Summary

An 11-year-old twin girl was admitted to hospital with a 24-hour history of profuse sweating, tachycardia, and hypertension after a single dose of imipramine. She was subsequently found to have a right adrenal pheochromocytoma. To our knowledge, this is the first reported case of tumour provocation by imipramine.

We thank Dr H. Simpson for his permission to report this case.

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Sucrase-isomaltase deficiency

A follow-up report

Sucrase-isomaltase deficiency is an autosomal recessive condition which usually presents in infancy with severe diarrhoea when sucrose is introduced into the diet. Sucrase deficiency was first described by Weijers *et al.* (1960); subsequently Anderson *et al.* (1962), using a qualitative method, reported deficiency of sucrase and isomaltase activities in duodenal biopsies of affected children. Burgess *et al.* (1964) reported 7 cases with quantitative estimations of mucosal disaccharidase activities. Four of these children were traced 10 years later and their progress is reported together with the progress of 5 children diagnosed more recently.

Patients and methods

All 9 children had presented with a typical history of chronic diarrhoea in infancy. In most cases this was severe and was associated with poor weight gain. The initial diagnosis of sucrase-isomaltase deficiency had been based on the following criteria. (1) A 'flat' sucrose tolerance test—i.e. after an oral sucrose load of 2g/kg the rise in blood glucose was <20 mg/100 ml (1.1 mmol/l). (2) A normal lactose and/or glucose tolerance test. (3) Histologically normal small intestinal biopsy. (4) Very low sucrase and palatinase activities while lactase was normal. Disaccharidases were measured by the method of Burgess *et al.* (1964). Palatinase was used as the substrate instead of isomaltase, as it is also hydrolysed by isomaltase.

In Case 9, the diagnosis was made at 6 weeks. It was based on carbohydrate tolerance tests and confirmed later by biopsy.

None of the children described by Burgess *et al.* (1964) had been followed up for more than a few months after diagnosis. Four, diagnosed between ages 13 months and 5 years, were seen again when aged between 11 and 16 years. Three could not be traced, 2 because they had left the country and one because he had been adopted.

Case 1 came to our notice because of school truancy and severe diarrhoea. He and his sister, Case 2, thought that sweet food made their symptoms worse but neither they nor their mother understood the implications of this nor knew their original diagnoses. Case 3 presented to the casualty department with episodes of diarrhoea and abdominal pain. Neither parents nor child had associated these symptoms with sugar ingestion. Cases 1 to 4 had repeat sucrose tolerance tests and Case 3 a repeat biopsy.

Five other children aged 6 weeks to 3 years at diagnosis were followed up until aged 3 to 9 years. Case 9 had a repeat sucrose tolerance test.

The symptoms of each child were recorded, a dietary history taken, and average daily sucrose and starch intake were calculated. As daily starch intake varied considerably, the figure given is only approximate. When distance from the hospital precluded a patient being seen, the parents completed a detailed questionnaire (Cases 6 and 8).

Results

All 9 children continued to have episodes of diarrhoea, often associated with abdominal pain, discomfort, or distension (Table). Most children had experienced some improvement after the first year of life and were able to eat small amounts of sucrose

Table Mucosal disaccharidase activities, and diet and symptoms at follow-up in 9 children with sucrose-isomaltase deficiency

Case	Age (years)	Disaccharidase activity in mucosa *				Sucrose tolerance test†	Follow-up		Symptoms
		Lactase	Sucrase	Palatinase	Maltase		Diet‡	Sucrose g/day	
1	5 15 ⁷ / ₁₂	18.2	0.48	0.24	6.7	Flat	Unrestricted. Has sugar in tea	50	Frequent severe diarrhoea with pain
2	1 ¹ / ₁₂ 11 ⁷ / ₁₂	19.8	0.24	1.1	46.4	Flat	Unrestricted	30	Episodes of severe diarrhoea
3	1 ² / ₁₂ 11 ⁶ / ₁₂	27.9	0.14	1.7	2.9	Flat	Dislikes sweets		
4	5 16 ⁸ / ₁₂	7.9	0.8	0.6	8.0	Flat	Likes jam	50	Episodes of abdominal pain and vomiting
5	6 ¹ / ₁₂ 4 ⁶ / ₁₂	15.5	0.29	—	13.2	Flat	Unrestricted	60	Occasional diarrhoea, always following high sucrose intake
6	1 ⁶ / ₁₂ 7 ⁸ / ₁₂	6.8	0	1.5	4.8	Flat	Dislikes sweets	20	Diarrhoea with abdominal pain if dietary sucrose less restricted
7	2 ¹⁰ / ₁₂ 8	8.2	0	0.3	4.8	Flat	Unrestricted, avoids sweets	0	Severe diarrhoea and abdominal pain if eats any sucrose
8	2 ¹⁰ / ₁₂ 9 ⁹ / ₁₂	0.5	0.8	0.2	1.3	Flat	Low sucrose, glucose for cooking	3.5	Severe diarrhoea if diet less restricted
9	6 weeks 3	0.3	0.6	0.4	4.7	Flat	Some fresh fruit	10	Diarrhoea if dietary sucrose less restricted
		3.5	0.6	—	4.2	Flat	Avoids pasta	20	Diarrhoea if dietary sucrose less restricted
						Flat	Very low sucrose		
						Flat	Glucose for cooking		
						Flat	Low sucrose, no added sugar		
						Flat	Low sucrose, no added sugar, some fresh fruit		

