



NET (neuroendocrine tumour) & NEC (neuroendocrine carcinoma)

Coding issues

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May 2021

Endocrine or neuro-endocrine?

- Endocrine cells release hormones into the blood but do not receive neural input
- Neuro-endocrine cells receive neural input (neurotransmitters from nerve cells) and release hormones into the blood
- NET = tumour of neuro-endocrine cells
- Neuro-endocrine cells can be found everywhere in the body where there is epithelium, so not in the CNS, bone or soft tissues

Hormone production

- As neuro-endocrine tumours can produce (a large variety of) hormones, patients can have all kinds of symptoms related to the hormone production
- 'carcinoid syndrome' (serotonin production)

NET or NEC

- NET=neuro-endocrine tumour
- NEC=neuro-endocrine carcinoma
- Both are cancers with neuro-endocrine characteristics, but they have different behaviour and prognosis
 - NET is relatively indolent
 - NEC is more aggressive
- Hormone production in NEC is rare
- Because of the differences patients receive different treatments
- NET should never be coded as NEC or the other way around

Most common sites of NET/NEC

Lungs (mostly NEC)

Gastro-intestinal tract

- ileum/jejunum
- appendix
- pancreas

Thyroid gland

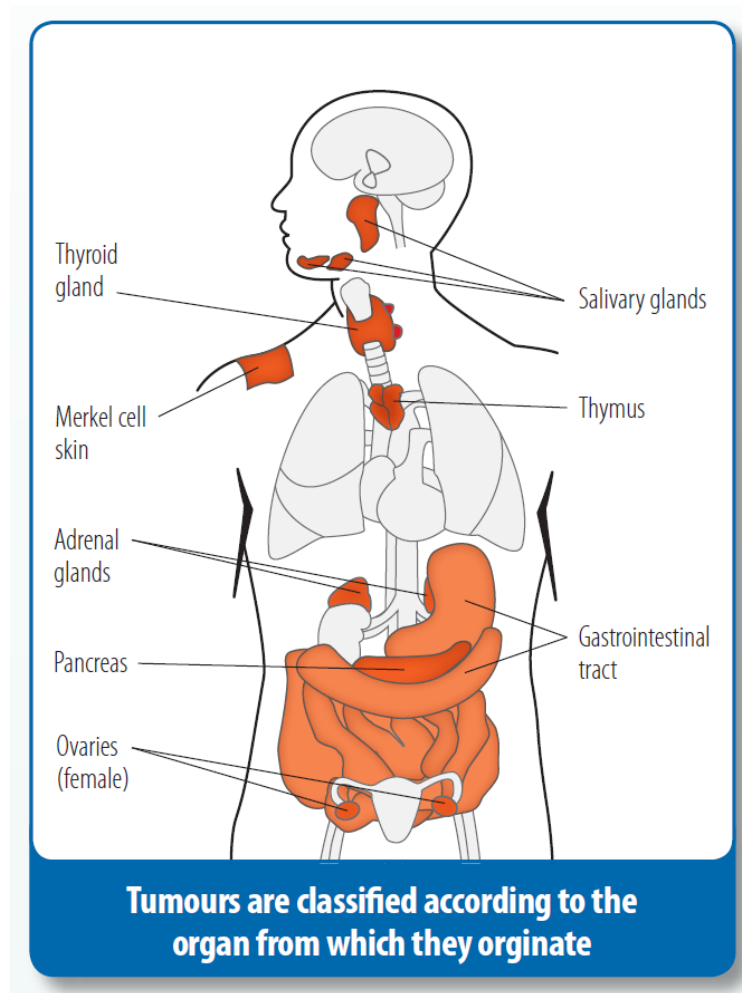
Skin

Salivary glands

Thymus

Adrenal gland (cortex)

