LRI Children's Hospital

Coeliac disease

Staff relevant to:	UHL Nursing & Medical staff caring for Children diagnosed with Coeliac disease within the Children's Hospital
Team approval date:	April 2025
Version:	6
Revision due:	April 2030
Reviewed by:	S. Pande, K. Devlin, E. Buick
Trust Ref:	C82/2006

Contents

1. Introduction and who this guideline applies to

Definition

Coeliac disease is an immune mediated systemic disorder that occurs in genetically predisposed people where the ingestion of gluten leads to damage in the small intestine. The prevalence of coeliac disease in the UK is estimated to be 1:100. There should be a low threshold for investigating both symptomatic children and those with associated conditions. The estimated lifetime prevalence of coeliac disease in first degree relatives of someone with coeliac disease is approximately 10%.

When someone with coeliac disease consumes gluten (a protein found in wheat, rye and barley), the body mounts an immune response that attacks the small intestine. This causes damage to the villi which can lead to malabsorption, faltering growth, and nutritional deficiencies in addition to other problems.

2. Guideline Standards and Procedures

2.1 Who to investigate

- First degree relatives of coeliac patients (10%)
- Asymptomatic children who have conditions associated with coeliac disease such as Type 1 Diabetes Mellitus (8%), Trisomy 21 (5-12%) and auto-immune thyroiditis.
- Faltering growth.
- Children with persisting GI symptoms such as abdominal pain, constipation, vomiting, chronic or intermittent diarrhoea
- Children with non-GI symptoms such as Dermatitis Herpetiformis, dental enamel hypoplasia of permanent teeth, short stature, chronic fatigue, irritability, and neuropathy.
- Delayed puberty
- Iron deficiency anaemia resistant to oral iron
- Selective IgA deficiency (2%)
- Unexplained abnormal LFTs
- Turner syndrome (4.1—8.1%)
- Williams syndrome (8.2%)

2.2 Investigation

Gluten Challenge

It is very important to check whether the child is on a gluten-containing diet before undertaking tTG screening test. Exclusion or reduction of gluten intake is very likely to cause an inaccurate result for blood tests, and biopsy if required.

It is recommended for cases of uncertain coeliac disease diagnosis that HLA typing should be performed prior to gluten challenge in order to eliminate children in whom the occurrence of coeliac disease is unlikely.

If gluten has already been excluded or reduced, it will need to be reintroduced to ensure accurate test results. As a general guideline, the recommendation is to eat gluten (10-15g per day) usually in at least two meals every day for at least 12 weeks before testing.

Investigation of symptomatic children

Ask for "Coeliac screen" on immunology form *(please see 3.2 Gluten Challenge)*

- IgA tTG (Human recombinant anti tissue transglutaminase antibody)
    - Very high sensitivity (92-100%) and specificity (91-100%)

- Anti-endomysial antibody (IgA) is done by the lab if tTG positive.
    - High sensitivity (88- 100%) and specificity (91-100%).
    - Less accurate in children under 2 years of age

If tTG is greater than the normal range, refer to Paediatric Gastroenterology for confirmation of coeliac disease which may require a biopsy. Please advise family not to reduce or exclude gluten at this stage unless told to do so by Paediatric Gastroenterology

If the level is 10x upper limit of normal or more and a second blood sample indicating EMA – IgA positivity is sufficient for diagnosis of coeliac disease via the no biopsy pathway. Please advise family not to reduce or exclude gluten at this stage unless told to do so by paediatric gastroenterology.

IgA deficiency will give a false negative IgA tTG result. Please note – in children with low total IgA concentrations, an IgG- based test (EMA) should be performed as a second step.

Refer to ESPGHAN guideline for Diagnosing coeliac disease 2020 and the flowchart in figure 1 of appendix for the full interpretation of results.

2.3 Screening of asymptomatic genetically predisposed children

Some children are at higher risk of developing coeliac disease (see 2.0); many of these children are screened regularly with tTG as part of routine blood tests.

