Classification of haematological malignancies based on ICD-O-3.2 (to be used for new registrations from 2020)

Haematological malignancies with malignant behaviour /3	Classification ICD-O-3.2	Topography and grade	COMMENTS
Mature lymphoid neoplasms			
Hodgkin lymphomas (HL)			
Hodgkin lymphoma, nodular lymphocyte predominant (NLPHL)	9659/3		
Classical Hodgkin lymphoma (cHL)			
Hodgkin lymphoma, nodular sclerosis, NOS (NS-cHL)	9663/3		
Hodgkin lymphoma, nodular sclerosis, cellular phase	9664/3		
Hodgkin lymphoma, nodular sclerosis, grade 1	9665/3		
Hodgkin lymphoma, nodular sclerosis, grade 2	9667/3		
Hodgkin lymphoma, mixed cellularity (MC-cHL)	9652/3		
Hodgkin lymphoma, lymphocyte-rich (LR-cHL)	9651/3		
Hodgkin lymphoma, lymphocyte depletion (LD-cHL)	9653/3		
Hodgkin lymphoma, NOS	9650/3 *(This code should be avoided)*		
Mature non-Hodgkin B-cell neoplasms			
Mature B-cell leukaemias and related lymphomas			
B-cell chronic lymphocytic leukaemia (CLL) / small lymphocytic lymphoma (SLL)	9823/3	Distinction between leukaemia and lymphoma with topography.[i]	
Other mature B-cell leukaemias			
B-cell prolymphocytic leukaemia	9833/3		
Hairy cell leukaemia	9940/3		**"BRAF V600E"**
Immunoproliferative diseases			
Waldenström macroglobulinemia	9761/3	C42.0	
Lymphoplasmacytic lymphoma	9671/3		
Heavy chain disease, NOS *(This code also includes Mu or Gamma or Alpha heavy chain disease)*	9762/3		
Immunoproliferative small intestinal disease	9764/3		
Immunoproliferative disease, NOS	9760/3 *(This code should be avoided)*		
Plasma cell neoplasms			
Plasma cell myeloma (PCM)	9732/3		
Plasma cell leukemia	9733/3		
Plasmacytoma, NOS *(This code also includes Solitary plasmacytoma of bone)*	9731/3	C40._-C41._	
Plasmacytoma, extramedullary	9734/3		
Marginal zone lymphomas			
Splenic marginal zone lymphoma (SMZL) *(This code also includes splenic lymphoma with villous lymphocytes)*	9689/3	C42.2	
Other marginal zone lymphoma (nodal / extranodal) *(This code also includes Mucosal-associated lymphoid tissue (MALT) lymphoma* *Broncus-associated lymphoid tissue (BALT) lymphoma* *Skin-associated lymphoid tissue (SALT) lymphoma* *Extranodal marginal zone lymphoma* *Nodal marginal zone lymphoma* *Primary choroidal lymphoma)*	9699/3	C69.3 for primary choroidal lymphoma	**"NMZL" for Nodal marginal zone lymphoma** **"MALT" for Extra nodal marginal zone lymphoma**

