The Christie NHS
NHS Foundation Trust

Colorectal and peritoneal oncology centre

Information for health professionals - palliative care and pseudomyxoma peritonei

What is it?
Pseudomyxoma Peritonei (PMP) is often a slowly progressive disease that produces extensive mucus accumulation within the abdomen and pelvis. There are two clinically distinct groups of peritoneal mucinous lesions. The first can be described as low-grade mucinous carcinoma peritonei (MCP-L) which would include mucin producing disease from an appendiceal adenoma to a well-differentiated mucinous carcinomatosis or well-differentiated variants of mucinous adenocarcinomas. The second group can be described as high-grade mucinous carcinoma peritonei (MCP-H) and would apply to either moderately or poorly differentiated adenocarcinomas.

In PMP, an adenoma arises within the appendix and as it grows it occludes the lumen of the appendix. This can be classified as a low-grade appendiceal mucinous neoplasm (LAMN). The appendix eventually ruptures, leaking mucous containing epithelial cells into the abdominal cavity leading to low-grade mucinous carcinoma peritonei. After the appendix decompresses, the perforation may reseal only to extrude more adenomatous epithelial cells at a later time. Sometimes neither a primary appendiceal tumour nor a normal appendix is apparent. In these cases it may be that the appendix has ruptured and has been obliterated by the developing fibrosis. Pseudomyxoma peritonei is often referred to as being a 'borderline malignant' condition. The tumour is not biologically aggressive because it does not metastasise via the lymphatics or blood stream like gastrointestinal adenocarcinomas. However, it is still a fatal process. The space required within the abdomen and pelvis for nutritional function eventually becomes replaced by mucinous tumour.

Most of these tumour cells are surrounded by fluid of varying consistency. Bulky cellular deposits are usually found within the omentum and beneath the right hemidiaphragm. Gravity creates a further accumulation of adenomucinous cells within the pelvis where the peritoneum reflects over the pelvic organs.

Common sites involved in tumour dispersion also include the stomach, the area around the terminal ileus and the rectosigmoid colon within the pelvis. All three of these sites are fixed to the retroperitoneum and are not free to move as a result of peristaltic activity. The peristaltic activity of the small bowel may prevent mucinous tumour implantation on these surfaces resulting in relative sparing of the small bowel.

What causes PMP?
For the majority of people with true pseudomyxoma an adenoma is found in the appendix. Like many other tumours, pseudomyxoma can occur in people who lead healthy lifestyles.

What are the signs and symptoms of advanced disease?
For women and men the most predominant feature is a gradual increase in abdominal girth. The increase in abdominal girth increases pressure on the gut and prevents the patient from eating normally. Despite this the patient often notes an increase in body weight. As one patient has commented:

"That's when I know its really sort of getting bigger as it were. My skirts go tighter and I get this feeling, especially say I have a meal and I stoop down after it, ooh, it comes up in my mouth"

Another patient complained of weight gain on his abdomen with associated weight loss on his arms and legs:

"*I am slim build, anyway my stomach just slowly got bigger and bigger and that's the only place I was putting weight on and I thought 'well hang on a minute that's a bit odd that''*

The pressure from disease compressing the GI tract can cause early satiety contributing to weight loss. This is especially marked with those patients in whom the stomach is encased with disease preventing distension. We recommend that patients with these symptoms eat small amounts of food frequently throughout the day. The type of foods may also need adjusting to include a higher calorie, lower fibre diet (PMP dietary advice can be found at: **www.christie.nhs.uk/patients/treatments/pmp/diet.aspx**)

Patients with advanced disease sometimes present with sub acute bowel obstruction. This can often initially resolve with conservative management and often requires hospitalisation/hospice management. This rarely leads to a surgical intervention.

As with more common cancers, fatigue is a predominate feature of advanced PMP.

PMP is a rare disease requiring specialist treatment. There are, however, no particular specialist requirements for continuing support other than good general palliative care when treatment becomes ineffectual. The colorectal and peritoneal oncology team can be contacted at the numbers below for advice and support if required. You can also contact The Christie supportive care team for advice on the management of refractory symptoms on **0161 446 3559**.

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 or would like to refer a patient, please contact **0161 446 8051.**

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 care at home and in hospital
- referral to specialist services

Who can contact us?
Any health care professional who requires information or advice and any patient coming for assessment or treatment for cytoreductive surgery and intraoperative intraperitoneal chemotherapy 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:

Consultants:	**Contact:**	
Miss S T O'Dwyer	Eve Kennerley	**0161 446 8311**
Mr M S Wilson	Gill Harrison	**0161 446 3366**
Mr P E Fulford	Rebecca Brown	**0161 918 7352**
Prof A Renehan	Bev Tyrrell	**0161 918 2189**
Mr C R Selvasekar	John Lupea	**0161 918 2310**
Mr O Aziz	Laura Elliott	**0161 918 2057**
Miss A Minicozzi	Marion McKenna	**0161 918 2391**
Clinical Nurse Specialists:	**Contact:**	
Rebecca Halstead	**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**	
Sarah Wemyss	**0161 918 2097 / 07824 373 785** **sarah.wemyss@christie.nhs.uk**	
	Fax: **0161 918 7078**	
Service Manager: Hannah Rogers		

Further information

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

Other useful websites
www.pmpawareness.org (support network)

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.

We try to ensure that all our information given to patients is accurate, balanced and based on the most up-to-date scientific evidence. If you would like to have details about the sources used please contact **patient.information@christie.nhs.uk**

For more information about The Christie and our services, please visit **www.christie.nhs.uk** or visit the cancer information centres at Withington, Oldham or Salford.

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

The Christie NHS Foundation Trust
Wilmslow Road
Manchester M20 4BX
T: 0161 446 3000
www.christie.nhs.uk

The Christie Patient Information Service
February 2017 - Review February 2020

Health & care information you can trust

The Information Standard Certified Member

CHR/CPOC/558/26.02.08 Version 4