If this result is above normal range, but less than 3x upper limit of normal and the child is not symptomatic, this will need to be monitored but not treated with any diet change. Please bear in mind the symptoms of coeliac disease are very varied (see above).

If the result is >3x upper limit of normal please refer to Paediatric Gastroenterology for further investigation.

Please advise family not to reduce or exclude gluten at this stage unless told to do so by paediatric gastroenterology.

2.4 Criteria for Diagnosis

Diagnosis is made on following basis, but needs to be done under the care of Paediatric Gastroenterology. Once you have a positive screening tTG then please refer **ALL** cases to Paediatric Gastroenterology at this stage. Emphasize the need to stay on a **normal gluten containing diet** unless or until told to change by a paediatric gastroenterologist.

If tTG >10x normal on first test:

1) Lab will automatically also do antiendomyseal antibody (EMA).
2) If tTG- IgA is 10 times or above the upper limit of normal, and EMA positive this is sufficient for the diagnosis of coeliac disease. Refer to ESPGHAN guideline for the diagnosis of coeliac disease for the full interpretation of results, and appendix below detailing a flowchart for the diagnosis of coeliac disease.

If tTG is greater than normal but <10x normal:

1) No further blood testing appropriate as these children will need duodenal biopsies taken to confirm diagnosis. This is done via upper GI endoscopy under general anaesthetic.

2.5 Management

The management of coeliac disease is life-long adherence to a strict gluten free diet. Benefits of gluten free diet are:

- Resolution of symptoms
- Improved growth, resolution of faltering growth
- Likely improved height growth to expected centile
- Reversal of bone demineralisation if occurs before age 19 years.
- Resolution of micronutrient deficiencies
- Decreased rate of delayed puberty and menstrual problems
- Decreased rate of some intestinal cancers to normal population level
- Possible improved glycaemic control in those with type 1 diabetes
- Improvement in sense of physical and psychological wellbeing.

Once diagnosis is confirmed by Paediatric Gastroenterologist, the child is referred to UHL Paediatric Dietetic – Led Coeliac clinic.

Once referral to Senior Specialist Paediatric Coeliac Dietitian is received, the Dietitian arranges to meet family face to face in clinic or contacts them by telephone as soon as possible to advise regarding gluten free diet. A coeliac pack is posted to the family.
This includes:

- Coeliac UK - Coeliac Disease and Me booklet
- UHL Coeliac Disease in Children leaflet
- Written information on dietary calcium and iron intake
- Leicestershire County Council school meals form/ online link to form (if applicable).
- Coeliac UK – School Pack
- Leicestershire Coeliac UK group flyer
- Free sample cards for various gluten free brands and products

Depending on dietetic assessment, the child is usually followed up in Dietetic-Led Paediatric Coeliac clinic at 6 and then 12 months after diagnosis. Once tTG IgA levels are within

normal limits reviews are scheduled at 12-24 month intervals. A full dietetic review and discussion of adherence to gluten free diet is discussed at each review consultation. Routine coeliac bloods as per ESPGHAN guideline are undertaken at each review (6-24 monthly). Follow-up appointments can be brought forward from the above timeline if there are concerns from patient, parents/carers, health professionals, previous dietetic assessment, and above normal tTG-IgA level.

All families should be advised to join Coeliac UK.

Most children would benefit from an age appropriate over the counter multivitamin supplement which contains vitamin D, particularly over the autumn and winter months. Parents should be reminded to check that this is gluten free.

Temporary lactose intolerance may occur around the time of diagnosis. Dietetic advice is required. It is suggested to trial a low lactose diet in children with symptoms suggestive of lactose intolerance i.e., ongoing diarrhea, abdominal pain, or gassiness despite adherence to the gluten free diet.

Dexa scan- not routinely done in children unless clinically indicated.

Further information

Refer to the ESPGHAN guidelines for the diagnosis (2020) and management of coeliac disease (2022).

