Clinical Profile of Children with Celiac Disease in Gujarat

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Abstract

Introduction : Celiac Disease (CD), an autoimmune enteropathy, triggered by the ingestion of gluten in genetically susceptible individuals, is one of the commonest causes of malabsorption in the west. It is now well documented from north India where wheat is the staple diet. We report here 22 children of CD from Gujarat to bring the awareness amongst the pediatricians for its early diagnosis. The clinical presentation, serological tests & duodenal biopsy confirms the diagnosis. The results of Gluten Free Diet (GFD) are quite gratifying. **Methodology :** Retrospective analysis of presentation of children diagnosed to have CD was done from maintained database of CD patients of last 5 years. **Results :** Twenty two children were diagnosed to have CD at our centre in last 5 years. The age of presentation was from 14 months to 11 years. Short stature, pallor & chronic diarrhoea were the commonest features. Distention of abdomen, anorexia, pain in abdomen & oedema were other manifestations. Vomiting, voracious appetite, irritability & dermatitis were also noted in some cases. Rickets, rectal prolapse & clubbing were less common findings. Serological tests, besides routine investigations & duodenal biopsy confirmed the diagnosis. Gluten Free Diet (GFD) showed impressive results in 3 to 6 months time. **Conclusion :** CD is well documented in north India, but it also exists in Gujarat. With clinical presentation of stunted growth, chronic diarrhoea & unexplainable anaemia, one should think of CD. Some other less common & atypical features should also be kept in mind. The results of serological tests for CD are fairly reliable. Still, it is mandatory to confirm the diagnosis by duodenal biopsy. The results of GFD are quite rewarding. To emphasize for compliance of GFD & to provide the list of GFD to the parent & regular follow up are essential components of management.

Key Words: Celiac Disease, Chronic diarrhoea, Stunted growth

Introduction :

Celiac Disease (CD), an autoimmune enteropathy, triggered by the ingestion of gluten in genetically susceptible individuals, is one of the commonest causes of malabsorption in the west. It was believed to be rare in rest of the world. Since 1960, several Indian adults & children suffering from CD have, however, been reported. [1-3] It is now well documented from north India where wheat is the staple diet. [4-7]. Recently, CD has been reported also from south India. [8] However, there is no reporting of CD from Gujarat. We are reporting 22 cases of confirmed CD with characteristic features & good response to GFD. This is to emphasize that CD is existent in Gujarat.

There are three known components that play role in pathogenesis of CD. They are genetic back ground, environmental factor (gluten) & the immune system. CD represents a unique model of autoimmune disorder in which the triggering environmental factor (gluten) is known. If an individual has the genetic background & ingests gluten, the immune system is activated & as a result of immune reactions, the disease is set in with clinical manifestations. It appears that people without the known HLA genes do not have CD. Also, people having the known HLA haplotypes, but living in areas where gluten is not the part of their diet, do not have the disease. However, the significant world wide genetic mixture & the international distribution of gluten containing food spread the two important pathogenic factors throughout the world, therefore, CD should be considered everywhere. [9] Chronic diaarhoea, distention of abdomen, pain in abdomen, weight loss, oedema, anaemia, malabsorption syndrome, dermatitis & stunted growth are common presentations, either alone or in combination. Some of them have atypical or extraintestinal manifestations like rickets, dental enamel defects, aphthous stomatitis, refractory iron deficiency anaemia, dementia, ataxia, delayed puberty, infertility etc. The association of CD with other autoimmune disorders like diabetes mellitus & thyroiditis is known. Its association with Down's syndrome has also been reported. The clinical symptoms, serological testing, intestinal biopsy & response to GFD are important components for diagnosis of CD. GFD for the lifetime is quite gratifying. One should be aware regarding GFD (not containing wheat, oat or rye) for proper dietary advice to the patient.

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Material & Methods

We maintain an ongoing database of CD patients. Retrospective analysis of mode of presentation of children diagnosed to have CD was done. Patients were labeled as having CD as per the standard ESPGHAN criteria. Detailed history & physical examination with

special reference to anthropometric assessment was performed in all children at the time of diagnosis. With clinical diagnosis of CD, serological tests were performed besides routine investigations. The serological tests include Antigliadin antibody Ig G & Ig A (AGA), Endomysial antibody Ig A(EMA), Tissue Transglutaminase (TTG) & in some cases Antireticulin antibodies(ARA). The small intestinal biopsy was performed in all cases for histological study. Children diagnosed as CD were put on Gluten free diet (GFD). The parents were given the list of GFD. They were explained in details regarding the disease, importance of compliance of diet & prognosis. In the follow up, these children were assessed for details of diet (to ascertain the compliance & quality of food intake) & their clinical profile, especially anthropometry. Anthropometry (weight, height, mid arm circumference) was recorded by standard technique. Informed consent of all the subjects was obtained before collecting samples. The intestinal biopsy was performed by a pediatric gastroenterologist by a pediatric fiber optic endoscope after written consent & appropriate sedation. The biopsy material was examined by an expert histopathologist & graded according to standard score. All of them were investigated for diabetes mellitus & hypothyroidism.

Results

Twenty two children (14 males, 8 females) were diagnosed to have CD at our center as per the ESPGHAN criteria. [10] The youngest child was 14 months old & eldest one 11 years old. The mean age of presentation was 6.87 years.

