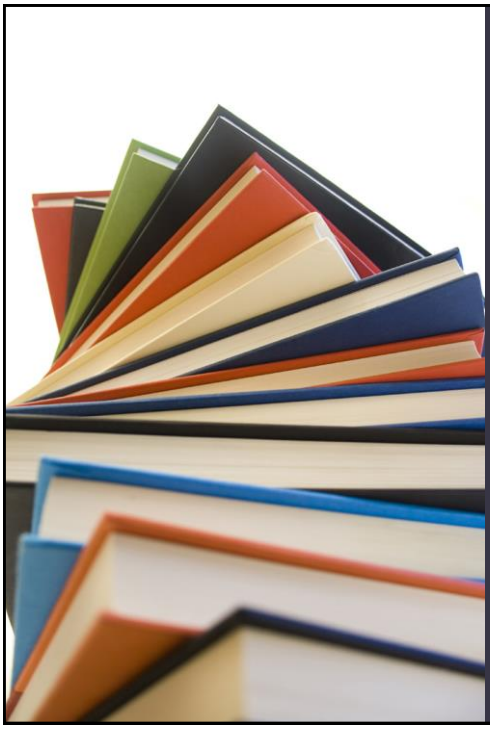


ICD-O-3.2 Coding Intensive

SEER

CDC Centers for Disease Control and Prevention

IACR International Association of Cancer Registries

2019-2020 FCDS Educational Webcast Series

9/22/2019

Steven Peace, CTR

NPCR National Program of Cancer Registries

FCDS

International Agency for Research on Cancer
World Health Organization

Florida HEALTH

ENCR European Network of Cancer Registries

1

ICD-O-3.2 Coding Intensive

CDC & Florida DOH Attribution

"Funding for this conference was made possible (in part) by the Centers for Disease Control and Prevention. The views expressed in written conference materials or publications and by speakers and moderators do not necessarily reflect the official policies of the Department of Health and Human Services, nor does the mention of trade names, commercial practices, or organizations imply endorsement by the US Government."

FCDS would also like to acknowledge the Florida Department of Health for its support of the Florida Cancer Data System, including the development, printing and distribution of materials for the 2019 FCDS Annual Conference and the 2019-2020 FCDS Webcast Series under state contract CODJU. The findings and conclusions in this series are those of the author(s) and do not necessarily represent the official position of the Florida Department of Health.

3

FLccSC LMS – CEU Quiz –FCDS IDEA

- Attendees must take and pass a 3-5 question CEU Quiz to get CEUs
- CEU Awards are Restricted to Attendees with a FLccSC LMS Account
- The CEU Quiz will be posted to FLccSC 1-2 hours after the webcast ends

- Only registered FLccSC Users will be given access to the CEU Quiz
- Florida attendees must have a Florida FLccSC Account to take the Quiz
- South Carolina attendees must have a South Carolina FLccSC Account
- New FLccSC States will follow similar instructions for the CEU Quiz

- Attendees can attend any of the live webcasts without receiving CEUs
- Recorded Sessions are also available for non-FLccSC Users – No CEUs

4

Outline

- Introduction to Session
- SEER is the Authority as ICD-O Standard Setter for United States
- Quick Review of ICD-O-3 Code & Behavior Update
- Quick Review Grade Coding Manual
- REMINDER: USE ICD-O-3 Updates with 2018/2019 Solid Tumor Rules
- ICD-O-3.2 @ IARC/WHO – where to get it & how to use it - DEMO!
- Annual Updates to ICD-O-5 and Solid Tumor Rules from 2021>
- Fast-Paced ICD-O-3 Case Vignette Coding
- Questions

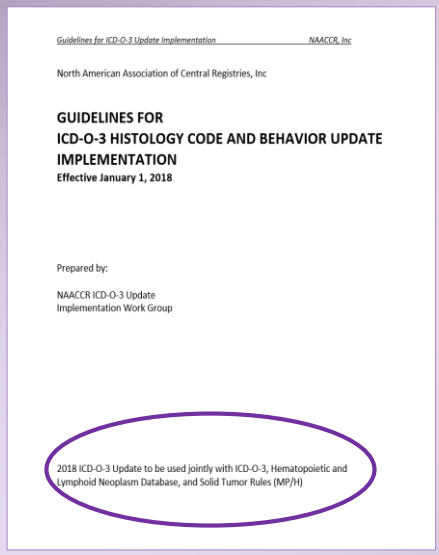
5

Introduction

- SEER is the U.S. Authority for ICD-O, Solid Tumor Rules, Heme DB & Rules
- CAnswer/Ask a Pathologist/CAP/AJCC Manual 8th ed. Are NOT Used in Place of SEER MP/H Rules & Do Not Overrule any SEER/WHO Histology Coding Instructions
- ICD-O is a World Standard for Cancer Registries to Code Primary Site, Histology, Behavior, Grade and Includes Rules for Using the International Classification
- SEER works closely with WHO/IARC/ICD-O to maintain this standard
- United States developed Solid Tumor Rules to be used with ICD-O when the ICD-O-3 was not keeping up with the release of WHO Classification 4th editions
- Beginning 2021 there will be ANNUAL Updates to ICD-O and Solid Tumors Rules
- Always go to SEER Inquiry when you have questions on Histology
- You may not be able to AJCC Stage some cases due to SEER/WHO/IARC Rules

6

ICD-O-3 Code & Behavior Updates



Guidelines for ICD-O-3 Update Implementation *NAACCR, Inc*

North American Association of Central Registries, Inc

GUIDELINES FOR
ICD-O-3 HISTOLOGY CODE AND BEHAVIOR UPDATE IMPLEMENTATION
Effective January 1, 2018

Prepared by:

NAACCR ICD-O-3 Update
Implementation Work Group

2018 ICD-O-3 Update to be used jointly with ICD-O-3, Hematopoietic and Lymphoid Neoplasm Database, and Solid Tumor Rules (MP/H)

December 1, 2017
Summary of changes covered in the 2018 ICD-O-3 Update:

The 2018 ICD-O-3 Update Guidelines includes comprehensive tables listing all changes to ICD-O-3 effective for cases diagnosed 1/1/2018 forward. The guidelines also provide background on the project and issues encountered during review of the WHO Classifications of Tumors. Issues not covered in the 2018 update include reportability of GIST and histology codes with terms that include the words "high grade neoplasia" or "high grade dysplasia" or "severe dysplasia" in digestive system sites.

On an international level, the need was recognized in 2010 for updating the morphology section to accurately code contemporary diagnoses described in the terms of the fourth editions of the World Health Organization's Classifications of Hematopoietic and Lymphoid Neoplasms, Tumors of the Central Nervous System, and Tumors of the Digestive System. In September 2011, the International Agency for Research on Cancer (IARC) and the World Health Organization (WHO) released the document *Updates to the International Classification of Diseases for Oncology, third edition (ICDO-3).*

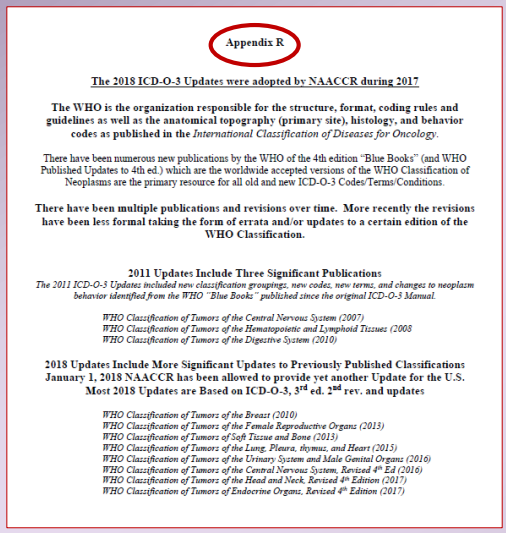
Important information for lung cases: Per WHO 4th Ed Tumors of Lung: In 2011, a new IASLC/ATS/ERS classification of lung adenocarcinoma proposed significant changes to the 2004 WHO classification for resected tumors, including discontinuing the terms bronchioloalveolar carcinoma (BAC).

Beginning with cases diagnosed 1/1/2018 forward, bronchioloalveolar carcinoma (BAC) is no longer the preferred term.

Currently in ICD-O-3, when a topography (C code) is listed in parentheses next to the morphology term, it indicates morphology is most common to that site. It may occur in other sites as well. Many of the new codes, terms, and behaviors listed in this update are site-specific and do not apply to all sites. Applicable C codes will be noted next to the term in **bold** font. These site- and histology-specific combinations will not be added to the "Impossible combination" edit. However, if a site other than the one listed with the morphology code is assigned, the result will be an edit requiring review. This is Interfield Edit 25.

7

ICD-O-3 Code & Behavior Updates



Appendix R

The 2018 ICD-O-3 Updates were adopted by NAACCR during 2017

The WHO is the organization responsible for the structure, format, coding rules and guidelines as well as the anatomical topography (primary site), histology, and behavior codes as published in the *International Classification of Diseases for Oncology*.

There have been numerous new publications by the WHO of the 4th edition "Blue Books" (and WHO Published Updates to 4th ed.) which are the worldwide accepted versions of the WHO Classification of Neoplasms are the primary resource for all old and new ICD-O-3 Codes/Terms/Conditions.

There have been multiple publications and revisions over time. More recently the revisions have been less formal taking the form of errata and/or updates to a certain edition of the WHO Classification.

2011 Updates Include Three Significant Publications
The 2011 ICD-O-3 Updates included new classification groupings, new codes, new terms, and changes to neoplasm behavior identified from the WHO "Blue Books" published since the original ICD-O-3 Manual.

WHO Classification of Tumors of the Central Nervous System (2007)
WHO Classification of Tumors of the Hematopoietic and Lymphoid Tissues (2008
WHO Classification of Tumors of the Digestive System (2010)

2018 Updates Include More Significant Updates to Previously Published Classifications
January 1, 2018 NAACCR has been allowed to provide yet another Update for the U.S. Most 2018 Updates are Based on ICD-O-3, 3rd ed. 2nd rev. and updates

WHO Classification of Tumors of the Breast (2010)
WHO Classification of Tumors of the Female Reproductive Organs (2013)
WHO Classification of Tumors of Soft Tissue and Bone (2013)
WHO Classification of Tumors of the Lung, Pleura, thymus, and Heart (2015)
WHO Classification of Tumors of the Urinary System and Male Genital Organs (2016)
WHO Classification of Tumors of the Central Nervous System, Revised 4th Ed (2016)
WHO Classification of Tumors of the Head and Neck, Revised 4th Edition (2017)
WHO Classification of Tumors of Endocrine Organs, Revised 4th Edition (2017)

FCDS DAM - Appendix R

- NAACCR Guidelines for ICD-O-3 Update
- ICD-O-3 Codes/Terms – Alpha Order
- ICD-O-3 Codes/Terms – Morphology Order
- 2018 ICD-O-3 Updates in Table Format
- 1/10/2018 Summary of Changes
- 4/4/2018 Summary of Changes
- 8/22/2018 Errata

The 2018 ICD-O-3 Update is to be used jointly with the ICD-O-3 Book including All Errata and 2011 Updates, the Hematopoietic & Lymphoid Neoplasm Database, and the Solid Tumor Rules

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ICD-O-3.2 Code & Behavior Updates

- Please note that not all ICD-O updates are approved for us in U.S
- Not all the codes in the first ICD-O were approved in 1977.
- There were at least two or three Provisional ICD-O-2 Coding Manuals before we had the green book in its official format.
- ICD-O-3 was the end of the 2nd edition WHO Classification Series.
- There were some issues not approved in the U.S. in ICD-O-3.1.
- There are still some issues not approved in the U.S. in ICD-O-3.1.
- Similarly, there are a few issues not yet approved in the U.S. for ICD-O-3.2 but we are working on them before we begin ICD-O-5.
- The IARC ICD-O committee has indicated future ICD-O books will reflect the edition of WHO Classification of Neoplasms (aka; the Blue Books) included in the new version. The next version will be ICD-O-5 and will include changes from 5th edition Blue Books.

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ICD-O-3.2 Code & Behavior Updates

- **From: <iacr@iarc.fr>**
  Date: October 8, 2018 at 11:54:58 AM EDT
  To: <glevin@med.miami.edu>, <dlee@med.miami.edu>
  Subject: ANUSAFLO: Announcement from the IARC/WHO ICD-O Committee; IACR 2018 Reminder
- **Dear Colleagues,**

  The IARC/WHO ICD-O Committee has updated the currently recommended ICD-O-3.1 classification. The new version, ICD-O-3.2, will be recommended for use from 2019. These documents are available and will remain open for feedback until 1 November 2018. Please visit the IACR website (newsflash) for more details:

  http://www.iacr.com.fr/index.php?option=com_content&view=article&id=149:icd-o-3-2&catid=80:newsflashes&Itemid=545

  After the consultation period, the final version will be locked and ICD-O-3.2 pdf generated.

  Reminder: Registrations are still open for IACR 2018 Arequipa, Peru this 12-15 November 2018. Details here: www.iacr2018.org

  With thanks and best regards,

  the IACR Secretariat
  www.iacr.com.fr
  www.iacr2018.org
  iacr@iarc.fr

- **ICD-O will treat modified copies of ICD-O tables, manuals and files as copyright infringement if altered or published by AJCC, CAP, or others without approval by IARC/WHO/ICD-O Committee – even for North America.**

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ICD-O-3.2 Code & Behavior Updates

- **WHO Classification of Tumor Books**
  - WHO will release two 5th Ed Blue Books *per year*
  - 5th Ed Tumors of the Eye and 5th Ed Tumors of Digestive System will be released in 2019
  - Schedule for remaining 5th Ed's not currently available
- **2019 forward: ICD-O Work Group process for recommending implementation of ICD-O updates**
  - Involve CAP pathologists in reviewing blue books
  - NCI contractor will create documents comparing the approved version of ICD-O and 5th Ed BB
  - Routine communication with WHO/IARC ICD-O committee chair
  - Work group members will review and discus changes
  - Identify possible issues with 5th Ed changes and send to ICD-O committee chair
  - Issues pertaining to reportability changes and recommendations to implement/adopt new codes, terms, behavior changes will be sent to the ***NAACCR Mid-level Tactical Group*** for approval. Reportability issues will no longer be sent to TAG
- **The Work Group is charged with developing and implementing a procedure for yearly updates based on future 5th Ed BB's.**
  - **Includes dissemination of changes for staging, solid tumor, site/type validation list, registry software vendors, etc.**
- **Current activities (Chair)**
  - Reviewing 5th Ed Endocrine Tumors, Skin Tumors, and Hematopoietic Lymphoid Tumors
  - Creating document for work group to review
  - Ordered 5th Ed Tumors of the Eye
  - Will review ICD-O-3.2 to see if any of the issues we submitted to the committee have been addressed
- **The ICD-O group will start meeting again this summer. I would like to ask that members who have the WHO blue books or have access to them, help with the review and rational prep. If any of you do not wish to continue as a member of the work group, please let me know.**

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ICD-O-3.2 Code & Behavior Updates

- GI Tract High Grade Dysplasia - still not reportable and do not use 8148/2
  - Why is this a problem in the United States
- Not all Thymoma are approved as 'malignant' – yet…still need to vote
- Borderline Ovary/Peritoneal/Fallopian Tube Cancers
- Lobular Carcinoma In Situ – is it cancer or not – treat it or not
- Some of the Site-Associated Cancers are not clearly marked
- Some of the Site-Restricted Cancers are not clearly marked
- Thyroid Classification is still a little dicey
- Hepato-Pancreato-Biliary Cancers are still a bit dicey, too
- There are 143 Histology Terms with the word 'papillary' in them – CAUTION
- Confusion Still over low/high grade pseudomyxoma peritoneii – what if they just call it metastatic mucinous adenocarcinoma of appendix?
- So, there are still outstanding issues – EDITS will take care if many of these.

