



GUIDELINES FOR THE DIAGNOSIS AND MANAGEMENT OF COELIAC DISEASE IN ADULTS

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These guidelines have been published by the Clinical Resource Efficiency Support Team (CREST), which is a small team of health care professionals established under the auspices of the Central Medical Advisory Committee in 1988. The aims of CREST are to promote clinical efficiency in the Health Service in Northern Ireland, while ensuring the highest possible standard of clinical practice is maintained.

The guidelines have been produced by a sub-group of health care professionals chaired by Dr William Dickey. CREST wishes to thank them and all those who contributed in any way to the development of these guidelines.

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1. INTRODUCTION

The purpose of this document is to give detailed practical guidance on the diagnosis and management of coeliac disease. It is intended to be of relevance to general practitioners, secondary care clinicians of all specialties, dietitians and nurses.

Coeliac disease affects around 1% of the Northern Ireland population. Although there have been major advances in our understanding of coeliac disease over the last ten years these have not been communicated effectively to healthcare professionals. Patient surveys indicate that coeliac disease is frequently unrecognised, resulting in delayed diagnosis or misdiagnosis, unnecessary or repeated investigations and consultations, and persisting ill-health with risk of complications in the longer term. Even after diagnosis, there is wide variation in management and follow-up. Increased awareness of the condition, in conjunction with improved diagnostic methods, will have a significant impact on this common condition.

2. DEFINITION

The key features of coeliac disease are:

- 1) It is an autoimmune condition
- 2) triggered in genetically predisposed individuals by
- 3) the consumption of gluten, a protein in wheat, and similar proteins in barley and rye (though individually distinct, all are commonly referred to as “gluten”).

These cereal proteins form complexes with the autoantigen of coeliac disease, tissue transglutaminase.

Coeliac disease characteristically causes inflammatory changes in the proximal small intestine (duodenum, jejunum) and biopsy of this area remains mandatory in diagnosis. However, coeliac disease can affect any organ system.

The inheritance of coeliac disease is closely linked to HLA antigens (DQ2 in over 90%, DQ8 in most of the remainder) but other unspecified genetic factors are involved.

The majority of patients have serum autoantibodies (to endomysium and tissue transglutaminase) which form the basis for useful diagnostic tests.

Coeliac disease is unique among autoimmune conditions as removal of an extrinsic factor (gluten) typically results in clinical improvement.

3. EPIDEMIOLOGY

Screening studies indicate that the prevalence of coeliac disease in Western Europe, and in populations with European ancestry including the Americas and Australasia, is of the order of 1:100 [1]. In first-degree relatives of coeliac patients the prevalence is

around 1:10, and the background prevalence is higher where other autoimmune diseases are present (Table 1). It is clear that diagnosis is frequently delayed until long after the onset of symptoms [2] and many patients remain undiagnosed: this reflects the many non-specific ways in which coeliac disease can present, and also the erroneous beliefs that it is rare and a condition primarily of children who “grow out of it” in their teens. Despite gluten ingestion from infancy, patients may present for the first time at any age. The majority of patients are over 40 at diagnosis, with first presentation in the elderly well recognised [3]. Both sexes are affected but most series report an excess of females, who are more likely to seek medical advice for symptoms and are at greater risk of anaemia. Occasionally a “trigger” event such as gastroenteritis, pregnancy or childbirth can be identified before the onset of symptoms.

Table 1. CONDITIONS ASSOCIATED WITH AN INCREASED PREVALENCE OF COELIAC DISEASE

Autoimmune Type 1 diabetes mellitus Autoimmune thyroid disease Primary biliary cirrhosis Autoimmune hepatitis Sjogren syndrome Addison’s disease
Others Selective IgA deficiency Down syndrome

4. PRESENTATION

The “classical” presentation of coeliac disease reflects its effect on the proximal small bowel with consequences of malabsorption: steatorrhoea, abdominal cramps, bloating and weight loss. In practice, a minority of patients report diarrhoea as a symptom [4]. The stereotype of the underweight coeliac patient no longer applies: a high proportion of patients are overweight [5]. There is no doubt that many patients with symptoms attributed to irritable bowel syndrome actually have coeliac disease [6].

Coeliac disease has been shown to cause motility problems in the gastrointestinal tract. This means that upper gastrointestinal symptoms like nausea and reflux may be prominent, and also, paradoxically, constipation.

Gastrointestinal symptoms may be minor or absent with the primary presentation originating elsewhere. Some of these presentations may be attributed directly to malabsorption (e.g. anaemia). Others, such as dermatitis herpetiformis, reflect the systemic autoimmune nature of the condition.