Follicular lymphoma and related lymphoma			
Follicular lymphoma (FL)			
Follicular lymphoma, NOS *(This code also includes Follicular lymphoma, pediatric type and Follicular lymphoma, testicular type)*	9690/3	C62.9 for follicular lymphoma, testicular type	**Ped-FL for Follicular lymphoma, pediatric type**
Follicular lymphoma, grade 1 *(This code also includes Follicular lymphoma, duodenal type)*	9695/3	C17.0 for Follicular lymphoma, duodenal type	
Follicular lymphoma, grade 2	9691/3		
Follicular lymphoma, grade 3A or Follicular lymphoma, grade 3B *(This code also includes Large B-cell lymphoma with IRF4 rearrangement)*	9698/3		**"Gr3A" for Follicular lymphoma, grade 3A , "Gr3B" for Follicular lymphoma, grade 3B, "LBCL IRF4+" pour Large B-cell lymphoma with IRF4 rearrangement**
Primary cutaneous follicle centre lymphoma	9597/3	C44._	
Mantle cell lymphoma (MCL)	9673/3		
Diffuse large B-cell lymphoma and related large B-cell lymphomas			
Diffuse large B-cell lymphoma (DLBCL), NOS *(This code also includes DLBCL germinal centre B-cell (GCB) subtype, DLBCL activated B-cell (ABC) subtype, Primary DLBCL of CNS, Primary cutaneous DLBCL, leg type, Vitroretinal lymphoma, EBV-positive DLBCL, DLBCL associated with chronic inflammation High grade B-cell lymphoma with MYC and BCL2 and/or BCL6 rearrangements High grade B-cell lymphoma, NOS)*	9680/3	C70._-C71._-C72._ for primary DLBCL of CNS Primary cutaneous DLBCL, leg type can have another location than the leg (C44.7) C69 for vitroretinal lymphoma	**"GCB" for DLBCL GCB, "ABC" for DLBCL ABC, "PCNSL" for Primary DLBCL of CNS, "leg type" for Primary cutaneous DLBCL, leg type, "EBV+" for EBV-positive DLBCL. For High grade B-cell lymphoma with or without MYC / BCL2 / BCL6 rearrangements: "HGBL MYC & BCL2" or "HGBL MYC & BCL6" or "HGBL MYC & BCL2 & BCL6" or "HGBL NOS" or "HGBL BCL2" or "HGBL BCL6" or "HGBL BCL2 & BCL6".**
Other related large B-cell lymphomas			
T-cell/histiocyte rich large B-cell lymphoma	9688/3		
Primary mediastinal large B-cell lymphoma	9679/3	C38.3	
ALK-positive large B-cell lymphoma	9737/3		
Lymphomatoid granulomatosis, grade 3	9766/3		
Intravascular large B-cell lymphoma	9712/3	C49._	
Primary effusion lymphoma (PEL)	9678/3		
Plasmablastic lymphoma	9735/3		
HHV8-positive diffuse large B-cell lymphoma	9738/3		
Burkitt lymphoma / leukaemia (BL) *(This code also includes Burkitt-like lymphoma with 11q aberration)*	9687/3	Distinction between leukaemia and lymphoma with topog	**"BLL 11q" for Burkitt-like lymphoma with 11q aberration**

