Cancer information fact sheet

Understanding Appendix Cancer and Pseudomyxoma Peritonei (PMP)

A guide for people affected by cancer

This fact sheet has been prepared to help you understand more about appendix cancer and pseudomyxoma peritonei (PMP).

Many people look for support after being diagnosed with a cancer that is rare or less common than other cancer types. This fact sheet includes information about how these cancers are diagnosed and treated, as well as where to go for additional information and support services.

Many people feel shocked and upset when told they have cancer. You may experience strong emotions after a cancer diagnosis, especially if your cancer is rare or less common like appendix cancer or PMP. A feeling of being alone is usual with rare cancers, and you might be worried about how much is known about your type of cancer as well as how it will be managed. You may also be concerned about the cancer coming back after treatment. Linking into local support services (see last page) can help overcome feelings of isolation and will give you information that you may find useful.

About the appendix

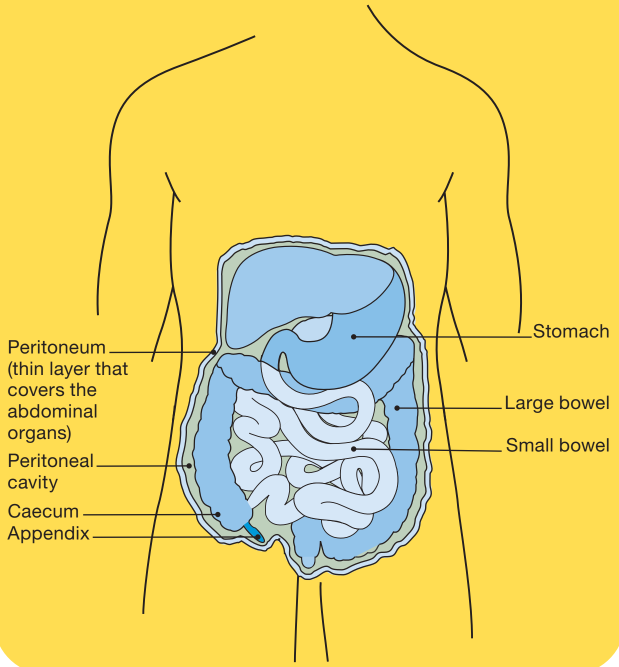
The appendix is a small tube that usually sits on the lower right side of the abdomen. It hangs off the end of the caecum, which is a pouch at the start of the large bowel. The appendix does not have a clear function, but is thought to store gut bacteria and may play a role in the immune system to help prevent disease.

What is appendix cancer?

Appendix cancer occurs when cells in the appendix become abnormal and keep growing and form a mass or lump called a tumour.

The type of cancer is defined by the particular cells that are affected and can be benign (non-cancerous) or malignant (cancerous). Malignant tumours have the potential to spread to other parts of the body through the blood stream or lymph vessels and form another tumour at a new site. This new tumour is known as secondary cancer or metastasis.

The abdomen



Types of appendix cancer

The most common types include:

Mucinous adenocarcinoma	These start in epithelial cells that line the inside of the appendix. They can produce mucin (a jelly-like substance) and spread to other parts of the body, including the peritoneum, which is a sheet of tissue that lines and protects organs in the abdomen (belly).
Neuroendocrine tumours (NETs)	These form in neuroendocrine cells inside the appendix. The neuroendocrine system is a network of glands and nerve cells that make hormones and release them into the bloodstream to help control normal body functions. Appendiceal NETs are often found at the tip of the appendix. See our *Understanding Neuroendocrine Tumours* fact sheet for information or contact NeuroEndocrine Cancer Australia (neuroendocrine.org.au).
Goblet cell carcinoma (GCC)	These have features of both a NET and adenocarcinoma but behave more like an adenocarcinoma, which can be more aggressive.
Colonic-type adenocarcinoma	These may behave like colon (large bowel) cancer and are often found at the base of the appendix.

For copies of Cancer Council fact sheets visit your local website (see last page for details).

What is PMP?

Pseudomyxoma peritonei (PMP) is a rare tumour that grows slowly and causes a build-up of mucin (a jelly-like substance) in the abdomen and pelvis, giving rise to the name “jelly belly”. Several other diseases may also be associated with “jelly belly” including mucinous adenocarcinoma, or may resemble features of PMP including mucinous tumours in the bowel.

