Changes in ICD-O3.2

Otto Visser

21 April 2021

European Commission

ICD-O-3.2

- Information about ICD-O-3.2 can be found at the website if the International Association of Cancer Registries (www.iacr.com.fr)
- ICD-O-3.2 will be available in print in the first half of 2021
- Its use is recommended for use from (year of incidence) 2020:
  - If a code for a specific entity changes, use the 'old' code before 2020 and the 'new' code as of 2020
  - If a code for a specific entity is missing in ICD-O-3.1 (e.g. for 'NUT carcinoma' M8023/3), that code may be used before 2020.

Content of the lesson

- Why changes?
- New terms and synonyms
- Changes in behaviour codes
- Changes in morphology codes
- New morphology codes

Why changes?

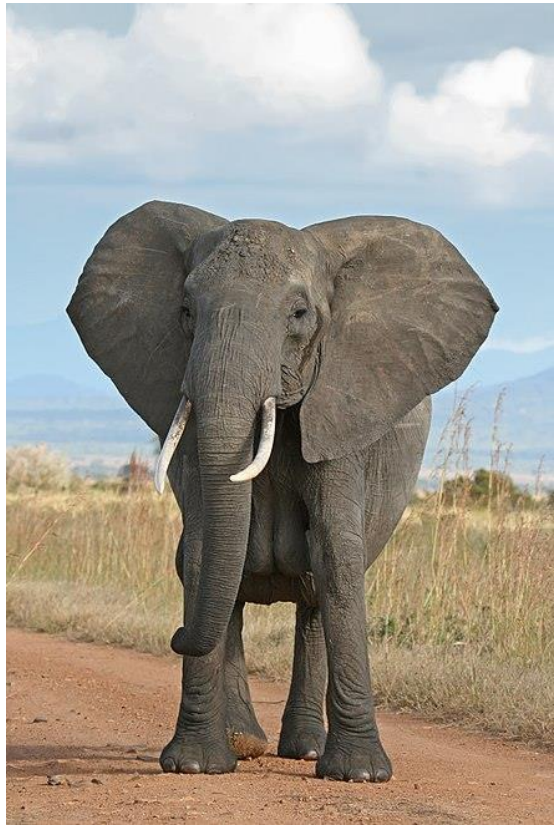
European Commission

Elephants

African elephant

Asian elephant

Elephants



African bush elephant

African forest elephant

Why changes?

- Research has shown that two entities are in fact the same entity with different appearance or that one entity is in fact a combination of different entities
- With new diagnostic techniques (cytogenetics, molecular diagnostics) subcategories have been defined in existing diagnoses, for example AML with mutated RUNX1 or IDH-mutant glioblastoma
- New evidence has become available regarding the risk of recurrence or distant metastasis which has led to a change in behaviour code
- Terminology changes over time

New (preferred) terms and synonyms

European Commission

New (preferred) terms and synonyms

- Many new terms and synonyms have been introduced
- There are too many to mention them all
- The purpose of the synonyms is to help registrars in finding the correct morphology code, even if an other term than the preferred one has been used

New (preferred) terms and synonyms

For example:

- In the past ‘carcinoid’ (morphology code 8240/3) was a commonly used diagnosis, but now ‘neuroendocrine tumour (NET)’ is the preferred term. As many pathologists still use ‘carcinoid’ it is mentioned as a synonym of ‘NET’.
- ‘Bronchiolo-alveolar carcinoma’ (morphology code 8250/3) of the lung is now called ‘lepidic adenocarcinoma’. Therefore, ‘lepidic adenocarcinoma’ is the preferred term and ‘bronchiolo-alveolar carcinoma’ is a synonym.

