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The incidence of childhood cancer in New Zealand 2010 - 2014

A report from
The New Zealand Children's Cancer Registry

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Contents

Executive Summary

When the New Zealand Children's Cancer Registry (NZCCR) published our 2000-2009 incidence report in 2014,[1] we envisaged that it would be the first of many reports to our patients and their families, clinicians, researchers, and other stakeholders. Here we report our analysis of new NZCCR registrations for the subsequent five year period of between 2010 and 2014. To aid comparisons we have replicated our previous report wherever possible, including grouping according to the International Classification of Childhood Cancers (ICCC), age group, sex, and ethnicity.

There were 762 new childhood cancer registrations in the five years between 2010 and 2014, giving an overall New Zealand child cancer incidence rate of 167 per million per year (age standardised to the World Standard Population). Overall incidence is not significantly higher than New Zealand's 2000-2009 incidence rate (IR) of 155 per million per year but there has, however, been a noticeable increase in incidence for boys (from 159 to 183 per million) and for 0-4 year olds (from 202 to 244 per million). These increases can be attributed, in part, to the recent re-classification of Langerhans cell histiocytosis as a malignancy, which more frequently occurs in males and in those under the age of five.

Overall cancer incidence among Maori children (IR: 160 per million, 95% CI: 137-183) is comparable with incidence among non-Maori (IR: 170 per million, 95% CI: 156-184). One in four (24%) of children diagnosed with cancer in the 2010-2014 were of prioritised Maori ethnicity.

Consistent with the 2000-2009 analysis, the most common cancers diagnosed in New Zealand children between 2010 and 2014 were leukaemias (one third of all cancers, 51 cases per year) and central nervous system tumours (21% of all cancers, 32 cases per year). Acute lymphoblastic leukaemias (ALL) accounted for over one in four of all new child cancer registrations within the study period. Haematological malignancies (i.e. leukaemias and lymphomas) accounted for half of all cancers diagnosed in boys but just 37% of girls.

By age group, children aged 0-4 years were significantly more likely to be diagnosed with cancer (382 cases, IR: 244 per million) than those aged 5-9 years (180 cases, 124 per million) or 10-14 years (197 cases, 132 per million). Half of all childhood cancers in the period were diagnosed in children under the age of five, compared to 44% in the 2000-2009 period. In accordance with our 2000-2009 report, we identified a significantly higher incidence of ALL, neuroblastoma, retinoblastoma and renal tumours among 0-4 year olds while malignant bone tumours and 'other epithelial neoplasms', such as melanomas, were most commonly diagnosed in older children aged 10-14 years.

The best way is improve the quality of registry data is through its continual use. The NZCCR is able to provide comprehensive and accurate diagnostic and treatment information because it is accessed on a daily basis for individual patient care by Late Effects Assessment Programme clinicians and because we cooperate with the New Zealand Cancer Registry (NZCR) to validate the data held by each registry. Our collaboration with the NZCR to verify our 2010-2014 registrations showed that the NZCCR's case ascertainment had increased since 2000-2009 to 94% and also identified further improvements that could be made to New Zealand's paediatric oncology referral pathways and our own registration practices. With two registries covering childhood cancers, New Zealand is uniquely positioned to conduct such activities and we thank the NZCR for their valuable contribution to this report.

1 Introduction

1.1 Childhood cancer in New Zealand

Although child cancers account for less than one percent of all cancers diagnosed each year cancer is nevertheless a significant health issue for New Zealand.[2] Cancer remains the second most common cause of death, after traffic accidents, for 1-14 year olds[3] and it is estimated that one in five hundred children will be diagnosed with cancer before the age of 15 years.[4] 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 paediatric oncology 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 NZCR,[5] the data collected for each patient is necessarily limited. 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)[6] which allows New Zealand child cancer incidence and survival to be directly compared with international benchmarks. The NZCCR is now under the governance of the POSG's successor, the National Child Cancer Network (NCCN). It holds verified demographic and treatment information for all New Zealand children diagnosed with cancer since January 2000.

