d-Lactic acidosis in a ten months old infant with short bowel syndrome: Early suspicion equals early treatment

Acidosis d-láctica en un niño de diez meses afecto de un síndrome de intestino corto: la rápida sospecha equivale a un rápido tratamiento

Dear Editor

Metabolic acidosis is a disorder of the acid–base balance which may be due to bicarbonate losses, deficient acid elimination by the kidney, exogenous intake of acids or an endogenous increased acid production. Within this last group, lactic acidosis should be noted, because of its frequency, morbidity and potential mortality. Lactic acidosis, which is generally caused by accumulation of the l-lactic isomer, can be congenital or secondary, the latter is in relation to tissue hypoxia (type A) or to hepatic, renal or oncologic diseases, intense exercise, seizures or toxics (type B). Less common are the situations where the cumulative isomer is the d-lactic, which have occasionally been reported in patients suffering from short bowel syndrome (SBS), presenting with a metabolic acidosis, increased anion GAP and neurological symptoms.

We present a case of a ten month old infant with SBS secondary to gastrosisch and intestinal necrosis. He had undergone surgery three times during the neonatal period with a remaining small bowel of 30 cm preserving the ileocecal valve. The patient’s diet was composed of extensively hydrolysed lactose-free formula, gluten-free cereal, vegetables and chicken. Feeding was done mostly through nasogastric tube, with some participation of oral feeding. Metronidazole 20 mg/kg/day and ceftizime 8 mg/kg/day were administered alternatively the first week of each month as intestinal decontamination protocol. The month before admission, abundant stool production with decreased consistency associated to weight loss was present. Coincidentally, the decontamination protocol was not followed and 48 h prior to admission, he developed a respiratory syncytial virus respiratory tract infection. On admission, the anthropometric data showed severe malnutrition: weight 4.41 kg (<P1, –5.2 DE), height 63 cm (<P1, –4.62), based on Fernández et al., 2011 standards. Shukla nutritional index was 50.65%. Poor general condition, severe malnutrition and dehydration signs were appreciated. Tachypnea, altered level of consciousness and generalized hypotonia were also present. Severe metabolic acidosis was noted (pH 6.8, bicarbonate 4.3 mmol/L) and the anion GAP was increased (27 mEq/L) with normal lactic levels (0.7 mmol/L). Ketonuria was absent. Correction therapy was started with intravenous fluids containing bicarbonate and oral metronidazole. Prior to this, a venous blood sample in a lithium heparin tube was immediately centrifuged. Plasma was frozen and sent for d-lactic acid analysis to a reference laboratory (Birmingham Children’s Hospital, Birmingham, through Reference Laboratory, Barcelona), confirming the suspected diagnosis (d-lactic acid > 6 mmol/L). The outcome was satisfactory with normalization of neurological symptoms and gradual improvement of the acid–base balance.

D-Lactic acidosis usually originates from an accumulation of d-lactic secondary to an elevated synthesis by gastrointestinal tract bacteria. Production of d-lactic is minimal under normal conditions and is easily metabolized by the mitochondrial d-lactate dehydrogenase. However, in the SBS, a high carbohydrate intake may generate an increased production and subsequent accumulation of d-lactate because of the bacterial overgrowth. This entity should be suspected in patients associating neurological symptoms and metabolic acidosis with elevated anion GAP without increasing l-lactic acid, as in our case. We could not find in the literature any report involving such a young infant. We highlight that in our routine blood analysis, only l-lactic acid is detected. When this condition is suspected, it should be confirmed by a special measurement of serum or plasma d-lactic concentration. Normal values of d-lactic acid in blood are undetectable, being those above 3 mmol/L pathological.

The treatment is based on pathogenic flora elimination (administration of oral nonabsorbable antibiotics as metronidazole, neomycin or vancomycin), correction of the acidosis and carbohydrate restriction diet. Administration of probiotics is controversial. Bifidobacterium breve and Lactobacillus casei are theoretically non d-lactate producing flora and have been occasionally used as adjuvant therapy with antibiotics, in order to replace the pathogenic flora. However, two cases of d-lactic acidosis in patients with SBS attributed to the administration of probiotics containing d-lactate producing strains (Lactobacillus acidophilus, Lactobacillus buiharicus and Bifidobacterium infantis) have been reported. Probiotics must be carefully selected in children with SBS. There are few data available related to the ideal maintenance therapy due to the lack of controlled studies. Avoiding fast-acting carbohydrate diets, periodical decontamination or the use of non d-lactate producing probiotics are different options which should be individualized in patients at risk of developing d-lactic acidosis.