Changes in behaviour codes

European Commission

Changes in behaviour code: from /0 to /2

Classification of squamous intraepithelial neoplasia (squamous dysplasia):

- Old classification: grade I, grade II, grade III
- New classification: low grade, high grade

old	morphology	new	morphology
Squamous intraepithelial neoplasia, grade I	8077/0	Squamous intraepithelial neoplasia, low grade	8077/0
Squamous intraepithelial neoplasia, grade II	8077/0	Squamous intraepithelial neoplasia, high grade	8077/2
Squamous intraepithelial neoplasia, grade III	8077/2		

Grade II is reclassified as high grade and therefore changes from 8077/0 to 8077/2

Changes in behaviour code: from /0 to /2

Sites with squamous dysplasia include:

- Cervix (CIN)
- Anus (AIN)
- Vulva (VIN)
- Vagina (VAIN)
- Esophagus

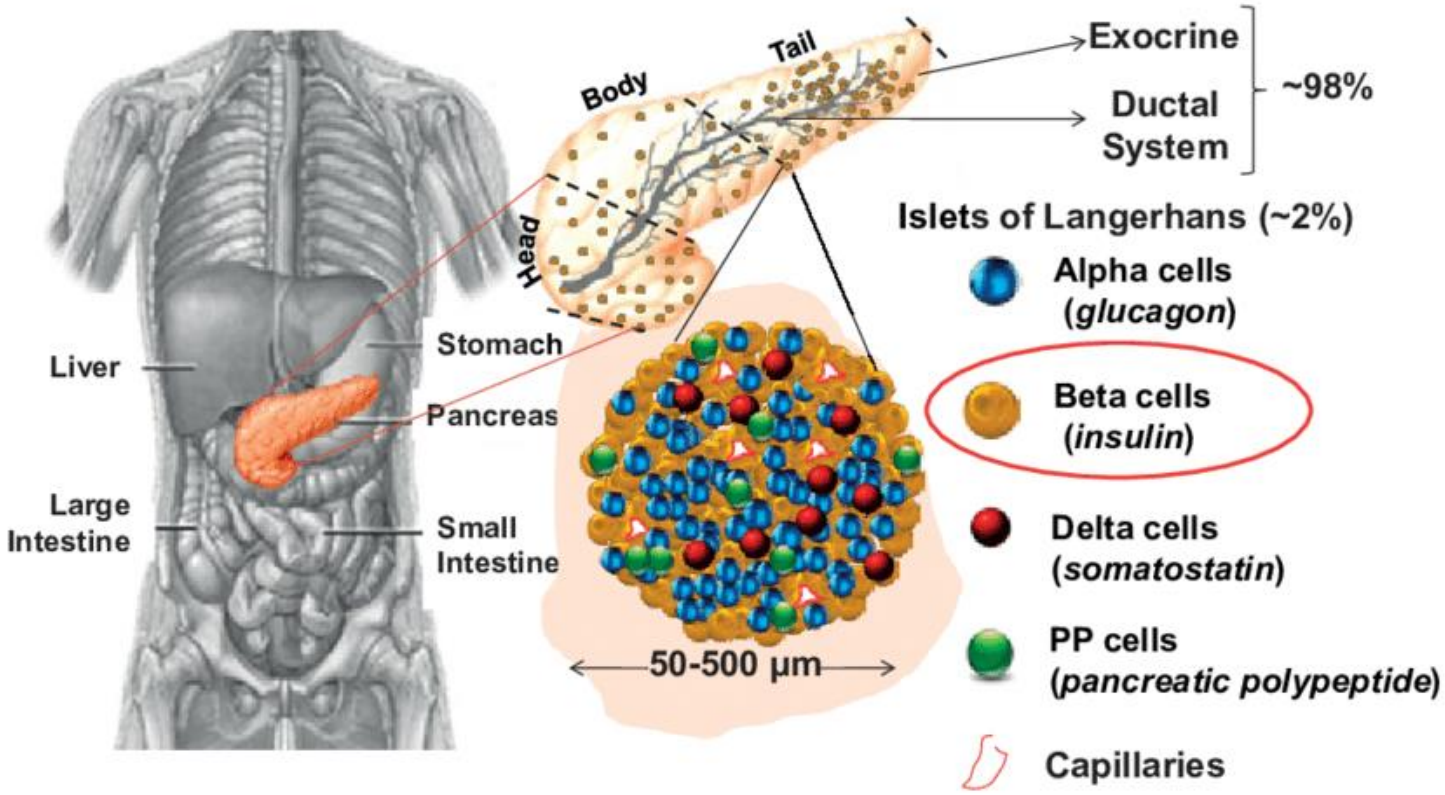
Changes in behaviour code: from /0 or /1 to /3