Ovaries



G1NET

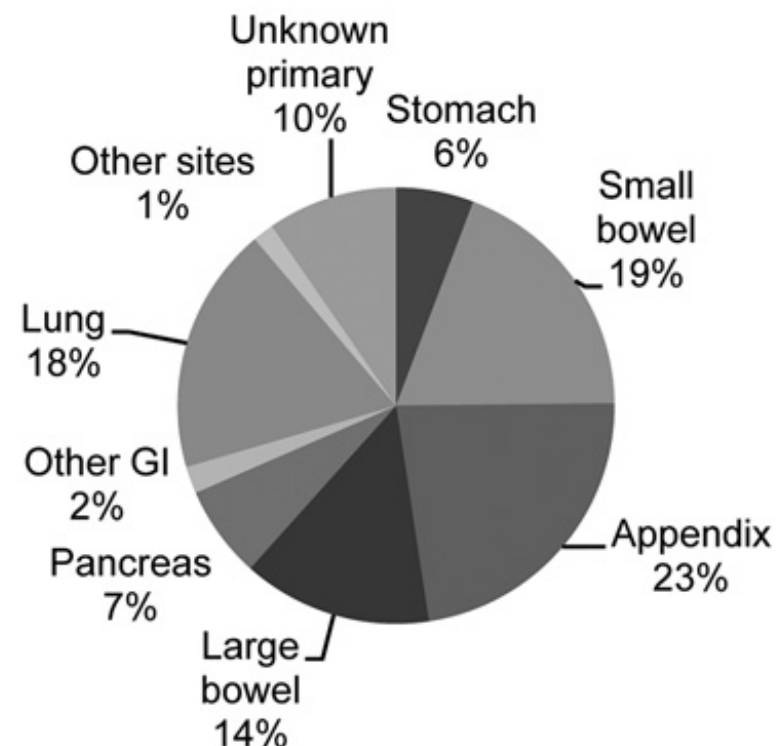
Grade 1

- NET grade 1
- carcinoid
- well differentiated NET
- NEC grade 1

→ Code as M8240/3(1)

G1NET also includes the islet cell tumours of the pancreas

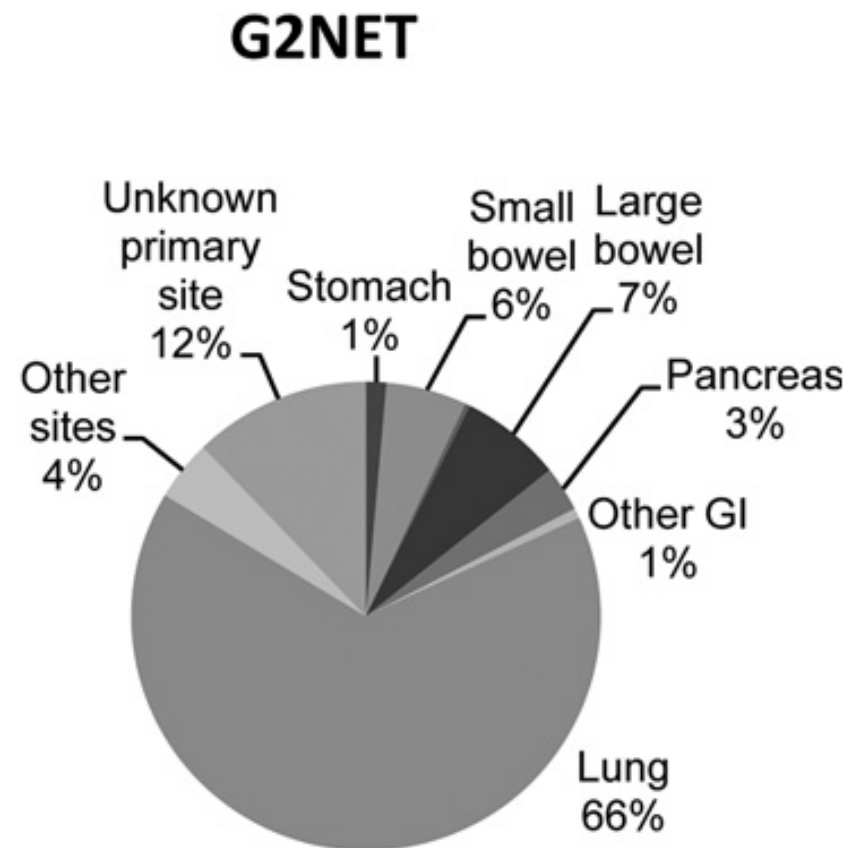
G1NET



G2NET

Grade 2

- NET grade 2
 - atypical carcinoid
 - moderately differentiated NET
 - NEC grade 2
- Code as M8249/3(2)



