) (2E63)

2E65.0 Lobular carcinoma in situ of breast

2E65.1 Lobular carcinoma in situ of breast, pleomorphic subtype

2E65.2 Ductal carcinoma in situ of breast

Exclusions: Atypical ductal hyperplasia of breast (2F75)

2E65.3 Ductal carcinoma in situ of breast, comedo subtype

2E65.4 Mixed ductal and lobular carcinoma in situ of breast

The co-existence of ductal and lobular carcinoma in situ in the breast, without evidence of stromal invasion.

2E65.5 Paget disease of nipple

Paget disease of the nipple describes a rare presentation of breast cancer, seen most frequently in women aged 50-60, manifesting with nipple drainage and itching, erythema, crusty, excoriated nipple, thickened plaques and hyperpigmentation (less frequently). It is due to tumour cells invading the nipple-areola complex and represents 1%-3% of all new breast cancer diagnoses.

2E65.Y Other specified carcinoma in situ of breast

2E65.Z Carcinoma in situ of breast, unspecified

2E66 Carcinoma in situ of cervix uteri

Exclusions: melanoma in situ of cervix (2E63)

severe dysplasia of cervix NOS (GA13.1)

2E66.0 Cervical Intraepithelial neoplasia grade II

A condition of the cervix, caused by chronic infection of the cervix with human papillomavirus. This condition is characterised by premalignant transformation and moderate dysplasia of the cervix confined to the basal two thirds of the squamous epithelial cells on the surface of the tissue. Confirmation is by the Papanicolaou smear test followed by a biopsy of any abnormal cell growth.

Inclusions: moderate cervical dysplasia

2E66.1 Cervical intraepithelial neoplasia grade III

A condition of the cervix, caused by chronic infection with human papillomavirus. This condition is characterised by premalignant transformation and severe dysplasia of the cervix that spans more than two thirds of the squamous epithelial cells on the surface of the tissue. Confirmation is by the Papanicolaou smear test followed by a biopsy of any abnormal cell growth.

Inclusions: severe cervical dysplasia and carcinoma in-situ

2E66.Y Other specified carcinoma in situ of cervix uteri

2E66.Z Carcinoma in situ of cervix uteri, unspecified

2E67 Carcinoma in situ of other or unspecified genital organs

Exclusions: Melanoma in situ neoplasms (2E63)

2E67.0 Carcinoma in situ of endometrium

2E67.1 Carcinoma in situ of vulva

Exclusions: severe dysplasia of vulva NOS (GA13.1)

2E67.10 Vulvar intraepithelial neoplasia

A precancerous neoplastic process characterised by dysplasia and maturation abnormalities of the vulvar squamous epithelium. There is no evidence of invasion. It is associated with human papillomavirus infection and is classified as low or high grade.

2E67.11 Vulvar Paget disease

An uncommon intraepithelial malignant neoplasm of eccrine or apocrine origin, arising from the vulva. It usually affects post-menopausal women. In approximately 10-20% of the cases there is an associated anorectal, or urothelial carcinoma or a skin appendage adenocarcinoma identified. It presents as a red, eczematous lesion. Microscopically, it is characterised by the presence of the typical Paget cells which are large, round cells with abundant cytoplasm and prominent nuclei.

2E67.2 Carcinoma in situ of vagina

2E67.20 Vaginal intraepithelial neoplasia grade II

A condition of the vagina, characterised by lesions of the squamous vaginal intraepithelial cells, leading to dysplasia, varying degrees of atypia, and abnormal cell growth within two thirds of the vaginal skin. This condition may be associated with human papillomavirus infection. Confirmation is by the Papanicolaou smear test followed by a biopsy of any abnormal cell growth.

Inclusions: moderate vaginal dysplasia

2E67.21 Vaginal intraepithelial neoplasia grade III

A condition of the vagina, characterised by lesions of the squamous vaginal intraepithelial cells and carcinoma in situ, leading to dysplasia, varying degrees of atypia, and abnormal cell growth within the full thickness of the vaginal skin. This condition may be associated with human papillomavirus infection. Confirmation is by the Papanicolaou smear test followed by a biopsy of any abnormal cell growth.

Inclusions: severe vaginal dysplasia and carcinoma in-situ

2E67.2Y Other specified carcinoma in situ of vagina

2E67.2Z Carcinoma in situ of vagina, unspecified

2E67.3 Carcinoma in situ of other or unspecified female genital organs

2E67.4 Carcinoma in situ of penis

This comprises both squamous carcinoma in situ and extramammary Paget disease of the penis. The former is an uncommon precancerous disease of penile skin. Lesions usually appear on the glans or inner aspect of the foreskin and are almost always found in uncircumcised men. If left untreated, 10-30% of cases develop into invasive squamous cell carcinoma of the penis. When it affects the skin of the shaft or prepuce it is commonly called Bowen disease. If it affects the glans or inner surface of the prepuce it may also be referred to as penile intraepithelial neoplasia (or in the past as erythroplasia of Queyrat). Extramammary Paget disease of penis is a rare form of carcinoma in situ involving penile skin or glans penis.

Coded Elsewhere: Extramammary Paget disease of penis (2E64.1)

2E67.40 Squamous cell carcinoma in situ of skin of penis

Squamous cell carcinoma affecting the skin of the prepuce or of the shaft of the penis and commonly called Bowen disease. HPV infection and chronic exposure to psoralen photochemotherapy are predisposing factors.

