GRELL & ENCR 2025 Scientific Meeting
Porto, Portugal

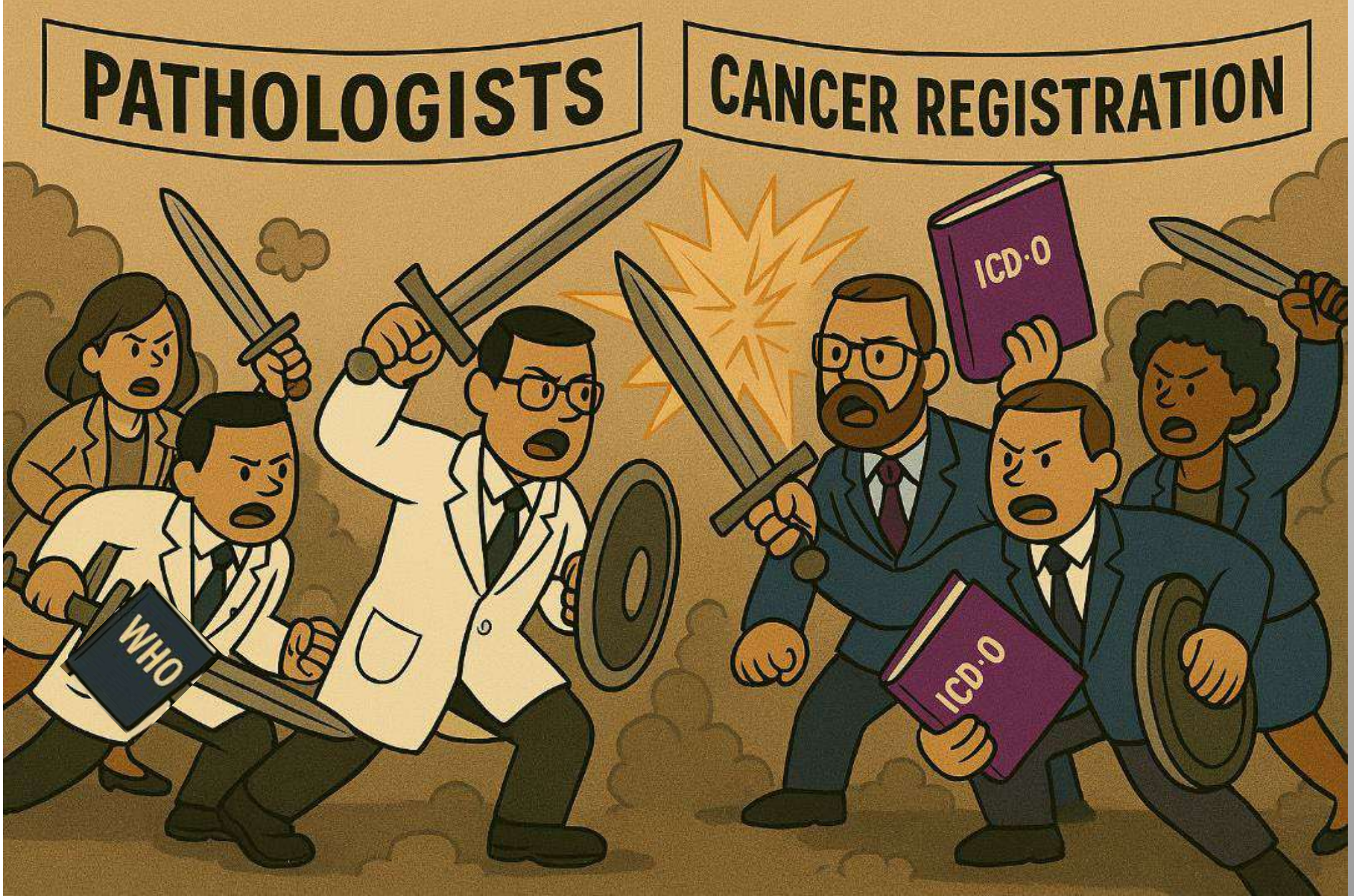
27-30 MAY

ICD-O4

Dr. Brian Rous, Cambridge, UK

Note:

ICD-O4 has not been finalised at the time of this presentation, so any ICD-O4 codes presented may differ in the final version.



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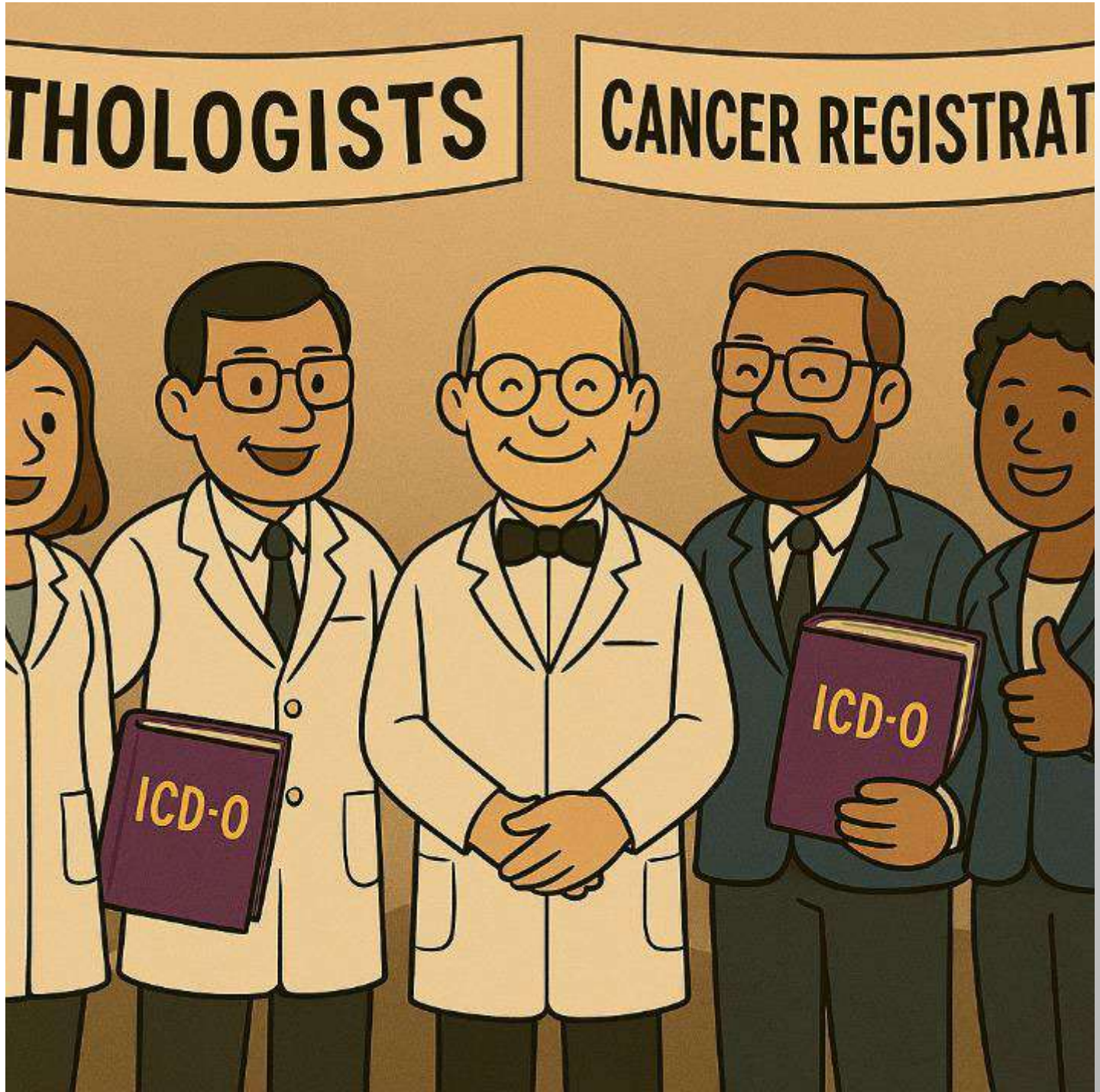

