

Recommendations for the coding of multiple primary tumours

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Background

Multiple primary tumours (MPTs) are defined as a diagnosis of two or more independent primary tumours in a patient, not being an extension, dissemination or recurrence of another primary tumour.

The occurrence of MPTs has increased due to the increase of long-term survival of patients with cancer, ageing of the population and improvement of diagnostic techniques.

Studies on MPTs could be a starting point for investigating possible shared aetiologies (lifestyle, environmental and/or genetic risk factors) and/or late effects of cancer treatment. In addition, these studies are important for clinical follow-up having implication for screening, prevention strategies and counselling.

The proportion of MPTs in a population-based cancer registries (PBCRs) depends on how long the registry has been in operation and the definition of MPTs.

The current <u>International Rules for Multiple Primary Cancers</u> for reporting incidence and survival will be followed for the purposes of international comparison of cancer incidence, including the updated table <u>Groups of malignant neoplasms considered to be histologically different.</u>

The rules given in this document are for recording MPT, defining the minimum number of tumours to be collected and registered in the European cancer registries.

Urothelial tumours, central nervous system tumours and haematological malignancies are excluded as there are specific ENCR Recommendations for these tumours.

Aims of the Recommendation

The aims of this recommendation are:

- To define a time window for recording MPTs in the same site and with the same histology.
- To define tumour sites (including laterality and 'large' organs) to be considered a single site for recording MPTs, including rules for unspecified sites.
- To define morphology codes to be considered as histologically different for recording MPTs, including rules for unspecified morphologies.
- To guide the MPTs registration for invasive tumours after non-invasive tumours or non-invasive tumours after invasive tumours.





Entering into Force

The new Recommendations for coding of multiple primary tumours is published on the website on ##-##-2025. These recommendations should be applied to all tumours with an incidence date as of 1-1-2026, but may also be applied to earlier dates.





General principles

- These rules are applicable to solid tumours, excluding urothelial tumours (see the recommendations for urothelial tumours) and central nervous system tumours (see the recommendations for CNS tumours)
- Metachronous tumours or tumours at different sites or tumours from different morphology groups should be registered as separate MPTs (unless the behaviour code is different).
- For synchronous tumours not considered MPTs i.e. multiple tumours with the same tumour type at the same site the following rules are valid:
 - if more than one subsite is involved code at CXX.8
 - if more than one specific morphology is present use a combination code, e.g. 8255/3, 8522/3, 8523/3 or 8524/3, or if a combination code is not available a more general code, e.g. register 8070/3 if one tumour is keratinizing squamous cell carcinoma (8071/3) and the other one non-keratinizing squamous cell carcinoma (8072/3)
 - if there is a combination of a specific morphology and an unspecific one the specific one prevails, e.g. a combination of serous adenocarcinoma (8441/3) and adenocarcinoma, NOS (8140/3) should be registered as 8441/3
 - the TNM and the tumour characteristics of the most unfavourable one are registered (e.g. higher grade, largest tumour size, etc.); the characteristics do not necessarily refer to the same tumour
- The following situations are not considered MPTs:
 - A contiguous tumour that involves, invades, or bridges adjacent or connecting sites or subsites
 - o A metastatic tumour
 - A recurrent tumour

Synchronous or metachronous

Two tumours are considered synchronous if the occurrence interval between them is within four months and metachronous tumours if the occurrence interval is greater than four months.

Systemic or multicentric cancers

The following synchronous systemic (or multicentric) cancers potentially involving several different sites or different laterality are only registered once:

- Colorectal cancer in a patient with familial polyposis coli
- Nephroblastoma
- Neuroblastoma





The following systemic (or multicentric) cancers potentially involving several different organs are only registered once in any individual:

