Pancreatic Cancer UK

Family history of pancreatic cancer

This fact sheet explains family history of pancreatic cancer. This isn't common, but sometimes pancreatic cancer can run in families. If you are worried about a family history of pancreatic cancer and how this might affect you, speak to your GP.

You can also speak to our specialist nurses on our confidential Support Line about any questions you have about family history of pancreatic cancer. Call free on **0808 801 0707** or email **nurse@pancreaticcancer.org.uk**

Contents

Key facts

- Sometimes cancers are said to 'run in the family'. This means there is a faulty gene in the family that is linked to a type of cancer.
- Genes carry information that controls how our bodies work. We inherit genes from our parents.
- Occasionally, changes in genes mean that someone is more likely to get a disease. These changes are called a fault or mutation. These faults may be inherited from a parent, though they are more often caused by other things like smoking.
- In most cases pancreatic cancer doesn't run in families.
- If pancreatic cancer does run in a family (familial pancreatic cancer – see page 3), there may be two or more first degree relatives with pancreatic cancer. Or there may be three or more relatives with pancreatic cancer on the same side of the family. First degree relatives are your parents, brother, sister or child.
- Between 5 and 10 in 100 pancreatic cancers (5-10%) may be caused by a rare genetic condition. These conditions are sometimes called family cancer syndromes (see page 4).
- Pancreatic cancer can also run in families if a family cancer syndrome has been diagnosed, and there is at least one family member with pancreatic cancer.
- Most of the family cancer syndromes linked to pancreatic cancer increase the risk of pancreatic ductal adenocarcinoma (the most common type of pancreatic cancer).
- A very small number of family cancer syndromes can increase the risk of pancreatic neuroendocrine cancers (also known as pancreatic neuroendocrine tumours or NETs).
- Hereditary pancreatitis is a rare type of pancreatitis that runs in families. Pancreatitis is inflammation of the pancreas. People with hereditary pancreatitis may be more likely to get pancreatic cancer (see page 6).
- If you think pancreatic cancer runs in your family, speak to your doctor. If you do have a high risk, you may be able to have monitoring (see page 7).

What are genes?

Genes carry the information that controls our appearance and how our bodies work. Genes come in pairs. We inherit one copy of each gene from our mother and the other from our father. We all have tiny differences in our genes that make us individual. For example, genes determine the colour of our eyes and hair.

Faulty genes

Occasionally, there may be changes in genes which mean that someone is more likely to get a disease. This type of change in a gene is often called a fault or mutation. Having a faulty gene doesn't always mean someone will develop cancer.

Changes in genes can be inherited from a parent. Changes to genes can also happen during our lifetime, and these changes aren't inherited from our parents. They may be caused by a random mistake when a cell divides. Or they may be caused by other things, such as smoking. These are much more common causes of cancer than inherited genetic faults.

Read more about the different risk factors for pancreatic cancer on our website at: **pancreaticcancer.org.uk/riskfactors**

Hereditary pancreatic cancers

On the next few pages, there is information about pancreatic cancer that runs in families (familial pancreatic cancer) and the small number of genetic conditions linked to a higher risk of pancreatic cancer.

Familial pancreatic cancer

Familial pancreatic cancer is pancreatic cancer that runs in families. Working out whether a family has this higher risk is not easy, but strong signs are:

- families with two or more first degree relatives (parent, brother, sister or child) with pancreatic cancer
- families with three or more relatives with pancreatic cancer on the same side of the family.

It is possible for families to have several cases of pancreatic cancer just by chance. But the more cases there are in a family, the more likely it is to be familial pancreatic cancer.

Pancreatic cancer can also run in families if a known family cancer syndrome (see below) has been diagnosed, and you have at least one family member with pancreatic cancer.

The chance that your family has familial pancreatic cancer is very low. Signs are:

- relatives diagnosed with pancreatic cancer under the age of 60
- more than two family members under 60 with pancreatic cancer
- people with pancreatic cancer in more than one generation on the same side of the family.

