

AUSTRALIA & NEW ZEALAND

LIVER TRANSPLANT REGISTRY



From the Combined Registries
of the Australian and New Zealand
Liver Transplant Centres



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STATISTICAL METHODS

Kaplan-Meier survival curves have been produced using SPSS® for Windows™ Release 12.0.1, SPSS Inc.

ACKNOWLEDGMENT

The Cancer Registry is maintained at Transplantation Services, Royal Prince Alfred Hospital, Sydney. Report prepared by Pamela Dilworth, Dr Deborah Verran, Dr Graham Stewart, Patrick Tang.

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Preface

We are pleased to present the 17th Report of the Australia and New Zealand Liver Transplant Registry (ANZLTR). This report contains data to the 31st December 2005 and analyses the cumulative data since the establishment of the first liver transplantation unit in Australia or New Zealand in 1985. This is the first to be based on data for the calendar year and brings the ANZLTR into line with all other organ transplant registries in Australia and New Zealand.

The Australia and New Zealand Liver Transplant Registry (ANZLTR) is a collaborative effort of the liver transplantation centres in Australia (Adelaide, Brisbane, Melbourne, Perth, Sydney) and New Zealand (Auckland). The Registry is supervised by the Management Committee who are involved in the ongoing supervision of the development of the Registry. The members of the Management Committee are listed on the inside cover.

Donor data have been supplied by the Australia and New Zealand Organ Donor Registry and we thank them for their collaboration.

The Editors would also like to thank the staff of all the Liver Transplant Units who now contribute their data by direct entry into the ANZLTR database. A full list of the Units and their contact information can be found in Appendix I. In particular we are grateful to the efforts of Pamela Dilworth, Program Manager for her continuing contribution to the maintenance of the Cancer Registry which is based at the Royal Prince Alfred Hospital, Sydney and who together with Dr Deborah Verran and Dr Graham Stewart prepare the Cancer Report.

The registry now has some financial support and we are grateful to the Commonwealth Department of Health and Aging for their financial contribution.

Comments are always welcome and should be forwarded to the Coordinating Centre at the contact information listed on the inside cover as should requests for further copies of this Report. The report is now also available on the ANZLTR public web site www.anzltr.org from where the report can be downloaded. Slides are available on request from the Coordinating Centre.

Stephen Lynch Glenda Balderson

Summary

Page

- 5. Between January 1985 and 31st December 2005, 2692 orthotopic liver transplants (OLT) were performed in Australia and New Zealand on 2498 patients 2033 adult patients (> 15 years) [81%] and 465 children [19%]. The median age of all recipients was 45.5 years. The ages ranged from 24 days to 73.1 years. There is a significant difference in gender distribution between children (M=46.5%) and adults (M=62.5%)
- 6. There was a decrease in the total number of new adult patients transplanted in 2005 compared with 2004 but an increase in the number of paediatric patients.
- 7. The trend to increasing age of adult recipients in recent years continued and the overall adult median age is now 48.7 years. The median age of new adult recipients in 2005 was 52.8 years.
- 8-9. Fewer transplants were performed in 2005 then in 2004. Split grafts now make a significant contribution to the total number of paediatric transplants performed providing 15 of 33 [45%] grafts in 2005 and 86 of 532 [16%] overall. In children, other reduced size grafts have been used in 266 [50%] cases including 13 living donor grafts. One child has been treated with liver cell implantation. Of adult patients, 121 have received reduced size grafts 97 split liver grafts (including 1 as auxiliary graft), 24 other reduced size grafts (1 as auxiliary graft) and 2 living donor grafts. One domino transplant of a whole liver has been performed.
- 10-11 Overall chronic viral hepatitis (CVH) is the most common primary indication for liver transplantation. In children biliary atresia (BA) is the most common primary disease. In adults chronic viral hepatitis is the primary disease in 28% of recipients. Full details of specific diagnoses categories by age group are listed in the Appendices Metabolic disorders (Appendix II), Other diseases (Appendix III), Fulminant Hepatic Failure (Appendix IV).
- 12-14. The predicted increase in the proportion of adult patients requiring transplantation for CVH is evident in recent data. In the era 2000-04, 34% of adult patients had a primary diagnosis of CVH, 25% with Hepatitis C, 8% Hepatitis B and 2% both Hepatitis B,C,D. In 2005 38% of new adult patients had a diagnosis of CVH. When patients with either primary or secondary diagnosis of Hepatitis B,C or both are included, the overall incidence of CVH in new adult patients in 2005 was 50%.
- 15. Current 1 year patient survival of all patients is 86%, 79% at 5 years and 69% at 10 years. Children had a significantly better survival rate then adults.
- 16. Whilst older children had superior survival then infants and babies, older adult recipients (60-65 and >65 years) had poorer outcomes.
- 17-18. Patient survival in 2000-04 cohort shows continued improvement in outcome for the first 5 years compared with earlier cohorts. This is seen in both children and adults.
- 19. The type of primary graft, (whole, reduced or split liver), had no effect on patient survival in either children or adults.
- 20. Children weighing < 8 kg at the time of transplant had inferior early survival compared to heavier patients.
- 21. Adult patients transplanted for biliary atresia had the best long term survival while those whose primary disease was malignancy had a significantly lower survival rate. Longer term survival for patients transplanted for Hepatitis C was also lower.

Summary

- 22. In children, patient survival was similar for all disease groups. There were no differences in survival between adults and children transplanted for fulminant hepatic failure [acute and subacute] with 5 year survival of 73%.
- 23. Recent cohorts of adult patients with a primary diagnosis of hepatitis B show a significantly improved survival which is not seen in adult patients with hepatitis C as primary disease. Patient transplanted for malignancy continue to have a poor outcome.
- 24. Sixteen patients have received a living donor graft, 12 children and 3 adults as a primary graft and one child as a second graft. Graft survival is 70% at 10 years.
- 25. Graft survival was significantly worse in second and third grafts.
- 26. Both split and other reduced grafts had lower graft survival in the early post-transplant years in both children and adults but had an improving longer term outcome particularly for split grafts.
- 27. Vascular complications and rejection were the commonest indications for retransplantation. Ten percent of retransplants were due to poor early graft function. Recurrent disease was the indication for retransplantation in 10% of cases [3% PSC,PBC and 7% HBV,HCV].
- 28-29. Overall, sepsis is the most frequent cause of death. Full details of Miscellaneous and Other graft failure deaths are listed in Appendix V. Forty-six percent of all deaths occurred within 6 months of transplant. Early graft failure was due to poor or no early graft function. By 1 year malignancy and graft failure from recurrent disease or chronic rejection cause most deaths.
- 30. There was a decline in number of cadaveric donors in 2005 resulting in fewer transplants then 2004. Fourteen cadaveric grafts were split in 2005.
- 31. Donor age has significantly increased in recent years. Long term graft survival was lower in grafts from donors aged 61-65y but not those aged over 65y.
- 32-33 The numbers of patients waiting for transplant continues to increase. At the end of 2005 141 patients were awaiting a transplant up from 117 at 31st December 2004. Delistings due to death, becoming too ill or tumour (HCC) progression increased from 6% in 2004 to 11% of patients listed in 2005. Waiting times are increasing with some patients waiting years to receive a graft.
- 34-35. Four hundred and thirty eight patients (18%) have had a pre- or post-transplant cancer. One hundred and nine (4%) of patients were transplanted for liver malignancy and 28 [26% of these patients] have died from this cancer. The type of liver tumour had no influence on survival.
- 36-37. Two hundred and forty five patients had liver cancer as a secondary diagnosis with hepatocellular carcinoma the most common. However those with cholangiocarcinoma had significantly poorer survival.
- 38. De novo non skin cancers (123) have developed in 119 (5%) patients and 49 [41%] have died from this cancer. Cancers of the alimentary tract and lymphoma predominate. Lower GI cancers account for 60% of alimentary tract cancers.
- 39. Patients with either de novo non skin cancers or liver cancers have significantly worse long term survival.
- 40. Alimentary tract cancers are the most common irrespective of the pre transplant liver disease.
- 41. Two hundred and eighty five (11%) patients have developed 1619 skin cancers with 148 patients having multiple skin cancer types. The cumulative risk of diagnosis on any cancer post transplant is approaching 40% by 20 years.

