Belgian Cancer Registry

Registratie van hematologische maligniteiten: Documentatie

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Staging van de lymfomen
(Hodgkin en non-Hodgkin behalve cutane lymfomen)
Ann-Arbor (geen TNM)

Table 5.5.1. - Ann Arbor staging system classification for lymphomas (Cotswolds revision)[11,12]

Stage	Definition
I	Involvement of a single lymph node region or lymphoid structure (e.g., spleen, thymus, or Waldeyer's ring)
II	Involvement of two or more lymph node regions on the same side of the diaphragm (mediastinum is a single site; hilar lymph nodes are lateralized); the number of anatomic sites should be indicated by suffix (e.g., II_3)
III	Involvement of lymph node regions or structures on both sides of the diaphragm
III_1	With or without splenic, hilar, celiac or portal hepatic nodes
III_2	With para aortic, iliac or mesenteric nodes
IV	Involvement of extranodal site(s) beyond these designated E (extranodal)
Annotations	
A	No symptoms
B	Fever, drenching sweats or weight loss
X	Bulky disease: >1/3 width of mediastinum at T5-6, or >10 cm
E	Involvement of a single extranodal site contiguous with or proximal to the known nodal site of disease
CS	Clinical stage
PS	Pathologic stage

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Staging van myelomen
Salmon-Dury-classificatie

Stage	Needed criteria	Hb (g/dl)	Pic IgG (g/l)	Pic IgA (g/l)	PBJ (g/24h)	Ca (mg/l)	Bone lesions	Estimation of the tumoral mass (cell number)
I	Alle	> 10	< 50	< 30	< 4	<= 120	≤ 1	$< 0{,}6.10^{12}$
II	Noch I noch III							$\geq 6.10^{12}$
III		< 8,5	> 70	> 50	> 12	> 120	meerdere	$>1{,}2.10^{12}$

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Staging van myelomen
International staging system (ISS)

ISS stage	Factors
I	Serum β_2-microglobulin < 3.5 mg/L AND Serum albumin ≥ 3.5 g/dL
II	Not ISS stage I or III
III	Serum β_2-microglobulin ≥ 5.5 mg/L

RISS Stage Group	Factors
I	Serum β_2-microglobulin < 3.5 mg/L AND Serum albumin ≥ 3.5 g/dL AND Cytogenetics are considered “not high risk” * AND LDH levels are normal
II	Not RISS stage I or III
III	Serum bβ_2-microglobulin ≥ 5.5 mg/L AND Cytogenetics are considered “high-risk”* AND/OR LDH levels are high

High risk cytogenetics :
(detected by iFISH)

-del(17p)
AND/OR
-translocation t(4;14)
AND/OR
-translocation t(14;16)

Gedrag borderline /1

	ICD-O-3 (/1)	Label New entities with classification of 2016/2017	Comments
Myeloid malignancies			
Chronic myeloid disorders			
Mastocytosis / Mast cell diseases	9740/1	Cutaneous mastocytosis	
	9741/1	Indolent systemic mastocytosis	
Acute myeloid leukemias (AML) and related precursor neoplasms			
Myeloid proliferations associated with Down	9898/1	transient abnormal myelopoiesis associated with Down syndrome	
Lymphoid malignancies			
Mature B-cell neoplasms			
Mature B-cell leukemias	9823/1	Monoclonal B-cell lymphocytosis (MBL), CLL-type	
	9823/1	Monoclonal B-cell lymphocytosis (MBL), non-CLL-type	9591/1
Immunoproliferative diseases / Waldenström macroglobulinaemia	9761/1	IgM monoclonal gammopathy of undetermined significance	
Plasma cell neoplasms (PCN)	9765/1	non-IgM monoclonal gammapathy of undetermined significance (MGUS)	
	9769/1	Primary amyloidosis	"amyloidosis"
	9769/1	Light chain and heavy chain deposition diseases	"LC/HV deposition"
Follicular lymphoma (FL)	9695/1	In situ follicular neoplasia	
Mantle cell lymphoma	9673/1	In situ mantle cel neoplasia	
Diffuse Large B-cell lymphoma (DLBCL) and related	9680/1	EBV-positive mucocutaneous ulcer	
	9766/1	Lymphomatoid granulomatosis, grade 1, 2	
		Multicentric Castelman disease (MCD)	"MCD"
	9738/1	HHV8-positive germinotropic lymphoproliferative disorder	
Mature T-cell and NK-cell neoplasms			
Leukemic presentation		Chronic active EBV infection of T- and NK-cell type, systemic form	"CAEBVTNK"
	9725/1	Hydroa vacciniforme-like lymphoproliferative disorder	"Hydroa"
		Severe mosquito bite allergy	"Mosquito bite A"
Extra-nodal non cutaneous presentation	9702/1	Indolent T-cell lymphoproliferative disorder of the gastrointestinal tract	(cf topo code)
Cutaneous presentation	9718/1	Lymphomatoid papulosis	
	9709/1	Primary cutaneous CD4-positive small/medium T-cell lymphoproliferative disorder	
Immunodeficiency-associated lymphoproliferative disorders			
Post-transplant lymphoproliferative disorders (PTLD)		Florid follicular hyperplasia	"FH"
	9971/1	Polymorphic Post transplant lymphoproliferative disorder (PTLD)	
Histiocytic and dendritic cell neoplasms			
Histiocytic and dendritic cell neoplasms	9751/1	Langerhans cell histiocytosis, NOS	"NOS"
	9751/1	Langerhans cell histiocytosis,monostatic	"monostatic"
	9751/1	Langerhans cell histiocytosis, polystatic	"polystatic"
		Disseminated juvenile xanthogranuloma	"DJX"

