# Pancreatic Cancer U K

# Types of pancreatic cancer

There are different types of pancreatic cancer. This fact sheet explains the different types, including the most common type, pancreatic ductal adenocarcinoma, neuroendocrine tumours, and rarer types of pancreatic cancer.

If you have any questions about anything you read in this fact sheet, speak to your medical team.



You can also speak to our specialist nurses on our confidential Support Line. Call free on **0808 801 0707** or email **nurse@pancreaticcancer.org.uk** 

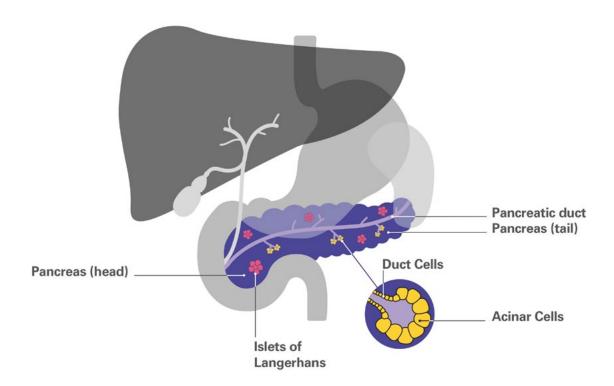
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There are different types of pancreatic cancer. They are divided into two main groups, based on the different types of cells found in the pancreas.

- Exocrine tumours start in the exocrine cells, where enzymes are made. These enzymes help to break down (digest) food. 95 out of 100 (95%) pancreatic cancers are exocrine tumours. The most common type of pancreatic cancer, pancreatic ductal adenocarcinoma (PDAC), is an exocrine tumour (see page 3).
- Neuroendocrine cancers start in the neuroendocrine cells. These cells make hormones that help to control how our bodies work. For example, in the pancreas they make insulin and glucagon that help to control blood sugar levels. You may hear neuroendocrine cancers called pancreatic neuroendocrine tumours, pancreatic NETs, PancNETs or pancreatic neuroendocrine neoplasms (pNENs). Read more on page 4.

## Diagram showing the anatomy of the pancreas



The pancreas can also be affected by pancreatic cysts (see page 8), or by other cancers that grow close to it or inside it (see page 10).

# **Exocrine tumours**

## Pancreatic ductal adenocarcinoma (PDAC)

Pancreatic ductal adenocarcinoma (PDAC) is a type of exocrine pancreatic cancer. It is the most common type of pancreatic cancer. About 95 out of 100 (95%) of all pancreatic cancers are pancreatic ductal adenocarcinoma (PDAC).

PDAC develops from cells lining small tubes in the pancreas called ducts (duct cells in the diagram on page 2). These carry the digestive juices, which contain enzymes, into the main pancreatic duct and then on into the duodenum (first part of the small intestine).

PDAC can grow anywhere in the pancreas, though it is most often found in the head of the pancreas. Symptoms can include tummy and back pain, weight loss, changes to your poo and jaundice.



Read more about the symptoms of PDAC on our website at: pancreaticcancer.org.uk/symptoms

### Rare exocrine cancers

#### Acinar cell carcinoma

About 1 out of 100 pancreatic cancers (1%) are acinar cell carcinoma. Acinar cell carcinoma is more common in men. It develops in the acinar cells at the end of the ducts (see diagram on page 2), which make the digestive enzymes. Symptoms can include tummy pain, weight loss, and feeling and being sick (nausea and vomiting).

### Solid pseudopapillary neoplasms

Solid pseudopapillary neoplasms grow mostly in the body and tail of the pancreas. They are more common in younger women and are also the most common pancreatic tumour in children. Symptoms can include a lump in the tummy, tummy pain, weight loss and sickness.

#### **Pancreatoblastoma**

This rare type of pancreatic cancer mostly affects children. It is extremely rare in adults.



## Questions to ask your doctor

What type of cancer do I have in my pancreas?

Is it an exocrine or neuroendocrine cancer?

Where in my pancreas is the cancer?

How is this type of cancer treated?



If you have any questions about your cancer, speak to your medical team. You can also speak to our specialist nurses on our Support Line.

# Pancreatic neuroendocrine cancers

Pancreatic neuroendocrine cancers are also known as pancreatic neuroendocrine tumours, pancreatic NETs or PancNETs. You may also hear them called pancreatic neuroendocrine neoplasms (pNENs).

