factors, such as platelets, fibrinogen, factors VII and X, have been described (Dossetor, Gutelius, and Kendall, 1966) as well as a rise and alteration in plasma lipids. In addition, a decrease in fibrinolysis may be present (Cotton, 1967).

Hypovolaemia is a usual finding in the nephrotic syndrome, and may be made worse by diuretic therapy. The increase in haematocrit, the vasoconstriction, and sluggish circulation tend to increase the likelihood of intravascular coagulation. Coagulation is also promoted by *corticosteroid therapy* in both nephrotic and non-nephrotic patients (Ozsoylu, Strauss, and Diamond, 1962). Some coagulation factors are increased, particularly factor VIII. It is not surprising that thrombotic incidents in nephrotic patients were noted, for example by Addis (1949). A curious anomaly has been the relatively frequent observation of pulmonary artery thrombosis in children (Levin, Zamit, and Schman, 1967).

Finally there is little doubt that there is a risk of *trauma* to the femoral artery during attempts at femoral vein puncture. This would be particularly likely to occur in a shocked, nephrotic patient where venoconstriction and oedema would make it difficult to obtain a sample. Goldbloom, Hillman, and Santulli (1967) reported three nephrotic children receiving corticosteroids in whom femoral vein puncture was followed by thrombosis of the femoral artery and amputation, with death in one case. Femoral artery thrombosis has also been reported after femoral vein puncture in non-nephrotic infants (Nabseth and Jones, 1963). It is difficult to assess the amount of risk; thousands of femoral vein punctures are performed in children every year without incident (McKay, 1966), and probably some dozens of these are on nephrotic children. However, spontaneous arterial thrombotic episodes in children who do not suffer from embolism are extremely rare. It seems reasonable to us that femoral vein puncture should be avoided in nephrotic children unless the sample is vital and cannot be obtained elsewhere.

In view of the good results obtained with cyclophosphamide in children with the 'minimal change' lesion and the nephrotic syndrome (Moncrieff *et al.*, 1969), it seems reasonable to add thrombotic episodes to the indications for transferring patients from corticosteroid therapy to this drug. Our patient had, in any case, severe steroid toxicity and increasing resistance to treatment.

Summary

A 2½-year-old girl with the nephrotic syndrome is described. During a relapse she developed

femoral artery thrombosis leading to a through knee amputation. At the time of the incident the patient was hypotensive with a raised blood urea, was receiving corticosteroids, and had bilateral femoral vein punctures to obtain blood for electrolytes. All these factors could have contributed. We conclude that femoral vein puncture should be avoided where possible in patients with the nephrotic syndrome, and that thromboembolic incidents are an indication for the use of cyclophosphamide in these patients.

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Malignant Sacrococcygeal Teratoma—A Problem in Diagnosis

Sacrococcygeal teratoma occurs in about 1 in 40,000 births (Gelb *et al.*, 1964), and presents at birth as a sacrococcygeal mass which may vary considerably in size. It is predominantly a female condition. Failure to recognize and treat adequately surgically at this time will result in a fatal outcome in what should be primarily a benign condition.

Berry, Keeling, and Hilton (1969) reported 58 sacrococcygeal teratomas seen at The Hospital for Sick Children, Great Ormond Street, London, over a 35-year period up to 1969. 10 of these were or

became malignant, an overall incidence of 17%. 8 of these received delayed treatment, the remainder had incomplete excision.

In the first half of 1970, a further four malignant sacrococcygeal teratomas have been treated. All came late to operation and all died. These 4 cases are now reported in detail with special reference to their early history.

Case Reports

Case 1. A female Indian child was noted at birth to have a pigmented skin lesion overlying her sacrum. This lesion was observed at regular intervals and noted to be growing with the child. There was no note of a rectal examination having been performed. She was referred to The Children's Hospital at the age of 1 year for a neurological opinion as a case of spinal dysraphism because of difficulty in walking, associated with a large swelling, covered with hairy pigmented skin, overlying the sacrum and extending into the left buttock. There was anterior displacement and rotation of the anus with a postanal dimple (Fig.). The pigmented area was firmly fixed to the underlying mass which could be felt surrounding the anorectal canal and extending into the pelvis. At laparotomy, large necrotic left iliac nodes were removed, and the mass bulging through the pelvic floor was mobilized. The main sacral mass, which was poorly encapsulated, was removed with difficulty. Pathology reported a malignant papillary growth resembling choroid plexus. In spite of radiotherapy, the child developed ascites with liver secondaries, and she was dead 3 months after operation.

FIG.—Case 1, showing swelling of left buttock and sacral area, which is covered by an area of hairy pigmented skin.

Case 2. This child was referred to The Children's Hospital at the age of 20 months from another hospital where she had been attending with a sacral birthmark. From 3 months, this had been associated with a distinct swelling which had been noted on rectal examination. In the 4 months before admission, this swelling had got much bigger and she had developed a limp and altered bowel habit. Appearances were similar to the first case, except that the skin, which contained numerous dilated veins, was fixed over a wider area. X-rays of the chest and spine were negative, but the sacrum was displaced by a large soft tissue mass. At operation the tumour was found to be widely infiltrating, and complete removal was impossible. Pathology reported a malignant tumour composed of polymorphic cells arranged in sheets and tubules. Three weeks after operation, she developed secondary glandular involvement in both inguinal regions. These glands subsided with radiotherapy, but she gradually developed ascites and was dead almost three months after operation, with liver secondaries.

Case 3. A 4-year-old girl had been seen at the age of 2 because of a fall in which she had injured her sacrum. At that time, a sacrococcygeal swelling was noted, but was said to have resolved spontaneously. There was no record of a rectal examination having

been performed. She was referred from another hospital where she had presented with a large sacrococcygeal mass. A biopsy had been taken and reported as a myxofibromatous tumour, probably malignant. She had a very large tumour, occupying the right buttock and sacral area. The skin was firmly fixed and irregular, with areas of increased vascularity. The anorectal canal was a rigid tube surrounded by tumour. A chest x-ray was clear. At operation, total excision was impossible as the tumour was densely adherent to rectum and infiltrating the glutei. Pathology confirmed a malignant lesion composed of sheets of undifferentiated cells with necrotic areas and foci of atypical bone. Radiotherapy was given postoperatively. Two months later she developed a fungating mass at the site of excision. Further radiotherapy and chemotherapy was given, but the child developed liver and lung metastases and died.

Case 4. Presented at the age of 2½ years with the story that she had a sacral swelling from the age of 2 months. The parents had watched the swelling grow with the child. She had been seen and examined at 3 months but no treatment was advocated at that time. She was referred from another hospital, where she had presented with a large hot painful swelling of her left buttock. This was treated initially as an abscess with

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.

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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.