

*Corso di aggiornamento annuale per operatori
dei Registri Tumori*

10 settembre 2012

GLI AGGIORNAMENTI DI ICD-O-3

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2010: the WHO/IARC ICDO Update Committee was established.

1.9.2011 UPDATES WHO/IARC

This document provides a listing of all official additions, changes, and revisions to the International Classification of Diseases for Oncology, third edition (ICDO3) and has been approved by the IARC/WHO Committee for ICDO3.

The changes become valid from 1 January 2012.

Relevant changes in other language versions of ICDO and in related tools will also have to be made and disseminated by the appropriate authority.

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Gli obiettivi principali

- Adeguare la nomenclatura alla terminologia più moderna
- Inserire nuove entità nosologiche
- Adeguare i codici di comportamento alle nuove conoscenze scientifiche

Riferimento: Blue Books IARC/WHO

Lo schema

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/			
Related term=not indented	behavior combination	Bold indicates preferred term		
Synonym = indented				
New code and term	8077/0	Squamous intraepithelial neoplasia, low grade		
New synonym	8077/0	Squamous intraepithelial neoplasia, grade I		
New synonym	8077/0	Squamous intraepithelial neoplasia, grade II		
New related term	8077/0	Anal intraepithelial neoplasia, low grade (C21.1)		
New related term	8077/0	Cervical intraepithelial neoplasia, low grade (C53.)		
New related term	8077/0	Esophageal squamous intraepithelial neoplasia (dysplasia), low grade (C15.)		
New preferred term	8077/2	Squamous intraepithelial neoplasia, high grade		
Move former preferred term to synonym	8077/2	Squamous intraepithelial neoplasia, grade III		<i>Unbold and indent former preferred term</i>
New related term	8077/2	Esophageal squamous intraepithelial neoplasia (dysplasia), high grade (C15.)		
New code and term	8148/0	Glandular intraepithelial neoplasia, low grade		
New synonym	8148/0	Glandular intraepithelial neoplasia, grade I		
New synonym	8148/0	Glandular intraepithelial neoplasia, grade II		
New related term	8148/0	Biliary intraepithelial neoplasia, low grade		

Nuovo codice e termine

Termine che non è più quello principale

Senza indentatura: nuovo termine correlato
 Con indentatura: nuovo sinonimo

Neoplasie squamose intraepiteliali

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/	Bold indicates preferred term		
Related term=not indented	behavior			
Synonym = indented	combina- tion			
New code and term	8077/0	Squamous intraepithelial neoplasia, low grade		
New synonym	8077/0	Squamous intraepithelial neoplasia, grade I		
New synonym	8077/0	Squamous intraepithelial neoplasia, grade II		
New related term	8077/0	Anal intraepithelial neoplasia, low grade (C21.1)		
New related term	8077/0	Cervical intraepithelial neoplasia, low grade (C53._)		
New related term	8077/0	Esophageal squamous intraepithelial neoplasia (dysplasia), low grade (C15._)		
New preferred term	8077/2	Squamous intraepithelial neoplasia, high grade		
Move former preferred term to synonym	8077/2	Squamous intraepithelial neoplasia, grade III		<i>Unbold and indent former preferred term</i>
New related term	8077/2	Esophageal squamous intraepithelial neoplasia (dysplasia), high grade (C15._)		

CIN 1-2

**BASSO GRADO
GRADO I E II**

/0 – nuovo codice

**ALTO GRADO
GRADO III**

/2

CIN3

Inserita sede esofago (anche displasia, a basso o alto grado)

Neoplasie ghiandolari intraepiteliali

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/behavior combination	Bold indicates preferred term		
Related term=not indented				
Synonym = indented				
New code and term	8148/0	Glandular intraepithelial neoplasia, low grade		
New synonym	8148/0	Glandular intraepithelial neoplasia, grade I		
New synonym	8148/0	Glandular intraepithelial neoplasia, grade II		
New related term	8148/0	Biliary intraepithelial neoplasia, low grade		
New related term	8148/0	Esophageal glandular dysplasia (intraepithelial neoplasia), low grade (C16._)		
New preferred term	8148/2	Glandular intraepithelial neoplasia, high grade		
Move former preferred term to synonym	8148/2	Glandular intraepithelial neoplasia, grade III		<i>Unbold and indent form preferred term</i>
New synonym	8148/2	Flat intraepithelial neoplasia, high grade		
New related term	8148/2	Flat intraepithelial glandular neoplasia, high grade (C24.1)		
New synonym	8148/2	Flat intraepithelial neoplasia (dysplasia), high grade (C24.1)		
New related term	8148/2	Biliary intraepithelial neoplasia, high grade		
New synonym	8148/2	Biliary intraepithelial neoplasia, grade 3 (BIIN-3)		
New related term	8148/2	Esophageal glandular dysplasia (intraepithelial neoplasia), high grade (C16._)		
New synonym	8148/2	Esophageal intraepithelial neoplasia, high grade (C16._)		