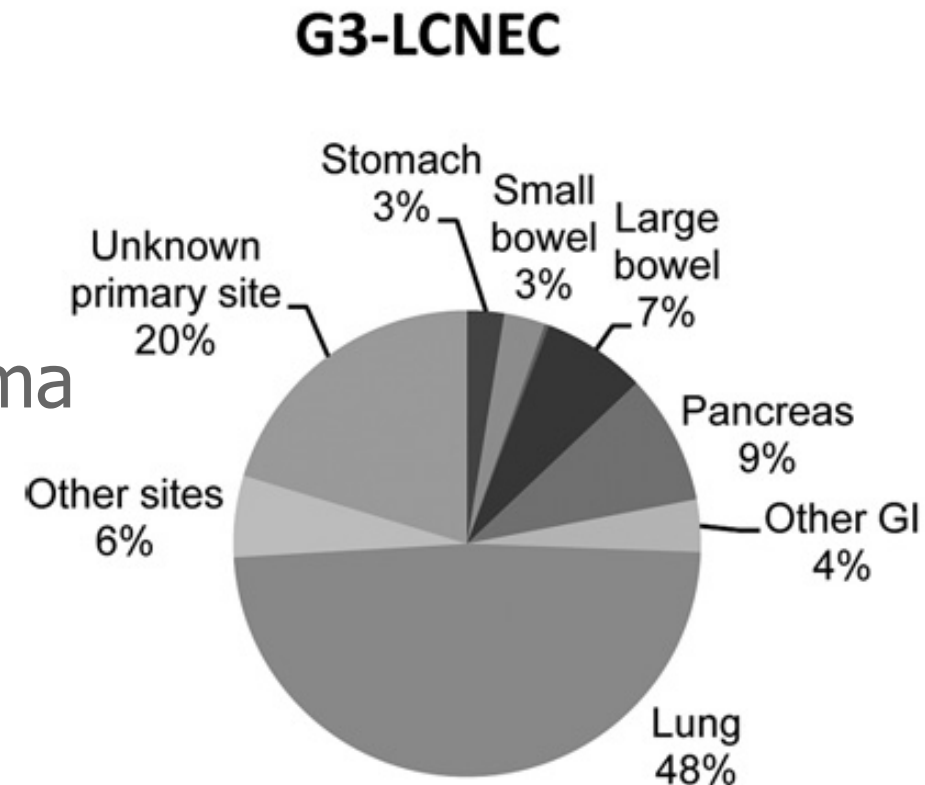
Other NETs

- NET, no grade specified → code as 8240/39
- NET, G3 → code as 8249/33

G3-LCNEC

Grade 3

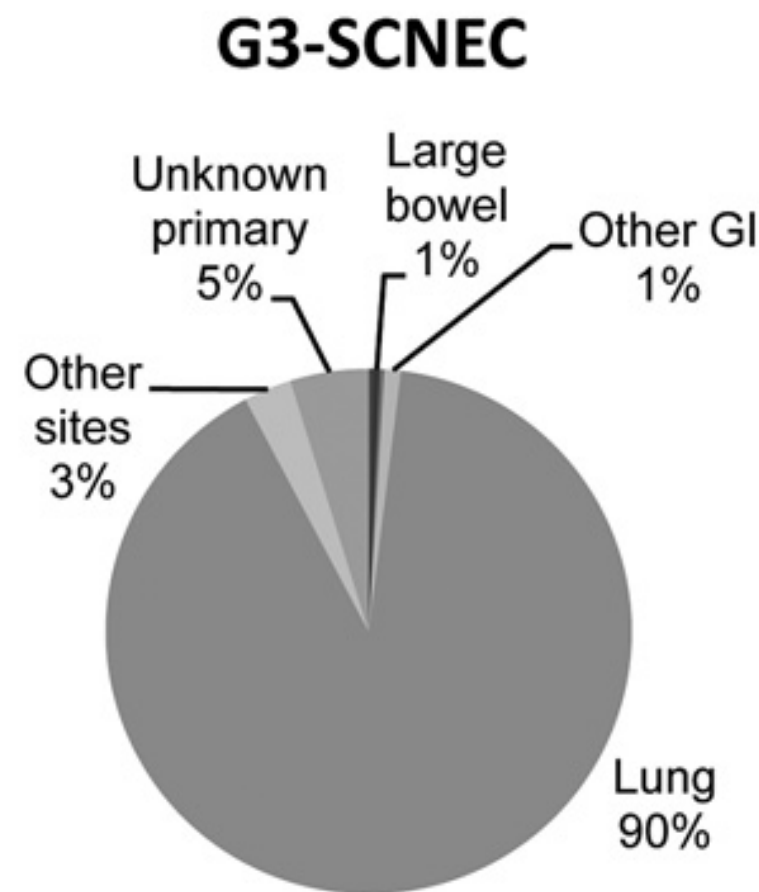
- large cell neuroendocrine carcinoma
 - grade 3 large cell neuroendocrine carcinoma
 - poorly differentiated neuroendocrine carcinoma
 - grade 3 neuroendocrine carcinoma
- Code as M8013/3(3)



G3(4)-SCNEC

Grade 4

- small cell neuroendocrine carcinoma
 - Grade 3 small cell neuroendocrine carcinoma
 - undifferentiated neuroendocrine carcinoma
 - small cell carcinoma
- Code as M8041/3(4)



Other NEC

Neuroendocrine carcinoma, grade unknown → Code as M8246/3

Try to avoid code 8246

Never code NET as 8246 or NEC as 8240

NET/NEC of the skin

NET or NEC (all grades) primary to the skin
→ Code as M8247/3 (Merkel cell carcinoma)

Merkel cell carcinoma is the most aggressive skin cancer!



