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Child Cancer Survival in New Zealand 2005 - 2014

A report from
The New Zealand Children's Cancer Registry

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- The Clinical Research Associates (CRAs) responsible for submitting data to the NZCCR. The quality of data held by the registry is due to the thoroughness and accuracy of these CRAs.

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Executive Summary

The 2005-2014 child cancer survival analysis replicates the methodology for first outcome analysis of the New Zealand Children Cancer Registry (NZCCR) 2000-2009. Cancer registrations for children under the age of 15 were primarily sourced from the NZCCR and also included cross-matching with the New Zealand Cancer Registry and Mortality Collection.

We are pleased to report continued improvements in the number of New Zealand children surviving childhood cancer. The five-year relative survival estimate for children diagnosed between 2005 and 2014 was 84%, an increase of 3% from the previously reported 2000-2009 period. One-year, three-year, and ten-year survival estimates were 91%, 85% and 82% respectively.

Compared to the 2000-2009 period, the greatest improvements in five-year survival were seen for neuroblastoma (+9% to 73%) and malignant bone tumours (+11% to 79%). Survival estimates were greater than 90% for retinoblastoma (100%), germ cell tumours (98%), lymphomas (96%), renal tumours (96%), and 'other malignant epithelial neoplasms' (90%) and leukaemia survival also increased from 85% to 89%. Improvements in five year survival were not seen for soft tissue sarcomas or central nervous system tumours (both 71%) during this time.

By age group, five-year survival estimates were unchanged for 0-4 year olds (83%) but improvements were seen for those aged 5-9 years (+4% to 84%) and 10-14 years (+6% to 84%).

There were no overall survival differences by sex although there was a noteworthy (but not statistically significant) difference in five-year survival for girls diagnosed with bone tumours (65%) compared to boys (89%).

There were 240 deaths recorded within this cohort of 1409 children. CNS tumours accounted for 38% of all deaths recorded with leukaemias accounting for an additional one in four cases (25%).

Although the overall ethnic survival differences were not yet statistically significant, there was a sizeable gap in five-year survival for Maori (79%) and Pacific Peoples (79%) compared to non-Maori/non-Pacific Peoples (86%). The survival improvements made since the 2000-2009 period are more evident for non-Maori/non Pacific children (+4%) than Maori (+2%), with five-year survival actually declining for Pacific Peoples in 2005-2014 (-3%).

The biggest five-year survival differences according to prioritised ethnicity were seen for those aged 10-14 years, with the survival gap between non-Maori/non-Pacific Peoples (88%) and Maori (75%) reaching statistical significance. Compared to 10-14 year olds diagnosed between 2000 and 2009, dramatic five-year survival gains were seen for non-Maori/non-Pacific Peoples (+9%) but not for Maori (+1%) or Pacific Peoples (-6%).

Small annual case numbers limits our ability to make ethnic comparisons by tumour group or disease staging at diagnosis. However, five-year survival for the most common childhood cancer, acute lymphoblastic leukaemia, remains consistent across the three prioritised ethnic groups, ranging from 91 to 93%.

New Zealand's overall five-year survival (84%) is comparable to the child cancer survival estimates published by countries recognised as world leaders in children's cancer research and treatment. These include Australia (84%), Canada (83%), the United States (83%), Switzerland (88%), and Germany (85%). Comparisons across diagnostic groups indicate that further survival gains are most likely achievable for New Zealand children diagnosed with bone tumours, soft tissue sarcomas, CNS tumours and neuroblastoma.

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1 Introduction

1.1 Childhood cancer in New Zealand

Although child cancers account for less than one percent of all cancers diagnosed each year,[1] cancer is nevertheless a significant health issue for New Zealand. Cancer remains the second most common cause of death, after traffic accidents, for 1-14 year olds[2] and it is estimated that around one in five hundred children will be diagnosed with cancer before the age of 15 years.[3] The majority of childhood cancers require intensive treatments conducted over an extended period, placing considerable stress on the child and their whanau. Many child cancer survivors will experience long-term adverse health effects as a result of their cancer and treatments.

Childhood cancer is developmental in origin, arising in growing and developing tissues and caused by the corruption of developmentally regulated genes. Hence, the natural patterns and types of cancer seen in children are very different to cancers seen in adults, which are strongly influenced by age, lifestyle and environmental risk factors. The relative rarity of childhood cancers, their unique biology and response to therapy, combined with the special needs of the child and their family mandates specialised care in dedicated child cancer treatment centres.

New Zealand has two specialist childhood cancer centres; the Starship Blood and Cancer Centre in Auckland and the Children's Haematology/Oncology Centre (CHOC) based in Christchurch Hospital. These two centres work closely with dedicated regional shared care services to ensure children can receive as much of their treatment as close to home as possible.

1.2 NZCCR background and purpose

The need to record and report the pattern of cancer seen in New Zealand children was first recognised over 40 years ago when, Dr David Becroft, a paediatric pathologist, began recording new cases of cancer presenting to the Princess Mary Hospital for Children. By the 1980s, members of the Paediatric Oncology Co-ordinating Committee of the Paediatric Society were registering all cases referred to each of their five children's cancer tertiary centres. This was due, in no small part, to the dogged determination of Dr Margaret Lewis, a Wellington paediatrician and academic who was passionate about establishing a nationwide children's cancer registry. When the Ministry of Health established the National Paediatric Oncology Steering Group (POSG) in 1999, one of the goals they were set was to establish a specific national children's cancer registry to provide contemporary data on the diagnosis and long-term outcome of all New Zealand children.

Although diagnostic pathology laboratories are mandated by law to report all cancer diagnoses to the New Zealand Cancer Registry (NZCR), the data collected for each patient is necessarily limited. The New Zealand Children's Cancer Registry was established in 2002 at the request of the Ministry of Health (MOH) to provide more detailed information about childhood cancer incidence, treatment, and outcomes in New Zealand. In addition to the data fields routinely collected by the NZCR, the NZCCR collects detailed clinical information regarding the stage of the disease, the treatments given, and treatment-related late effects for all children receiving treatment in a paediatric oncology setting. The NZCCR also classifies all cancers according to the International Classification of Childhood Cancers (ICCC-3)[4], which allows New Zealand child cancer survival to be directly compared with international benchmarks. The registry is now under the governance of the National Child Cancer Network (NCCN). It holds verified demographic and treatment information for all children diagnosed with cancer in New Zealand since the 1st of January 2000.

The NZCCR serves multiple functions. At an individual patient level, the information collected and held by the registry is later accessed by the Late Effects Clinical Nurse Specialist to produce the patient's end of treatment summary and surveillance plan. It is therefore a "living" registry. At a national and international level, the registry provides anonymised datasets used for service delivery planning, research, and statistical reporting purposes. The NZCCR has approval from the Health and Disability Ethics Committee for the ongoing collection and analysis of registry data. Ongoing analysis of the NZCCR is central to evaluating the health outcomes for New Zealand children with cancer; identifying the spectrum of cancers diagnosed, tracking improvements in survival over time, and assessing whether any disparities exist in treatment outcomes.

1.3 Registration processes

All data is initially entered onto the NZCCR by a Clinical Research Associate at each specialist paediatric oncology centre. The data is verified and electronically sent to the NZCCR national database. Access to the NZCCR is controlled by personal login and password and staff can only access patient data entered by their own centre to ensure privacy and confidentiality. Parents are informed of the NZCCR via a parent information sheet and have the opportunity to opt out of the registry at any time. The NZCCR Working Group, reporting directly to the NCCN, is charged with the ongoing administration of the registry. The NZCCR Working Group has representatives from both paediatric oncology treatment centres including the NZCCR Analyst, Clinical Research Associates, and Consultant Haematologists/Oncologists.

1.4 NZCCR registration criteria

In order to ensure that the NZCCR provides an accurate workload model for service delivery planning, all patients who receive cancer treatment in a paediatric oncology centre are registered on the NZCCR.

However, not all NZCCR registrations are included in New Zealand child cancer incidence and outcome statistics. For example, 15-18 year olds diagnosed with cancer in New Zealand may receive their care in a paediatric oncology setting when this is judged to be in the best interests of the adolescent and their family, but these adolescents are not included in New Zealand child cancer statistics. Also, non-New Zealand residents, or children who were diagnosed with cancer and treated overseas before emigrating or returning to New Zealand are also excluded. Finally, children requiring paediatric haematology / oncology involvement for diseases not currently included in the ICCC-3[4] are excluded from any overall analyses.

The criteria for inclusion in any NZCCR incidence and outcome statistics are as follows;

- *The child was aged less than 15 years old at diagnosis*
- *The child was diagnosed and received treatment in New Zealand*
- *The child had New Zealand residency at the time of diagnosis*
- *The child's diagnosis is included in the ICCC-3[4]*

1.5 The International Classification of Childhood Cancers (ICCC-3)

The first internationally accepted childhood cancer classification system was developed by Jillian Birch and Henry Marsden in 1987[5] and was used for generating international comparisons for the International Incidence of Childhood Cancer, Volume 1, published by the International Association for Research on Cancer (IARC).[6] While adult cancers are classified according to the location in the body where the cancer originates, the International Classification of Childhood Cancers recognises that for childhood cancers it is the tissue of origin which best predicts the tumour behaviour and dictates the required treatment. The ICCC[4], now in its' third revision, is the standard for the presentation of international data on childhood cancer incidence and survival, accepted by the World Health Organisation (WHO), IACR and the United States Surveillance Epidemiology and End Results (SEER). The NZCCR classifies all registrations according to the ICCC-3.

The ICCC-3[4] contains 12 diagnostic groups (see Table 1.5), which are further divided into 47 diagnostic subgroups. Appendix AII provides full details of the ICCC-3 based on the International Statistical Classification of Diseases for Oncology, 3rd Edition (ICD-O-3) site and histology[7]. According to the ICCC-3[4], a cancer diagnosis must be that of a primary **malignant** neoplasm in order to be registered. However, there is one important exception: **all** intracranial and intraspinal neoplasms (including benign tumours or those of uncertain behaviour) are included. This is due to non-malignant intracranial and intraspinal neoplasms having similar prognoses, clinical symptoms, and late effects to malignant neoplasms.[4] The inclusion of non-malignant intracranial/intraspinal neoplasm mostly concerns 'diagnostic group III: central nervous system and miscellaneous intracranial and intraspinal neoplasms', but very occasionally a child may be diagnosed with a benign intracranial/intraspinal germ cell tumour (diagnostic group Xa) which also meets ICCC-3[4] criteria for cancer registration.

Table 1.5 International Classification of Childhood Cancer (ICCC-3)[4] diagnostic groups

Group	Title (the abbreviated title used throughout this report is highlighted in bold)
I.	**Leukaemias,** myeloproliferative diseases, and myelodysplastic diseases
II.	**Lymphomas** and reticuloendothelial neoplasms
III.	Central nervous system (**CNS**) **tumours** and miscellaneous intracranial and intraspinal neoplasms
IV.	**Neuroblastoma** and other peripheral nervous cell tumours
V.	**Retinoblastoma**
VI.	**Renal tumours**
VII.	**Hepatic tumours**
VIII.	**Malignant bone tumours**
IX.	**Soft tissue sarcomas** and other extraosseous sarcomas
X.	**Germ cell tumours,** trophoblastic tumours, and neoplasms of gonads
XI.	**Other malignant epithelial** neoplasms and malignant melanomas
XII.	**Other and unspecified** malignant neoplasms

1.6 The purpose and structure of this report

The purpose of this report is to provide a comprehensive analysis of the survival of childhood cancer in New Zealand for the period of the 1st of January 2005 to the 31st of December 2014. A second companion report released in August 2017 contains incidence data for the 2010-2014 time period. In order to aid comparisons, the methodology and reporting structure replicates the previously published 2000-2009 report[8] wherever possible.

Chapter Two provides a description of the methodology used in this analysis. It details the data selection and verification process and the way in which child cancer survival has been calculated and reported.

Chapter Three contains survival data for children aged less than 15 years first diagnosed with cancer between the 1st of January 2005 and the 31st of December 2014, with follow-up to the 31st of December 2016. Cancer survival is reported by ICCC-3[4] diagnostic group and subgroup according to sex, age at diagnosis, and prioritised ethnicity. It is expressed in terms of relative survival, which accounts for the underlying survival of the general child population within the same time period. Following the international standard, the survival reported is predominantly five-year survival.

Chapter Four provides an overview of the survival probabilities pertaining to each ICCC-3[4] diagnostic group and each diagnostic subgroup, where sample size allows. Due to the small number of cases that were assigned to 'diagnostic group XII: other and unspecified malignant neoplasms', no specific analyses were undertaken for this group.

2 Methodology

2.1 Data selection, validation, and conversion

Ongoing approval for the collection, analysis and publication of NZCCR data was approved by the Multi-region Ethics Committee (ethics ref: MEC/11/EXP/134) in December 2011.

Prior to any data analysis of the NZCCR, a rigorous data validation process took place. All registrations not meeting NZCCR inclusion criteria for child cancer statistics (such as those patients initially diagnosed overseas and those children over the age of 15 treated in a paediatric centre) were excluded from the final dataset. All anomalies were investigated and any remaining data gaps were filled. The NZCCR and NZCR collaborated to validate all registrations held by the NZCCR and the NZCR for the 2010-2014 period. The 2005-2009 registrations had already been matched with the NZCR prior to the 2000-2009 incidence and survival analyses but these earlier registrations were also re-checked against the national Mortality Collection to ensure that the NZCCR had correctly recorded all dates of death.

Most differences identified in the NZCR and NZCCR data matching exercise were explained by the different registration criteria used. An important area covered by the ICCC are the benign/low grade CNS tumours. There is international agreement that these classes of tumour should be registered in children as they require significant intervention and are associated with significant morbidity and some deaths.[5] Non-malignant central nervous system tumours are registered on the NZCCR however, many international cancer registries, including the NZCR, register malignant CNS tumours only (i.e. those tumours with a behaviour code of '3: primary malignant neoplasm'). This resulted in an additional 55 non-malignant CNS tumours registered by the NZCCR but not the NZCR in the 2010-2014 period. Also, the NZCCR adopted the first revision of the ICD-O-3[9] as from the 1st January 2010 while the NZCR implemented it on 1st January 2014. This lead to the NZCCR's inclusion of an additional 16 Langerhans cell histiocytosis (LCH) cases, which had been re-classified as a malignancy in the revision. The audit identified 24 cases registered in error by the NZCR, most commonly children who had been initially diagnosed in the Pacific Islands but who were subsequently flown to New Zealand for part of their treatment.

Data matching with the NZCR led to the identification of 43 child cancer cases which were unknown to the NZCCR but which met our registration criteria. Although the referral of all child cancer cases to paediatric oncology services is strongly encouraged, 19 cases in this time period were not referred to a specialist paediatric oncology centre; 5 carcinomas, 5 melanomas, 4 CNS tumours, 3 sarcomas and 2 germ cell tumours. In addition, five children were diagnosed at autopsy or within one week of their date of death. Nineteen cases were known to the paediatric centres but missed by the NZCCR due to human error; the most common cause was a misunderstanding regarding the requirement to register myelodysplastic syndrome cases. In summary, the review highlighted that the NZCCR had improved in case ascertainment since 2000-2009 (now at 94%), but some improvements were identified to further improve both New Zealand's paediatric oncology referral pathways and NZCCR registration practices.

2.2 Prioritised ethnicity

According to MOH ethnicity data protocols, individuals may select up to three ethnic groups that they identify with. When a prioritised ethnicity system is used, each respondent is assigned to a single ethnic group using a priority system; Maori, Pacific Peoples, and non-Maori/non-Pacific Peoples (European, Asian, Middle Eastern, Latin American, 'Not Elsewhere Reported' and 'Not Stated/Unknown'). Assigning a single ethnicity simplifies the data as the ethnic group populations sum to the total New Zealand population, but there are limitations with prioritisation; an increasing number of New Zealand children and young people identify with more than one ethnic group and the use of prioritised ethnicity goes against the principle of self-identification. However, prioritised output is often used in the health and disability sector to ensure that Maori and Pacific Peoples, whose health status is lower on average than that of other New Zealanders, are not swamped by the European group.[10]

Cancer registrations which had no ethnicity recorded were assigned to the 'non-Maori/non-Pacific Peoples' prioritised ethnic group. In the cases of 'unknown' ethnicity, the fact that no ethnicity data was collected may suggest that the person was diagnosed with a cancer which had an excellent prognosis, requiring minimal treatment and little, if any, involvement with the public health system. Although there was a very small number of 'unknown' ethnicity overall, the inclusion of the 'unknowns' within the 'non-Maori/non-Pacific Peoples' ethnic group has potentially slightly over-inflated this group's cancer survival.

Table 2.2 shows the New Zealand child population according to the 2006 census by prioritised ethnicity, and also by sex and age group.

Table 2.2 New Zealand's child population by age group, sex, and prioritised ethnicity, 2006 census data

	0-4 years		5-9 years		10-14 years		Total 0-14 years	
	2006 census population[a]	%	2006 census population[a]	%	2006 census population[a]	%	2006 census population[a]	%
Sex								
Male	140 382	51.0	146 535	51.1	157 113	51.3	**444 030**	51.2
Female	134 697	49.0	139 956	48.9	148 893	48.7	**423 546**	48.8
Prioritised Ethnicity								
Maori	66 423	24.2	66 771	23.3	66 726	21.8	**199 920**	23.0
Pacific Peoples	25 176	9.2	25 365	8.9	24 996	8.2	**75 537**	8.7
Non-Maori/non-Pacific Peoples	183 477	66.7	194 352	67.8	214 290	70.0	592 119	68.3
Asian Peoples	*21 279*	*7.7*	*22 935*	*8.0*	*26 268*	*8.6*	*70 482*	*8.1*
Other (MELAA and other ethnicities)[a]	*2 094*	*0.7*	*2 244*	*0.8*	*2 409*	*0.8*	*6 747*	*0.8*
European / NZ European	*148 707*	*54.1*	*158 265*	*55.2*	*174 360*	*57.0*	*481 332*	*55.5*
Not elsewhere included[b]	*11 397*	*4.1*	*10 908*	*3.8*	*11 253*	*3.7*	*33 558*	*3.9*
Total 2006 census population[c]	**275 076**		**286 491**		**306 009**		**867 576**	

[a] MELAA: Middle Eastern, Latin American and African.
[b] Includes 'response unidentifiable', 'response outside scope' and 'not stated'.
[c] Statistics NZ applies random rounding to base 3 to census outputs. As each value in a table is rounded independently, the marginal totals can differ very slightly from the corresponding sum of the rows or columns.

2.3 Survival calculations

A range of measures can be used to report cancer survival. The most commonly used measures of survival when reporting data from population-based cancer registries are observed survival and relative survival. Relative survival ratios are calculated by dividing the observed survival (i.e. the proportion of people who remain alive following their cancer diagnosis) by the expected survival of a group from the general population which is comparable with respect to age, sex, and the time period under investigation. Relative survival does not require information about the actual cause of death and allows comparisons between different areas with different population structures. Two approaches are commonly used for calculating relative survival; the cohort method and the period method. This study uses the period method, which provides more accurate survival predictions for recently diagnosed patients.[11]

Date of death was sourced from the New Zealand Children's Cancer Registry and also from the Ministry of Health national collections. The final date of follow-up was the 31st of December 2016, and those who were still alive at that date were censored. To avoid bias, those rare cases where the cancer diagnosis was based on death certificate or autopsy only and children who had a survival time of zero days were excluded. Expected survival data was calculated according to the Ederer II method using life-tables for the total New Zealand resident population. These tables are produced by Statistics New Zealand. The observed survival and expected survival data were used to calculate estimated cancer survival ratios using the Stata® MP14.2 statistical software package.

Although relative survival should technically be expressed as a ratio, we have chosen to convert the ratios to a percentage. This report is intended for a wide audience and we consider that using percentages makes the report easier for the general reader to follow. Also, within this report we are making comparisons with survival data published by other cancer registries, all of which have expressed relative survival as a percentage. Note that it is possible for relative survival to be greater than 100%. That is, those children diagnosed with a particular cancer may have survival which is actually better than the survival for the general child population. For example, five-year relative survival for children diagnosed with retinoblastoma was 100.2%, as there was not a single death recorded among this group of patients within the study period. While we have produced tables showing cumulative relative survival for up to 10 years, following standard cancer survival methodology we focus primarily on five-year relative survival.

2.4 Confidence intervals and statistical significance

A confidence interval (CI) is used to report the level of accuracy of statistical estimates. The reported 95% confidence intervals can be interpreted as indicating that there is a 95% probability that the true cancer survival lies somewhere within the reported lower and upper values. If two statistics have non-overlapping 95% confidence intervals, they are necessarily significantly different at the $p<0.05$ level. Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.

