CASE REPORT

CONGENITAL OBLITERATION OF THE BILE DUCTS*

BY

REGINALD WILSON, B.A., M.D., D.C.H.

John Thomson¹ has shown that the clinical condition entitled congenital obliteration of the bile ducts may depend upon a number of somewhat different anatomical deformities. From his study he concluded that the pathological process was primarily a developmental arrest. He described aplasia and narrowing of the ducts which he believed caused obstruction. This he thought gave rise to the cirrhosis of the liver which is so constantly found at autopsy. Opposing this view Rolleston^{2,3} later suggested that there was a placental toxæmia which had its effect upon the foetal liver causing a cholangitis. This was a descending process which resulted in an inflammatory thickening of the duct walls, and thus a stenosis.

Case record.

A. E., a female child, was six months of age when admitted to the Westminster Hospital. The complaints were jaundice, boils on the buttocks, and failure to gain weight. The jaundice had had its onset very insidiously in the first weeks of life. The infection was a more recent affair. The parents are both well and their Wassermanns are negative. They have four other children, three of these were slightly jaundiced during the neonatal period, but in none did it persist.

EXAMINATION revealed an emaciated yellow child, with an enlarged liver and spleen. There were staphylococcal abscesses of the buttocks. The stool was usually almost white, but was occasionally streaked with bile. This was shown not to be urinary contamination, although the urine did contain large amounts of bile. The blood Wassermann was negative.

PROGRESS. The infection gradually subsided, but the degree of jaundice did not alter, and the child never gained weight during the subsequent eight weeks in Hospital. Measles and a complicating bronchopneumonia finally caused death.

PATHOLOGICAL FINDINGS. The skin, sclerae and all organs were deeply bile-stained. There were numerous healed scars on the buttocks. The lungs showed bronchopneumonia. The liver was firm and its dark green surface had a granular appearance. Bile could not be squeezed from

* The study of this case was undertaken during the tenure of the Wander Research Scholarship at the Westminster Hospital and I am indebted to Dr. Reginald Lightwood for special facilities for investigation supported by the Thomas Smythe Hughes Medical research fund,

FIG. 1.—Common bile duct to show thick wall and narrow lumen.

FIG. 2.—Drawing of the liver with dissected cystic duct showing its vacuolated condition.

FIG. 3.—Microphotograph of the liver showing coarse fibrosis of the portal tract and small atrophic bile ducts.

FIG. 4.—Wall of the cystic duct showing its thickness and honeycombing.

the ampulla of Vater when pressure was exerted on the small gall bladder. A firm bristle could, with difficulty, be passed from the ampulla along the common duct into the hepatic duct; the walls of both of these were very thick (fig. 1).

On opening the gall bladder it was found to be thickened, and to contain a little greenish mucus. The first part of the cystic duct was carefully opened by cutting away the superior surface. No opening from the gall bladder could be discovered, and the thick-walled duct had its lumen honey-combed with bands of tissue (see fig. 2). No continuous lumen could be found till the lower third of the duct was reached. For the sake of comparison the bile duct system of a new-born infant, who had died of an injury, was dissected. The spiral valves of Heister were demonstrated and the cystic duct could be probed with ease.

MICROSCOPIC EXAMINATION of the liver showed a widespread fibrosis, irregularly arranged but chiefly affecting Glisson's capsule (fig. 3). The bile ducts were small and atrophic, and surrounding them were aggregations of small round cells. There was much bile staining. Sections through the cystic duct showed it to be devoid of the normal columnar epithelium. The muscle coat was atrophic and the lining epithelium was composed of flattened polyhedral cells two to four cells deep. There appeared to be one main lumen, but the walls showed smaller spaces, the lining of which was similar to the main lumen (fig. 4). There were no glands in the wall, and the signs of inflammation were lacking.

Discussion.

Keith⁴ states that the common bile duct, the gall bladder and the cystic duct are developed from the hind part of the hepatic diverticulum of the fore gut. The hepatic ducts arise from the solid liver outgrowth. The lumen of the gall bladder and ducts is occluded by an epithelial proliferation during the second month. In normal development this condition becomes vacuolated, the vacuoles coalesce and the lumen becomes established.

Such an arrest in the vacuolated stage would account for the situation in this case. Careful comparison of the gross and microscopic anatomy between this and the normal case emphasized the pathological significance of the vacuolated condition of the cystic duct, which may represent an arrest in the process of canalization.

Summary.

A case is presented in which:—

1. The gross and microscopic anatomical findings suggest that jaundice was due to a partial stenosis of the common bile duct. The cystic duct shows evidence of arrest at an early stage of foetal development.
2. An unusually long period—eight months—ensued before death from intercurrent infection took place.

I wish to thank Dr. Donald Paterson for permission to publish this case.

REFERENCES.

1. Thomson, J., *Edin. Med. Jour.*, Edin., 1891, XXXVII, 532.
2. Rolleston, H. D., *Brit. Med. J.*, London, 1907, ii, 947.
3. Rolleston, H. D., & Hayne, L. B., *ibid.*, 1901, i, 758.
4. Keith, A., *Human Embryology*, fifth edition, London, 1933, 308.