antibiotics, but failed to respond. A biopsy was then taken and this was reported as malignant. The wound failed to heal and the mass got bigger. When seen at the Children's Hospital, she had a large sacrococcygeal mass which was hard and irregular. The biopsy site was ulcerating. An irregular pelvic mass could be felt both rectally and abdominally. No attempt at excision was made, and radiotherapy was given with some reduction in the tumour size. Within three weeks of stopping radiotherapy, bilateral pulmonary metastases appeared together with an increase in the size of the pelvic mass. In spite of further radiotherapy and chemotherapy, the child was dead five months after her initial biopsy.

Discussion

The baby born with a large sacrococcygeal teratoma presents no diagnostic problem and, with complete excision, should have an excellent prognosis. Where the lesion is small at birth, diagnosis appears to be difficult, in spite of the presence of a simple skin lesion such as a haemangioma or hairy pigmented naevus situated over the sacrococcygeal area. The importance of a proper rectal examination at this stage cannot be overstressed, in particular careful palpation of the coccyx (Donnellan and Swenson, 1968). It is further noted by Conklin and Abell (1967) that a sacrococcygeal teratoma is a germ cell neoplasm located anterior to the sacrum and coccyx and posterior to the rectum.

Gross, Clatworthy, and Meeker (1951) suggested that sacrococcygeal teratomas might pursue two separate courses: (1) persist as benign encapsulated lesions and grow with the child, or (2) one element of the teratoma during this quiescent period may suddenly become malignant and invade widely; the latter mode of behaviour being commonly shown by the exceedingly small tumours. Evidence suggests that this 'quiescent period' may be anything from a few months to a year. In comparing malignancy rates, Waldhausen et al. (1963) showed this to be 7% for those patients less than 4 months of age, but 42% for those in an age group 4 months to 15 years. They suggest a neonatal malignancy rate of about 4% but it is possible that this is very much less.

The 4 cases of malignant sacrococcygeal teratoma presented here illustrate well the behaviour pattern of the small sacral tumour. They are presented in an attempt to encourage early diagnosis during the 'quiescent period', when surgical intervention might be life saving.

Conclusions and Summary

Any 'birthmark' or lump in the sacral region merits a rectal examination and the consideration of the diagnosis of sacrococcygeal teratoma. The 'quiescent period' in the growth of these tumours makes early diagnosis of vital importance. Four illustrative cases are described.

I wish to thank Mr. H. H. Nixon for permission to report these cases submitted under his care, and also for helpful advice and criticism.

References


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Jejunal Disaccharidase Activities in Children with Marasmus and with Kwashiorkor Response to Treatment

Pathological changes in the intestine in protein-calorie malnutrition have been documented both in humans and in experimental animals. The extent of the functional defects associated with the structural changes varies, but in children it has been shown that lactose intolerance is an important factor in the production of diarrhoea (Bowie, Barbezat, and Hansen, 1967). Defects in glucose absorption and in sucrose hydrolysis have also been documented (James, 1970), and these findings are consistent with the concept of a generalized damage to intestinal structure and function in malnutrition.

Cook (1967) suggested that a primary genetic deficiency of lactase might predispose children to malnutrition, and showed that lactase deficiency persisted in children tested 4 to 10 years after treatment for kwashiorkor (Cook and Lee, 1966).

The present work assesses the incidence of disaccharidase deficiency in Jamaican children with marasmus and with kwashiorkor before and after a relatively prolonged period of treatment in hospital.
Methods

Children, aged 6 to 24 months, with clinical evidence of moderate or severe protein-calorie malnutrition, were admitted to a metabolic ward for investigation. All had a history of inadequate diet for at least 3 months before admission, and most had had recurrent episodes of diarrhoea often unaccompanied by fever or vomiting. Only one child had such severe diarrhoea on admission that he required intravenous therapy. The other children received frequent oral glucose-water with graduated feeds of either a modified milk preparation (Lactogen) with additional sucrose, or a non-lactose formula (Sobee). Children were studied during the first 2 to 3 weeks after admission, when they had received oral feeds containing sucrose and maltose for at least 3 days.

Jejunal biopsies were taken with a paediatric Watson capsule using a technique which simplified and shortened the procedure (James, 1968). Biopsies were only taken when the children were free of oedema.

The position of the biopsy capsule was checked by x-ray with image intensification, and biopsies were taken from the jejunum within 5 cm of the duodeno-jejunal flexure. The biopsy specimen was divided in two, and one half was retained for pathological studies. The remainder was rinsed with ice-cold saline, blotted, and weighed on an electromagnetic balance. The biopsy was then stored in a small sealed container in dry ice until the disaccharidase activity of the specimen was measured. This was done within 24 hours of the time of biopsy.

Disaccharidase activities were measured on the biopsy specimen by the method of Burgess et al. (1964). The hydrolysis of disaccharides has been shown to be linear under the conditions used. The activity of the disaccharidases was expressed in terms of the wet weight of the mucosal tissue. The use of DNA as a more acceptable reference standard was not possible because of the limited material available.