PMP often starts in the appendix but can also start in other organs such as the large bowel and ovary. While it doesn’t spread to other parts of the body, PMP can put pressure on important organs as it continues to grow and this may cause problems.

How common are appendix cancer and PMP?

Appendix cancer is rare with 0.12 cases per 1,000,000 people each year being reported for primary malignancies (cancer that first develops in the appendix).[1] The most common types are seen in middle-aged people, with the typical age at diagnosis about 40 to 60 years. There is an almost equal risk for males and females for mucinous adenocarcinoma and GCC.[1] Colonic-type adenocarcinoma is diagnosed slightly more often in men and appendiceal NETs are diagnosed more often in women.[1]

PMP is also rare with about 1 or 2 cases per 1,000,000 people each year.[2] It is more likely to be diagnosed in people aged 40 years or over. Women may be diagnosed slightly more often and at an earlier stage than men, after a mass or lump is found in their ovary.

What are the risk factors?

The causes of appendix cancer and PMP are not known. There are no clear risk factors and neither appear to run in families. Increasing age, however, can increase the risk of appendix cancer.

What are the symptoms?

Appendix cancer may not cause symptoms in its early stages. However, some people may experience symptoms such as:

- appendicitis (lower right abdominal pain)
- gradual increase in waist size
- build-up of fluid in the abdomen
- bloating
- changes in bowel habits
- hernia
- ovarian mass or lump.

PMP is also difficult to detect, and symptoms may take a while to develop. Symptoms that some people may experience include:

- abdominal or pelvic pain
- gradual increase in waist size
- bloating
- changes in bowel habits
- hernia
- loss of appetite.

Diagnosis

Appendix cancer is often found during abdominal surgery for a different condition or after an appendectomy (surgical removal of the appendix) for a suspected case of appendicitis. Similarly, PMP is often discovered when investigating a different condition.

If your doctor thinks that you may have appendix cancer or PMP, they will perform a physical examination and carry out certain tests. If the results suggest that you may have appendix cancer or PMP, your doctor will refer you to a specialist who will carry out more tests. These may include:

- **Blood tests –** including a full blood count to measure your white blood cells, red blood cells, platelets and tumour markers (chemicals produced by cancer cells)

- **CT (computerised tomography) or MRI (magnetic resonance imaging) scans –** special machines are used to scan and create pictures of the inside of your body. Before the scan you may have an injection of dye (called contrast) into one of your veins, which makes the pictures clearer. During the scan, you will need to lie still on an examination table. For a CT scan, the table moves in and out of the scanner which is large and round like a doughnut; the scan takes about 10 minutes. For an MRI scan, the table slides into a large metal tube that is open at both ends; the scan takes a little longer, about 30–90 minutes to perform. Both scans are painless.

- **Ultrasound scan –** soundwaves are used to create pictures of the inside of your body. For this scan, you will lie down and a gel will be spread over the affected part of your body. A small device called a transducer is moved over the area. The transducer sends out soundwaves that echo when they encounter something dense, like an organ or tumour. The ultrasound images are then projected onto a computer screen. An ultrasound is painless and takes about 15–20 minutes.

- **Diagnostic laparoscopy –** a thin tube with a camera on the end (laparoscope) is inserted under sedation into the abdomen to view inside the cavity

- **Biopsy –** removal of some tissue from the affected area for examination under a microscope. The biopsy may be done in one of two ways. In a core needle biopsy, a local anaesthetic is used to numb the area, then a thin needle is inserted into the tissue under ultrasound or CT guidance. An open or surgical biopsy is done under general anaesthesia. The surgeon will cut through the skin and use a tiny instrument with a light and camera (laparoscope) to view the affected area and use another instrument to take a tissue sample.

Finding a specialist

Rare Cancers Australia have a knowledgebase directory of health professionals and cancer services across Australia: rarecancers.org.au.

Pseudomyxoma Survivor have a directory of PMP surgeons and specialists in Australia: pseudomyxomasurvivor.org/pmp-specialists/pmp-surgeons-and-specialists-australia/

Understanding Appendix Cancer and Pseudomyxoma Peritonei (PMP)

Treatment

You will be cared for by a multi-disciplinary team of health professionals during your treatment for appendix cancer or PMP. These may include a surgeon, medical oncologist (to prescribe and coordinate a course of systemic therapy which includes chemotherapy), radiation oncologist (to prescribe and coordinate a course of radiation therapy), nurse and allied health professionals such as a social worker, psychologist or counsellor, dietitian, physiotherapist and occupational therapist.