Inclusions: Bowen disease of skin of penis

2E67.41 Squamous cell carcinoma in situ of mucocutaneous epithelium of penis

Inclusions: Penile intraepithelial neoplasia of inner preputial epithelium

Penile intraepithelial neoplasia of glans penis

2E67.5 Carcinoma in situ of prostate

High grade prostatic intraepithelial neoplasia characterised by the presence of severe architectural and cytologic abnormalities.

Inclusions: high grade prostatic intraepithelial neoplasia

Exclusions: low grade dysplasia of prostate (GA91.6)

2E67.6 Carcinoma in situ of other or unspecified male genital organs

2E68 Carcinoma in situ of bladder

Stage 0is includes: Tis, N0, M0. Tis: Carcinoma in situ: flat tumour. N0: No regional lymph node metastasis. M0: No distant metastasis.

2E69 Carcinoma in situ of other or unspecified urinary organs

2E6A Carcinoma in situ of the eye or ocular adnexa

2E6A.0 Carcinoma in situ of the conjunctiva

Exclusions: Melanoma in situ of conjunctiva (2E63.1)

2E6A.1 Carcinoma in situ of the cornea

2E6A.Y Carcinoma in situ of other and unspecified part of the eye and adnexa

2E6B Carcinoma in situ of thyroid and other endocrine glands

Exclusions: Carcinoma in situ of ovary (2E67.3)

Carcinoma in situ of testis (2E67.6)

2E6Y Carcinoma in situ of other specified site

2E6Z Carcinoma in situ of unspecified site

Benign neoplasms, except of lymphoid, haematopoietic, central nervous system or related tissues (BlockL1‑2E8)

A neoplasm which is characterised by the absence of morphologic features associated with malignancy (severe cytologic atypia, tumour cell necrosis, and high mitotic rate). Benign neoplasms remain confined to the original site of growth and only rarely metastasize to other anatomic sites.

Benign mesenchymal neoplasms (BlockL2‑2E8)

2E80 Benign lipomatous neoplasm

A benign tumour composed of adipose (fatty) tissue. The most common representative of this category is the lipoma.

2E80.0 Lipoma

2E80.00 Superficial subcutaneous lipoma

A benign well-circumscribed mesenchymal neoplasm composed of mature adipocytes and commonly known as a lipoma.

2E80.01 Deep subfascial lipoma

Deep subfascial lipomata are benign neoplasms of adipose tissue which arise deep to the deep fascia and have a tendency to infiltrate between and into muscle. They may occur at any body site and may cause diagnostic difficulty. They are well recognised to occur on the forehead beneath the frontalis muscle (frontalis-associated lipoma).

Inclusions: Frontalis-associated lipoma

Infiltrating lipoma of soft tissue

Intramuscular lipoma of soft tissue

2E80.02 Deep internal or visceral lipoma

2E80.0Y Lipoma, other specified site

2E80.0Z Lipoma, unspecified site

2E80.1 Lipoblastoma

2E80.Y Other specified benign lipomatous neoplasm

2E80.Z Benign lipomatous neoplasm, unspecified

2E81 Benign vascular neoplasms

Exclusions: Blue naevus (2F20)

Pigmented naevus (2F20)

Coded Elsewhere: Lobular capillary haemangioma (2F26)

2E81.0 Neoplastic haemangioma

A benign localised vascular neoplasm usually occurring in infancy and childhood. It is characterised by the formation of capillary-sized or cavernous vascular channels. The majority of cases are congenital.

Coded Elsewhere: Pulmonary sclerosing haemangioma (2F00.Y)

2E81.00 Umbilical cord haemangioma

tumour composed of thin walled blood vessels lined by endothelium present within the cord

2E81.01 Conjunctival haemangioma or haemolymphangioma

2E81.0Y Neoplastic haemangioma of other specified site

2E81.0Z Neoplastic haemangioma, unspecified

2E81.1 Benign lymphatic neoplasms

Benign circumscribed or diffuse neoplasms of lymphatic vessels. They are are much less common than lymphatic malformations and are distinguished from the latter by proliferative growth and the potential to become widely disseminated.

Exclusions: Lymphatic malformations (LA90.1)

2E81.10 Disseminated lymphangiomatosis

A rare disorder characterised by widespread proliferation of aberrant lymphatic vessels which typically infiltrate vital organs in the thorax and abdomen.

2E81.11 Acquired progressive lymphangioma

Acquired progressive lymphangioma is a benign localised but slowly progressive tumour of lymphatic vessels that typically presents as reddish or bruise‐like plaques on the abdominal wall, thigh or calf of young adolescents.

Exclusions: Lymphatic malformations (LA90.1)

2E81.1Y Other specified benign lymphatic neoplasms

2E81.1Z Benign lymphatic neoplasms, unspecified

2E81.2 Benign vascular neoplasms of infancy and childhood

The commonest benign vascular neoplasm of infancy is infantile haemangioma. Less common neoplasms are congenital haemangioma, spindle cell haemangioma, tufted angioma and kaposiform haemangioendothelioma.