- Kaposi sarcoma
- Mesothelioma

Topography

- Table 1 gives an overview of all tumour sites that should be considered as different sites, e.g. C17.0 ('duodenum') and C17.1 ('small intestine') are different sites, while C53 and C54 are the same site ('uterus').
- Different laterality results in a different tumour site for the salivary glands, nasal cavity, middle ear, accessory sinuses, lung, extremities, breast, testis, kidney, renal pelvis, ureter, eye and adrenal gland, but not for the ovary and the fallopian tube. Laterality should be coded as follows:
 - Left 1Right 2Midline 3Bilateral 4Unknown 9

Midline (3) is applicable to tumours of the nasal cavity (C30.0), while bilateral (4) is applicable to synchronous bilateral tumours of the ovary (C56.9) and fallopian tube (C57.0), as well as for synchronous bilateral nephroblastoma and neuroblastoma.

- Subsites are considered as separate sites in a number of 'large' organs, such as colon and skin, as well as for ICD-O subsites representing separate anatomic organs, such as nasal cavity and middle ear.
- If the first three digits of the topography code are the same, overlapping and unspecified topography codes are considered as the same tumour site, e.g. C17.0 and C17.8 as well as C18.0 and C18.9 are considered as the same tumour site.
- Combinations of (mostly ill-defined) sites that are not to be considered as different sites are mentioned in table 2

Table 1: Tumour sites to be considered as different

Tumour site	Topography codes
lip	C00.0-2, C00.6
oral cavity	C00.3-5, C02-C04, C05.0, C06
left salivary gland	C07-C08, left
right salivary gland	C07-C08, right
oropharynx	C01, C05.1-2, C09-C10
nasopharynx	C11
hypopharynx	C12-13
oesophagus	C15
stomach	C16





Tumour site	Topography codes
duodenum	C17.0
small intestine	C17.1-3
appendix	C18.1
right colon	C18.0, C18.2-3
transverse colon	C18.4
left colon	C18.5-C18.7
rectum	C19, C20
anus	C21
liver	C22
gallbladder	C23, C24.4
bile ducts	C24.0-3
pancreas	C25
left nasal cavity	C30.0, left
right nasal cavity	C30.0, right
left middle ear	C30.1, left
right middle ear	C30.1, right
left accessory sinuses	C31, left
right accessory sinuses	C31, right
larynx	C32
trachea	C33
left lung	C34, left
right lung	C34, right
thymus & mediastinum	C37-C38
skin/soft tissue/bone of head & neck	C41.0-1, C44.0-4, C47.0, C49.0, C69.6, C76.0
skin/soft tissue/bone of trunk	C41.2-4, C44.5, C47.3-6, C49.3-6, C76.1-3, C76.7
skin/soft tissue/bone of left upper limb	C40.0-1, C44.6, C47.1, C49.1, C76.4, left
skin/soft tissue/bone of right upper limb	C40.0-1, C44.6, C47.1, C49.1, C76.4, right
skin/soft tissue/bone of left lower limb	C40.2-3, C44.7, C47.2, C49.2, C76.5, left
skin/soft tissue/bone of right lower limb	C40.2-3, C44.7, C47.2, C49.2, C76.5, right
left breast	C50, left
right breast	C50, right
vulva	C51
vagina	C52
uterus	C53-C55
ovary, fallopian tube & peritoneum	C48.1-8, C56, C57.0
placenta	C58
penis	C60
prostate and male urethra	C61 and C68.0
left testis	C62, left
right testis	C62, right
epididymis	C63.0
spermatic cord	C63.1
scrotum	C63.2
seminal vesicle, tunica vaginalis	C63.7
left kidney	C64, left
right kidney	C64, right
left renal pelvis & ureter	C65 and C66, left
epididymis spermatic cord scrotum seminal vesicle, tunica vaginalis left kidney right kidney	C63.0 C63.1 C63.2 C63.7 C64, left C64, right



Tumour site	Topography codes
right renal pelvis & ureter	C65 and C66, right
bladder	C67
female urethra	C68.0
left eye	C69.0-5, left
right eye	C69.0-5, right
meninges	C70
cerebrum	C71.0-4
ventricles	C71.5
cerebellum & brain stem	C71.6-7
spinal cord & cauda equina	C72.0-1
optic chiasm	C72.3, midline
left cranial nerves	C72.2-5, left
right cranial nerves	C72.2-5, right
thyroid gland & parathyroid gland	C73.9, C75.0
left adrenal gland	C74, left
right adrenal gland	C74, right
pituitary gland & craniopharyngeal duct	C75.1-2
pineal gland	C75.3