3. Education and Training

None

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
1) Audit – done 2018, aim to redo in 3 yrs 2) Abnormal screening bloods sent to gastroenterology consultants monthly	Compliance with the guideline confirmed after audit	A Willmott S Pande	Every 3 years Monthly	Local dept and national poster presentation at BSPGHAN annual meeting

5. Supporting References

1. Mearin, Maria Luisa et al. “ESPGHAN Position Paper on Management and Follow-up of Children and Adolescents With Celiac Disease.” Journal of pediatric gastroenterology and nutrition vol. 75,3 (2022): 369-386. doi:10.1097/MPG.0000000000003540
2. Husby, Steffen et al. “European Society Paediatric Gastroenterology, Hepatology and Nutrition Guidelines for Diagnosing Coeliac Disease 2020.” Journal of pediatric gastroenterology and nutrition vol. 70,1 (2020): 141-156. doi:10.1097/MPG.0000000000002497

6. Key Words

Coeliac disease (CD), Gluten, Paediatric gastroenterology

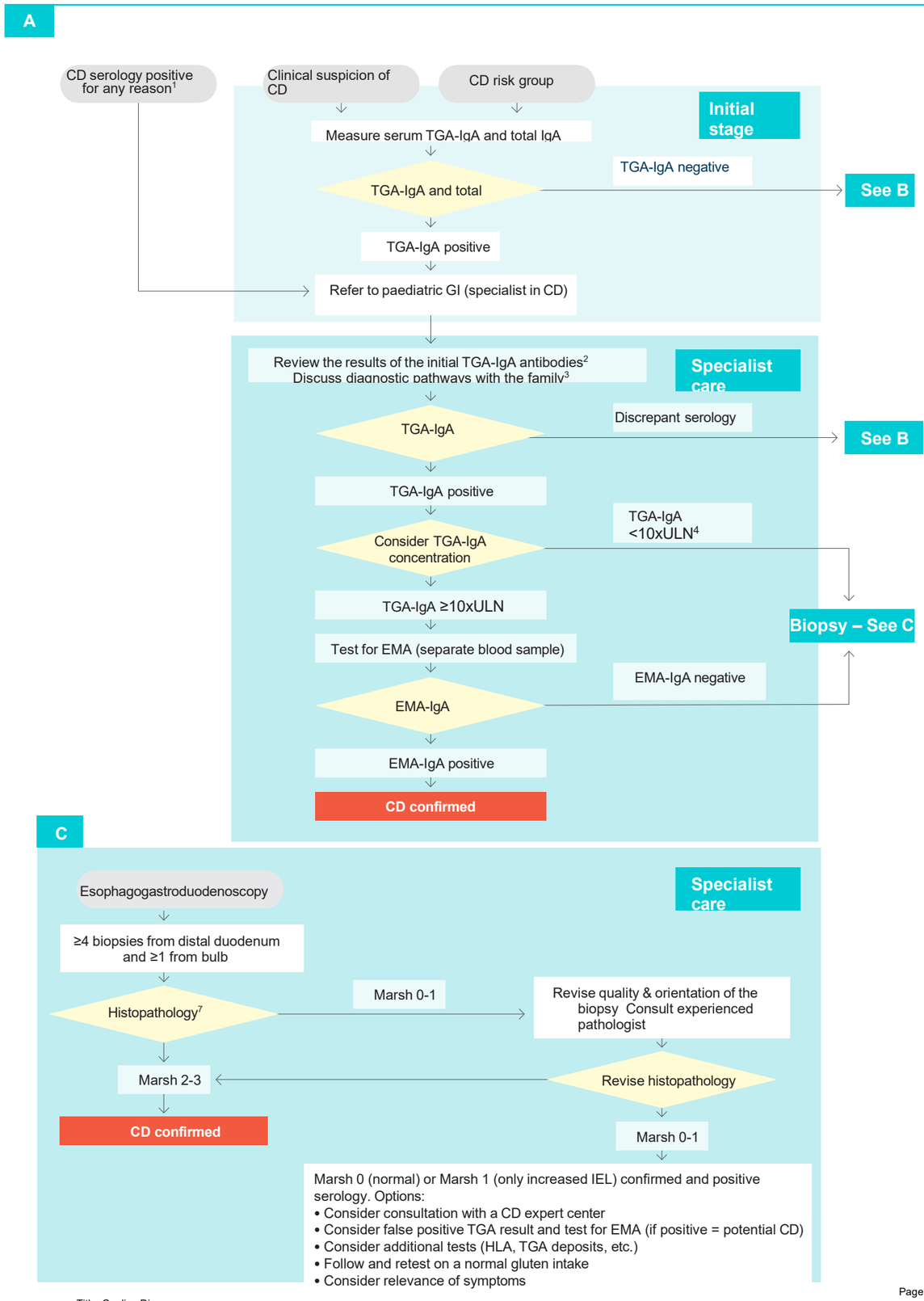
The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