Hematologische maligniteiten in de context van een immunodeficiëntie

ICD-O-3 (/3)	Label	Commentaire
classified according to the lymphoid neoplasm (including Hodgkin) to which they correspond → assign the respective ICD-O code	Lymphoproliferative diseases associated with primary immune disorders	"PID"
	Lymphomas associated with HIV infection	"HIV"
	Monomorphic PTLD (B- and T/NK-cell types)	"PTLD"
	Other iatrogenic immunodeficiency-associated lymphoproliferative disorders	"Other iatrogenic ID"

Nieuwe entiteiten 2017 met bestaande codes

ICD-O-3 (/3)	New entities 2016/2017 with existing codes	Comments
9680	*DLBCL (NOS, CNS, leg-type, EBV+)*	
9680	High Grade B-cell lymphoma with MYC and BCL2 and/or BCL6 rearrangements	"HGBCL MYC & BCL2" "HGBCL MYC & BCL6" "HGBCL MYC & BCL2 & BCL6"
9680	High Grade B-cell lymphoma, NOS	"HGBCL NOS" "HGBCL BCL2" "HGBCL BCL6" "HGBCL BCL2 & BCL6"
9687	*Burkitt lymphoma/leukemia*	
9687	Burkitt-like lymphoma with 11q aberration	"BLL 11q"
9698	*Follicular lymphoma*	
9698	Large B-cell lymphoma with IRF4 rearrangement	"LBCL IRF4+"
9699	*Marginal zome lymphoma (nodal, extranodal)*	*Cf C*
9699	Paediatric nodal MZL	"Ped NMZL"
9831	*T-cell large franular lymphocytic leukemia*	
9831	Chronic lymphoproliferative disorder of NK cells	"NK-CLPD"
9985	*MDS with multilineage dysplasia*	
9985	Refractory cytopenia of childhood	"Ped RC"

Entiteiten met nieuwe codes in 2017 → voor 2019

ICD-O-3 (/3)	New entities 2016/2017 with new codes	Comments
9702	Anaplastic large cell lymphoma (ALCL), *ALK*-negative	9715
9702	Breast implant-associated anaplastic large cell lymphoma	9715 C50
9766	Lymphomatoid granulomatosis, grade 3	
9811	B-lymphoblastic leukaemia/lymphoma *BCR-ABL1*-like	9819
9861	AML with mutated *NPM1*	9877
9861	AML with biallelic mutation of *CEBPA*	9878
9861	AML with mutated *RUNX1*	9879
9861	AML with t(9;22)(q34.1;q11.2); *BCR-ABL1*	9912
classified according to the myeloid neoplasms to which they correspond → assign the respective ICD-O code	Myeloid and lymphoid neoplasms with *PCM1-JAK2*	9968
	Erdheim-Chester disease	9749

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AML with recurrent genetic abnormalities

