Diagnosis and Treatment of Celiac Disease in Children

Clinical Practice Guideline Summary

PURPOSE:

This clinical practice guideline summary was developed to assist the primary and specialist medical provider in the diagnosis and treatment of celiac disease in children. Recommendations are based on an integration of a comprehensive and systematic review of the medical literature combined with expert opinion.

The following sections summarize the conclusions and recommendations of the NASPGHAN Celiac Disease Guideline Committee on the value of diagnostic tests and treatment modalities commonly used for the management of celiac disease, and how those strategies can be applied to clinical situations. The complete guideline can be downloaded from www.naspghan.org.

DEFINITION:

Celiac Disease: an immune-mediated enteropathy caused by a permanent sensitivity to gluten in genetically susceptible individuals

PREVALENCE IN CHILDREN BETWEEN 2.5 AND 15 YEARS OF AGE:

3 to 13 per 1000 children or approximately 1 in 300 to 1 in 80 children. Therefore, in a pediatric practice of 1500 children there are probably between 5 and 20 children with CD, either diagnosed or undiagnosed.

GASTROINTESTINAL MANIFESTATIONS OF CELIAC DISEASE IN CHILDREN:

- Diarrhea with failure to thrive
- Abdominal pain
- Vomiting
- Constipation
- Abdominal distension

CDHNF
Children's Digestive Health and Nutrition Foundation

NASPGHAN
North American Society for Pediatric Gastroenterology, Hepatology and Nutrition

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The North American Society of Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) represents more than 1000 pediatric gastroenterologists predominantly located in the United States, Canada and Mexico. NASPGHAN strives to improve the care of infants, children and adolescents with digestive disorders by promoting advances in clinical care, research and education.

The Children's Digestive Health and Nutrition Foundation (CDHNF) was established by NASPGHAN to promote research and education that will improve the health of children with digestive and nutritional disorders.

CDHNF/NASPGHAN guidelines on the Diagnosis and Treatment of Celiac Disease in Children will be published in the Journal of Pediatric Gastroenterology and Nutrition ©2005; Volume 40, Number 1 (Jan): pages 1-19. Complete guidelines can also be found on the following websites: ***www.cdhnf.org*** *and* ***www.naspghan.org.***

TABLE 1 - *Non-gastrointestinal manifestations of celiac disease*

A) Manifestations for which there is strong to moderate evidence of an association with celiac disease
Dermatitis herpetiformis
Dental enamel hypoplasia of permanent teeth
Osteopenia and osteoporosis
Short stature
Delayed puberty
Iron-deficient anemia unresponsive to treatment with oral iron *(well documented in adults only)*
B) Manifestations for which the evidence is less strong
Hepatitis (elevated liver enzymes)
Arthritis
Epilepsy with occipital calcifications

TABLE 2 - *Conditions associated with an increased prevalence of celiac disease*

Type 1 diabetes	
Autoimmune thyroiditis	
Down Syndrome	
Turner Syndrome	
Williams Syndrome	
Selective IgA deficiency	
First degree relatives of celiac patients	

TESTING IS RECOMMENDED FOR CHILDREN WITH:

- Diarrhea and failure to thrive
- Persistent GI symptoms including recurrent abdominal pain, anorexia, constipation and vomiting
- Dermatitis herpetiformis
- Dental enamel hypoplasia of permanent teeth
- Osteoporosis
- Short stature
- Delayed puberty
- Iron-deficient anemia resistant to oral iron
- Asymptomatic children with conditions associated with an increased prevalence of Celiac Disease *(See Table 2)*

It is recommended that testing of asymptomatic children who belong to groups at risk begin around 3 years of age provided they have had an adequate gluten containing diet for at least one year prior to testing. It is also recommended that asymptomatic patients with negative serological tests who belong to groups at risk be considered for repeat testing at intervals.

HOW TO TEST?

