

# AUSTRALIA & NEW ZEALAND

LIVER TRANSPLANT REGISTRY



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

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Brisbane, QLD, AUSTRALIA Editors: S.V. Lynch, G.A. Balderson

#### STATISTICAL METHODS

Kaplan-Meier survival curves have been produced using SPSS® for Windows™ Release 15.0, SPSS Inc.

#### **ACKNOWLEDGMENT**

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

Director: Professor G.W McCaughan

All queries to: Dr Deborah Verran Email deborah@email.cs.nsw.gov.au

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DATA TO 31/12/2006

#### Preface

We are pleased to present the 18<sup>th</sup> Report of the Australia and New Zealand Liver Transplant Registry (ANZLTR). This report contains data to the 31<sup>st</sup> December 2006 and analyses the cumulative data since the establishment of the first liver transplantation unit in Australia or New Zealand in 1985.

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 front page.

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, prepares 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 front page 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 2006, 2886 orthotopic liver transplants (OLT) were performed in Australia and New Zealand on 2677 patients, 2188 adult patients (> 15 years) [82%] and 489 children [18%]. The median age of all recipients was 46 years. The ages ranged from 24 days to 73.1 years. There is a significant difference in gender distribution between children (M=47%) and adults (M=63%)
- 6. There was a decrease in the total number of new paediatric patients transplanted in 2006 compared with 2005 but an increase in the number of new adult patients.
- 7. The trend to increasing age of adult recipients in recent years continued and the overall adult median age is now 48.9 years. The median age of new adult recipients in 2005-06 was 51.6 years.
- 8-9. Only three more transplants were performed in 2006 then in 2005. Split grafts now make a significant contribution to the total number of paediatric transplants performed providing 12 of 27 [44%] grafts in 2006 and 96 of 560 [17%] overall. In children, other reduced size grafts have been used in 276 [49%] cases including 17 living donor grafts. One child has been treated with liver cell implantation. Of adult patients, 136 have received reduced size grafts 108 split liver grafts (including 1 as auxiliary graft), 24 other reduced size grafts (1 as auxiliary graft) and 4 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). The number of patients transplanted for non alcoholic fatty liver disease [ NAFLD/NASH] continued to increase with 9 new patients in 2006 (Appendix III).
- 12-14. Fewer adult patients with a primary diagnosis of Hepatitis B but more with a diagnosis of Hepatitis C were transplanted in 2005-06 compared with the previous era.- 2000-04, 35% primary diagnosis CVH [25% Hepatitis C, 8% Hepatitis B and 2% Hepatitis B,C,D]; in 2005-06, 33% primary diagnosis CVH [27% Hep C, 4% Hep B, 2% Hep B/C/D]. 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 2005-06 was 42%.
- 15. Current 1 year patient survival of all patients is 87% at 1 year, 79% at 5 years and 70% 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 longer term 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 or hepatitis virus coinfections 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.
- 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 sub-acute] with 5 year survival of 74%.

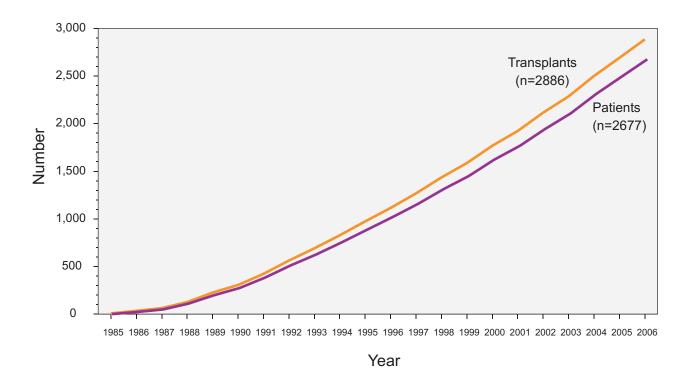
### Summary

- 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.
- Twenty two patients have received a living donor graft, 15 children and 5 adults as a primary graft and two children as a second or third graft. Patient survival was 90% and graft survival 85%.
- 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. Thirteen percent of retransplants were due to poor early graft function. Recurrent disease was the indication for retransplantation in 9% of cases [3% PSC, PBC and 6% 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-two 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 slight rise in the in number of cadaveric donors in 2006 resulting an increase in transplants from 2005. Twelve 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.
- The numbers of patients waiting for transplant remains high. At the end of 2006, 132 patients were awaiting a transplant compared with 145 at 31st December 2005. Delistings due to death, becoming too ill or tumour (HCC) progression were 10%. Forty-two patients were listed as urgent in 2006 [16 Category 1 and 26 Category 2]. In 2006 the majority of urgently listed patients received a timely transplant.
- Waiting times continue to increase with some patients waiting years to receive a graft. Blood group O patients tend to have the longest waiting times.
- Five hundred and fifteen patients (18%) have had a pre- or post-transplant cancer.

  One hundred and twenty eight (4%) of patients were transplanted for liver malignancy (Primary Malignancy) and thirty (23% of these patients) died from this cancer.
- 38-40 Two hundred and sixty two patients had a liver cancer as a secondary diagnosis (Secondary Malignancy) and thirty six [14% of these patients] have died from this cancer. Hepatocellular carcinoma was the most common secondary malignancy (230), however those with cholangiocarcinoma (24) had significantly poorer survival.
- 41-44 De novo non skin cancers (136) have developed in 130 (5%) patients and 52 [40%] have died from this cancer. Cancers of the alimentary tract and lymphoma predominate. Lower GI cancers account for 60% of alimentary tract cancers. Patients with either de novo non skin cancers or liver cancers have significantly worse long term survival. Alimentary tract cancers are the most common irrespective of the pre transplant liver disease.
- Three hundred and ten (11%) patients have developed 1927 skin cancers with 182 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

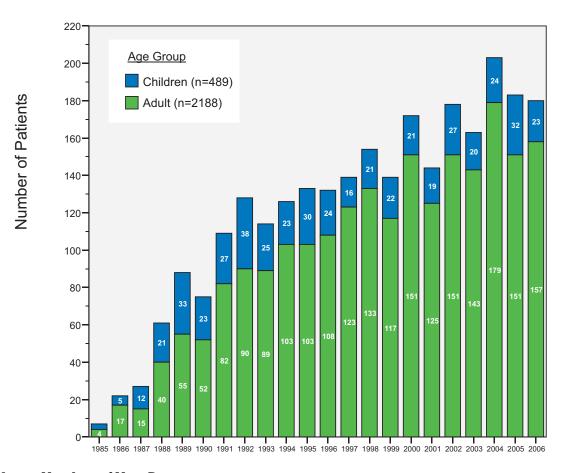


#### **Summary Statistics - Age and Gender**

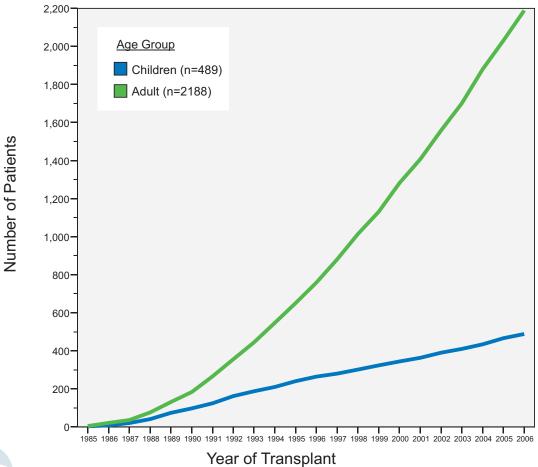
#### **ALL PATIENTS**

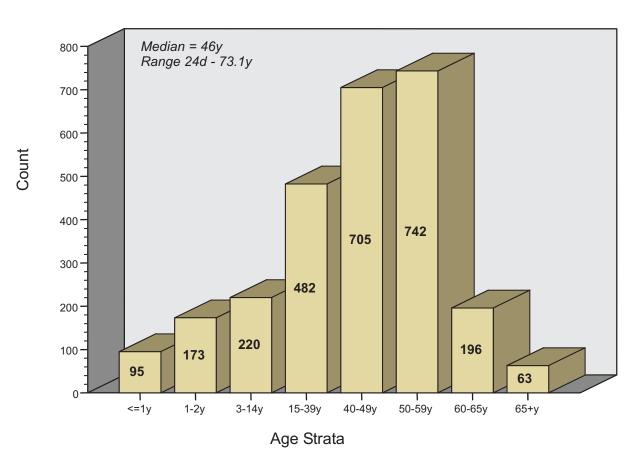
	Children	Adults	Total			
Patients	489	2188	2677			
Age						
Mean ± SD	4.4 ± 4.2	47.2 ± 11.8	39.4 ± 19.8			
Median	2.5y	48.9y	46y 24d - 73.1y			
Range	24d -14.9y	15.0 - 73.1y				
Gender	Gender					
Female	259 (53%)	814 (37%)	1073 (40%)			
Male	230 (47%)	1375 (63%)	1604 (60%)			
Surviving	385 (79%)	1610 (74%)	1995 (75%)			



