Types of pancreatic cancer fact sheet

This fact sheet is for anyone diagnosed with pancreatic cancer who would like to find out more about the different types of pancreatic cancer. It contains information on exocrine and endocrine pancreatic cancer as well as information about other cancers associated with the pancreas.

Introduction

Pancreatic cancers are grouped into two main types:

- exocrine tumours originating in cells in the exocrine pancreas (produces pancreatic juices containing enzymes)
- endocrine tumours originating from endocrine (hormone producing) cells.

They are also grouped according to where they are in the pancreas. Cancer can grow anywhere in the pancreas, although about 65% of tumours start in the head of the pancreas, 30% in the body and tail, and 5% can involve the whole pancreas.

The exact type of cancer is determined by a pathologist looking at a tissue sample under a microscope.

Exocrine cancers

Exocrine tumours, which occur in the exocrine cells of the pancreas, are the most common form of pancreatic cancer. These tumours account for well over 95% of all pancreatic cancers, and can occur anywhere along the length of the pancreas. Pancreatic ductal adenocarcinoma (PDAC) is the most common type, making up about 90% of all exocrine tumours. Other types of exocrine tumour are rare.
Pancreatic ductal adenocarcinoma (PDAC)

Pancreatic ductal adenocarcinomas develop from cells lining the ducts that carry the digestive juices into the main pancreatic duct and then on into the duodenum (small bowel). They can grow anywhere in the pancreas, although most often they are found in the head of the pancreas. There are several very rare variants of PDAC, including adenosquamous carcinoma and colloid carcinoma.

Pancreas showing ductal cells

![Pancreas diagram](image)

Rare exocrine cancers

**Acinar cell carcinoma**
This rare cancer (no more than 1-2% of pancreatic cancers) develops in the acinar cells that produce and secrete the digestive enzymes. These tumours can produce excessive amounts of pancreatic lipase, the enzyme secreted to digest fats. The level of pancreatic lipase can be measured in the blood. In
these cases the tumours may cause distinct symptoms including unusual skin rashes and joint pain. Acinar cell carcinomas occur mostly in older men.

**Intraductal papillary mucinous neoplasm with invasive carcinoma**
Pancreatic cancer may arise from an intraductal papillary mucinous neoplasm (IPMN). IPMNs form in the main pancreatic duct and/or in its side branches. They can be single or multiple and can occur in the head, body and tail of the pancreas. The tumours grow finger-like, or papillary, projections into the duct and secrete a large amount of mucus which can cause the duct to expand. IPMNs occur most often in older people and are more common in men than women. IPMNs may be benign (non-cancerous) but they can become malignant (cancerous).

**Mucinous cystic neoplasm with invasive carcinoma**
Pancreatic cancer may arise from a mucinous cystic neoplasm (MCN). MCNs are cystic tumours that usually occur in the body or tail of the pancreas. They account for around 1% of exocrine tumours and are almost exclusively found in middle-aged women. The cysts are usually filled with thick mucus. MCNs may be benign but they can become malignant.

**Pancreatoblastoma**
This extremely rare type of pancreatic cancer is predominantly found in children. They can be associated with Beckwith-Wiedemann syndrome.

**Serous cystadenocarcinoma**
These extremely rare tumours develop from benign cysts called serous cystadenomas, which contain watery fluid.

**Solid pseudopapillary neoplasm**
These tumours can occur anywhere in the pancreas. Parts of the tumour are solid and parts are cystic, with finger-like, or papillary, projections. They are mostly found in younger women and are usually not aggressive.

**Pancreatic Neuroendocrine tumours (NETs)**
Pancreatic neuroendocrine tumours start in the hormone producing cells of the pancreas. They account for less than 5% of all pancreatic tumours and are considered rare. These tumours are also known as neuroendocrine tumours (NETs) or islet cell tumours. Some endocrine tumours occur in people with the genetic condition Multiple Endocrine Neoplasia, type 1
(MEN1) or in von Hippel-Lindau (VHL) Syndrome, but most are not related to inherited syndromes (i.e. are sporadic).

In the healthy pancreas endocrine cells (also called islet cells or Islet of Langerhans cells) produce hormones including insulin, glucagon and somatostatin. These hormones regulate different functions in the body, such as blood sugar levels. When the cells grow abnormally they form neuroendocrine tumours.

Neuroendocrine tumours tend to grow much more slowly than exocrine tumours. They may be functioning or non-functioning.

- Functioning tumours overproduce hormones causing a clinical syndrome. They give rise to different symptoms depending on which hormone is being produced.
- Non-functioning tumours secrete hormones but do not produce a clinical syndrome. This means they do not cause specific symptoms.

Endocrine tumours can be benign (non-cancerous) or malignant (cancerous). The behaviour of pancreatic neuroendocrine tumours is usually determined by the size of the tumour, how far the tumour has spread, and how fast the tumour is growing.

**Functioning endocrine tumours**

**Gastrinomas (Zollinger-Ellison Syndrome)**
These tumours occur in the head of the pancreas and the duodenum. They overproduce a hormone called gastrin, which can cause peptic ulcers in the stomach or duodenum. This can result in severe pain, bleeding (causing black, tarry stools) and diarrhoea. Gastrinomas are the second most common of the different functioning endocrine tumours.

