

Optimal care pathway for people with neuroendocrine tumours

FIRST EDITION



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Endorsed by



Australian Government
Cancer Australia



Statement of acknowledgement

We acknowledge the Traditional Owners of Country throughout Australia and their continuing connection to the land, sea and community. We pay our respects to them and their cultures and to Elders past, present and emerging.

This work is available from the Cancer Council website <www.cancer.org.au/OCP>.

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Welcome and introduction

On behalf of the optimal care pathways team and contributors, welcome to the first edition of the optimal care pathway for people with neuroendocrine tumours.

Each patient affected with cancer may have a different experience depending on their background and the type of tumour. Neuroendocrine tumours exemplify this variability, as different neuroendocrine tumours arise from different organs and may behave in very different ways. This variability makes it all the more important to outline the principles of high-quality care for neuroendocrine tumours, which remain similar across different grades of neuroendocrine tumours, and if followed are likely to result in the best possible outcomes for patients, families and carers.

We are proud to be part of the excellent Australian healthcare system, but we also recognise that pathways for cancer patients may be complex. These often span multiple health professionals for neuroendocrine tumours (including surgeons, radiation oncologists, medical oncologists, endocrinologists, nuclear medicine physicians, general practitioners, allied health professionals, nurses and managers of cancer services), extending across public and private systems. This complexity in accessing care has led us to develop the optimal care pathways. These pathways describe an integrated model of care incorporating both the technical aspects of care and the delivery of such care. They provide a national standard for the high-quality cancer care that all Australians should expect.

The optimal care pathways are not intended to replace clinical guidelines. The choice of treatment for an individual is a professional responsibility based on patient factors and preferences, practice guidelines and emerging data. This optimal care pathway was developed in 2021-22, at a time when the global COVID-19 pandemic was challenging the Australian healthcare sector in an unprecedented way. The pandemic led to rapid practice change, including greater uptake of telehealth. Where appropriate, learnings have informed review of the pathways. We hope that this optimal care pathway will optimise the care for patients affected by neuroendocrine tumours.

The optimal care pathways should be read and understood by all health professionals involved in cancer care, as well as trainees in all relevant disciplines. We also recommend them to patients and their carers, in the hope that they may guide discussions with their healthcare team. There are specific versions of the optimal care pathways for consumers, the Guides to best cancer care, available in different languages.

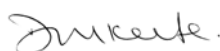
The optimal care pathways are endorsed by Cancer Australia and have Australia-wide acceptance and government support.

We would like to reiterate our thanks to everyone involved in developing this optimal care pathway – the Expert Working Group and additional reviewers. It is our sincere hope that it will point to the patient-centred nature of the cancer experience, and consequently help smooth the difficult road that patients and carers walk with a cancer diagnosis.



Dr David Chan

Chair, Optimal Care Pathway for people with
Neuroendocrine Tumours Expert Working Group



Professor Dorothy Keefe PSM MD

Chief Executive Officer,
Cancer Australia

Summary

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

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.

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

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 pheochromocytoma/paranglioma 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's¹ 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.

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**.

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.

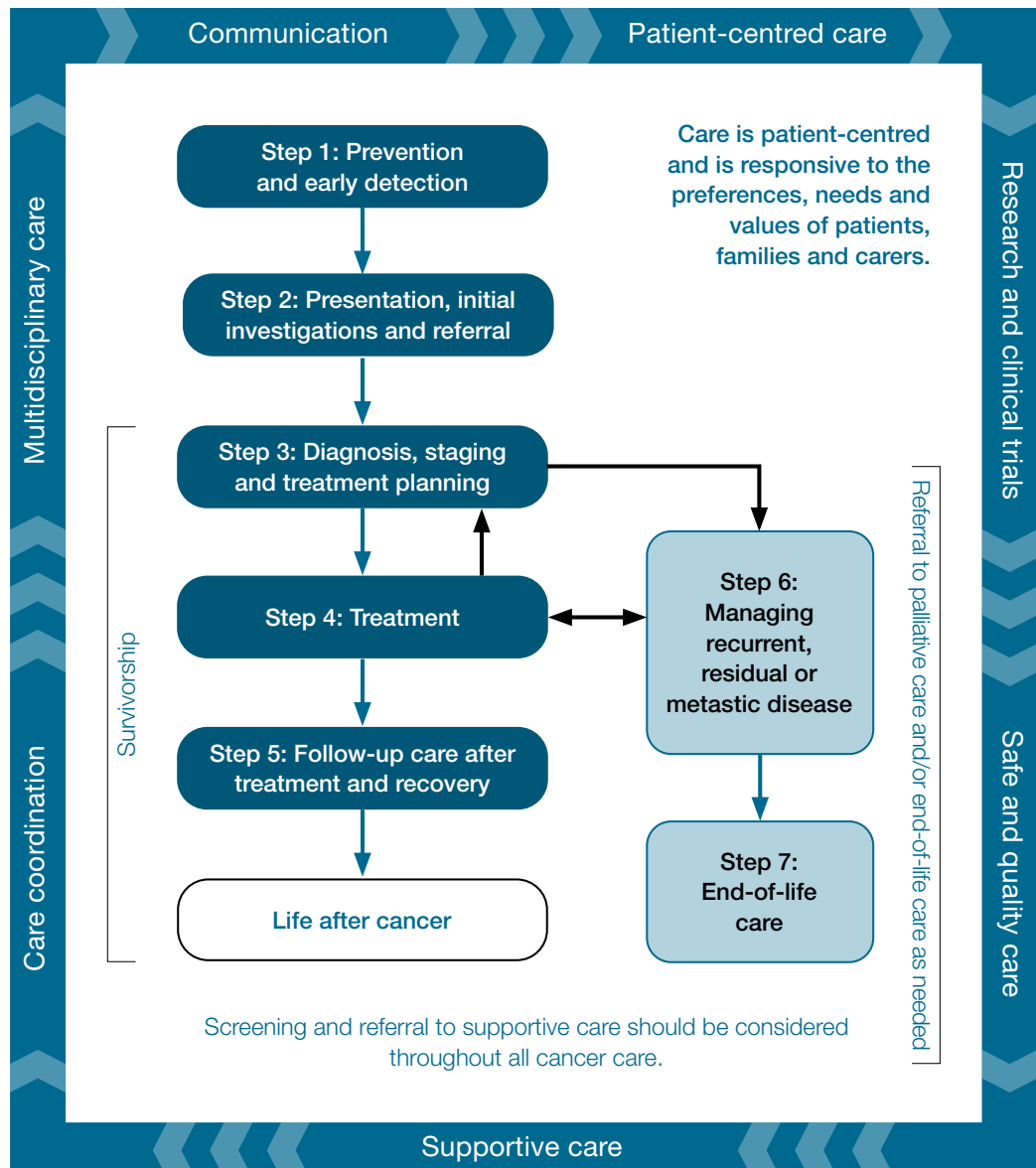
Intent of the optimal care pathways

Optimal care pathways map seven key steps in cancer care. Each of these steps outlines nationally agreed best practice for the best level of care. While the seven steps appear in a linear model, in practice, patient care does not always occur in this way but depends on the particular situation (e.g. the type of cancer, when and how the cancer is diagnosed, prognosis, management, the patient's decisions and their physiological response to treatment).

The principles underpinning optimal care pathways always put patients at the centre of care throughout their experience and prompt the healthcare system to deliver coordinated care.

The optimal care pathways do not constitute medical advice or replace clinical judgement, and they refer to clinical guidelines and other resources where appropriate.

Figure 1: The optimal care pathway



Optimal care pathway resources

There are three resources for each pathway: an optimal care pathway, a quick reference guide for health professionals and a guide to best cancer care for patients, carers and families.

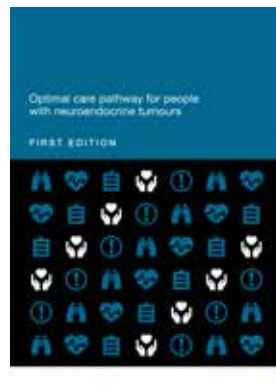
Optimal care pathways

This optimal care pathway is designed for **health professionals and health services**. However, **patients and carers** may find useful information in this version to help understand the processes their treating health professionals are following.

This resource aims to:

- assist health professionals to provide optimal care and support to patients with cancer, their families and carers
- provide optimal timeframes for delivering evidence-based care
- emphasise the importance of communication and collaboration between health providers and people affected by cancer
- assist and inform new health professionals or trainees who are entering the cancer care workforce
- provide value to health systems to identify gaps in current cancer services, bring about quality improvement initiatives and improve how services are planned and coordinated. Adherence to the pathways should be measured wherever possible.

Visit the Cancer Council website <www.cancer.org.au/OCP> to view the optimal care pathways.



Quick reference guides

The quick reference guides are for **health professionals and health services**. They provide a summary of each optimal care pathway for health professionals and patients.

The quick reference guides include:

- optimal timeframes within which tests or procedures should be completed
- checklists with indicators related to recommendations in the optimal care pathway.

Visit the Cancer Council website <www.cancer.org.au/OCP> to view the quick reference guide for this optimal care pathway.



Guides to best cancer care

The guides to best cancer care are consumer resources that help patients understand the optimal cancer care that should be provided at each step. Carers, family and friends may also find the guides helpful.

The guides to best cancer care:

- include optimal timeframes within which tests or procedures should be completed
- include prompt questions to support patients to understand what might happen at each step of their cancer journey and to consider what questions to ask
- provide information to help patients and carers communicate with health professionals
- are available in several languages.

Visit the Cancer Council's website <www.cancercareguides.org.au> to view the guides to best cancer care.



Optimal care pathway for Aboriginal and Torres Strait Islander people with cancer

The *Optimal care pathway for Aboriginal and Torres Strait Islander people with cancer* provides a tool to help reduce disparities and improve outcomes and experiences for Aboriginal and Torres Strait Islander people with cancer. This resource can be used in conjunction with the optimal care pathway for each cancer type.

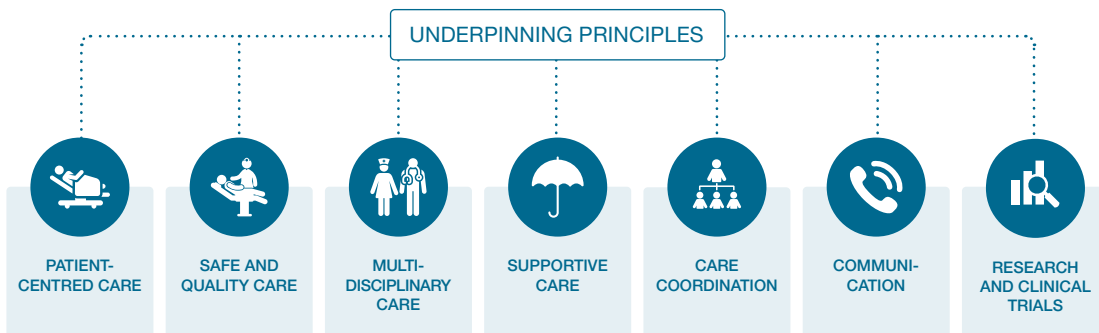
Visit the Cancer Australia website <<https://www.canceraustralia.gov.au/affected-cancer/atsi/resources-health>> to view the optimal care pathway for Aboriginal and Torres Strait Islander people.



Principles of the optimal care pathway

The seven principles of care define appropriate and supportive cancer care that is the right of all patients and the right of those caring for and connected with them.

Figure 2: The seven principles underpinning the optimal care pathway



Principle 1: Patient-centred care



Patient-centred care informs and involves patients in their care and respects and responds to the preferences, needs and values of patients, families and carers.

A patient-centred focus increases the experience and satisfaction of patients, their families and carers, and staff, as well as safety and cost-effectiveness (ACSQHC 2019a).

Patient-centred care means:

- patients are informed and involved in decisions about their cancer and the treatment, post-treatment and recovery program ahead
- patients, their families and carers are provided with access to appropriate and accessible health information
- respect for the cultural and religious beliefs of patients and their families is demonstrated when discussing the diagnosis of cancer
- active communication is used to engage patients, their families and carers in the care process – an essential step for patients to be informed
- care processes are mutually beneficial for patients and providers
- special needs are addressed – for example, the needs of people with disabilities or mental health issues.

Informed choice and consent

An informed patient has greater confidence and competence to manage their cancer journey.

Health professionals are responsible for enabling patients to make informed choices according to their preferences, needs and values. Patients should be provided with:

- individualised and timely information and guidance about their treatment
- details of their care, including the advantages and disadvantages of each treatment, the associated potential side effects, the likely outcomes on their performance status (how well a patient is able to carry out activities of daily life) and subsequently their quality of life and any financial implications, at each stage of the pathway (ACSQHC 2020).

Health professionals have a legal responsibility to obtain consent for all procedures from either the patient or their substitute decision-maker if they are not deemed competent.

Referral choices and informed financial consent

Patients have the right to receive the information they need to be able to make an informed decision on where to be referred for treatment. Treating specialists and practitioners should clearly explain the costs or how to find out the costs of services, tests and treatment options upfront to avoid consumers experiencing 'bill shock'.

At the time of referral, the patient's general practitioner or other referring doctor should discuss the different 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. Referral decisions influence the care patients receive along the pathway and the direct and indirect costs they and their carers may incur. Different referrals have different costs:

- referral to a public hospital, which may involve some costs
- initial referral to a private specialist with associated costs, with the option of ongoing treatment in a public hospital at any time
- referral to a patient's choice of practitioner for immediate and ongoing private hospital management with associated costs.

Patients should be made aware that even though public hospital health care is 'free' to all Australian citizens and most permanent residents of Australia, there are still associated direct costs such as:

- over-the-counter medication and prescriptions
- wound dressings
- travel costs
- parking fees
- tests that are not covered by Medicare.

A cancer diagnosis and treatment may affect a patient's or carer's income. This is an indirect cost associated with cancer. Social work support is essential to help patients and their families deal with this issue. Patients should be advised not to undergo private care with significant out-of-pocket expenses if financially constrained. Specialists in private practice need to explain costs at the start of each new treatment to acknowledge the cumulative out-of-pocket expenses that patients can incur.

For more information on informed financial consent see Cancer Council's 'Standard for informed financial consent' <<https://www.cancer.org.au/health-professionals/resources/informed-financial-consent>>.

Financial counselling services can provide advice on dealing with financial difficulties. These services can be accessed publicly (via social workers at hospitals, financial counsellors at neighbourhood houses or rural financial aid), privately or through cancer support services such as local charity groups or social work services.

For practical and financial assistance, patients may consider Cancer Council's financial services <<https://www.cancer.org.au/about-cancer/patient-support/practical-and-financial-assistance.html>>.

Shared care

Shared care between a cancer specialist and primary care health professional is delivered in two or more settings by two or more professionals. The primary care provider is usually a general practitioner but can include nurses and allied health practitioners. Shared care can be delivered throughout the care pathway including during treatment, follow-up care, survivorship care and end-of-life care.

Shared care offers several advantages to patients, including the potential for treatment closer to home and more efficient care with less duplication and greater coordination. Evidence comparing shared care and specialised care indicates equivalence in outcomes including recurrence rate, cancer survival and quality of life (Cancer Research in Primary Care 2016).

Telehealth can enable efficient shared care and should be explored for all patients. Patients in some rural or remote locations may access specialists via Medicare Benefit Scheme funded telehealth consultations. General practitioners working in rural or remote locations should be aware of specialist multidisciplinary teams with facilities to reduce the travel burden and costs for patients.

Principle 2: Safe and quality care



Hospitals and health professionals are responsible for providing safe and quality care.

Health professionals need to have appropriate training and experience to undertake treatment for neuroendocrine tumours (NETs). Patients should be reviewed at a cancer centre with expertise in NETs.

Safe and high-quality care is care provided by appropriately trained and credentialed health professionals who undertake regular quality reviews of their performance, contribute to regular audits of their care and are actively involved in continuing professional development. Hospitals and clinics must have the equipment, staff numbers, policies and procedures in place to support safe and high-quality care for cancer patients. Patients should be offered the safest options for care, which may include using telehealth (Cancer Australia 2020).

Hospital quality committees should ensure all health care is informed by evidence, and health professionals and health service managers (including executives) have a responsibility to evaluate and monitor their practice. Optimal care pathways provide a framework to help evaluate and monitor practice over time. Services should be routinely collecting relevant minimum datasets to support benchmarking, quality care and service improvement. Hospital committees and health professional peak bodies should be auditing this process (ACSQHC 2017, 2020).

The Australian Council on Health Standards <<https://www.achs.org.au/>> has created a set of indicators that helps hospitals conform to appropriate standards.

Data collection and data linkage play a pivotal role in informing care and improving outcomes for people with NETs. Data linkage facilitates a person-centred, evidence-based approach to understanding the health and wellbeing of patients (CVDL 2022). There are several important purposes of data linkage such as measuring patient outcomes and understanding the needs and clinical pathways of patients to design more effective services (ACSQHC 2011; Safer Care Victoria 2022; WHO 2020).

Patient-reported experience and outcome measures

Patient-reported experience measures (PREMs) and patient-reported outcome measures (PROMs) should be incorporated into routine cancer care.

PREMs are used to obtain patients' views and observations on aspects of healthcare services they have received (AIHW 2018). Patient experience data is collected for specific services and then relayed to service providers to instigate improvements in patient services (ACSQHC 2019b).

The Australian Hospital Patient Experience Question Set (AHPEQS) is a tool used to assess patient experiences of treatment and care in a private or public hospital. AHPEQS helps to improve the safety and quality of health care by allowing organisations to understand the patient's perspective (ACSQHC 2019b; AIHW 2018).

PROMs measure aspects of a person's health status such as symptoms, quality of life and needs and are collected directly from patients either online, via a smartphone or through paper-based means.

Collecting PROMs, and then instigating an appropriate clinical response, has been shown to prolong survival, reduce health system use and improve patients' quality of life. While there are many sets of PROMs questions that are relevant to any cancer patient, specific questions can be tailored to particular cancer types, populations or different phases of cancer care.

Principle 3: Multidisciplinary care



Multidisciplinary care is an integrated team approach that involves all relevant health professionals discussing all relevant treatment options and making joint recommendations about treatment and supportive care plans, taking into account the personal preferences of patients.

Multidisciplinary care improves patient outcomes. Cancer Australia's 'Principles of multidisciplinary care' provides a flexible definition, allowing services to vary implementation according to cancer type and the service location. The principles stipulate:

- a team approach that involves core disciplines that are integral to providing good care, including general practice, with input from other specialties as required
- communication among team members about treatment planning and plans for follow-up
- access to the full therapeutic range for all patients, regardless of geographical remoteness or size of institution
- care delivery in accordance with nationally agreed standards
- patient involvement in decisions about their care (Cancer Australia 2019a).

In addition to these principles, treatment teams should consider clinical trial participation for all eligible patients.

Multidisciplinary meetings, often called MDMs, should be based on the principles outlined above.

For more information on the principles of multidisciplinary care and the benefits of adopting a multidisciplinary approach, see Cancer Australia's 'Principles of multidisciplinary care' <www.canceraustralia.gov.au/clinical-best-practice/multidisciplinary-care/all-about-multidisciplinary-care/principles-multidisciplinary-care>.

Principle 4: Supportive care



Supportive care is a vital part of any cancer treatment program. Supportive care deals with issues that emerge for patients, families and carers from the effects of the cancer diagnosis and its treatment. It is made up of all the services, information and resources patients may need to meet their physical, psychological, social, information and spiritual needs from the time of diagnosis.

Supportive care may be 'patient-defined' and based on unmet needs. It is a core component of evidence-based clinical care and its benefits are well established. All cancer patients and their carers should be formally supported and have access to understandable, relevant information about the medical, practical and emotional aspects of the cancer and its treatment (Fitch 2008). The wishes and needs of the patient, their family and their carers should determine the level of support provided. Supportive care is a standard or routine aspect of cancer care and the treatment team should make patients aware of this.

Supportive care should begin from the time of diagnosis and continue throughout the cancer pathway.

For health professionals, supportive care involves:

- screening and assessing patients and families for their supportive care needs
- providing patients with access to a range of multidisciplinary support services, groups and therapies designed to assist them to live with cancer and its treatment and optimise recovery
- optimising referral pathways to community support organisations (cancer-related non-government, not-for-profit and charities) that provide services to cancer survivors – these address many of the care-navigation, psychosocial and information needs of cancer survivors and those affected by cancer (Australian Cancer Survivorship Centre 2019)
- being aware of and delivering culturally appropriate care.

All members of the multidisciplinary team have a role in providing supportive care along the care pathway, with special attention at transition points.

Supportive care involves routinely and systematically assessing patients to determine their needs. Health professionals can use a variety of validated screening tools for this task (see box on page 13). Clinical review and individual assessment are still required to ensure all patient concerns are identified.



More information

For information and resources on supportive care visit:

- NeuroEndocrine Cancer Australia website <www.neuroendocrine.org.au>
- Healthdirect website <www.healthdirect.gov.au/neuroendocrine-cancers#resources-support>
- WeCan website <www.wecan.org.au>.

Validated screening tools

- National Comprehensive Cancer Network Distress Thermometer and Problem Checklist
- Supportive Care Needs Assessment Tool for Indigenous People (SCNAT-IP)
- Edmonton Symptom Assessment Tool (ESAT) – Neuroendocrine Tumours.

Key review points

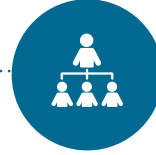
The treatment team should assess patients for supportive care needs at these key stages:

- initial presentation or diagnosis (first three months)
- the beginning of treatment or a new phase of treatment
- change in prognosis
- if a patient is found to have a germline genetic mutation predisposing to cancer
- end of treatment
- throughout survivorship
- diagnosis of recurrence
- change in or development of new symptoms
- palliative care
- end-of-life care
- other time points based on clinical judgement.

The team also needs to decide whether the patient requires ongoing referral to supportive care services. Access to services can be through general practice–led chronic disease management plans, team care arrangements and mental health plans. Community support services also have a role to play.