In general, the more cases involved in calculating the estimate, the smaller the confidence interval. For some cancer diagnostic groups and subgroups there were very few cases recorded for children in New Zealand within the ten year period; this is reflected in the wide 95% confidence intervals which are reported alongside. Therefore, **any between-group differences in the incidence or survival reported, or any differences in comparison to other published data, should be interpreted extremely cautiously.**

2.5 Survival comparisons

In Section 3.5 child cancer survival for the 2005-2014 period is compared with earlier published New Zealand data.[8,12,13] In Section 3.6 our childhood cancer survival is compared with survival data reported from Australia,[14] Switzerland,[15] Canada,[16] Germany,[17] and the United States.[18] These countries were selected as they had published childhood cancer survival probabilities by ICCC-3 for 0-14 year olds for a comparable time period. Such comparisons are vital as they describe the progress made in our child cancer survival over time, and allow us to place New Zealand's survival in the context of what is being achieved internationally. However, the data must be interpreted cautiously as the survival probabilities reported may be influenced by differences in timing, population composition, registry data quality, and completeness.[19]

Studies with a range of different survival calculation methods have been included in these comparisons. Although it would be ideal if all studies had calculated survival using the same methodology, it should be noted that mortality in the general child population in industrialised nations is low, so child cancer survival does not typically show much variability according to whether observed survival, cause-specific survival (where non-cancer related deaths are censored), or relative survival has been used.[11]

2.6 Deaths within the cohort

In this report we present charts showing the proportionate number of deaths which were recorded within this study population, by ICCC-3[4] diagnostic group. Note that the numbers of deaths reported for this cohort are not the same as the child cancer mortality for 2005-2014, as the figures do not include children who died within this period but who had been diagnosed prior to the year 2005. However, the raw numbers and percentages do provide an *indication* of the diagnostic groups which are the leading cause of death among New Zealand children diagnosed with cancer. As cause of death was retrieved from the NZCCR and the Mortality Collection, it can be confirmed that in all but three cases the primary cause of death was attributed to the child's cancer and/or treatment. Within this particular analysis, the three individuals who died of unrelated causes were not included within the total deaths for their respective diagnostic groups.

3 Childhood Cancer Survival

The following section reports the relative survival for those New Zealand children (0-14 years) diagnosed with cancer between the 1st of January 2005 and the 31st of December 2014, with follow-up to the 31st of December 2016. Relative survival is calculated by dividing the observed survival (i.e. the proportion of people who remain alive following their cancer diagnosis) by the expected survival of a group from the general population which is comparable with respect to age, sex, and the time period under investigation (see Section 2.3 for further details).

For some diagnostic groups and subgroups there were very few cases recorded. In such cases, the true survival cannot be reliably estimated and this is reflected in the wide 95% confidence intervals which are reported alongside. Therefore, any between-group differences in survival should be interpreted cautiously. It should also be noted that confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.

3.1 Overall childhood cancer survival

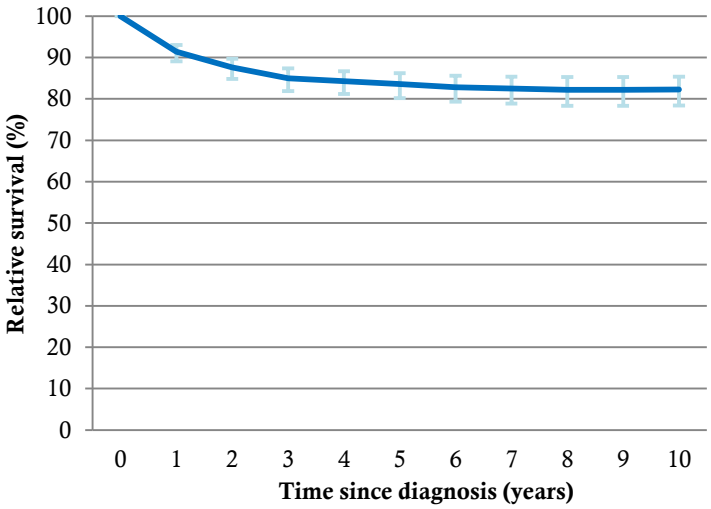
3.1.1 Childhood cancer relative survival by time since diagnosis

There were 1409 new childhood cancer registrations in the ten years between 2005 and 2014[a]. Overall cancer relative survival for children was 91.4% at one year, 85.0% at three years, and 83.6% at five years (see Table 3.1.1 and Figure 3.1.1). There continued to be a slight decline in relative survival (-1.3%) between five years and ten years of follow-up. Survival estimates were higher than those reported for the 2000-2009 period; one year survival increased by 2.0%, three year survival increased by 1.9%, five year survival increased by 2.9% and 10 year survival increased by 3.8%.

Table 3.1.1 Childhood cancer relative survival by time since diagnosis, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	1409	91.4	(89.8 - 92.8)
2	1287	87.6	(85.8 - 89.2)
3	1233	85.0	(83.0 - 86.8)
4	1078	84.3	(82.3 - 86.2)
5	935	83.6	(81.5 - 85.5)
6	782	82.8	(80.6 - 84.7)
7	651	82.5	(80.3 - 84.5)
8	527	82.2	(80.0 - 84.2)
9	418	82.2	(80.0 - 84.3)
10	302	82.3	(80.0 - 84.3)

Figure 3.1.1 Childhood cancer relative survival by time since diagnosis, New Zealand, 2005-2014



[a] Note that this figure excludes five cases which were notified by death certificate/autopsy only which, according to standard practice, are included in cancer incidence but excluded from survival figures.

3.1.2 Survival by ICCC diagnostic group

Table 3.1.2 shows that one-year relative survival ranged from 77.9% for hepatic tumours to 100.1% for retinoblastoma (i.e. survival for this diagnostic group was slightly higher than for the general child population). The greatest decline in survival between one and three years was seen for soft tissue sarcomas (-15.7%) and neuroblastoma (-12.1%).

Table 3.1.2 One-year, three-year, five-year and ten-year cumulative relative survival by ICCC diagnostic group, New Zealand, 2005-2014

ICCC-3 diagnostic group		Total cases	One-year cumulative relative survival (95% CI)		Three-year cumulative relative survival (95% CI)		Five-year cumulative relative survival (95% CI)		Ten-year cumulative relative survival (95% CI)	
	All childhood cancers	**1409**	**91.4**	**(89.8 - 92.8)**	**85.0**	**(83.0 - 86.8)**	**83.6**	**(81.5 - 85.5)**	**82.3**	**(80.0 - 84.3)**
I.	Leukaemias	476	95.4	(93.1 - 97.0)	90.3	(87.2 - 92.7)	88.7	(85.3 - 91.3)	86.4	(82.5 - 89.5)
II.	Lymphomas	149	96.7	(92.2 - 98.6)	95.4	(90.5 - 97.8)	95.5	(90.5 - 97.9)	95.7	(90.7 - 98.1)
III.	CNS tumours	295	80.7	(75.7 - 84.8)	72.5	(67.0 - 77.3)	70.5	(64.8 - 75.4)	69.0	(63.2 - 74.1)
IV.	Neuroblastoma	102	86.5	(78.1 - 91.8)	74.4	(64.7 - 81.9)	73.3	(63.4 - 80.9)	73.4	(63.5 - 81.0)
V.	Retinoblastoma	41	100.1	[a]	100.2	[a]	100.2	[a]	100.3	[a]
VI.	Renal tumours	53	98.2	(87.5 - 99.9)	96.3	(85.7 - 99.2)	96.4	(85.7 - 99.2)	96.4	(85.8 - 99.3)
VII.	Hepatic tumours	18	77.9	(51.2 - 91.2)	72.4	(45.7 - 87.6)	72.4	(45.7 - 87.6)	[b]	[b]
VIII.	Malignant bone tumours	79	92.4	(83.9 - 96.5)	82.2	(71.8 - 89.1)	79.0	(67.9 - 86.7)	79.2	(68.1 - 86.9)
IX.	Soft tissue sarcomas	91	89.1	(80.6 - 94.0)	73.4	(62.9 - 81.3)	70.6	(59.7 - 79.0)	68.7	(57.3 - 77.6)
X.	Germ cell tumours	50	98.2	(86.8 - 99.9)	98.2	(86.8 - 99.9)	98.3	(86.9 - 100.0)	98.5	(87.1 - 100.2)
XI.	Other malignant epithelial	53	96.2	(85.8 - 99.1)	90.3	(78.2 - 95.9)	90.4	(78.3 - 96.0)	87.3	(73.0 - 94.5)
XII.	Other & unspecified	2	100.0	[a]	100.1	[a]	100.1	[a]	[b]	[b]

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.
[b] Ten-year relative survival could not be calculated as no cases had a full ten years of follow-up.

3.1.3 Five-year relative survival by ICCC diagnostic group and subgroup

Table 3.1.3 shows that by diagnostic group, the highest five-year survival probabilities were recorded for retinoblastoma (100.2%, 41 cases), germ cell tumours (98.3%, 50 cases), renal tumours (96.4%, 53 cases), and lymphomas (95.5%, 149 cases). For the first time New Zealand five-year child cancer survival for all 12 ICCC diagnostic groups exceeded 70%, albeit marginally for central nervous system tumours (70.5%, n=295) and soft tissue sarcomas (70.6%, n=91).

Reporting overall survival by diagnostic group can be misleading, as survival can vary considerably according to diagnostic subgroup. By diagnostic subgroup, acute lymphoblastic leukaemia survival (ALL; 92.6%, n=362) was significantly higher than the survival probability for acute myeloid leukaemia (AML; 73.5%. n=76) in the same period. Among the CNS diagnostic subgroups, five-year survival for 'intracranial & intraspinal embryonal tumours' (such as medulloblastoma) and 'other gliomas' were significantly poorer than for astrocytomas, 'ependymomas & choroid plexus tumours' and 'other specified intracranial and intraspinal neoplasms' which all had survival estimates which exceeded 80%.

Table 3.1.3 Five-year relative survival by ICCC diagnostic group and subgroup, New Zealand, 2005-2014

	ICCC-3 diagnostic group/subgroup	Total cases	Five-year relative survival (95% CI)	
	All childhood cancers	**1409**	**83.6**	**(81.5 - 85.5)**
I.	**Leukaemias, myeloproliferative & myelodysplastic diseases**	**476**	**88.7**	**(85.3 - 91.3)**
I(a)	*Lymphoid leukaemias*	362	92.6	(89.2 - 95.0)
I(b)	*Acute myeloid leukaemias*	76	73.5	(62.0 - 82.1)
I(c)	*Chronic myeloproliferative diseases*	7	85.8	(33.4 - 98.0)
I(d)	*Other myeloproliferative diseases*	17	94.3	(65.1 - 99.3)
I(e)	*Other & unspecified leukaemia*	14	64.4	(34.4 - 83.5)
II.	**Lymphoma & reticuloendothelial neoplasms**	**149**	**95.5**	**(90.5 - 97.9)**
II(a)	*Hodgkin lymphomas*	51	100.1	[a]
II(b)	*Non-Hodgkin lymphomas (excl. Burkitt lymphomas)*	49	94.0	(82.3 - 98.1)
II(c)	*Burkitt lymphomas*	22	86.4	(63.5 - 95.5)
II(d)	*Miscellaneous lymphoreticular neoplasms*	26	96.4	(75.9 - 99.7)
II(e)	*Unspecified lymphomas*	1	100.1	[a]
III.	**Central nervous system & intracranial/intraspinal neoplasms**	**295**	**70.5**	**(64.8 - 75.4)**
III(a)	*Ependymomas & choroid plexus tumours*	29	89.6	(70.9 - 96.7)
III(b)	*Astrocytomas*	117	82.1	(73.9 - 88.0)
III(c)	*Intracranial & intraspinal embryonal tumours*	67	52.9	(40.0 - 64.2)
III(d)	*Other gliomas*	41	46.0	(30.3 - 60.4)
III(e)	*Other specified intracranial & intraspinal neoplasms*	32	89.8	(71.1 - 96.7)
III(f)	*Unspecified intracranial & intraspinal neoplasms*	9	33.4	(7.9 - 62.5)
IV.	**Neuroblastoma & other peripheral nervous cell tumours**	**102**	**73.3**	**(63.4 - 80.9)**
IV(a)	*Neuroblastoma & ganglioneuroblastoma*	102	73.3	(63.4 - 80.9)
IV(b)	*Other peripheral nervous cell tumours*	-	-	-
V.	**Retinoblastoma**	**41**	**100.2**	**[a]**
VI.	**Renal tumours**	**53**	**96.4**	**(85.7 - 99.2)**
VI(a)	*Nephroblastoma & other non-epithelial renal tumours*	53	96.4	(85.7 - 99.2)
VI(b)	*Renal carcinomas*	-	-	-
VI(c)	*Unspecified malignant renal tumours*	-	-	-
VII.	**Hepatic tumours**	**18**	**72.4**	**(45.7 - 87.6)**
VII(a)	*Hepatoblastoma*	12	83.6	(48.3 - 95.9)
VII(b)	*Hepatic carcinomas*	6	50.1	(11.1 - 80.5)
VII(c)	*Unspecified malignant hepatic tumours*	-	-	-
VIII.	**Malignant bone tumours**	**79**	**79.0**	**(67.9 - 86.7)**
VIII(a)	*Osteosarcomas*	38	78.1	(60.7 - 88.5)
VIII(b)	*Chondrosarcomas*	1	100.3	[a]
VIII(c)	*Ewing tumours & related bone sarcomas*	32	80.5	(61.3 - 90.8)
VIII(d)	*Other specified malignant bone tumours*	8	75.1	(31.5 - 93.2)
VIII(e)	*Unspecified malignant bone tumours*	-	-	-

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.

Table 3.1.3 (cont.) Five-year relative survival by ICCC diagnostic group and subgroup, New Zealand, 2005-2014

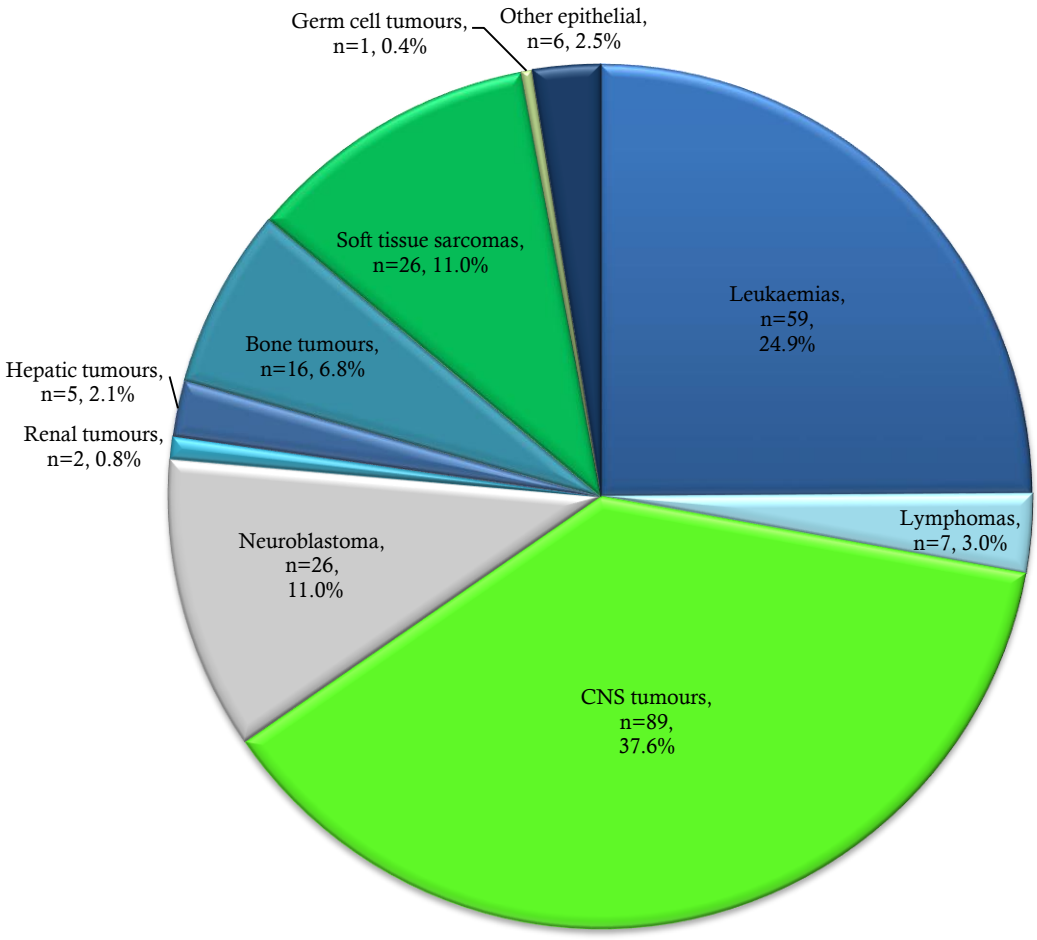
	ICCC-3 diagnostic group/subgroup	Total cases	Five-year relative survival (95% CI)	
IX.	**Soft tissue and other extraosseous sarcomas**	**91**	**70.6**	**(59.7 - 79.0)**
IX(a)	*Rhabdomyosarcomas*	48	66.0	(50.5 - 77.7)
IX(b)	*Fibrosarcomas & other fibrous neoplasms*	6	83.6	(27.4 - 97.7)
IX(c)	*Kaposi sarcomas*	-	-	-
IX(d)	*Other specified soft tissue sarcomas*	28	69.8	(48.0 - 83.9)
IX(e)	*Unspecified soft tissue sarcomas*	9	89.1	(43.4 - 98.6)
X.	**Germ cell tumours, trophoblastic tumours & neoplasms of gonads**	**50**	**98.3**	**(86.9 - 100.0)**
X(a)	*Intracranial & intraspinal germ cell tumours*	20	95.1	(69.6 - 99.4)
X(b)	*Malignant extracranial & extragonadal germ cell tumours*	14	100.5	[a]
X(c)	*Malignant gonadal germ cell tumours*	15	100.3	[a]
X(d)	*Gonadal carcinomas*	-	-	-
X(e)	*Other & unspecified malignant gonadal tumours*	1	[b]	[b]
XI.	**Other malignant epithelial neoplasms & malignant melanomas**	**53**	**90.4**	**(78.3 - 96.0)**
XI(a)	*Adrenocortical carcinomas*	3	66.7	(5.4 - 94.6)
XI(b)	*Thyroid carcinomas*	7	100.1	[a]
XI(c)	*Nasopharyngeal carcinomas*	1	100.3	[a]
XI(d)	*Melanomas*	21	95.4	(70.8 - 99.5)
XI(e)	*Skin carcinomas*	-	-	-
XI(f)	*Other & unspecified carcinomas*	21	84.8	(59.6 - 94.9)
XII.	**Other & unspecified malignant neoplasms**	**2**	**100.1**	**[a]**
XII(a)	*Other specified malignant tumours*	2	100.1	[a]
XII(b)	*Other unspecified malignant tumours*	-	-	-

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.
[b] Five-year relative survival could not be calculated as no cases had a full five years of follow-up.

3.1.4 Cancer-related deaths in children first diagnosed with cancer between 2005 and 2014

240 deaths were recorded within the cohort during the follow up period (up to December 31st 2016) of which all but 3 were directly attributable to the child's cancer and treatment. CNS tumours, with an overall survival probability of 70.5%, accounted for the largest number of deaths (89 deaths, 37.6%, see Figure 3.1.4), up from 32% for the 2000-2009 period. Childhood leukaemias had a five-year survival probability of 88.9% which was higher than overall childhood cancer survival of 83.6%. However, due to the greatest number of childhood cancers being diagnosed in this group, leukaemias nevertheless accounted for one in four of all deaths within the cohort during the study period (59 deaths, 24.9%).