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Morphology

Do we *really* need a new coding system?

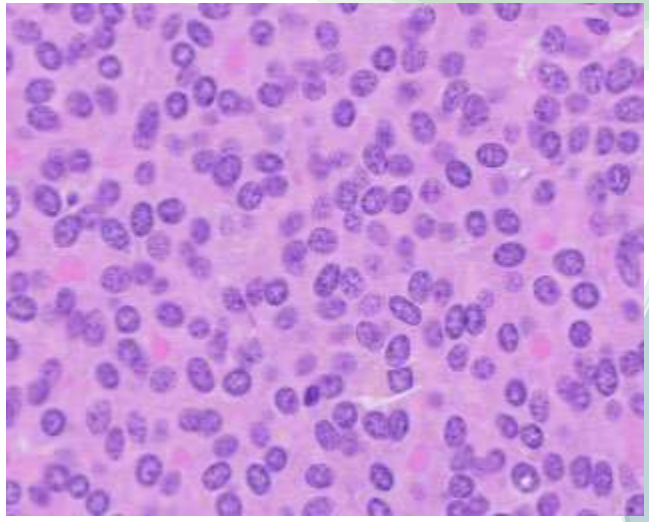
9680/3	Diffuse large B-cell lymphoma, NOS
9680/3	Diffuse large B-cell lymphoma, centroblastic subtype
9680/3	Diffuse large B-cell lymphoma, anaplastic subtype
9680/3	DLBCL, germinal-centre B-cell subtype
9680/3	Diffuse large B-cell lymphoma, activated B-cell subtype
9680/3	DLBCL with *MYC* and *BCL6* rearrangements
9680/3	Diffuse large B-cell lymphoma / high-grade B-cell lymphoma with *MYC* and *BCL2* rearrangements (DLBCL/HGBL-*MYC*/*BCL2*)
9680/3	EBV-positive diffuse large B-cell lymphoma
9680/3	DLBCL associated with chronic inflammation
9680/3	Primary large B-cell lymphoma of immune-privileged sites†
9680/3	Primary large B-cell lymphoma of the CNS
9680/3	Primary large B-cell lymphoma of the vitreoretina
9680/3	Primary large B-cell lymphoma of the testis
9680/3	Primary cutaneous diffuse large B-cell lymphoma, leg type
9680/3	High-grade B-cell lymphoma, NOS
9680/3	High-grade B-cell lymphoma, NOS, with *MYC* and *BCL6* rearrangements

Options

- Do nothing
- Use another coding system
  - SNOMED
  - ICD11
- Create a new coding system
  - Start from scratch
  - Build on ICD-O3
    - Expand from just using 8 and 9 as first digit?
    - Extra-digit
    - Letters/numbers

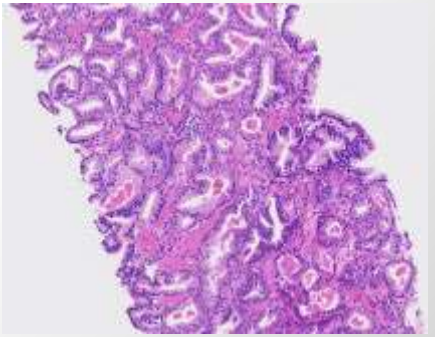
SNOMED CT
The global language of healthcare

Well-differentiated neuroendocrine tumor grade 2 of lung (disorder)
SCTID: 1287175002



2C25.4 Carcinoid or other malignant neuroendocrine neoplasms of bronchus or lung
XH51K1 Neuroendocrine tumour, grade 2

Evolution of neoplasm coding



09.8 Adenocarcinoma NOS

MOTNAC
1951

ACS

MOTNAC
1968

ACS

SNOP
1965
Sections 8,9
Neoplasms
CAP

8143 Adenocarcinoma NOS

ICD-O
1976

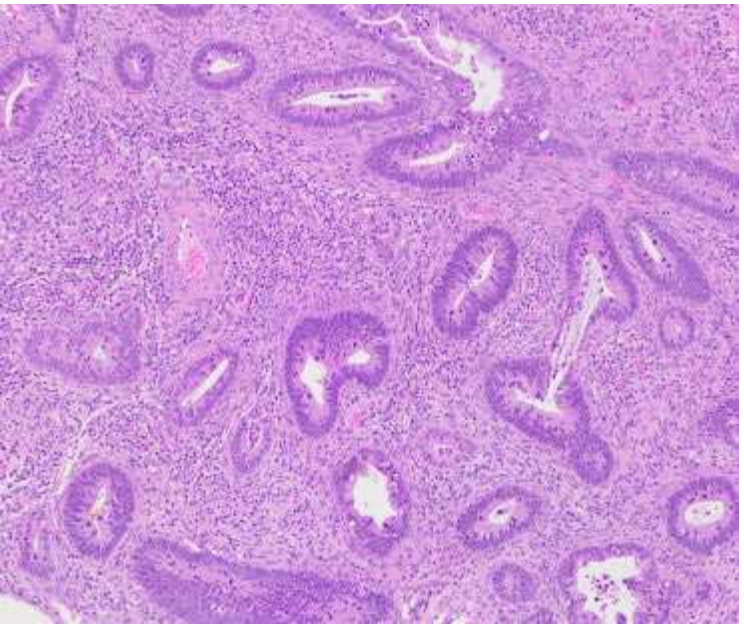
WHO

SNOMED
Morphology
Neoplasms
1977
CAP

8140/3 Adenocarcinoma, NOS

New morphology coding structure

81400/3



This digit is alphanumeric

Will one extra digit be enough?