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ICD-O-3.2 New Histology Codes with New Terms - Only

Status	Histology Value	Behavior	Preferred	label	Reportable	Comments
New term & code	8023	3	FALSE	Midline carcinoma of children and young adults with NUT rearrangement (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	TRUE	NUT carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	FALSE	NUT midline carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8054	3	FALSE	Condylomatous carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8054	3	TRUE	Warty carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8085	3	TRUE	Squamous cell carcinoma, HPV-positive (C01.9, 09.9,C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8086	3	TRUE	Squamous cell carcinoma, HPV-negative (C01.9, C09.9, C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8158	1	FALSE	ACTH-producing tumor	N	Not reportable for all years
New term & code	8158	1	TRUE	Endocrine tumor, functioning, NOS	N	Not reportable for all years
New term & code	8163	3	FALSE	Adenocarcinoma, pancreatobiliary-type (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8163	3	TRUE	Pancreatobiliary-type carcinoma (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8256	3	TRUE	Minimally invasive adenocarcinoma, non-mucinous (C34._)	Y	
New term & code	8257	3	TRUE	Minimally invasive adenocarcinoma, mucinous (C34._)	Y	
New term & code	8265	3	TRUE	Micropapillary carcinoma, NOS (C18._, C19.9, C20.9, C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8265	3	FALSE	Micropapillary adenocarcinoma (C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8339	3	TRUE	Follicular thyroid carcinoma (FTC), encapsulated angoinvasive (C73.9)	Y	
New term & code	8474	3	TRUE	Seromucinous carcinoma (C56.9)	Y	
New term & code	8509	2	TRUE	Solid papillary carcinoma in situ (C50._)	Y	
New term & code	8509	3	TRUE	Solid papillary carcinoma with invasion (C50._)	Y	
New term & code	8519	2	TRUE	Pleomorphic lobular carcinoma in situ (C50._)	Y	ICD-O-3 rule F DOES NOT APPLY to code 8519. Invasive pleomorphic lobular carcinoma is coded 8520/3
New term & code	8552	3	TRUE	Mixed acinar ductal carcinoma	Y	Cases diagnosed prior to 1/1/2018 use code 8523/3
New term & code	8594	1	TRUE	Mixed germ cell sex cord-stromal tumor, unclassified (C48.2, C56.9, C57.9)	N	Not reportable for all years
New term & code	8714	3	FALSE	Malignant perivascular epithelial cell tumor	Y	
New term & code	8714	3	TRUE	PEComa, malignant	Y	
New term & code	8714	3	FALSE	Perivascular epithelioid cell tumor, malignant	Y	
New term/behavior	8815	1	TRUE	Solitary fibrous tumor/hemangiopericytoma Grade 2 (CNS) (C71._)	Y	Reportable for CNS ONLY
New term & code	8975	1	TRUE	Calcifying nested epithelial stromal tumor (C22.0)	N	Not reportable for all years
New term & code	9045	3	TRUE	Biphenotypic sinonasal sarcoma (C30.0, C31.0-C31.3, C31.8, C31.9)	Y	
New term & code	9086	3	TRUE	Germ cell tumors with associated hematological malignancy (C37.9)	Y	
New term & code	9137	3	TRUE	Intimal sarcoma	Y	
New term & code	9137	3	FALSE	Pulmonary artery intimal sarcoma	Y	
New term & code	9385	3	TRUE	Diffuse midline glioma, H3 K27M-mutant (C71._)	Y	
New term & code	9395	3	TRUE	Papillary tumor of pineal region (C75.3)	Y	Cases diagnosed prior to 1/1/2018 use code 9361/3
New term & code	9396	3	TRUE	Ependymoma, RELA fusion-positive (C71._)	Y	
New term & code	9425	3	TRUE	Pilomyxoid astrocytoma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9421/3
New term & code	9431	1	TRUE	Angiocentric glioma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9432	1	TRUE	Pituicytoma (C75.1)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9445	3	TRUE	Glioblastoma, IDH-mutant (C71._)	Y	
New term & code	9475	3	TRUE	Medulloblastoma, WNT-activated (C71._)	Y	
New term & code	9476	3	TRUE	Medulloblastoma, SHH-activated and TP53 mutant (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 3 (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 4 (C71._)	Y	
New term & code	9477	3	TRUE	Medulloblastoma, non-WNT/non-SHH (C71._)	Y	
New term & code	9478	3	FALSE	Embryonal tumor with multilayered rosettes C19MC-altered (C71._)	Y	
New term & code	9478	3	TRUE	Embryonal tumor with multilayered rosettes, NOS (C71._)	Y	
New term & code	9509	1	FALSE	Diffuse leptomeningeal glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	TRUE	Papillary glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	FALSE	Rosette-forming glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9542	3	TRUE	Epithelioid malignant peripheral nerve sheath tumor (C47.0-C47.6, C47.8, C47.9)	Y	

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IARC/WHO and ICD-O-3.2

http://www.iacr.com.fr/index.php?option=com_content&view=article&id=149:icd-o-3-2&catid=80:newsflashes&Itemid=545

ICD-O-3.2

Created on Tuesday, 23 April 2019 14:05

ICD-O-3.2

The IARC/WHO ICD-O Committee[1] has updated the draft ICD-O-3.1 classification, with new morphology codes and terms from the 4th series of WHO Classification of Tumours (Blue Books). The IACR Working Group on ICD-O Updates[2] has compiled a listing of additio changes and revisions between ICD-O-3.1 and ICD-O-3.2 as a reference materi registries.

Both documents have been revised according to the comments received during the consultation period and the final Excel tables are available for download in our Support for registries pages.

The ICD-O-3.2 book in pdf format is in preparation. We would like to thank all registries and individuals for comments provided to the draft versions.

[1] Ian Cree, Jacques Ferlay, Robert Jakob, Brian Rous, Reiko Watanabe, Valerie White, Ariana Znaor
[2] Atul Budukh, Jacques Ferlay, Keri Green, Tomohiro Matsuda, Brian Rous, Ariana Znaor

ICD-O-3

INTERNATIONAL CLASSIFICATION OF DISEASES FOR ONCOLOGY

Third edition Edited by A. Fritz, C. Percy, A. Jack, K. Shanmugaratnam, L. Sobin,D.M. Parkin and S. Whelan

This publication is now available online: http://codes.iarc.fr

ICD-O-3.2 TABLES

The IARC/WHO ICD-O Committee[1] has updated the draft ICD-O-3.1 classification, with new morphology codes and terms from the 4th series of WHO Classification of Tumours (Blue Books). The new version, **ICD-O-3.2, is recommended for use from 2020**. The IACR Working Group on ICD-O Updates[2] has compiled a listing of additions, changes and revisions between ICD-O-3.1 and ICD-O-3.2 as a reference material for cancer registries.

Both documents have been revised according to the comments receive the consultation period and the final tables are available for download here:

A LISTING OF ALL ADDITIONS, CHANGES AND REVISIONS TO THE ICD-O-3 REVISION (ICDO-O-3.1) FOR ICD-O-3.2

ICDO- THIRD EDITION, SECOND REVISION MORPHOLOGY

The ICD-O-3.2 book in pdf format is in preparation. We thank all the individuals and institutions/organizations that provided comments to the draft versions. Their contributions will be acknowledged in the ICD-O-3.2 book, while the individual replies will be provided via email.

[1] Ian Cree, Jacques Ferlay, Robert Jakob, Brian Rous, Reiko Watanabe, Valerie White, Ariana Znaor

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ICD-O-3.2 – complete histology table

http://www.iacr.com.fr/index.php?option=com_content&view=article&id=149:icd-o-3-2&catid=80:newsflashes&Itemid=545

International Agency for Research on Cancer
World Health Organization

ICD-O- Third Edition, Second Revision Morphology

ICDO3.2	Level	Term	Code reference	obs	See also	See r	Includes	Excludes
8173/3	Preferred	Hepatocellular carcinoma, spindle cell variant	(C22.0)					
8173/3	Synonym	Hepatocellular carcinoma, sarcomatoid	(C22.0)					
8174/3	Preferred	Hepatocellular carcinoma, clear cell type	(C22.0)					
8175/3	Preferred	Hepatocellular carcinoma, pleomorphic type	(C22.0)					
8180/3	Preferred	Combined hepatocellular carcinoma and cholangiocarcinoma	(C22.0)					
8180/3	Synonym	Hepatocholangiocarcinoma	(C22.0)					
8180/3	Synonym	Mixed hepatocellular and bile duct carcinoma	(C22.0)					
8190/0	Preferred	Trabecular adenoma						
8190/3	Preferred	Trabecular adenocarcinoma						
8190/3	Synonym	Trabecular carcinoma						
8191/0	Preferred	Embryonal adenoma						
8200/0	Preferred	Eccrine dermal cylindroma	(C44._)					
8200/0	Related	Cylindroma of skin	(C44._)					
8200/0	Related	Cylindroma of breast	(C50._)					
8200/0	Related	Turban tumor	(C44.4)					
8200/3	Preferred	Adenoid cystic carcinoma						
8200/3	Synonym	Cylindroma, NOS		[obs]				(except of skin or breast
8200/3	Synonym	Adenocarcinoma, cylindroid		[obs]				
8200/3	Synonym	Adenocystic carcinoma						
8200/3	Related	Bronchial adenoma, cylindroid	(C34._)	[obs]				
8200/3	Related	Thymic carcinoma with adenoid cystic carcinoma-like features	(C37.9)					
8201/2	Preferred	Cribriform carcinoma in situ	(C50._)					
8201/2	Synonym	Ductal carcinoma in situ, cribriform type	(C50._)					
8201/3	Preferred	Cribriform carcinoma, NOS						
8201/3	Synonym	Ductal carcinoma, cribriform type	(C50._)					
8201/3	Related	Cribriform comedo type carcinoma	(C18._, C19.9, C20.9)					
8201/3	Synonym	Adenocarcinoma, cribriform comedo type	(C18._, C19.9, C20.9)					

Don't forget to Demonstrate How to Use

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ICD-O-3.2 – changes only

IACR
International Association of Cancer Registries

International Agency for Research on Cancer
World Health Organization

A LISTING OF ALL ADDITIONS, CHANGES, AND REVISIONS TO THE INTERNATIONAL CLASSIFICATION OF DISEASES FOR ONCOLOGY, THIRD EDITION, 1st REVISION (ICD-O-3.1) FOR ICD-O-3.2
compiled by the IACR Working Group on ICD-O updates
(Atul Budukh, Keri Green, Jacques Ferlay, Tomohiro Matsuda, Brian Rous, Ariana Znaor; with contribution by Reiko Watanabe, IARC), approved by IARC/WHO ICD-O Committee

Status	Level	Histology	Behavior	Term
New related term	Related	8013	3	Combined large cell neuroendocrine carcinoma
New related term	Related	8020	3	Carcinoma, poorly differentiated, NOS
New related term	Related	8020	3	Anaplastic undifferentiated carcinoma
New related term	Related	8020	3	Dedifferentiated carcinoma
New code and term	Preferred	8023	3	Nuclear protein in testis (NUT) associated carcinoma
New synonym	Synonym	8023	3	NUT carcinoma
New synonym	Synonym	8023	3	NUT midline carcinoma
New synonym	Synonym	8041	3	Small cell carcinoma pulmonary type
New related term	Related	8044	3	Small cell carcinoma, hypercalcemic type (C56.9)
New code and term	Preferred	8054	0	Warty dyskeratoma
Change of code (was 8051/3)	Preferred	8054	3	Warty carcinoma
Change of code (was 8051/3)	Synonym	8054	3	Condylomatous carcinoma
New related term	Related	8054	3	Warty-basaloid carcinoma
New behavior code and term	Preferred	8070	0	Actinic keratosis
New behavior code and term	Related	8070	0	Arsenical keratosis
New behavior code and term	Related	8070	0	PUVA keratosis
New behavior code and term	Preferred	8071	2	Differentiated intraepithelial neoplasia
New behavior code and term	Related	8071	2	Differentiated penile intraepithelial neoplasia (PeIN) (C60._)

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IACR International Association of Cancer Registries

International Agency for Research on Cancer
World Health Organization

A LISTING OF ALL ADDITIONS, CHANGES, AND REVISIONS TO THE INTERNATIONAL CLASSIFICATION OF DISEASES FOR ONCOLOGY, THIRD EDITION, 1st REVISION (ICD-O-3.1) FOR ICD-O-3.2
compiled by the IACR Working Group on ICD-O updates
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with contribution by Reiko Watanabe, IARC), approved by IARC/WHO ICD-O Committee

Status	Level	Histology	Behavior	Term
New related term	Related	8140	3	Parathyroid carcinoma (C75.0)
New related term	Related	8140	3	Acinar adenocarcinoma of prostate (C61.9)
New synonym	Synonym	8144	3	Adenocarcinoma, enteric
New synonym	Synonym	8144	3	Mucinous carcinoma, intestinal type
New term	Preferred	8150	0	Pancreatic neuroendocrine microadenoma (C25.4)
Move to synonym	Synonym	8150	0	Pancreatic endocrine tumor, benign (C25.4)
New preferred term	Preferred	8150	3	Pancreatic neuroendocrine tumor, nonfunctioning (C25.4)
Change of behavior code (from 1)	Synonym	8150	3	Pancreatic endocrine tumor, NOS (C25.4)
Change of behavior code (from 0)	Related	8150	3	Islet cell adenoma (C25.4)
Change of behavior code (from 0)	Related	8150	3	Islet cell adenomatosis (C25.4)
Change of behavior code (from 0)	Related	8150	3	Nesidioblastoma (C25.4)
Change of behavior code (from 1)	Related	8150	3	Islet cell tumor, NOS (C25.4)
Move to related	Related	8150	3	Islet cell adenocarcinoma (25.4)
Move to related	Related	8150	3	Islet cell carcinoma (C25.4)
Change of behavior code (from 0)	Preferred	8151	3	Insulinoma, NOS (C25.4)
Change of behavior code (from 0)	Synonym	8151	3	Beta cell adenoma (C25.4)
Change of behavior code (from 1)	Preferred	8158	3	ACTH-producing tumor
Change of behavior code (from 1)	Related	8158	3	Endocrine tumor, functioning, NOS
New related term	Related	8200	3	Thymic carcinoma with adenoid cystic carcinoma-like features (C37.9)
Change of wording	Preferred	8213	0	Serrated adenoma, NOS (C18._)
Change of wording	Related	8213	0	Sessile serrated adenoma, NOS
New synonym	Synonym	8230	3	Solid adenocarcinoma, NOS
New preferred term	Preferred	8240	3	Neuroendocrine tumor, NOS
Move to synonym	Synonym	8240	3	Carcinoid tumor, NOS
Move to synonym	Synonym	8240	3	Neuroendocrine carcinoma, low grade
Move to synonym	Synonym	8240	3	Neuroendocrine carcinoma, well-differentiated
Change of wording	Related	8240	3	Neuroendocrine tumor, grade 1
Move to synonym	Synonym	8240	3	Typical carcinoid
New synonym	Synonym	8244	3	Mixed carcinoid and adenocarcinoma

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ICD-O-3 Code & Behavior Updates

Site-Restricted/Associated Codes – Many for Only One Site
Appendix R of FCDS DAM

Status	ICD-O-3 Morphology Code	Term	Reportable Y/N	Comments
New Term	8720/3	Meningeal melanoma **(C70. _, C71. _)**	Y	
New Term	8575/3	Metaplastic carcinoma of no special type **(C50. _)**	Y	
New Term	8571/3	Metaplastic carcinoma with chondroid differentiati **0. _)**	Y	
New Term	8571/3	Metaplastic carcinoma with osseous differentiati **0. _)**	Y	
New Term	8575/3	Metaplastic carcinoma with other types mes al differentiation **(C50. _)**	Y	
New Term	8120/3	Microcystic urothelial carcinoma **(C65.9, C6 , C67. _, C68. _)**	Y	
New code/term	8265/3	Micropapillary adenocarcinoma **(C34. _)**	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New code/term	8265/3	**Micropapillary carcinoma, NOS (C18. _, C19.9, C20.9, C34. _)**	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New code/term	8023/3	Midline carcinoma of children and young adults with NUT rearrangement **(C30.0, C31.9, C34. _)**	Y	
New code/term	8257/3	**Minimally invasive adenocarcinoma, mucinous (C34. _)**	Y	
New code/term	8256/3	**Minimally invasive adenocarcinoma, non-mucinous (C34. _)**	Y	

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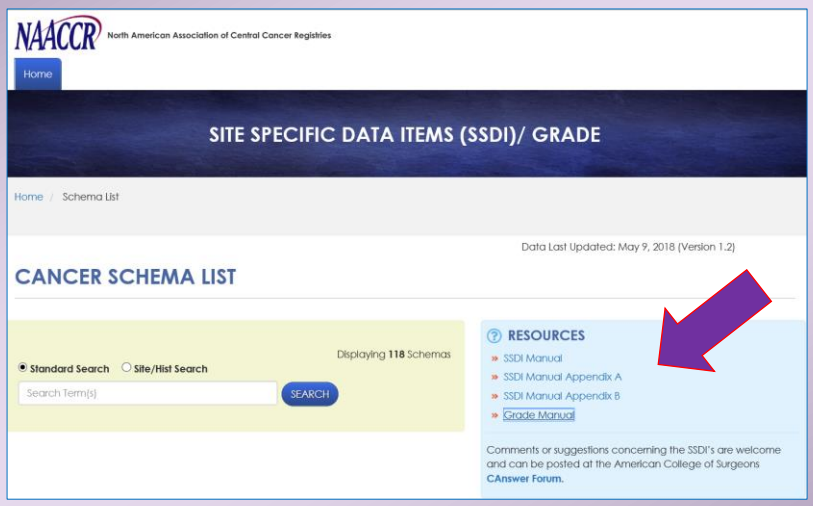
Back to Coding Grade



https://apps.naaccr.org/ssdi/list/

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Back to Coding Grade

- **Clinical Grade** - the grade of a solid primary tumor before any treatment. Treatment may include surgical resection, systemic therapy, radiation therapy, or neoadjuvant therapy. NOTE: All surgical procedures are not treatment, e.g. TURB and endoscopic biopsies.