Table 2. PRESENTATIONS OF COELIAC DISEASE

Gastrointestinal symptoms due to malabsorption	Diarrhoea, steatorrhoea Abdominal cramps, borborygmi Abdominal bloating, distension Excessive flatulence/flatus Weight loss
Gastrointestinal symptoms due to dysmotility	Heartburn, regurgitation Dysphagia, vomiting Epigastric pain Constipation
Haematological	Any combination of iron, B12 or folic acid deficiency Hyposplenism
Liver	Abnormal liver biochemistry: usually raised aspartate and alanine transaminase (AST, ALT)
Skin and mucus membranes	Dermatitis herpetiformis Alopecia Aphthous mouth ulcers
Rheumatological	Arthralgia, arthritis
Bone	Osteoporosis Osteomalacia, rickets (rare) Defective tooth enamel
Gynaecological [7]	Late menarche, early menopause Infertility Recurrent miscarriage
Neurological [8]	Ataxia Partial seizures Migraine Peripheral neuropathy
Other	Short stature Chronic fatigue Depression “muzzy head”

5. DIAGNOSIS

5.1 Introduction

It is crucial that confirmation of the diagnosis is made **before** starting a gluten-free diet and duodenal biopsy remains mandatory.

However, the non-specific symptoms of many coeliac patients mean that endoscopy with biopsy as the initial investigation for all is impracticable and serological testing should be the first investigation with duodenal biopsy for those who are seropositive. Serological testing by general practitioners has resulted in a considerable increase in diagnosis rates [4].

As false negative serology occurs in 5-10% of cases with current antibody tests [4], biopsy should be performed in patients with a high suspicion of coeliac disease (family history, “classic” symptoms) and in patients undergoing upper gastrointestinal endoscopy (OGD) as part of the investigation of anaemia even if antibody testing is negative.

Small bowel barium x-rays have poor sensitivity for coeliac disease and there is no place for tests of malabsorption (e.g. d-xylose, Schilling test) which are non-specific.

5.2 Serological Testing

In Northern Ireland, all serological assays for coeliac disease are performed at the Regional Immunology Laboratory, Kelvin Laboratories, Royal Hospitals, Belfast.

Testing for antiendomysial antibodies (EmA) and for antitransglutaminase antibodies using human recombinant antigen (TGA) are both highly sensitive and specific for coeliac disease.

At present IgA class TGA is used for initial testing. If TGA is positive, serum is then tested automatically for IgA EmA: positive testing for either antibody should prompt referral for biopsy.

5.2.1. IgA deficiency in coeliac disease

Coeliac disease occurs more frequently in patients with IgA deficiency than in the general population [9]. In such patients IgA TGA level will be 0. All such patients have their total immunoglobulin A levels measured automatically. Those found to be IgA deficient will be further tested for IgG class TGA and EmA.

TGA and EmA can be used not only for diagnosis but also for monitoring of dietary compliance: seroconversion occurs rapidly with strict gluten exclusion.

Conversely, if serology is to be meaningful in the diagnosis of coeliac disease patients must be taking a gluten containing diet for at least 3 months.

It is essential to provide a relevant clinical history on the request form for proper interpretation of results (i.e. symptoms and whether the request is for diagnosis or for monitoring dietary compliance).

Negative antibody tests do not preclude positive serology and development of coeliac disease in the future.

5.2.2. False positive anti transglutaminase antibodies

Positive TGA have been detected in patients with chronic liver disease, renal disease, monoclonal gammopathy and autoimmune disease (e.g. rheumatoid arthritis and systemic lupus erythematosus).

5.2.3. Positive endomysial antibodies without villous atrophy

A minority of patients with positive EmA have normal duodenal biopsies. Most will develop villous atrophy on follow-up biopsy if left untreated or will clinically respond to gluten exclusion [10]. Some patients with positive EmA but negative biopsies may therefore warrant a gluten-free diet.

5.2.4. False negative serology

Around 5-10% of coeliac patients are negative for both antibody tests despite normal IgA levels. Biopsy should be performed in patients whose clinical presentation is strongly suggestive, even if seronegative.

