BRIEF REPORT

Auckland paediatric liver transplant experience 1990-2000

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Abstract

Aims. New Zealand is establishing its own Paediatric Liver Transplant Service. However there have been no readily available data on the experience of New Zealand paediatric transplant recipients to date. The aim of our study was to determine numbers and indications for transplant at present, current outcomes and to estimate the likely demand for the service in the future.

Methods. A retrospective search of computerised records was performed on children cared for at Starship Hospital from 1990 to 2000.

Results. Seventeen children received eighteen transplants. The indication for transplantation was biliary atresia in the majority of patients (11/17, 65%). A higher proportion of Maori and Pacific Island children received transplants than would be expected from their proportion in the population (59 vs 29%, p<0.01). Significant and often multiple complications occurred post transplantation in the majority of children, but overall outcomes were good.

Conclusions. A New Zealand Paediatric Liver Transplant Program is likely to perform about six transplants per year.

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The first paediatric liver transplant was performed in 1963 on a three year old with biliary atresia.[1] With improved perioperative management, surgical techniques and immunosuppression resulting in improved survival,[2,3] the indications for transplantation have expanded, and liver transplantation has become a treatment option for an increasing number of children.[4]

The first New Zealand child received a liver transplant in 1988. Since then most New Zealand children have been transplanted at the Queensland Liver Transplant Service, Brisbane. In 2000 the first paediatric liver transplant was performed in New Zealand and more are planned. However, there has been no registry of New Zealand paediatric recipients and therefore no readily available information on the likely demand for this service in New Zealand, or the workload implications. The aim of this study was to estimate likely demand, the indictions for transplantation, complications and outcome. These data would be useful to evaluate any changes with the establishment of a New Zealand paediatric liver transplant service.

Methods

The Queensland Liver Transplant Service were asked to provide details of New Zealand children transplanted. Recipients cared for at Starship Hospital were identified from computerised records of inpatient, day stay and outpatient attendances from July 1990 to April 2000. Data not available from the medical record were sought from the Queensland Liver Transplant Unit database or the New Zealand Liver Transplant Trust.

Results

A total of 54 New Zealand children have been referred to the Queensland liver transplant service. 47 liver transplants were performed and eight patients died post transplantation. Eighteen of these 47 transplants (38%) in seventeen children came from the Auckland region. This report concerns the seventeen children identified from searching the medical records as receiving their post-transplant care at Starship Hospital. These thirteen boys and four girls received eighteen transplants. One child required a second transplant for autoimmune hepatitis. One child was transplanted in New Zealand and the remainder in Brisbane. The median age at transplant was 2.4 years, (range 0.8-15). The median length of followup was 3.7 years (range 0.2-12).

The main indication for liver transplantation was biliary atresia (11/17, 65%). Other indications included one each of: α_1 antitrypsin deficiency, cryptogenic cirrhosis, hepatopulmonary syndrome, hepatoblastoma, non-syndromic bile duct paucity and fulminant hepatic failure.

Six Maori and four Pacific Island children received liver transplants. This is a significantly higher number than would be expected from the population, (59% versus 29% in the population under 15 years, $p< 0.01$[5]). Eight of these children received a transplant for biliary atresia, one for neonatal hepatitis and one for hepatoblastoma.

At the time of review fifteen children were more than one year post transplantation and sixteen were still alive (94%). The one year survival rate for child and graft was 93%. One child died at the transplant center six months post transplantation from multiple complications including chronic rejection, chronic lung disease, prematurity and cerebral atrophy.

Synthetic liver dysfunction and jaundice occurred in fifteen children prior to transplantation. The two children without synthetic liver dysfunction had hepatoblastoma and hepatopulmonary syndrome. Other complications of chronic liver disease included recurrent bleeding varices (3), cholangitis (8), ascites (5) and encephalopathy (1).