The diagnosis of celiac disease is diagnosed definitively by biopsy of the small intestinal mucosa. Serological (blood) tests are used for case finding (screening) to determine who is likely to have celiac disease when biopsied. Selective IgA deficiency is a factor when using serological tests.

SEROLOGICAL TESTING

- Measure TTG (tissue transglutaminase) for initial blood testing. TTG is the IgA antibody to human recombinant tissue transglutaminase.
- Measurement of endomysial antibody (IgA antibody to endomysium, EMA) may also be reliable but is subject to added cost and interpretation error.
- AGA IgA and AGA IgG tests are no longer recommended as initial testing due to the inferior accuracy of antigliadin antibody tests (AGA).

IgA Deficiency

- Individuals with CD who are also IgA deficient will not have abnormally elevated levels of TTG IgA. When testing for CD in children with symptoms suspicious for CD, measurement of quantitative serum IgA can facilitate interpretation when the TTG IgA is low.
- In individuals with known selective IgA deficiency and symptoms suggestive of CD, testing with TTG IgG is recommended.

Intestinal biopsy

- When the serum TTG is elevated it is recommended that a small intestinal biopsy be performed.
- It is currently recommended that confirmation of the diagnosis of CD requires an intestinal biopsy in all cases.
- Even if serological tests for CD are negative, a small intestinal biopsy may be useful in symptomatic children (particularly when they are 0 to 2 years old) with chronic diarrhea, FTT, a positive family history of CD or IgA deficiency to identify the unusual case of serology negative CD or other intestinal mucosal disorders. [RBC2]

WHO TO TREAT?

- A gluten-free diet is recommended for all symptomatic children with intestinal histopathological abnormalities that are characteristic of Celiac Disease.
- A gluten-free diet is also recommended for asymptomatic children who have a condition associated with Celiac Disease and characteristic histological findings on small intestinal biopsy.

BENEFITS OF TREATMENT:

There is evidence that treatment with a gluten-free diet:

- Resolves symptoms of persistent diarrhea and poor weight gain
- Reverses reduced bone mineralization
- Can prevent the onset of osteoporosis, decrease the incidence of spontaneous abortions in fertile females, lower the incidence of low birth weight infants, decrease the risk of cancer and avoid other consequences of delayed diagnosis.

HOW TO TREAT?

- A gluten-free diet for life
- Treatment should only be started after diagnosis is confirmed by intestinal biopsy
- American Dietetic Association guidelines for the treatment of Celiac Disease have been endorsed by the NASPGHAN Celiac Disease Guideline Committee
- Ingesting even small amounts of gluten can lead to mucosal changes on intestinal biopsy
- Management of a gluten-free diet is an ongoing collaboration between the patient, health care professional and dietician
- Dietary lactose restrictions are not usually necessary

HOW TO MONITOR?

- Periodic visits for assessment of symptoms, growth, physical examination and adherence to the gluten-free diet
- Measurement of TTG 6 months after treatment with a gluten-free diet is begun, and then approximately once a year if the patient has no symptoms
- Measurement of TTG at any time after starting a gluten-free diet if the patient has persistent or recurring symptoms
- Studies show that about 45% to 80% of children adhere to a gluten free diet

