Isolated fructose malabsorption

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Abstract

Manipulation of carbohydrate intake was used to treat severe, recurrent d-lactic acidosis in a patient with short bowel syndrome. Dietary carbohydrare composition was determined after assessment of d-lactic acid production from various carbohydrate substrates by faecal flora in vitro. This approach may be preferable to repeated courses of antibiotics.

D-lactic acidosis is a well recognised, but rare, complication of short bowel syndrome and intestinal bypass surgery. To date, seven children have been reported in whom this disorder occurred as a complication of short bowel syndrome. Non-absorbed carbohydrate is fermented by colonic organisms, but lactobacilli when present, unlike most other bacteria, produce d-lactic acid, which cannot be metabolised by d-lactic dehydrogenase. Absorption leads to a severe metabolic acidosis. Treatment has been directed at altering the colonic flora with antibiotics, or by administering a standard bacterial flora orally.

Recently we have successfully treated a girl with this disorder by dietary carbohydrate manipulation alone.

Case report

After massive small bowel resection for a spontaneous small intestinal volvulus, which followed a vigorous session of disco dancing, a 9 year old girl was left with 14 cm of jejunum beyond the duodenojejunal flexure; the ileocaecal valve was preserved. After prolonged parental nutrition she was discharged nine months later on a normal diet together with energy supplements given as sip feeds.
Six months later, after visiting a funfair and eating large amounts of sucrose rich foods, she presented with severe ataxia, slurred speech, and confusion. Biochemical investigations showed a severe metabolic acidosis (hydrogen ion concentration 80 mmol/l, carbon dioxide pressure 2·3 kPa, and bicarbonate 8 mmol/l) with a high plasma d-lactate concentration (9·1 mmol/l; normally not detected); l-lactate was normal (1·1 mmol/l; normal range 0·6–2·2) and ethanol not detected. She was treated with intravenous sodium bicarbonate and recovered rapidly.

During the next year she had several identical episodes that were related to eating large amounts of sucrose. Eighteen months after her operation episodes were occurring two to three times per month and she was exhibiting growth failure.

Nutritional support was provided by a whole protein feed (delivered nasogastrically by continuous infusion) in which the carbohydrate was provided as glucose oligosaccharides (Isocal, Mead-Johnson). Increasing the external feed to provide more than 30% of her energy requirement, however, again resulted in d-lactic acidosis.

Stool culture showed a pure growth of lactobacilli, but treatment with a variety of broad spectrum oral antibiotics was ineffective. This led us to investigate the d-lactate producing capabilities of her colonic flora in relation to various carbohydrate substrates in vitro, with a view to manipulating her dietary carbohydrate.

**Methods**

Two species of lactobacilli (Lactobacillus fermentum and Lactobacillus acidophilus) were identified in the patient’s stool. Their ability to produce acid from different carbohydrate substrates was assessed in DeMan, Rogosa, and Sharpe’s (MRS) lactobacillus broth in the presence and absence of a variety of carbohydrates (20 g/l). After incubation at 37°C for 48 hours, pH was tested with 2% bromocresol purple. Acid production was deemed to be present if the solution turned yellow (pH <6). Both organisms isolated from our patient fermented glucose, lactose, maltose, and sucrose, but there was no acid production from starch.

Further studies were undertaken to confirm that acid production in our test did indeed reflect the ability of lactobacilli to produce d-lactic acid from differing carbohydrate substrates.

Four different species of lactobacilli, including those of the patient, were therefore cultured as before, and the pH tested after 48 hours. After a further five days incubation, cultures were filtered and analysed blind for d-lactic acid using an enzymatic kit for l-lactate determination, substituting d-lactate dehydrogenase (Boehringer Mannheim).

**Results**

The results are shown in the table. Concentrations of d-lactate up to 141 mmol/l were obtained. A positive bromocresol purple reaction was present after 48 hours incubation in all solutions in which the d-lactic acid concentrations exceeded 14 mmol/l at seven days, with no false positives or negatives. This confirmed that the bromocresol purple reaction was a valid screen for d-lactic acid production by lactobacilli.

**DIETARY MANIPULATION**

Our investigations confirmed the absence of d-lactic acid production from starch by the patient’s Lactobacillus spp. The diet was therefore altered, such that carbohydrate was provided mainly in polymeric form, using Osmolite (Abbott Laboratories) as a nasogastric tube feed. This resulted in a dietary carbohydrate profile as follows: monosaccharide 1%, disaccharide 6%; trisaccharide 8%, tetrasaccharide 7%, and pentasaccharide (and above) 78%.