- Islet cell tumours of the pancreas
- Skin appendage tumour: aggressive digital papillary adenoma
- Thymoma
- Granulosa cell tumour, adult type
- Paraganglioma & pheochromocytoma
- Gastrointestinal stromal tumour
- Epithelioid hemangioendothelioma
- Clear cell odontogenic tumour

Islet cell tumours of the pancreas

Islet cell tumours are neuroendocrine tumours (NETs) of the islets of Langerhans in the pancreas:

- Non-functioning pancreatic NETs
- Functioning pancreatic NETs



Islet cell tumours of the pancreas

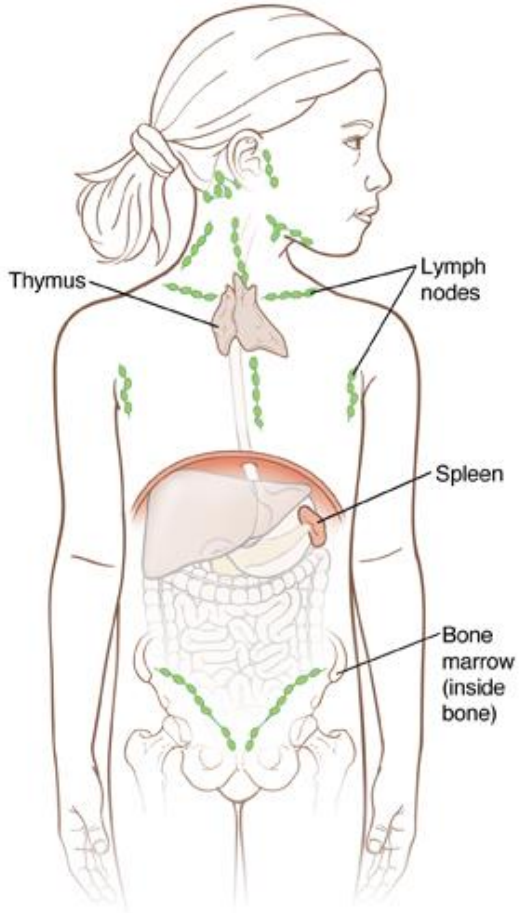
- Functioning tumours are associated with clinical syndromes caused by abnormal secretion of hormones by the tumour cells
- Non-functioning tumours are not associated with clinical syndromes, but they may secrete hormones that do not cause symptoms
- Non-functioning tumours <5 mm (microadenomas) are considered benign

Islet cell tumours of the pancreas

tumour	morphology
Pancreatic NET, non-functioning	8150/3
Pancreatic NET, functioning:	
- insulin-producing (insulinoma)	8151/3
- glucagon-producing (glucagonoma)	8152/3
- gastrin-producing (gastrinoma)	8153/3
- VIPoma	8155/3
- somatostatin-producing (somatostatinoma)	8156/3
- ACTH-producing	8158/3
- serotonin-producing	8241/3

Thymomas

A thymoma is a tumour of the thymus composed of neoplastic epithelial cells of the thymus epithelium as well as non-neoplastic thymocytes. Thymocytes are hematopoietic stem cells from the bone marrow that develop into T-lymphocytes in the thymus.



Thymomas

- Depending on the proportion of thymocytes and resemblance to cells from the cortex or medulla, different subtypes of thymoma (A, B) are recognized.
- All thymomas are considered malignant except:
  - Microscopic thymoma (8580/0)
  - Micronodular thymoma with lymphoid stroma (8580/1)

Thymomas

tumour	morphology of epithelial cells	proportion of thymocytes	morphology
Thymoma, NOS	?	?	8580/3
Thymoma, type A (medullary)	medullary	+	8581/3
Thymoma, type B1 (lymphocyte-rich)	cortical	+++	8583/3
Thymoma, type B2 (cortical)	cortical	++	8584/3
Thymoma, type B3 (epithelial)	cortical	+	8585/3
Thymoma, type AB (mixed)	both		8582/3

