Appendix

Overview / Tests / Follow Up

The Appendix

The appendix can be found near the junction where the large and small bowel meet. It is a small thin tube, approximately 4 inches long. Normally, the appendix sits in the lower right abdomen. It has no clearly identified function in humans.

NENs of the appendix are uncommon and usually only discovered during tests or treatments for other conditions, for example, histology review following appendicectomy.

Appendiceal NENs, less than 1cm, completely removed by surgery, often do not need any further treatment or follow up - they are essentially cured. However, if the tumour is larger, shows evidence of spread, is found at the base of the appendix or the appendix has ‘burst’, you may need further follow up and / or treatment.

For NET, this may include further surgery.

For NEC, this may include further surgery +/- chemotherapy.

NET or Neuroendocrine Tumour is neuroendocrine cancer with well-differentiated cells that usually has a slow to moderate growth rate.

NEC or Neuroendocrine Carcinoma is neuroendocrine cancer with poorly-differentiated cells that grow more rapidly.
Diagnosis for Appendix

nb Most appendiceal NENs are diagnosed AFTER removal or during test for something else

Blood / Urine Tests

- Full blood count
- Liver and kidney function
- CEA, Ca19-9, Ca-125.
- Chromogranin A
- Urinary or serum 5HiAA (serotonin).

Scans

- CT chest & abdomen and / or CT chest &MRI abdomen
- Octreotide (SPECT) or Gallium-Dotatate PET/CT.
- Colonoscopy or CT colonoscopy/enterolysis may be helpful to exclude other bowel tumours.

Pathology

- Differentiation and cellular morphology
- Synaptophysin
- Chromogranin
- CEA.

Treatment for Appendix

For all patients, there are many things to consider in planning treatments. Your treatment will be personalised to you and the type of NEN you have.

Even if you have a diagnosis that sounds the same as another patient, your treatment and follow up plan may be different.

Your care team will discuss your treatment options with you - giving you both written and verbal information - to help you make an informed choice. Together you can agree on the most appropriate treatment for you.

Information about the treatments that are used in NET and NEC can be found in the NPF Handbook - Your Guide to Living with Neuroendocrine Cancer - www.netpatientfoundation.org

There is consensus agreement that all Neuroendocrine Cancer patients should be reviewed by a Specialist Neuroendocrine Cancer MDT.

www.netpatientfoundation.org
Follow-up for Appendix
As per national and international guidelines nb local policy may vary

Following appendicectomy:

- **Less than 1cm, completely resected (R0):**
  Well-differentiated neuroendocrine tumour (WDNET), Grade 1 (Ki67 <2%), lymph node negative with no evidence of further disease
  - NO follow up required beyond routine post-op check.

- **More than 1cm, less than 2cm, completely resected (R0):**
  - WDNET, Grade 1, NO evidence of mesoappendiceal or angio-invasion or further disease
  - A single post-op CT chest/abdomen - if clear, NO further follow up required
  - If there is any evidence of further disease (e.g. enlarged locoregional lymph nodes), a right hemicolectomy and lymph node assessment is recommended.

If further surgery is not undertaken or other risk factors* are present then longer-term follow up is recommended.

Risk factors* include tumour found at base of appendix, mesoappendiceal invasion > 3mm, grade 2 (Ki67 3-20%) or evidence of vascular invasion.

- **More than 2cm:**
  - Grade 1 or 2, with or without angioinvasion or deep mesoappendiceal infiltration and / or lymph node involvement
  - CT chest/abdo/pelvis 6-12 monthly with annual Chromogranin A for a minimum 7 - 8 years.

Following right hemicolectomy:

- If no evidence of residual disease - no lymph nodes or metastases: NO further follow up is required
- Where there is evidence of lymph node and / or distant metastases - follow up as per >2cm
- Biochemistry (CEA, Ca19-9, Ca 125 +/- Chromogranin A) may be used a surrogate markers
- Colorectal surveillance as per national Improving Outcome Guidance (IOG) protocols.
A big part of meeting with your doctors, or specialist nurse, is to make sure you get the information you need to understand what’s happening, so that you can make an informed choice about your care. Asking questions can be difficult, especially if you’re feeling nervous, confused, frightened or struggling to understand what you are being told. You might want to know as much as possible straight away or prefer to take things in small amounts at your own pace.

**Suggestions that may help:**

- Prepare a list of questions that are important to you
- Ask for simple explanations - do not be worried about asking your nurse or doctor to repeat what they have said
- Take someone with you or ask if you can record the conversation. Many mobile phones have a record function or an app you can download
- Ask for a copy of any letters sent to your GP and/or other care team(s)
- If you have a nurse specialist - keep in touch. They can be a great source of information and support for you.

**Example questions:**

- Who can I call if I have any questions? Who is my main point of contact?
- Who will be involved in my care?
- What are the treatment options for me? How might they affect me?
- How often will I need to have scans and tests?
- Are there any flags or warning signs I need to look out for?

Further information about making the most of your consultations can be found in our handbook: www.netpatientfoundation.org

**REFERENCES**


Guidance for Management of the Appendix (Bitesize Guidance) Clinical Practice www.ukinets.org