

Short reports

Severe lactose intolerance with lactosuria and vomiting

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SUMMARY An infant with lactose intolerance is described. A breast-fed infant developed vomiting at 3 weeks, and became dehydrated. Lactosuria, aminoaciduria, and liver damage were present. A milk-free diet led to rapid recovery. At 6 months a normal diet was well tolerated.

Severe lactose intolerance, first described by Durand,¹ is, unlike congenital alactasia, not usually accompanied by diarrhoea. Though it is a serious disease, which can cause death if untreated, the disorder is transient and on a milk-free diet all symptoms disappear, with the establishment later of normal tolerance to milk, as documented in the following report.

Case report

The patient was the first child of nonconsanguineous parents. The father's uncle has severe diabetes mellitus. The boy was born at term, birthweight 3.5 kg, and was breast fed. In the third week he vomited after nearly every feed. Stools were normal or constipated.

On admission at 7 weeks his weight was 3.63 kg, he was pale and dehydrated; the liver was 1 cm below the costal margin. Urine tests for protein and ketone bodies were weakly positive; urobilinogen and glucose oxidase tests were repeatedly negative, with positive results to Nylander and Fehling tests; no cells or casts. The sugar was identified chromatographically as lactose, with a concentration of 2 g/l (5.85 mmol/l). Aminoaciduria was present, particularly cystathionine. Galactose-1-phosphate uridyl transferase in erythrocytes was normal. The blood findings, serum electrolytes, alkaline phosphatase, urea, blood lipids, acid base status, serum proteins with their electrophoretic fractions, α -1-antitrypsin, and urinary 17-ketosteroids were normal. HBsAg negative. Blood glucose 67 mg/100 ml (3.72 mmol/l), and the oral glucose tolerance test

were normal. Bilirubin was 1.0 mg/100 ml (17.1 μ mol/l), thymol turbidity test was 2.0 units, aspartate transaminase 65, alanine transaminase 135, γ -glutamyltranspeptidase 300, and lactate dehydrogenase 317 U/l with the increased fractions 1, 4, and 5. Optical media were clear, the lenses were transparent, and the fundi normal. On x-ray examination there was free passage through the oesophagus; the duodenogastric reflux could not be demonstrated. A milk-free regimen resulted in disappearance of the vomiting, lactosuria, and aminoaciduria, and in the normalisation of serum transaminases. Cautious and gradual addition of milk from the sixth month was tolerated well, the boy thrived excellently on normal diet. At age 9 months his weight was 8.8 kg.

The infant's severe condition initially did not allow a lactose tolerance test to be carried out, but at age 5 months the test was normal and the assay of lactose in the intestinal mucosa was also normal.

Discussion

This potentially lethal form of lactose intolerance is characterised by vomiting and dehydration, beginning soon after birth. The absence of diarrhoea in most cases differentiates it clinically from congenital lactase deficiency.²⁻³ Lactosuria is the most striking feature, together with aminoaciduria and renal tubular acidosis; there is liver damage with bleeding tendency in some cases. On a lactose-free diet for several weeks or months, milk tolerance becomes normal. Arakawa *et al.*,⁴ and later Berg *et al.*⁵ and Arashima *et al.*,⁶ showed that intestinal lactase activity is normal, with a normal glucose response to lactose loading even in the acute phase of the disease. In the child reported by Berg *et al.*⁵ lactosuria and vomiting disappeared when milk formula was given intraduodenally.

A gastrogenic origin of the disorder with abnormal absorption of disaccharides is likely,⁵ lactose passing through an abnormally permeable gastric mucosa and leading to lactosuria. The defect of gastric mucosa appears to be temporary. It is not known

whether the damage to the liver cell and renal tubule is caused by lactose or by another unknown substance. Lactose is not normally found in the blood and its presence may have toxic effects similar to fructose-1-phosphate in fructose intolerance, and galactose-1-phosphate in galactosaemia.⁶ Russo *et al.*⁷ and Hirashima *et al.*⁸ noted cataracts in children with lactosuria and in some of their relatives.

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Typhoid glomerulonephritis

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SUMMARY 15 patients with typhoid glomerulonephritis were studied and compared with a group of children with poststreptococcal nephritis. Useful criteria distinguishing the two diseases are given. The diseases may present in a similar manner and therefore it is important to remember typhoid as a cause of glomerulonephritis in endemic areas or in patients travelling from endemic areas.

Although both typhoid fever and poststreptococcal glomerulonephritis (PSGN) are common among the underprivileged in South Africa, rarely does the former present as nephritis.¹ The clinical findings of acute PSGN have been well described but those of typhoid nephritis have not. Failure to distinguish one from the other affects management and may determine the outcome. We therefore report 15 cases of typhoid glomerulonephritis comparing them with a matched group of children with PSGN, and put forward criteria helpful in distinguishing the two diseases.

Patients and methods

A study of 15 patients with typhoid glomerulo-

nephritis admitted to King Edward VIII Hospital, Durban, and observed by one or both of us between 1973 and 1978 was carried out. 15 patients matched for race, age, and sex, and admitted to the same hospital in 1978 with PSGN were randomly selected as the comparison group. The age range of patients was 2–13 years. 10 were girls.

A diagnosis of glomerulonephritis was made on the clinical criteria of periorbital and peripheral oedema, oliguria with evidence of hypertension in nearly all cases, together with haematuria, casts, albumin <2 g/l, and leucocyturia in some patients. An antistreptolysin O titre (ASOT) of >200 U/l was used to confirm the diagnosis of poststreptococcal disease. (93% of children with PSGN at this hospital have a positive response to ASOT).²

A diagnosis of typhoid was made as follows: *Salmonella typhi* was cultured from blood in 5 patients, in blood and stools in 2, in blood and urine in 2, in urine alone in one patient, and in stool alone in one patient. The Widal test was positive (*S. typhi* O titre >320) in 9 patients. Six of these 9 patients had negative blood cultures. However, in the presence of pyrexia >39°C, abdominal findings of pain, tenderness and guarding, with constipation or diarrhoea, and an initial leucopenia (absolute neutrophil count