Liver transplantation for treatment of biliary atresia

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When portoenterostomy (PE) was developed throughout the world after Morio Kasai’s description of the technique in 1968, undeniable progress was achieved in results of treatment of the so-called “uncorrectable” cases of biliary atresia (BA). Latest reports on long-term outcome do support evidence that an acceptable quality of life could thus be achieved for a few patients. But in spite of further improvement in results due to earlier diagnosis and operation, early or late liver failure remains a potential threat, with this disease of unknown etiology.

Since 1983, liver transplantation (LT) is an accepted mode of therapy for end-stage liver disease, and in children, BA has become rapidly one of the commonest indications. In the 1991 updated European Transplant Liver Registry, among all patients less than 15 years of age submitted to LT, nearly half (572/1162) suffered from BA. Our program of LT, initiated in 1988, presently includes 53 BA cases from a total of 92 children.

Compared to other indications, BA is the most challenging condition for LT. Most of these children are small, in poor nutritional condition in spite of specific care, exposed to esophageal bleeding, ascitis, and various infections. The transplant operation is complicated by previous surgery, portal hypertension, graft reduction and technical constraints to avoid postoperative arterial or portal thrombosis.

Leaving out the question of the yet rather short cyclosporin era (8 years), with unknown effects of both life-long immunosuppression and aging of the liver beyond childhood, two conclusions should be drawn: 1/ the good overall results of LT for BA compare favorably with those of PE, 2/ for the present time, all should be done at first to save the native liver.