The clinical profile of the cases at the time of presentation is depicted in Table I. Short stature, pallor & chronic diarrhoea were the commonest features in our patients. Distention of abdomen, anorexia, pain in abdomen & oedema were other common manifestations. Vomiting, voracious appetite, irritability & dermatitis were also noted in some cases. Rickets, rectal prolapse & clubbing were less common findings. Two patients presented with constipation, although chronic diarrhoea was the most common feature in our patients.

Table 1 : Clinical Features of Children with celiac Disease (n = 22)

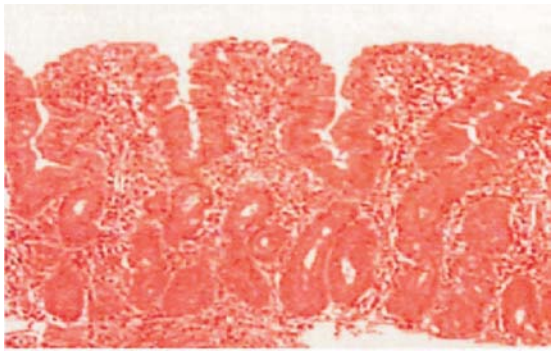
Clinical Feature	Number	Percentage (%)
Chronic diarrhoea	20	(90.9)
Abdominal distention	16	(72.7)
Pain in abdomen	10	(45.4)
Stutned Growth	22	(100)
Pallor (anaemia)	22	(100)
Anorexia	14	(63.6)
Voracious appetite	4	(18.1)
Vomiting	8	(36.3)
Oedema	12	(54.5)
Irritability	7	(31.8)
Constipation	4	(18.2)
Rectal prolapse	2	(9)
Rickets	4	(18.2)
Dermatitis	6	(27.2)
Clubbing	4	(18.2)

Routine investigations showed anaemia in almost every child. Thrombocytosis was noted in few patients. It may be due to hyposplenism which is known characteristic of CD. Hypoalbuminemia was noted in patients with oedema. Amongst the serological tests for CD, AGA & TTG were positive in all the patients. EMA was positive in more than two third patients. ARA was performed in 6 patients & was found positive in two patients. Duodenal biopsy showed the changes of villous atrophy in all patients. Twenty patients showed subtotal villous atrophy & two moderate villous atrophy. The investigations for diabetes mellitus & hypothyroidism were negative in all the patients. Still, regular follow up investigations for these two conditions are performed.

Table 2: Serological & histopathological data of children with Celiac Disease (n = 22)

Investigation		Number	Percentage (%)
Anti Gliadin Antibodies (AGA)		22	(100)
Endomysial Antibodies (EMA)		16	(72.7)
Tissue Transglutaminase antibodies (TTG)		22	(100)
Anti Reticulin Antibodies (ARA)		2	(33.3) (Performed in 6 patients)
Duodenal Biopsy	Subtotal villous atrophy	20	(90.9)
	Moderate villous atrophy	02	(9)

Figure I: Histopathalogical changes in duodenal biopsy (subtotal atrophy)



Institution of GFD showed impressive results on follow up. There was dramatic improvement in clinical picture, anthropometric & hematological parameters. Most of the patients showed recovery within 3 to 6 months, some of them were late.

Discussion

Although CD was once thought to be rare in India, several recent reports have cleared this misconception. [6][11] However, there is not a single documented report of CD from Gujarat. We are reporting 22 cases from our centre. Diarrhoea, vomiting & failure to thrive are reported to be the most common presenting symptoms in children with CD. Main difference in the clinical features of CD as highlighted by the present study as compared to west are chronic diarrhoea, stunted growth & anaemia. These manifestations are features of more severe disease & correlated with later age of diagnosis. Distention of abdomen, pain in abdomen, anorexia, oedema & irritability are commonly considered as manifestations of malnutrition at our set up. Some of them do not show recovery in spite of proper diet, at this juncture one should think of CD. Iron deficiency anaemia which is not responding to iron therapy for more than 3 months is the crucial point to consider CD. Short stature may be the sole presentation of CD. Atypical presentations of CD like constipation, rickets & prolapse of rectum should be kept in mind.

The widespread availability of highly sensitive & specific serological tests has resulted in gratifying results. Still, endoscopy & duodenal biopsy is mandatory to confirm the histopathological diagnosis of CD. [10][11]

Several reports from our country indicate non compliance or doubtful compliance after diagnosis of CD, resulting in poor response to therapy. [5] We have found that after diagnosis of CD, it is imperative to have the counseling of the parents, the patient & probably the whole family. It should be explained to them in details regarding condition, importance of compliance of GFD for life time. It should be emphasized that a small liberty for a while can also result in failure of therapy. They should be provided written list of food items which can be consumed by the child. When they are not sure regarding absence of gluten in the food, it should not be consumed. Regular follow up & repeated counseling are corner stones of the successful management.

Clinical, anthopometic & hematological response is impressive after institution of GFD within 3 to 6 months period. Atypical presentations may take longer time to respond. [6]

Conclusion

Our study suggests that CD is a common entity in countries like India especially in the northern parts where wheat is the staple diet. CD is also existing in Gujarat, provided the clinicians are aware & carry out proper work up in a suggesting case scenario. The serological tests have good sensitivity & specificity, still endoscopy & histopathological examination of small intestinal biopsy is mandatory. GFD leads to dramatic improvement in clinical, anthropometric & hematological status. Regular follow up & counseling is recommended to ensure dietary compliance.

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