See Appendices A, B and C for more information on supportive care and the specific needs of people that may arise.

Principle 5: Care coordination



Care coordination is the responsibility of every professional, both clinical and non-clinical, who works with patients, their families and carers.

Seamless care coordination is essential for patients to successfully navigate the complex health system. Care coordination is a comprehensive approach to achieving continuity of care for patients. It aims to ensure care is delivered in a systematic, connected and timely way that promotes efficiency and reduces the risk of duplication and over-servicing to meet the medical and personal needs of patients.

Care coordination includes:

- proactive and timely communication with patients, their families and carers
- treatment plans, survivorship care plans and/or advance care directives
- coordinated appointments to ensure timely diagnosis, treatment and survivorship care
- appropriate tests and results being available to the treating team so treatment decisions can be made
- medical records being available to all members of the treating team and at scheduled appointments
- translation or interpreter services arranged if the patient/carer is from a non-English speaking background or has difficulty communicating due to a physical disability
- practical support such as transport, accommodation, advance care planning and financial support
- referral and access to supportive care
- access to clinical trials
- access to telehealth for people in rural and remote areas and for managing vulnerable patients.

Care coordination brings together different health professionals, teams and health services. It also encompasses MDMs, multidisciplinary assessment clinics, supportive care screening and assessment, referral practices, data collection, common protocols, information for patients and individual clinical treatment.

Care coordination should cross the acute and primary care interface and should aim to achieve consistency of care through clear communication, linkages and collaborative integrated care planning.

Care coordination can be facilitated through electronic health record management such as My Health Record. My Health Record is a secure online database that helps with data collection and care coordination (My Health Record 2019).

Formal care coordination through appointed care coordinators plays an important role in managing and supporting patients through the health system. The availability of dedicated care coordinators varies across states and territories according to the complexity of care required and local service capacity and resourcing.

Principle 6: Communication



Everyone employed in the healthcare system is responsible for ensuring the communication needs of patients, their families and carers are met.

Good and open communication is a key principle of care for cancer patients. This includes communication between oncology and primary care health professionals and with patients. General practitioners should be involved in care from the point of diagnosis, and patients should be encouraged to maintain a relationship with their general practitioner through all stages of cancer care. Communication should be regular and timely.

Attendance of a family member or carer at clinical appointments is beneficial for many patients, as the family member or carer can provide informational and emotional support. General practitioners and clinicians should encourage and support the involvement of family members and carers by providing an inclusive and supportive consultation environment (Laidsaar-Powell et al. 2018a). Laidsaar-Powell et al. provide evidence-based guidance on how to support family member or carer involvement in consultations (Laidsaar-Powell et al. 2018a, 2018b).

Every person with cancer will have different communication needs, including cultural and language differences. When anyone involved in treatment communicates with patients, they should be truthful and transparent but aware of cultural and psychological sensitivities. In communicating with patients, healthcare providers should undertake to:

- empower patients to be active in treatment discussions
- use professionally trained interpreters if required – for example, when communicating with people from culturally diverse backgrounds whose primary spoken language is not English and for people with a hearing impairment (visit the Translating and Interpreting Services website <www.tisnational.gov.au> for more information on interpreter and language services)
- use culturally sensitive and appropriate forums of communication for people from culturally diverse backgrounds and Aboriginal and Torres Strait Islander people, as appropriate
- provide appropriate information for people from culturally diverse backgrounds
- provide information on community-based supportive care services and resources to patients and their families and carer
- identify the patient's substitute treatment decision-maker to ensure they are involved in relevant discussions
- ensure patients, their families or their carers have the opportunity to ask questions
- seek consent before conveying information between health professionals or healthcare teams or with family and carers
- be respectful if a patient seeks a second opinion from another health professional
- ensure patients do not have to convey information between areas of care (it is the provider's and healthcare system's responsibility to transfer information between areas of care)
- communicate in plain language (avoiding complex medical terms and jargon)
- ensure information is communicated at a level relevant to the patient's health literacy and that of their families and carers (ACSQHC 2020)

- use tools, diagrams and aids as appropriate (Gilligan et al. 2017)
- ensure the patient is aware of how to access electronic patient information, where appropriate
- allow enough time for communication, especially when conveying complex or sensitive information such as an initial diagnosis
- check the patient's and/or their family or carer's understanding by asking the patient and/or their family or carer to say in their own words what has been conveyed.

Healthcare providers should also consider offering patients a question prompt list before a consultation and recordings or written summaries of their consultations afterwards. Question prompt lists are effective in improving communication and the psychological and cognitive outcomes of cancer patients. Recordings or summaries of key consultations improve patients' recall of information and satisfaction (Hack et al. 2012). Written care plans, treatment summaries, survivorship care plans and advance care directives are effective records and communication tools.

Communication skills training programs that use role-play to develop skills and observe patient interactions to provide feedback, should be available to health professionals at every level of practice (Gilligan et al. 2017).

Communication skills training programs and resources can be found on the following websites:

- Australian Commission on Safety and Quality in Healthcare, Communicating for safety resource portal <<https://c4sportal.safetyandquality.gov.au>>
- state and territory Cancer Councils <www.cancer.org.au/about-us/state-and-territory-councils/> for the relevant council
- eviQ <<https://education.eviq.org.au>>
- VITAL talk <www.vitaltalk.org>.

Telehealth has become an increasingly acceptable alternative to face-to-face consultations. When using telehealth, the team must consider what is best for the patient, including the patient's preferences of the patient. A face-to-face consultation should be the first option, if it is safe, when delivering critical diagnosis information, a change in therapy or prescribing intensive treatment. If this is not an option, a video consultation should be considered, and the patient should be encouraged to have a support person with them to assist (Cancer Australia 2020).

Principle 7: Research and clinical trials



Research and clinical trials play an important role in establishing the efficacy and safety of diagnostic, prognostic and therapeutic interventions, as well as establishing the role of psychological, supportive care and palliative care interventions (Sjoquist & Zalberg 2013).

Clinical trials are the foundation for improved cancer outcomes, allowing new treatments to be tested and offering patients access to potentially more effective therapies than otherwise available to them.

Clinical trials are available for multiple types of cancer and may be a valuable option for people with rare, difficult-to-treat conditions for which there may be limited evidence about how the condition is best treated or managed (Australian Clinical Trials 2015).

Treating specialists and multidisciplinary teams should be aware of or search for clinical trials that may be suitable for their patients. Specialists should be willing to refer appropriate patients to other treating centres to participate in research or clinical trials at any stage of the care pathway and be willing to discuss the pros and cons of participating in such trials. Any member of the multidisciplinary team can encourage cross-referral between clinical trials centres. Possible ineligibility to participate in a clinical trial should be discussed with the patient. Acknowledge disappointment and offer support in this instance.

Health services should strive to implement policies and procedures that facilitate equitable access to clinical trials for all patients, including culturally diverse patients, regional patients and those from Aboriginal or Torres Strait Islander communities.

The use of telehealth technology, such as the Australasian Tele-trial Model, hopes to improve access to trials for patients being treated in rural and regional areas (COSA 2016).

Australian Cancer Trials is a national clinical trials database. It provides information on the latest clinical trials in cancer care, including trials that are recruiting new participants. Search for a trial <www.australiancancertrials.gov.au> via its website.

Education and training

Research and clinical trials provide an opportunity to educate health professionals who are in training. Cancer centres may be affiliated with teaching hospitals, universities or research groups to promote higher education or to develop the academic workforce, leading to more sustainable practice. Specialists should be encouraged to take up and retain active membership to professional societies and organisations that can assist with professional development opportunities.

Summary – optimal timeframes

Evidence-based guidelines, where they exist, should inform timeframes. Treatment teams need to recognise that shorter timeframes for appropriate consultations and treatment can promote a better experience for patients. Three steps in the pathway specify timeframes for care (Figure 3). They are designed to help patients understand the timeframes in which they can expect to be assessed and treated, and to help health services plan care delivery in accordance with expert-informed time parameters to meet the expectation of patients. These timeframes are based on expert advice from the Neuroendocrine Tumours Working Group.

Figure 3: Timeframes for care

Step in pathway	Care point	Timeframe
Presentation, initial investigations and referral	Signs and symptoms	Presenting symptoms should be promptly and clinically triaged by a general practitioner
	Initial investigations initiated by GP	Where this is a strong suspicion of neuroendocrine tumours, investigations should be conducted within 2 weeks of the initial GP appointment
	Referral to specialist	All patients with a suspected or proven neuroendocrine tumour should be referred to an appropriate specialist within 1 week of completing initial investigations
Diagnosis, staging and treatment planning	Diagnosis and staging	Diagnostic investigations should be completed within 2 weeks of the initial specialist appointment
	Multidisciplinary team meeting and treatment planning	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
Treatment	Surgery	Timeframe for surgery will be based on investigation and staging of the neuroendocrine tumour and surgery intent
	Localised radiation therapy	Treatment should start as soon as possible for symptomatic patients
	Systemic radiation therapy (peptide receptor radionuclide therapy [PRRT])	When PRRT is necessary (for patients who have sufficient expression of somatostatin receptors that have progressed on somatostatin analogues or where initial PRRT is considered appropriate), treatment should start as soon as possible
	Systemic therapy	When active treatment is considered necessary, treatment should start within 4 weeks of the treatment decision
	Liver directed therapy	When active treatment is considered necessary, treatment should start within 4 weeks of the treatment decision

Optimal care pathway

Seven steps of the optimal care pathway

- Step 1: Prevention and early detection
- Step 2: Presentation, initial investigations and referral
- Step 3: Diagnosis, staging and treatment planning
- Step 4: Treatment
- Step 5: Care after initial treatment and recovery
- Step 6: Managing recurrent, residual or metastatic disease
- Step 7: End-of-life care

Neuroendocrine neoplasms (NENs) are a family of tumours defined by specific morphological features. They are separated into neuroendocrine tumours (NETs) and neuroendocrine carcinomas (NECs) which, by definition, are high-grade. The general term to refer to NENs was NETs until very recently and it is still known by this name to most non-NEN specialists; therefore, NETs will be used as the preferred term for this optimal care pathway.

The incidence of NETs has historically been thought to be very low and consequently they were categorised as a 'rare cancer' (NECA 2021). This is no longer the case, with more than 5,000 new patients estimated to be diagnosed this year in Australia (Cancer Australia 2021). In 2021 it was estimated that 3.4 per cent of all new cancer diagnoses will be neuroendocrine cancers, and their incidence now exceeds cancers such as bladder, liver, ovarian, brain and kidney cancers (AIHW 2021).

The increase in the incidence of NETs may partly be attributed to greater awareness of the disease and the development of improved imaging techniques. In 2017, there were 4,228 new cases of NETs diagnosed in Australia compared with more than 5,000 estimated in 2021 (AIHW 2021). Both the incidence of NETs and the survival rates are increasing (Dasari et al. 2017). Earlier diagnosis, improved diagnostic procedures and optimal treatments are the keys to improved survival rates and ongoing symptom management and improved quality of life.

More than 10,000 Australians diagnosed between 2012 and 2016 currently live with NETs, and more than 22,000 people are still living who had been diagnosed with NETs in the 35 years from 1982 to 2016 (AIHW 2021). NETs are the second most prevalent gastrointestinal malignancy after colorectal (bowel) cancer (NECA 2021).

According to the latest Australian Institute of Health and Welfare (AIHW) figures, in 2021, the average Australian is estimated to have a one in 56 chance of being diagnosed with a NET by the age of 85 (AIHW 2021). Despite the surprisingly high occurrence and high morbidity rates, NETs remain under-represented in local cancer research.

Survival rates vary significantly depending on the grade and location of the primary tumour. However, the overall survival for NETs is almost always better than for carcinomas arising at the same site. For example, pancreatic NETs have a 69 per cent five-year survival compared with 6.7 per cent for the more common pancreatic adenocarcinoma (AIHW 2021).

NETs can occur in people of all ages, and affect men and women equally. The most common sites for NETs are the stomach, lung and pancreas (Michael et al. 2021).

This optimal care pathway covers gastrointestinal NETs, lung NETs (known as carcinoid tumours; not small cell lung cancer), thymic NETs, thyroid NETs (known as medullary carcinoma), paragangliomas and pheochromocytomas. Both high-grade and low-grade NETs will be considered in this pathway.

Step 1: Prevention and early detection

This step outlines recommendations for the prevention and early detection of NETs.

Evidence shows that not smoking, avoiding or limiting alcohol intake, eating a healthy diet, maintaining a healthy body weight, being physically active, being sun smart and avoiding exposure to oncoviruses or carcinogens may help reduce general cancer risk (Cancer Council Australia 2018).

1.1 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 the risk is greater, and genetic screening and regular reviews need to be in place to detect early asymptomatic cancer (refer to section 1.3).

1.2 Risk factors

The risk factors for developing NETs include (Cancer Centre 2021; Roswell Park 2021; Department of Health and Ageing 2013):

- hereditary conditions such as multiple endocrine neoplasia type 1 (MEN-1), multiple endocrine neoplasia type 2 (MEN-2), von Hippel-Lindau disease (VHL), phacomatoses (neurocutaneous syndromes)
- genetic disorders associated with multiple tumours – for example, tuberous sclerosis and neurofibromatosis
- conditions that affect stomach acid, such as 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.

1.3 Surveillance for patients with hereditary NETs

Some people have hereditary conditions that may predispose them to forming a NET. These people should be surveilled for syndrome-specific diagnosis and monitored annually. Hereditary conditions include (Rogel Cancer Centre University of Michigan 2021):

- multiple endocrine neoplasia (MEN)
 - MEN-1 – at risk of developing pituitary adenomas (PitNETs), pancreatic neuroendocrine tumours (pNETs) and hyperparathyroidism
 - MEN type 2A (RET gene mutation) – at risk of developing medullary thyroid cancer, hyperparathyroidism and pheochromocytoma, noting that some families will only express medullary thyroid cancer
 - MEN type 2B (also known as MEN-3) (RET) – at risk of developing medullary thyroid cancer at a very young age, pheochromocytoma and mucosal neuromas of the lips, tongue and eyelids
 - MEN type 4 (CDKN1B gene mutation) – at risk of developing pituitary tumours, pancreatic NETs and hyperparathyroidism
 - MEN type 5 (MAX) – at risk of developing pheochromocytomas and pituitary NETs
- VHL– at risk of developing pheochromocytomas, central nervous system and retinal haemangioblastomas, inner ear tumours, kidney tumours and pancreatic NETs
- succinate dehydrogenase gene complex (SDHA/B/C/D) mutations / paraganglioma syndrome – increased risk of pheochromocytomas, paragangliomas, stomach gastrointestinal stromal tumours, kidney cancers and PitNETs
- fumarate hydratase mutations, which cause hereditary leiomyomatosis and renal cell carcinoma syndrome characterised by renal carcinomas, benign smooth muscle tumours of the skin and uterus and, occasionally, pheochromocytoma and paragangliomas
- TMEM127 mutations – increased risk of pheochromocytomas, paragangliomas and renal cell cancers.

For more information visit:

- pheochromocytoma/paraganglioma panel testing <<https://www.eviq.org.au/cancer-genetics/adult/genetic-testing-using-cancer-gene-panels/3601-paraganglioma-pheochromocytoma-panel-testi>>
- VHL disease risk management <<https://www.eviq.org.au/cancer-genetics/adult/risk-management/397-vhl-von-hippel-lindau-disease-risk-managem>>.

1.4 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. General practitioners should have a strong clinical suspicion of patients who present with a combination of symptoms, or persistent symptoms (see section 2.1 for symptoms).

1.4.1 Screening recommendations

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

Step 2: Presentation, initial investigations and referral

This step outlines the process for the general practitioner to initiate the right investigations and refer to the appropriate specialist in a timely manner. The types of investigations the general practitioner undertakes will depend on many factors, including access to diagnostic tests, the availability of medical specialists and patient preferences.

2.1 Signs and symptoms

The following signs and symptoms for NETs are not specific, but where persistent or suspicious further investigation could be warranted:

- 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.

For gastrointestinal NETs, more specific symptoms for the NET location are:

- watery diarrhoea
- cramping
- intermittent abdominal pain
- flushing
- asthma-like wheezing
- bowel obstruction
- flushing, diarrhoea and dyspnoea – classic triad symptoms are seen in less than 20 per cent of cases.

For pancreatic NETs, more specific symptoms for the NET location are:

- epigastric or back pain
- peptic ulcer disease
- diarrhoea
- intermittent hypoglycemic episodes (low blood sugar)
- diabetes
- rash.

For lung NETs, more specific symptoms for the NET location are:

- wheezing
- cough
- dyspnoea
- haemoptysis
- recurrent chest infections/pneumonia.

For paragangliomas and pheochromocytomas, more specific symptoms for the NET location are:

- hypertension, often paroxysmal
- headache
- heavy sweating for no known reason
- a strong, fast or irregular heartbeat
- tremor
- pallor.

The presence of multiple signs and symptoms indicates an increased risk of NETs.

2.1.1 Timeframe for general practitioner consultation

Presenting symptoms should be promptly and clinically triaged by a general practitioner.

2.2 Assessments by the general practitioner

General practitioner examinations and investigations should include:

- taking of a medical history and a physical examination
- full blood count, B12 and serum iron, liver function tests (LFTs) and renal function, thyroid function, calcium, cholesterol and C-reactive protein (CRP)
- imaging tests, such as ultrasound, chest x-ray (CXR), computed tomography (CT) scans
- referral for endoscopy/colonoscopy or bronchoscopy depending on imaging result.

Tumour markers – for example, chromogranin A and 24-hour urinary 5HIAA – should be used with caution in the pre-diagnosis stage given the number of common false positives (e.g. chromogranin A may be falsely elevated in the context of proton pump inhibitor usage or renal failure).

See table 1 for specific investigations for the NET location.

Table 1: Specific investigations for the NET location

Gastroenteropancreatic NET	Lung NET	Paraganglioma / Pheochromocytoma
Full blood count LFTs Renal function Calcium/vitamin D CRP B12 and serum iron Ultrasound CXR CT Endoscopy/colonoscopy Chromogranin A (optional as can have false-positives as stated above)	Full blood count LFTs Renal function Calcium/vitamin D CRP Ultrasound CXR CT Bronchoscopy Chromogranin A (optional as can have false-positives as stated above)	Full blood count LFTs Renal function Calcium/vitamin D Plasma metanephrine, normetanephrine and 3-methoxytyramine or Urinary metanephrine/ normetanephrines (catecholamines) CT MRI

2.2.1 Timeframe for completing investigations

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

Investigations could be sequential. The appropriate sequence of investigations may vary. When in doubt, formal specialist consultation is advised.

2.3 Initial referral

If the cancer diagnosis is confirmed or the results are inconsistent or indeterminate, the general practitioner must refer the patient to an appropriate specialist (e.g. medical oncologist, gastroenterologist or respiratory physician) to make the diagnosis.

Patients should be enabled to make informed decisions about their choice of specialist and health service. General practitioners should make referrals in consultation with the patient after considering the clinical care needed, cost implications (see referral options and informed financial consent on page 9), waiting periods, location and facilities, including discussing the patient's preference for health care through the public or the private system.

Referral for suspected or diagnosed NETs should include the following essential information to accurately triage and categorise the level of clinical urgency:

- important psychosocial history and relevant medical history
- family history, current symptoms, medications and allergies
- results of current clinical investigations (imaging and pathology reports)
- results of all prior relevant investigations
- notification if an interpreter service is required.

Many services will reject incomplete referrals, so it is important that referrals comply with all relevant health service criteria.

If access is via online referral, a lack of a hard copy should not delay referral.

The specialist should provide timely communication to the general practitioner about the consultation and should notify the general practitioner if the patient does not attend appointments.

Aboriginal and Torres Strait Islander patients will need a culturally appropriate referral. To view the optimal care pathway for Aboriginal and Torres Strait Islander people with cancer and the corresponding quick reference guide, visit the Cancer Australia website <<https://www.canceraustralia.gov.au/affected-cancer/atsi/resources-health>>. Download the consumer resources – *Checking for cancer* and *Cancer* from the Cancer Australia website <<https://www.canceraustralia.gov.au/affected-cancer/atsi/resources-people>>.

2.3.1 Timeframe for referring to a specialist

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

2.4 Support and communication

2.4.1 Supportive care

The patient's general practitioner should consider an individualised supportive care assessment where appropriate to identify the needs of an individual, their carer and family. Refer to appropriate support services as required. See validated screening tools mentioned in Principle 4 'Supportive care'.

A number of specific needs may arise for patients at this time:

- assistance for dealing with the emotional distress and/or anger of dealing with a potential cancer diagnosis, anxiety/depression, interpersonal problems and adjustment difficulties
- management of physical symptoms such as diarrhoea
- monitoring needs for appropriate dietary support such as anti-diarrhoeal medication
- encouragement and support to increase levels of exercise (Cormie et al. 2018; Hayes et al. 2019).

For more information refer to the National Institute for Health and Care Excellence 2015 guidelines, *Suspected cancer: recognition and referral* <www.nice.org.uk/guidance/ng12/chapter/Recommendations-on-patient-support-safety-netting-and-the-diagnostic-process>.

For additional information on supportive care and needs that may arise for different population groups, see Appendices A, B and C.

2.4.2 Communication with patients, carers and families

The general practitioner is responsible for:

- providing patients with information that clearly describes to whom they are being referred, the reason for referral and the expected timeframes for appointments
- requesting that patients notify them if the specialist has not been in contact within the expected timeframe
- considering referral options for patients living rurally or remotely
- supporting the patient while waiting for the specialist appointment. NeuroEndocrine Cancer Australia NET Nurse support line 1300 287 363 or support online <<https://neuroendocrine.org.au/net-nurse/>> and Cancer Council nurses (via 13 11 20) are available to act as a point of information and reassurance during the anxious period of awaiting further diagnostic information.



More information

Refer to Principle 6 'Communication' for communication skills training programs and resources.