PIN 1-2

**BASSO GRADO
GRADO I E II**

/0 – nuovo codice

**ALTO GRADO
GRADO III**

/2 PIN3

Inserite sedi esofago (anche displasia), vie biliari, ampolla di Vater

Tumori pancreatici endocrini

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/behavior combination	Bold indicates preferred term		
New preferred term	„ < †€/ ∴	Pancreatic endocrine tumor, benign (C^Yo. _)		
Move former preferred term to synonym	8150/0		Islet cell adenoma (C25. _)	<i>Unbold and indent former preferred term</i>
New related term	„ < †€/ ∴	Pancreatic microadenoma (C ^Y o. _)		
New preferred term	„ < †€/ ∴	Pancreatic endocrine tumor, NOS (C^Yo. _)		
Move former preferred term to synonym	„ < †€/ ∴		Islet cell tumor, NOS (C ^Y o. _)	<i>Unbold and indent former preferred term</i>
New preferred term	8150/3	Pancreatic endocrine tumor, malignant		
Move former preferred term to synonym	„ < †€/ ∴		Islet cell carcinoma (C ^Y o. _)	<i>Unbold and indent former preferred term</i>
New related term	8150/3	Pancreatic endocrine tumor, nonfunctioning		
New preferred term	8154/3	Mixed pancreatic endocrine and exocrine tumor, malignant (C25. _)		
New related term	8154/3	Mixed endocrine and exocrine adenocarcinoma (C25. _)		
New synonym for related term	8154/3		Mixed islet cell and exocrine adenocarcinoma (C25. _)	<i>Unbold former preferred term</i>
New related term	8154/3	Mixed acinar-endocrine-ductal carcinoma		

- cambi di termine preferito
- nuovi termini correlati

Altri tumori endocrini

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
Code change; new related term	8152/1	Enteroglucagonoma, NOS		<i>Was 8157/1; now related term for glucagonoma</i>
New related term	8152/1	L-cell tumor		
New related term	8152/1	Glucagon-like peptide-producing tumor		
New related term	8152/1	Pancreatic peptide and pancreatic peptide-like peptide within terminal tyrosine amide producing tumor		
New synonym for related term	8152/1	PP/PYY producing tumor		
Code change; new related term	8152/3	Enteroglucagonoma, malignant		<i>Was 8157/3; now related term for glucagonoma, malignant</i>
Delete code and term	8157/1	Enteroglucagonoma, NOS		<i>Term recoded as 8152/1</i>
Delete code and term	8157/3	Enteroglucagonoma, malignant		<i>Term recoded as >>>>/></i>
New term and code	8158/1	Endocrine tumor, functioning, NOS		
New related term	8158/1	ACTH-producing tumor		

Enteroglucagonoma: non più codice autonomo
Rientra nel codice del Glucagonoma, insieme a nuove nosologie simili

Nuovo codice per il tumore endocrino funzionante e il tumore secernente ACTH

Neoplasie pancreatobiliari

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
New term and code	8163/0	Pancreatobiliary neoplasm, non-invasive		
New synonym	8163/0		Noninvasive pancreatobiliary papillary neoplasm with low grade dysplasia	
New synonym	8163/0		Noninvasive pancreatobiliary papillary neoplasm with low grade intraepithelial neoplasia	
New term and code	8163/2	Papillary neoplasm, pancreatobiliary-type, with high grade intraepithelial neoplasia (C24.1)		
New synonym	8163/2		Noninvasive pancreatobiliary papillary neoplasm with high grade dysplasia (C24.1)	
New synonym	8163/2		Noninvasive pancreatobiliary papillary neoplasm with high grade intraepithelial neoplasia (C24.1)	
New term and code	8163/3	Pancreatobiliary-type carcinoma (C24.1)		
New synonym	8163/3		Adenocarcinoma, pancreatobiliary type (C24.1)	

NUOVI CODICI E NOSOLOGIE SOLO SU BASE MORFOLOGICA

Displasia a basso/alto grado = neoplasia intraepiteliale a basso/alto grado

I codici /2 e /3 sono riferiti alla papilla di Vater (C24.1)

Il codice /2 fa riferimento ad una neoplasia papillare (e le forme non papillari ?)

Se descritta la neoplasia di tipo pancreatobiliare della papilla di Vater con displasia ad alto grado usare lo stesso codice

Altre neoplasie ghiandolari

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
New related term	8201/3	Cribriform comedo-type carcinoma (C18._, C19.9, C20.9)		Nuovi termini del carcinoma cribriforme
New synonym	8201/3	Adenocarcinoma, cribriform comedo-type (C18._, C19.9, C20.9)		
New synonym to primary term	8213/0	Traditional serrated adenoma		Nuovi termini dell'adenoma serrato
New related term	8213/0	Sessile serrated adenoma		
New related term	8213/0	Sessile serrated polyp		
New related term	8213/0	Traditional sessile serrated adenoma		
New term	8213/3	Serrated adenocarcinoma		Nuovo codice e termine
New synonym	8263/0	Tubulo-papillary adenoma		Nuovo termine dell'adenoma tubulo villosa
New code and term	8265/3	Micropapillary carcinoma, NOS (C18._, C19.9, C20.9)		Nuovo codice e termine a sede colo-rettale
New related term	8290/0	Spindle cell oncocytoma (C75.1)		Nuovo termine dell'adenoma ossifilo a sede ipofisaria

