Short-Term Corticosteroids for Celiac Crisis in Infants

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We report two infants with celiac crisis who continued to have persistent secretory diarrhea despite gluten and lactose free diet and supportive parenteral nutrition. The children were given corticosteroid therapy. After a five-day oral prednisone in the dose of 2 mg/kg/daily, both patients rapidly recovered.

Key words: *Celiac crisis, Corticosteroids, Infants.*

Celiac crisis (CC) is a rare and life-threatening complication of celiac disease (CD) [1-4]. Although possible at all ages, it is most often seen in children younger than two years of age [1-5]. Beside the usual measures, to achieve a successful recovery, glucocorticoid therapy is also occasionally necessary [1,2,5-7].

CASE REPORT

We present two infants with severe celiac crisis who required a short-term administration of steroids. The diagnosis of celiac disease was based on the criteria of the European Society of Pediatric Gastroenterology, Hepatology and Nutrition [8], and that of celiac crisis on the acutization of chronic diarrhea followed by severe dehydration, metabolic acidosis, refusal of meals, abdominal distension, hypoproteinemic edema and a marked decrease of bodyweight [1]. Stool examination for bacterial infection, Rota-Adeno virus latex agglutination test, ova and parasites, as well as duodenal fluid for *Giardia lamblia*, were negative in both cases.

Case 1. An 8-month-old male infant presented with severe dehydration, abdominal distension, generalized edema and marked malnutrition caused by a persistent two-month diarrhea which intensified a week prior to admission. Glutin had been introduced in the diet at 3 months of age and diarrhea had started 3 months later. The child was lethargic, with cold peripheries. Rectal temperature was 35.9°C, heart rate was 142/min, respiration 40/min and BP 50/30 mmHg. The child was malnourished with bodyweight 33% of the expected. There was perianal erythema and erosive changes. Stool pH was 5 and stool reducing substances was 1.5% (Clinitest 3+). Antibodies to tissue transglutaminase (ATTG) IgA was 68 U/mL.

Case 2. A 9-month-old female infant presented due to the worseningof a one and half-month progressive diarrhea followed by severe dehydration, abdominal distension, periorbital and pedal edema and malnutrition. This child had a rectal temperature of 36.8^{0}C, HR 144/min, RR 42/min and BP 55/30 mmHg. The child also had acute severe malnutrition. ATTG IgA was 88 U/mL. She also had gluten introduction in diet at 3 months and onset of diarrhea at 7.5 months.

Following correction of water, electrolyte and acid-base disturbances, both patients were started on supportive parenteral nutrition. After a week of treatment, the biopsy of small bowel mucosa was performed using the Watson's modification of Crosby's capsule for children with double port. In both children the mucosa, viewed under the dissecting microscope, was flat (Marsh score IIIc).

Although the condition of both infants stabilized under the applied therapy, due to persistent secretory

TABLE I BLOOD LABORATORY VALUES ON ADMISSION

Case	Sodium (mmol/L)	Potassium (mmol/L)	pH	Creatinin (μmol/L)	Calcium (mmol/L)	Magnesium (mmol/L)	Phosphate (mmol/L)	Albumin (g/L)	Hb (g/L)
1	126	2.1	7.22	133	1.02	0.61	0.62	18	93
2	128	2.4	7.24	108	1.1	0.64	0.66	24	71

diarrhea requiring a continual intravenous fluid substitution and also perpetual anorexia, in case 1 on the 10th and in case 2 on the 11th day after admission, we started oral prednisone therapy in the dose of 2 mg/kg/ daily. After five days, both infants showed improvement in diarrhea and improved appetite. During the next week, prednisone therapy was gradually decreased and finally stopped. Under a strict gluten free diet, a two-week lactose free diet and four-month addition of iron, folic acid and multivitamin preparations, both patients achieved full recovery. After six-month treatment, serum transaminase activity was also normal. By provocation gluten tolerance test, in case 1 aged 5.8 years and in case 2 aged 5.6 years, we confirmed the diagnosis of celiac disease.

DISCUSSION

Although celiac crisis usually resolves to rehydratation, correction of metabolic imbalance, gluten and lactose free diet, and supportive parenteral nutrition, the rapid recovery of the patient may sometimes fail [1,2]. In cases resistant to conventional therapy, several authors have reported positive experience with the use of intravenous corticosteroid therapy based on its anti-inflam-matory effects, as well as positive influence on the bowel epithelium maturation [5-7].

We report two infants with CC resistant to conventional therapy. Both were breastfed for a short time and were early exposed to gluten. Also, in both, the disease was of long duration. In none of the cases, celiac crisis was precipitated with gastrointestinal or extraintestinal infection. Although being aware of side effects, the lack of response to gluten and lactose free diet and supportive parenteral nutrition prompted us to use prednisone. Additionally, the lack of gastrointestinal and extraintestinal infections made such therapeutic approach easier. Short-term, administration of corticosteroids was justified in both cases, with no side effects. However, this option should only be exercised in cases who are resistant to standard therapy.

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