Mature T-cell and NK-cell neoplasms			
Primary cutaneous T-cell lymphomas			
Mycosis fungoïdes / Sezary syndrome			
Mycosis fungoides (MF)	9700/3		
Sézary syndrome (SS)	9701/3		
Other primary cutaneous T-cell lymphoma			
Primary cutaneous anaplastic large cell lymphoma	9718/3	C44._	
Primary cutaneous γδ T-cell lymphoma	9726/3	C44._	
Cutaneous T-cell lymphoma, NOS *(This code also includes* *Primary cutaneous CD8 - positive aggressive epidermotropic cytotoxic T-cell lymphoma and* *Primary cutaneous acral CD8 - positive T-cell lymphoma)*	9709/3	C44._	**"aggressive" for Primary cutaneous CD8 positive aggressive epidermotropic cytotoxic T-cell lymphoma,** **"acral" for Primary cutaneous acral CD8 positive T-cell lymphoma**
Peripheral NK/T-cell lymphomas			
Nodal PNK/TCL			
Peripheral NK/T-cell lymphoma, NOS (PTCL, NOS) *(This code also includes Follicular T-cell lymphoma,* *Nodal peripheral T-cell lymphoma with T follicular helper phenotype,* *lymphoepithelioid lymphoma,* *Peripheral T-cell lymphoma, NOS or Mature T-cell lymphoma, NOS)*	9702/3		**"PTCL" for Peripheral T-cell lymphoma, NOS,** **"Follicular" for Follicular T-cell lymphoma, "TFH" for Nodal peripheral T-cell lymphoma with T follicular helper phenotype**
Anaplastic large cell lymphoma (ALCL), ALK-positive *(This code also includes ALCL, NOS (ALK not tested))*	9714/3		**"NOS" if ALK no tested**
Anaplastic large cell lymphoma (ALCL), ALK-negative *(This code also includes Breast implant-associated anaplastic large cell lymphoma)*	9715/3	C50._ for Breast implant-associated anaplastic large cell lymphoma	
Angioimmunoblastic T-cell lymphoma (AILD)	9705/3		
Leukaemic PNK/TCL			
T-cell prolymphocytic leukaemia (T-PLL)	9834/3		
Adult T-cell leukaemia / lymphoma (HTLV1 positive) (ATLL)	9827/3		
T-cell large granular lymphocytic leukaemia (TLGLL) *(This code also includes chronic lymphoproliferative disorder of NK cells)*	9831/3		**"T-LGL" for T-cell large granular lymphocytic leukaemia (T-LGL),** **"NK-CLPD" for Chronic lymphoproliferative disorder of NK cells**
Systemic EBV-positive T-cell lymphoproliferative disease of childhood	9724/3		
Aggressive NK-cell leukaemia	9948/3		
Extra-nodal PNK/TCL			
Hepatosplenic T-cell lymphoma	9716/3		
Intestinal T-cell lymphoma *(This code also includes Enteropathy-associated T-cell lymphoma (EATCL) and* *Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITCL))*	9717/3		
Extranodal NK/T-cell lymphoma, nasal and nasal-type	9719/3		
Subcutaneous panniculitis-like T-cell lymphoma	9708/3		

