



Original Research Article

Review of clinical presentation of children with celiac disease - A retrospective study

Sunil Pathak¹, Avinash Bothra¹, Manish Rasanian¹, Dipika Bhil^{1,*}, Nimya Mary¹, Prerna Dogra¹¹Dept. of Pediatrics, Smt. B.K. Shah Medical Institute & Research Centre, Vadodara, Gujarat, India

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ABSTRACT

Introduction: Celiac disease is a chronic immune mediated enteropathy, produced by the gluten ingestion in susceptible people, where gluten act as environmental trigger. Celiac disease has global prevalence of 1%. Its presentation ranges from simple anaemia, gastrointestinal manifestation, short stature to neurological manifestations. The high level of suspicion helps in prevention of chronic complications of the disease.

Objective: To identify diverse clinical manifestations of children with celiac disease.

Materials and Methods: Children with previously established or newly diagnosed celiac disease, during (2011-2019) were recruited. Data was collected retrospectively from medical records and clinical notes, and subsequently analysed.

Result: The mean age was 7.90 years (3-15 years). 59.09% (13) patients were females. 90.90% (20) patients had gastrointestinal symptoms and anemia and 81.81% (18) had short stature. Other symptoms include lower-limb edema (1), seizure (1), dental caries (1), anasarca (1), abdominal Koch (1), rashes (1), lower limb weakness (1). Biopsy was positive in 7/8 patient.

Conclusion: Knowledge of diverse clinical presentations will help in increased clinician's awareness and high level of suspicion. It will enable an early diagnosis and management.

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1. Introduction

Celiac disease (CD) is a chronic small intestine immune-mediated disorder elicited by the ingestion of gluten in genetically predisposed individuals. The disease is associated with human leukocyte antigen (HLA) DQ2 and DQ8 haplotypes.¹ In the continued presence of gluten, CD is self-perpetuating. Given the proven role of gluten in causing inflammation and autoimmunity, CD represents a unique example of immune mediated disease for which early diagnosis and dietary treatment can prevent severe, sometimes life threatening complications.^{2,3} The universal screening is not currently being advised. However,

awareness should be raised and there should be a low threshold for investigating both symptomatic children and those with associated conditions, as it is known that approximately 90% of cases remain undiagnosed.^{1,4} The CD is commonly detected in patients with diarrhoea, but the clinical manifestations of CD are numerous and vary from none (asymptomatic) to a broad spectrum of gastrointestinal symptoms and extra-intestinal manifestations.¹ These different modes of presentation, lead experts to elaborate the Oslo Classification,⁵ which subdivides CD into symptomatic CD, includes the "classical" and "non-classical" presentations, versus the "asymptomatic" or "subclinical" CD. The classic CD presents with signs and symptoms of malabsorption and the non-classic CD is characterized by other gastrointestinal symptoms apart from

* Corresponding author.

E-mail address: dipika7299@gmail.com (D. Bhil).

diarrhoea and extra-intestinal symptoms. The subclinical CD is below the threshold of clinical detection. There is also the “potential CD” describing the patients at risk of developing the disease in the future.

This study was conducted retrospectively to examine the clinical presentation of celiac disease in paediatric patients, admitted to a tertiary medical centre in the years 2011-2019. Our main goal was to identify the symptoms and clinical phenotype of these patients at presentation.

2. Materials and Methods

This retrospective observational study assessed all the paediatric patients diagnosed as celiac disease, presented to Dhiraj hospital from 2011 to 2019. Data from each selected patient was obtained retrospectively from medical records of paediatric gastroenterology clinic and were scanned for patients with celiac disease in the span of last 9 years. Total 22 patients record were found. Their demographic characteristics, symptoms, associated diseases, type of clinical presentation, family history, serology tests requested at presentation and their results, histopathology findings if done, diet pattern, the clinical and growth response after treatment were recorded and put in excel sheet for further analysis.

