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Jaundice in Early Infancy*

THE SURGICAL ASPECTS

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In this paper those conditions which cause jaundice in early infancy and are of direct interest to the surgeon are considered. While the treatment of this group is essentially surgical, an accurate diagnosis can only be made after a careful consideration of all aspects of the case in order to exclude the commoner causes of jaundice in early infancy and in particular to differentiate between the two principal causes of obstructive jaundice in young infants.

INSPISSATED BILE SYNDROME

Its relative importance to the surgeon may be appreciated from the figures of Hsia *et al.* (1952), who found 53 cases in a series of 156 examples of obstructive jaundice in young infants.

It is proposed to consider this condition in more detail when the clinical management of biliary atresia is discussed.

CONGENITAL BILIARY ATRESIA

Our clinical material in this group consists of five infants who were treated in the Memorial Hospital, Bulawayo, between October, 1957, and

June, 1958. All these cases were proven at operation to have congenital biliary atresia. All were African infants, and it is felt that the incidence of the disease may be higher than that reported by Moore (1953) in the United States of one in 20-30,000 births. The largest series is that of Gross (1953). The condition is thus one of great rarity and the personal experience of one surgeon must be limited.

There are few references to the condition in the British literature to this condition, and the first successfully treated case was reported by Beavan and Duncan (1945). Ladd (1928) was the first surgeon to treat the condition successfully. Atresia of the bile ducts may occur in any or all parts of the biliary tree.

It will be appreciated that in only some 18 per cent. of cases was it possible to treat the condition. The distribution of the disease in the present group of cases is shown in Fig. 2. This shows that in only one case was it possible to perform an anastomosis.

The aetiology of the atresia has not been proven in this condition, but it is thought to result from a failure to recanalise the ducts after a period of epithelial condescence in early foetal life. There may be associated congenital abnormalities, and in the present group two infants had a situs inversus which was limited to the abdominal viscera.

The principal pathological features in these cases may be correlated with the inability of the liver to excrete bile into the alimentary tract

Table 1
ONE HUNDRED AND FIFTY-SIX CASES OF OBSTRUCTIVE JAUNDICE IN INFANCY

Biliary atresia	94 or 60 per cent.	Four successful.
Inspissated bile in erythroblastosis	23 or 15 per cent.	All alive.
Inspissated bile of unknown aetiology	30 or 19 per cent.	Eighteen alive.
Other causes	9 or 6 per cent.	

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and the secondary effects that ensue. Jaundice is present from a very early age and has been noted at birth. It is usually steadily progressive and the serum bilirubin may reach a high level. It is the commonest presenting symptom and sign. The infants in the present group were not brought to hospital until they were four to four and a half months old. None were the children of educated parents, so that an accurate estimate of the age of the infant or of the duration of the jaundice was not possible. Jaundice in the African is easily missed and it will only be possible to detect early cases in the African if the conjunctivae of all babies are inspected at birth and at routine post-natal clinics.

The serum bilirubin is initially low relative to the levels found in haemolytic jaundice at the same age (Hsia *et al.*, 1952).

An infant of two months commonly presents with a level of 5 to 10 mg. per cent., and the level thenceforth tends to show a progressive rise on serial estimation. There is usually an equal rise in the direct and indirect levels. The highest level found in the present group was 13 mg. per cent. in an infant of seven months.

The stools are constantly acholic and it is only in the advanced case that there is slight bile staining of the surface of the stool by the discoloured mucous of the alimentary tract. Even then the stool retains its pale colour and is characteristic of the condition. A trace of bile in the stool makes it very unlikely that the cause of the jaundice is biliary atresia. Bile is present in the urine and there is a complete lack of urobilinogen in the urine. The general nutrition of the child is normally well maintained and in only one of our cases was there serious evidence of impaired nutrition. Spontaneous haemorrhage as a result of vitamin K deficiency is rare and the response of the prothrombin index to parenteral administration of vitamin K is normally good.

The liver becomes progressively enlarged and may reach relatively enormous proportions. In one of our cases it reached to the pelvic brim and by its mere size was producing respiratory embarrassment. The liver is smooth and has a firm rounded edge. The enlargement is usually symmetrical, but the presence of a situs inversus may lead to confusion. The colour is a

