

CONGENITAL HYPERBILIRUBINEMIA

Pages with reference to book, From 310 To 310

Sir,

The concept of congenital hyperbilirubinemia, its frequency and its benign nature is not clear in our country. These patients are being investigated as acute and chronic liver disease and managed as such. This approach causes considerable anxiety and inconvenience to the patient and their relatives. A retrospective analysis was undertaken to know the behaviour and long term prognosis of these patients. Of 3629 patients attending the outpatient's department of PMRC Research Centre, Jinnah Postgraduate Medical Centre with various liver diseases, 21 (0.58%) had congenital hyperbilirubinemia. Of these, 4 had conjugated and 17 unconjugated hyperbilirubinemia. Amongst the unconjugated, 10 had Gilbert's syndrome and 7 Crigler Najjar Syndrome Type II. All the cases of conjugated hyperbilirubinemia and cases with splenomegaly¹ were excluded from the study because of incomplete investigations. Mean age at first detection of jaundice was 19 years for Gilbert's syndrome and 10 years for Crigler Najjar syndrome Type II. Male preponderance was noted in both the diseases, ratio being 9:1 and 2.5:1, respectively. In contrast no sex difference was noted by other workers¹. Presenting symptoms in majority of the cases were repeated episodes of jaundice along with fatigue, malaise, anorexia, nausea and vomiting. Few cases had pruritus, fever with rigors and passage of dark coloured urine. All cases of Crigler Najjar and 7 of 10 cases of Gilbert's Syndrome were jaundiced at the time of examination. Liver was just palpable in 6 cases except in one where it was palpable 5cm below the costal margin. Liver biopsy was done in 6 patients; it was normal in all except mild cholestasis and round cell infiltration in one case. Of 17 cases, 10 were followed from 1-13 years. No complication was found in any case; however, icteric sclera persisted intermittently in most cases which increased with physical activity, intercurrent infections and warm weather. Response to phenobarbitone was good in 5 cases, variable in 3 and none in 2 cases. Multiplicity of symptoms and failure to elicit any common pattern of complaints indicates that symptoms may be unrelated to the disorder of bilirubin metabolism. Symptoms are likely to develop after the patients become aware of jaundice and; therefore, are probably anxiety related. Thus if a patient has congenital hyperbilirubinemia with a normal health all that is required is reassurance.

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