All the patients whose diagnosis was recorded as celiac disease were included in the study (based on serology tTG-IgA alone or serology plus histopathology). Histological findings in small intestine biopsies, taken from the bulb or the second duodenal portion, were classified according to the Modified Marsh Classification by Oberhuber.⁶

2.1. Statistical method

Categorical data were presented as percentages and quantitative data were presented using average and range value.

3. Result

Total 22 patients with celiac disease were isolated from records of paediatric gastroenterology clinic in the span of last 9 years (2011-2019). The mean age of the group on presentation was 7.9 (3 to 15 yrs.) yrs. Maximum patients presented between the age of 7-10 years. 13 (59.09%) patients were females while 9 (40.90%) were males. Majority of the patients, 18 (81.91%) were from the state of MP, while rest 4 (18.19%) patients were from Gujarat.

Figure 1 shows that majority, 20 (90.90%) of the patients presented with GI symptoms. Even the GI symptoms were diverse. 9 (40.90%) patients presented with the complaint of generalized weakness. 7 (31.81%) patients presented with the complaints of failure to gain height and weight. Other spectrum of presentation observed in the group were seizures, dental caries, generalized swelling, skin rash and

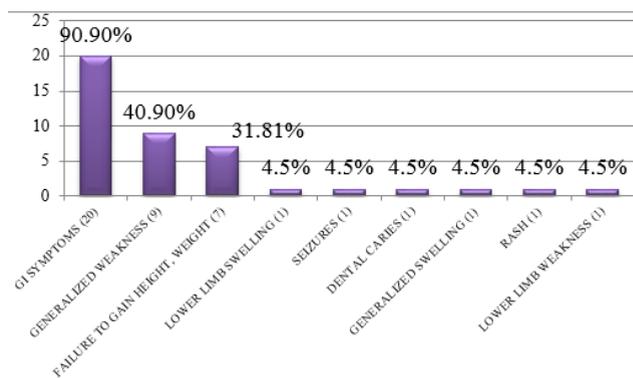


Fig. 1: Symptoms and clinical profile of children with celiac disease

lower limb weakness in 1(4.5%) each of patients.

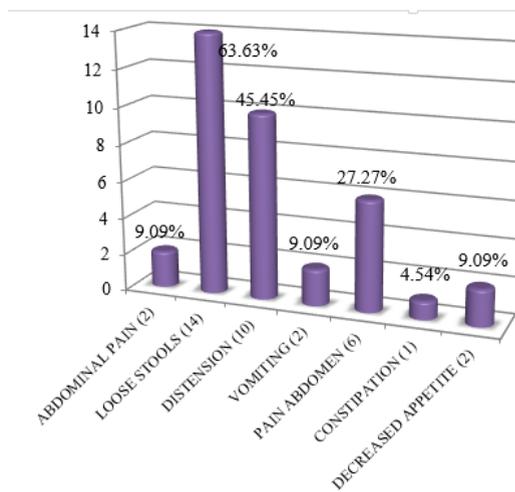


Fig. 2: Gastro-intestinal symptoms of children with celiac disease.

Among gastro intestinal complaints majority 14 (63.63%) patients had loose stools as presenting complaint. Abdominal distension was seen in 10 (45.54%) patients. 6 (27.27%) had history of pain abdomen on presentation and 2 (9.09%) patients presented with abdominal pain, vomiting and decreased appetite each. 1 patient had constipation as presenting complaint (Figure 2).

Table 1: General physical finding of children with celiac disease.

S. No.	Clinical feature	N(%)
1.	Pallor	18(81.81%)
2.	Short stature	18(81.81%)
3.	Anasarca	1(4.5%)
4.	Clubbing	1(4.5%)
5.	Lymphadenopathy	1(4.5%)

On examination, pallor was seen in 18(81.81%) of the patients. 18(81.81%) had short stature. Anasarca, clubbing

and lymphadenopathy was found in one patient each.

Table 2: Laboratory investigation of children with celiac disease.