2E81.20 Focal infantile haemangioma

Infantile haemangioma is a common benign vascular neoplasm which develops in about 4% of infants. It appears within weeks of birth as a blanched, blushed, or telangiectatic area that then rapidly proliferates for several months before entering a prolonged process of involution lasting up to 12 years, leaving a residual variably prominent scar. A solitary focal tumour is seen in about 85% of cases. Over half of cases are located on the head and neck. Complications include bleeding, infection, ulceration and, in tumours situated close to the eye, amblyopia.

Inclusions: Strawberry naevus

2E81.21 Multifocal infantile haemangioma

Infantile haemangioma is multifocal in up to 25% of cases with numbers ranging from a few to many dozens. If more than 5 cutaneous tumours are present there is an increased risk of associated internal haemangiomatosis, especially of the liver.

2E81.2Y Other specified benign vascular neoplasms of infancy and childhood

2E81.2Z Benign vascular neoplasms of infancy and childhood, unspecified

2E81.Y Other specified benign vascular neoplasms

2E81.Z Benign vascular neoplasms, unspecified

2E82 Benign chondrogenic tumours

2E82.0 Benign chondrogenic tumours of bone or articular cartilage of limbs

2E82.1 Benign chondrogenic tumours of bone or articular cartilage of other specified sites

2E82.Z Benign chondrogenic tumours, site unspecified

2E83 Benign osteogenic tumours

A neoplasm arising from the bone or articular cartilage that does not invade adjacent tissues or metastasize to other anatomic sites. Representative examples include benign fibrous histiocytoma of bone, osteoma, osteoblastoma, chondroblastoma, and enchondroma.

2E83.0 Benign osteogenic tumours of bone or articular cartilage of skull or face

2E83.1 Benign osteogenic tumours of bone or articular cartilage of lower jaw

2E83.2 Benign osteogenic tumours of bone or articular cartilage of vertebral column

Exclusions: Benign osteogenic tumour of sacrum (2E83.4)

2E83.3 Benign osteogenic tumours of bone or articular cartilage of ribs, sternum or clavicle

2E83.4 Benign osteogenic tumours of bone or articular cartilage of pelvic bones, sacrum or coccyx

2E83.5 Benign osteogenic tumours of bone or articular cartilage of limbs

2E83.Y Benign osteogenic tumour of other specified site

2E83.Z Benign osteogenic tumour of unspecified site

2E84 Benign fibrogenic or myofibrogenic tumour

2E84.0 Benign fibrogenic or myofibrogenic tumour of skin

2E84.Y Benign fibrogenic or myofibrogenic tumour of other specified sites

2E84.Z Benign fibrogenic or myofibrogenic tumour, site unknown

2E85 Benign fibrohistiocytic tumour

Exclusions: Benign neoplasm of peripheral nerves or autonomic nervous system (BlockL2‑2E9)

Benign lymphatic neoplasms (2E81.1)

Benign lipomatous neoplasm (2E80)

Haemangioma (2E81)

Benign neoplasm of uterine ligament, any (2F31)

Benign vascular neoplasms (2E81)

Leiomyoma of uterus (2E86.0)

Benign neoplasm of connective tissue of breast (2F30)

2E85.0 Benign fibrohistiocytic tumour of soft tissues of limbs

2E85.1 Benign fibrohistiocytic tumour of retroperitoneum or peritoneum

Exclusions: Benign lipomatous neoplasm (2E80)

Benign neoplasm of mesothelial tissue (2F10)

2E85.2 Benign fibrohistiocytic tumour of skin

2E85.Y Benign fibrohistiocytic tumour of other specified sites

2E85.Z Benign fibrohistiocytic tumour, site unspecified

2E86 Benign smooth muscle or skeletal muscle tumour

2E86.0 Leiomyoma of uterus

A well-circumscribed benign smooth muscle neoplasm characterised by the presence of spindle cells with cigar-shaped nuclei, interlacing fascicles, and a whorled pattern.

Exclusions: Leiomyoma of ovary (2E86.1)

Leiomyoma of fallopian tube (2E86.1)

Leiomyoma of broad ligament (2E86.1)

Leiomyoma of vagina (2E86.1)

Leiomyoma of vulva (2E86.1)

Benign non-mesenchymal neoplasms of uterus (2F31)

2E86.1 Leiomyoma of other or unspecified sites

2E86.2 Rhabdomyoma

2E86.Y Other specified benign smooth muscle or skeletal muscle tumour

2E86.Z Benign smooth muscle or skeletal muscle tumour, unspecified

2E87 Benign gastrointestinal stromal tumour

2E88 Benign endometrial stromal nodule

2E89 Benign mesenchymal tumours of uncertain differentiation

2E89.0 Benign tumours of uncertain differentiation, bone or cartilage

2E89.1 Benign tumours of uncertain differentiation, soft tissue

2E89.Y Benign mesenchymal tumours of uncertain differentiation of other specified site

2E89.Z Benign mesenchymal tumours of uncertain differentiation of unspecified site

2E8A Other mixed or unspecified benign mesenchymal tumours

2E8Y Benign neoplasm of mesothelial tissue, other specified organs

2E8Z Benign mesenchymal neoplasms, unspecified

Benign non-mesenchymal neoplasms (BlockL2‑2E9)

2E90 Benign neoplasm of lip, oral cavity or pharynx

2E90.0 Benign neoplasm of lip

A neoplasm without malignant characteristics arising from the lip.