The NZCCR serves multiple functions. At an individual patient level, the information collected and held by the registry is later accessed and updated 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 oncology/haematology involvement for diseases not currently included in the ICCC-3, are excluded from any overall analyses.

The criteria for inclusion in 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*

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[7] 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).[8] 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[6] 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[5] 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.[9] According to the ICCC-3[6] 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.[5] The inclusion of non-malignant intracranial and intraspinal neoplasms mostly concerns 'diagnostic group III: central nervous system and miscellaneous intracranial and intraspinal neoplasms', but occasionally a child may be diagnosed with a benign intracranial/intraspinal germ cell tumour (diagnostic group Xa) which also meets ICCC-3 criteria for cancer registration.

Table 1.5 Diagnostic groups of the International Classification of Childhood Cancers, 3rd edition (ICCC-3)[6]

Diagnostic 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 an updated analysis of the incidence of childhood cancer in New Zealand for the period of the 1st of January 2010 to the 31st of December 2014. In order to aid comparisons, the methodology and reporting structure replicates the previously published 2000-2009 report[1] wherever possible. As this was a smaller cohort than for the previous analysis, incidence was only reported for two prioritised ethnic groups; Maori and non-Maori. It is our intention to conduct future analyses with a larger cohort specifically to look at ethnic variations in childhood cancer and assess survival by ethnicity.

Chapter 3 contains incidence data for children aged less than 15 years first diagnosed with cancer between the 1st of January 2010 and the 31st December 2014. Cancer incidence is reported by ICCC-3 diagnostic group and subgroup according to sex, age at diagnosis, and prioritised ethnicity. It is expressed in terms of the raw number of new cases diagnosed in the period, the proportion of total cancers reported, and the age-standardised incidence rate. Incidence rates have been age-standardised to the World Standard Population, and are expressed per million population per year. The 95% confidence interval, which expresses the degree of accuracy associated with the estimated incidence rate, is also reported in each table. Incidence rates (and corresponding 95% confidence intervals) were censored where there were only a small number of cases diagnosed within the time period. Further details regarding the methodology used in this report is provided in Chapter 2.

Chapter 4 provides an overview of the incidence pertaining to each ICCC-3 diagnostic group and for 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. Finally, the NZCCR and NZCR collaborated to validate all registrations held by the NZCCR and the NZCR for the study period.

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 is 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 included on the NZCCR but not the NZCR. Also, the NZCCR adopted the first revision of the ICD-O-3[9] on the 1/1/2010 while the NZCR delayed implementation until 1/1/2014. 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 by 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. 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.[11]

In the previously published ten year analysis, three prioritised ethnicity groupings were used: Maori, Pacific Peoples and non-Maori/non-Pacific Peoples. Due to the small number of cases recorded over this five year period for Pacific children (an average of 15 cases per year), it was decided to include only prioritised Maori and Non-Maori in this report (see Table 2.3.2). The NZCCR Working Group acknowledge the importance of identifying ethnic variations in cancer incidence in New Zealand's child population, and this will be addressed in future analyses with a larger cohort.

Table 2.2. Population by age group and sex in New Zealand, 2010-2014 Census

Prioritised ethnicity	Population base[a]	%
Maori	232 050	25.5
Non-Maori	677 656	74.5
Total	**909 700[b]**	**100.0**

[a] Average population estimate as at 30 June 2010-2014, Stats NZ
[b] Totals do not due to the rounding of annual estimates

2.3 Incidence calculations

Incidence is defined as the number of new primary cancer cases diagnosed in a specified population, for example 0-4 year olds or males aged 0-14 years, during a specified time period (usually one year). Due to the relatively small number of cases diagnosed annually, cancer incidence is usually expressed as a rate per 100,000 or 1,000,000 population per year.