Diagnostic Algorithm for Serological Testing in Coeliac Disease

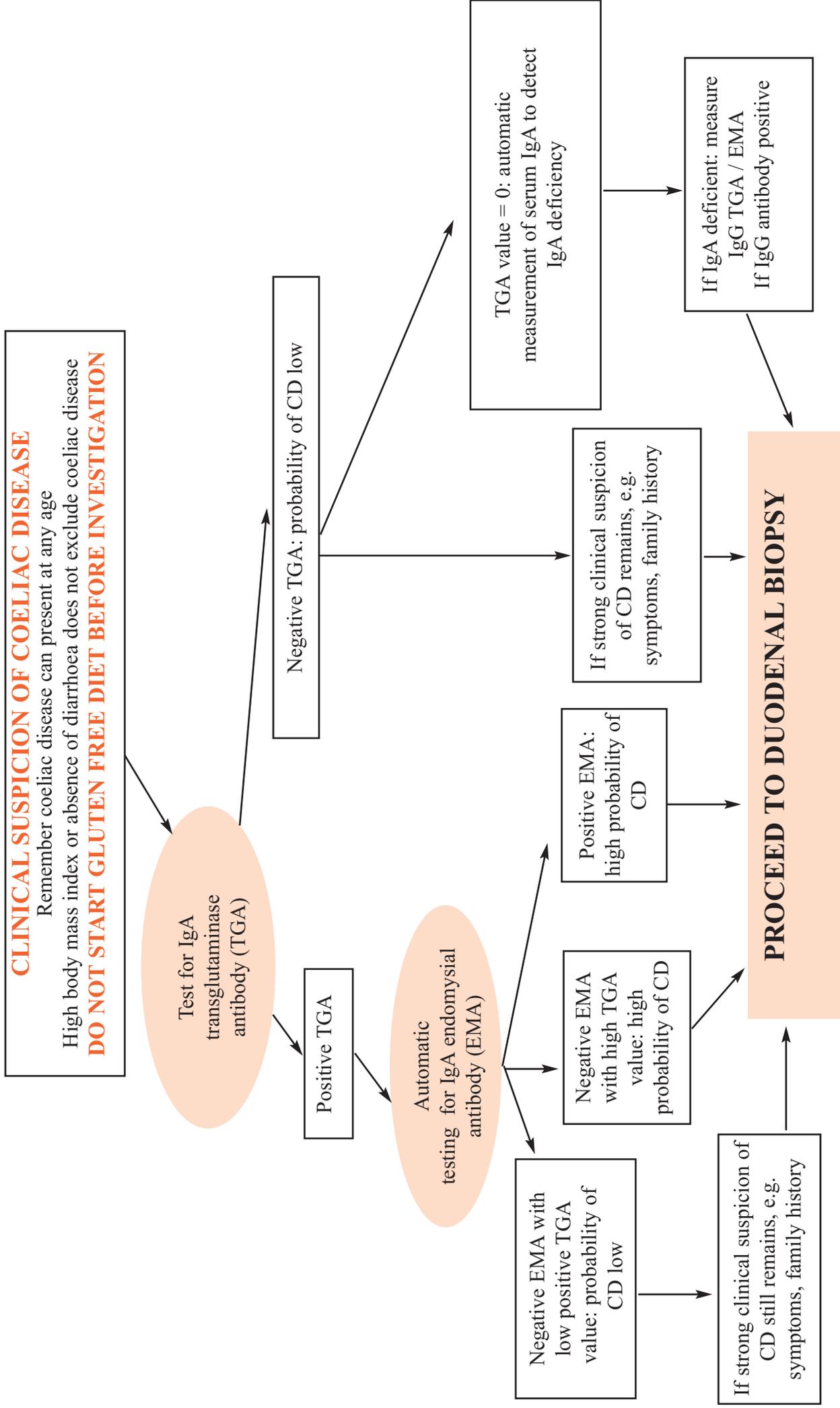


Figure 1.

5.3. Endoscopic Recognition of Coeliac Disease

Many patients with upper GI symptoms due to coeliac disease (Table 2) will be referred for OGD as their initial investigation. There is no evidence that routine duodenal biopsy in all patients undergoing OGD for dyspeptic symptoms is effective [11] and this practice would have major resource implications, although the yield may be higher in patients with no evidence of ulcer or reflux disease or who have not responded to acid suppression. Biopsies may also be worthwhile in groups with an increased prevalence of coeliac disease (type 1 diabetes mellitus, thyroid disease).

The duodenum should be studied closely in all patients undergoing OGD as a high proportion of patients with high-grade villous atrophy have obvious stigmata which should prompt duodenal biopsy [12]. These include:

- “Mosaic”, nodular or fissured mucosa
- Scalloped duodenal folds
- Duodenal fold loss
- Visible submucosal blood vessels
- Multiple erosions in the second part of duodenum.

These markers are best seen in the second part of duodenum and may be accentuated by instillation of water. They are not present in all patients with villous atrophy or with milder forms of gluten sensitive enteropathy and their absence should not prevent biopsy if clinically indicated.

5.4. Histological Abnormalities Associated with Coeliac Disease

Duodenal biopsies are taken by forceps during OGD. In routine clinical practice, suction biopsy devices (Watson, Crosby capsules) which pass to the small bowel by peristalsis are obsolete.

Traditionally, coeliac disease was only diagnosed when there was severe (total, subtotal) villous atrophy on small bowel biopsy. We now use the Marsh classification [13] to define gluten enteropathy:

- 0: Normal
- I: Increased intraepithelial lymphocyte counts (> 30 per 100 enterocytes)
- II: Intraepithelial lymphocytes plus increase in crypt depth (crypt hyperplasia)
- III: Above plus villous atrophy: the “classic” coeliac lesion.
- IV: Villous atrophy without lymphocytes: rare; typically unresponsive to diet.

5.4.1. Assessment of biopsies

Accurate assessment of the patient with suspected coeliac disease depends on several factors. It is important that adequate numbers of biopsies (at least 3) are taken, distal to the duodenal bulb. Biopsies should be orientated. This can be done on card or filter paper at the time of sampling, but submission of biopsies free floating in formalin and subsequent orientation in the laboratory by biomedical staff is equally acceptable. This ensures proper representation of mucosa, appropriate orientation, with

minimisation of artefact. This is most important in early lesions, patchy disease or in cases where there is only an excess of intraepithelial lymphocytes (Marsh I).