Exclusions: Benign neoplasm of skin of lip (BlockL3‑2F2)

2E90.1 Benign neoplasm of tongue

Abnormal growth, without malignant characteristics, of the cells that comprise the tongue.

2E90.2 Benign neoplasm of floor of mouth

2E90.3 Benign neoplasm of other or unspecified parts of mouth

Exclusions: Benign neoplasm of nasopharyngeal surface of soft palate (2E90.6)

benign odontogenic neoplasms (2E83.0)

mucosa of lip (2E90.0)

2E90.4 Benign neoplasm of tonsil

Exclusions: benign neoplasm of pharyngeal tonsil (2E90.6)

benign neoplasm of lingual tonsil (2E90.1)

benign neoplasm of tonsillar pillars (2E90.5)

benign neoplasm of tonsillar fossa (2E90.5)

2E90.5 Benign neoplasm of oropharynx

A neoplasm of the oropharynx which is characterised by the absence of morphologic features associated with malignancy (severe cytologic atypia, tumour cell necrosis, and high mitotic rate). Benign neoplasms remain confined to the original site of growth and only rarely metastasize to other anatomic sites.

Exclusions: Benign neoplasm of epiglottis, NOS (2F00)

Benign neoplasm of epiglottis, suprahyoid portion (2F00)

2E90.6 Benign neoplasm of nasopharynx

A neoplasm of the nasopharynx which is characterised by the absence of morphologic features associated with malignancy (severe cytologic atypia, tumour cell necrosis, and high mitotic rate). Benign neoplasms remain confined to the original site of growth and only rarely metastasize to other anatomic sites.

2E90.7 Benign neoplasm of hypopharynx

2E90.8 Benign neoplasm of pharynx, unspecified

2E91 Benign neoplasm of major salivary glands

Exclusions: Benign neoplasms of minor salivary glands NOS (2E90.3)

2E91.0 Benign neoplasm of parotid gland

2E91.1 Benign neoplasm of other specified major salivary glands

2E91.Z Benign neoplasm of major salivary glands, unspecified

2E92 Benign neoplasm of digestive organs

A neoplasm of other and/or ill-defined parts of the digestive system which is characterised by the absence of morphologic features associated with malignancy (severe cytologic atypia, tumour cell necrosis, and high mitotic rate). Benign neoplasms remain confined to the original site of growth and only rarely metastasize to other anatomic sites.

2E92.0 Benign neoplasm of oesophagus

A non-metastasizing neoplasm arising from the esophageal wall.

Coded Elsewhere: Benign mesenchymal tumour of oesophagus (2E8Y)

2E92.1 Benign neoplasm of stomach

A non-metastasizing neoplasm arising from the gastric wall.

Coded Elsewhere: Benign mesenchymal tumour of stomach (2E8Y)

2E92.2 Benign neoplasm of duodenum

A non-metastasizing neoplasm arising from the wall of the duodenum.

Coded Elsewhere: Benign mesenchymal tumour of duodenum (2E8Y)

2E92.3 Benign neoplasm of other or unspecified parts of small intestine

Exclusions: Benign neoplasm of duodenum (2E92.2)

Coded Elsewhere: Benign mesenchymal tumour of small intestine (2E8Y)

2E92.4 Benign neoplasm of the large intestine

A non-metastasizing neoplasm arising from the wall of the colon and rectum.

Coded Elsewhere: Benign mesenchymal tumour of large intestine (2E8Y)

2E92.40 Polyposis syndrome

Intestinal polyposis syndromes can be divided, based on histology, into the broad categories of familial adenomatous polyposis (FAP), hamartomatous polyposis syndromes, and other rare polyposis syndromes, such as hereditary-mixed polyposis syndrome (HMPS).

Coded Elsewhere: Gardner syndrome (LD2D.3)

Peutz-Jeghers syndrome (LD2D.0)

Cronkhite-Canada syndrome (LD27.01)

Familial adenomatous polyposis (2B90.Y)

Juvenile gastrointestinal polyposis (2B90.Y)

2E92.4Y Other specified benign neoplasm of the large intestine

2E92.4Z Benign neoplasm of the large intestine, unspecified

2E92.5 Benign neoplasm of anus or anal canal

Primary benign tumour that forms in tissues lining the anus and anal canal.

Exclusions: Benign neoplasm of perianal skin (BlockL3‑2F2)

Benign neoplasm of anal margin (BlockL3‑2F2)

Benign neoplasm of anal skin (BlockL3‑2F2)

2E92.6 Benign neoplasm of gallbladder, extrahepatic bile ducts or ampulla of Vater

Coded Elsewhere: Benign mesenchymal tumour of gallbladder, extrahepatic bile ducts or ampulla of Vater (2E8Y)

Adenoma of bile ducts (2E92.6)

2E92.7 Benign neoplasm of liver or intrahepatic bile ducts

Coded Elsewhere: Focal nodular hyperplasia of liver (DB99.Y)

Haemangioma of liver (2E81.0Y)

2E92.8 Benign neoplasm of pancreas

A non-metastasizing neoplasm arising from the pancreas.