Table 2: Tumour site combinations of unspecified and specified topography codes to be considered as the same site

Tumour site	Unspecified topography codes	Specified topography codes
oral cavity & pharynx	C14	C00-C13
gastrointestinal tract	C26	C15-C25
respiratory tract	C39	C30-C38
peritoneum	C48.1-2	C16-C26
female genital organs	C57.1-8	C51-C56, C57.0
male genital organs	C63.8-9	C60-C63.7
urinary tract	C68.8-9	C64-C68.1
endocrine glands	C75.8-9	C73-C75.5
ill-defined sites of head & neck	C76.0	C00-C13, C30-C32, C73
ill-defined sites of thorax	C76.1	C15, C33-C39, C50
ill-defined sites of abdomen	C76.2	C16-C26
ill-defined sites of pelvis	C76.3	C51-C68
other/overlapping ill-defined site	s C76.7-8	Any code
lymph nodes	√C77	Any code
unknown primary site	C80	Any code

Example 1

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C16.9	8140/3	15-5-2021, C16.9, 8140/3
2	31-6-2021	C76.2	8140/3	

Example 2





Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C80.9	8070/3	15-5-2021. C34.1. 8070/3
2	31-6-2021	C34.1	8070/3	

Example 3

Tumour	Incidence date	Topography	Morphology	Tumours to be registered
1	15-5-2021	C02.1	8070/3	15-5-2021, C02.1, 8070/3
2	31-6-2021	C09.0	8070/3	31-6-2021, C09.0, 8070/3

Example 4

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C18.0	8140/3	15-5-2021, C18.8, 8140/3
2	31-6-2021	C18.2	8140/3	

Example 5

Tumour	Incidence date	Topography	Morphology	Tumours to be registered
1	15-5-2021	C04.9	8070/3	15-5-2021, C04.9, 8070/3
2	31-6-2021	C10.9	8070/3	31-6-2021, C10.9, 8070/3

Example 6

Tumour	Incidence date	Topography	Morphology	Tumours to be registered
1	15-5-2021	C34.3 R	8070/3	15-5-2021, C34.3 R, 8070/3
2	31-6-2021	C34.1 L	8070/3	31-6-2021, C34.1 L, 8070/3

Example 7

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C09.9	8070/3	15-5-2021. C10.9. 8071/3
2	31-6-2021	C01.9	8071/3	== = ====, = 10.0, 00.1,

Example 8

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C53.1	8070/3	15-5-2021, C53.1, 8070/3
2	31-6-2021	C54.1	8070/3	

Example 9

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C44.6 R	8800/3	15-5-2021. C49.1 R. 8800/3
2	31-6-2021	C49.1 R	8800/3	13 3 1311, 3334

Example 10

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C02.1	8070/3	15-5-2021, C06.9, 8070/3
2	31-6-2021	C04.9	8070/3	

Morphology





Table 3 gives an overview of specified morphology groups that should be considered as different. For example, adenocarcinoma (8140/3; group 3) and large cell neuroendocrine carcinoma (8013/3; group 5) are in different morphology groups and therefore considered as different, while round cell sarcoma (8803/3; group 16) and Ewing sarcoma (9364/3; group 16) are in the same group and therefore not considered as different.

