One-hour blood-xylose in cystic fibrosis

The 1-hour blood-xylose test was sometimes performed (Schaad et al., 1975, 1978) in children with cystic fibrosis (CF) during initial diagnostic procedures. We noticed that in these patients the 1-hour blood-xylose values tended to be higher than in age-matched controls.

Rolles et al. (1973) stated that in a group of 21 children with CF the mean of the blood-xylose values was higher than in controls.

Although we do not believe that this test helps in the diagnosis of CF we reinvestigated this phenomenon in the present study.

Materials and methods

The 1-hour blood-xylose values of 26 patients with proved CF were compared with those obtained in a control group comprising 102 children of similar ages and weights. In the control group the test was done because malabsorption was suspected but in all these patients a morphologically normal jejunal biopsy was found and all other tests for small intestinal absorptive function including fat balance were normal. In patients with CF pulmonary symptoms were present and each exhibited some degree of pancreatic insufficiency with steatorrhoea. The diagnosis was confirmed by using the sweat test. The 1-hour blood-xylose test was performed according to Rolles et al. (1973). 5 g D-xylose in 100 ml water was given to the patient in the fasted state and exactly one hour later the venous blood sample was drawn. D-xylose was measured by the method of Roe and Rice (1948) as modified by Colombo (1978).

Results and discussion

The Table shows that there is a statistically significant difference between mean 1-hour blood-xylose values in the two groups and that patients with CF had higher blood-xylose concentrations.

Because individual values overlapped the test is of no diagnostic use for identifying children with CF.

We cannot explain the increased 1-hour blood-xylose values in patients with CF. We suspect however that this phenomenon could be owing to decreased metabolism of xylose in the liver or to poor urinary clearance. The increased permeability of the small intestinal mucosa to certain sugars suggested by Gibbons (1969) is another explanation. We believe that further investigations of the small intestinal function in CF would be of interest.

Within the CF group we found no correlation of the 1-hour blood-xylose concentration with weight, age, or severity of pulmonary symptoms. The possibility that the high 1-hour blood-xylose values may be caused by hypovolaemia is unlikely since plasma volume was measured in several children with CF and it was not different from that in age-matched controls (O. Oetiker, personal communication 1976).

Summary

One-hour blood-xylose concentrations after an oral xylose load were measured in children with cystic fibrosis (CF) and healthy controls. The mean of the 1-hour blood-xylose values was significantly increased in the group with CF. The finding confirms an earlier observation by Rolles et al. (1973). Its significance is not at present understood but it suggests that small intestinal function should be further investigated in CF.

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References


Theophylline toxicity in a neonate

Theophylline and its derivatives have been widely used in the successful management of recurrent apnoeic attacks in preterm infants (Kuzemko and Paala, 1973; Shannon et al., 1975). Recently there have been reports of less beneficial metabolic sequelae of theophylline administration. Nobel and Light (1977) reported a diuresis in such an infant, theophylline infusion significantly raised blood glucose in 2 infants (Aranda and Dupont, 1976), and small infants may metabolise theophylline less efficiently than do adults (Aranda et al., 1976). We report a pair of twins, one of whom developed hyperglycaemia and glycosuria while receiving theophylline; these abnormalities were corrected within 48 hours of stopping theophylline. The other twin also received theophylline, but at a smaller dose, and did not develop hyperglycaemia or glycosuria.

Case histories

Twins were born to a 24-year-old Caucasian primigravida. Pregnancy was uncomplicated but spontaneous onset of labour occurred at 30 weeks' gestation and delivery of both twins was normal. Both twins were transferred to this unit from a referral hospital at 36 hours.

Twin 1 was a girl weighing 880 g. Apgar scores were 9 and 10 at 1 and 5 minutes respectively. Short apnoeic attacks occurred on the first day and were initially treated by stimulation and increased ambient O₂ concentration. Treatment with oral choline theophyllinate at 2 mg/kg per day was begun on the 2nd day and a maximum plasma level of 4 μg/ml one hour after the dose was recorded.

No increase in dosage was necessary and no apnoeic attacks took place after the 7th day. Theophylline treatment was discontinued on the 28th day. No hyperglycaemia or glycosuria was recorded. The only metabolic abnormality was a nonrespiratory acidosis treated with oral sodium bicarbonate.

Twin 2 was a boy weighing 880 g; Apgar scores were 7 and 10 at 1 and 5 minutes respectively. Early symptoms of respiratory distress developed and despite ambient O₂ concentration of 40–50%, recurrent apnoeic attacks occurred after 15 hours. At 30 hours intubation and intermittent positive pressure ventilation (IPPV) were required. The initial clinical and x-ray findings were compatible with hyaline membrane disease and IPPV was continued for 3 days; this was followed by a further 9 days of continuous positive airways pressure (CPAP). Apnoeic attacks initially resolved by the 10th day but were again present by the 20th day. On this occasion they were associated with copious pharyngeal and tracheal secretions and radiological evidence of pulmonary consolidation. Serratia marcescens and Escherichia coli were grown from tracheal aspirates on separate occasions and appropriate treatment was given. An enterobacter septicaemia occurred on the 14th day and was treated with a 7-day course of flucloxacillin and gentamicin.

IPPV was again necessary from day 27 to day 34 followed by a period of nasal CPAP. Apnoeic attacks were not recorded after day 42. Oral choline theophyllinate was given from the 3rd day at an initial dosage of 2 mg/kg per day. Because of inadequate control of apnoeic attacks despite the intermittent use of CPAP the dosage was increased by 1 mg/kg per day at approximately weekly intervals. Plasma levels were checked at similar intervals one hour after the dose and maximum plasma levels of 4 μg/ml were obtained. Hyperglycaemia and glycosuria occurred on two occasions (Figure). The first episode was related to the use of 10% dextrose IV in addition to a small dose of theophylline. During the second episode on the 34th day the infant was clear of infection and receiving 230 ml/kg of expressed breast milk daily. Maximum urine sugar concentration was 1% and blood glucose 15 mmol/l (270 mg/100 ml). On this occasion the abnormalities were corrected within 36 hours of stopping theophylline. A metabolic acidosis, responsive to bicarbonate, and hyponatraemia were also recorded. The hyponatraemia resolved with the introduction of sodium supplements, but choline theophyllinate was also discontinued at this time.

The infant was discharged to the referral hospital on the 45th day weighing 1·56 kg.

Discussion

Choline theophyllinate has been found of value in the management of apnoea of prematurity (Kuzemko and Paala, 1973; Shannon et al., 1975), but recent reports have suggested that it is not without risk (Aranda and Dupont, 1976; Aranda et al., 1976;