Changes in biopsies from patients with coeliac disease are characteristically most severe proximally and decrease in severity distally. In the untreated case, the duodenum is always involved, the proximal jejunum usually and the ileum only in severe cases. The wide disparity in severity of gut symptoms among patients may reflect this variation in the length of small bowel involved.

5.4.2. Histology of untreated disease

The examining pathologist should have an appropriate knowledge and understanding of normal small bowel mucosa, which has a villous height-crypt depth ratio of 3-5:1 depending on the anatomical location of the biopsy. Intraepithelial lymphocyte density is normally less than 30 per 100 enterocytes [13]. At least three biopsies, examined through at least three levels, should be assessed. The biopsy report should refer to each of the three mucosal components- villi, crypts and intraepithelial lymphocytes- involved in gluten enteropathy in a structured fashion, using the Marsh classification as appropriate.

While in western European adults the presence of villous atrophy is highly specific for coeliac disease the finding of increased intraepithelial lymphocytes only is less so and should be correlated with clinical presentation, family history of coeliac disease and serological markers.

The presence of positive TGA and/or EmA plus biopsy abnormalities is sufficient to make a diagnosis of coeliac disease. There is no requirement for gluten challenge after a period of treatment if these investigations are done before dietary restriction.

Other causes of villous atrophy are well-described [14] but are rare in Northern Ireland and beyond the remit of this document. However they should be considered in the patient with negative serology pre-treatment who does not respond clinically to gluten exclusion.

5.4.3. Histology of treated disease

Despite good dietary compliance and clinical response, the duodenal mucosa may take months or even years to regain normal architecture and some patients show persistent inflammatory and architectural changes. There is no general agreement on whether routine follow-up biopsies should be performed. If planned, these should be deferred for at least two years after treatment is started as many patients do not show villous recovery at shorter intervals [15].

5.5 Additional Investigations in Coeliac Disease

All patients at diagnosis should have:

- Full blood count
- Ferritin
- Vitamin B12 and folate
- Liver and bone biochemistry

There is an association between coeliac disease and hyposplenism. While this appears to be much less common than previously supposed and pneumococcal vaccination in the UK is routinely given to all patients over 65 or with risk factors, it would seem reasonable to check for blood markers of hyposplenism (e.g. Howell Jolly bodies on blood film) as a one-off at initial diagnosis.

British Society of Gastroenterology guidelines recommend routine DEXA scanning in all coeliac patients [16]. Further studies are needed to determine whether subsets of coeliac patients at low risk who do not require scanning can be identified, particularly as the risk of low impact fracture associated with coeliac disease appears to be relatively low [17].

During discussion of the diagnosis, patients should be made aware of the increased risk of the condition in immediate relatives, who may wish to put themselves forward for testing.

5.6 Genetic testing

Some 90% of coeliac patients have the DQ2 heterodimer and most of the remainder are DQ8 positive. DQ2 positivity is of limited value in diagnosis as it is present in some 30% of the Northern Ireland population, but it may have a role in determining which relatives of coeliac patients are at risk of developing coeliac disease in the future. Currently, HLA-DQ2/DQ8 testing is not available in Northern Ireland.

6. MALIGNANCY IN COELIAC DISEASE

A number of malignant conditions including small bowel lymphoma, small bowel adenocarcinoma, and oesophageal and pharyngeal squamous carcinoma have been described as occurring more commonly in coeliac disease. Early studies showing a high risk of malignancy date from when coeliac disease was considered only in very ill patients and are not applicable to the coeliac population in general. Recent large, population based studies indicate that the increased risk of malignancy associated with coeliac disease is much less than previously thought with figures for non-Hodgkin lymphoma of 2.6 – 6.3. While there is an excess mortality in coeliac disease (hazard ratio 2.09), which is due to malignancy, this is confined to the first year after diagnosis and then falls to 1.17 [18]. A Northern Ireland study of small bowel lymphoma [19] suggests that this translates into an absolute risk of only 1:1000 based on the local prevalence of coeliac disease. The risk (standardized incidence ratio) associated with coeliac disease for cancer overall in one large series of 12 000 patients was 1.3, lymphoma 5.9, small bowel adenocarcinoma 10, oesophageal cancer 4.2, oropharyngeal cancer 2.3 and colorectal cancer 1.5 [20]. In conclusion, current studies indicate that the absolute risk of malignancy in clinically diagnosed coeliac disease is much less than previously supposed. There is evidence that adherence to a gluten-free diet reduces this risk within five years to that of the general population.

