Ashford and St. Peter's Hospitals
NHS Foundation Trust

Coeliac Disease- diagnosis and management

Author: Dr Emily Taylor ST6 Paediatric Trainee

Supervisor: Dr Alka Thakur

Contact details: alka.thakur@nhs.net

Guideline History		
Date	**Comments**	**Approved By**
June 2020	Ratified	Paediatric Guidelines Group
February 2024	BSPGHAN and NICE guidelines reviewed – no changes made	Paediatric Guidelines Group

Contents

1. Guideline
    a. Introduction
    b. Diagnosis & diagnostic pathways
    c. Treatment
    d. Clinic management and follow up
2. Dietetic Standard Operating Procedure for newly diagnosed paediatric coeliac patients
3. Supporting References
4. Supporting Trust Guidelines
5. Signposting and advice for families
6. Guideline Governance
    a. Scope
    b. Purpose
    c. Duties and Responsibilities
    d. Approval and Ratification
    e. Dissemination and Implementation
    f. Review and Revision Arrangements
    g. Equality Impact Assessment
    h. Document Checklist

Coeliac Disease

Introduction

Coeliac disease is an immune mediated systemic condition in which dietary proteins, (glutens), activate an abnormal mucosal response causing chronic inflammation and villous atrophy to the lining of the small intestine [NICE, 2015].

It is strongly dependent on the human leukocyte antigen (HLA)-DQ2(95% cases) and DQ8 (5% cases) haplotypes.

The prevalence of CD is estimated to be 1:100 in the UK, and 90% of cases are thought to be undiagnosed, therefore low threshold for investigating is advised.

Once diagnosed, coeliac disease can be treated very successfully with a lifelong gluten free (GF) diet. This requires the complete avoidance of wheat, barley, rye and, for a small number of people, oats.

'BSPGHAN recommends that all patients with suspected CD should have their diagnosis established by a paediatric gastroenterologist and their follow-up under the care of a paediatric gastroenterologist or a paediatrician with a special interest in CD, with access to appropriately skilled paediatric dietetic services'

The following guideline has been adapted from the European Society Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN); New Guidelines for the Diagnosis of Paediatric CD 2020.

Clinical features

There is a wide range of presenting symptoms, and can be asymptomatic- picked up on screening.

- **Gastrointestinal -**indigestion, diarrhoea, abdominal pain, bloating, constipation, nausea and vomiting.
- **Non-gastrointestinal-**
- Dermatological- dermatitis herpetiformis, apthous stomatitis
- Endocrine- delayed puberty, amenorrhoea, recurrent miscarriage
- Neurological- peripheral neuropathy
- Growth/Ortho- failure to thrive, short stature, arthralgia
- Haematological- anaemia, unexplained iron, B12 or folate deficiency.
- Hepatic- deranged LFT's

Diagnosis

Increased risk of CD

- Type I diabetes (≥ 8%)
- Selective IgA deficiency (1.7%–7.7%)
- Down (5%–12%), Williams (8.2%) and Turner (4.1%–8.1%) Syndromes
- Autoimmune thyroiditis (~15%)
- Autoimmune liver disease
- Unexplained raised transaminases without known liver disease
- Intussusception
- Dermatitis herpetiformis
- Relatives of coeliac patient:
  - First-degree relative (~10%)
  - HLA-matched sibling (~30%–40%)
  - Monozygotic twin (~70%)