Figure 3.1.4 Cancer-related deaths in New Zealand children first diagnosed with cancer between 2005 and 2014[a]



[a] Follow up to the 31st of December 2016

3.2 Childhood cancer relative survival by age at diagnosis

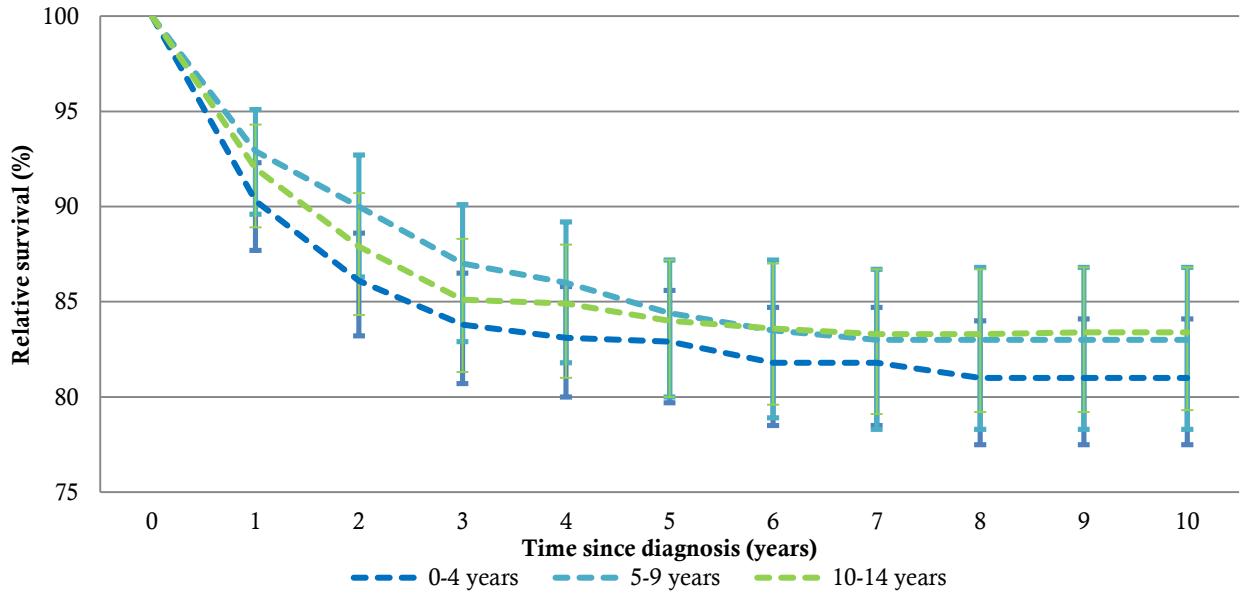
3.2.1 Childhood cancer relative survival, by age group and time since diagnosis

There were no significant differences with respect to relative survival by age group at diagnosis. One-year survival was consistently around 90% and three-year survival ranged between 87.0% for those aged 5-9 years to 83.8% for the 0-4 year age group (see Table 3.2.1 and Figure 3.2.1). There was little further decline in relative survival for any age group between 5 and 10 years of follow-up. Compared with the 2000-2009 period, five-year survival estimates were unchanged for the 0-4 year group (82.9% c.f. 82.8%) but improvements were seen for those aged 5-9 (84.4% c.f. 79.9%) and 10-14 years (84.0 c.f. 78.4%).

Table 3.2.1 Childhood cancer cumulative relative survival by age group and time since diagnosis, New Zealand, 2005-2014

Time since diagnosis (years)	0-4 years			5-9 years			10-14 years		
	n	Cumulative relative survival (95% CI)		n	Cumulative relative survival (95% CI)		n	Cumulative relative survival (95% CI)	
1	648	90.3	(87.7 - 92.3)	349	92.9	(89.6 - 95.1)	412	92.0	(88.9 - 94.3)
2	584	86.1	(83.2 - 88.6)	324	90.0	(86.3 - 92.7)	379	87.9	(84.3 - 90.7)
3	557	83.8	(80.7 - 86.5)	314	87.0	(82.9 - 90.1)	362	85.1	(81.3 - 88.3)
4	489	83.1	(80.0 - 85.8)	270	86.0	(81.8 - 89.2)	319	84.9	(81.0 - 88.0)
5	419	82.9	(79.7 - 85.6)	233	84.4	(80.0 - 87.9)	283	84.0	(80.0 - 87.2)
6	336	81.8	(78.5 - 84.7)	200	83.5	(78.9 - 87.2)	246	83.6	(79.6 - 87.0)
7	270	81.8	(78.5 - 84.7)	167	83.0	(78.3 - 86.7)	214	83.3	(79.1 - 86.7)
8	211	81.0	(77.5 - 84.0)	136	83.0	(78.3 - 86.8)	180	83.3	(79.2 - 86.7)
9	168	81.0	(77.5 - 84.1)	108	83.0	(78.3 - 86.8)	142	83.4	(79.2 - 86.8)
10	129	81.0	(77.5 - 84.1)	77	83.0	(78.3 - 86.8)	96	83.4	(79.3 - 86.8)

Figure 3.2.1 Overall child cancer relative survival by age group and time since diagnosis, New Zealand, 2005-2014



3.2.2 Five-year relative survival by age group and ICCC diagnostic group and selected subgroup

There were no statistically significant differences in survival estimates by age group (see Table 3.2.2). Overall leukaemia survival was almost identical for each age quintile, although AML survival for 0-4 year olds were around 15% lower than survival for school age children. Compared to other age groups, the 5-9 year age group recorded the best survival for CNS tumours (79.5%), malignant bone tumours (85.1%) and soft tissue sarcomas (83.1%).

Table 3.2.2 Five-year relative survival by age group and ICCC diagnostic group and selected subgroup, New Zealand, 2005-2014

		0-4 years			5-9 years			10-14 years		
		Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)	
	All childhood cancers	**648**	**82.9**	**(79.7 - 85.6)**	**349**	**84.4**	**(80.0 - 87.9)**	**412**	**84.0**	**(80.0 - 87.2)**
I.	Leukaemias	246	88.2	(83.4 - 91.7)	131	89.0	(81.7 - 93.5)	99	89.6	(81.3 - 94.4)
I(a)	*Lymphoid leukaemias*	194	94.2	(89.7 - 96.8)	105	90.4	(82.2 - 94.9)	63	91.6	(80.7 - 96.5)
I(b)	*Acute myeloid leukaemias*	34	64.9	(46.4 - 78.4)	17	81.2	(52.1 - 93.6)	25	79.8	(57.9 - 91.2)
II.	Lymphomas	37	94.8	(80.2 - 98.8)	34	94.2	(78.5 - 98.6)	78	96.3	(88.7 - 98.9)
II(a)	*Hodgkin lymphomas*	2	100.1	[a]	10	100.1	[a]	39	100.2	[a]
II(b)	*Non-Hodgkin lymphomas*	8	100.1	[a]	11	81.9	(44.8 - 95.2)	30	96.8	(78.7 - 99.7)
III.	CNS tumours	116	63.5	(53.9 - 71.6)	104	79.5	(70.2 - 86.1)	75	69.0	(57.0 - 78.2)
III(b)	*Astrocytoma*	41	87.9	(73.3 - 94.9)	43	88.4	(74.3 - 95.1)	33	66.7	(47.9 - 80.0)
III(c)	*Intracranial & intraspinal embryonal neoplasms*	26	26.7	(11.7 - 44.4)	27	80.3	(58.7 - 91.4)	14	49.6	(22.3 - 72.1)
IV.	Neuroblastoma	86	76.6	(65.9 - 84.3)	14	64.3	(34.4 - 83.4)	1	[b]	[b]
V.	Retinoblastoma	39	100.2	[a]	1	100.1	[a]	1	100.1	[a]
VI.	Renal tumours	44	97.9	(85.1 - 99.9)	7	85.8	(33.4 - 97.9)	2	100.1	[a]
VII.	Hepatic tumours	13	92.6	(56.8 - 99.2)	4	25.1	(0.9 - 66.6)	1	[b]	[b]
VIII.	Malignant bone tumours	4	50.0	(5.8 - 84.6)	20	85.1	(60.4 - 95.0)	55	78.9	(64.7 - 87.8)
VIII(a)	*Osteosarcomas*	-	-	-	9	89.0	(43.3 - 98.4)	29	74.9	(54.1 - 87.3)
VIII(c)	*Ewing tumours*	3	66.7	(5.4 - 94.6)	8	87.6	(38.7 - 98.2)	21	79.6	(53.8 - 92.0)
IX.	Soft tissue sarcomas	39	68.3	(50.8 - 80.8)	19	83.1	(56.0 - 94.3)	33	66.4	(47.5 - 79.9)
IX(a)	*Rhabdomyosarcomas*	26	67.8	(45.4 - 82.6)	9	100.1	[a]	13	37.8	(13.4 - 62.5)
X.	Germ cell tumours	19	100.5	[a]	6	100.1	[a]	25	96.2	(75.0 - 99.6)
XI.	Other malignant epithelial	5	80.1	(20.4 - 97.0)	8	87.6	(38.7 - 98.1)	40	92.4	(77.9 - 97.6)
XII.	Other and unspecified	-	-	-	1	100.1	[a]	1	[b]	[b]

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.
[b] Five-year relative survival could not be calculated as no cases had a full five years of follow-up.

3.3 Childhood cancer relative survival by sex

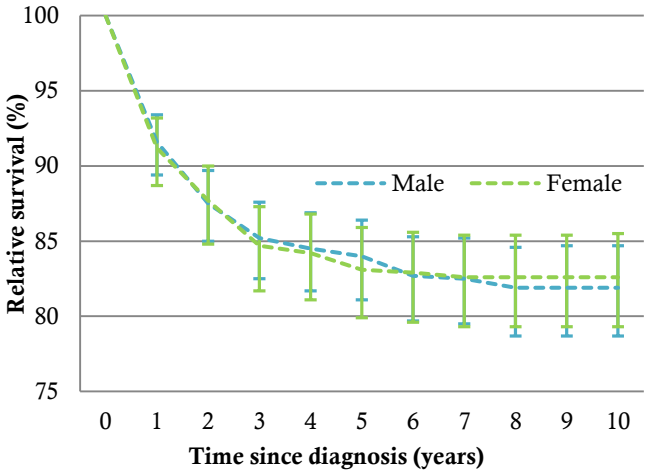
3.3.1 Childhood cancer cumulative relative survival by sex and time since diagnosis

In 10 years of follow up there was less than a single percentage difference in survival estimates for boys and girls (see Table 3.3.1 and Figure 3.3.1).

Table 3.3.1 Childhood cancer cumulative relative survival by sex and time since diagnosis, New Zealand, 2005-2014

Time since diag-nosis (years)	Male			Female		
	n	Cumulative relative survival (95% CI)		n	Cumulative relative survival (95% CI)	
1	765	91.6	(89.4 - 93.4)	644	91.2	(88.7 - 93.2)
2	700	87.5	(85.0 - 89.7)	587	87.7	(84.8 - 90.0)
3	669	85.2	(82.5 - 87.6)	564	84.7	(81.7 - 87.3)
4	590	84.5	(81.7 - 86.9)	488	84.2	(81.1 - 86.8)
5	512	84.0	(81.1 - 86.4)	423	83.1	(79.9 - 85.9)
6	433	82.7	(79.7 - 85.3)	349	82.9	(79.6 - 85.6)
7	359	82.5	(79.5 - 85.2)	292	82.6	(79.3 - 85.4)
8	278	81.9	(78.7 - 84.6)	249	82.6	(79.3 - 85.4)
9	206	81.9	(78.7 - 84.7)	212	82.6	(79.3 - 85.4)
10	149	81.9	(78.7 - 84.7)	153	82.6	(79.3 - 85.5)

Figure 3.3.1 Childhood cancer cumulative relative survival by sex and time since diagnosis, New Zealand, 2005-2014



3.3.2 Five-year relative survival by sex and ICCC diagnostic group and subgroup

Table 3.3.2 shows that there were no statistically significant differences in five-year survival probabilities for males compared to females. One noteworthy (non-significant) survival difference was the poorer survival for females diagnosed with bone tumours (64.9% for females c.f. 89.0% for males), this is due in part to there not being a single death recorded among those males diagnosed with Ewing sarcomas. The opposite was seen in soft tissue sarcoma survival, with female survival (80.0%) higher than for males (62.8%).

Table 3.3.2 Five-year survival by sex and ICCC diagnostic group and subgroup, New Zealand, 2005-2014

	ICCC-3 diagnostic group/subgroup	Male			Female		
		Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)	
	All childhood cancers	**765**	84.0	(81.1 - 86.4)	**644**	83.1	(79.9 - 85.9)
I.	**Leukaemias, myeloproliferative & myelodysplastic diseases**	**271**	**87.8**	**(83.1 - 91.3)**	**205**	**89.9**	**(84.6 - 93.4)**
I(a)	*Lymphoid leukaemias*	209	91.8	(86.9 - 95.0)	153	93.6	(88.0 - 96.7)
I(b)	*Acute myeloid leukaemias*	41	68.3	(51.6 - 80.2)	35	79.8	(62.1 - 89.9)
I(c)	*Chronic myeloproliferative diseases*	5	100.1	[a]	2	50.0	(0.6 - 91.1)
I(d)	*Other myeloproliferative diseases*	8	87.6	(38.8 - 98.3)	9	100.2	[a]
I(e)	*Other & unspecified leukaemias*	8	75.2	(31.6 - 93.3)	6	50.1	(11.1 - 80.6)

Table 3.3.2 (cont.) Five-year survival by sex and ICCC` diagnostic group and subgroup, New Zealand, 2005-2014

	ICCC-3 diagnostic group/subgroup	Male			Female		
		Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)	
II.	**Lymphoma & reticuloendothelial neoplasms**	**96**	**97.0**	**(90.8 - 99.1)**	**53**	**92.6**	**(81.2 - 97.2)**
II(a)	*Hodgkin lymphomas*	28	100.2	[a]	23	100.1	[a]
II(b)	*Non-Hodgkin lymphomas (excl. Burkitt lymphomas)*	32	93.9	(77.4 - 98.6)	17	94.2	(65.1 - 99.3)
II(c)	*Burkitt lymphomas*	15	93.4	(61.3 - 99.1)	7	71.5	(25.8 - 92.0)
II(d)	*Miscellaneous lymphoreticular neoplasms*	20	100.2	[a]	6	83.5	(27.4 - 97.7)
II(e)	*Unspecified lymphomas*	1	100.1	[a]	-	-	-
III.	**Central nervous system & intracranial/intraspinal neoplasms**	**154**	**70.6**	**(62.7 - 77.2)**	**141**	**70.3**	**(61.8 - 77.2)**
III(a)	*Ependymomas & choroid plexus tumours*	13	92.2	(55.5 - 99.1)	16	87.6	(58.7 - 96.9)
III(b)	*Astrocytomas*	54	89.0	(77.0 - 95.0)	63	76.2	(63.6 - 85.0)
III(c)	*Intracranial & intraspinal embryonal tumours*	48	49.6	(34.7 - 62.8)	19	61.1	(34.7 - 79.5)
III(d)	*Other gliomas*	18	55.7	(30.6 - 74.9)	23	38.7	(19.5 - 57.8)
III(e)	*Other specified intracranial & intraspinal neoplasms*	15	86.8	(56.5 - 96.7)	17	92.7	(58.0 - 99.0)
III(f)	*Unspecified intracranial & intraspinal neoplasms*	6	33.5	(4.6 - 67.8)	-	-	-
IV.	**Neuroblastoma & other peripheral nervous cell tumours**	**49**	**73.4**	**(58.5 - 83.8)**	**53**	**73.3**	**(59.0 - 83.3)**
IV(a)	*Neuroblastoma & ganglioneuroblastoma*	49	73.4	(58.5 - 83.8)	53	73.3	(59.0 - 83.3)
IV(b)	*Other peripheral nervous cell tumours*	-	-	-	-	-	-
V.	**Retinoblastoma**	**21**	**100.2**	**[a]**	**20**	**100.2**	**[a]**
VI.	**Renal tumours**	**19**	**94.8**	**(67.6 - 99.5)**	**34**	**97.2**	**(81.0 - 99.7)**
VI(a)	*Nephroblastoma & other non-epithelial renal tumours*	19	94.8	(67.6 - 99.5)	34	97.2	(81.0 - 99.7)
VI(b)	*Renal carcinomas*	-	-	-	-	-	-
VII.	**Hepatic tumours**	**9**	**78.0**	**(36.6 - 94.2)**	**9**	**66.8**	**(28.2 - 88.0)**
VII(a)	*Hepatoblastoma*	6	83.7	(27.4 - 97.9)	6	83.5	(27.4 - 97.7)
VII(b)	*Hepatic carcinomas*	3	66.7	(5.4 - 94.6)	3	33.4	(0.9 - 77.6)
VIII.	**Malignant bone tumours**	**46**	**89.0**	**(75.3 - 95.4)**	**33**	**64.9**	**(45.2 - 79.1)**
VIII(a)	*Osteosarcomas*	24	82.9	(60.4 - 93.3)	14	70.2	(38.5 - 87.7)
VIII(b)	*Chondrosarcomas*	1	100.3	[a]	-	-	-
VIII(c)	*Ewing tumours & related bone sarcomas*	17	100.1	[a]	15	59.7	(31.3 - 79.6)
VIII(d)	*Other specified malignant bone tumours*	4	75.1	(12.8 - 96.2)	4	75.1	(12.8 – 96.1)
VIII(e)	*Unspecified malignant bone tumours*	-	-	-	-	-	-
IX.	**Soft tissue and other extraosseous sarcomas**	**50**	**62.8**	**(47.3 - 74.9)**	**41**	**80.0**	**(63.7 - 89.5)**
IX(a)	*Rhabdomyosarcomas*	29	65.1	(44.7 - 79.6)	19	67.7	(41.6 - 84.1)
IX(b)	*Fibrosarcomas & other fibrous neoplasms*	3	67.0	(5.4 - 95.0)	3	100.1	[a]
IX(c)	*Kaposi sarcomas*	-	-	-	-	-	-
IX(d)	*Other specified soft tissue sarcomas*	12	44.5	(14.5 - 71.3)	16	87.7	(58.7 - 96.9)
IX(e)	*Unspecified soft tissue sarcomas*	6	83.6	(27.4 - 97.7)	3	100.1	[a]
X.	**Germ cell tumours, trophoblastic tumours & neoplasms of gonads**	**26**	**96.5**	**(76.0 - 99.8)**	**24**	**100.2**	**[a]**
X(a)	*Intracranial & intraspinal germ cell tumours*	10	90.1	(47.4 - 98.7)	10	100.1	[a]
X(b)	*Malignant extracranial & extragonadal germ cell tumours*	7	100.7	[a]	7	100.1	[a]
X(c)	*Malignant gonadal germ cell tumours*	9	100.4	[a]	6	100.1	[a]
X(d)	*Gonadal carcinomas*	-	-	-	-	-	-
X(e)	*Other & unspecified malignant gonadal tumours*	-	-	-	1	[b]	[b]
XI.	**Other malignant epithelial neoplasms & malignant melanomas**	**23**	**95.8**	**(73.1 - 99.5)**	**30**	**86.1**	**(66.9 - 94.6)**
XI(a)	*Adrenocortical carcinomas*	2	50.1	(0.6 - 91.1)	1	[b]	[b]
XI(b)	*Thyroid carcinomas*	3	100.2	[a]	4	100.1	[a]
XI(c)	*Nasopharyngeal carcinomas*	1	100.3	[a]	-	-	-
XI(d)	*Melanomas*	10	100.2	[a]	11	91.0	(50.9 - 98.8)
XI(e)	*Skin carcinomas*	-	-		-	-	
XI(f)	*Other & unspecified carcinomas*	7	100.2	[a]	14	76.1	(42.3 - 91.7)
XII.	**Other & unspecified malignant neoplasms**	**1**	**100.1**	**[a]**	**1**	[b]	[b]

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.
[b] Five-year relative survival could not be calculated as no cases had a full five years of follow-up.

3.4 Childhood cancer relative survival by prioritised ethnicity

The following section reports child cancer survival by prioritised ethnicity; Maori, Pacific Peoples, and all others (labelled as 'non-Maori/non-Pacific Peoples'). As noted earlier, any between-group differences must be interpreted with caution as often there were few cases diagnosed within the time period and any differences may be within the bounds of chance variation. Also, any differences in overall child cancer survival by ethnicity may be explained by a number of other factors such as a higher incidence of certain cancers, which may have a more favourable/unfavourable prognosis, within a particular ethnic group.