Neuroendocrine cancers develop in neuroendocrine cells, which are found in different organs or parts of the body. Neuroendocrine cells make hormones that help to control how our bodies work.

Pancreatic neuroendocrine cancers start in cells in the pancreas called Islet of Langerhans cells (see diagram on page 2). These cells make hormones, including insulin and glucagon, which control the sugar levels in the blood. There are two main types of pancreatic neuroendocrine cancer:

- neuroendocrine tumours (NETs), which tend to grow slowly
- neuroendocrine carcinomas (NECs), which grow faster and cause symptoms similar to PDAC.

We use the term pancreatic neuroendocrine cancer for both. But remember that they behave differently, so speak to your doctor about whether your cancer is a NET or NEC.

We don't know why most neuroendocrine cancers happen. But a very small number of rare inherited genetic conditions are linked to pancreatic neuroendocrine cancer.



Read more about family cancer syndromes on our website at: pancreaticcancer.org.uk/familyhistory

Neuroendocrine Cancer UK have more detailed information about the different neuroendocrine cancers, including pancreatic neuroendocrine cancer. Find their details on page 14.

Pancreatic neuroendocrine cancers may be called functioning or non-functioning. This depends on whether they produce higher levels of hormones, which cause symptoms. Most are non-functioning. This means that they don't produce more hormones than normal.

## Non-functioning pancreatic neuroendocrine cancers

Most pancreatic neuroendocrine cancers (60-90%) are called non-functioning pancreatic neuroendocrine cancers. They don't make higher levels of hormones. Rarely, they can cause symptoms that are similar to the symptoms of pancreatic ductal adenocarcinoma. These include pain, weight loss, jaundice and diarrhoea.

Non-functioning neuroendocrine cancers may be harder to detect and diagnose than functioning neuroendocrine cancers. This is because they don't cause specific symptoms. They are often diagnosed during tests for another problem.

## **Functioning pancreatic neuroendocrine cancers**

Some pancreatic neuroendocrine cancers produce higher levels of certain hormones, which can cause specific symptoms. These are called functioning neuroendocrine cancers. Up to 30 in 100 (30%) of pancreatic neuroendocrine cancers are functioning.

#### Insulinomas

Insulinomas can occur anywhere in the pancreas. They make too much of the hormone insulin, causing low blood sugar levels. Low blood sugar levels can cause symptoms like dizziness, light headedness, sweating, hunger, confusion and irritability.

#### **Gastrinomas**

Gastrinomas are found in the pancreas and the duodenum (first part of the small intestine). They produce too much of a gut hormone called gastrin. High levels of gastrin means that too much stomach acid is made.

Symptoms may include chest and/or tummy pain, acid reflux, heartburn, diarrhoea and tiredness. These combined symptoms are called Zollinger-Ellison syndrome. Excess gastrin may also cause irritation of the stomach or duodenum, leading to ulcers and bleeding.

Up to 30 in 100 gastrinomas (30%) are linked to a family cancer syndrome called multiple endocrine neoplasia type 1 (MEN1).

## **Glucagonomas**

Glucagonomas are mostly found in the body and tail of the pancreas. They make too much of the gut hormone glucagon, which helps to control blood sugar levels.

Symptoms include a distinct type of skin rash (redness and blisters), particularly on the face, tummy, bottom and feet. Glucagonomas can also cause diabetes (high blood sugar levels), diarrhoea (runny poo), weight loss, changes in mood, anaemia (low levels of red blood cells), blood clots, and a sore mouth and tongue.

#### **VIPomas**

VIPomas are more common in the tail of the pancreas. They make too much of a hormone called vasoactive intestinal peptide (VIP). VIP normally helps to neutralise stomach acid and control how quickly food passes through the bowel.

Symptoms include lots of watery diarrhoea, dehydration, low levels of potassium in the blood (hypokalaemia), tummy pain, bloating, and a flushed face.

#### Other functioning neuroendocrine cancers

Other functioning pancreatic neuroendocrine cancers include:

- Somatostatinomas which produce too much somatostatin. This can lead to symptoms including gallstones, steatorrhoea (pale, oily, poo), anaemia, tummy pain, high blood sugar levels and jaundice.
- ACTHomas which produce too much ACTH (adrenocorticotropic hormone). This can cause symptoms of Cushing's disease, including weight gain, easy bruising, anaemia, depression, increased risk of infection and darkened skin.
- PTHrPomas which produce too much PTHrP (Parathyroid hormone-related protein). This can cause symptoms including high calcium levels, tummy pain, feeling and being sick, constipation, bone pain, osteoporosis, fatigue, and confusion.

