Unarousable child with a short bowel

Luisa Cortellazzo Wiel ,[1] Ester Conversano,[1] Giorgia Carlone ,[1] Grazia Di Leo,[2]
Giulia Gortani,[2] Chiara Zanchi,[2] Egidio Barbi[2]

UNAROUSABLE CHILD WITH SHORT BOWEL

A 4-year-old boy was admitted with progressive lethargy of a few hours' duration and no other symptoms. His medical history was relevant for short bowel syndrome (SBS), following neonatal volvulus, with residual bowel length of 23 cm and intact ileocecal valve. He had similar self-limiting episodes in the past, after weaning parenteral nutrition, especially after eating large meals. The day before, he had consumed a large amount of apples.

Arterial blood gas (ABG) analysis showed metabolic acidosis with normal lactacidaemia (pH 7.09, pCO_2 19 mm Hg, pO_2 101 mm Hg, HCO3 5.8 mmol/L, BE −24, anion gap 29.4, chloride 116 mmol/L, L-lactate level 4 mmol/L).

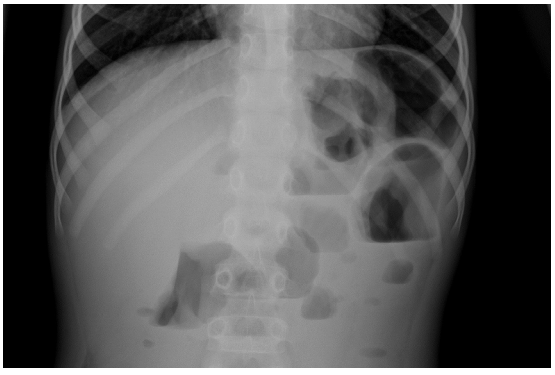

Figure 1 Abdominal distension with gas and bloating.

On admission, the child could be awakened, but he was confused with slurred speech (Glasgow Coma Scale 14), with a body temperature of 37 C°, a heart rate of 125 beats/min and a respiratory rate of 38 breaths/min. The abdomen was distended, without guarding and with normal bowel sounds. Blood glucose levels were normal, as well as white blood cell count, liver and kidney function test and C reactive protein. An abdominal ultrasound ruled out an intussusception. An abdominal X-ray was performed too (see figure 1).

QUESTIONS

1. Which is the most likely diagnosis?
   - A. Encephalitis
   - B. D-lactic acidosis
   - C. Dehydration with third space fluid collection and acidosis
   - D. Hereditary fructose intolerance.
2. How is this diagnosis confirmed?
   - A. D lactic dosage
   - B. Breath test for bacterial overgrowth
   - C. Urine organic acid dosage
   - D. Search for reductive substances in the stools.
3. How should this patient be managed?
   - A. Intravenous fluids to facilitate D-lactic excretion
   - B. Restrict carbohydrates in the diet
   - C. Intravenous bicarbonates
   - D. Antibiotic treatment to reduce bowel bacterial overgrowth.

Answers can be found on page 197.

ANSWERS TO THE QUESTIONS ON PAGE 196

1. THE ANSWER IS B

On the ground of history, clinical presentation and ABG, a D-lactic acidosis was suspected.

D-lactate is an isomer of L-lactate, which is normally undetectable in human peripheral blood, because of the lack of an human specific enzyme (D-lactate dehydrogenase) for its production. It is produced by the fermentation of non-absorbed carbohydrate by bacterial flora in the colon into organic acids.

D-lactic acidosis occurs in patients with SBS, due to the higher amount of simple carbohydrate delivered to the colon and fermented by D-lactate-producing lactobacilli.[1] Clinical presentation consists of an abrupt onset of altered mental status, slurred speech, nystagmus and ataxia, the so called 'D-lactate encephalopathy', occasionally progressing to a frank but reversible coma.[2] Consistent with metabolic acidosis, Kussmaul breathing and vomiting may be present. Abdominal distension with gas and bloating is a common finding and abdominal X-ray could be a clue to the diagnosis.[3]

ABG shows metabolic acidosis with high serum anion gap and normal lactacidaemia, as most laboratories routinely perform only L-lactate measurement. D-lactic acidosis is reported to be often misdiagnosed as alcoholic or other forms of encephalopathy, making its true incidence unknown.[4] Differential diagnosis can rely on the mnemonic of 'KILU', which stands for ketoacidosis (DKA), ingestion of ethylene glycol, methanol and salicylate, L-lactacidosis and uraemia. The differential diagnosis includes others causes of high-serum anionic gap metabolic acidosis in children, such as ketotic hyperglycaemic in DKA and increased L-lactate levels in dehydration; both conditions are not hyperchloremic and improve with intravenous fluid therapy. Instead, in the proximal and distal renal tubular acidosis, bicarbonate levels are low, with an hyperchloremic metabolic acidosis with normal plasmatic anion gap. In all the former clinical conditions, history and recurrence of symptoms are substantially different from D-lactic acidosis.[5 6]

2. THE ANSWER IS A

D-Lactic acidosis is conventionally defined as a blood D-lactate concentration above 3 mmol/L. Greater concentrations have been related to brain damage, although a linear correlation between serum concentration and outcomes has not been described.

The diagnosis can be challenging, requiring specific essays for D-lactic acid, which is not always available in the emergency setting; thus a high index of suspicious is needed in front of the clinical picture of an acute-onset encephalopathy in a patient with SBS. Furthermore, in patients with SBS, breath test could be useful to detect proximal bowel bacterial overgrowth, which acts as a contributing factor for the occurrence of D-lactic acidosis.[7] A urine organic acids dosage will rule out an organic aciduria, which is not associated as a rule to SBS.

3. ALL, A, B, C AND D ARE CORRECT

The cornerstone of acute management consists on the cessation of carbohydrate intake with the aim of preventing the further production of D-lactic acid, 'starving' the colonic bacteria. Intravenous fluids, avoiding lactated Ringer's solutions, are useful to facilitate renal clearance. Intravenous bicarbonate should be given slowly in case of altered mental status, and then orally to correct metabolic acidosis. Poorly absorbed antibiotics, such as aminoglycosides or rifaximin, are used to control the bowel bacterial overgrowth.

Long-term approach relies primarily on the restriction of monosaccharides and disaccharides, which are major risk factors for bowel bacterial overgrowth and D-lactic acidosis, associated with the presence of colon without continent ileocecal valve[8]; moreover, other long-term preventive measures include avoidance of exogenous fermented foods, such as yoghurts and pickles.

Once recognised and properly managed, D-lactic acidosis has a favourable outcome, without occurrence of residual deficits.

PATIENT OUTCOME

The patient displayed restoration of mental status and improved metabolic equilibrium within 8 hours. An empirical course of antibiotic decontaminating treatment was started with metronidazole and rifaximin.

Luisa Cortellazzo Wiel ,[1] **Ester Conversano,**[1] **Giorgia Carlone** ,[1] **Grazia Di Leo,**[2] **Giulia Gortani,**[2] **Chiara Zanchi,**[2] **Egidio Barbi**[2]
[1]Department of Medicine, Surgery, and Health Sciences, University of Trieste, Trieste, Italy
[2]Paediatrics, IRCCS Materno Infantile Burlo Garofolo, Trieste, Italy

Correspondence to Dr Giorgia Carlone, Department of Medicine, Surgery and Health Sciences, University of Trieste, Trieste, Friuli-Venezia Giulia, Italy; giorgiacarlone@gmail.com

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ORCID iDs
Luisa Cortellazzo Wiel http://orcid.org/0000-0003-0069-560X
Giorgia Carlone http://orcid.org/0000-0002-2926-5462

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