Clinicopathological progress in hepatic tumors of children

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Hepatoblastoma (HB) and hepatocellular carcinoma (HCC) comprise 2/3 of primary hepatic tumors of infants and children. While excision is the treatment of choice, studies of the Children's Cancer Study Group (CCSG) revealed only 60% of stage 1 HB/HCC patients to be disease free at 24 months. Completely resected pure fetal histology HB 24 month prognosis is superior (92%) to that of embryonal, macrotrabecular, small cell (anaplastic) histologic variants (57%). The proportion of embryonal histology influencing unfavorable outcome in predominantly fetal HB is unknown. DNA aneuploidy in embryonal HB may indicate poor prognosis. In advanced stage HB pure fetal histology has not been demonstrated to influence survival; however differentiated mesenchymal elements and the absence of mitotic activity are associated with improved survival.

Of 196 CCSG HB and HCC 68% were advanced stage at presentation. Adjuvant chemotherapy rendered 59% of 34% unresectable tumors amenable to removal. HB had a better prognosis than HCC but survival was not equivalent to stage 1 tumors.

Pathogenetic considerations of HB describe heteromorphism of chromosomes 2 and 20. Patients with HB and the gene for familial adenomatosis polyposis coli may be identifiable by congenital hypertrophy of retinal pigment epithelium; their tumors may be less aggressive than sporadic HB.