

Paediatric liver transplantation

Dr Jerome Loveland
Paediatric surgeon

Dr Jean Botha
Liver transplant surgeon,
Johannesburg

Background

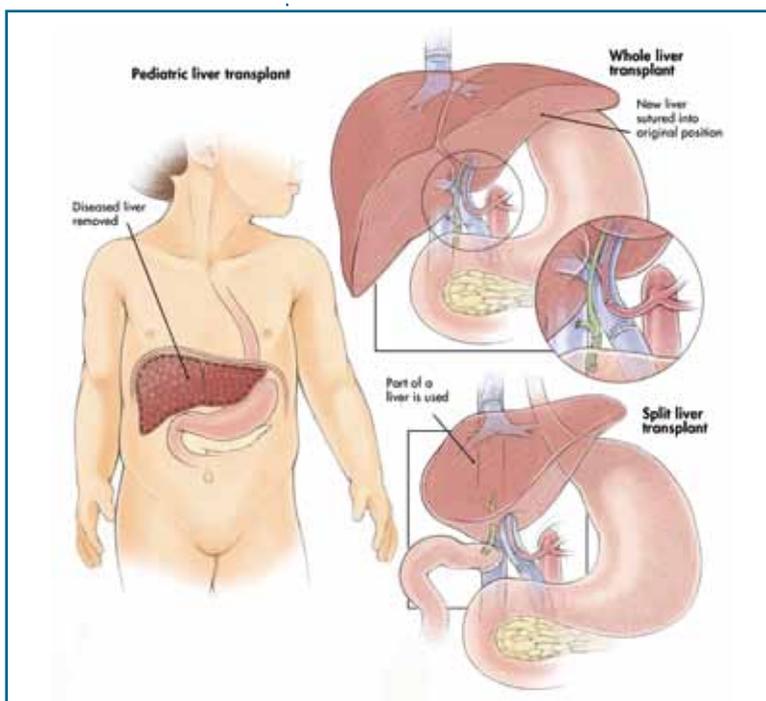
The Charlotte Maxeke Johannesburg Academic Hospital and Wits Donald Gordon Medical Centre (Wits DGMC), operating as a single enterprise through the University of the Witwatersrand, have established themselves as the leading centre in liver, kidney and simultaneous kidney-pancreas transplantation in Southern Africa, offering these services to both the adult and paediatric populations. Profs Myburgh and Du Plessis established renal transplantation at the University of the Witwatersrand in the 1960s, and the unit was also transiently involved in liver transplantation during its inception in the late 1980s. The division has run a highly successful renal programme in both the provincial and private sectors for many years, including a strong paediatric programme, which includes both deceased and related living donor components. The ambition to add liver transplantation to the ambit of the unit

was not supported by national government, which did not see the need for a second liver transplant unit in the academic sector in SA, feeling that the unit at Groote Schuur Hospital in Cape Town was adequate.

However, the university's ambition to expand coincided fortuitously with the establishment of the Wits DGMC, a public-private partnership between the University of the Witwatersrand and the private healthcare sector, which was identified as an ideal environment to launch Johannesburg's liver transplant unit. Initially catering only for the private patients (a strong criticism), the first liver transplant was performed in August 2004, and after much negotiating, both funded and indigent state patients are currently listed on a common waiting list, thereafter transplanted according to blood group and severity of disease. The unit has subsequently grown exponentially, developing into a highly successful programme that offers a near complete spectrum of transplant solutions to an ever-increasing recipient pool.

In excess of 200 patients have now received a liver transplant, including 34 children. In addition to the core renal and liver transplant programmes, the unit runs a simultaneous kidney-pancreas transplant programme, which has performed in excess of 80 transplants to date, and has introduced the laparoscopic harvest procedure into the related living donor (RLD) kidney transplant programme. We have performed 65 laparoscopic RLD harvests, and the procedure has undoubtedly been rapidly assimilated as the technique of choice.