Tumori neuroendocrini

E' UNO DEI CAMBIAMENTI PRINCIPALI PER LA REGISTRAZIONE

Status	ICD-O-3	Term	Indent 1	
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
Behavior code change ; delete code and term	8240/1	Carcinoid tumor, NOS, of appendix (C18.1)		Code changed to >>>>/>
Behavior code change ; delete code and term	8240/1	Carcinoid, NOS, of appendix (C18.1)		Code changed to 8240/3
Wording change	8240/3	Carcinoid tumor, NOS		Delete "(except of appendix M-8240/1)" Delete "(except of appendix M-8240/1)"
Wording change	8240/3	Carcinoid, NOS		
New related term	8240/3	Neuroendocrine tumor, grade 1		
New related term	8240/3	Neuroendocrine carcinoma, low grade		
New related term	8240/3	Neuroendocrine carcinoma, well-differentiated		
New preferred term	8244/3	Mixed adenoneuroendocrine carcinoma		
Move former preferred term to synonym	8244/3	Composite carcinoid		Unbold and indent former preferred term
New synonym	8244/3	Combined/mixed carcinoid and adenocarcinoma		
New synonym	8244/3	MANEC		
New synonym	8249/3	Neuroendocrine tumor, grade 2		
New related term	8249/3	Neuroendocrine carcinoma, moderately differentiated		

I Carcinoidi dell'appendice diventano tutti /3
si allinea la codificazione al TNM 7
Rimangono /1 i carcinoidi argentaffini e ad incerta malignità potenziale

Nuovi termini del carcinoma tipico

Nuovi termini del carcinoma atipico

Da notare che
 Tumore neuroendocrino grado 1= carcinoma neuroendocrino a basso grado o G1
 = **CARCINOIDE TIPICO 8240/3**
 Tumore neuroendocrino grado 2= carcinoma neuroendocrino G2
 = **CARCINOIDE ATIPICO (ALTRO CODICE!) 8249/3**
 Carcinoma neuroendocrino G3 = 8246/3

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
New related term	8453/0	Intraductal papillary-mucinous tumor with low grade dysplasia (C25._)		
New synonym	8453/0	Intraductal papillary-mucinous neoplasm with low grade dysplasia (C25._)		
Code change; new related term	8453/0	Intraductal papillary-mucinous tumor with moderate dysplasia (C25._)		Was 8453/1
New synonym	8453/0	Intraductal papillary-mucinous neoplasm with moderate dysplasia (C25._)		
New related term	8453/0	Intraductal papillary-mucinous tumor with intermediate dysplasia (C25._)		
Delete code and term	8453/1	Intraductal papillary-mucinous tumor with moderate dysplasia (C25.1)		Code changed to 8453/0
New related term	8453/2	Intraductal papillary mucinous neoplasm with high grade dysplasia		
New related term	8453/3	Intraductal papillary mucinous neoplasm with an associated invasive carcinoma		
New related term	8470/0	Mucinous cystic tumor with low grade dysplasia (C25._)		
New related term	8470/0	Mucinous cystic neoplasm with low-grade intraepithelial neoplasia (C22._)		
New related term	8470/0	Mucinous cystic neoplasm with intermediate-grade intraepithelial neoplasia (C22._)		
New related term	8470/0	Mucinous cystic neoplasm with low-grade dysplasia (C25._)		
New related term	8470/0	Mucinous cystic neoplasm with intermediate-grade dysplasia (C25._)		
Code change; new related term	8470/0	Mucinous cystic tumor with moderate dysplasia (C25._)		Was 8470/1
New related term	8470/0	Mucinous cystic tumor with intermediate dysplasia (C25._)		
Delete code and term	8470/1	Mucinous cystic tumor with moderate dysplasia (C25._)		Code changed to 8470/0
New related term	8470/2	Mucinous cystic tumor with high-grade dysplasia (C25._)		
New synonym	8470/2	Mucinous cystic neoplasm with high-grade intraepithelial neoplasia (C22._)		
New synonym	8470/2	Mucinous cystic neoplasm with high-grade dysplasia (C25._)		
New related term	8470/3	Mucinous cystic tumor with an associated invasive carcinoma (C25._)		
New synonym	8470/3	Mucinous cystic neoplasm with an associated invasive carcinoma (C25._)		
New code and term	8480/1	Low grade appendiceal mucinous neoplasm (C18.1)		
New related term	8490/3	Poorly cohesive carcinoma		

Neoplasie cistiche mucinose

Soprattutto nuovi termini riportati alla displasia

Passano da /1 a /0

Nuovo codice e termine (appendice)

