

UPDATE

Module 1664

This module covers:

- Causes and symptoms of coeliac disease
- Diagnosis in adults and children
- Management using a gluten-free diet
- Components of a gluten-free diet

JULY >>**Gastrointestinal**

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* Online-only module for Update and Update Plus subscribers

Clinical guide to coeliac disease

Nicola Crawford-Taylor

Coeliac disease is an autoimmune condition characterised by inflammation of the mucosa of the small intestine. It is caused by the ingestion of gluten, the protein fraction in the cereals wheat, barley and rye. Some people with coeliac disease are also sensitive to gluten-like proteins, called avenins, found in oats. The only treatment is a lifelong gluten-free diet.

Coeliac disease affects around one in 100 people in the UK, although only about 10 to 15 per cent of those with the condition are clinically diagnosed. There are approximately half a million people in the UK who have undiagnosed coeliac disease. The condition affects all ethnic groups, and is common not just in Europe, but also southern Asia, the middle east, north-west and east Africa, and South America.

Coeliac disease has a genetic component, and is strongly associated with a number of genetic mutations that affect the human leukocyte antigens (HLA) DQ2 and DQ8. HLA-DQ genes are responsible for the development of the immune system. There is a one in 10 chance that a first degree relative of someone with coeliac disease will also have the condition. There are no agreed recommendations for screening relatives, but genetic screening can exclude the possibility of coeliac disease in relatives of those with the condition.

Coeliac disease is more common among people who have other autoimmune diseases, such as type 1 diabetes and autoimmune thyroid disease. Serological testing for coeliac disease should be offered to children or adults who have any of the symptoms or conditions that are associated with coeliac disease.

The Nice guideline on the recognition and assessment of coeliac disease contains a full list of associated conditions.¹

Presenting symptoms

Symptoms can vary from person to person and range from mild to severe. Common



1 in 100
people are affected by coeliac disease in the UK

10 to 15%
of sufferers clinically diagnosed

2 to 5%
do not respond to a gluten-free diet

symptoms of coeliac disease include:

- chronic or intermittent diarrhoea
- failure to thrive or faltering growth in children
- persistent or unexplained gastrointestinal symptoms, including nausea and vomiting
- prolonged fatigue
- recurrent abdominal pain, cramping or distension
- sudden or unexpected weight loss
- unexplained iron deficiency anaemia, or other unspecified anaemia.

The symptoms of coeliac disease and irritable bowel syndrome (IBS) are similar and include bloating, flatulence, diarrhoea and/or constipation. In both conditions symptoms may be intermittent and associated with periods of stress. Coeliac disease can, therefore, be misdiagnosed as IBS. The Nice guideline on the diagnosis of IBS recommends that any individual suspected of having IBS should also have a blood test for coeliac disease.²

Diagnosis in adults and children

The immune system response in coeliac disease involves the production of immunoglobulin A (IgA) antibodies, directed against an enzyme normally present in the intestines called tissue transglutaminase (tTG). Endomysial antibodies (EMA) are also produced against a type of tTG found in the endomysium, a layer of connective tissue that ensheaths muscle fibres.

Adult patients should be offered serological testing for IgA tTG or EMA. If the result is positive, they should then be referred to a gastroenterologist for an intestinal biopsy to confirm or exclude coeliac disease.

New guidelines on the diagnosis of coeliac disease in children have been published by the British Society of Paediatric Gastroenterology, Hepatology and Nutrition (BSPGHAN) and Coeliac UK. The guidance recommends that in some cases, a biopsy may not be required.

In symptomatic cases where IgA tTG results are greater than 10 times the normal upper limit, diagnosis can be confirmed by either small bowel biopsy or a blood test for IgA EMA and genetic testing. If IgA EMA is positive, and either HLA DQ2 or DQ8 present, the diagnosis may be confirmed without the need for a biopsy.

These new approaches will aid more accurate diagnosis, help children get treated more quickly and should bring savings to the NHS.

Management options

Once an individual is medically diagnosed with coeliac disease, the treatment is a strict, lifelong gluten-free diet, which should be supervised by their healthcare team.

Gluten is found in wheat, barley and rye; obvious sources in the diet include bread, pasta, breakfast cereals, flour, pizza bases, cakes and biscuits. Gluten can also be found in soups, sauces, ready meals and processed foods.

A gluten-free diet consists of:

- naturally gluten-free foods such as meat, fish, fruit and vegetables, rice, potatoes, pulses (peas, beans and lentils) and dairy foods
- processed foods that do not contain gluten, such as ready meals and soups
- gluten-free substitute foods such as gluten-free bread, flour, pasta, crackers and biscuits.