Exclusions: Benign neoplasm of endocrine pancreas (2E92.9)

2E92.9 Benign neoplasm of endocrine pancreas

Inclusions: Islet cell tumour

benign neoplasm of islets of Langerhans

2E92.A Benign neoplasm of ill-defined site within the digestive system

2E92.Y Benign neoplasm of other specified digestive organs

2E92.Z Benign neoplasm of unspecified digestive organs

Benign neoplasm of respiratory or intrathoracic organs (BlockL3‑2F0)

2F00 Benign neoplasm of middle ear or respiratory system

2F00.0 Middle ear endocrine tumour

2F00.1 Recurrent respiratory papillomatosis

Recurrent respiratory papillomatosis is a rare respiratory disease characterised by the development of exophytic papillomas, affecting the mucosa of the upper aero-digestive tract (with a strong predilection for the larynx), caused by an infection with human papilloma virus. Symptoms at presentation may include hoarseness, chronic cough, dyspnoea, recurrent upper respiratory tract infections, pneumonia, dysphagia, stridor, and/or failure to thrive.

2F00.2 Laryngeal endocrine tumour

2F00.Y Other specified benign neoplasm of middle ear or respiratory system

2F00.Z Benign neoplasm of middle ear or respiratory system, unspecified

2F01 Benign neoplasm of intrathoracic organs

2F0Y Benign neoplasms of other specified respiratory and intrathoracic organs

2F0Z Benign neoplasms of respiratory and intrathoracic organs, unspecified

2F10 Benign neoplasm of mesothelial tissue

A benign neoplasm arising from mesothelial cells. It is characterised by the formation of glandular and tubular patterns. It can occur in several anatomic sites including the pleura, peritoneum, and epididymis.

Benign cutaneous neoplasms (BlockL3‑2F2)

Abnormal growth of the cells that comprise the tissues of the skin, without any evidence of malignancy.

Coded Elsewhere: Benign vascular neoplasms of infancy and childhood (2E81.2)

2F20 Benign cutaneous melanocytic neoplasms

Inclusions: Mole

Pigmented naevus

Benign melanocytic naevus

2F20.0 Common acquired melanocytic naevus

2F20.00 Multiple benign melanocytic naevi

The presence of multiple benign melanocytic naevi (often taken as more than 20 naevi >2mm in diameter), an independent risk factor for the development of melanoma with highest risk associated with highest numbers of naevi (>100).

2F20.0Y Other specified common acquired melanocytic naevus

2F20.1 Atypical melanocytic naevus

Solitary or multiple, slightly raised, pigmented lesions with irregular borders, usually measuring more than 0.6cm in greatest dimension. Morphologically, there is melanocytic atypia and the differential diagnosis from melanoma may be difficult. Patients are at an increased risk for the development of melanoma.

Inclusions: Dysplastic naevus, unspecified

2F20.2 Congenital melanocytic naevus

Congenital melanocytic naevi are circumscribed areas of skin pigmentation present at birth as a result of abnormal intrauterine proliferation of melanocytes within the dermis, the epidermis or both. They may range in size from a few millimetres to many centimetres in diameter. If their projected or final adult maximal diameter is greater than 20 cm they are termed giant congenital melanocytic naevi.

2F20.20 Giant congenital melanocytic naevus

A congenital melanocytic naevus (CMN) with a predicted or final adult maximal diameter of 400 mm or more. Giant CMNs are commonly centred on the dorsal surface of the body between the vertex and the buttocks but may occur elsewhere; they may be associated with multiple smaller satellite naevi (congenital or tardive), hypertrichosis, lipomas or benign proliferative nodules. There is a risk of pre-pubertal melanoma within giant CMN or the central nervous system (CNS). Leptomeningeal melanocytosis or focal neuromelanosis, found in 10-15% of cases, is often associated with other CNS tumours, hydrocephalus, epilepsy, arachnoid cysts, or Dandy-Walker malformation.

2F20.2Y Other specified congenital melanocytic naevus

2F20.2Z Congenital melanocytic naevus, unspecified

2F20.3 Generalised eruptive melanocytic naevi

This phenomenon describes the rapid simultaneous appearance of multiple melanocytic naevi, often hundreds in number, on previously uninvolved sun-exposed skin. The phenomenon has been linked to immunosuppression, particularly in renal transplant recipients and in individuals receiving cancer chemotherapy, and may be considered a more advanced counterpart of generalised eruptive lentiginosis.

Exclusions: Multiple benign melanocytic naevi (2F20.00)

Generalized eruptive lentiginosis (ED61)

2F20.Y Other specific types of melanocytic naevus

2F20.Z Melanocytic naevus, unspecified

2F21 Benign keratinocytic acanthomas

A group of benign discrete epidermal proliferative disorders including seborrhoeic keratosis and clear cell acanthoma.

2F21.0 Seborrhoeic keratosis

Seborrhoeic keratoses are very common benign neoplasms of epidermal keratinocytes which increase in prevalence and number with age. They are commonly multiple and are very variable in shape and colour. Because of the sometimes intense pigmentation they are frequently mistaken for melanocytic tumours.

Inclusions: Basal cell papilloma

Seborrheic wart

2F21.Y Other specified benign keratinocytic acanthomas

2F22 Benign neoplasms of epidermal appendages

A range of benign neoplasms arising from the hair follicle, its associated glands or from sweat glands.

