

Colorectal and peritoneal oncology centre

Goblet cell adenocarcinoma of the appendix

(previously goblet cell carcinoid)

Goblet cell adenocarcinoma of the appendix

Goblet cell adenocarcinoma of the appendix is a rare cancer, occurring in approximately 1 to 2 people per million. The goblet cell adenocarcinomas cover a range of cancers with different types of cells within them. These cancers can behave differently depending on their types. These cancers can spread from the appendix to other parts of the abdomen and have secondary growths that attach to other organs or the inner lining of the abdominal and pelvic cavity which is called the peritoneum. These are called peritoneal metastases. These growths can produce a quantity of a jelly-like substance called 'mucin' or 'mucinous ascites' which can cause swelling of the abdomen. A subset of these cancers can also spread via the lymph nodes and blood to other areas of the body although this is less likely than spread within the peritoneum.

How is cancer of the appendix diagnosed?

It is usually diagnosed as an unsuspected finding during surgery for other reasons, typically an appendicectomy for suspected appendicitis. It may also present as abdominal pain or swelling.

Symptoms of Goblet Cell Adenocarcinoma of the appendix

The primary cancer may have no symptoms. The progression of this disease can be difficult to detect. There are few early symptoms of cancer of the appendix spreading into the abdomen. In most cases the spread to the abdomen is discovered at surgery or on scans.

A few patients have pain in the abdomen and some notice an increase in the size of their abdomen.

Treatment

The internationally accepted treatment for this condition is called Cytoreduction and Hyperthermic IntraPeritoneal Chemotherapy (HIPEC). As one of the 2 national centres that treat cancers arising from the appendix in the UK, this treatment is performed routinely here at The Christie. We do about 155 to 160 such operations a year. We are a recognised training centre for other surgeons by the European Society of Surgical Oncology (ESSO) and the Peritoneal Surface Oncology Group International (PSOGI).

Cytoreduction

The treatment for goblet cell adenocarcinoma that we can offer will vary depending on the extent that the cancer has spread. There is now a large amount of evidence that if the size and spread of the cancer is limited then a special surgical technique called 'cytoreductive surgery' where all the visible disease is removed can be helpful.



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This treatment can involve major surgery to remove organs in the abdomen and the peritoneum. This process removes the cancer as well as removing areas that are at risk of developing disease. This is a long procedure and takes a whole day.

HIPEC

Once the cytoreduction surgery has been performed, heated chemotherapy drugs are then put into the abdomen while you are still under the anaesthetic to kill any cancer cells that cannot be seen. This is a process called hyperthermic intraperitoneal chemotherapy or 'HIPEC'. Using a chemotherapy drug in a heated fluid that is circulated through the abdominal cavity helps to destroy and remove any cancer cells that could otherwise grow into further tumours. The surgeon puts 4 tubes into the side of the abdomen and a machine/pump is used to circulate and heat the chemotherapy which is mixed with fluid. After up to 90 minutes the chemotherapy is washed out and the abdomen is closed in the normal manner.

Two regimens of chemotherapy are used. Most commonly, Mitomycin C is heated to 42 degrees centigrade and put into your abdomen for 90 minutes. The second regime uses a combination drug given to you by an intravenous drip (5-Fluorouracil) as well as the heated abdominal chemotherapy (Oxaliplatin) for 30 minutes.

Side effects

Whilst this is a, commonly performed procedure here at The Christie, cytoreduction and HIPEC is a significant operation to have and consequently complications/side effects can occur. Most of the side effects are from having the operation rather than as a direct result of having the chemotherapy (HIPEC).

In our experience of performing over 1,200 procedures since 2002, the risk of serious complication occurs in about 11% of cases. Such a complication may be in the form of bleeding or infection and may require a return to the oncology critical care unit (OCCU) or theatre. Minor complications e.g. mild infections, wound problems or clots in the calf veins (DVT), occur in about 22% of cases, but these are often managed quickly and easily.

There is a risk of death as a result of this treatment, but our rate is less than 1% within 90 days of surgery in all patients treated.

Side effects from chemotherapy agents (as opposed to the surgery) are minimal but may include:

- Nausea and vomiting (sickness) the severity of this varies from person to person. Anti-sickness medication will be given if necessary. After the operation you will have a tube in your nose that goes into your stomach, which will also help to stop you being sick.
- Prone to infection the chemotherapy in your abdomen may cause a temporary drop in your immune system. This is manifested by a reduction in the white cells in your blood (neutropenia). This is rare and is self-limiting after a few days.
- Delayed healing the chemotherapy may slightly delay the healing process within your abdomen after the surgery.
- Lethargy/tiredness some chemotherapy may make you feel tired and lacking in energy. This, in combination with major surgery, will mean you will need plenty of time to rest and it will be a while before you reach your usual energy levels.
- Diarrhoea this may become a problem. However, it is more likely to be from the surgery itself rather than the chemotherapy.