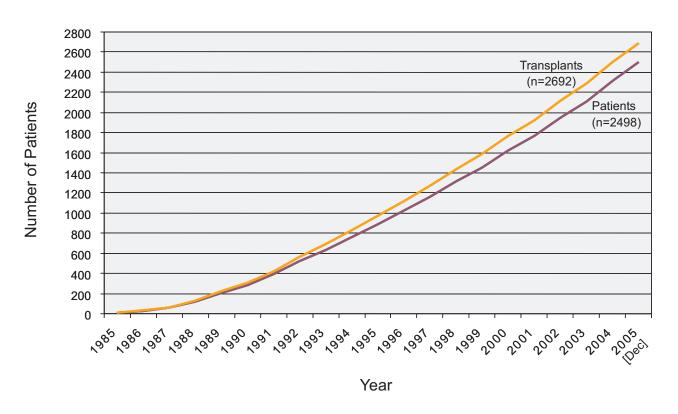


Section 1

Demographic Data







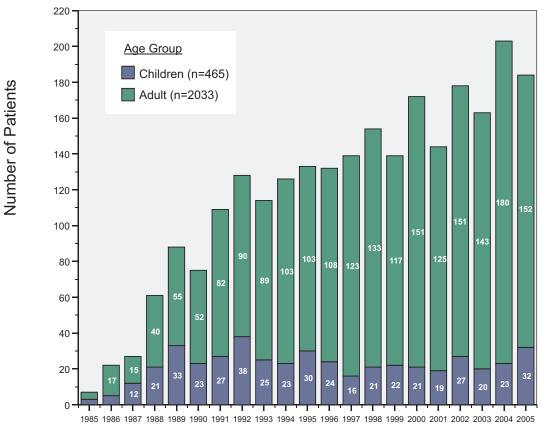
Summary Statistics - Age and Gender

ALL PATIENTS

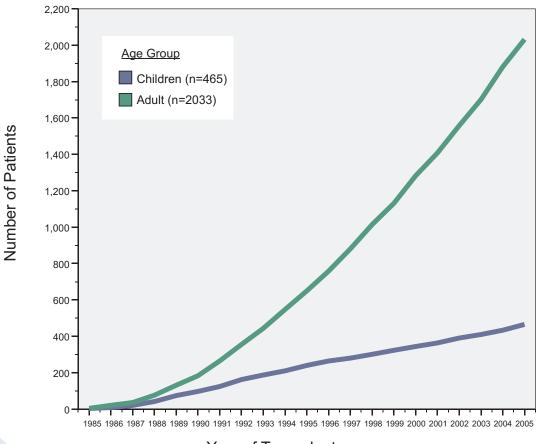
| | Children | Adults | Total | | | | |
|-----------|-------------|--------------|-------------|--|--|--|--|
| Patients | 465 | 2033 | 2498 | | | | |
| Age | je | | | | | | |
| Mean ± SD | 4.4 ± 4.2 | 47 ± 11.9 | 39.1 ± 19.8 | | | | |
| Median | 2.5y | 48.7y | 45.5y | | | | |
| Range | 24d -14.9y | 15.0 - 73.1y | 24d - 73.1y | | | | |
| Gender | | | | | | | |
| Female | 249 (53.5%) | 761 (37.5%) | 1010 (40%) | | | | |
| Male | 216 (46.5%) | 1272 (62.5%) | 1488 (60%) | | | | |
| Surviving | 366 (79%) | 1506 (74%) | 1872 (75%) | | | | |



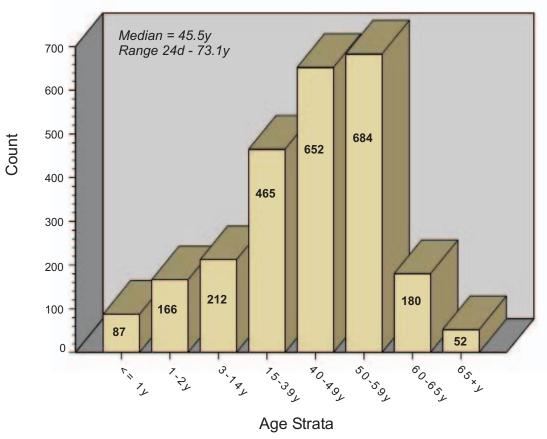
SECTION 1 : DEMOGRAPHIC DATA



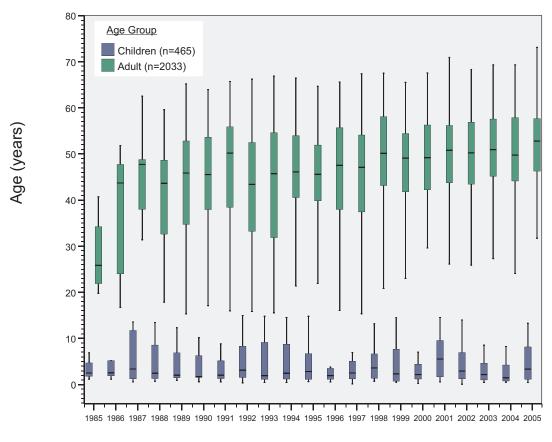
Cumulative Number of New Patients





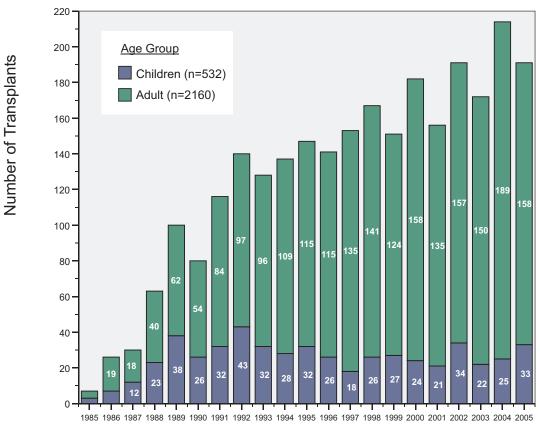


Age at Primary Transplant by Year

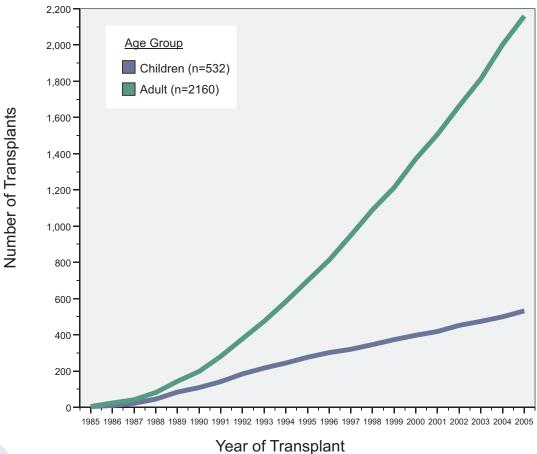




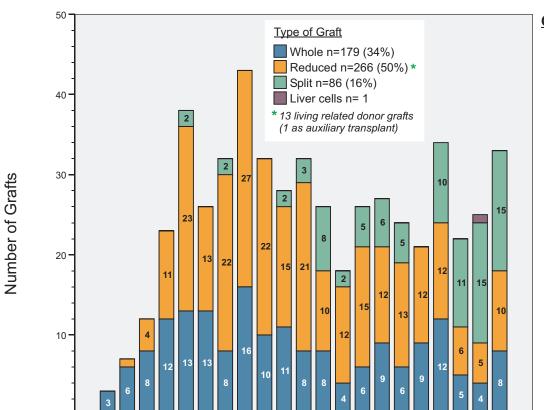




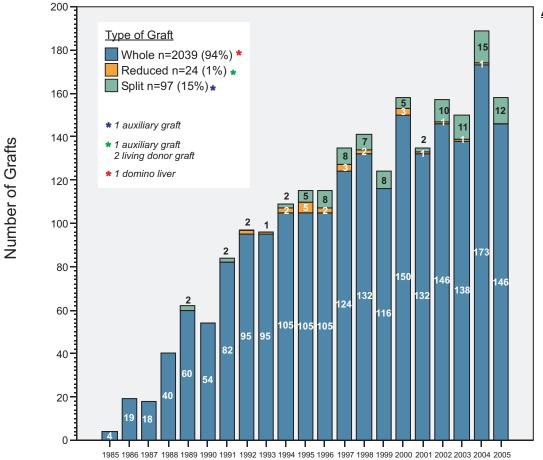
Cumulative Number of Transplants







Children (n = 532)



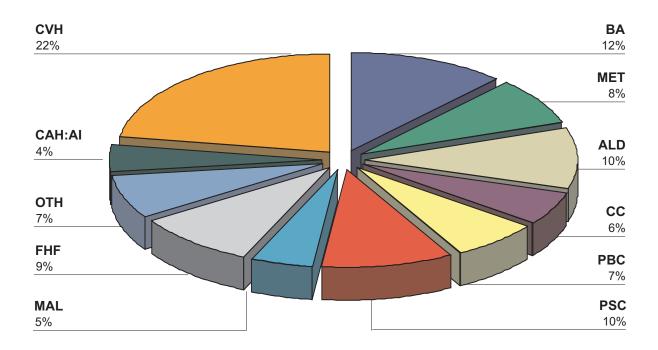
Adults (n = 2160)

