

returned to normal within 6 hours. After hyperammonemia resolved, carnitine dosage was reduced to 30 mg/kg/day and protein-restricted diet was discontinued. The patient's ammonia levels remained within normal limits. At 3 years of age, the patient has no neurodevelopmental abnormalities.

The initial NCG dosage for treatment of acute hyperammonemia ranges between 100 to 250 mg/kg/day [2,3]. After the acute episode, the lowest reported effective daily dosage is 15 mg/kg/day [4]. Our patient had no hyperammonemia episodes under NCG treatment with a dose of 10 mg/kg/day. However, it seems that a lower dose is not enough during illness, as he was using a dose of 20 mg/kg/day when he developed hyperammonemia along with an infection. NCG therapy appears to correct the metabolic defect; therefore, dietary protein can be increased to 2-3 g/kg/day in some patients [2,5]. In our child – after the NCG dosage was raised to 30 mg/kg/day – protein restriction was totally removed but the ammonia levels remained normal. We conclude that NCG is effective for controlling hyperammonemia in NAGS deficiency; at a much lower dose, except during acute infections.

ERTUGRUL KIYKIM AND TANYEL ZUBARIOGLU

*Istanbul University Cerrahpasa Medical Faculty,
Department of Pediatrics,
Division of Nutrition and Metabolism, Istanbul, Turkey.
rtugrulkiykim@hotmail.com*

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Steroids in Celiac Crisis: Doubtful Role !

Celiac crisis is characterized by severe diarrhea, dehydration, hypokalemia, hyponatremia, hypomagnesaemia, hypocalcaemia, and hypoproteinemia. Although seen in all ages, it is most often seen in children younger than two years [1]. Apart from the usual supportive care, glucocorticoid therapy is usually required to achieve a successful recovery [2-4]. We present three patients who presented with celiac crisis. Despite adequate care and early institution of steroids the outcome was unfavourable in two of them.

First was a 9-year-old boy, and second a 4-year-old boy; both presented with history of recurrent diarrhea, weight loss and abdominal pain. Both patients were lethargic, emaciated, dehydrated and hypotensive. Serum level of tissue transglutaminase (TTG) IgA antibodies was >200 IU/mL in both children. Endoscopy in second patient revealed flattened duodenal folds with scalloped margins, and partial villous atrophy. Both these patients received full supportive care, including intravenous

hydro-cortisone. First patient died due to disseminated intra-vascular coagulation, and second did not improve; the parents got him discharged against medical advice.

Third patient was a 5-year-old girl, known case of celiac disease, who presented with worsening diarrhea, and weight loss, pedal edema. TTG levels were 190 IU/mL. This child received full supportive care; she gradually improved and was discharged after 3 weeks.

The reason why some patients with celiac disease have a much severe course is unclear. A combination of varied mucosal inflammation, immune activation and disruption of normal patterns of motility is likely [5]. The possible precipitating cause of crisis in our patients were severe malnutrition, hypoproteinemia, infection and late diagnosis. Corticosteroids are indicated in celiac crisis to reduce the mucosal inflammation, restore brush border epithelium enzymes and cause positive influence on the bowel epithelium maturation [1,3,4]. However, two of our patients deteriorated on steroids; third improved despite receiving no steroids. Use of steroids, especially with a probability of underlying sepsis, could be counterproductive. Further, steroids can exaggerate hypokalemia by causing kaliuresis. The role of steroids in celiac crisis needs further evaluation.

SHALU GUPTA AND KAPIL KAPOOR

Department of Pediatrics
LHMC and Kalawati Saran Children Hospital,
New Delhi, India; drshalugupta@yahoo.co.in

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Neonatal Meningitis due to *Neisseria meningitidis* Serogroup Y

Neonatal meningococcal meningitis is very rare; among the existing 13 serogroups of *Neisseria meningitidis*, serogroups B, C and Y have been reported in neonatal meningococcal meningitis [1-3]. Here we report isolation of *N. meningitidis* serogroup Y from the cerebrospinal fluid of a neonate which to the best of our knowledge is the first report from India. This could alert the clinicians to keep an index of suspicion for this bacterium in the etiology of neonatal meningitis.

A 38-week-gestation male baby, born by Caesarean section to a 32 year old multigravida in a district hospital with a birth weight of 2500 grams, presented on 14th day of life with symptoms of lethargy and refusal to feed. He had no history of seizure, apnea, or fever. On examination he had bulging anterior fontanel and hypotonia. He was afebrile and no skin rash or petechiae was seen. Sepsis screen was found positive with a C-reactive protein value of 2.76 mg/dL (Normal: 0.5mg/dL). The total leucocyte count was 4485 cells/mm³. Blood culture was negative. The CSF biochemical and cytological parameters revealed protein 65 mg/dL, sugar 35 mg/dL, and a cell count of 4cells/mm³ (all lymphocytes). No bacteria were seen on direct Gram staining. CSF culture showed growth of *Neisseria meningitidis* whose identification was based on colony appearance, gram stain and biochemical tests. Identification was confirmed using polymerase chain reaction targeting the conserved regulatory gene *crg A* using established primers [4]. Slide agglutination with *N. meningitidis* antisera (Difco, BD Diagnostics) confirmed it as *N. meningitidis* Serogroup Y. Nasopharyngeal swabs

from both parents were negative for the bacterium. The neonate recovered following treatment with amikacin for 7 days and piperacillin-tazobactam for 21 days.

To the best of our knowledge there exists only one previous case of neonatal meningitis associated with sepsis due to Serogroup Y who had a favourable outcome [3]. Previous authors have remarked on the importance of a lumbar puncture to avoid missing the diagnosis in this age group [5]. In the present case, even though the CSF biochemical parameters were not suggestive of meningitis, culture was helpful and demonstrated the existence of this rare pathogen.

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UTPALA DEVI AND JAGADISH MAHANTA

Regional Medical Research Centre,
NE Region (ICMR), Post Box-105
Dibrugarh-786001, Assam, India
jmahanta@gmail.com

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