Children remained in hospital for at least 2 months on an optimal calorie and protein intake, and were then retested when they had reached the expected weight for a North American child of the same height (Nelson, 1959). At this stage they had regained their normal body proportions but were still small for their age.

Results

Table I shows that 8 of 19 children when malnourished had jejunal lactase levels below 2 units and 4 of these were less than 1. After these 8 children had been treated in hospital there was a marked increase in lactase activities in 7.

When the lactase levels were very low then the activities of both sucrase and maltase were also decreased in the malnourished children, e.g. the first few children in Table I.

In Table II the disaccharidase activities were grouped according to the clinical diagnosis. The jejunal disaccharidases in those children fulfilling the clinical criteria of marasmus were higher than those malnourished children with oedema. This was not a reflection of dilution of the jejunal enzymes by mucosal oedema for the children were not biopsied until free of oedema.

With treatment there was a much greater increase in sucrose and maltase activities than in lactase, and the increase was seen in both groups of children.

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Marasmus</th>
<th>Lactase</th>
<th>Sucrase</th>
<th>Maltase</th>
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<tr>
<td>M</td>
<td>R</td>
<td>M</td>
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<td>—</td>
<td>4-2</td>
<td>—</td>
<td>7-8</td>
</tr>
</tbody>
</table>

| Mean ± SEM | 2-7 ± 0-7 | 3-6 ± 0-7 | 4-4 ± 0-7 | 9-1 ± 1-4 |
| p | <0-05 | <0-005 | <0-005 |

*Marasmic children denoted by +.
†Gastroenteritis one week before the second biopsy.

| Table II

Disaccharidase Activities of Jejunal Mucosa (umoles disaccharide hydrolysed/min at 37° C per g wet weight) of Malnourished (M) and Recovered Children (R) Grouped According to Clinical Diagnosis on Admission

<table>
<thead>
<tr>
<th>Marasmus</th>
<th>Marasmus-kwashiorkor and Kwashiorkor</th>
</tr>
</thead>
<tbody>
<tr>
<td>M (8)</td>
<td>R (6)</td>
</tr>
<tr>
<td>Lactase</td>
<td>4-1 ± 1-4*</td>
</tr>
<tr>
<td>Sucrase</td>
<td>5-2 ± 1-1</td>
</tr>
<tr>
<td>Maltase</td>
<td>22-0 ± 4-4</td>
</tr>
</tbody>
</table>

*Means ± SEM. Figures in brackets indicate number in each group.
Discussion

It is clear that not all malnourished children have lactase deficiency and that Jamaican children with marasmus are likely to have normal lactase values. A greater number of biopsies is required to substantiate the finding that children with kwashiorkor are particularly prone to develop disaccharidase deficiencies, but the observed differences between the two nutritional groups are in keeping with the more normal appearances of biopsies from marasmic children than from those with kwashiorkor (Brunser et al., 1966).

These findings support the concept that protein depletion per se can produce disaccharidase deficiency, presumably by reducing the supply of amino acids available for enzyme synthesis. Lactase is most severely affected and maltase least, suggesting that a normal level of lactase activity in the jejunum may require much more protein synthesis than does normal maltase activity as the epithelial cells migrate up the jejunal villi.

The malnourished children received a diet containing sucrose and maltose as well as protein for at least 3 days before the biopsies were obtained. This time interval would allow any immediate effects of substrate induction to have occurred. The increase in the sucrase and maltase activities after treatment in hospital is often much more than the 50% increase in the activity of these enzymes which may result after feeding the disaccharides. Protein repletion and villous repair were presumably the main factors producing this large increase in disaccharidase levels.

The increase in lactase activity during treatment though marked in several cases, was smaller than the increase in sucrase and maltase. The slower rate of recovery of lactase was similar to that seen in tropical sprue and in other conditions where there is generalized damage to the intestine.

The increase in the lactase activity with treatment makes it clear that a genetic defect in lactase production cannot account for the incidence of lactose intolerance in these malnourished children of Negroid stock, and the normal level of lactase in the marasmic children is against genetic predisposition to lactase deficiency leading to marasmus in these children.

Most children admitted to hospital with protein-calorie malnutrition remain for a much shorter time than those treated in this series. It seems probable, therefore, that children with protein deficiency and a damaged intestine, if returned early to a nutritionally inadequate environment, may fail to recover normal levels of disaccharidase activities and remain with permanent sugar intolerance especially affecting lactose.

Summary

Eight of 19 malnourished Jamaican children had a jejunal lactase activity below 2 units together with reduced sucrase and maltase levels. Children with kwashiorkor were particularly prone to disaccharidase deficiencies, whereas marasmic children tended to have normal levels.

Treatment in hospital for an average of 3 months produced a marked rise in sucrase and maltase activities in both nutritional groups. 7 of the 8 children with lactase deficiency responded well to treatment with a rise in lactase activity. It was concluded that protein deficiency rather than genetic factors led to lactase deficiency in these children.

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