



# Goblet Cell Carcinoid Tumours

NET Patient Foundation





## Goblet Cell Carcinoid Tumours

These tumours start in the appendix and display features of both a neuroendocrine tumour (NET) and a more aggressive form of cancer called an adenocarcinoma.


One person in every 1,500 who have surgery for acute appendicitis is found to have the tumour.

Neuroendocrine tumours originate from cells called enterochromaffin cells that play a part in the hormonal system of the body, while adenocarcinomas are cancers that start in cells that line the inside of organs and secrete substances, in this case mucus.

Under a microscope, goblet cell tumours are found to contain features of both carcinoid tumours, the most common form of neuroendocrine tumour, and cells originating from the lining of the appendix, called epithelial cells.

The name goblet cell tumour refers to the shape of the cells as they appear under a microscope. The epithelial cells in this case are shaped like miniature wine goblets.

*For further information about carcinoid tumours see the NET Patient Foundation booklet on Carcinoid Tumours.*



## What is known about these tumours?

Around a quarter of all carcinoid tumours originate in the appendix, although the vast majority start elsewhere in the intestinal system.

Goblet cell tumours form a rare subgroup of carcinoid tumours found in the appendix. They were first described and recognised as a different condition as recently as 1969.

Some 20% of patients have a family history of colon cancer.

The tumours are most often found in people over 50, and usually at the time of an operation for another condition, or the removal of the appendix. They usually start in the tip or base of the appendix. In some cases they can cause a bowel obstruction as the tumour infiltrates into the small or large intestine from the appendix.

The current evidence suggests that these tumours behave in a more aggressive way than ordinary carcinoid tumours and are more likely to spread (metastasise) beyond the appendix into the abdomen. If the tumour does spread the most common area affected is the space inside the abdomen (the peritoneal cavity). In women the tumour can spread to the ovaries and the womb, and in some cases this type of cancer can be misdiagnosed as classic ovarian cancer.

## What are the symptoms?

Appendicitis, or complications from a burst appendix, are the most common first symptoms.

However, you may also suffer other problems such as diarrhoea, severe lower abdominal pain, or chronic vague abdominal pain.

## How is it diagnosed?

The tumour in the vast majority of cases is found incidentally during appendix surgery (for acute appendicitis). A sample of the removed appendix will be sent to the pathology lab for analysis by a histopathologist.

To find out more about your tumour you may also be asked to have a:

- **Blood test.** Doctors will be looking for carcinoid tumour markers such as chromogranin A.
- **A 5-HIAA urine test.** Normally, an amino acid called tryptophan, is used by the body to manufacture niacin and certain proteins. In carcinoid patients it can get diverted to produce higher than normal levels of serotonin in the body. This excess can be picked up by a 24-hour urine test called 5-HIAA that stands for 5-hydroxyindoleacetic acid.
- **A PET scan.** This stands for positron emission tomography, and is a specialised scan that produces three-dimensional coloured images of substances functioning within the body tissues. It can provide information about whether the cancer has spread.
- **An octreotide scan (OctreoScan) -** This is a useful diagnostic test for many neuroendocrine tumours, including carcinoid tumours, that have special somatostatin receptors in their surfaces. Octreotide binds with these receptors, and when it is combined with a mildly radioactive ingredient and injected via a vein in the arm it can help reveal the site of tumours under a special scan. The tumours appear 'lit up' on the screen.
- **A CT or MRI scan -** Both computerised tomography (CT) or magnetic resonance imaging (MRI) of the pancreas can determine the position and size of tumours, and regular scans can be useful to find out their rate of growth.

## How is it treated?

The two key treatments are surgery and chemotherapy.

**Surgery:** Because this type of tumour is more aggressive than the usual carcinoid tumours found in the appendix, you will also be asked to consider a bigger operation to remove part of your large intestine to reduce the risk of the disease spreading. This procedure is called a hemicolectomy.

If the spread is to the ovaries then you will be advised to have a hysterectomy. This operation may also be recommended by some doctors even if there is no spread, as a preventative measure.

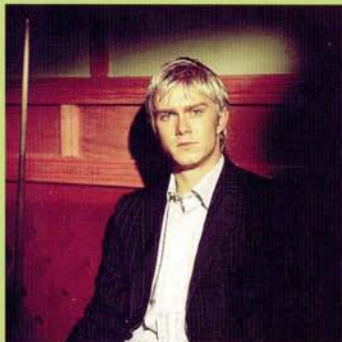
In the case of spread to the abdomen you may be advised to have a right hemicolectomy, or other 'debulking' surgery to remove as much tumour spread as possible.

Occasionally some people have to adjust the type of foods they eat after a hemicolectomy in order to keep their bowels regular. Please talk to one of the nurses or doctors about this if you are in doubt.

**Chemotherapy:** Chemotherapy can help to stabilise tumour growth. Your NET specialist will talk to you about the treatment combinations that research has shown to be the most effective. At the time of writing a common combination is 5- fluorouracil plus cisplatin and streptozotocin.

After diagnosis and treatment you will be asked back for regular monitoring and CT scans to monitor your health.

This booklet is part of the Paul Hunter Information Library, a series of patient friendly publications about neuroendocrine tumours, and related conditions treatments and tests, produced in memory of the late snooker player Paul Hunter, 1979-2006.



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NET Patient Foundation  
incorporating  
Living with Carcinoid



Help and Support for those with 'The Quiet Cancer'  
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The NET Patient Foundation supports people diagnosed with neuroendocrine tumours and their families.

For further information and to make contact telephone 0800 434 6476  
or visit our website:  
[www.netpatientfoundation.com](http://www.netpatientfoundation.com)

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