UK and Ireland Neuroendocrine Tumour Society

UKINETS bitesize guidance
Management of NETs of the Appendix

Pathological Features*
- NET 1-2cm and G2 or G3
- NET >2cm
- R1 or R2 resection

Yes

Radiological Staging
- CT or MRI to stage
- functional imaging if regional or distant metastases suspected

Distant metastases confirmed: see guidance on advanced midgut NETs

No

- Low G2**
- < 2cm
- no radiological evidence of residual disease

- 1-2cm and R1 and G1/low G2
- no radiological evidence of residual disease

- >2cm
- 1-2cm or R1/2 and high G2/G3
- radiological evidence of residual local/nodal disease

NET MDT consider role of further surgery***

Oncological right hemicolectomy

Discharge

No residual disease

- Nodal metastasis
- High G2 or G3

CT surveillance
- annual for 3 years
- 2 yearly thereafter (until at least 8 years follow up completed)

*Histological diagnosis must be made by histopathologist with expertise in NETs

**Low G2 generally considered with Ki67 index <10%

***ENETs guidelines recommend right hemicolectomy when primary resection is R1

PLEASE REFER TO ADDITIONAL GUIDANCE NOTES BELOW

Preparation date March 2024 - Review date March 2026

For more information, please visit our website: www.ukinets.org

UKINETS bitesize guidance
Management of NETs of the Appendix

Neuroendocrine neoplasms of the appendix are mainly diagnosed incidentally following surgery, most ofter after appendicectomy with a rate of 3-8/1000 appendicectomies.

Simple appendicectomy is adequate treatment for most tumours.

Tumours greater than 2cm in diameter have a great risk of nodal involvement and right hemicolectomy is therefore recommended.

ENETs recommend right hemicolectomy in cases of R1 excision. These guidelines recommend MDT discussion about need for further surgery

For tumours measuring between 1 and 2 cm in diameter, the main risk factor for nodal involvement is a high grade (higher G2 >10%). The presence of deep mesoappendix invasion (>3mm), angioinvasion, perineural invasion, and serosal involvement are of uncertain significance but have been associated with lymph node metastasis in some studies.

Lymph node metastasis has not been shown to impact overall survival at date of writing guidance.

Somatostatin receptor scintigraphy is unlikely to be helpful for surveillance and should be reserved for patients with suspected residual/recurrent disease.

Serum markers such as chromogranin are not useful for surveillance after surgery for aNET.

The frequency, nature and duration of follow up is poorly evidence based, and long term recurrence rates are low. Current guidelines recommend imaging for at least 8 years.

UKINETS bitesize guidance
Management of NETs of the Appendix

References

1. Moertel CG, Weiland LH, Nagorney DM, Dockerty MB. Carcinoid tumour of the appendix: treatment and prognosis. N Eng J Med 1987; 317: 1699-1701
2. Stinner B and Rothmund M. Neuroendocrine tumours (carcinoids) of the appendix. Best Pract Res Clin Gastroenterol 2005; 19: 729-738
3. Brighi N, La Rosa S, Rossi G, et al. Morphological Factors Related to Nodal Metastases in Neuroendocrine Tumors of the Appendix: A Multicentric Retrospective Study. Ann Surg 2018 doi: 10.1097/SLA.0000000000002939
4. Boudreaux JP, Klimstra DS, Hassam MM, et al. The NANETS consensus guideline for the diagnosis and management of neuroendocrine tumours. Well differentiated neuroendocrine tumours of the jejunum, ileum, appendix and cecum. Pancreas 2010; 39: 753-766
5. Pape U-F, Niederle B, Costa F, et al. ENETS consensus guidelines for neuroendocrine neoplasms of the appendix (excluding goblet cell carcinomas). Neuroendocrinology 2016; 103: 144-152
6. Niere I, Smith LL, Thurairasa D, et al. Systematic review and meta-analysis of appendiceal carcinoid tumors in children. Pediatr Blood Cancer 2018; 65: e27069. doi 10.1002/pbc.27069
7. Kaltsas G, Walter T, Knigge U et al. ENETS 2023 guidance paper for appendices neuroendocrine tumours (aNET). J Neuroendocrinol. 2023; 1-14.
8. Pawa N, Clift AK, Omani H et al. Surgical management of patients with neuroendocrine neoplasms of the appendix: appendectomy or more. Neuroendocrinol. 2018; 106: 242-251.
9. Nesti C, Brautigam K, Benavent M et al. Hemicolectomy versus appendectomy for patients with appendiceal NETs 1-2cm in size: a retrospective, Europe wide, pooled cohort study. Lancet Oncol. 2023; 24: 187-94.

Authors: Zahir Soonawalla, Thomas Armstrong
Preparation date March 2024. Review date March 2026