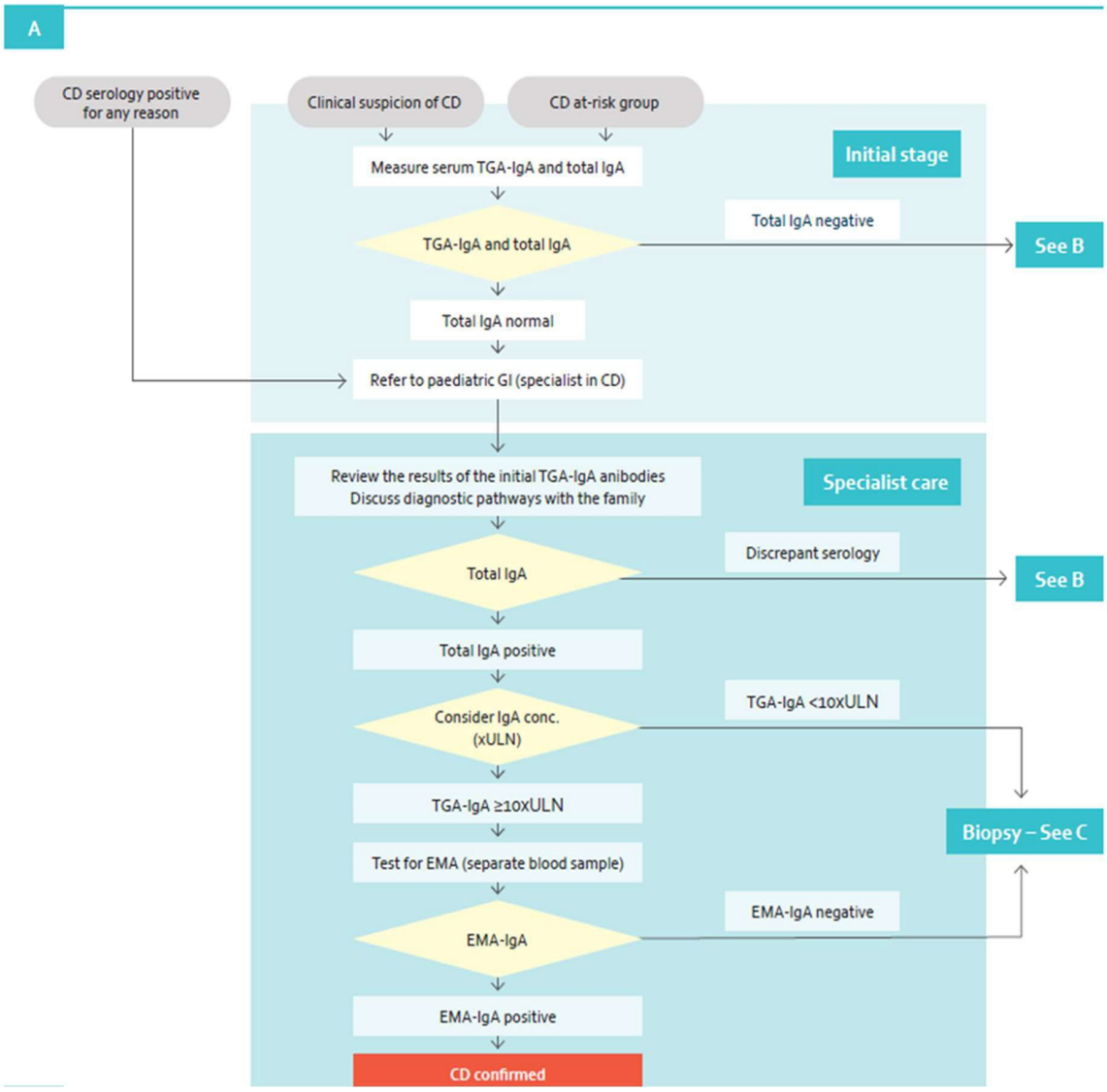
Initial investigations

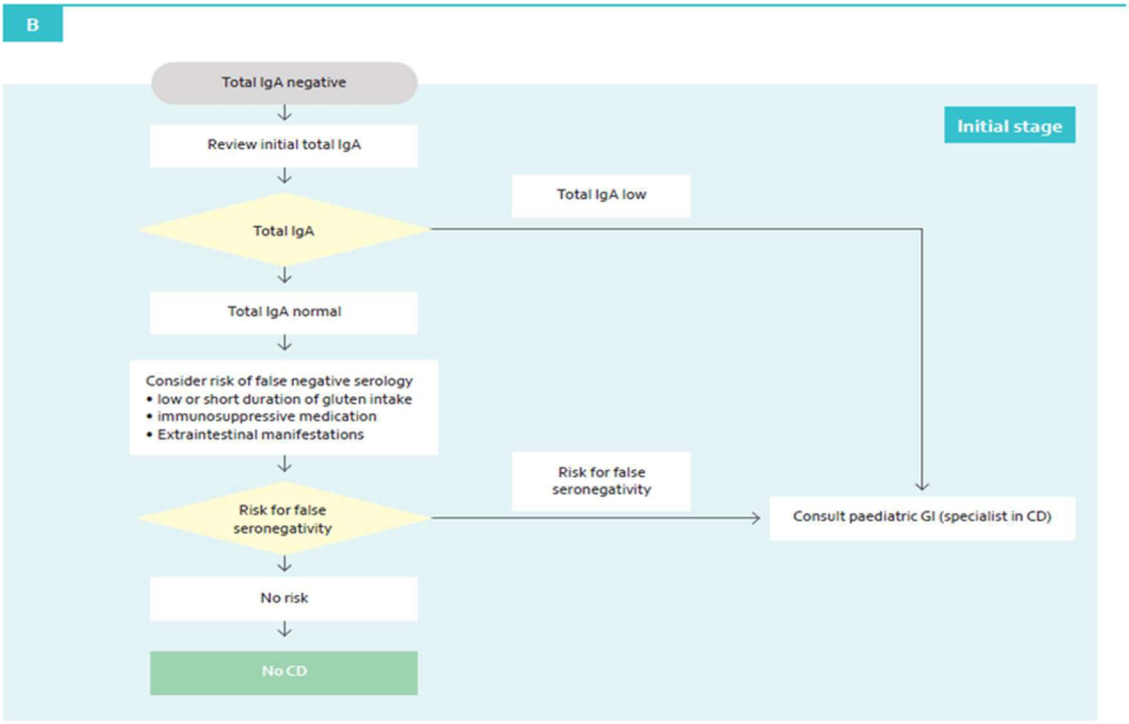
Child should be on gluten containing diet for **3 months** prior to testing.