- **Pathological Grade** - the grade of a solid primary tumor that has been surgically resected and for which no neoadjuvant therapy was administered. If AJCC pathological staging is being assigned, the tumor must have met the surgical resection requirements in the AJCC manual. This may include the grade from the clinical workup, as all information from diagnosis (clinical staging) through the surgical resection is used for pathological staging.

- **Post-Therapy Grade** - the grade of a solid primary tumor that has been resected following neoadjuvant therapy. If AJCC post-therapy staging is being assigned, the tumor must have met the surgical resection requirements for yp in the AJCC manual. Neoadjuvant therapy must meet guidelines or standards, and not be that given for variable or unconventional reasons as noted in the AJCC manual.

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Back to Coding Grade

The tables for grade have been re-structured for 2018. There may be a combination of numeric and alphabetic codes within the same table, according to this template.

Template for a Cancer-Specific Grade Table

Code	Grade Description
1	Site-specific grade system category
2	Site-specific grade system category
3	Site-specific grade system category
4	Site-specific grade system category
5	Site-specific grade system category
L	Low grade
H	High grade
M	Site-specific grade system category
S	Site-specific grade system category
A	Well differentiated
B	Moderately differentiated
C	Poorly differentiated
D	Undifferentiated and anaplastic
8	Not applicable (Hematopoietic neoplasms only)
9	Grade cannot be assessed; Unknown
Blank	(Post-therapy only)

Codes 1-5, L, H, M, S, and 9 all represent AJCC recommended grading systems.

Codes 1-5 are applicable for the AJCC-recommended grading systems. Not all grade tables will have five codes; most will have three or four. GX is coded to 9.

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Back to Coding Grade

General Instructions for the Time Frames for Grade

The three new grade data items reflect the points in time in the patient's care when grade may be assessed. These are similar to the time frames used for assigning AJCC TNM staging.

Grade Clinical

For the Grade Clinical data item, record the grade of a solid primary tumor before any treatment. Treatment may include surgical resection, systemic therapy, radiation therapy, or neoadjuvant therapy. All surgical procedures are not treatment, e.g. TURB and endoscopic biopsies.

Grade Pathological

For the Grade Pathological data item, record the grade of a solid primary tumor that has been surgically resected and for which no neoadjuvant therapy was administered. If AJCC pathological staging is being assigned, the tumor must have met the surgical resection requirements in the AJCC manual. This may include the grade from the clinical workup, as all information from diagnosis (clinical staging) through the surgical resection is used for pathological staging.

Grade Post-Therapy

For the Grade Post-Therapy data item, record the grade of a solid primary tumor that has been resected following neoadjuvant therapy. If AJCC post-therapy staging is being assigned, the tumor must have met the surgical resection requirements for yp in the AJCC manual. Neoadjuvant therapy must meet guidelines or standards, and not be that given for variable or unconventional reasons as noted in the AJCC manual.

This data item corresponds to the yp staging period only.

Clinical Grade - There is grade information before any treatment – from bx.

Pathological Grade - Grade information is from surgical resection of primary site – only.

Post-Therapy Grade - Grade information is from surgery following pre-surgical neo-adjuvant therapy.

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Back to Coding Grade

Cancer Registry Coding of the Cell Indicator or Grade for Hematopoietic and Lymphoid Neoplasms (9590-9992)

Historically the cell lineage indicator (B-cell, T-cell, Null cell, NK-cell) was collected in the Grade data item. Cell lineage indicator/grade for hematopoietic and lymphoid neoplasms will no longer be collected for cases diagnosed 1/1/2018 and forward.

Note: *The Lymphoma Ocular Adnexa chapter in the AJCC manual has a defined grading system for the follicular histologies. Grade is to be assigned to these according to the Lymphoma Ocular Adnexa chapter, chapter 71. The primary sites and follicular histologies included in chapter 71 are as follows.*

- *Applicable primary sites: C441, C690, C695, C696*
- *Applicable histologies: 9690/3, 9691/3, 9695/3, 9698/3*
- *Grade for all other histologies collected in the Lymphoma Ocular Adnexa chapter will be coded to 9*

For all other cases with histologies 9590/3-9992/3, the three grade fields should be coded '8' for not applicable.

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2021 – All Bets are off…

More Data Requirements – NPCR/SEER/CoC

Annual Updates to Solid Tumor Rules

Annual Updates - ICD-O-5

New Research to Add New SSDI and Text Requirements in Diagnostics (Imaging and Histology), Biomolecular Genetics, Lab Tests, Anti-Neoplastic Agents, Radiation Therapy Techniques, Target Agents, etc.

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What you need - right now

- ICD-O-3 Purple Book
- ICD-O-3.2 Excel Table
- 2018 FCDS DAM – Appendix R – ICD-O-3 Updates for 2018
  - New ICD-O-3.2 Histology Codes – Slide #9
- Grade Coding Manual v1.7
  - Schema ID List from Grade Coding Manual v1.7
- 2018 Solid Tumor Rules – July 2019 Revision
- Hematopoietic Database – Live Version
- Internet Access

ICD-O-3 Primary Site
ICD-O-3 Histology
ICD-O-3 Behavior
ICD-O-3 Clinical Grade
ICD-O-3 Pathologic Grade

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Case #1

Upper Outer Quadrant Left Breast Mass, Excision: Papillary carcinoma, solid type with microinvasion. Tumor is completely excised. 2 sentinel lymph nodes negative for tumor.

ICD-O-3 Primary Site	C50.4 (breast – upper outer quadrant)
ICD-O-3 Histology	8509 (solid papillary carcinoma, invasive)
ICD-O-3 Behavior	3 (invasive)
ICD-O-3 Clinical Grade	9 (unknown)
ICD-O-3 Path Grade	9 (unknown)

WHY is the histology code 8509 instead of micropapillary adenocarcinoma (8265) or microinvasive adenocarcinoma (8257) or invasive papillary/micropapillary carcinoma of breast (8507) or some other histology?? Solid Type Papillary - 8509

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ICD-O-3.2 New Histology Codes with New Terms - Only

Status	Histology Value	Behavior	Preferred	label	Reportable	Comments
New term & code	8023	3	FALSE	Midline carcinoma of children and young adults with NUT rearrangement (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	TRUE	NUT carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	FALSE	NUT midline carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8054	3	FALSE	Condylomatous carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8054	3	TRUE	Warty carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8085	3	TRUE	Squamous cell carcinoma, HPV-positive (C01.9, 09.9,C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8086	3	TRUE	Squamous cell carcinoma, HPV-negative (C01.9, C09.9, C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8158	1	FALSE	ACTH-producing tumor	N	Not reportable for all years
New term & code	8158	1	TRUE	Endocrine tumor, functioning, NOS	N	Not reportable for all years
New term & code	8163	3	FALSE	Adenocarcinoma, pancreatobiliary-type (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8163	3	TRUE	Pancreatobiliary-type carcinoma (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8256	3	TRUE	Minimally invasive adenocarcinoma, non-mucinous (C34._)	Y	
New term & code	8257	3	TRUE	Minimally invasive adenocarcinoma, mucinous (C34._)	Y	
New term & code	8265	3	TRUE	Micropapillary carcinoma, NOS (C18._, C19.9, C20.9, C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8265	3	FALSE	Micropapillary adenocarcinoma (C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8339	3	TRUE	Follicular thyroid carcinoma (FTC), encapsulated angoinvasive (C73.9)	Y	
New term & code	8474	3	TRUE	Seromucinous carcinoma (C56.9)	Y	
New term & code	8509	2	TRUE	Solid papillary carcinoma in situ (C50._)	Y	
New term & code	8509	3	TRUE	Solid papillary carcinoma with invasion (C50._)	Y	
New term & code	8519	2	TRUE	Pleomorphic lobular carcinoma in situ (C50._)	Y	ICD-O-3 rule F DOES NOT APPLY to code 8519. Invasive pleomorphic lobular carcinoma is coded 8520/3
New term & code	8552	3	TRUE	Mixed acinar ductal carcinoma	Y	Cases diagnosed prior to 1/1/2018 use code 8523/3
New term & code	8594	1	TRUE	Mixed germ cell sex cord-stromal tumor, unclassified (C48.2, C56.9, C57.9)	N	Not reportable for all years
New term & code	8714	3	FALSE	Malignant perivascular epithelial cell tumor	Y	
New term & code	8714	3	TRUE	PEComa, malignant	Y	
New term & code	8714	3	FALSE	Perivascular epithelioid cell tumor, malignant	Y	
New term/behavior	8815	1	TRUE	Solitary fibrous tumor/hemangiopericytoma Grade 2 (CNS) (C71._)	Y	Reportable for CNS ONLY
New term & code	8975	1	TRUE	Calcifying nested epithelial stromal tumor (C22.0)	N	Not reportable for all years
New term & code	9045	3	TRUE	Biphenotypic sinonasal sarcoma (C30.0, C31.0-C31.3, C31.8, C31.9)	Y	
New term & code	9086	3	TRUE	Germ cell tumors with associated hematological malignancy (C37.9)	Y	
New term & code	9137	3	TRUE	Intimal sarcoma	Y	
New term & code	9137	3	FALSE	Pulmonary artery intimal sarcoma	Y	
New term & code	9385	3	TRUE	Diffuse midline glioma, H3 K27M-mutant (C71._)	Y	
New term & code	9395	3	TRUE	Papillary tumor of pineal region (C75.3)	Y	Cases diagnosed prior to 1/1/2018 use code 9361/3
New term & code	9396	3	TRUE	Ependymoma, RELA fusion-positive (C71._)	Y	
New term & code	9425	3	TRUE	Pilomyxoid astrocytoma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9421/3
New term & code	9431	1	TRUE	Angiocentric glioma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9432	1	TRUE	Pituicytoma (C75.1)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9445	3	TRUE	Glioblastoma, IDH-mutant (C71._)	Y	
New term & code	9475	3	TRUE	Medulloblastoma, WNT-activated (C71._)	Y	
New term & code	9476	3	TRUE	Medulloblastoma, SHH-activated and TP53 mutant (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 3 (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 4 (C71._)	Y	
New term & code	9477	3	TRUE	Medulloblastoma, non-WNT/non-SHH (C71._)	Y	
New term & code	9478	3	FALSE	Embryonal tumor with multilayered rosettes C19MC-altered (C71._)	Y	
New term & code	9478	3	TRUE	Embryonal tumor with multilayered rosettes, NOS (C71._)	Y	
New term & code	9509	1	FALSE	Diffuse leptomeningeal glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	TRUE	Papillary glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	FALSE	Rosette-forming glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9542	3	TRUE	Epithelioid malignant peripheral nerve sheath tumor (C47.0-C47.6, C47.8, C47.9)	Y	

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Use Both Appendix R and ICD-O-3.2

Ovary: Uterus, cervix, left tube and left ovary showing serous neoplasm of low malignant potential (serous borderline tumor) Micropapillary Type, 2cm in Size. No Surface involvement identified. 8 pelvic nodes negative, FIGO Stage IB

Is this Case Reportable?

What is Histology/Behavior?

But, it states low malignant potential (/1)?

Does borderline /1 conflict with non-invasive /2 code?

Which Reference(s) Must You Use? Why?

How can the stage = FIGO IB and behavior of tumor is /1 or /2?

Or is this really a malignant neoplasm with behavior = /3?

Reportability determined by terminology used: non-invasive versus borderline.

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Use Both Appendix R and ICD-O-3.2

This is not reportable – 8460/2 – serous borderline tumor, micropapillary variant.
ICD-O-3.2 does not tell you if something is reportable or not. It just gives a code.

8460/2	8460	2	Preferred	Serous borderline tumor, micropapillary variant	(C56.9)

BUT, Appendix R does tell you that this is reportable…see notes below…

Status	ICD-O-3 Morphology Code	Term	Reportable Y/N	Comments
New code/term	8552/3	**Mixed acinar ductal carcinoma**	Y	Cases diagnosed prior to 1/1/2018 use code 8523/3
New code/term	8594/1	**Mixed germ cell sex cord-stromal tumor, unclassified (C48.2, C56.9, C57.9)**	N	
New Term	8254/3	**Mixed invasive mucinous and non-mucinous adenocarcinoma (C34. _)**	Y	
New Term	8482/3	Mucinous carcinoma, gastric type **(C53. _)**	Y	
New Term	8144/3	Mucinous carcinoma, intestinal type **(C53. _)**	Y	
New Term	8470/3	Mucinous cystic tumor with associated invasive carcinoma **(C25. _)**	Y	
New Term	8480/3	Mucinous tubular and spindle cell carcinoma **(C64.9)**	Y	
New Term	8933/3	Mullerian adenosarcoma **(C54. _, C55.9)**	Y	
Behavior Code/term	8811/1	Myxoinflammatory fibroblastic sarcoma (MIFS) **(C49. _)**	N	
New Term	8120/3	Nested urothelial carcinoma **(C65.9, C66.9, C67. _, C68. _)**	Y	
New Term	8041/3	Neuroendocrine carcinoma, poorly differentiated **(C50. _)**	Y	
New Term	8246/3	Neuroendocrine tumor, well differentiated **(C50. _)**	Y	
New Term	8343/2	Non-invasive EFVPTC **(C73.9)**	Y	Cases diagnosed 1/1/2017 forward
New Term	8343/2	Non-invasive encapsulated follicular variant of papillary thyroid carcinoma (non-invasive EFVPTC) **(C73.9)**	Y	Cases diagnosed 1/1/2017 forward
New Term	8343/2	Non-invasive follicular thyroid neoplasm with papillary-like nuclear features (NIFTP) **(C73.9)**	Y	Cases diagnosed 1/1/2017 forward
New Term	8343/2	Non-invasive FTP **(C73.9)**	Y	Cases diagnosed 1/1/2017 forward
Behavior Code/term	8460/2	Non-invasive low grade serous carcinoma **(C56.9)**	Y	
New Term	8500/2	Non-invasive mammary carcinoma **(C50. _)**	Y	
New code/term	8023/3	**NUT carcinoma (C30.0, C31.9, C34. _)**	Y	

Status	ICD-O-3 Morphology Code	Term	Reportable Y/N	Comments
New code/term	9137/3	Pulmonary artery intimal sarcoma	Y	
Behavior Code/term	8842/3	Pulmonary myxoid sarcoma with EWSR1-CREB1 translocation **(C34. _)**	Y	
New Term	8312/3	Renal cell carcinoma, unclassified **(C64.9)**	Y	
New Term	8510/3	Renal medullary carcinoma **(C64.9)**	Y	
New code/term	9509/1	Rosette-forming glioneuronal tumor **(C71. _)**	Y	
New Term	8500/3	Salivary duct carcinoma **(C06.9, C08.9)**	Y	
New Term	8840/3	Sclerosing epithelioid fibrosarcoma	Y	
New Term	8912/3	Sclerosing rhabdomyosarcoma	Y	
New code/term	8474/3	**Seromucinous carcinoma (C56.9)**	Y	
Behavior Code/term	8460/2	Serous borderline tumor-micropapillary variant **(C56.9)**	N	Not reportable for 2018
Behavior Code/term	8441/2	Serous endometrial intraepithelial carcinoma **(C54. _, C55.9)**	Y	
Behavior Code/term	8441/2	Serous tubal intraepithelial carcinoma **(C57.0)**	Y	
Behavior code/term	8213/3	**Serrated adenocarcinoma (C18.0, C18.2, C18.9, C19.9, C20.9)**	Y	
New Term	8041/3	Small cell carcinoma pulmonary type **(C56.9)**	Y	
New Term	8044/3	Small cell carcinoma, hypercalcemic type **(C56.9)**	Y	
New code/term	8509/2	**Solid papillary carcinoma in situ (C50. _)**	Y	
New code/term	8509/3	**Solid papillary carcinoma with invasion (C50. _)**	Y	
New Term	8815/0	Solitary fibrous tumor/hemangiopericytoma Grade 1 (CNS) **(C71. _)**	Y	Reportable for CNS
Behavior code/term	8815/1	**Solitary fibrous tumor/hemangiopericytoma Grade 2 (CNS) (C71. _)**	Y	Reportable for CNS ONLY

Case #2

Subareolar Breast Mass, Excision: Intraductal carcinoma (DCIS) with cribriform, clear cell, micropapillary, and lobular features. Tumor is completely excised. 2 sentinel lymph nodes negative for tumor.

