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ABSENT INTRAHEPATIC PORTAL VEINS ARE ASSOCIATED WITH LIVER TUMOURS IN PATIENTS WITH CONGENITAL PORTOSYSTEMIC SHUNTS

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Aims: Congenital porto-systemic shunts predispose to the formation of both benign and malign liver tumours. The aim of this study was to outline the use of radiological investigations and to assess the significance of absent portal veins in those investigations.

Methods: Single-centre retrospective cohort study of all patients with congenital portosystemic shunts referred from 1990 to 2016. Radiological investigations were reviewed by two consultant radiologists for the presence of intrahepatic portal veins (IHPV). Groups were compared for their association with liver tumours, serum ammonia, and surgical closure of the shunt using appropriate non-parametric tests. Data are quoted as median (IQR). A P value of 0.05 was considered significant.

Results: 46 patients were investigated for congenital portosystemic shunts at a median age of 8 months (IQR 1 month – 14 years). Ultrasound was used to investigate 41/46 (89%), CT in 25/46 (53%), MRI in 29/46 (63%), and 15/46 (33%) underwent a portal venogram. Only 9/46 (19%) had only one modality of imaging.

IHPV were identified in 15 (33%) of which 11 (75%) had associated liver tumours compared to 10/31(32%) in those without IHPV (P = 0.01). Median ammonia level at diagnosis was similar in both groups 84 (IQR 62-98) vs. 98 (IQR 79-109) μmol/L respectively (P = 0.16).

20 patients underwent surgical closure of the shunt at a median age of 8 (3 – 13) years with 6 (30%) requiring a staged approach. Primary closure was possible 4/8 (50%) patients with undetectable IHPV compared to 10/12 (84%) of those with detectible IHPV (P=0.13).

Conclusions: Lack of IHPV on multimodal imaging and not the level of plasma ammonia suggests a risk factor for development of liver tumours in patients with congenital porto-systemic shunts.