ICD-O-3 (/3)	New entities 2016/2017 without codes		Comments
	Cytogenetics		
9865	AML with t(6;9)(p23;q34); DEK-NUP214		
9866	Acute promyelocytic leukemia t(15;17) (q22;q11-12) ; PML/RARA (FAB M3)		
9869	AML with inv(3)(q21.3q26.2) or t(3;3)(q21.3;q26.2); MECOM/GATA2 (MECOM=EVI1) (RPN1-EVI1)		
9871	AML with inv(16)(p13.1q22) or t(16;16)(p13.1q22); CBFbeta-MYH11 (FAB M4Eo / Acute myelomonocytic leukemia with abnormaleosinophils)		
9896	AML with t(8,21)(q22,q22.1) ; RUNX1-RUNX1T1 (RUNX1=AML1=CBFalpha) (RUNX1T1=ETO)		
9897	AML with t(9;11)(p21.3;q23.3); KMT2A-MLLT3 (KMT2A=MLL)		
9897	AML with 11q23/MLL/KMT2A anomalies		"Other KMT2A" "KMT2A NOS"
9911	AML (megakaryoblastic) with t(1;22)(p13.3;q13.1); RBM15-MKL1		
	Gene mutations (new entities 2017 → 2019)		
9861	AML with t(9;22)(q34.1;q11.2); BCR-ABL1		9912
9861	AML with mutated NPM1		9877
9861	AML with biallelic mutation of CEBPA		9878
9861	AML with mutated RUNX1		9879

Nieuwe entiteiten 2017 zonder code Myeloïde HM met kiemlijnmutatie

ICD-O-3 (/3)	New entities with classification of 2016/2017 without codes	commentaires
9861	AML with germline CEBPA mutation	"GL CEPBA"
classified according to the myeloid neoplasms to which they correspond → assign the respective ICD-O code	Myeloid neoplasms with germline DDX41 mutation	"GL DDX41"
	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 GATA2 mutation	"GL GATA2"
	Myeloid neoplasms with germline predisposition associated with inherited bone marrow failure syndromes and telomere biology disorders	"GL BMF" "GL TELO" ou "GL nom du gène : TERC, TERT,..."

Entiteiten zonder specifieke codes : Acute leukemieën

ICD-O-3 (/3)	Label		
9805	Mixed-phenotype acute leukaemia, NOS, rare types	inclut : "MPAL B/T"	
9805	Acute leukaemias of ambiguous lineage, NOS	inclut "Ind AL" (indifférenciée)	
9727	NK-lymphoblastic leukaemia/lymphoma	"NK ALL/LL"	*A différencier de :* *- blastic plasmacytoid dendritic cell neoplasm - 9727* *- NK/T cell lymphoma, nasal et nasal-type 9719* *- NK/T LGL - 9831*

Entiteiten zonder specifieke codes: Plasmocytaire neoplasmen

ICD-O-3	Label	
classified according to the plasma cell neoplasms to which they correspond → assign the respective ICD-O code	POEMS Syndrome	"POEMS"
	TEMPI syndrome	"TEMPI"

Verschillende entiteiten voor dezelfde code: agressieve lymfomen

ICD-O-3 (/3)	Label	Comments
9680	Diffuse Large B-cell lymphoma (DLBCL), NOS	
9680	Germinal center B-cell subtype	"GCB"
9680	Activated B-cell subtype	"ABC", "non-GCB"
9680	Primary DLBCL of the CNS	"PCNSL" (+ code Topo)
9680	Primary cutaneous DLBCL, leg type [not only on legs]	"leg-type" (+ code Topo)
9680	EBV+ DLBCL, NOS	"EBV+"
9680	DLBCL associated with chronic inflammation Fibrin-associated DLBCL	
9680	High Grade B-cell lymphoma with MYC and BCL2 and/or BCL6 rearrangements	"HGBCL MYC & BCL2" "HGBCL MYC & BCL6" "HGBCL MYC & BCL2 & BCL6"
9680	High Grade B-cell lymphoma, NOS	"HGBCL NOS" "HGBCL BCL2" "HGBCL BCL6" "HGBCL BCL2 & BCL6"

Eosinofiele neoplasmen

ICD-O-3 (/3)	Label	Comments
9965	Myeloid and lymphoid neoplasms with *PDGFRA* rearrangement	
9966	Myeloid and lymphoid neoplasms with *PDGFRB* rearrangement	
9967	Myeloid and lymphoid neoplasms with *FGFR1* rearrangement	
classified according to the myeloid neoplasms to which they correspond → assign the respective ICD-O code	Myeloid and lymphoid neoplasms with *PCM1-JAK2*	9968
9964	Chronic eosinophilic leukemia, NOS	**Bij verstek !**

