

CORELLATING CLINICAL FINDINGS WITH LIVER HISTOLOGY AT INITIAL PRESENTATION IN BILIARY ATRESIA

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Aim: Age at presentation and total serum bilirubin have been shown to impact on the outcome of Portoenterostomy (PE) for Biliary Atresia (BA). The aim of this study was to correlate both these factors with the severity of liver disease on histology.

Methods: We maintain a prospective data base of all children undergoing PE for BA at our institutions since February 2013 to the present date. For purposes of this study, we used data from 45 consecutive children with BA. All PE's are performed by a single surgeon and all wedge biopsy specimens are reported on by a single pathologist. A numerical liver injury score was computed by assigning scores to 3 grades of fibrosis (Ishak), presence or absence of Ductal plate malformation (DPM), Giant cell transformation, extramedullary hematopoiesis, inflammation in the portal plate, lobular inflammation, ductular proliferation and canalicular bile stasis. The total score was correlated to the age at presentation and the initial total bilirubin level using standard statistical methods.

Main results: The average age at presentation was 79 days (range 23-139 days). The average total bilirubin was 11.1 mg/dl (range 7.1- 18.1 mg/dl). 4 children had grade 1, 22 children had grade 2 and 19 children had grade 3 fibrosis. 14 children had DPM, 22 children had extramedullary hematopoiesis. 21 children had giant cell transformation. All children had ductular proliferation, canalicular bile stasis, inflammation of the portal plate and lobular inflammation. The average score was 7.5 (range 6-10). Total bilirubin correlated significantly with a higher liver injury score ($p < 0.05$). However, age at presentation did not correlate with liver injury score

Conclusion: In our study the total bilirubin at presentation reflects the severity of liver damage in infants with BA. However, age does not correlate and hence older children do not necessarily have severe liver damage.