Primary Carcinoma Arising in Normal Liver in a Boy aged 7

J S Kirkham MB (for H H Nixon FRCS)

History: This previously fit boy of healthy family presented with two days of abdominal pain following two months of loss of weight. There was no history of serious illness. His general health was good. A punch biopsy of his palpably enlarged liver suggested hepatoblastoma or malignant hepatoma, and he was transferred to the Hospital for Sick Children, Great Ormond Street.
On examination: The only abnormal finding was a grossly enlarged liver with smooth edge and regular surface.
Pre-operative investigations: Chest X-ray clear. Blood count and complete blood chemistry normal.
Operation (1.1.64)*:* Left hemihepatectomy and partial right hemihepatectomy. A pale, firm tumour occupying the bulk of the liver apart from the right half of the right lobe was resected.
Post-operative course: Recovery was uneventful and methotrexate was given, initially 2·5 mg daily later increased to 3·75 mg daily. He has remained well with no evidence of recurrence, and with normal liver function tests except for moderately raised serum transaminase levels.
Pathology: The tumour had not penetrated the liver capsule but had extended along portal tract blood vessels. It consisted of pleomorphic cells with some characteristics of liver parenchyma, but no embryonic mesenchyme. The diagnosis was pleomorphic hepatic carcinoma. Apart from the tumour the liver was normal. Fig 1 shows the deeply staining pleomorphic tumour cells apparently bursting through a capsule into normal liver tissue.

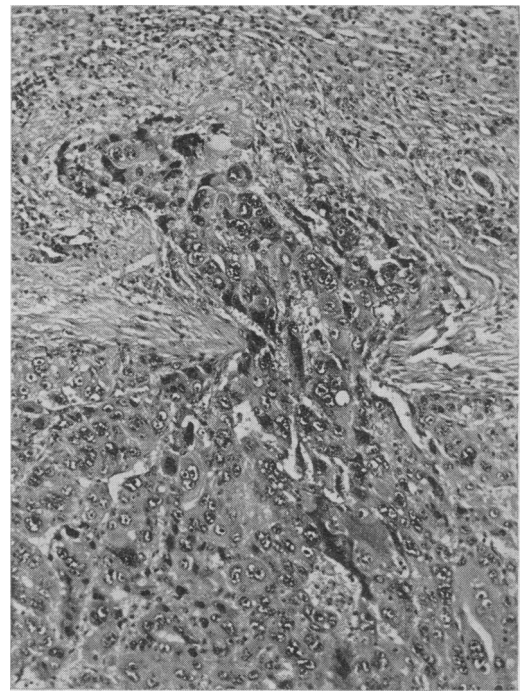

Fig 1 *The darkly staining pleomorphic tumour cells in the lower zone appear to be bursting through the pale capsule into normal liver tissue.* ×95

Comment
The presentation of such a tumour raises certain points of interest:
(1) Large advanced liver tumours in children may often be operable, though assessment of metastasis or the likelihood of recurrence is even more difficult with primary carcinoma than with hepatoblastoma, owing to its even greater rarity.
(2) Partial resection of the liver is complicated anatomically by the fact that the middle hepatic vein territory includes adjacent parts of both right and left halves of the glissonian system.
(3) In this patient the estimated one-fifth of normal liver bulk remaining has proved adequate.
(4) Though most primary malignant hepatic tumours of childhood are hepatoblastomas divided into three types by Willis (1962), the histological picture in this patient was of an adult type of primary hepatic carcinoma and it has been classed as such, though Willis stresses that no true distinction can be made between a hepatomatous type of hepatoblastoma, and an adult type of primary hepatic carcinoma arising in childhood.

REFERENCE
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Sucrose and Isomaltose Intolerance in Siblings

T M Barratt MB
(for Professor A A Moncrieff CBE MD FRCP)

D J (4 years) and G J (2 years) have recently been investigated for chronic diarrhœa. They are male, and the only children of healthy parents. In both brothers the diarrhœa dated from birth; in the light of later investigations this is rather surprising, for both were breast fed. The stools were described as bulky and foul smelling. The diarrhœa was variable, and was more severe in the younger child, who had socially incapacitating fæcal incontinence. There were no abnormal physical signs, both children being of normal stature.

