Frequency of Common Clinical Presentations of Celiac Disease in Children

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Abstract:

Celiac disease is an immune mediated gluten sensitive enteropathy which occurs in genetic susceptible individuals. Clinically most of children present with gastrointestinal manifestations and extra intestinal manifestations. Its prevalence is 0.5 to 1% in general population. Various factors may be involved in its pathogenesis including both environmental and genetic factors. Gliadin is a fraction of gluten in wheat, rye and barley which is the inciting stimuli and plays an important role, that should be avoided strictly in diet and kept on lifelong gluten free diet after the disease has been diagnosed on the basis of small bowl biopsy or raised titer of anti-tissue transglutaminase levels. One hundred patients fulfilling inclusion criteria that are diagnosed cases of celiac disease on basis of raised Anti TGA or proven small bowel biopsy from 6 month to 14 year of age were included. The study was done in The Children's Hospital & The Institute of Child Health, Lahore. One hundred patients were included in this study with age range from 6 months to 14 years with a median age of 3 years. 46 were male patients and 54 were female. The most common clinical presentation in all cases of my study was diarrhea (81%), followed by failure to thrive (79%), edema (36%), clubbing (35%).It is concluded that most common clinical manifestation of celiac disease is diarrhea in our population followed by failure to thrive, edema and clubbing.

Key words: Celiac disease, Frequency, Children, age, male and female

Introduction

Celiac disease is an immune mediated enteropathy that occurs in genetic susceptible individuals [1], and is triggered by a well-identified dietary factors and the auto antigen. The disease primarily affects the small intestine, where it progressively leads to flattening of the small intestinal mucosa. Based on a number of studies from Europe and the United States, it has been estimated that the prevalence of celiac disease in children between 2.5 and 15 yr in the general population ranges from 3 to 13/1,000 children or ≈1/300 to 1/80 children.[1] Three cereals contain gluten and are toxic for patients with celiac disease: wheat, rye, and barley. Celiac disease can occur at any stage in life; Classic Gastrointestinal pediatric cases usually appear in children aged 9-18 months. The variation in the symptom onset possibly depends on the amount of gluten in the diet and other environmental factors, such as duration of breast feeding. In fact if gluten is introduced during breast feeding, the symptoms tend to be less often GI related and tend to appear later in life.[2] Evidence suggests that celiac disease is under diagnosed in children, Serologic testing has the potential to detect otherwise undiagnosed disease. Evidence is also accumulating that daily ingestion of wheat, rye, and barley results in long-term extra intestinal sequelae in subjects with undiagnosed or untreated celiac disease.[3,4] Early detection of the disease and subsequent dietary elimination of gluten might be the appropriate method for reducing complications later in life.

The common presenting features, especially in toddlers with malabsorption are diarrhea, abdominal distention, and failure to gain weight, with a fall in growth chart percentiles. Physical findings include the loss of the subcutaneous fat, wasting of muscles and the loose skin folds. The nutritional

effects of malabsorption are more pronounced in toddlers, because of the limited energy reserves and higher proportion of calorie intake being used for weight gain and linear growth. In older children, malnutrition may result in growth retardation, as commonly seen in children with late diagnosis of celiac disease. If untreated, linear growth slows and with prolonged malnutrition death may follow due to its complications. The assessment of nutrition is an important part of clinical evaluation in children with malabsorptive disorders, such as Long-term calcium and vitamin D malabsorption can lead to reduced bone mineral density with increased risk of bone fractures and Vitamin K malabsorption can result in coagulopathy. Severe protein-losing enteropathy is often associated with malabsorption syndromes (celiac disease, intestinal lymphangiectasia) and causes hypoalbuminemia and edema. Other nutrient deficiencies include iron malabsorption causing microcytic anemia and low reticulocyte count, low serum folate levels in conditions associated with mucosal atrophy, and low serum vitamin A and vitamin E concentration in fat malabsorption.[1] CD has been considered a disease of childhood, manifesting during the first years of life as a gastrointestinal syndrome that consists of diarrhea, abdominal distention and failure to thrive. This concept has, however been challenged in recent years, as it became increasingly evident that, first CD can affect individuals of any age, and second patients may present with symptoms that once were considered highly unusual for the diagnosis of CD.[5] Among children, these non-classical forms include atypical GI findings such as abdominal pain, vomiting or constipation as well as a number of extraintestinal problems like iron deficiency, altered bone metabolism, short stature and unexplained elevation of transaminases[6,7]. Because of variability in presentation we aimed to study the presentation of celiac disease in our population.