While general opinion is often that transplantation is a purely surgical endeavour, the surgery is, in fact, only one component in an intricate process carried out by an enormous team of dedicated staff. It is this team that proudly defines our unit. Comprising donor and recipient transplant coordinators, psychologists, physiotherapists, allied medical professionals and an enormous administra-



Paediatric liver transplantation

tive staff, the team works continually on education of the public, donor recruitment and management, diagnosis of, and care during, end-stage disease, and work-up for the transplant itself. Pre- and post-operatively, a highly organised in-hospital component takes over, directing the medical and nursing care of the recipient. From a medical perspective, an essential yet less publicised aspect of the unit is the numerous ancillary aspects of medical care, including renal replacement therapy (dialysis), obligatory vascular and peritoneal access for the former, management of endocrine complications of chronic renal failure and diabetes, as well as the multidisciplinary management of end stage liver disease.

Paediatric liver transplantation

In the absence of related living donors, recipients listed in Johannesburg have had to rely on deceased donors as their sole source of organs, and until recently this remained one of the biggest limiting factors in our programme. The recent 'relaunch' of our paediatric liver programme highlights the work done to overcome many of the challenges that we face, having recently recruited a paediatric hepatologist back to SA and set up a sustainable paediatric intensive care service, both of which were factors in previously halting our progress. Finally, in addition to offering a deceased donor service, we are in the process of establishing a related living donor programme.

Thomas Starzl performed the first human liver transplant in a two-year-old child with biliary atresia in 1963. The patient died in the operating room of uncontrolled haemorrhage, and it was only four years later in 1967 that Starzl performed the first successful liver transplant. Since then, the technical aspects of transplantation have been developed and refined, and together with enormous advances in immunosuppression, five-year survival after transplantation approximates 80 per cent.

The evolution of paediatric liver transplantation has focussed on the refinement of surgical techniques to counteract the critical shortage of deceased donor organs, thereby expanding the available donor pool. This shortage is most profound in children, who require smaller grafts; thus availability of appropriately size-matched organs is obviously significantly less. Given the low number of paediatric donors, up to 50%

of children would die on the waiting list before receiving a transplant. To alleviate the pressure on deceased donor organs for young recipients, 'reduced' and then 'split' deceased graft liver transplants were performed in the 1980s, followed by the development of related living donation liver transplantation (LDLT).

The development of these techniques has almost eliminated waiting list mortality in children. Of note is that 80% of paediatric deaths caused by liver disease occur in children younger than two years old. LDLT offers several advantages over deceased donation, including reduction in the time on the waiting list, procurement under optimal conditions from a healthy donor, a shorter cold ischaemia time and elective scheduling of the operation. Results after LDLT are optimised as the process allows for transplantation to take place before the onset of life threatening complications and severe nutritional failure.

Liver cirrhosis secondary to biliary atresia is by far the most common cause of end-stage liver disease in the paediatric population, accounting for over 50% of the indications for liver transplantation in children. Our initial experience is consistent with this trend. As it happens, the first living donor liver transplant, in which segments II and III were procured from the mother and transplanted into a child with biliary atresia, was reported in 1988. The procurement usually involves removing the left lateral segment (segments II and III) along with the left branch of the portal vein, left hepatic artery and left hepatic vein. The recipient operation is similar to that of implantation of a cadaveric split left graft, where the IVC is preserved and the left hepatic vein is anastomosed to the recipient IVC, and the left portal vein of the graft is anastomosed to the main portal vein in the recipient.

The arterial anastomosis is slightly more difficult as only the left hepatic artery is available; this is usually anastomosed to the recipient common hepatic artery. The biliary anastomosis is technically difficult because it is performed at the level of the right or left hepatic duct. Biliary reconstruction is most commonly in the form of a Roux-en-Y hepaticojejunostomy due to the size mismatch of the ducts and the anatomical position and orientation of the left hepatic duct conferred by the size of the graft. In recipients transplanted for biliary atresia, the portal vein is typically hypo-plastic and narrow



The Wits Donald Gordon Medical Centre is a Private Academic Hospital providing quality multidisciplinary care!

The centre was established to enhance training of specialist and sub-specialists.