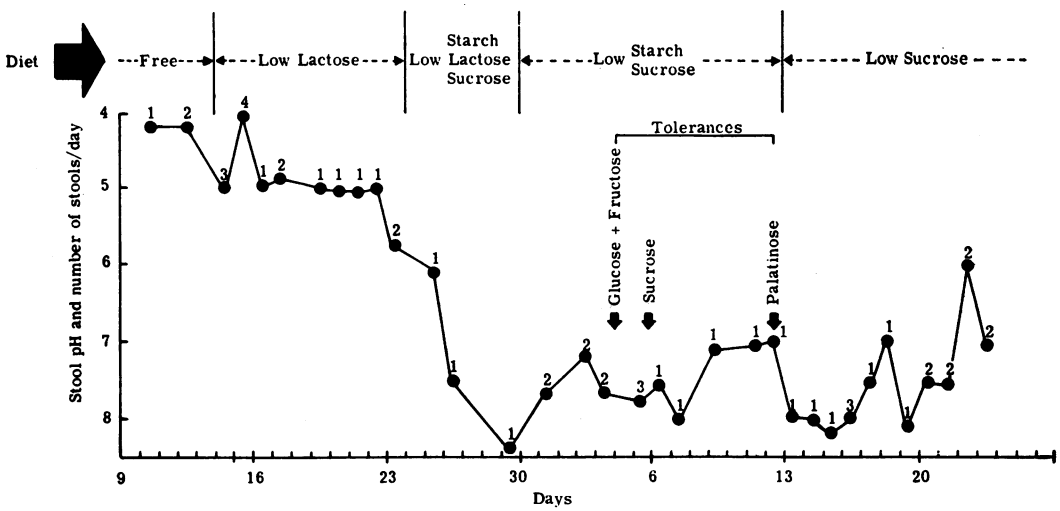

Fig 1 *D J. Stool pH and frequency*

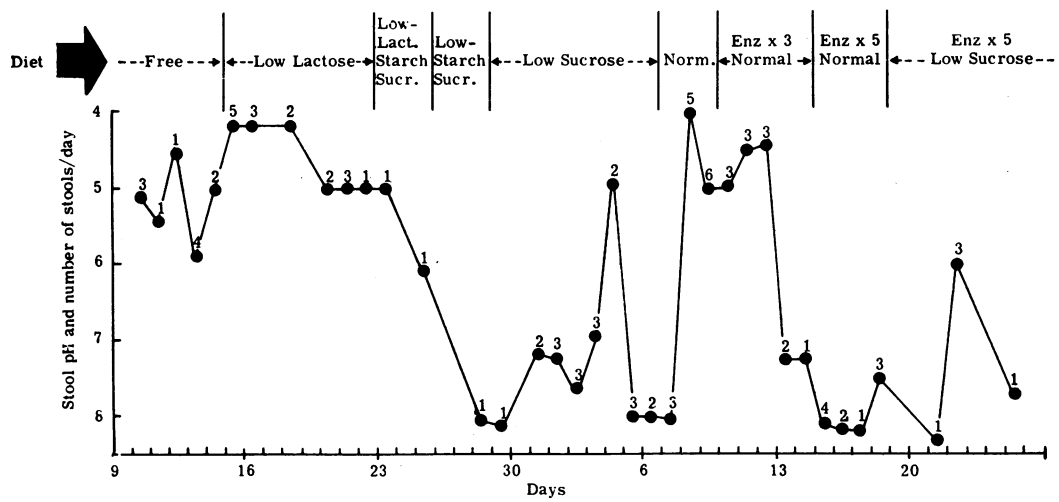

Fig 2 *G J. Stool pH and frequency*

Initial investigations showed a mild steatorrhœa in D J (90% fat absorption), but not in G J (95%). In both, the transit time of a carmine marker was under two hours. Other routine investigations of chronic diarrhœa were uninformative. The twenty-four-hour fæcal lactic acid excretion was more than 1 g in both sibs, greatly in excess of the normal value of 35 mg quoted by Weijers & Van de Kamer (1963). In both, the stool pH was found to fluctuate between 4·0 and 6·0, and this simple investigation, which can be performed by the nursing staff with indicator paper, subsequently proved to be the most useful tool in the unravelling of these cases.

In view of these findings, a disaccharide intolerance was suspected. It is not possible to perform disaccharide tolerances whilst the diarrhœa is in full flood, but the symptoms must be brought under control by the empirical use of various sugar-free diets before the investigations can proceed. Because the diarrhœa dated from birth, alactasia was suspected, but there was no response to a lactose-free diet (*see* Figs 1 and 2).