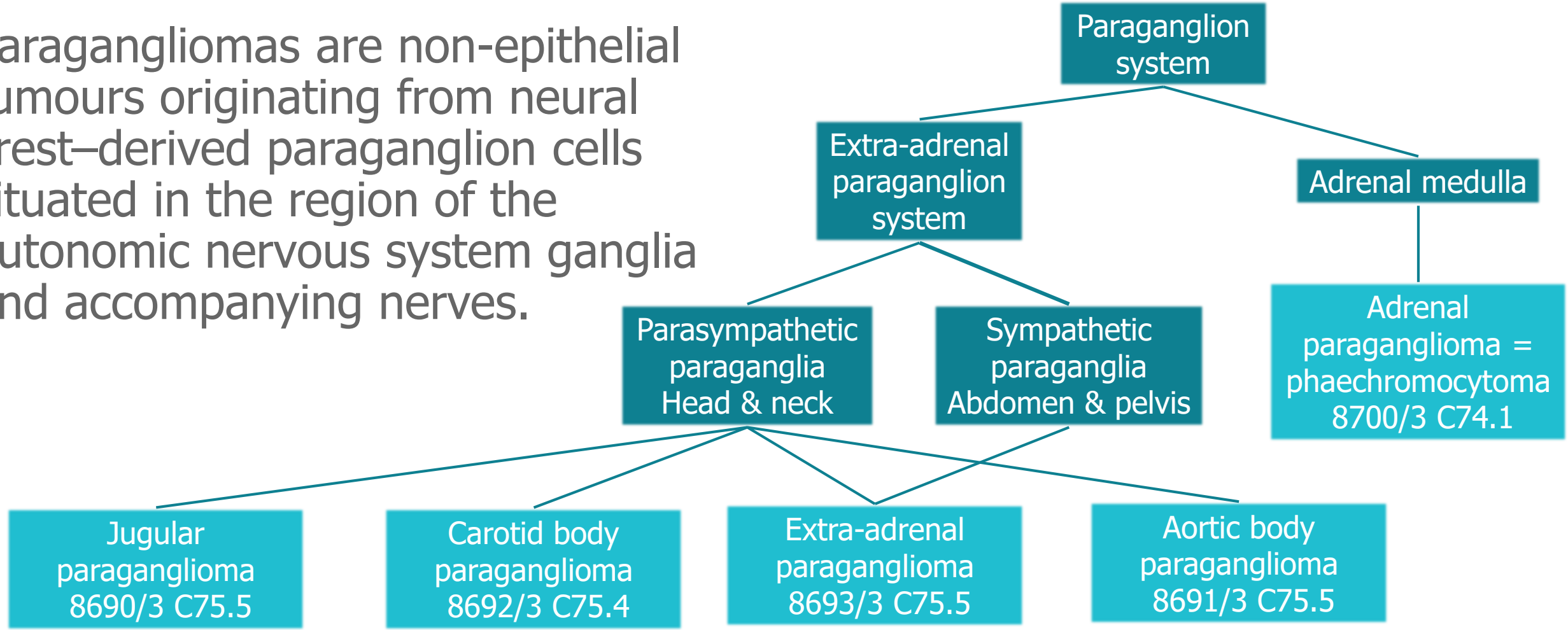
Granulosa cell tumour

- Granulosa cell tumour is the most common sex cord-stromal tumour of the ovary
- There are two types:
  1. Adult type (all ages [median age 57 years]; M8620/3)
  2. Juvenile type (age range 0-35 years [median 15 years], M8622/1)

- Adult granulosa cell tumours have a propensity for late recurrence up to 20 years after diagnosis and therefore they are now all classified as malignant

Paraganglioma & pheochromocytoma

Paragangliomas are non-epithelial tumours originating from neural crest–derived paraganglion cells situated in the region of the autonomic nervous system ganglia and accompanying nerves.



Spinal paraganglioma

- Paraganglioma of the cauda equina (C72.1) or filum terminale (C72.0) of the spinal cord is a tumour of the central nervous system
- Corresponds histologically to CNS grade I
- Therefore still coded as 8680/1

Gastrointestinal stromal tumour (GIST)

- The potency for recurrence or metastases of GIST is dependent of
  - size of the tumour
  - number of mitoses
- Small tumours without mitosis were considered benign (/0), large tumours or tumours with many mitoses were considered malignant (/3); the intermediate group was considered borderline malignant (/1).
- It is now recommended to register all GIST as malignant (/3), but it is still relevant to register the tumour size and the number of mitosis

Other tumours

- Skin appendage tumour: aggressive digital papillary adenoma (8408/3)
- Epithelioid haemangioendothelioma, NOS (9133/3)
- Clear cell odontogenic tumour (9341/3)

