

Colorectal and peritoneal oncology centre

Pseudomyxoma peritonei (PMP)

What is pseudomyxoma peritonei?

Pseudomyxoma peritonei (PMP) is a rare, often slow-growing tumour usually starting from the appendix. It produces large amounts of a jelly-like substance (mucus) that collects in the abdomen. The tumour does not spread through the blood but stays in the abdomen increasing in size. This can cause problems with moving and breathing and affects the body's ability to absorb food properly. This disease can be described as a low grade or slow growing cancer.

What causes PMP?

Most people with PMP have a tumour (adenoma) in the appendix. Like many other tumours, this can develop in people who lead healthy lifestyles. There is no clear association within families. It does not appear to be an inherited condition.

Signs and symptoms

For both women and men, the most common symptom is a slow increase in the size of the waist. The increase in the size of the abdomen puts pressure on the gut and prevents people from eating normally. Despite this, people often notice an increase in body weight. The symptoms can be non-specific and are often misdiagnosed.

How is it diagnosed?

PMP is difficult to diagnose. It is often an unexpected finding during investigations of discomfort or swelling in the abdomen. These investigations usually involve an ultrasound or CT scan or you may have had an operation on your abdomen. Women with PMP may sometimes have their diagnosis confused with ovarian cancer and it is not unusual that women are seen by a gynaecologist at first. The ovarian tumour is often the presenting symptom or sign and is sometimes assumed to be the original site. Doctors do not always recognise that the appendix is a possible source of a mucinous tumour.

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 PMP in the UK, this treatment is performed routinely here at The Christie. We do approximately 150 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 we can offer will vary depending on the extent that the PMP has spread. There is now a large amount of evidence that if the size and spread of the PMP is limited then a special surgical technique called 'cytoreductive surgery' where all the visible disease is removed can be helpful. This treatment can involve major surgery to remove organs in the abdomen and the peritoneum. This process removes the PMP as well as removing areas that are at risk of developing disease. This is a long procedure and we set aside the whole day in order to perform it.

HIPEC

Once the cytoreduction has been performed, heated chemotherapy drugs are then put into the abdomen while you are still under the anaesthetic to kill any tumour cells that cannot be seen. This is a process called hyperthermic intraperitoneal chemotherapy or 'HIPEC'. Using a chemotherapy drug in a heated fluid circulated through the abdominal cavity helps to destroy and remove any tumour 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 routine, 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).

The surgery has serious complications reported in the international literature, however our own complication rates compare very favourably to this. In our experience of performing over 1200 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 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.

Unfortunately 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 transient drop in your protection against infection. This is manifested by a reduction in the white cells in your blood (neutropenia). In fact we rarely see this complication and it has always been 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.

What are the benefits of cytoreductive surgery and HIPEC?

The benefits of treatment will depend on whether the surgeon can remove all the visible disease and give intraperitoneal chemotherapy. The aim would be to cure the disease or reduce the chances of the disease coming back.

Are there any alternatives to cytoreductive surgery and HIPEC?

All treatments carry risks as well as benefits and thinking about possible options can help you decide what to do. The team will discuss alternative treatments with you, which may include the following:

Watch and wait: We monitor the situation closely. If the tumour continues to grow we may suggest that you have chemotherapy or surgery.

Debulking surgery: This is when an attempt is made to remove disease that is causing or may cause problems in the near future. This would not be to cure the disease but to deal with particular symptoms.

Chemotherapy: A chemotherapy trial has been carried out for patients unsuitable for cytoreduction. One of the drugs used is the same used in theatre after cytoreductive surgery, Mitomycin C. The other drug comes in a tablet form and is called capecitabine. These drugs are known to be relatively well-tolerated. Patients are monitored with regular follow up including blood tests and CT scans at a dedicated clinic.

What happens if you decide not to have cytoreductive surgery?

There is a risk that your tumour may continue to grow and any symptoms you may have could get worse. You can discuss what to do next with your consultant or specialist nurse.

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) about any fertility concerns prior to your surgery.

- For men, it may be possible to store sperm.
- For women, it may be possible to remove eggs, fertilise them with sperm and store them.
- Or it may 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 (seat area). 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 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 a 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 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 on page 6. 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 PMP. Please ask your CNS for the date of the next event.

Visiting times

When in the oncology critical care unit (OCCU) it is open visiting but only 2 - 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 - 12 weeks.

Please follow these general advice points:

- You should not attempt to drive, lift or move heavy objects, do heavy housework (for example vacuuming, 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 for your hospital stay, 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 - 6 weeks following your stay in hospital and you will then be reviewed at regular intervals.

The colorectal and peritoneal oncology centre

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 our service 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. Any patient coming for assessment or treatment for cytoreductive surgery and intraoperative intraperitoneal chemotherapy for colorectal cancer can contact the service themselves or by referral from another health care professional. We are also 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

Professor S T O'Dwyer

Mr M S Wilson

Professor A G Renehan

Mr C R Selvasekar

Mr O Aziz

Mr H W Clouston

Mr J Wild

Secretary

0161 446 8311

0161 446 3366

0161 918 2189

0161 918 2310

0161 918 2057

0161 918 2391

0161 918 7352

Clinical nurse specialists:

Rebecca Halstead (lead)

0161 918 7096 or 07766 780952
rebecca.halstead@christie.nhs.uk

Rachel Connolly

0161 918 7859 or 07785 725629
rachel.connolly@christie.nhs.uk

Lisa Wardlow

0161 918 7183 or 07826 892213
lisa.wardlow@christie.nhs.uk

Amanda Coop

0161 918 2097 or 07824 373785
amanda.coop@christie.nhs.uk
Fax: 0161 918 7078

If your key worker is not available, please leave a message on the answering machine with your name, date of birth and telephone number. All messages will be responded to as quickly as possible, but this may not always be on the same day.

If you have any problems after your operation, please contact the surgical oncology ward on **0161 446 3860**.

Further information

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

Christie information

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

Complementary therapy and smoking cessation

There is an outpatient drop-in service at The Christie on Tuesday and Thursday from 4pm. Contact 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 the road from The Christie on Kinnaird Road. Open for drop in Monday to Friday 9:00am - 5:00pm. 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 Cancer Support publish booklets which are free and available on their website www.macmillan.org.uk

Hospital chaplaincy

Situated in department 57, on the first floor 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)

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

Travel insurance

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

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.



Contact The Christie Hotline for
urgent support and specialist advice
The Christie Hotline: 0161 446 3658
Open 24 hours a day, 7 days a week