While there is no evidence that routine screening for lymphoma by small bowel barium studies and CT scans in all coeliac patients is of benefit, there should be a strong suspicion in patients with significant and persisting weight loss, symptoms which fail to improve, or who initially respond then deteriorate despite compliance with the gluten-free diet. As conventional imaging using small bowel barium studies

and CT scanning has limited sensitivity for small bowel neoplasia, diagnosis is often delayed even if suspected. A few centres provide enteroscopy which may permit earlier, non-operative diagnosis, although diagnosis may still need to rely on laparoscopy or laparotomy. Capsule endoscopy, currently available at two centres in Northern Ireland, represents a new imaging method which may permit earlier diagnosis.

7. CONSIDERATION OF OTHER PATHOLOGY

The diagnosis of coeliac disease does not exclude other, coincidental pathology, particularly in older patients. In patients over 40 with iron deficiency or diarrhoea, around 10% will have colon polyps or colorectal cancer whether or not coeliac disease is present [21] and these patients should be considered for colonic investigation.

Failure to respond to a gluten-free diet should prompt consideration of other pathology, although deliberate or inadvertent exposure to gluten is the most likely explanation: persistently positive coeliac serology supports this. Patients with coeliac disease have a higher prevalence of:

- lactose intolerance: lactase is contained in the tips of normal villi and a lactose-free as well as gluten-free diet may be needed in the early stages of treatment. This is uncommon and usually temporary;

- pancreatic insufficiency;

- microscopic (lymphocytic or collagenous) colitis.

Recent studies [22] indicate that there is also a higher prevalence of ulcerative colitis and Crohn's disease among coeliacs.

Failure of symptoms and histology to respond to a gluten-free diet in the absence of other pathology (having excluded deliberate or inadvertent non-compliance) is defined as refractory sprue and this may require steroid or other immunosuppressive therapy.

8. MANAGEMENT OF COELIAC DISEASE

8.1 Gluten-Free Diet

The gluten-free diet aims to exclude the relevant proteins in wheat (gliadin), rye (secalin), and barley (hordein) and in ingredients derived from these cereals which trigger the abnormal immunological response of coeliac disease.

It is imperative that a gluten-free diet is **NOT** commenced prior to serological testing or a biopsy being taken. Problems arise particularly when patients are advised to exclude gluten by alternative practitioners without specific and appropriate investigations for coeliac disease beforehand, when diagnosis of coeliac disease is then rendered difficult.

Strict life-long adherence to a gluten-free diet is essential, to:

- Improve symptoms and general well being
- Achieve a nutritionally balanced diet
- Promote growth and development in children
- Normalise gut mucosa, and
- Reduce the risk of long-term complications

Gluten exclusion is difficult as it is found in bread, biscuits, pastries, pasta, beer, many breakfast cereals and processed foods e.g. sausages, soups and sauces. In addition wheat flour is commonly used as a processing aid, binder, filler and as a carrier for flavourings and spices. Contamination with gluten can also occur during storage, processing or manufacturing.

Most non-cereal foods that have not undergone processing, such as fresh meat, fish, eggs, milk, fruit and vegetables, can be safely included in a gluten-free diet.

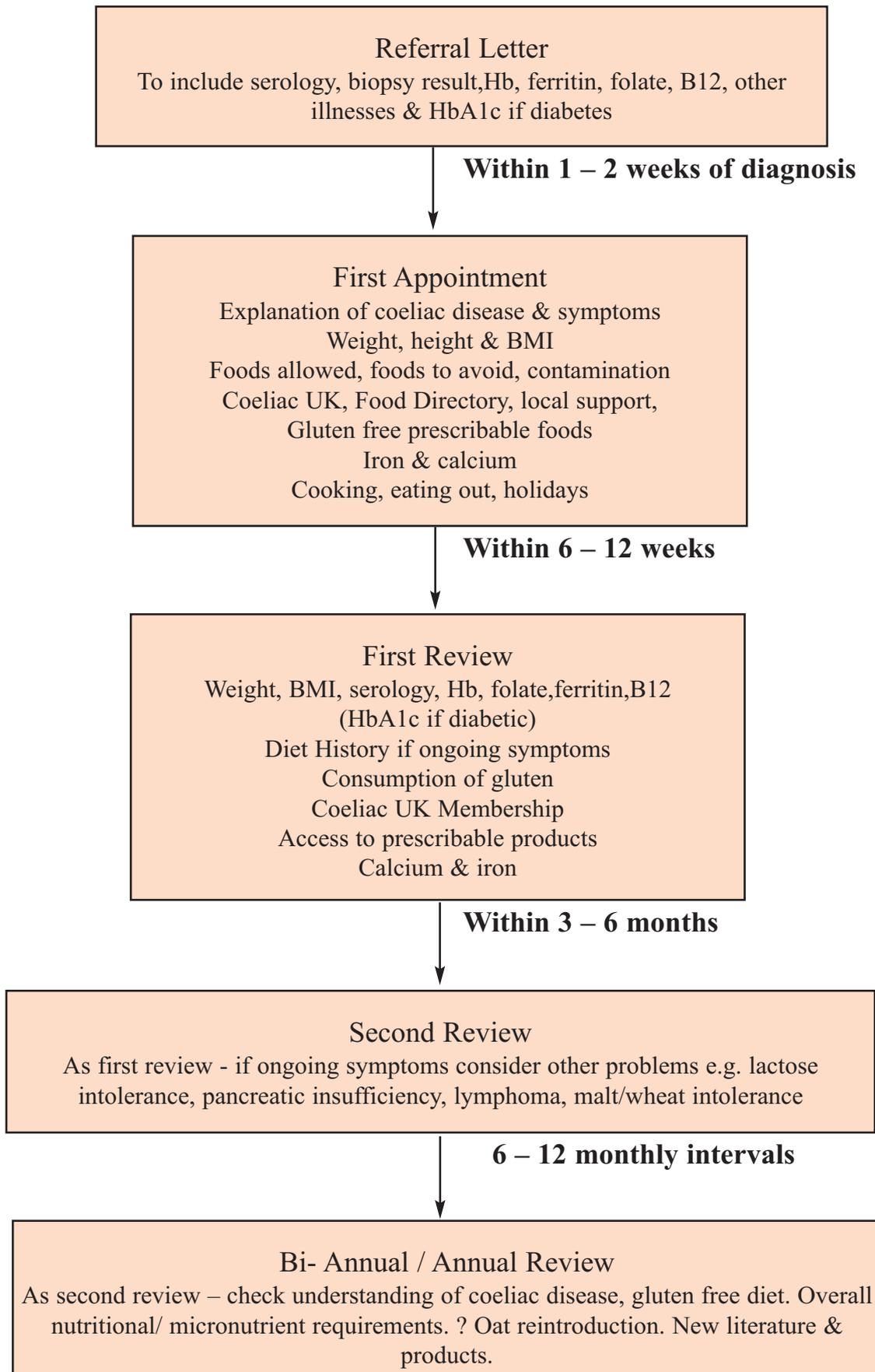
A gluten-free diet is a major undertaking and requires expert dietetic advice.