'Hairy cell' - leukemie

ICD-O-3 (/3)	Label	Comments
9940	**Hairy cell leukaemia**	Si mutation: "BRAF V600E"
9591	**Hairy cell leukaemia variant**	"v-HCL"
9591	*Splenic B-cell leukaemia/lymphoma, unclassifiable*	"SBL,NOS"
9591	*Splenic diffuse red pulp small B-cell lymphoma*	"SDRPSBL"

Nuttige afkortingen: check codes

AITL = LAI	AngioImmunoblastic T-cell Lymphoma, Lymphome Angio-Immunoblastique	9705/3
ALCL	Anaplastic Large Cell Lymphoma	9714/3 if ALK+ 9702/3 if ALK-
ALL = LLA	Acute Lymphoid Leukaemia, Leucémie Lymphoïde Aiguë	9835/3
AML = LMA	Acute Myeloid Leukaemia, Leucémie Myéloïde Aiguë	9861/3
ANLL	Acute Non-Lymphocytic Leukaemia	9861/3
c-ALCL	Cutaneous ALCL	9718/3
CLL = LLC	Chronic Lymphoid Leukaemia, Leucémie Lymphoïde Chronique	9823/3
CML = LMC	Chronic Myeloid Leukaemia, Leucémie Myéloïde Chronique	9863/3
CMML =LMMC	Chronic MyeloMonocytic Leukaemia, Leucémie MyéloMonocytaire Chronique	9945/3
DLBCL	Diffuse Large B-Cell Lymphoma	9680/3
ET = TE	Essential Thrombocythemia, Thrombocythémie Essentielle	9962/3
HCL	Hairy Cell Leukaemia	9940/3
HL	Hodgkin lymphoma	9650/3
LRCHL	Lymphocyte-Rich Classical Hodgkin Lymphoma	9651/3
MBL	Monoclonal B-cell Lymphocytosis (CLL-type MBL)	Not to be registered*
MCL	Mantle Cell Lymphoma	9673/3
MDS = SMD	MyeloDysplastic Syndrome, Syndrome MyéloDysplasique	9989/3

Nuttige afkortingen: check codes

MF	Mycosis Fungoides	9700/3
MPN	MyeloProliferative Neoplasms	9960/3
MPS = SMP	MyeloProliferative Syndrome, Syndrome MyéloProlifératif	
MZL	Marginal Zone Lymphoma	9699/3
NHL = LNH	Non-Hodgkin Lymphoma, Lymphome malin Non-Hodgkinien	9591/3
PTLD	Post-Transplant Lymphoproliferative Disorder - Polymorphic (/1 → not to be registered) - Monomorphic (/3 → to be registered)	9971/X
PV	Polycythemia Vera (= Vaquez)	9950/3
RA = AR	Refractory Anemia, Anémie Réfractaire	9980/3
RAEB	Refractory Anemia with Excess Blasts, Anémie Réfractaire avec Excès de Blastes	9983/3
RARS	Refractory Anemia with Ringed Sideroblasts, Anémie Réfractaire avec Sidéroblastes en Couronne	9982/3
RCMD	Refractory Cytopenia with Multilineage Dysplasia, Cytopénie Réfractaire avec Dysplasie Multilignée	9985/3
SLVL	Splenic Lymphoma with Villous Lymphocytes	9689/3
T-LGL = LGL-T	T-cell Large Granular Lymphocytic Leukaemia, Leucémie à Grands Lymphocytes granuleux T	9831/3

