Celiac Disease in the Pediatric Patient
What is celiac disease?
Celiac disease is a common chronic condition that affects about 1% of the general population. It occurs in genetically predisposed individuals and is triggered by the ingestion of products that contain wheat, barley or rye, collectively known as “gluten.” Ingestion of gluten initiates an inflammatory cascade in the small intestine that causes progressive destruction of the intestinal villi.

Symptoms of Celiac Disease
Symptoms are highly variable and may be gastrointestinal or non-gastrointestinal as shown in the table.

- Symptoms may occur singly or in combination.
- Age of onset can be any time from infancy to late in adulthood.
- Young children tend to have predominantly gastrointestinal symptoms while older children and adults are more likely to have non-gastrointestinal manifestations initially.

Because of the variable manifestations, a high index of suspicion for the disease and liberal use of screening tests is needed to avoid unnecessary delays in diagnosis.

<table>
<thead>
<tr>
<th>Gastrointestinal Manifestations</th>
<th>Non-gastrointestinal Manifestations</th>
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</thead>
<tbody>
<tr>
<td>• Diarrhea (chronic)</td>
<td>• Fatigue</td>
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<tr>
<td>• Abdominal pain/discomfort</td>
<td>• Unexplained weight loss</td>
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<tr>
<td>• Abdominal distension/bloating</td>
<td>• Anemia</td>
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<tr>
<td>• Excessive intestinal gas</td>
<td>• Short stature/growth failure</td>
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<tr>
<td>• Weight loss/poor weight gain</td>
<td>• Pubertal delay</td>
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<tr>
<td>• Constipation</td>
<td>• Dental enamel defects</td>
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<tr>
<td>• Chronic nausea/vomiting</td>
<td>• Skin rashes (dermatitis herpetiformis)</td>
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<td></td>
<td>• Elevated liver enzymes (unexplained)</td>
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<td></td>
<td>• Arthritis</td>
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Associated Conditions
Some people are at increased risk for celiac disease because of an associated condition. These individuals may have no symptoms or very minor complaints but when investigated are found to have the intestinal damage. Groups at increased risk include:

- First-degree relatives of an index case
- Type 1 diabetics
- Down syndrome
- Turner syndrome
- Selective IgA deficiency
- Other autoimmune diseases (autoimmune hepatitis, thyroiditis)
**Diagnosing Celiac Disease**

Blood tests looking for specific antibodies that are found when the disease is active can be used to screen for celiac disease. Recommended tests include:

- Tissue transglutaminase antibody (TTG)
- Endomysium antibody (EMA)
- Deamidated gliadin peptide antibodies (DGP)

The TTG-IgA antibody provides the most cost-effective and reliable test to screen for celiac disease. It can be combined with measurement of serum total IgA level to make sure the patient does not have selective IgA deficiency. Those with selective IgA deficiency should be screened using either the DGP-IgG or TTG-IgG antibody test. (See algorithm.)

In children less than 2 years of age, the TTG and EMA are less reliable, so testing in this age group should use a combination of the TTG-IgA and DGP-IgG.

Serological testing for celiac disease is only reliable if the child is eating a regular gluten-containing diet. **Do not start a gluten-free diet until the diagnosis of celiac disease is confirmed by a gastroenterologist.**

**Confirming the Diagnosis**

Serological tests are not sufficient to confirm diagnosis on their own. Confirmation requires a small intestinal biopsy sample to look for the characteristic histological changes that occur in celiac disease.

**Algorithm for Diagnosing Celiac Disease**

![Algorithm Diagram]

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* For children less than 2 years of age, request both TTG-IgA and DGP-IgG initially.
** If the clinical suspicion for celiac disease is high, refer to pediatric GI specialist even if the serological tests are negative.
Who should be tested for celiac disease?
In children with typical GI symptoms (diarrhea, weight loss, abdominal distension), testing for celiac disease antibodies should be one of the first tests ordered. For those with less typical symptoms, consider testing if no other cause for the symptoms can be identified. Testing can be considered in those at increased risk for celiac disease even if they are asymptomatic.

Can celiac disease be confused with other conditions?
Because the manifestations of celiac disease are so variable, it can be confused with many other conditions. Two conditions that cannot be differentiated from celiac disease on clinical grounds alone are:

- Wheat allergy
- Non-celiac gluten sensitivity

Non-celiac gluten sensitivity is defined as a condition causing symptoms that improve on removal of gluten from the diet in the absence of positive tests for wheat allergy or celiac disease. Hence it can only be considered as a possible diagnosis after both celiac disease and wheat allergy have first been excluded.

Differentiating celiac disease from these conditions is important as the dietary restrictions may not be as strict as they are for celiac disease and hence less burdensome to the family.

Treating Celiac Disease
Treatment entails adherence to a strict gluten-free diet for life. In most cases, this will result in complete symptom resolution and recovery from the intestinal damage.

Following a gluten-free diet is not easy. It has quality of life implications and increases the cost of food significantly. Therefore it is essential to confirm the diagnosis before prescribing this lifelong dietary change.

Because gluten is found in many processed foods, newly diagnosed patients should be referred to a nutritionist with special expertise in celiac disease for education on gluten avoidance and optimizing nutrition. It is also recommended to refer all patients to a celiac support group.

The Gluten-Free Gang, is a support group in Central Ohio for those with celiac disease or gluten intolerance and their families and friends. Learn more about this support group at GlutenFreeGang.org.

Celiac Disease Center at Nationwide Children's Hospital
The Celiac Disease Center at Nationwide Children's provides comprehensive care for the diagnosis and treatment of children with celiac disease. Learn more at NationwideChildrens.org/GI.