

SIGNIFICANCE AND MANAGEMENT OF BILE LAKES AFTER PORTOENTEROSTOMY FOR BILIARY ATRESIA

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Aim of the study: Bile lakes are severe and incompletely characterized complication after portoenterostomy (PE) for biliary atresia (BA). We investigated relationships between bile lakes, cholangitis and clinical outcome as well as the surgical management of bile lakes at our institution.

Methods: Medical records and imaging studies of all patients who had undergone PE for BA at our institution during 1987-2016 (N = 62) were retrospectively reviewed after ethical approval. Cholangitis was defined as febrile illness without any other identified cause treated with antibiotics. Mann-Whitney U and Fischer's exact test were used for data analyses.

Main Results: Overall, median of 3.0 episodes of cholangitis occurred in 48 (77%) patients. 36 (58%) patients had ≥ 2 cholangitis episodes. Bile lakes occurred in 8 (13%) patients. All patients with bile lakes had ≥ 2 cholangitis episodes as opposed to 52% in those without bile lakes ($P=0.016$). Frequency of cholangitis episodes/patient was ~ 5 times higher among the patients with bile lakes (**Figure**). Among the patients with bile lakes, the first cholangitis occurred exceptionally early, median 34 (IQR 105) days after PE, and bile lakes were diagnosed 216 (IQR 147) days after PE. Before 1995, two patients underwent transcutaneous drainage of bile lakes and both died within a year of PE. Six more recent patients underwent Roux-en-Y bile lake-jejunostomy with reduction of yearly cholangitis rate/patient from 8.8 (1.7-15) to 1.1 (0-4.4), $P=0.028$. Four of the patients have survived 8.5 (range, 1.9-17) years jaundice-free with their native livers, and two received liver transplant 1.3 and 4.9 years after the operation. Clearance of jaundice, PE age, other anomalies, BA type, and gender were not related to cholangitis episodes.

Conclusion: Bile lakes were the major risk factor for recurrent cholangitis. Bile lake-jejunostomy successfully reduced recurrent cholangitis episodes and extended native liver survival in patients with bile lakes.

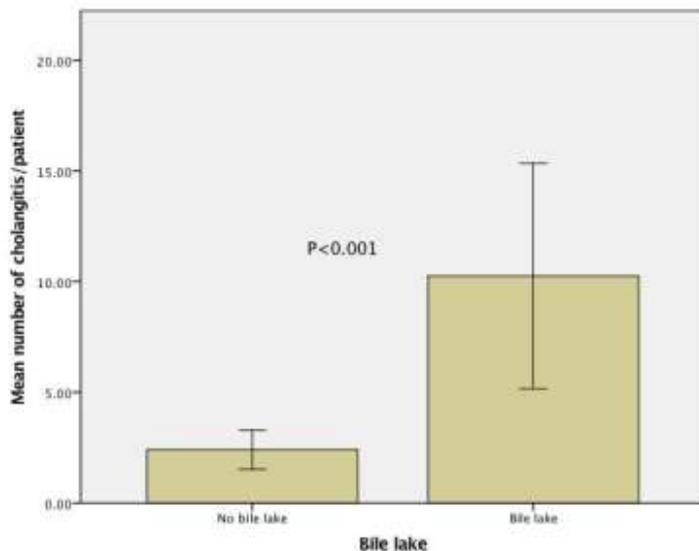


Figure. Mean (95% CI) number of cholangitis episodes/patient according to the presence of bile lakes.