Discussion

Intramural haematoma may be subserosal, intramucosal, or submucosal, or may involve the entire thickness of the bowel wall as in this case. There was no reason to believe that the haematoma was secondary to a haematological disorder and investigations for a bleeding or a coagulation defect proved to be negative. The haematoma may have been secondary to injury to blood vessels by direct trauma or by a shearing stress, as in patients with non-penetrating abdominal trauma (Bailey and Akers, 1965).

Direct injury to a blood vessel seems an unlikely cause in this case. Histological examination of the biopsy specimen did not reveal any large vessel, as has been reported by McDonald (1966). One would also expect the bleeding to occur into the lumen of the bowel, rather than intramurally, in such circumstances.

A shearing stress, with indirect injury to blood vessels, seems more likely. There had been considerable delay before the capsule passed from the duodenum to the jejunum. This can be attributed to the malrotation, which may also have caused some hindrance to withdrawal of the capsule back into the duodenum. It is conceivable that during withdrawal the jejunum may have been sheared, with tearing of minute vessels in its wall, with extravasation and formation of the haematoma.

The clinical picture in this case was typical of intramural haematoma, with signs of jejunal obstruction, an upper abdominal mass, and a delay of 48 hours between injury and onset of symptoms. Though conservative non-operative treatment has occasionally been employed in patients with external abdominal trauma (Bailey and Akers, 1965), Stewart *et al.* (1970) recommend exploratory laparotomy in most cases.

While intestinal biopsy has rendered radiological examination of the alimentary tract in true coeliac disease unnecessary, Anderson (1966) suggests that barium studies should be performed in the atypical case to exclude abnormalities such as malrotation which may occasionally be responsible for the symptoms. Leslie and Matheson (1965) described 4 cases of failure to thrive which they considered secondary to intestinal malrotation. It appears that if intestinal malrotation is demonstrated radiologically, it should be treated surgically and must be considered a contraindication to peroral biopsy.

Finally Noonan's syndrome is usually associated with multiple extracardiac anomalies (Noonan, 1968), but the association with an intestinal anomaly such as malrotation has not been reported previously.

Summary

An intramural jejunal haematoma developed after peroral biopsy in a 4-month-old baby who had failed to thrive, and who had malrotation of the intestines. Surgical treatment of the haematoma was successful.

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Aetiology of Transient Neonatal Diabetes

In 1970 Ferguson and Milner reported the occurrence of transient neonatal diabetes mellitus in two male sibs and discussed the possible roles of insulin resistance and insulinopenia in the pathogenesis of the condition. The mother of these boys became pregnant again and the opportunity was taken to study the third infant prospectively to see if it would develop transient diabetes and if so to investigate further the cause of this condition.

Case Report

A male infant was born on 20 June 1970 at 40 weeks' gestation, birthweight 2040 g. The mother, aged 27 years, was slightly obese but had a normal pregnancy, apart from low urinary oestriol excretions: 4.0 mg/24 hr (28 weeks), 6.8 mg/24 hr (32 weeks), 8.3 mg/24 hr (36 weeks), 7.9 mg/24 hr (38 weeks), 9.3 mg/24 hr (39 weeks), 6.1 mg/24 hr (40 weeks). Labour was induced at term and fetal distress (slowing of the fetal heart and meconium-stained liquor) resulted in a low forceps delivery. The Apgar score at 1 minute was 6, and regular respirations were established by 6 minutes. Physical examination revealed no abnormality apart from loose wrinkled skin which was meconium stained.

The placenta weighed 500 g and had diffuse areas of calcification.

Maternal and umbilical blood specimens were collected at delivery. All urine was collected for the first 6 days of life and was stored immediately at 4 °C until 24-hour collections were complete. The volume was then measured and an aliquot stored at -20 °C. At the age of 24 hours an intravenous glucose tolerance test was performed by injecting glucose 0.5 g/kg via the umbilical vein and collecting blood samples before and at 2, 10, 30, and 60 minutes after the injection. All blood samples were heparinized. Plasma was separated immediately and stored at -20 °C until the glucose concentration was determined by a glucose oxidase method (Trinder, 1969) and the insulin concentration by immunoassay (Hales and Randle, 1963). The pH of the urine samples was adjusted to 7.4 and the osmolality to 280 mOsm/litre before urinary insulin concentrations were determined. Urinary creatinine was measured by the method of Bonsnes and Taussky (1945).

The maternal plasma glucose and insulin concentrations at the time of delivery were 138 mg/100 ml and 35 μ U/ml. The umbilical cord plasma glucose was 128 mg/100 ml and plasma insulin, 17 μ U/ml. When the baby was 24 hours old the plasma glucose had risen to 260 mg/100 ml, but the plasma insulin level was < 6 μ U/ml. There was no response in plasma insulin levels to the intravenous glucose challenge (Table I).