Nuovo termine del carcinoma a cellule ad anello con castone

Neoplasie duttali, lobulari e a cellule acinose

Soprattutto nuovi termini rapportati a displasia e grado basso e intermedio/alto, e a sedi specifiche (dotti biliari intra ed extraepatici, colecisti)

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combina- tion	Bold indicates preferred term		
New related term	8503/0	Intraductal papillary neoplasm, NOS		
New related term	8503/0	Intraductal papillary neoplasm with low grade intraepithelial neoplasia (C22._, C24.0)		
New synonym	850/30	Intraductal papillary neoplasm with intermediate grade neoplasia (C22._, C24.0)		
New related term	8503/0	Intracystic papillary neoplasm with low grade intraepithelial neoplasia (C23.9)		
New synonym	8503/0	Intracystic papillary neoplasm with intermediate grade intraepithelial neoplasia (C23.9)		
New synonym	8503/0	Intraglandular papillary neoplasm with low grade intraepithelial neoplasia (C22.1, C24.0)		
New related term	8503/0	Intraductal tubular-papillary neoplasm, low grade		
New related term	8503/2	Intraductal papillary neoplasm with high grade intraepithelial neoplasia		
New synonym	8503/2	Intraductal papillary neoplasm with high grade dysplasia		
New synonym	8503/2	Intraductal papillary tumor with high grade intraepithelial neoplasia		
New synonym	8503/2	Intraductal papillary tumor with high grade dysplasia		
New synonym	8503/2	Intracystic papillary neoplasm with high grade intraepithelial neoplasia (C23.9)		
New synonym	8503/2	Intracystic papillary tumor with high grade intraepithelial neoplasia (C23.9)		
New synonym	8503/2	Intracystic papillary tumor with high grade dysplasia (C23.9)		
New related term	8503/2	Intraductal tubular-papillary neoplasm, high grade		
New related term	8503/3	Intraductal papillary neoplasm with associated invasive carcinoma		
New synonym for related term	8503/3	Intracystic papillary neoplasm with associated invasive carcinoma (C23.9)		
New term and code	8552/3	Mixed acinar-ductal carcinoma		Nuovo codice e termine

Da notare che i tumori intracistici papillari di altre sedi potrebbero essere ricondotti al codice 8504
E i carcinomi intracistici papillari invasivi della colecisti al 8504/3

Neoplasie fibromatose e complesse miste e stromali

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
New related term	8811/0	Plexiform fibromyxoma		
New related term	8970/3	Hepatoblastoma, epithelioid (C22.0)		
New related term	8970/3	Hepatoblastoma, mixed epithelial-mesenchymal (C22.0)		
New term and code	8975/1	Calcifying nested epithelial stromal tumor (C22.0)		

Nuovo codice e termine riferito al fegato

Gliomi e neoplasie neuroepiteliomatose

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
New term and code	9395/3	Papillary tumor of the pineal region		Nuovo codice e termine
New term and code	9425/3	Pilomyxoid astrocytoma		
New term and code	9431/1	Angiocentric glioma		
New term and code	9432/1	Pituicytoma		
New related term	9471/3	Medulloblastoma with extensive nodularity		
New related term	9474/3	Anaplastic medulloblastoma		
New related term	9506/1	Extraventricular neurocytoma		
New term and code	9509/1	Papillary glioneuronal tumor		Nuovo codice e termine
New related term	9509/1	Rosette-forming glioneuronal tumor		

EMATOLINFOPOIETICO

E' IL CAPITOLO CON MAGGIORI INNOVAZIONI

di fatto viene recepito l'ultimo Blue Book IARC

Nuove classificazioni = NUOVE NOSOLOGIE

- **diagnostica citogenetica**
- **conoscenza della biochimica del tumore**
- **immunoistochimica**
- **differenze cliniche (epidemiologia, terapia, sopravvivenza)**

- * Qualche codificazione è di difficile comprensione logica
- * Nel tracciato c'è qualche errore.

NB In campo oncoematologico molte diagnosi sono entrate nell'uso clinico prima dell'ufficializzazione, per cui sarebbe meglio aggiornare il datawarehouse ed utilizzarlo prima possibile.

Poche sono peraltro le modifiche di forte impatto.

LINFOMI MALIGNI, NAS O DIFFUSI

Nuovi termini del LNH NAS, o a cellule B o diffuso
MA

- **LA LEUCEMIA A CELLULE CAPELLUTE VARIANTE ERA CLASSIFICATA COME 9940/3**
mantenere la classificazione come 9940/3 (non è stata cancellata la voce)?

