056

DETERIORATION IN NATIVE LIVER FUNCTION IN LONG-TERM SURVIVORS OF PORTOENTEROSTOMY FOR BILIARY ATRESIA IS VIRTUALLY A DEATH SENTENCE IN JAPAN

Takanori Ochi[1], Hiroki Nakamura[1], Momoko Wada[1], Tsuyoshi Tamura[1], Tadaharu Okazaki[1], Masahiko Urao[1], Yoichi Ishizaki[2], Seiji Kawasaki[2], Mureo Kasahara[3], Koichi Mizuta[4], Geoffrey J. Lane[1], Atsuyuki Yamataka[1]

[1]Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, Tokyo, Japan, [2]Department of Hepatobiliary-Pancreatic Surgery, Juntendo University School of Medicine, Tokyo, Japan, [3]Organ Transplantation Center, National Center for Child Health and Development, Tokyo, Japan, [4]Transplant Surgery Division, Department of Surgery, Jichi Medical University, Tochigi, Japan

Aim: We reviewed our post-portoenterostomy biliary atresia (BA) patients who required liver transplantation (LTx) for sudden deterioration in native liver (NL) function focusing on the fate of long-term survivors.

Methods: Indications for LTx, age at LTx, type of donor, and outcome were compared for patients less than 18 years old (U18; n=17) and long-term survivors aged 18 or older (18+; n=13) at the time of LTx. All patients initially achieved jaundice clearance (JC; TBil<1.2mg/dL).

Results: The table summarizes our findings. In U18, donors were living (LD) in 14, performed at a mean of 6.3 years and cadaveric (CD) in 3, performed at a mean of 1.3 years. All CDLTx were performed overseas after a waiting period of less than 2 months. All U18 cases were clinically stable after a mean of 13.2 years. In 18+, LDLTx (n=8) were performed at a mean of 27.7 years with distinctly different LD. One LD case died from graft versus host disease soon after LTx. CD were indicated for lack of age/size appropriate LD (n=5). Four died while awaiting LTx at a mean of 30.0 years after a mean of 1.4 years from the onset of sudden NL deterioration at a mean of 28.6 years. The remaining 37.2 years old case has been on a waiting list for CD for more than 6 months. Seven LDLTx cases in 18+ were clinically stable after a mean of 6.7 years. At the time of review, 24/30 (U18: n=17/17; 18+: n=7/13) were alive, 5/30 had died (U18: n=0/17; 18+: n=5/13), and 1/30 was awaiting LTx. Mortality rates were 0/17 (0%) in U18 and 5/13 (38.5%) in 18+ (p = .006).

Conclusion: Our results highlight the extremely grave prognosis for long-term BA patients requiring LTx when 18 or older because of poor donor availability in Japan.