

## **Celiac disease: A comprehensive current review**

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### **ABSTRACT**

Celiac disease is an autoimmune disorder that's triggered when you eat gluten. It is also known as Celiac Sprue, nontropical sprue, or gluten sensitive enteropathy. Gluten is a protein in wheat, barley, rye, and other grains. It's what makes dough elastic and gives bread its chewy texture. When someone with celiac disease eats something with gluten, their body overreacts to the protein and damages their villi, small finger-like projections found along the wall of their small intestine. When villi are injured, small intestine cannot properly absorb nutrients from food. Eventually, this can lead to malnourishment, as well as loss of bone density, miscarriage, infertility or even neurological diseases or certain cancers. If celiac disease isn't better after at least a year without gluten, it's called refractory or nonresponsive celiac disease. Most people with celiac disease never know that they have it. Researchers think that as few as 20% of people with the disease get the right diagnosis. The damage to intestine is very slow, and symptoms are so varied that it can take years to get a diagnosis. Celiac disease isn't the same thing as gluten intolerance or gluten insensitivity. People with gluten intolerance may have some of the same symptoms and may want to avoid gluten. But they don't show an immune response or damage to the small intestine.

**Key Words :** Celiac disease, Autoimmune, tTG-IgA Test (Tissue transglutaminase IgA test), Villi, Malabsorption, Prevalence, Gluten intolerance, Gluten insensitivity

### **INTRODUCTION**

#### **What is celiac disease ?:**

Celiac disease is an autoimmune disorder that's triggered when you eat gluten. It is also known as Celiac Sprue, nontropical sprue, or gluten sensitive enteropathy. Gluten is a protein in wheat, barley, rye, and other grains. It's what makes dough elastic and gives bread its chewy texture. When someone with celiac disease eats something with gluten, their body overreacts to the protein and damages their villi, small finger-like projections found along the wall of their small intestine. When villi are injured, small intestine cannot properly absorb nutrients from food. Eventually, this can lead to malnourishment, as well as loss of bone density, miscarriage, infertility or even neurological diseases or certain cancers.

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don't show an immune response or damage to the small intestine.

### **Prevalence in India:**

In India, celiac disease is suspected to be more prevalent in the north Indian population where wheat is primarily grown and forms the staple cereal. These states would include Punjab, Haryana, Uttar Pradesh, Maharashtra, Rajasthan, Bihar, Uttaranchal, Madhya Pradesh and Gujarat.

So far, it was considered non-existent in south India but there is now evidence that it is being diagnosed in the native population of South India too though it is rarer than in the North.

Studies show that the prevalence of celiac disease has been doubling every 20 years. It is not clear why there has been an increase in the number of cases but the reason could be many- an overall increase in the consumption of wheat, usage of the new strain of wheat which is considered more antigenic, increased use of antibiotics etc.

The diagnosis rate of celiac disease though is extremely low presently, only about 5%. That would mean out of 100 people affected with celiac disease, 95 are not aware of this condition. This has therefore been popularly depicted in the form of an iceberg where the visible part i.e. the number of diagnosed cases is minuscule (3 – 5 %) whereas most of the cases are submerged, i.e., not yet diagnosed.

### **How does Celiac disease develop :**

Three factors have been identified to be prerequisites for celiac disease to develop: A gene, Consumption of gluten and a trigger.

1. Presence of a gene: 99 percent of celiac disease patients all over the world carry the gene identified as HLA DQ2 and / or HLA DQ8. Approximately 30 to 40 % of the population in India and the world carry one or both of these genes out of which about 1- 5 % are expected to develop celiac disease.

2. Consumption of Gluten: Gluten ( meaning glue in Latin) is a storage protein found in many grains. Gluten found in the three grains- wheat, barley and rye is antigenic and responsible for the development of celiac disease. About 5-10% of the celiac disease population also reacts to the storage protein found in oats in the same way as they do to gluten of wheat, barley and rye.