Section 2

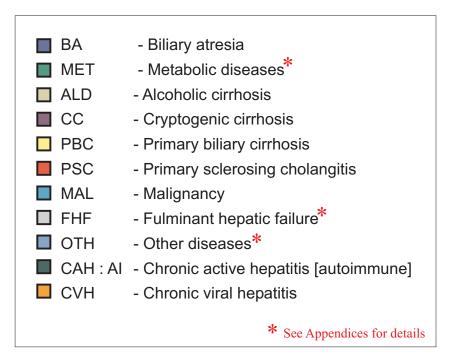
Primary Diagnosis





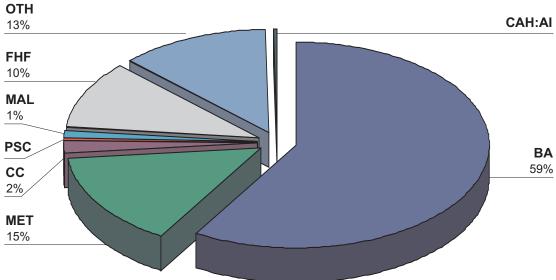


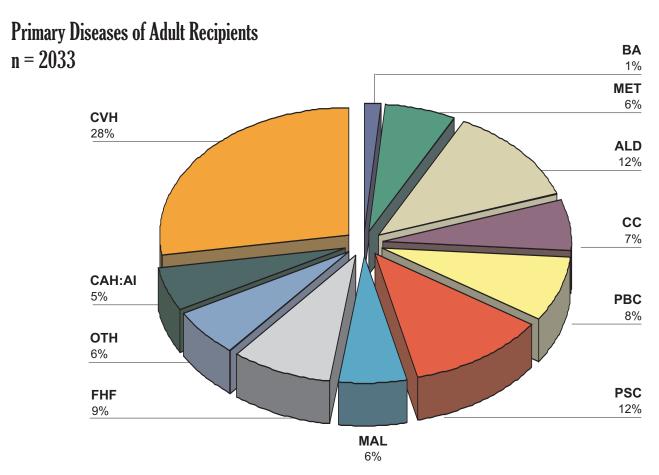
Diagnosis Group

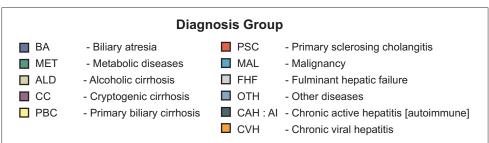


Primary Diseases of Children n = 465

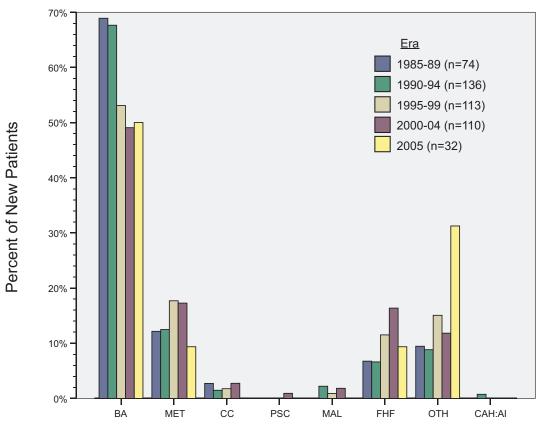




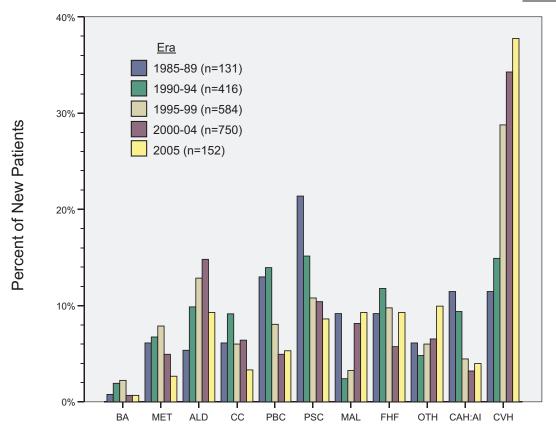




Children (n=465)



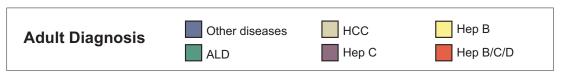
Adults (n = 2033)

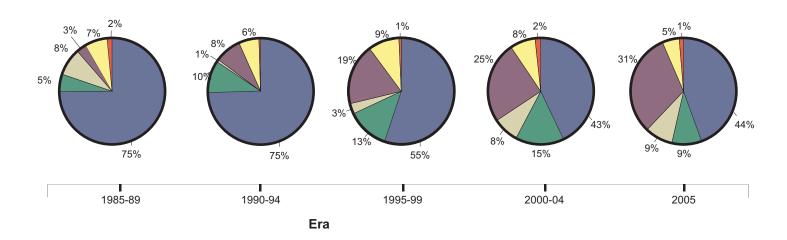


Primary Diagnosis Group

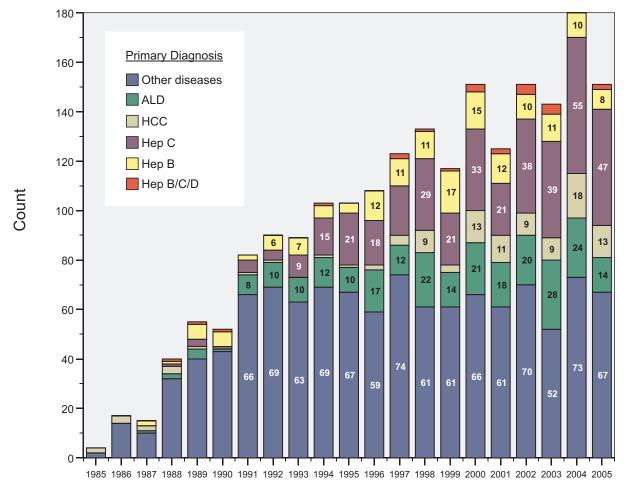








Adult Primary Diagnosis by Year



Year of Transplant

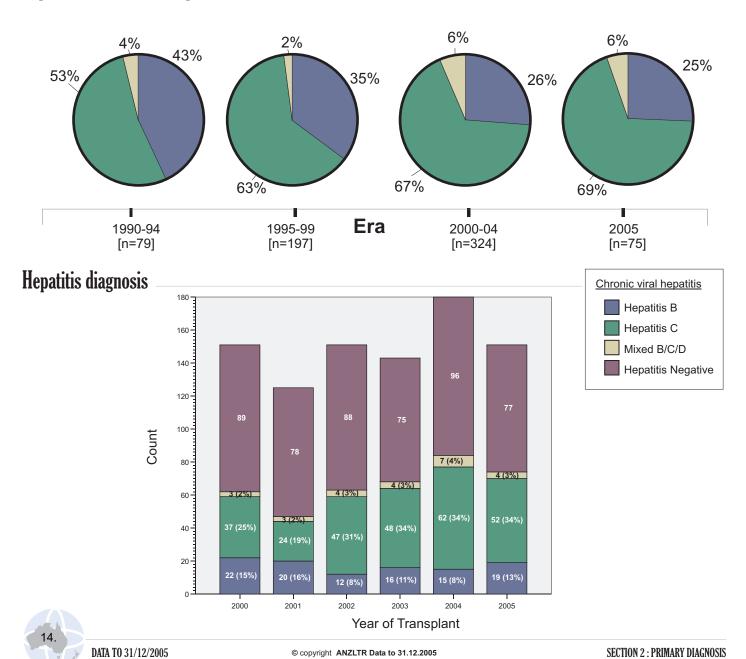


Chronic Viral Hepatitis as Primary or Secondary Diagnosis in Adult Patients



| | | | Secondary / Tertiary diagnosis | | | | | |
|-------------------|-----------------|------|--------------------------------|-------------|------------------|-----|-----|--|
| ra | | n= | Hepatitis C | Hepatitis B | Hepatitis B,C | НСС | ALD | |
| Primary Diagnosis | Hepatitis C | 381 | | 2 | | 73 | 89 | |
| | Hepatitis B | 156 | 3 | | | 42 | 4 | |
| | Hepatitis | 23 | | | | 2 | 2 | |
| | BD/BC/BCD | | | | | | | |
| rim | HCC + cirrhosis | 106 | 43 | 38 | 4 | | 4 | |
| P | ALD | 248 | 9 | 2 | | 21 | | |
| | Other | 1119 | 11 | 3 | | 29 | 15 | |
| | TOTAL | 2033 | | | | | | |

Type of Chronic Viral Hepatitis in Adult Patients

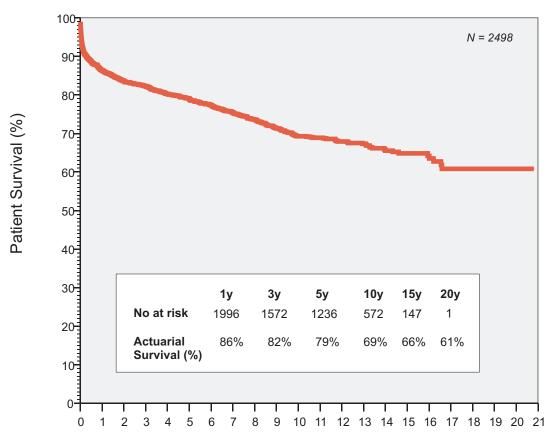


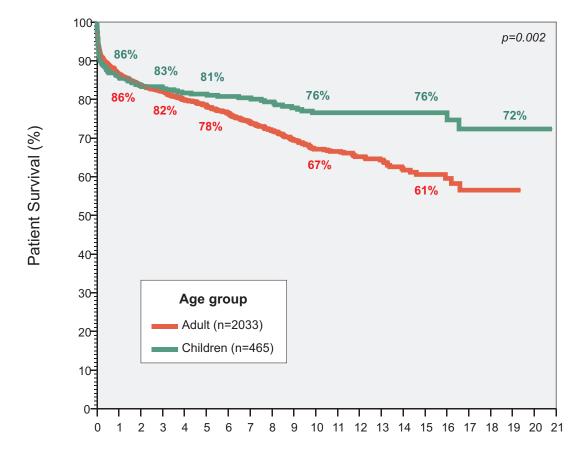


Section 3

Patient Survival

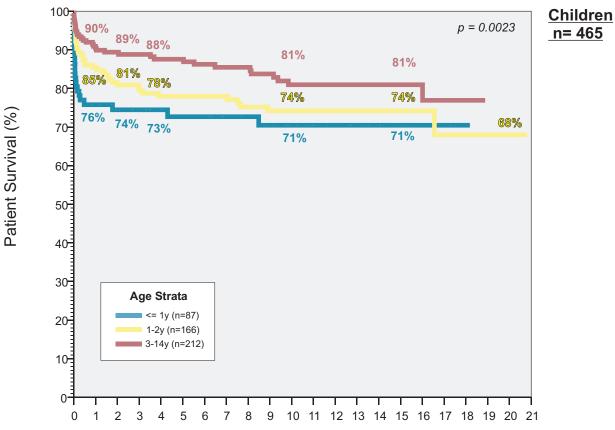


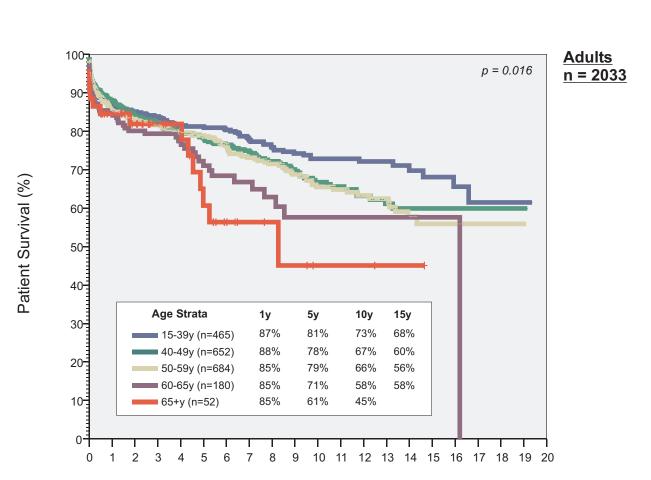




Time Post-transplant (years)

