Case Report

Recurrent D-lactic acidosis in a child with short bowel syndrome

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D-lactic acidosis is a rare complication in children with short bowel syndrome. It results from fermentation of dietary carbohydrate by luminal bacteria in the small bowel caused by bacterial overgrowth. We present the case of a 14-year-old boy who had been diagnosed with short bowel syndrome from surgical treatment of midgut volvulus five years previously. His nutritional status was maintained by total parenteral nutrition and enteral feeding as tolerated. During hospitalization, episodic confusion and hyperpnea developed. The investigation showed severe metabolic acidosis with serum bicarbonate of 9 mmol/L and a wide anion gap. The serum D-lactic acid was 11.21 mmol/L. There was no evidence of renal or hepatic failure. Therefore, D-lactic acidosis from enteral carbohydrate overload was diagnosed. The treatment was correction of metabolic acidosis by sodium bicarbonate infusion and carbohydrate restriction. The results of the therapy were satisfactory. Early detection and appropriate treatment is necessary to avoid morbidity and mortality following this complication of short bowel syndrome.

Key Words: lactic acidosis, short bowel syndrome.

Introduction

Short bowel syndrome is defined as congenital or postnatal loss of at least 50% of the small bowel, with or without loss of a portion of the large intestine. It can lead to severe malabsorption and malnutrition. Congenital short bowel syndrome usually occurs as a condition of malrotation or gastroschisis in utero. Most cases of postnatal loss of the bowel are due to surgical resection of the small intestine from Crohn disease, trauma or other small bowel diseases.[1]

The consequences of short bowel syndrome are numerous, including bacterial overgrowth, growth failure, micronutrient deficiency and metabolic derangements. D-lactic acidosis is rare in children with short bowel syndrome. It results from fermentation of dietary carbohydrate by luminal bacteria in the small bowel caused by bacterial overgrowth. It was first described in adults in 1979[2] and in children in 1980.[3] Following this initial report, several other cases of this syndrome in children have been described.[4-13] The present reports D-lactic acidosis in a patient with short bowel syndrome in a university teaching hospital in Thailand.

Case report

A fourteen-year-old boy was admitted with a problem of vomiting and poor oral intake. The child had a history of short bowel syndrome from surgical treatment of midgut volvulus five years previously. The surgical procedure was jejunal and ileal resection with end to end anastomosis (15.2cm of jejunal and 10.2cm of ileal bowel remaining with preserved ileocaecal valve). Following the surgery, he was on total parenteral nutrition and received enteral feeding as tolerated. His regular diet was low fat and low fiber. Each meal was small, but he was fed frequently. His stool was semi-formed, with two to three defaecations per day. He developed protein-energy malnutrition and required parenteral nutrition three days per week at the community hospital, including a micronutrient supplement which consisted of zinc solution, ferrous sulfate and multivitamins. Two months prior to admission, antibiotics (cotrimoxazole 25mg/kg/day and metronidazole 30mg/kg/day) were prescribed alternately every month for the treatment of bacterial overgrowth in his gastrointestinal tract. He was admitted to the hospital because of vomiting, poor oral intake and severe malnutrition. Physical examination revealed stable vital signs, body weight 20 kg, height 131cm, wasted, stunted appearance and hypopigmented hair. He had good consciousness and mild dehydration.

The remainder of the examination was unremarkable. The initial laboratory findings were within normal limits. He was put on total parenteral nutrition as detailed in Table 1. Oral intake was provided as tolerated. His eating habits gradually improved. On the twelfth day following admission, he developed lethargy, mental confusion and slurred and incoherent speech after a large meal. Hyperpnea

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Accepted 14 January 2005

Table 1. Total parenteral nutrition

Volume (ml/kg/day)	75.0
Energy (Kcal/kg/day)	77.0
Carbohydrate (g/kg/day)	15.0
Protein (g/kg/day)	2.0
Fat (g/kg/day)	2.0
Sodium (mmol/kg/day)	3.0
Potassium (mmol/kg/day)	3.0
Chloride (mmol/kg/day)	3.0
Acetate (mmol/kg/day)	3.0
Phosphorus (mmol/kg/day)	0.5
Calcium (mmol/kg/day)	0.5
Magnesium (mmol/kg/day)	0.2
Zinc (µg/day)	100.0
Trace element (ml/day)	1.5
Multivitamin	§