3. A Trigger: A trigger initiates the disease at a particular time but has not been ascertained so far. There are several that have been suspected though, including adenovirus infection, other viral infections, emotional and surgical trauma and many more.

All three factors have to be present for celiac disease to develop. The trigger can get activated at any age, from infancy to even 90 years.

### **Symptoms of Celiac disease:**

Celiac disease is a multisystem multi-organ disorder impacting people differently. There are more than 200 signs and symptoms which have so far been associated with celiac disease. Children more often exhibit the Classical symptoms ( mostly gastrointestinal) whereas in adults, the symptoms are mostly atypical or extra- intestinal. Non – bloody diarrhoea, weight loss and iron deficiency anaemia are the most common symptoms in adults. Onset of symptoms can happen at any age. In India, most of the diagnosed cases are adults. Thus, notion that this is a childhood disease, no longer holds true.

The understanding of symptoms is evolving with time, *i.e.* exhibit no symptoms but may still have high tTG values and even intestinal damage; celiac disease was never considered in an obese individual earlier but about 5- 20% of all patients with celiac disease are now reported to be obese.

The list of Symptoms of Celiac disease are :

Classical Symptoms : The Classical symptoms are gastrointestinal. These were so far considered as the typical presentation of celiac disease.

- Chronic diarrhoea ( non- bloody)
- Constipation (5% of patients)
- Recurrent diarrhoea
- Vomiting
- Foul- smelling stool
- Abdominal bloating/ Pain
- Failure to thrive
- Weight loss
- Recurrent intussusceptions

**Atypical symptoms:**

These are mostly extra- intestinal such as:

- Short Stature
- Iron deficiency anaemia
- Inability to concentrate
- Dermatitis Herpetiformis
- Mouth ulcers
- Dental enamel defects
- Chronic liver disease
- Osteopenia
- Osteoporosis
- Migraines
- Tingling or numbness in hands or feet
- Delayed puberty
- Infertility
- Frequent miscarriages
- Alopecia
- Arthritis
- And many more

Unlike what was believed earlier, about one – third of all new patients present with atypical symptoms today.

**People at risk:**

Some individuals are more susceptible to developing celiac disease than others.

1. Relatives of individuals with celiac disease- Since celiac disease is a genetic condition, first degree relatives, *i.e.*, parents, siblings and children of the patients with celiac disease have higher chances of developing it than others. Grandparents, cousins, uncles, aunts too are more susceptible to developing this condition than the general population, though not as high as the first degree relatives.

2. People with other autoimmune conditions- Autoimmune conditions tend to occur together. It has been seen that people with other autoimmune conditions like Type 1 diabetes mellitus, Thyroid disorders, Addison’s disease and Autoimmune hepatitis are more likely to develop celiac disease.

3. People with other genetic conditions- People with other genetic conditions like Down syndrome, Williams syndrome and Turner’s syndrome.

**How is Celiac disease Diagnosed?:**

The diagnosis of celiac disease is based upon the following:

1. Blood Tests

2. Biopsy
3. Confirmation by a positive response to gluten free diet.

**Celiac disease treatment:**

The only treatment for celiac disease presently is a strict gluten free diet for life. There is no medication needed except for supplements for nutrition deficiencies, which need to be taken for 6-12 months after diagnosis.

The intestine starts healing almost immediately after gluten is removed from the diet though it may take up to two years for it to heal completely. As the intestines start to recover, there is visible improvement in symptoms within 2-4 weeks, especially gastrointestinal symptoms.

tTG value too starts to reduce, though it may take 2 years to normalize. One must remember however, that a normal tTG value does not mean that the individual has been cured but only that the disease is under control with gluten free diet. These patients, their family and friends do deserve a pat on their back but does not give them a green signal to restart gluten!

Compliance to a strict gluten free diet is thus critical. Accidental or frequent ingestion of even small amounts of gluten can cause short term symptoms and long term damage. Read some of our patients' experiences with non-compliance to the diet. In rare instances, refractory celiac disease occurs where the intestine does not heal despite all precautions.