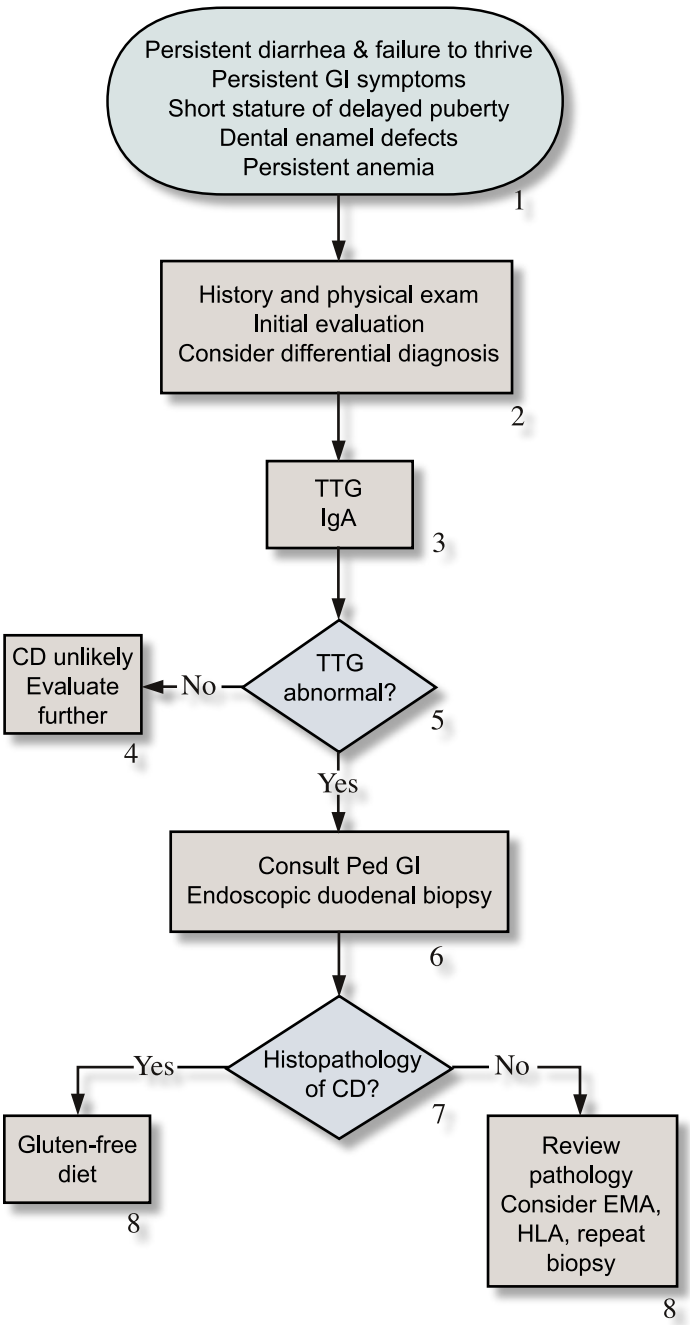
GUIDELINE SUMMARY DISCLAIMER:

The guidance in this report does not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

ALGORITHMS:

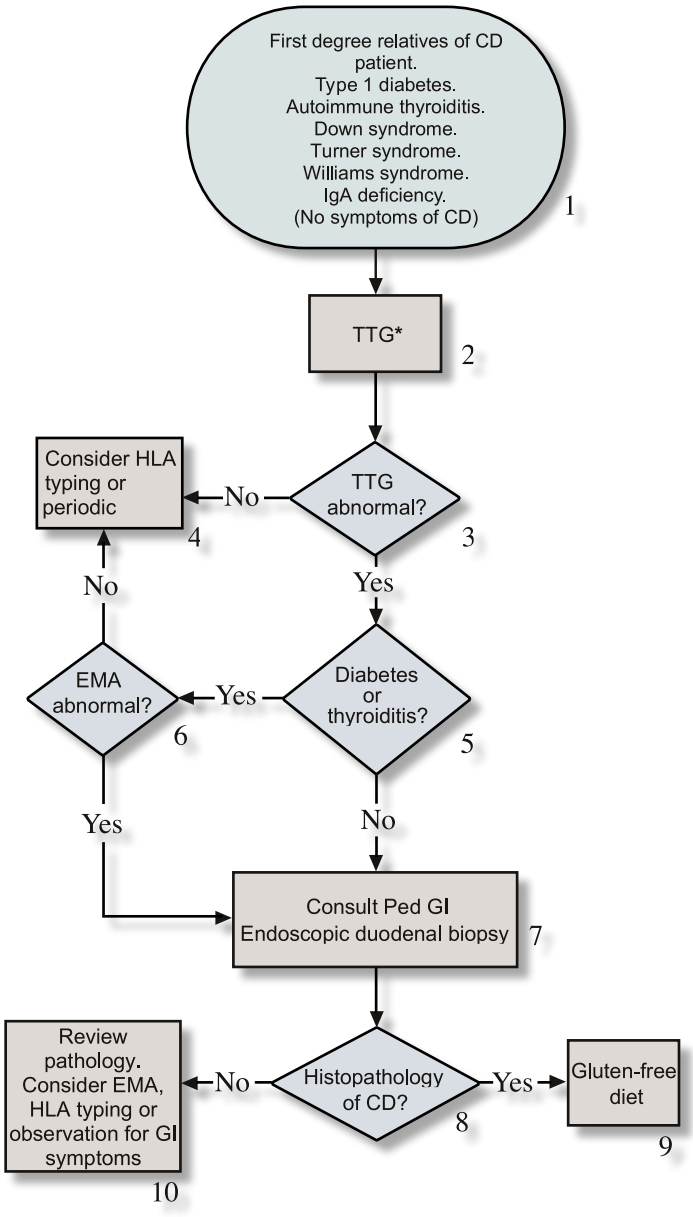
Algorithm for the evaluation for celiac disease in a child with symptoms.