• **Ppomas** which produce too much pancreatic polypeptide. This can cause symptoms including diarrhoea, abdominal pain and weight loss.

## Finding out more

Speak to your medical team with any questions about pancreatic neuroendocrine cancers and how they are treated.

Neuroendocrine Cancer UK have more detailed information and support around diagnosing, treating, managing and living with pancreatic neuroendocrine cancer. They have a specialist nurse helpline, online and local support groups, an online community and a dedicated neuroendocrine cancer counselling team. Find their contact details on page 14.



## Questions to ask your doctor or nurse

What type of pancreatic neuroendocrine cancer do I have?

Is it a neuroendocrine tumour or a neuroendocrine carcinoma?

Is it functioning or non-functioning?

How is this type of neuroendocrine cancer treated?

# Pancreatic cysts

Different types of cysts can affect the pancreas. A cyst is a sac filled with fluid. These cysts are usually not cancerous (benign) but some can become cancerous (malignant).

If you have a pancreatic cyst you should be offered a CT and/or MRI/MRCP scan to check that it's not cancer. You may be referred for surgery if the scan shows that:

- you have jaundice and cysts in the head of the pancreas
- the cyst may have a solid part to it, which may suggest that the cyst could be cancer
- the main pancreatic duct is very large.

The NICE guidelines for pancreatic cancer have made recommendations about how pancreatic cysts should be checked for cancer. There are also clinical trials looking at pancreatic cysts.



Read more of the NICE guidelines on our website at: pancreaticcancer.org.uk/nice

Find out more about tests, such as a CT or MRI/MRCP, at: pancreaticcancer.org.uk/tests

## Intraductal papillary mucinous neoplasms

Intraductal papillary mucinous neoplasms (IPMNs) are cysts that are usually not cancerous, although they can become cancerous. IPMNs can develop in the main pancreatic duct (see diagram on page 2) or the smaller ducts in the pancreas. They are more common in people over 50.

IPMNs often don't cause any symptoms and may be found during a scan for another reason. If there are symptoms, these can include tummy pain, weight loss, sickness and jaundice.

Surgery may be an option, but it depends on several things, including the size of the IPMN, how quickly it is growing, and any changes found during regular monitoring.

## Mucinous cystic neoplasms

Mucinous cystic neoplasms (MCNs) are cysts that are usually not cancerous but can become cancerous. MCNs are usually found in the body or tail of the pancreas. They usually affect women.

MCNs often don't cause any symptoms, although some people may have tummy pain or a lump in the tummy. Surgery may be an option, but this depends on several things, including the size of the MCN and how quickly it is growing.

## Serous cystadenomas (also known as serous cystic neoplasms)

Serous cystadenomas (SCAs) are non-cancerous cysts. They may be found anywhere in the pancreas, and mostly affect women over 50. SCAs often don't cause any symptoms and are only found during a scan for another reason. If they do cause symptoms, these may include tummy pain, a lump in the tummy or, rarely, jaundice.

If there are no symptoms, no treatment is needed. If there are symptoms, surgery may be an option.



#### Questions to ask your doctor

What type of pancreatic cyst do I have?

Does it need any treatment?

Will it be monitored to check for any changes?

Could the cyst become cancerous?



If you have any questions about pancreatic cysts, speak to your doctor. You can also speak to our specialist nurses on our free Support Line.

# Other cancers linked with the pancreas

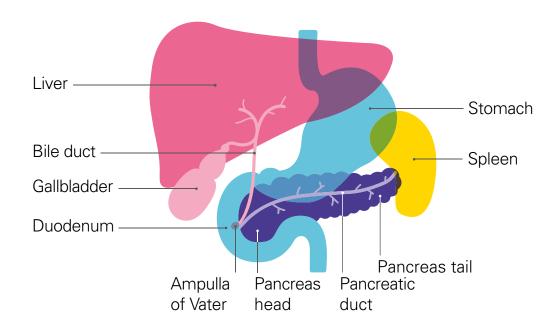
Some cancers grow in structures that are close to or inside the pancreas, such as the bile duct. They cause similar symptoms to pancreatic cancer. You may be offered an operation to remove the cancer, such as a Whipple's operation.