Other lymphoid neoplasms			
B-cell lymphoma, unclassifiable, with features intermediate between DLBCL and classical Hodgkin lymphoma (DLBCL/CHL)	9596/3		
Lymphoid neoplasms, NOS			
Malignant lymphoma, NOS	9590/3 *(This code should be avoided)*	Grade and topography should be specified.	
Malignant lymphoma, non-Hodgkin, NOS *(This code also includes Hairy cell leukemia variant, Splenic B-cell lymphoma/leukemia, unclassifiable and Splenic diffuse red pulp small B-cell lymphoma)*	9591/3 *(This code should be avoided)*	Grade and topography should be specified.	**"v-HCL" for the Hairy cell leukemia variant , "SBL,NOS" for the Splenic B-cell lymphoma/leukemia, unclassifiable, "SDRPSBL" for the Splenic diffuse red pulp small B-cell lymphoma**
Lymphoid leukemia, NOS	9820/3 *(This code should be avoided)*	Grade should be specified.	
Prolymphocytic leukemia, NOS	9832/3 (*This code should be avoided)*	Grade should be specified.	

Precursor neoplasms			
Precursor lymphoid neoplasms (PLN) or lymphoblastic leukaemia (ALL) / lymphoma			
B-cell PLN or lymphoblastic leukaemia / lymphoma			
B-cell PLN with recurrent genetic abnormalities			
B-cell PLN with t(9;22)(q34.1;q11.2); *BCR-ABL1*	9812/3	Distinction between leukaemia and lymphoma with topography.	
B-cell PLN with t(v;11q23.3); *KMT2A* rearranged	9813/3	Distinction between leukaemia and lymphoma with topography.	
B-cell PLN with t(12;21)(p13.2;q22.1); *ETV6-RUNX1*	9814/3	Distinction between leukaemia and lymphoma with topography.	
B-cell PLN with Hyperdiploidy	9815/3	Distinction between leukaemia and lymphoma with topography.	
B-cell PLN with Hypodiploidy	9816/3	Distinction between leukaemia and lymphoma with topography.	
B-cell PLN with t(5;14)(q31.1;q32.1); *IGH-IL3*	9817/3	Distinction between leukaemia and lymphoma with topography.	
B-cell PLN with t(1;19)(q23;p13.3); *TCF3-PBX1*	9818/3	Distinction between leukaemia and lymphoma with topography.	
B-cell PLN, BCR-ABL1-like	9819/3	Distinction between leukaemia and lymphoma with topography.	
B-cell PLN or lymphoblastic leukaemia / lymphoma, NOS *(This code also includes B lymphoblastic leukemia/lymphoma with iAMP21)*	9811/3	Distinction between leukaemia and lymphoma with topography.	
T-cell and NK-cell PLN or lymphoblastic leukaemia / lymphoma	9837/3	Distinction between leukaemia and lymphoma with topography.	
PLN or lymphoblastic leukaemia / lymphoma, NOS and related neoplasms		Distinction between leukaemia and lymphoma with topography.	
Blastic plasmacytoid dendritic cell neoplasm *(This code also includes Precursor cell lymphoblastic lymphoma, NOS)*	9727/3	Grade and topography should be specified.	
Precursor cell lymphoblastic leukemia, NOS	9835/3 *(This code should be avoided)*	*Grade should be specified.*	
Acute myeloid leukaemias and related precursor neoplasms			
Acute myeloid leukaemias with recurrent genetic abnormalities			
AML with t(8;21)(q22;q22.1); *RUNX1-RUNX1T1*	9896/3		
AML with inv/t(16;16)(p13.1;q22); *CBFB-MYH11*	9871/3		
Acute promyelocytic leukaemia with t(15;17)(q22;q11-q12) or variant *RARA* translocation	9866/3		
AML with t(v;11q23.3); *KMT2A* rearranged	9897/3		**"KMT2A-MLLT3", "Other KMT2A", "KMT2A NOS"**
AML with t(6;9)(p23;q34.1); *DEK-NUP214*	9865/3		
AML with inv/t(3;3)(q21.3;q26.2); *GATA2, MECOM*	9869/3		
AML with t(1;22)(p13.3;q13.1); *RBM15-MKL1*	9911/3		
AML with with t(9;22)(q34.1;q11.2); *BCR-ABL1*	9912/3		
AML with mutated *NPM1*	9877/3		
AML with biallelic mutation of *CEBPA*	9878/3		
AML with mutated *RUNX1*	9879/3		
Acute myeloid leukaemias with specific conditions			
AML with myelodysplasia-related changes	9895/3		
Therapy-related myeloid neoplasm	9920/3		
Myeloid leukaemia associated with Down syndrome	9898/3		

Other AML and related precursor neoplasms			
Other AML according to the FAB classification			
AML with minimal differentiation (FAB M0)	9872/3		
AML without maturation (FAB M1)	9873/3		
AML with maturation (FAB M2)	9874/3		
Acute myelomonocytic leukaemia (FAB M4)	9867/3		
Acute monocytic leukaemia (FAB M5)	9891/3		
Acute erythroid leukaemia (FAB M6)	9840/3		
Acute megakaryoblastic leukaemia (FAB M7)	9910/3		
Acute basophilic leukaemia	9870/3		
Other related myeloid precursor neoplasms			
Acute panmyelosis with myelofibrosis	9931/3		
Myeloid sarcoma	9930/3	Topography should be specified (Never use C42.1).	
Acute myeloid leukaemias, NOS	9861/3 *(This code should be avoided)*		
Acute leukaemias of ambiguous lineage			
Acute undifferentiated leukaemia *(This code also includes Acute leukaemia, NOS)*	9801/3		**"AUL" for acute undifferentiated leukaemia based on comprehensive CMF "NOS" (justifty why)**
Mixed phenotype acute leukaemia with t(9;22)(q34.1;q11.2); *BCR-ABL1*	9806/3		
Mixed phenotype acute leukaemia with t(v;11q23.3); *KMT2A* rearranged	9807/3		
Mixed phenotype acute leukaemia B/myeloid, NOS	9808/3		
Mixed phenotype acute leukaemia T/myeloid, NOS	9809/3		
Acute biphenotypic leukaemia, NOS	9805/3 *(This code should be avoided)*		