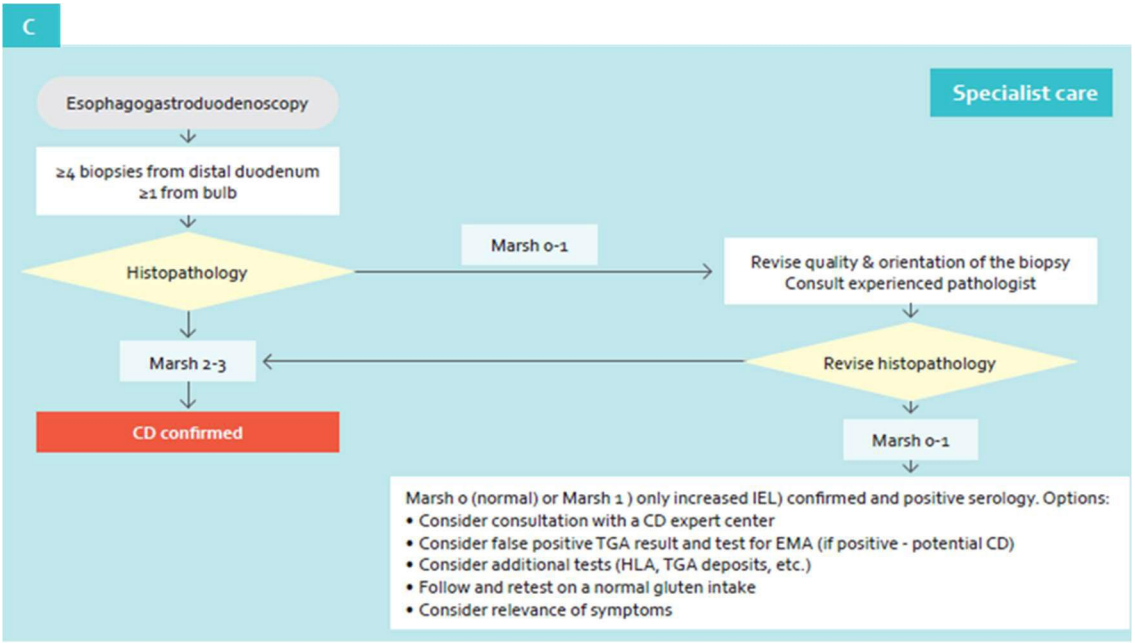
If severe symptoms, ie coeliac crisis, discuss with tertiary centre for consideration of bringing forward investigations.

