Optimal care pathway for people with neuroendocrine tumours

Quick reference guide

The optimal care pathways describe the standard of care that should be available to all cancer patients treated in Australia. The pathways support patients and carers, health systems, health professionals and services, and encourage consistent optimal treatment and supportive care at each stage of a patient's journey. Seven key principles underpin the guidance provided in the pathways: patient-centred care; safe and quality care; multidisciplinary care; supportive care; care coordination; communication; and research and clinical trials.

This quick reference guide provides a summary of the *Optimal care pathway for people with neuroendocrine tumours* (NETs).

Please note that not all patients will follow every step of the pathway.

Support: Assess supportive care needs at every step of the pathway and refer to appropriate health professionals or organisations.

Step 1: Prevention and early detection

Prevention

The causes of most NETs are not fully understood, and there is currently no clear prevention strategy. However, when there is a history of hereditary conditions, such as those listed below, the risk is greater, and genetic surveillance and regular reviews need to be in place to detect early asymptomatic cancers.

Risk factors

The risk factors for developing NETs include:

- hereditary conditions such as multiple endocrine neoplasia type 1 (MEN-1), multiple endocrine neoplasia type 2 (MEN-2), von Hippel-Lindau disease (VHL) and phacomatoses (neurocutaneous syndromes)
- genetic disorders associated with multiple tumours (e.g. tuberous sclerosis and neurofibromatosis)
- conditions that affect stomach acid (e.g. pernicious anaemia and chronic atrophic gastritis)
- age – although NETs can occur at any age, there are some age groups that specific NETs can occur in, such as appendiceal NETs in a younger age group.

Early detection

Increased awareness among health professionals is paramount to enable early diagnosis. There is often a prolonged delay in diagnosis because features are non-specific. GPs should have a strong clinical suspicion of patients who present with a combination of symptoms or persistent symptoms listed in Step 2.

People with a family history of hereditary disorders should be referred to a familial cancer service, geneticist or oncologist for genetic screening.

Screening and surveillance recommendations

There is no national screening program for non-hereditary NETs.

Refer to the optimal care pathway for people with NETs for hereditary conditions that may predispose people to forming a NET.

Checklist

- ❑ Referral to a familial cancer service, geneticist or oncologist for genetic screening if there is a known history of hereditary NETs
- ❑ Recent weight changes discussed and the patient's weight recorded
- ❑ Alcohol intake discussed and recorded and support for reducing alcohol consumption offered if appropriate
- ❑ Smoking status discussed and recorded and brief smoking cessation advice offered to smokers
- ❑ Physical activity recorded
- ❑ Referral to a dietitian considered
- ❑ Referral to a physiotherapist or exercise physiologist considered
- ❑ Education on being sun smart considered

Step 2: Presentation, initial investigations and referral

The following signs and symptoms should be investigated:

- abdominal pain
- bloating
- repeated dry flushing on the face and neck
- diarrhoea, even while not eating
- wheezing/bronchoconstriction (asthma-like symptoms)
- episodes of hypotension or palpitations
- unexplained right-sided heart disease
- unexplained weight loss
- fatigue.

Refer to the optimal care pathway for people with NETs for more specific symptoms for the NET location.

Checklist

- ❑ Signs and symptoms recorded
- ❑ Investigation as per suspected site of NET
- ❑ Supportive care needs assessment completed and recorded, and referrals to allied health services actioned as required

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Support: Assess supportive care needs at every step of the pathway and refer to appropriate health professionals or organisations.

Step 2: Presentation, initial investigations and referral continued

Initial investigations include:

- taking of a medical history and a physical examination
- full blood count, B12 and serum iron, LFTs and renal function, thyroid, calcium, cholesterol and CRP
- imaging tests (e.g. ultrasound, CXR, CT scans)
- referral for endoscopy/colonoscopy or bronchoscopy depending on imaging result.

Refer to the optimal care pathway for people with NETs for specific investigations of the NET location.

Referral options

At the referral stage, the patient's GP or other referring doctor should advise the patient about their options for referral, waiting periods, expertise, if there are likely to be out-of-pocket costs and the range of services available. This will enable patients to make an informed choice of specialist and health service.

Communication

The GP's responsibilities include:

- explaining to the patient and/or carer who they are being referred to and why
- supporting the patient and/or carer while waiting for specialist appointments
- informing the patient and/or carer that they can contact:
  - Cancer Council on 13 11 20
  - NET Nurse at NeuroEndocrine Cancer Australia on 1300 287 363.