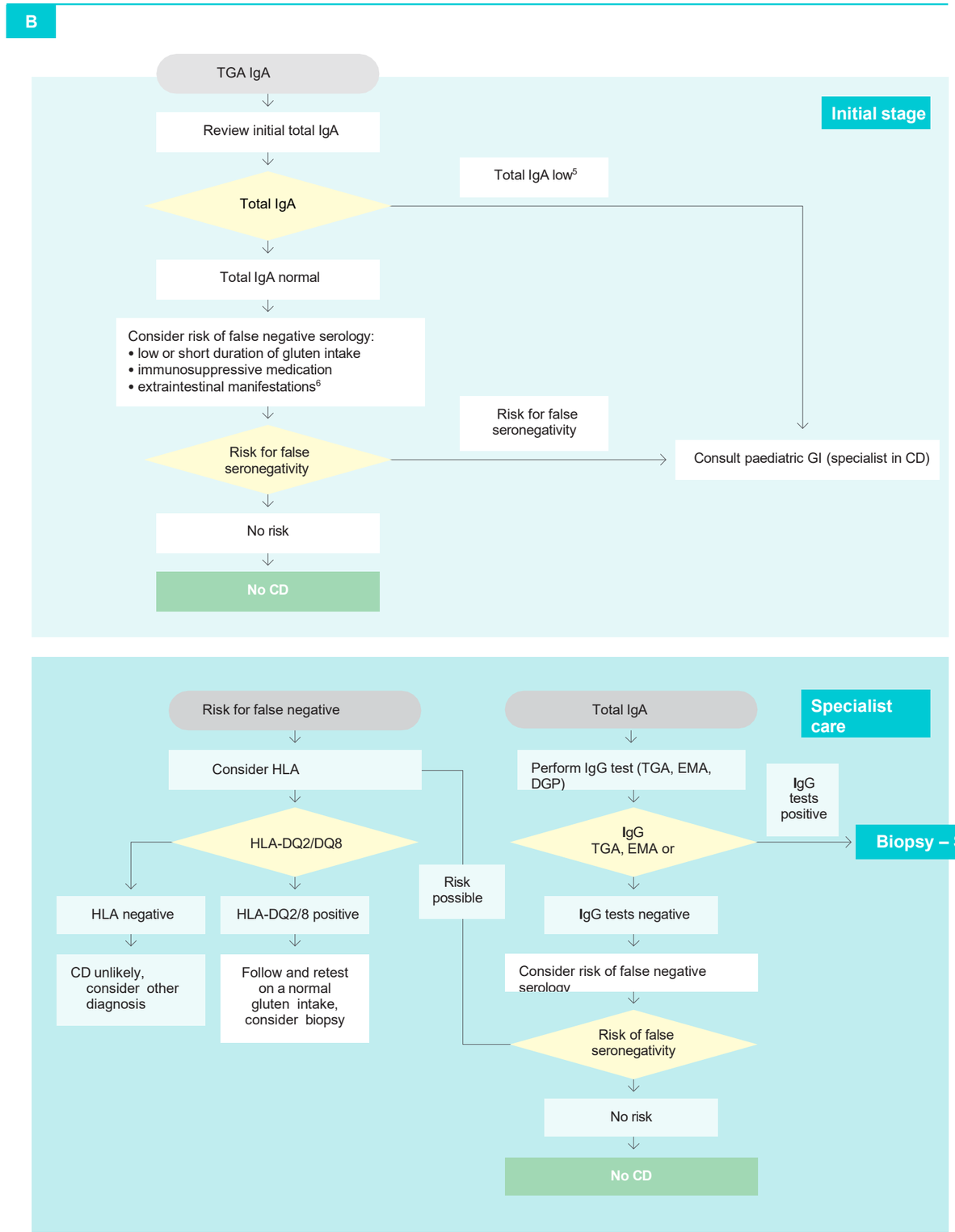
CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title) **2019 A Willmott - Consultant** Previous version; Y Lakhani, Dr A Willmott, Dr M Green, Sara Mcdowell July 2006, Aug 2009 May 2016: Dr A Willmott, Dr S Pande, Kristian Bravin	**Executive Lead** Chief Nurse
Details of changes made during review (March 2025): Addition to page 2: further examples of people to investigate 'chronic or intermittent diarrhoea' , and chronic fatigue, irritability, and neuropathy. Clarification on children with uncertain coeliac disease diagnosis further testing for HLA should be done prior to gluten challenge. Clarification of gluten challenge amount '10-15g /day', and update to 12 weeks before re test of TGA. Addition to page 3 – updates to the diagnosis of coeliac disease as per ESPGHAN guidelines for diagnosing coeliac disease 2020. "Please note – in children with low total IgA concentrations, an IgG- based test (EMA) should be performed as a second step". Only require one positive tTG IgA above 10 times upper limit of normal and a separate positive EMA blood sample test to serologically confirm the diagnosis of coeliac disease. Redactions from page 3, HLA typing no longer necessary for diagnosis of coeliac disease. Redaction of "Please note - in young children (<2 years) we have a lower threshold to confirm with duodenal biopsy if atypical presentation, even with higher tTG. Refer to Joint BSPGHAN and Coeliac UK guideline for full interpretation of results". All children can be diagnosed the same through serum and biopsy pathways as indicated in ESPGHAN 2020 guideline. Addition to page 4: Review times stretched to 24 months as clinically indicated and agreed with parents/carers as per guidance from ESPGHAN