Nosologicamente è una variante prolinfocitica di HCL, ma ad essa biologicamente non correlata (diverso immunofenotipo ; resistenza alla terapia)

Status	ICD-O-3	Term	Indent 1
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/	Bold indicates preferred term	
Related term=not indented	behavior	combination	
Synonym = indented	tion		
New related term	9591/3	Splenic B-cell lymphoma/leukemia, unclassifiable	
New related term	9591/3	Splenic diffuse red pulp small B-cell lymphoma	
New related term	9591/3	Hairy cell leukemia variant	
New related term	9596/3	B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma	
New term and code	9597/3	Primary cutaneous follicle centre lymphoma	

Nuovo termine del Linfoma Composito Hodgkin e NH

Nuovo codice e termine
Linfoma cutaneo primitivo dei centri follicolari

Da notare che i linfomi splenici sono ripartiti in 3 codici:

- * epatosplenici a cellule T (o gamma-delta) 9716/3
- * della zona marginale splenica (o splenico a linfociti villosi) 9689/3
- * splenico a piccoli linfociti B diffuso della polpa rossa (o splenico B-cell linfoma/leucemia) 9591/3

LINFOMI MALIGNI a cellule B mature

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
New related term	9680/3	Diffuse large B-cell lymphoma associated with chronic inflammation		Formerly pyothorax-associated lymphoma
New related term	9680/3	B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma		
New related term	9680/3	EBV positive diffuse large B-cell lymphoma of the elderly		
New related term	9680/3	Primary diffuse large B-cell lymphoma of the CNS (C70._, C71._, C72._) Primary cutaneous DLBCL, leg type (C44.7)		
New related term	9680/3			
Delete code and term	9680/3	T-cell/histiocyte rich large B-cell lymphoma		Code changed to 9688/3
Code change; delete code and term	9684/3	Plasmablastic lymphoma		Code changed to 9735/3
Code restored	9688/3	T-cell/histiocyte rich large B-cell lymphoma		Was 9688/3 in ICD-O-2
New synonym	9698/3	Follicular lymphoma, grade 3A		
New synonym	9698/3	Follicular lymphoma, grade 3B		
New synonym of Mucosal associated lymphoid tissue lymphoma	9699/3	Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue		

Passa ai tumori plasmacellulari

Ritorna al codice ICD-O-2

Le forme che trovano nuova specifica codificazione sono
 * LNH a grandi cellule B T-cell/histiocyte rich (ritorno a codice precedente)
 •Linfoma a cellule B non classificabile con caratteristiche intermedie tra linfoma a grandi cellule B diffuso e linfoma di Burkitt (non si privilegia il codice più alto)

Le altre forme erano già facilmente classificabili

LINFOMI MALIGNI a cellule T e NK mature

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/behavior combination	Bold indicates preferred term		La ricollocazione del linfoma intravascolare a grandi cellule B pare scorretta dal punto di vista classificativo perché il gruppo è quello dei LNH T/NK. Essendo un codice autonomo senza sinonimi e termini correlati, ne è possibile il recupero a posteriori in caso di correzioni; va codificato il grado 6.
New related term	9702/3	Anaplastic large cell lymphoma, ALK negative		
New related term	9709/3	Primary cutaneous CD8-positive aggressive epidermotropic cytotoxic T-cell lymphoma		
New related term	9709/3	Primary cutaneous CD4-positive small/medium T-cell lymphoma		
Code restored	9712/3	Intravascular large B-cell lymphoma (C49.9)		Was 9712/3 in ICD-O-2
New related term	9714/3	Anaplastic large cell lymphoma, ALK positive		
New preferred term	9716/3	Hepatosplenic T-cell lymphoma		
Move former preferred term to synonym	9716/3	Hepatosplenic $\gamma\delta$ (gamma-delta) cell lymphoma		Unbold and indent former preferred term
New synonym	9719/3	Extranodal NK/T-cell lymphoma, nasal type		

Il Linfoma anaplastico a grandi cellule ALK- non va codificato come anaplastico, ma come Linfoma a cellule T mature.

I Linfomi Anaplastici a Grandi Cellule ALK+ invece esprimono l'antigene glicoproteico di superficie CD30/Ki-1. Nell'età pediatrica circa l'80% di questi linfomi presenta una tipica traslocazione cromosomica t(2;5)(p23;q35), da cui origina la proteina chimerica NPM-ALK/p80, che possiede attività tirosina chinasi. Perciò solo questi rimangono nel gruppo dei LNH Ki-1+ o CD30+

(NPM=nucleofosmina ALK= Anaplastic Lymphoma Kinase – presenti nel citoplasma)

LINFOMI MALIGNI linfoblastici a cellule progenitrici

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/	Bold indicates preferred term		
Related term=not indented	behavior combination			
Synonym = indented				
New term and code	9724/3	Systemic EBV positive T-cell lymphoproliferative disease of childhood		
New term and code	9725/3	Hydroa vacciniforme-like lymphoma		NUOVO codice e termine
New term and code	9726/3	Primary cutaneous gamma-delta T-cell lymphoma		
New related term	9727/3	Blastic plasmacytoid dendritic cell neoplasm		
New related term	9727/3	Blastic NK cell lymphoma [obs]		

Da notare:

- la **Malattia linfoproliferativa dell'infanzia a cellule T EBV+** è una nosologia prima classificabile come NAS 9760/3
- la **Neoplasia blastica delle cellule dendritiche plasmacitoidi** è una nosologia prima classificabile come Tumore delle cellule dendritiche 9757/3
- il **Linfoma a cellule T gamma-delta primitivo della cute** prima era classificabile come 9709/3 (cutaneo a cellule T)
- il **Linfoma blastico a cellule NK** prima era classificabile come 9719/3 (Linfoma a cellule T/NK)

