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Abstracts selected for presentation at the XX Annual Meeting of ESPGAN, Lisbon, Portugal 24–26 June, 1987

1 INCIDENCE OF CIRRHOSIS IN CHILDREN WITH CHRONIC HEPATITIS. THE VALUE OF LAPAROSCOPY P. Vajro, P. Hadchouel, M. Hadchouel, O. Bernard, D. Alagille. Hépatologie Pédiatrique INSERM U 56 and Département Pediatrie, Hôpital Bicêtre, 94270 Bicêtre, France.

Incidence of cirrhosis (CIR) in children with chronic hepatitis (CH) varies greatly according to previous reports. CIR complicating CH is most often macronodular, this being a cause of sampling error in the interpretation of needle biopsy. We report here on 92 children with CH who were investigated, at admission, for the presence of CIR by the combined use of laparoscopy and needle liver biopsy between 1975 and 1985. - 46 children had hepatitis B virus-related CH. CIR was present in 15 (33 %). Laparoscopy showed nodules in 14 ; liver histology showed definite signs of CIR in 8, severe signs of aggressivity (A) in 5, moderate A in 5 and no signs of A in 5. Signs of active viral replication were present in 5 of 11 children studied. 6 patients had CIR less than 12 months after the first sign of liver disease. - 46 children had autoimmune hepatitis. CIR was present in 41 (89 %). Laparoscopy showed nodules in all. Histology showed definite signs of CIR in 16, probable or possible CIR in 7 others, severe A in 19, moderate A in 16 and no A in 6. CIR was present in 10 of 10 children studied 2 to 5 months after the first sign of liver disease. - These results indicate that in children with CH (1) combined use of laparoscopy and biopsy is twice as much reliable than biopsy alone for the diagnosis of CIR (2) true incidence of CIR is high (3) CIR occurs early irrespective of etiology.

2 SCLEROSING CHOLANGITIS IN CYSTIC FIBROSIS

Hjelte L*, Gabrielsson N**, Strandvik B*
Departments of Pediatrics* and Diagnostic Radiology**, Karolinska Institute, Huddinge University Hospital, Stockholm, Sweden.

Three (3.0%) of the 102 patients with cystic fibrosis (CF) attending our CF-center were shown to have ERCP-findings indicating sclerosing cholangitis. All had pulmonary symptoms. Two of them, both females (aged 17 and 20 years), presented with abdominal pain while a 26 years old male was asymptomatic. One of the females also had an unspecific colitis. Two of the patients had persistently altered liver function tests with increase of serum concentration of transaminases and glutamyltransferase while one normalized her laboratory findings. This patient, who was the only one with a microgall bladder, showed no progress during a three years follow up. Liverbiopsies revealed portal fibrosis, bile duct proliferation and an inflammatory reaction and in one case heavy steatosis. Congenital anomalies, biliary calculi, previous operative trauma and malignancy was not found.

Primary sclerosing cholangitis is usually related to inflammatory bowel diseases which may be associated with a wide variety of extraintestinal lesions including pulmonary disease. Gastrointestinal manifestations such as liver and biliary diseases are well known complications to CF. To the best of our knowledge this is the first report of sclerosing cholangitis in this disease.

3 CHARACTERIZATION OF THE ANTIGEN RECOGNIZED BY THE ANTI-LIVER KIDNEY MICROSOME ANTIBODIES (LKMA). F. ALVAREZ, C. DELEMOS-CHIARANDINI, K. PARADIS, O. BERNARD. INSERM U 56, 94275 Bicêtre, France and Dept Cell Biology NY U Medical Center, New-York.

A subgroup of children with autoimmune hepatitis is defined by the presence in the serum of high titers of LKMA, usually detected by immunofluorescence (IF). We have studied the sera of 5 such children to characterize the antigen recognized by LKMA. Negative controls were provided by sera from children with smooth muscle antibody positive chronic hepatitis, Wilson's disease and Hepatitis B related chronic active hepatitis. Cell fractionation of rat liver and immunoblot analysis showed the antigen to be a protein of 50000 molecular weight, present in high concentrations in smooth microsome subfractions, integrated in the microsomal membranes, and exposed on their cytoplasmic side; the antigen is not glycosylated and does not form homo or heteropolymers by disulfide bonds. Immunoelectronmicroscopy of rat liver showed the antigen to be detectable only in endoplasmic reticulum membranes of hepatocytes and not in other liver cell types. Cell fractionation and immunoblot of human liver detected a 48000 MW protein with characteristics similar to that of rat liver. An ELISA technique was developed, using rat liver microsomes as substrate, that proved to be most useful for diagnosis and follow-up studies, being more sensitive and cheaper than IF. These results may prove useful both for improving detection and care of these children and for understanding the mechanisms of autoimmune hepatitis.

4 D-PENICILLAMINE INCREASES SURVIVAL IN PRE-ICTERIC INDIAN CHILDHOOD CIRRHOSIS (ICC). MS Tanner, SA Bhave, IR John, AN Pandit, Dept of Child Health, University of Leicester,UK and Dept Pediatrics, KEM Hospital, Poona, India.

Interim encouraging results of a D-penicillamine trial in ICC (presented to ESPGAN 1984) are now confirmed. 30 children with biopsy-proven ICC who had not yet developed jaundice or ascites were treated double blind with: D-penicillamine (Group P, n=10); penicillamine + prednisolone 2 mg/kg/d for 4 weeks then 5 mg/d (Group PP, n=10); or placebo (Group Plac). Entry parameters were found to be comparable between groups. 9/10 placebo treated children died, median survival 58 days; 1 survives to date (2.7y). 5 in Group P, and 5 in Group PP survive after 1.3-4.0y. Life table analysis (Breslow) showed a significant therapeutic benefit (P vs Plac p=0.01; PP vs Plac p=0.03) but no difference between active treatment groups (p=0.71). Inclusion of a further 13 children treated non-blind did not introduce entry differences between groups but increased significance of survival differences: P'(n=15) vs Plac'(n=14) p=0.002; PP'(n=14) vs Plac' p=0.002. 14/29 P' or PP' treated children survive, are well and have follow-up liver biopsies showing inactive micronodular cirrhosis in 5, portal fibrosis in 6, and minimal abnormalities in 3. Conclusions: D-penicillamine increased survival in pre-icteric ICC from 1/14 to 14/29, with reversal of cirrhosis in 9, and prednisolone conferred no additional benefit.