§ contains thiamine mononitrate 3.2mg, riboflavin 3.6mg, nicotinamide 40mg, pyridoxine 4mg, pantothenic acid 15mg, ascorbic acid 100mg, biotin 60µg, folic acid 0.4mg, cyanocobalamin 5µg, retinol 690µg, calciferol 10µg, alphatocopherol 6.4mg and phytomenadion 200µg

Table 2. Laboratory findings at each episode of mental confusion and metabolic acidosis

	First episode	Second episode	Third episode	Fourth episode
Bicarbonate (mmol/L)	6.00	9.00	7.00	10.00
Anion gap (mmol/L)	24.75	28.20	24.68	24.62
Blood for pH	7.26	7.23	-	7.28
Base excess (mmol/L)	-20.90	-19.10	-	-12.60
Plasma D-lactate (mmol/L)	-	11.21	-	-
Plasma L-lactate (mmol/L)	-	2.89	1.55	1.44

was also observed. He had no fever and no focal neurological abnormality. The laboratory investigation showed severe metabolic acidosis with serum bicarbonate of 6mmol/L, pH of 7.26 and base excess of -20.9 mmol/L (Table 2). Serum sodium and potassium were both normal. The anion gap was calculated as 24.75 mmol/L. There was no evidence of renal or hepatic failure. He was given sodium bicarbonate infusion. Thiamine deficiency could not be excluded, therefore a high dose of thiamine was also administered intravenously. Enteral feeding was discontinued. He recovered in 24 hours, and a regular diet was reintroduced 24 hours after this recovery.

Four days later, while still on the high dose of thiamine, a recurrent episode of mental confusion and hyperpnea occurred after a large meal with similar clinical and biochemical disturbance. D-lactic acidosis was suspected and confirmed by a markedly elevated level of D-lactic acid (11.21 mmol/L) (Table 2). These findings were consistent with D-lactic acidosis from enteral carbohydrate overload. Enteral feeding was discontinued and sodium bicarbonate was given to correct the metabolic acidosis. There was improvement of neurological symptoms and resolution of acidosis over the next 12 to 24 hours. A carbohydrate-restricted diet was re-introduced 24 hours after his mental status had returned to normal.

Two additional episodes occurred at the fortieth and fiftieth days after admission with similar clinical and biochemical disturbances (Table 2). The treatment for each episode was correction of the metabolic acidosis by sodium bicarbonate infusion and the child rapidly recovered. He was eventually discharged with body weight of 28kg and height of 137cm. Ongoing prevention of bacterial overgrowth was in the form of alternate oral metronidazole and co-trimoxazole for ten days in each month. Small and frequent enteral feeding was recommended. Over the following 14 months he remained well without further symptoms.

Discussion

D-lactic acidosis is an uncommon complication of short bowel syndrome. It was first described as a disease of humans in 1979,[2] although it had been known as a disease of ruminants for decades.[14] Following this, there were many further reports of this phenomenon resulting from short bowel syndrome secondary to a variety of causes, with the cases of fourteen children reported from 1980-1999 (Table 3). The present case is the first reported case in Thailand of D-lactic acidosis in a child due to short bowel syndrome from midgut volvulus.

D-lactic acidosis is defined as the condition of metabolic acidosis accompanied by an increase in serum D-lactic acid.[1] Lactic acid is classified into L- and D-lactic acid. Both compounds are produced from pyruvate by the enzyme lactate dehydrogenase (LDH) during glycolysis. The enzyme LDH is specific to each form of lactic acid; the production of L- and D-lactic acid requires L- and D-LDH respectively. Mammals have only the enzyme L-LDH, thus only L-lactic acid is normally found in the blood of humans, although small amounts of D-lactic may be found in human blood and excreted in the urine. This D-lactic acid is produced through the methyl-glyoxal pathway or bacterial fermentation in the gastrointestinal tract. Other sources are some kinds of fermented foods such as yoghurt and sauerkraut. Lactate Ringer Solution and peritoneal dialysate also can contain both L- and D-lactic acid.[15]