2F23 Benign dermal fibrous or fibrohistiocytic neoplasms

Benign dermal neoplasms due to abnormal proliferation of fibroblasts, myofibroblasts or primitive mesenchymal cells.

2F23.0 Dermatofibroma

A common benign skin tumour which presents as a firm dermal papule or nodule, most commonly on the lower limbs. Histologically it is characterised by coarse, haphazardly arranged collagen bundles and a variable cellular infiltrate including fibrocytes.

2F23.Y Other specified benign dermal fibrous or fibrohistiocytic neoplasms

2F24 Benign cutaneous neoplasms of neural or nerve sheath origin

2F25 Cherry angioma

Inclusions: Campbell de Morgan spot

Senile angioma

2F26 Lobular capillary haemangioma

Historically called pyogenic granuloma, this is a common benign proliferation of capillary blood vessels which may be induced by trauma or by certain drugs. It presents as one or more bright red papules or nodules often located around the mouth or on a terminal phalanx in relation to the nail. Bleeding, ulceration and crusting frequently occur. BRAF mutations within vascular endothelial cells may be present, indicating that this is, in at least a proportion of cases, a true neoplastic process.

Inclusions: Lobular capillary haemangioma of skin

2F2Y Other specified benign cutaneous neoplasms

2F2Z Benign cutaneous neoplasm of unspecified type

2F30 Benign neoplasm of breast

A non-metastasizing neoplasm arising from the breast parenchyma.

Exclusions: Benign neoplasm of skin of breast (BlockL3‑2F2)

Lipoma (2E80.0)

2F30.0 Tubular adenoma of breast

A benign, well circumscribed neoplasm that arises from the breast. It is composed entirely of tubular structures that contain epithelial and myoepithelial cells.

2F30.1 Lactating adenoma of breast

A tubular type adenoma of the breast in which, during pregnancy and lactation, the epithelial cells show extensive secretory changes.

2F30.2 Intraductal papilloma of breast

A benign papillary neoplasm that arises anywhere in the ductal system of the breast. It is characterised by fibrovascular structures lined by benign epithelial and myoepithelial proliferations. Intraductal breast papillomas are classified as central, when they arise in large ducts, or peripheral, when they arise in the terminal ductal lobular units.

2F30.3 Benign phyllodes tumour of breast

A usually unilateral, benign and well circumscribed biphasic neoplasm that arises from the breast. It usually affects middle-aged women. It is characterised by the presence of a double layer of epithelial cells that are arranged in clefts, surrounded by a cellular, monomorphic spindle cell mesenchymal component. Mitoses are rare. Necrotic changes may be present in large tumours.

2F30.4 Fibromatosis of breast

2F30.5 Fibroadenoma of breast

A benign tumour of the breast characterised by the presence of stromal and epithelial elements. It presents as a painless, solitary, slow growing, firm, and mobile mass. It is the most common benign breast lesion. It usually occurs in women of childbearing age. The majority of fibroadenomas do not recur after complete excision. A slightly increased risk of developing cancer within fibroadenomas or in the breast tissue of patients previously treated for fibroadenomas has been reported.

2F30.6 Extensive adenomatosis of nipple

Rare benign nipple condition presenting as pruritus, burning or pain symptoms with clinical signs showing a nipple which appears ulcerated, crusting, scaling, indurated and erthymatous. Differential diagnosis: Paget, psoriasis, etc

2F30.Y Other specified benign neoplasm of breast

2F30.Z Benign neoplasm of breast, unspecified

2F31 Benign non-mesenchymal neoplasms of uterus

Other non-malignant tumours of the uterus not detailed elsewhere.

Exclusions: Leiomyoma of uterus (2E86.0)

2F31.0 Benign non-mesenchymal neoplasm of uterus, cervix uteri

2F31.00 Cervical Intraepithelial neoplasia grade I

2F31.0Y Other specified benign non-mesenchymal neoplasm of uterus, cervix uteri

2F31.0Z Benign non-mesenchymal neoplasm of uterus, cervix uteri, unspecified

2F31.1 Benign non-mesenchymal neoplasm of uterus, corpus uteri

2F31.2 Benign non-mesenchymal neoplasms of uterus, other parts

2F32 Benign neoplasm of ovary

A non-metastasizing neoplasm that arises from the ovary. Representative examples include serous cystadenoma, mucinous cystadenoma, clear cell adenofibroma, benign Brenner tumour, thecoma, and fibroma.

Coded Elsewhere: Struma ovarii (5A02.Y)

2F32.0 Cystic teratoma

A condition of the ovary, caused by abnormal proliferation due to genetic mutations, abnormal growth or division of germ cells. This condition is characterised by a benign ovarian neoplasm, and abdominal pain, mass or swelling, or abnormal uterine bleeding, and may lead to ovarian torsion or cystic rupture. Confirmation is by imaging.

2F32.1 Ovarian fibroma

A condition of the ovary, caused by abnormal proliferation due to genetic mutations, abnormal growth or division of cells. This condition is characterised by a benign sex chord ovarian tumour. Confirmation is by imaging.

