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- HEPATOLOGY

Abstract Title:

Long-term outcome of biliary atresia into adult life

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ABSTRACT: Background:Biliary Atresia(BA) is the single commonest cause of neonatal cholestasis leading to cirrhosis,portal hypertension and liver failure and is the main indication for pediatric liver transplant(LT). Aim:Evaluate the long-term outcome of children with BA transitioning to adult life Subjects & Methods:Records of patients of BA managed over a period of 34 years(1980-2014) at a single institution were retrospectively reviewed .Patients with more than 10 years of follow-up were included in the study.Data collection included demographics,age at Kasai Portoenterostomy(KPE),associated malformations, survival with native liver or post-LT,mortality, current education/work/marital/family status. Results: 493 BA patients were managed during this period(260 F & 233 M).Median age at kasai was 53 days(range:7-183 days). 92 % had isolated BA while 8 % had BA polysplenia malformation syndrome.332 patients were included in this study (1980 – 2004). 11 patients were lost to follow-up. Median patient survival is 17.3 yrs(0.32- 34.6) & median survival with native liver is 2.25 yrs(0.07-34.6). 53 patients(16.5%) died in pediatric care; 26 with their native livers & 27 after LT.135 patients(50.3%) are still in pediatric care(Group A).57 are surviving with their native liver(A1) while 78 children have been transplanted(A2).7 patients are awaiting transplant

in Group A1.133(49.6 %) patients were transferred to adult services(Group B); 49 with native livers(B1) and 84 after LT(B2). 28 patients in group B1 had portal hypertension(PH);20 treated with beta blockers,esophageal banding or shunts. 9 patients transferred to adult services with native liver(B1) subsequently required LT & 7 are listed for LT due to decompensated liver disease.6 patients in group B2 required retransplant. After transfer to adult care, 3 patients in Group B1 died(one due to ruptured splenic aneurysm & 2 due to decompensated liver disease) while 5 patients in Group B2 died from post-transplant lymphoproliferative disorders(PTLD),Hepatopulmonary syndrome, ruptured psoas cyst and bleeding & chronic rejection).Out of 268 patients in this series, majority participated in normal school education while 32(12 %) required special needs support. 29 transferred went to university, 18 obtained non-vocational qualifications and 33 joined various training courses. Conclusion: Improved medical and surgical techniques have improved the outcome and quality of life for patients with BA, allowing them to live into adult life, complete their education & function as useful members of the society

Key Words:
Biliary atresia,liver transplant, transition to adult