This case is a reminder of the importance of suspecting this rare entity in SBS affected children presenting with neurological symptoms and metabolic acidosis where routine blood tests do not detect the increased anion. In these situations an early treatment should be mandatory.

References

Bedside lung ultrasound in paediatric intensive care

Utilidad de la ecografía pulmonar a pie de cama en cuidados intensivos pediátricos

Dear Editor,

In recent years, the use of point-of-care ultrasound (PoCUS), understood as the bedside ultrasound examination of the patient by the physician in charge, has been increasing in paediatric patients for the investigation of pulmonary, pleural and diaphragmatic disease, following its use in adult patients, in whom the technique has been developing for more than a decade, and whose ultrasound semiotics can be safely extrapolated to the paediatric age group. The unossified costal cartilage and sternum and thinner adipose subcutaneous tissue in children provide ideal acoustic windows.1,2

There are limitations to plain chest radiography, such as poor image quality, the presence of artefacts, the time required to obtain the image and the exposure to ionising radiation. There are also limitations to chest computerised tomography (CT), the gold standard for the diagnosis of respiratory pathology, including its high cost, reduced availability, higher exposure to radiation and difficulty involved in transporting the patient outside the unit.3 Thus, PoCUS is emerging as the ideal diagnostic tool in the paediatric intensive care unit (PICU): quick, non-invasive, repeatable, offering real-time information and without exposure to radiation, with sensitivities and specificities that approximate those of computed tomography (CT).4,5

We present five clinical cases of common diseases in the PICU in which PoCUS was a useful diagnostic tool and guided changes to the therapeutic approach.

Case 1

Infant aged 2 months admitted for bronchiolitis. After being intubated, the patient had difficulty with oxygenation and ventilation, leading to suspicion of a secondary pneumothorax. PoCUS: sonographic signs of right-sided pneumothorax (absence of lung sliding, stratosphere sign, absence of B lines, presence of lung point). After insertion of a pleural drainage tube, the resolution of pneumothorax was detected by ultrasound. The use of PoCUS prevented diagnostic and treatment delays (Fig. 1).

Case 2

Male aged 5 years with pleural effusion associated with right lower lobe pneumonia on the fifth day of antibiotic treatment. PoCUS: right-sided pleural effusion (quad sign) and consolidation of the underlying lung parenchyma (tissue-like sign or hepatisation and bronchograms). A pleural drainage tube was placed, and the draining fluid had normal characteristics. These findings led to performance of a contralateral PoCUS, which revealed a small volume of pleural effusion and consolidation of the underlying lung parenchyma. Following the sonographic diagnosis of bilateral pneumonia, testing for atypical pathogens was requested, the results of which were positive for Mycoplasma pneumoniae and adenovirus. PoCUS was useful in guiding the thoracocentesis and the aetiologic diagnosis, and in finding evidence of consolidation that had not been detected by plain radiography (Fig. 1).

Case 3

Male aged 17 years with a history of operated tetralogy of Fallot admitted to the unit for respiratory difficulty and cyanosis following resection of a nasal polyp. At admission, the patient presented with mild tachycardia, tachypnoea, cyanosis, intercostal and suprasternal retractions and rales. PoCUS: pattern of coalescing B lines in anterior regions of both lungs (bilateral diffuse interstitial syndrome). In light of these findings, the tachycardia was reassessed, leading to diagnosis of atrial flutter with 2:1 conduction. The use of PoCUS allowed the diagnosis of acute pulmonary oedema secondary to arrhythmia in a patient with a prior history of ventriculotomy and a difficult-to-interpret ECG (Fig. 1).

Case 4

Male aged 7 years that had received a diagnosis of severe Ebstein’s anomaly and admitted following surgical closure of atrial septal defect, tricuspid valve repair and a biderectorial Glenn procedure. The patient had a complicated...