Checklist continued

- ❑ Patient notified of support services (e.g. Cancer Council and NeuroEndocrine Cancer Australia)
- ❑ Referral options discussed with the patient and/or carer including cost implications.

Timeframe

Where this is a strong suspicion of NETs, investigations should be conducted **within 2 weeks** of the initial GP appointment.

All patients with a suspected or proven NET should be referred to an appropriate specialist **within 1 week** of completing initial investigations.

Step 3: Diagnosis, staging and treatment planning

Diagnosis and staging

- Biochemical markers – measurement of serum chromogranin A may be appropriate. Specific hormonal assessment will depend on symptomology of the primary NET.
- Anatomical (e.g. CT, MRI) and functional imaging (68Ga-DOTATATE PET/CT, 18F-FDG PET) as indicated.
- Biopsy – histopathological diagnosis (grade and differentiation). Biopsies should be reviewed by a pathologist with experience in NETs.

Genetic testing

Approximately 10–15% of all pancreatic neuroendocrine tumors (pNETs) are associated with MEN-1 and up to 80 per cent of patients with MEN-1 will develop pNETs – **refer to the optimal care pathway for people with NETs for more information.**

Find out more about phaeochromocytoma/paraganglioma panel testing, MEN-2 and von Hippel-Lindau disease risk management on the eviQ website <www.eviq.org.au/cancer-genetics/adult>.

Treatment planning

All newly diagnosed patients should be presented at an appropriate neuroendocrine tumour multidisciplinary meeting, with all appropriate investigation results, **within 4 weeks** of diagnosis to develop the patient's management plan. The level of discussion may vary, depending on the patient's clinical and supportive care factors.

Research and clinical trials

Consider enrolment where available and appropriate. Search for a trial <www.australiancancertrials.gov.au>.

Communication

The lead clinician's[1] responsibilities include:

- discussing a timeframe for diagnosis and treatment options with the patient and/or carer
- explaining the role of the multidisciplinary team in treatment planning and ongoing care
- encouraging discussion about the diagnosis, prognosis, advance care planning and palliative care while clarifying the patient's wishes, needs, beliefs and expectations, and their ability to comprehend the communication
- providing appropriate information and referral to support services as required
- communicating with the patient's GP about the diagnosis, treatment plan and recommendations from multidisciplinary meetings.

Checklist

- ❑ Diagnosis confirmed
- ❑ Full histology obtained
- ❑ Performance status and comorbidities measured and recorded
- ❑ Patient discussed at an MDM and decisions provided to the patient and/or carer
- ❑ Clinical trial enrolment considered
- ❑ Supportive care needs assessment completed and recorded and referrals to allied health services actioned as required
- ❑ Patient referred to support services (e.g. Cancer Council and NeuroEndocrine Cancer Australia) as required
- ❑ Treatment costs discussed with the patient and/or carer

Timeframe

Diagnostic investigations should be completed **within 2 weeks** of the initial specialist appointment.

1 Lead clinician – the clinician who is responsible for managing patient care.
The lead clinician may change over time depending on the stage of the care pathway and where care is being provided.

Support: Assess supportive care needs at every step of the pathway and refer to appropriate health professionals or organisations.

Step 4: Treatment

Establish intent of treatment

- Curative
- Anti-cancer therapy to improve quality of life and/or longevity without expectation of cure
- Symptom palliation

Surgery: The surgical procedure undertaken will depend on the location(s) of the NET and treatment intent. Surgery may be curative, de-bulking for symptom control or palliative.

No treatment / active surveillance (watch and wait): No treatment may be suitable for some NET patients if the NET is not causing symptoms or problems, there is little disease, the disease is stable or the tumour is low grade (G1).

Localised radiation therapy: Patients with oligometastatic disease, a dominant or critically strategic site of progression or highly symptomatic metastases may benefit from radiation therapy.

Peptide receptor radionuclide therapy (PRRT): Patients with metastatic disease who have progressed following first-line somatostatin analogues (SSAs) may benefit from systemic radionuclide therapy (or PRRT).

Systemic therapy:

- SSAs are the most common first-line treatment of G1/G2 NETs. They have antisecretory and antiproliferative effects.
- Oral targeted therapy: Molecular targeted therapy for mTOR and multitargeted pathways.
- Chemotherapy is an option for NET patients with pancreatic, bronchial or high-grade (G2/G3) NETs. It can be used in combination with PRRT and adjuvant to surgery.

Liver-directed therapy: Targeted therapy with radiation or chemotherapy directly to liver metastases may be indicated for some patients.

Immunotherapy is investigational for NETs and is an emerging therapy.