- 26 letters and 9 digits – so 35x more codes.

- What about all the different combinations of molecular abnormalities tumours?
  - The WHO Classification does not intend to list or molecular combinations, it aims to classify tumours and only uses molecular features where they signify biological uniqueness.

- ICD-O is not aiming to code all molecular data
  – if this is desired it should be coded through other mechanism.

ICD-O4 Guiding framework

1. Each distinct entity should have a unique code
2. Morphology and Topography codes should align to ICDO3.2 as much as possible.
3. Changes should reflect codes and terminology used in the WHO classification 5th series.

Synonyms & Related terms

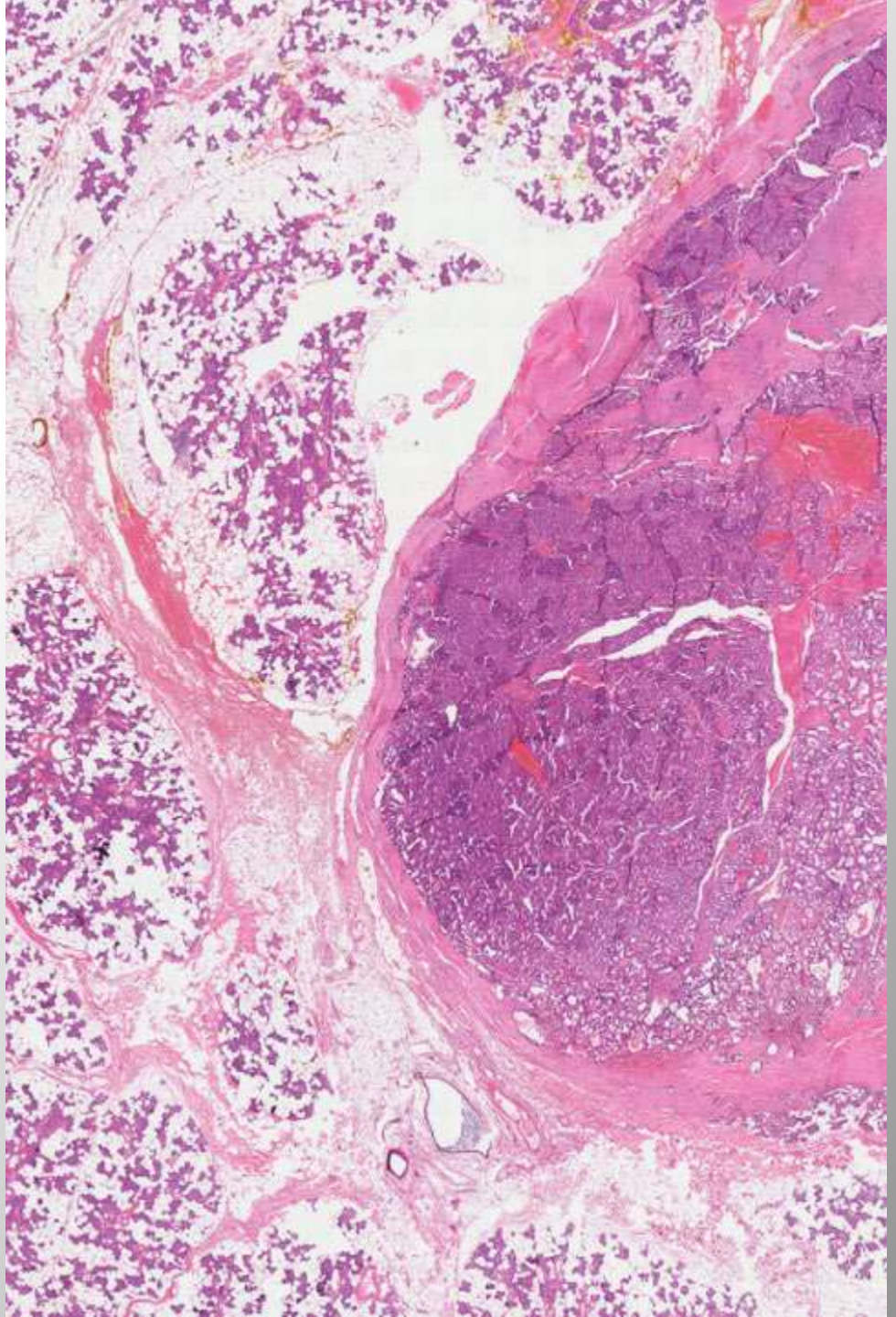
- Synonym – an alternative name for the same entity
- Related term - not a synonym, but coded the same

C07.9 Parotid gland

Parotid, NOS

Stensen duct

Parotid gland duct



ICDO3.1 and ICDO3.2 unfortunately used Related terms for distinct entities

97270/3 **9727/3 Precursor cell lymphoblastic lymphoma, NOS** *(see also M-9835/3)* [obs]
Malignant lymphoma, lymphoblastic, NOS *(see also M-9835/3)*
Lymphoblastoma [obs]
Malignant lymphoma, convoluted cell [obs]

97271/3 Blastic NK cell lymphoma [obs]