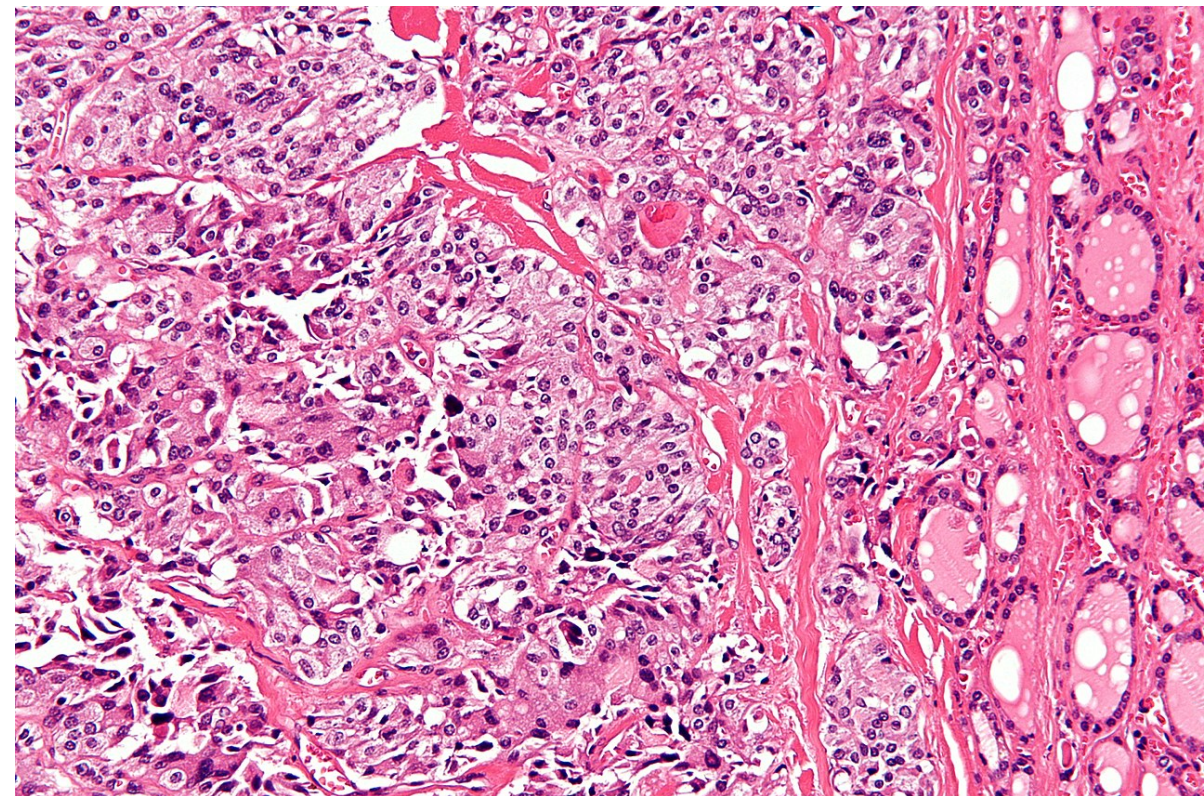
NET of the ovary

- Mature teratoma or dermoid cysts of the ovary contain all kinds of tissues
- One of these tissues may undergo malignant transformation → teratoma/dermoid cyst with malignant transformation (9084/3)
- If the malignancy in the teratoma/dermoid cyst is a NET it is called 'strumal carcinoid' which is coded as 9091/1

NET/NEC of the thyroid

NET or NEC (all grades) primary to the thyroid

→ Code as M8345/3 (Medullary carcinoma of the thyroid)



Functional (=hormone producing) NET

Code to the specific morphology in case of hormone production

- Functioning tumours:
 - insulinoma → 8151
 - glucagonoma → 8152
 - gastrinoma → 8153
 - vipoma → 8155
 - somatostatinoma → 8156
 - serotonin producing tumour → 8241
 - other/ACTH-producing tumour 8158
- Non-functioning tumours (8240 / 8150)

NET/NEC of the pancreas (topography C25.4)

- NET grade 1 (8240) / Islet cell tumour (8150)
- NET grade 2 (8249)
- Large cell neuroendocrine carcinoma (8013)
- Small cell carcinoma (8041)

New morphology terms & codes in 2nd revision of ICD-O-3

Code	Term
8150/3	Pancreatic neuroendocrine tumour, nonfunctioning (C25.4) Pancreatic endocrine tumour, NOS (C25.4)
8158/3	Endocrine tumour, functioning, NOS

All neuroendocrine tumours of the pancreas are considered malignant, unless they are very small (< 0.5 cm) and are non-functioning



EXERCISES



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