Step 3: Diagnosis, staging and treatment planning

Step 3 outlines the process for confirming the diagnosis and stage of cancer and for planning subsequent treatment. The guiding principle is that interaction between appropriate multidisciplinary team members should determine the treatment plan.

3.1 Specialist diagnostic work-up

The treatment team, after taking a thorough medical history and making a thorough medical examination of the patient, should undertake the following investigations under the guidance of a specialist.

- Biochemical markers – measurement of serum chromogranin A may be appropriate. Specific hormonal assessment will be 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.

3.1.1 Timeframe for completing investigations

All diagnostic investigations should be completed **within two weeks** of the initial specialist appointment.

3.1.2 Genetic testing (family risk)

Hereditary susceptibility should be considered in any patient with a NET. The risk of a heritable cause is particularly high for people with medullary thyroid cancer, pheochromocytoma and paraganglioma, for whom genetic counselling and testing should be routinely performed.

MEN-1 should be considered in those with a gastrinoma or multifocal gastroenteropancreatic neuroendocrine cancer (GEP-NET) at any age; patients with a GEP-NET before age 40 years; and patients with a bronchial or thymic NET at any age.

In MEN, approximately 10–15 per cent of all pNETs are associated with MEN-1, and up to 80 per cent of patients with MEN-1 will develop pNETs (O’Shea & Druce 2017).

For the exact tumours related to each MEN subtype, refer to section 1.3.

Other potential genetic causes include mutations in VHL, SDHA/B/C/D, fumarate hydratase and TMEM127 – refer to section 1.3 for more information.

Other rare genetic causes of pheochromocytoma and paraganglioma include KIF1B, MDH2, GOT2, DLST and SLC25A11.

Anyone diagnosed with cancer should have a detailed personal and family cancer history taken. Consult relevant guidelines <<https://www.eviq.org.au/cancer-genetics/referral-guidelines>> to determine if referral to a familial cancer service is appropriate.

A familial cancer service assessment can determine if genetic testing is appropriate. Genetic testing is likely to be offered when there is at least a 10 per cent chance of finding a causative ‘gene error’ (pathogenic gene variant; previously called a mutation). Usually testing begins with a variant search in a person who has had cancer (a diagnostic genetic test). If a pathogenic gene variant is identified, variant-specific testing is available to relatives to see if they have or have not inherited the familial gene variant (predictive genetic testing).

Medicare funds some genetic tests via a Medicare Benefits Schedule (MBS) item number but most are not. Depending on the personal and family history, the relevant state health system may fund public sector genetic testing.

Pre-test counselling and informed consent is required before any genetic testing. In some states the treating team can offer 'mainstream' diagnostic genetic testing, after which referral is made to a familial cancer service if a pathogenic gene variant is identified. The familial cancer service can provide risk management advice, facilitate family risk notification and arrange predictive genetic testing for the family.

Visit the Centre for Genetics Education website <www.genetics.edu.au/individuals-and-families/cancer-in-the-family> for basic information about cancer in a family.

For detailed information and referral guidelines for NETs risk assessment and consideration of genetic testing, see *When should genetic testing be performed in patients with neuroendocrine tumours* <<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5849652/>>.

Find more information on MEN-1 genetic testing <<https://www.eviq.org.au/cancer-genetics/adult/genetic-testing-for-heritable-pathogenic-variants/1428-men1-and-cdkn1b-genetic-testing>>, risk management <<https://www.eviq.org.au/cancer-genetics/adult/risk-management/192-men1-risk-management#evidence-for-risk-management-guidelines>> and lifetime risk <<https://www.eviq.org.au/cancer-genetics/adult/risk-management/192-men1-risk-management#lifetime-risk-of-cancer-tumour>>.

For more information on MEN-2, visit the eviQ website <<https://www.eviq.org.au/cancer-genetics/adult/genetic-testing-for-heritable-pathogenic-variants/1429-ret-men2-genetic-testing>>.

The succinate dehydrogenase (SDH) complex genes are a family of genes (SDHA, SDHB, SDHC, SDHD and SDHAF2). For more information visit the eviQ website <<https://www.eviq.org.au/cancer-genetics/adult/genetic-testing-using-cancer-gene-panels/3601-paraganglioma-phaeochromocytoma-panel-testing>>.

3.1.3 Pharmacogenetics

Pharmacogenetics describes how individual genetic differences can lead to differences in the way certain medicines interact with the body. These interactions can affect the effectiveness of medications and any side effects. Applying pharmacogenetics to treatment planning may help patients to be prescribed the most appropriate treatment at the optimal dose from the beginning of treatment (NHMRC 2013).

3.2 Grading and Staging

Grading and staging is a critical element in treatment planning and should be clearly documented in the patient's medical record.

The pathological biopsy should be reported or reviewed by a pathologist with expertise in NETs. NETs are graded pathologically based on the mitotic count (expressed as mitoses per 2mm²), the Ki-67 proliferative index and the presence of necrosis. Different cut-offs of Ki-67 index and mitotic rate are used to grade tumours of different sites, and for some sites the presence of necrosis increases the grade.

Other critical elements in the pathological reporting of NETs are:

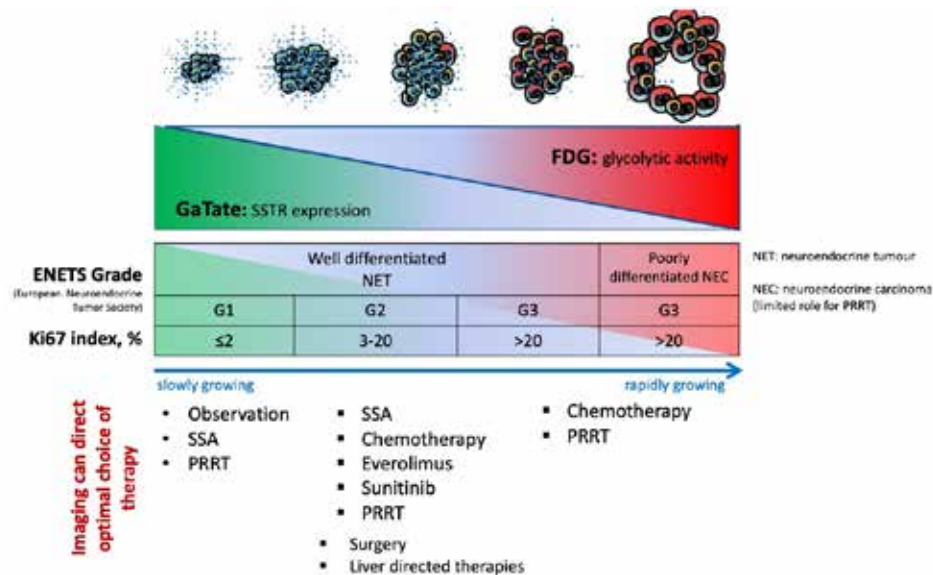
- tumour site and size
- multifocality
- lymphovascular and perineural invasion
- extent of local invasion
- surgical margins
- nodal status
- presence of background disease.

In addition to mitotic count, Ki-67 index and the reporting of tumour necrosis (subject to site), which are considered mandatory in all NETs, other ancillary tests that may be useful in selected circumstances include (Perren et al. 2017):

- DAXX/ATRX immunohistochemistry (loss favours pancreatic origin)
- p53/pRb immunohistochemistry (p53 mutation and loss of RB1 favours NEC over NET)
- SSTR2 (may be used to predict positivity on 68Ga-DOTATATE PET/CT scans)
- MGMT immunohistochemistry or promoter methylation (potential predictor of response to temozolomide)
- immunohistochemistry for pancreatic hormone expression in context of a clinical hormonal syndrome (e.g. insulin and glucagon)
- SDHB immunohistochemistry (loss of expression makes syndromic disease due to germline mutation of one of the SDH genes highly likely).

See figure 4 (below) for classification of neuroendocrine tumour with corresponding imaging features and treatment options.

Figure 4: Classification of neuroendocrine tumour with corresponding imaging features and treatment options.



* GaTate: 68Ga-DOTATATE PET/CT; a specialised Gallium 68-dotatate tracer is injected and binds to somatostatin receptors on the cell surface of neuroendocrine cells (I-Med Radiology Network 2020). The presence of these receptors enables appropriate treatment choice.

* FDG: 18F-FDG PET/CT: The most common radiotracer is F-18 fluorodeoxyglucose (FDG), a molecule similar to glucose. Cancer cells are more metabolically active and may absorb glucose at a higher rate. 18-Fluorodeoxyglucose (18F-FDG) positron emission tomography (PET) avidity in NETs has been associated with higher grade disease (Chan et al. 2020). The presence of active uptake of 18F-FDG PET/CT enables appropriate treatment choice.

Source: Hofman & Hicks 2012

Classifying NETs into one of the 4 stages is site-specific and should be in the form of the tumour, nodes and metastases (TNM) system – refer to the 8th edition of the American Joint Committee on Cancer's Staging Manual.

Staging for NETs may involve these tests:

- CT scan
- MRI
- 68Ga-DOTATATE PET/CT
- 18F-FDG PET/CT
- 18F-DOPA (for metastatic MTC, and for certain pheochromocytomas)
- I-123-MIBG or I-131 MIBG imaging (for pheochromocytomas and paragangliomas) – choice of imaging depends on availability and local preference.

Note: Pheochromocytomas/paragangliomas should never be biopsied.



More information

Visit the Cancer Institute New South Wales website for information about understanding the stages of cancer <www.cancer.nsw.gov.au/understanding-cancer/stages-of-cancer>.

3.3 Performance status

Patient performance status is a central factor in cancer care and should be clearly documented in the patient's medical record.

Performance status should be measured and recorded using an established scale such as the Karnofsky scale or the Eastern Cooperative Oncology Group (ECOG) scale. The ECOG scale is the most used scale for NETs.

3.4 Treatment planning

3.4.1 Key considerations beyond treatment recommendations

A number of factors should be considered at this stage:

- the patient's overall condition, life expectancy, personal preferences and decision-making capacity
- discussing the multidisciplinary team approach to care with the patient
- appropriate and timely referral to an MDM
- pregnancy and fertility
- support with travel and accommodation
- teleconferencing or videoconferencing as required.

3.4.2 Timing for multidisciplinary team planning

The multidisciplinary team should meet to discuss newly diagnosed patients before definitive treatment so that a treatment plan can be recommended. Patients should be presented at an appropriate NET multidisciplinary meeting, with all appropriate investigation results, **within four 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. Some patients with non-complex cancers may not be discussed by a multidisciplinary team; instead the team may have treatment plan protocols that will be applied if the patient's case (cancer) meets the criteria. If patients are not discussed at an MDM, they should at least be named on the agenda for noting. The proposed treatment must be recorded in the patient's medical record and should be recorded in an MDM database where one exists.

Teams may agree on standard treatment protocols for non-complex care, facilitating patient review (by exception) and associated data capture.

Results of all relevant tests and access to images should be available for the MDM. Information about the patient's concerns, preferences and social and cultural circumstances should also be available.

3.4.3 Responsibilities of the multidisciplinary team

The multidisciplinary team requires administrative support in developing the agenda for the meeting, for collating patient information and to ensure appropriate expertise around the table to create an effective treatment plan for the patient. The MDM has a chair and multiple lead clinicians. Each patient case will be presented by a lead clinician (usually someone who has seen the patient before the MDM). In public hospital settings, the registrar or clinical fellow may take this role. A member of the team records the outcomes of the discussion and treatment plan in the patient history and ensures these details are communicated to the patient's general practitioner. The team should consider the patient's values, beliefs and cultural needs as appropriate to ensure the treatment plan is in line with these.

3.4.4 Members of the multidisciplinary team for NETs

The multidisciplinary team should be composed of the core disciplines that are integral to providing good care. Team membership should reflect both clinical and supportive care aspects of care. Pathology and radiology expertise are essential.

See Appendix E for a list of team members who may be included in the multidisciplinary team for NETs.

Core members of the multidisciplinary team are expected to attend most MDMs either in person or remotely via virtual mechanisms. Additional expertise or specialist services may be required for some patients. An Aboriginal and Torres Strait Islander cultural expert should be considered for all patients who identify as Aboriginal or Torres Strait Islander.

3.4.5 Responsibilities of individual team members

The general practitioner who made the referral is responsible for the patient until care is passed to another practitioner who is directly involved in planning the patient's care.

The general practitioner may play a number of roles in all stages of the cancer pathway including diagnosis, referral, treatment, shared follow-up care, post-treatment surveillance, coordination and continuity of care, as well as managing existing health issues and providing information and support to the patient, their family and carer.

A nominated contact person from the multidisciplinary team may be assigned responsibility for coordinating care in this phase. Care coordinators are responsible for ensuring there is continuity throughout the care process and coordination of all necessary care for a particular phase (COSA 2015). The care coordinator may change over the course of the pathway.

The lead clinician is responsible for overseeing the activity of the team and for implementing treatment within the multidisciplinary setting.

3.5 Research and clinical trials

Patients should be encouraged to participate in research or clinical trials where available and appropriate.

For more information visit the Cancer Australia website <www.australiancancertrials.gov.au>.

3.6 Support and communication

3.6.1 Prehabilitation

Cancer prehabilitation uses a multidisciplinary approach combining exercise, nutrition and psychological strategies to prepare patients for the challenges of cancer treatment such as surgery, systemic therapy and radiation therapy. Team members may include anaesthetists, oncologists, surgeons, haematologists, clinical psychologists, exercise physiologists, physiotherapists and dietitians, among others.

Patient performance status is a central factor in cancer care and should be frequently assessed. All patients should be screened for malnutrition using a validated tool, such as the Malnutrition Screening Tool (MST). The lead clinician may refer obese or malnourished patients to a dietitian preoperatively or before other treatments begin.

Patients who currently smoke should be encouraged to stop smoking before receiving treatment. This should include an offer of referral to Quitline in addition to smoking cessation pharmacotherapy if clinically appropriate.

Evidence indicates that patients who respond well to prehabilitation may have fewer complications after treatment. For example, those who were exercising before diagnosis and patients who use prehabilitation before starting treatment may improve their physical or psychological outcomes, or both, and this helps patients to function at a higher level throughout their cancer treatment (Cormie et al. 2017; Silver 2015).

For patients with NETs, including low-grade NETs, who will initially undergo close observation in a 'watch and wait' approach, the multidisciplinary team should consider these specific prehabilitation assessments and interventions for treatment-related complications or major side effects:

- conducting a physical and psychological assessment to establish a baseline function level
- identifying impairments and providing targeted interventions to improve the patient's function level (Silver & Baima 2013)
- reviewing the patient's medication to ensure optimisation and to improve adherence to medicine used for comorbid conditions
- referral to a psycho-oncology service to improve the patient's knowledge and motivation towards healthy self-care behaviours and to emotionally regulate throughout the course of the disease and treatments.

Rehabilitation programs have considerable potential to enhance physical function while undergoing cancer treatment.

3.6.2 Fertility preservation and contraception

Cancer and cancer treatment may cause fertility problems. This will depend on the age of the patient, the type of cancer and the treatment received. Infertility can range from difficulty having a child to the inability to have a child. Infertility after treatment may be temporary, lasting months to years, or permanent (AYA Cancer Fertility Preservation Guidance Working Group 2014).

Patients need to be advised about and potentially referred for discussion about fertility preservation before starting treatment and need advice about contraception before, during and after treatment. Patients and their family should be aware of the ongoing costs involved in optimising fertility. Fertility management may apply in both men and women. Fertility preservation options are different for men and women and the need for ongoing contraception applies to both men and women.

The potential for impaired fertility should be discussed and reinforced at different time points as appropriate throughout the diagnosis, treatment, surveillance and survivorship phases of care. These ongoing discussions will enable the patient and, if applicable, the family to make informed decisions. All discussions should be documented in the patient's medical record.



More information

See the Cancer Council website <www.cancervic.org.au/living-with-cancer/common-side-effects/fertility/fertility-overview.html> for more information.

3.6.3 Supportive care

See validated screening tools mentioned in Principle 4 'Supportive care'.

A number of specific challenges and needs may arise for patients at this time:

- assistance for dealing with psychological and emotional distress while adjusting to the diagnosis; treatment phobias; existential concerns; stress; difficulties making treatment decisions; anxiety or depression or both; psychosexual issues such as potential loss of fertility and premature menopause; history of sexual abuse; and interpersonal problems
- strong emotions such as anger at the medical community for the delays and misunderstanding of the diagnosis, confusion, fear and hopelessness about what the future holds
- management of physical symptoms such as pain and fatigue (Australian Adult Cancer Pain Management Guideline Working Party 2019)
- malnutrition or undernutrition, identified using a validated nutrition screening tool such as the MST (note that many patients with a high BMI [obese patients] may also be malnourished [WHO 2018])
- referral to a dietitian for management of dietary needs and management of symptoms such as abdominal pain, diarrhoea and flushing – patients may benefit from using a food and symptom diary <<https://neuroendocrine.org.au/other-resources/>>.
- explaining the role of diagnostic tools such as PET scans and funding
- support for families or carers who are distressed with the patient's cancer diagnosis
- support for families/relatives who may be distressed after learning of a genetically linked cancer diagnosis
- specific spiritual needs that may benefit from the involvement of pastoral/spiritual care.

Additionally, palliative care may be required at this stage.

For more information on supportive care and needs that may arise for different population groups, see Appendices A, B and C.

3.6.4 Communication with patients, carers and families

In discussion with the patient, the lead clinician should undertake the following:

- establish if the patient has a regular or preferred general practitioner and if the patient does not have one, then encourage them to find one
- provide written information appropriate to the health literacy of the patient about the diagnosis and treatment to the patient and carer and refer the patient to the Guide to best cancer care (consumer optimal care pathway) for NETs, as well as to relevant websites and support groups such as NeuroEndocrine Cancer Australia NET Nurse support line 1300 287 363 or support online <<https://neuroendocrine.org.au/net-nurse/>>
- provide a treatment care plan including contact details for the treating team and information on when to call the hospital
- refer the patient to the NeuroEndocrine Cancer Australia *Treatment and wellness plan* <<https://neuroendocrine.org.au/treatment-wellness-care-plan>>
- discuss a timeframe for diagnosis and treatment with the patient and carer
- discuss the benefits of multidisciplinary care and gain the patient's consent before presenting their case at an MDM
- provide brief advice and refer to Quitline (13 7848) for behavioural intervention if the patient currently smokes (or has recently quit), and prescribe smoking cessation pharmacotherapy, if clinically appropriate
- recommend an 'integrated approach' throughout treatment regarding nutrition, exercise and minimal or no alcohol consumption among other considerations
- communicate the benefits of continued engagement with primary care during treatment for managing comorbid disease, health promotion, care coordination and holistic care
- where appropriate, review fertility needs with the patient and refer for specialist fertility management (including fertility preservation, contraception, management during pregnancy and of future pregnancies)
- be open to and encourage discussion about the diagnosis, prognosis (if the patient wishes to know) and survivorship and palliative care while clarifying the patient's preferences and needs, personal and cultural beliefs and expectations, and their ability to comprehend the communication
- encourage the patient to participate in advance care planning including considering appointing one or more substitute decision-makers and completing an advance care directive to clearly document their treatment preferences. Each state and territory has different terminology and legislation surrounding advance care directives and substitute decision-makers.

3.6.5 Communication with the general practitioner

The lead clinician has these communication responsibilities:

- involving the general practitioner from the point of diagnosis
- ensuring regular and timely communication with the general practitioner about the diagnosis, treatment plan and recommendations from MDMs and inviting them to participate in MDMs (consider using virtual mechanisms)
- supporting the role of general practice both during and after treatment
- discussing shared or team care arrangements with general practitioners or regional cancer specialists, or both, together with the patient.



More information

Refer to Principle 6 'Communication' for communication skills training programs and resources.

Step 4: Treatment

Step 4 describes the optimal treatments for NETs, the training and experience required of the treating clinicians and the health service characteristics required for optimal cancer care.

All health services must have clinical governance systems that meet the following integral requirements:

- identifying safety and quality measures
- monitoring and reporting on performance and outcomes
- identifying areas for improvement in safety and quality (ACSQHC 2020).

Step 4 outlines the treatment options for NETs. Get detailed clinical information on treatment types <<https://www.cancer.net/cancer-types/neuroendocrine-tumors/types-treatment>>, practice guidelines <<https://www.esmo.org/guidelines/endocrine-and-neuroendocrine-cancers/gastroenteropancreatic-neuroendocrine-neoplasms>> and treatment protocols <<https://www.eviq.org.au/medical-oncology/upper-gastrointestinal/neuroendocrine>>.

4.1 Treatment intent

The intent of treatment can be defined as one or more of the following:

- curative
- anti-cancer therapy to improve quality of life and/or longevity without expectation of cure
- symptom palliation.

The treatment intent should be established in a multidisciplinary setting, documented in the patient's medical record and conveyed to the patient and carer as appropriate.

The potential benefits need to be balanced against the morbidity and risks of treatment.

The lead clinician should discuss the advantages and disadvantages of each treatment and associated potential side effects with the patient and their carer or family before treatment consent is obtained and begins so the patient can make an informed decision. Supportive care services should also be considered during this decision-making process. Patients should be asked about their use of (current or intended) complementary therapies (see Appendix D).

Timeframes for starting treatment should be informed by evidence-based guidelines where they exist. The treatment team should recognise that shorter timeframes for appropriate consultations and treatment can promote a better experience for patients.

Initiate advance care planning discussions with patients before treatment begins (this could include appointing a substitute decision-maker and completing an advance care directive). Formally involving a palliative care team/service may benefit any patient, so it is important to know and respect each person's preference (AGDH 2021).

4.2 Treatment options

4.2.1 Surgery

Some patients may benefit from surgery. The decision to proceed with surgery is often complex and must be tailored to each patient's disease. Surgery may be undertaken with curative intent, to palliate symptoms, to prevent symptoms, or to prolong survival (Gangi & Anaya 2020).