Gastrinomas can be associated with a hereditary syndrome called Multiple Endocrine Neoplasia, type 1 (MEN-1) where several tumours develop in different endocrine glands, including the pancreas.

**Glucagonomas**
These tumours overproduce a hormone called glucagon, which normally helps to regulate the amount of glucose (sugar) in the blood. Symptoms include a specific type of skin rash (redness, blistering and scabbing),
anaemia (lack of red blood cells), weight loss and inflammation inside the cheeks and lips. Glucagonomas are most often found in the tail of the pancreas. The tumours often spread outside the pancreas (metastasise), commonly to the liver. Glucagonomas mostly affect post-menopausal women.

**Insulinomas**
These tumours overproduce the hormone insulin, which can lead to symptoms of hypoglycaemia (low blood sugar), such as weakness, loss of energy, dizziness and drowsiness. They are the most common type of sporadic functioning endocrine tumours. Insulinomas tend to occur in middle age and are slightly more common in women than men. Insulinomas can occur in MEN-1 but less commonly than gastrinomas.

**Somatostatinomas**
These tumours overproduce a hormone called somatostatin. This causes gallstones, diabetes, diarrhoea and steatorrhoea (fatty stools which are large, pale, oily, floating and smelly). They are very rare. They occur more commonly in women and in the head of the pancreas.

**VIPomas**
These tumours overproduce a hormone called vasoactive intestinal polypeptide (VIP). This causes a great deal of watery diarrhoea and sometimes flushing of the face. This condition is also known as watery diarrhoea and hypokalaemia achlorhydria (WDHA) or Verner-Morrison Syndrome after the doctors who discovered it. VIPomas occur most frequently in the tail of the pancreas. They are more common in women than men.

**Non-functioning endocrine tumours**

Non-functioning pancreatic endocrine tumours can be harder to detect and diagnose. This is because although they produce certain hormones this is not enough to cause a noticeable collection of symptoms. They are often diagnosed in the course of tests for another problem. Most are found in the head of the pancreas.

**Finding out more**

The NET Patient Foundation is a national charity supporting the neuroendocrine cancer community and has more information on diagnosing and treating NETs and offers support for patients affected by Pancreatic NETs - [www.netpatientfoundation.org](http://www.netpatientfoundation.org)
Questions to ask

- What type of pancreatic cancer do I have?
- Is it an exocrine or endocrine tumour?
- Where in my pancreas is the cancer?

Other cancers associated with the pancreas

There are some cancers that occur in the pancreas are not, strictly speaking, pancreatic cancers. Rather they are associated with the pancreas.

Ampullary cancer

This is also known as a tumour of the ampulla of Vater. It occurs in an area called the ampulla of Vater (after the anatomist who described it). This is a fleshy nipple where the pancreatic duct and bile duct meet and empty into the duodenum. Because of their position at the exit of the bile duct these tumours commonly cause jaundice by restricting or blocking the flow of bile. This means they can be picked up at an early stage, making surgery easier.

Intra-pancreatic bile duct cancer

Bile duct cancer (cholangiocarcinoma) can occur in any part of the bile duct. If it occurs where it passes through the pancreas it is known as intra-pancreatic bile duct cancer. Bile duct cancer is almost always adenocarcinoma and it can be confused with pancreatic cancer, particularly as it causes similar symptoms such as jaundice.

Because these tumours are hard to distinguish from pancreatic cancers and behave in the same way, they are often treated the same. The best treatment is surgery. Doctors aren’t certain how effective chemotherapy is for this type of cancer so clinical trials are taking place to find out.

Lymphoma of the pancreas

Lymphoma is a cancer of the body’s lymphatic system. It can occur anywhere in the body including, rarely, the pancreas. Lymphomas in the pancreas may
be primary or secondary (metastatic) tumours. You can find out more about lymphoma and how it is treated at the Lymphoma Association - http://www.lymphomas.org.uk

Metastatic (secondary) cancers

Sometimes when cancer is found in the pancreas it has actually spread (metastasised) there from another part of the body. Although spread to the pancreas is rare it does occur with the following primary cancers, among others: lung cancer, renal cell carcinoma, breast cancer, colon cancer and melanoma. People with cancers that have spread to the pancreas from another part of the body will be treated by the team specialising in the particular primary cancer.

Questions to ask

- 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 available from Pancreatic Cancer UK

- What is pancreatic cancer?
- Signs and symptoms of pancreatic cancer
- Diagnosing pancreatic cancer

You can find this information on our website www.pancreaticcancer.org.uk/publications

Glossary

You can find an A to Z of some of the common medical words that you may hear when you are finding out about pancreatic cancer on our website – www.pancreaticcancer.org.uk/glossary

This fact sheet has been produced by the Support and Information Team at Pancreatic Cancer UK. It has been reviewed by healthcare professionals and people affected by pancreatic cancer.
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acknowledgement of the health professionals who reviewed the booklet are
available on our website - www.pancreaticcancer.org.uk

Pancreatic Cancer UK makes every effort to make sure that its services
provide up-to-date, unbiased and accurate information about pancreatic
cancer. We hope that this information will add to the medical advice you have
received and help you to take part in decisions related to your treatment and
care. Please do continue to talk to your doctor, specialist nurse or other
members of your care team if you are worried about any medical issues.

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