Material and Methods

This study was conducted at the Department of Gastroenterology in the Children's Hospital and the Institute of Child Health .It is a tertiary care hospital for all sorts of patients including those with GI disorders. This study was conducted over 6 months and comprised of 100 patients per selection criteria. Only admitted patients of celiac disease in gastroenterology Department fulfilling inclusion criteria were included in the study. Patients were stabilized initially, After informed and written consent from parents, biodata of patients including name, age, sex, address were noted, detailed history of clinical presentations including failure to thrive, diarrhea and physical examination for clubbing and edema was performed with their due respect and data was recorded on a Proforma.

Data Analysis Procedure

All the taken Data was entered in SPSS version 11. The variables analyzed were included Mean and Standard Deviation of age calculated, Male and female ratio was calculated. Frequencies and percentages of qualitative variables like gender, failure to thrive, loose motion, edema and clubbing calculated and presented in tabulated form.

Results

One hundred patients were included in this study, out of 100 patients 46 cases were male and 54 were female. Male to female ratio was 1:1.2.

The percent disease frequency was more in patient age group of 06 months to 04 years (65%), in 4-7 years age group was (31%) and 8-14 years age group (4%) was investigated. Further results showed that disease present were found more

in small age grouped and as the age is increased the disease was decreased (Table 01).

The most common clinical presentation in all cases in this study was diarrhea (75.9%), followed by failure to thrive (74.0%), edema (42.6%), clubbing (27.8%) in female patients. Similarly in male patients the most common clinical presentation in all cases in this study was diarrhea (87.0%), followed by failure to thrive (85.0%), edema (32.6%), clubbing (43.4%). In female and males, the frequencies and percentages of clinical manifestations has been given in table 2 and 3 respectively.
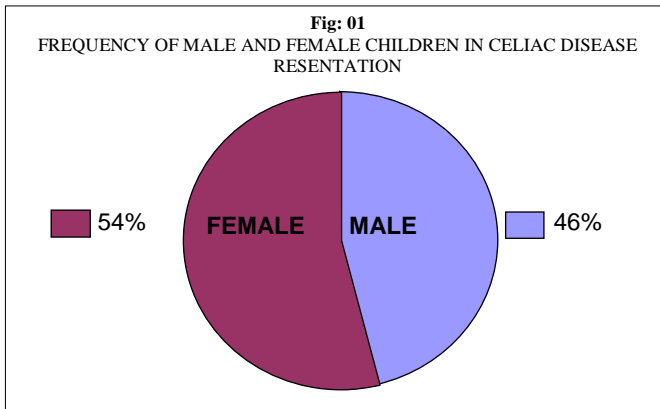

Fig: 01
FREQUENCY OF MALE AND FEMALE CHILDREN IN CELIAC DISEASE RESENTATION

Table 1: Age Distribution (n*=100)

Age in Years	Frequency	Percentage
6 month - <4 years	65	65%
4 year – 7 years	31	31%
8 year -14 years	04	4%
Mean ± Standard Deviation	3.44 +1.91	

*n=Total no. of patients.

Table No.2 Frequency of Common Clinical Manifestations of Celiac Disease in Female Patients. *(n=54)

S.NO.	CLINICAL MANIFESTATIONS	+FREQUENCY	PERCENTAGE
1.	Diarrhea	41	75.9%

2.	Failure to thrive	40	74.0%
3.	Edema	23	42.6%
4.	Clubbing	15	27.8%
Mean and SD of age. 3.07 ±1.52 SD			

*n = total no. of female children with celiac disease.

+   =   frequency of positive cases presented with single or combined manifestations.

SD =   Standard Deviation

Table No.3 Common Clinical Manifestations of Celiac Disease in Male Patients. *(n=46)

SNO.	CLINICAL MANIFESTATIONS	+FREQUENCY	PERCENTAGE
1.	Diarrhea	40	87%
2.	Failure to thrive	39	85%
3.	Edema	15	32.6%
4.	Clubbing	20	43.4%
Mean and SD of age. 5.4 ±1.67 SD			

*n = total no. of male children with celiac disease.