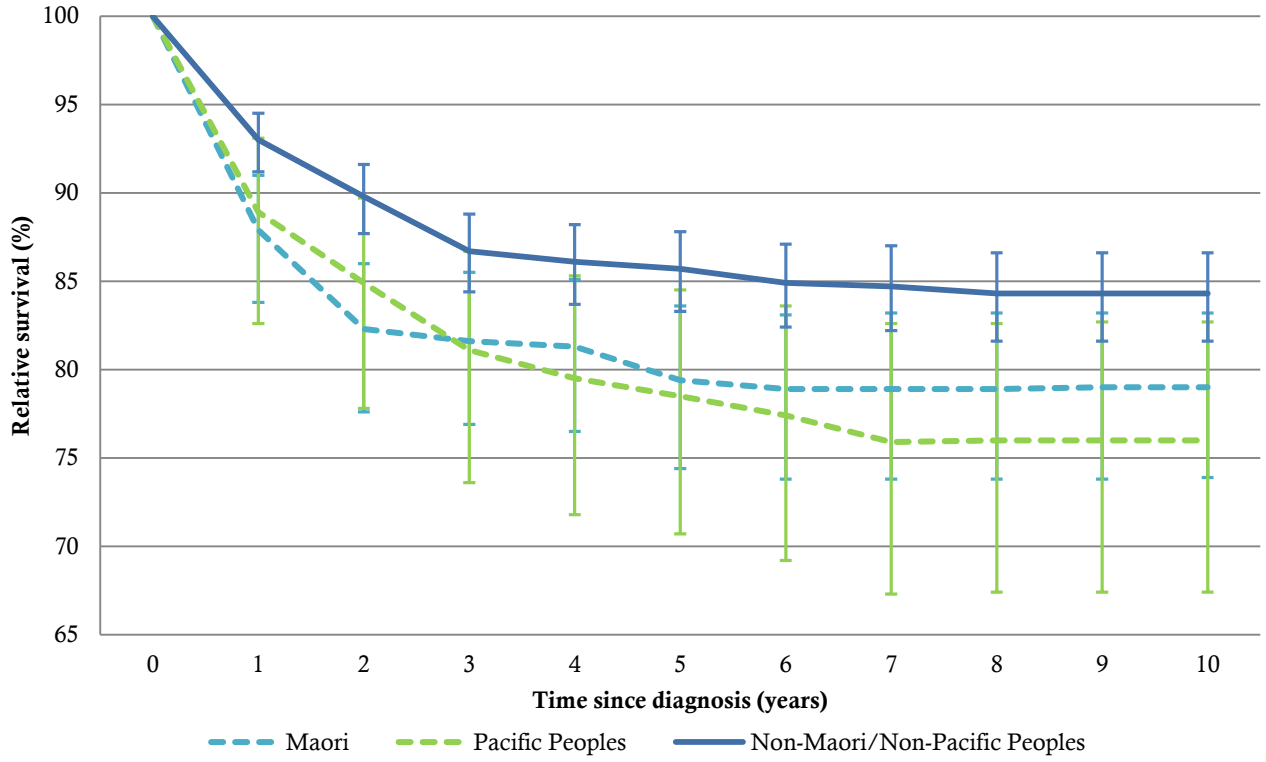
3.4.1 Childhood cancer cumulative relative survival, by prioritised ethnicity and time since diagnosis

In terms of overall cancer survival, there were no statistically significant differences according to ethnicity at any point of follow-up (see Table 3.4.1 and Figure 3.4.1). However, there is a noteworthy survival gap of 5% or more for Maori and Pacific Peoples compared with non-Maori/non-Pacific Peoples for most follow up periods. Five-year survival ranged from 78.5% for Pacific Peoples to 85.7% for non-Maori/non-Pacific children. Compared to the 2000-2009 period, survival improvements were more evident for non-Maori/non-Pacific Peoples where five-year survival increased by 4.0% to 85.7% than for Maori (increasing by 2.5% to 79.4%). With the exception of 1 year of follow-up, survival estimates for Pacific Peoples were lower for the 2005-2014 period than for the earlier 2000-2009 period with five-year survival dropping by 2.9% to 78.5%.

Table 3.4.1 Childhood cancer cumulative relative survival, by prioritised ethnicity and time since diagnosis, New Zealand, 2005-2014

Time since diagnosis (years)	Maori			Pacific Peoples			Non-Maori/non-Pacific Peoples		
	n	Cumulative relative survival (95% CI)		n	Cumulative relative survival (95% CI)		n	Cumulative relative survival (95% CI)	
1	320	87.9	(83.8 - 91.0)	144	88.9	(82.6 - 93.1)	945	93.0	(91.2 - 94.5)
2	281	82.3	(77.6 - 86.0)	128	84.8	(77.8 - 89.7)	878	89.8	(87.7 - 91.6)
3	263	81.6	(76.9 - 85.5)	122	81.1	(73.6 - 86.7)	848	86.7	(84.4 - 88.8)
4	234	81.3	(76.5 - 85.1)	103	79.5	(71.8 - 85.3)	741	86.1	(83.7 - 88.2)
5	194	79.4	(74.4 - 83.6)	90	78.5	(70.7 - 84.5)	651	85.7	(83.3 - 87.8)
6	154	78.9	(73.8 - 83.1)	76	77.4	(69.2 - 83.6)	552	84.9	(82.4 - 87.1)
7	129	78.9	(73.8 - 83.2)	57	75.9	(67.3 - 82.6)	465	84.7	(82.2 - 87.0)
8	104	78.9	(73.8 - 83.2)	48	76.0	(67.4 - 82.6)	375	84.3	(81.6 - 86.6)
9	86	79.0	(73.8 - 83.2)	36	76.0	(67.4 - 82.7)	296	84.3	(81.6 - 86.6)
10	55	79.0	(73.9 - 83.2)	27	76.0	(67.4 - 82.7)	220	84.3	(81.6 - 86.6)

Figure 3.4.1 Childhood cancer relative survival by prioritised ethnicity and time since diagnosis, New Zealand, 2005-2014



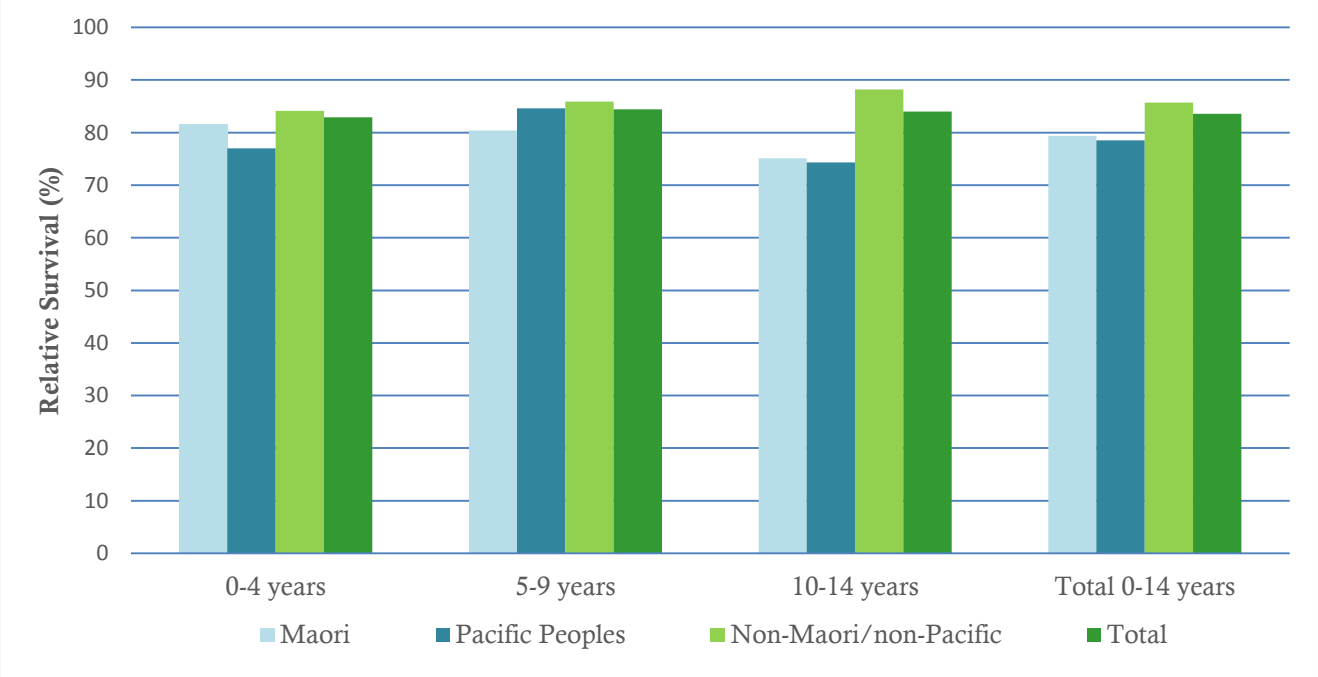
3.4.2 Five-year relative survival for all childhood cancers by age group and prioritised ethnicity

The largest five-year survival differences according to prioritised ethnicity were seen for those aged 10-14 years at diagnosis; survival for Maori (75.1%) was significantly lower than for non-Maori/non-Pacific Peoples (88.2%), while survival for Pacific 10-14 year olds was even lower at 74.3% (see Table 3.4.2). Although survival for Maori 10-14 year olds had actually improved slightly from the 2000-2009 period (+0.9%) it was only in the non-Maori/non-Pacific group where dramatic survival gains were evident (+8.9%, from 79.3% to 88.2%).

Table 3.4.2 Five-year relative survival for all childhood cancers by age group and prioritised ethnicity, New Zealand, 2005-2014

	0-4 years			5-9 years			10-14 years			Total 0-14 years		
	Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)	
Maori	146	81.5	(74.1 - 87.0)	88	80.4	(69.6 - 87.7)	86	75.1	(64.3 - 83.1)	320	79.4	(74.4 - 83.6)
Pacific Peoples	53	77.0	(62.8 - 86.3)	46	84.6	(70.3 - 92.4)	45	74.3	(58.0 - 85.0)	144	78.5	(70.7 - 84.5)
Non-Maori/ non-Pacific Peoples	449	84.1	(80.3 - 87.2)	215	85.9	(80.3 - 90.0)	281	88.2	(83.7 - 91.5)	945	85.7	(83.3 - 87.8)
Total	**648**	**82.9**	**(79.7 - 85.6)**	**349**	**84.4**	**(80.0 - 87.2)**	**412**	**84.0**	**(80.0 - 87.2)**	**1409**	**83.6**	**(81.5 - 85.5)**

Figure 3.4.2 Five-year relative survival for all childhood cancers by age group and prioritised ethnicity, New Zealand, 2005-2015



3.4.3 Five-year relative survival by prioritised ethnicity and ICCC diagnostic groups and selected subgroups

As modern treatment of ALL is complex, prolonged, and requires open access to health care, we have previously used it as an indicator disease. For the 2000-2009 period, five-year relative survival for ALL across the three ethnic groups was almost identical; 89.8% for Maori, 88.0% for Pacific Peoples, and 89.4% for 'All Others'. The closeness in these survival figures was taken as an indication that New Zealand is achieving equitable outcomes for children diagnosed with cancer regardless of ethnicity. For the 2005-2014 period, leukaemia survival for all three prioritised ethnic groups improved further to range from 90.9% for Pacific Peoples to 93.2% for non-Maori/non-Pacific children. Maori had the poorest survival for bone and soft tissue sarcomas and none of the seven Maori children diagnosed with 'other gliomas' survived.

Table 3.4.3 Childhood cancer five-year relative survival by prioritised ethnicity, New Zealand, 2005-2014

ICCC-3 diagnostic group/ subgroup		Maori			Pacific Peoples			Non-Maori/non-Pacific Peoples		
		Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)	
	All childhood cancers	**320**	**79.4**	**(74.3 - 83.6)**	**144**	**78.5**	**(70.7 - 84.5)**	**945**	**85.7**	**(83.3 - 87.8)**
I.	Leukaemias	114	84.4	(75.8 - 90.1)	50	85.7	(72.1 - 93.0)	312	90.7	(86.8 - 93.5)
I(a)	*Lymphoid leukaemias*	78	91.4	(81.4 - 96.1)	35	90.9	(74.1 - 97.0)	249	93.2	(89.1 - 95.8)
I(b)	*Acute myeloid leukaemias*	28	67.7	(47.0 - 81.8)	8	62.6	(23.0 - 86.2)	40	79.8	(63.6 - 89.5)
II.	Lymphomas	28	89.4	(70.5 - 96.6)	15	93.4	(61.3 - 99.1)	106	97.3	(91.6 - 99.2)
II(a)	*Hodgkin lymphomas*	8	100.1	[a]	4	-	[b]	39	100.2	[a]
II(b)	*Non-Hodgkin lymphomas (excl. Burkitt lymphoma)*	8	87.6	(38.8 - 98.3)	4	100.1	[a]	37	94.7	(80.2 - 98.8)
III.	CNS Tumours	78	71.6	(60.0 - 80.3)	31	52.8	(33.1 - 69.2)	186	72.9	(65.8 - 78.8)
III(b)	*Astrocytomas*	29	86.3	(67.4 - 94.7)	10	59.3	(24.3 - 82.5)	78	83.4	(73.1 - 90.1)
III(c)	*Intracranial & intraspinal embryonal tumours*	26	64.5	(42.6 - 79.9)	8	37.6	(8.7 - 67.5)	33	48.4	(30.7 - 64.0)
III(d)	*Other gliomas*	7	0.0	[a]	7	57.2	(17.2 - 83.8)	27	54.9	(34.3 - 71.5)
IV.	Neuroblastoma	22	67.9	(44.0 - 83.4)	4	75.3	(12.8 - 96.4)	76	74.8	(63.3 - 83.2)
V.	Retinoblastoma	8	100.2	[a]	7	100.2	[a]	26	100.2	[a]
VI.	Renal tumours	5	100.2	[a]	5	100.3	[a]	43	95.5	(82.7 - 99.0)
VII.	Hepatic tumours	5	80.3	(20.5 - 97.2)	2	-	[b]	11	82.0	(44.9 - 95.4)
VIII.	Malignant bone tumours	17	66.3	(35.1 - 85.2)	14	78.7	(47.3 - 92.7)	48	83.1	(69.0 - 91.3)
VIII(a)	*Osteosarcomas*	7	71.5	(25.8 - 92.1)	8	75.1	(31.5 - 93.2)	23	82.1	(58.9 - 93.0)
VIII(c)	*Ewing tumour*	9	63.7	(23.5 - 86.9)	5	100.1	[a]	18	83.2	(56.4 - 94.3)
IX.	Soft tissue sarcomas	14	46.8	(18.7 - 70.9)	9	89.0	(43.3 - 98.4)	68	72.8	(60.2 - 82.0)
IX(a)	*Rhabdomyosarcomas*	7	57.2	(17.2 - 83.8)	5	80.1	(20.4 - 97.0)	36	65.9	(47.6 - 79.1)
X.	Germ cell tumours	18	94.7	(66.8 - 99.5)	4	100.1	[a]	28	100.3	[a]
XI.	Other malignant epithelial	10	90.1	(47.4 - 98.7)	3	66.8	(5.4 - 94.7)	40	92.4	(78.0 - 97.6)
XII.	Other & unspecified malignant neoplasms	1	100.1	[a]	-	-	-	1	-	[b]

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.
[b] Five-year relative survival could not be calculated as no cases had a full five years of follow-up.

3.5 New Zealand childhood cancer survival over time

The following section compares New Zealand child cancer five-year survival from 2005-2014 with survival for those children diagnosed from 1961-1970, 1990-1993, and 2000-2009. Due to the methodological differences between studies and New Zealand's relatively small population, caution should be taken when making comparisons between the survival probabilities reported for each time period. Confidence intervals and survival probabilities by diagnostic subgroup were not available for the 1961-1970 cohort.

Overall child cancer survival in New Zealand has improved from 28% in 1961-1970, to 66% in 1990-1993, to 81% in 2000-2009 to 84% in 2005-2014 (see Table 3.5). Children diagnosed with leukaemia in the 1961 to 1970 period had only a 6% chance of surviving five years, the poorest survival of any diagnostic group. However, leukaemia survival has shown a dramatic improvement since the 1960s. The overall survival for childhood leukaemia in 2005-2014 was 89%, a further improvement on the 85% achieved in 2000-2009.

Five-year survival estimates for children diagnosed with neuroblastoma have increased by 7% between 2000-2009 and 2005-2014, likely reflecting the advancements made in treating high-risk neuroblastoma patients during this time. 5-year survival for bone tumours is now back to what was reported in 1990-1993, having shown a 17% drop in 2000-2009. In contrast to most diagnostic groups, five-year survival for CNS tumours have remained unchanged from their 2000-2009 estimate of 71% while survival for soft tissue sarcomas has dropped slightly from 73% to 71%.

Table 3.5 Five-year survival for childhood cancer in New Zealand over time

ICCC-3 diagnostic group/subgroup		1961-1970[11]		1990-1993[12]			2000-2009			2005-2014		
		Total cases	Five-year observed survival	Total cases	Five-year cause-specific survival (95% CI)		Total cases	Five-year relative survival (95% CI)		Total cases	Five-year relative survival (95% CI)	
	All childhood cancers	**1002**	**28**	**409**	**66**	**(62 - 71)**	**1321**	**81**	**(78 - 83)**	**1409**	**84**	**(82 - 86)**
I.	Leukaemias	345	6	144	65	(57 - 73)	455	85	(81 - 88)	476	89	(85 - 91)
I(a)	*Lymphoid leukaemias*	-	-	*111*	*70*	*(62 - 79)*	*352*	*89*	*(85 - 92)*	*362*	*93*	*(89 - 95)*
I(b)	*Acute myeloid leukaemias*	-	-	*26*	*50*	*(31 - 69)*	*78*	*69*	*(57 - 79)*	*76*	*74*	*(62 - 82)*
II.	Lymphomas	82	33	37	89	(79 - 99)	114	93	(86 - 97)	149	96	(91 - 98)
II(a)	*Hodgkin lymphomas*	-	-	*14*	*93*	*(79 - 100)*	*36*	*97*	*(82 - 100)*	*51*	*100*	[a]
II(b)	*Non-Hodgkin lymphomas*	-	-	*23*[b]	*87*	*(73 - 100)*	*58*	*95*	*(85 - 98)*	*49*	*94*	*(82 – 98)*
III.	CNS tumours	217	33[c]	80	49[c]	(38 - 60)	283	71	(65 - 76)	295	71	(65 - 75)
IV.	Neuroblastoma	36	19	20	35	(14 - 56)	87	66	(55 - 76)	102	73	(63 - 81)
V.	Retinoblastoma	[d]	[d]	12	100	(74 - 100)	39	100	[a]	41	100	[a]
VI.	Renal tumours	66	33	25	80	(64 - 96)	61	97	(87 - 99)	53	96	(86 - 99)
VII.	Hepatic tumours	[d]	[d]	7	86	(60 - 100)	13	69	(37 - 87)	18	72	(46 - 88)
VIII.	Malignant bone tumours	54	28	20	79	(61 - 97)	72	67	(54 - 77)	79	79	(68 - 87)
VIII(a)	*Osteosarcomas*	-	-	*12*	*91*	*(74 - 100)*	*37*	*67*	*(49 - 80)*	*38*	*78*	*(61 - 89)*
VIII(c)	*Ewing tumours*	-	-	*7*	*57*	*(20 - 94)*	*28*	*61*	*(38 - 78)*	*32*	*81*	*(61 - 91)*
IX.	Soft tissue sarcomas	[d]	[d]	23	45	(25 - 66)	94	73	(62 - 81)	91	71	(60 - 79)
IX(a)	*Rhabdomyosarcomas*	-	-	*12*	*42*	*(14 - 70)*	*50*	*71*	*(55 - 82)*	*48*	*66*	*(51 - 78)*
X.	Germ cell tumours	[d]	[d]	23	83	(67 - 98)	57	97	(87 - 99)	50	98	(87 - 100)
XI.	Other malignant epithelial	[d]	[d]	16	81	(62 - 100)	42	85	(69 - 93)	53	90	(78 - 96)

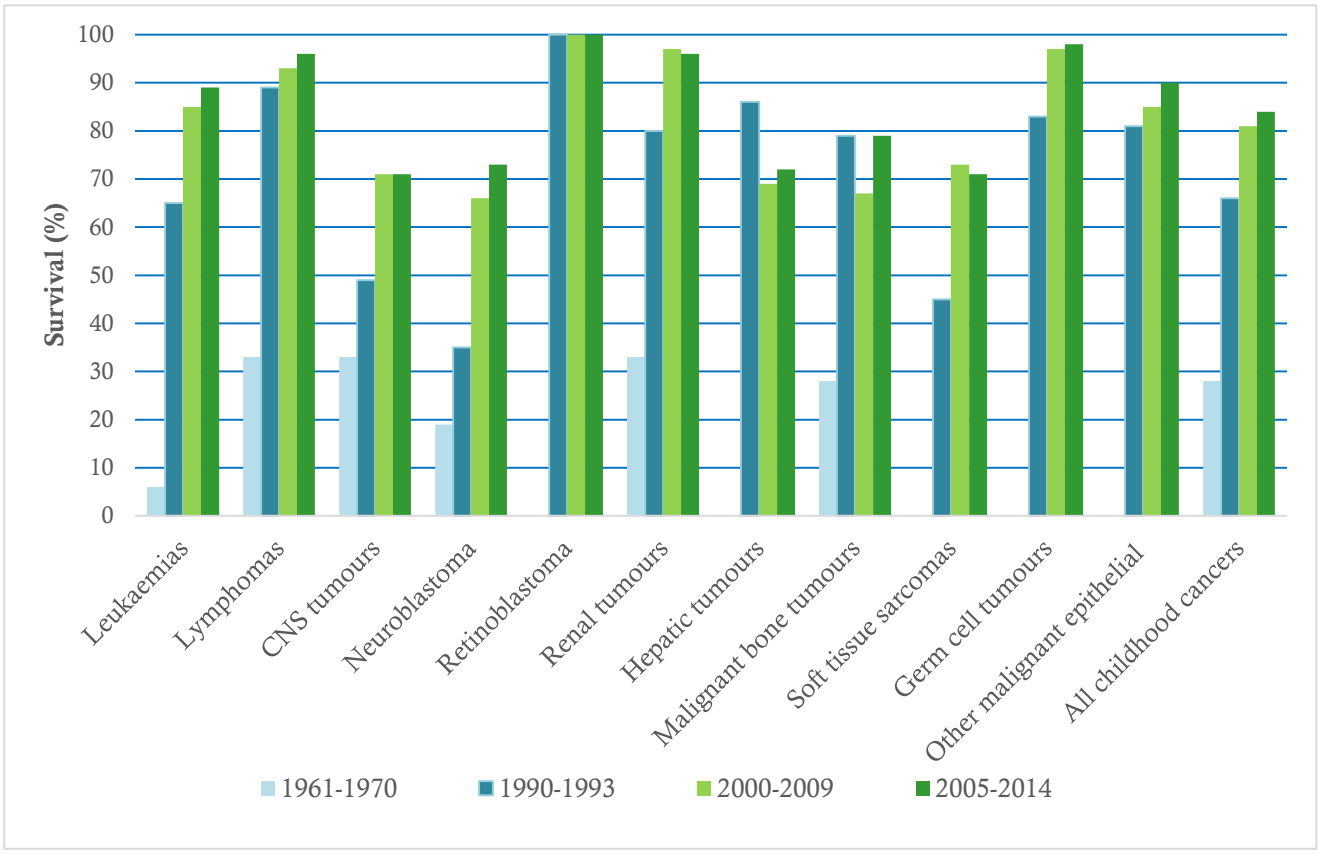
[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.