If you think pancreatic cancer runs in your family, speak to your doctor. If you do have a high risk of familial pancreatic cancer, you may be able to have monitoring (see page 7).

Family cancer syndromes

Family cancer syndromes are rare genetic conditions. Only between 5 and 10 in 100 pancreatic cancers (5-10%) may be caused by one of these conditions.

Most of the family cancer syndromes linked to pancreatic cancer increase the risk of pancreatic ductal adenocarcinoma. This is the most common type of pancreatic cancer.

There are also rare family cancer syndromes that are linked to an increased risk of pancreatic neuroendocrine cancers. You may hear these cancers called pancreatic neuroendocrine tumours or NETs. Read more about this on page 10.

Peutz-Jeghers syndrome

This causes lots of non-cancerous growths (polyps) in the digestive system, and dark spots on the hands and face. 9 out of 10 cases (90%) are caused by a fault in a gene called STK11. This condition mainly increases the risk of bowel and breast cancer but may also increase the risk of pancreatic cancer.

The NICE guidelines for pancreatic cancer (see page 7) recommend monitoring for pancreatic cancer if you have Peutz-Jeghers syndrome.

Faults in the BRCA2 and BRCA1 genes

Both men and women can have faults in the BRCA2 and BRCA1 genes. Faults in these genes greatly increase the risk of breast and ovarian cancer. They also increase the risk of other cancers, including prostate and skin cancer (melanoma).

In families with one or more cases of pancreatic cancer (but few cases of breast or ovarian cancer), a fault in the BRCA2 gene increases the risk of pancreatic cancer. In families with breast and ovarian cancer, a fault in the BRCA2 gene slightly increases the risk of pancreatic cancer.

There's less evidence that a fault in the BRCA1 gene increases the risk of pancreatic cancer. The NICE guidelines (see page 7) recommend monitoring for pancreatic cancer if you have a fault in BRCA2 or BRCA1 and one or more first degree relatives (parent, brother, sister or child) with pancreatic cancer.

Most pancreatic, breast, ovarian, prostate and skin cancers aren't caused by faults in BRCA2 or BRCA1. So if someone in your family has one of these cancers, it does not mean you have a higher risk of developing one of them.

BRCA Umbrella have an online forum for people with a fault in a BRCA gene. Find their contact details on page 13.

Faults in the PALB2 gene

PALB2 is a gene linked to the BRCA2 gene. A fault in this gene increases the risk of breast cancer and slightly increases the risk of pancreatic cancer.

The NICE guidelines (see page 7) recommend monitoring for pancreatic cancer if you have a fault in PALB2 and one or more first degree relatives with pancreatic cancer.

Familial atypical multiple mole and melanoma syndrome (FAMMM)

This is a condition caused by a fault in genes called CDKN2A (p16) and CDK4. People with FAMMM have large numbers of unusual moles. They also have a higher risk of skin cancer (melanoma) and pancreatic cancer.

The NICE guidelines (see page 7) recommend monitoring for pancreatic cancer if you have a fault in CDKN2A (p16) and one or more first degree relatives with pancreatic cancer.

Lynch syndrome

Lynch syndrome is caused by faults in one of several genes – MLH1, MSH2, MSH6 or PMS2. Lynch syndrome increases the risk of bowel cancer. It also slightly increases the risk of some other cancers, including pancreatic cancer. You may hear of Lynch I or Lynch II. Lynch II is the form of the syndrome that may be linked with pancreatic cancer.

The NICE guidelines (see page 7) recommend that monitoring for pancreatic cancer should be considered if you have Lynch syndrome and any first degree relatives with pancreatic cancer.

Lynch Syndrome UK support people with the condition. Find their contact details on page 13.

Li-Fraumeni syndrome

This is usually caused by a fault in a gene called TP53. It increases the risk of several cancers, including breast cancer, brain tumours and leukaemia (a type of blood cancer). It may also increase the risk of pancreatic cancer.

Hereditary pancreatitis

Pancreatitis is inflammation of the pancreas. It can cause severe tummy pain, which often needs treating in hospital. Over time it may damage the pancreas, which can cause problems, for example with digesting food.