ICD-O-3 Primary Site	C50.1 (central breast - subareolar)
ICD-O-3 Histology	8500 (ductal carcinoma, NST)
ICD-O-3 Behavior	2 (non-invasive/in-situ)
ICD-O-3 Clinical Grade	9 (unknown)
ICD-O-3 Path Grade	9 (unknown)

3. **DCIS/Carcinoma NST in situ** has a major classification change.
   A. Subtypes/variant, architecture, pattern, and features **ARE NOT CODED**. The majority of in situ tumors will be coded to DCIS **8500/2.**
   B. It is very important to code the grade of all DCIS.
      i. Code grade as designated in current AJCC Manual, SEER Coding Manual, and COC Coding Manual.
      ii. The current breast **WHO** edition emphasizes coding the **grade** of tumor rather than the **subtype/variant**.
      iii. The WHO editions are used internationally by pathologists to keep their nomenclature and histology identification current.
      iv. Over time, **subtypes/variants** will be diagnosed **less frequently.**

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Case #3

Descending Colon @ 75cm: Biopsy followed by complete resection - adenocarcinoma, poorly differentiated, with mucinous features arising in a large 8cm adenomatous polyp with invasion through the muscularis mucosae into the pericolic fat. 4 of 8 mesenteric lymph nodes are positive for metastatic adenocarcinoma. pT3 pN2a

ICD-O-3 Primary Site	C18.6 (descending colon)
ICD-O-3 Histology	8140 (adenocarcinoma, NOS)
ICD-O-3 Behavior	3 (invasive/malignant)
ICD-O-3 Clinical Grade	3 (poorly differentiated)
ICD-O-3 Path Grade	3 (poorly differentiated)

Polyps are now **disregarded** when coding histology. For example, adenocarcinoma in an adenomatous polyp is coded as adenocarcinoma 8140. For the purposes of determining multiple primaries, tumors coded as adenocarcinoma in a polyp for pre-2018 cases should be treated as adenocarcinoma 8140.

Code the histology described as **differentiation** or **features/features of ONLY** when there is a specific ICD-O code for the "NOS with _____ features" or "NOS with _____ differentiation".
Note: Do not code differentiation or features when there is no specific ICD-O code.

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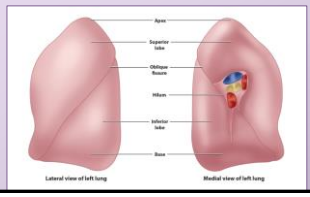
Case #4

Apex Left Lung, biopsy: minimally invasive mucinous adenocarcinoma. Wedge resection shows well-differentiated mucinous adenocarcinoma. 3 mediastinal nodes negative. No further workup.

ICD-O-3 Primary Site	C34.1 (apex is at the top of the lung)
ICD-O-3 Histology	8257 (new histology code just for lung)
ICD-O-3 Behavior	3 (any invasion is invasive cancer)
ICD-O-3 Clinical Grade	9 (no grade given on biopsy)
ICD-O-3 Path Grade	1 (well-differentiated on resection)

Used to be called Mucinous Broncho-alveolar Carcinoma



Now called Minimally Invasive Mucinous Adenocarcinoma

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ICD-O-3.2 New Histology Codes with New Terms - Only

Status	Histology Value	Behavior	Preferred	label	Reportable	Comments
New term & code	8023	3	FALSE	Midline carcinoma of children and young adults with NUT rearrangement (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	TRUE	NUT carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	FALSE	NUT midline carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8054	3	FALSE	Condylomatous carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8054	3	TRUE	Warty carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8085	3	TRUE	Squamous cell carcinoma, HPV-positive (C01.9, 09.9,C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8086	3	TRUE	Squamous cell carcinoma, HPV-negative (C01.9, C09.9, C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8158	1	FALSE	ACTH-producing tumor	N	Not reportable for all years
New term & code	8158	1	TRUE	Endocrine tumor, functioning, NOS	N	Not reportable for all years
New term & code	8163	3	FALSE	Adenocarcinoma, pancreatobiliary-type (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8163	3	TRUE	Pancreatobiliary-type carcinoma (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8256	3	TRUE	Minimally invasive adenocarcinoma, non-mucinous (C34._)	Y	
New term & code	8257	3	TRUE	Minimally invasive adenocarcinoma, mucinous (C34._)	Y	
New term & code	8265	3	TRUE	Micropapillary carcinoma, NOS (C18._, C19.9, C20.9, C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8265	3	FALSE	Micropapillary adenocarcinoma (C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8339	3	TRUE	Follicular thyroid carcinoma (FTC), encapsulated angoinvasive (C73.9)	Y	
New term & code	8474	3	TRUE	Seromucinous carcinoma (C56.9)	Y	
New term & code	8509	2	TRUE	Solid papillary carcinoma in situ (C50._)	Y	
New term & code	8509	3	TRUE	Solid papillary carcinoma with invasion (C50._)	Y	
New term & code	8519	2	TRUE	Pleomorphic lobular carcinoma in situ (C50._)	Y	ICD-O-3 rule F DOES NOT APPLY to code 8519. Invasive pleomorphic lobular carcinoma is coded 8520/3
New term & code	8552	3	TRUE	Mixed acinar ductal carcinoma	Y	Cases diagnosed prior to 1/1/2018 use code 8523/3
New term & code	8594	1	TRUE	Mixed germ cell sex cord-stromal tumor, unclassified (C48.2, C56.9, C57.9)	N	Not reportable for all years
New term & code	8714	3	FALSE	Malignant perivascular epithelial cell tumor	Y	
New term & code	8714	3	TRUE	PEComa, malignant	Y	
New term & code	8714	3	FALSE	Perivascular epithelioid cell tumor, malignant	Y	
New term/behavior	8815	1	TRUE	Solitary fibrous tumor/hemangiopericytoma Grade 2 (CNS) (C71._)	Y	Reportable for CNS ONLY
New term & code	8975	1	TRUE	Calcifying nested epithelial stromal tumor (C22.0)	N	Not reportable for all years
New term & code	9045	3	TRUE	Biphenotypic sinonasal sarcoma (C30.0, C31.0-C31.3, C31.8, C31.9)	Y	
New term & code	9086	3	TRUE	Germ cell tumors with associated hematological malignancy (C37.9)	Y	
New term & code	9137	3	TRUE	Intimal sarcoma	Y	
New term & code	9137	3	FALSE	Pulmonary artery intimal sarcoma	Y	
New term & code	9385	3	TRUE	Diffuse midline glioma, H3 K27M-mutant (C71._)	Y	
New term & code	9395	3	TRUE	Papillary tumor of pineal region (C75.3)	Y	Cases diagnosed prior to 1/1/2018 use code 9361/3
New term & code	9396	3	TRUE	Ependymoma, RELA fusion-positive (C71._)	Y	
New term & code	9425	3	TRUE	Pilomyxoid astrocytoma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9421/3
New term & code	9431	1	TRUE	Angiocentric glioma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9432	1	TRUE	Pituicytoma (C75.1)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9445	3	TRUE	Glioblastoma, IDH-mutant (C71._)	Y	
New term & code	9475	3	TRUE	Medulloblastoma, WNT-activated (C71._)	Y	
New term & code	9476	3	TRUE	Medulloblastoma, SHH-activated and TP53 mutant (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 3 (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 4 (C71._)	Y	
New term & code	9477	3	TRUE	Medulloblastoma, non-WNT/non-SHH (C71._)	Y	
New term & code	9478	3	FALSE	Embryonal tumor with multilayered rosettes C19MC-altered (C71._)	Y	
New term & code	9478	3	TRUE	Embryonal tumor with multilayered rosettes, NOS (C71._)	Y	
New term & code	9509	1	FALSE	Diffuse leptomeningeal glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	TRUE	Papillary glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	FALSE	Rosette-forming glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9542	3	TRUE	Epithelioid malignant peripheral nerve sheath tumor (C47.0-C47.6, C47.8, C47.9)	Y	

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Case #5

Breast UOQ mass excision: mixed squamous cell carcinoma and small cell carcinoma, extending to the excisional margin with rare foci of high grade ductal carcinoma in situ, micropapillary, papillary and solid architectures, nuclear grade 2/3 identified. Tumor size 7.5 x 6.8 x 6cm. No nodes identified.

ICD-O-3 Primary Site	C50.4 (UOQ Breast)
ICD-O-3 Histology	8045 (combined small cell carcinoma - small cell carcinoma combined with squamous cell carcinoma
ICD-O-3 Behavior	3
ICD-O-3 Clinical Grade	9 (high grade and nuclear grade for insitu)
ICD-O-3 Path Grade	9 (high grade and nuclear grade for insitu)

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Case #5

Table 2: Histology Combination Codes

Table 2 does not have an entry for this combination. So, you check back to basic ICD-O-3 codes and find the existing 8045/3 code combined small cell carcinoma which includes small cell carcinoma combined with squamous cell carcinoma – no specific rule in ICD-O-3 Book or in the Solid Tumor Rules – Basic ICD-O-3 coding – don't forget the basics of coding – they still apply!!

Code a **combination code** when there are **two histologies** (two components) within a single tumor and the majority histology is unknown/not documented.
Note 1: Use Table 2 in the Equivalent Terms and Definitions to identify valid combination codes.
Note 2: The rules are hierarchical, so the tumors are **NOT** a NOS/NST and a single subtype/variant.
Note 3: The diagnosis may be two subtypes/variants and the pathologist may mention the presence of duct/carcinoma NST. Ignore the mention of carcinoma NST.
Note 4: **Do not** use a combination code when the second histology is described as **features or differentiation** unless it is part of the preferred term.
Note 5: The histologies may be identified as:
- Mixed histologies
- Combination histologies
- Histology 1 **AND** histology 2
- Histology 1 **WITH** histology 2

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Case #6

Resection RUL Lung shows well differentiated lepidic predominant invasive bronchoalveolar adenocarcinoma with mucinous features, neg margins

ICD-O-3 Primary Site	C34.1 (Upper lobe, right lung)
ICD-O-3 Histology	8250 (lepidic adenocarcinoma)
ICD-O-3 Behavior	3 (malignant)
ICD-O-3 Clinical Grade	9 (no clinical grade – no biopsy)
ICD-O-3 Path Grade	1 (well differentiated)

Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Adenocarcinoma 8140 **Note 1:** Mucinous adenocarcinoma for lung only is coded as follows: • **8253/3*** when o Behavior unknown/not documented (use staging form to determine behavior when available) o Invasive • **8257/3*** when o Microinvasive o Minimally invasive • **8253/2*** when o Preinvasive o In situ	Adenocarcinoma NOS Adenocarcinoma in situ **8140/2** Adenocarcinoma invasive **8140/3** Adenocarcinoma, non-mucinous, NOS	Acinar adenocarcinoma/adenocarcinoma, acinar predominant **(for lung only) 8551*** Adenoid cystic/adenocystic carcinoma **8200** Colloid adenocarcinoma **8480** Fetal adenocarcinoma **8333** Lepidic adenocarcinoma/adenocarcinoma, lepidic predominant **8250/3*** Mucinous carcinoma/adenocarcinoma **(for lung only)** in situ **8253/2*** invasive **8253/3*** minimally invasive **8257/3*** microinvasive **8257/3*** preinvasive **8253/2***

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Case #7

Right Occipital Brain Mass, biopsy: high grade neuroendocrine carcinoma, large cell type.

- Immunohistochemistry stains for CK 7, CK 20, TTF-1, P40, synaptophysin, chromogranin, and CD 56.
- CK 7: strongly and diffusely positive
- CK 20: negative
- TTF-1: strongly and diffusely positive
- P40: negative
- Synaptophysin: strongly and diffusely positive
- Chromogranin: strongly and focally positive
- CD 56: strongly and diffusely positive

ICD-O-3 Primary Site	C80.9 (unknown primary)
ICD-O-3 Histology	8013 (large cell neuroendocrine carcinoma
ICD-O-3 Behavior	3
ICD-O-3 Clinical Grade	9 (never code grade from metastatic site)
ICD-O-3 Path Grade	9 (never code grade from metastatic site)

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Case #7

General Grade Coding Instructions for Solid Tumors

Listed below are general guidelines for coding all three new grade data items.

1. Code the grade from the primary tumor only
   a. Do NOT code grade based on metastatic tumor or recurrence. In the rare instance that tumor tissue extends contiguously to an adjacent site and tissue from the primary site is not available, code grade from the contiguous site
   b. If primary site is unknown, code grade to 9.
2. If there is more than one grade available for an individual grade data item (i.e. within the same time frame)
   a. Priority goes to the recommended AJCC grade listed in the applicable AJCC chapter
      i. If none of the specified grades are from the recommended AJCC grade system, record the highest grade
   b. If there is no recommended AJCC grade, code the highest grade
3. In situ and/or combined in situ/invasive components:
   a. If a grade is given for an in situ tumor, code it. Do NOT code grade for dysplasia such as high-grade dysplasia.
   b. If there are both in situ and invasive components, code only the grade for the invasive portion even if its grade is unknown.
4. Systemic treatment and radiation can alter a tumor's grade. Therefore, it is important to code clinical grade based on information prior to neoadjuvant therapy even if grade is unknown during the clinical timeframe. Grade can now be collected in grade post-therapy cases when grade is available from post-neoadjuvant surgery

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Case #8

Right Lung Hilus FNA: NSCLC (non-small cell carcinoma of lung).

ALK+, EGRF+, ROS1 neg, PD-L1 positive – favor adenocarcinoma of probable large cell type

ICD-O-3 Primary Site	C34.0 (lung, hilum)
ICD-O-3 Histology	8012 (large cell adenocarcinoma)
ICD-O-3 Behavior	3 (malignant)
ICD-O-3 Clinical Grade	9 (no grade noted on FNA)
ICD-O-3 Path Grade	9 (no resection of primary tumor)

NSCLC is usually adenocarcinoma, squamous cell carcinoma, or large-cell carcinoma. See the instructions for coding histology when NSCLC is the diagnosis.