TABLE I

Plasma Glucose and Insulin Levels during Intravenous Glucose Tolerance Test

Time (min)	Plasma Glucose (mg/100 ml)	Plasma Insulin (μ U/ml)
0	260	< 6
2	540	8
10	374	6
30	358	< 6
60	440	6

Despite the hyperglycaemia, glucosuria was not noted until the third day. Treatment with soluble insulin 2U/day in divided doses began on the fourth day of life. The urinary insulin excretion for the first 6 days of life is presented in Table II. On days 1 and 3 insulin was not detected in the urine and the excretion of endogenous insulin during the first 3 days of life, expressed as a function of the creatinine excretion, could not have been greater than low normal by the criteria of Lowy and Schiff (1968). Thereafter insulin excretion increased markedly due to insulin therapy.

During the early weeks of life the baby developed a mild gastroenteritis associated with *Esch. coli* O55 in the stools, which delayed hospital discharge. During this time insulin requirements rose to a maximum of 10U/day but fell with recovery from the gastroenteritis. He was discharged from hospital on 4U soluble insulin daily aged 3 months. Insulin therapy was discontinued

TABLE II

Urinary Insulin Excretion Before and After Insulin: Treatment Started on Day 4

Day of Life	Urine Volume (ml)	Insulin (μ U/day)	Creatinine (mg/day)	Insulin/Creatinine (μ U/mg)
1	45	< 113	21.6	< 5.2
2	128	96	25.6	3.7
3	162	< 81	22.5	< 3.6
4	142	582	17.1	33.8
5	345	300	15.2	19.7
6	222	< 111	8.9	< 12.5

at 4½ months and glucosuria finally disappeared at age 5 months. After discharge from hospital he thrived and developed normally and when last seen aged 7 months he weighed 6.53 kg.

Discussion

The birth of a third infant to a woman who had previously born two sons suffering from transient neonatal diabetes mellitus provided a unique opportunity to study the postnatal course of the disease prospectively. Two explanations have been offered for this condition: insulin deficiency or insulin resistance. The evidence from the present study confirmed the viewpoint expressed previously (Ferguson and Milner, 1970) that transient neonatal diabetes is due to delay in functional maturation of the fetal β -cell.

At the age of 24 hours the baby had plasma insulin levels at the lower limit of detection, despite being hyperglycaemic. No rise in plasma insulin occurred in response to a glucose challenge. Urinary insulin excretion was within or below the low normal range during the first three days of life. Once insulin therapy started, the urinary insulin excretion rose considerably and became similar to that of normal newborn infants on the fifth day (Lowy and Schiff, 1968). The urinary creatinine excretion fell from day 4 to 6, raising the possibility of incomplete collections at this time. If the collections were incomplete the urinary insulin excretion was underestimated.

If transient neonatal diabetes were due to insulin resistance, increased insulin secretion would be expected and thereby increased urinary insulin excretion. Since the infant failed to respond to intravenous glucose with a rise in plasma insulin levels and urinary excretion was low, deficient insulin secretion was held to be responsible. In considering temporary insulinopenia as a mechanism for the pathogenesis of neonatal diabetes, it must be remembered that the human fetal β -cell is unresponsive to glucose when a variety of other stimuli

cause insulin release *in vitro* (Milner, Ashworth, and Barson, 1971). Infants with transient diabetes may have β -cells which are unresponsive to all stimuli or which, as in the fetus, are unresponsive to glucose alone. In the former case, the cell might be morphologically immature but in the latter case morphological normality would be expected.

Summary

An infant, whose two elder brothers had suffered from transient neonatal diabetes mellitus, developed the same condition. An intravenous glucose tolerance test at the age of 24 hours was diabetic and caused no rise in plasma insulin levels. Urinary insulin excretion during the first three days of life, before insulin therapy began, was within or below the low normal range. These findings support the view that transient neonatal diabetes is due to a delay in β -cell maturation.

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Transient Metastatic Calcification Complicating Renal Failure in an Infant

Ectopic calcification is a well-recognized complication of chronic renal insufficiency in adults (Black, 1967), whereas in infancy it is rare. The purpose of this report is to document the occurrence

of transient generalized soft tissue calcification in association with chronic renal insufficiency in a 2-month-old child.

Case History

Male infant, 3.55 kg, born to healthy parents after a normal pregnancy. He was fed on full-cream dried milk. At 2 weeks he was admitted to the local hospital with a history of weight loss and vomiting. *Esch. coli* was isolated from culture of blood and urine. At this time a random serum calcium was 12.0 mg/100 ml. Treatment with kanamycin and cloxacillin was started and his clinical condition improved. An intravenous pyelogram failed to opacify the renal tract when the blood urea was 284 mg/100 ml. No ectopic soft tissue calcification was visible. By the age of 5 weeks he had gained weight, though urine was still infected with *Esch. coli* and the blood urea was 184 mg/100 ml. Treatment was continued at home with nalidixic acid. When 8 weeks old, he was readmitted with a short history of vomiting and twitching and was transferred to this hospital. On admission he was a normal looking infant and his developmental age was in keeping with his chronological age. His weight was below 3rd centile. Systolic blood pressure was 100 mmHg. The genitalia were normal and he had a good urinary stream. A hard intracutaneous nodule was palpable over the anterior aspect of the left tibia (0.5 cm in diameter) and several similar nodules were palpable in the occipital region.

Fig. 1 shows the concentration of blood urea, serum

FIG. 1.—The concentrations of urea, calcium, phosphorus, and alkaline phosphatase between the ages of 2 and 6 months.