TUMORI PLASMACELLULARI

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/	Bold indicates behavior		
Related term=not indented	combination	preferred term		
Synonym = indented				
New synonym	9734/3		Extraosseous plasmacytoma	
New term and code	9735/3	Plasmablastic lymphoma		
New term and code	9737/3	ALK positive large B-cell lymphoma		
New term and code	9738/3	Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease		

Nuovi codici e termini peraltro relativi a LNH a cellule B mature

Linfoma plasmablastico: maschi HIV+ o immunosoppressi, EBV+, markers plasmacellulari, presenza di ipermutazione dei geni IG. Precedentemente classificato come 9684/3

Linfoma a grandi cellule B insorto in Malattia di Castleman multicentrica HHV-8 associata:
 L'HHV8 (human herpes virus 8), presente nella maggioranza di casi di MC in soggetti HIV+ e in un terzo circa dei casi di MC in soggetti HIV-, si localizza in immunoblasti e linfociti CD20+ e induce la produzione di un omologo virale dell'interleuchina-6 (vIL6), che facilita la comparsa dei linfomi B e dei linfomi a effusione primaria.

Nelle MC multicentriche si rileva un'iperplasia linfatica angiofollicolare policlonale, spesso sotto forma plasmacellulare, associata anche a gammopatia monoclonale IgM e poliadenopatie periferiche. Concorrono quindi 3 diagnosi: MC multicentrica, HHV8+ e LNH a grandi cellule B (generalmente plasmablastico senza ipermutazione dei geni IG)

Il **LNH a grandi cellule B ALK+** (prima 9680/3) è un linfoma di grandi cellule B immunoblastiche-like talvolta con differenziazione plasmablastica

TUMORI MASTOCITARI

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
New related term	9740/1	Cutaneous mastocytosis		
New related term	9740/1	Urticaria pigmentosa		
New related term	9740/1	Diffuse cutaneous mastocytosis		
New synonym	9740/1	Solitary mastocytoma of skin		
New synonym	9740/1	Extracutaneous mastocytoma		
New term and code	9741/1	Indolent systemic mastocytosis		
New related term	9741/3	Systemic mastocytosis with associated hematological clonal non-mast cell disorder		
New related term	9741/3	Systemic mastocytosis with AHNMD		
New related term	9741/3	Aggressive systemic mastocytosis		

Nuovo codice e termine

La Mastocitosi sistemica prima era sempre da attribuire al 9741/3 (termine Malattia sistemica del tessuto mastocitico)
Adesso è 9714/1 se è specificato che è una forma indolente.

NEOPLASIE DEGLI ISTIOCITI E DELLE CELLULE LINFOIDI ACCESSORIE

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
Notes added	9751/1	Langerhans cell histiocytosis, NOS [obs] (use 9751/3)		
Notes added	9751/1	Langerhans cell granulomatosis [obs] (use 9751/3)		
Note added	9751/1	Histiocytosis X, NOS [obs] (use 9751/3)		
				Use this code for all types of Langerhans cell histiocytosis, including the former 9751/1 through 9754/3 terms.
Behavior code change	9751/3	Langerhans cell histiocytosis, NOS		
Notes added	9752/1	Langerhans cell histiocytosis, unifocal [obs] (use 9751/3)		
Notes added	9752/1	Langerhans cell granulomatosis, unifocal [obs] (use 9751/3)		
Notes added	9752/1	Langerhans cell histiocytosis, mono-ostotic [obs] (use 9751/3)		
Notes added	9753/1	Langerhans cell histiocytosis, multifocal [obs] (use 9751/3)		
Notes added	9753/1	Langerhans cell histiocytosis, poly-ostotic [obs] (use 9751/3)		
Note added	9753/1	Hand-Schuller-Christian disease [obs] (use 9751/3)		
Notes added	9754/3	Langerhans cell histiocytosis, disseminated [obs] (use 9751/3)		
Notes added	9754/3	Langerhans cell histiocytosis, generalized [obs] (use 9751/3)		
Notes added	9754/3	Letterer-Siwe disease [obs] (use 9751/3)		
Notes added	9754/3	Acute progressive histiocytosis X [obs] (use 9751/3)		
Note added	9754/3	Nonlipid reticuloendotheliosis [obs] (use 9751/3)		
New related term	9757/3	Indeterminate dendritic cell tumor		
New term and code	9759/3	Fibroblastic reticular cell tumor		

Nuovo sistema di codifica – caso particolare

Nuovo codice e termine

Anche se hanno mantenuto il codice originario, il WHO indica di codificare tutte le forme di Istiocitosi neoplastiche comprese da 9751/1 a 9754/3 come **9751/3**.
 Quindi le forme a comportamento /1 entrano in incidenza

LEUCEMIE, NAS

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3	9806/3	Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1		
Related term=not indented	9807/3	Mixed phenotype acute leukemia with t(v;11q23); MLL rearranged		
Synonym = indented	9808/3	Mixed phenotype acute leukemia, B/myeloid, NOS		
	9809/3	Mixed phenotype acute leukemia, T/myeloid, NOS		

Sono tutti nuovi termini e codici di leucemia acuta bifenotipica, basati su morfologia.