Note 5: For those sites/histologies which have recognized **biomarkers**, the biomarkers are most frequently used to target treatment. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries and/or histologic type. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

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Case #8

The 2015 WHO Classification of Neoplasms of the Lung and the work of the International Association for the Study of Lung Cancer (IASLC) states; "One of the great advances in the past decade in lung cancer diagnosis and treatment is the concept of personalized medicine, where therapeutic decisions are based on the specific histologic and genetic characteristics of the patient's tumor.

This has given a new importance for pathologists to classify NSCLC further into specific pathologic subtypes (e.g., adenocarcinoma versus squamous cell carcinoma) as this determines eligibility for certain types of molecular testing and therapeutic strategies.

NSCLC can be squamous cell carcinoma, adenocarcinoma or large cell carcinoma or even large cell neuroendocrine carcinoma – and any of the subtypes of these NOS terms/types. NSCLC just means that the histology is NOT small cell carcinoma…so, it is not even a real histology by itself.

NSCLC has NEVER been an actual histology – it was only introduced because so many physicians were using it back in the day that registrars needed a code. All it means is that the tumor is 'not small cell carcinoma'…which means it could be anything else…just not small cell.

When pathologists use the NSCLC terminology, they are supposed to run genetic tests to rule out adenocarcinoma or squamous cell carcinoma, NOS. You are never supposed to change a histology to 'fit' the rules of another manual or reference - so, if the genetic tests are run and find nothing and rerun and nothing - then use 8046/3 per H3 Rule.

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Case #9

Right Lung FNA: NSCLC (non-small cell carcinoma of lung). No molecular testing performed

ICD-O-3 Primary Site	C34.9
ICD-O-3 Histology	8046 (FCDS will Override/FORCE the histology)
ICD-O-3 Behavior	3
ICD-O-3 Clinical Grade	9
ICD-O-3 Path Grade	9

IMPORTANT: WHO and IASLC both recognize that the percentage of cases diagnosed as NSLCL can be as high as 30%-50% in some institutions and the NSCLC, NOS diagnosis has only been rising over time. The importance of distinguishing from NSCLC to specific type of NSCLC is critical to determine major therapeutic implications based on molecular type and characteristics that distinguish squamous cell carcinoma from adenocarcinoma from small cell carcinoma lung cancer. *NOTE: DO NOT CHANGE TO 8010 as noted in CAnswer. If you do, we completely lose track of pathologist continued use of 8046/3.*

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TABLE 2.1. Currently Recommended Molecular Testing for NSCLC

BIOMARKER	TEST DETECTS	WHEN	TECHNOLOGY	RECOMMENDATIONS	EVIDENCE	CANCER TYPE
ALK	Gene fusion	Metastatic workup	FISH, NGS, RT-PCR[a]	Response to oral ALK TKIs; alectinib has improved efficacy over crizotinib in first line	High-level, wide acceptance	Adenocarcinoma, large cell, NSCLC NOS
	Fusion protein expression		IHC[b]			
		Together with *EGFR* testing in "never smokers" or small/mixed histology specimens		Response to oral ALK TKIs, eg, crizotinib	Lower level, wide acceptance	Squamous cell
EGFR T790M	Mutation	Metastatic workup	NGS, multiple mutation testing	Resistant to EGFR TKIs	High-level, wide acceptance	Adenocarcinoma, large cell, NSCLC NOS
EGFR exon 21 (L858R, L861), exon 20 (S768I), exon 18 (G719X, G719)	Mutation	Metastatic workup	NGS, multiple mutation testing	Sensitive to EGFR TKIs	High-level, wide acceptance	Adenocarcinoma, large cell, NSCLC NOS
					Lower level, wide acceptance	Squamous cell
EGFR exon 19	Deletion	Metastatic workup	NGS, multiple mutation testing	Sensitive to EGFR TKIs	High-level, wide acceptance	Adenocarcinoma, large cell, NSCLC NOS
					Lower level, wide acceptance	Squamous cell
EGFR exon 20 7p12	Insertion mutation	Metastatic workup	NGS, multiple mutation testing	Likely resistant to EGFR TKIs	High-level, wide acceptance	Adenocarcinoma, large cell, NSCLC NOS
					Lower level, wide acceptance	Squamous cell
ROS1	Fusion rearrangement	Metastatic workup	NGS, FISH, RT-PCR	Responsive to ROS1 TKIs	Lower level, wide acceptance	Adenocarcinoma, large cell, squamous cell, NSCLC NOS
PD-L1	Protein expression ≥50%	Metastatic workup	NGS, multiple mutation testing	Response to pembrolizumab in first-line; FDA approved treatment[15]	Lower level, wide acceptance	Adenocarcinoma, large cell, NSCLC, squamous cell NOS
KRAS	Mutation	Metastatic workup	Gene sequencing	Resistance to EGFR TKIs. Gives poor prognosis compared with *KRAS* wt	Lower level, wide acceptance	All NSCLC
BRAF	Mutation, V600E	Metastatic workup	NGS, pyrosequencing, AS-PCR	Emerging targeted agents[18]; responsive to combined BRAF and MEK inhibition	Lower level, wide acceptance	All NSCLC
HER2	Mutation	Anytime	NGS, multiple mutation testing	Emerging targeted agents[20]	Lower level, limited acceptance	All NSCLC
MET	Amplification, mutation	Anytime	NGS, FISH	Emerging targeted agents[21]	Lower level, wide acceptance	All NSCLC
RET	Fusion, rearrangement	Anytime	NGS, FISH, RT-PCR	Emerging targeted agents[22,23]	Lower level, wide acceptance	All NSCLC

Abbreviations: AS-PCR, allele-specific polymerase chain reaction; FDA, US Food and Drug Administration; FISH, fluorescence in situ hybridization; IHC, immunohistochemistry; NGS, next-generation sequencing; NOS, not otherwise specified; NSCLC, non-small cell lung cancer; PD-L1, programmed death 1 ligand; RT-PCR, reverse transcription-polymerase chain reaction; TKIs, tyrosine kinase inhibitors; wt, wild type.
[a]FISH is the US Food and Drug Administration-approved method for *ALK* gene rearrangement. NGS and RT-PCR currently are not used widely in clinical practice.
[b]IHC can be used as a good alternative to FISH.[24]

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NSCLC Advances in Diagnostic Tests

- 2015 WHO Classification from the International Association for the Study of Lung Cancer (IASLC) states:
- "One of the great advances in the past decade in lung cancer diagnosis and treatment is the concept of personalized medicine, where therapeutic decisions are based on the specific histologic and genetic characteristics of the patient's tumor. This has given a new importance for pathologists to classify NSCLC further into specific pathologic subtypes (e.g., adenocarcinoma versus squamous cell carcinoma) as this determines eligibility for certain types of molecular testing and therapeutic strategies.
- Until the past decade, there have been no therapeutic implications to classify the NSCLC tumors further, so little attention was been given to the distinction of adenocarcinoma and squamous cell carcinoma in small tissue samples.
- This situation changed dramatically with the discovery of several therapeutic options that are only approved for treatment of patients with specific histologic types. Discovery that epidermal growth factor (EGFR) mutations and anaplastic lymphoma kinase (ALK) rearrangements are effective targets for EGFR tyrosine kinase inhibitors or ALK inhibitors in patients with advanced lung adenocarcinoma has not only revolutionized therapeutic strategies, but transformed clinical practice for pathologists.
- Further, the IASLC has provided new criteria for the diagnosis of lung cancer based on small biopsies and cytology as the clinical importance for identification of specific treatment options by histologic type rather than just NSCLC has risen in importance and needs to be managed carefully for molecular testing.

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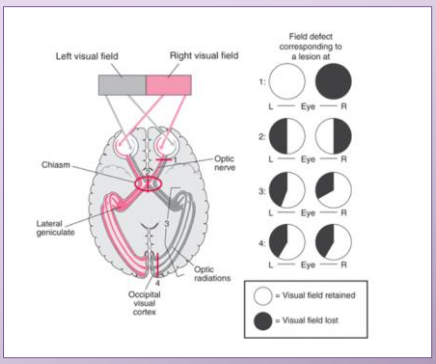
Case #10

Optic Nerve Biopsy – optic pathway glioma in patient with neurofibromatosis type 1 (NF1) and near total vision loss

ICD-O-3 Primary Site	C72.3 (optic nerve)
ICD-O-3 Histology	9421
ICD-O-3 Behavior	1
ICD-O-3 Clinical Grade	9
ICD-O-3 Path Grade	9



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Optic Pathway Glioma

- Optic pathway glioma is now classified along with Optic glioma/pilocytic astrocytoma and is classified as borderline malignancy according to Solid Tumor Rules with histology 9421/1.
- Optic pathway gliomas (OPGs) occur in 15%-20% of children with neurofibromatosis type 1 (NF1), leading to visual deficits in fewer than half of these individuals. The goal of chemotherapy is to preserve vision, but vision loss in NF1-associated OPG can be unpredictable.
- Neurofibromatosis type 1 (NF1) is one of the most common brain tumor predisposition syndromes, in which affected children are prone to the development of low-grade gliomas. While NF1-associated gliomas can be found in several brain regions, the majority arise in the optic nerves, chiasm, tracts, and radiations (optic pathway gliomas; OPGs).
- Owing to their location, 35-50% of affected children present with reduced visual acuity. Unfortunately, despite tumor stabilization following chemotherapy, vision does not improve in most children..
- The implementation of these models for drug discovery and validation has galvanized molecularly-targeted clinical trials in children with NF1-OPG. Future research focused on defining the cellular and molecular factors that underlie optic glioma development and progression also has the potential to provide personalized risk assessment strategies for this pediatric population.

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Case #11

Biopsy of Scalp – spindle cell neoplasm favor malignant fibrous histiocytoma, low grade

ICD-O-3 Primary Site	C49.0 (soft tissue scalp) – NOT SKIN of SCALP
ICD-O-3 Histology	8830 (MFH – malignant fibrous histiocytoma)
ICD-O-3 Behavior	3
ICD-O-3 Clinical Grade	L
ICD-O-3 Path Grade	9

Code	Grade Description
1	WHO Grade I: Circumscribed tumors of low proliferative potential associated with the possibility of cure following resection
2	WHO Grade II: Infiltrative tumors with low proliferative potential with increased risk of recurrence
3	WHO Grade III: Tumors with histologic evidence of malignancy, including nuclear atypia and mitotic activity, associated with an aggressive clinical course
4	WHO Grade IV: Tumors that are cytologically malignant, mitotically active, and associated with rapid clinical progression and potential for dissemination
L	Stated as "low grade" NOS
H	Stated as "high grade" NOS
A	Well differentiated
B	Moderately differentiated
C	Poorly differentiated
D	Undifferentiated, anaplastic
9	Grade cannot be assessed; Unknown

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MFH Soft Tissue of Scalp – NOT SKIN

- Tumor is not a spindle cell carcinoma or melanoma of skin
- Tumor is soft tissue sarcoma of Head and Neck
- There is no AJCC Chapter for this tumor in this anatomic location
- You must assign SS2018 – You cannot assign AJCC TNM 8th ed.

Primary Site	Histology	AJCC ID	Description
C000-C148, C150, C153, C158, C300-C329, C470, C490, C722, C724-C725, C739, C750-C755, C758-C759	8711, 8800-8802, 8810-8811, 8815, 8825, 8832-8833, 8840, 8850, 8852, 8854, 8858, 8890, 8901, 8912, 9040-9041, 9043, 9133, 9136, 9180, 9251, 9364, 9540, 9542, 9561, 9580	40	Soft Tissue Sarcoma of the Head and Neck
C470, C490	8000-8700, 8720-8790, 9700-9701	XX	Other Soft Tissue Sarcoma of the Head and Neck
C000-C148, C150, C153, C158, C300-C329, C470, C490	8710, 8712-8714, 8803, 8812-8814, 8820-8824, 8826-8831, 8834-8836, 8841-8842, 8851, 8853, 8855-8857, 8860-8881, 8891-8898, 8900, 8902-8905, 8921, 8932-8934, 8950-8973, 8975, 8981, 8983-8990, 9000-9016, 9030, 9042, 9045-9110, 9121-9132, 9135, 9137, 9141-9175, 9181-9230, 9240-9250, 9252-9363, 9365-9539, 9541, 9550-9560, 9562-9571, 9582	XX	Other Soft Tissue Sarcoma of the Head and Neck

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Case #12

Appendix Biopsy and Biopsy Peritoneal Nodule – low grade appendiceal mucinous neoplasm (LAMN) with loss of lamina propria and muscularis mucosae and submucosal fibrosis. Low grade pseudomyxoma peritoneii.

Is this case Not Reportable?
Could this be a carcinoid tumor?
Mucin not usually associated with carcinoid and no mention NET.

No. of Responses	Confined to mucosa	Dissecting Mucin	Pushing invasion	Infiltrative Invasive	Signet Ring Cells
11	?	Low-grade mucinous neoplasm (LAMN)		Mucinous adenocarcinoma	
5	Adenoma	Low-grade mucinous neoplasm (LAMN)		Mucinous adenocarcinoma	
8	?	?	Low-grade mucinous adenocarcinoma	High-grade mucinous adenocarcinoma	High-grade mucinous adenocarcinoma with signet ring cells
6	?	?	Low-grade mucinous adenocarcinoma	High-grade mucinous adenocarcinoma	
2	Adenoma			Adenocarcinoma	

Rule clarification: Pseudomyxoma peritonei (accumulation of mucin-secreting tumor cells in the abdominal or pelvic cavity) now has a **two-tiered system** (WHO 2010) that classifies pseudomyxoma peritonei as either **high-grade** or **low-grade** (see below). Pseudomyxoma peritonei is usually associated with **mucinous** tumors of the appendix and is rarely associated with ovarian mucinous tumors.