8.1.1 Dietetic Management

Initial dietetic advice and regular reviews are essential to improve understanding and adherence of the diet and to ensure a balanced diet is achieved to promote long-term health. Annual dietetic review is important to ensure that even experienced coeliac patients do not inadvertently consume gluten. An annual review is also essential to promote an overall balanced diet and discuss any nutritional issues or possible deficiencies which may arise [23].

Figure 2.

Dietary Management of Coeliac Disease



The dietary objectives are to ensure that people with coeliac disease:

- Exclude all dietary sources of gluten
- Know which foods and ingredients are naturally free from gluten
- Substitute gluten-containing foods and ingredients with gluten-free alternatives to improve dietary acceptability and nutritional adequacy
- Have nutritional deficiencies identified and treated, e.g. iron/folate/ vitamin B12 deficiency, osteoporosis
- Have appropriate management of associated conditions which require dietary intervention, e.g. type 1 diabetes mellitus, lactose intolerance and pancreatic insufficiency
- Consume a balanced diet which helps to maintain health and protect against disease
- Have ongoing life-long dietetic support

8.1.2 Coeliac UK

Coeliac UK (www.coeliac.org.uk) is the leading charity providing support for coeliac patients. Professional membership is also available. Membership, which is free, provides essential information on suitable products and other valuable information on coping with a gluten-free diet, e.g. eating out and holidays. A telephone information line is provided. A Food and Drink Directory is produced annually which lists gluten-free foods. Coeliac UK provides monthly updates via their website, e-mail and BBC Ceefax which are essential as manufacturing changes can alter gluten-free status. All the food and drinks listed are guaranteed by manufacturers to contain less than 200ppm (parts per million) gluten, which is defined by the international Codex Standard and considered a safe level for the majority of people with coeliac disease. Sweeteners derived from gluten-containing cereals (e.g. glucose syrups, maltodextrin and dextrose) have very low gluten levels, are considered safe for most coeliacs, and are exempt from labelling. However, some coeliacs may be unable to tolerate even small amounts of gluten in food and require foods which are labelled wheat and gluten free.

8.1.3 Food labelling

The Crossed Grain symbol sometimes found on packaging guarantees the product to be gluten-free. The symbol is owned by Coeliac UK and used under license by food producers.

However, new food labelling regulations EC Directive (2003/89/EC) mean all food manufacturers have to declare which allergens are present in the food. All products containing gluten or wheat are highlighted as 'contains gluten' or 'contains wheat'. Exceptions include unpackaged foods. It must always be remembered food labelled 'wheat free' is not necessarily gluten-free as rye and barley may be present.

8.1.4 Oats

Until recently, the oat protein avenin was thought to have the same effects as gluten derived from wheat, barley and rye. Oats were therefore traditionally excluded from a gluten-free diet. There is now evidence that many people with coeliac disease can tolerate pure uncontaminated oats. However, commercially available oats may be

contaminated with other grains, and a minority of coeliacs (5%) are sensitive to pure oats. Coeliac UK currently advise that a moderate amount (up to 50g, i.e. one reasonable serving) of pure (uncontaminated) oats per day can be consumed by most coeliacs without risk. Before oats are introduced patients should have a dietetic review and have normal serology. After oats are introduced patients must have regular review for evidence of clinical, biochemical and serological deterioration. Serology should be rechecked after 3 months.