Hereditary pancreatitis is pancreatitis that runs in families. It is rare. It is linked to a fault in the PRSS1 gene. People who have this faulty gene have a very high chance of developing pancreatitis. The pancreatitis starts in early childhood and can keep coming back.

People with hereditary pancreatitis may be more likely to get pancreatic cancer. About 2 in 5 (40%) people with hereditary pancreatitis may develop pancreatic cancer at some point in their lives. The risk may be higher for people who smoke and people who have diabetes.

If you have hereditary pancreatitis, speak to your doctor. You may be able to have monitoring for pancreatic cancer. Read more about monitoring on page 7.

Hereditary pancreatitis may lead to chronic (long term) pancreatitis. But in most cases chronic pancreatitis is caused by other things that are not inherited. Chronic pancreatitis may also increase the risk of pancreatic cancer, although the risk is low.

What should I do if I think I have a family history of pancreatic cancer?

If you think you might have a family history of pancreatic cancer, talk to your doctor, especially if you have any worrying symptoms. Give them as much information as possible about any pancreatic cancer or family cancer syndromes in your family. Ask whether you should be referred:

- to a specialist in pancreatic disease
- to the regional genetics service (see page 8)
- to the EUROPAC study (see below).

Is screening available for people with a family history of pancreatic cancer?

Screening aims to identify people who will get a disease before they get any symptoms. The aim is to pick up cancers early, which means treatment may be more successful. At the moment there is no screening programme in the UK for pancreatic cancer because there isn't a test for pancreatic cancer that is reliable and accurate enough. But NICE guidelines (see below) do recommend monitoring for people at higher risk of pancreatic cancer. This includes some people with a family history.

People with a family history of pancreatic cancer may also be able to take part in the EUROPAC study. This study monitors people with a family history of pancreatic cancer (pancreatic ductal adenocarcinoma). It is also monitoring people with hereditary pancreatitis. It aims to develop a way to screen for pancreatic cancer in people who may be at higher risk.

Search for EUROPAC using our clinical trial finder on our website at: **pancreaticcancer.org.uk/clinicaltrials**

What do the NICE guidelines recommend?

The National Institute for Health and Care Excellence (NICE) guidelines for pancreatic cancer recommend that monitoring for pancreatic cancer (pancreatic ductal adenocarcinoma) should be offered to people with:

- hereditary pancreatitis and a fault in the PRSS1 gene
- faults in the BRCA1, BRCA2, PALB2 or CDKN2A (p16) genes and one or more first degree relatives with pancreatic cancer
- Peutz-Jeghers syndrome.

NICE also recommend that monitoring for pancreatic cancer should be considered for people with:

- two or more first degree relatives with pancreatic cancer, across two or more generations
- Lynch syndrome and any first degree relatives with pancreatic cancer.

Monitoring means having regular MRI/MRCP (magnetic resonance cholangio-pancreatography) or EUS (endoscopic ultrasound) scans. People with hereditary pancreatitis should be offered CT scans rather than MRI/MRCP or EUS.

If you think you fit any of these groups, speak to your doctor about being monitored.

Read more of the NICE guidelines for pancreatic cancer on our website at: **pancreaticcancer.org.uk/NICE**

Find out more about tests, such as an MRI/MRCP or EUS, at: **pancreaticcancer.org.uk/tests**

Can I have a genetic test for pancreatic cancer?

People with familial pancreatic cancer may be referred to a specialist genetics clinic. If you have a genetic condition in the family that is linked to a higher risk of pancreatic cancer (family cancer syndrome), you may also be referred.

The genetics clinic will work out how likely it is that there is a faulty gene in the family. Depending on this risk assessment you may be:

- offered a genetic test if you do have a higher risk and if a test is appropriate
- told you have a higher risk but that genetic testing isn't appropriate, based on your family history or because you have no living relative with pancreatic cancer who can be tested first
- told that you aren't at any higher risk than the general population.