- Bloods: FBC, ferritin, vitamin D, liver and renal profile bloods, folate, iron, vitamin B_{12}, thyroid function, Total IgA (deficiency if <0.05g/l) and IgA anti-tTG antibodies

Diagnostic Tool for Symptomatic children and At-Risk Groups

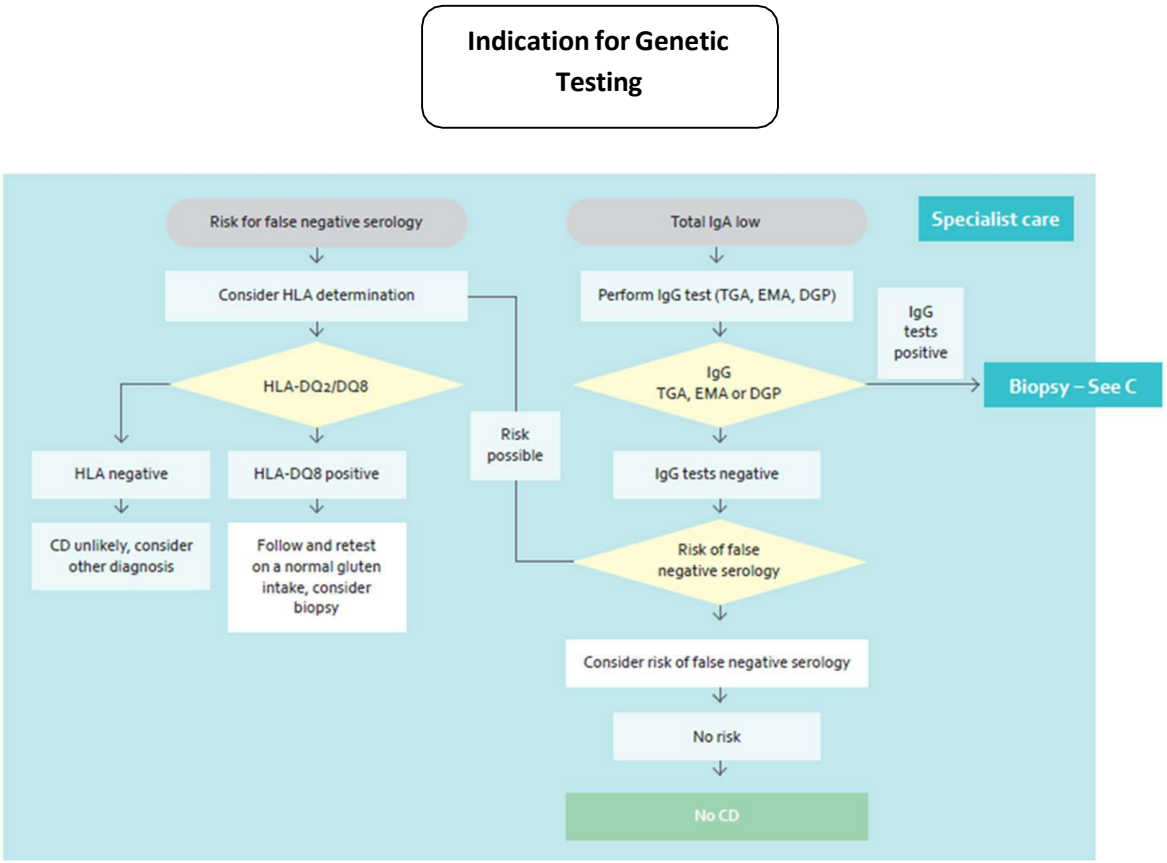



B
Total IgA negative
Review initial total IgA
Total IgA
Total IgA low
Total IgA normal
Consider risk of false negative serology
• low or short duration of gluten intake
• immunosuppressive medication
• Extraintestinal manifestations
Risk for false seronegativity
Risk for false seronegativity
Consult paediatric GI (specialist in CD)
No risk
No CD
Initial stage