A gluten-free diet is a complete treatment for coeliac disease, helping to improve symptoms and absorption of key nutrients such as iron, calcium and folate, and reducing the risk of health complications. Diagnosis of coeliac disease and adoption of a gluten-free diet has also been shown to improve quality of life to levels comparable with the general population.

The time it takes for someone to feel better on a gluten-free diet can vary. Many people feel better within a few days and symptoms like nausea, diarrhoea and bloating usually clear up within a few weeks. Some symptoms may take longer to improve, or an individual may find that one symptom gets better before another. The time it takes for the gut damage to heal completely can vary between six months and two years.

A small number of people with coeliac disease do not respond to treatment with a gluten-free diet, or they may respond initially, but symptoms return at a later date. This is known as refractory coeliac disease, and it is estimated to affect around two to 5 per cent of those with coeliac disease. The most common symptoms of refractory coeliac disease are constant severe diarrhoea, sudden unexpected weight loss, ongoing anaemia and fatigue.

Complications of untreated coeliac disease include cancer of the small bowel and osteoporosis. Following a strict gluten-free diet will help reduce the already low risk of developing cancer and also help reduce problems from other conditions associated with coeliac disease. However, the increased risk of osteoporosis remains due to less effective absorption. For this reason it is recommended that people with coeliac disease have between 1,000mg and 1,500mg of calcium a day.

The charity Coeliac UK recommends that all patients with coeliac disease are vaccinated against pneumococcal infection. This is because some people with coeliac disease have reduced functioning of the spleen, and are at risk of developing overwhelming pneumococcal sepsis. Since 2006, the pneumococcal vaccine has been included in the childhood immunisation programme, so those born ▶

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Dermatitis herpetiformis

Dermatitis herpetiformis (DH) is a skin manifestation of coeliac disease that affects one in 10,000 people. It commonly occurs as a rash on the elbows, forearms, knees and buttocks, although it can occur anywhere on the body. A skin biopsy is used for diagnosis. Intestinal biopsy nearly always shows flattening of the intestinal villi (seen in coeliac disease) in DH patients, although the gastrointestinal symptoms characteristic of coeliac disease are not necessarily present

before then will require a one-off vaccination.

Patients with coeliac disease should be reviewed on an annual basis by their gastroenterologist, dietitian or GP, as well as being assessed for antibody levels and nutritional deficiencies.

Gluten-free foods

In the UK, people with coeliac disease and dermatitis herpetiformis (left) are eligible for gluten-free food on prescription. A list of foods available on prescription can be found in the Monthly Index of Medical Specialties (MiMs), the BNF and the Drug Tariff. Coeliac UK also has a Food and Drink Directory, which may be more useful, and easier to access, for patients.

National guidelines for professionals have been produced in collaboration with Coeliac UK, the Primary Care Society of Gastroenterology (PCSG), BSPGHAN and the British Dietetic Association (BDA). They set out a reasonable amount of gluten-free staple food recommended for an individual per month. These recommendations are based upon:

- a review of consumption data from national diet and nutrition surveys to establish a benchmark for a reasonable amount of gluten-free food on prescription
- the fact that people with coeliac disease include naturally gluten-free staple foods (potatoes, rice) as well as gluten-free specialist foods in their diet
- a consideration of the eatwell plate³ model for balanced eating.

These guidelines recommend that:

- staple foods such as bread (including fresh bread), pasta, flour, crackers and crispbreads and pizza bases listed by the Advisory Committee on Borderline Substances (ACBS) should remain available
- cake mixes should no longer be available, and sweet biscuits should be considered only in exceptional circumstances on clinical advice.

The role of oats in a gluten-free diet has been debated for many years. Most people with coeliac disease can eat oats, but many oats are harvested and milled in the same place as wheat, barley and rye, which makes them unsuitable due to cross contamination. A very small number of people with coeliac disease are still sensitive to uncontaminated oats because they are sensitive to the gluten-like proteins called avenins, which are found in oats.

It is recommended that oats are avoided in the first six months to a year after diagnosis to allow the gut to heal and for an individual to become symptom free. Gluten-free oats can then be introduced under the guidance and monitoring of the healthcare team.

References

1. NICE Clinical Guideline 86: *Coeliac disease: Recognition and assessment of coeliac disease*, May 2009, nice.org.uk/CG86

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2. NICE Clinical Guideline 61: *Irritable bowel syndrome in adults: Diagnosis and management of irritable bowel syndrome in primary care*, February 2008, nice.org.uk/guidance/CG61

3. The eatwell plate, NHS Choices, nhs.uk/Livewell/Goodfood/Pages/eatwell-plate.aspx

5 minute test

■ Sign up to take the 5 Minute Test and get your answers marked online: chemistanddruggist.co.uk/update

Take the 5 Minute Test

1. The only treatment for coeliac disease is a lifelong gluten-free diet.

True or false?