Changes in behaviour code: from /3 to /1

- Hydroa vacciniforme-like lymphoma (9725/3 → 9725/1)
- Primary cutaneous CD4+ small/medium T-cell lymphoma (9709/3→9709/1)
- Polymorphic post transplant lymphoproliferative disorder (9971/3 → 9971/1)
- Dermatofibrosarcoma protuberans
- Primary cutaneous CD30+ T-cell lymphoproliferative disorder/lymphomatoid papulosis
- Langerhans histiocytosis, NOS/mono-ostotic/poly-ostotic

Dermatofibrosarcoma

old	morphology	new	morphology
Dermatofibrosarcoma protuberans, NOS	8832/3	Dermatofibrosarcoma protuberans, NOS	8832/1
Dermatofibrosarcoma protuberans, (fibrosarcomatous)	8832/3	Dermatofibrosarcoma protuberans, (fibrosarcomatous)	8832/3

Primary cutaneous CD30+ T-cell disorders

old	morphology	new	morphology
Primary cutaneous CD30+ T-cell lymphoproliferative disorder/lymphomatoid papulosis	9718/3	Primary cutaneous CD30+ T-cell lymphoproliferative disorder/lymphomatoid papulosis	9718/1
Primary cutaneous anaplastic (CD30+) large cell lymphoma	9718/3	Primary cutaneous anaplastic (CD30+) large cell lymphoma	9718/3

Langerhans cell histiocytosis

	ICD-O ICD-O-2	ICD-O-3	ICD-O-3.1	ICD-O-3.2
Langerhans cell histiocytiosis, NOS	-	9751/1	9751/3	9751/1
Langerhans cell histiocytiosis, unifocal/mono-ostotic		9752/1		
Langerhans cell histiocytiosis, multifocal/poly-ostotic		9753/1		
Langerhans cell histiocytiosis, disseminated/multisystem (Letterer-Siwe disease)	9722/3	9754/3		9751/3