2F32.2 Meigs' Syndrome

Benign Syndrome including ovarian fibroma, ascites and pleural effusion

2F32.3 Serous ovarian cystadenoma

2F32.Y Other specified benign neoplasm of ovary

2F32.Z Benign neoplasm of ovary, unspecified

2F33 Benign neoplasm of other or unspecified female genital organs

A non-metastasizing neoplasm that arises from the female reproductive system. Representative examples include uterine corpus leiomyoma, endocervical polyp, and benign ovarian germ cell tumour.

2F33.0 Vulvar intraepithelial neoplasia, grade I, usual type, HPV-associated

A condition of the vulva, characterised by lesion of the squamous vulvar intraepithelial cells, leading to dysplasia, varying degrees of atypia, and carcinoma in situ of the cells. This condition is associated with smoking and immunosuppression, chronic vulvar irritation, or infection with human papillomavirus or herpes simplex virus type 2. Confirmation is by tissue biopsy.

Exclusions: Vulvar intraepithelial neoplasia grade II-III, usual type, HPV-associated (2E67.10)

Vulvar intraepithelial neoplasia grade III, simplex, differentiated type, non-HPV associated (2E67.10)

Vulvar intraepithelial neoplasia, HPV independent (2F33)

2F33.1 Vaginal intraepithelial neoplasia grade I

A condition characterised by lesions of the squamous vaginal intraepithelial cells, leading to dysplasia, varying degrees of atypia, and abnormal cell growth within one third of the vaginal skin. This condition may be associated with human papillomavirus infection. Confirmation is by the Papanicolaou smear test followed by a biopsy of any abnormal cell growth.

2F33.Y Benign neoplasm of other specified female genital organs

2F33.Z Benign neoplasm of unspecified female genital organs

2F34 Benign neoplasm of male genital organs

A non-metastasizing neoplasm that arises from the male reproductive system. Representative examples include benign prostate phyllodes tumour, benign Sertoli cell tumour, seminal vesicle cystadenoma, and epididymal adenomatoid tumour.

2F35 Benign neoplasm of urinary organs

A non-metastasizing neoplasm that arises from the organs that comprise the urinary system. Representative examples include renal oncocytoma, bladder inverted papilloma, and urothelial papilloma.

2F36 Benign neoplasm of eye or ocular adnexa

Exclusions: Benign neoplasm of optic nerve (2A02.3)

Benign neoplasm of skin of eyelid (BlockL3‑2F2)

Coded Elsewhere: Conjunctival haemangioma or haemolymphangioma (2E81.01)

Seborrhoeic keratosis (2F21.0)

2F36.0 Benign neoplasm of uvea

Abnormal growth of the cells of the choroid without malignant characteristics.

Coded Elsewhere: Haemangioma of choroid (2E81.0Y)

2F36.1 Benign neoplasm of iris

2F36.2 Benign neoplasm of ciliary body

2F36.3 Teratoma of orbit

This is an encapsulated tumour with tissue or organ components resembling normal derivatives of all three germ layers. This diagnosis is of the cavity or socket of the skull in which the eye and its appendages are situated.

2F36.4 Cysts of eyelid

Coded Elsewhere: Epidermoid cyst (EK70.0)

2F36.Y Other specified benign neoplasm of eye or ocular adnexa

2F36.Z Benign neoplasm of eye or ocular adnexa, unspecified

2F37 Benign neoplasm of endocrine glands

Exclusions: Benign neoplasm of endocrine pancreas (2E92.9)

Benign neoplasm of thymus (2F01)

Benign neoplasm of ovary (2F32)

Benign neoplasm of testis (2F34)

Benign neoplasm of hypothalamus (2A00.5)

2F37.0 Non-secreting pituitary adenoma

2F37.Y Other specified benign neoplasm of endocrine glands

2F37.Z Benign neoplasm of endocrine glands, unspecified

2F3Y Benign non-mesenchymal neoplasms of other specified site

2F3Z Benign non-mesenchymal neoplasms of unspecified site

2F5Y Other specified benign neoplasms, except of lymphoid, haematopoietic, central nervous system or related tissues

2F5Z Benign neoplasms, except of lymphoid, haematopoietic, central nervous system or related tissues, unspecified

Neoplasms of uncertain behaviour, except of lymphoid, haematopoietic, central nervous system or related tissues (BlockL1‑2F7)

A neoplasm displaying morphologic, phenotypic, or genotypic characteristics that are clearly not benign but do not permit the establishment of a definitive diagnosis of malignancy. Such neoplasms may or may not eventually have a more aggressive clinical course. Representative examples include lymphoproliferations of uncertain malignant potential (e.g., lymphomatoid granulomatosis and lymphomatoid papulosis), borderline ovarian epithelial neoplasms (e.g., borderline ovarian endometrioid tumour and borderline ovarian mucinous tumour), borderline exocrine pancreatic neoplasm (e.g., pancreatic borderline intraductal papillary-mucinous neoplasm), and primary borderline peritoneal epithelial neoplasm.