C
Esophagogastroduodenoscopy
≥4 biopsies from distal duodenum
≥1 from bulb
Histopathology
Marsh 0-1
Revise quality & orientation of the biopsy
Consult experienced pathologist
Revise histopathology
Marsh 2-3
CD confirmed
Marsh 0-1
Marsh 0 (normal) or Marsh 1) only increased IEL) confirmed and positive serology. Options:
• Consider consultation with a CD expert center
• Consider false positive TGA result and test for EMA (if positive - potential CD)
• Consider additional tests (HLA, TGA deposits, etc.)
• Follow and retest on a normal gluten intake
• Consider relevance of symptoms
Specialist care

If tTG raised (more than 10 times upper limit of normal), and EMA positive, then diagnosis of CD is confirmed; HLA typing and Duodenal biopsy are not required.

Asymptomatic children follow the same pathway as symptomatic children i.e. If tTG≥10 times upper limit of normal with positive EMA then they can be labelled as CD. If tTG < 10 times upper limit of normal then they need duodenal biopsies for confirming CD. There is no value of doing HLA testing as first line in asymptomatic children or as second line testing in symptomatic patients. HLA testing is only indicated under CD specialist care for patients at risk for false negativity.



If diagnostic uncertainty refer for endoscopy and duodenal biopsies including the duodenal cap to identify seronegative CD and other mucosal disorders is required.

If biopsy required referral to Evelina Childrens Hospital Gastroenterology team is required- please discuss with Dr Thakur.

Differentials

See chronic abdominal pain guideline

Non coeliac gluten sensitivity - Reproducible adverse outcome to ingesting gluten, symptomatic or on small intestinal biopsy in whom coeliac disease has been excluded. Potential causes include non IgE mediated food allergy.

Treatment

Gluten Free Diet- life long
Benefits:

- Symptom resolution
- Reversed bone demineralisation.
- Resolution of micronutrient deficiencies and better height gain.
- Decreased rate of delayed puberty, menstrual problems, subfertility, spontaneous abortions and low birth weight babies.
- Decreased rate of some intestinal cancers to normal population levels.
- Possible prevention of onset of other autoimmune conditions (evidence conflicting).

Oats
Approximately 5% with CD are sensitive to Gluten free oats.
For this reason wait until tissue transglutaminase antibody levels normalise before reintroducing uncontaminated oats to the diet.
Patients advised to cease gluten free oats in diet if they become symptomatic or if tTG levels rise at next blood test.

There is no risk of dietary deficiencies on a standard GFD.

Clinic management and follow up

Aims

- Enable patients to independently manage their CD
- Strict GF diet to reduce risk of complications
- Identify and minimise complications and deficiencies

Children should be reviewed within 1 month of diagnosis, and followed up within 6 months. Dependent on recent biochemistry and the family's needs the child will be reviewed annually thereafter. If patients continue to be symptomatic then a more urgent review will be scheduled.

Height and weight
Annual bloods- FBC, ferritin, tTG and vitamin D, LFT, U&E, Folate, iron, vitamin B_{12}, thyroid function and specific IgE should be checked where there is a specific clinical concern.
Dietetic input- from day one of diagnosis. Discuss adherence to diet. See SOP.

Transition

Transition should be discussed in annual review clinic when patients approach the age of 12 years old. Transition will occur between the ages of 16-18 years of age. Prior to transitioning into adult services, the patient should be seen by the paediatric gastroenterologist, who will arrange for transition into appropriate adult services.

When should a patient be discussed with the gastroenterologist?

- Poor response to a strict gluten-free diet
- Unexplained weight loss or poor growth on a strict gluten-free diet
- Persistent diarrhoea or blood in stools
- Onset or on-going unexplained abdominal pain
- Persistent gastrointestinal symptoms
- Persistent nutritional deficiencies
- Other clinical or social concerns

Routine Gluten re-challenge is not required if diagnosis is secure.
Gluten challenges should not be completed routinely and only undertaken under the recommendation of the consultant gastroenterologist in cases where there is diagnostic uncertainty. Timing of the gluten challenge should be carefully considered on a case-by-case basis and it is not advised in children under 5 years of age or during periods of rapid growth such as pubertal growth spurt.