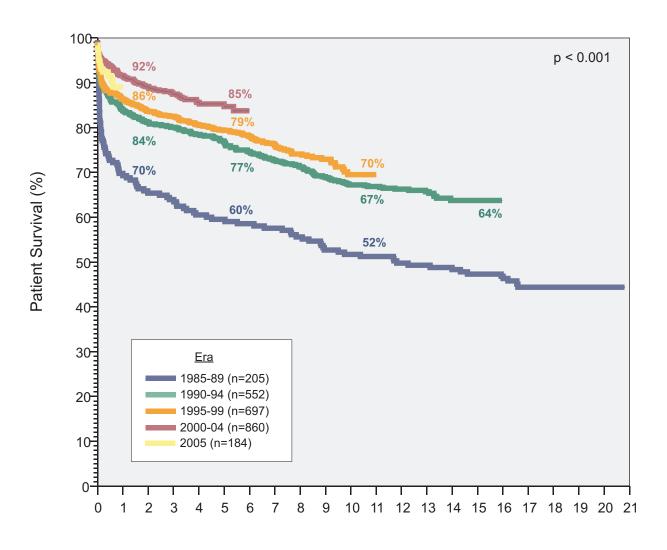




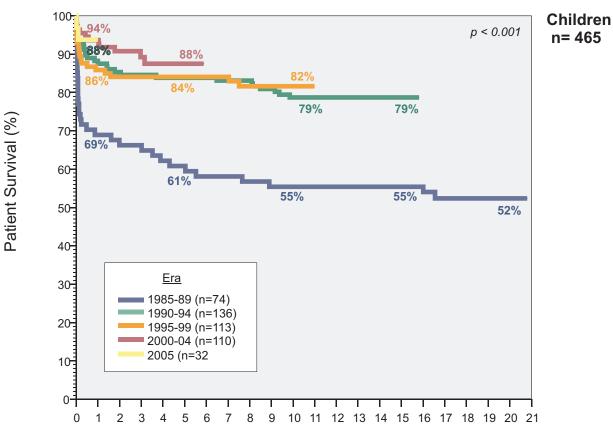


Time Post-transplant (years)

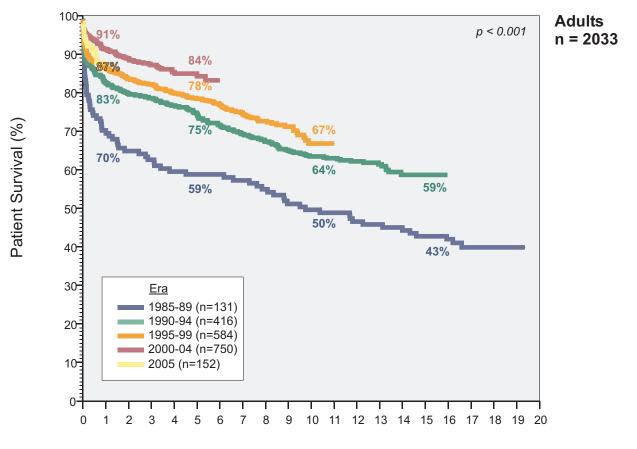




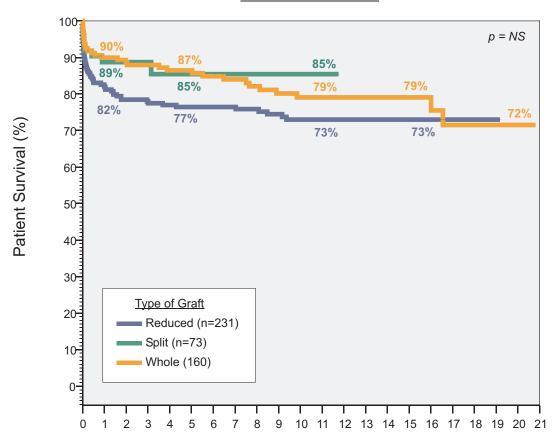
Time Post-transplant (years)



