



# Enregistrement des hémopathies malignes : Documentation

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## Staging des lymphomes (Hodgkin et non Hodgkinien sauf lymphomes cutanés) Ann-Arbor (pas TNM)

Table 5.5.1. - Ann Arbor staging system classification for lymphomas (Cotswolds revision)<sup>11,12</sup>

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 <sub>2</sub> )
III	Involvement of lymph node regions or structures on both sides of the diaphragm
III <sub>1</sub>	With or without splenic, hilar, celiac or portal hepatic nodes
III <sub>2</sub>	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



## Staging des myélomes classification de Salmon-Dury

Stade	critères nécessaires	Hb (g/dl)	Pic IgG (g/l)	Pic IgA (g/l)	PBJ (g/24h)	Ca (mg/l)	Lésion osseuses	Estimation de la masse tumorale (nb de cellules)
I	Tous	> 10	< 50	< 30	< 4	<= 120	≤ 1	< 0,6.10 <sup>12</sup>
II	Ni I ni III							≥ 6.10 <sup>12</sup>
III	Un seul	< 8,5	> 70	> 50	> 12	> 120	Plusieurs	>1,2.10 <sup>12</sup>

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## Staging des myélomes International staging system (ISS)

ISS stage	Factors
I	Serum $\beta_2$ -microglobulin < 3.5 mg/L AND Serum albumin $\geq$ 3.5 g/dL
II	Not ISS stage I or III
III	Serum $\beta_2$ -microglobulin $\geq$ 5.5 mg/L

RISS Stage Group	Factors
I	Serum $\beta_2$ -microglobulin < 3.5 mg/L AND Serum albumin $\geq$ 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 $\beta_2$ -microglobulin $\geq$ 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)

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## Comportement borderlines / 1

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

## Hémopathies malignes dans contexte de déficit immunitaire

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"



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## Nouvelles entités 2017 avec codes existants

ICD-O-3 (/3)	New entities 2016/2017 with existing codes	Commentaire
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 zone lymphoma (nodal, extranodal)	Cf C
9699	Paediatric nodal MZL	"Ped NMZL"
9831	T-cell large granular lymphocytic leukemia	
9831	Chronic lymphoproliferative disorder of NK cells	"NK-CLPD"
9985	MDS with multilineage dysplasia	
9985	Refractory cytopenia of childhood	"Ped RC"

## Entités avec nouveaux codes 2017 → pour 2019

ICD-O-3 (/3)	New entities 2016/2017 with new codes	Commentaires
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	Commentaire
<b>Cytogenetics</b>		
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=EV11) (RPN1-EV11)	
9871	AML with inv(16)(p13.1q22) or t(16;16)(p13.1q22); CBFbeta-MYH11 (FAB M4Eo / Acute myelomonocytic leukemia with abnormal eosinophils)	
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-MLL3 (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	
<b>Gene mutations (new entities 2017 → 2019)</b>		
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

## Nouvelles entités 2017 sans codes HM myéloïdes avec mutation germinale

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..."

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## Entités sans codes spécifiques : Acute leukemias

ICD-O-3 (/3)	Label		
9805	Mixed-phenotype acute leukaemia, NOS, rare types	includ : "MPAL B/T"	
9805	Acute leukaemias of ambiguous lineage, NOS	includ "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

## Entités sans codes spécifiques : Plasmocyte neoplasms

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"

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## Plusieurs entités pour un même code : lymphomes agressifs

ICD-O-3 (/3)	Label	Commentaire
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"

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## Eosinophilic neoplasms

ICD-O-3 (/3)	Label	Commentaire
9965	Myeloid and lymphoid neoplasms with <i>PDGFRA</i> rearrangement	
9966	Myeloid and lymphoid neoplasms with <i>PDGFRB</i> rearrangement	
9967	Myeloid and lymphoid neoplasms with <i>FGFR1</i> rearrangement	
classified according to the myeloid neoplasms to which they correspond → assign the respective ICD-O code	<b>Myeloid and lymphoid neoplasms with <i>PCM1-JAK2</i></b>	<b>9968</b>
9964	Chronic eosinophilic leukemia, NOS	<b>Par défaut !</b>

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## Hairy cell leukemia

ICD-O-3 (/3)	Label	Commentaire
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"

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## Useful abbreviations: check codes

AITL = LAI	Angioimmunoblastic T-cell Lymphoma, Lymphome Anglo-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

## Useful abbreviations: 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 Blastés	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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## Basic Terminology

Suffix	Meaning	Prefix	Meaning
-cyte	Cell	a- an-	lack, without, absent
-emia	Blood	aniso-	unequal, dissimilar
-itis	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

## Codes ICO3 Rev non utilisés par la WHO mais utiles « en dépannage »

ICD-O-3 (/3)	Label	
9590	Malignant lymphoma	A EVITER
9591	NHL, NOS	A EVITER
9650	HL, NOS	A EVITER
9733	Plasma cell leukemia	Plasma cell leukemia
9750	Malignant histiocytosis (obs but kept for ECD)	Erdheim-Chester disease
9760	Immunoproliferative disease, NOS (if possible : use 9761 or 9762)	A EVITER
9800	Leukaemia, NOS	A EVITER
9801	Acute leukaemia, NOS	A EVITER
9820	Lymphoid leukemia, NOS	A EVITER
9832	Prolymphocytic leukemia, NOS	A EVITER
9835	ALL/AL, NOS	A EVITER
9860	Myeloid leukaemia, NOS	A EVITER
9805	Acute leukaemia, ambiguous lineage (use by preference 9806/9807/9808/9809)	Cf M/L
9863	CML, NOS	A EVITER
9897	AML with 11q23 abnormalities	Cf other than t(9;11)
9960	Chronic myeloproliferative neoplasms, NOS	



## 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 chromosome 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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