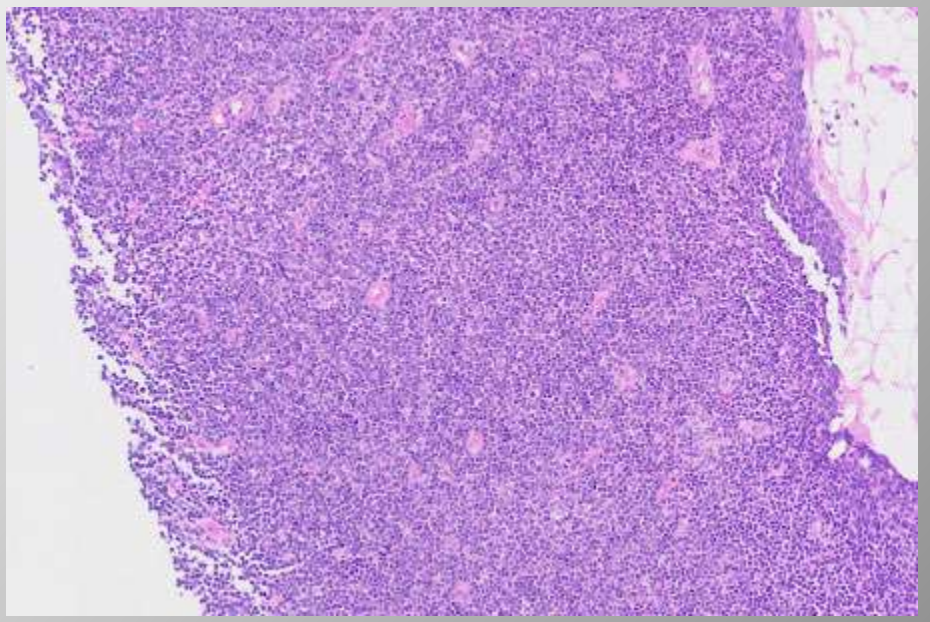
97272/3 Blastic plasmacytoid dendritic cell neoplasm

98110/3

ICD-O coding
9811/3 B-lymphoblastic leukaemia/lymphoma

98370/3

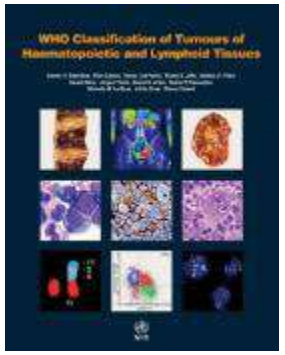
ICD-O coding
9837/3 T-lymphoblastic leukaemia/lymphoma, NOS



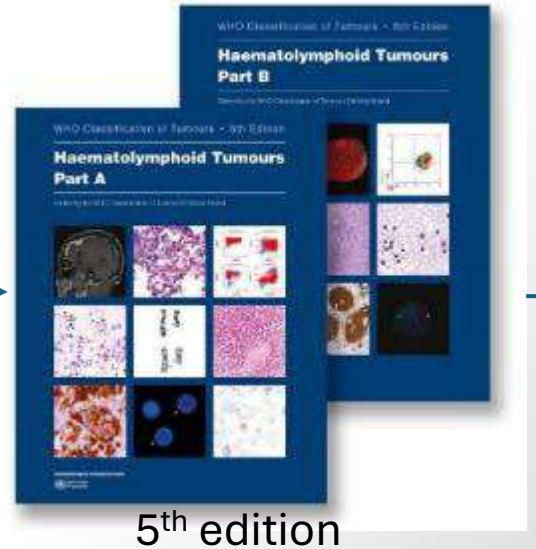
~~8044/3 – Small cell carcinoma, intermediate type~~

80440/3 8044/3 – Small cell carcinoma, hypercalcaemic type (C56.9)

Relationship between WHO Blue Books and ICD-O

ICD-O3.1 → 4th edition 

→ **Anaplastic large cell lymphoma, ALK-negative** 9715/3* → ICD-O3.2

ICD-O3.2 → 

5th edition
→ **Clonal cytopenia of undetermined significance** 9980/1* → ICD-O4

Adding new terms from WHO BB 5th series

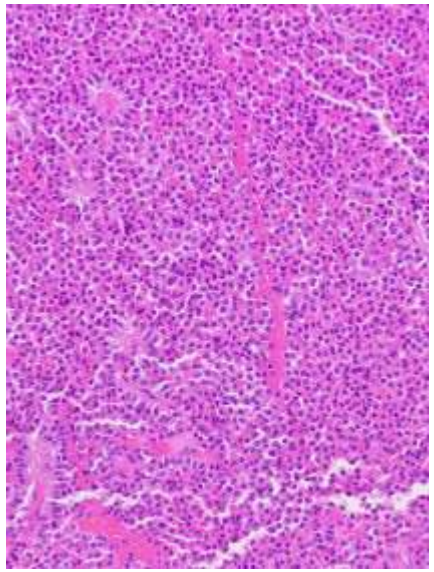
For both entities and sub-types:

- Adding new preferred terms where terminology has changed
- Adding recommended terms
- Not adding ‘Not recommended terms’ (but not removing if already exists).

Mammosomatotroph PitNET/adenoma

Definition

Mammosomatotroph pituitary neuroendocrine tumour (PitNET)/adenoma is a well-differentiated PitNET composed of PIT1-lineage adenohypophysial cells with mammosomatotroph differentiation.



ICD-O coding

8280/3 Mammosomatotroph PitNET/adenoma

82803/3

ICD-11 coding

2D12.Y Other specified malignant neoplasms of other endocrine glands or related structures

Related terminology

Acceptable: mammosomatotroph tumour.

Not recommended: growth hormone and prolactin–positive PitNET/adenoma; growth hormone and prolactin–producing PitNET/adenoma; growth hormone and prolactin–producing tumour/adenoma.

Subtype(s)

None

Keeping coding decisions made in 5th series of Blue Books:

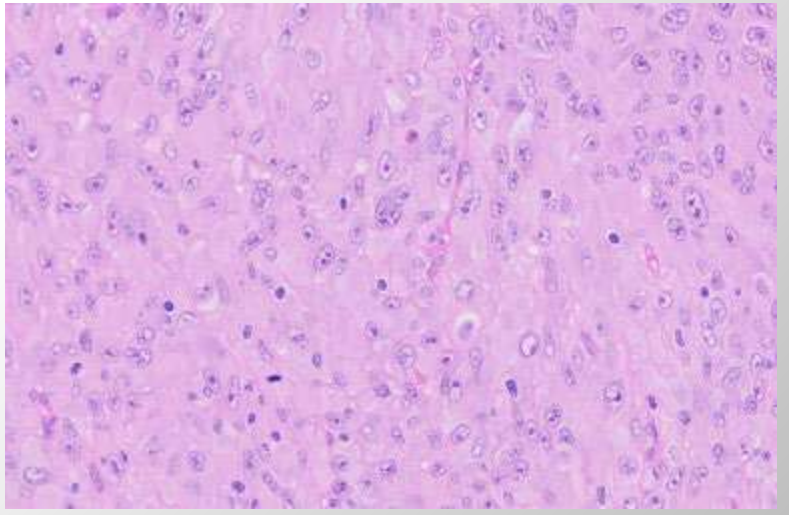
- Some codes look out of place because of the decision to maintain coding endorsed in 5th series of WHO classification.
- "**Hepatoblastoma, hepatocellular neoplasm, NOS**"
  - Previously thought to be interemediate or combined biology between Hepatocellular carcinoma and Hepatoblastoma.
  - Now considered a sub-type of Hepatoblastoma.
  - "Must not have a HCC related code and cannot share a code with hepatoblastoma"
  - Ended up with 8000/3. 80001/3