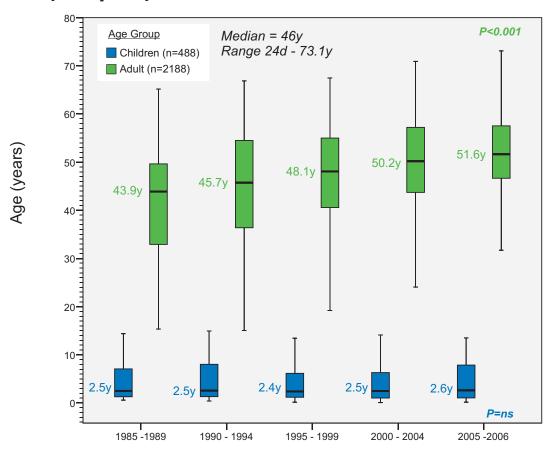


#### **Cumulative Number of New Patients**



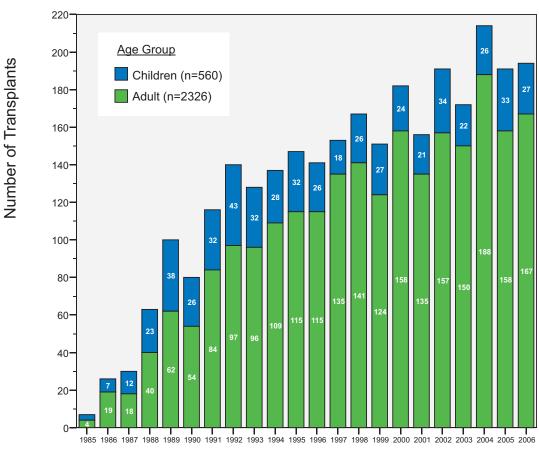


#### Age at Primary Transplant by Era

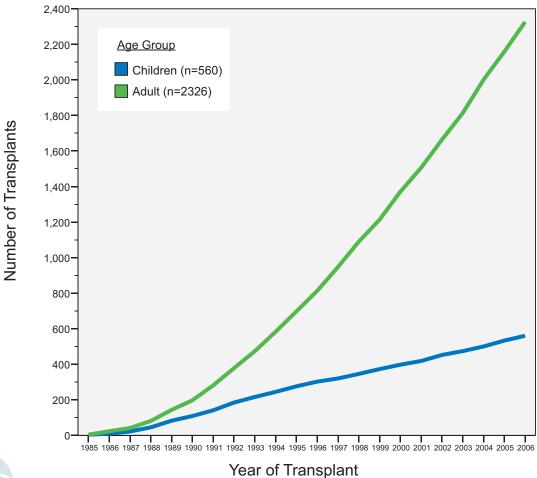




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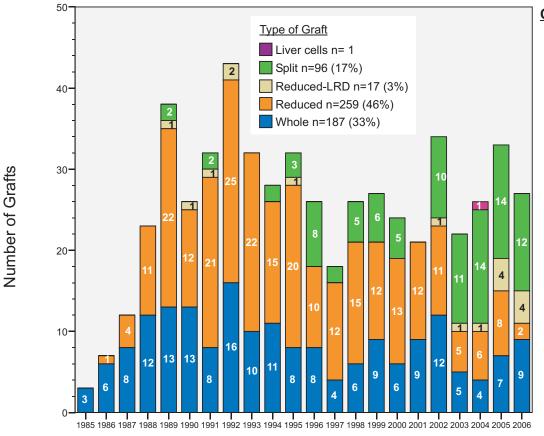


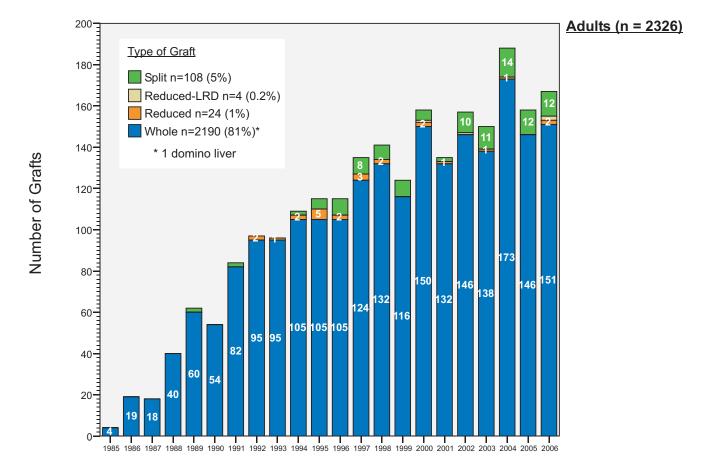
#### **Cumulative Number of Transplants**





Children (n = 560)

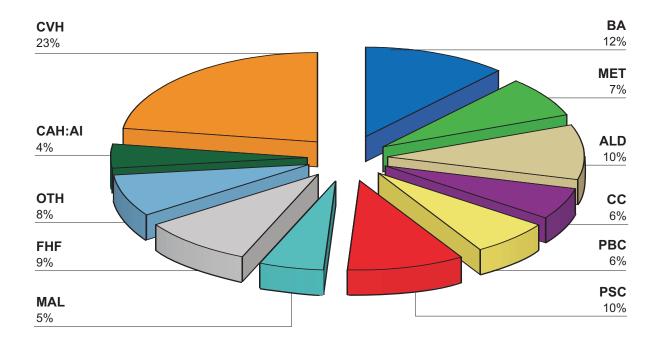




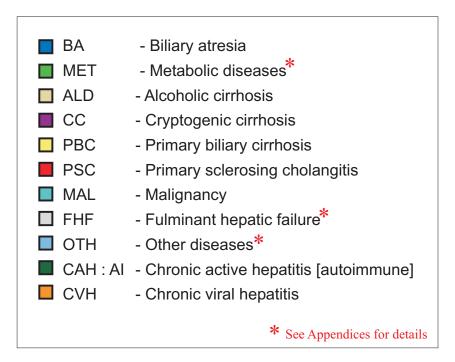
## Section 2

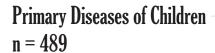
**Primary Diagnosis** 





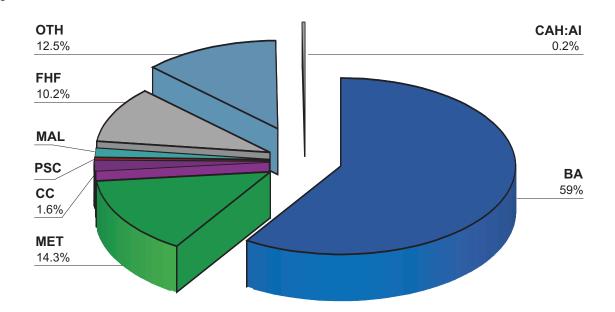
#### **Diagnosis Group**

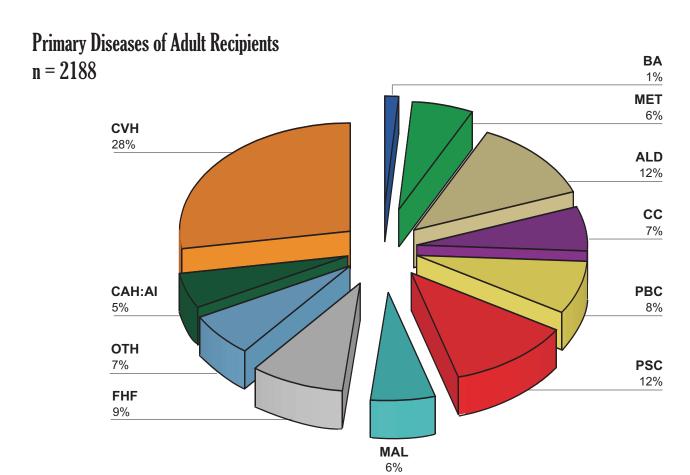


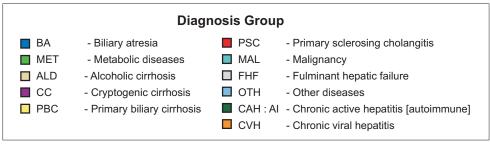


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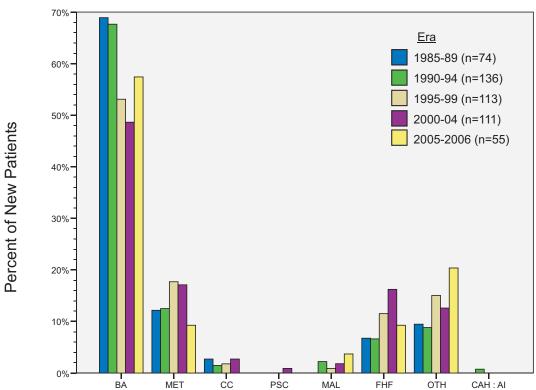




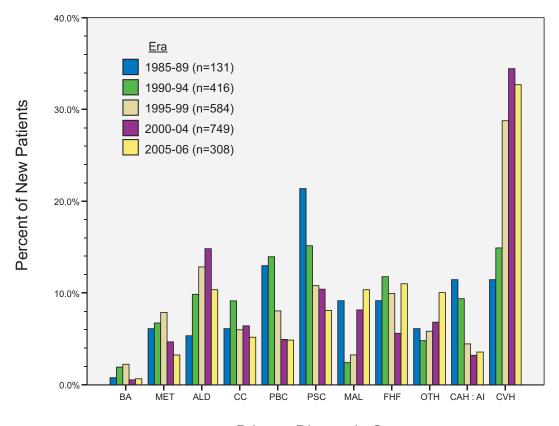


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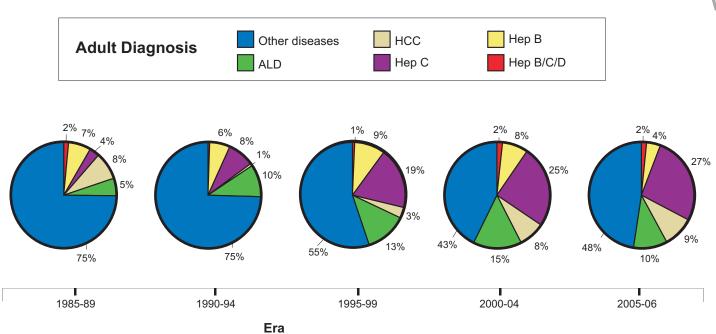
#### Children (n=489)