2F70 Neoplasms of uncertain behaviour of oral cavity or digestive organs

2F70.0 Neoplasms of uncertain behaviour of lip, oral cavity or pharynx

2F70.1 Neoplasms of uncertain behaviour of stomach

2F70.2 Neoplasms of uncertain behaviour of small intestine

2F70.3 Neoplasms of uncertain behaviour of colon

2F70.4 Neoplasms of uncertain behaviour of rectum

2F70.5 Neoplasms of uncertain behaviour of liver, gallbladder or bile ducts

2F70.Y Neoplasms of uncertain behaviour of oral cavity and digestive organs, other specified site

2F70.Z Neoplasms of uncertain behaviour of oral cavity and digestive organs, unspecified site

2F71 Neoplasms of uncertain behaviour of middle ear, respiratory or intrathoracic organs

2F71.0 Neoplasms of uncertain behaviour of thymus

2F71.1 Neoplasms of uncertain behaviour of larynx

2F71.2 Neoplasms of uncertain behaviour of pleura

2F71.3 Neoplasms of uncertain behaviour of trachea, bronchus or lung

2F71.4 Neoplasms of uncertain behaviour of mediastinum

2F71.Y Neoplasms of uncertain behaviour of middle ear, respiratory and intrathoracic organs, other specified site

2F71.Z Neoplasms of uncertain behaviour of middle ear, respiratory and intrathoracic organs, unspecified site

2F72 Neoplasms of uncertain behaviour of skin

2F72.0 Keratoacanthoma

Keratoacanthoma is a relatively common keratinocytic epidermal tumour which shows resemblances to squamous cell carcinoma of the skin, from which it may be difficult to distinguish either clinically or histopathologically. It is characterised by rapid growth over a few weeks to months, followed by spontaneous resolution over 4-6 months. Because it is not possible to predict its benign behaviour with complete certainty during its initial growth phase, the designation "Well-differentiated squamous cell carcinoma (keratoacanthoma type)" is also used.

2F72.1 Spitzoid tumour of uncertain malignant potential

A spindle cell and epithelioid cell melanocytic neoplasm in which there are sufficient features distinguishing it from a benign Spitz naevus to cast doubt on its benign nature. These atypical features include development in adult life, asymmetry, large diameter (>6 and especially >10 mm), significant thickness (particularly subcutaneous extension), lack of “maturation” and nodule formation, cytological atypia and a high mitotic rate.

2F72.2 Melanocytic naevus with severe melanocytic dysplasia

Melanocytic naevus with severe melanocytic dysplasia is a histopathological diagnosis based on the presence of severe cytological atypia, defined as enlarged, spindle- and epithelioid-shaped melanocytes with hyperchromatic nuclei (typically at least twice the size of those of basal keratinocytes) and distinct nucleoli. Such naevi tend to be irregular in size and pigmentation and to have been excised because of concern that they may represent early melanoma.

2F72.Y Other specified neoplasms of uncertain behaviour of skin

2F73 Neoplasms of uncertain behaviour of retroperitoneum

2F74 Neoplasms of uncertain behaviour of peritoneum

2F75 Neoplasms of uncertain behaviour of breast

2F76 Neoplasms of uncertain behaviour of female genital organs

2F77 Neoplasms of uncertain behaviour of male genital organs

2F78 Neoplasms of uncertain behaviour of urinary organs

2F79 Neoplasms of uncertain behaviour of eye or ocular adnexa

2F7A Neoplasms of uncertain behaviour of endocrine glands

2F7A.0 Multiple polyglandular tumours

Coded Elsewhere: Carney complex (5A70.Y)

Von Hippel-Lindau disease (5A75)

2F7A.Y Other specified neoplasms of uncertain behaviour of endocrine glands

2F7A.Z Neoplasms of uncertain behaviour of endocrine glands, unspecified

2F7B Neoplasms of uncertain behaviour of bone or articular cartilage

2F7C Neoplasms of uncertain behaviour of connective or other soft tissue

2F7Y Neoplasms of uncertain behaviour of other specified site

2F7Z Neoplasms of uncertain behaviour of unspecified site

Neoplasms of unknown behaviour, except of lymphoid, haematopoietic, central nervous system or related tissues (BlockL1‑2F9)

2F90 Neoplasms of unknown behaviour of oral cavity or digestive organs

2F90.0 Neoplasms of unknown behaviour of colon

2F90.1 Neoplasms of unknown behaviour of rectum

2F90.Y Neoplasms of unknown behaviour of oral cavity and digestive organs, other specified site

2F90.Z Neoplasms of unknown behaviour of oral cavity and digestive organs, unspecified site

2F91 Neoplasms of unknown behaviour of middle ear, respiratory or intrathoracic organs

2F91.0 Neoplasms of unknown behaviour of larynx

2F91.1 Neoplasms of unknown behaviour of trachea, bronchus or lung

2F91.Y Neoplasms of unknown behaviour of other specified middle ear, respiratory or intrathoracic organ

2F92 Neoplasms of unknown behaviour of skin

2F93 Neoplasms of unknown behaviour of retroperitoneum

2F94 Neoplasms of unknown behaviour of peritoneum

2F95 Neoplasms of unknown behaviour of breast

2F96 Neoplasms of unknown behaviour of female genital organs

2F97 Neoplasms of unknown behaviour of male genital organs

2F98 Neoplasms of unknown behaviour of urinary organs

2F99 Neoplasms of unknown behaviour of eye or ocular adnexa

2F9A Neoplasms of unknown behaviour of endocrine glands

2F9B Neoplasms of unknown behaviour of bone or articular cartilage

2F9C Neoplasms of unknown behaviour of connective or other soft tissue

2F9Y Neoplasms of unknown behaviour of other specified site

2F9Z Neoplasms of unknown behaviour of unspecified site