When the cancer is detected early, is small and has not metastasised, it is treated with curative intent. Completely removing the entire tumour is the standard treatment, when possible. Most patients with localised NETs are successfully treated with surgery alone. If the tumour can be removed in its entirety, then surgery may cure the cancer (NECA 2019a).

The surgical procedure undertaken will depend on the location(s) of the NET and treatment intent. It may involve different surgical specialties if disease involves more than one organ system.

Patients with NETs of the intestine may develop complications such as bowel obstruction or ischaemia. This can sometimes necessitate emergency surgery, even in the presence of unresectable metastases. Asymptomatic intestinal NETs in the presence of widespread metastases may not require any surgery. When curative intent surgery is undertaken, locoregional lymph nodes should be removed along with the GEP-NET for adequate staging and longer term disease control.

Metastases to the liver from NETs in any part of the body may potentially be managed with surgery, either with curative intent, or to improve survival if more than 70 per cent of the disease can be removed. Overall, surgery for GEP-NET is guided by the grade and degree of differentiation of the disease.

For patients with localised paraganglioma and pheochromocytoma, surgery is usually undertaken to completely remove the tumour, and for pheochromocytoma is usually an adrenalectomy (removal of one or both adrenal glands). Pre-operative control of blood pressure (typically with alpha-blockade) is essential for all functioning pheochromocytomas and paragangliomas. At the time of surgery, tissue and lymph nodes will be checked for metastases, and may be removed. If both adrenal glands are removed, lifelong corticosteroid and mineralocorticoid replacement therapy will be required.

Cardiac or thoracic surgery should be considered for patients with lung NETs, metastases to the lungs, or with carcinoid heart disease who need a cardiac valve replacement.

People who have developed carcinoid syndrome are at risk of experiencing a carcinoid crisis during surgery. These patients should already be treated with somatostatin analogues (SSAs), but if not, consideration should be given to commencing them prior to surgery. To avoid major complications from a carcinoid crisis, the anaesthetic team must be fully aware of this risk before surgery so they can have treatment on hand to control the symptoms.

Intravenous octreotide is usually given before surgery to prevent carcinoid crisis. See section 6.5 for more information on carcinoid crisis.

Palliative surgery may be offered to patients when the tumour or tumours have spread or become too large to remove completely. Palliative surgery aims to 'de-bulk' the tumour, which could relieve some symptoms.

Timeframe for starting treatment

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

Training and experience required of the surgeon

Fellow of the Royal Australian College of Surgeons (or equivalent) with adequate training and experience that enables institutional credentialing and agreed scope of practice in this area.

Documented evidence of the surgeon's training and experience, including their specific (sub-specialty) experience with NETs and procedures to be undertaken, should be available.

Different surgeons may be involved such as colorectal, hepatobiliary, and cardiothoracic.

There is strong evidence to suggest that institutions with a high volume of NET resections have better clinical outcomes for complex cancer surgery (Toomey et al. 2016).

Health service characteristics

To provide safe and quality care for patients having surgery, health services should have these features:

- critical care support
- 24-hour medical staff availability
- 24-hour operating room access and intensive care unit
- diagnostic imaging
- pathology
- nuclear medicine imaging.

4.2.2 No treatment / active surveillance

No treatment, active surveillance or watchful waiting, may be suitable for some NET patients especially if the NET is not causing symptoms or problems, there is little disease, the disease is stable, or the tumour is low grade (G1). In some circumstances, poor general health or complications secondary to treatments may also make further NET treatment inadvisable.

4.2.3 Localised radiation therapy

NETs are usually radiosensitive tumours. A number of patients may benefit from radiation therapy if they have oligometastatic disease, a dominant or critically strategic site of progression or highly symptomatic metastases (e.g. in bone):

- External beam radiation therapy can be used in selected patients with painful skeletal metastases, particularly when peptide receptor radionuclide therapy (PRRT) is unavailable or contraindicated.
- Stereotactic body radiation therapy may be considered for selected patients with solitary liver or lung metastasis, as an alternative to surgery.
- Proton beam therapy – this has not been specifically evaluated in NETs.
- Radioembolisation (selective internal radiation therapy) (see section 4.2.6) – this procedure is performed by an interventional radiologist, with the assistance of a nuclear medicine specialist licensed to administer radionuclides.

External beam radiation therapy for symptom palliation can be given to almost all patients, regardless of their overall health and performance status. More invasive procedures (e.g. radioembolisation) are not generally provided as part of end-of-life care but are offered to patients with advanced metastatic disease if it is anticipated that local disease control will improve overall symptoms or quality of life.

Timeframe for starting treatment

Treatment should start **as soon as possible** for symptomatic patients.

Training and experience required of the appropriate specialists

Fellow of the Royal Australian and New Zealand College of Radiologists or Royal Australasian College of Physicians (or equivalent) with adequate training and experience that enables institutional credentialing and agreed scope of practice in NETs.

The training and experience of the radiation oncologist, interventional radiologist and nuclear medicine specialist should be documented.

Health service unit characteristics

To provide safe and quality care for patients having external beam radiation therapy or stereotactic body radiation therapy, health services should have these features:

- linear accelerator capable of image-guided radiation therapy
- dedicated CT planning
- access to MRI and PET imaging
- automatic record-verify of all radiation treatments delivered
- a treatment planning system
- trained medical physicists, radiation therapists and nurses with radiation therapy experience
- coordination for combined therapy with systemic therapy, especially where facilities are not co-located
- participation in Australian Clinical Dosimetry Service audits
- an incident management system linked with a quality management system.

For patients undergoing radioembolisation, health services should have these features:

- sterile, licensed radiopharmaceutical dispensing facilities with adequate storage of radioactive waste
- interventional radiology facilities (digital subtraction angiography) and appropriately trained and credentialed radiologists with experience in hepatic artery anatomy and radioembolisation techniques, authorised for use of unsealed radiation sources by the relevant state or territory licensing authority
- trained medical physicists and nurses with nuclear medicine and interventional radiology experience
- capacity for concurrent or sequential trans-catheter chemo-embolisation
- capacity for post-treatment inpatient admission for pain relief if required
- trained nuclear medicine specialists for dose calculation and treatment planning (together with interventional radiologist, referring clinician and medical physicist)
- SPECT or PET facilities for pre-treatment dosimetric planning and post-therapy dose distribution and isodose contouring.

4.2.4 Systemic radiation therapy – peptide receptor radionuclide therapy

Patients with metastatic disease who have progressed following first-line SSAs may be suitable for systemic radionuclide therapy (or PRRT). A major criteria for selection is sufficient expression of somatostatin receptors as demonstrated on molecular imaging, typically with ⁶⁸Ga-DOTATATE PET/CT scan. These provide the suitable target for systemically administered radiopharmaceutical. Imaging with ¹⁸F-FDG PET provides additive in-vivo evaluation of grade with the ideal situation being high somatostatin receptor expression (ideally Krenning score 3-4) without discordant FDG avidity.

PRRT can be delivered concurrently with chemotherapy when appropriate, such as in patients with extensive tumour burden, pNETs, rapid disease progression or those with higher grade disease at diagnosis (GI Cancer Institute 2021). This treatment option should only be recommended following patient review at an appropriate NET multidisciplinary team. Some expert centres may use PRRT as first-line in selected cases.

There are several centres around Australia with expertise to deliver PRRT for NETs. PRRT is usually an outpatient (day admission) therapy (refer to Appendix E). An infusion of amino acids is administered at the time of PRRT to help protect the kidneys from nephrotoxicity. The radiopharmaceutical itself is usually infused over 15–20 minutes following appropriate premedication.

PRRT is most often given as an induction course of four treatments, with each dose/cycle separated by six to eight weeks.

During the recent NETTER-1 trial (Strosberg et al. 2017), patients were able to tolerate standard doses of ¹⁷⁷Lu-DOTATATE without any significant decline in renal function compared with patients who received high-dose SSA therapy. Both amino acid infusions and intravenous radiopharmaceuticals can result in nausea and so antiemetic medication is given routinely to reduce the risk of nausea and vomiting.

In case of eventual future NET progression, the patient's multidisciplinary team should consider repeating PRRT treatment as salvage therapy. In many countries, including Australia, PRRT can continue to be offered, with appropriate intervals, providing there was initial objective and symptomatic treatment response.

PRRT is also suited to patients with poorly controlled NET symptoms, even when there is only stable or slowly progressive tumour burden.

There are several different radionuclides that can be used for PRRT. Those currently used in Australia include:

- ¹⁷⁷Lu-DOTATATE is currently the radiopharmaceutical of choice.
- Yttrium-90 (Y-90 or ⁹⁰Y) was the original isotope used in PRRT but is now largely replaced by ¹⁷⁷Lu-DOTATATE due to its lower cost and toxicity.
- Actinium-225 (Ac-225 or ²²⁵Ac) is the newest radionuclide used in PRRT. It is still an investigational agent but its compassionate use in Australia is slowly increasing.
- Copper-67 (Cu-67 or ⁶⁷Cu) is one of the additional emerging radionuclides. At this time of writing, it is not funded in Australia, but trials are underway.

Monitoring of PRRT side effects

PRRT is well tolerated and can be used in nearly all patients, including elderly patients. Due to its renal excretion, moderate renal impairment at baseline (GFR < 30 ml/min/1.73m²) is generally considered an absolute contraindication to PRRT, while it should be used with caution in those with milder renal impairment (GFR < 50 mL/min). Nephrotoxicity from PRRT is more likely in those with baseline renal impairment, and so close monitoring of renal function is required.

PRRT is also myelotoxic and so should be used cautiously in patients with impaired bone marrow reserve who have previously received combination chemotherapy, particularly with regimens containing alkylating agents, and in patients who are also receiving concurrent, radio-sensitising chemotherapy at the time of their PRRT. The nadir count is usually at three to four weeks after treatment, so close monitoring is recommended. Accurate CTCAE grading of cytopenias will guide scheduling and dosing. An increase in the interval between treatments and reduction in the radiation dose of PRRT may be required in such patients.

A potential uncommon but serious side effect of PRRT is carcinoid crisis (Kaltsas et al. 2017). This can be a worsening of carcinoid symptoms or could precipitate a carcinoid crisis. Acute intravenous administration of octreotide has been reported to provide rapid reversal of a carcinoid crisis, and the current focus of carcinoid therapy is to prevent mediator release with octreotide prophylaxis. This has largely replaced the use of other drugs for acute treatment. See section 6.5 for more information on carcinoid crisis.

Timeframes for starting treatment

When PRRT is necessary (for patients who have sufficient expression of somatostatin receptors that have progressed on SSAs or where initial PRRT is considered appropriate), treatment should start **as soon as possible**.

Training and experience required of the appropriate specialists

Fellow of the Royal Australian and New Zealand College of Radiologists or Royal Australasian College of Physicians (or equivalent) with adequate training and experience that enables institutional credentialing and agreed scope of practice in NETs.

The training and experience of the nuclear medicine specialist should be documented.

Health service characteristics

To provide safe and quality care for patients having PRRT, health services should have these features:

- sterile radiopharmaceutical manufacturing facilities, which are licensed by the relevant state or territory authority, with access to full radiopharmaceutical quality control techniques
- a shielded room in which to deliver radionuclide therapies, authorised for such purposes by the relevant state or territory body – this room should contain emergency call provisions and medical gases with capacity for patient resuscitation if required (e.g. carcinoid crisis)
- on-site nuclear medicine specialist, medical physicist and nursing staff with training/expertise in PRRT
- capacity for immediate administration of SC or IV octreotide fluid resuscitation in the event of worsening carcinoid symptoms or crisis, as well as overnight admission for those at high risk of carcinoid crisis
- on-site gamma camera facilities for post-therapy and, where available, dosimetric scans, nuclear medicine technologist staff with training/expertise in post-therapy and dosimetric scanning and available medical physics support
- capacity for inpatient or intensive care transfer in the event of a medical emergency (e.g. carcinoid crisis).

4.2.5 Systemic therapy

Systemic therapies that should be considered for patients are:

- adjuvant chemotherapy for some patients with resected high-grade NETs
- SSAs
- concurrent chemotherapy / PRRT
- chemotherapy for patients with neuroendocrine carcinoma
- chemotherapy for patients with advanced high-grade / unresectable high-grade NETs
- PRRT for patients with well differentiated NETs – refer to section 4.2.4
- targeted therapies.

Medications for less common subtypes of NETs include proton pump inhibitors (gastrinomas), diazoxide (insulinomas), alpha blockers (paragangliomas and pheochromocytomas) and dopamine receptor agonists (ectopic cushings in bronchial carcinoids).

Somatostatin analogues

SSAs are the mainstay of medical treatment for most patients with NETs. Two studies show the growth delay effect with SSAs—PROMID (Rinke et al. 2016) and CLARINET (Caplin et al. 2021).

SSAs are the most common first-line treatment of G1/G2 NETs. They have antisecretory and antiproliferative effects. Depot injections of SSAs (octreotide, lanreotide) are available to control some symptoms caused by NETs. Injections of these analogues can stop the overproduction of hormones (e.g. serotonin) that cause symptoms such as flushing and diarrhoea. These are usually given every four weeks – sometimes more frequently (two or three weekly).

Short-acting octreotide may be considered to control symptoms for two to three days until a correct dose of long-lasting SSA can be prescribed. In some cases, short-acting SSA may be used ongoingly to treat breakthrough symptoms of severe diarrhoea and flushing in patients with active/severe carcinoid syndrome.

Once commenced, SSA therapy is usually continued for life unless there are intolerable poorly tolerated side effects, even when second-line therapies have been instituted.

Molecular targeted therapies

An mTOR inhibitor (everolimus) has been developed to treat advanced NETs of the gastrointestinal tract, lung and pancreas. This drug can help slow down the growth of these tumours in some patients, but it does not usually shrink tumours.

Sunitinib, a drug with multiple targets, is also effective in slowing down the growth of pNETs.

Other targeted therapies for NETs are being researched in clinical trials. They include drugs that interfere with new blood vessel formation or with specific survival pathways of cancer cells.

Chemotherapy

Chemotherapy may be considered for some patients, especially for NET patients with pancreatic, bronchial or high-grade (G2/G3) NETs. Not all NETs respond equally to chemotherapy, therefore careful selection is required to improve the chance of response and avoid unnecessary side effects and toxicity. The histological grade and the primary site of the tumour (how it looks under the microscope after biopsy or operation) may help determine the type of treatment that is recommended. Although there is little data to guide practice, chemotherapy may sometimes be recommended after surgery (adjuvant chemotherapy) (NCCN 2021).

PRRT has now emerged as an effective therapy for paragangliomas/phaeochromocytomas, although combination chemotherapy may sometimes be used in some cases for patients with phaeochromocytomas and paragangliomas.

Timeframes for starting treatment

When active treatment is considered necessary, treatment should start **within four weeks** of the treatment decision.

Training and experience required of the appropriate specialists

Medical oncologists and endocrinologists must have training and experience of this standard:

Fellow of the Royal Australian College of Physicians (or equivalent) with adequate training and experience that enables institutional credentialing and agreed scope of practice in NETs (ACSQHC 2015).

Documented evidence of the medical oncologist and endocrinologist's training and experience, including their specific (sub-specialty) experience with NETs should be available.

There is strong evidence to suggest that institutions with a high volume of NET patients have better clinical outcomes due to comprehensive multidisciplinary management (Magi et al. 2019; Tsoli et al. 2018).

Cancer nurses should have accredited training in these areas:

- anti-cancer treatment administration
- specialised nursing care for patients undergoing cancer treatments, including side effects and symptom management
- the handling and disposal of cytotoxic waste (ACSQHC 2020).

Systemic therapy should be prepared by a pharmacist whose background includes this experience:

- adequate training in systemic therapy medication, including dosing calculations according to protocols, formulations and/or preparation.

Some therapies may be delivered by a general practitioner or nurse with training and experience that enables credentialing and agreed scope of practice within this area. This should be in accordance with a detailed treatment plan or agreed protocol, and with communication as agreed with the medical oncologist or endocrinologist with NET interest or as clinically required.

Health service characteristics

To provide safe and quality care for patients having systemic therapy, health services should have these features:

- a clearly defined path to emergency care and advice after hours
- access to full-suite diagnostic pathology including basic haematology and biochemistry, NET-specific biochemistry, experienced histopathology and molecular pathology and advanced imaging
- cytotoxic drugs prepared in a pharmacy with appropriate facilities
- occupational health and safety guidelines regarding handling of cytotoxic drugs, including preparation, waste procedures and spill kits (eviQ 2019)
- guidelines and protocols to deliver treatment safely (including dealing with extravasation of drugs)
- coordination for combined therapy with radiation therapy, especially where facilities are not co-located
- coordination for combined medical and systemic radiation therapy
- appropriate molecular pathology access

Lead NET team characteristics

To provide safe and quality care for patients, the lead NET team should have these features:

- significant case volume
- documented expertise in managing NETs
- ideally involved in ongoing NET research
- multidisciplinary NET coordinated care.

4.2.6 Liver directed therapy

Hepatic artery embolisation (HAE) and transcatheter arterial chemoembolisation (TACE) may benefit patients if the NET tumour has spread to the liver.

Radioembolisation (selective internal radiation therapy) may benefit patients with liver metastases that cannot be removed with surgery.

Embolisation may benefit patients with pheochromocytomas and paragangliomas.

Ablation therapy (radiofrequency or cryoablation) may benefit patients with tumours specific regions (e.g. in the liver).

Timeframes for starting treatment

When active treatment is considered necessary, treatment should start **within four weeks** of the treatment decision.

Training and experience required of the appropriate specialists

Interventional radiologist (Fellow of the Royal Australian and New Zealand College of Radiologists (or equivalent)) with adequate training and experience in liver-directed therapies and institutional credentialing and agreed scope of practice in NETs.

Tier B (advanced) interventional radiology competency (as defined by the Royal Australian and New Zealand College of Radiologists / Interventional Radiology Society of Australasia) is recommended following fellowship training sufficient to obtain European Board of Interventional Radiology or equivalent standard.

4.2.7 Immunotherapy

Immunotherapy is a rapidly evolving therapeutic field in different types of cancer. However, the role of immunotherapy in managing NETs is still investigational.

Immunotherapy for NETs management is currently being evaluated in several studies using innovative clinical trial designs, such as basket or umbrella trials, either prescribed alone or in combination with other therapies.

4.2.8 Emerging therapies

The key principle for precision medicine is prompt and clinically oriented communication and coordination with an accredited NETs multidisciplinary team. Tissue analysis is integral for access to emerging therapies and, as such, tissue specimens should be treated carefully to enable additional histopathological or molecular diagnostic tests in certain scenarios.

4.3 Palliative care

Early referral to palliative care can improve the quality of life for people with cancer and in some cases may be associated with survival benefits (Haines 2011; Temel et al. 2010; Zimmermann et al. 2014). This is particularly true for cancers with poor prognosis.

The lead clinician should ensure patients receive timely and appropriate referral to palliative care services. Referral should be based on need rather than prognosis. Emphasise the value of palliative care in improving symptom management and quality of life to patients and their carers.

The 'Dying to Talk' resource may help health professionals when initiating discussions with patients about future care needs (see 'More information'). Ensure that carers and families receive information, support and guidance about their role in palliative care (Palliative Care Australia 2018).

Patients, with support from their family or carer and treating team, should be encouraged to consider appointing a substitute decision-maker and to complete an advance care directive.

Refer to step 6 for a more detailed description of managing patients with recurrent, residual or metastatic disease.



More information

These online resources are useful:

- Advance Care Planning Australia <www.advancecareplanning.org.au>
- Care Search <www.caresearch.com.au/Caresearch/>
- Dying to Talk <www.dyingtotalk.org.au>
- the Palliative Care resource kit <www.health.gov.au/health-topics/palliative-care>
- Palliative Care Australia (for patients and carers) <www.palliativecare.org.au>.

4.4 Research and clinical trials

The team should support the patient to participate in research or clinical trials where available and appropriate. Many emerging treatments are only available on clinical trials that may require referral to certain trial centres.

For more information visit the Cancer Australia website <www.australiancancertrials.gov.au>.

4.5 Support and communication

4.5.1 Supportive care

See validated screening tools mentioned in Principle 4 'Supportive care'.

A number of specific challenges and needs may arise for patients at this time:

- assistance for dealing with emotional and psychological issues, including body image concerns, fatigue, quitting smoking, traumatic experiences, existential anxiety, treatment phobias, anxiety/depression, interpersonal problems and sexuality concerns
- potential isolation from normal support networks, particularly for rural patients who are staying away from home for treatment
- decline in mobility or functional status as a result of treatment
- assistance with beginning or resuming regular exercise with referral to an exercise physiologist or physiotherapist (COSA 2018; Hayes et al. 2019)
- explaining sequencing of treatment and the need for different treatments at different times such as surgery, liver-directed therapy, PRRT and SSAs
- surgical recovery including diabetes education where relevant
- screening, assessment and management of flare up of symptoms associated with carcinoid syndrome, which may include flushing diarrhoea, palpitations, anxiety, wheezing and bloating abdominal pain
- management of treatment side effects associated with liver directed therapies such as fatigue, nausea, vomiting and pain around the liver
- administration of medication, which may be before, during and after surgery, to keep blood pressure and heart rate controlled
- management of side effects associated with chemotherapy or molecular targeted therapies such as mouth sores, lowering of blood counts, diarrhoea, nausea, vomiting, fatigue and high blood pressure
- management of side effects related to PRRT such as nausea, fatigue and lowering of blood counts
- monitoring of long-term toxicity from PRRT – this involves a 5 per cent risk of myelodysplastic syndrome, which may sometimes progress to acute leukaemia.

Early involvement of general practitioners may lead to improved cancer survivorship care following acute treatment. General practitioners can address many supportive care needs through good communication and clear guidance from the specialist team (Emery 2014).

Patients, carers and families may have these additional issues and needs:

- financial issues related to loss of income (through reduced capacity to work or loss of work) and additional expenses as a result of illness or treatment
- advance care planning, which may involve appointing a substitute decision-maker and completing an advance care directive
- legal issues (completing a will, care of dependent children) or making an insurance, superannuation or social security claim on the basis of terminal illness or permanent disability.