Keeping coding decisions made in 5th series of Blue Books:

ICD-O coding

8120/3 Invasive urothelial carcinoma
8120/3 Conventional urothelial carcinoma
8120/3 Urothelial carcinoma with squamous differentiation
8120/3 Urothelial carcinoma with glandular differentiation
8120/3 Urothelial carcinoma with trophoblastic differentiation
8120/3 Nested urothelial carcinoma
8120/3 Large nested urothelial carcinoma
8120/3 Tubular and microcystic urothelial carcinomas
8131/3 Micropapillary urothelial carcinoma
8082/3 Lymphoepithelioma-like urothelial carcinoma
8122/3 Plasmacytoid urothelial carcinoma
8031/3 Giant cell urothelial carcinoma
8120/3 Lipid-rich urothelial carcinoma
8120/3 Clear cell (glycogen-rich) urothelial carcinoma
8120/3 Sarcomatoid urothelial carcinoma
8020/3 Poorly differentiated urothelial carcinoma

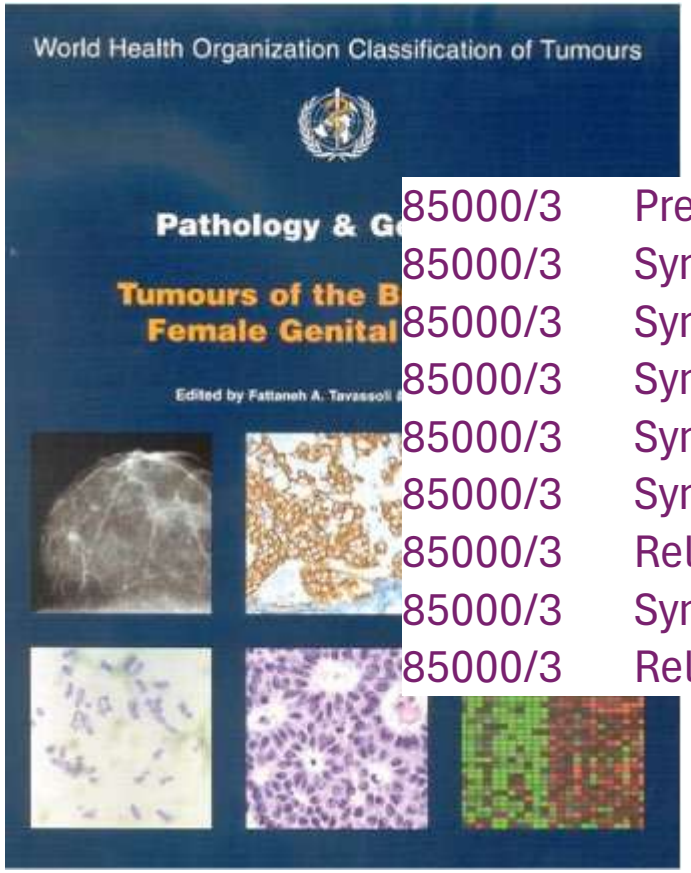
80204/3



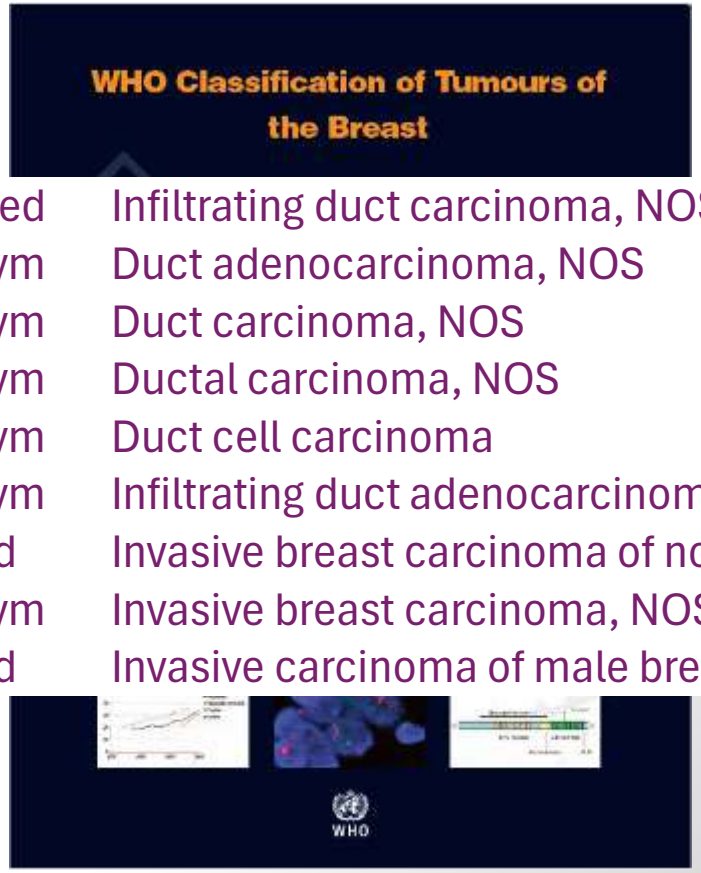
"The poorly differentiated urothelial carcinoma subtype encompasses carcinomas lacking morphological features that point to a urothelial origin. Immunohistochemistry is necessary to establish epithelial differentiation in such tumours."