Follow-up visits are thus essential to evaluate the response to gluten free diet. Follow-ups are recommended after 2 weeks, 3 months, 6 months of diagnosis and thereafter annually. Refer to the gluten free health card for the tests which one must undergo at the time of follow-up and for keeping track of one's health.

Biopsy need not be repeated if the individual is doing well on gluten free diet. Only in certain cases – where the diagnosis was not clear, response is not as expected or the diagnosis had taken place before the age of 2 years – the doctor may consider repeating the biopsy.

**Gluten free diet:**

Gluten (meaning glue in Latin) is a storage protein found in several grains. Gluten found in the three grains – wheat, barley and rye is antigenic and responsible for the development of celiac disease. About 5-10% of the celiac disease population also reacts to the storage protein found in oats in the same way as they do to gluten of wheat, barley and rye. 20 parts per million, i.e. 20 mg of gluten per kg of food, is the generally accepted limit of gluten in 'gluten free' labelled food items in most parts of the world.

**Safe and unsafe items:**

'Which food items are safe and which are not?' is the most critical question for individuals with celiac disease and for their families. The good news is that 3 out of 4 basic food groups are completely safe for a gluten free diet. The bad news – that wheat finds its way in many food items, especially packaged products.

**Safe items:**

Except wheat, barley, rye, oats and their hybrid grains, all naturally occurring food items are inherently gluten free. This would make many food items which you are eating presently gluten free in their natural form. Some of these items are listed below.

**Grains** \*: Most of these alternate grains are easily available in India and had been used traditionally for many years but fallen out of favour over the last few decades.

- Rice (all varieties and colours)
- Millets including pearl millet (bajra), sorghum (jowar), finger millet (ragi)

- Amaranth (ramdaana /cholai / rajgira)
- Buckwheat (kuttu)
- Corn (makka)
- Fresh vegetables
- Fresh fruits
- Plain meat and fish
- Plain milk
- Daals/ Pulses \*
- Arrowroot, tapioca, sago(sabudana), soya, water chestnut (singhara) \*
- Plain nuts and seeds \*
- Egg
- Whole spices (sabut masala) \*
- Salt, Sugar
- Tea, Coffee
- Vegetable oil
- Honey

\*Flours and/or starches made out of these.

**Note:**

- Heeng, though inherently gluten free, could have gluten in commercial packaging in the form of wheat flour as an anticaking agent. Till we have information from the manufacturers, it is advisable to avoid using heeng.
- Many of these flours are commercially available in India but care needs to be taken as most are usually milled in factories where wheat is also milled. Hence the chance of cross contamination with wheat is extremely high in them. It is recommended that one grinds these flours at home using an electric grinder.
- It is also recommended that you grind spice/masala powders in your own grinder or in a shop where wheat is not used. Do also remember NOT to use the grinder meant for gluten free flours and spices for grinding any gluten containing item.
- Packaged and processed products could be safe if declared gluten free as per laid down international standards.

**Unsafe items**

Gluten can be present in a food product in many ways:

- **As an ingredient** – If it has wheat (or any of its forms such as maida, daliya, sooji), barley, rye or oats as one of the ingredients
- As the source of one of the ingredients, additives or as a processing aid.
- **Due to cross – contamination** - It is important to keep in mind that gluten protein is very resistant to alteration and no amount of cooking or method of cooking alters its toxic potential for a celiac.

**FAQs on Celiac disease:**

Can one be cured of Celiac disease? With the medical knowledge available presently, celiac disease cannot be cured. But the good news is that you can control this condition and all the symptoms related to it if you adhere to a strict gluten free diet.

Is it advisable for other members in the family to also stop eating gluten once one member is diagnosed with celiac disease? It would be advisable for other members of the family to get screened for celiac disease and only then stop eating gluten altogether, if needed. Please remember that if one chooses to switch to gluten free diet without diagnosis, then one will have to follow it strictly like an

individual who has celiac disease.