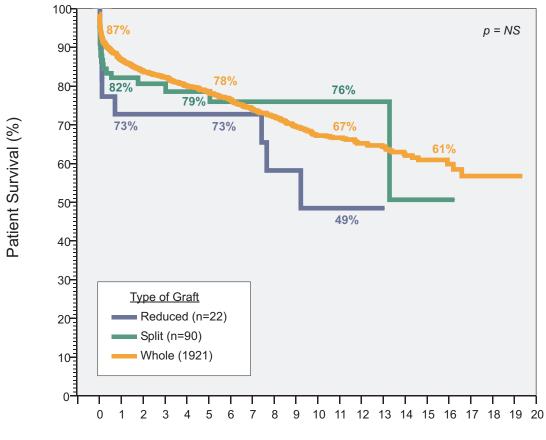
Patient Survival - Adults

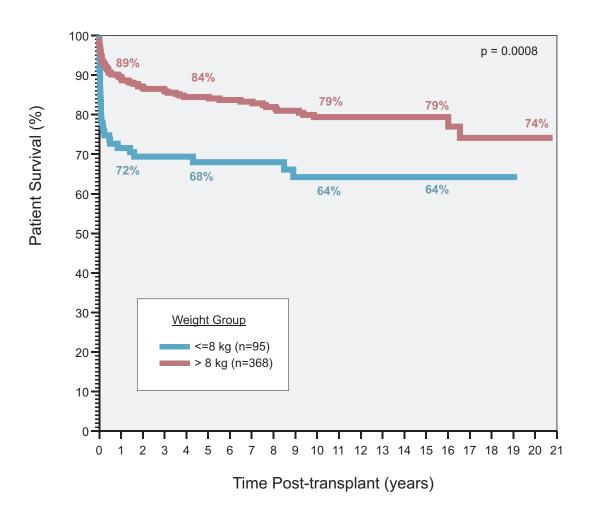


Children - n = 464

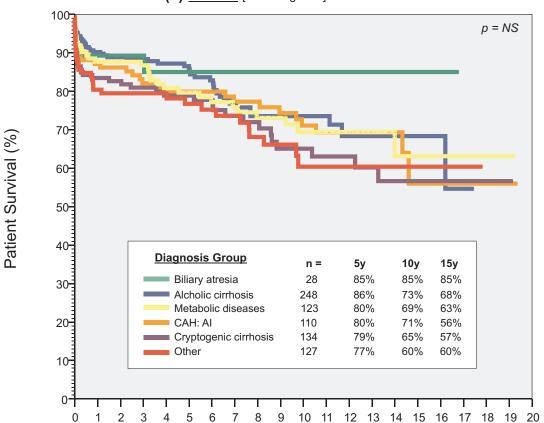


Adults - n = 2033

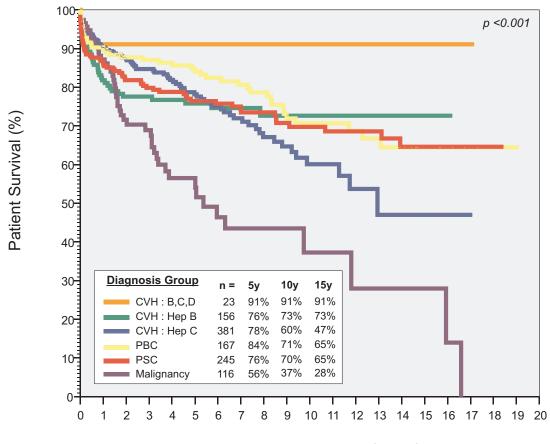




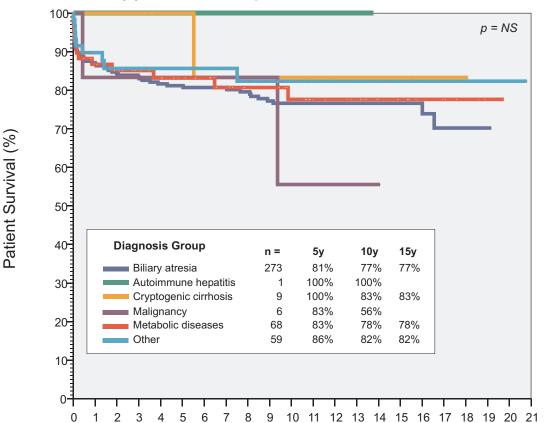
(1) Adults [excluding FHF] - n=770



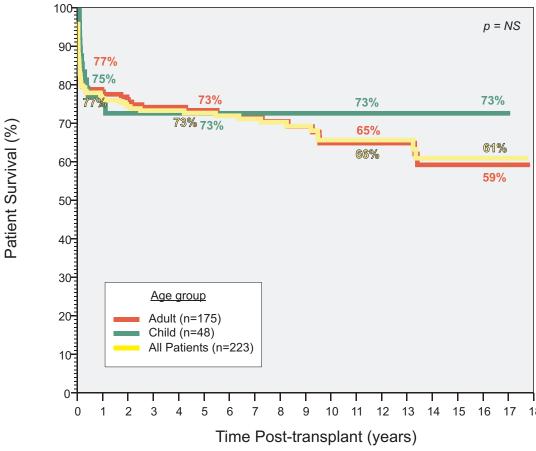
(2) Adults [excluding FHF] - n=1088

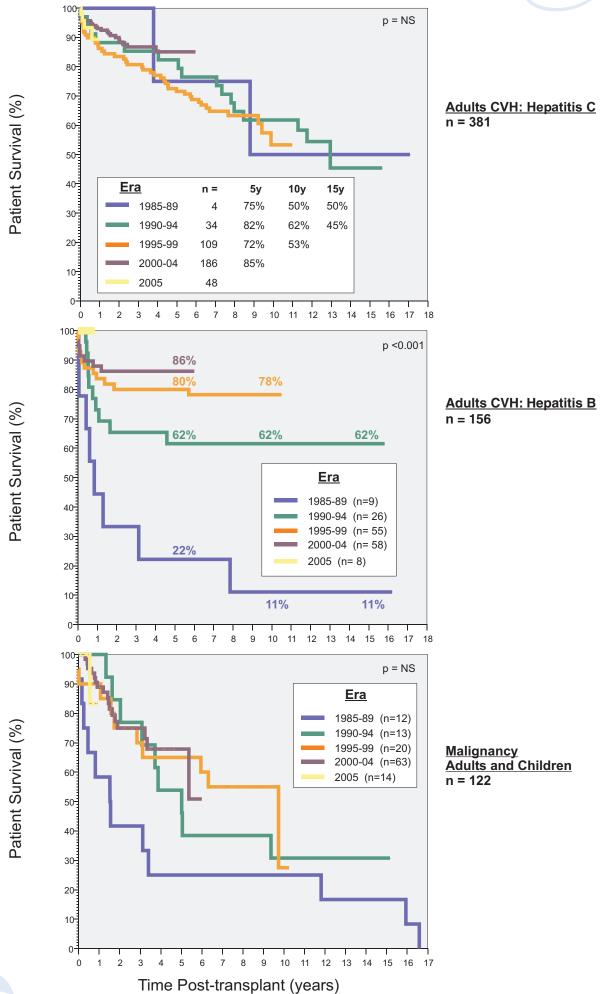


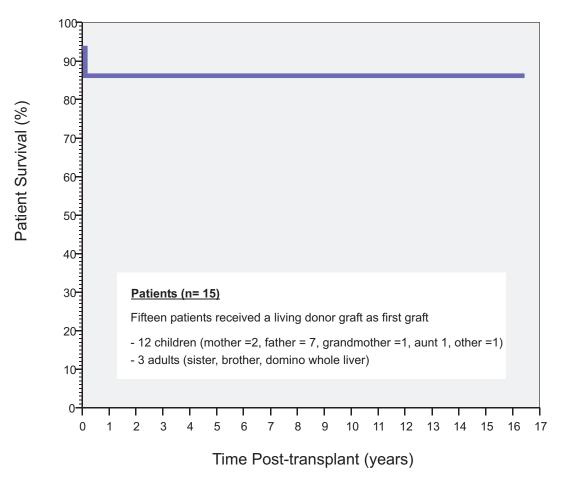
(3) Paediatric recipients [excluding FHF] - n=417

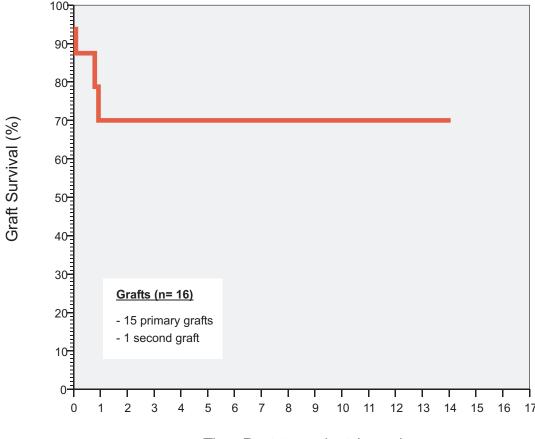


(4) Fulminant hepatic failure (n=223)









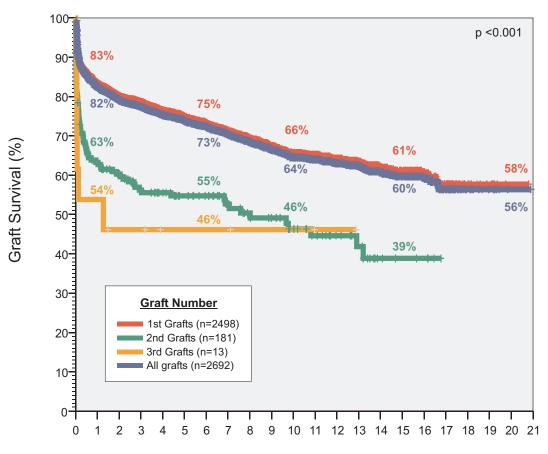
Time Post-transplant (years)

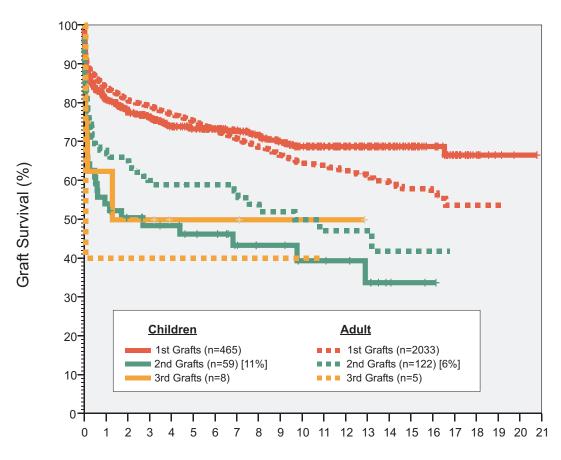


Section 4

Graft Outcome

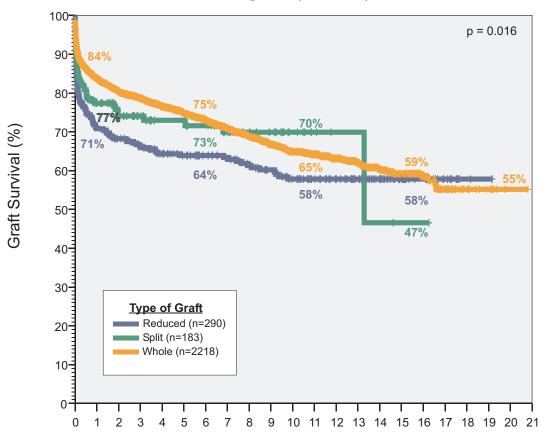


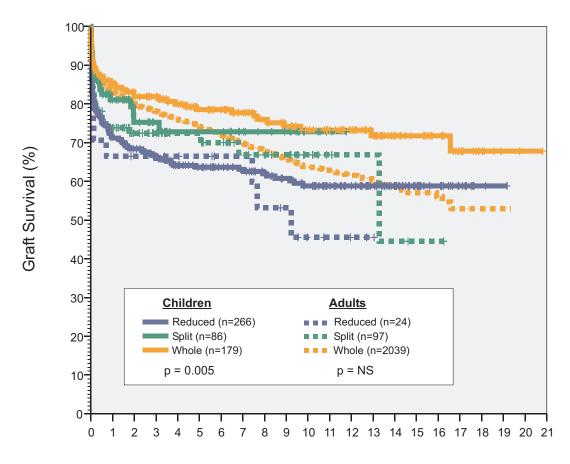






All grafts (n = 2691)

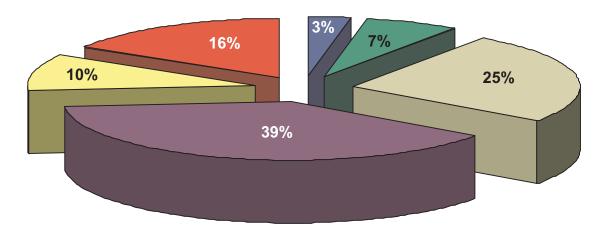


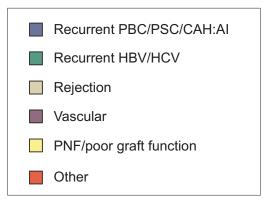


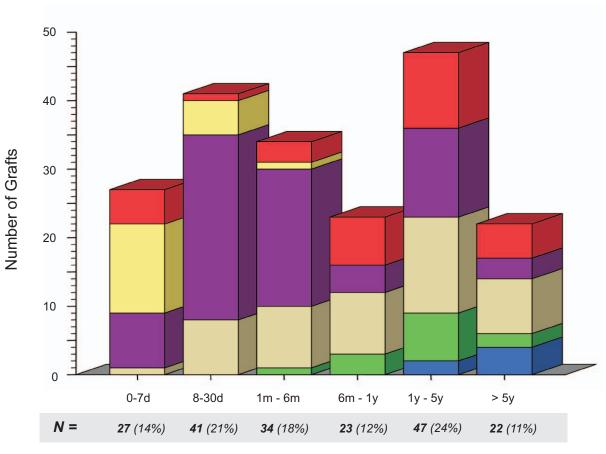
Indication for Retransplantation

 $\mathbf{n} = 194$ (181 2nd grafts, 13 3rd grafts)