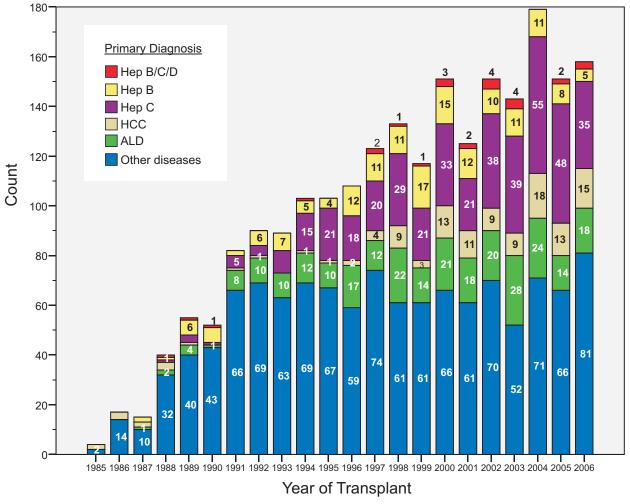
#### <u>Adults (n = 2188)</u>



Primary Diagnosis Group



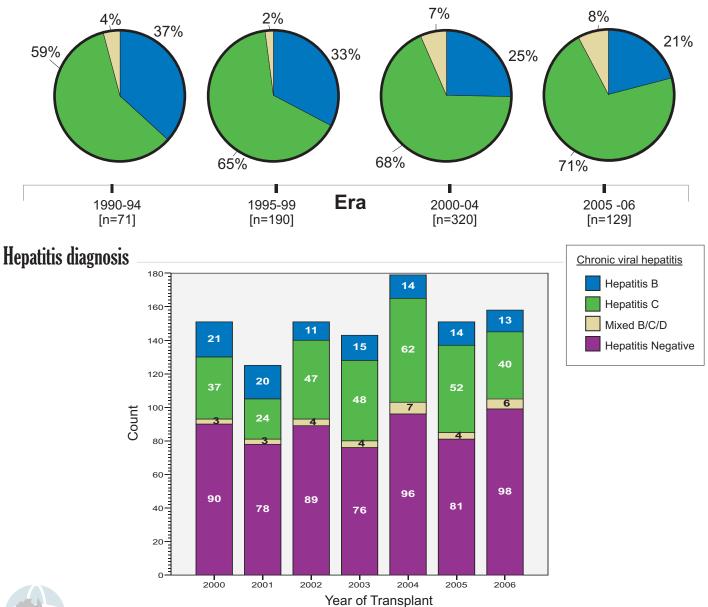
#### Adult Primary Diagnosis by Year



#### Chronic Viral Hepatitis as Primary or Secondary Diagnosis in Adult Patients

			Secondary / Tertiary diagnosis					
Primary Diagnosis		n=	Hepatitis C	Hepatitis B	Hepatitis B,C	НСС	ALD	
	Hepatitis C	416		5		82	96	
	Hepatitis B	162	4			44	4	
	Hepatitis	26				2	5	
	BD/BC/BCD							
	HCC + cirrhosis	121	49	47	4		11	
	ALD	266	10	2		25		
	Other	1197	11	3		35	19	
	TOTAL	2188						

#### Type of Chronic Viral Hepatitis in Adult Patients

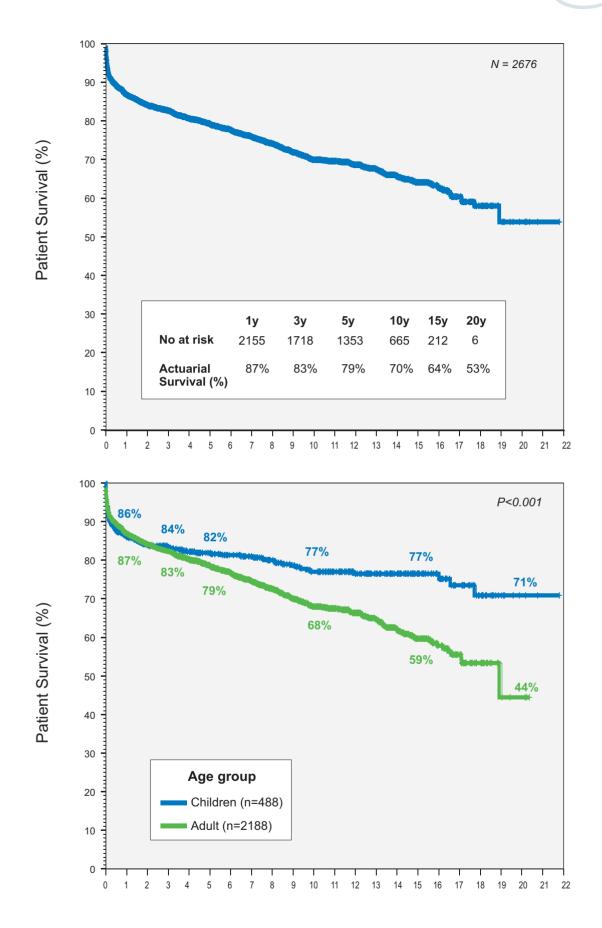


**SECTION 3: PATIENT SURVIVAL** 

## Section 3

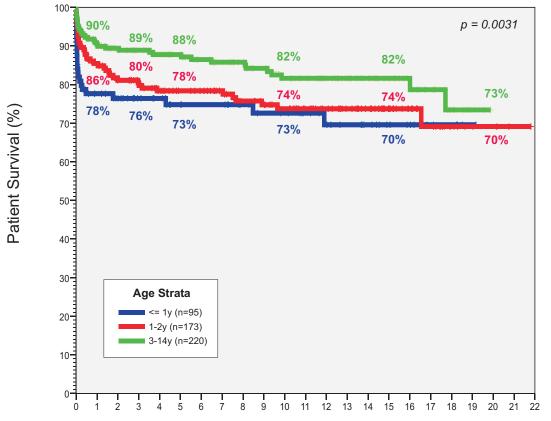
Patient Survival



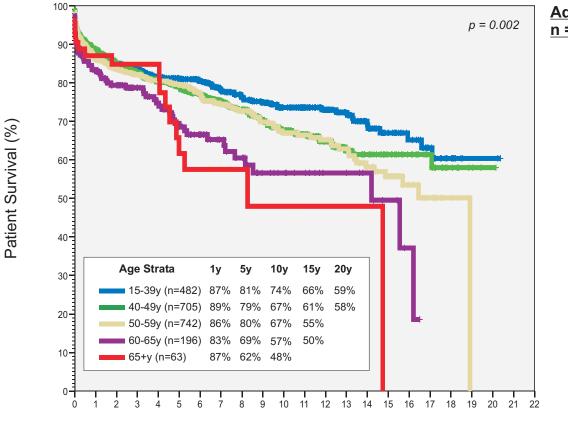


Time Post-transplant (years)



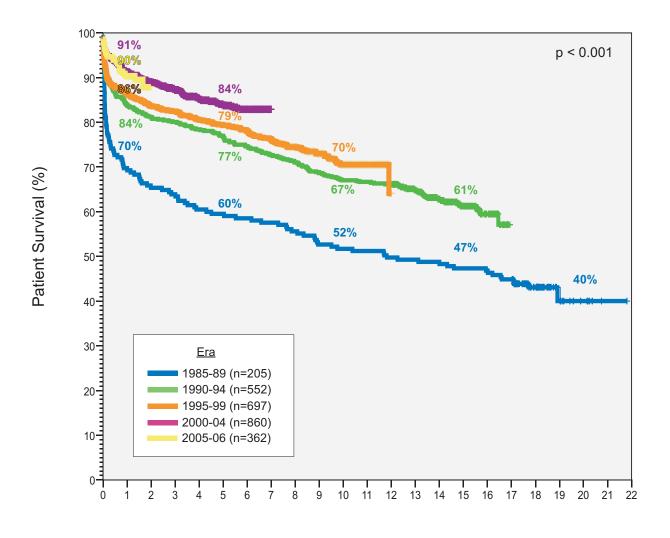


Children n= 488



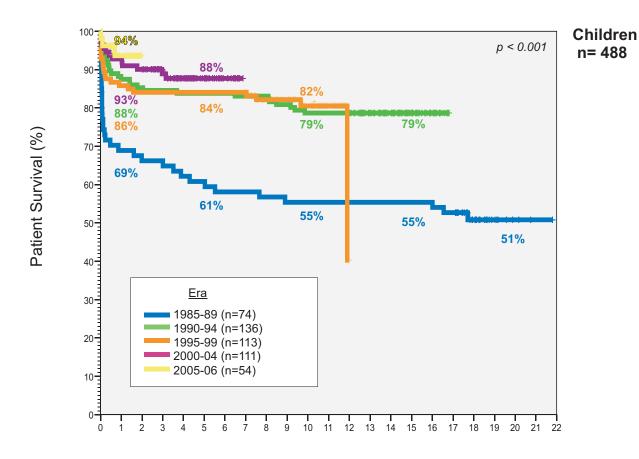
<u>Adults</u> n = 2188

Time Post-transplant (years)