Genetic tests look for faults in the few genes that we know can be linked to familial pancreatic cancer. Testing is usually first offered to someone in the family who has developed cancer. If a genetic fault is found, relatives who don't have cancer can be offered a blood test to look for the same genetic fault.

If you are referred to a genetics clinic, you should first be offered a genetic consultation with a genetic counsellor or genetics doctor. They will provide information about an inherited condition, and the risk of developing it or passing it on. This helps to prepare you for what it means if you find out you have a faulty gene that greatly increases your risk of pancreatic cancer.

If you are offered a genetic test after you have talked to a specialist, it is up to you to decide whether to have it. You can take as much time as you need to decide.

If the test shows that you have a fault in a gene that may cause pancreatic cancer, you should then be told about regular monitoring. This may be through the EUROPAC research study.

If you are referred to a genetics clinic it is helpful to find out the following information beforehand:

- how everyone in the family is related to you and to each other
- how old each relative with pancreatic cancer is and their age when they were diagnosed
- what other cancers have been diagnosed in the family.

Questions to ask

To get the most out of your genetics consultation you might also want to think beforehand about questions to ask the genetic counsellor. You might find these questions helpful.

- What is my risk of getting a particular cancer and what is the general population risk?
- How accurate are the tests that you do?
- Can I do anything to reduce my risk?
- Who else in my family can be offered testing to see if they are at greater risk?
- What about the risk to my children?
- Will a test and monitoring be automatically offered to me?
- If not, what should I do?
- What support will I receive once I get the results of the test?

Hereditary pancreatic neuroendocrine cancers

Pancreatic neuroendocrine cancers start in neuroendocrine cells in the pancreas that produce hormones. You may also hear them called pancreatic neuroendocrine tumours or pancreatic NETs. Only about 5 in 100 (5%) pancreatic cancers are neuroendocrine cancers.

Some neuroendocrine tumours may be linked to a family cancer syndrome. We explain the different syndromes below. If your family is affected by one of these, ask your doctor what screening and support is available. Or contact Neuroendocrine Cancer UK, who have information and support for anyone affected by hereditary neuroendocrine cancers. Find their contact details on page 13.

Read more about pancreatic neuroendocrine cancers on our website at: **pancreaticcancer.org.uk/nets**

Multiple endocrine neoplasia disorders

These are inherited conditions that cause tumours in more than one endocrine gland. Endocrine glands produce hormones – the pancreas is an endocrine gland. Some types of multiple endocrine neoplasia can cause pancreatic neuroendocrine tumours.

Multiple endocrine neoplasia type 1 (MEN1) is a rare condition caused by faults in the MEN1 gene. Tumours may develop in different glands, including the pancreas, the parathyroid gland and the pituitary gland. Up to three quarters (75%) of people with MEN1 may develop pancreatic neuroendocrine cancer.

People with MEN1 should have regular monitoring (screening) to check for tumours. Check-ups should include annual blood tests and MRI, EUS and/or CT scans every 1-3 years. Read more about screening and genetic testing for pancreatic cancer on pages 7 and 8.

Multiple endocrine neoplasia type 4 (MEN4) may be even rarer than MEN1, although there has not been much research into it. It is caused by faults in the CDNK1B gene. It can cause tumours in the pancreas, and in the pituitary and parathyroid glands. Pancreatic neuroendocrine tumours are less common in people with MEN4 than in people with MEN1.

The Association for Multiple Endocrine Neoplasia Disorders (AMEND) offer information and support. Find their contact details on page 13.

Find out more about tests, such as an MRI scan, CT scan or EUS, on our website at: **pancreaticcancer.org.uk/tests**

von Hippel-Lindau (VHL) syndrome

von Hippel-Lindau (VHL) syndrome is caused by a fault in the VHL gene. It can cause tumours to develop in different places in the body, including the central nervous system, kidneys, eyes and pancreas. Most tumours linked with VHL are not cancerous.

Around half of people with VHL may have cysts in the pancreas, which don't usually cause any symptoms. Some cysts can become cancerous so cysts should be assessed to check the risk of them becoming cancer.