Chronic myeloid neoplasms	
Myeloproliferative neoplasms (MPN)	
Chronic myeloid leukaemia	
Chronic myeloid leukemia; t(9;22)(q34;q11); *BCR-ABL1* positive (CML/LMC)	9875/3
Chronic myeloid leukemia, NOS	9863/3 *(This code should be avoided)*
Myeloproliferative neoplasms *BCR-ABL1* negative and related neoplasms	
Polycythaemia vera (PV)	9950/3
Essential thrombocythaemia (TE/ET)	9962/3
Primary myelofibrosis (PMF/MFP)	9961/3
Other MPN and related neoplasms	
Chronic neutrophilic leukaemia (CNL/LNC)	9963/3
Myeloid/lymphoid neoplasm with *PDGFRA* rearrangement	9965/3
Myeloid neoplasm with *PDGFRB* rearrangement	9966/3
Myeloid/lymphoid neoplasm with *FGFR1* abnormalities	9967/3
Myeloid/lymphoid neoplasm with *PCM1-JAK2*	9968/3
Chronic eosinophilic leukaemia, NOS (CEL/LCE)	9964/3
Myeloproliferative neoplasm, NOS	9960/3 *(This code should be avoided)*
Mast cell neoplasms	
Malignant mastocytosis *(This code also includes Systemic mastocytosis with an associated haematological neoplasm* *Aggressive systemic mastocytosis)*	9741/3
Mast cell leukaemia	9742/3
Mast cell sarcoma	9740/3
Myelodysplastic syndrome (MDS)	
Myelodysplastic syndrome (MDS) with single or multilineage dysplasia	
MDS with single lineage dysplasia (MDS-SLD) *(This code also includes Refractory anemia -without sideroblasts-,* *Refracrory neutropenia,* *Refractory thrombocytopenia)*	9980/3
MDS with multilineage dysplasia (MDS-MLD) *(This code also includes Refractory cytopenia of childhood)*	9985/3
MDS with ring sideroblasts	
MDS with ring sideroblasts and single lineage dysplasia (MDS-RS-SLD) *(This code also includes* *Refractory anemia with ring sideroblasts associated with marked thrombocytosis* *Myelodysplastic/myeloproliferative neoplasm with ring sideroblasts and thrombocytosis* *MDS with ring sideroblasts, NOS)*	9982/3
MDS with ring sideroblasts and multilineage dysplasia (MDS-RS-MLD)	9993/3
MDS with excess blasts (MDS-EB)	9983/3
MDS with isolated del(5q)	9986/3
MDS, NOS	9989/3

Myelodysplastic/myeloproliferative neoplasms (MDS/MPN)		
Chronic myelomonocytic leukaemia (CMML)	9945/3	
Other myelodysplastic/myeloproliferative neoplasm		
Juvenile myelomonocytic leukaemia (JMML)	9946/3	
Atypical chronic myeloid leukaemia, *BCR-ABL1* negative (aCML)	9876/3	
Myelodysplastic/myeloproliferative neoplasm, unclassifiable *(This code also includes Myeloproliferative neoplasm, unclassifiable)*	9975/3 *(This code should be avoided)*	
Other leukaemias, NOS		
Myeloid leukemia, NOS	9860/3 *(This code should be avoided)*	
Leukemia, NOS	9800/3 *(This code should be avoided)*	
Histiocytic and dendritic cell neoplasms		
Langerhans cell histiocytosis, disseminated	9751/3	
Langerhans cell sarcoma	9756/3	
Histiocytic sarcoma	9755/3	
Dendritic cell tumour *(This code also includes Indeterminate dendritic cell tumour and Interdigitating dendritic cell sarcoma)*	9757/3	**"indeterminate" for Indeterminate dendritic cell tumour, "interdigitating" for Interdigitating dendritic cell sarcoma**
Follicular dendritic cell sarcoma	9758/3	
Fibroblastic reticular cell tumour	9759/3	
Erdheim-Chester disease	9749/3	
Malignant histiocytosis, NOS	9750/3 *(This code should be avoided)*	

