

In the third patient there was an improvement, though the xylose level remained low (0.60 mmol/l).

Discussion

The association between the presence of rotavirus and adenovirus in the lumen of the small intestine and a low blood-xylose suggests strongly that both these viruses can cause small intestinal dysfunction. Utilization of xylose by micro-organisms in the small intestine is not a real possibility; bacterial counts were low and these viruses do not metabolize xylose.

It is tempting to speculate that xylose malabsorption is the result of mucosal damage by the virus (Bishop *et al.*, 1973), but we have no proof of this because jejunal biopsy was not performed. The data indicate the importance of seeking pathogens in the intestinal fluid as well as in stools. We do not know the explanation of the symptoms in those infants where virus was found in the faeces only, but it is possible that the demonstrated viruses were not true pathogens.

This evidence strongly supports the earlier epidemiological studies implicating adenovirus as a causative agent in acute infantile gastroenteritis. The technique of intraduodenal xylose absorption may provide a valuable indicator of intestinal dysfunction in gastroenteritis caused by other agents when intestinal biopsies are considered unethical.

Summary

The absorption of D-xylose infused into the duodenum was assessed in infants with acute gastroenteritis. 1-hour blood-xylose levels were low in 6 patients found to harbour rotavirus in the small intestinal aspirate. Normal levels (>1.26 mmol/l) were obtained in the absence of virus particles in the small intestine in a further 6 patients: in 3 of these adenovirus or rotavirus was recovered from the stools.

Three patients with adenovirus in the small intestinal juice and ill with acute gastroenteritis also had low xylose levels. This finding supports earlier epidemiological studies that adenovirus may be a causative agent of acute infantile gastroenteritis. The association between virus in the small intestine and xylose malabsorption may indicate mucosal damage. Formal proof of this is awaited.

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J. MAVROMICHALIS,* N. EVANS, A. S. MCNEISH,†
A. S. BRYDEN, H. A. DAVIES, and T. H. FLEWETT
*Institute of Child Health Birmingham, and Regional
Virus Laboratory, East Birmingham Hospital.*

Present addresses: *University of Thessaloniki, Greece.
†Dept. of Child Health, Leicester Royal Infirmary, Leicester LE1 5WW.

Correspondence to Dr. N. Evans, Royal Alexandra Hospital for Sick Children, Dyke Road, Brighton, Sussex BN1 3JN.

Hypoglycaemia in congenital adrenal hyperplasia

It is well known that salt-wasting occurs in many children with congenital adrenal hyperplasia (CAH) due to 21-hydroxylase deficiency and paediatricians are familiar with the salt-losing crises which can arise in such patients. Though hypoglycaemia has only occasionally been reported in the disorder (White and Sutton, 1951; Wilkins, 1965) we suspect that this complication is more common than is generally recognized and give the case histories of 2 young children with CAH who became acutely ill as a result of hypoglycaemia.

Case reports

Clinical details of the 2 patients with CAH due to 21-hydroxylase deficiency are summarized in the Table, together with results of relevant investiga-

Table *Clinical and biochemical details*

	Case 1	Case 2
Age (yrs)	1·8	2·5
Sex	M	F
History of salt-loss	Yes	Yes
Family history	No sibs	2 sibs died aged 1 month; necropsy diagnosis of CAH
Glucocorticoid treatment (mg/m ² per 24 h)	Cortisone acetate 30	Hydrocortisone 14
Mineralocorticoid treatment	Percorten 25 mg 1M monthly	Percorten 25 mg 1M monthly
Investigations on admission		
Blood glucose (mmol/l)	1·0	0·3
Plasma Na	132	145
" K	4·9	4·7
" Cl	98	112
" HCO ₃	16	15
" urea	8·3	2·2

Conversion: SI to traditional units—Glucose: 1 mmol/l ≈ 18 mg/100 ml. Na, K, Cl, HCO₃: 1 mmol/l = 1 mEq/l. Urea: 1 mmol/l ≈ 6 mg/100 ml.

tions. Both patients had shown salt-loss in early infancy but appeared to be stabilized on glucocorticoid and mineralocorticoid treatment.

Case 1. He was well until the day of admission to hospital. He did not eat lunch at his nursery school and became drowsy during the afternoon. On arrival at hospital he was barely rousable. Investigations showed hypoglycaemia (Table) with normal plasma electrolytes and he responded rapidly to treatment with intravenous glucose and hydrocortisone.

Case 2. She developed diarrhoea and vomiting and though her cortisol treatment was doubled she had a generalized convulsion 12 hours after the onset of her illness. On admission to hospital she was shocked with peripheral cyanosis and it was assumed that she was in a salt-losing crisis. Treatment with intravenous saline was given while awaiting the results of investigations. These showed profound hypoglycaemia (Table) with normal plasma electrolytes. Intravenous glucose rapidly corrected her hypoglycaemia but she suffered a respiratory arrest and developed a hemiplegia 12 hours after admission. Over the next 5 days she made a full recovery.

Discussion

Hypoglycaemia is a well-recognized complication of hypoadrenal states, such as Addison's disease.

Recent work indicates that it is due to impaired gluconeogenesis and occurs when liver glycogen is depleted. Alanine, which is released from muscle and converted to pyruvate in the liver, is probably the most important substrate for gluconeogenesis (Felig, 1973). This mechanism is dependent on corticosteroids, and Permutt *et al.* (1972) have shown that plasma alanine is reduced during fasting in untreated patients with Sheehan's syndrome but returns to normal with glucocorticoid therapy.