Discussion with your doctor will help you decide on the best treatment for your cancer depending on:

- the type of cancer you have
- where it is in your body
- whether or not the cancer has spread (stage of disease)
- your age, fitness and general health
- your preferences.

The main treatments for appendix cancer and PMP are surgery and chemotherapy. These can be given alone or in combination and are an effective treatment with a little over 60% of patients receiving both cytoreductive surgery and HIPEC (see below) surviving beyond 10 years. PMP may not be treated straight away if the tumour is small and growing slowly; in this case it will be observed and monitored regularly, an approach known as active surveillance.

Surgery

Surgery is the main treatment for appendix cancer, especially for people with early-stage disease who are otherwise in good health. The type of operation depends on the location and stage of the tumour.

PMP is usually treated with surgery: either cytoreductive surgery followed by chemotherapy (HIPEC – see next page) when aiming to cure PMP, or if the cancer cannot be treated effectively debulking surgery may be used instead to remove as much of the tumour as possible to reduce symptoms. Debulking surgery may be done again if the tumour grows back.

If part of the bowel is removed during surgery, the surgeon will usually join it back together. If this isn't possible, you may need a stoma where the end of the intestine is brought through an opening (the stoma) made in your abdomen and stitched onto the skin to allow faeces to be removed from the body and collected in a bag. The stoma may be temporary (where the operation is reversed later on) or permanent, depending on the amount of bowel that has been removed.

If you need a stoma, the surgeon will refer you to a stomal therapy nurse before surgery. These are nurses with special training in stoma care. They can answer your questions about adjusting to life with a stoma. For more information visit the Australian Association of Stomal Therapy Nurses at stomaltherapy.com or call Cancer Council 13 11 20.

› For a free copy of Cancer Council's booklet *Understanding Surgery* visit your local Cancer Council website or call 13 11 20.

Types of surgery

Appendectomy	Surgery to remove the appendix. Often used for early stage appendiceal NETs
Hemicolectomy	Surgery to remove a small part of the large bowel next to appendix; surrounding lymph nodes and blood vessels may also be removed during the procedure. Often used for appendiceal NETs at risk of spreading or appendix cancers that are not neuroendocrine
Cytoreductive surgery (CRS or peritonectomy)	Surgery to remove all visible tumour from the abdominal cavity; part of the bowel and other organs including gallbladder, spleen, stomach and kidney may also be removed. In females the uterus, ovaries and fallopian tubes may be removed; in males the seminal vesicles may be severed. Often used for late-stage appendix cancer and PMP Chemotherapy (see next page) may be used after the surgery.

Chemotherapy

Chemotherapy (sometimes just called "chemo") is the use of drugs to kill or slow the growth of cancer cells. You may have one chemotherapy drug, or a combination of drugs. This is because different drugs can destroy or shrink cancer cells in different ways.

Your treatment will depend on your situation and the type of cancer you have. It may also be used to help stop your cancer coming back after surgery. Your medical oncologist will discuss your options with you.

There are different types of chemotherapy used to treat appendix cancer and PMP:

- **Local chemotherapy –** where the chemotherapy drugs are delivered directly to the cancer. When placed directly in the abdomen it is called intraperitoneal chemotherapy (see below).

- **Systemic chemotherapy –** where the chemotherapy drugs enter the bloodstream and travel throughout the body to target rapidly dividing cancer cells in the organs and tissues. This type of chemotherapy is given through a drip into a vein (intravenously) or as a tablet that is swallowed.

Types of intraperitoneal chemotherapy

HIPEC (heated intraperitoneal chemotherapy)	The chemotherapy drug is heated to around 40°C (body temperature is about 37°C) to increase its effectiveness and placed directly in the abdomen to kill any tumour cells that remain after surgery or help control ascites (build-up of fluid); typically removed after 30–90 minutes
EPIC (early post-operative intraperitoneal chemotherapy)	The chemotherapy drug is delivered to the abdomen the day after surgery using an access port (small plastic device); continued for several days. Usually used after HIPEC when able to be tolerated

› For a free copy of Cancer Council's booklet *Understanding Chemotherapy* visit your local Cancer Council website or call 13 11 20.

Radiation therapy

Radiation therapy (also known as radiotherapy) uses high energy rays to destroy cancer cells. It may be used for appendix cancer when it has spread to other parts of the body, such as the bone. Radiation therapy can shrink the cancer and relieve symptoms.