In the subsequent 30 months there have been no further episodes of d-lactic acidosis and no antibiotic treatment. Growth has resumed and the patient has remained well in full time education.

**Discussion**

D-lactic acidosis should be suspected in any patient with short bowel syndrome with recurrent acidosis, particularly when complicated by abnormal neurological signs. In 1983 Perlmutter et al showed that a reduction in dietary carbohydrate in children with short gut syndrome led to a reduction in plasma d-lactate.
concentration.\textsuperscript{1} Dahlquist \textit{et al} were able to precipitate an episode of D-lactic acidosis in a patient with intestinal bypass surgery, by using a 25-08 MJ (6000 kcal) load containing 54% carbohydrate.\textsuperscript{2} Similarly, Rosenthal and Pesce described a recurrence of abnormal neurological signs, together with high D-lactic concentrations, in a patient in whom enteral feeds were substantially increased.\textsuperscript{3}

Our studies show the importance of the nature of the dietary carbohydrate substrate in the production of D-lactic acidosis. Manipulation of dietary carbohydrate with strict control of monosaccharides and oligosaccharides was successful in our patient and preferable to repeated courses of broad spectrum antibiotics, which may alter the colonic flora in such a way as to impair valuable colonic salvage of non-absorbed nutrients.


Atopic eczema, hyponatraemia, and hypoalbuminaemia

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Abstract

We describe an infant with atopic eczema, treated with homoeopathic medicines, who presented with erythema and limb oedema. Concentrations of urinary and plasma sodium and plasma albumin were low. On conventional treatment he made a satisfactory recovery.

Atopic eczema affects 5–10% of children under the age of 5 years.\textsuperscript{1} We report an infant with atopic eczema, treated inappropriately with homoeopathic medicines, who became seriously ill with deterioration of the eczema and associated metabolic complications.

Case report

A 6 month old boy presented to casualty with a one week history of generalised erythematous weeping skin and pronounced oedema of the limbs. His parents had refused hospital admission when seen four days previously. Since 1 month of age his skin had been dry and eczematous. Homoeopathic medicines were prescribed for his eczema by a registered homoeopath. The eldest of his three brothers who had mild eczema in infancy had received similar treatment. Conventional treatment was repeatedly declined by the family, apart from a one week hospital admission at 4 months of age with bronchiolitis and eczema. He had received nine different homoeopathic medicines before admission including a six centesimal (10\textsuperscript{-12}) dilution of trace metals (iron and arsenic).

Biochemical investigations showed low plasma and urinary sodium concentrations of 121 mmol/l and 10 mmol/l respectively and a low plasma albumin of 11 g/l (no proteinuria). Activities of liver transaminases and alkaline phosphatase and concentrations of plasma urea and potassium were normal.

Treatment was given with 0.45% sodium chloride solution, repeated albumin infusions, and intravenous fluclaxacillin and benzylpenicillin. Intensive local skin care was started with potassium permanganate in short baths. An ointment containing 1% hydrocortisone and 3% cloquinol (Vioform-Hydrocortisone; Ciba) was applied and then changed to one containing 0.0125% flurandrenolone and 3% cloquinol (Haelan C; Dista) applied twice daily to affected areas. A cream containing 1% hydrocortisone and 2% miconazole nitrate (Daktacort; Janssen) was applied three times daily to the perineum, and Diprobase cream moisturiser (Kirby-Warrick) was used on all areas of the body every two hours. Skin swabs initially grew \textit{Staphylococcus aureus} and \textit{β} haemolytic \textit{Streptococcus Lancefield} group A. Repeat swabs also grew klebsiella and \textit{Escherichia coli} and thus the antibiotics were changed to ceftazidime and gentamicin.

The weeping, erythema, and oedema of his skin gradually subsided and progress was satisfactory, apart from an episode of rotavirus gastroenteritis. He was discharged after three weeks with normal plasma sodium and albumin concentrations. On review at 10 months of age, he is thriving with mild eczema, which is now maintained on daily baths using an additive containing 5% acetylated wool alcohols and 63.7% liquid paraffin (Olatan Emollient; Stiefel), 1% hydrocortisone ointment, and Diprobase cream.
Dietary management of D-lactic acidosis in short bowel syndrome.

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