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Letter to Editor

Anaesthesia Section

A Rare Cause of Sudden Onset-Severe Metabolic Acidosis in Paediatric Surgical Patients- Organic Acidemia

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Sir,

A one-year-old male, weighing 7 kg, suddenly developed recurrent vomiting, tachypnea and became lethargic 6 hours after cicumcision. Circumcision was done under sedation using intravenous midazolam and fentanyl. He was afebrile and blood sugar was normal at the time of presentation. On systemic examination, chest was clear, no murmur heard on auscultation of heart. Abdomen was soft, non tender with no organomegaly. On blood gas analysis, there was severe metabolic acidosis (pH <6.9) with increased anion gap [Table/Fig-1]. Intubation done and put on mechanical ventilation to decrease work of breathing.

To rule out aetiologies of metabolic acidosis, blood was sent to measure levels of lactate, pyruvate, ammonia and chloride level along with complete blood count and serum procalcitonin level. After that, we started 0.45% DNS infusion as a maintenance fluid after giving some boluses. Inj. ceftriaxone 750 mg i.v. once a day started. Sodium bicarbonate infusion started as there was severe metabolic acidoses along with hypotension with nil urine output. After 1 hour, vitals became stable along with improvement in urine output. Urine was send to evaluate renal response of metabolic acidosis and to evaluate urinary organic acid profile. ABG repeated after 6 hours showing slight improvement in acidosis but still pH was<7.0. Complete blood count and serum procalcitonin was within normal limit. Urinary anion gap was negative while organic acids were found on analysis by Gas chromatography/mass spectrometry (GC/MS). With these investigation findings, we concluded that it could be a case of organic acidemia. We started tab thiamine 300 mg daily, tab biotin 10 mg 6th hourly, capsule carnisure (L-carnitine) 200 mg 8th hourly through Ryle's tube and hydroxycobalamine (vitamine B12) 1 mg intramuscularly(IM) daily. There was significant improvement after 12 hours of treatment [Table/Fig-2].

Sodium bicarbonate infusion stopped after 24 hrs of infusion. He was off ventilator on 2nd day and discharged to ward after 3 days of ICU admission. He was discharged to home with continuation of oral cofactor therapy and regular follow-up to detect any developemental delays.

FiO_2	0.4
pH	6.93
pCO_2	9 mmHg
pO_2	108 mmHg
Na+	138 mmol/L
K+	5.1 mmol/L
Ca+	1.33 mmol/L
Glucose	101 mg/dl
Lactate	0.6 mmol/L
Haematocrit	36%
HCO_3	3.0 mmol/L

FiO_2	0.3
pH	7.25
pCO_2	14 mmHg
pO_2	100 mmHg
Na+	156 mmol/L
K+	2.8 mmol/L
Ca+	1.23 mmol/L
Glucose	94 mg/dl
Lactate	0.5 mmol/L
Haematocrit	31%
HCO_3	10.0 mmol/L

[Table/Fig-1]: ABG on 1st day.
[Table/Fig-2]: ABG on 2nd day after initiation of adjuvant therapy.

From the literature, it has been found that Inborn Errors of Metabolism (IEM) are an important cause of acute illness in newborns and infants. IEM are disorders in which there is a block at some point in the normal metabolic pathway caused by a genetic defect of a specific enzyme [1].

While the diseases individually are rare, they collectively account for a significant proportion of neonatal and childhood morbidity and mortality. Diagnosis is important not only for treatment and prognostication but also for genetic counselling and antenatal diagnosis in subsequent pregnancies. Organic acidemias should be considered in the differential diagnosis of any sick child along with common acquired causes such as sepsis, hypoxic-ischemic encephalopathy, duct-dependant cardiac lesions, congenital adrenal hyperplasia and congenital infections. Children with an organic acidemia are susceptible to metabolic decompensation during episodes of increased catabolism, such as intercurrent illness, trauma, surgery, or prolonged episodes of fasting [2]. Metabolic investigations should be initiated as soon as the possibility is considered. The outcome of treatment of many IEM is directly related to the rapidity with which problems are detected and appropriate management instituted.

Complete blood count, arterial blood gases and electrolytes, blood glucose, plasma ammonia, arterial blood lactate, liver function tests, urine ketones and CT/MRI are the first line investigations in suspected cases of IEM [3].

In most cases, treatment needs to be instituted empirically without a specific diagnosis.

To reduce the formation of toxic metabolites, we can decrease availability of particular substrate for metabolism but we have to provide adequate calories parenterally as well. By means of haemodialysis, we can enhance the excretion of toxic metabolites. Co-factor therapy should be instituted for specific disease even if diagnosis is not established. Along with all these things supportive care is very important like treatment of seizures (avoid sodium valproate as it may increase ammonia levels), maintenance of euglycaemia and normothermia, fluid, electrolyte & acid-base balance, treatment of infection, mechanical ventilation if required.

Appropriate immediate treatment not only improves survival but also reduces the chances of neurodevelopmental sequelae.

Although IEM are rare findings but think of IEM in sick children in parallel with other common conditions such as sepsis so that it could be treated timely.

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