In asymptomatic patients, a challenge period of 3 months is recommended unless symptoms develop with a minimum period of 4-6 weeks in symptomatic patients (Murch et al. 2013). The suggested intake over this period is 10-15g gluten/day (Murch et al. 2013). Parents/families of children undertaking the challenge will be provided a letter outlining the above.

In order to have enough gluten for an accurate test, it is recommended that the child consumes 10-15g gluten per day for at least 6 weeks before the test is done. This roughly equates to 2-3 gluten-containing meals per day. During this time, one can include any foods containing gluten in the diet.

To get an idea of the quantity needed to be consumed, to reach 10-15g gluten per day, the following foods contain 2-3g of gluten.

Examples of portions of these gluten containing foods:

1 medium slice of bread

1 weetabix

2 rusks/digestive biscuits

4 tablespoons of cooked pasta

If symptoms become unmanageable, please do not stop the challenge immediately but get in contact the paediatric team.

2. Dietetic Standard Operating Procedure for newly diagnosed Paediatric Coeliac Patients

Diagnosis

Patient Identified as new coeliac using ASPH Coeliac Procedures

Child referred to Dietetics and prioritised an general outpatient clinic appointment

During Outpatient Appointment

Ensure child and parents understand the diagnosis of Coeliac Disease and what it is: provide education if uncertainty

Consider current weight: height anthropometry and trend in the last few years

Diet history and tailor dietary advice to individual family- consider additional patient factors such as any additional dietary restrictions

Advise on GFD – foods to avoid (wheat, barley, oats and rye) and other foods to include freely as alternatives - explanations of food labelling

Consider nursery/school meals

Consider cross contamination risks e.g. sharing spreads/ cutlery/ toasters with other family members

Advice on Oats: Establish child on a strict gluten free diet initially excluding ALL oats. After a year when tTg is re-tested and antibodies have normalised (and weight gain is optimal), then gluten free oats can be considered for reintroducing into the diet. Patients will be advised to stop gluten free oats again if they become symptomatic or tTg levels start to rise again

Consider clubs for example brownies, scouts etc.
How to manage social situations, eating out and family holidays
Advice on optimising calcium and iron and Vitamin D intake.
Advice on avoidance of 'very low gluten foods' and foods that have a 'may contain' label
Advice on Coeliac UK and 'Food and Drink Directory' and signpost to Coeliac UK membership for regular updates?
Discuss GF prescribing advice from GP

Follow-up

Follow-up will be offered at 6 monthly intervals initially for the first year and thereafter annually. Additional input can be provided as required, for example difficulties with dietary compliance

3. Supporting References

European Society Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN); New Guidelines for the Diagnosis of Paediatric CD 2020.

Available at:https://paediatrie.at/images/AGLeiter/Gastroenterologie/new-guidelines-for-the-diagnosis-of-paediatric-coeliac-disease-espghan-advice-guide-2020-ver11.pdf

National Institute for Health and Care Excellence (NICE) clinical guideline NG20. Coeliac Disease: recognition, assessment and management. September 2015

Paediatric Gastroenterology Guideline: Management of Children and young persons with Coeliac Disease- Rita Shergill-Bonner , Dr Vora, Dr Mohamed Mutalib, Dr Jochen Kammermeier, Annina Whipp. April 2020

4. Supporting relevant trust guidelines

Chronic abdominal pain in children
https://ashfordstpeters.net/Guidelines_Paediatrics/Abdominal%20Pain%20-%20Chronic%20Dec%202019.pdf

5. Signposting and advice for families

https://www.coeliac.org.uk/home/

https://www.bda.uk.com/resource/wheat-free-diet.html

4. Guideline Governance

a. Scope

This guideline in relevant to all staff caring for all children from 0-18 years old across the emergency department, inpatient ward and outpatient department.

b. Purpose

i. This guidelines aims to facilitate a common approach to the management of children. At times deviation from the guideline may be necessary, this should be documented and is the responsibility of the attending consultant.

ii. This guideline is subject to regular review to ensure ongoing evidence based practice.

c. Duties and Responsibilities

What is expected from the health care professionals using this guideline to look after children age 0-18 years old.

d. Approval and Ratification

This guideline will be approved and ratified by the Paediatric Guidelines Group.