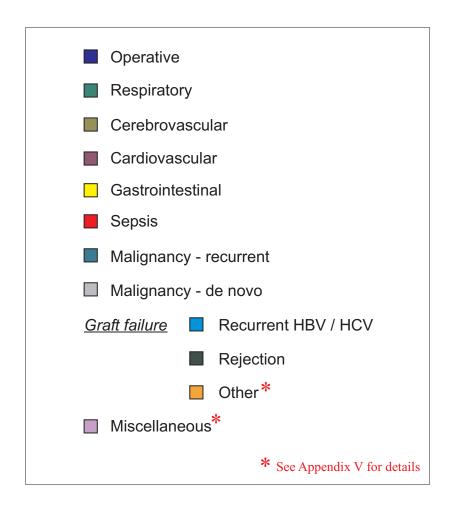
Section 5

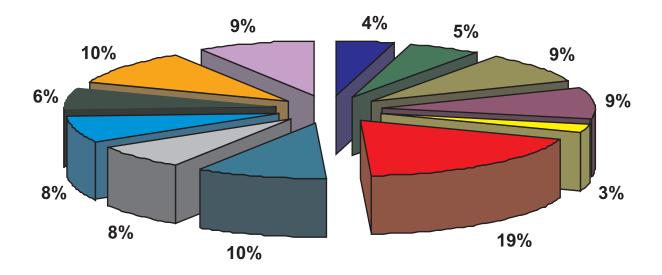
Cause of Patient Death



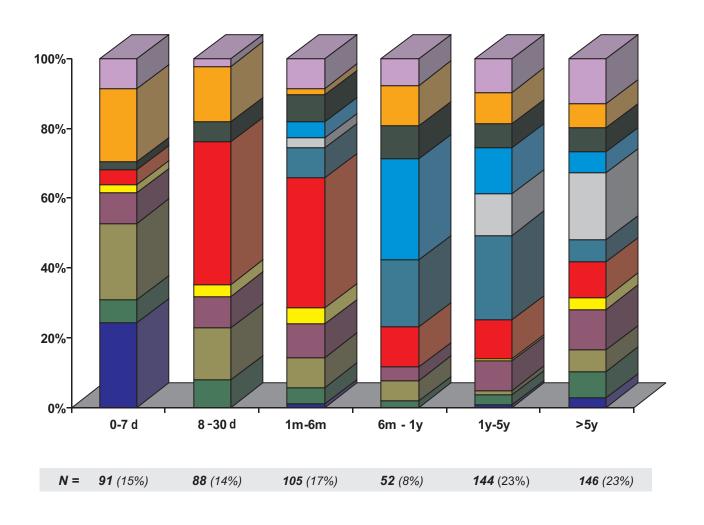


All Patients n = 626













Section 6

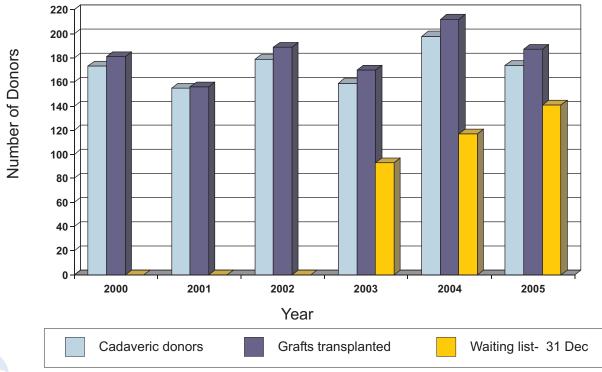
Donor Information

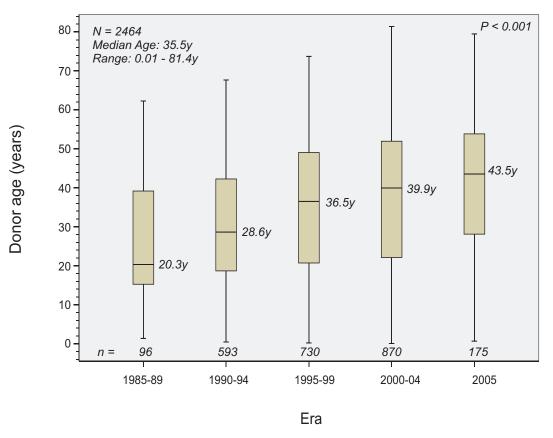




| | QLD | NSW/ACT | VIC/TAS | SA/NT | WA | NZ | TOTAL |
|------|-----|---------|---------|-------|----|----|-------|
| 1990 | 22 | 27 | 16 | 5 | | 7 | 77 |
| 1991 | 29 | 35 | 20 | 6 | 8 | 11 | 109 |
| 1992 | 43 | 32 | 18 | 9 | 8 | 24 | 134 |
| 1993 | 28 | 40 | 25 | 12 | 6 | 16 | 127 |
| 1994 | 29 | 39 | 23 | 12 | 10 | 21 | 134 |
| 1995 | 29 | 44 | 24 | 17 | 8 | 21 | 143 |
| 1996 | 26 | 37 | 19 | 17 | 10 | 24 | 133 |
| 1997 | 31 | 49 | 19 | 19 | 8 | 22 | 148 |
| 1998 | 29 | 44 | 27 | 22 | 13 | 27 | 162 |
| 1999 | 15 | 31 | 31 | 29 | 11 | 27 | 144 |
| 2000 | 26 | 51 | 26 | 24 | 12 | 34 | 173 |
| 2001 | 37 | 40 | 26 | 14 | 9 | 29 | 155 |
| 2002 | 34 | 42 | 38 | 24 | 11 | 30 | 179 |
| 2003 | 34 | 32/3 | 29/2 | 13 | 15 | 31 | 159 |
| 2004 | 30 | 49/4 | 35/1 | 26/1 | 17 | 35 | 198 |
| 2005 | 24 | 36/8 | 38/2 | 17/3 | 25 | 21 | 174 |

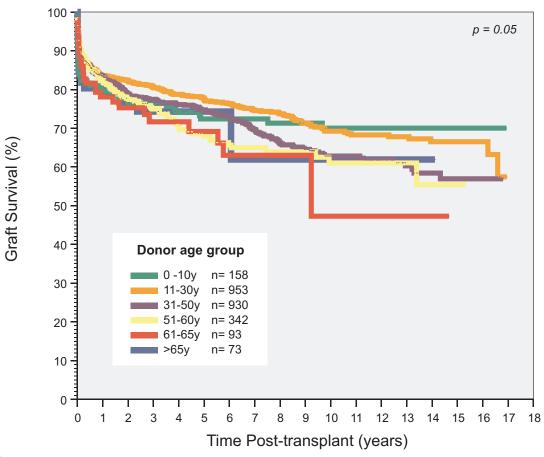
Grafts from cadaver donors





Graft Survival by Donor Age

N = 2464



Section 7

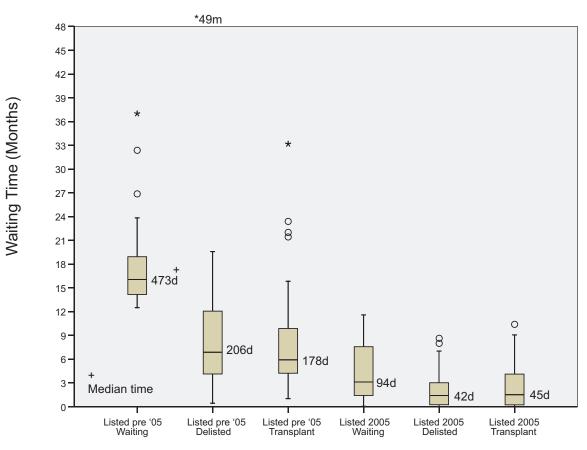
Waiting List





| Activity 2004 | | Activity 2005 | | | |
|---|-------------------------|----------------------------|------------------------|----------------|----------------------|
| Listed at 31/12/2003 Listed in 2004 TOTAL | 93 279 372 | | Listed at 31/12/04 117 | Listed in 2005 | TOTAL 2005 404 |
| OUTCOME | | | | | |
| Transplant in 2004 | 214 [58%] | Transplant in 2005 | 76 | 115 | 191 [47%] |
| Delisted in 2004 | 41[10%] | Delisted in 2005 | 20 | 52 | 72 [18%] |
| Died on list | 14) | Died on list | 6 | 20 | 26) |
| Too sick | 8 6% | Too sick | 2 | 7 | 9 \ [11%] |
| Tumour progression | 2) | Tumour progression | 1 | 8 | 9) |
| Improved | 8 | Improved | 5 | 10 | 15 |
| Other | 9 | Other | 6 | 7 | 13 |
| Still listed at 31/12/2004 | 117 [32%] | Still listed at 31/12/2005 | 21 | 120 | 141 [35%] |

Waiting Time by Outcome



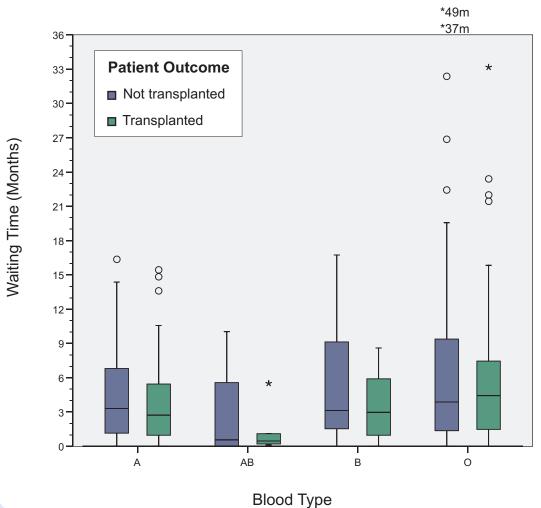




| | Blood Group | | | | | |
|------------------|-------------|-----------|----------|---------|-------|--|
| | Α | 0 | В | AB | TOTAL | |
| n= | 148 (37%)* | 191 (47%) | 56 (14%) | 9 (2%) | 404 | |
| Not transplanted | 76 | 108 | 25 | 4 | 213 | |
| Transplanted | 72 (49%)** | 83 (44%) | 31 (55%) | 5 (56%) | 191 | |

[%] of total number listed

Waiting Time by Outcome & Blood Group



^{** %} of blood group

Section 8

Liver Transplantation and Cancer



Types of Cancer in Liver Transplant Recipients N=2497



| AT Tx | | |
|--------------------------------|-----|---|
| TX FOR LIVER CA | 109 | (4%) |
| SECONDARY LIVER CA | 244 | (10%) |
| TOTAL | 353 | (14%) |
| POST Tx | | |
| RECURRENT LIVER CA | 64 | (2.7% of all pts, 19% of pts with Ca at Tx) |
| DE NOVO CA | 118 | (5%) 122 Ca |
| SKIN CA | 253 | (10%) 1620 Ca |
| TOTAL | 438 | (18%) |
| MULTIPLE CA | 67 | |
| TRANSFERRED FROM DONOR | 2 | |
| DEVELOPED NON SKIN CA < 90DAYS | 7 | |