A course of radiation therapy needs careful planning. During your first consultation you will meet with a radiation oncologist. At this session you will lie on an examination table and have a CT scan in the same position you will be placed in for treatment. The information from this session will be used by your specialist to work out the treatment area and how to deliver the right dose of radiation. Radiation therapists will then deliver the course of radiation therapy as set out in the treatment plan.

Radiation therapy does not hurt and is usually given in small doses over a period of time to minimise side effects.

› For a free copy Cancer Council's booklet *Understanding Radiation Therapy* visit your local Cancer Council website or call 13 11 20.

Clinical trials

Your doctor or nurse may suggest you take part in a clinical trial. Doctors run clinical trials to test new or modified treatments and ways of diagnosing disease to see if they are better than current methods. For example, if you join a randomised trial for a new treatment, you will be chosen at random to receive either the best existing treatment or the modified new treatment. Over the years, trials have improved treatments and led to better outcomes for people diagnosed with cancer.

For more information, visit australiancancertrials.gov.au or contact the Appendix Cancer/Pseudomyxoma Peritonei Research Foundation (ACPMP): acpmp.org. For more information on appendiceal NET clinical trials contact:

- NeuroEndocrine Cancer Australia: neuroendocrine.org.au
- Australasian Gastro-Intestinal Trials Group (AGITG): gicancer.org.au

› For a free copy Cancer Council's booklet *Understanding Clinical Trials and Research* visit your local Cancer Council website or call 13 11 20.

Understanding Appendix Cancer and Pseudomyxoma Peritonei (PMP)

Complementary therapies and integrative oncology

Complementary therapies are designed to be used alongside conventional medical treatments (such as surgery, chemotherapy and radiation therapy) and can increase your sense of control, decrease stress and anxiety, manage fatigue and improve your mood.

Complementary therapy	Clinically proven benefits
acupuncture	reduces chemotherapy-induced nausea and vomiting; improves quality of life
aromatherapy	improves sleep and quality of life
art therapy, music therapy	reduce anxiety and stress; manage fatigue; aid expression of feelings
counselling, support groups	help reduce distress, anxiety and depression; improve quality of life
hypnotherapy	reduces pain, anxiety, nausea and vomiting
massage	improves quality of life; reduces anxiety, depression, pain and nausea
meditation, relaxation, mindfulness	reduce stress and anxiety; improve coping and quality of life
qi gong	reduces anxiety and fatigue; improves quality of life
spiritual practices	help reduce stress; instil peace; improve ability to manage challenges
tai chi	reduces anxiety and stress; improves strength, flexibility and quality of life
yoga	reduces anxiety and stress; improves general wellbeing and quality of life

Some Australian cancer centres have developed “integrative oncology” services where evidence-based complementary therapies are combined with conventional treatments to create patient-centred cancer care that aims to improve both wellbeing and clinical outcomes.

Let your doctor know about any therapies you are using or thinking about trying, as some may not be safe or evidence-based.

- For a free copy of Cancer Council’s booklet *Understanding Complementary Therapies* visit your local Cancer Council website or call 13 11 20.

Alternative therapies are therapies used *instead* of conventional medical treatments. These are unlikely to be scientifically tested and may prevent successful treatment of the cancer. Cancer Council does not recommend the use of alternative therapies as a cancer treatment.

Nutrition and exercise

If you have been diagnosed with appendix cancer or PMP, both the cancer and treatment will place extra demands on your body. Research suggests that eating well and exercising can benefit people during and after cancer treatment.

Eating well and being physically active can help you cope with some of the common side effects of cancer treatment, speed up recovery, improve sleep, and help improve your quality of life by giving you more energy, keeping your muscles strong, helping you maintain a healthy body weight and boosting your mood.

You can discuss individual nutrition and exercise plans with health professionals such as dietitians, exercise physiologists and physiotherapists.

- For free copies of Cancer Council’s booklets on *Nutrition and Cancer* and *Exercise for People Living with Cancer* visit your local Cancer Council website or call 13 11 20.

Side effects of treatment

All treatments can have side effects. The type of side effects that you may have will depend on the type of treatment and where in your body the cancer is. Some people have very few side effects and others have more. Your specialist team will discuss all possible side effects, both short and long-term (including those that have a late effect and may not start immediately), with you before your treatment begins.

One issue that is important to discuss before you undergo treatment is fertility, particularly if you want to have children in the future. This is especially the case if during your abdominal or pelvic surgery:

- your seminal vesicles are severed (in males)
- your uterus, ovaries and/or fallopian tubes are removed (in females).