8020/3 Carcinoma, undifferentiated, NOS

Carcinoma, poorly differentiated, NOS



85000/3	Preferred	Infiltrating duct carcinoma, NOS
85000/3	Synonym	Duct adenocarcinoma, NOS
85000/3	Synonym	Duct carcinoma, NOS
85000/3	Synonym	Ductal carcinoma, NOS
85000/3	Synonym	Duct cell carcinoma
85000/3	Synonym	Infiltrating duct adenocarcinoma
85000/3	Related	Invasive breast carcinoma of no special type
85000/3	Synonym	Invasive breast carcinoma, NOS
85000/3	Related	Invasive carcinoma of male breast



Invasive ductal carcinoma, NOS

2003

Invasive carcinoma of no special type

2011

Invasive breast carcinoma of no special type

"Special morphological patterns"

2019

the frequency found in endometrioid and clear cell carcinomas, further supporting their close relationship {2053}.

Prognosis and predictive factors

These tumours are associated with a good outcome even in the presence of peritoneal implants {474,1612,1655,1656,1743}.

Seromucinous carcinoma

Definition

A carcinoma composed predominantly of serous and endocervical-type mucinous epithelium. Foci containing clear cells and areas of endometrioid and squamous differentiation are not uncommon.

ICD-O code 8474/3

Synonyms

Endocervical-type mucinous and mixed epithelial carcinomas of Müllerian type

Epidemiology

These tumours are quite uncommon and therefore data on the epidemiology are not available.

Clinical Features

The mean age in one reported series {1743} was 45 years. Most patients presented with a pelvic mass and 57% of the women had peritoneal endosalpingiosis.

Macroscopy

The mean tumour size in one series was 12 cm and over half the tumours were bilateral {1743}. Tumours were unilocular or multilocular and contained solid areas. Papillary excrescenses were present on the inner lining of the cysts and on the surface.

Histopathology

Tumours are generally papillary and display epithelial stratification closely resembling serous tumours. The most common pattern of invasion is cribriform and

84740/3 Seromucinous carcinoma [obs]

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Seromucinous carcinoma

Seromucinous carcinoma

Seromucinous carcinoma was included in the previous classification and was defined as a carcinoma composed predominantly of serous and endocervical-type mucinous epithelium, often with foci containing clear cells and areas of endometrioid and squamous differentiation { 25723110 }. It has been removed from this classification because this is a poorly reproducible diagnosis and there is significant morphological overlap with other tumour types, especially endometrioid carcinoma. Immunohistochemical and molecular studies have also suggested that most cases represent unusual morphological patterns of endometrioid carcinoma { 28125452 }; therefore, seromucinous carcinoma is now considered a subtype of endometrioid carcinoma and is discussed in that section (see section *Endometrioid carcinoma of the ovary*).

Chronic myeloid leukaemia

Chronic myeloid leukaemia

Definition

Chronic myeloid leukaemia (CML) is a myeloproliferative neoplasm defined by the *BCR::ABL1* fusion gene and characterized by neutrophilic granulocytosis.

ICD-O coding

9875/3 Chronic myeloid leukaemia

9863/3 Chronic myeloid leukemia, NOS

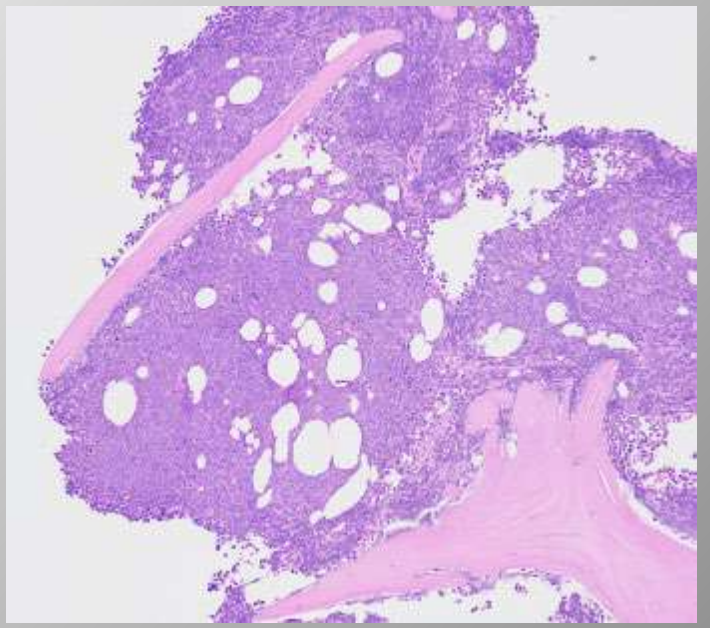
Chronic granulocytic leukemia, NOS
Chronic myelocytic leukemia, NOS
Chronic myelogenous leukemia, NOS

98630/3 Chronic myeloid leukaemia, NOS (excludes if stated to be BCR::ABL1, M-98750/3)

9875/3 Chronic myelogenous leukemia, BCR/ABL positive

Chronic granulocytic leukemia, BCR/ABL
Chronic granulocytic leukemia, Philadelphia chromosome (Ph1) positive
Chronic granulocytic leukemia, t(9;22) (q34;q11)
Chronic myelogenous leukemia, Philadelphia chromosome (Ph1) positive
Chronic myelogenous leukemia, t(9;22) (q34;11)

98750/3 Chronic myeloid leukaemia, BCR::ABL1 positive



“Carcinoma”

Often used by pathologists in WHO classification when they mean Adenocarcinoma.

Carcinoma of the gallbladder

Definition

Carcinoma of the gallbladder is a malignant epithelial neoplasm arising in the gallbladder from biliary epithelium.

ICD-O coding

8140/3 Adenocarcinoma NOS
8070/3 Squamous cell carcinoma NOS
8020/3 Carcinoma, undifferentiated, NOS

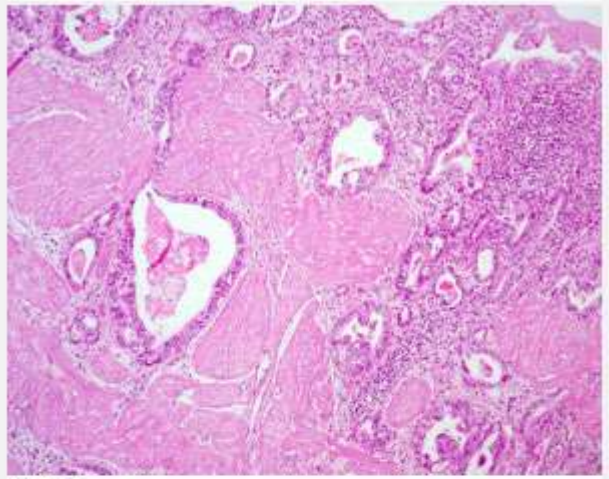
81400/3 Preferred Adenocarcinoma, NOS
81400/3 Related Carcinoma of the gallbladder