Cancer Council's 13 11 20 information and support line can assist with information and referral to local support services.

NeuroEndocrine Cancer Australia offers online nurse support, a telephone support service (1300 287 363) and a private Facebook community where many patients converse.

For more information on supportive care and needs that may arise for different population groups, see Appendices A, B and C.

4.5.2 Rehabilitation

Rehabilitation may be required at any point of the care pathway. If it is required before treatment, it is referred to as prehabilitation (see section 3.6.1).

All members of the multidisciplinary team have an important role in promoting rehabilitation. Team members may include occupational therapists, speech pathologists, dietitians, social workers, psychologists, physiotherapists, exercise physiologists and rehabilitation specialists.

To maximise the safety and therapeutic effect of exercise for people with cancer, all team members should recommend that people with cancer work towards achieving, and then maintaining, recommended levels of exercise and physical activity as per relevant guidelines. Exercise should be prescribed and delivered under the direction of an accredited exercise physiologist or physiotherapist with experience in cancer care (Vardy et al. 2019). The focus of intervention from these health professionals is tailoring evidence-based exercise recommendations to the individual patient's needs and abilities, with a focus on the patient transitioning to ongoing self-managed exercise.

Other issues that may need to be dealt with include managing cancer-related fatigue, improving physical endurance, achieving independence in daily tasks, optimising nutritional intake, returning to work and ongoing adjustment to cancer and its sequels. Referrals to dietitians, psychosocial support, return-to-work programs and community support organisations can help in managing these issues.

4.5.3 Communication with patients, carers and families

A lead clinician should be nominated to be the principal contact person for each NET patient, ideally by consensus among the multidisciplinary team. This lead clinician may change occasionally, depending on the patient's circumstance, but will most often be a medical oncologist with experience in NETs.

The lead or nominated clinician should take responsibility for these tasks:

- discussing treatment options with patients and carers, including the treatment intent and expected outcomes, and providing a written version of the plan and any referrals
- providing patients and carers with information about the possible side effects of treatment, managing symptoms between active treatments, how to access care, self-management strategies and emergency contacts
- encouraging patients to use question prompt lists and audio recordings, and to have a support person present to aid informed decision making
- initiating a discussion about advance care planning and involving carers or family if the patient wishes
- ensuring active and ongoing communication with the patient's GP.

4.5.4 Communication with the general practitioner

The general practitioner plays an important role in coordinating care for patients, including helping to manage side effects and other comorbidities, and offering support when patients have questions or worries. For most patients, simultaneous care provided by their general practitioner is very important.

The lead clinician, in discussion with the patient's general practitioner, should consider these points:

- the general practitioner's role in symptom management, supportive care and referral to local services
- using a chronic disease management plan and mental health care management plan
- how to ensure regular and timely two-way communication about:
 - the treatment plan, including intent and potential side effects
 - supportive and palliative care requirements
 - the patient's prognosis and their understanding of this
 - enrolment in research or clinical trials
 - changes in treatment or medications
 - the presence of an advance care directive or appointment of a substitute decision-maker
 - recommendations from the multidisciplinary team.



More information

Refer to Principle 6 'Communication' for communication skills training programs and resources.

Step 5: Care after initial treatment and recovery

The term ‘cancer survivor’ describes a person living with cancer, from the point of diagnosis until the end of life. For NETs this would include both patients with resected disease having ongoing follow-up, as well as residual disease who need ongoing treatment and monitoring. Survivorship care in Australia has traditionally been provided to patients who have completed active treatment and are in the post-treatment phase. But there is now a shift to provide survivorship care and services from the point of diagnosis to improve cancer-related outcomes.

Cancer survivors may experience inferior quality of life and cancer-related symptoms for up to five years after their diagnosis (Jefford et al. 2017). Distress, fear of cancer recurrence, fatigue, obesity and sedentary lifestyle are common symptoms reported by cancer survivors (Vardy et al. 2019).

Due to an ageing population and improvements in treatments and supportive care, the number of people surviving cancer is increasing. International research shows there is an important need to focus on helping cancer survivors cope with life beyond their acute treatment. Cancer survivors often face issues that are different from those experienced during active treatment for cancer and may include a range of issues, as well as unmet needs that affect their quality of life (Lisy et al. 2019; Tan et al. 2019).

Physical, emotional and psychological issues include fear of cancer recurrence, cancer-related fatigue, pain, distress, anxiety, depression, cognitive changes and sleep issues (Lisy et al. 2019). Late effects may occur months or years later and depend on the type of cancer treatment. Survivors and their carers may experience impacted relationships and practical issues including difficulties with return to work or study and financial hardship. They may also experience changes to sex and intimacy. Fertility, contraception and pregnancy care after treatment may require specialist input.

The Institute of Medicine, in its report *From cancer patient to cancer survivor: Lost in transition*, describes the essential components of survivorship care listed in the paragraph above, including interventions and surveillance mechanisms to manage the issues a cancer survivor may face (Hewitt et al. 2006). Access to a range of health professions may be required including physiotherapy, occupational therapy, social work, dietetics, clinical psychology, fertility and palliative care. Coordinating care between all providers is essential to ensure the patient’s needs are met.

Cancer survivors are more likely than the general population to have and/or develop comorbidities (Vijayvergia & Denlinger 2015). Health professionals should support survivors to self-manage their own health needs and to make informed decisions about lifestyle behaviours that promote wellness and improve their quality of life (Australian Cancer Survivorship Centre 2016; Cancer Australia 2017).

5.1 Transitioning from active treatment

The transition from active treatment to post-treatment care is critical to long-term health. In some cases, people will need ongoing, hospital-based care, and in other cases a shared follow-up care arrangement with their general practitioner may be appropriate. This will vary depending on the type and stage of cancer and needs to be planned.

Shared follow-up care involves the joint participation of specialists and general practitioners in the planned delivery of follow-up and survivorship care. A shared care plan is developed that outlines the responsibilities of members of the care team, the follow-up schedule, triggers for review, plans for rapid access into each setting and agreement regarding format, frequency and triggers for communication.

Different scenarios need to be defined in transitioning from treatment. This will include resected patients, as well as patients with stable disease and unstable disease. Treatment is often ongoing and sequential.

A designated member of the multidisciplinary team (most commonly nursing or medical staff involved in the patient's care) should provide the patient with a needs assessment and treatment summary and develop a survivorship care plan in conjunction with the patient. This should include a comprehensive list of issues identified by all members of the multidisciplinary team involved in the patient's care and by the patient. These documents are key resources for the patient and their healthcare providers and can be used to improve communication and care coordination.

The **treatment summary** should cover, but is not limited to:

- the diagnostic tests performed and results
- diagnosis including stage, prognostic or severity score
- tumour characteristics
- treatment received (types and dates)
- current toxicities (severity, management and expected outcomes)
- interventions and treatment plans from other health providers
- potential long-term and late effects of treatment
- supportive care services provided
- follow-up schedule
- contact information for key healthcare providers.

5.2 Follow-up care

Patients with NETs may need multiple courses of therapy over their lifetime, given the relapsing-remitting nature of NETs. Monitoring for late effects and second malignancies is required, and primary prevention and risk minimisation strategies are encouraged.

Specific monitoring for sequelae of therapy should include:

- after surgery resulting in short gut syndrome – dietary intervention is required due to malabsorption
- after and/or during continuous systemic treatment – review and physical examination every three to six months for two years, and every six to 12 months subsequently (ESMO 2014; Knigge et al. 2017)
- follow-up echocardiography annually for patients who have carcinoid heart disease (they have an increased risk of cardiac valve dysfunction)
- after PRRT monitoring of renal function and risk of myelodysplastic syndrome
- monitoring for recurrence after surgery (Singh et al. 2018) – there are no specific follow-up guidelines because resected GEP-NETs are complex and emphasise closer follow-up for the first three years; however, it is recommended for follow-up to be for up to 10 years for fully resected small-bowel and pNETs and also identify clinical situations where follow-up is not required.

Responsibility for follow-up care should be agreed between the lead clinician, the general practitioner, relevant members of the multidisciplinary team and the patient. This is based on guideline recommendations for post-treatment care, as well as the patient's current and anticipated physical and emotional needs and preferences.

Evidence comparing shared follow-up care and specialised care indicates equivalence in outcomes including recurrence rate, cancer survival and quality of life (Cancer Research in Primary Care 2016).

Ongoing communication between healthcare providers involved in care and a clear understanding of roles and responsibilities is key to effective survivorship care.

In particular circumstances, other models of post-treatment care can be safely and effectively provided such as nurse-led models of care (Monterosso et al. 2019). Other models of post-treatment care can be provided in these locations or by these health professionals:

- in a shared care setting
- in a general practice setting
- by non-medical staff
- by allied health or nurses
- in a non-face-to-face setting (e.g. by telehealth).

A designated member of the team should document the agreed survivorship care plan. The survivorship care plan should support wellness and have a strong emphasis on healthy lifestyle changes such as a balanced diet, a non-sedentary lifestyle, weight management and a mix of aerobic and resistance exercise (COSA 2018; Hayes et al. 2019).

This survivorship care plan should also cover, but is not limited to:

- what medical follow-up is required (surveillance for recurrence or secondary and metachronous cancers, screening and assessment for medical and psychosocial effects)
- model of post-treatment care, the health professional providing care and where it will be delivered
- care plans from other health providers to manage the consequences of cancer and cancer treatment
- wellbeing, primary and secondary prevention health recommendations that align with chronic disease management principles
- rehabilitation recommendations
- available support services
- a process for rapid re-entry to specialist medical services for suspected recurrence.

Prognosis

Many patients have a promising prognosis from low-grade NETs. Patients may need follow-up over a long period to monitor for progression or recurrence. Patients with incurable low-grade (grade 1) disease often survive for many (10+) years (NECA 2019a).

In contrast, patients with high-grade (grade 3) disease (high tumour burden that is aggressive) have poor progression-free survival, with an average survival time measured in the range of many months to a few years, despite best treatment (NECA 2019a).

There is a big variation in prognosis – there is a lot of variation in outcomes and no ‘magic number’ for a particular patient. Some patients find discussion of ranges in prognosis (best case / worse case / expected scenarios) very helpful, while others do not (NECA 2019a).

Prognoses based on the available information are a rough estimate because new treatments and insights can improve care and hence prognosis for all NET patients (NECA 2019a).

Table 2 provides a general guide for evaluating prognostic factors and outcomes for patients with NETs.

Table 2: Prognostic factors and outcomes for patients with neuroendocrine tumours

Factor	Good prognosis	Poor prognosis
Disease resectability	Resected	Metastatic
Site	Appendiceal/rectal	Pancreas
Grade	Low-grade (e.g. G1)	High-grade (e.g. G3)
Nuclear imaging	Significant 68Ga-DOTATATE PET based avidity, lack of 18F-FDG PET/CT avidity	Significant 18F-FDG PET avidity, lack of 68Ga-DOTATATE PET/CT avidity
Disease course	Well controlled on initial therapies	Short duration of control on initial therapies
Performance status	Good	Bad
Disease volume / number of sites	Low	High

Survivors generally need regular follow-up, often for at least 10 years for most resected NETs and lifelong for those with incurable disease. The survivorship care plan therefore may need to be updated to reflect changes in the patient’s clinical and psychosocial status and needs.

Processes for rapid re-entry to hospital care should be documented and communicated to the patient and relevant stakeholders.

Care in the post-treatment phase is driven by predicted risks (e.g. the risk of recurrence, developing late effects of treatment and psychological issues) as well as individual clinical and supportive care needs. It is important that post-treatment care is based on evidence and is consistent with guidelines. Not all people will require ongoing tests or clinical review and may be discharged to general practice follow-up.

The lead clinician should discuss (and general practitioner reinforce) options for follow-up at the start and end of treatment. It is critical for optimal aftercare that the designated member of the treatment team educates the patient about the symptoms of recurrence.

General practitioners (including nurses) can:

- connect patients to local community services and programs
- manage long-term and late effects
- manage comorbidities
- provide wellbeing information and advice to promote self-management
- screen for cancer and non-cancerous conditions.



More information

Templates and other resources to help with developing treatment summaries and survivorship care plans are available from these organisations:

- Australian Cancer Survivorship Centre
- Cancer Australia – Principles of Cancer Survivorship
- Cancer Council Australia and states and territories
- Clinical Oncology Society of Australia – Model of Survivorship Care
- eviQ – Cancer survivorship: introductory course
- mycareplan.org.au
- South Australian Cancer Service – Statewide Survivorship Framework resources
- American Society of Clinical Oncology – guidelines.

5.2.1 Preventing recurrence

Not smoking, eating a healthy diet, being sun smart, avoiding or limiting alcohol intake, being physically active and maintaining a healthy body weight may help health in general and potentially reduce the risk of other cancers, although there is relatively little evidence in NETs.

Encourage and support all cancer survivors to reduce modifiable risk factors for recurrence as well as other chronic diseases. Ongoing coordination of care between providers should also deal with any comorbidities, particularly ongoing complex and life-threatening comorbid conditions.

5.3 Research and clinical trials

Comparative data from randomised controlled trials are lacking in NETs. Therefore, treatment decisions are based on phase II trial data and patient/physician preferences. However, increasing active research and prospective trials are becoming available.

Support cancer survivors to participate in research or clinical trials where they are available and appropriate. These might include studies to understand survivors' issues, to better manage treatment side effects, or to improve models of care and quality of life.

For more information visit the Cancer Australia website <www.australiancancertrials.gov.au>.

5.4 Support and communication

5.4.1 Supportive care

Patients with NETs may experience multiple lines of treatment over many years and therefore benefit from reassessment of their supportive care needs:

- on an ongoing basis during monitoring
- prior to each subsequent line of therapy.

See validated screening tools mentioned in Principle 4 'Supportive care'. Additionally, the 'Cancer Survivors Unmet Needs (CaSun)' is another screening tool that may help health professionals to identify the unmet needs of NETs patients during survivorship.

A number of specific challenges and needs may arise for cancer survivors:

- management of symptoms such as fatigue, muscle weakness, intermittent abdominal pain, diarrhoea, skin rashes, headaches, anxiety and depression – a working relationship with the patient's NET specialist is essential to manage side effects and guide treatment
- nutritional needs and guidance for dietary intake
- feelings of a 'stolen' identity – the patient's identity being replaced with a focus on the disease – consider referral to a psychologist and NeuroEndocrine Cancer Australia's guide for health professionals (NECA 2019b)
- financial and employment issues (e.g. loss of income and assistance with returning to work, and the cost of treatment, travel and accommodation)
- appointing a substitute decision-maker and completing an advance care directive
- legal issues such as completing a will.

For more information on supportive care and needs that may arise for different population groups, see Appendices A, B and C.

5.4.2 Rehabilitation and recovery

Rehabilitation may be required at any point of the care pathway from the pre-treatment phase through to disease-free survival and palliative care (Cormie et al. 2017).

Issues that may need to be dealt with include managing cancer-related fatigue, coping with cognitive changes, improving physical endurance, achieving independence in daily tasks, returning to study or work and ongoing adjustment to cancer and its sequels.

Exercise is a safe and effective intervention that improves the physical and emotional health and wellbeing of cancer patients. Exercise should be embedded as part of standard practice in cancer care and be viewed as an adjunct therapy that helps counteract the adverse effects of cancer and its treatment.

Cancer survivors may find referral to specific cancer rehabilitation, optimisation programs or community-based rehabilitation appropriate and beneficial. Other options include referral to allied health supports through team care arrangements and mental health plans. Some community support organisations (cancer-related non-government, not-for-profit and charities) provide services to cancer survivors.

5.4.3 Communication with patients, carers and families

The lead clinician (themselves or by delegation) should take responsibility for these tasks:

- explaining the model of post-treatment care and the roles of health professionals involved in post-treatment care including the role of general practice
- explaining the treatment summary and follow-up care plan
- discussing the development of a shared follow-up and survivorship care plan where a model of shared follow-up care has been agreed
- discussing how to manage any of the physical, psychological or emotional issues identified
- providing information on the signs and symptoms of recurrent disease
- providing a survivorship care plan with information on secondary prevention and healthy living
- providing contact details of the care team involved
- providing clear information about the role and benefits of palliative care and advance care planning.

5.4.4 Communication with the general practitioner

The lead clinician should ensure regular, timely, two-way communication with the general practitioner about:

- the patient's progress
- MDM outcomes
- the follow-up care plan
- potential late effects
- supportive and palliative care requirements
- any shared care arrangements
- clarification of various roles in patient care
- a process for rapid re-entry to medical services for patients with suspected recurrence or if there are other concerns.



More information

Refer to Principle 6 'Communication' for communication skills training programs and resources.

Step 6: Managing recurrent, residual or metastatic disease

Patients who present with recurrent, residual or metastatic disease should be managed by a multidisciplinary team and offered timely referral to appropriate physical, practical and emotional support.

Step 6 is concerned with managing recurrent or local residual and metastatic disease. The likelihood of recurrence depends on many factors usually related to the type of cancer, the stage of cancer at presentation and the effectiveness of treatment. Some cancers cannot be eradicated even with the best initial treatment. But controlling disease and disease-related symptoms is often possible, depending on the clinical situation.

6.1 Signs and symptoms of metastatic disease

Some patients will have metastatic disease on initial presentation. Others may present with symptoms of recurrent disease after a previous cancer diagnosis. Access to the best available therapies, including clinical trials, as well as treatment overseen by a multidisciplinary team, are crucial to achieving the best outcomes for anyone with metastatic disease.

Signs and symptoms will depend on the type of cancer initially diagnosed and the location of metastatic disease. They may be discovered by the patient or by surveillance in the post-treatment period. New symptoms or changes in symptoms may include (Cancer.Net 2019):

- increase in facial flushing
- increased diarrhoea
- wheezing
- heart problems such as palpitations, difficulty breathing, becoming easily tired
- increased abdominal pain
- bloating
- bone pain.

6.2 Managing metastatic disease

In a suspected recurrence or progression, re-staging of relapsed or refractory disease should be performed with:

- bloods
- 24-hour urinary 5HIAA if carcinoid syndrome
- contrast CT of chest/abdomen/pelvis or MRI
- Ga-68-DOTATATE PET/CT (or similar)
- FDG PET/CT.

Managing metastatic disease is complex and should therefore involve all the appropriate specialties in a multidisciplinary team including palliative care where appropriate. From the time of diagnosis, the team should offer patients appropriate psychosocial care, supportive care, advance care planning and symptom-related interventions as part of their routine care. The approach should be personalised to meet the patient's individual needs, values and preferences. The full complement of supportive care measures as described throughout the optimal care pathway and in Appendices A, B and C should be offered to assist patients and their families and carers to cope. These measures should be updated as the patient's circumstances change.

Survivorship care should be considered and offered at an early stage. Many people live with advanced cancer for many months or years. As survival is improving in many patients, survivorship issues should be considered as part of routine care. Health professionals should therefore be ready to change and adapt treatment strategies according to disease status, prior treatment tolerance and toxicities and the patient's quality of life, in addition to the patient's priorities and life plans.

6.3 Multidisciplinary team

If there is an indication that a patient's cancer has returned or progressed, care should be provided under the guidance of a treating specialist. Each patient should be referred back to the original multidisciplinary team. The multidisciplinary team may include new members such as palliative care specialists.

6.4 Treatment

Treatment will depend on the location, extent of recurrent or residual disease, previous management and the patient's preferences.

In managing people with NETs, treatment may include these options (refer to Step 4 for further detail):

- change in SSA – regimen and/or dose
- PRRT
- localised progression – consider liver-directed therapy and radiation therapy for bone metastasis
- widespread and systemic recurrence – consider systemic therapy – chemotherapy, targeted therapies, immunotherapy (remains investigational)
- pre-screen patient for any currently recruiting or imminent clinical trial
- palliation.

The potential goals of treatment should be discussed, respecting the patient's cultural values. Wherever possible, written information should be provided.

Encourage early referral to clinical trials or accepting an invitation to participate in research.

6.5 Associated conditions

Carcinoid syndrome

When GEP-NETs metastasise, the most common site for metastatic tumours ('secondaries') is the liver. Other areas of spread can include the bones, the lungs and the lymphatic system. Many metastatic NETs can cause symptoms due to over-production of hormones. The most common is carcinoid syndrome, which can occur in approximately 10 per cent of patients and is caused when an excess of hormones such as serotonin, histamine and somatostatin are produced (NORD 2021). The symptoms of carcinoid syndrome vary and can often be highly individual.

Typical symptoms include (NECA 2020a):

- flushing
- diarrhoea
- faecal urgency
- wheezing
- abdominal pain
- pellagra (niacin deficiency). Although rare, pellagra presents as a rash, dark pigmentation on the skin, a swollen mouth and a bright red tongue. Patients can experience vomiting and diarrhoea, headache, fatigue, depression, disorientation, confusion or memory loss.

Carcinoid crisis

Sometimes patients with functioning NETs may suffer a particularly severe episode of carcinoid syndrome triggered by stressors, general anaesthetic or treatments. Early recognition of a carcinoid crisis enables preventative measures. Symptoms include (NECA 2020a):

- intense flushing
- diarrhoea
- abdominal pain
- wheezing
- palpitations
- low or high blood pressure
- altered mental state and, in extreme cases, coma.

Without treatment, the complications can be life-threatening, but if the patient is having any procedures the NET specialist should ensure the patient is comprehensively monitored and may give an infusion of an SSA (octreotide) as a preventative measure as per institutions policy.

NeuroEndocrine Cancer Australia provides a fact sheet named Carcinoid Crisis and Anaesthesia <<https://neuroendocrine.org.au/other-resources/>>, and a wallet card for patients <<https://neuroendocrine.org.au/other-resources/>>, which gives advice on treatment in the event of carcinoid crisis.