D-lactic acidosis is the result of the overproduction and absorption of D-lactic acid from the intestine, impaired D-lactic acid metabolism and colonic bacterial flora such as species lactobacillus that produce D-lactic acid. The malabsorption syndrome with increasing delivery of food particles to the colon can produce osmotic diarrhea which results in inadequate substrate to produce D-lactic acid. Hence, colonic stagnation might be another important contributing factor. In the present case, the patient had a large dinner before he developed the symptoms. It was possible to believe that the large amount of carbohydrate intake could not be absorbed in the small bowel, leading to increased delivery to the colon with fermentation by colonic bacteria to L- and D-lactic acid which caused D-lactic acidosis.

The clinical manifestations of D-lactic acidosis are recurrent episodes of encephalopathy and metabolic acidosis occurring in patients with short bowel syndrome.

Table 3. Previous case reports of D-lactic acidosis in children

Patient number	Author	Age (year)	Underlying disorder
1	Schoorel EP[3]	3.00	thrombosis of mesenteric vessels
2	Satoh T[4]	2.75	volvulus
3	Satoh T[4]	5.00	volvulus
4	Perlmutter DH[5]	1.50	malrotation/volvulus
5	Perlmutter DH[5]	5.00	volvulus
6	Rosenthal P[6]	2.50	midgut volvulus
7	Haan E[7]	8.00	volvulus
8	Scully TB[8]	16.00	trauma, leading to resection
9	Mayne AJ[9]	9.00	spontaneous volvulus
10	Gurevitch J[10]	1.67	malrotation/volvulus
11	Koletzko S[11]	9.00	intestinal resection
12	Bongaerts G[12]	1.00	no record
13	Bongaerts G[12]	3.00	no record
14	Day AS[13]	5.00	gastroschisis

Neurological abnormalities include confusion, dizziness, ataxia, dysarthria, headache, weakness, visual disturbance and behavioral changes. These symptoms last from a few hours to several days.[13]

The neurological abnormalities caused from D-lactic acid itself, and unknown compounds produced along with the D-lactic acid, can be toxic to the brain. Metabolic acidosis always occurs. The serum anion gap is usually wide. The present case had both neurological abnormallities and metabolic acidosis.

Physicians should consider the possibility of D-lactic acidosis when a patient presents with acidosis and one or more of any of the following clues: increased serum anion gap with normal lactic acid levels measured by conventional technique, presence of short bowel or other forms of malabsorption syndrome, acidosis that is preceded by oral intake and which improves when oral intake is discontinued, and neurological abnormalities. The diagnosis is confirmed by a high level of serum and/or urine D-lactic acid. Some reports have shown normal values of serum D-lactic acid (Table 4), but the exact normal value of serum D-lactic acid is unknown.

The serum D-lactic acid level in the present case was 11.21 mmol/L, which was markedly high, making D-lactic acidosis a definite diagnosis.

The treatment of D-lactic acidosis includes diminishing the amount of substrate available for colonic bacterial fermentation by prolonged oral fasting and use of low-carbohydrate diets, intravenous alimentation, oral antibiotics (such as metronidazole, neomycin, vancomycin, kanamycin and ampicillin) to eradicate the abnormal colonic flora, and correction of the current metabolic acidosis by sodium bicarbonate infusion.

Table 4. The normal value of serum D-lactic acid

Study	Normal value of serum D-lactic acid
Bongaerts G[12]	not found
Uribarri J[15]	≤ 3.0 mmol/L
Jover R[16]	< 0.5 mmol/L

Mayne[9] studied the dietary management of D-lactic acidosis in short bowel syndrome and showed that the manipulation of dietary carbohydrate with strict control of monosaccharides and oligosaccharides was successful in patients with D-lactic acidosis. The outcome of the present case was satisfactory. He did not have any recurrence of D-lactic acidosis after discharge. Prevention should be suggested to all patients, which includes avoidance of excess carbohydrates and prevention of colonic bacterial overgrowth.

Conclusion

D-lactic acidosis should be considered in patients with short bowel syndrome who develop encephalopathy and/or metabolic acidosis. Early detection and appropriate treatment with a low-carbohydrate diet are necessary to avoid morbidity and mortality.

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