All of these listed are only possible side effects. You may not experience any of them. Most of the chemotherapy stays in the abdomen rather than going into the blood, therefore most of the potential side effects will relate to the abdomen, for example, being prone to infection in that area, and delayed healing.

Fertility

HIPEC may affect your fertility. If you have not yet had a family or have not completed it please discuss with your consultant and/or clinical nurse specialist (CNS) any fertility concerns prior to your surgery.

- For men, it may be possible to store sperm.
- For women, it may possible to remove eggs, fertilise them with sperm and store them.
- It may also be possible to remove eggs and freeze them so they may be fertilised later.

If you have intercourse post-surgery please ensure you use contraception for the first 6 weeks.

Stomas

After the surgery you might need a stoma. A stoma is where a piece of bowel is brought to the surface of the abdomen. The opening is called a stoma and your bowel motions (stools) pass out of the opening into a bag. This can be temporary but may not be in all cases and this will be discussed further with your surgeon. You will see a stoma nurse prior to the procedure who will support you and provide you with information. They will put a mark on each side of your abdomen in a place suitable for where you position your clothing. It is normal to mark potential positions for stomas before surgery, but this does not mean that one will definitely be created. The prospect of a stoma can be quite daunting and upsetting but your CNS and stoma nurse will ensure they support you through this. Stoma nurses - **0161 445 5001**

Preparation for surgery

You will be asked to come along to a pre-operative assessment clinic where you will meet a nurse who will check you are medically prepared for your operation.

You will have a MRSA (Methicillin Resistant Staphylococcus Aureus) test before your admission to hospital. This involves taking a simple swab of your nose, throat and perineum. This is a routine procedure carried out on all patients who are admitted to The Christie for surgery. You can get more detailed information about MRSA from the infection control team, or ask one of the nurses.

We may ask you to have some pre-operative tests to assess your fitness prior to the surgery. This would include a breathing test (pulmonary function) and an echocardiogram (ultrasound scan of your heart). You may also be asked to have a cardio-pulmonary exercise test (CPX) where you will be asked to ride on a stationary bike breathing into a face mask.

The Christie has an online surgery school which can help you get ready for surgery and will also provide information for post-surgery. You can find this on **www.christie.nhs.uk/surgeryschool.**

If you are particularly anxious about your surgery we have a complementary therapy team who you can be referred to for 'hypno calm' prior to surgery. Please ask your CNS if this is something you would like to be referred for.

If you are a smoker, it would benefit you greatly to stop smoking or cut down before you have your operation. This could reduce the risk of chest problems as smoking makes your lungs sensitive to anaesthetic. If you need help/support in stopping smoking we have a smoking cessation team you can contact **0161 719 7175** or you can contact your GP.

Before you come into hospital for your operation, try to organise things ready for when you go home. If you have a freezer, stock it up with easy-to-prepare food. Arrange for relatives and friends to do your heavy work such as changing your bedding, vacuuming and gardening and to look after your children if necessary.

Support

We know that people who have had a cancer diagnosis can be affected in a variety of ways. For some people there will be physical concerns while others may have emotional or spiritual needs. Practical and financial worries can come to the fore during cancer investigations and treatment and these can put significant strain on people. Most people want to be well informed and involved in decision making but people's information needs vary too.

Everyone is an individual and has their own concerns and needs. You will be offered an holistic needs assessment (HNA) by your CNS which is to help you to identify any concerns or needs that you might have so that we can then support you better. You will then be given a summary of your discussion in the form of a care plan. If you are not offered a an HNA please ask your CNS who will be able to go through this with you.

Your CNS is there to support you throughout your journey and if you have any questions/concerns or would just like to go through things again please do contact them on the number provided below. If appropriate and with your consent, we can refer you to a trained counsellor. We have a counsellor and a consultant psycho-oncologist who specialise in problems relating to cancer. If you feel you may be helped by these services please talk to your CNS to discuss a referral.

We hold an annual patient day in the auditorium at The Christie to support patients with a diagnosis of appendiceal adenocarcinoma. Please ask your CNS for the date of the next event.

Visiting times

When in the OCCU, it is open for visiting but only 2 to 3 visitors are allowed at the bedside at one time. Once on the ward, visiting is 2:00pm 8:00pm and meal times are protected (12:00pm - 1:00pm and 5:00 - 6:00pm).

Consent to treatment

The colorectal and peritoneal oncology team will discuss the treatment that is recommended for you and explain how it will affect you.

We will ask you to sign a consent form agreeing to accept the treatment that you are being offered. The basis of the agreement is that you have had The Christie's written description of the proposed treatment and that you have been given an opportunity to discuss any concerns. You are entitled to request a second opinion from another doctor who specialises in treating this cancer. You can ask your own consultant or your GP to refer you. Your consent may be withdrawn at any time before or during this treatment. Should you decide to withdraw your consent then a member of your treating team will discuss the possible consequences with you.

Leaving hospital and coping at home

Recovery time after your operation varies from person to person. You may find it takes several weeks to feel better. You can expect a gradual improvement over the next 6 to 12 weeks.

