

The Gut in Scleroderma

Symptoms

The symptoms may begin with heartburn, difficulty in swallowing as the oesophagus becomes damaged, and large bowel disorder leading to severe constipation. Then, with small bowel dysfunction, diarrhoea, malabsorption, and possibly nausea, vomiting, and regurgitation with gastric involvement. Fortunately not every patient will develop this kind of involvement but these are the most serious complications that can occur.

In the early stage of scleroderma, one loses the function of the sphincter that separates the stomach from the oesophagus, allowing acid, bile and food to regurgitate back into the oesophagus. This leads to heartburn and sometimes regurgitation of material into the mouth. This is perhaps the most common symptom.

The Oesophagus

Most patients present with symptoms from the oesophagus, the tube joining the mouth to the stomach. Amongst these is the pain caused by acid from the stomach passing upward into the oesophagus (acid reflux), giving rise to heartburn or a recurrent cough, particularly at night. Treatment with drugs which reduce acid production, called proton pump inhibitors (PPIs e.g. omeprazole), are often required to be taken for life. Resistant cases may require double the recommended dose for a few weeks. Ranitidine taken at night may be added and other drugs that suppress acidity may also be required.

Difficulty in swallowing solid foods is a common complaint. This may be helped by taking plenty of fluids with each meal. Domperidone which increases the movement of the oesophagus is sometimes prescribed. Patients should avoid large meals late at night and raising the head of the bed is also helpful.

Persistent acid reflux may change the lining of the oesophagus in a few patients to a degree that may predispose it to malignant changes. These patients may have to undergo repeat endoscopy every two years with a small biopsy of the gastro-oesophageal junction to make sure it is not becoming cancerous.

What is a Stricture?

A stricture is a tight circumferential band of scar tissue in the oesophagus, which often results from recurrent acid reflux and may partially obstruct swallowing. This is treated by dilatation or stretching, a procedure usually carried out under sedation as an outpatient, by passing a fiberoptic tube down the oesophagus, (an endoscopy), and using a balloon to open up the stricture. Use of PPIs has reduced the frequency of strictures in scleroderma.

The Mouth

A dry mouth is a common problem due to the reduced production of saliva that can occur in scleroderma. This may lead to early gum disease and tooth decay, therefore regular visits to the dental hygienist are helpful. The condition can be helped by the use of a salivary replacement in the form of spray or lozenges. Sugar free chewing gums may also be helpful.

A smaller mouth opening, which often accompanies this disease is caused by a general tightening of the skin over the face, making lip and mouth movements, as well as oral hygiene, difficult. Reduced mobility of the tongue may affect chewing, swallowing and sometimes speech. Mouth stretching exercises and facial grimacing are probably the best treatment for this problem. Sometimes injections that increase lip bulk can be helpful but collagen injections are generally not recommended.

The Stomach

A common stomach problem is a feeling of fullness occurring after a few mouthfuls of food. This is often associated with abdominal discomfort. Drugs such as domperidone, metoclopramide and erythromycin may be helpful to increase the movements of the bowel.

Bleeding sometimes occurs at the junction of the oesophagus and stomach, or from the stomach itself, from dilated blood vessels. The latter can be treated with a laser but if the bleeding occurs surreptitiously, without being noticed by the patient, the blood count will fall causing anaemia. This will lead to shortness of breath and a feeling of weakness particularly on exertion.

The Small Intestine

Problems of the small intestine may give rise to pain, bloating of the abdomen or diarrhoea due to stagnation of the contents of the gut allowing overgrowth of normal bacterial organisms. Treatment includes Imodium and antibiotics often given in rotation. The small intestine is responsible for absorbing all valuable nutrients from the diet. Even if this organ fails completely, as it does in only a very small number of patients, feeding can still be continued using the intravenous route. Occasionally some patients may have another coexistent disease such as Coeliac disease. This mimics the symptoms of small intestinal disease. Diagnosis of this condition is aided by a simple blood test.

The Large Intestine

Large intestinal problems are common. These include constipation, difficulty with evacuation, faecal urgency and incontinence. The latter is a particularly disturbing symptom and is often not admitted by the patient as they feel socially embarrassed. It is important to investigate these problems as the more common condition of diverticular disease may present with similar symptoms and requires different treatment. Cancer of the colon can present with these symptoms but it is often accompanied by a sudden change in bowel habit. This needs to be excluded by further tests including special X-rays and colonoscopy.