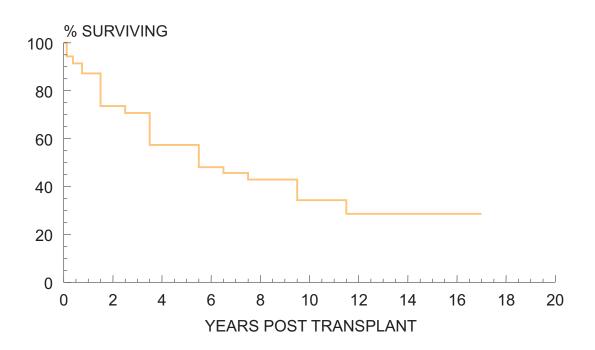
Primary Liver Cancer N = 2497

| | No | DIED | DIED OF THIS CANCER |
|-------------------------------------|----------|-----------------------------|-----------------------------|
| Hepatocellular Ca | 91 | 32 | 18 (20%) |
| Lamellar Variant | 5 | 4 | 2 (40%) |
| Carcinoid | 4 | 4 | 4 (100%) |
| Hepatoblastoma | 4 | 2 | 1 |
| Endocrine | 2 | 2 | 2 |
| Angiosarcoma | 1 | 1 | 1 |
| Cholangiocarcinoma | 1 | 0 | 0 |
| Epitheloid Haemangioendothelioma | 1 | 0 | 0 |
| TOTALS | 109 (4%) | 45 (41% of pts with PCa) | 28 (26% of pts with PCa) |

N = 2497

n = 109 (4%) with cancer

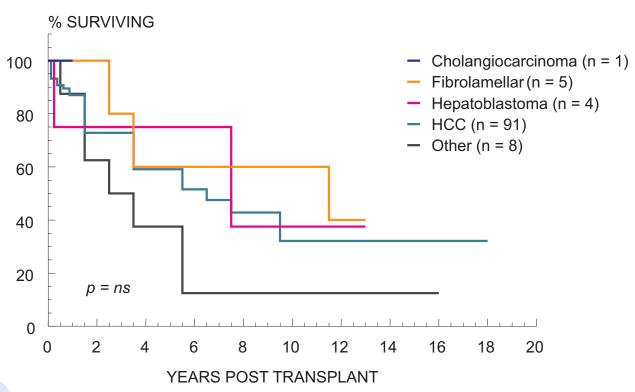




Primary Liver Cancer All Patients

N = 2497

n = 109 (4%) with cancer





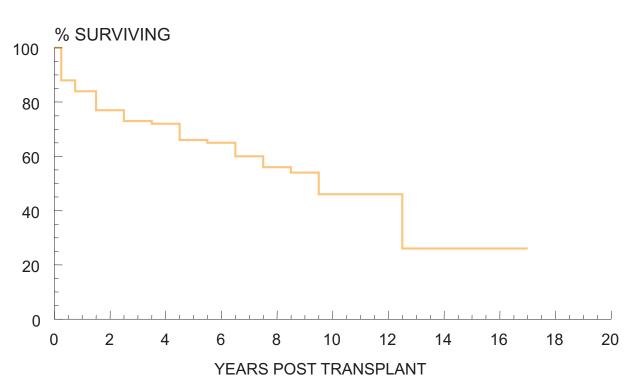
| | | | DIED | DIED OF THIS CANCER |
|------------------------|--------------------|--------------------------|---------------------------|-------------------------|
| HEPATOCELLULAR C | HEPATOCELLULAR CA* | | 58 | 20 (9%) |
| CHOLANGIO CA | | 24 | 16 | 13 (54%) |
| ADENOCARCINOMA | | 3 | 2 | 0 |
| HEPATOBLASTOMA* | HEPATOBLASTOMA* | | 1 | 0 |
| FIBROLAMELLAR | FIBROLAMELLAR | | 1 | 0 |
| ANGIOSARCOMA | | 1 | 1 | 1 |
| EPITHELOID HAEMANGIOCA | | 1 | 0 | 0 |
| | TOTALS | 245* in 244 pts (10%) | 79% (32% of pts with Sca) | 34 (14% of pts with Sca |

* 1 patient had 2 secondary malignancies

Secondary Liver Cancer

N = 2497

n = 244 pts (10%)

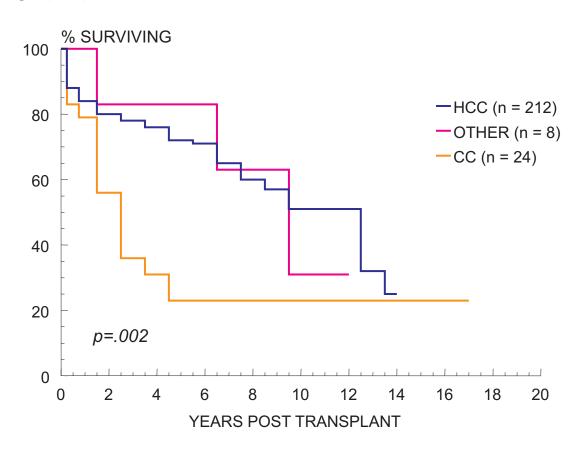


Secondary Liver Cancer All Patients



n = 244 pts (10%)

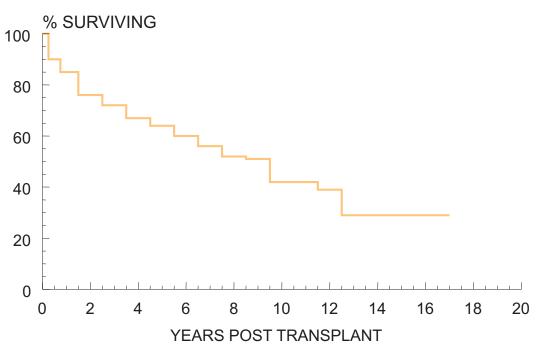




Primary and Secondary Liver Cancer

$$N = 2497$$

 $n = 353 (14\%)$

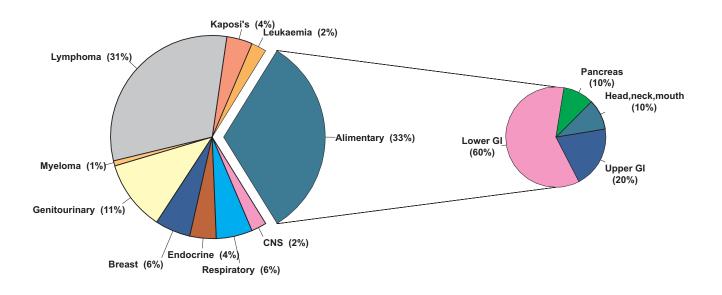




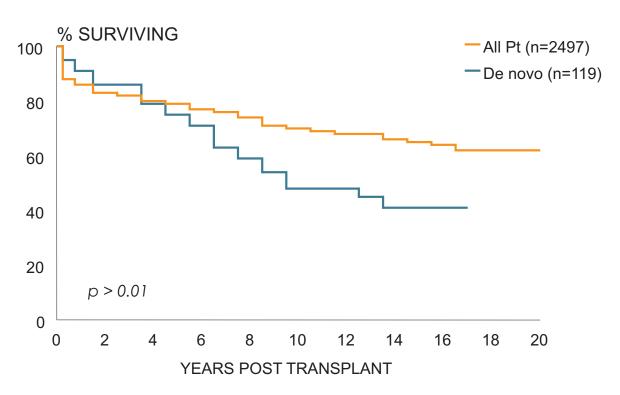
| | No | Male | Female | Age of pts (yrs) | Time to diagnosis (mths) | Died of This Cancer |
|------------------|-----------------------|------|--------|------------------|--------------------------|-------------------------|
| Alimentary* | 40 | 26 | 14 | 12.6 - 74.8 | 3 - 173 | 19 (48%) |
| Lymphoma* | 38 | 21 | 17 | 1.5 - 65.2 | 1 - 182 | 17 (48%) |
| Leukaemia | 3 | 1 | 2 | 2.9 - 49.5 | 16 - 44 | 0 |
| Kaposi's | 5 | 4 | 1 | 32.1 - 64.6 | 2 - 48 | 0 |
| Genitourinary* | 14 | 6 | 8 | 38.5 - 70.5 | 2 - 164 | 2 |
| Breast | 7 | - | 7 | 39.1 - 62.8 | 11 - 189 | 2 |
| Endocrine | 5 | 2 | 3 | 35.8 - 69.5 | 47 - 144 | 2 |
| Respiratory | 7 | 4 | 3 | 44.7 - 61.1 | 37 - 111 | 5 |
| CNS | 3 | 1 | 2 | 16.5 - 75 | 66 - 174 | 2 |
| Multiple Myeloma | 1 | - | 1 | 67 - 67 | 6 - 6 | 0 |
| Total | *123 ca in 119 pts | 65 | 58 | 1.5 - 75 | 1 - 189 | 49 (41% of pts with Ca) |

Eight patients also had secondary liver malignancy; * 3 patients had 2 de novo malignancies

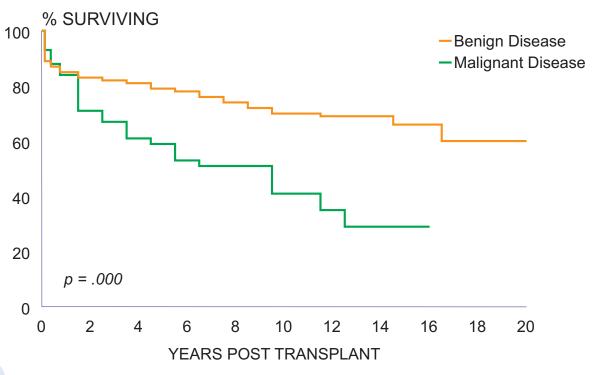
De Novo Non Skin Cancer





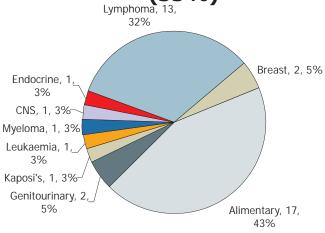


Patient Survival Benign Disease vs Primary or Secondary Liver Malignacy N = 2497

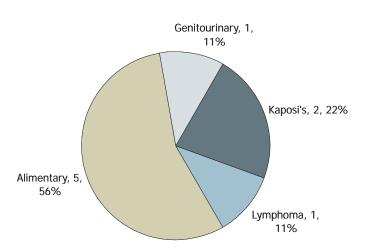




PSC + Auto-immune - 39
(33%)
Lymphoma, 13,



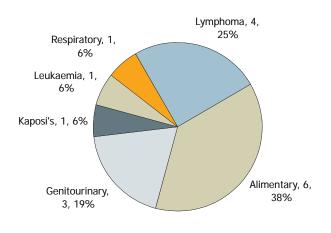
HBV - 9 (8%)

