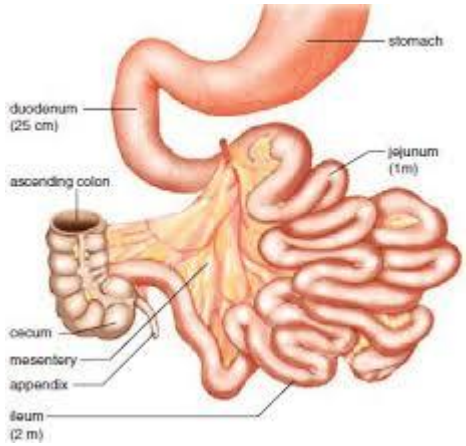
Lynch Syndrome UK
Registered charity no. 161325
www.lynch-syndrome-uk.org
facebook closed group – 1000+ support & information
email team@lynch-syndrome-uk.org

SMALL BOWEL CANCERS IN LYNCH SYNDROME

The Cancer Research UK website has very good information. It is in the section called "small bowel cancer."
http://www.cancerresearchuk.org/about-cancer/small-bowel-cancer

Small bowel cancers are more common in people with Lynch syndrome than in those that do not have it. It seems to affect more people with MLH1, MSH2 and Epcam than people with MSH6 and PMS2.



The small bowel consists of the **duodenum** which is just below the stomach, the **jejunum** which follows on from that and the **small intestines** which are a mass of coiled narrow tubes before the food enters the colon.

The small bowel contains food which is fairly liquid and it absorbs nutriments to sustain the body and releases enzymes and vitamins to absorb and digest food.

Small bowel cancers in Lynch syndrome are not as common as colon and rectal cancers but they are difficult to diagnose and frequently do not produce tell-tale symptoms. That means they can grow to a later stage before being discovered.

In Lynch syndrome, most of the small bowel cancers (at least 50% of them) occur in the duodenum. The rest are spread through the jejunum and small intestines, fewer as you go down.

40% of small bowel cancers are adenocarcinomas.

Statistics

"How many cases of small bowel cancer occur each year in the UK?"

1500

"How many of those are due to Lynch syndrome?"

Probably 10% of those

"What is the risk of getting small bowel cancer for people with Lynch syndrome?"

The risk of getting small bowel cancer for people with MLH1 and MSH2 is approximately **5%** by the age of 70 years. That means that out of 100 people with those LS gene mutations, 5 will get small bowel cancer by that age. This is not regarded as substantial risk and therefore in the UK, screening of the small bowel is not routinely done.

Family history

There is no published evidence that Lynch syndrome small bowel cancer clusters in families, so risk should not be increased just because an LS relative has had this cancer. But there may be other sporadic (or non-LS gene mutations) that influence the formation of cancers and some families do have screening of relatives. Some do have some form of upper gastrointestinal tract (GIT) screening and it may reassure relatives to have at least some part of the small bowel looked at regularly. (see screening)

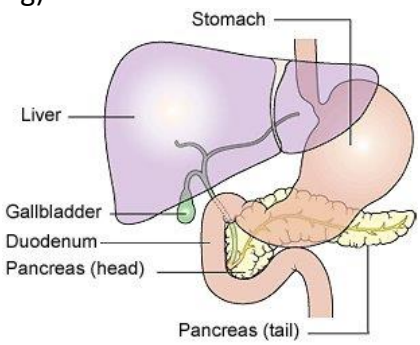


Diagram showing the position of the pancreas

Symptoms

(With thanks to Cancer Help UK for pictures)

Glossary …..Motions = stools/poo/faeces

The duodenum and jejunum are small tubes through which a fairly mushy collection of food and liquid passes through. A cancer there causes symptoms of blockage, pain behind the bottom of the breast bone, bleeding into the small bowel and blockage.

If the cancer occurs in the duodenum at or close to the head of the pancreas, that will produce additional symptoms cause by blockage of the hepatic and pancreatic ducts. Those are the entry points for various enzymes and liquids – bile from the liver and gall bladder and other enzymes such as amylase from the pancreas.

Blockage there might cause, jaundice, indigestion, carbohydrate intolerance and intermittent pain that is worsened by eating. Diarrhoea will often be caused by food malabsorption – producing fatty, smelly motions, pale if there is a halt of bile into the gut but dark black if there is bleeding.