If the older child has celiac disease, can parents do anything to prevent it in their younger child? Preventing celiac disease is impossible as per our understanding of this condition today. Parents could though keep in mind the following:

- *Weaning tips* – Breast-feeding your baby for one year, introducing solids between 4 and 6 months of age under the cover of breast-feeding, introducing wheat between 6 to 7 months of age under the cover of breast feeding.

- Rotavirus vaccine for the younger child
- Ensuring to test the younger child at 2 years of age and an annual clinical assessment thereafter (or earlier if there are any symptoms).

Can I do anything during pregnancy to prevent celiac disease for my unborn child? Celiac disease is a genetic condition and the genes one inherits cannot be altered. But you can help your baby in terms of food allergy prevention and several other medical benefits by eating a wholesome diet, breast-feeding your baby for one year and introducing solids between 4 and 6 months of age under the cover of breast-feeding. Wheat should also be introduced under the cover of breast feeding, between 6 and 7 months of age. It appears that the duration of breast feeding is more critical in allergy prevention than just the exclusivity of breast feeding.

#### **FAQs about gluten free diet:**

Is gluten free diet inferior in any way to gluten containing diet? Gluten free diet does not lack any nutrients and is absolutely healthy provided a balanced diet is consumed. However, most experts would recommend micronutrient supplements like iron, calcium and vitamins at the time of diagnosis of celiac disease as most celiacs are found deficient in these at the time of diagnosis.

Is gluten free food expensive? No. All fresh and natural ingredients which can be used for preparing a gluten free meal are locally grown and easily available in India. Many of these would be used presently in the kitchen too. However, packaged gluten free food products are sold at a premium worldwide. It is advised that you learn how to prepare at least some of these at home. Learning to bake a bread, biscuits and cake at home would be helpful if you have a celiac child at home.

#### **Myths and Facts about Celiac disease:**

Celiac disease and gluten free diet are both relatively new terms in India. There are many false notions around these which baffle new patients and their families. Here are some facts to dispel some of these myths.

#### **Celiac disease is a childhood disease:**

**FACT:** Celiac disease can develop anytime between infancy and 90 years of age. Initially only children were being diagnosed with this condition but now more and more adults are being diagnosed all over the world including in India.

#### **Celiac disease is a “western” disease:**

**FACT:** The prevalence rate of 1% in India is the same as in other parts of the world. However, the diagnosis rate and awareness of the condition are extremely low here, hence the perception that it is a western disease.

Celiac disease is to be suspected only in case of gastro-intestinal problems.

**FACT:** Till few years back, almost all patients presented with gastrointestinal symptoms but today, about one third of children and most adults present with extra – intestinal symptoms. Celiac disease is now established as a multi-system disorder and hence should be suspected even if symptoms are not gastro-intestinal.

**About gluten free food:**

Only wheat needs to be avoided on a gluten free diet.

**FACT:** Apart from wheat, barley, rye, oats and products made out of them in any form need to be avoided too.

Gluten free food is not available in India.

**FACT:** India has many gluten free grains which are grown locally which are hence available in abundance and at reasonable prices – Sorghum (jowar), bajra, makki( maize flour), ragi, kuttu, cholai( Amaranth)etc. Daals( pulses), rice and vegetables form part of our everyday food. Hence, gluten free offers many flavors in India. In fact, world over, Indian cuisine is perceived as one of the most popular cuisines by the gluten intolerant community! Information about gluten in packaged products is however not available and hence the perception that gluten free food is difficult to obtain in India.

**Conclusion:**

Celiac disease is a unique autoimmune disorder in which the environmental precipitant, gluten, is known. Originally considered a rare malabsorption syndrome of childhood, celiac disease is now recognized as a common condition that may be diagnosed at any age and that affects many organ systems. This Review discusses the pathogenesis, diagnosis, and management of the disease.

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