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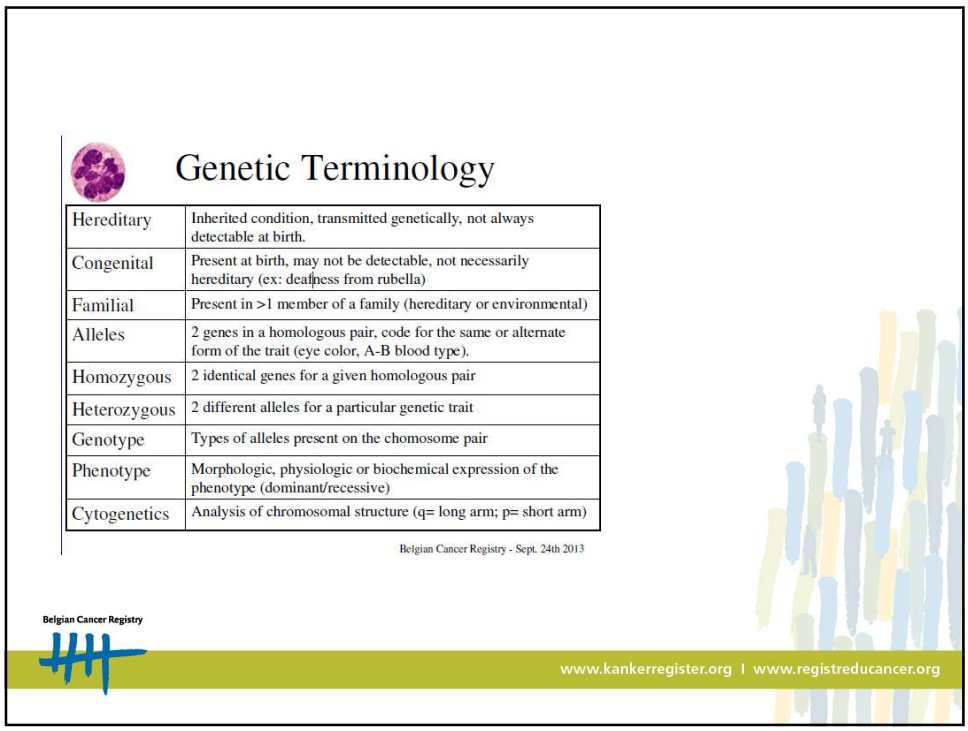
Basis Terminologie

Suffix	Meaning	Prefix	Meaning
-cyte	Cell	a- an-	lack, without, absent
-emia	Blood	aniso-	unequal, dissimilar
-it is	Inflammation	cyt-	Cell
-lysis	destruction, dissolving	dys-	abnormal, difficult, bad
-oma	swelling, tumor	erythro-	Red
-opathy	Disease	hemo- /hemato-	pertaining to blood
-osis	abnormal increase	hypo-	decreased, deficient
-penia	decreased, deficiency	hyper-	increased, extreme
-phil(ic)	attracted to	iso-	same, equal, alike
-plasia (plastic)	cell production or repair	leuk(o)-	White
-poiesis	cell production, formation and development	meta-	change, after, next
-poietin	stimulates production	macro-	large, long
		mega-	large, giant
		micro-	Small
		myel(o)-	from bone marrow
		pan-	all, overall
		phago-	eat, ingest
		poikilo-	varied, irregular
		poly-	Many
		schis-	Split
		scler-	Hard
		splen-	Spleen
		thromb(o)-	clot, thrombus

Rev ICDO3-codes niet gebruikt door WHO maar nuttig bij het oplossen van problemen

ICD-O-3 (/3)	Label	
9590	Malignant lymphoma	**Te vermijden**
9591	NHL, NOS	**Te vermijden**
9650	HL, NOS	**Te vermijden**
9733	Plasma cell leukemia	Plasma cell leukemia
9750	Malignant histiocytosis (obs but keept for ECD)	Erdheim-Chester disease
9760	Immunoproliferative disease, NOS (if possible : use 9761 or 9762)	**Te vermijden**
9800	Leukaemia, NOS	**Te vermijden**
9801	Acute leukaemia, NOS	**Te vermijden**
9820	Lymphoid leukemia, NOS	**Te vermijden**
9832	Prolymphocytic leukemia, NOS	**Te vermijden**
9835	ALL/AL, NOS	**Te vermijden**
9860	Myeloid leukaemia, NOS	**Te vermijden**
9805	Acute leukaemia, ambiguous lineage *(use by preference 9806/9807/9808/9809)*	Cf M/L
9863	CML, NOS	**Te vermijden**
9897	AML with 11q23 abnormalities	Cf other than t(9;11)


Genetic Terminology
Hereditary
Inherited condition, transmitted genetically, not always detectable at birth.
Congenital
Present at birth, may not be detectable, not necessarily hereditary (ex: deafness from rubella)
Familial
Present in >1 member of a family (hereditary or environmental)
Alleles
2 genes in a homologous pair, code for the same or alternate form of the trait (eye color, A-B blood type).
Homozygous
2 identical genes for a given homologous pair
Heterozygous
2 different alleles for a particular genetic trait
Genotype
Types of alleles present on the chomosome pair
Phenotype
Morphologic, physiologic or biochemical expression of the phenotype (dominant/recessive)
Cytogenetics
Analysis of chromosomal structure (q= long arm; p= short arm)
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