At this stage, sucrose was identified chromatographically in the stools of D J (but not of G J). Removal of starch and sucrose from the diet produced a prompt remission, and the replacement of lactose in the diet did not cause diarrhœa. Starch was removed for a theoretical reason. Auricchio *et al.* (1961) showed that in cases of sucrose intolerance starch products, particularly isomaltose (which is an end-product of the amylytic degradation of starch), are also tolerated badly. In this dietary-induced remission, carbo-

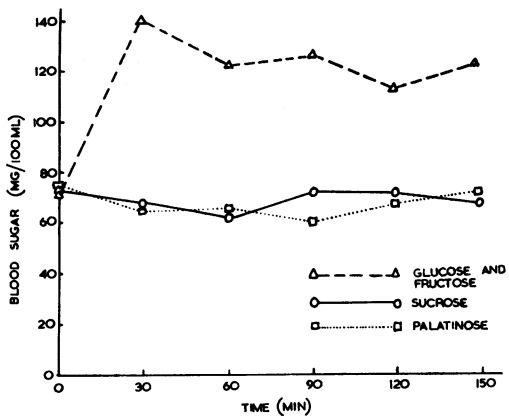

Fig 3 *Carbohydrate tolerances in D J. Dose = 50 g per sq. m surface area. Blood sugar measured as total reducing substances*

hydrate tolerances were performed on D J (Fig 3). Sucrose loading did not affect the blood sugar level, but an equivalent dose of glucose and fructose was followed by a sharp rise. Palatinose is an artificial disaccharide that is a good substrate for isomaltase, and was first used by Auricchio *et al.* (1961) to demonstrate isomaltose intolerance. There was no rise in blood sugar following oral palatinose.

These findings implied a deficiency of sucrase (invertase) and isomaltase; treatment was therefore initiated with a low sucrose diet (5 g/day). D J was symptom free on this regime, but G J still had diarrhœa, and in his case the treatment was supplemented with an oral preparation of sucrase.

In view of the satisfactory therapeutic response, it was decided not to attempt to demonstrate the enzyme defect directly. The disaccharidases are located within the intestinal epithelial cells, and are not found in the succus entericus. Their distribution in human adult small intestinal mucosa has been studied by Dahlqvist (1962). Anderson *et al.* (1963) have developed an elegant technique for the assessment of disaccharidase activity in duodenal biopsy specimens, and have demonstrated the absence of sucrase and isomaltase in two siblings similar to D J and G J.

The one puzzling feature in these sibs is the history of the onset of diarrhœa before any sucrose was present in the diet. Lifshitz & Holman (1964) have recently described a case in which lactase deficiency was apparently associated with that of sucrase and isomaltase. This possibility was not explored, but it seems unlikely that there can now be a significant deficiency of lactase in these boys.

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Auricchio S, Prader A, Mürset G & Witt G (1961) *Helv. pædiat. acta* **16**, 483
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Lifshitz F & Holman G H (1964) *J. Pediat.* **64**, 34
Weijers H A & Van de Kamer J H (1963) *Acta pædiat., Stockh.* **52**, 329

Congenital Multilocular Mesenteric Cyst with Chylous Ascites

A A Cunningham MD

This case was previously shown on November 23, 1962 (*see Proceedings*, 1963, **56**, 295).

A B, born 11.12.60

This boy, handicapped by the large abdomen and recurrent chest infections, has continued to make good progress with conservative treatment. Height 94 cm (average for age 96·5 cm); weight 40 lb 4 oz (average for age 34–35 lb). He talks and behaves in a manner which compares favourably with other children of the same age and he seems to be of average intelligence. The testes are undescended; buccal sex chromatin smear showed the normal male cell pattern.

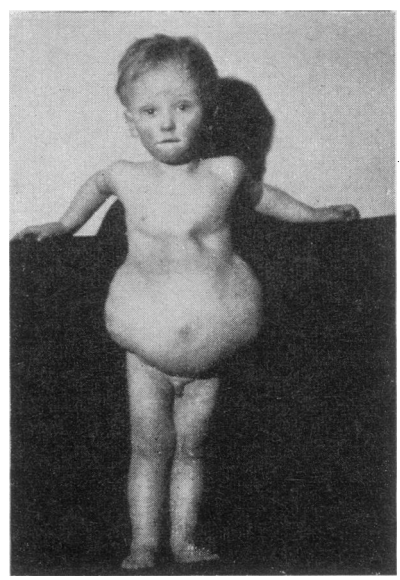

Fig 1

The abdomen with its very thin muscles is pendulous and prominent (Fig 1) and there is a large tumour mass extending transversely but situated mostly on the left side. There is no gut distension and bowel movements are regular without aperients.

The mother now buys large trousers and adapts them neatly to provide abdominal support. The child can walk up to 400 yards at a time and is learning to climb stairs unaided.