I primi due prevedono diagnosi mieloide con citogenetica e linfoide generica

I secondi due prevedono diagnosi di cellularità linfoide e mieloide generica.

Secondo le regole correnti di registrazione, in caso di doppia diagnosi precisa, prevale il codice contenente l'informazione citogenetica (e non il codice più alto)

LEUCEMIE LINFOLDI

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/	Bold indicates preferred term		
Related term=not indented	behavior combination			
Synonym = indented				
New term and code	9811/3	B lymphoblastic leukemia/lymphoma, NOS		
New term and code	9812/3	B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1		
New term and code	9813/3	B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged		
New term and code	9814/3	B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1)		
New term and code	9815/3	B lymphoblastic leukemia/lymphoma with hyperdiploidy		
New term and code	9816/3	B lymphoblastic leukemia/lymphoma with hypodiploidy (Hypodiploid ALL)		
New term and code	9817/3	B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH		
New term and code	9818/3	B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A-PBX1 (TCF3-PBX1)		
Behavior code change	9831/3	T-cell large granular lymphocytic leukemia		<i>Was 9831/1</i>
New related term	%pfi< / ::	Chronic lymphoproliferative disorder of NK cells		
New related term	%pfi, / ::	T lymphoblastic leukemia/lymphoma		

La leucemia linfocitica a grandi cellule T granulari diventa /3 (anche i sinonimi)

Le leucemie linfoidi comprendono adesso anche l'asse 981 (981-983), che contiene tutte le forme di leucemia linfoblastica/linfoma a cellule B (oltre la citogenetica, interessa anche la ploidia)

La leucemia linfoblastica/linfoma a cellule T viene inserita non con codice autonomo, ma come termine correlato alla Leucemia linfoblastica a cellule progenitrici T

Solo quando non è specificata la linea cellulare si continua ad usare il 9835/3

Passa a /3 anche l'unica leucemia linfocitica a cui ICD-O-3 aveva attribuito il comportamento /1

LEUCEMIE MIELOIDI

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/behavior combination	Bold indicates preferred term		
Related term=not indented				
Synonym = indented				
New related term	9861/3	Acute myeloid leukemia with mutated NPM1		
New related term	9861/3	Acute myeloid leukemia with mutated CEBPA		
New term and code	9865/3	Acute myeloid leukemia with t(6;9)(p23;q34); DEK-NUP214		
New term and code	9869/3	Acute myeloid leukemia with inv(3)(q21q26.2) or t(3;3)(q21;q26.2); RPN1-EVI1		
New related term	9891/3	Acute monoblastic and monocytic leukemia		
New preferred term	9895/3	Acute myeloid leukemia with myelodysplasia-related changes		
Move former preferred term to synonym	<<<</::	Acute myeloid leukemia with multilineage dysplasia		<i>Unbold and indent former preferred term</i>
New synonym	9896/3	Acute myeloid leukemia with t(8;21)(q22;q22); RUNX1-RUNX1T1		
New synonym	9897/3	Acute myeloid leukemia with t(9;11)(p22;q23); MLLT3-MLL		
New term and code	9898/1	Transient abnormal myelopoiesis		
New term and code	9898/3	Myeloid leukemia associated with Down Syndrome		
New term and code	9911/3	Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); RBM15-MKL1		
New preferred term	9920/3	Therapy related myeloid neoplasm		
Move former preferred term to synonym	9920/3	Therapy-related acute myeloid leukemia, NOS		<i>Unbold and indent former preferred term</i>

Tra i nuovi termini spiccano

- 3 nosologie basate su citogenetica, di cui 1 è una LMA megacarioblastica
- la Leucemia mieloide associata alla sindrome di Down (si intende sempre acuta)
- la Mielopoiesi anormale transitoria (tipica dei neonati Down) con comportamento /1

Il codice 9895/3 è sempre più chiaramente riferito a LMA con/in mielodisplasia

DISORDINI MIELOPROLIFERATIVI CRONICI

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3 Related term=not indented Synonym = indented	Bold indicates a new code/ behavior combination	Bold indicates preferred term		
New preferred term	9960/3	Myeloproliferative neoplasm, NOS		
Move former preferred term to synonym	9960/3	Chronic myeloproliferative disease, NOS		<i>Unbold and indent former preferred term</i>
Code change; new synonym	9960/3	Myeloproliferative disease, NOS		<i>Was 9975/1</i>
New preferred term	9961/3	Primary myelofibrosis		
Move former preferred term to synonym	9961/3	Myelosclerosis with myeloid metaplasia		<i>Unbold and indent former preferred term</i>
New preferred term	9964/3	Chronic eosinophilic leukemia, NOS		
Move former preferred term to synonym	9964/3	Hypereosinophilic syndrome		<i>Unbold and indent former preferred term</i>
New term and code	9965/3	Myeloid and lymphoid neoplasms with PDGFRA rearrangement		
New term and code	9966/3	Myeloid neoplasms with PDGFRB rearrangement		
New term and code	9967/3	Myeloid and lymphoid neoplasms with FGFR1 abnormalities		