*Expressed as μmol of substrate hydrolysed/g mucosa per min. Normal range—lactase 0.2–16.2; sucrase 1.3–20.2; palatinase 1.3–6.9; maltase 3.2–77.†Flat = rise in blood glucose $< 20 \text{ mg}/100 \text{ ml}$ (1.1 mmol/l).

‡Diet when first assessed at follow-up.

without upset. Case 4 had noticed further improvement in his sucrose tolerance at about age 10 years. Cases 6 and 7 were still on very low sucrose diets (0 and 3.5 g daily) and rapidly developed diarrhoea if they did not adhere strictly to their diets. Three children (Cases 1, 2, and 3) were eating an unrestricted diet with 30 to 50 g/day of sucrose but diarrhoea was causing concern. They showed a marked improvement in symptoms when advised to restrict their sucrose intake to 10 to 20 g/day. Case 4 was eating a relatively unrestricted diet and had the least symptoms. He had only occasional bouts of diarrhoea, induced by food with a very high sucrose content.

Starch intake was low to average and restricted only because of association with food containing sucrose. Starch appeared to cause diarrhoea in Case 5 only.

The repeat sucrose tolerance test performed in 5 cases was abnormal in each instance. Sucrase activity was very low in the repeat biopsy in Case 3.

The children were all of normal height and weight although the weight of Case 1 was below that predicted for his height.

Discussion

Completely asymptomatic children with sucrase-isomaltase deficiency have been reported (Kerry and Townley, 1965; Antonowicz *et al.*, 1972). However, in this series it has been our experience that if a patient presents in childhood with symptoms it is likely that these will persist into adolescence and continued restriction of dietary sucrose will be necessary. We, therefore, disagree with the opinion recently expressed in an editorial (*British Medical Journal*, 1977) that most older children tolerate normal amounts of sucrose. Ament *et al.* (1973) found that fruit and vegetables containing more than 2% sucrose caused diarrhoea in their patients. The amount of sucrose tolerated by each of our patients varied and most found their limit by trial and error. Dietary starch rarely caused symptoms, a finding similar to that of other investigators.

Antonowicz *et al.* (1972), reporting cases followed up for 6 years, found that sucrase and palatinase activities in biopsies remained very low, and our findings are in agreement.

It seems advisable that some long-term contact be maintained with children who have sucrase-isomaltase deficiency. This will ensure that each individual as he gets older understands his condition, and dietary advice can be given when necessary. Children lost to follow-up may gradually return to a normal diet and remain unaware of the cause of their diarrhoea.

The marked improvement in sucrose tolerance with age occasionally reported (Auricchio *et al.*, 1963), probably occurs only rarely in the homozygous state but may be more common in heterozygous carriers of the condition who may have subnormal levels of sucrase and isomaltase (Burgess *et al.*, 1964; Kerry and Townley, 1965). The 9 children reported here are considered to be homozygous for the condition as in each case sucrase activity was considerably below normal range.

Summary

Nine children with sucrase-isomaltase deficiency were assessed up to 10 years after diagnosis. All children continued to have episodes of diarrhoea associated with sucrose ingestion. Sucrose tolerance tests showed that malabsorption of sucrose persists into adolescence.

Three older patients were unaware of their condition and were eating normal diets with unrestricted amounts of sucrose. They complained of gastrointestinal symptoms which improved after sucrose restriction.

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