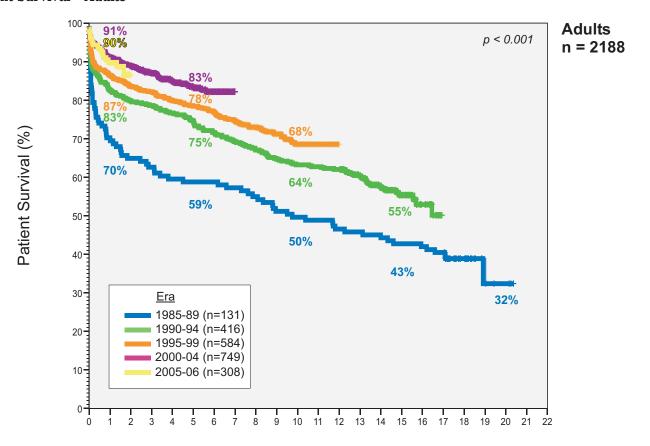


Time Post-transplant (years)





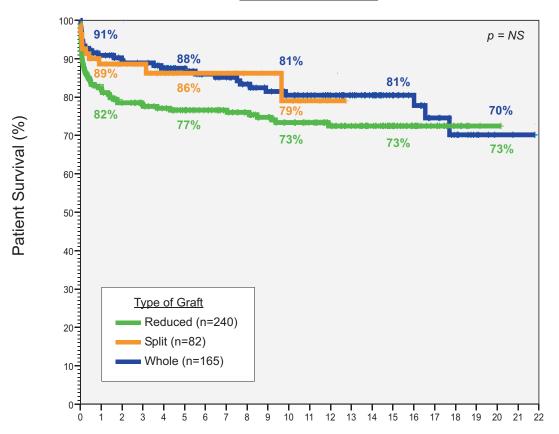
#### **Patient Survival - Adults**



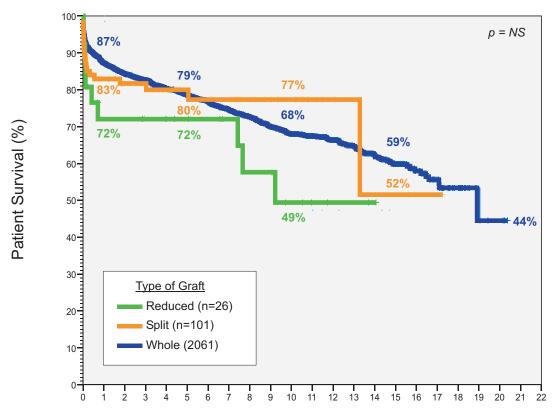
Time Post-transplant (years)



#### Children - n = 488

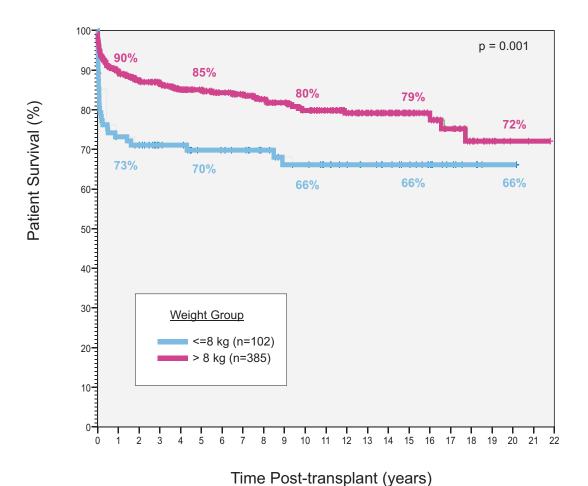


#### Adults - n = 2188



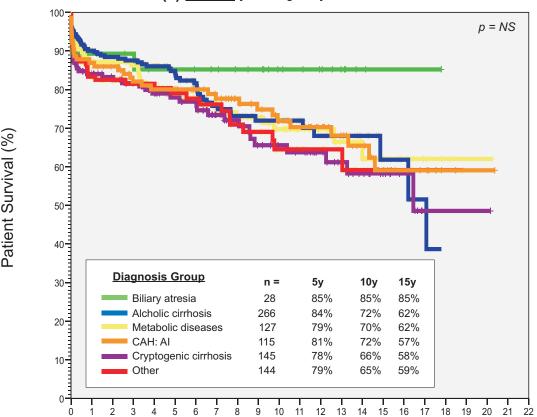
Time Post-transplant (years)



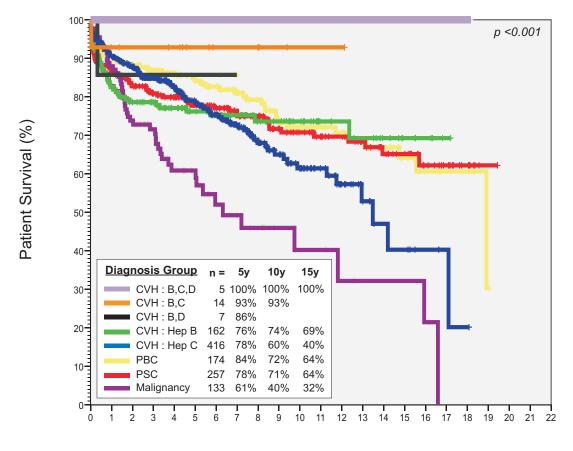


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#### (1) Adults [excluding FHF] - n=825



#### (2) Adults [excluding FHF] - n=1168

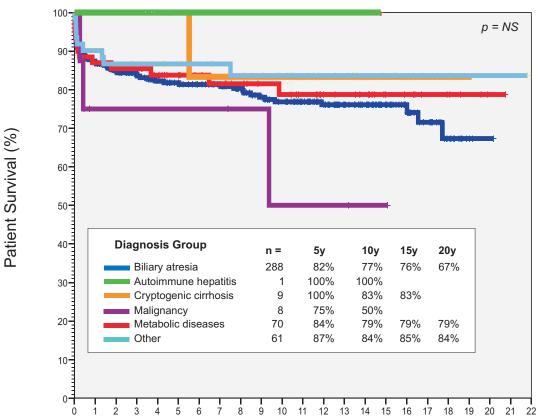


Time Post-transplant (years)

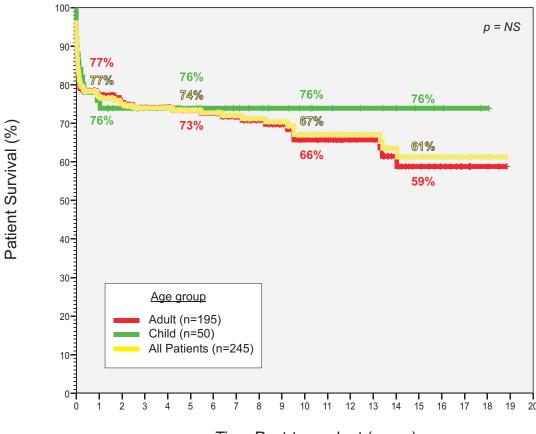
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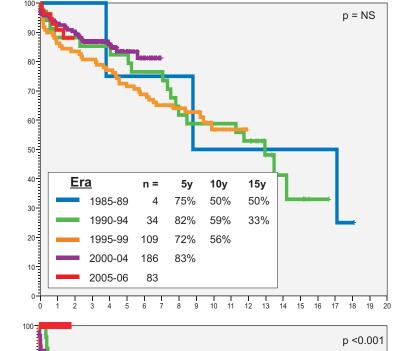


#### (4) Fulminant hepatic failure (n=245)

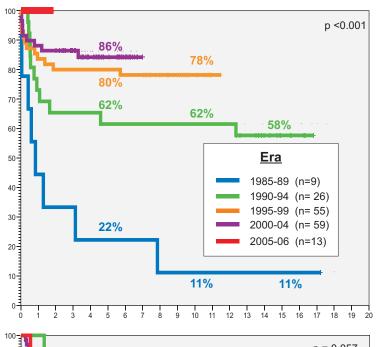


Time Post-transplant (years)

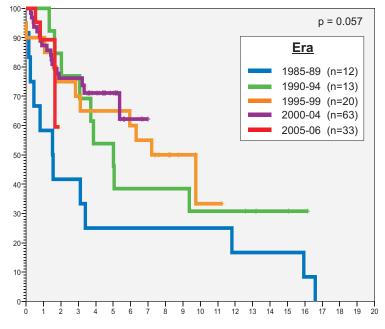




Adults CVH: Hepatitis C n = 416



**Adults CVH: Hepatitis B** n = 162



Malignancy Adults and Children n = 141

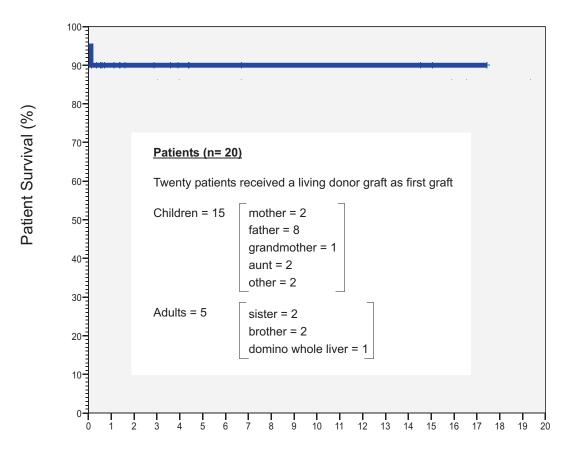


DATA TO 31/12/2006

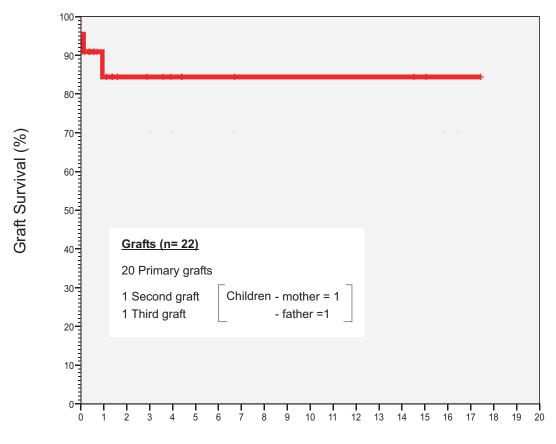
Patient Survival (%)

