

# Liver transplantation in children

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Editorial

Till recently the option of liver transplantation (LT) was not available in developing world due to the prohibitive costs, lack of expertise and sophisticated infrastructure.<sup>1,2</sup> Even though adult liver transplantation already has started in our country. The first ever liver transplant surgery in Bangladesh was done at Birdem Hospital on 3 June, 2010, which is indeed a milestone. The first recipient (42) and donor (29) are now living a normal life. Second liver transplantation also successfully done in BIRDEM, on August 6, 2011. Both the donor (38), and recipient (35) are now in good health. These cases prove that liver transplantation is possible in Bangladesh by using indigenous skills and infrastructure. Though it is difficult to start paediatric LT in our country but not impossible and now it is the optimum necessity to start paediatric LT here to save many child's life with liver failure.

Paediatric LT is now a routine treatment for end stage liver failure in developed countries with excellent 1 to 5 year survival.<sup>3</sup> It is not indicated if an acceptable alternative is available or if contraindications are present (eg, some cases of malignancy, terminal conditions, when expected outcome is poor.)

The first human liver transplant was performed in 1963 by a surgical team led by Dr. Thomas Starzl<sup>4</sup> of Denver, Colorado, United States. Dr. Starzl performed several additional transplants over the next few years before the first short-term success was achieved in 1967 with the first one-year survival post transplantation. Despite the development of viable surgical techniques, liver transplantation remained experimental through the 1970s, with one year patient survival in the vicinity of 25%. The introduction of ciclosporin by Sir Roy Calne markedly improved patient's outcomes, and during 1980s liver transplantation (with appropriate indications) was recognised as a standard clinical treatment for both adult and paediatric patients.<sup>5</sup> Since then, the evolution of immunosuppression and the development of new surgical approaches have led to the

establishment of 100 transplant centers in the United States. Surgeons currently perform more than 500 pediatric transplantations per year.<sup>6</sup> The first report of successful Living Donor Liver Transplant (LDLT) was by Dr. Christoph Broelsch at the University of Chicago Medical Center in November 1989, when two-year-old Alyssa Smith received a portion of her mother's liver.<sup>7</sup> Now LT also has been successfully extended to neonates.<sup>8</sup>

About 50% of the paediatric patients who require a liver transplant have biliary atresia. Other disease states that progress to end-stage liver disease among paediatric patients which require liver transplantation include metabolic disorders, progressive intrahepatic cholestasis and acute poisoning etc.

Examples of metabolic derangements include Wilson disease, alpha 1-antitrypsin deficiency, tyrosinemia and hemochromatosis. Other metabolic disease states leading to hepatic dysfunction include<sup>9</sup> Crigler-Najjar syndrome, Metabolic respiratory chain deficiencies, Familial hyper-cholesterolemia etc.

Donor liver for LT comes from two sources

a) Deceased Donor : A liver from a deceased donor must be transplanted in to the recipient within 12-18 hours. A team of surgeons and anaesthesiologists performs an operation to remove the liver from the donor. The liver is then preserved and packed for transport. These procedures are performed by using standard surgical practices and sterile techniques. Upon completion of the operation, the incisions are closed, and the donor's body is prepared for funeral or cremation.

b) Living Donor : A living person can donate a part of his/her liver (right or left lobe) to one of his/her relative. Eligibility of a person for liver donation is judged after detailed examination and tests by the transplant team, where donor safety is considered first. The criteria for a liver donation include:

- being in good health

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- having a blood type that matches or is compatible with the recipient's
- having a charitable desire of donation without financial motivation
- being between 18 and 60 years old
- being of similar or bigger size of liver than the recipient

It may be possible to remove up to 70% of the liver from a healthy living donor without harm in most cases. The donor's liver will regenerate approaching 100% function within 4-6 weeks, and will almost reach full volumetric size with recapitulation of the normal structure soon thereafter. Generally, living donors do not have any restrictions or specific medications or special diet as a result of liver donation.

The transplanted portion will reach full function and the appropriate size in the recipient as well, although it will take longer time than the donor.<sup>4</sup>

Virtually all liver transplants are done in an orthotopic fashion, that is, the native liver is removed and the new liver is placed in the same anatomic location. The transplant operation can be conceptualized as consisting of the hepatectomy (diseased liver removal) phase, the anhepatic (no liver) phase, and the postimplantation phase. This operation is critical and can take several hours is about 12 to 14 hours.

Following liver transplant surgery, patients frequently remain on a ventilator for the first 24-48 hours. Patients are moved out of the pediatric ICU (PICU) in a few days, depending on their recovery. Reintroduction of oral intake can begin within the week following surgery. Typically, hospital stays range from 1-2 weeks.

Immunosuppressive drugs are started immediately after liver transplant to prevent rejection. It must be maintained for the rest of the life. Initial cost is high for first few months, but subsequently reduces to one or two medicine at the end of the year and single medicine in 2-4 years time, which needs to be taken life long. Liver functions and drug level also needs to be evaluated repeatedly.

Prior to discharge of the patient the transplant team provides follow-up care and medication instructions. The patient's and caregivers' questions are answered, and signs of rejection are discussed with the patient in an age-appropriate manner and with the family. The patient and family should be instructed to continue a rehabilitation program that includes exercise, proper nutrition and the continuation of immunosuppression and other medications.

Most children with LT can now be expected to have long term survival. The dramatic increase in the survival rate throughout the 1990's in most International Centers has been sustained with most reporting one year actuarial

survival rates of >90% in elective patients and >70% in acute liver failure. Long term survival figures for 10-15 years are >80%.<sup>10,11</sup>

The success of paediatric liver transplantation in developed countries has increased the awareness of need for such procedures in the developing country like Bangladesh. For LT sufficient professional and public education is needed to develop organ donation. Living related liver donation is the only realistic option in our country in the absence of cadaveric donors. It is also essential to have the proper resources to develop the surgical, medical and multidisciplinary support necessary for the success of LT. Also, there is a need for professional education with regard to early and appropriate referral to optimize child's survival.

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