› For a free copy of Cancer Council's booklet *Fertility and Cancer* visit your local Cancer Council website or call 13 11 20. To speak to a health professional about treatment side effects call 13 11 20.

Common side effects

Surgery: general	Bleeding, damage to nearby tissue and organs (including nerves), drug reactions, pain, infection after surgery, blood clots, weak muscles (atrophy), lymphoedema
Surgery: CRS or peritonectomy	Bowel leaks and slow return to normal bowel function, dehydration, loss of fertility (severing of seminal vesicles or removal of uterus, ovaries or fallopian tubes)
Chemotherapy	Fatigue, loss of appetite, nausea, bowel issues such as constipation or diarrhoea, hair loss, mouth sores, skin and nail problems, increased chance of infections, loss of fertility
Radiation therapy	Fatigue, loss of appetite, nausea, bowel issues such as diarrhoea, abdominal cramps and excess wind, bladder issues, hair loss, dry mouth, skin problems, lymphoedema, loss of fertility

Making decisions about treatment

It can be difficult to know which treatment is best for you. It is important to speak with a specialist team before making your decision. Ask them to give you a plan of your treatment options, including information about side effects. Some people prefer to seek several opinions before feeling confident to go ahead with the treatment. If you are confused or want to check anything, ask your specialist questions. This will make sure you have all the information you need to make decisions about treatment and your future that you are comfortable with.

You may have to attend many appointments. It's a good idea to take someone with you. They will be able to listen, ask questions and remember what the doctor says. It may help to take a list of questions with you, take notes or ask the doctor if you can record the discussion (many mobile phones have a recording function or you can use the CAN.recall app – visit rarecancers.org.au for more information). There are some suggestions for questions you could ask at the end of this sheet.

› Cancer Council's podcast *Making Treatment Decisions* can be downloaded from your local Cancer Council website. For a free copy of Cancer Council's booklet *Cancer Care and Your Rights* visit your local Cancer Council website or call 13 11 20.

Looking after yourself

There is no right way to feel if you have been diagnosed with a cancer such as appendix cancer or PMP. Feeling a range of emotions is normal and you may feel overwhelmed, anxious, fearful, angry, sad or lonely. Many people need emotional support before, during and after treatment. Adjusting to living with visible scars, changes to your physical appearance, changes to your lifestyle and changes to bodily functions can be hard and take time. If this is affecting you or likely to affect you, it's important to seek help.

It can help to talk things through with a counsellor, psychologist, friend or family member. Talk to your medical team or call Cancer Council 13 11 20 about what support services are available.

› For a free copy of Cancer Council's booklet *Emotions and Cancer* visit your local Cancer Council website or call 13 11 20.

Practical and financial support

There are many services that can help you manage with practical or financial issues caused by appendix cancer and PMP. Benefits, pensions and hardship programs can help pay for prescription medicines (for example the Australian Government's Pharmaceutical Benefits Scheme [PBS]), transport and parking costs or utility bills. Ask the hospital social worker which services are available in your local area and if you are eligible to receive them.

For additional income, you may be able to access your superannuation early in certain circumstances, or claim on insurance policies such as income protection, trauma, and total and permanent disability (TPD).

Managing your ability to work or study, particularly during cancer treatment, is important to consider and will depend on your personal situation.

› For free copies of Cancer Council's booklets on *Cancer and Your Finances* and *Cancer, Work & you* visit your local Cancer Council website or call 13 11 20.

✓ If you need legal or financial advice, you should talk to a qualified professional about your situation. Cancer Council offers free legal and financial services in some states and territories for people who can't afford to pay – call **13 11 20** to ask if you are eligible.

Life after treatment

Once your treatment has finished, you will have regular check-ups to confirm that the cancer hasn't come back. Ongoing surveillance for appendix cancer and PMP involves a schedule of ongoing scans and physical examinations. It's important to let your doctor know immediately of any health problems between visits.

Some cancer centres work with patients to develop a "survivorship care plan" which usually includes a summary of your treatment, sets out a clear schedule for follow-up care, lists any symptoms to watch out for and possible long-term side effects, identifies any medical or psychosocial problems that may develop and suggests ways to adopt a healthy lifestyle going forward. Maintaining a healthy body weight, eating well and being physically active are all important.