Papillary thyroid carcinoma

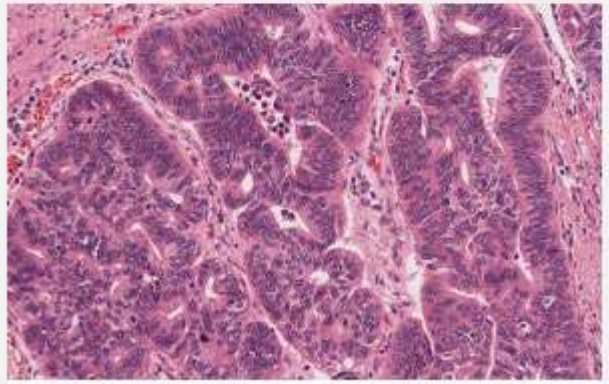
Definition

Papillary thyroid carcinoma (PTC) is a malignant tumour of follicular cell derivation characterized by distinct nuclear features. PTC diagnosis requires either papillary or solid/trabecular architecture, or invasive growth in follicular-patterned tumours.

82600/3 Preferred Papillary adenocarcinoma, NOS
82600/3 Related Papillary thyroid carcinoma, NOS



#978
Carcinoma of the gallbladder



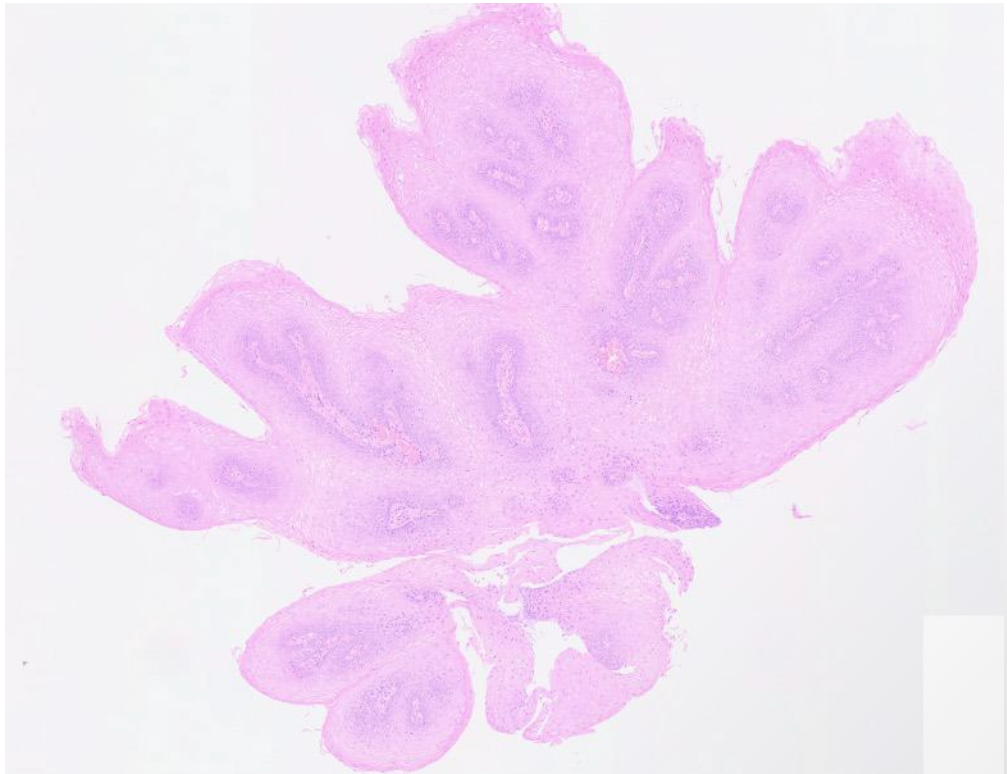
#979
Carcinoma of the gallbladder

https://tumourclassification.iarc.who.int/chaptercontent/31/110
https://tumourclassification.iarc.who.int/chaptercontent/53/44

Not otherwise specified

8052/0 Squamous cell papilloma, NOS
Keratotic papilloma
Squamous papilloma

8053/0 Squamous cell papilloma, inverted



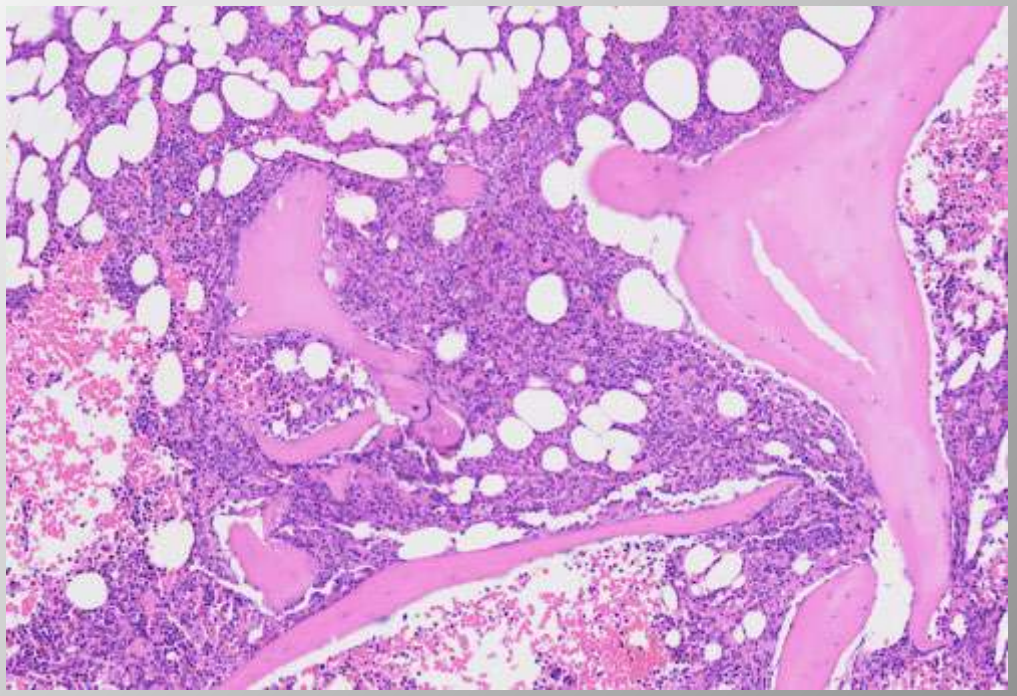
- “NOS” is printed after topographic and morphologic terms that appear elsewhere in ICD-O with an additional modifying word or phrase.