Patient Survival (%)





Time Post-transplant (years)



Time Post-transplant (years)

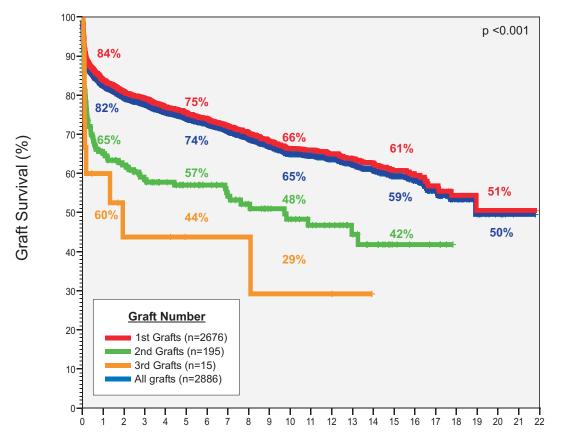


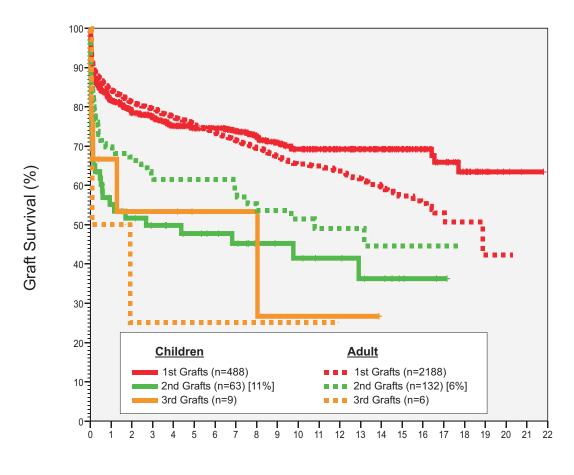
## Section 4

Graft Outcome

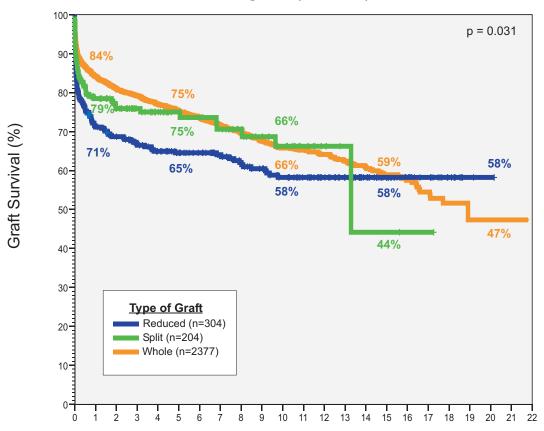


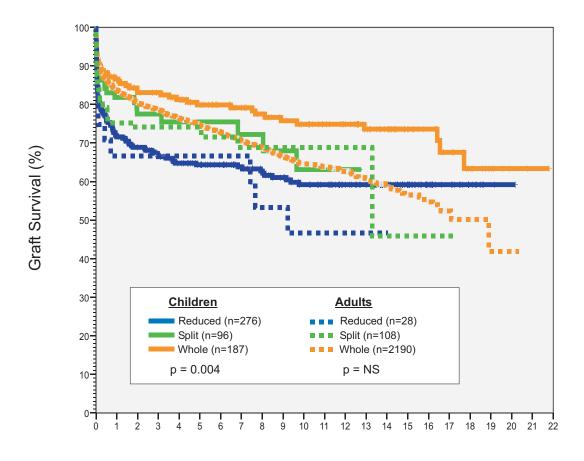






#### **All grafts (n = 2886)**

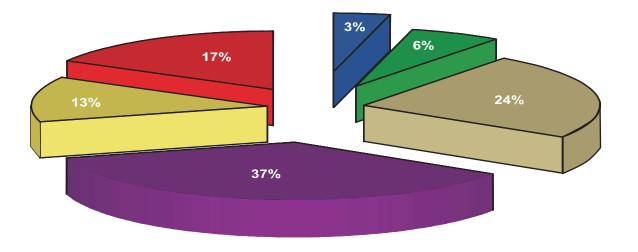


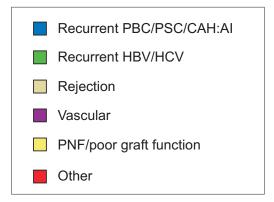


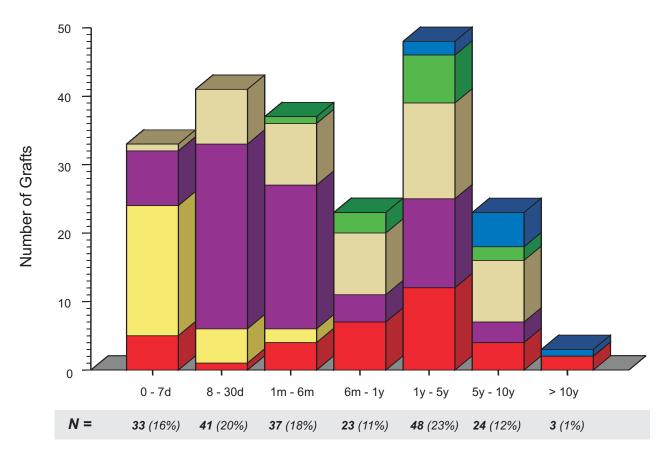
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**n** = **209** (194 2nd grafts, 15 3rd grafts)





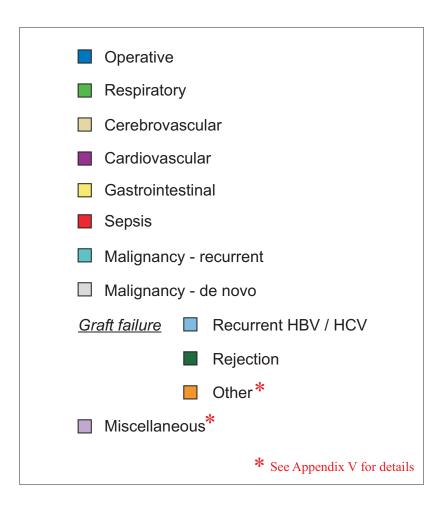


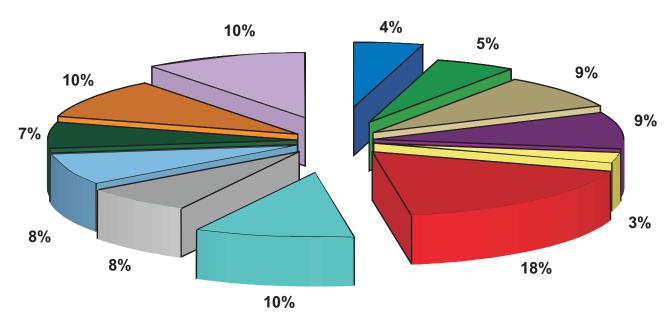
## Section 5

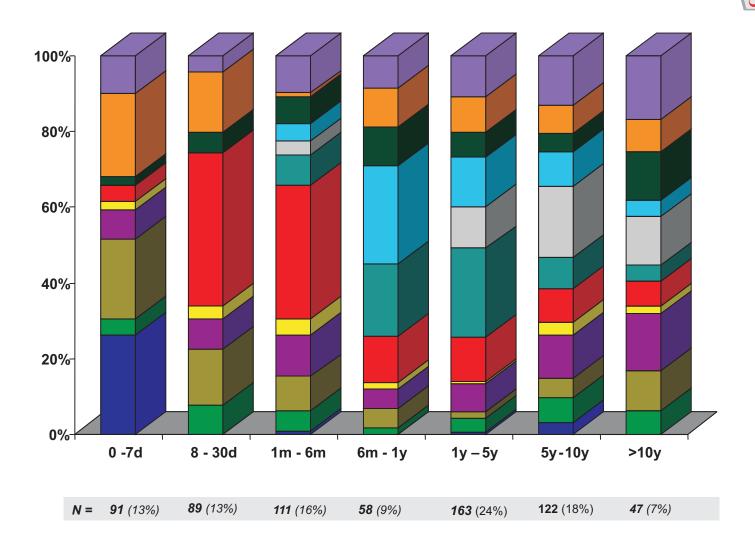
**Cause of Patient Death** 



#### All Patients n = 681









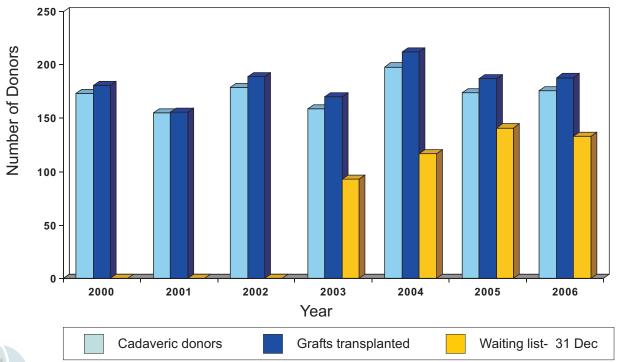
# Section 6

**Donor Information** 

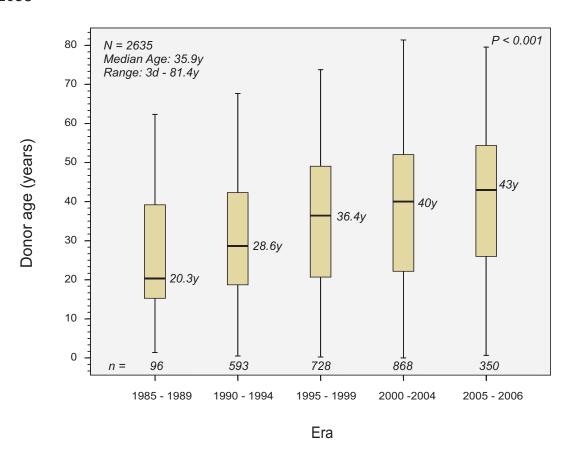


	QLD	NSW/ACT	VIC/TAS	SA/NT	WA	NZ	TOTAL
1990	22	27	16	5		7	77
1991	29	35	20	6	8	11	109
1992	43	32	18	9	8	24	134
1993	28	40	25	12	6	16	127
1994	29	39	23	12	10	21	134
1995	29	44	24	17	8	21	143
1996	26	37	19	17	10	24	133
1997	31	49	19	19	8	22	148
1998	29	44	27	22	13	27	162
1999	15	31	31	29	11	27	144
2000	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
2006	28	34/3	39/6	25	17	24	176