Up to 20 out of 100 people (20%) with VHL may develop one or more pancreatic neuroendocrine tumours. These tumours are usually non-functioning. This means they don't produce more hormones than normal and generally don't cause symptoms.

Screening for people with faults in the VHL gene should begin from early childhood. People diagnosed with VHL and those with a faulty VHL gene should have an ultrasound or MRI scan of their tummy every year, starting from their mid-teens. These scans check for tumours in the kidneys, adrenal glands, pancreas and pelvis. Read more about screening and genetic testing for pancreatic cancer (on pages 7 and 8).

VHL UK/Ireland provide information about support for people with von Hippel-Lindau syndrome. Find their contact details on page 13.

Further information and support

Pancreatic Cancer UK services

We are here for everyone affected by pancreatic cancer.

Our specialist nurses are here to talk now

If your world has been turned upside down by a pancreatic cancer diagnosis, we are here to talk now. We can answer your questions, recommend practical steps and provide the emotional support you and those close to you need, when you need it most.

Call free on **0808 801 0707** or email **nurse@pancreaticcancer.org.uk**

Expert information

Our free information covers everything about pancreatic cancer to help you understand your diagnosis, ask questions, make decisions and live as well as you can.

Go to: **pancreaticcancer.org.uk/information**

Download or order our free publications at:
pancreaticcancer.org.uk/publications or call **0808 801 0707**

Our online forum

The forum is a supportive online space where everyone affected by pancreatic cancer can be there for each other at any time.

Go to: **forum.pancreaticcancer.org.uk**

Living with Pancreatic Cancer Online Support Sessions

Our online support sessions are hosted by our specialist pancreatic cancer nurses and will give you the chance to connect with others who have also been diagnosed.

Go to: **pancreaticcancer.org.uk/supportsessions**

Real life stories

Read other people's experiences of pancreatic cancer to find out how they coped with their diagnosis and treatment and their tips on looking after themselves.

Go to: **pancreaticcancer.org.uk/stories**

Useful organisations

Association for Multiple Endocrine Neoplasia Disorders (AMEND)
www.amend.org.uk
Offers support and information for people affected by multiple endocrine neoplasia disorders.

BRCA Umbrella
brcaumbrella.ning.com
Provides practical and emotional support to men and women with a fault in the BRCA gene or at high risk of BRCA related cancers. This includes an online support forum and information.

Cancer Research UK
www.cancerresearchuk.org
Helpline: 0808 800 4040 (Mon-Fri 9am-5pm)
Information for anyone affected by cancer.

Lynch Syndrome UK
www.lynch-syndrome-uk.org
Provides people and families affected by Lynch syndrome with information and support.

Macmillan Cancer Support
www.macmillan.org.uk
Support Line: 0808 808 00 00 (Everyday, 8am-8pm)
Provides practical, medical and financial support for anyone affected by cancer.

Maggie's Centres
www.maggies.org
Centres around the UK and online offer free, comprehensive support for anyone affected by cancer.

Neuroendocrine Cancer UK
www.neuroendocrinecancer.org.uk
Helpline: 0800 434 6476 (Tues-Thurs 10am-4pm)
Provides information and support for people affected by neuroendocrine cancers.

VHL UK/Ireland
vhl-uk-ireland.org
Provides information about support for people with von Hippel-Lindau syndrome.

Pancreatic Cancer UK

This fact sheet has been produced by the Support and Information Team at Pancreatic Cancer UK.

We make every effort to make sure that our services provide up-to-date, accurate information about pancreatic cancer. We hope this will add to the medical advice you have had, and help you make decisions about your treatment and care. This information should not replace advice from the medical team – please speak to your doctor, nurse or other members of your medical team about any questions.

Email us at **publications@pancreaticcancer.org.uk** for references to the sources of information used to write this fact sheet.

Give us your feedback

We hope you have found this information helpful. We are always keen to improve our information, so let us know if you have any comments or suggestions. Email us at **publications@pancreaticcancer.org.uk** or write to our Information Manager at the address below.

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