+   =   frequency of positive cases presented with single or combined Manifestations.

SD =   Standard Deviation.

Discussion:

Celiac disease is a chronic enteropathy caused by intolerance to gluten. Population-based studies, using serologic screening, have indicated that the prevalence of celiac disease in Caucasian populations is 0.5%–1%. Many studies have been conducted on clinical manifestations of celiac disease in different countries. According to Kliegmann and Hal B. Jenson Failure to thrive was the most commonest clinical presentation 85.7%, followed by diarrhea 71.3%, [8] while according to Kuloglu Z, kansu A, Ensari A, diarrhea was the most common clinical presentation in children of turkey,[9] and according to Poddar U, Thapa BR, Singh K in India majority of patients presented with diarrhea 84%.[10] In this study in our population the diarrhea was the most common clinical manifestation (81%),so our study goes in favour of study by Ensari A and kansu A in turkey children, and Singh K in India. According to

Kuloglu Z and Ensari on clinical features of 109 patients of celiac disease, the failure to thrive was the second most common clinical manifestation.[9] while according to Poddar U, and Singh K on clinical features of celiac disease in India on children The majority (84%) presented with diarrhea other features were failure to thrive in 91%, anemia in 84%, wasting in 87%, and stunting in 60% of cases. [10] According to Kliegmann, and Hal B. Jenson, failure to thrive is commonest presentation (85.7 %), [8] while in my study the failure to thrive was the second common clinical presentation (79%) among all patients, my study favours the kuloglu Z, Ensari in Turkish children, and Poddar U and Singh K in Indian children. According to Kliegmann, and Hal B. Jenson, the less common clinical manifestations were edema 33.3%, and clubbing 26%.[8] while in our study edema (36%) and clubbing (35%) were the less common manifestations , so these clinical features are in favour with study by Kliegmann and hall B.

Although the most common manifestation of children with celiac disease was the diarrhea in my study, the possible reasons for this manifestation could be poverty, poor adherence to gluten free diet, poor follow up visits, other infectious causes, though these causes should be sort out and considered in differentials.

This study concluded that common clinical presentations of celiac disease which should not be overlooked by physicians and warrants for further work up for to rule out celiac disease in patients with chronic diarrhea with typical history , failure to thrive, edema and clubbing. Screening for celiac disease has been recommended for those who are at more risk such as patient with history of being a first degree relative, diabetes mellitus, autoimmune infection such as thyroiditis, and those with suspected or diagnosed case of Down syndrome, turner syndrome, and William syndrome. Anti-endomysium IgA antibody and anti-tissue transglutaminase IgA antibody tests are highly sensitive and specific in identifying individuals with

celiac disease. Although now anti tissue transglutaminase (TGA) has replaced anti- endomysium antibodies due to its simplicity. The negative test result doesn't exclude the diagnosis of celiac disease because some patients with IgA deficiency can give the false positive test so serum IgA level must be monitored before labeling. The definitive diagnosis is off course the small intestine biopsy, as none of the available serologic tests are 100% reliable. Biopsy pieces are taken from multiple areas because of possible patchy involvement of intestinal mucosa. The characteristic histopathological changes include partial or total villous atrophy, increased number of intraepithelial lymphocytes, crypt elongation and decreased villous/crypt ratio, intraepithelial lymphocyte mitotic index >0.2%, decreased height of epithelial cells.

The only treatment for celiac disease is lifelong exclusion of gluten. This requires a wheat, barley, and rye-free diet, and treatment of micro/macronutrients deficiencies with nutritional rehabilitation.

Conclusion:

This study demonstrated the common clinical presentations of celiac disease in pediatric population in patient with history of chronic diarrhea, failure to thrive, edema and clubbing.
This study concluded that:
1. Most common clinical manifestation is diarrhea in our population followed by failure to thrive, edema and clubbing, while in western countries common presentation Failure to thrive is the most common followed by diarrhea, edema and clubbing.
2. Female children were affected more than male children, while in different studies and literature view there is slight predominant female celiac patient with slight difference.

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