The Rectum and Anus

A few patients develop a rectal prolapse where the lining of the bowel is extruded through the anus. This condition can be treated with an operation but patients need to be screened for heart and lung disease before having an anaesthetic. Surgical treatment of prolapse does not always involve opening the abdomen. Patients who are already incontinent before their operation, may not regain their continence after this procedure, although they are usually cured of their prolapse symptoms. Incontinence is sometimes resolved with simple remedies such as control of diarrhoea, anal plugs and occasionally various surgical procedures such as sacral nerve stimulation. Only rarely, if the patient's symptoms are intolerable, will a colostomy be suggested.

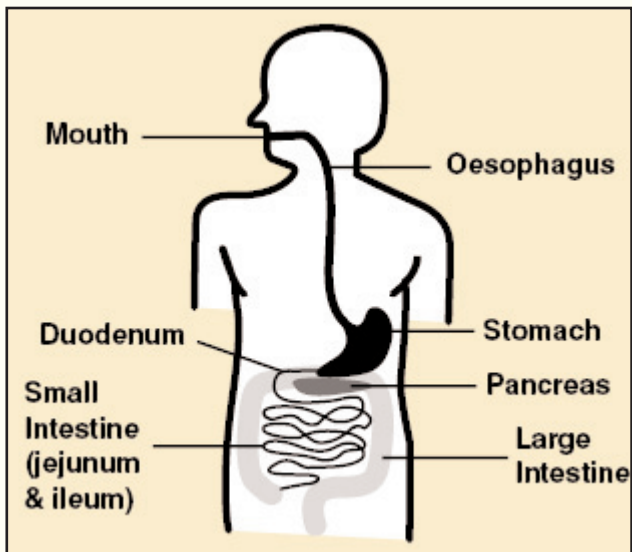


Diagram showing the intestinal tract

Questions

Are gut problems common in scleroderma?

Yes, gut problems in scleroderma are common. Symptoms are often only reluctantly admitted by the patient. They require early investigation and are amenable to treatment.

What is an intestinal obstruction?

Patients should be aware of a situation where they may suddenly develop signs of abdominal pain, vomiting and inability to pass a motion. This is referred to as pseudo-obstruction and should not normally be treated with an operation. The patient should be referred to a centre used to dealing with scleroderma where they will be given the appropriate medical treatment.

What is Barrett's Oesophagus?

Barrett's oesophagus is a complication in all patients with reflux, but especially complicated in scleroderma patients. It occurs in about 10% of all patients with prominent reflux symptoms, but in almost 40% of patients because of inadequate early therapy. Barrett's tissue is what is called a metaplastic tissue and is seen in many patients with scleroderma. The change in cells lining the oesophagus can be pre-cancerous and so follow-up with regular endoscopy is recommended to ensure that any malignant change is detected as early as possible. Sometimes laser treatment is recommended.

Malabsorption

Malabsorption is the inability of the bowel to absorb food. It can occur in scleroderma for several reasons. Firstly, the bowel can become thickened and so absorption is more difficult. Secondly, there can be a deficiency of digestive enzymes. Thirdly, and possibly most often, the slow contraction of the small intestine in scleroderma means that there is a tendency for the bowel contents to stagnate. Bacteria then overgrow and compete for nutrients and cause diarrhoea. This complication is often accompanied by bloating and can be diagnosed with a hydrogen breath test and treated with antibiotics. Resistance of bacteria to these antibiotics can be a problem over long treatment periods.

The Liver

There is a small group of patients who develop an associated condition of the liver called primary biliary cirrhosis. In this condition, the patient presents with tiredness and skin irritation. Diagnosis is by a simple blood test and liver biopsy. This complication may contribute to malabsorption of vital food products and can lead to thinning of the bones. Very rarely will this require liver transplantation.

The Pancreas

Occasionally scleroderma patients produce insufficient juice from the pancreatic gland. This may exacerbate their gut symptoms but can be treated by taking Creon, a pancreatic substitute.



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