e. Dissemination and Implementation

i. This guideline will be uploaded to the trust intranet 'Paediatric Guidelines' page and thus available for common use.
ii. This guideline will be shared as part of ongoing education within the Paediatric Department for both medical and nursing staff.
iii. All members of staff are invited to attend and give comments on the guideline as part of the ratification process.

f. Review and Revision Arrangements

a. This policy will be reviewed on a 3 yearly basis.
b. If new information comes to light prior to the review date, an earlier review will be prompted.
c. Amendments to the document shall be clearly marked on the document control sheet and the updated version uploaded to the intranet. Minor amendments will be ratified through the Paediatric Guidelines Group. A minor amendment would consist of no major change in process, and includes but is not limited to, amendments to documents within the appendices.

g. Equality Impact Assessment

Background • Who was involved in the Equality Impact Assessment
Author and the supervising consultants
Methodology • A brief account of how the likely effects of the policy was assessed (to include race and ethnic origin, disability, gender, culture, religion or belief, sexual orientation, age) • The data sources and any other information used • The consultation that was carried out (who, why and how?)
All groups of staff and patients were taken into consideration and there is no bias towards or against any particular group.
Key Findings • Describe the results of the assessment • Identify if there is adverse or a potentially adverse impacts for any equalities groups
There is no evidence of discrimination.
Conclusion • Provide a summary of the overall conclusions
There is no evidence of discrimination.
Recommendations • State recommended changes to the proposed policy as a result of the impact assessment • Where it has not been possible to amend the policy, provide the detail of any actions that have been identified • Describe the plans for reviewing the assessment
This guideline is appropriate for use.

h. Document Checklist

To be completed (electronically) and attached to any document which guides practice when submitted to the appropriate committee for approval or ratification.

Title of the document: Coeliac Disease – Diagnosis and Management

Policy (document) Author: Dr Emily Taylor

Executive Director: Dr Alka Thakur

		Yes/No/ Unsure/NA	Comments
1.	**Title**		
	Is the title clear and unambiguous?	**Yes**	
	Is it clear whether the document is a guideline, policy, protocol or standard?	**Yes**	
2.	**Scope/Purpose**		
	Is the target population clear and unambiguous?	**Yes**	
	Is the purpose of the document clear?	**Yes**	
	Are the intended outcomes described?	**Yes**	
	Are the statements clear and unambiguous?	**Yes**	
3.	**Development Process**		
	Is there evidence of engagement with stakeholders and users?	**Yes**	
	Who was engaged in a review of the document (list committees/ individuals)?		**Paediatric Guideline Group 18/01/21**
	Has the policy template been followed (i.e. is the format correct)?	**Yes**	
4.	**Evidence Base**		
	Is the type of evidence to support the document identified explicitly?	**Yes**	

		Yes/No/ Unsure/NA	Comments
	Are local/organisational supporting documents referenced?	**N/A**	
5.	**Approval**		
	Does the document identify which committee/group will approve/ratify it?	**Yes**	
	If appropriate, have the joint human resources/staff side committee (or equivalent) approved the document?	**N/A**	
6.	**Dissemination and Implementation**		
	Is there an outline/plan to identify how this will be done?	**Yes**	
	Does the plan include the necessary training/support to ensure compliance?	**Yes**	
7.	**Process for Monitoring Compliance**		
	Are there measurable standards or KPIs to support monitoring compliance of the document?	**Yes**	
8.	**Review Date**		
	Is the review date identified and is this acceptable?	**Yes**	
9.	**Overall Responsibility for the Document**		
	Is it clear who will be responsible for coordinating the dissemination, implementation and review of the documentation?	**Yes**	
10.	**Equality Impact Assessment (EIA)**		
	Has a suitable EIA been completed?	**Yes**	

Committee Approval (Neonatal Guidelines Committee)			
If the committee is happy to approve this document, please complete the section below, date it and return it to the Policy (document) Owner			
Name of Chair	**Dr Claire Mitchell**	**Date**	**08/03/2024**
Ratification by Management Executive (if appropriate)			
If the Management Executive is happy to ratify this document, please complete the date of ratification below and advise the Policy (document) Owner			
Date: n/a			