Please follow these general advice points:

- You should not attempt to drive, lift or move heavy objects, do heavy housework (for example hoovering, changing the beds) or start digging the garden until a minimum of 6 weeks following your operation.
- Getting back to work will depend on what type of job you do. Please ask if you are unsure. The ward should provide you with a sick note, please ask before you go home. Your GP can then supply you with further sick notes.
- Getting back to normal activities and exercise will depend on you. It is safe to gently increase your levels of physical activity providing it is comfortable to do so.

Follow up after surgery

Your CNS will contact you once you are discharged home to check you are recovering well. We will then see you in The Christie outpatient clinic 4 to 6 weeks following your stay in hospital and you will then be reviewed at regular intervals.

The colorectal and peritoneal oncology centre (CPOC)

The colorectal and peritoneal oncology centre has an international reputation for treating advanced and early colorectal cancer, appendix tumours, peritoneal tumours, anal cancer and tumours within the pelvis.

If you have a query regarding our service, please contact **0161 446 8051** or visit The Christie website **www.christie.nhs.uk/cpoc**

The service will provide:

- ongoing advice and support for patients, their partners and families
- information and advice about treatment and treatment options
- a point of contact should problems arise
- a link with other health care professionals involved in your care at home and in hospital
- referral to specialist services.

Who can contact us?

Any health care professional who needs information or advice and any patient coming for assessment or treatment (for cytoreductive surgery and intraoperative intraperitoneal chemotherapy for appendiceal cancer) can contact the service themselves. They can also be referred by another health care professional. We are happy to speak to partners, friends and family, providing the patient has given consent.

If you know the name of your consultant, please contact their secretary directly.

| Consultant | Secretary |
|-----------------------|---------------|
| Professor S T O'Dwyer | 0161 446 8311 |
| Mr M S Wilson | 0161 446 3366 |
| Professor A G Renehan | 0161 918 2189 |
| Mr C R Selvasekar | 0161 918 2310 |
| Mr O Aziz | 0161 918 2057 |
| Mr H W Clouston | 0161 918 2391 |
| Mr J Wild | 0161 918 7352 |

Clinical nurse specialists:

| Rebecca Halstead (lead) | 0161 918 7096 / 07766 780952 rebecca.halstead@christie.nhs.uk |
|-------------------------|---|
| Rachel Connolly | 0161 918 7859 / 07785 725629 rachel.connolly@christie.nhs.uk |
| Lisa Wardlow | 0161 918 7183 / 07826 892213 lisa.wardlow@christie.nhs.uk |
| Amanda Coop | 0161 918 2097 / 07824 373785 amanda.coop@christie.nhs.uk |

Fax: 0161 918 7078

There will be a CNS available between the hours of 07:30am to 6:00pm.

Further information

For information about the colorectal and peritoneal oncology centre visit www.christie.nhs.uk/cpoc

Cancer information centre

The cancer information centre stocks a wide range of booklets free to patients, their families and carers and offers a free confidential service for anyone affected by cancer. **Telephone 0161 446 8100.**

Complementary therapy and smoking cessation

There is an outpatient drop-in service at The Christie on Tuesday and Thursday. Contact the service directly by calling **0161 446 8236 or 0161 918 7175**.

Maggie's centre

The centre provides a full programme of practical and emotional support, including psychological support, benefits advice, nutrition and head care workshops, relaxation and stress management. Maggie's Manchester is directly across from The Christie on Kinnaird Road. Contact Maggie's on **0161 641 4848** or email **manchester@maggiescentres.org**

Macmillan Cancer Support

This is a national charity offering advice and support. Call the freephone helpline **0808 808 0000** (Monday to Friday, 9:00am to 8:00pm) or if you are hard of hearing, use the text phone **0808 808 0121**. Macmillan publish booklets which are free and available on their website **www.macmillan.org.uk**

Hospital chaplaincy

Situated in department 57, on the first floor above the dining room and next to the education centre. We have a Christian chapel, a Muslim prayer room and a multi-faith quiet space. If you would like someone to visit you on the ward please ask a member of staff.

Useful websites: www.pseudomyxomasurvivor.org (charity and patient forum).

Travel insurance

For help with travel insurance please see **www.medicaltravelcompared.co.uk** or ask your CNS for advice.

We do have a very active research and education programme into colorectal and peritoneal cancer here at The Christie and if you would like further information, please visit our website **www.christie.nhs.uk/cpoc** or ask your specialist nurse.

If you need information in a different format, such as easy read, large print, BSL, braille, email, SMS text or other communication support, please tell your ward or clinic nurse.

The Christie is committed to producing high quality, evidence based information for patients. Our patient information adheres to the principles and quality statements of the Information Standard. If you would like to have details about the sources used please contact **patient.information@christie.nhs.uk**

For information and advice visit the cancer information centres at Withington, Oldham or Salford. Opening times can vary, please check before making a special journey.



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Open 24 hours a day, 7 days a week