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
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Original written by: Reviewed by:	Y Lakhani, A Willmott, M Green & S Mcdowell S. Pande, K. Devlin
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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 is 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.

This guideline is relevant to UHL Nursing & Medical staff caring for Children diagnosed with Coeliac disease within the Children's Hospital.

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, diarrhoea
- Children with non-GI symptoms such as Dermatitis Herpetiformis, dental enamel hypoplasia of permanent teeth, short stature.
- 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.

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 some gluten in at least two meals every day for at least six weeks before testing.

Investigation of symptomatic children

Ask for "Coeliac screen" on immunology form *(please see 2.2 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 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 on two separate tests, antiendomyseal antibody is positive and genetic test (HLA) is positive, biopsy may not be necessary for diagnosis. Please advise family not to reduce or exclude gluten at this stage unless told to do so by paediatric gastroenterology.

IgA level is routinely tested as part of coeliac screen. IgA deficiency will give a false negative IgA tTG result in coeliac disease and in this instance IgG tTG will be tested although is not as sensitive as a marker of coeliac disease as IgA tTG. We recommend duodenal biopsies in children with IgA deficiency if Coeliac disease is suspected from symptoms, family history, other autoimmune conditions, genetic conditions etc. Please note, temporary IgA deficiency is common in children and it is worth repeating Coeliac blood test with IgA level before referring children for duodenal biopsies as IgA level may recover.

Refer to Joint BSPGHAN and Coeliac UK guideline for full interpretation of results.

2.3 Screening of asymptomatic genetically predisposed children

Some children are at higher risk of developing coeliac disease (see 2.1); 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 care of Paediatric Gastroenterology. Once you have a positive screening tTG then please refer **ALL** cases to Paediatric Gastroenterology at this stage, emphasising 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.
2. A second blood test including repeat tTG and HLA typing for coeliac disease is required.

If all three results meet the criteria for diagnosis by blood test, then it is possible to diagnose without a biopsy. Refer to Joint BSPGHAN and Coeliac UK guideline for full interpretation of results.

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.

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.

2.5 Management

The management of coeliac disease is life-long adherence to a 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 also 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
- Leicester City Council / Leicestershire County Council school meals 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 12 months after diagnosis and at 12-18 month intervals thereafter with full dietetic review and discussion of adherence to gluten free diet. Routine coeliac bloods as per Joint BSPGHAN and Coeliac UK guideline are undertaken at each review (6-12 monthly).

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. Persistent lactose intolerance requires further assessment to exclude poor dietary compliance to gluten free diet or a separate issue of cow's milk protein intolerance.

Dexa scan- not routinely done in children unless concern.

Further information

Refer to the Joint BSPGHAN and Coeliac UK guidelines for the diagnosis and management of coeliac disease in children (2013).

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
Abnormal screening bloods sent to gastro consultants monthly	Audit	A Willmott S Pande	Every 3 years	Local dept and national poster presentation at BSPGHAN annual meeting

5. Supporting References

1. Joint BSPGHAN and Coeliac UK guidelines for the diagnosis and management of coeliac disease in children Arch Dis Child 2013 98: 806-811 originally published online August 28, Simon Murch, Huw Jenkins, Marcus Auth, et al.
2. Europ https://www.coeliac.org.uk/document-library/122-https://www.coeliac.org.uk/document-library/122-bspghan/?&&type=rfst&set=true#cookie-widget – accessed March 2022

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) **S Pande – Consultant** **K Devlin - Senior Specialist Dietitian**	**Executive Lead** Chief Nurse
Details of Changes made during review: Updated introduction to include prevalence and symptoms Updated IgA & duodenal biopsy results review and actions Added reference to coeliac.uk. throughout Gluten challenge – to ensure that 6 weeks (previously 4 weeks) with at least two gluten containing meals per day are consumed Removed reference to Type 1 diabetes being treated with a GFD Dexa scan not to be offered routinely Section 2.5 – management- updated health improvements once following GFD and added reference to coeliac pack for further information	