[b] Also includes Burkitt lymphoma.

[c] Excludes non-malignant CNS tumours.

[d] For the 1961-1970 period, survival for diagnostic groups V, VII, IX, X and XI were combined under 'other sites'. Five-year observed survival for the 202 children diagnosed with one of these five tumour groups was 59%. Diagnostic group XII: 'other and unspecified' was not reported in the 1961-1970 or 1990-1993 studies.

Figure 3.5 Changes in New Zealand childhood cancer five-year survival over time, by diagnostic group



3.6 International comparisons of childhood cancer survival

The following section places New Zealand's child cancer survival in the context of what is being achieved internationally. Childhood cancer survival is reported from Australia,[14] Switzerland,[15] Canada,[16] Germany,[17] and the United States (SEER).[18] These developed countries were selected as they had published childhood cancer survival probabilities by ICCC-3 for 0-14 year olds for a comparable time period. However, the data must be interpreted cautiously as any differences in survival estimates may be influenced by variations in registry inclusion criteria and survival calculation methodologies used. Please refer to Section 2.5 for further details about the data which is reported in this section.

Table 3.6 shows that New Zealand's overall childhood cancer five-year relative survival (84%) is comparable with survival rates published by countries recognised as world leaders in children's cancer research and treatment. New Zealand ranked highly for leukaemia, lymphoma, germ cell tumour and renal tumour survival. At 73%, New Zealand's neuroblastoma survival estimate had improved since 2000-2009 but remained below that reported by Switzerland (80%), Germany (79%), and the United States (79%). Switzerland's figures demonstrated that further survival gains are potentially achievable for New Zealand children diagnosed with bone tumours, soft tissue sarcomas and CNS tumours.

Table 3.6 International comparisons of five-year survival for childhood cancer

ICCC-3 diagnostic group/subgroup		NZ 2005-2014	Australia 2004-2013[14]	Switzerland 2004-2013[15]	Canada 2004-2008[16]	Germany 2005-2014[17]	US (SEER) 2007-2013[18]
		relative survival	relative survival	observed survival	observed survival	observed survival	relative survival
	Overall childhood cancers	**84**	**84**	**88**	**83**	**85**	**83**
I.	Leukaemias	89	88	88	88	89	86[a]
II.	Lymphomas	96	94	95	92	94	93
III.	CNS tumours	71	74	74	74	77	73[b]
IV.	Neuroblastoma & other peripheral nervous cell tumours	73	74	80	77	79	79
V.	Retinoblastoma	100	98	94	94	97	95
VI.	Renal tumours	96	90	94	84	93	92
VII.	Hepatic tumours	72	72	80	68	77	79
VIII.	Malignant bone tumours	79	79	85	70	[c]	74
IX.	Soft tissue sarcomas	71	77	80	72	73	75
X.	Germ cell tumours	98	90	94	91	94	92
XI.	Other malignant epithelial	90	92	[c]	94	[c]	93

[a] Excludes myelodysplastic syndromes.
[b] Excludes Benign CNS tumours
[c] Not reported

4 Childhood Cancer Survival by Diagnostic Group

The following section describes cancer survival for each of the ICCC-3 diagnostic subgroups in turn. Each subsection begins with a description of the diagnostic group, including the defining characteristics of the group, the initial presenting symptoms, the conditions associated with increased risk, and the typical course of treatment. For some diagnostic groups and subgroups there were very few cases recorded, and in such cases the true survival cannot be reliably estimated; this is reflected in the wide 95% confidence intervals which are reported alongside. In such cases, any between-group differences in survival or any differences in comparison to other published data should be interpreted extremely cautiously. It should also be noted that confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.

4.1 Leukaemias, myeloproliferative diseases, and myelodysplastic diseases

Leukaemias arise from malignant transformation of haematopoietic stem cells in the bone marrow. Leukaemic blasts cause clinical symptoms by impairing normal bone marrow function and by dissemination through the blood into distant sites. There are two main leukaemia cell types; lymphoblastic leukaemias are derived from lymphoid precursor cells and myeloid leukaemias are derived from myeloid precursor cells. Leukaemias are further classified as either acute or chronic. The majority of childhood leukaemias are acute, with acute lymphoblastic leukaemia (ALL) accounting for about 80% of all childhood leukaemias, acute myeloid leukaemia (AML) accounting for about 15%, and the remainder being various chronic leukaemias and other myeloproliferative diseases. The aetiology of leukaemia remains unclear; some cases are familial or are associated with other genetic diseases but 95% of cases are sporadic with no predisposing condition.

The prognosis and treatment of childhood acute leukaemia depends on the leukaemia type, the age at diagnosis, the presence or absence of involvement of the central nervous system or testes, and the presence of specific cytomolecular genetic features. All acute leukaemias require treatment with multi-agent chemotherapy, some patients may be treated with radiotherapy, and a smaller number will undergo a bone marrow transplant. ALL treatment lasts approximately 26 months for girls and 38 months for boys, with differing intensity of treatment depending on the leukaemia subtype, the patient's age and response to initial therapy. AML is treated with an intense course of chemotherapy over six months, with much of that time spent in hospital. New Zealand's two children's cancer treatment centres are members of the Children's Oncology Group (COG), a collaborative clinical trial group that runs clinical trials across the USA, Canada, Australia and New Zealand. Where possible, children diagnosed with ALL and AML are treated on one of the COG leukaemia clinical trials.

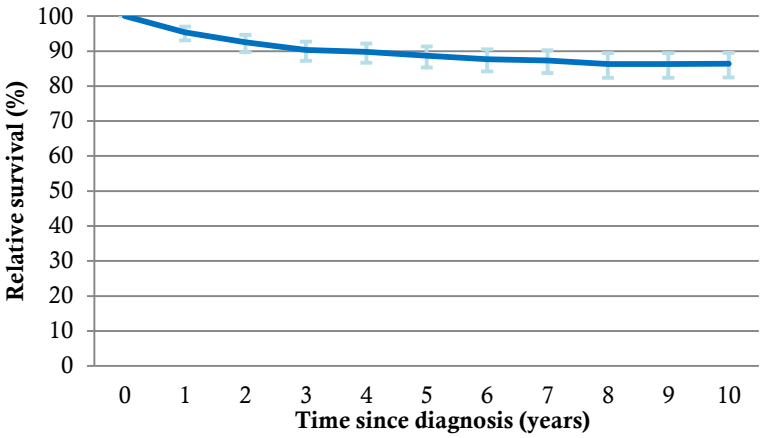
4.1.1 Childhood leukaemias cumulative relative survival by time since diagnosis

Although not yet statistically significant, survival probabilities for children diagnosed with leukaemia between 2005 and 2014 were consistently above the survival reported for the 2000-2009 period. Five-year survival was at 88.7%, an increase of 3.7% while the 10-year survival increased by 5.8% to 86.4%.

Table 4.1.1 Childhood leukaemias cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	476	95.4	(93.1 - 97.0)
2	454	92.5	(89.7 - 94.6)
3	440	90.3	(87.2 - 92.7)
4	381	89.8	(86.7 - 92.2)
5	338	88.7	(85.3 - 91.3)
6	282	87.7	(84.2 - 90.5)
7	237	87.3	(83.7 - 90.2)
8	193	86.3	(82.4 - 89.4)
9	151	86.3	(82.4 - 89.4)
10	102	86.4	(82.5 - 89.5)

Figure 4.1.1 Childhood leukaemias cumulative relative survival, New Zealand, 2005-2014



4.1.2 Leukaemia survival by sex, age group, and prioritised ethnicity

There were no significant differences in five-year relative survival by sex, age group or prioritised ethnicity. However, five-year relative survival for ALL (92.6%) was significantly higher than for AML (73.5%), (see Table 4.1.3).

Table 4.1.2 Childhood leukaemias survival by sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total leukaemias	**476**	**100.0**	**88.7**	**(85.3 - 91.3)**
Diagnostic subgroup				
I(a) Lymphoid leukaemias	362	76.1	92.6	(89.2 - 95.0)
I(b) Acute myeloid leukaemias	76	16.0	73.5	(62.0 - 82.1)
I(c) Chronic myeloproliferative diseases	7	1.5	85.8	(33.4 - 98.0)
I(d) Other myeloproliferative diseases	17	3.6	94.3	(65.1 - 99.3)
I(e) Other & unspecified leukaemia	14	2.9	64.4	(34.4 - 83.5)
Sex				
Male	271	56.9	87.8	(83.1 - 91.3)
Female	205	43.1	89.9	(84.6 - 93.4)
Age group				
0-4 years	246	51.7	88.2	(83.4 - 91.7)
5-9 years	131	27.5	89.0	(81.7 - 93.5)
10-14 years	99	20.8	89.6	(81.3 - 94.4)
Prioritised ethnicity				
Maori	114	23.9	84.4	(75.8 - 90.1)
Pacific Peoples	50	10.5	85.7	(72.1 - 93.0)
Non-Maori/non-Pacific Peoples	312	65.5	90.7	(86.8 - 93.5)

4.1.3 ALL and AML survival by sex, age group, and prioritised ethnicity

Between 2000-2009 and 2005-2014 five-year survival for ALL improved by 3.2% and survival for AML improved by 4.3%. Table 4.1.3 shows that there were no significant differences in outcomes for either of the two main childhood leukaemia groups according to sex, age group or ethnicity. Five-year survival probabilities for ALL were above 90% for all three prioritised ethnic groups; 91.4% for Maori, 90.9% for Pacific Peoples, and 93.2% for non-Maori/non-Pacific Peoples.

Table 4.1.3 Childhood ALL and AML five-year relative survival by sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Acute lymphoblastic leukaemia				Acute myeloid leukaemias			
	Cases	%	Five-year relative survival (95% CI)		Cases	%	Five-year relative survival (95% CI)	
Total cases	**362**	**100**	**92.6**	**(89.2 - 95.0)**	**76**	**100**	**73.5**	**(62.0 - 82.1)**
Sex								
Male	209	57.7	91.8	(86.9 - 95.0)	41	53.9	68.3	(51.6 - 80.2)
Female	153	42.3	93.6	(88.0 - 96.7)	35	46.1	79.8	(62.1 - 89.9)
Age group								
0-4 years	194	53.6	94.2	(89.7 - 96.8)	34	44.7	64.9	(46.4 - 78.4)
5-9 years	105	29.0	90.4	(82.2 - 94.9)	17	22.4	81.2	(52.1 - 93.6)
10-14 years	63	17.4	91.6	(80.7 - 96.5)	25	32.9	79.8	(57.9 - 91.2)
Prioritised ethnicity								
Maori	78	21.5	91.4	(81.4 - 96.1)	28	36.8	67.7	(47.0 - 81.8)
Pacific Peoples	35	9.7	90.9	(74.1 - 97.0)	8	10.5	62.6	(23.0 - 86.2)
Non-Maori/non-Pacific Peoples	249	68.8	93.2	(89.1 - 95.8)	40	52.6	79.8	(63.6 - 89.5)

4.2 Lymphomas and reticuloendothelial neoplasms

Lymphomas arise from the malignant transformation of primitive lymphoid stem cells in the developing lymphatic system. They spread to involve adjacent and distant lymph nodes, and may involve other locations such as the spleen, bone marrow, bones, and brain. Lymphomas are divided into two distinct categories, Hodgkin lymphomas (HL) and non-Hodgkin lymphomas (NHL), and, like leukaemias, may be acute or chronic. In children and young people, most lymphomas are acute and high grade, with chronic or low grade lymphomas being more common in older adults. The diagnostic group also includes lymphoreticular neoplasms such as Langerhans cell histiocytosis (LCH).

Hodgkin lymphomas arise in small proportion of lymphoid cells within a lymph node and tend to spread to adjacent lymph nodes and nodal regions. Common symptoms at diagnosis include progressive painless lymph node swelling, fever, weight loss and lethargy.

The non-Hodgkin lymphomas are a heterogeneous group of diseases. In children, most NHLs are acute lymphoblastic lymphoma and have a clinical behaviour similar to acute lymphoblastic leukaemia. The most common NHLs in children are T-cell lymphoblastic lymphoma and Burkitt lymphoma. T-cell lymphoblastic lymphoma is treated in a similar manner to ALL, while Burkitt lymphoma is treated with a very intense course of multi-agent chemotherapy of varying duration depending on disease status. Most children in New Zealand who are diagnosed with lymphoma will be treated according to a clinical trial or according to a disease-specific clinical protocol.

In LCH, too many Langerhans cells are produced and build up where they can form tumours. LCH can appear as a single lesion or can be multisystem, affecting bones and organs such as the liver, lung, brain and skin. It is more commonly diagnosed in boys and children under the age of 5. Treatment ranges from observation to systemic chemotherapy depending on the extent of disease and involvement of "high risk" organs. There have been differing opinions among experts on whether LCH is best classified as an immune dysfunction or as a cancer but for many years in New Zealand LCH cases have been routinely referred to paediatric oncology centres. In the revised ICD-O-3,[9] which the NZCCR adopted from 1/1/2010, LCH was reclassified from a 'tumour of uncertain behaviour' to a malignancy and it now is included within the diagnostic subgroup 'II(d): miscellaneous lymphoreticular neoplasms'. Previously, this diagnostic subgroup was rarely used, with only a single case registered in the 2000-2009 period.

4.2.1 Childhood lymphomas cumulative relative survival by time since diagnosis

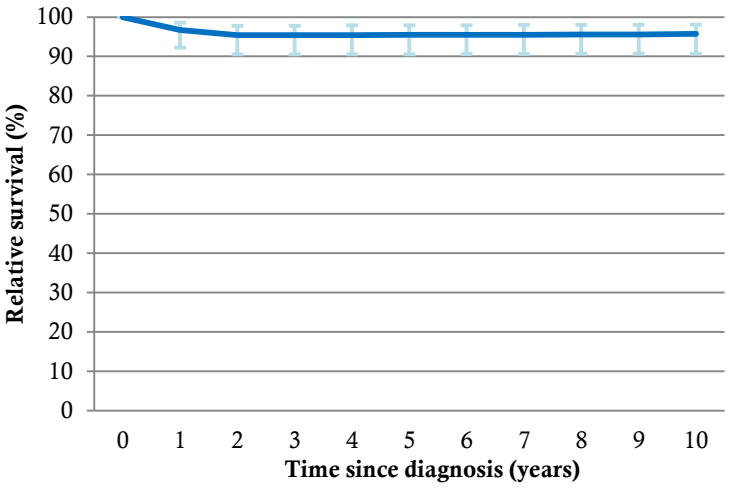
Five-year relative survival for lymphomas (95.5%) was significantly higher than child cancer survival overall (83.6%). Within the cohort, five deaths were recorded during the first year of follow-up and two deaths were recorded within the following year. No additional deaths occurred during the remaining eight years of follow-up (see Table 4.2.1 and Figure 4.2.1).

The slight improvement in five-year survival between 2000-2009 (92.9%) and 2005-2014 (95.5%) most likely reflects the recent inclusion of LCH in the revised ICD-O-3, which typically has an excellent prognosis.

Table 4.2.1 Childhood lymphomas cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	149	96.7	(92.2 - 98.6)
2	144	95.4	(90.5 - 97.8)
3	142	95.4	(90.5 - 97.8)
4	125	95.4	(90.5 - 97.9)
5	108	95.5	(90.5 - 97.9)
6	93	95.5	(90.6 - 97.9)
7	80	95.5	(90.6 - 98.0)
8	62	95.6	(90.7 - 98.0)
9	48	95.6	(90.7 - 98.1)
10	36	95.7	(90.7 - 98.1)

Figure 4.2.1 Childhood lymphomas cumulative relative survival by time since diagnosis, New Zealand, 2005-2014



4.2.2 Lymphoma survival by diagnostic subgroup, sex, age group, and prioritised ethnicity

There were no significant differences in five-year relative survival for lymphoma according to diagnostic group, sex, age group or ethnicity (see Table 4.2.2).

Table 4.2.2 Childhood lymphomas five-year relative survival by diagnostic subgroup, sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total lymphomas and reticuloendothelial neoplasms	**149**	**100.0**	**95.5**	**(90.5 - 97.9)**
Diagnostic subgroup				
II(a) Hodgkin lymphomas	51	34.2	100.1	[a]
II(b) Non-Hodgkin lymphomas (except Burkitt lymphoma)	49	32.9	94.0	(82.3 - 98.1)
II(c) Burkitt lymphoma	22	14.8	86.4	(63.5 - 95.5)
II(d) Miscellaneous lymphoreticular neoplasms	26	17.4	96.4	(75.9 - 99.7)
II(e) Unspecified lymphomas	1	0.7	100.1	[a]
Sex				
Male	96	64.4	97.0	(90.8 - 99.1)
Female	53	35.6	92.6	(81.2 - 97.2)
Age group				
0-4 years	37	24.8	94.8	(80.2 - 98.8)
5-9 years	34	22.8	94.2	(78.5 - 98.6)
10-14 years	78	52.3	96.3	(88.7 - 98.9)
Prioritised ethnicity				
Maori	28	18.8	89.4	(70.5 - 96.6)
Pacific Peoples	15	10.1	93.4	(61.3 - 99.1)
Non-Maori/non-Pacific	106	71.1	97.3	(91.6 - 99.2)

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.

4.2.3 Hodgkin and non-Hodgkin lymphomas survival by sex, age group, and prioritised ethnicity

Five-year relative survival for children diagnosed with Hodgkin lymphomas between 2005 and 2014 was at 100.1%. There were no differences in five-year relative survival for non-Hodgkin lymphomas according to sex, age group or ethnicity (see Table 4.2.3). This is as expected given the very small number of deaths which occurred among those diagnosed with lymphoma within the study period.

Table 4.2.3 Childhood Hodgkin and non-Hodgkin lymphomas five-year relative survival by sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Hodgkin lymphomas				Non-Hodgkin lymphomas (excluding Burkitt lymphomas)			
	Cases	%	Five-year relative survival (95% CI)		Cases	%	Five-year relative survival (95% CI)	
Total cases	**51**	**100.0**	**100.1**	[a]	**49**	**100.0**	**94.0**	**(82.3 - 98.1)**
Sex								
Male	28	54.9	100.1	[a]	32	65.3	93.9	(77.4 - 98.6)
Female	23	45.1	100.1	[a]	17	34.7	94.2	(65.1 - 99.3)
Age group								
0-4 years	2	3.9	100.1	[a]	8	16.3	100.1	[a]
5-9 years	10	19.6	100.1	[a]	11	22.4	81.9	(44.8 - 95.2)
10-14 years	39	76.5	100.2	[a]	30	61.2	96.8	(78.7 - 99.7)
Prioritised ethnicity								
Maori	8	15.7	100.1	[a]	8	16.3	87.6	(38.8 - 98.3)
Pacific Peoples	4	7.8	-	[b]	4	8.2	100.1	[a]
Non-Maori/non-Pacific Peoples	39	76.5	100.1	[a]	37	75.5	94.7	(80.2 - 98.8)

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.
[b] Five-year relative survival could not be calculated as no cases had a full five years of follow-up.