Laboratory investigation	Patients (n=22)
Serum tTG- IgA (positive)	22
Duodenal biopsy positive for celiac disease	7

Upper GI Endoscopy and Duodenal Biopsy was done in only 8/22 patients. It was positive for Celiac disease in 7/8 patients while 1 was indeterminate. Mean Hemoglobin of the patients on presentation came out to be 8.75 gm/dl (range 2.9 to 12.5 gm/dl). 20 (90.90%) patients had hemoglobin level less than 11gm/dl. 1(4.5%) patient had severe anemia, 12 had moderate and 7 patients had mild anemia at presentation.

Out of 22 patients only 8 patients were followed up for 3 months, 6 patients till 6 months, 3 patients till 9 months and 2 patients till 12 months post diagnosis. Average weight gain at 3, 6, 9 and 12 months were 935 gm, 3953 gm, 3980 gm and 4790 gm respectively. Similarly, height gain at 6 months and 12 months were 3.75 cm and 8.3 cm respectively. We could not retrieve data longer than one year follow up in any of the patients.

4. Discussion

The children included in this retrospective study presented with a wide variety of clinical features. We found an increased prevalence of coeliac disease amongst male compared to females. In our study, we found that majority of patients were from the state of MP, this could be attributed to the fact that MP is primarily a wheat eating state. Study by Yacha et al reported that prevalence of celiac disease is eight times less in Gujarat as compared to Punjab. He attributed this to genetic difference in two population.⁷ But it may also be due to dietary pattern of two population, as staple diet of people in Punjab, Haryana, Rajasthan, Madhya Pradesh and western Uttar Pradesh is mainly wheat based while in Gujarat it is mixed wheat and rice.

Considering that the variable clinical picture of coeliac disease is related to genetic and immunological base, the age of onset may influence the clinical presentation. In our study, mean age at diagnosis was 7.9 years (3-15 years). In a study by Yacha et al., he reported mean age at diagnosis between 6.3- 8.6 years (2.5- 14 years).⁸ We found that maximum number of patients presented between the ages of 7-10 years.

Similar to study by Bharadiya et al. Diarrhoea (63%), Abdominal distension (45%) and abdominal pain (27%) were the most common gastrointestinal symptoms at presentation and short stature was the extra-intestinal finding.⁸ Study by Gracinda et al, found Abdominal pain and diarrhoea as major presenting symptoms.¹ While Study by Bhattacharya M et al, found Abdominal pain and

abdominal distension as main gastro-intestinal symptoms and short stature as extra-intestinal finding at presentation.⁷

All the patients were investigated for complete blood count at the time on presentation and anaemia was found in 49% of total. Prevalence of Anemia among patients with celiac disease reportedly varied from 23% to 90% in different studies.^{1,7,8}

Serological markers provide simple yet non-invasive initial screening test especially for the individual who presents with atypical clinical manifestation. In our study, the diagnosis was based mainly on clinical suspicion and confirmation by Serum tTG- IgA. Although, Small bowel biopsy remains the gold standard for the diagnosis of coeliac disease. In our study, What-ever biopsy was done, it was done in other hospital due to non-availability of gastro-physician, that sometimes become too costly affair to our patients. Biopsy was done in only 8 out of 22 patients. In 7 out of 8 patients biopsy findings were consistent with celiac disease, while in one patient finding was indeterminate.

Most of the patients in our study were from far distance (Madhya Pradesh). Getting long term follow up is a difficult for poor parents. That is one of the reasons of lost to long term follow in current study. Availability of at least good counselling centre nearby, where patients can take regular counselling can keep them motivated and aware for importance of gluten free diet. Patients can be monitored there for growth and development, and adherence to gluten free diet.

5. Conclusion

This study outlines the diverse clinical presentations of paediatric celiac disease. A high level of suspicion and Increased clinician's awareness will enable an early diagnosis and treatment, with subsequent improvement in symptoms and nutritional status.

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7. Conflicts of Interest

No conflicts of interest.

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Avinash Bothra, Assistant Professor

Manish Rasanias, Professor

Dipika Bhil, Assistant Professor

Nimya Mary, Resident

Prerna Dogra, Resident

Author biography

Sunil Pathak, Associate Professor

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