Hematological malignancies with associated conditions	Classification ICD-O-3.2	
Myeloid neoplasms with germline predisposition	To be classified according to the neoplasm to which they correspond with the respective ICD-O3 code	**"GL Ohter" If Myeloid neoplasms with other germline predisposition than those listed below**
Myeloid neoplasms with germline predisposition without a pre-existing disorder or organ dysfunction		
AML with germline *CEBPA* mutation		**"GL CEPBA"**
Myeloid neoplasms with germline *DDX41* mutation		**"GL DDX41"**
Myeloid neoplasms with germline predisposition and pre-existing platelet disorders		
Myeloid neoplasms with germline *RUNX1* mutation		**"GL RUNX1"**
Myeloid neoplasms with germline *ANKRD26* mutation		**"GL ANKRD26"**
Myeloid neoplasms with germline ETV6 mutation		**"GL ETV6"**
Myeloid neoplasms with germline predisposition associated with other organ dysfunction		
Myeloid neoplasms with germline GATA2 mutation		**"GL GATA2"**
Myeloid neoplasms with germline predisposition associated with inherited bone marrow failure syndromes and telomere biology disorders		**"GL BMF" "GL TELO" or "GL name of mutated gene: TERC, TERT,..."**
Immunodeficiency-associated lymphoproliferative disorders		
Lymphoproliferative diseases associated with primary immune disorders (PID)		**"PID"**
Lymphomas associated with HIV infection		**"HIV"**
Post-transplant lymphoproliferative disorders (PTLD), Monomorphic		**"PTLD"**
Other iatrogenic immunodeficiency-associated lymphoproliferative disorders		**"Other iatrogenic ID"**

Haematological disorders with uncertain behaviour /1	Classification ICD-O-3.2	Topography	
Mature lymphoid disorders			
Mature B-cell disorders			
Monoclonal B-cell lymphocytosis, CLL type	9823/1		
Monoclonal B-cell lymphocytosis, NOS / non-CLL-type	9591/1		
Non-IgM monoclonal gammopathy of undetermined significance *(This code also includes monoclonal gammopathy of undetermined significance, NOS)*	9765/1		
IgM monoclonal gammopathy of undetermined significance	9761/1		
Immunoglobulin deposition disease / Primary amyloidosis / Systemic light chain disease	9769/1		***"amyloidosis" for Primary amyloidosis, "LC deposition" for Systemic light chain disease***
In situ follicular neoplasia	9695/1		
In situ mantle cell neoplasia	9673/1		
EBV positive mucocutaneous ulcer	9680/1		
Angiocentric immunoproliferative lesion / Lymphomatoid granulomatosis, NOS (grade 1 or grade 2)	9766/1		
Polymorphic post-transplant lymphoproliferative disorder, NOS	9971/1		
Lymphoproliferative disorder, NOS	9970/1		
Mature T-cell and NK-cell disorders			
Hydroa vacciniforme-like lymphoproliferative disorder	9725/1		
Angioimmunoblastic lymphadenopathy	9767/1		
Indolent T-cell lymphoproliferative disorder of gastrointestinal tract	9702/1		
Primary cutaneous CD30 positive T-cell lymphoproliferative disorder/ Lymphomatoid papulosis	9718/1	(C44._)	
Primary cutaneous CD4 positive small/medium T-cell lymphoproliferative disorder	9709/1	(C44._)	
T-gamma lymphoproliferative disease	9768/1		
Chronic myeloid disorders			
Transient abnormal myelopoiesis	9898/1		
Mastocytoma, NOS	9740/1		
Indolent systemic mastocytosis	9741/1		
Histiocytic and dendritic cell disorders			
Langerhans cell histiocytosis, NOS / monostotic / polystotic	9751/1		**"NOS" for Langerhans cell histiocytosis, NOS, "monostatic" for Langerhans cell histiocytosis monostotic, "polystotic" for Langerhans cell histiocytosis polystotic**

[i] *For the correct classification of the following malignancies, additional inclusion criteria are applied based on topography:*

- The ICD-O-3 histology codes corresponding with "B-cell chronic lymphocytic leukaemia", "Burkitt leukaemia" and "lymphoblastic leukaemia" are combined with topography codes C42.1.

- The ICD-O-3 histology codes corresponding with "Small lymphocytic lymphoma", "Burkitt lymphoma" and "lymphoblastic lymphoma" are combined with all topography codes other than C42.0-C42.1.

In grey codes and / or names of entities which should be avoided in favor of a more specific code

The other ICD-O-3 codes which are not mentioned in this list should not be used for registration of new diagnoses ov haematological malignancy.