4.3 Central nervous system tumours and miscellaneous intracranial and intraspinal neoplasms

Tumours of the central nervous system can arise from any structure in the brain, its adjacent coverings and the spinal cord. Within the brain, cancers may develop within primitive neuron-like cells (the embryonal tumours), or the supporting structures such as glial tissue (gliomas), or as remnants of primitive developmental structures (germ cell tumours). This heterogeneous group of tumours vary from relatively benign tumours such as pilocytic astrocytoma, to highly malignant and metastatic tumours such as medulloblastoma and atypical teratoid/rhabdoid tumours. It is likely each group of brain and spinal tumour has a different origin, but some tumours are known to occur in association with familial cancer predisposition syndromes such as Neurofibromatosis type 1, or in association of inherited syndromes such as Gorlin and Li Fraumeni Syndrome.

The treatment and prognosis for a CNS tumour depends on the histological type, its location, the presence or absence of metastatic spread, and the age of the child at diagnosis. Most CNS tumours require expert neurosurgical resection, with many needing further treatment with chemotherapy and/or radiotherapy.

The ICD-O-3[9] used by the New Zealand Cancer Registry (and many other mandatory international cancer registries) classify the benign and low grade gliomas (such as juvenile pilocytic astrocytoma) as non-malignant, so these tumours are not recorded or reported in New Zealand cancer statistics. However, benign and low-grade gliomas in children represent a unique clinical challenge and often require treatment that is similar to malignant CNS tumours and may be associated with long-term morbidity. For this reason, the International Childhood Cancer Classification (ICCC[4]) has included these tumours and, by consensus, international childhood cancer registries record and report on the incidence of non-malignant CNS tumours.

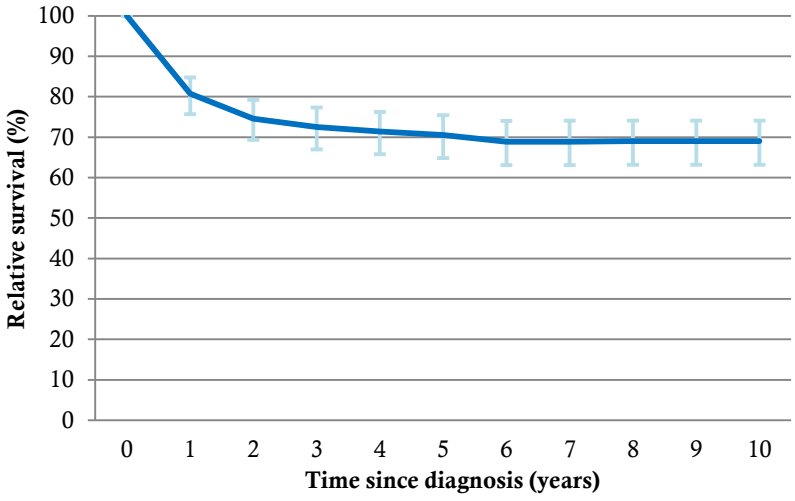
4.3.1 Childhood CNS tumours cumulative relative survival by time since diagnosis

92 deaths were recorded for the 295 children diagnosed with a central nervous system tumour during the follow up period, with many of these (n=57, 62.0%) occurring within the first year following diagnosis (see Table 4.3.1 and Figure 4.3.1). No survival improvements were seen for children diagnosed with CNS tumours in 2005-2014 compared with those diagnosed between 2000 and 2009.

Table 4.3.1 Childhood CNS tumours cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	295	80.7	(75.7 - 84.8)
2	238	74.6	(69.3 - 79.2)
3	220	72.5	(67.0 - 77.3)
4	196	71.4	(65.8 - 76.2)
5	170	70.5	(64.8 - 75.4)
6	148	68.9	(63.1 - 74.0)
7	119	69.0	(63.1 - 74.1)
8	95	69.0	(63.2 - 74.1)
9	77	69.0	(63.2 - 74.1)
10	62	69.0	(63.2 - 74.1)

Figure 4.3.1 Childhood CNS tumours cumulative relative survival by time since diagnosis, New Zealand, 2005-2014



4.3.2 CNS tumour survival by diagnostic subgroup, sex, age group, and prioritised ethnicity

There were substantial differences in five-year survival for CNS tumours according to diagnostic subgroup. Five-year survival probabilities for ependymomas (89.6%), astrocytomas (82.1%) and 'other specified' (89.8%) were significantly higher than for intracranial and intraspinal embryonal tumours (52.9%) and 'other gliomas' (46.0%). There were no sex differences in survival. Children aged 5-9 years at the time of diagnosis had the highest survival at 79.5%.

Table 4.3.2 Childhood CNS tumours five-year relative survival by diagnostic subgroup, sex, age group, and prioritised ethnicity, New Zealand, 2005–2014

	All CNS tumours			
	Total cases	%	Five-year relative survival (95% CI)	
Total cases	**295**	**100.0**	**70.5**	**(64.8 - 75.4)**
Diagnostic subgroup				
III(a) Ependymomas and choroid plexus tumours	29	9.8	89.6	(70.9 - 96.7)
III(b) Astrocytomas	117	39.7	82.1	(73.9 - 88.0)
III(c) Intracranial and intraspinal embryonal tumours	67	22.7	52.9	(40.0 - 64.2)
III(d) Other gliomas	41	13.9	46.0	(30.3 - 60.4)
III(e) Other specified intracranial and intraspinal neoplasms	32	10.8	89.8	(71.1 - 96.7)
III(f) Unspecified intracranial and intraspinal neoplasms	9	3.1	33.4	(7.9 - 62.5)
Sex				
Male	154	52.2	70.6	(62.7 - 77.2)
Female	141	47.8	70.3	(61.8 - 77.2)
Age group				
0-4 years	116	39.3	63.5	(53.9 - 71.6)
5-9 years	104	35.2	79.5	(70.2 - 86.1)
10-14 years	75	25.4	69.0	(57.0 - 78.2)
Prioritised ethnicity				
Maori	78	26.4	71.6	(60.0 - 80.3)
Pacific Peoples	31	10.5	52.8	(33.1 - 69.2)
Non-Maori/non-Pacific Peoples	186	63.1	72.9	(65.8 - 78.8)

4.3.3 Astrocytoma and embryonal tumour survival by sex, age group, and prioritised ethnicity

Diagnostic group III(c): Intracranial and intraspinal embryonal tumours, the majority of which were medulloblastoma, had a five-year survival of 52.9% in 2005-2014 compared to 66.1% in 2000-2009. Survival for those diagnosed when they were 5-9 years of age (80.3%) was significantly higher than those who were diagnosed before the age of 5 (26.7%). Five-year survival for those diagnosed with astrocytomas, such as juvenile pilocytic astrocytoma, increased from 77.7% in 2000-2009 to 82.1% in 2005-2014.

Table 4.3.3 Astrocytoma and embryonal tumour five year relative survival by sex, age group, and prioritised ethnicity, New Zealand, 2005–2014

	Astrocytomas				Intracranial & intraspinal embryonal tumours			
	Total cases	%	Five-year relative survival (95% CI)		Total cases	%	Five-year relative survival (95% CI)	
Total cases	117	100.0	82.1	(73.9 - 88.0)	67	100.0	52.9	(40.0 - 64.2)
Sex								
Male	54	46.2	89.0	(77.0 - 95.0)	48	71.6	49.6	(34.7 - 62.8)
Female	63	53.8	76.2	(63.6 - 85.0)	19	28.4	61.1	(34.7 - 79.5)
Age group								
0-4 years	41	35.0	87.9	(73.3 - 94.9)	26	38.9	26.7	(11.7 - 44.4)
5-9 years	43	36.8	88.4	(74.3 - 95.1)	27	40.3	80.3	(58.7 - 91.4)
10-14 years	33	28.2	66.7	(47.9 - 80.0)	14	20.9	49.6	(22.3 - 72.1)
Prioritised ethnicity								
Maori	29	24.8	86.3	(67.4 - 94.7)	26	38.8	64.5	(42.6 - 79.9)
Pacific Peoples	10	8.5	59.3	(24.3 - 82.5)	8	11.9	37.6	(8.7 - 67.5)
Non-Maori/non-Pacific Peoples	78	66.7	83.4	(73.1 - 90.1)	33	49.3	48.4	(30.7 - 64.0)

4.4 Neuroblastoma and other peripheral nervous cell tumours

Neuroblastoma is a heterogeneous group of cancers that arise from primitive neural crest cells within the sympathetic nervous system. Malignant neuroblastoma most commonly originates in the adrenal glands, or from adjacent abdominal sympathetic nerves, but tumours may arise anywhere along the sympathetic chain from the neck, chest, abdomen and the pelvis. These tumours often present as an otherwise asymptomatic abdominal masses. They vary from benign fully differentiated solid tumours, to highly malignant undifferentiated and metastatic cancers. The aetiology of neuroblastoma remains to be determined; most are sporadic but occasional tumours are associated with familial syndromes.

As many of the early warning signs of neuroblastoma, such as fatigue, pain, loss of appetite, and fever, mimic those of other common childhood illnesses, these tumours may grow to a very large size before becoming clinically apparent. The prognosis and treatment of neuroblastoma depends on the patient's age, the tumour site and histology, the presence of specific molecular features, and the extent of disease at diagnosis. Malignant neuroblastoma requires very aggressive treatment with multi-agent chemotherapy, surgery, radiotherapy, stem cell transplantation, differentiation therapy and immunotherapy. Recent developments in treatment have significantly improved the prognosis for children with advanced stage neuroblastoma. In New Zealand most children are treated according to an international clinical trial.

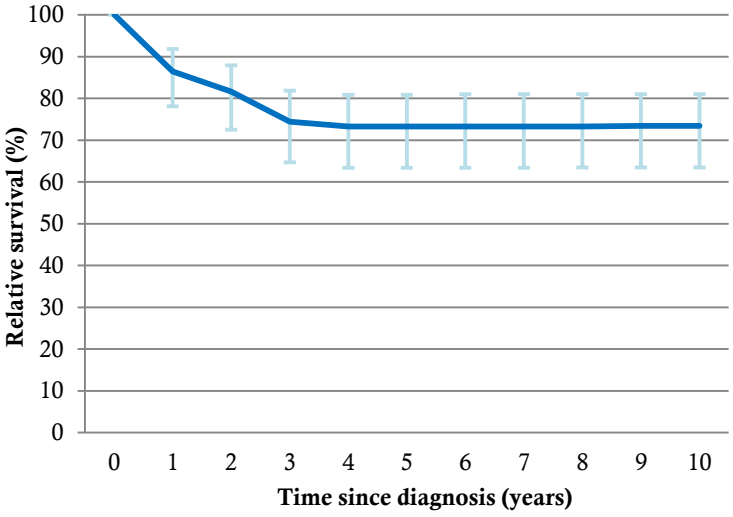
4.4.1 Childhood neuroblastoma cumulative relative survival by time since diagnosis

Within the cohort of 102 children diagnosed with 'neuroblastoma and other peripheral nervous cell tumours', 27 died during the follow-up period, including 14 within one year of their initial diagnosis. However, as noted above, a neuroblastoma patient's expected survival is heavily dependent on a number of known risk factors. Table 4.4.1 and Figure 4.4.1 show that there was little further decline in survival after five years following diagnosis. Five year survival improved from 66.1% in 2000-2009 to 73.3% in 2005-2014.

Table 4.4.1 Childhood neuroblastoma cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	102	86.5	(78.1 - 91.8)
2	88	81.6	(72.5 - 87.9)
3	83	74.4	(64.7 - 81.9)
4	70	73.3	(63.4 - 80.9)
5	58	73.3	(63.4 - 80.9)
6	45	73.3	(63.4 - 81.0)
7	34	73.3	(63.4 - 81.0)
8	29	73.3	(63.5 - 81.0)
9	24	73.4	(63.5 - 81.0)
10	19	73.4	(63.5 - 81.0)

Figure 4.4.1 Childhood neuroblastoma cumulative relative survival by time since diagnosis, New Zealand, 2005-2014



4.4.2 Neuroblastoma survival by diagnostic subgroup, sex, age group, and prioritised ethnicity

Neuroblastoma was the only type of peripheral nervous cell tumour diagnosed within the study period. There were no major differences in five-year survival according to sex, age group or prioritised ethnicity.

Table 4.4.2 Childhood neuroblastoma five-year relative survival by sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total neuroblastoma & other peripheral nervous cell tumours	**102**	**100**	**73.3**	**(63.4 - 80.9)**
Diagnostic subgroup				
IV(a) Neuroblastoma & ganglioneuroblastoma	102	100	73.3	(63.4 - 80.9)
IV(b) Other peripheral nervous cell tumours	-	-	-	
Sex				
Male	49	48.0	73.4	(58.5 - 83.8)
Female	53	52.0	73.3	(59.0 - 83.3)
Age group				
0-4 years	86	84.3	76.6	(65.9 - 84.3)
5-9 years	14	13.7	64.3	(34.4 - 83.4)
10-14 years	1	1.0	[b]	[b]
Prioritised ethnicity				
Maori	22	21.6	67.9	(44.0 - 83.4)
Pacific Peoples	4	3.9	75.3	(12.8 - 96.4)
Non-Maori/non-Pacific Peoples	76	74.5	74.8	(63.3 - 83.2)

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.
[b] Five-year relative survival could not be calculated as no cases had a full five years of follow-up.

4.5 Retinoblastoma

Retinoblastoma forms from primitive retinal cells in the eye and often progresses rapidly to fill the entire posterior chamber of the orbit. Although rare, it is the most common type of eye cancer in children. Retinoblastoma may be sporadic or occur in association with familial mutations in the retinoblastoma gene (RB1). Sporadic retinoblastoma most commonly involves one eye (unilateral RB) and most cases do not have a germline mutation of the RB gene. Hereditary retinoblastoma develops in children inheriting a germline mutation of the RB gene; it may be unilateral, bilateral and in rare cases also involve the pineal gland (trilateral RB). Patients with hereditary RB are at long-term risk of developing additional cancers and second cancers.

The most common presentation of RB is when parents notice a white pupil (leucoria) instead of the typical "red eye" seen in photos taken with a flash. Other symptoms can include squinting, crossed eyes, eye swelling and redness, and double vision. Management of retinoblastoma requires an expert ophthalmology assessment of the affected and unaffected eye and access to high quality diagnostic imaging, expert diagnostic pathology and molecular genetics. Most children are diagnosed with retinoblastoma before they are five years old. Provided the cancer has not spread beyond the eye, retinoblastoma has one of the best survival rates of all childhood cancers.

4.5.1 Childhood retinoblastoma cumulative relative survival by time since diagnosis

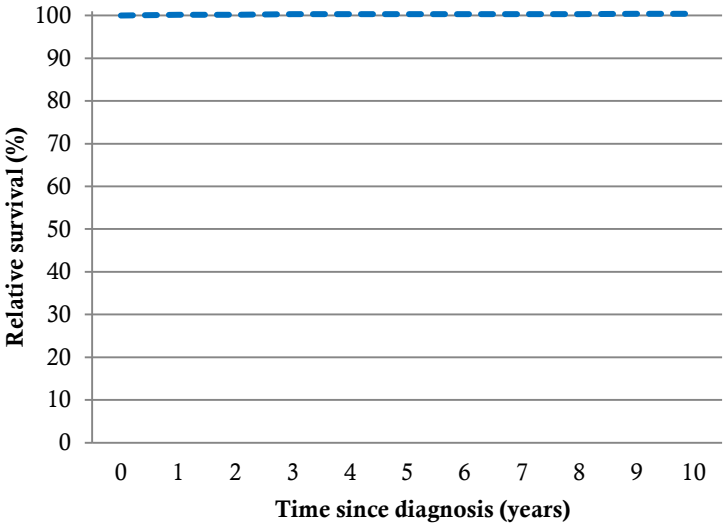
The survival of children diagnosed with retinoblastoma is similar to the survival of New Zealand's general child population. Not a single death was recorded among the 41 children diagnosed with retinoblastoma during the 2005-2014 period.

Table 4.5.1 Childhood retinoblastoma cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	41	100.1	[a]
2	41	100.2	[a]
3	41	100.2	[a]
4	38	100.2	[a]
5	31	100.2	[a]
6	26	100.2	[a]
7	18	100.2	[a]
8	13	100.2	[a]
9	11	100.3	[a]
10	10	100.3	[a]

[a] Confidence intervals cannot be calculated in instances where no deaths were recorded.

Figure 4.5.1 Childhood retinoblastoma cumulative relative survival by time since diagnosis, New Zealand, 2005-2014



4.5.2 Retinoblastoma survival by sex, age group, and prioritised ethnicity

As there was not a single death recorded among children diagnosed with retinoblastoma in New Zealand between 2000 and 2009, it follows that there were therefore no differences according to sex, age group, or ethnicity (see Table 4.5.2).

Table 4.5.2 Childhood retinoblastoma five-year relative survival by sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total retinoblastoma	**41**	**100.0**	**100.2**	[a]
Sex				
Male	21	51.2	100.2	[a]
Female	20	48.8	100.2	[a]
Age group				
0-4 years	39	95.1	100.2	[a]
5-9 years	1	2.4	100.1	[a]
10-14 years	1	2.4	100.1	[a]
Prioritised ethnicity				
Maori	8	19.5	100.2	[a]
Pacific Peoples	7	17.1	100.2	[a]
Non-Maori/non-Pacific Peoples	26	63.4	100.2	[a]

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.

4.6 Renal tumours

Renal tumours, or malignancies of the kidney, represent around 6% of cancer diagnoses among children younger than 15 years of age. Nephroblastoma (also known as Wilms' tumour) is the most common form of childhood renal cancer. Other primary renal tumours in children include rhabdoid tumour of the kidney, clear cell sarcoma of the kidney, renal carcinoma and rare cases of intra-renal rhabdomyosarcoma and neuroblastoma. While most Wilms' tumours are sporadic, some occur in association with specific developmental disorders such as the Beckwith Wiedemann syndrome, WAGR syndrome and Denys-Drash syndrome. At diagnosis, most Wilms' tumours are unilateral but about 7% are bilateral.

Wilms' tumours arise in the developing kidney from primitive malignant clusters of cells termed nephrogenic rests. These form during growth of the kidney in utero and transform into malignant tumours during post-natal growth and development. Wilms' tumours usually present in children under the age of five with an abdominal mass but occasionally present with pain and haematuria. Treatment usually involves nephrectomy and pre and/or post-operative chemotherapy. Some cases also require radiotherapy. Children in New Zealand are treated according to an international collaborative clinical trial through SIOP or COG.

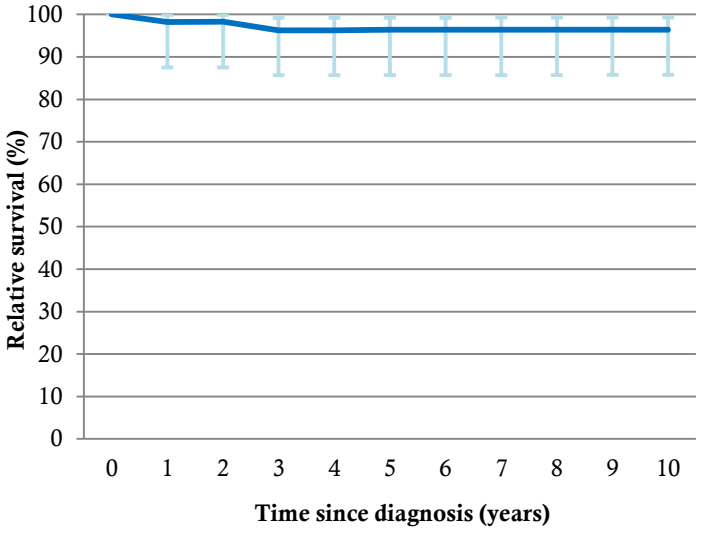
4.6.1 Childhood renal tumours cumulative relative survival by time since diagnosis

Between 2005 and 2014 in New Zealand renal tumours had one of the best survival probabilities of all ICCC-3 diagnostic groups. Of the 53 cases diagnosed within the time period, only two deaths was recorded, one within the first year following diagnosis and one in the third year.

Table 4.6.1 Childhood renal tumours cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	53	98.2	(87.5 - 99.9)
2	52	98.3	(87.5 - 99.9)
3	52	96.3	(85.7 - 99.2)
4	48	96.3	(85.7 - 99.2)
5	42	96.4	(85.7 - 99.2)
6	33	96.4	(85.7 - 99.2)
7	27	96.4	(85.7 - 99.3)
8	20	96.4	(85.7 - 99.3)
9	14	96.4	(85.8 - 99.3)
10	11	96.4	(85.8 - 99.3)

Figure 4.6.1 Childhood renal tumours relative survival by time since diagnosis, New Zealand, 2005-2014



4.6.2 Renal tumour survival by diagnostic subgroup, sex, age group, and prioritised ethnicity

Nephroblastoma (which is commonly known as Wilms' tumour) accounted for all 61 renal tumours diagnosed between 2005 and 2014. Renal carcinomas are rare in children and none were diagnosed within the 10-year period.

