tests which assess distal tubular function (Broberger Winberg, and Zetterström, 1960). The disorder may appear at different ages and have variable rates of progression, even within the same family, but it usually takes a rapid course in childhood, while a more protracted course is common in adults (Axelsson and Ödlund, 1968).

The aetiology of nephronophthisis is obscure. Bacterial infection is not considered to play a role in its pathogenesis. It has been postulated (Mongeau and Worthen, 1967) that the tubular damage might be caused by a ‘toxin’ resulting from an inborn metabolic error.

Summary

A sporadic example of an uncommon congenital nephropathy, previously described as medullary cystic disease or familial juvenile nephronophthisis, is reported. The patient, a 4½-year-old boy, had developed polyuria and polydipsia in early infancy followed by manifestations of azotaemic osteodystrophy. He also had a congenital nystagmus. His blood pressure was normal. Investigations revealed vasopressin-resistant hypotension and minimal salt wasting but otherwise normal urinalyses. The renal histology was typical of nephronophthisis.

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Short Reports

Haemagglutination Reaction in Indian Childhood Cirrhosis Indicating Viral Aetiology

Indian childhood cirrhosis or infantile cirrhosis has a special lure for Indian investigators as it is a disease peculiar to Indian children. Its aetiology still remains an enigma. Several possibilities have been postulated, but a viral aetiology is generally favoured. Attempts to identify the hypothetical virus by histopathological (Achar and Raju, 1951) or electron microscopical means or by animal transmission experiments have been inconclusive. Morrison and Hoyt (1957) described a haemagglutination test with greater specificity for viral hepatitis. This was further confirmed by its higher positivity in adult cirrhosis of viral aetiology, as compared to the non-viral group of cirrhosis (Morrison et al., 1961). Their results showed that the application of the haemagglutination tests in Indian childhood cirrhosis could prove to be an indirect method of investigating the relation of viruses with this type of cirrhosis in children.

Material and Method

One hundred cases of infantile cirrhosis and 25 normal controls were investigated. The disease was diagnosed primarily on the clinical features of a firm enlarged liver, splenomegaly, jaundice, and ascites, with other associated features of liver failure; where possible, liver biopsy was done (38 cases). The controls were healthy children belonging to the same age group as the cirrhosis cases; none had suffered from jaundice, or had any family history of cirrhosis; normal liver function tests were recorded in all.

Haemagglutination test. The procedure adopted is a modification of the original Hoyt and Morrison test as described by Morrison et al. (1960). The conventional scoring (+ to +++) was used in the readings.

Observations and analysis. One hundred cases of Indian childhood cirrhosis—87 males and 13 females in the age group 7 months to 4 years—were studied. 7 cases were in the early stage of the disease, 54 presented in the intermediate stage, and 39 in the late stage. According to the clinical course of the disease, 41 cases belonged to the subacute group, 43 to the acute, and 16 were of the fulminant type.

The haemagglutination test was positive in 83 out of the 100 cases as against 2 of the 25 controls (Fig. 1). These results are highly significant. The positivity of the test was directly related to the severity of the clinical course and to the stage of the disease (Fig. 2), though there was no significant correlation between the haemagglutination titres and the clinical picture, except in the acute and fulminant cases. However, there was no
correlation between the degree of positivity of the haemagglutination reaction and the extent of derangement of the liver function tests.

Discussion

The frequent history of fever and the presence of leucocytosis and splenic enlargement suggested an infective factor in the aetiology of Indian childhood cirrhosis. Stokes et al. (1951) reported a case in a newborn who subsequently developed cirrhosis of the liver. Virus B was shown in the mother and infant. Similar cases have been reported by other workers. Similarities in the histopathology of the liver between fatal cases of infective hepatitis and Indian childhood cirrhosis (Achar and Raju, 1951) and the presence of eosinophilic intracellular hyaline bodies identical to the Mallory bodies were also considered pointers towards a viral affliction. The haemagglutination test considered to have a higher specificity to the virus was carried out in this series with a view to eliciting evidence, if any, for a viral aetiology in Indian childhood cirrhosis. The positivity of the test in 83 cases of cirrhosis, with higher titres in the acute and fulminant varieties, is highly significant. Hoyt and Morrison (1956) postulated that the haemagglutinating agent present in the series of their positive cases could be the virus itself. This was further supported by Rubin, Kemp, and Bennet (1957). If this is accepted our results provide evidence in favour of a viral aetiology. However, Hoyt, Morrison, and Levine (1961) later described the nature of the haemagglutinating agent as an antibody γ-globulin. We have noted a positive correlation between haemagglutination titres and the globulin levels in the sera of our cases of Indian childhood cirrhosis (to be published). From the present study one can only conclude that the high positivity of the haemagglutination reaction in Indian childhood cirrhosis favours a viral aetiology, if the haemagglutinating agent is the virus itself; or an antibody γ-globulin may be responsible for this reaction.