- In the WHO classification, there is a general dislike for using NOS and it is generally reserved for a tumour that does not fit into a more specific category e.g. renal cell carcinoma, NOS

NOS / Unclassifiable

- WHO 4 & 4.1
  - [Myeloproliferative neoplasm, NOS -9960/3]
  - Myeloproliferative neoplasm, unclassifiable -9975/3

- WHO 5
  - Myeloproliferative neoplasm, NOS – 9975/3



99600/3 Myeloproliferative neoplasm, NOS (See also M-99750/3)
99750/3 Myeloproliferative neoplasm, NOS (unclassifiable)

Aligning with ICDO3.2: Neuroendocrine neoplasms

801-804 Epithelial neoplasms, NOS

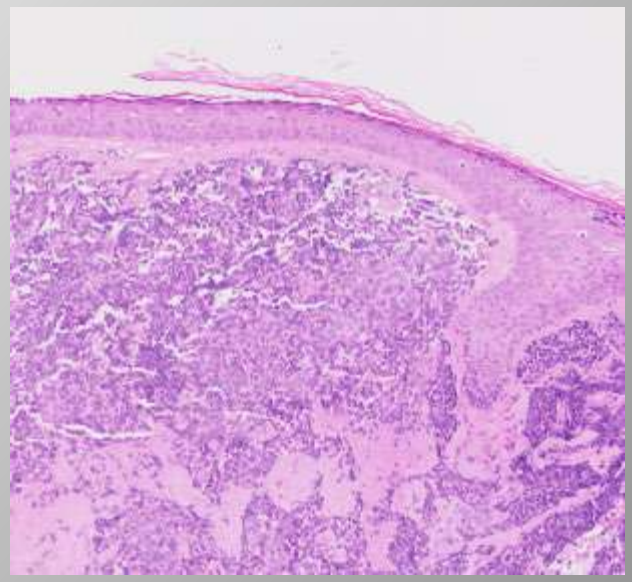
80130/3 large cell neuroendocrine carcinoma, NOS
80410/3 small cell neuroendocrine carcinoma

906-909 Germ cell neoplasms

90911/1 Strumal carcinoid

814-838 Adenomas and adenocarcinomas

82401/3 Neuroendocrine tumour, grade 1
82460/3 Neuroendocrine carcinoma
82470/3 Merkel cell carcinoma
82491/3 Neuroendocrine tumour, grade 2



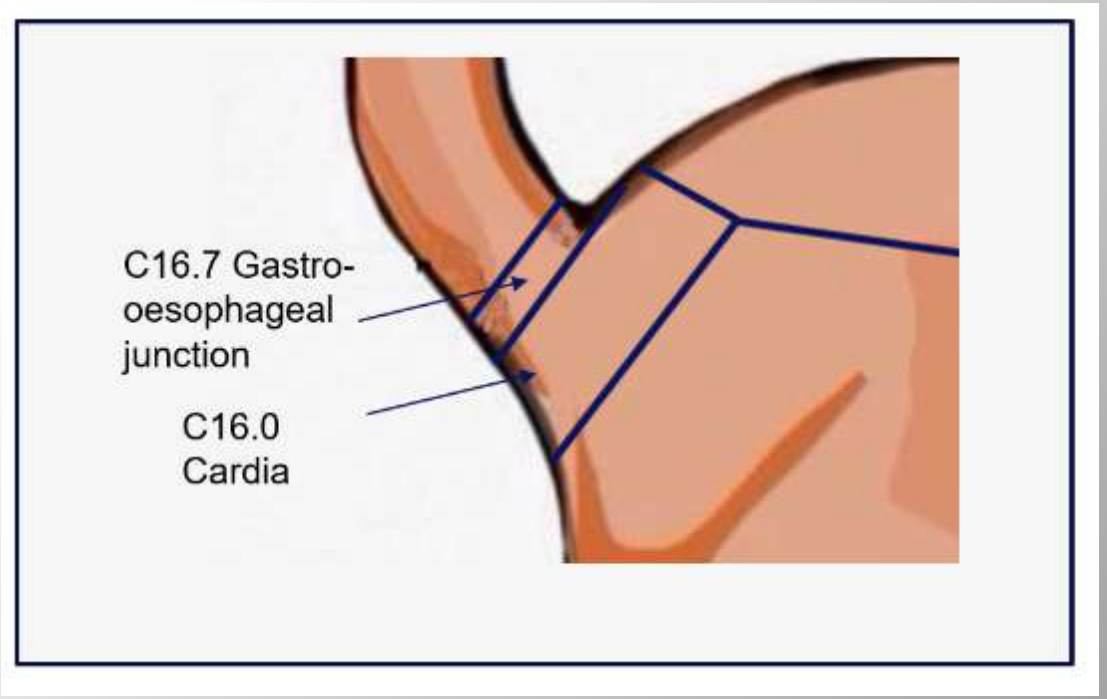
868-871 Paragangliomas and glomus tumors

86930/3 Extra-adrenal paraganglioma, NOS
87000/3 Phaeochromocytoma

Topography

New code for Gastro-oesophageal junction

- GOJ previously included in C16.0 with gastric cardia, but staged with oesophageal tumours.
- Options:
  - Move to C15
    - Logical in terms of staging and sequence
  - **Move to another point in C16 (C16.7)**
    - Keeps in C16 so counts of C15 and C16 are unaltered



New code for Peri-anal skin

- Peri-anal skin previously included in C44.5 with skin of trunk, but staged with anal cancers.
- Options:
  - **Move to C21 (C21.3)**
    - Logical in terms of staging and sequence
  - Move to another point in C44
    - Keeps in C44 so counts of C44 and C21 are unaltered

Small note about “Topography with optional extra digit”

- These are largely further adaptations to accommodate identify staging system by tumour site/morphology.

- Ileocecal valve staged with cecum for carcinomas.
- Ileocecal valve staged with ileum (C17.2) for NET.

C18.0	**Cecum**
	Ileocecal valve
	Ileocecal junction

C18.00	**Cecum**
C18.01	Ileocecal valve
C18.01	Ileocecal junction