In congenital adrenal hyperplasia, deficiency of the enzyme 21-hydroxylase leads to impaired cortisol biosynthesis, but hypoglycaemia has seldom been reported in the condition. Wilkins (1965) noted a tendency to hypoglycaemia in only 2 out of 140 patients with CAH. Our experience suggests that this complication is more common than is reported in the literature. Another of our patients with CAH, aged 1·2 years, became drowsy, limp, and grey during an attack of tonsillitis. Her mother, a nurse, gave her hydrocortisone by injection and sweetened drinks while travelling to hospital. On arrival, she had recovered completely. No investigations were carried out but her rapid response to treatment suggests that her symptoms were due to hypoglycaemia. All our patients had severe CAH with salt-loss and while such cases may be more prone to hypoglycaemia, the child described by White and Sutton (1951) was not a salt-loser.

It is important that those involved in the care of children with CAH are aware of this potentially lethal complication and appreciate that emergency treatment with intravenous glucose may be necessary in a patient with the disorder who suddenly or unexpectedly collapses. Treatment with glucagon injection is unlikely to be effective as hepatic glycogen reserves will almost certainly be depleted in such a patient. Emergency treatment with intravenous glucose may not always be possible and in this situation an injection of hydrocortisone (50–100 mg) should be given without delay *before* arrangements are made to transfer the child to hospital. We recommended that the parents of a young child with salt-losing CAH should keep an emergency supply of hydrocortisone injection at home for use by their general practitioner (or the parents themselves) during any illness associated with repeated vomiting or increasing drowsiness.

Summary

Two young children with salt-losing congenital adrenal hyperplasia developed profound hypoglycaemia. In one child hypoglycaemia occurred after a prolonged fast and in the other it was precipitated by infection. This complication may be

more common than the literature suggests, and emergency treatment with glucose or hydrocortisone, given by injection, should be given to any child with the disorder who suddenly or unexpectedly collapses.

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J. MACKINNON and D. B. GRANT

The Hospital for Sick Children, Great Ormond Street, London WC1N 3JH.

Correspondence to Dr. D. B. Grant.

Addendum

Since the preparation of this report, Case 2 has developed recurrent convulsions which are not associated with hypoglycaemia and are almost certainly related to brain damage sustained at the time of her hypoglycaemic episode.

Gastroschisis treated with lyophilized dura

At the Royal Alexandra Hospital for Children, Sydney, it has usually been possible to manage babies with gastroschisis by reduction and primary suture after skin mobilization, muscle stretching, and transverse muscle division as recommended by Savage and Davey (1971). Our results with covering patches or pouches of silastic sheeting as described by Schuster (1967) have not been satisfactory. There has been unacceptable loss of peritoneal fluid at the edge of the silastic sac and necrosis of bowel has occurred under the silastic sheeting. We report 2 patients who demonstrate these complications and advantages of the use of lyophilized dura to replace the silastic sheeting.

Case reports

Case 1. A female weighing 1.6 kg at birth was admitted at 2 hours of age with a typical gastroschisis. There was an abdominal wall defect to the

right of the umbilicus through which prolapsed the small and large bowel, stomach, and a small amount of spleen. Operation was performed urgently and an attempt was made to replace the abdominal contents by extensive mobilization of skin and division of muscle to gain skin cover for primary suture. However, ventilation was difficult and inadequate even before suturing was started. The defect was therefore bridged and closed with a pouch of silicone sheeting and a gastrostomy was established. Over the next 2 days there was considerable fluid loss around the edges of the silicone pouch, and so on the third day of life the silicone sheeting was removed. There were some pin-head sized areas of dubious blood supply on the exposed gut. This gut was disturbed as little as possible and a patch of lyophilized dura was stitched to the under-surface of the skin at the edge of the defect using mattress sutures. A central venous catheter was also inserted through the right internal jugular vein for total parenteral nutrition. Postoperatively the exposed dura was painted daily with mercurochrome solution.

After 10 days a small discharge appeared at the right side of the dural patch which had mummified and lifted off the underlying tissues. The exposed dura was therefore excised without anaesthesia. An enteric fistula was present but the rest of the exposed bowel was covered by a layer of healthy granulation tissue. Peripherally the dura was left intact where it had become vascularized to seal off the peritoneal cavity. Swabs taken from the exposed area grew no organisms on culture. The skin edge was then painted with mercurochrome solution and silver sulphadiazine cream was smeared over the exposed granulation tissue. Total parenteral nutrition was continued because of the fistula in the bowel. After 4 weeks the defect had closed except for the fistula. At 2 months of age the abdomen was explored and a length of 8 cm of atretic small bowel with adjacent fistulae was excised and anastomosis performed to restore intestinal continuity. Apparently the fistulae were precipitated by the atretic bowel. Subsequently the child was weaned gradually off intravenous feeding which had continued for 107 days. The child at 3 years of life is developing normally. The muscle layer of the abdomen appears to have closed as no defect edge can be felt and the abdominal contour is regular.

Case 2. A male infant with a birthweight of 2.4 kg was admitted on the day of birth under the care of a colleague who unsuccessfully attempted to close the gastroschisis defect after skin mobilization and muscle division. A silastic sac was therefore attached to the edge of the defect. The child's postoperative