Table 4.6.2 Childhood renal tumours five-year relative survival by sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total renal tumours	**53**	**100**	**96.4**	**(85.7 - 99.2)**
Diagnostic subgroup				
VI(a) Nephroblastoma & other non-epithelial renal tumours	53	100	96.4	(85.7 - 99.2)
VI(b) Renal carcinomas	-	-	-	
VI(c) Unspecified malignant renal tumours	-	-	-	
Sex				
Male	19	35.8	94.8	(67.6 - 99.5)
Female	34	64.2	97.2	(81.0 - 99.7)
Age group				
0-4 years	44	83.0	97.9	(85.1 - 99.9)
5-9 years	7	13.2	85.8	(33.4 - 97.9)
10-14 years	2	3.8	100.1	[a]
Prioritised ethnicity				
Maori	5	9.4	100.2	[a]
Pacific Peoples	5	9.4	100.2	[a]
Non-Maori/non-Pacific Peoples	43	81.1	95.5	(82.7 - 99.0)

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.

4.7 Hepatic tumours

Primary liver tumours are rare in children. The two most common subgroups diagnosed are hepatoblastoma and hepatocellular carcinoma. Although representing a small proportion of the total number of cases diagnosed, some genetic conditions, such as Beckwith-Wiedemann syndrome, are associated with an increased risk of developing hepatoblastoma, while hepatocellular carcinoma, more commonly diagnosed in adulthood, is associated with a prior history of hepatitis and a number of chronic liver conditions. Hepatic tumours may present with non-specific symptoms including abdominal distension, pain, a palpable mass, weight loss, and jaundice.

Hepatocellular carcinoma can be resistant to chemotherapy and is typically treated in adults with surgery alone but children with hepatocellular carcinoma are usually offered chemotherapy to achieve resectability. The prognosis for hepatoblastoma depends on the histological subtype, the level of tumour marker, the extent of tumour in the liver, and the presence or absence of metastatic spread. Hepatoblastoma is usually treated with pre-operative chemotherapy, surgical resection of the tumour or liver transplantation, and post-operative chemotherapy. Children in New Zealand with hepatoblastoma are currently treated according to the SIOPEL international cooperative clinical trial. For those with localised and resectable disease the overall prognosis is excellent.

4.7.1 Childhood hepatic tumours cumulative relative survival by time since diagnosis

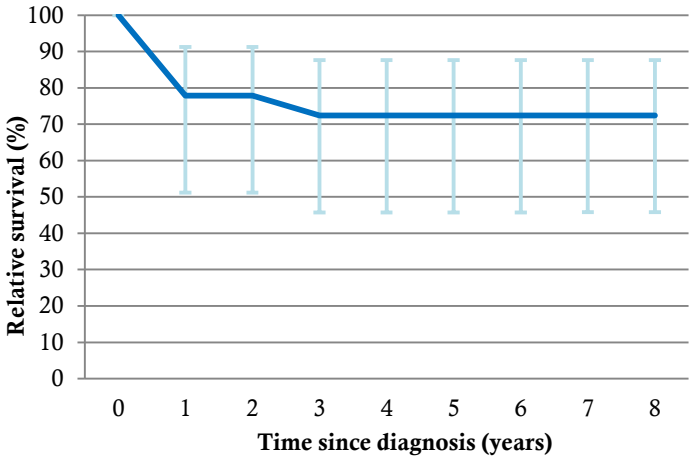
Of the 18 children who were diagnosed with a hepatic tumour between 2005 and 2014, five died during the follow-up period. In four of the five cases the death occurred within one year of the initial diagnosis (see Table 4.7.1 and Figure 4.7.1).

Table 4.7.1 Childhood hepatic tumours cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)[a]	n	Cumulative relative survival (95% CI)	
1	18	77.9	(51.2 - 91.2)
2	14	77.9	(51.2 - 91.2)
3	14	72.4	(45.7 - 87.6)
4	13	72.4	(45.7 - 87.6)
5	9	72.4	(45.7 - 87.6)
6	7	72.4	(45.7 - 87.6)
7	5	72.4	(45.8 - 87.6)
8	3	72.4	(45.8 - 87.6)

[a] Nine and ten year relative survival estimates could not be calculated as no cases had more than 8 years of follow up

Figure 4.7.1 Childhood hepatic tumours cumulative relative survival by time since diagnosis, New Zealand, 2005-2014



4.7.2 Hepatic tumour survival by diagnostic subgroup, sex, age group, and prioritised ethnicity

Of the five hepatic tumour deaths recorded, two were cases of hepatoblastoma and three were hepatic carcinomas. Most deaths occurred in children over the age of five.

Table 4.7.2 Childhood hepatic tumours five-year relative survival by diagnostic subgroup, sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total hepatic tumours	**18**	**100.0**	**72.4**	**(45.7 - 87.6)**
Diagnostic subgroup				
VII(a) Hepatoblastoma	12	66.7	83.6	(48.3 - 95.9)
VII(b) Hepatic carcinomas	6	33.3	50.1	(11.1 - 80.5)
VII(c) Unspecified malignant hepatic tumours	-	-	-	-
Sex				
Male	9	50.0	78.0	(36.6 - 94.2)
Female	9	50.0	66.8	(28.2 - 88.0)
Age group				
0-4 years	13	72.2	92.6	(56.8 - 99.2)
5-9 years	4	22.2	25.1	(0.9 - 66.6)
10-14 years	1	5.6	0.0	[a]
Prioritised ethnicity				
Maori	5	27.8	80.3	(20.5 - 97.2)
Pacific Peoples	2	11.1	[b]	[b]
Non-Maori/non-Pacific Peoples	11	61.1	82.0	(44.9 - 95.4)

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.
[b] Five-year relative survival could not be calculated as no cases had a full five years of follow-up.

4.8 Malignant bone tumours

The two most common types of primary malignant bone tumour in children are osteosarcomas and Ewing sarcomas. Osteosarcomas originate in the osteoid tissue and usually grow in the long bones of the leg, often directly above the knee joint. Ewing sarcomas arise from primitive mesenchymal elements in the bone or, less often, in soft tissue (those which originate in soft tissue are classified as a soft tissue sarcoma rather than a malignant bone tumour according to the ICCC-3). Ewing sarcomas may develop in any bone but most commonly develop in the long bones or bones of the central axis, including vertebrae, ribs, sternum, clavicle and pelvis.

Most primary bone tumours are sporadic but osteosarcoma can rarely develop in association with the Li Fraumeni syndrome, Rothmund Thomson syndrome, and in children with germline retinoblastoma mutations. Ewing sarcoma is nearly always sporadic with few know risk factors. Most primary bone tumours present as a painful progressive swelling of a bone in a teenager; these grow slowly and are often diagnosed as a soft tissue injury before the correct diagnosis is established. The peak age of onset is 14 years, coinciding with the pubertal growth spurt. The prognosis and treatment depends on the tumour histology, its location and extent of disease. All primary bone tumours require expert treatment with chemotherapy, surgery, and, less often, radiotherapy.

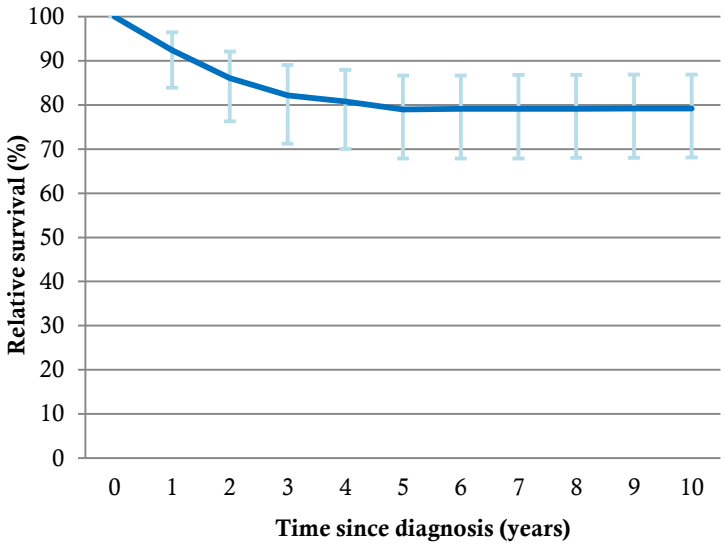
4.8.1 Childhood malignant bone tumours cumulative relative survival by time since diagnosis

Of the 17 deaths recorded within the cohort of 79 children diagnosed with a malignant bone tumour between 2005 and 2014, all occurred within the first five years of follow-up (see Table 4.8.1). Survival probabilities were consistently higher than those reported for the 2000-2009 period, with five-year survival at 79.0% (c.f. 66.8%) and ten-year survival at 79.2% (c.f. 58.6%).

Table 4.8.1 Childhood malignant bone tumours cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	79	92.4	(83.9 - 96.5)
2	73	86.1	(76.3 - 92.1)
3	68	82.2	(71.8 - 89.1)
4	61	80.8	(70.1 - 88.0)
5	50	79.0	(67.9 - 86.7)
6	40	79.1	(67.9 - 86.7)
7	37	79.1	(67.9 - 86.8)
8	27	79.1	(68.0 - 86.8)
9	22	79.2	(68.0 - 86.9)
10	15	79.2	(68.1 - 86.9)

Figure 4.8.1 Childhood malignant bone tumours relative survival by time since diagnosis, New Zealand, 2005-2014



4.8.2 Malignant bone tumour survival by diagnostic subgroup, sex, age group, and prioritised ethnicity

In 2000-2009 five-year survival for childhood malignant bone tumours was significantly below survival for childhood cancer overall (66.8% c.f. 80.7%) but in 2005-2014 this difference had reduced to less than 5% (79.0% c.f. 83.6%). Table 4.8.2 shows that survival was considerably higher for males (89.0%) compared to females (64.9%), although this did not reach statistical significance. Five-year survival was similar for the two main types of bone tumours diagnosed in children; Ewing tumours (80.5%) and osteosarcomas (78.1%).

Table 4.8.2 Childhood malignant bone tumours five-year relative survival by diagnostic subgroup, sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total malignant bone tumours	**79**	**100.0**	**79.0**	**(67.9 - 86.7)**
Diagnostic subgroup				
VIII(a) Osteosarcomas	38	48.1	78.1	(60.7 - 88.5)
VIII(b) Chondrosarcomas	1	1.3	100.3	[a]
VIII(c) Ewing tumours & related bone sarcomas	32	40.5	80.5	(61.3 - 90.8)
VIII(d) Other specified malignant bone tumours	8	10.1	75.1	(31.5 - 93.2)
VIII(e) Unspecified malignant bone tumours	-	-	-	-
Sex				
Male	46	58.2	89.0	(75.3 - 95.4)
Female	33	41.8	64.9	(45.2 - 79.1)
Age group				
0-4 years	4	5.1	50.0	(5.8 - 84.6)
5-9 years	20	25.3	85.1	(60.4 - 95.0)
10-14 years	55	69.6	78.9	(64.7 - 87.8)
Prioritised ethnicity				
Maori	17	21.5	66.3	(35.1 - 85.2)
Pacific Peoples	14	17.7	78.7	(47.3 - 92.7)
Non-Maori/non-Pacific Peoples	48	60.8	83.1	(69.0 - 91.3)

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period

4.8.3 Osteosarcomas and Ewing tumours survival by sex, age group, and prioritised ethnicity

Table 4.8.3 shows that the differences in survival by sex were most marked for Ewing tumours of the bone, with all those who died within the period being female (five-year survival: 59.7%).

Table 4.8.3 Childhood osteosarcoma and Ewing tumour five-year relative survival by sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Osteosarcomas				Ewing tumour and related bone sarcomas			
	Cases	%	Five-year relative survival (95% CI)		Cases	%	Five-year relative survival (95% CI)	
Total cases	**38**	**100.0**	**78.1**	**(60.7 - 88.5)**	**32**	**100.0**	**80.5**	**(61.3 - 90.8)**
Sex								
Male	24	63.2	82.9	(60.4 - 93.3)	17	53.1	100.1	[a]
Female	14	36.8	70.2	(38.5 - 87.7)	15	53.1	59.7	(31.3 - 79.6)
Age group								
0-4 years	-	-	-	-	3	9.4	66.7	(5.4 - 94.6)
5-9 years	9	23.7	89.0	(43.3 - 98.4)	8	25.0	87.6	(38.7 - 98.2)
10-14 years	29	76.3	74.9	(54.1 - 87.3)	21	65.6	79.6	(53.8 - 92.0)
Prioritised ethnicity								
Maori	7	18.4	71.5	(25.8 - 92.1)	9	28.1	63.7	(23.5 - 86.9)
Pacific Peoples	8	21.1	75.1	(31.5 - 93.2)	5	15.6	100.1	[a]
Non-Maori/non-Pacific Peoples	23	60.5	82.1	(58.9 - 93.0)	18	56.3	83.2	(56.4 - 94.3)

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period

4.9 Soft tissue and other extraosseous sarcomas

Soft tissue sarcomas are amongst the most diverse and challenging of all childhood cancers. They arise from malignant precursor cells in tissue of mesenchymal origin; cells that normally produce muscle, fibrous tissue, fat, blood vessels and other supporting tissue. Therefore, they can they can develop in any location and with highly varied histology. While there are over 50 different histological subtypes, the most common soft tissue sarcoma diagnosed in children are rhabdomyosarcomas, which account for over half of all cases diagnosed.

Rhabdomyosarcomas most commonly develop in the abdomen, trunk, head and neck, and in the extremities. The protean nature of these tumours makes them difficult to diagnose and they may present late and have disseminated by the time of diagnosis. As with other solid tumours of childhood, the prognosis and treatment depends on the location, histology, and extent of spread of the tumour. The malignant sarcomas all require multi-agent chemotherapy, surgery, and many need local radiotherapy. This class of tumour is challenging to diagnose and manage, and their treatment is associated with significant long-term treatment-related toxicity.

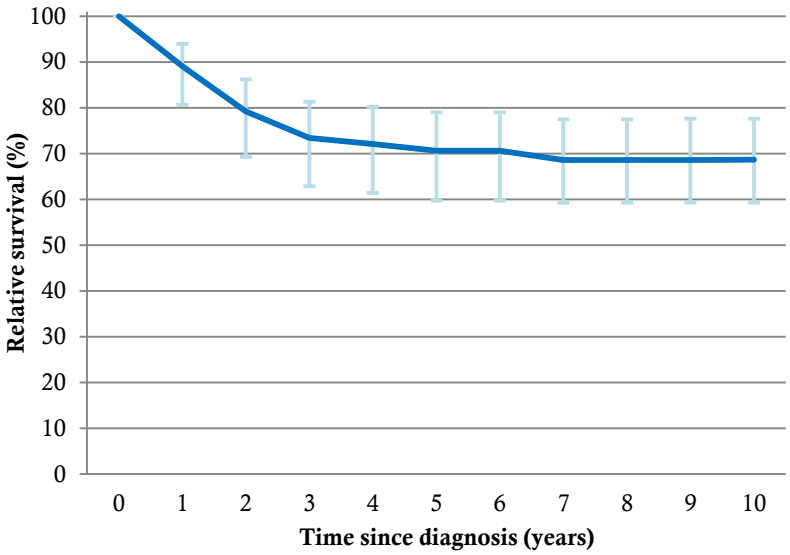
4.9.1 Childhood soft tissue sarcomas cumulative relative survival by time since diagnosis

Table 4.9.1 and Figure 4.9.1 show that most soft tissue sarcoma deaths within this cohort occurred within three years of diagnosis. Three-year survival for soft tissue sarcomas was 73.4%, 11.6% below the three-year survival for childhood cancers overall.

Table 4.9.1 Childhood soft tissue sarcomas cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	91	89.1	(80.6 - 94.0)
2	81	79.2	(69.3 - 86.2)
3	72	73.4	(62.9 - 81.3)
4	58	72.1	(61.4 - 80.2)
5	52	70.6	(59.7 - 79.0)
6	43	70.6	(59.7 - 79.0)
7	36	68.6	(57.2 - 77.5)
8	33	68.6	(57.3 - 77.5)
9	29	68.6	(57.3 - 77.6)
10	22	68.7	(57.3 - 77.6)

Figure 4.9.1 Childhood soft tissue sarcomas cumulative relative survival by time since diagnosis, New Zealand, 2005-2014



4.9.2 Soft tissue sarcoma survival by sex, age group, and prioritised ethnicity

Five-year survival for soft tissue sarcomas (70.6%) was significantly less than for childhood cancers overall (83.6%). Although not statistically significant, soft tissue sarcomas five-year survival was higher for girls than boys (80.0% c.f. 62.8%) and for 5-9 year olds (83.1%) compared to younger (68.3%) and older children (66.4%), (see Table 4.9.2). By prioritised ethnicity, the highest five year survival was recorded for Pacific children (89.0%), almost twice that of Maori (46.8%).

Table 4.9.2 Childhood soft tissue sarcomas five-year relative survival by diagnostic group, sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total cases	**91**	**100.0**	**70.6**	**(59.7 - 79.0)**
Soft tissue and other extraosseous sarcomas				
IX(a) Rhabdomyosarcomas	48	52.7	66.0	(50.5 - 77.7)
IX(b) Fibrosarcomas & other fibrous neoplasms	6	6.6	83.6	(27.4 - 97.7)
IX(c) Kaposi sarcomas	-	-	-	-
IX(d) Other specified soft tissue sarcomas	28	30.8	69.8	(48.0 - 83.9)
IX(e) Unspecified soft tissue sarcomas	9	9.9	89.1	(43.4 - 98.6)
Sex				
Male	50	54.9	62.8	(47.3 - 74.9)
Female	41	45.1	80.0	(63.7 - 89.5)
Age group				
0-4 years	39	42.9	68.3	(50.8 - 80.8)
5-9 years	19	20.9	83.1	(56.0 - 94.3)
10-14 years	33	36.3	66.4	(47.5 - 79.9)
Prioritised ethnicity				
Maori	14	15.4	46.8	(18.7 - 70.9)
Pacific Peoples	9	9.9	89.0	(43.3 - 98.4)
Non-Maori/non-Pacific Peoples	68	74.7	72.8	(60.2 - 82.0)

4.9.3 Soft tissue sarcoma survival by diagnostic subgroup

Rhabdomyosarcomas was the main type of soft tissue sarcoma diagnosed in children between 2005 and 2014 and had a five-year survival probability of 66.0%, down from 73.1% for the 2000-2009 period. Rhabdomyosarcoma is most commonly diagnosed in those under the age of five. Survival was poorest for older children aged 10-14 years (37.8%) compared to 0-4 year olds (67.8%) and 5-9 year olds (100.1%).

Table 4.8.3 Childhood rhabdomyosarcoma five-year relative survival by sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Rhabdomyosarcomas			
	Total cases	%	Five-year relative survival (95% CI)	
Total cases	**48**	**100.0**	**66.0**	**(50.5 - 77.7)**
Sex				
Male	29	60.4	65.1	(44.7 - 79.6)
Female	19	39.6	67.7	(41.6 - 84.1)
Age group				
0-4 years	26	54.2	67.8	(45.4 - 82.6)
5-9 years	9	18.8	100.1	[a]
10-14 years	13	27.1	37.8	(13.4 - 62.5)
Prioritised ethnicity				
Maori	7	14.6	57.2	(17.2 - 83.8)
Pacific Peoples	5	10.4	80.1	(20.4 - 97.0)
Non-Maori/non-Pacific Peoples	36	75.0	65.9	(47.6 - 79.1)

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period

4.10 Germ cell tumours, trophoblastic tumours, and neoplasms of gonads

Germ cell tumours are the archetypal embryonic tumour. They all develop from primitive tissue remnants of embryonal tissues and may form in the developing gonads (gonadal germ cell tumours), or in regions of the chest, abdomen, and brain, where germ cell elements can persist beyond foetal development. Many germ cell tumours are benign (and therefore not classified by ICCC[4]).

The presenting symptoms of germ cell tumours will vary considerably depending on the site; a boy with a gonadal germ cell tumour may develop a painless mass in the scrotum, while a child with an intracranial germ cell tumour may experience difficulty with movement or exhibit personality changes. The most common treatment for germ cell tumours is surgery, sometimes in conjunction with chemotherapy. Malignant germ cell tumours respond well to chemotherapy and even metastatic disease has an excellent long-term prognosis.