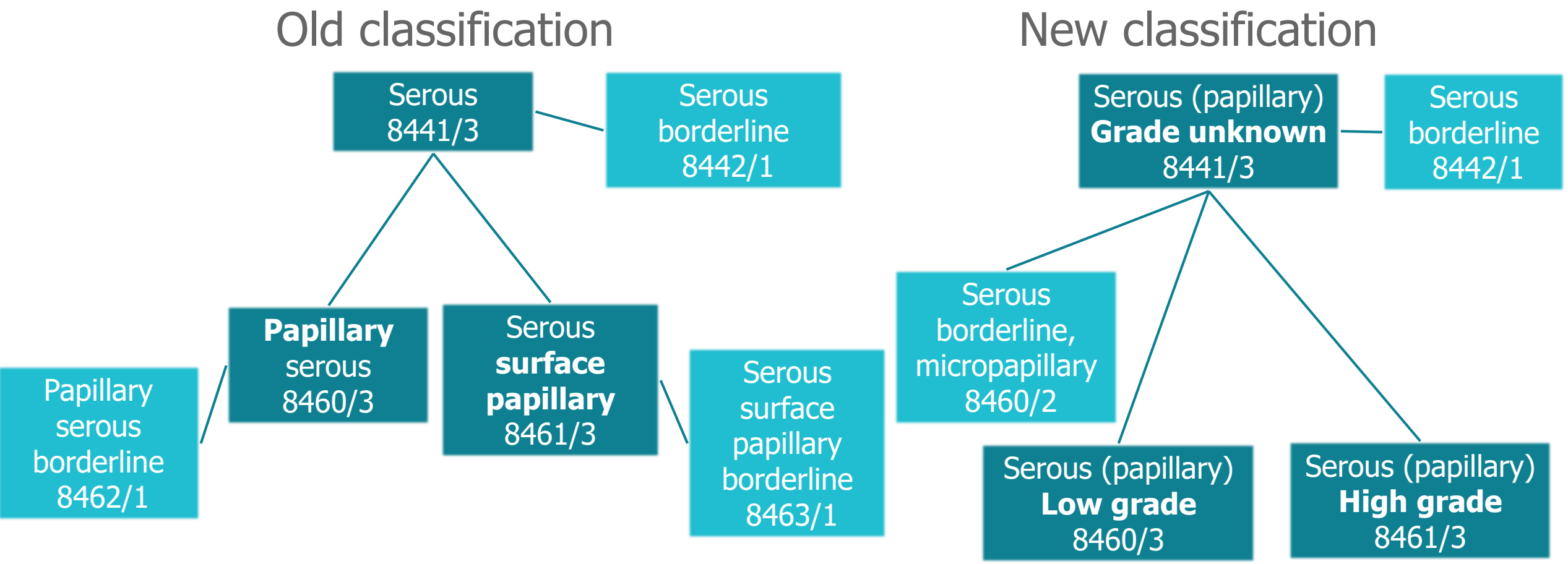
Change of morphology codes

European Commission

Change in morphology codes

tumour/malignancy	old code	new code
Warty carcinoma / condylomatous carcinoma	8051/3	8054/3
Papillary serous (cyst)adenocarcinoma Serous surface papillary carcinoma	8460/3 8461/3	8441/3 Serous (cyst)adenocarcinoma
Cystic hypersecretory carcinoma Cystic hypersecretory carcinoma, intraductal	8508/3 8508/2	8500/2 DCIS
Hemangiopericytoma, malignant	9150/3	8815/3 solitary fibrous tumour
Ewing sarcoma	9260/3	9364/3 PNET
Refractory neutropenia Refractory thrombocytopenia	9991/3 9992/3	9980/3 MDS with single lineage dysplasia

Serous (cyst)adenocarcinoma of the ovary



One code for lymphoma and leukaemia

malignancy	old code	new code
Burkitt cell leukaemia	9826/3	9687/3 Burkitt lymphoma/leukaemia
Precursor B-cell lymphoblastic leukaemia (B-ALL) Precursor B-cell lymphoblastic lymphoma (B-LBL)	9836/3 9728/3	9811/3 B lymphoblastic leukaemia/lymphoma
Small lymphocytic lymphoma (SLL)	9670/3	9823/3 Chronic lymphocytic leukaemia/SLL
T lymphoblastic lymphoma	9729/3	9837/3 T lymphoblastic leukaemia/lymphoma

- Non-leukaemic cases without bone marrow infiltration should be coded at the primary site of the tumour (mostly lymph nodes [C77], but extranodal sites occur)
- Other cases should be coded at bone marrow (C42.1)

New morphology codes

European Commission

New morphology codes: carcinoma

tumour	morphology	most common sites
NUT associated carcinoma (NUT midline carcinoma) Rearrangement of the NUT (= NUclear protein in Testis) gene	8023/3	Midline (nasal cavity, mediastinum, lung)
Squamous cell carcinoma, HPV-positive Squamous cell carcinoma, HPV-negative	8085/3 8086/3	Oropharynx: tonsil (C09), base of tongue (C01)
Minimally invasive adenocarcinoma, non-mucinous Minimally invasive adenocarcinoma, mucinous	8256/3 8257/3	Lung (C34)
Follicular carcinoma, encapsulated, angioinvasive	8339/3	Thyroid (C73)
Seromucinous carcinoma	8474/3	Ovary (C56)
Solid papillary carcinoma in situ Solid papillary carcinoma with invasion Lobular carcinoma in situ, pleomorphic	8509/2 8509/3 8519/2	Breast (C50)