#### **Grafts from cadaver donors**

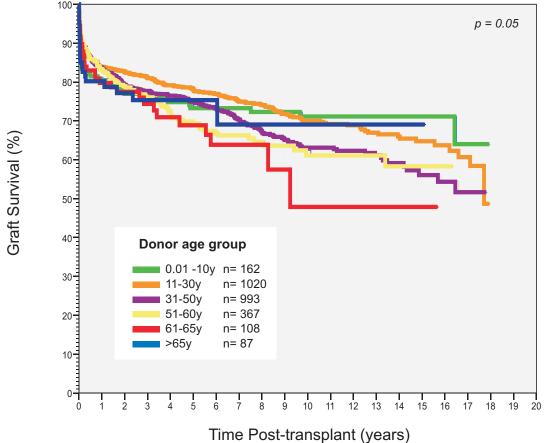






#### Graft Survival by Donor Age

N = 2737



# Section 7

Waiting List





Activity	2004	2005		2006	
Listed at 1 January New listings	93 279	117 291	145 -	- 258	TOTAL 2006
TOTAL	372	408	145	258	403
		OUTCOM	E		
Transplant	214 [58%]	191 [47%]	80	114	194 [48%]
Delisted	41 [10%]	72 [18%]	37	40	77 [19%]
Died on list	14)	26	5	13	18 ງ
Too sick	8 \ 6%	9 } 11%	9	4	13 \[ [10%]
Tumour progression	2	9 )	4	4	8)
Improved	8	15	8	8	16
Other	9	13	11	11	22*
Still listed at 31 Dec	117 [32%]	145 [35%]	28	104	132 [33%]

<sup>[\*</sup>Other: Further investigations 11; New medical complication - 7; Declined Tx - 2, Other treatment - 1, Ongoing alcohol - 1]

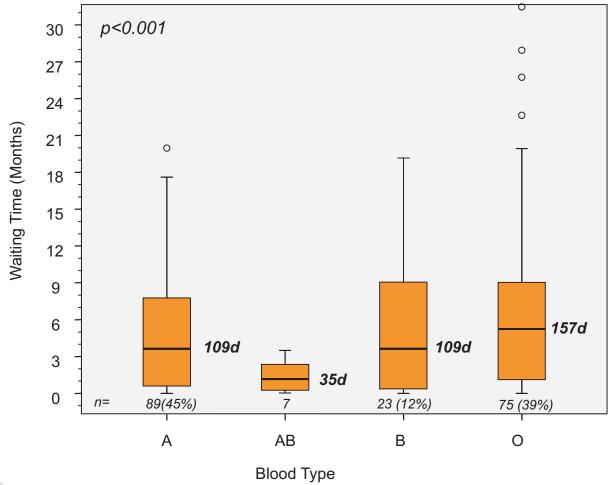
#### **Outcome of Urgent Listing**

	CAT	EGORY 1	CATEGORY 2		
OUTCOME	<b>2005</b> (n=14)	<b>2006</b> (n=16)	<b>2005</b> (n=31)	<b>2006</b> (n=26)	
TRANSPLANTED	4 64%	12) 88%	20 68%	21) 88%	
IMPROVED	<sub>5</sub>	2	1 5	2	
DIED	5	2	10	2	
OTHER TREATMENT	-	-	-	1	

		Blood Group           A         O         B         AB         TOTAL					
	Α						
n=	151 (37.5%)	186 (46%)	56 (14%)	10 (2.5%)	403		
Not transplanted	62	111	33	3	209		
Transplanted	89 (59%) <sup>**</sup>	75 (40%)	23 (41%)	7 (70%)	194		

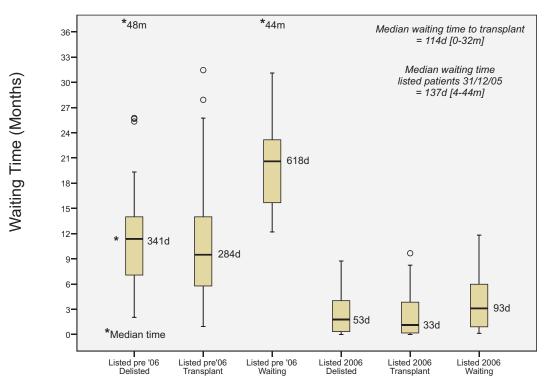
<sup>%</sup> of total number listed

#### Waiting Time to Transplant 2006



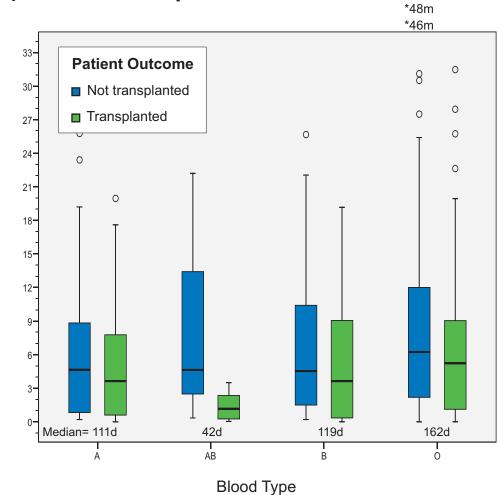
<sup>\*\* %</sup> of blood group





**Patient Outcome** 

#### Waiting Time by Outcome & Blood Group



Waiting Time (Months)

# Section 8

Liver Transplantation and Cancer

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## Cancer in Liver Transplant Recipients N = 2677



At Tx		
Tx for Liver Ca	128	(5%)
Liver Ca as a Secondary Diagnosis	262	(10%) 263 Ca
Total	387	(14%)
Post Tx		
Recurrent Liver Ca	75	(2.8% of all pts, 19% of pts with Ca at Tx)
De Novo Ca	130	(5%) 136 Ca
Skin Ca	310	(11%)
Total	515	(18%)
Multiple Ca	67	
Transferred from Donor	2	
<b>Developed non skin Ca &lt; 90days</b>	8	

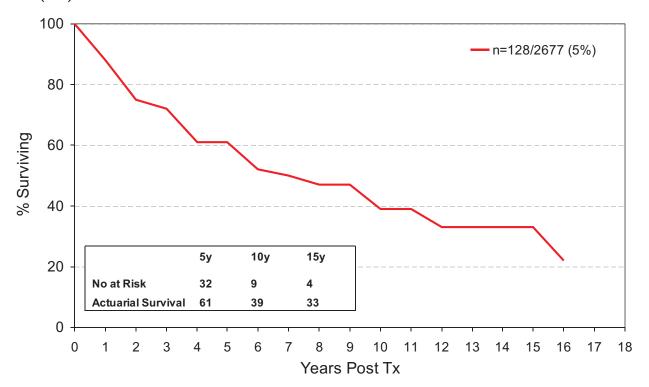
Primary Liver Cancer - N = 2677

TYPE OF CA	No	DIED	DIED OF THIS CA
HEPATOCELLULAR CA	109	34	19 (18%)
LAMELLA VARIANT	5	4	2 (40%)
HEPATOBLASTOMA	5	2	1 (20%)
CARCINOID	4	4	4 (100%)
CHOLANGIOCARCINOMA	1	1	1 (100%)
ANGIOSARCOMA	1	1	1 (100%)
EPITHELOID HAEMANGIOENDOTHELIOMA	1	0	0
GASTRINOMA	1	1	1 (100%)
PANCREATIC ISLET CELL	1	1	1 (100%)
TOTALS	128 (5% of pts)	48 (38% of those with PCa)	30 (23% of those with PCa)

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Primary Liver Cancer N = 2677

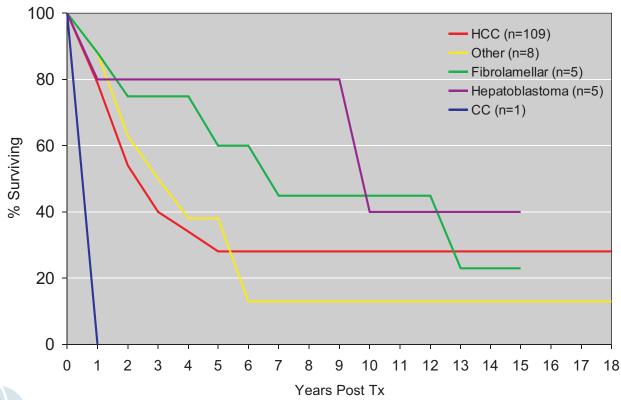
n = 128 (5%)