Carcinoid heart disease

Up to 20 per cent of patients with carcinoid syndrome present with carcinoid heart disease and, without treatment, can develop right heart failure (NECA 2020b). With SSAs, the progression of carcinoid heart disease is significantly slowed and other symptoms of heart failure may be managed with diuretics. Some patients with carcinoid heart disease may be suitable for cardiac surgery to replace the leaking valves (NECA 2020b). ECG and chest x-ray may provide clues to the diagnosis of carcinoid heart disease, but the most sensitive test is echocardiography of the heart. Echocardiography should be performed regularly to monitor the function of the heart in patients with functional neuroendocrine cancers (NECA 2020b). Urinary 5HIAA is a useful companion biomarker that measures serotonin excretion and also ProBNP (Grozinsky-Glasberg et al. 2015). While early valvular replacement can be complex, this should be managed in a specialist centre with experience in managing carcinoid valvular disease.

6.6 Advance care planning

Advance care planning is important for all patients with a cancer diagnosis but especially those with advanced disease. Patients should be encouraged to think and talk about their healthcare values and preferences with family or carers, appoint a substitute decision-maker and consider developing an advance care directive to convey their preferences for future health care in the event they become unable to communicate their wishes (AGDH 2021).



More information

Refer to section 4.3 'More information' for links to resources.

Refer patients and carers to Advance Care Planning Australia <www.advancecareplanning.org.au> or to the Advance Care Planning National Phone Advisory Service on 1300 208 582.

6.7 Palliative care

Early referral to palliative care can improve the quality of life for people with cancer and in some cases may be associated with survival benefits (Haines 2011; Temel et al. 2010; Zimmermann et al. 2014). The treatment team should emphasise the value of palliative care in improving symptom management and quality of life to patients and their carers. Refer to section 4.3 for more detailed information.

The lead clinician should ensure timely and appropriate referral to palliative care services. Referral to palliative care services should be based on the patient's need and potential for benefit, not prognosis.



More information

Refer to the end of section 4.3 'Palliative care' for links to resources.

6.8 Research and clinical trials

The treatment team should support the patient to participate in research and clinical trials where available and appropriate.

For more information visit the Cancer Australia website <www.australiancancertrials.gov.au>.

6.9 Support and communication

6.9.1 Supportive care

See validated screening tools mentioned in Principle 4 'Supportive care'.

A number of specific challenges and needs may arise at this time for patients:

- assistance for dealing with emotional and psychological distress resulting from fear of death or dying, existential concerns, anticipatory grief, communicating wishes to loved ones, interpersonal problems and sexuality concerns
- potential isolation from normal support networks, particularly for rural patients who are staying away from home for treatment
- cognitive changes as a result of treatment and disease progression such as altered memory, attention and concentration (a patient may appoint someone to make medical, financial and legal decisions on their behalf – a substitute decision-maker – before and in case they experience cognitive decline)
- management of physical symptoms including dietary issues (reduction in food intake and avoidance of food types due to symptoms)
- decline in mobility or functional status as a result of recurrent disease and treatments (referral to physiotherapy or occupational therapy may be required)
- coping with hair loss and changes in physical appearance (refer to the Look Good, Feel Better program – see 'Resource list')
- appointing a substitute decision-maker and completing an advance care directive
- financial issues as a result of disease recurrence such as gaining early access to superannuation and insurance
- legal issues (completing a will, care of dependent children) and making an insurance, superannuation or social security claim on the basis of terminal illness or permanent disability.

6.9.2 Rehabilitation

Rehabilitation may be required at any point of the metastatic care pathway, from preparing for treatment through to palliative care. Issues that may need to be dealt with include managing cancer-related fatigue, improving physical endurance, achieving independence in daily tasks, returning to work and ongoing adjustment to cancer and its sequels.

Optimising nutrition (maintaining a healthy weight and food intake/diet) during and post-treatment is important to avoid further complications such as declining nutritional status (malnutrition), negative social impacts and reduced quality of life. Patients with a NET often change their diet due to NET symptoms and therefore require support and education to ensure they are meeting their dietary needs. Referral to a dietitian for assessment and nutrition support should be considered for symptomatic patients and those reporting weight loss or a change in their diet.

Exercise is a safe and effective intervention that improves the physical and emotional health and wellbeing of cancer patients. Exercise should be embedded as part of standard practice in cancer care and be viewed as an adjunct therapy that helps counteract the adverse effects of cancer and its treatment.

6.9.3 Communication with patients, carers and families

The lead clinician should ensure there is adequate discussion with patients and carers about the diagnosis and recommended treatment, including treatment intent and possible outcomes, likely adverse effects, prognosis (refer to step 5) and the supportive care options available.



More information

Refer to Principle 6 'Communication' for communication skills training programs and resources.

Step 7: End-of-life care

Step 7 is concerned with maintaining the patient's quality of life and meeting their health and supportive care needs as they approach the end of life, as well as the needs of their family and carers.

Some patients with advanced cancer will reach a time when active treatment is no longer appropriate. The team needs to share the principles of a palliative approach to care when making decisions with the patient and their family or carer. End-of-life care is appropriate when the patient's symptoms are increasing and functional status is declining.

7.1 Multidisciplinary palliative care

If the treatment team does not include a palliative care member, the lead clinician should consider referring the patient to palliative care services, with the general practitioner's engagement. This may include inpatient palliative unit access (as required).

The multidisciplinary team may consider seeking additional expertise from these professionals:

- clinical psychologist
- clinical nurse specialist or practitioner
- social worker
- palliative medicine specialist
- pain specialist
- pastoral or spiritual carer
- bereavement counsellor
- music therapist
- art therapist
- cultural expert
- Canteen for children of parents with cancer <www.canteen.org.au/young-people/my-parent-has-cancer/>.

The team might also recommend that patients access these services:

- home and community-based care
- specialist community palliative care workers
- community nursing.

If the patient does not already have an advance care directive in place, a designated member of the treatment team should encourage them to develop one in collaboration with their family or carer (AGDH 2021).

It is essential for the treatment team to consider the appropriate place of care, the patient's preferred place of death and the support needed for the patient, their family and carers.

The treatment team should also ensure that carers and families receive the information, support and guidance about their role according to their needs and wishes (Palliative Care Australia 2018).



More information

The treatment team can refer patients and carers to these resources:

- Palliative Care Australia <www.palliativecare.org.au>
- Advance Care Planning Australia <www.advancecareplanning.org.au> or to Advance Care Planning Australia's National Advisory Service on 1300 208 582.

7.2 Research and clinical trials

Clinical trials may help improve palliative care and in managing a patient's symptoms of advanced cancer (Cancer Council Victoria 2019). The treatment team should support the patient to participate in research and clinical trials where available and appropriate.

For more information visit the Cancer Australia website <www.australiancancertrials.gov.au>. See 'Resource list' for additional clinical trial databases.

7.3 Support and communication

7.3.1 Supportive care

See validated screening tools mentioned in Principle 4 'Supportive care'.

A number of specific challenges and needs may arise for patients at this time:

- assistance for dealing with emotional and psychological distress from anticipatory grief, fear of death or dying, anxiety/depression and interpersonal problems
- management of physical symptoms associated with carcinoid syndrome
- decline in mobility or functional status, affecting the patient's discharge destination (a referral to physiotherapy, exercise physiology, occupational therapy or social work may be needed)
- appointing a substitute decision-maker and completing an advance care directive
- legal issues (completing a will, care of dependent children) and making an insurance, superannuation or social security claim on the basis of terminal illness or permanent disability
- specific support for families where a parent is dying and will leave behind bereaved children or adolescents, creating special family needs
- arranging a funeral.

These services and resources can help:

- referral to 13 11 20 for Cancer Council Australia's Pro Bono Program for free legal, financial, small business accounting and workplace assistance (subject to a means test)
- *Sad news sorry business* (Queensland Health 2015) for the specific needs of Aboriginal and Torres Strait Islander people.

For more information on supportive care and needs that may arise for different population groups, see Appendices A, B and C.

7.3.2 Communication with patients, carers and families

The lead clinician is responsible for:

- being open to and encouraging discussion with the patient about the expected disease course, considering the patient's personal and cultural beliefs and expectations
- discussing palliative care options, including inpatient and community-based services as well as dying at home and subsequent arrangements
- providing the patient and carer with the contact details of a palliative care service
- referring the patient to palliative care in the community according to the carer's wishes.

7.3.3 Communication with the general practitioner

The lead clinician should discuss end-of-life care planning to ensure the patient's needs and goals are met in the appropriate environment. The patient's general practitioner should be kept fully informed and involved in major developments in the patient's illness path.

<insert icon> **More information**

For support with communication skills and training programs, see these sources:

- Sad news sorry business <www.health.qld.gov.au/__data/assets/pdf_file/0023/151736/sorry_business.pdf>
- Principle 6 'Communication'.

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Our thanks to the following health professionals, consumer representatives, stakeholders and organisations consulted in developing this optimal care pathway.

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Advance Care Planning Australia

Allied Health Professions Australia

Australasian Association of Nuclear Medicine Specialists

Australasian Chapter of Palliative Medicine, Royal Australia College of Physicians

Australasian Gastro-Intestinal Trials Group

Australian and New Zealand Society of Neuroradiology

Australian and New Zealand Society of Palliative Care

Australian Cancer Survivorship Centre

Australian College of Nursing

Australian Medical Association

Australian Society of Medical Imaging and Radiation Therapy

Cancer Nurses Society of Australia

Clinical Oncology Society of Australia – Gastrointestinal Cancers Group & Neuroendocrine Tumours Group

Commonwealth Neuroendocrine Tumour Research Collaborative (CommNETs)

Gastroenterological Society of Australia

Interventional Radiology Society of Australasia

Medical Oncology Group of Australia

NeuroEndocrine Cancer Australia

Oncology Social Workers Australia and New Zealand

Royal Australasian College of Physicians

Royal Australasian College of Surgeons

Royal Australian and New Zealand College of Radiologists

Royal Australian College of General Practitioners

Royal College of Pathologists of Australasia

Governance – project steering committee representation

Cancer Australia

Cancer Council Victoria

Other stakeholders consulted to provide feedback include relevant Cancer Council committees and networks, Integrated Cancer Services, Primary Health Networks and several health services.

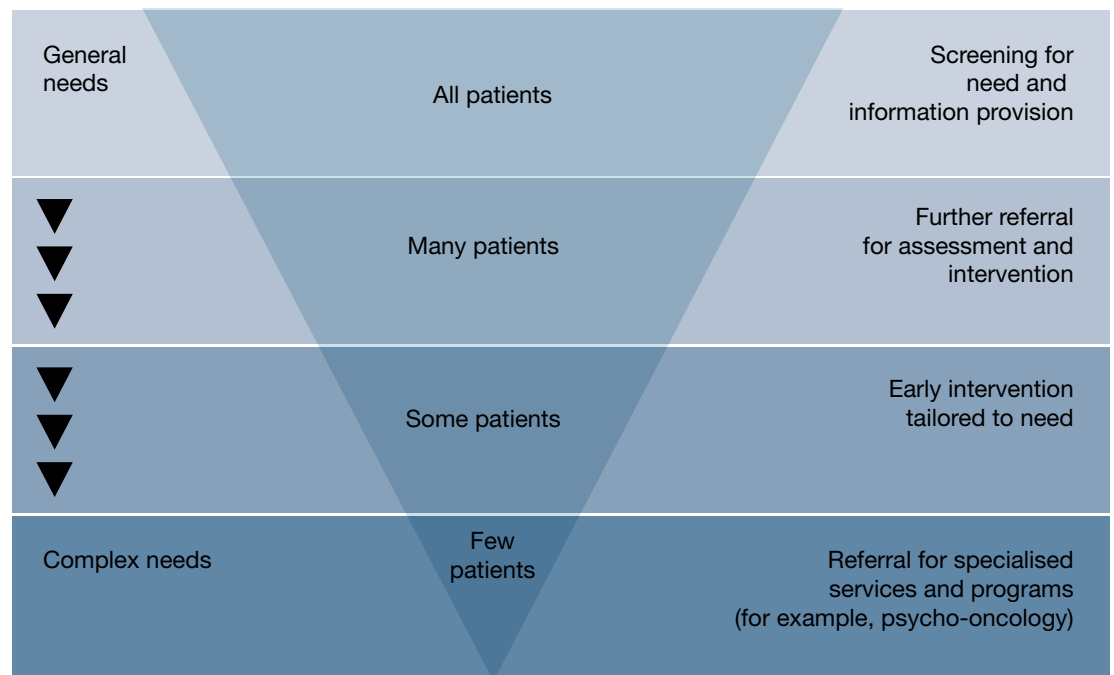
Appendix A: Supportive care domains

Supportive care in cancer refers to the following five domains:

- the physical domain, which includes a wide range of physical symptoms that may be acute, relatively short lived or ongoing, requiring continuing interventions or rehabilitation
- the psychological domain, which includes a range of issues related to the patient's mental health wellbeing and personal relationships
- the social domain, which includes a range of social and practical issues that will affect the patient, carer and family such as the need for emotional support, maintaining social networks and financial concerns
- the information domain, which includes access to information about cancer and its treatment, recovery and survivorship support services and the health system overall
- the spiritual domain, which focuses on the patient's changing sense of self and challenges to their underlying beliefs and existential concerns (Palliative Care Victoria 2019).

Fitch's (2000) model of supportive care recognises the variety and level of intervention required at each critical point as well as the need to be specific to the individual patient (Figure A1). The model targets the type and level of intervention required to meet patients' supportive care needs.

Figure A1: Fitch's tiered approach to supportive care



Appendix B: Psychological needs

Consider a referral to a psychologist, psychiatrist, pastoral/spiritual care practitioner, social worker, specialist nurse or a relevant community-based program if the patient has these issues:

- displaying emotional cues such as tearfulness, distress that requires specialist intervention, avoidance or withdrawal
- being preoccupied with or dwelling on thoughts about cancer and death
- displaying fears about the treatment process or the changed goals of their treatment
- displaying excessive fears about cancer progression or recurrence
- worrying about loss associated with their daily function, dependence on others and loss of dignity
- becoming isolated from family and friends and withdrawing from company and activities that they previously enjoyed
- feeling hopeless and helpless about the effect that cancer is having on their life and the disruption to their life plans
- struggling to communicate with family and loved ones about the implications of their cancer diagnosis and treatment
- experiencing changes in sexual intimacy, libido and function
- struggling with the diagnosis of metastatic or advanced disease
- having difficulties quitting smoking (refer to Quitline on 13 7848) or with other drug and alcohol use
- having difficulties transitioning to palliative care.

Additional considerations that may arise for the multidisciplinary team include:

- support for the carer – encourage referrals to psychosocial support from a social worker, psychologist or general practitioner
- referral to an exercise physiologist or physiotherapist as a therapeutic approach to prevent and manage psychological health
- referral to wellness-after-cancer programs to provide support, information and offer strategies.

Appendix C: Special population groups

The burden of cancer is not evenly spread across Australia. People experiencing socioeconomic disadvantage, Aboriginal and Torres Strait Islander communities, culturally diverse communities, people living with a disability, people with chronic mental health or psychiatric concerns and those who live in regional and rural areas of Australia have poorer cancer outcomes.

Aboriginal and Torres Strait Islander people

Cancer is the third leading cause of burden of disease for Aboriginal and Torres Strait Islander people. While Australia's cancer survival rates are among the best in the world, Aboriginal and Torres Strait Islander people continue to experience a different pattern of cancer incidence and significant disparities in cancer outcomes compared with non-Indigenous Australians.

For Aboriginal and Torres Strait Islander people, health and connection to land, culture, community and identity are intrinsically linked. Health encompasses a whole-of-life view and includes a cyclical concept of life–death–life.

The distinct epidemiology of cancer among Aboriginal and Torres Strait Islander people, and unique connection to culture, highlight the need for a specific optimal care pathway for Aboriginal and Torres Strait Islander people with cancer. Ensuring this pathway is culturally safe and supportive is vital to tackling the disparities for Aboriginal and Torres Strait Islander people.

Published in 2018, the *Optimal care pathway for Aboriginal and Torres Strait Islander people with cancer* provides guidance to health practitioners and service planners on optimal care for Aboriginal and Torres Strait Islander people with cancer across the cancer continuum.

In addition to the key principles underpinning cancer-specific pathways, these are the key concepts that are fundamental to Aboriginal and Torres Strait Islander health:

- providing a holistic approach to health and wellbeing
- providing a culturally appropriate and culturally safe service
- acknowledging the diversity of Aboriginal and Torres Strait Islander peoples
- understanding the social determinants and cultural determinants of health (Cancer Australia 2015).

To view the *Optimal care pathway for Aboriginal and Torres Strait Islander people with cancer*, visit the Cancer Australia website <<https://www.canceraustralia.gov.au/affected-cancer/atsi/resources-health>>. To view the consumer resources – Checking for cancer and Cancer, visit the Cancer Australia website <<https://www.canceraustralia.gov.au/affected-cancer/atsi/resources-people>>.

Culturally diverse communities

For people from culturally diverse backgrounds in Australia, a cancer diagnosis can come with additional complexities, particularly when English proficiency is poor. In many languages there is not a direct translation of the word 'cancer', which can make communicating vital information difficult. Perceptions of cancer and related issues can differ greatly in people from culturally diverse backgrounds and this can affect their understanding and decision making after a cancer diagnosis. In addition to different cultural beliefs, when English language is limited there is potential for miscommunication of important information and advice, which can lead to increased stress and anxiety for patients.

A professionally trained interpreter (not a family member or friend) should be made available when communicating with people with limited English proficiency. Navigation of the Australian healthcare system can pose problems for those with a non-Anglo culture, and members of the treatment teams should pay particular attention to supporting these patients.

The Australian Cancer Survivorship Centre has developed a glossary of more than 700 cancer terms in nine different languages. The multilingual glossary has been designed as a resource for professional translators, interpreters and bilingual health professionals working in the cancer field. The glossary is a unique tool that enables language professionals with access to accurate, consistent and culturally appropriate terminology.

Visit the Peter Mac website <www.petermac.org/multilingualglossary> to see the glossary.

People with disabilities

Disability, which can be physical, intellectual or psychological, may have existed before the cancer diagnosis or may be new in onset (occurring due to the cancer treatment or incidentally). Adjusting to life with a disability adds another challenge to cancer care and survivorship.

Several barriers prevent people with disabilities from accessing timely and effective health care (AIHW 2017):

- physical limitations
- competing health needs
- the trauma of undergoing invasive procedures
- potential barriers associated with obtaining informed consent
- failure to provide assistance with communication
- lack of information
- discriminatory attitudes among healthcare staff.

In caring for people with disabilities and a cancer diagnosis, the Australian Institute of Health and Welfare disability flag should be used at the point of admittance to correctly identify and meet the additional requirements of a person with disability. Facilities should actively consider access requirements, and health practitioners should make reasonable adjustments where required.

Patients aged between seven and 65 years who have a permanent or significant disability may be eligible for support or funding through the National Disability Insurance Scheme (National Disability Insurance Agency 2018). More information can be found on the NDIS website <www.ndis.gov.au>.

Patients aged 65 years or older (50 years or older for Aboriginal or Torres Strait Islander people) may be eligible for subsidised support and services through aged care services. An application to determine eligibility can be completed online over the phone. More information can be found at the My Aged Care website <www.myagedcare.gov.au>.



More information

'Talking End of Life' is a resource that shows how to teach people with intellectual disability about end of life. It is designed for disability support workers but is also helpful for others including families, health professionals and educators.

To view the resource, visit the Talking End of Life website <www.caresearch.com.au/tel/tabid/4881/Default.aspx>.

Older people with cancer

Planning and delivering appropriate cancer care for older people can present a number of challenges. This could also be true for frail people or those experiencing comorbidities. Effective communication between oncology and geriatrics departments will help facilitate best practice care, which takes into account physiological age, complex comorbidities, risk of adverse events and drug interactions, as well as the implications of cognitive impairment on suitability of treatment and consent (Steer et al. 2009).

At a national interdisciplinary workshop convened by the Clinical Oncology Society of Australia, it was recommended that people over the age of 70 undergo some form of geriatric assessment, in line with international guidelines (COSA 2013; palliAGED 2018). Screening tools can be used to identify those patients in need of a comprehensive geriatric assessment (Decoster et al. 2015). This assessment can be used to help determine life expectancy and treatment tolerance and guide appropriate referral for multidisciplinary intervention that may improve outcomes (Wildiers et al. 2014).

Frailty is not captured through traditional measures of performance status (e.g. ECOG) and includes assessment in the domains of:

- function
- comorbidity
- presence of geriatric syndromes
- nutrition
- polypharmacy
- cognition
- emotional status
- social supports.

Adolescents and young adults

In recent years, adolescent and young adult oncology has emerged as a distinct field due to lack of progress in survival and quality-of-life outcomes (Ferrari et al. 2010; Smith et al. 2013). The significant developmental change that occurs during this life stage complicates a diagnosis of cancer, often leading to unique physical, social and emotional effects for young people at the time of diagnosis and throughout the cancer journey (Smith et al. 2012).

In caring for young people with cancer, akin to the comorbidities that require specific care in the older cancer population, the treatment team needs to pay careful attention to promoting normal development (COSA 2014). This requires personalised assessments and management involving a multidisciplinary, disease-specific, developmentally targeted approach that adheres to the following principles:

- understanding the developmental stages of adolescence and supporting normal adolescent health and development alongside cancer management
- understanding and supporting the rights of young people
- communication skills and information delivery that are appropriate to the young person
- meeting the needs of all involved, including the young person, their carers and their family
- working with educational institutions and workplaces
- considering survivorship and palliative care needs.

An oncology team caring for an adolescent or young adult with cancer should be able to demonstrate these specific areas of expertise:

- be able to ensure access to expert adolescent and young adult health providers who have knowledge specific to the biomedical and psychosocial needs of the population
- understand the biology and current management of the disease in the adolescent and young adult age group
- consider participating in research and clinical trials for each patient
- engage in proactive discussion and management of fertility preservation, late effects of treatment, ongoing need for contraception, and psychosocial and psychosexual needs
- provide treatment in an environment that is friendly to adolescents and young adults.