Table 3: Solid tumour types (morphology groups) to be considered as different

Morphology group	Morphology codes
Squamous and transitional cell carcinoma	8051-8086, 8120-8131
2. Basal cell carcinoma	8090-8110
3. Adenocarcinoma	8140-8149, 8160-8163, 8190-8221, 8250-8552, 8570-8576,
	8940-8941, 9110
4. Other specific carcinomas	8023, 8170-8180, 8230-8231, 8560-8562, 8580-8589
5. Neuroendocrine neoplasms	8013, 8041-8045, 8150-8158, 8240-8249
6. Specialized gonadal neoplasms	8590-8671
7. Paraganglioma	8680-8700
8. Melanoma	8720-8790
9. Adipocytic sarcoma	8850-8881
10. (Myo)fibroblastic sarcoma	8810-8827, 8832-8842
11. Vascular & perivascular sarcoma	8710-8711, 9120-9133, 9150-9170
12. Smooth muscle sarcoma	8890-8898
13. Skeletal muscle sarcoma	8900-8921, 8991
14. Stromal sarcoma	8930-8931, 8935-8936
15. Peripheral nerve sheath sarcoma	9540-9580
16. Round cell sarcoma	8803, 9260, 9364-9365
17. Osteosarcoma	9180-9200
18. Chondrosarcoma	9210-9242
19. Odontogenic sarcoma	9270-9342
20. Sarcoma of uncertain differentiation	9040-9045, 9261, 9581
21, Mesothelioma	9050-9055
22. Germ cell tumours	9060-9110
23. Kaposi sarcoma	9140
24. Chordoma	9370-9373

A combination of an unspecified morphology and a specific morphology should be considered as the same tumour type (table 4). For example, non-small cell carcinoma (8046/3) and adenocarcinoma (8140/3) are considered as the same tumour type.

Table 4: Solid tumour type combinations to be considered as the same tumour type

Tumour type	Unspecified morphology codes	Specified morphology codes
Unspecified carcinomas	8010-8012, 8014-8015, 8020-8022,	8013, 8041-8045, 8051-8589, 8940-8941,
	8030-8035	9110
Non-small cell carcinoma	8046	8013, 8045, 8051-8589, 8940-8941, 9110
Unspecified papillary carcinoma	8050	8052, 8130, 8260, 8340-8344, 8347,
		8408, 8450-8463, 8471, 8473, 8503, 8504
Unspecified sarcomas	8800-8802, 8804-8806, 8830, 8990	8803, 8810-8827, 8832-8931, 8935-8936,
		8991, 9040-9044, 9120-9342





Tumour type	Unspecified morphology codes	Specified morphology codes
Mixed neoplasms	8933-8934, 8950-8983, 9000-9030	Any code
Unspecified cancers	8000-8005	Any code

Example 1

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C16.3	8140/3	15-5-2021. C16.3. 8140/3
2	31-6-2021	C76.2	8000/3	15 5 1511, 610.0, 61.0, 6

Example 2

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C16.3	8140/3	15-5-2021, C16.3, 8140/3
2	31-6-2021	C16.9	8000/3	

Example 3

Tumour	Incidence date	Topography	Morphology	Tumours to be registered
1	15-5-2021	C22.1	8160/3	15-5-2021, C22.1, 8160/3
2	31-6-2021	C25.1	8140/3	31-6-2021, C25.1, 8140/3

Example 4

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C34.1 R	8046/3	15-5-2021. C34.1 R. 8551/3
2	31-6-2021	C34.9 R	8551/3	

Example 5

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C49.1	9140/3	15-5-2021. C49.1. 9140/3
2	31-6-2021	C05.1	9140/3	

Example 6

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C49.0	8802/3	15-5-2021. C49.0. 8855/3
2	31-6-2021	C76.0	8855/3	, , , , , , , , , , , , , , , , , , , ,

Example 7

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C73.9	8050/3	15-5-2021. C73.9. 8260/3
2	31-6-2021	C73.9	8260/3	

Tumours with different behaviour codes





Non-invasive (/2) tumour after invasive tumour (/3)

A non-invasive tumour (synchronous or metachronous) after an invasive tumour at the same site and with the same tumour type does not have to be registered.

Invasive (/3) tumour after non-invasive tumour

A synchronous (i.e. within 4 months) invasive tumour after a non-invasive tumour at the same site and with the same tumour type should be registered as one tumour with the incidence date of the non-invasive tumour.