**Primary Liver Cancer Survival** 

N = 2677

n = 128 (5%)

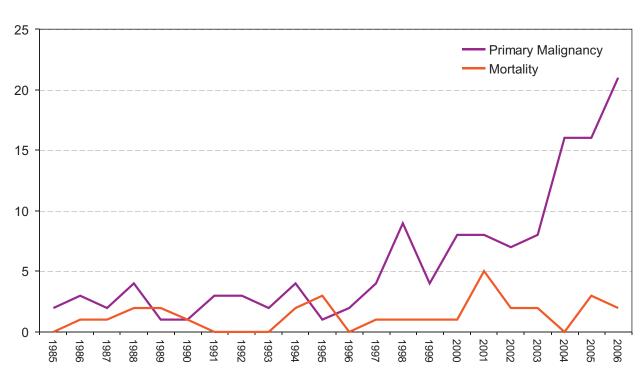


		<b>5</b> y	10yr	15yr
	n	24	5	2
HCC (n=110)	% Surviving	23	38	38
	n	4	2	2
Other (n=8)	% Surviving	38	13	13
	n	4	3	2
Hepatoblastoma (n=5)	% Surviving	80	40	40
	n	4	4	2
Fibrolamellar (n=5)	% Surviving	60	60	40
CC (n=1)	n	0		
	% Surviving	0		

**Primary Liver Cancer Incidence and Mortality** 

N = 2677

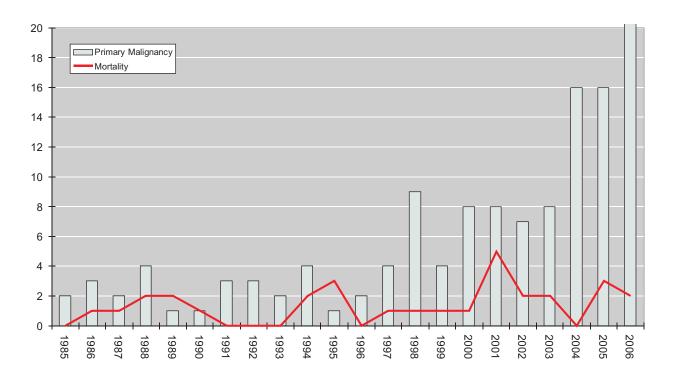
n = 128 (5%)



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N = 2677

n = 128 (5%)

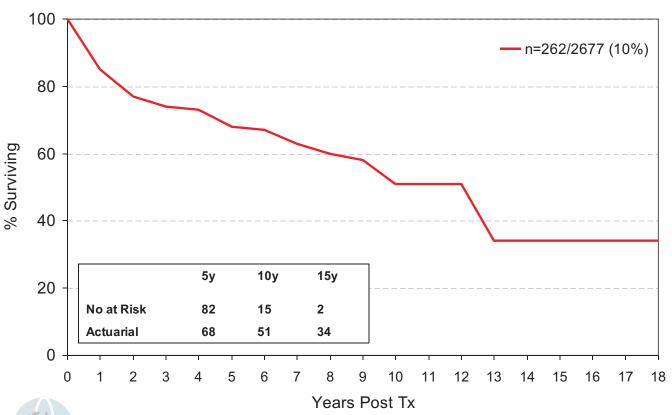


#### **Overall Survival**

#### Liver Cancer as a Secondary Diagnosis

N = 2677

n = 262 (10%)



36 (14% of pts

with SCa)

	No	Died	Died of This Cancer
HEPATOCELLULAR CA*	230	62	21 (9%)
CHOLANGIO CA	24	16	13 (54%)
ADENOCARCINOMA	3	3	0
HEPATOBLASTOMA*	2	1	0
FIBROLAMELLAR	2	1	1
ANGIOSARCOMA	1	1	1
EPITHELOID HAEMANGIOCA	1	0	0

263\* in 262

pts (10%)

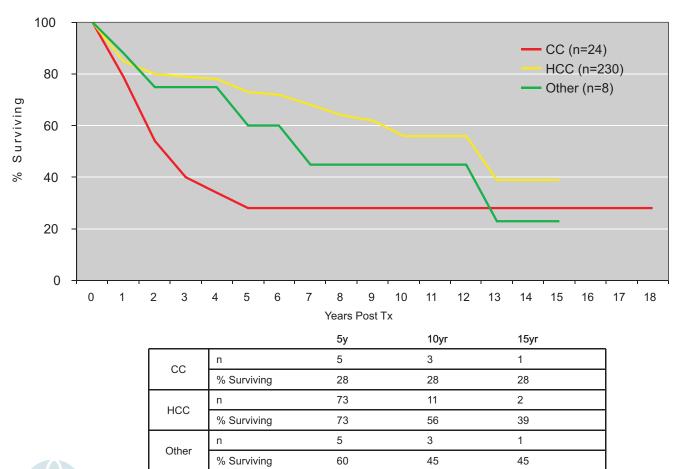
#### \* 1 patient had 2 secondary malignancies

84 (33% of pts

with SCa)

## Liver Cancer as a Secondary Diagnosis N = 2677

**Total** 



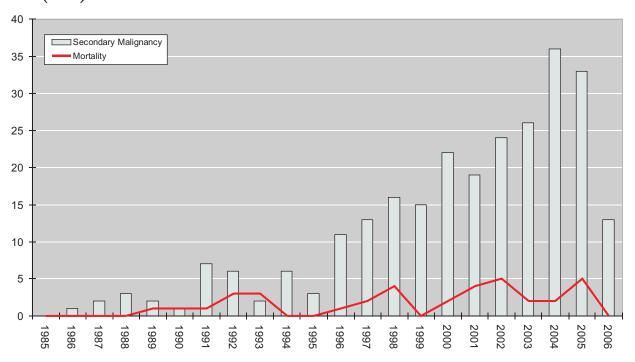
**Liver Cancer (Secondary Diagnosis) Incidence and Mortality** 

18<sup>TH</sup> ANZLT REGISTRY

CLICK HERE to return to Contents page

N = 2677

n = 262 (10%)



Patient Survival
Benign Disease vs Primary or Secondary Liver Malignancy
N = 2677





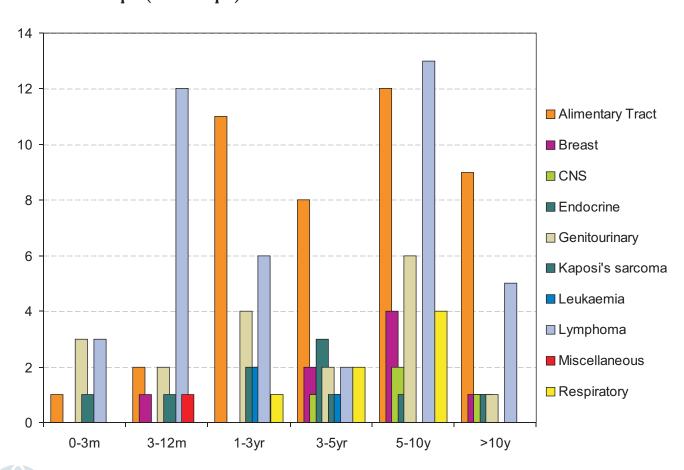
	No	Male	Female	Age of pts (yrs)	Time to diagnosis (mths)	Died of This Cancer
Alimentary*	43	29	14	12.6 – 74.8 (m 57)	3 – 173 (m 59)	22 (48%)
Lymphoma*	41	25	17	1.5 – 65.2 (m 45)	1 – 182 (m 40)	17 (48%)
Genitourinary*	18	10	8	38.5 – 70.5 (m 59)	2 – 164 (m 29)	2 (11%)
Breast	8	-	8	39.1 – 62.8 (m 50)	11 – 189 (m 79)	2 (25%)
Respiratory	7	4	3	44.7 – 61.1(m 56)	37 – 111(m 65)	5 (71%)
Kaposi's	5	4	1	32.1 – 64.6 (m 50)	2 – 48 (m 19)	0
Endocrine	5	2	3	35.8 – 69.5 (m 59)	47 – 144 (m 67)	2 (40%)
CNS	4	2	2	16.5 – 75 (m 47)	66 – 174 (m 93)	2 (50%)
Leukaemia	3	1	2	2.9 – 49.5 (m 33)	16 – 44 (m 30)	0
Multiple Myeloma	1	-	1	67 – 67 (m 67)	6 – 6 (m 6)	0
Total	*136 ca in 130 pts	77	59	1.5 – 75 (m 52)	1 – 189 (m 53)	52 (40% of pts with Ca)

Eight patients also had secondary liver malignancy; \* 5 patients had 2 de novo malignancies

#### **De Novo Non Skin Cancer Incidence**

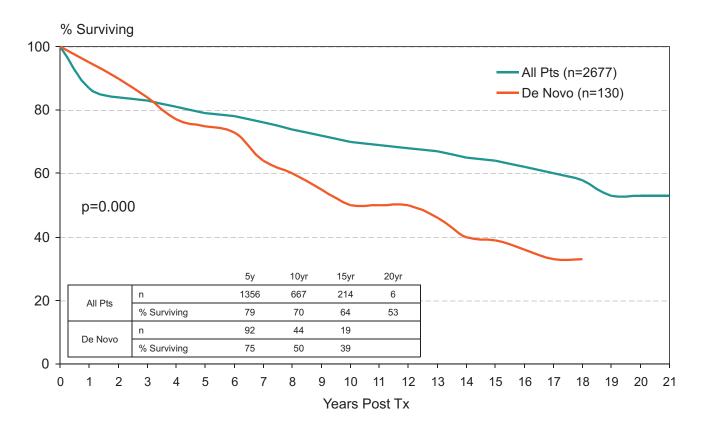
N = 2677

136 cancers in 130 pts (5% of all pts)