Symptoms in small bowel cancer can include any or all the following: -

Indigestion – vague pain behind the breast bone or just below it, going through to back. Might spread round on the LHS. May be worse with eating and volume of food may have to be reduced.

Pain in epigastrium – the "pit of the stomach" may end up as continuous, not relieved by medications. Can end up as relentless back pain at level of the breasts

Nausea and vomiting – starting as feelings of sickness will eventually increase to being unable to keep food down. Vomiting immediately after food occurs later if the lumen of the small bowel becomes occluded.

Diarrhoea can be an early feature due to food malabsorption if cancer is in the duodenum at site of pancreatic and hepatic ducts. Stools are loose, smelly, pale or dark black from altered blood.

Jaundice – caused by blockage of the hepatic ducts – dark yellow urine, yellow skin and eyes, pale motions.

Paleness and tiredness due to anaemia is as a result of bleeding into the small bowel
Weight loss – unexplained by dieting

Spread and staging in small bowel cancer in Lynch syndrome

All the small bowel is very thin walled so cancers do tend to break through into surrounding tissues quite quickly. Lynch syndrome cancers tend to have *better* prognosis than ordinary "sporadic" cancers until there is spread elsewhere. If it goes into the blood stream it will pass to the local gastrointestinal lymph nodes and the liver.

Lynch syndrome cancers tend to have a better prognosis (at least 10% better by 10 years) because they produce a particular level of "immune response" such a higher level of T-cell lymphocyte infiltration than usual.

Stages

The lower the stage the better. Treatments vary from stage to stage so don't worry if the picture is rather mixed at first.
Macmillan has a good description of staging in small bowel cancer and other information…
http://www.macmillan.org.uk/information-and-support/bowel-cancer/small-bowel#291454

Investigations

Gastoscopy – endoscope into the stomach
Duodenoscopy – endoscope into the duodenum
CT scan
MRI scan
Capsule video-endoscopy – a capsule with a video on it, swallowed and x ray pictures recorded in real time

Treatment
Treatment for small bowel cancer invariably means surgery. If additional treatment is required that will usually be chemotherapy – either before your surgery to shrink the cancer or afterwards to mop all the lumps of cancer cells that have not been identified at surgery or afterwards as a means of mopping up all those and the cancers cells that might have spread through the blood stream. Make sure that you ask whether Lynch syndrome specific chemotherapy will help – it will depend on the histology of your cancer.
Radiotherapy might be used later in the illness to shrink localized areas of cancer.

Survival
Don't read survival figures for small bowel cancer!! They are mostly out of date and don't apply to Lynch syndrome. They can look extremely gloomy so regard your treatment as being successful from the word go. Depression is the enemy of your immune system and you should be positive but also realistic. Get your treatment done as soon as possible and concentrate on getting better.

Screening
At present the UK does not do routing screening of the small bowel in Lynch syndrome. That is because there is not a satisfactory method that is both safe and effective. The following methods are undertaken by some gastroenterologists in some areas. The intervals are usually 1-2 years.

Gastroscopy and "look through"
Because at least 50% of small bowel cancers in LS are in the duodenum, some LS mutation carriers (most often MLH1 and MSH2) with a family history are offered gastroscopy (looking into the stomach) then pushing the scope through into the duodenum for a "look through". This will only pick up duodenal cancers but will catch about 50% of small bowel cancers so is a compromise.

Duodenoscopy
Is similar to the above. It just looks at the duodenum but is a better look.
"Video-cam"
You swallow a special pill – a large pill shaped video cam and carry around a receiver for 24 hours. It then takes a few days for a radiologist to read the pictures.

None of these tests are perfect and some people have had clear results but then a cancer has appeared soon afterwards. (A false negative)

Other people have had false positives – they have been told they have cancer but haven't got it. Therefore if you have MLH1 or MSH2 and have a relative who has had a Lynch related small bowel cancer ask to be referred to a gastroenterologist to discuss whether screening is advised and available at your hospital. You might need to be referred to a teaching hospital if you have a Pill-cam.

Prevention

Take aspirin or be on the CaPP3 trial
Don't eat a very hot spicy type of diet (as in Korea)
Probably will help to keep alcohol at sensible levels
Don't smoke
Make sure your Helicobacter pylori status is checked and treated if positive.