Summary

The haemagglutination test of Hoyt and Morrison was recorded in 100 cases of Indian childhood cirrhosis and was positive in 83 cases as against 2 out of 25 controls. The high rate of positivity favours a viral aetiology.

References


Insulin Studies in Temporary Neonatal Hyperglycaemia

Very few insulin studies have been performed in infants with temporary neonatal hyperglycaemia. Lewis and Mortimer (1964) used the rat diaphragm method and were unable to demonstrate any insulin-like activity in a patient before treatment was started. Ferguson (1967) carried out plasma insulin studies from birth in one such infant. The immunoreactive insulin levels were considered to be at the upper limit of normal in that infant. Unfortunately, no further insulin studies were possible, after the patient recovered from the diabetic state, as he died shortly afterwards in renal failure as a result of polycystic disease of the kidneys.

Gentz (1969), also using the radioimmunoassay technique, reported levels of 6 and 17 μU/ml plasma immunoreactive insulin before treatment in 2 affected infants. Both these infants had developed symptomatic hypoglycaemia before the development of the hyperglycaemia.

A case is reported here where plasma immunoreactive insulin studies were carried out before treatment was started, and repeated again at the age of 1 month, and at the age of 4 months.

Case History

The patient, a female infant weighing 1890 g, was born by spontaneous vaginal delivery in October 1969, three weeks before the expected date of delivery. The mother was 30 years old, in good health, and this was her ninth pregnancy. She had four children who were alive and well. Four other pregnancies had aborted spontaneously during the first 16 weeks of pregnancy. During this pregnancy she had been well apart from some vaginal bleeding at the eighth week. The infant gasped and cried immediately at birth, and only required mucus extraction and facial oxygen for resuscitation. The Apgar score was 7 at 2 minutes. She showed the appearances of placental insufficiency, with wrinkled skin and lack of subcutaneous fat. During the first 2 days of life there was some vomiting, but artificial feeding with Ostermilk No. 1 was quickly established, and by the eighth day she had almost regained her birthweight.

On the eighth day of life generalized twitching of both arms and legs was noted. A provisional diagnosis of tetany was confirmed, the serum calcium being 3.0 mEq/l., phosphate 10.5 mg/100 ml, and the alkaline phosphatase 14·5 KA units/100 ml. She was treated with calcium chloride 300 mg orally every 6 hours for 3 days and the convulsions ceased. Feeding continued satisfactorily during this time but she was noted to be losing weight. On the eleventh day there was a rapid deterioration in her general condition, the respiration was acidotic in type, and severe acidosis was confirmed biochemically. The pH was 6.87, Pco2 22·0 mm Hg, standard bicarbonate less than 5·0 mEq/l., and the base deficit 22·0 mEq/l. Convulsive twitching again developed on this day. The metabolic acidosis was regarded as due, at least in part, to over dosage with calcium chloride. Her general condition rapidly improved over the next few hours as the acidosis was corrected with intravenous 8·4% sodium bicarbonate, but she continued to have convulsive twitching for another three days in spite of intramuscular pheno-barbitone 15 mg 6-hourly. Twitching finally ceased in the fourteenth day of life. On the thirteenth day of life, however, she was found to have a marked glycosuria, and the true blood glucose was 350 mg/100 ml. There was no ketonuria, but a trace of albumin was present in the urine. A urine chromatogram showed only glucose to be present. After a 4-hour fast a glucose tolerance test with insulin studies was performed after the oral administration of 5 g glucose. The glucose tolerance test was diabetic in type (Table I). She was given 5 units of soluble insulin, twice that day, which promptly controlled the hyperglycaemia, and there was marked improvement in her general condition. The following day, 4 units of soluble insulin were required on two occasions. On the third day of treatment only 2 units of soluble insulin were required, and 1 unit of soluble insulin was given 3 days later. After this time no further insulin was required. On the seventeenth day of life the urine was free of reducing substances, the fasting true blood glucose was 57 mg/100 ml, and a transfusion of 75 ml whole blood was given with further improvement in her condition. Over the next few days good progress was maintained and an excellent weight gain was established. The urine remained free of reducing substances, and on the twenty-sixth day of life a fasting true blood glucose after a 31-hour fast was 54 mg/100 ml,

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Downloaded from http://adc.bmj.com/ on October 20, 2017 - Published by group.bmj.com
Haemagglutination reaction in Indian childhood cirrhosis indicating viral aetiology.
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Arch Dis Child 1971 46: 390-392
doi: 10.1136/adc.46.247.390

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