(Full Algorithm Shown)



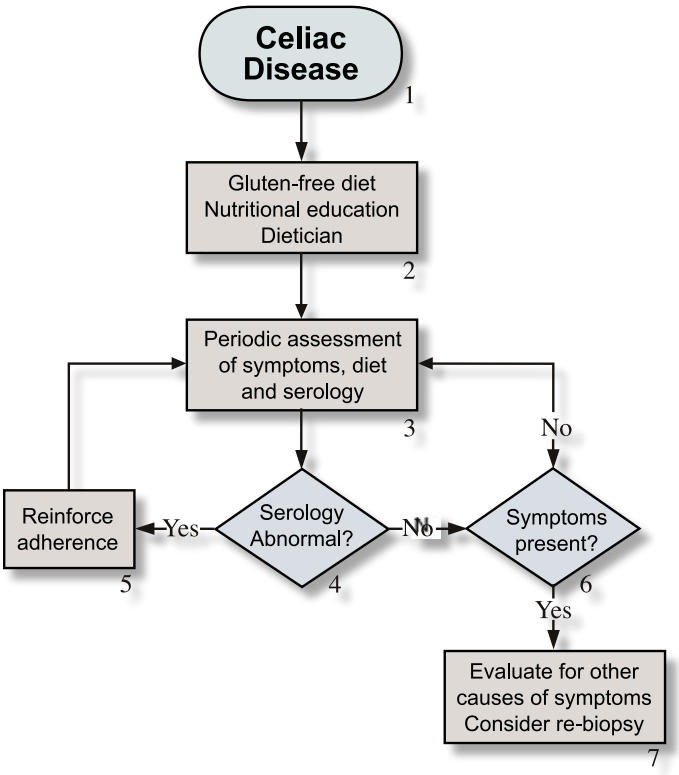
Algorithm for the evaluation of an asymptomatic child belonging to a group at-risk for celiac disease. *For patients with selective IgA deficiency measurement of TTG-IgG is recommended.

(Full Algorithm Shown)



Algorithm for the treatment and monitoring of a child with celiac disease.

(Full Algorithm Shown)



Children's Digestive Health and Nutrition Foundation

CDHNF President
George D. Ferry, MD

CDHNF Celiac Education Campaign Chair
Alessio Fasano, MD

NASPGHAN Celiac Disease Guideline Authors:
Ivor Hill, MD
Richard B. Colletti, MD
Martha Dirks, MD
Alessio Fasano, MD
Stefano Guandalini, MD
Edward Hoffenberg, MD
Karoly Horvath, MD
Gregory Liptak, MD
Joseph Murray, MD
Mitchell Pivor, MD
Ernest Seidman, MD

Guideline Summary Developed by:
Michelle Pietzak, MD
Richard Colletti, MD