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Biliary atresia: the Brazilian experience

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ABSTRACT

Objective: To evaluate epidemiological, clinical and prognostic characteristics of children with biliary atresia.

Methods: Data regarding portoenterostomy, liver transplantation (LTx), age at last follow-up and survival were collected from the records of patients followed up in six Brazilian centers (1982-2008) and compared regarding decades of surgery. **Results:** Of 513 patients, 76.4% underwent portoenterostomy [age: 60-94.7 (82.6 ± 32.8) days] and 46.6% underwent LTx. In 69% of cases, LTx followed portoenterostomy, whereas in 31% of cases LTx was performed as the primary surgery. Patients from the Northeast region underwent portoenterostomy later than infants from Southern (p = 0.008) and Southeastern (p = 0.0012) Brazil, although even in the latter two regions age at portoenterostomy was higher than desirable. Over the decades, LTx was increasingly performed. Overall survival was 67.6%. Survival increased over the decades (1980s vs. 1990s, p = 0.002; 1980s vs 2000s, p < 0.001; 1990s vs. 2000s, p < 0.001). The 4-year post-portoenterostomy survival, with or without LTx, was 73.4%, inversely correlated with age at portoenterostomy (80, 77.7 and 60.5% for ≤ 60, 61-90 and > 90 days, respectively). Higher survival rates were observed among transplanted patients (88.3%). The 4-year native liver survival was 36.8%, inversely correlated with age at portoenterostomy (54, 33.3 and 26.6% for ≤ 60, 61-90 and > 90 days, respectively). **Conclusions:** This multicenter study showed that late referral for biliary atresia is still a problem in Brazil, affecting patient survival. Strategies to enhance earlier referral are currently being developed aiming to decrease the need for liver transplantation in the first years of life.

(Key words: Biliary atresia, portoenterostomy, hepatic, surgery, diagnosis, differential, prognosis).

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