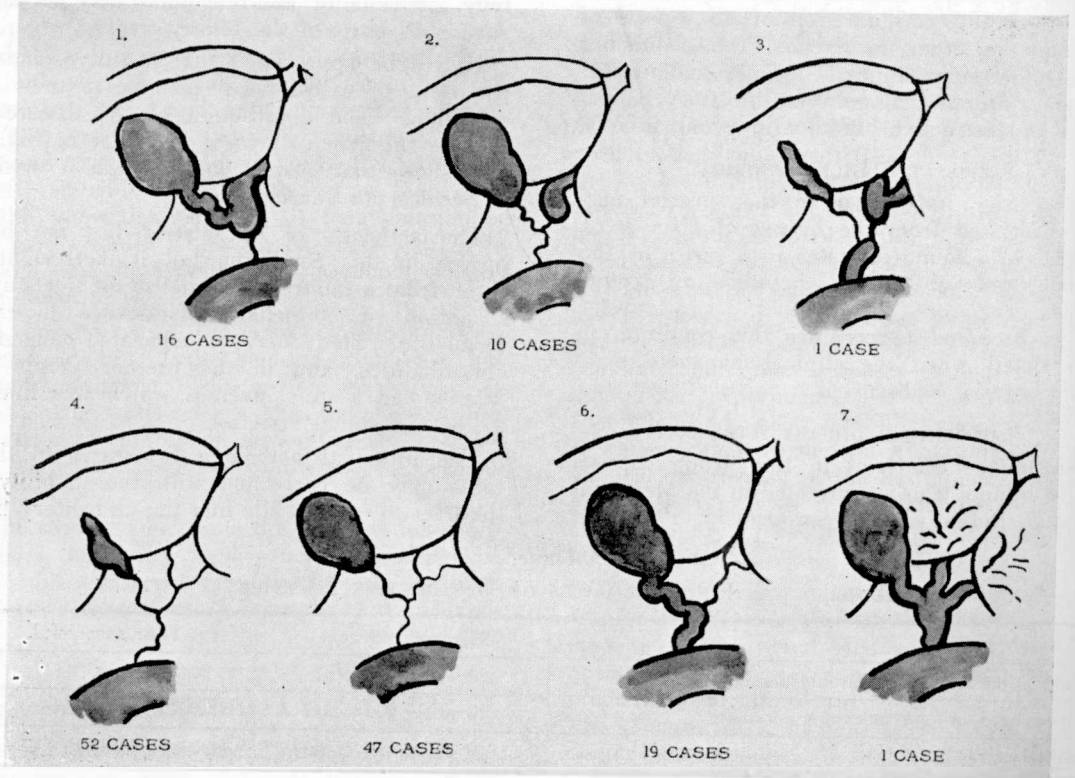


Fig. 1—Gross (1953).

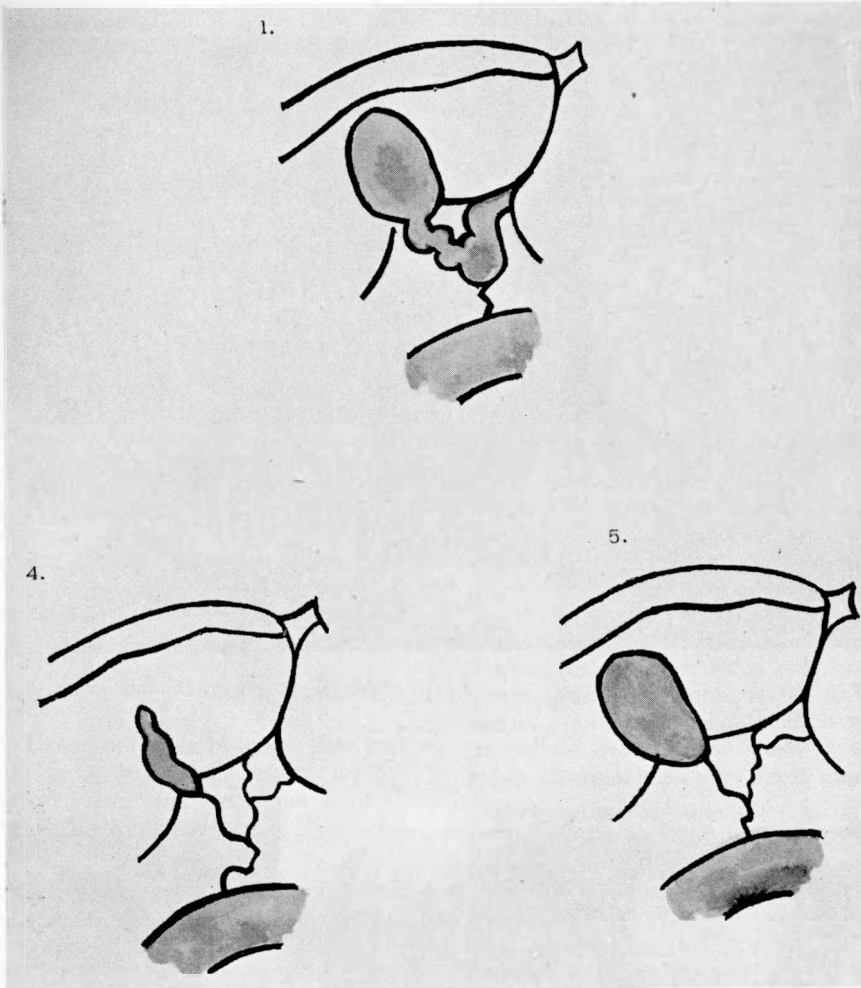


Fig. 2—No. 1, one case; No. 4, two cases; No. 5, two cases.

dark greenish-brown with small orange flecks. The histological changes are numerous and important, but a detailed consideration is beyond the scope of this paper. Complete absence of the intra-hepatic ducts was a feature of two of our five cases. In the others there was a well marked peri-portal fibrosis with numerous poorly formed ducts, which were not significantly dilated. The liver cells may show degenerative change. The onset of biliary cirrhosis is said to be early in these cases and may be apparent in infants of two months (Gross, 1953), but it is not invariable even in the presence of a purely extra-hepatic atresia (Myers *et al.*, 1956). The presence of moderate cirrhosis is compatible with a good long-term result (Gross, 1953) (Fig. 3).

When the biliary cirrhosis is more severe the features of portal hypertension may obtrude: splenomegaly, ascites and even oesophageal varices. In one of our cases there was evident ascites and in two a markedly enlarged spleen. In the light of this experience and that of others, there seems to be no indication for the use of radical operations such as Longmire's (intra-hepatic cholangio-jejunostomy) in this condition in view of the absence of significant dilatation of the intra-hepatic ducts.

The natural history of the condition is of great interest in that these children may survive for many months or even years. The condition is invariably fatal if untreated and the usual cause of death is intercurrent infection or hepatic failure. These facts would appear to

has preferred cholecysto-jejunosotomy with a Roux-en-Y anastomosis. In the solitary operable case in the present group, a cholecysto-jejunosotomy was performed (Fig. 4).

SUMMARY

The pathology, clinical features and treatment of obstructive jaundice in young infants are reviewed after a study of five cases in African children in the Memorial Hospital, Bulawayo.

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