Uncontaminated oat products are listed as an appendix in the Coeliac UK Food and Drink Directory.

8.1.5 Malt

Barley derived malt is used as a flavouring in some maize and rice-based products such as breakfast cereals. In some brands, particularly of corn flakes, the gluten content has recently been shown to exceed the accepted threshold of 200 ppm defined by the international Codex standard. These brands have been removed from the Coeliac UK Food and Drink Directory.

8.1.6. Weaning

Gluten should not be introduced into any baby's diet before six months (in general, solids are not recommended until six months). Delaying the introduction of gluten after this time appears to be of no benefit.

Breastfeeding is recommended and there is evidence that this can at least delay the onset of coeliac disease. Formula milks are gluten free.

8.2 Dietary Treatment and Prevention of Osteoporosis

- Coeliacs should consume a minimum of 1500mg/day calcium, most of which should be of high bioavailability such as that from milk and dairy products. If adequate daily calcium intake cannot be achieved, supplemental calcium (500-1000mg/day) should be considered
- An adequate intake of vitamin D: supplements may be advisable in elderly people who have little exposure to sunlight and in some ethnic groups
- Avoidance of smoking and excessive alcohol intake
- Regular weight bearing exercise

8.3 Prescriptions

As people medically diagnosed with coeliac disease cannot eat foods made from wheat flour special gluten free bread, flour mixes, pasta, pizza bases, biscuits and crackers are available on Advisory Committee on Borderline Substances (ACBS) prescriptions. These products are important in providing a gluten-free balanced diet and improving adherence to the dietary restrictions.

A complete list of prescribable foods can be found in Coeliac UK's Food and Drink Directory, the British National Formulary (BNF) and the Monthly Index of Medical Specialities (MIMS). Luxury items such as cakes, fancy biscuits and seasonal foods like mince pies can be bought at some supermarkets, pharmacies, health food shops or by mail order.

8.3.1 Accessing Gluten Free Food in Primary Care

Gluten free food is prescribed, generally at the request of hospital physicians or dietitians, by general practitioners. It is ordered and supplied using a HPSS prescription through a community pharmacy. The Drug Tariff governs what is allowable on HPSS prescriptions in primary care in Northern Ireland. The current Drug Tariff contains a wide variety of gluten free foods approved under the Advisory Committee on Borderline Substances, which are prescribable for gluten intolerance.

If patients are not entitled to free prescriptions a pre-payment certificate, lasting either 4 months or 12 months is recommended.

“Gluten-free foods: a prescribing guide” lists guidelines recently produced and endorsed by the British Society of Paediatric Gastroenterology, Hepatology and Nutrition, the British Dietetic Association, the Primary Care Society for Gastroenterology, and Coeliac UK. They have been produced to assist general practitioners, dietitians, pharmacists and practice nurses in the decision making process when prescribing gluten-free foods. The guide includes background information about coeliac disease, as well as stating the minimum recommendations and advice when prescribing the quantities and varieties of gluten-free foods available on prescription. Quantities are calculated on a “unit” basis and are age- and gender- dependent. The guide is not intended to replace a full dietetic assessment but to provide an example of the minimum requirements for those needing a gluten free diet. Copies are available from Good Relations Healthcare, Holborn Gate, 26 Southampton Buildings, London WC2A 1PQ (Tel 020 7861 3030) and may be viewed in summary form on the Primary Care Society for Gastroenterology website (<http://www.pcs.org.uk>).

8.3.2 Issues with the Current Supply Route

Some of the permitted gluten-free foods have short expiry dates and this can lead to problems if adjustments in treatment are made. It can also mean that the initial prescription may take some time before actually being filled.

GP computer systems have an array of food available and unless the actual specification of the product is known, difficulties can arise with the incorrect product being chosen. This can lead to confusion and delays in issuing repeat prescriptions.

9. CURRENT SERVICE PROVISION

Diagnostic rates for coeliac disease have increased year on year across Northern Ireland. It is estimated that around 500 people are newly diagnosed with coeliac disease each year (Regional Immunology Laboratory data). Increased awareness of the condition is likely to increase diagnosis rates. Based on a background prevalence of 1%, there are an estimated 17,000 people with coeliac disease in Northern Ireland (NI Population: 1.7 million in 2004).

Increased awareness of the condition will impact on the first instance on laboratory services through increased numbers of requests for coeliac serology both at initial diagnosis and during follow-up to monitor dietary compliance. Recently, point-of-care assays for TGA have been shown to have high sensitivity and specificity for coeliac disease [24] and may have a future role

Before commencing treatment, patients with coeliac disease require endoscopy with biopsy for confirmation of the diagnosis. This may be difficult in some areas where there is a long waiting list for OGD. Recognising that patients with upper gastrointestinal symptoms suggestive of sinister pathology require fast-tracking, consideration needs to be given to a similar priority system for patients with suspected coeliac disease, particularly if positive serology has been obtained in primary care. Such patients could in most cases be brought directly for endoscopy/biopsy, without the need for prior outpatient consultation, if the general practitioner has already explained the rationale.