Only five children had an uncomplicated post transplantation course (Table 1). Common complications included biliary strictures (intra and extra hepatic) (6), vascular complications (hepatic artery thromboses (2), hepatic artery stricture (1), portal vein (2) and inferior vena cava thrombosis (1)) and infection (cholangitis (7), bacterial sepsis (1), viral (7)). Cholangitis, infection, thrombosis, and biliary strictures frequently occurred in the same child. Complications occurring less frequently included gastrointestinal protein loss, recurrent duodenal ulcer, autoimmune hepatitis and short stature. Complications presumed secondary to immunosuppressive medications included decreased glomerular filtration rate (7), renal impairment (3) and hypertension (3). Two children require anti-hypertensive medication. All children of school age were participating in a full time school program.

Discussion

This study provides the first collated information on the likely service demands for a New Zealand Paediatric liver transplant

Table 1. Complications of liver transplant. Multiple complications commonly occurred in the same patient.

Patient	Cholangitis	Infection	Thrombosis	Biliary stricture	Other
1	Cholangitis	Bacterial sepsis			
2		HHV6, EBV hepatitis			Autoimmune hepatitis
3			HA thrombosis	Biliary stricture	Gastrointestinal protein loss
4	Cholangitis		HA thrombosis	Biliary stricture	Recurrent duodenal ulcer
5		EBV, HHV6		Biliary stricture	
6					
7	Cholangitis			Biliary Stricture	
8					
9	Cholangitis	HHV6 hepatitis	PV thrombosis		Autoimmune hepatitis
10		EBV			
11	Cholangitis		HA stricture	Biliary stricture	
12	Cholangitis	CMV			
13					
14			IVC thrombosis		
15			PV thrombosis		
16					
17					Short stature

HA = hepatic artery, PV = portal vein, IVC = inferior vena cava, EBV = Epstein-Barr virus, HHV6 = Human herpes virus 6, CMV = cytomegalovirus.

service. Children in our study represent approximately one third of New Zealand paediatric liver transplants. Therefore a New Zealand service could expect to perform liver transplants on up to six children annually. Families transplanted in Queensland moved to Australia for 14 - 71 weeks (median 24 weeks). With transplants performed in New Zealand that time should be shorter and less disruptive.

This study may have underestimated the total number of New Zealand paediatric liver transplant recipients. The majority of transplants occurred in Brisbane, but we cannot exclude the possibility that individual health boards in New Zealand referred to other centres. Children cared for in Auckland may also not be representative of those elsewhere in the county. Although this was a retrospective review of medical records, it is unlikely that we missed significant complications. It is also unlikely that we missed earlier cases because we could not search computerised records prior to 1990, since outpatient records allowed us to detect the first New Zealand child to receive a liver transplant in 1988.

Biliary atresia was the indication for transplantation in the majority of our children (65%). This is consistent with the indications reported elsewhere.[6,7] However, there was an excess of Maori or Pacific Island children amongst Auckland paediatric transplant recipients. This over-representation has previously been reported in patients with biliary atresia (58%),[8] and eight of the ten Maori and Pacific Island patients in our review had biliary atresia. We speculate that this may lead to a higher number of transplants in New Zealand than might be predicted on a population basis.

In this study survival one year post transplantation was 93% for child and graft. This is similar to one year survival rates in larger series (now approach 90%) with four to eight year survival rates of 70-85%.[6,9-12] We were concerned that managing transplant recipients in a setting geographically isolated from the transplant center may have adversely affected outcome, but our data suggest that this is not the case.

Although most complications associated with significant mortality occur in the first few days and weeks post transplant, many important complications develop later. Our finding that more than two thirds of children have significant complications post transplantation is consistent with that of larger centers elsewhere.[9] Currently when New Zealand transplant recipients develop complications, management questions are predominantly directed to their transplant center. We predict that with the establishment of the New Zealand paediatric transplant program, more referrals of existing transplant recipients will be made to Starship Hospital, resulting in an increase in workload out of proportion to the number of transplants performed.

This review provides the first collated information about New Zealand paediatric liver transplant recipients, who have had a high survival rate and a good quality of life. The main indication for transplantation was biliary atresia. There was over-representation of Maori and Pacific Island recipients. Most recipients experienced significant pre and post transplantation complications. A New Zealand Paediatric Liver Transplant Program is likely to perform six transplants per year and the workload may increase out of proportion to the number of transplants. This study provides baseline data for ongoing monitoring of the New Zealand Paediatric Liver Transplant Program.

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