Ci sono 3 nuovi termini basati su diagnosi citogenetica, di cui 2 riguardano neoplasie sia linfoidi che mieloidi (le forme linfoidi esulano dalla categoria gerarchica)

Le Malattie mieloproliferative NAS, prima inserite tra gli altri disturbi ematologici con comportamento /1 (entravano in incidenza solo quelle con la specifica "croniche") cambiano codice e diventano 9960/3.

Se è esplicitata la dicitura "NON CLASSIFICABILE" (significato diverso da NAS), diventano 9975/3 (Neoplasia mieloproliferativa NAS 9960/3 vs. Neopl. mieloproliferativa non classificabile 9975/3)

ALTRI DISORDINI EMATOLOGICI

Status	ICD-O-3	Term	Indent \	Comment
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/	Bold indicates behavior		
Related term=not indented	combination	preferred term		
Synonym = indented				
New term and code	9971/1	Post transplant lymphoproliferative disorder, NOS		
New synonym	9971/1	PTLD, NOS		
New term and code	9971/3	Polymorphic post transplant lymphoproliferative disorder		
Delete code and term	9975/1	Myeloproliferative disease, NOS [obs]		Code changed to 9960/3
New code and term	9975/3	Myeloproliferative neoplasm, unclassifiable		
New synonym	9975/3	Myelodysplastic/myeloproliferative neoplasm, unclassifiable		

Le malattie mieloproliferative non classificabili sono le patologie miste mielodisplastiche/ mieloproliferative diverse dalle Leucemie Mielomonocitiche Croniche (codici da 9940/3 a 9948/3)

Compaiono con il codice 9971 anche 2 nuove nosologie post-trapianto:

i Disordini linfoproliferativi NAS (comportamento /1) (early lesions)

i Disordini linfoproliferativi polimorfi (comportamento /3) (neoplasie multiple LNH B-cell, T-cell e LH)

I Disordini linfoproliferativi monomorfi post-trapianto prendono il codice della specifica nosologia.

NB i Disordini linfoproliferativi NAS NON post- trapianto sono 9970/1 (= malattia linfoproliferativa NAS)

i disordini linfoproliferativi cronici a cellule NK sono 9831/3 (leucemie a grandi cell. T granulari)

altre malattie linfoproliferative con codici specifici: primitiva cutanea a cellule T CD30+ (9718/3), sistemica a cellule T dell'infanzia EBV+ (9724/3), t-gamma (9768/3)

Diagnosi di disordini (o malattie) linfoproliferativi NAS o cronici a cellule B non post-trapianto non hanno codice specifico > 9970/1

SINDROMI MIELODISPLASTICHE

Status	ICD-O-3	Term	Indent 1	Comment
Bold indicates change from what is printed in ICD-O-3	Bold indicates a new code/	Bold indicates preferred behavior		
Related term=not indented	combination	term		
Synonym = indented				
New synonym	9982/3		Refractory anemia with ring sideroblasts associated with marked thrombocytosis	
New synonym	9985/3		Refractory cytopenia of childhood	
New synonym	9986/3		Myelodysplastic syndrome with isolated del (5q)	
New synonym	9989/3		Myelodysplastic syndrome, unclassifiable	
New term and code	9991/3	Refractory neutropenia		
New term and code	9992/3	Refractory thrombocytopenia		

Due nuove nosologie: Neutropenie e Trombocitopenie refrattarie.

Viene loro assegnato un codice 999, per cui il raggruppamento gerarchico delle MDS è adesso 998-999

PER CONCLUDERE

Le variazioni di maggior impatto sulla registrazione sono le seguenti:

- Abbiamo a disposizione un codice per le CIN 1 e 2 (e simili) e non è più necessario ricorrere al codice SNOMED
- In parallelo è rinforzata la relazione tra neoplasia intraepiteliale e displasia:
se a basso grado =/0 se ad alto grado =/2

PER CONCLUDERE - 2

- I carcinoidi dell'appendice non sono più diversi dagli altri e quindi diventano /3
- Tumori e carcinomi neuroendocrini di grado diverso hanno codice diverso

PER CONCLUDERE - 3

- Le neoplasie ematologiche, salvo pochissime eccezioni, sono tutte /3 e sono aumentate le nosologie codificabili in maniera precisa
- Anche diversi codici sono cambiati



*Grazie per l'ascolto
&
Grazie per l'attenzione...*

Riferimenti:

WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues 4th Edition – IARC Lyon 2008

<http://www.who.int/classifications/icd/updates/ICDO3Updates2011.pdf>