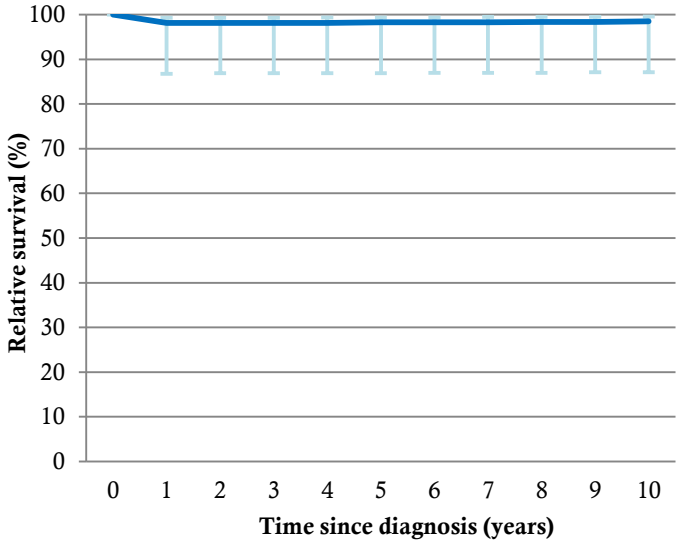
4.10.1 Childhood germ cell tumours cumulative relative survival by time since diagnosis

The germ cell tumour group of cancers has one of the best survival probabilities of all childhood cancers. Within the cohort of 50 children diagnosed between 2000 and 2009, only one death was recorded. This occurred within one year of their diagnosis (see Table 4.10.1 and Figure 4.10.1).

Table 4.10.1 Childhood germ cell tumours cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	50	98.2	(86.8 - 99.9)
2	49	98.2	(86.8 - 99.9)
3	49	98.2	(86.8 - 99.9)
4	45	98.2	(86.9 - 100.0)
5	39	98.3	(86.9 - 100.0)
6	35	98.3	(86.9 - 100.0)
7	32	98.3	(86.9 - 100.1)
8	28	98.4	(87.0 - 100.1)
9	21	98.4	(87.0 - 100.2)
10	13	98.5	(87.1 - 100.2)

Figure 4.10.1 Childhood germ cell tumours cumulative relative survival by time since diagnosis, New Zealand, 2005-2014



4.10.2 Germ cell tumour survival by diagnostic subgroup, sex, age group, and prioritised ethnicity

There was little difference in germ cell tumours survival according to diagnostic subgroup, sex, age group, or ethnicity, which is to be expected given that childhood germ cell tumour have an excellent overall survival. Table 4.10.2 shows that the single death within the cohort was recorded for an older boy diagnosed with an intracranial and intraspinal germ cell tumour.

Table 4.10.2 Childhood germ cell tumours five-year relative survival by diagnostic subgroup sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total germ cell tumours	**50**	**100.0**	**98.3**	**(86.9 – 100.0)**
Diagnostic subgroup				
X(a) Intracranial & intraspinal germ cell tumours	20	40.0	95.1	(69.6 - 99.4)
X(b) Malignant extracranial & extragonadal germ cell tumours	14	28.0	100.5	[a]
X(c) Malignant gonadal germ cell tumours	15	30.0	100.3	[a]
X(d) Gonadal carcinomas	-	-	-	-
X(e) Other & unspecified malignant gonadal tumours	1	2.0	[b]	[b]
Sex				
Male	26	52.0	96.5	(76.0 - 99.8)
Female	24	48.0	100.2	[a]
Age group				
0-4 years	19	38.0	100.5	[a]
5-9 years	6	12.0	100.1	[a]
10-14 years	25	50.0	96.2	(75.0 - 99.6)
Prioritised ethnicity				
Maori	18	36.0	94.7	(66.8 - 99.5)
Pacific Peoples	4	8.0	100.1	[a]
Non-Maori/non-Pacific Peoples	28	56.0	100.3	[a]

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.
[b] Five-year relative survival could not be calculated as no cases had a full five years of follow-up.

4.11 Other malignant epithelial neoplasms and malignant melanomas

Cancers of epithelial origin are the most common cancers seen in adults and are termed carcinomas. Most adult carcinomas arise in a specific location – such as the breast, colon, prostate, lung and pancreas, and are associated with older age and environmental and lifestyle factors. This class of cancer is rare in childhood but they begin to make an appearance in the young adult age group.

The most common malignant epithelial cancer seen in children is melanoma. These may arise in children where there is a family history of melanoma, or in association with congenital melanocytic nevi. Although sun exposure plays less of a role in the development of melanoma in children than in adults, childhood melanoma incidence rates are nevertheless higher in countries with high ultra-violet radiation, such as New Zealand. Melanoma is usually treated with complete removal by surgery and monitored carefully due to the possibility of the cancer spreading to other parts of the body and the increased risk of developing another melanoma in later life.

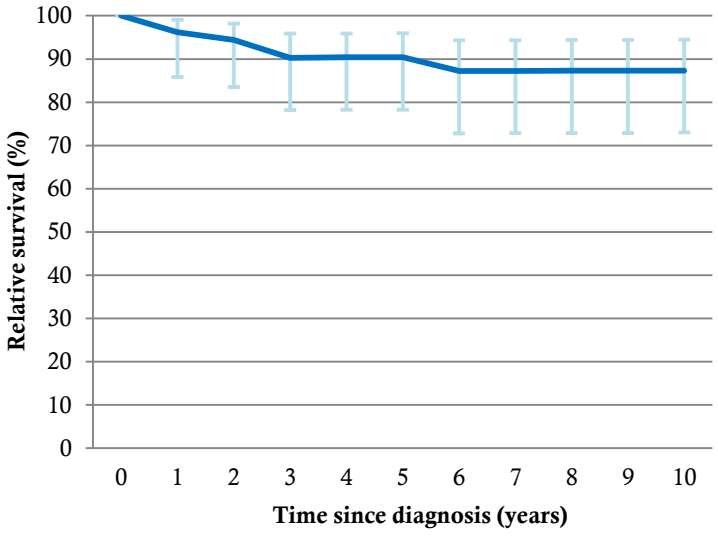
4.11.1 Other childhood malignant epithelial neoplasms cumulative relative survival by time since diagnosis

Within the cohort, 53 children were diagnosed with a cancer from the diagnostic group 'other malignant epithelial neoplasms and malignant melanomas'. Five-year survival was over 90%, with 10 year survival at 87.3% (see Table 4.11.1 and Figure 4.11.1).

Table 4.11.1 Childhood malignant epithelial neoplasms cumulative relative survival, New Zealand, 2005-2014

Time since diagnosis (years)	n	Cumulative relative survival (95% CI)	
1	53	96.2	(85.8 - 99.1)
2	51	94.4	(83.5 - 98.2)
3	50	90.3	(78.2 - 95.9)
4	41	90.4	(78.3 - 95.9)
5	37	90.4	(78.3 - 96.0)
6	29	87.2	(72.8 - 94.3)
7	25	87.2	(72.9 - 94.3)
8	23	87.3	(72.9 - 94.4)
9	19	87.3	(72.9 - 94.4)
10	12	87.3	(73.0 - 94.5)

Figure 4.11.1 Childhood malignant epithelial neoplasms cumulative relative survival, New Zealand, 2005-2014



4.11.2 Other malignant epithelial neoplasm survival by diagnostic subgroup, sex, age group, and prioritised ethnicity

The diagnostic group 'other malignant epithelial neoplasms and malignant melanomas' includes a diverse range of tumours and the survival probabilities are known to vary considerably. During the follow-up period, one child diagnosed with melanoma died (five-year relative survival for melanomas, the main specified diagnostic subgroup, was 95.4%). The remaining deaths were recorded for one child diagnosed with an adrenocortical carcinoma and three of the 21 children diagnosed with an 'other and unspecified carcinomas' (see Table 4.11.2).

Table 4.11.2 Childhood malignant epithelial neoplasms five-year survival by diagnostic subgroup, sex, age group, and prioritised ethnicity, New Zealand, 2005-2014

	Total cases	%	Five-year relative survival (95% CI)	
Total malignant epithelial neoplasms & melanomas	**53**	**100.0**	**90.4**	**(78.3 - 96.0)**
Diagnostic subgroup				
XI(a) Adrenocortical carcinomas	3	5.7	66.7	(5.4 - 94.6)
XI(b) Thyroid carcinomas	7	13.2	100.1	[a]
XI(c) Nasopharyngeal carcinomas	1	1.9	100.3	[a]
XI(d) Melanomas	21	39.6	95.4	(70.8 - 99.5)
XI(e) Skin carcinomas	-	-	-	-
XI(f) Other & unspecified carcinomas	21	39.6	84.8	(59.6 - 94.9)
Sex				
Male	23	43.4	95.8	(73.1 - 99.5)
Female	30	56.6	86.1	(66.9 - 94.6)
Age group				
0-4 years	5	9.4	80.1	(20.4 - 97.0)
5-9 years	8	15.1	87.6	(38.7 - 98.1)
10-14 years	40	75.5	92.4	(77.9 - 97.6)
Prioritised ethnicity				
Maori	10	18.9	90.1	(47.4 - 98.7)
Pacific Peoples	3	5.7	66.8	(5.4 - 94.7)
Non-Maori/non-Pacific Peoples	40	75.5	92.4	(78.0 - 97.6)

[a] Confidence intervals cannot be calculated in instances where there were either no deaths or no survivors within the period.

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Appendices

AI Abbreviations

ALL	Acute lymphoblastic leukaemia
AML	Acute myeloid leukaemia
AYA	Adolescents and young adults
CI	Confidence Interval
CHOC	Children's Haematology and Oncology Centre
CNS	Central nervous system
COG	Children's Oncology Group
HL	Hodgkin lymphoma
IARC	International Association for Research on Cancer
ICCC-3	International Classification of Childhood Cancer, Third edition
ICD-O-3	International Statistical Classification of Diseases for Oncology, Third edition
LCH	Langerhans cell histiocytosis
MELAA	Middle Eastern, Latin American and African
MOH	Ministry of Health
NCCN	National Child Cancer Network
NHL	Non-Hodgkin lymphoma
NZCCR	New Zealand Children's Cancer Registry
NZCR	New Zealand Cancer Registry
POSG	Paediatric Oncology Steering Group
RB	Retinoblastoma
SEER	Surveillance Epidemiology and End Results (U.S. Cancer Statistics)
SIOP	Societe Internationale d'Oncologie Pediatrique (International Society of Paediatric Oncology)
SIOPEL	Societe Internationale d'Oncologie Pediatrique – Epithelial Liver Tumour Study Group (recently renamed as the International Childhood Liver Tumors Strategy Group)
WHO	World Health Organisation

AII International Classification of Childhood Cancer, Third Edition (ICCC-3)[5]

The ICCC-3 classifies childhood cancers according to ICD-O-3 histology and site. Cancers are classified into 12 main diagnostic groups, which are further split into 47 subgroups. The following table assigns the morphology and topography codes of ICD-O-3 to the ICCC-3 main diagnostic groups and subgroups.

Table AII International Classification of Childhood Cancer, Third Edition[5]

Diagnostic group/subgroup	ICD-O-3 histology	ICD-O-3 site
I. Leukaemias, myeloproliferative diseases & myelodysplastic diseases		
(a) Lymphoid leukaemias	9820, 9823, 9826, 9827, 9831-9837, 9940, 9948	C000-C809
(b) Acute myeloid leukaemias	9840, 9861, 9866, 9867, 9870-9874, 9891, 9895-9897, 9910, 9920, 9931	C000-C809
(c) Chronic myeloproliferative diseases	9863, 9875, 9876, 9950, 9960-9964	C000-C809
(d) Myelodysplastic syndrome and other myeloproliferative diseases	9945, 9946, 9975, 9980, 9982-9987, 9989	C000-C809
(e) Unspecified and other specified leukaemias	9800, 9801, 9805, 9860, 9930	C000-C809
II. Lymphomas and reticuloendothelial neoplasms		
(a) Hodgkin lymphomas	9650-9655, 9659, 9661-9665, 9667	C000-C809
(b) Non-Hodgkin lymphomas (except Burkitt lymphoma)	9591, 9670, 9671, 9673, 9675, 9678-9680, 9684, 9689-9691, 9695, 9698-9702, 9705, 9708, 9709, 9714, 9716-9719, 9727-9729, 9731-9734, 9760-9762, 9764-9769, 9970	C000-C809
(c) Burkitt lymphoma	9687	C000-C809
(d) Miscellaneous lymphoreticular neoplasms	9740-9742, 9750, 9754-9758	C000-C809
(e) Unspecified lymphomas	9590, 9596	C000-C809
III. Central Nervous System and miscellaneous intracranial and intraspinal neoplasms		
(a) Ependymomas and choroid plexus tumour	9383, 9390-9394	C000-C809
(b) Astrocytomas	9380	C723
	9384, 9400-9411, 9420, 9421-9424, 9440-9442	C000-C809
(c) Intracranial and intraspinal embryonal tumours	9470-9474, 9480, 9508	C000-C809
	9501-9504	C700-C729
(d) Other gliomas	9380	C700-C722,C724-C729, C751, C753
	9381, 9382, 9430, 9444, 9450, 9451, 9460	C000-C809
(e) Other specified intracranial and intraspinal neoplasms	8270-8281, 8300, 9350-9352, 9360-9362, 9412, 9413, 9492, 9493, 9505-9507, 9530-9539, 9582	C000-C809
(f) Unspecified intracranial and intraspinal neoplasms	8000-8005	C700-C729, C751-C753
IV. Neuroblastoma and other peripheral nervous cell tumours		
(a) Neuroblastoma and ganglioneuroblastoma	9490, 9500	C000-C809
(b) Other peripheral nervous cell tumours	8680-8683, 8690-8693, 8700, 9520-9523	C000-C809
	9501-9504	C000-C699, C739-C768, C809
V. Retinoblastoma	9510-9514	C000-C809

Table AII (cont.) International Classification of Childhood Cancer, Third Edition[5]

Diagnostic group/subgroup	ICD-O-3 histology	ICD-O-3 site
VI. Renal tumours		
(a) Nephroblastoma and other non-epithelial renal tumours	8959, 8960, 8964-8967	C000-C809
	8963, 9364	C649
(b) Renal carcinomas	8010-8041, 8050-8075, 8082, 8120-8122, 8130-8141, 8143, 8155, 8190-8201, 8210, 8211, 8221-8231, 8240, 8241, 8244-8246, 8260-8263, 8290, 8310, 8320, 8323, 8401, 8430, 8440, 8480-8490, 8504, 8510, 8550, 8560-8576	C649
	8311, 8312, 8316-8319, 8361	C000-C809
(c) Unspecified malignant renal tumours	8000-8005	C649
VII. Hepatic tumours		
(a) Hepatoblastoma	8970	C000-C809
(b) Hepatic carcinomas	8010-8041, 8050-8075, 8082, 8120-8122, 8140, 8141, 8143, 8155, 8190-8201, 8210, 8211, 8230, 8231, 8240, 8241, 8244-8246, 8260-8264, 8310, 8320, 8323, 8401, 8430, 8440, 8480-8490, 8504, 8510, 8550, 8560-8576	C220, C221
	8160-8180	C000-C809
(c) Unspecified malignant hepatic tumours	8000-8005	C220, C221
VIII. Malignant bone tumours		
(a) Osteosarcomas	9180-9187, 9191-9195, 9200	C400-C419, C760-C768, C809
(b) Chondrosarcomas	9210, 9220, 9240	C400-C419, C760-C768, C809
	9221, 9230, 9241-9243	C000-C809
(c) Ewing tumour and related sarcomas of bone	9260	C400-C419, C760-C768, C809
	9363-9365	C400-C419
(d) Other specified malignant bone tumours	8810, 8811, 8823, 8830	C400-C419
	8812, 9250, 9261, 9262, 9270-9275, 9280-9282, 9290, 9300-9302, 9310-9312, 9320-9322, 9330, 9340-9342, 9370-9372	C000-C809
(e) Unspecified malignant bone tumours	8000-8005, 8800, 8801, 8803-8805	C400-C419
IX. Soft tissue & other extraosseous sarcomas		
(a) Rhabdomyosarcomas	8900-8905, 8910, 8912, 8920, 8991	C000-C809
(b) Fibrosarcomas, peripheral nerve sheath tumours, and other fibrous neoplasms	8810, 8811, 8813-8815, 8821, 8823, 8834-8835	C000-C399, C440-C768, C809
	8820, 8822, 8824-8827, 9150, 9160, 9491, 9540-9571, 9580	C000-C809
(c) Kaposi sarcoma	9140	C000-C809

Table AII (cont.) International Classification of Childhood Cancer, Third Edition[5]

Diagnostic group/subgroup	ICD-O-3 histology	ICD-O-3 site
(d) Other specified soft tissue sarcomas	8587, 8710-8713, 8806, 8831-8833, 8836, 8840-8842, 8850-8858, 8860-8862, 8870, 8880, 8881, 8890-8898, 8921, 8982, 8990, 9040-9044, 9120-9125, 9130-9133, 9135, 9136, 9141, 9142, 9161, 9170-9175, 9231, 9251, 9252, 9373, 9581	C000-C809
	8830	C000-C399, C440-C768, C809
	8963	C000-C639, C659-C699, C739-C768, C809
	9180, 9210, 9220, 9240	C490-C499
	9260	C000-C399, C470-C759
	9364	C000-C399, C470-C639, C659-C699, C739-C768, C809
	9365	C000-C399, C470-C639, C659-C768, C809
(e) Unspecified soft tissue sarcomas	8800-8805	C000-C399, C440-C768, C809
X. Germ cell tumours, trophoblastic tumours, and neoplasms of gonads		
(a) Intracranial and intraspinal germ cell tumours	9060-9065, 9070-9072, 9080-9085, 9100, 9101	C700-C729, C751-C753
(b) Malignant extracranial and extragonadal germ cell tumours	9060-9065, 9070-9072, 9080-9085, 9100-9105	C000-C559, C570-C619, C630-C699, C739-C750, C754-C768, C809
(c) Malignant gonadal germ cell tumours	9060-9065, 9070-9073, 9080-9085, 9090, 9091, 9100, 9101	C569, C620-C629
(d) Gonadal carcinomas	8010-8041, 8050-8075, 8082, 8120-8122, 8130-8141, 8143, 8190-8201, 8210, 8211, 8221-8241, 8244-8246, 8260-8263, 8290, 8310, 8313, 8320, 8323, 8380-8384, 8430, 8440, 8480-8490, 8504, 8510, 8550, 8560-8573, 9000, 9014, 9015	C569, C620-C629
	8441-8444, 8450, 8451, 8460-8473	C000-C809
(e) Other and unspecified malignant gonadal tumours	8590-8671	C000-C809
	8000-8005	C569, C620-C629
XI. Other malignant epithelial neoplasms and malignant melanomas		
(a) Adrenocortical carcinomas	8370-8375	C000-C809
(b) Thyroid carcinomas	8010-8041, 8050-8075, 8082, 8120-8122, 8130-8141, 8190, 8200, 8201, 8211, 8230, 8231, 8244-8246, 8260-8263, 8290, 8310, 8320, 8323, 8430, 8440, 8480, 8481, 8510, 8560-8573	C739
	8330-8337, 8340-8347, 8350	C000-C809
(c) Nasopharyngeal carcinomas	8010-8041, 8050-8075, 8082, 8083, 8120-8122, 8130-8141, 8190, 8200, 8201, 8211, 8230, 8231, 8244-8246, 8260-8263, 8290, 8310, 8320, 8323, 8430, 8440, 8480, 8481, 8500-8576	C110-C119

Table AII (cont.) International Classification of Childhood Cancer, Third Edition[5]

Diagnostic group/subgroup	ICD-O-3 histology	ICD-O-3 site
(d) Malignant melanomas	8720-8780, 8790	C000-C809
(e) Skin carcinomas	8010-8041, 8050-8075, 8078, 8082, 8090-8110, 8140, 8143, 8147, 8190, 8200, 8240, 8246, 8247, 8260, 8310, 8320, 8323, 8390-8420, 8430, 8480, 8542, 8560, 8570-8573, 8940, 8941	C440-C449
(f) Other and unspecified carcinomas	8010-8084, 8120-8157, 8190-8264, 8290, 8310, 8313-8315, 8320-8325, 8360, 8380-8384, 8430-8440, 8452-8454, 8480-8586, 8588-8589, 8940, 8941, 8983, 9000, 9010-9016, 9020, 9030	C000-C109, C129-C218, C239-C399, C480-C488, C500-C559, C570-C619, C630-C639, C659-C729, C750-C768, C809
XII. Other and unspecified malignant neoplasms		
(a) Other specified malignant tumours	8930-8936, 8950, 8951, 8971-8981, 9050-9055, 9110	C000-C809
	9363	C000-C399, C470-C759
(b) Other unspecified malignant tumours	8000-8005	C000-C218, C239-C399, C420-C559, C570-C619, C630-C639, C659-C699, C739-C750, C754-C809

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