Clinical trials: Many emerging therapies are only available by participating in clinical trials.

Palliative care:

Early referral to palliative care can improve quality of life and in some cases survival. Referral should be based on need, not prognosis. For more, visit the Palliative Care Australia website <www.palliativecare.org.au>.

Communication

The lead clinician and team's responsibilities include:

- discussing treatment options with the patient and/or carer including the intent of treatment as well as risks and benefits
- discussing advance care planning with the patient and/or carer where appropriate
- communicating the treatment plan to the patient's GP
- helping patients to find appropriate support for programs where appropriate to improve treatment outcomes.

Checklist

- ❑ Intent of treatment established
- ❑ Risks and benefits of treatments discussed with the patient and/or carer
- ❑ Treatment plan discussed with the patient and/or carer
- ❑ Treatment plan provided to the patient's GP
- ❑ Treating specialist has adequate qualifications, experience and expertise
- ❑ Supportive care needs assessment completed and recorded, and referrals to allied health services actioned as required
- ❑ Early referral to palliative care considered
- ❑ Advance care planning discussed with the patient and/or carer

Timeframe

Surgery: Timeframe for surgery will be based on investigation and staging of the NET and surgery intent.

Localised radiation therapy: Treatment should start **as soon as possible** for symptomatic patients.

PRRT: When PRRT is necessary, treatment should start **as soon as possible**.

Systemic therapy: When active treatment is necessary, treatment should start **within 4 weeks**.

Liver directed therapy: When active treatment is necessary, treatment should start **within 4 weeks**.

Support: Assess supportive care needs at every step of the pathway and refer to appropriate health professionals or organisations.

Step 5: Care after initial treatment and recovery

Provide a treatment and follow-up summary to the patient, carer and GP outlining:

- the diagnosis, including tests performed and results
- tumour characteristics
- treatment received (types and date)
- current toxicities (severity, management and expected outcomes)
- interventions and treatment plans from other health professionals
- potential long-term and late effects of treatment and care of these
- supportive care services provided
- a follow-up schedule, including tests required and timing
- contact information for key healthcare providers who can offer support for lifestyle modification
- a process for rapid re-entry to medical services for suspected recurrence.

Communication

The lead clinician's responsibilities include:

- explaining the treatment summary and follow-up care plan to the patient and/or carer
- informing the patient and/or carer about secondary prevention and healthy living
- discussing the follow-up care plan with the patient's GP.

Checklist

- ❑ Treatment and follow-up summary provided to the patient and/or carer and the patient's GP
- ❑ Supportive care needs assessment completed and recorded and referrals to allied health services actioned as required
- ❑ Patient-reported outcome measures recorded

Step 6: Managing recurrent, residual or metastatic disease

Detection

Most residual or recurrent disease will be detected via routine follow-up or by the patient presenting with symptoms.

Treatment

Evaluate each patient for whether referral to the original multidisciplinary team is appropriate. Treatment will depend on the location and extent of disease, previous management and the patient's preferences.

Advance care planning

Advance care planning is important for all patients but especially those with advanced disease. It allows them to plan for their future health and personal care by thinking about their values and preferences. This can guide future treatment if the patient is unable to speak for themselves.

Survivorship and palliative care

Survivorship and palliative care should be addressed and offered early. Early referral to palliative care can improve quality of life and in some cases survival. Referral should be based on need, not prognosis.

Communication

The lead clinician and team's responsibilities include:

- explaining the treatment intent, likely outcomes and side effects to the patient and/or carer and the patient's GP.

Checklist

- ❑ Treatment intent, likely outcomes and side effects explained to the patient and/or carer and the patient's GP
- ❑ Supportive care needs assessment completed and recorded and referrals to allied health services actioned as required
- ❑ Advance care planning discussed with the patient and/or carer
- ❑ Patient referred to palliative care if appropriate
- ❑ Routine follow-up visits scheduled

Step 7: End-of-life care

Palliative care

Consider a referral to palliative care. Ensure an advance care directive is in place.

Communication

The lead clinician's responsibilities include:

- being open about the prognosis and discussing palliative care options with the patient
- establishing transition plans to ensure the patient's needs and goals are considered in the appropriate environment.

Checklist

- ❑ Supportive care needs assessment completed and recorded, and referrals to allied health services actioned as required
- ❑ Patient referred to palliative care
- ❑ Advance care directive in place

Visit our guides to best cancer care webpage <www.cancercareguides.org.au> for consumer guides. Visit our OCP webpage <www.cancer.org.au/OCP> for the optimal care pathway and instructions on how to import them into your GP software.

Published in June 2022.