

Liver dysfunction in steady state sickle cell disease,
by Taiwo Kotila et al.

Although patients with sickle cell anemia (SCA) may present liver dysfunction, the extent of involvement of the liver in this condition is unclear. The hepatic complications of SCA disorders can be related to hemolysis and/or to the outcome of massive transfusion regimen such as HBV or HCV infection. The paper by Kotila addresses this topic by analyzing a sufficiently large number of patients with SCA and assessing liver function by the common tests. The conclusion is that liver is minimally involved in the SCA since the majority of cases showed normal ALT/AST values; the alteration in alkaline phosphatase may be due to extra-hepatic involvement. Exception was the serum bilirubin which was associated to the hyperhemolysis and correlated with the degree of hepatic enlargement. In the presence of normal liver function serum unconjugated bilirubin never exceeded 3.0 mg/dL (60 μ M) in spite of severe degree of red cell destruction. Data reported, although unfortunately very limited, shows that a total bilirubin below that threshold was found in subjects with normal liver dimension while bilirubin it climbed as the dimension of the liver increased, suggesting liver damage. It is therefore tempting to speculate that bilirubin may be a nice indicator of liver damage (due to whatever reason) and may be used to predict whether the liver function is reduced. It would be interesting to correlate serum bilirubin with either liver dimensions and ALT/AST/Alkaline Phosphatase values. It would be not surprising that the simple, clinically relevant observation of the color of the sclerae will be more indicative than more sophisticated, expensive tests.

Autoimmune-type chronic active hepatitis in children. A report of 23 cases at a Hospital in Northwestern Mexico,
by Norberto Sotelo and Guillermo López.

Autoimmune Hepatitis (AI) is a disease where immunologic “dysfunction” is at the basis of liver damage. In spite of its recognition more than 50 years ago, the clinical spectrum, the pathogenesis and most important the natural history is still less than clear. Several classifications have been proposed but with the time it has become clear that some clinical and laboratory pictures were difficult to fit in predefined boxes. If AI is rare in adults, this situation is even more rare in children. This is why the series reported by Sotelo and López is of relevance. Over a period of 30 years, 23 babies with clinical and histological abnormalities compatible with the diagnosis of AI were retrospectively recruited and the outcome followed after treatment with prednisone, either alone or in combination with azathioprine. The outcome of patients who followed the treatment was better than that of those who discontinued the drug(s). Of particular importance was the observation that 30% of children who stopped the treatment ended up with a cirrhosis in a rather short period of time. Unfortunately the treatment was associated with severe side effects pointing to the need of a balanced assessment of the pros and cons before starting a treatment in young patients. As the drugs proven to be effective are basically the same used when the AI was discovered 50 years ago, the need for a better, less harmful treatment is evident.

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