- o if more than one subsite is involved code at CXX.8
- o if more than one specific morphology is present use a combination code, e.g. 8255/3, 8522/3, 8523/3 or 8524/3

A metachronous (i.e. after 4 months or later) invasive tumour after a non-invasive tumour at the same site and with the same tumour type should be registered as a subsequent primary tumour.

Example 1

Tumour	Incidence date Topography		Morphology	Tumour to be registered
1	15-5-2021	C50.3, left	8500/3	15-5-2021, C50.3, 8500/3
2	31-6-2022	C50.5, left	8500/2	

Example 2

Tumour	Incidence date	Topography	Morphology	Tumours to be registered
1	15-5-2021	C50.3, left	8500/2	15-5-2021, C50.3, 8500/2
2	31-6-2022	C50.5, left	8500/3	31-6-2022, C50.5, 8500/3

Difficult situations

Adjacent anatomical structures

Special attention should be paid to tumours with the same tumour type at site combinations that do not necessarily represent the same site, but relate to adjacent anatomical structures and might refer to the same primary tumour, e.g. 'lip' and 'skin of lip' or 'duodenum' and head of pancreas'. MPTs should only be registered if there is clinical evidence that the tumour sites represent separate tumours. The topography codes concerned are in table 5.

Table 5: Tumour sites potentially referring to the same tumour

Site 1	Topography codes	Site 2	Topography codes
lip	C00.0-2, C00.6	skin of lip	C44.0
cardia, GOJ	C16.0, C16.7	(lower) oesophagus,	C15.1-9, C16.1-6
		stomach	





Site 1	Topography codes	Site 2	Topography codes
duodenum	C17.0	bile ducts, pancreas	C24, C25
small intestine	C17.1-3	colon	C18
transverse colon	C18.4	splenic or hepatic flexure	C18.3, C18.5
sigmoid colon	C18.7	rectum, rectosigmoid	C19, C20
anus	C21	rectum	C20
bile ducts	C24.0-3	intrahepatic bile ducts, gall bladder, pancreas	C22.1, C23, C24.4, C25
nasal cavity	C30.0	skin of nose	C44.3
middle ear	C30.1	skin of ear	C44.2
accessory sinuses	C31	nasopharynx, nasal cavity	C11, C30.0
lung	C34	trachea	C33
placenta	C58	uterus	C54, C55
left kidney	C64, left	left renal pelvis	C65, left
right kidney	C64, right	right renal pelvis	C65, right
left eye	C69.0-5, left	skin of left eyelid	C44.1, left
right eye	C69.0-5, right	skin of right eyelid	C44.1, right
adrenal gland	C74	peripheral nerves, retroperitoneum	C47, C48.0

Metastatic or recurrent tumour or MPT

If two tumour lesions could be considered as two MPTs but also as metastatic or recurrent disease, code as metastatic or recurrent disease, unless there is clinical evidence of MPTs.

Example 1

Tumour	Procedure	Date	Site	Morphology	Tumour to be registered
1	Biopsy	15-5-2021	nose	8070/3	15-5-2021, C30.0, 8070/3
2	Resection	30-5-2021	nasal cavity	8070/3	13 3 2021, 630.0, 6070/3

Example 2

Tumour	Procedure	Date	Site	Morphology	Tumour to be registered
1	Biopsy	15-5-2021	trachea	8041/3	15-5-2021. C34.0. 8041/3
2	Biopsy	30-5-2021	left main bronchus	8041/3	13 3 2021, 634.0, 6041/3

Example 3

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C64.9, left	8312/3	15-5-2021, C64.9, 8312/3,
2	31-6-2021	C34.9, right	8010/3	metastatic

Example 4

Tumour	Incidence date	Topography	Morphology	Tumour to be registered
1	15-5-2021	C64.9, left	8312/3	15-5-2021, C64.9, 8312/3
2	31-6-2022	C34.9, right	8010/3	Recurrent tumour





Appendix 1: Working Group Members

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