coeliac disease management and follow up (2022). Redaction from Page 4: Leicester City Schools no longer have a school meal form, as school meal dietitian no longer in place. Page 5: Redaction of references changed to updated guidelines from ESPGHAN: European Society Paediatric Gastroenterology, Hepatology and Nutrition Guidelines for Diagnosing Coeliac Disease (2020). ESPGHAN Position Paper on Management and Follow up of Children and Adolescents With Celiac Disease (2022). Page 8: Addition of ESPGHAN flowchart for the diagnosis of coeliac disease, in formulated appendix.	

APPENDIX- Algorithm for the diagnosis of coeliac disease (ESPGHAN, 2020)

See next page

FIGURE 1 (Algorithm for the diagnosis of coeliac disease) (ESPGHAN, 2020))





Title: Coeliac Disease
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Footnotes

1. Other than TGA-IgA, including point-of-care tests and DGP. 2. Check the value also in relation to the cut-off and repeat the test if questionable or borderline. No need to retest if done with validated assay with calibration curve. Test with conventional TGA-IgA test if positive POCT and TGA has not been measured quantitatively. 3. Convey the message that the diagnosis of coeliac disease with or without biopsy confirms the need for a lifelong gluten-free diet and that re-evaluation after introduction of the diet would need prolonged re-exposure to gluten with a series of further investigations. 4. If TGA-IgA is only borderline positive confirm sufficient gluten intake and considerer re-testing of TGA-IgA and EMA. 5. Low for age or <0.2 g/L above the age of 3 years. 6. For example, dermatitis herpetiformis, in which serology is frequently negative. 7. The cut-off for normal numbers of IEL is >25 cells/100 enterocytes.