People experiencing socioeconomic disadvantage

In general, people from lower socioeconomic groups are at greater risk of poor health, have higher rates of illness, disability and death, and live shorter lives than those from higher socioeconomic groups (AIHW 2016). People experiencing socioeconomic disadvantage are less likely to participate in screening programs, more likely to be obese, less likely to exercise and much more likely to smoke, which are all risk factors for cancer. In 2010–2014 age-standardised cancer incidence rates were higher in the lowest socioeconomic areas compared with the highest socioeconomic areas for all cancers combined (Cancer Australia 2019b).

Socioeconomic status and low health literacy are closely correlated. Therefore, effective communication with patients and carers is particularly important given the prevalence of low health literacy in Australia (estimated at 60 per cent of Australian adults) (ACSQHC 2014).

Consideration should be taken for cancer patients experiencing socioeconomic disadvantage to reduce their risk of being underserved for health care.

People with chronic mental health or psychiatric concerns

A diagnosis of cancer may present additional challenges to people who have pre-existing chronic mental health or psychiatric concerns, resulting in exacerbation of their mental health symptoms. This may include heightened anxiety, worsening depression or thoughts of self-harm.

As poor adjustment and coping can affect treatment decisions, people who are known to have a mental health diagnosis need psychosocial assessment in the oncology setting to formulate a plan for ongoing support throughout treatment.

Psychosocial support can assist with challenges in communicating with health professionals, enhance understanding of the treatment journey, ensure capacity for consent to treatment options and improve compliance with treatment requests. A referral for psychosocial support from a health professional to the psycho-oncology team can ensure these patients are provided with targeted interventions or referrals to community-based services that may mitigate problems associated with the impacts of social isolation that frequently accompany chronic mental ill-health.

Many patients with chronic mental health problems may be well known to external service providers. Psycho-oncology health professionals can form meaningful partnerships with existing service providers to optimise patient care throughout treatment and beyond.

Drug use disorders fall within the area of mental health conditions. People who are opiate dependent may have specific and individual requirements regarding pain management and their own preference for type of opiate prescribed or used.

Sexually and gender diverse groups

People who identify as sexually or gender diverse may have unique needs following a cancer diagnosis. Sexually or gender diverse identities include (but are not limited to) people who identify as lesbian, gay, bisexual or transgender, collectively 'LGBT'. There is no universally agreed upon initialism to describe this community, with other terms such as queer/questioning (Q), intersex (I), asexual (A) and pansexual (P) often included, as well as a plus symbol (+) indicating inclusivity of other identities not explicitly mentioned.

Sexual orientation and gender identity are relevant across the entire spectrum of cancer care, from prevention to survivorship and end-of-life care. LGBT people are less likely to participate in cancer screening, and some segments of the LGBT community exhibit elevated rates of specific cancer risk factors – for example, higher rates of smoking and alcohol use. Regarding treatment, there may be unique factors relevant to LGBT people that may affect decision making. Additionally, the LGBT population experiences higher rates of anxiety, depression and stressful life circumstances, and may be at risk of inferior psychosocial outcomes following a cancer diagnosis. LGBT people are also more likely to be estranged from their families of origin, and for older people, less likely to have adult children who may provide support and care.

Barriers to care for LGBT people include past negative interactions with healthcare systems, experiences or fear of discrimination and harassment in healthcare settings, assumptions of cisgender/heterosexual identity, lack of recognition or exclusion of same-sex partners from care, and a lack of relevant supportive care and information resources.

To provide safe and appropriate care for LGBT people with cancer, healthcare providers should:

- display environmental cues to show an inclusive and safe setting for LGBT patients
- avoid assumptions about the sexual orientation or gender identity of patients and their partners
- facilitate positive disclosure of sexual orientation or gender identity
- include same-sex/gender partners and families of choice in care
- be aware of relevant supportive care and information resources
- provide non-judgemental, patient-centred care.

Appendix D: Complementary therapies

Complementary therapies may be used together with conventional medical treatments to support and enhance quality of life and wellbeing. They do not aim to cure the patient's cancer. Instead, they are used to help control symptoms such as pain and fatigue (Cancer Council Australia 2019).

The lead clinician or health professional involved in the patient's care should discuss the patient's use (or intended use) of complementary therapies not prescribed by the multidisciplinary team to assess safety and efficacy and to identify any potential toxicity or drug interactions.

The lead clinician should seek a comprehensive list of all complementary and alternative medicines being taken and explore the patient's reason for using these therapies and the evidence base. A transparent and honest discussion that is free from judgement should be encouraged.

While some complementary therapies are supported by strong evidence, others are not. For such therapies, the lead clinician should discuss their potential benefits and use them alongside conventional therapies (NHMRC 2014).

If the patient expresses an interest in using complementary therapies, the lead clinician should consider referring patients to health providers within the multidisciplinary team who have expertise in the field of complementary and alternative therapies (e.g. a clinical pharmacist, dietitian or psychologist) to assist them to reach an informed decision. Costs of such approaches should be part of the discussion with the patient and considered in the context of evidence of benefit.

The lead clinician should assure patients who use complementary therapies that they can still access a multidisciplinary team review and encourage full disclosure about therapies being used.



More information

See the Clinical Oncological Society of Australia's position statement *Use of complementary and alternative medicine by cancer patients* <www.cosa.org.au/publications/position-statements.aspx>.

Appendix E: Members of the multidisciplinary team for neuroendocrine tumours

The multidisciplinary team may include the following members:

- endocrinologist*
- interventional radiologist*
- medical oncologist*
- nuclear medicine specialist*
- nurse coordinator*
- pathologist*
- surgeon*
- Aboriginal health practitioner, Indigenous liaison officer or remote general practitioner
- cardiologist
- clinical trials Staff
- dietitian
- exercise physiologist / physiotherapist
- fertility specialist
- gastroenterologist
- general practitioner
- geneticist
- nuclear medicine technologist
- psychiatrist
- psychologist
- palliative care specialist
- radiation oncologist
- respiratory physician
- social worker
- spiritual/pastoral care

* Denotes core members. Core members of the multidisciplinary team are expected to attend most multidisciplinary team meetings either in person or remotely.

Comprehensive neuroendocrine multidisciplinary teams

European Neuroendocrine Tumour Society (ENETS) Accredited Centres of Excellence (Neuroendocrine Multidisciplinary Teams):

- Peter MacCallum Cancer Centre, Melbourne, Vic
- Royal North Shore Hospital, St Leonards, NSW.

Non-ENETS Accredited Neuroendocrine Multidisciplinary Teams:

- Royal Brisbane and Women's Hospital, Brisbane, Qld
- Fiona Stanley Hospital, Murdoch, WA
- The Queen Elizabeth Hospital, Adelaide, SA.

Resource list

For patients, families and carers

Advance Care Planning Australia

Advance Care Planning Australia provides national advance care planning resources for individuals, families, health professional and service providers. Resources include a national advisory service, information resources, a legal forms hub and education modules.

- Telephone: 1300 208 582
- Website <www.advancecareplanning.org.au>

Australian Cancer Survivorship Centre

The Australian Cancer Survivorship Centre has developed information resources and events to help people move from initial treatment to post treatment and beyond, including those receiving maintenance treatments. While they do not provide clinical advice, they connect with a range of providers to enable improved care.

- Telephone: (03) 8559 6220
- Website <www.petermac.org/cancersurvivorship>

Australian Commission on Safety and Quality in Health Care

The Australian Commission on Safety and Quality in Health Care has developed a resource for patients and carers explaining the coordination of care that patients should receive from their health service during cancer treatment. The resource is called *What to expect when receiving medication for cancer care* <<https://www.safetyandquality.gov.au/publications-and-resources/resource-library/what-expect-when-receiving-medication-cancer-care>>.

Beyond Blue

Beyond Blue provides information about depression, anxiety and related disorders, as well as about available treatment and support services.

- Telephone: 1300 22 4636
- Website <www.beyondblue.org.au>

Cancer Australia

Cancer Australia is a specialist agency within the Australian Government's Health portfolio, providing national leadership in cancer control across all cancers, for all Australians.

Cancer Australia's purpose is to minimise the impact of cancer, address disparities, and improve the health outcomes of people affected by cancer in Australia by providing national leadership in cancer control.

Cancer Australia achieves this by developing and promoting evidence-based best practice cancer care; providing consumer and health professional cancer information; funding priority cancer research; and strengthening national cancer data capacity.

Cancer Australia provides accessible, evidence-based information about cancer for people affected by cancer, carers and their families through the Cancer Australia websites, resource library and video content.

- Website <www.canceraustralia.gov.au>

Cancer Council's Cancer Information and Support Service

Cancer Council 13 11 20 is a confidential telephone support service available to anyone affected by cancer. This service acts as a gateway to evidence-based documented, practical and emotional support available through Cancer Council services and other community organisations. Calls will be answered by a nurse or other oncology professional who can provide information relevant to a patient's or carer's situation. Health professionals can also access this service.

- Telephone: 13 11 20 – Monday to Friday, 9.00am to 5.00pm (some states have extended hours)
- Website <www.cancer.org.au/about-us/state-and-territory-councils/>

Cancer Council's Cancer Connect

Cancer Connect is a free and confidential telephone peer support service that connects someone who has cancer with a specially trained volunteer who has had a similar cancer experience.

A Connect volunteer can listen with understanding and share their experiences and ways of coping. They can provide practical information, emotional support and hope. Many people newly diagnosed with cancer find this one-to-one support very beneficial.

For more information on Cancer Connect call Cancer Council on 13 11 20.

Canteen

Canteen helps adolescents, young adults and parents to cope with cancer in their family. Canteen offers individual support services, peer support services and a youth cancer service, as well as books, resources and useful links.

- Telephone: 1800 835 932 to talk to a health professional about information and support for young people or 1800 226 833 for other enquiries
- Website <www.canteen.org.au/>

Clinical trial information

For a collection of clinical trials available in Australia see the following sources of information:

- Australian clinical trials <www.australianclinicaltrials.gov.au/>
- Australian New Zealand Clinical Trials Registry <www.anzctr.org.au/>
- ClinicalTrials.gov <www.clinicaltrials.gov/>.

CanEAT pathway

A guide to optimal cancer nutrition for people with cancer, carers and health professionals.

- Website <<https://education.eviq.org.au/courses/supportive-care/malnutrition-in-cancer>>
- Website <<https://www.cancer.nsw.gov.au/general-cancer-information/cancer-support/coping-with-cancer/physical-wellbeing/eating-well>>

Guides to best cancer care

The short guides help patients, carers and families understand the optimal cancer care that should be provided at each step. They include optimal timeframes within which tests or procedures should be completed, prompt lists to support patients to understand what might happen at each step of their cancer journey and to consider what questions to ask, and provide information to help patients and carers communicate with health professionals.

The guides are located on an interactive web portal, with downloadable PDFs available in multiple languages.

- Website <www.cancercareguides.org.au>

Look Good, Feel Better

A free national community service program, run by the Cancer Patients Foundation, dedicated to teaching cancer patients how to manage the appearance-related side effects caused by treatment for any type of cancer.

- Telephone: 1800 650 960
- Website <<https://lgfb.org.au>>

Quitline

Quitline is a confidential, evidence-based telephone counselling service. Highly trained Quitline counsellors use behaviour change techniques and motivational interviewing over multiple calls to help people plan, make and sustain a quit attempt.

Quitline is a culturally inclusive service for all, and Aboriginal counsellors are also available. Health professionals can refer patients to Quitline online or via fax.

- Telephone: 13 7848
- Website <www.quit.org.au> or the relevant website in your state or territory.

For health providers

Australian Cancer Survivorship Centre

The Australian Cancer Survivorship Centre provides expertise in survivorship care, information, support and education. Its purpose is to support and enable optimal survivorship care.

- Telephone: (03) 8559 6220
- Website <www.petermac.org/cancersurvivorship>

Australian Commission on Safety and Quality in Health Care

The Australian Commission on Safety and Quality in Health Care has developed a guide for clinicians containing evidence-based strategies to support clinicians to understand and fulfil their responsibilities to cancer patients. This guide is particularly relevant to steps 3 to 6 of the optimal care pathway. The guide is titled *NSQHS Standards user guide for medication management in cancer care for clinicians* <<https://www.safetyandquality.gov.au/publications-and-resources/resource-library/nsqhs-standards-user-guide-medication-management-cancer-care-clinicians>>.

Cancer Australia

Cancer Australia provides evidence-based information for health professionals including guidance, cancer learnings, cancer guides, reports, resources, videos, posters and pamphlets.

- Website <www.canceraustralia.gov.au>
- Website <www.canceraustralia.gov.au/cancer-types/neuroendocrine-tumours/overview>

Cancer Council Australia

Information on prevention, research, treatment and support provided by Australia's peak independent cancer authority.

- Website <www.cancer.org.au>

CanEAT pathway

A guide to optimal cancer nutrition for people with cancer, carers and health professionals.

- Website <<https://education.eviq.org.au/courses/supportive-care/malnutrition-in-cancer>>
- Website <<https://patients.cancer.nsw.gov.au/coping-with-cancer/physical-wellbeing/eating-well>>.

eviQ

A clinical information resource providing health professionals with current evidence-based, peer-maintained, best practice cancer treatment protocols and information relevant to the Australian clinical environment.

- Website <www.eviq.org.au>

National Health and Medical Research Council

Information on clinical practice guidelines, cancer prevention and treatment.

- Website <www.nhmrc.gov.au>

NeuroEndocrine Cancer Australia

NeuroEndocrine Cancer Australia (NECA) is a not-for-profit medical charity providing support to Australians living with neuroendocrine cancers. NECA is governed by five pillars: patient support, education, research, awareness and advocacy. These pillars have been created to give neuroendocrine cancer patients, their families and friends, a voice in the community and access to the care and treatment they deserve.

- Telephone: 1300 287 363
- Website <www.neuroendocrine.org.au>

Glossary

advance care directive – voluntary person-led document that focus on an individual's values and preferences for future health and medical treatment decisions, preferred outcomes and care. They are completed and signed by a competent person. They are recognised by specific legislation (statutory) or common law (non-statutory). Advance care directives can also appoint the substitute decision-maker(s) who can make decisions about health or personal care on the individual's behalf if they are no longer able to make decisions themselves. Advance care directives focus on the future health care of a person, not on the management of his or her assets. They come into effect when an individual loses decision-making capacity.

advance care planning – the process of planning for future health and personal care, where the person's values, beliefs and preferences are made known so they can guide decision making at a future time when that person cannot make or communicate their decisions.

alternative therapies – treatments used in place of conventional medical treatment.

care coordinator – the health provider nominated by the multidisciplinary team to coordinate patient care. The care coordinator may change over time depending on the patient's stage in the care pathway and the location and care in which care is being delivered.

complementary therapies – supportive treatment used in conjunction with conventional medical treatment. These treatments may improve wellbeing and quality of life and help people deal with the side effects of cancer.

end-of-life care – includes physical, spiritual and psychosocial assessment, and care and treatment, delivered by health professionals and ancillary staff. It also includes support of families and carers and care of the patient's body after their death.

immunotherapy – a type of cancer treatment that helps the body's immune system to fight cancer. Immunotherapy can boost the immune system to work better against cancer or remove barriers to the immune system attacking the cancer.

indicator – a documentable or measurable piece of information regarding a recommendation in the optimal care pathway.

informed financial consent – the provision of cost information to patients, including notification of likely out-of-pocket expenses (gaps), by all relevant service providers, preferably in writing, before admission to hospital or treatment (Commonwealth Department of Health 2017).

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

metastatic disease – cancer that has spread from the part of the body where it started (the primary site) to other parts of the body.

multidisciplinary care – an integrated team approach to health care in which medical and allied health providers consider all relevant treatment options and collaboratively develop an individual treatment plan for each patient.

multidisciplinary team – comprises the core disciplines that are integral to providing good care. The team is flexible in approach, reflects the patient's clinical and psychosocial needs and has processes to facilitate good communication.

multidisciplinary team meeting – a meeting of health professionals from one or more clinical disciplines who together make decisions about recommended treatment of patients.

optimal care pathway – the key principles and practices required at each stage of the care pathway to guide the delivery of consistent, safe, high-quality and evidence-based care for all people affected by cancer.

performance status – an objective measure of how well a patient can carry out activities of daily life.

primary care health professional – in most cases this is a general practitioner but may also include general practice nurses, community nurses, nurse practitioners, allied health professionals, midwives, pharmacists, dentists and Aboriginal health workers.

spiritual care – the aspect of humanity that refers to the way individuals seek and express meaning and purpose and the way they experience their connectedness to the moment, to self, to others, to nature, and to the significant or sacred.

substitute decision-maker – a person permitted under the law to make decisions on behalf of someone who does not have competence or capacity.

supportive care – care and support that aims to improve the quality of life of people living with cancer, cancer survivors and their family and carers and particular forms of care that supplement clinical treatment modalities.

survivorship – an individual is considered a cancer survivor from the time of diagnosis, and throughout their life; the term includes individuals receiving initial or maintenance treatment, in recovery or in the post-treatment phase.

survivorship care plan – a formal, written document that provides details of a person's cancer diagnosis and treatment, potential late and long-term effects arising from the cancer and its treatment, recommended follow-up, surveillance, and strategies to remain well.

References

- Australian Adult Cancer Pain Management Guideline Working Party 2019, *Australian adult cancer pain management guideline: 'Cancer pain management in adults'*, Cancer Council Australia, Sydney, viewed 6 June 2019, <<https://wiki.cancer.org.au/australiawiki/index.php?oldid=191646>>.
- Australian Cancer Survivorship Centre 2016, 'Survivorship care planning', viewed 2 October 2020, <https://www.petermac.org/sites/default/files/page/downloads/Survivorship_Care_Planning.pdf>.
- Australian Cancer Survivorship Centre 2019, *Community support organisations' cancer survivorship care consensus statement*, viewed 10 February 2020, <https://www.petermac.org/sites/default/files/media-uploads/NGO_ConsensusStatement.pdf>.
- Australian Clinical Trials 2015, 'Potential benefits and risks', National Health and Medical Research Council, Department of Industry, Innovation and Science, Australian Government, Canberra, viewed 24 July 2019, <<https://www.australianclinicaltrials.gov.au/why-be-part-clinical-trial/potential-benefits-and-potential-risks>>.
- Australian Commission for Safety and Quality in Health Care (ACSQHC) 2015, *Credentialing health practitioners and defining their scope of clinical practice: a guide for managers and practitioners*, ACSQHC, Sydney, viewed 18 February 2020, <<https://www.safetyandquality.gov.au/publications-and-resources/resource-library/credentialing-health-practitioners-and-defining-their-scope-clinical-practice-guide-managers-and-practitioners>>.
- Australian Commission for Safety and Quality in Health Care (ACSQHC) 2020, '*NSQHS Standards user guide for medication management in cancer care*', ACSQHC, Sydney, viewed 16 April 2020, <<https://www.safetyandquality.gov.au/publications-and-resources/resource-library/nsqhs-standards-user-guide-medication-management-cancer-care>>.
- Australian Commission on Safety and Quality in Health Care (ACSQHC) 2011, *Patient-centred care: Improving quality and safety through partnerships with patients and consumers*, ACSQHC, Sydney, viewed 17 March 2022.
- Australian Commission on Safety and Quality in Health Care (ACSQHC) 2014, *Health literacy: taking action to improve safety and quality*, ACSQHC, Sydney, viewed 18 February 2020, <<https://www.safetyandquality.gov.au/sites/default/files/migrated/Health-Literacy-Taking-action-to-improve-safety-and-quality.pdf>>.
- Australian Commission on Safety and Quality in Health Care (ACSQHC) 2017, *National Safety and Quality Health Service Standards guide for hospitals*, ACSQHC, Sydney, viewed 18 February 2020, <<https://www.safetyandquality.gov.au/wp-content/uploads/2017/12/National-Safety-and-Quality-Health-Service-Standards-Guide-for-Hospitals.pdf>>.
- Australian Commission on Safety and Quality in Health Care (ACSQHC) 2019a, *Person-centred care*, ACSQHC, Sydney, viewed 15 May 2020, <<https://www.safetyandquality.gov.au/our-work/partnering-consumers/person-centred-care>>.
- Australian Commission on Safety and Quality in Health Care (ACSQHC) 2019b, *Australian Hospital Patient Experience Question Set*, ACSQHC, Sydney, viewed 25 March 2020, <<https://www.safetyandquality.gov.au/our-work/indicators-measurement-and-reporting/australian-hospital-patient-experience-question-set>>.
- Australian Government Department of Health (AGDH) 2021, *National framework for advance care planning documents*, AGDH, viewed 24 March 2022, <<https://www.health.gov.au/resources/publications/national-framework-for-advance-care-planning-documents>>.

Australian Institute of Health and Welfare (AIHW) 2016, *Australia's health 2016*, Australia's health series no. 15. Cat. no. AUS 199, AIHW, Canberra.

Australian Institute of Health and Welfare (AIHW) 2017, *Access to health services by Australians with disability*, viewed 20 February 2020, <<https://www.aihw.gov.au/reports/disability/access-health-services-disability/contents/content>>.

Australian Institute of Health and Welfare (AIHW) 2018, *Australia's health 2018*, Australia's health series no. 16. Cat. no. AUS 221, AIHW, Canberra.

Australian Institute of Health and Welfare (AIHW) 2021, Cancer data in Australia, Cancer summary data visualization: neuroendocrine tumours, viewed 9 December 2021, <<https://www.aihw.gov.au/reports/cancer/cancer-data-in-australia/contents/cancer-summary-data-visualisation>>.

AYA Cancer Fertility Preservation Guidance Working Group 2014, *Fertility preservation for AYAs diagnosed with cancer: guidance for health professionals*, Cancer Council Australia, Sydney, viewed 20 July 2020, <https://wiki.cancer.org.au/australia/COSA:AYA_cancer_fertility_preservation/Introduction>.