2. Coeliac disease is uncommon in those of Asian or African origin.

True or false?

3. Coeliac disease is more common among people with other autoimmune diseases.

True or false?

4. NICE recommends that patients with suspected IBS are tested for coeliac disease.

True or false?

5. Patients with refractory coeliac disease show a good response to treatment with a gluten-free diet.

True or false?

6. Complications of untreated coeliac disease include cancer of the small bowel and osteoporosis.

True or false?

7. People with coeliac disease should ensure

they have 1,000 to 1,500mg of calcium per day.

True or false?

8. Pneumococcal vaccination is recommended for patients with coeliac disease.

True or false?

9. Patients with coeliac disease are advised to avoid naturally gluten-free foods due to cross-contamination risks.

True or false?

10. Most people with coeliac disease are sensitive to proteins in oats as well.

True or false?

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Tips for your CPD entry on coeliac disease

Reflect What are the symptoms of coeliac disease? How is coeliac disease diagnosed? Which foods are available on prescription for those suffering from coeliac disease?

Plan This article describes coeliac disease and contains information about its symptoms, diagnosis and management. The availability of gluten-free foods on prescription and the suitability of oats for patients with coeliac disease is also discussed.

Act Read the Update article and the suggested reading (below), then take the 5 Minute Test (above). Update and Update Plus subscribers can then access their answers and a pre-filled CPD logsheet online at chemistanddruggist.co.uk/mycpd.

Find out more about coeliac disease on the NHS Choices website at tinyurl.com/coeliac1

Find out more about the amounts of gluten free foods patients should receive on prescription on the Coeliac UK website, which also has a list of prescribable products

tinyurl.com/coeliac2

tinyurl.com/coeliac3

Read more about gluten free recipes and other dietary advice for patients with coeliac disease on the Coeliac UK website at

tinyurl.com/coeliac4

Find out more about dermatitis herpetiformis on the British Association of Dermatologists website at

tinyurl.com/coeliac5

Evaluate Are you now confident in your knowledge of the symptoms, diagnosis and management of coeliac disease? Could you give advice to patients about a gluten-free diet and the foods available on prescription?



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- Linden has recently dealt with a patient who was suffering from achalasia, a disorder where the muscles in the lower oesophageal sphincter do not relax appropriately on swallowing, leading to a build up of food. He wonders whether this is another case and he decides to refer Mr Roberts to his GP as a matter of urgency.

Mr Roberts returns to the pharmacy some months later and Linden asks him how he is doing. He tells Linden that he has been to the hospital for tests. He had a barium swallow and a pharyngeal pouch has been diagnosed.

What is a pharyngeal pouch?

Linden has not heard of a pharyngeal pouch and decides to investigate using the internet. He discovers that the condition is also called Zenker's diverticulum and occurs when the pharyngeal mucosa herniates just above the upper sphincter of the oesophagus (the cricopharyngeal part of the inferior pharyngeal constrictor muscle).

Herniation results in an out-pouching of the pharyngeal mucosa to form a balloon (or diverticulum).

For many people, the condition causes no problems, but if the hernia increases in size, more severe symptoms can occur.

The main symptoms are dysphagia and regurgitation of undigested food. In the early stage of the condition, dysphagia is usually only for solids, although dysphagia for liquids can develop in time.

Acute symptoms may also present, such as the feeling of a bolus of food in the throat, gagging on food, and coughing while eating. Sometimes nocturnal regurgitation of undigested food can occur. Patients may also complain of a hoarse voice or halitosis.

Complications include significant weight loss and aspiration pneumonia.

The condition is thought to be caused by uncoordinated contraction, and/or spasm and/or impaired relaxation of the cricopharyngeal muscle. The condition has an incidence of about one in 100,000 cases each year in the UK, occurring mainly in people over 50 years old, with the peak age for treatment being in patients aged 70 years.

Mr Roberts has been told that he needs to have an operation. He asks Linden again whether indigestion remedies will help him in the meantime.

Is there any medication that Lynden could safely sell Mr Roberts?

Lynden reads that there is no pharmacological treatment for this condition. The retention of orally consumed medications in the diverticulum can make them ineffective. Usually patients are managed surgically. Previously, surgery involved an incision in the neck following general anaesthetic – clearly risky for the older age group of patients affected by this condition. However, increasingly endoscopy is used if the patient meets certain criteria (see www.nice.org.uk/IPG22).