Established units of excellence include:

- Transplant Unit
- Paediatric and Adult Oncology
- Critical Care (ICU)
- Medical and Surgical Gastroenterology
- Endovascular Surgery
- Geriatric Medicine
- Arthroplasty
- The Wendy Applebaum Institute for Women's Health



Wits University
Donald Gordon
Medical Centre

Patient-centred. Independent. Academic.

MEDICLINIC 



Paediatric liver transplantation

as a result of recurrent cholangitis and the previous Kasai portoenterostomy. This usually means that the portal vein needs to be dissected back to the splenoportal junction and may require the use of vein grafts.

Conversely, the hepatic artery is usually unexpectedly large by comparison to the size of the child, making the arterial anastomosis relatively straightforward. Because children with biliary atresia have usually had prior abdominal surgery, operative blood loss and the risk of iatrogenic enterotomies is higher than in other recipients. There may also be associated cardiac and intestinal anomalies that need to be documented prior to transplantation so that optimal postoperative care can be rendered. While the left lateral segment (segments II and III), is a viable solution for many children, even this may be too large for children weighing less than 10kg. Monosegment transplantation appears to be a satisfactory option for such infants, where indicated. Either segment II or III can be transplanted with satisfactory results in very small children.

Primary non-function of a liver transplant is, thankfully, rare, but is nevertheless a devastating complication and needs to be recognised early so that appropriate management can be instituted and retransplantation offered. Similarly, hepatic artery thrombosis usually also leads to massive hepatic necrosis and allograft failure also necessitating retransplantation. If identified early, arterial reconstruction can be attempted with variable results. Portal vein thrombosis usually does not result in graft loss but needs to be corrected by thrombectomy and anastomotic revision when detected in the

early post transplant period. Particular to left lateral segment grafts is the increased risk of problems with the hepatic venous anastomosis which can sometimes result in an acute Budd Chiari syndrome; attention to technique will usually avoid this complication.

Biliary complications occur in 10-20% of paediatric living donor liver recipients. Bile leaks from either the cut surface of the liver or from the anastomosis are the most common. Cut surface leaks are mostly self-limiting and can be managed conservatively with drainage only, while anastomotic leaks may require reoperation and anastomotic revision. Anastomotic strictures can present in a delayed fashion and are usually managed by percutaneous methods and occasionally by revision of the anastomosis.

The typical living donor is usually a parent or first degree relative between the ages of 18 and 55 years with a compatible blood type. The donor undergoes thorough medical and psychological evaluation after which detailed imaging (CT or MRI) is performed to evaluate the potential graft size as well as vascular and biliary anatomy. Children are well served by receiving a left lateral segment graft.

Donor safety is the overriding concern and has been excellent after left lateral segmentectomy, and the risk of donor death after this procedure is 0.02-0.05%, a risk approaching that of donating a kidney. LDLT has been widely debated from both societal and ethical perspectives and has become an accepted procedure worldwide, especially for paediatric recipients. Donor mortality and morbidity rates are low, and recipient survival rates are between 80 and 90% at one year in experienced centres.

Conclusion

In addition to the RLD liver transplant arm (aimed initially at the paediatric population), current plans include expanding our unit to include a non-heart beating donor programme, and broadening the pancreas programme, looking at using a deceased donor pancreas with a simultaneous RLD kidney, pancreas after kidney and pancreas alone. In addition we are the first centre in South Africa to develop and institute a formal two-year surgical 'sub-specialty' training programme, covering all aspects of solid organ transplantation. We continue to work actively on the above projects.

The organ itself is the ultimate 'scarce resource'. Education of the public around the importance of organ donation remains an important focus in an attempt to produce a larger donor pool, particularly as SA runs an 'opt in' as opposed to 'opt out' donor policy, as in Spain, where all residents are donors unless otherwise specified. What defines the success of our unit is the smooth functioning of a large and diverse team, all of whom are experts in their respective fields. Probably our greatest attribute though is the fact that these skills are broadly distributed to more than a single individual in a particular discipline, allowing all aspects of the unit to function seamlessly in a continuum, allowing a deserving patient population continuous access to the organs that they require.