While serological testing allows selection of patients for duodenal biopsy, increased referrals following positive serology (TGA +/- EmA) are likely to have an impact on histopathology workload.

Once diagnosed, patients with coeliac disease need to be given high priority, with their first dietetic consultation within two weeks. The British Society of Gastroenterology (http://www.bsg.org.uk/clinical_prac/guidelines/coeliac.htm) recommend all patients should be then reviewed at 2-3 months and again at 6 months, followed by an annual review for all coeliac patients to assess progress, nutritional status and dietary compliance. At present the Dietetic Service is unable to offer all patients new and review appointments within the timescale recommended. Care is provided by a number of senior dietitians but there are no specialist dietitians for coeliac disease in Northern Ireland. In contrast, in Britain, a number of specialist dietitians run dietetic led coeliac clinics.

In addition to clinical input, there are an increasing number of requests for input into local support groups, supermarket tours and cookery classes.

9.1 Recommendations for Staffing

New patients require 45 minutes to one hour for their first dietetic appointment, and 30 minute follow-up appointments thereafter. If the British Society of Gastroenterology guidelines were followed and all patients were receiving appropriate follow up, at least 1 whole time equivalent specialist dietitian would be required in each of the new 5 recommended Health & Social Care Trusts. Additional specialist dietitians are required for paediatric services. If dietetic led clinics evolve advanced practitioner dietitians will be required to lead the service.

9.2 Recommendations for Prescribing

Consideration should be given to a “unit” based method of prescribing (see. 8.3.1) which will reduce general practice workload and improve access and choice for patients.

10. IMPLEMENTATION AND AUDIT

10.1 Implementation of the Guidelines

Implementation of the guidelines is the responsibility of each Trust and is an essential part of clinical governance.

Mechanisms should be put in place to ensure that the care provided is reviewed against the guideline recommendations and the reasons for any differences assessed and, where appropriate, addressed.

10.2 Resource Implications of Implementing the Guidelines

There will be resource implications with regard to serological and histological testing and DEXA scanning as well as workload for gastroenterologists and dietitians as diagnosis rates increase. Part of the increased workload will arise from training needs and increasing awareness as well as direct patient care.

10.3 Audit

Trusts and primary care organisations should carry out clinical audit. Service providers should create a database of coeliac patients to facilitate audit and recall. Audit parameters may include those suggested below relating to detection, diagnosis, primary and secondary care and dietetics

10.3.1. Detection

There should be increased awareness of the various clinical presentations of coeliac disease which differ from the “classical” picture and of at risk groups such as patients with family history or with insulin dependent diabetes. Audit should consider the changing clinical presentation of diagnosed cases and of those undergoing serological testing.

10.3.2 Diagnosis

Audit standards may include the number of patients who have recorded evidence of:

- Serological testing as first line investigation
- Duodenal biopsy if serological testing is positive
- Full blood count
- Ferritin
- Vitamin B12 and folate
- Time to biopsy confirmation

Audit of serological testing by cipher number may identify variation in testing between practices.

10.3.3 Management of Coeliac Disease

Medical

Audit standards may include number of patients who had documented evidence of:

- Blood markers checked for hyposplenism
- Osteoporosis assessment
- Relevant investigations: regular full blood count, ferritin, B12, folate checks
- Patients with hyposplenism offered relevant vaccination
- Annual medical review

Dietetics

Audit standards may include number of patients who had documented evidence of:

- First appointment within two weeks
- Offer of a review 2-3 months after diagnosis
- Offer of a review at 6 months
- An annual dietetic review
- Monitoring of body mass index, diet, compliance and membership of Coeliac UK

Training

Audit standards should assess quality of training for dietitians and physicians in primary and secondary care.

11. INTERNET RESOURCES AND REFERENCES

11.1 Guidelines on Coeliac Disease for Medical Practitioners

British Society of Gastroenterology Interim Guidelines for the Management of patients with Coeliac Disease, 2004 (currently undergoing revision)

http://www.bsg.org.uk/clinical_prac/guidelines/ceeliac.htm

The Primary Care Society for Gastroenterology has guidelines aimed at general practitioners on its website <http://www.pcs.org.uk>.

WGO-OMGE (World Gastroenterology Organisation) Practice Guideline: Celiac Disease, February 2005 http://omge.org/globalguidelines/guide13/g_data13_en.htm

National Institutes of Health Consensus Development Conference Statement, June 28–30, 2004

<http://consensus.nih.gov/2004/2004CeliacDisease118html.htm>

11.2 Resources for Patients

Coeliac UK <http://www.coeliac.org.uk>

Coeliac Society of Ireland <http://www.coeliac.ie>

Celiac Disease and Gluten-free Diet Support Center at Celiac.com. An American patient site

<http://www.celiac.com>

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