Read more about surgery to remove pancreatic cancer in our fact sheet: Surgery to remove pancreatic cancer Or on our website at: pancreaticcancer.org.uk/surgery

Less commonly, cancer in other parts of the body can spread to the pancreas. Strictly speaking, these cancers are not pancreatic cancers.

## Diagram of the pancreas and surrounding organs



## **Ampullary cancer**

This is found in the ampulla of Vater. This is the area where the pancreatic duct and bile duct meet and empty into the duodenum (see diagram on page 10). These tumours may block the bile duct. This means they often cause jaundice by blocking the flow of bile. They may also block the pancreatic duct, which affects the flow of enzymes into the duodenum.

## Bile duct cancer (cholangiocarcinoma)

Bile duct cancer can occur in any part of the bile duct. The bile duct is a tube that carries bile from the liver to the duodenum (the first part of the small intestine). If bile duct cancer occurs where it passes through the pancreas it is called intrapancreatic bile duct cancer or distal cholangiocarcinoma.

Bile duct cancer can be confused with pancreatic cancer, as it causes similar symptoms, such as jaundice.

AMMF is a charity that supports people with bile duct cancer. Find their contact details on page 14.

## **Duodenal cancer**

The duodenum is the first part of the small intestine and is next to the pancreas. Duodenal cancer (cancer of the duodenum) has symptoms similar to pancreatic cancer. It can block food moving through the duodenum to the rest of the bowel. It can also block the bile duct, causing jaundice.

Sometimes duodenal cancer can be removed without removing the pancreas. But if surgery is possible, the head of the pancreas is often also removed. This is a Whipple's operation, which is used to treat early pancreatic cancer.

## Lymphoma of the pancreas

Lymphoma is a cancer of the body's lymphatic system, which is part of the immune system. It can happen anywhere in the body, including the pancreas, although this is rare. It is called primary pancreatic lymphoma.

Lymphoma Action have more information on lymphoma of the pancreas. Find their contact details on page 14.

## **Metastatic (secondary) cancers**

Sometimes cancer found in the pancreas has spread (metastasised) to the pancreas from another part of the body. This is unusual. But sometimes kidney cancer, bowel cancer, melanoma (skin cancer), breast cancer and lung cancer can spread to the pancreas.

Cancer that has spread from another part of the body is still called by its original name. For example, kidney cancer that has spread to the pancreas will still be kidney cancer, not pancreatic cancer. This means that if you have cancer that has spread to the pancreas, the medical team specialising in that particular cancer will treat you, rather than a team that specialises in pancreatic cancer. But pancreatic surgeons may also be involved, as surgery is sometimes a treatment option.

Macmillan Cancer Support and Cancer Research UK have more information about these cancers.



## Questions to ask your doctor

What type of cancer do I have?

How is this cancer affecting my pancreas?

Who will treat me for this type of cancer?

Where can I find out more about treatment for this type of cancer?

# 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**

## AMMF – The Cholangiocarcinoma Charity

## ammf.org.uk

Email: info@ammf.org.uk

Provides information and support to people with bile duct cancer.

#### **Cancer Research UK**

www.cancerresearchuk.org

Helpline: 0808 800 4040 (Mon-Fri 9am-5pm) Information for anyone affected by cancer.

#### **Healthtalk**

## www.healthtalk.org

Personal experiences presented in written, audio and video formats, including people talking about pancreatic cancer.

## **Lymphoma Action**

www.lymphoma-action.org.uk

**Helpline: 0808 808 5555** (Mon-Fri 10am-3pm)

Provides information and support for people affected by lymphoma, including lymphoma of the pancreas.

## **Macmillan Cancer Support**

www.macmillan.org.uk

**Support Line: 0808 808 00 00** (Every day, 8am-8pm)

Provides practical, medical and financial support for anyone affected by cancer.

## **Maggie's Centres**

www.maggies.org

Telephone: 0300 123 1801

Centres around the UK and online offer free practical, emotional and social support for anyone affected by cancer.

### **Neuroendocrine Cancer UK**

www.neuroendocrinecancer.org.uk

**Call free on 0800 434 6476** (Tues-Thurs 10am-4pm)

Provides information and support for people affected by neuroendocrine cancers.

## Pancreatic Cancer U K

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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