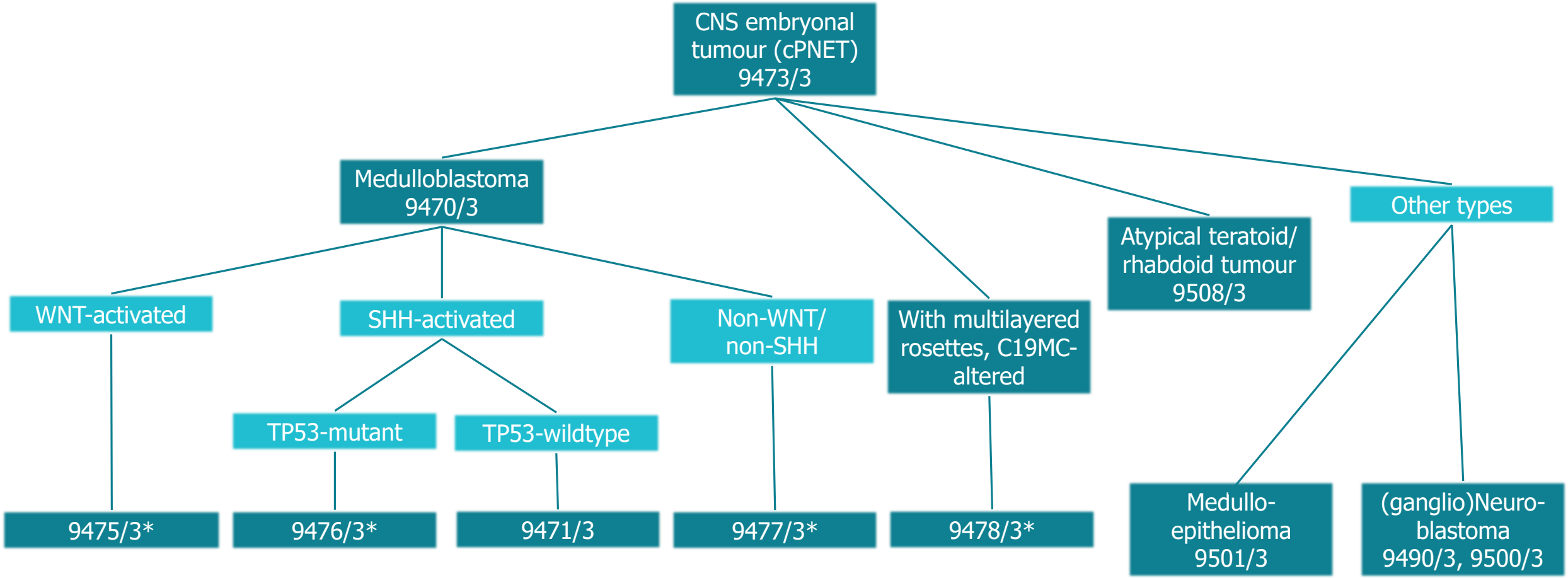
New morphology codes: non-epithelial tumours

tumour	morphology	most common sites
Perivascular epithelioid tumour (PEComa), malignant	8714/3	Uterus, kidney, retroperitoneum
Biphenotypic sinonasal sarcoma	9045/3	Nasal cavity (C30.0), sinuses (C31)
Intimal sarcoma	9137/3	Heart, pulmonary artery, other arteries
Malignant peripheral nerve sheath tumor, epithelioid	9542/3	Superficial trunk & extremities
Germ cell tumor with associated haematological malignancy (both evolve from a common shared precursor)	9086/3	Mediastinum

New morphology codes: central nervous system

tumour	morphology	most common sites
Pituitary blastoma	8273/3	Pituitary gland (C75.1)
Diffuse midline glioma, H3 K27M-mutant	9385/3	Brain stem (C71.7), spinal cord (C72.0)
Ependymoma, RELA fusion-positive	9396/3	Brain (C71)
Glioblastoma, IDH-mutant	9445/3	Cerebrum (C71)
Medulloblastoma, WNT-activated, NOS Medulloblastoma, SHH-activated and TP53 mutant Medulloblastoma, non-WNT/non-SHH	9475/3 9476/3 9477/3	Cerebellum (C71.6)
Embryonal tumor with multilayered rosettes with C19MC alteration	9478/3	Brain (C71)

Classification of embryonal tumours of the CNS



*new codes in ICD-O-3 2nd revision

New morphology codes: haematological malignancies

malignancy	morphology
Anaplastic large cell lymphoma, ALK negative	9715/3
Erdheim-Chester disease	9749/3
B-lymphoblastic leukaemia/lymphoma, BCR-ABL1–like	9819/3
Acute myeloid leukaemia with mutated NPM1 Acute myeloid leukaemia with biallelic mutation of CEBPA* Acute myeloid leukaemia with mutated RUNX1 Acute myeloid leukaemia with BCR-ABL1	9877/3 9878/3 9879/3 9912/3
Myeloid and lymphoid neoplasm with PCM1-JAK2	9968/3
Myelodysplastic syndrome with ring sideroblasts and multilineage dysplasia	9993/3

* Monoallelic mutation of CEPBA does *not* qualify for 9878/3

Questions?

Please be aware that any question on coding can be submit at the website of the ENCR:
https://www.encr.eu/ask-an-expert

European Commission