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# Case Report: Celiac disease with celiac crisis.

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## Abstract

**Celiac crisis is a life threatening and very rare complication of Celiac disease. Clinically, it is characterized by severe diarrhoea, dehydration, abdominal distension and metabolic disturbances like hypokalaemia, hypomagnesaemia, hypocalcaemia, hypoproteinemia and shock. A 5-year-old boy with chronic diarrhoea, weight loss, abdominal distension, pedal oedema, severe undernutrition and vomiting was presented to paediatric unit. He was diagnosed with celiac disease presenting with celiac crisis. Child was treated with short course corticosteroid therapy and rapidly recovered over a period of 5 days.**

**Keywords:** Celiac disease, Celiac crisis, Corticosteroid

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## Introduction

Celiac disease (CD) is a disease of the small intestine caused by an immune response to ingested gluten. This response results in characteristic damage to the villi, leading to malabsorption [1]. CD is manifested by a variety of clinical signs and symptoms that may begin in either childhood or adult life. Some individuals are completely asymptomatic [2]. Celiac crisis (CC) is a rare and life threatening complication of celiac disease (CD).

## Case

A 5-years-old-boy was brought to our hospital with complaints of weight loss, vomiting, weaknessand chronic diarrhoea, abdominal distension and pedal oedema.

From the history, it was learnt that similar complaints had begun from five months of his life, when supplementary nutrition was started. On physical examination, body weight and height were 12.0 kg (-3 SD), 99 cm (-3 SD) respectively. Child was severely undernourished (WHO Criteria). Vital signs: heart rate-126/min, respiratory rate-42/min, afebrile, CRT-6sec, blood pressure -74/40 mm Hg. The subcutaneous fat tissue was markedly decreased. Pink-brown coloured desquamates macular rash were present on abdomen and back. Abdominal distension was observed.Child was severely dehydrated in metabolic acidosis (pH 7.12, $pCO^2$ 34 mmhg, $HCO^{3-}$ 12.4, $pO^2$ 84 mmhg). On laboratory examination, complete blood count analysis was normal except mild anaemia (Haemoglobin 10.0 g/ dl, MCV 78fl, MCH 25, MCHC 32, RDW30). Erythrocyte sedimentation rate was normal and C reactive protein was negative too.Other biochemical examinations were normal except hyponatremia (123 mEq/L), hypokalaemia (2.2 mEq/L), and hypoalbuminemia (1.5 g/L). Liver and renal function tests were in normal range. (SGPT-19.2 IU/L, S.Creatinine-1.29 mg/dl, Blood Urea-73.3 mg/dl, Serum bilirubin total, direct and indirect were 0.5, 0.3,0.2 mg/dl respectively) Thyroid function test and parathyroid hormone levels were in normal range. However, ferritin level (10 ng/ mL) was found to be low. The serologies for human immunodeficiency virus, brucella, salmonella, hepatitis markers were negative. Cortisol and adrenocorticotrophic hormone levels were measured as normal (41.8 ng/ mL, 17.3 pg/mL respectively). The abdomen and urinary systems' ultrasound examination and urine analysis were normal. The amount of urinary protein excretion for 24 hours was less than 4 $mg/m^2$ /hour. Serum folate and vitamin B12 levels were normal. Urine and blood culture were negative. The serologic markers for celiac disease were positive (tissue transglutaminase immunoglobulin (Ig) A and G, anti-gliadinIg A and G, anti endomysiumIg A). Therefore, endoscopy was performed and biopsy was taken. The examination of intestinal biopsy revealed CD. Villous atrophy with hyperplasia of the crypts and increased intraepithelial lymphocyte count was found on examination of biopsy.The diagnosis of celiac disease was based on the criteria of the European Society of Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN) [3]. Patient was initially treated for metabolic acidosis and shock with fluid resuscitation. Later, patient was started on inotropes and short term corticosteroid therapy with hydrocortisone. Patient re-

sponded to this treatment in the span of 4 to 5 days, and after 5 days inotropes and corticosteroid were stopped. Gluten free diet was continued when child was stabilized. Child responded with a satisfactory gain in weight and appetite, cessation of diarrhoea, reduction of pedal oedema and abdominal distension and correction of biochemical and metabolic derangement.

## Discussion

Toddlers and young children classically present with chronic diarrhoea, vomiting, poor appetite, abdominal distension, abdominal pain, irritability, and failure to thrive sometime after the introduction of gluten in the diet (4). More frequently, the child with CD presents with subtle gastrointestinal symptoms such as constipation. The child may also present with non-gastrointestinal symptoms (e.g., short stature) or be asymptomatic but have a parent with CD. Some children may be simply cranky or have sleep disturbance (5). In general, CD should be included in the differential diagnosis of most patients seen in a paediatric gastroenterology practice. Our patient had classic symptom of celiac disease (weight loss, vomiting, weakness, and chronic diarrhoea, abdominal distension). The symptoms had started after supplementary nutrition, which included gluten. The diagnosis of CD is established by positive results of serological testing and evidence of characteristic histopathology on intestinal biopsy. Characteristic histologic features of CD include varying degrees of villous atrophy, with hyperplasia of the crypts and increased intraepithelial lymphocyte count. Grading of the endoscopic results was done according to the Marsh criteria (6). Most symptomatic patients have partial, subtotal or total villous atrophy, which are Marsh type 3 lesions. Our patient was categorised under type 3, grade B1.

Positive identification of these abnormalities leads to a presumptive diagnosis of CD and institution of a gluten-free diet. Clear clinical improvement while the patient is following the diet yields a definitive diagnosis. Celiac crisis is the term that has been applied to patients with CD of acute onset that is severe enough to be potentially fatal. It may arise in patients with established CD or it may be the initial presentation of their disease [7]. Clinically, it is characterized by severe diarrhoea, dehydration and metabolic disturbances like hypokalaemia, hypomagnesaemia, hypocalcaemia, hypoproteinemia [7]. Various precipitating factors identified for crisis are severe malnutrition, infections, hypoproteinemia, and poor compliance to gluten free diet, bacterial overgrowth in setting of altered motility in CD and anticholinergic drugs.

Celiac crisis may not respond to a gluten-free diet alone. Some cases in literature, authors have used corticosteroids for variable periods. In severely ill children with celiac crisis, the use of corticosteroids hascaused a dramatic improvement [9-12]. In 1952, Anderson and di'SantAgnese followed the clinical course of 58 children with CD (9). They observed 35 episodes of celiac crisis and 3 fatalities among these patients. In 1951 Adlersberg et al reported that the use of corticosteroids in adults with CD was effective therapy but relapse occurred when treatment was withdrawn [10]. But over the time it was realized that early recognition of CD and then gluten free diet in these patients is quite helpful to tilt the balance. In our patient precipitating factors for celiac crisis were found to be hypoproteinemia and severe malnutrition. Steroid was started which showed a considerable improvement of signs and symptoms. In conclusion, in some patients, the lesions of CD may be extensive and may therefore result in more severe symptoms.

In conclusion, we emphasize that gastrointestinal symptoms, which are best known in celiac disease, metabolic disturbance and abdominal distension should preoccupy celiac crisis and besides supporting treatment, corticosteroids might be used for treatment.

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