- **High**-grade pseudomyxoma peritonei is **malignant** /3
- **Low**-grade pseudomyxoma peritonei is **not malignant** /1

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Case #13

Right lower lobe lung, wedge – grade1 minimally invasive adenocarcinoma with predominantly lepidic pattern, non-mucinous

ICD-O-3 Primary Site	C34.3
ICD-O-3 Histology	8256 (non-mucinous adenoca, minimally invasive)
ICD-O-3 Behavior	3
ICD-O-3 Clinical Grade	1
ICD-O-3 Path Grade	1

Rule H2 Code **non-mucinous** adenocarcinoma as follows:
- **8256/3 when**
  - Microinvasive
  - Minimally invasive

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ICD-O-3.2 New Histology Codes with New Terms - Only

Status	Histology Value	Behavior	Preferred	label	Reportable	Comments
New term & code	8023	3	FALSE	Midline carcinoma of children and young adults with NUT rearrangement (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	TRUE	NUT carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	FALSE	NUT midline carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8054	3	FALSE	Condylomatous carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8054	3	TRUE	Warty carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8085	3	TRUE	Squamous cell carcinoma, HPV-positive (C01.9, 09.9,C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8086	3	TRUE	Squamous cell carcinoma, HPV-negative (C01.9, C09.9, C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8158	1	FALSE	ACTH-producing tumor	N	Not reportable for all years
New term & code	8158	1	TRUE	Endocrine tumor, functioning, NOS	N	Not reportable for all years
New term & code	8163	3	FALSE	Adenocarcinoma, pancreatobiliary-type (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8163	3	TRUE	Pancreatobiliary-type carcinoma (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8256	3	TRUE	Minimally invasive adenocarcinoma, non-mucinous (C34._)	Y	
New term & code	8257	3	TRUE	Minimally invasive adenocarcinoma, mucinous (C34._)	Y	
New term & code	8265	3	TRUE	Micropapillary carcinoma, NOS (C18._, C19.9, C20.9, C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8265	3	FALSE	Micropapillary adenocarcinoma (C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8339	3	TRUE	Follicular thyroid carcinoma (FTC), encapsulated angoinvasive (C73.9)	Y	
New term & code	8474	3	TRUE	Seromucinous carcinoma (C56.9)	Y	
New term & code	8509	2	TRUE	Solid papillary carcinoma in situ (C50._)	Y	
New term & code	8509	3	TRUE	Solid papillary carcinoma with invasion (C50._)	Y	
New term & code	8519	2	TRUE	Pleomorphic lobular carcinoma in situ (C50._)	Y	ICD-O-3 rule F DOES NOT APPLY to code 8519. Invasive pleomorphic lobular carcinoma is coded 8520/3
New term & code	8552	3	TRUE	Mixed acinar ductal carcinoma	Y	Cases diagnosed prior to 1/1/2018 use code 8523/3
New term & code	8594	1	TRUE	Mixed germ cell sex cord-stromal tumor, unclassified (C48.2, C56.9, C57.9)	N	Not reportable for all years
New term & code	8714	3	FALSE	Malignant perivascular epithelial cell tumor	Y	
New term & code	8714	3	TRUE	PEComa, malignant	Y	
New term & code	8714	3	FALSE	Perivascular epithelioid cell tumor, malignant	Y	
New term/behavior	8815	1	TRUE	Solitary fibrous tumor/hemanglopericytoma Grade 2 (CNS) (C71._)	Y	Reportable for CNS ONLY
New term & code	8975	1	TRUE	Calcifying nested epithelial stromal tumor (C22.0)	N	Not reportable for all years
New term & code	9045	3	TRUE	Biphenotypic sinonasal sarcoma (C30.0, C31.0-C31.3, C31.8, C31.9)	Y	
New term & code	9086	3	TRUE	Germ cell tumors with associated hematological malignancy (C37.9)	Y	
New term & code	9137	3	TRUE	Intimal sarcoma	Y	
New term & code	9137	3	FALSE	Pulmonary artery intimal sarcoma	Y	
New term & code	9385	3	TRUE	Diffuse midline glioma, H3 K27M-mutant (C71._)	Y	
New term & code	9395	3	TRUE	Papillary tumor of pineal region (C75.3)	Y	Cases diagnosed prior to 1/1/2018 use code 9361/3
New term & code	9396	3	TRUE	Ependymoma, RELA fusion-positive (C71._)	Y	
New term & code	9425	3	TRUE	Pilomyxoid astrocytoma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9421/3
New term & code	9431	1	TRUE	Angiocentric glioma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9432	1	TRUE	Pituicytoma (C75.1)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9445	3	TRUE	Glioblastoma, IDH-mutant (C71._)	Y	
New term & code	9475	3	TRUE	Medulloblastoma, WNT-activated (C71._)	Y	
New term & code	9476	3	TRUE	Medulloblastoma, SHH-activated and TP53 mutant (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 3 (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 4 (C71._)	Y	
New term & code	9477	3	TRUE	Medulloblastoma, non-WNT/non-SHH (C71._)	Y	
New term & code	9478	3	FALSE	Embryonal tumor with multilayered rosettes C19MC-altered (C71._)	Y	
New term & code	9478	3	TRUE	Embryonal tumor with multilayered rosettes, NOS (C71._)	Y	
New term & code	9509	1	FALSE	Diffuse leptomeningeal glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	TRUE	Papillary glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	FALSE	Rosette-forming glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9542	3	TRUE	Epithelioid malignant peripheral nerve sheath tumor (C47.0-C47.6, C47.8, C47.9)	Y	

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Case #14

Biopsy right tonsil with squamous cell carcinoma. p16+, HPV+

ICD-O-3 Primary Site	C09.9 (tonsil, right)
ICD-O-3 Histology	8085 (HPV+ squamous cell carcinoma)
ICD-O-3 Behavior	3 (malignant)
ICD-O-3 Clinical Grade	9 (unknown)
ICD-O-3 Path Grade	9 (unknown)

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ICD-O-3.2 New Histology Codes with New Terms - Only

Status	Histology Value	Behavior	Preferred	label	Reportable	Comments
New term & code	8023	3	FALSE	Midline carcinoma of children and young adults with NUT rearrangement (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	TRUE	NUT carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	FALSE	NUT midline carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8054	3	FALSE	Condylomatous carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8054	3	TRUE	Warty carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8085	3	TRUE	Squamous cell carcinoma, HPV-positive (C01.9, 09.9,C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8086	3	TRUE	Squamous cell carcinoma, HPV-negative (C01.9, C09.9, C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8158	1	FALSE	ACTH-producing tumor	N	Not reportable for all years
New term & code	8158	1	TRUE	Endocrine tumor, functioning, NOS	N	Not reportable for all years
New term & code	8163	3	FALSE	Adenocarcinoma, pancreatobiliary-type (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8163	3	TRUE	Pancreatobiliary-type carcinoma (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8256	3	TRUE	Minimally invasive adenocarcinoma, non-mucinous (C34._)	Y	
New term & code	8257	3	TRUE	Minimally invasive adenocarcinoma, mucinous (C34._)	Y	
New term & code	8265	3	TRUE	Micropapillary carcinoma, NOS (C18._, C19.9, C20.9, C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8265	3	FALSE	Micropapillary adenocarcinoma (C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8339	3	TRUE	Follicular thyroid carcinoma (FTC), encapsulated angoinvasive (C73.9)	Y	
New term & code	8474	3	TRUE	Seromucinous carcinoma (C56.9)	Y	
New term & code	8509	2	TRUE	Solid papillary carcinoma in situ (C50._)	Y	
New term & code	8509	3	TRUE	Solid papillary carcinoma with invasion (C50._)	Y	
New term & code	8519	2	TRUE	Pleomorphic lobular carcinoma in situ (C50._)	Y	ICD-O-3 rule F DOES NOT APPLY to code 8519. Invasive pleomorphic lobular carcinoma is coded 8520/3
New term & code	8552	3	TRUE	Mixed acinar ductal carcinoma	Y	Cases diagnosed prior to 1/1/2018 use code 8523/3
New term & code	8594	1	TRUE	Mixed germ cell sex cord-stromal tumor, unclassified (C48.2, C56.9, C57.9)	N	Not reportable for all years
New term & code	8714	3	FALSE	Malignant perivascular epithelial cell tumor	Y	
New term & code	8714	3	TRUE	PEComa, malignant	Y	
New term & code	8714	3	FALSE	Perivascular epithelioid cell tumor, malignant	Y	
New term/behavior	8815	1	TRUE	Solitary fibrous tumor/hemanglopericytoma Grade 2 (CNS) (C71._)	Y	Reportable for CNS ONLY
New term & code	8975	1	TRUE	Calcifying nested epithelial stromal tumor (C22.0)	N	Not reportable for all years
New term & code	9045	3	TRUE	Biphenotypic sinonasal sarcoma (C30.0, C31.0-C31.3, C31.8, C31.9)	Y	
New term & code	9086	3	TRUE	Germ cell tumors with associated hematological malignancy (C37.9)	Y	
New term & code	9137	3	TRUE	Intimal sarcoma	Y	
New term & code	9137	3	FALSE	Pulmonary artery intimal sarcoma	Y	
New term & code	9385	3	TRUE	Diffuse midline glioma, H3 K27M-mutant (C71._)	Y	
New term & code	9395	3	TRUE	Papillary tumor of pineal region (C75.3)	Y	Cases diagnosed prior to 1/1/2018 use code 9361/3
New term & code	9396	3	TRUE	Ependymoma, RELA fusion-positive (C71._)	Y	
New term & code	9425	3	TRUE	Pilomyxoid astrocytoma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9421/3
New term & code	9431	1	TRUE	Angiocentric glioma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9432	1	TRUE	Pituicytoma (C75.1)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9445	3	TRUE	Glioblastoma, IDH-mutant (C71._)	Y	
New term & code	9475	3	TRUE	Medulloblastoma, WNT-activated (C71._)	Y	
New term & code	9476	3	TRUE	Medulloblastoma, SHH-activated and TP53 mutant (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 3 (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 4 (C71._)	Y	
New term & code	9477	3	TRUE	Medulloblastoma, non-WNT/non-SHH (C71._)	Y	
New term & code	9478	3	FALSE	Embryonal tumor with multilayered rosettes C19MC-altered (C71._)	Y	
New term & code	9478	3	TRUE	Embryonal tumor with multilayered rosettes, NOS (C71._)	Y	
New term & code	9509	1	FALSE	Diffuse leptomeningeal glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	TRUE	Papillary glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	FALSE	Rosette-forming glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9542	3	TRUE	Epithelioid malignant peripheral nerve sheath tumor (C47.0-C47.6, C47.8, C47.9)	Y	

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Case #15

Left tonsil biopsy with well differentiated squamous cell carcinoma with keratinizing features. Should this histology be 8070/3, 8071/3 or 8086/3?

8086/3- Squamous cell carcinoma, HPV-negative (C01.9, 09.9, C10.2, C10.3, C10.8, C10.9, C31.0–C31.3, C31.9). 09.9 added to the topography codes eligible for this histology.

Do Not Assume HPV-Negative

LOOK for HPV Testing and/or p16 Testing.

Code 8070/3 – do not code features.

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Case #16

Biopsy right true vocal cord positive for squamous cell carcinoma. HPV test panel was negative for all types/subtypes. p16 testing was also negative.

- Is p16+ the same as HPV+ and if not, what is the difference?

8085/3- Squamous cell carcinoma, HPV-positive (C01.9, 09.9, C10.2, C10.3, C10.8, C10.9, C31.0–C31.3, C31.9). 09.9 added to the topography codes eligible for this histology.

- P16 (INK4a) is a protein used as a surrogate marker for 'high risk' HPV infection in H&N Squamous Cell Carcinoma. It is detected using IHC testing methods.
- HPV Type-16 DNA and RNA is detected using PCR methods on frozen tissue.
- P16+ patients have significant survival and disease free benefit
- P16+ and HPV16+ patients have greater improved survival/disease free interval
- Physicians should test for both p16 and HPV Type 16 to obtain the best information

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ICD-O-3.2 New Histology Codes with New Terms - Only

Status	Histology Value	Behavior	Preferred	label	Reportable	Comments
New term & code	8023	3	FALSE	Midline carcinoma of children and young adults with NUT rearrangement (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	TRUE	NUT carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	FALSE	NUT midline carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8054	3	FALSE	Condylomatous carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8054	3	TRUE	Warty carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8085	3	TRUE	Squamous cell carcinoma, HPV-positive (C01.9, 09.9,C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8086	3	TRUE	Squamous cell carcinoma, HPV-negative (C01.9, C09.9, C10.2, C10.3, C10.8, C10.9, C31.0-C31.3, C31.9)	Y	
New term & code	8158	1	FALSE	ACTH-producing tumor	N	Not reportable for all years
New term & code	8158	1	TRUE	Endocrine tumor, functioning, NOS	N	Not reportable for all years
New term & code	8163	3	FALSE	Adenocarcinoma, pancreatobiliary-type (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8163	3	TRUE	Pancreatobiliary-type carcinoma (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8256	3	TRUE	Minimally invasive adenocarcinoma, non-mucinous (C34._)	Y	
New term & code	8257	3	TRUE	Minimally invasive adenocarcinoma, mucinous (C34._)	Y	
New term & code	8265	3	TRUE	Micropapillary carcinoma, NOS (C18._, C19.9, C20.9, C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8265	3	FALSE	Micropapillary adenocarcinoma (C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8339	3	TRUE	Follicular thyroid carcinoma (FTC), encapsulated angoinvasive (C73.9)	Y	
New term & code	8474	3	TRUE	Seromucinous carcinoma (C56.9)	Y	
New term & code	8509	2	TRUE	Solid papillary carcinoma in situ (C50._)	Y	
New term & code	8509	3	TRUE	Solid papillary carcinoma with invasion (C50._)	Y	
New term & code	8519	2	TRUE	Pleomorphic lobular carcinoma in situ (C50._)	Y	ICD-O-3 rule F DOES NOT APPLY to code 8519. Invasive pleomorphic lobular carcinoma is coded 8520/3
New term & code	8552	3	TRUE	Mixed acinar ductal carcinoma	Y	Cases diagnosed prior to 1/1/2018 use code 8523/3
New term & code	8594	1	TRUE	Mixed germ cell sex cord-stromal tumor, unclassified (C48.2, C56.9, C57.9)	N	Not reportable for all years
New term & code	8714	3	FALSE	Malignant perivascular epithelial cell tumor	Y	
New term & code	8714	3	TRUE	PEComa, malignant	Y	
New term & code	8714	3	FALSE	Perivascular epithelioid cell tumor, malignant	Y	
New term/behavior	8815	1	TRUE	Solitary fibrous tumor/hemangiopericytoma Grade 2 (CNS) (C71._)	Y	Reportable for CNS ONLY
New term & code	8975	1	TRUE	Calcifying nested epithelial stromal tumor (C22.0)	N	Not reportable for all years
New term & code	9045	3	TRUE	Biphenotypic sinonasal sarcoma (C30.0, C31.0-C31.3, C31.8, C31.9)	Y	
New term & code	9086	3	TRUE	Germ cell tumors with associated hematological malignancy (C37.9)	Y	
New term & code	9137	3	TRUE	Intimal sarcoma	Y	
New term & code	9137	3	FALSE	Pulmonary artery intimal sarcoma	Y	
New term & code	9385	3	TRUE	Diffuse midline glioma, H3 K27M-mutant (C71._)	Y	
New term & code	9395	3	TRUE	Papillary tumor of pineal region (C75.3)	Y	Cases diagnosed prior to 1/1/2018 use code 9361/3
New term & code	9396	3	TRUE	Ependymoma, RELA fusion-positive (C71._)	Y	
New term & code	9425	3	TRUE	Pilomyxoid astrocytoma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9421/3
New term & code	9431	1	TRUE	Angiocentric glioma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9432	1	TRUE	Pituicytoma (C75.1)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9445	3	TRUE	Glioblastoma, IDH-mutant (C71._)	Y	
New term & code	9475	3	TRUE	Medulloblastoma, WNT-activated (C71._)	Y	
New term & code	9476	3	TRUE	Medulloblastoma, SHH-activated and TP53 mutant (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 3 (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 4 (C71._)	Y	
New term & code	9477	3	TRUE	Medulloblastoma, non-WNT/non-SHH (C71._)	Y	
New term & code	9478	3	FALSE	Embryonal tumor with multilayered rosettes C19MC-altered (C71._)	Y	
New term & code	9478	3	TRUE	Embryonal tumor with multilayered rosettes, NOS (C71._)	Y	
New term & code	9509	1	FALSE	Diffuse leptomeningeal glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	TRUE	Papillary glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	FALSE	Rosette-forming glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9542	3	TRUE	Epithelioid malignant peripheral nerve sheath tumor (C47.0-C47.6, C47.8, C47.9)	Y	

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Case #17

Biopsy followed by gross total excision left parietal lobe with WHO Grade 4 glioblastoma multiforme of IDH-wild type variety

ICD-O-3 Primary Site	C71.3 (parietal lobe of brain)
ICD-O-3 Histology	9440 (glioblastoma multiforme, NOS)
ICD-O-3 Behavior	3 (malignant)
ICD-O-3 Clinical Grade	4 (WHO Grade 4)
ICD-O-3 Path Grade	4 (WHO Grade 4)
Brain Molecular Markers	05 Glioblastoma, IDH wildtype (9440/3)

Code	Description
01	Diffuse astrocytoma, IDH-mutant (9400/3)
02	Diffuse astrocytoma, IDH-wildtype (9400/3)
03	Anaplastic astrocytoma, IDH-mutant (9401/3)
04	Anaplastic astrocytoma, IDH-wildtype (9401/3)
05	Glioblastoma, IDH-wildtype (9440/3)
06	Oligodendroglioma, IDH-mutant and 1 p/19q co-deleted (9450/3)
07	Anaplastic oligodendroglioma, IDH-mutant and 1p/19q co-deleted (9451/3)

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Case #18

Excision cerebellopontine angle with glioblastoma multiforme IDH-mutant type, WHO Grade 4

ICD-O-3 Primary Site	C71.6 (Cerebellum, NOS)
ICD-O-3 Histology	9445 (IDH-mutant type glioblastoma multiforme)
ICD-O-3 Behavior	3 (malignant)
ICD-O-3 Clinical Grade	4 (WHO Grade 4 – cannot tell if biopsy or resection)
ICD-O-3 Path Grade	4 (WHO Grade 4 – cannot tell if biopsy or resection)
Brain Molecular Marker	99 (IDH mutation is picked up in Histology Code)

NOTE: 9445 histology code is not in the Brain Molecular Markers Table. So?