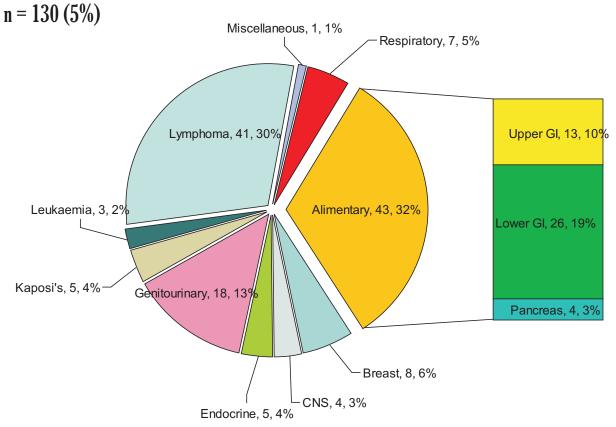
## De Novo Non Skin Cancer vs All Patients N = 2677





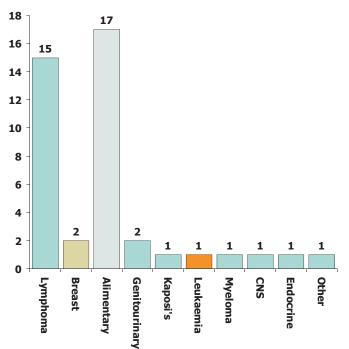




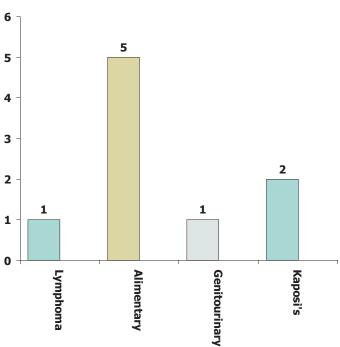






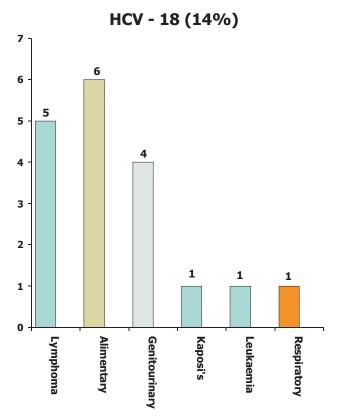


HBV - 9 (8%)

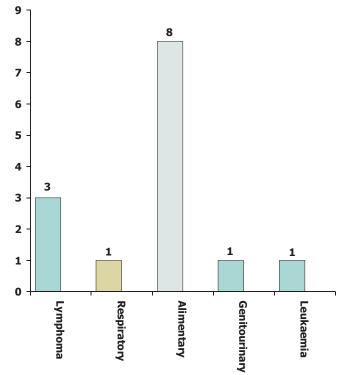


Pre Transplant Liver Disease and De Novo Non Skin Cancer

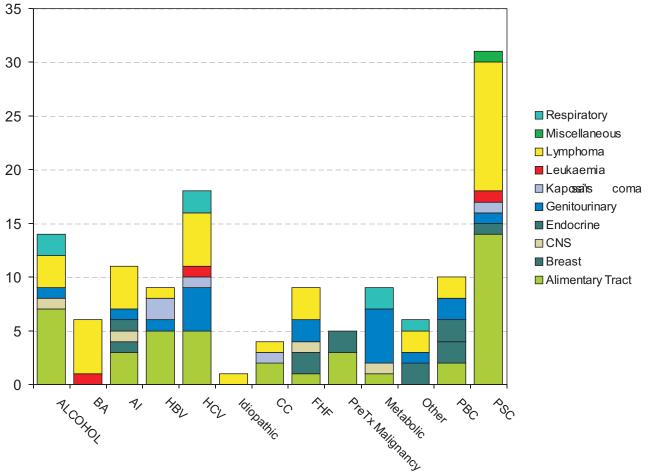
N = 2677



#### Alcohol - 14 (11%)







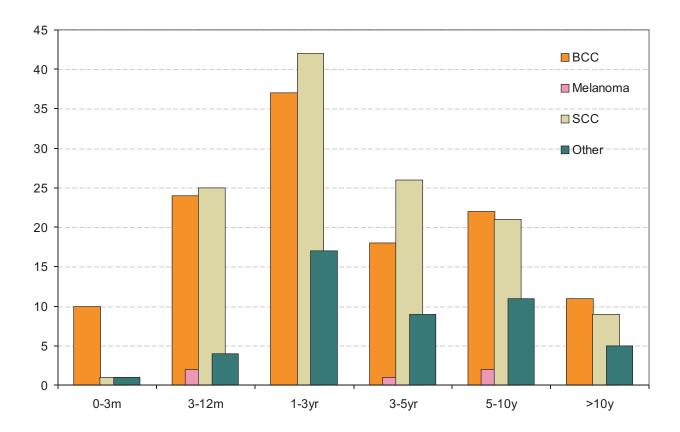
Skin Ca Post Ltx N= 2677

Type of Skin Cancer	Pts	Cancers
ВСС	228	531
SCC	246	759
Melanoma	15	15
Total	310 (11% of all pts)**	1927

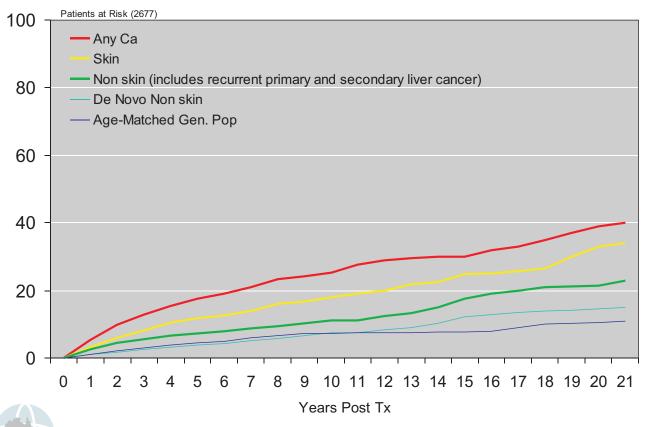
\*\* 182 pts had multiple skin cancer types



## Time to $1^{st}$ Skin Cancer Development N = 2677 310 (11% of all pts)



## Cumulative Risk of Diagnosis of Cancer Following Liver Tx. 1985-2006 N=2677





**APPENDIX** 

### 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	Total	
Primary Diagnosis	Child	Adult	
-1 Antitrypsin deficiency	30	39	69
Crigler-Najjar	4	1	5
Familial amyloid polyneuropathy	0	27	27
Glycogen storage disease	0	1	1
Haemochromatosis	2	23	25
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	26	33
Total	70	127	197

<sup>\*</sup> Bile acid synthesis disorder, Protein C deficiency, methylmalonic acidemia, familial immunodeficiency, mitochondrial disease

<sup>\*\*</sup> OTC deficiency 6; citrullinemia 4



## **Appendix III**

#### ANZLTR PRIMARY Diagnosis - Other by Age Group

	Age	Age group		
Primary Diagnosis	Child	Adult		
Alagille syndrome	22	1	23	
Alagille non-syndromic	2	0	2	
Benign liver tumour -Adenomatosis	0	1	1	
Benign liver tumour-Hemangioma	0	2	2	
Caroli's disease	1	12	13	
Choledocal cyst	1	2	3	
Cholestatic disease-Other	1	3	4	
Chronic Budd Chiari	1	24	25	
Congenital biliary fibrosis	1	1	2	
Ductopenia	0	3	3	
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	27	27	
Polycystic Liver disease	0	12	12	
Polycystic liver and kidney disease	0	4	4	
Progressive familial intrahepatic cholestasis(PFIC)	12	4	16	
Secondary biliary cirrhosis	1	9	10	
Secondary biliary cirrhosis - Hepatolithiasis	0	4	4	
Secondary biliary cirrhosis - Cystic fibrosis	7	11	18	
Other -specify	3	14	17	
Total	60	143	203	

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 group		Total
	Children	Adult	
Acute - Budd Chiari	0	2	2
Acute - Wilson's	4	11	15
Acute1 -AAT	2	0	2
Acute Autoimmune hepatitis	0	6	6
Acute Unknown / unspecified	33	58	91
Acute -Paracetamol	0	7	7
Acute -Other drugs	2	13	15
Acute Herbs / mushrooms	0	4	4
Acute - Hepatitis A	0	2	2
Acute - Hepatitis B	0	32	32
Acute - NonA-NonB	4	11	15
Acute - Hepatitis E	0	1	1
Acute - Post liver resection	1	1	2
Subacute - Wilson's	1	2	3
Subacute Autoimmune hepatitis	0	6	6
Subacute - Drug	0	3	3
Subacute - Unknown / unspecified	3	26	29
Subacute - Hepatitis A	0	2	2
Subacute - Hepatitis B	0	8	8
Total	50	195	245



### 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		3
Primary non function		19
Massive haemorrhagic necrosis		4
Recurrent disease		4
(ALD, PSC, CAH:AI)		
De novo Hep C		2
Biliary Complications		9
Other		10
(PNC, immune hepatitis, outflow obstruction)		

<u>Miscellaneous</u>	
AA 111	
Multiorgan failure	20
Renal Failure	13
Graft vs Host disease	5
Social	11
(accident, suicide,non-compliance, Rx withdrawn)	
Sudden death (cause unknown)	11
Other	7
(Hyperkalaemia, motor neurone disease	
diabetes complications, drug reaction, progression	
FAP)	