If you don't have a care plan, ask your specialist for a written summary of your cancer and treatment and make sure a copy is given to your GP and other health care providers.

› For a free copy of Cancer Council's booklet *Living Well After Cancer* visit your local Cancer Council website or call 13 11 20.

If the cancer comes back

For some people appendix cancer and PMP do come back after treatment, which is known as a recurrence. This is most likely to happen within the first five years after treatment. If the cancer does come back, treatment will depend on where the cancer has returned to in your body and may include a mix of surgery, chemotherapy and radiation therapy.

In some cases of advanced cancer, treatment will focus on managing any symptoms, such as pain, loss of appetite and improving your quality of life, without trying to cure the disease. This is called palliative treatment.

Palliative care can be provided in the home, in a hospital, in a palliative care unit or hospice, or in a residential aged care facility. Services vary, because palliative care is different in each state and territory.

When cancer is no longer responding to active treatment, it can be difficult to think about how and where you want to be cared for towards the end of life. However, it's essential to talk about what you want with your family and health professionals, so they know what is important to you. Your palliative care team can support you in having these conversations.

› For free copies of Cancer Council's booklets on *Understanding Palliative Care*, *Living with Advanced Cancer* and *Facing End of Life* visit your local Cancer Council website or call 13 11 20. You can also listen to our podcast series *The Thing About Advanced Cancer.*

Understanding Appendix Cancer and Pseudomyxoma Peritonei (PMP)

Questions for your doctor

Asking your doctor questions will help you make an informed choice. You may want to include some of the questions below in your list.

- How far has the cancer spread? What stage of cancer do I have?
- Have you treated this type of cancer before?
- What are the treatment options for me? What do you recommend and why?
- How will you consult with other specialists for my treatment?
- How long will treatment take?
- Is it possible to have surgery to try to cure my cancer?
- If I can have surgery, which type will I need and why?
- Do you use HIPEC only during surgery, or do you use EPIC afterwards for several days?
- What type of rehabilitation will I need after my surgery? How long will recovery take?
- What are the possible risks and side effects of my treatment? How will these be managed?
- Is this treatment covered by Medicare/private insurance? Will there be extra expenses?
- Are there any complementary therapies that might help me?

If you are thinking of taking part in a clinical trial, here are some questions you could ask:

- What are the possible benefits and risks to me?
- What is being tested and why?
- How many people will be involved in this trial?

Reference

1. McCusker M E, Coté T R, Clegg L X, Sobin L H. Primary malignant neoplasms of the appendix: a population-based study from the surveillance, epidemiology and end-results program, 1973–1998. Cancer. 2002;94(12):3307–3312.
2. Smeenk RM, van Velthuysen ML, Verwaal VJ, Zoetmulder FA. Appendiceal neoplasms and pseudomyxoma peritonei: a population based study. Eur J Surg Oncol. 2008;34:196–201.

A web-based resource for Australians with less common cancers project is a Cancer Australia Supporting people with cancer Grant initiative, funded by the Australian Government. Website: canceraustralia.gov.au

Acknowledgements

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Support services

Australia

- Cancer Council: visit your local Cancer Council website (see below for details) or call **13 11 20**
- Rare Cancers Australia**: rarecancers.org.au** or call **1800 257 600**
- NeuroEndocrine Cancer Australia: **neuroendocrine.org.au** or call **1300 287 363**
- Without a Ribbon **–** Fighting for rare cancer support: **withoutaribbon.org**
- Talk to a nurse, social worker or Cancer Council **13 11 20** about what is available in your area.

International

- Appendix Cancer/Pseudomyxoma Peritonei Research Foundation (ACPMP): **acpmp.org**
- Pseudomyxoma Survivor: **pseudomyxomasurvivor.org**

Where to get help and information

Call Cancer Council **13 11 20** for more information about appendix cancer and PMP. Trained health professionals can listen to your concerns, provide information, and put you in touch with local services and support groups. Ask for free copies of booklets that may be relevant to you, or download digital versions from your local Cancer Council website:

ACT	**actcancer.org**
NSW	**cancercouncil.com.au**
NT	**cancer.org.au/nt**
QLD	**cancerqld.org.au**
SA	**cancersa.org.au**
TAS	**cancertas.org.au**
VIC	**cancervic.org.au**
WA	**cancerwa.asn.au**
Australia	**cancer.org.au**

Cancer Council
13 11 20

For information and support on cancer-related issues, call Cancer Council **13 11 20**. This is a confidential service.