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ICD-O-3.2 New Histology Codes with New Terms - Only

Status	Histology Value	Behavior	Preferred	label	Reportable	Comments
New term & code	8023	3	FALSE	Midline carcinoma of children and young adults with NUT rearrangement (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	TRUE	NUT carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8023	3	FALSE	NUT midline carcinoma (C30.0, C31.9, C34._)	Y	
New term & code	8054	3	FALSE	Condylomatous carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8054	3	TRUE	Warty carcinoma (C60.0-C60.2, C60.9)	Y	Cases diagnosed prior to 1/1/2018 use code 8051/3 All other sites use 8051/3 2018 forward
New term & code	8085	3	TRUE	Squamous cell carcinoma, HPV-positive (C01.9, 09.9,C10.2, C10.3, C10.8, C10.9, C31.0–C31.3, C31.9)	Y	
New term & code	8086	3	TRUE	Squamous cell carcinoma, HPV-negative (C01.9, C09.9, C10.2, C10.3, C10.8, C10.9, C31.0–C31.3, C31.9)	Y	
New term & code	8158	1	FALSE	ACTH-producing tumor	N	Not reportable for all years
New term & code	8158	1	TRUE	Endocrine tumor, functioning, NOS	N	Not reportable for all years
New term & code	8163	3	FALSE	Adenocarcinoma, pancreatobiliary-type (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8163	3	TRUE	Pancreatobiliary-type carcinoma (C24.1)	Y	Cases diagnosed prior to 1/1/2018 use code 8255/3
New term & code	8256	3	TRUE	Minimally invasive adenocarcinoma, non-mucinous (C34._)	Y	
New term & code	8257	3	TRUE	Minimally invasive adenocarcinoma, mucinous (C34._)	Y	
New term & code	8265	3	TRUE	Micropapillary carcinoma, NOS (C18._, C19.9, C20.9, C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8265	3	FALSE	Micropapillary adenocarcinoma (C34._)	Y	Cases diagnosed prior to 1/1/2018 use code 8507/3. Code 8265 is not valid for C50._. Use 8507 for micropapillary adenocarcinoma in breast primaries
New term & code	8339	3	TRUE	Follicular thyroid carcinoma (FTC), encapsulated angioinvasive (C73.9)	Y	
New term & code	8474	3	TRUE	Seromucinous carcinoma (C56.9)	Y	
New term & code	8509	2	TRUE	Solid papillary carcinoma in situ (C50._)	Y	
New term & code	8509	3	TRUE	Solid papillary carcinoma with invasion (C50._)	Y	
New term & code	8519	2	TRUE	Pleomorphic lobular carcinoma in situ (C50._)	Y	ICD-O-3 rule F DOES NOT APPLY to code 8519. Invasive pleomorphic lobular carcinoma is coded 8520/3
New term & code	8552	3	TRUE	Mixed acinar ductal carcinoma	Y	Cases diagnosed prior to 1/1/2018 use code 8523/3
New term & code	8594	1	TRUE	Mixed germ cell sex cord-stromal tumor, unclassified (C48.2, C56.9, C57.9)	N	Not reportable for all years
New term & code	8714	3	FALSE	Malignant perivascular epithelial cell tumor	Y	
New term & code	8714	3	TRUE	PEComa, malignant	Y	
New term & code	8714	3	FALSE	Perivascular epithelioid cell tumor, malignant	Y	
New term/behavior	8815	1	TRUE	Solitary fibrous tumor/hemanglopericytoma Grade 2 (CNS) (C71._)	Y	Reportable for CNS ONLY
New term & code	8975	1	TRUE	Calcifying nested epithelial stromal tumor (C22.0)	N	Not reportable for all years
New term & code	9045	3	TRUE	Biphenotypic sinonasal sarcoma (C30.0, C31.0-C31.3, C31.8, C31.9)	Y	
New term & code	9086	3	TRUE	Germ cell tumors with associated hematological malignancy (C37.9)	Y	
New term & code	9137	3	TRUE	Intimal sarcoma	Y	
New term & code	9137	3	FALSE	Pulmonary artery intimal sarcoma	Y	
New term & code	9385	3	TRUE	Diffuse midline glioma, H3 K27M-mutant (C71._)	Y	
New term & code	9395	3	TRUE	Papillary tumor of pineal region (C75.3)	Y	Cases diagnosed prior to 1/1/2018 use code 9361/3
New term & code	9396	3	TRUE	Ependymoma, RELA fusion-positive (C71._)	Y	
New term & code	9425	3	TRUE	Pilomyxoid astrocytoma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9421/3
New term & code	9431	1	TRUE	Angiocentric glioma (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9432	1	TRUE	Pituicytoma (C75.1)	Y	Cases diagnosed prior to 1/1/2018 use code 9380/1
New term & code	9445	3	TRUE	Glioblastoma, IDH-mutant (C71._)	Y	
New term & code	9475	3	TRUE	Medulloblastoma, WNT-activated (C71._)	Y	
New term & code	9476	3	TRUE	Medulloblastoma, SHH-activated and TP53 mutant (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 3 (C71._)	Y	
New term & code	9477	3	FALSE	Medulloblastoma, group 4 (C71._)	Y	
New term & code	9477	3	TRUE	Medulloblastoma, non-WNT/non-SHH (C71._)	Y	
New term & code	9478	3	FALSE	Embryonal tumor with multilayered rosettes C19MC-altered (C71._)	Y	
New term & code	9478	3	TRUE	Embryonal tumor with multilayered rosettes, NOS (C71._)	Y	
New term & code	9509	1	FALSE	Diffuse leptomeningeal glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	TRUE	Papillary glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9509	1	FALSE	Rosette-forming glioneuronal tumor (C71._)	Y	Cases diagnosed prior to 1/1/2018 use code 9505/1
New term & code	9542	3	TRUE	Epithelioid malignant peripheral nerve sheath tumor (C47.0-C47.6, C47.8, C47.9)	Y	

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IDH-Wild Type and IDH-Mutant Type

- Before we had the ability to do gene testing, we knew only that we could use the size and shape and arrangements of brain tumor cells to assign a WHO Grade which was associated with varying prognosis based on the WHO Grade of the neoplasm.
- IDH mutant positive/negative is a new and additional way to classify these neoplasms in addition to WHO Grade. It is also a potential new target for delivering drugs or radiation directly to GBM tumor cells either with or without the mutation.
- IDH gene mutation (positive/negative) is a better predictor of survival than WHO Grade.
- The use of the term 'wild-type' has always sounded backwards to me.
- "Wild-type" classifications of a gene indicates that the tumor cells do not have the specific gene mutation (IDH in this case). So, IDH wild type GBM are tumors that do not have the IDH Mutation.
- GBM tumors with the IDH mutation are associated with the patient having a weaker natural immune response to the glioma which in turn reduces the aggressiveness of the GBM so patients live longer when they have the mutation. These patients can live 5-10 years with GBM.
- GBM with "unmutated" IDH prompts a stronger natural immune response that actually increases the tumors aggressiveness and negatively impacts survival. Patients with IDH-wild (unmutated) type GBM tumors carry a much poorer prognosis with median survival of only about 18 months.
- So, looking for this testing of IDH for GBM is really important to classify these tumors – huge differences in survival and different treatment plan.
- Again, they also hope that IDH can be a target for development of new drugs directed at the GBM cells to get them to convert to IDH mutant.

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Case #19

Radical Left Nephrectomy with kidney showing 7.5cm papillary clear cell renal cell carcinoma, Grade 2 of 4 (WHO/ISUP). No lymph nodes identified.

ICD-O-3 Primary Site	C64.9 (kidney, left)
ICD-O-3 Histology	8323 (clear cell papillary renal cell carcinoma)
ICD-O-3 Behavior	3 (malignant)
ICD-O-3 Clinical Grade	9 (no biopsy prior to resection of primary tumor)
ICD-O-3 Path Grade	2 (WHO/ISUP is preferred grading system)

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants
Nephroblastoma **8960**	Wilms tumor	
Neuroendocrine tumor (NET) **8240**	Carcinoid [OBS] Well-differentiated neuroendocrine tumor	Large cell neuroendocrine carcinoma/tumor **8013** Small cell neuroendocrine carcinoma **8041**
Renal cell carcinoma NOS **8312** *Note 1:* WHO, IARC, and CAP agree that sarcomatoid carcinoma is a pattern of differentiation, not a specific subtype, of renal cell carcinoma. *Note 2:* Sarcomatoid is listed in the CAP Kidney protocol under the header "features."	RCC Sarcomatoid carcinoma Sarcomatoid renal cell carcinoma Succinate dehydrogenase-deficient renal cell carcinoma (SDHD) Unclassified renal cell carcinoma	Acquired cystic disease-associated renal cell carcinoma/tubulocystic renal cell carcinoma **8316*** Chromophobe renal cell carcinoma (ChRCC) **8317** Clear cell papillary renal cell carcinoma **8323/3** *Note:* The 2016 WHO 4th Edition Classification of Tumors of the Urinary System and Male Genital Organs has reclassified this histology as a /1 because it is low nuclear grade and is now thought to be a neoplasm. This change was not implemented in the 2018 ICD-O update. Clear cell renal cell carcinoma (ccRCC) **8310** Collecting duct carcinoma **8319**

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Case #19

- The Note provided in Table 1, Column 3 for "Clear cell papillary renal cell carcinoma 8323/3," indicates this exact diagnosis is considered a borderline (/1) tumor per the 2016 WHO 4th Edition Classification of Tumors of the Urinary System and Male Genital Organs.
- However, the standard setters did not implement this change for 2018.
- This is still to be collected as a reportable, malignant (/3) tumor for cases diagnosed 2018 and later.
- In other words, this is to be considered a reportable tumor until the standard setters implement this change in behavior of a clear cell papillary renal cell carcinoma from 8323/3 to 8323/1.
- The note confirms that, although WHO has proposed a borderline behavior for this tumor (8323/1), this change has not been implemented by the standard setters yet. Therefore, diagnoses of exactly, "clear cell papillary renal cell carcinoma," should continue to be collected as 8323/3 until the standard setters have implemented this change.

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Case #20

Hepatic Flexure Tumor Resection shows 6cm large cell neuroendocrine carcinoma (NEC/NET). IHC stains positive for CD56, synaptophysin, CDX-2 and chromogranin). Ki-67 shows a high proliferative index (66%).

ICD-O-3 Primary Site	C18.3 (hepatic flexure)
ICD-O-3 Histology	8013 (large cell neuroendocrine carcinoma)
ICD-O-3 Behavior	3 (malignant)
ICD-O-3 Clinical Grade	9 (no biopsy prior to resection)
ICD-O-3 Path Grade	3 (Ki-67 index (%) is greater than 20)

Code	Grade Description
1	G1: Mitotic count (per 10 HPF) less than 2 AND Ki-67 index (%) less than 3
2	G2: Mitotic count (per 10 HPF) equal 2-20 OR Ki-67 index (%) equal 3-20
3	G3: Mitotic count (per 10 HPF) greater than 20 OR Ki-67 index (%) greater than 20
A	Well differentiated
B	Moderately differentiated
C	Poorly differentiated
D	Undifferentiated, anaplastic
9	Grade cannot be assessed (GX); Unknown

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BONUS CASE #1

Renal Pelvis with high grade urothelial carcinoma with squamous, sarcomatoid and osteoclast like giant cell features. Carcinoma invades into renal parenchyma and peripelvic adipose tissue. LVI is identified. Perineural invasion not identified. Extensive high gade urothelial carcinoma invades the perirenal adipose. 0/11 LNs.

Per synoptic report, histological type – urothelial carcinoma, sarcomatoid variant, urothelial carcinoma, giant cell variant, urothelial carcinoma with squamous differentiation.

Vesical pleomorphic giant cell carcinoma (PGCC) is a variant of urothelial carcinoma characterized by highly pleomorphic tumour with giant cells.

The association of PGCC with UC and an overlap in immunoexpression suggests that PGCC represents an extreme form of urothelial carcinoma de-differentiation

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BONUS CASE #1

- This is clearly a rare tumor. The Solid Tumor Rules Bladder Synonyms Table indicates histology is either 8122/3 sarcomatoid urothelial carcinoma or giant cell urothelial carcinoma (8031/3). The tumor is clearly pleomorphic in nature and appearance but still urothelial.
- Any available histology mixed code would lose the fact that it is urothelial carcinoma. (And, we ignore the squamous differentiation).
- Please code histology to giant cell urothelial carcinoma (8031/3) for the following reason in literature…"The association of pleomorphic giant cell carcinoma (PGCC) a variant of urothelial carcinoma and an overlap in immunoexpression suggests that PGCC represents an extreme form of urothelial carcinoma de-differentiation."
- This would make it more likely to behave poorly and be treated more aggressively due to recurrence likelihood as well as survival.

Histopathology: 2016 Mar; 68(4):533-40. doi: 10.1111/his.12785. Epub2015 Sep22.

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BONUS CASE #2

- NET/NEC/Neuroendocrine Tumors – found in multiple anatomic locations – lung, GI Tract including stomach, small intestine, pancreas (gastro-entero-pancreatic), colon, even the pituitary gland, parathyroid or adrenal gland as a pheochromocytoma and even mMrkel cell carcinoma is NET.
- So, we have many inconsistencies in naming, coding, grading and staging these neoplasms – they are all over the place.
- Grade and Histology Currently Described in Many Ways: High Grade/Low Grade/Mitotic Count/Mitotic Rate/Ki-67% – Histology includes: small cell carcinoma, carcinoid, large cell neuroendocrine carcinoma, NET, NEC, islet cell tumor, and other neuroendocrine neoplasms – some reportable/some not
- Case Description
  - Adenocarcinoma of Stomach on biopsy – signet ring cell features
  - Followed by Neo-Adjuvant chemo & Resection
  - Post-Treatment Resection: G3 well differentiated neuroendocrine carcinoma of GI origin
  - Tumor is 4cm in size
  - Mitotic Rate of 8 mitosis per 2 m2 & KI 67 80% (>20%)
  - invades into sub-serosal tissue without penetration of serosa

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Grade 07

Grade ID 07-Clinical Grade Instructions

Schema ID#	Schema ID Name	AJCC ID	AJCC Chapter
00290	NET Stomach	29	Neuroendocrine Tumors of the Stomach
00301	NET Duodenum	30	Neuroendocrine Tumors of the Duodenum and Ampulla of Vater
00302	NET Ampulla of Vater	30	Neuroendocrine Tumors of the Duodenum and Ampulla of Vater
00310	NET Jejunum and Ileum	31	Neuroendocrine Tumors of the Jejunum and Ileum
00320	NET Appendix	32	Neuroendocrine Tumors of the Appendix
00330	NET Colon and Rectum	33	Neuroendocrine Tumors of the Colon and Rectum
00340	NET Pancreas	34	Neuroendocrine Tumors of the Pancreas

Note 1: Clinical grade must not be blank.

Note 2: Assign the highest grade from the primary tumor assessed during the clinical time frame.

Note 3: Codes 1-3 take priority over codes A-D.

Note 4: Code 9 when
- Grade from primary site is not documented
- Clinical workup is not done (for example, cancer is an incidental finding during surgery for another condition)
- Grade checked "not applicable" on CAP Protocol (if available) and no other grade information is available

Note 5: If there is only one grade available and it cannot be determined if it is clinical, pathological, or after neo-adjuvant therapy, assign as a clinical grade and code unknown (9) for pathological grade, and blank for post-therapy grade.