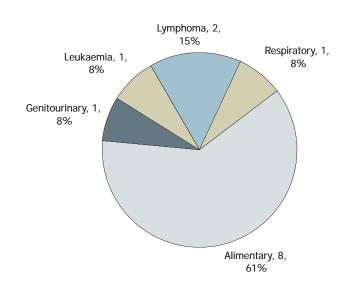


Pre Transplant Liver Disease and De Novo Non Skin Cancer

HCV - 16 (13%)

Alcohol - 13 (11%)



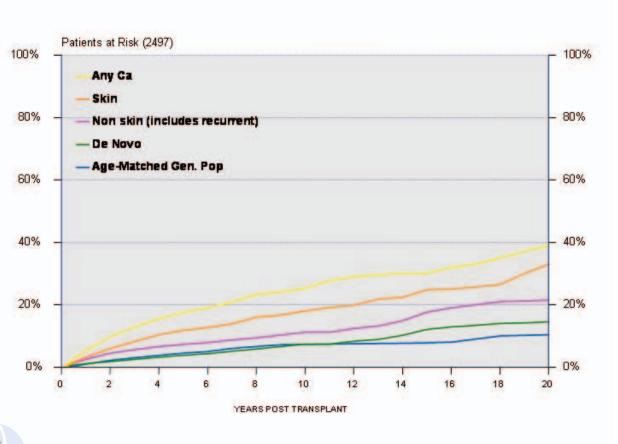




| TYPE OF SKIN CANCER | PATIENTS | CANCERS |
|------------------------|----------|---------|
| BCC | 163 | 458 |
| SCC | 171 | 612 |
| MELANOMA | 12 | 12 |
| TOTALS | 285** | 1619 |

** 148 pts had multiple skin cancer types

Cumulative Risk of Diagnosis of Cancer Following Liver Tx. 1985 - December 2005 N = 2497





Appendix I

Liver Transplant Units of Australia and New Zealand

and

and

and

Australian National Liver Transplant Unit

Royal Prince Alfred Hospital

Missenden Road

CAMPERDOWN NSW 2050

Email: anltu@cs.nsw.gov.au

http://www.cs.nsw.gov.au/Gastro/LiverTransplant/default.htm

The New Children's Hospital

Hawkesbury Road

WESTMEAD NSW 2145

Liver Transplant Unit

The Austin

Studley Road

HEIDELBERG VIC 3084

Royal Children's Hospital

Flemington Road

PARKVILLE VIC 3052

Queensland Liver Transplant Service

Princess Alexandra Hospital

Ipswich Road

WOOLLOONGABBA QLD 4102

Royal Children's Hospital

Bowen Bridge Road

HERSTON QLD 4029

South Australian Liver Transplant Unit

Flinders Medical Centre

Flinders Drive

BEDFORD PARK SA 5042

http://www.flinders.sa.gov.au/flinders_centre_for_digestive_health/

WA Liver Transplantation Service

Sir Charles Gardiner Hospital

Verdun Street

NEDLANDS WA 6009

New Zealand Liver Transplant Unit

Auckland Public Hospital

Park Road

Auckland

New Zealand

Http://www.nzliver.org/



Appendix II

ANZLTR PRIMARY Diagnosis Metabolic disorders by Age Group

| D.i | Age | Age group | | |
|---------------------------------|-------|-----------|-----|--|
| Primary Diagnosis | Child | Adult | | |
| -1 Antitrypsin deficiency | 28 | 36 | 64 | |
| Crigler-Najjar | 4 | 1 | 5 | |
| Familial amyloid polyneuropathy | 0 | 26 | 26 | |
| Glycogen storage disease | 0 | 1 | 1 | |
| Haemochromatosis | 1 | 23 | 24 | |
| Homozygous Hypercholesterolemia | 3 | 1 | 4 | |
| Indian childhood cirrhosis | 1 | 0 | 1 | |
| Other* | 7 | 1 | 8 | |
| Primary hyperoxaluria | 5 | 6 | 11 | |
| Tyrosinemia | 4 | 0 | 4 | |
| Urea cycle disorders** | 7 | 3 | 10 | |
| Wilsons disease | 7 | 24 | 31 | |
| Total | 67 | 122 | 189 | |

^{*} Bile acid synthesis disorder, Protein C deficiency, methylmalonic acidemia, familial immunodeficiency, mitochondrial disease

^{**} OTC deficiency 6; citrullinemia 4



Appendix III

ANZLTR PRIMARY Diagnosis - Other by Age Group

| | Age | group | Total |
|---|-------|-------|-------|
| Primary Diagnosis | Child | Adult | |
| Alagille syndrome | 21 | 1 | 22 |
| Alagille non-syndromic | 2 | 0 | 2 |
| Benign liver tumour -Adenomatosis | 0 | 1 | 1 |
| Benign liver tumour-Haemangioma | 0 | 2 | 2 |
| Caroli's disease | 1 | 10 | 11 |
| Choledocal cyst | 1 | 2 | 3 |
| Cholestatic disease-Other | 1 | 2 | 3 |
| Chronic Budd Chiari | 1 | 24 | 25 |
| Congenital biliary fibrosis | 1 | 1 | 2 |
| Ductopenia | 0 | 2 | 2 |
| Granulomatous hepatitis / sarcoidosis | 0 | 4 | 4 |
| Histiocytosis X | 4 | 0 | 4 |
| Liver Trauma | 0 | 1 | 1 |
| Neonatal hepatitis | 4 | 0 | 4 |
| Nodular regenerative hyperplasia | 0 | 4 | 4 |
| Non alcoholic fatty liver (NAFLD or NASH) | 0 | 18 | 18 |
| Polycystic Liver disease | 0 | 11 | 11 |
| Polycystic liver and kidney disease | 0 | 4 | 4 |
| Progressive familial intrahepatic cholestasis(PFIC) | 11 | 4 | 15 |
| Secondary biliary cirrhosis | 1 | 9 | 10 |
| Secondary biliary cirrhosis - Hepatolithiasis | 0 | 4 | 4 |
| Secondary biliary cirrhosis - Cystic fibrosis | 7 | 10 | 17 |
| Other -specify | 3 | 14 | 17 |
| Total | 58 | 128 | 186 |

[#] Vanishing bile duct syndrome
Haemangiotelangiectasia
Veno-occlusive disease
Chronic Active Hepatitis A
Non-cirrhotic portal hypertension
Kassabach-Merritt syndrome
Arterial-venous malformation
Hereditary haemorrhagic telengectasia / OWRD.





Appendix IV

ANZLTR PRIMARY Diagnosis Fulminant Hepatic Failure by Age Group

| Primary Diagnosis | Age gı | oup | Total |
|----------------------------------|----------|-------|-------|
| , g | Children | Adult | |
| Acute - Budd Chiari | 0 | 2 | 2 |
| Acute - Wilson's | 4 | 10 | 14 |
| Acute1 -AAT | 2 | 0 | 2 |
| Acute Autoimmune hepatitis | 0 | 5 | 5 |
| Acute Unknown / unspecified | 32 | 57 | 89 |
| Acute -Paracetamol | 0 | 6 | 6 |
| Acute -Other drugs | 2 | 11 | 13 |
| Acute Herbs / mushrooms | 0 | 3 | 3 |
| Acute - Hepatitis A | 0 | 2 | 2 |
| Acute - Hepatitis B | 0 | 28 | 28 |
| Acute - NonA-NonB | 4 | 8 | 12 |
| Acute - Hepatitis E | 0 | 1 | 1 |
| Acute - Post liver resection | 1 | 1 | 2 |
| Subacute - Wilson's | 1 | 2 | 3 |
| Subacute Autoimmune hepatitis | 0 | 5 | 5 |
| Subacute - Dug | 0 | 2 | 2 |
| Subacute - Unknown / unspecified | 2 | 25 | 27 |
| Subacute - Hepatitis A | 0 | 1 | 1 |
| Subacute - Hepatitis B | 0 | 7 | 7 |
| Total | 49 | 175 | 224 |



Appendix V

ANZLTR Causes of Patient death

| Graft failure - other | | |
|-----------------------------------|-------------|----|
| Vascular thrombosis | | 18 |
| Hepatic artery | 10 | |
| Portal vein | 7 | |
| Hepatic vein | 1 | |
| Non thrombotic infarction | | 2 |
| Primary non function | | 18 |
| Massive haemorrhagic necro | osis | 4 |
| Recurrent disease | | 3 |
| (ALD, PSC, CAH:AI) | | |
| De novo Hep C | | 2 |
| Biliary Complications | | 8 |
| Other | | 8 |
| (PNC, immune hepatitis, outflow o | bstruction) | |

| <u>Miscellaneous</u> | |
|--|----|
| | |
| Multiorgan failure | 19 |
| Renal Failure | 10 |
| Graft vs Host disease | 5 |
| Social | 10 |
| (accident, suicide,non-compliance, Rx withdrawn) | |
| Sudden death (cause unknown) | 6 |
| Other | 6 |
| (Hyperkalaemia,motor neurone disease | |
| diabetes complications, drug reaction) | |