Health-Related Quality of Life in Children With Biliary Atresia

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BACKGROUND: In children with biliary atresia (BA) who did not have surgery or in those who had an unsuccessful surgery, survival beyond three years of age is rare without liver transplantation (LT). Children with BA who had a successful surgery require long term follow up to detect complications associated with chronic liver disease (CLD).

OBJECTIVE: We investigated the quality of life (QoL) in children with BA, comparing it with children who have other forms of CLD, and with normal healthy children.

DESIGN/METHODS: Children with BA and other CLD, aged between 2 to 18 years old, who were followed up in the pediatric liver clinic at the University Malaya Medical Center, Kuala Lumpur from 1996 to 2010, were recruited. Health related QoL (HRQoL) was determined by using PedQL4.0 Generic Core scales, which is a 23-question instrument assessing the physical, emotional, social and school functioning domains.

RESULTS: A total of 44 children with BA (median age 7.4 years, range 2-18 years) were reviewed. Of these, 41 had Kasai surgery while three had primary LT without prior Kasai surgery. Another five children had LT after an unsuccessful surgery. The overall 2-year survival rate (native and transplanted liver) for BA improved from 40% in the years 1996-2005 to 52% in the years 2006-2010. The overall PedsQL score in children with BA who had a successful surgery was significantly lower than normal healthy control (85.6 vs. 91.6, P= 0.031). Further analysis of the PedsQL scores in the four domains failed to show any statistically significant difference. Presence of portal hypertension and frequent hospital admissions adversely affected the QoL in children with BA. No significant difference in the PedsQL scores between children who survived BA and those with CLD (85.6 vs. 87.1, P= 0.613), and those with CLD and healthy controls (87.1 vs. 91.6; P=0.07) was noted.

CONCLUSIONS: Children with BA have, on average, a 6-point lower total PedsQL score as compared to healthy controls. Children with BA who had a successful surgery required careful long term follow-up to monitor their quality of life, especially in those with portal hypertension and required frequent hospital admissions.