Code	Grade Description
1	G1: Mitotic count (per 10 HPF) less than 2 AND Ki-67 index (%) less than 3
2	G2: Mitotic count (per 10 HPF) equal 2-20 OR Ki-67 index (%) equal 3-20
3	G3: Mitotic count (per 10 HPF) greater than 20 OR Ki-67 index (%) greater than 20
A	Well differentiated
B	Moderately differentiated
C	Poorly differentiated
D	Undifferentiated, anaplastic
9	Grade cannot be assessed (GX); Unknown

Grade 02

Grade ID 02-Clinical Grade Instructions

Schema ID#	Schema ID Name	AJCC ID	AJCC Chapter
00111	Oropharynx (p16-)	11.1	Oropharynx (p16-)
00112	Hypopharynx	11.2	Hypopharynx
00150	Cutaneous Squamous Cell Carcinoma of Head and Neck	15	Cutaneous Squamous Cell Carcinoma of the Head and Neck
00180	Small Intestine	18	Small Intestine
00200	Colon and Rectum	20	Colon and Rectum
00220	Liver	22	Liver
00360	Lung	36	Lung
00370	Pleura	37	Malignant Pleural Mesothelioma
00640	Skin of Eyelid	64	Eyelid Carcinoma
00650	Conjunctiva	65	Conjunctival Carcinoma

Note 1: Clinical grade must not be blank.

Note 2: Assign the highest grade from the primary tumor assessed during the clinical time frame.

Note 3: G4 includes anaplastic.

Note 4: Code 9 when
- Grade from primary site is not documented
- Clinical workup is not done (for example, cancer is an incidental finding during surgery for another condition)
- Grade checked "not applicable" on CAP Protocol (if available) and no other grade information is available

Note 5: If there is only one grade available and it cannot be determined if it is clinical, pathological, or after neo-adjuvant therapy, assign as a clinical grade and code unknown (9) for pathological grade, and blank for post-therapy grade.

Code	Grade Description
1	G1: Well differentiated
2	G2: Moderately differentiated
3	G3: Poorly differentiated
4	G4: Undifferentiated
9	Grade cannot be assessed (GX); Unknown

Return to Grade Tables (in Schema ID order)

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BONUS CASE #3

- Pancreas body, trans-gastric FNA shows high grade adenocarcinoma with small cell carcinoma and squamous carcinoma dedifferentiation
- Pancreatic high grade adenocarcinoma – ignore minor component of dedifferentiation as noted in the FNA material.
- No special code just for adenocarcinoma with dedifferentiation
- "Dedifferentiation" is a term used to describe the process of tumor transformation from a high grade adenocarcinoma to a mixed tumor
- Case will be treated as an adenocarcinoma.
- There are no specific rules for this. The Registrar must think thru what the rules imply and use their own best judgement to determine the best histology code – or contact FCDS or SEER.

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BONUS CASE #4

- Code Hepatocellular, cholangiocarcinoma or adenocarcinoma?
- What does path report state? How do you know site/histo to confirm?
- Distinguishing biliary system cholangiocarcinoma (common bile duct, intrahepatic bile duct and extrahepatic bile duct) from primary liver tumor (hepatocellular carcinoma) or metastatic adenocarcinoma can be difficult. Differentiating primary cholangiocarcinoma from metastases (lung, esophagus, stomach, pancreas), often cannot be made histologically. Imaging is the main diagnostic modality for cholangiocarcinoma - not biopsy.
- Unfortunately, we are seeing physicians using these terms interchangeably more and more often - and they are definitely not the same. One is a bile duct tumor (90% adenocarcinoma with mucin production) and the other a liver parenchyma cancer. The bile ducts connect the liver to the gallbladder and small intestine - so, location is key to identifying a primary cholangiocarcinoma. They also have different causes and different treatment options

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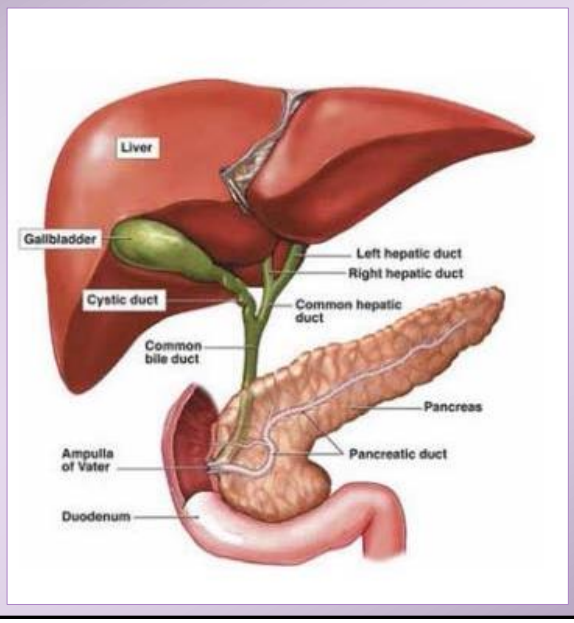
BONUS CASE #4



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BONUS CASE #4

- Primary sclerosing cholangitis associated with ulcerative colitis or inflammatory bowel disease is the most common risk factor for cholangiocarcinoma.
- Cholangiocarcinoma is less likely to be caused by or associated with Hepatitis B/C or cirrhosis.
- Hepatocellular carcinoma is nearly always associated with chronic liver disease caused by Hep B or C and/or cirrhosis.
- Combined hepatocellular and cholangiocarcinoma is rare - separate primary cancers.
- Cholangiocarcinoma almost always presents with bile duct obstruction (biliary obstruction) because the tumor originates in the bile duct and as it grows the tumor blocks the flow of bile which causes symptoms such as jaundice, itching, dark urine, white stools, weight loss, nausea and vomiting.

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BONUS CASE #4

- Imaging is the main diagnostic tool for cholangiocarcinoma.
- When cholangiocarcinoma is suspected, the radiologist will usually look for a single intrahepatic mass lesion with characteristics of a metastasis, a hilar stricture or distal bile duct obstruction, with or without a discernible mass. Abdominal ultrasound may show biliary ductal dilation related to the primary tumor and there is often biliary obstruction causing clinical symptoms.
- Contrast MRI is the optimal imaging study for suspected cholangiocarcinoma because it more clearly delineates hepatobiliary anatomy, extent of duct involvement, presence of liver mets, etc. However, MRI is not as effective in detecting distant mets to lungs and/or bone.
- Cholangiography is essential for assessing the extent of bile duct involvement and to determine whether or not the tumor is resectable.
- Immunohistochemistry panels including CK7, CK19, CK20, CDX-2, TTF-1, estrogen/progesterone receptors and PSA, can be helpful depending on clinical presentation. Cholangiocarcinoma is usually CK7 positive and CK20 negative.
- Other characteristics that may distinguish hepatocellular carcinoma from cholangiocarcinoma include presence or absence of mucin production (more likely cholangiocarcinoma when mucin production present) and expression of HepPar-1, CD10 and glypican-3 by HCC are useful.

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BONUS CASE #4

- Genetics: Inactivation of tumour suppressor genes; p53, Smad-4, bcl-2 and p16. Mutations in oncogenes including K-ras, p53, c-erbB-2 and c-neu.
- Below are a few questions you should ask yourself when trying to determine the correct primary site and/or histology for these cancers.

1. Do you have an abdominal CT that shows either a single liver mass or multiple liver masses or nodules?
2. Does the CT give any indication of tumor location?
3. Is it somewhere in the parenchyma of the liver or is it more closely associated with the bile duct system and hepatic artery?
   - If there are multiple tumors - then I would think about a liver primary or mets from another site - especially if patient has history of alcoholic liver.
   - The liver mass bx doesn't help us much since it just states - poorly diff carcinoma - stains ruled out GI Tract, lung, breast - then states "possible cholangiocarcinoma" - so, it is really not helpful at all.
   - TACE or Trans Arterial Chemotherapeutic Embolization for unresectable intrahepatic cholangiocarcinoma versus hepatocellular carcinoma can be used to treat a single large tumor or single lobe of the liver with only one source of blood that can be blocked by TACE. TACE is more often used for hepatocellular carcinoma than for cholangiocarcinoma due to anatomical considerations. Location of this tumor and the artery that was embolized may also give a clue as to whether this is primary liver neoplasm or primary bile duct neoplasm.

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BONUS CASE #5 – Information Only

- Pancreatic Tumors
  - hepatopancreatobiliary
- 2010 WHO Classification
- Watch the Behavior
  - /2 – non-invasive
  - /2 – not really in-situ
  - /3 – invasive
- Clinical Diagnosis
- Direct Visual Diagnosis
- Others are Imaging Dx
- Not Histological Proven

Reportable	ICD-O-3	Description
Yes	****/2	All Histologies with Behavior Code of /2 (in-situ)
Yes	****/3	All Histologies with Behavior Code of /3 (invasive)
Yes	8440/3	Cystadenocarcinoma of the pancreas
Yes	8150/3	Cystic Pancreatic Endocrine Neoplasm (CPEN)
Yes	8500/3	Infiltrating Duct Carcinoma of the pancreas
Yes	8503/2	Intraductal Oncocytic Papillary Neoplasm (IOPN) of the pancreas
Yes	8453/2	Intraductal Papillary Mucinous Neoplasms (IPMN) of the pancreas
Yes	8453/3	Intraductal Papillary Mucinous Neoplasm (IPNM) with invasive carcinoma
Yes	8503/2	Intraductal Tubule-Papillary Neoplasm (ITPN) of the pancreas
Yes	8503/3	Intraductal Tubule-Papillary Neoplasm (ITPN) with invasive carcinoma
Yes	8470/2	Mucinous Cystic Neoplasm (MCN) of the pancreas with high-grade dysplasia
Yes	8470/2	Non-invasive Mucinous Cystic Neoplasm (MCN) of the pancreas with high-grade dysplasia
Yes	8470/2	Mucinous Cystadenocarcinoma, non-invasive (MCN)
Yes	8470/3	Mucinous Cystadenocarcinoma of the pancreas
Yes	8470/3	Mucinous Cystic Neoplasm (MCN) of the pancreas with invasive carcinoma
Yes	8246/3	Neuroendocrine Carcinoma of the pancreas
Yes	8240/3	Neuroendocrine Tumor, Grade 1 (NET GR1) of the pancreas
Yes	8249/3	Neuroendocrine Tumor, Grade 2 (NET GR2) of the pancreas
Yes	8471/3	Papillary Mucinous Cystadenocarcinoma of the pancreas
Yes	8452/3	Solid Pseudo-Papillary Neoplasm (SPN) of the pancreas
Yes	8552/3*	Mixed acinar-ductal carcinoma
Yes	8163/2*	Papillary neoplasm, pancreatobiliary-type, with high grade intraepithelial neoplasia
Yes	8163/3*	Pancreatobiliary-type carcinoma
No	n/a	Histologies with Behavior Code of /0 (benign)
No	n/a	Histologies with Behavior Code of /1 (borderline)
No	n/a	Serous cystadenomas, solid and cystic papillary (Hamoudi) tumors, lympho-epithelial cysts and simple cysts are all benign and not reportable

* New histology codes not yet implemented in the U.S. are still reportable – use histology 8500 or 8140

References: 2010 WHO Classification of Tumours of the Pancreas; Pathologe. 2011 Nov;32 Suppl 2:332-6. doi: 10.1007/s00292-011-1515-2; Ann Surg. 2004 May; 239(5): 651–659), 2011 ICD-O-3 Updates, 2015 SEER Program Coding and Staging Manual, and NCI SEER Ask A SEER Registrar.

NOTE: May need to add D13.6 & K86.9 to Casefinding because many cases coded as benign neoplasms by medical coders.

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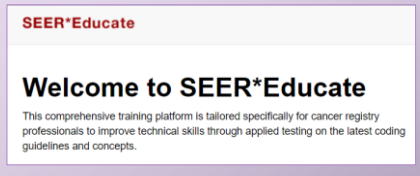
SEER Coding Drills for Dx Year 2018 Histologies

- SEER*Educate just released on August 30, 2019 - 110 hands-on coding exercises for coding primary site, histology, and behavior and to reinforce the use of 2018 ICD-O-3 Updates/ 2018 Solid Tumor Rules. Most cases show how to use the Solid Tumor Rules with New Codes
- Check personal coding skills and ability to follow the Solid Tumor Rules under the Training Menu/CTR Prep Tests/CTR Prep – Coding Drill – Dx 2018 Histology (Solid Tumors) on the SEER*Educate Website https://educate.fredhutch.org/
  - Colon, Rectosigmoid, and Rectum (10 cases)
  - Cutaneous Melanoma (10 cases)
  - Head & Neck (10 cases)
  - Kidney (10 cases)
  - Lung (20 cases)
  - Malignant CNS and Peripheral Nerves (10 cases)
  - Non-Malignant CNS (10 cases)
  - Urinary (10 cases)



DEMONSTRATE

Don't Forget to Demonstrate the Site

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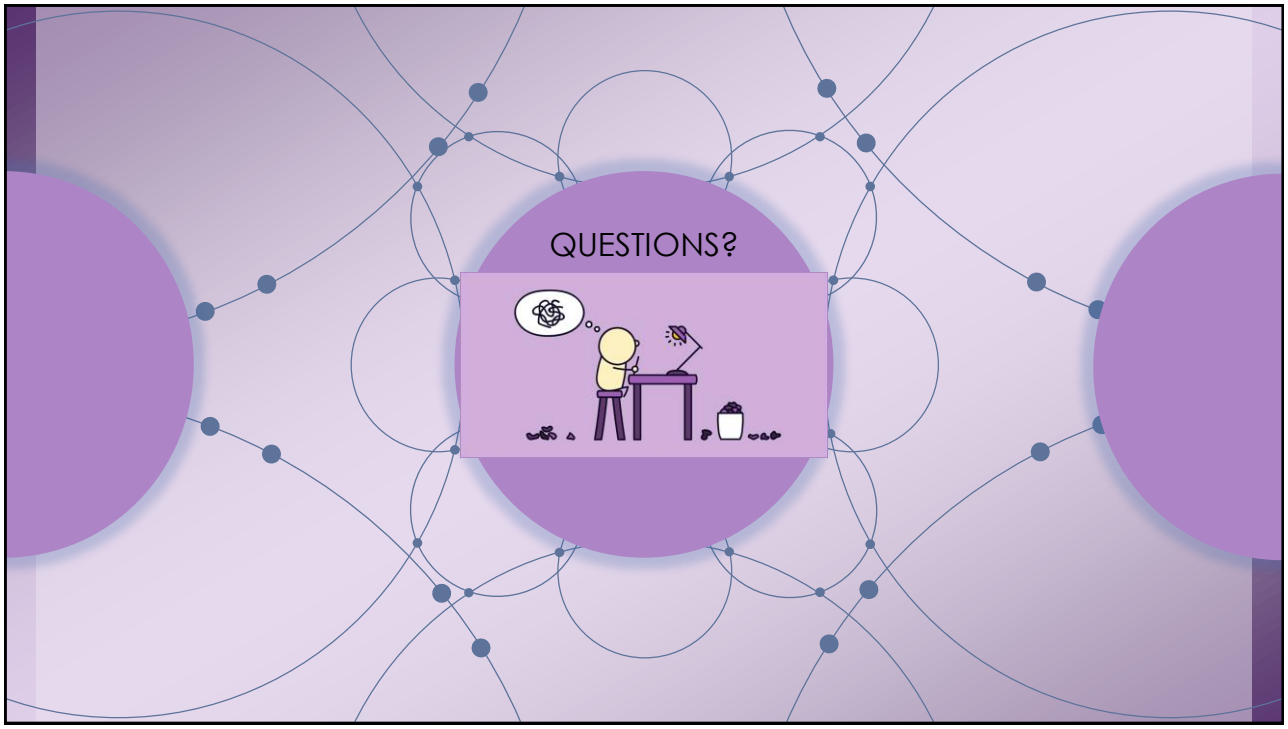
QUESTIONS?