Cancer Australia 2015, *National Aboriginal and Torres Strait Islander cancer framework*, Cancer Australia, Surry Hills, viewed 24 August 2017, <<https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/national-aboriginal-and-torres-strait-islander-cancer-framework>>.

Cancer Australia 2017, *Principles of cancer survivorship*, Australian Government, Sydney, viewed 18 February 2020, <https://canceraustralia.gov.au/sites/default/files/publications/principles-cancer-survivorship/pdf/pocs_-_principles_of_cancer_survivorship.pdf>.

Cancer Australia 2019a, *Principles of multidisciplinary care*, Australian Government, Sydney, viewed 18 July 2019, <https://canceraustralia.gov.au/system/tdf/guidelines/all_about_multidisciplinary_care.pdf?file=1&type=node&id=3551>.

Cancer Australia 2019b, 'Cancer incidence', viewed 18 February 2020, <<https://ncci.canceraustralia.gov.au/diagnosis/cancer-incidence/cancer-incidence>>.

Cancer Australia 2020, *Cancer care in the time of COVID-19: a conceptual framework for the management of cancer during a pandemic*, Australian Government, Sydney, viewed 3 September 2020, <<https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/cancer-care-time-covid-19-conceptual-framework-management-cancer-during-pandemic>>.

Cancer Australia 2021, Neuroendocrine tumours, viewed 9 December 2021 <<https://www.canceraustralia.gov.au/cancer-types/neuroendocrine-tumours/statistics>>.

Cancer Centre 2021, About Neuroendocrine tumors 2021, viewed 27 September 2021, <<https://www.cancercenter.com/cancer-types/neuroendocrine-tumors/about>>.

Cancer Council Australia 2018, 'Prevention', Cancer Council Australia, Sydney, viewed 18 July 2019, <<https://www.cancer.org.au/preventing-cancer/>>.

Cancer Council Australia 2019, 'Complementary and alternative therapies', Cancer Council Australia, Sydney, viewed 22 July 2019, <<https://www.cancer.org.au/about-cancer/treatment/complementary-therapies-and-cancer.html>>.

Cancer Council Victoria 2019, 'Palliative care', Cancer Council Victoria, viewed 7 October 2019, <https://www.cancervic.org.au/cancer-information/treatments/treatments-types/palliative_care/palliative-care-treatment.html>.

- Cancer Research in Primary Care 2016, *Principles statement: Shared care*, PC4 Shared Care Working Group, Melbourne, viewed 4 October 2019, <<http://pc4tg.com.au/wp-content/uploads/2016/07/PC4-Principles-Statement-shared-care-2016-1.pdf>>.
- Cancer.Net 2019, Neuroendocrine tumors: symptoms and signs, viewed 27 September 2021, <<https://www.cancer.net/cancer-types/neuroendocrine-tumors/symptoms-and-signs>>.
- Caplin ME, Pavel M, Phan AT, Cwikla JB, Sedlackova E, Thanh XMT, et al. 2021, 'Lanreotide autogel / depot in advanced enteropancreatic neuroendocrine tumours: final results of the CLARINET open-label extension study', vol. 71, no. 2, pp. 502–513.
- Chan DL, Bernard E, Schembri G, Roach, P, Johnson M, Pavlakis N, et al. 2020, 'High metabolic tumour volume on 18-fluorodeoxyglucose positron emission tomography predicts poor survival from neuroendocrine neoplasms', *Neuroendocrinology*, vol. 110, pp. 950–958.
- Clinical Oncology Society of Australia (COSA) 2013, Special Issue: COSA's 40th Annual Scientific Meeting, Cancer Care Coming of Age, 12–14 November 2013, Adelaide Convention Centre, *Asia-Pacific Journal of Clinical Oncology*, vol. 9, no. 3, pp. 61–98.
- Clinical Oncology Society of Australia (COSA) 2014, *Psychosocial management of AYAs diagnosed with cancer: guidance for health professionals*, COSA, Sydney, viewed 7 October 2019, <http://wiki.cancer.org.au/australia/COSA:Psychosocial_management_of_AYA_cancer_patients>.
- Clinical Oncology Society of Australia (COSA) 2015, *Cancer care coordinator: position statement*, COSA, Sydney, viewed 22 July 2019, <https://www.cosa.org.au/media/332296/cancer-care-coordinator-position-statement_final-endorsed-by-council_161115_cnsa-logo.pdf>.
- Clinical Oncology Society of Australia (COSA) 2016, *Australasian tele-trial model: access to clinical trials closer to home using tele-health a national guide for implementation*, v1.7, COSA, Sydney, viewed 18 July 2019, <<https://www.cosa.org.au/media/332325/cosa-teletrial-model-final-19sep16.pdf>>.
- Clinical Oncology Society of Australia (COSA) 2018, *COSA position statement on exercise in cancer care*, COSA, Sydney, viewed 22 July 2019, <<https://www.cosa.org.au/media/332488/cosa-position-statement-v4-web-final.pdf>>.
- Commonwealth Department of Health 2017, *Out-of-pocket expenses for private medical treatment (informed financial consent)*, Commonwealth of Australia, Canberra.
- Cormie P, Atkinson M, Bucci L, Cust A, Eakin E, Hayes S, et al. 2018, 'Clinical Oncology Society of Australia position statement on exercise in cancer care', *Medical Journal of Australia*, vol. 209, no. 4, pp. 184–187.
- Cormie P, Zopf EM, Zhang X, Schmitz KH 2017, 'The impact of exercise on cancer mortality, recurrence, and treatment-related adverse effects', *Epidemiologic Reviews*, vol. 39, no. 1, pp. 71–92.
- Dasari A, Shen C, Halperin D, Zhao B, Zhou S, Xu Y, et al. 2017, 'Trends in the prevalence, and survival outcomes in patients with neuroendocrine tumors in the United States' *JAMA Oncology*, vol. 3, no. 10, pp. 1335–1342.
- Decoster L, Van Puyvelde K, Mohile S, Wedding U, Basso U, Colloca G, et al. 2015, 'Screening tools for multidimensional health problems warranting a geriatric assessment in older cancer patients: an update on SIOG recommendations', *Annals of Oncology*, vol. 26, no.1, pp 288–300.
- Department of Health and Ageing 2013, *South Australian gastroenteropancreatic neuroendocrine tumours pathway*, Government of South Australia, Adelaide.

- Emery J 2014, 'Cancer survivorship – the role of the GP', *Australian Family Physician*, vol. 43, no. 8, pp. 521–525.
- eviQ 2019, 'Safe handling and waste management of hazardous drugs', Cancer Institute NSW, Sydney, viewed 22 July 2019, <<https://www.eviq.org.au/clinical-resources/administration-of-antineoplastic-drugs/188-safe-handling-and-waste-management-of-hazardou>>.
- Ferrari A, Thomas D, Franklin A, Hayes-Lattin B, Mascarini M, van der Graaf W, et al. 2010, 'Starting an adolescent and young adult program: some success stories and some obstacles to overcome', *Journal of Clinical Oncology*, vol. 28, no. 32, pp. 4850–4857.
- Fitch M 2000, 'Supportive care for cancer patients', *Hospital Quarterly*, vol. 3, no. 4, pp. 39–46.
- Fitch MI 2008, 'Supportive care framework', *Canadian Oncology Nursing Journal*, vol. 18, no. 1, pp. 6–14.
- Gangi A, Anaya DA 2020, 'Surgical principles in the management of small bowel neuroendocrine tumors', *Current Treatment Options in Oncology*, vol. 21, no. 11, Article 88.
- GI Cancer Institute 2021, Control Nets, viewed 9 December 2021, <<https://gicancer.org.au/clinical-trial/control-nets/>>.
- Gilligan T, Coyle N, Frankel RM, Berry DL, Bohlke K, Epstein, RM, et al. 2017, 'Patient–clinician communication: American Society of Clinical Oncology consensus guideline', *Journal of Clinical Oncology*, vol. 35, no. 31, pp. 3618–3623.
- Grozinsky-Glasberg S, Grossman AB, Gross DJ 2015, 'Carcinoid heart disease: from pathophysiology to treatment – “Something in the way it moves”', *Neuroendocrinology*, no. 101, pp. 263–273.
- Hack TF, Reuther DJ, Weir LM, Grenier D, Degner LF 2012, 'Promoting consultation recording practice in oncology: identification of critical implementation factors and determination of patient benefit', *Psycho-Oncology*, vol. 22, pp. 1273–1282.
- Haines IE 2011, 'Managing patients with advanced cancer: the benefits of early referral for palliative care', *Medical Journal of Australia*, vol. 194, no. 3, pp. 107–108.
- Hayes SC, Newton RU, Spence RR, Galvao DA 2019, 'The Exercise and Sports Science Australia position statement: exercise medicine in cancer management', *Journal of Science and Medicine in Sport*, vol. 22, no. 11, pp. 1175–1199.
- Hewitt M, Greenfield S, Stovall E 2006, *From cancer patient to cancer survivor: lost in transition*, National Academies Press, Washington.
- Hofman MS, Hicks RJ 2012, 'Changing paradigms with molecular imaging of neuroendocrine tumours', *Discovery Medicine*, vol. 14, no. 74, pp. 71–81.
- I-Med Radiology Network 2020, 68Ga Dotatate PET-CT scan, viewed 27 September 2021, <<https://i-med.com.au/68ga-dotatate-pet-ct-scan>>.
- Jefford M, Ward AC, Lisy K, Lacey K, Emery JD, Glaser AW, et al. 2017, 'Patient-reported outcomes in cancer survivors: a population-wide cross-sectional study', *Support Care Cancer*, no. 10, pp. 3171–3179.
- Kaltsas G, Caplin M, Davies P, Ferone D, Garcia-Carbonero R, Grozinsky-Glasberg S, et al. 2017, 'ENETS 2017 Consensus Guidelines for the Standards of Care in Neuroendocrine Tumors', *Neuroendocrinology*, vol. 105, no. 3, pp. 245–254.

Knigge U, Capdevila J, Bartsch D, Baudin E, Falkerby J, Kianmanesh R, et al. 2017, 'ENETS Consensus Recommendations for Standards of Care in Neuroendocrine Neoplasms: Follow-Up and Documentation', *Neuroendocrinology*, vol. 105, no.3, pp. 310-319.

Lidsaar-Powell R, Butow P, Boyle F, Juraskova 2018a, 'Facilitating collaborative and effective family involvement in the cancer setting: guidelines for clinicians (TRIO guidelines-1)', *Patient Education and Counseling*, vol. 101, no. 6, pp. 970–982.

Lidsaar-Powell R, Butow P, Boyle F, Juraskova 2018b, 'Managing challenging interactions with family caregivers in the cancer setting: guidelines for clinicians (TRIO guidelines-2)', *Patient Education and Counseling*, vol. 101, no. 6, pp. 983–994.

Lisy K, Langdon L, Piper A, Jefford M 2019, 'Identifying the most prevalent unmet needs of cancer survivors in Australia: a systematic review', *Asia-Pacific Journal of Clinical Oncology*, vol. 15, no. 5, pp. e68–e78.

Magi L, Mazzuca F, Rinzivillo M, Arrivi G, Pillozzi E, Prosperi et al. 2019, 'Multidisciplinary management of neuroendocrine neoplasia: a real-world experience from a referral center', *Journal of Clinical Medicine*, vol. 8, no. 6, p. 910.

Michael M, Thursfield V, te Marvelde L, Kong G, Hick RJ 2021, 'Incidence, prevalence and survival trends for neuroendocrine neoplasms in Victoria, Australia from 1982 to 2019. Based on site, grade and region 'Asia-Pacific Journal of Clinical Oncology, 1–12. doi: 10.1111/ajco.13671.

Monterosso L, Platt V, Bulsara M, Berg M 2019, 'Systematic review and meta-analysis of patient reported outcomes for nurse-led models of survivorship care for adult cancer patients', *Cancer Treatment Reviews Journal*, no. 73, pp. 62–72.

My Health Record 2019, 'What is My Health Record?' Australian Government, Sydney, viewed 17 December 2019, <<https://www.myhealthrecord.gov.au/for-you-your-family/what-is-my-health-record>>.

National Comprehensive Cancer Network (NCCN) 2021, *Guidelines Version 3: Neuroendocrine and adrenal*, viewed 27 September 2021, <<https://www.nccn.org/guidelines/guidelines-detail?category=1&id=1448>>.

National Disability Insurance Agency 2018, 'How the NDIS works', NDIS, Canberra, viewed 3 June 2019, <<https://www.ndis.gov.au/understanding/how-ndis-works>>.

National Health and Medical Research Council (NHMRC) 2013, *Personalised medicine and genetics*, viewed 3 June 2019 <<https://www.nhmrc.gov.au/about-us/publications/personalised-medicine-and-genetics>>.

National Health and Medical Research Council (NHMRC) 2014, *Talking with your patients about complementary medicine: a resource for clinicians*, NHMRC, Canberra, viewed 18 February 2020, <<https://www.caresearch.com.au/caresearch/ClinicalPractice/PatientConsiderations/ComplementaryTherapies/tabid/1258/Default.aspx>>.

National Organisation for Rare Diseases (NORD) 2021, Carcinoid syndrome, viewed 27 September 2021, <<https://rarediseases.org/rare-diseases/carcinoid-syndrome>>.

NeuroEndocrine Cancer Australia (NECA) 2019a, *Neuroendocrine tumours: a guide for patients*, viewed 27 September 2021, <<https://neuroendocrine.org.au/wp-content/uploads/2020/02/190822-NET-Patient-Booklet.pdf>>.

NeuroEndocrine Cancer Australia (NECA) 2019b, *A guide for health professionals*, viewed 27 September 2021, <<https://neuroendocrine.org.au/wp-content/uploads/2020/02/191009-UFD-NETs-forprofessionals.pdf>>.

- NeuroEndocrine Cancer Australia (NECA) 2020a, *Symptoms and signs: clinical presentation*, viewed 27 September 2021, <https://neuroendocrine.org.au/wp-content/uploads/2020/06/N5058-NECA_Symptoms_Download-v2.pdf>.
- NeuroEndocrine Cancer Australia (NECA) 2020b, *Carcinoid heart disease*, viewed 27 September 2021, <https://neuroendocrine.org.au/wp-content/uploads/2020/07/N5058-NECA_CarcinoidHeartdisease_Download-v1-1.pdf>.
- NeuroEndocrine Cancer Australia (NECA) 2021, *National action plan*, viewed 27 September 2021, <<https://neuroendocrine.org.au/get-involved/net-national-action-plan/>>.
- O'Shea T, Druce M 2017 'When should genetic testing be performed in patients with neuroendocrine tumours?' *Review of Endocrine and Metabolic Disorders*, vol. 18, no. 4, pp. 499– 515.
- palliAGED 2018, 'Needs Assessment', Flinders University, Bedford Park, viewed 1 October 2019, <<https://www.palliaged.com.au/tabid/4879/Default.aspx>>.
- Palliative Care Australia 2018, *National Palliative Care Standards*, 5th edn, Palliative Care Australia, Canberra, viewed 24 July 2019, <http://palliativecare.org.au/wp-content/uploads/dlm_uploads/2018/11/PalliativeCare-National-Standards-2018_Nov-web.pdf>.
- Palliative Care Victoria 2019, 'Spiritual care', Palliative Care Victoria, Melbourne, viewed 22 July 2019, <<https://www.pallcarevic.asn.au/healthcare-professionals/about-palliative-care/care/spiritual-care/>>.
- Perren A, Couvelard A, Scoazec JY, Costa F, Borbath I, Fave G, et al. 2017, 'ENETS Consensus Guidelines for the Standards of Care in Neuroendocrine Tumors: Pathology: Diagnosis and Prognostic Stratification', *Neuroendocrinology*, vol. 105, no. 3, pp. 196-200.
- Peter MacCallum Cancer Centre 2019, *Community support organisations' cancer survivorship care consensus statement*, viewed 10 February 2020, <https://www.petermac.org/sites/default/files/media-uploads/NGO_ConsensusStatement.pdf>.
- Queensland Health 2015, *Sad news sorry business: guidelines for caring for Aboriginal and Torres Strait Islander people through death and dying*, State Government of Queensland, Brisbane, viewed 22 July 2019, <https://www.health.qld.gov.au/__data/assets/pdf_file/0023/151736/sorry_business.pdf>.
- Rinke A, Wittenberg M, Schade-Brittinger C, Aminssadati B, Ronicke e, Gress TM, et al. 2016, 'Placebo-controlled, double-blind, prospective, randomized study on the effect of octreotide LAR in the control of tumor growth in patients with metastatic neuroendocrine midgut tumors (PROMID): results of long-term survival', *Neuroendocrinology*, vol. 104, no. 1, pp. 260–232.
- Rogel Cancer Centre University of Michigan 2021, 'Endocrine syndromes / endocrine tumor genetics', viewed 27 September 2021, <<https://www.rogelcancercenter.org/endocrine-cancer/familial-neuroendocrine-syndromes>>.
- Roswell Park 2021, *Prevention and detection of neuroendocrine tumors*, viewed 27 September 2021, <<https://www.roswellpark.org/cancer/neuroendocrine-tumors/prevention-detection>>.
- Safer Care Victoria 2021, *Data linkage plays a key role in COVID-19 pandemic response*, State of Victoria, Melbourne, viewed 4 March 2022, <<https://www.bettersafecare.vic.gov.au/news-and-media/data-linkage-plays-a-key-role-in-covid-19-pandemic-response>>.
- Silver JK 2015, 'Cancer prehabilitation and its role in improving health outcomes and reducing health care costs', *Seminars in Oncology Nursing*, vol. 31, no. 1, pp. 1–3.

- Silver JK, Baima J 2013, 'Cancer prehabilitation: an opportunity to decrease treatment-related morbidity, increase cancer treatment options, and improve physical and psychological health outcomes', *American Journal of Physical Medicine & Rehabilitation*, vol. 92, no. 8, pp. 715–727.
- Singh S, Moody L, Chan DL, Metz DC, Strosberg J, Asmis T, et al. 2018, 'Follow-up recommendations for completely resected gastroenteropancreatic neuroendocrine tumors', *JAMA Oncology*, vol. 4, no. 11, pp. 1597–1604.
- Sjoquist K, Zalcberg J 2013, 'Clinical trials – advancing cancer care', *Cancer Forum*, vol. 37, no. 1, pp. 80–88.
- Smith A, Bellizzi K, Keegan T, Zebrack B, Chen V, Neale A, et al. 2013, 'Health-related quality of life of adolescent and young adult patients with cancer in the United States: the Adolescent and Young Adult Health Outcomes and Patient Experience Study', *Journal of Clinical Oncology*, vol. 31, no. 17, pp. 2136–2145.
- Smith S, Case L, Waterhouse K, Pettitt N, Beddard L, Oldham J, et al. 2012, *A blueprint of care for teenagers and young adults with cancer*, Teenage Cancer Trust, Manchester, UK.
- Steer B, Marx G, Singhal N, McJannett M, Goldstein D, Prowse R 2009, 'Cancer in older people: a tale of two disciplines', *Internal Medicine Journal*, vol. 39, pp. 771–775.
- Strosberg J, El-Haddad G, Wolin E, Hendifar A, Yao J, Chasen B, et al. 2017, 'Trial of ¹⁷⁷Lu-Dotatate for midgut neuroendocrine tumors' *New England Journal of Medicine*, vol. 376, no. 2, pp. 125–135.
- Tan SY, Turner J, Kerin-Ayres K, Butler S, Deguchi C, Khatri S, et al. 2019, 'Health concerns of cancer survivors after primary anti-cancer treatment', *Support Cancer Care*, no. 10, pp. 3739–3747.
- The Centre for Victorian Data Linkage (CVDL) 2021, The Centre for Victorian Data Linkage overview, State of Victoria, Melbourne.
- Temel J, Greer J, Muzikansky A, Gallagher E, Admane S, Jackson V 2010, 'Early palliative care for patients with non-metastatic non-small cell lung cancer', *New England Journal of Medicine*, vol. 363, no. 8, pp. 733–742.
- Toomey PG, Teta AF, Patel KD, Ross SB, Rosemurgy AS 2016 'High-volume surgeons vs high volume hospitals; are best outcomes more due to who or where?' *The American Journal of Surgery*, vol. 211, no.1, pp. 59–63.
- Tsoli M, Chatzellis E, Koumariou A, Kolomodi D, Kaltsas G 2019 'Current best practice in the management of neuroendocrine tumors', *Therapeutic Advances in Endocrinology and Metabolism*, vol. 10. doi: 10.3390/jcm8060910.
- Vardy JL, Raymond JC, Koczwara B, Lisy K, Cohn RJ, Joske D, et al. 2019, 'Clinical Oncology Society of Australia position statement on cancer survivorship care', *Australian Journal of General Practice*, vol. 48, no.12, pp. 833–836.
- Vijayvergia N, Denlinger CS 2015, 'Lifestyle factors in cancer survivorship: where we are and where we are headed', *Journal of Personalized Medicine*, vol. 5, no. 3, pp. 243–263.
- Wildiers H, Heeren P, Puts M, Topinkova E, Maryska LG, Janssen-Heijnen MLG, et al. 2014, 'International Society of Geriatric Oncology Consensus on geriatric assessment in older patients with cancer', *Journal of Clinical Oncology*, vol. 32, no. 24, pp. 2595–2603.
- World Health Organization (WHO) 2018, 'Malnutrition', WHO, Geneva, viewed 18 July 2019, <<https://www.who.int/news-room/fact-sheets/detail/malnutrition>>.
- World Health Organization (WHO) 2020, *WHO report on cancer: setting priorities, investing wisely and providing care for all*, WHO, Geneva, viewed 17 March 2022.
- Zimmermann C, Swami N, Krzyzanowska M, Hannon B, Leighl N, Oza A, et al. 2014, 'Early palliative care for patients with advanced cancer: a cluster-randomised controlled trial', *Lancet*, vol. 383, no. 9930, pp. 1721–1730.

