Long-term postsurgical outcome of biliary atresia

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摘要

Abstract

Background/Purpose: A successful Kasai procedure is effective in creating biliary drainage and radically altering the natural history of infants with biliary atresia (BA). Since its introduction in the 1950s, long-term follow-up would appear to show that only 30% to 50% of patients have a good long-term prognosis despite initially good surgical outcome. The authors reviewed their experience in treating BA from 1968 to 1997 to assess long-term outcome.

Materials and Methods: The records of 163 patients treated surgically for BA from 1968 to 1997 were reviewed. Fortyeight (29%) were alive at the end of 1997, of whom, 14 had received liver transplants (LT). Surviving patients who had not undergone transplantation were divided into two groups according to clinical condition: group A, normal liver function without cholangitis (CG) and portal hypertension (PH) and group B, liver dysfunction with CG or PH. The study period was divided arbitrarily into three periods, 1968 to 1975 (period I, N = 34); 1976 to 1985 (period II, N = 81); 1986 to 1997 (period III, N = 48).

Results: Thirty-four patients were alive without LT at the end of 1997. There were eight patients (mean age, 16.3 ± 4.8 years) in group A, and 26 patients (mean age, 14.3 ± 7.6 years) in group B. Recently, four group A patients (mean age, 19.3 ± 1.9 years) shifted to group B because of sudden deterioration in condition involving severe CG with multiple bile lakes (n = 2), uncontrollable intestinal bleeding (n = 1), and liver atrophy (n = 1). Survival deteriorated with length of follow-up. There were three survivors from 34 patients treated in period I, 16 survivors from 81 patients treated in period II (three had LT), and 29 survivors from 48 patients treated in period III (11 had LT).

Conclusions: Although satisfactory bile drainage can be obtained with portoenterostomy, our data suggest that liver function can deteriorate progressively, with a possible turning point in late adolescence, indicating that as the length of follow-up increases, clinical assessment should be regular and

comprehensive. The timing of LT in postoperative BA patients with deteriorating liver function is a vital management issue