Incidence should not be confused with prevalence, which is defined as the number of people currently alive with a particular condition. Given that the treatment duration for many cancers is up to three years, the number of children receiving active treatment in New Zealand at any given year will be higher than the number of new cancer cases.

2.3.1 Age-specific incidence

Age-specific rates provide information on the cancer incidence in an age group relative to the total number of people at risk in that age group. Age-specific incidence rates for each year are calculated simply by dividing the number of cases diagnosed each year for each age group by the population for that same period. We have used the estimated resident population (as at June 30) reported annually by Statistics New Zealand.

$$\text{Incidence rate for period} = \frac{\text{Number of new cases over the specified period}}{\text{Person-years at risk over period}} \times 1{,}000{,}000$$

2.3.2 Age-standardised incidence

Since the risk of cancers varies by age group, it is common practice to age-standardise incidence rates to allow for more valid comparisons over time or between populations that have different age structures. The incidence rates presented in this report have been calculated by the direct age-standardisation method, where the age-specific incidence rates are multiplied by a standard population. The age-standardised rates reported throughout this document have been age-standardised to the World Standard population. Table 2.3.2 compares New Zealand's population with the World Standard Population. All age-standardised incidence calculations were conducted using SAS® software v9.3.

Confidence intervals for incidence rates were calculated assuming the cases were drawn from a Poisson distribution. As rates based on small numbers may be distorted due to random fluctuations, age standardised incidence rates were censored in sections 3 and 4 for rare diagnostic subgroups where there were fewer than ten cases reported.

Table 2.3.2 Population by age group and sex in New Zealand, 2010-2014

	Total[a]	NZ population weightings	World standard population weightings[b]
0-4 years	313 654	34.5	33.9
5-9 years	296 392	32.6	33.2
10-14 years	299 654	32.9	32.9
Total children 0-14 years	**909 700**	**100.0**	**100.0**

[a] Average population estimate as at 30 June 2010-2014, Stats NZ
[b] Retrieved from: https://seer.cancer.gov.stdpopulations/world.who.html

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 age-standardised rate 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.

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 five 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.**

3 Childhood Cancer Incidence

3.1 Overall incidence of childhood cancer, 2010-2014

There were 762 new childhood cancer registrations in the 5 years between 2010 and 2014 out of a total average resident child population of 909,700, giving an overall New Zealand child cancer age standardised incidence rate of 166.8 per million per year (95% CI: 155.0-178.7, see Table 3.1). This is at the upper end of what was reported in the International Incidence of childhood cancer, 2001-2010 for Oceania (152.9 per million) and other high income populations (e.g. between 143.8 and 170.9 per million for European regions).[12]

Although overall incidence is not significantly higher than New Zealand's 2000-2009 child cancer incidence rate of 154.5 per million per year (95% CI: 146.1-162.8) which we reported previously,[1] there are two groups where there have been noticeable increases in annual numbers. Firstly, between 2000-2009 and 2010-2014 the incidence rate for boys increased from 158.5 per million (95% CI: 146.7-170.4) to 183.2 per million (95% CI: 165.8-200.5). No such increases were seen for girls in these two time periods, with incidence rates stable at 150.2 (95% CI: 138.3-162.0) and 149.6 per million (95% CI: 133.5-165.7) respectively. Secondly, there was a significant increase in cancer incidence among the 0-4 year old population between the two time periods, from 202.4 per million in 2000-2009 (95% CI: 186.0-218.8) to 243.6 per million in 2010-2014 (95% CI: 219.2-268.0). Between 2000 and 2009, 43.9% of all child cancer cases were diagnosed in children under five years of age. In the 2010-2014 period, cancers in under five year olds now accounted for half (50.1%) of all childhood malignancies.

Overall cancer incidence among Maori children (IR: 160.3 per million, 95% CI: 137.3-183.3) is comparable with incidence among non-Maori (IR: 170.0 per million, 95% CI: 156.1-183.9). Nearly one in four (24.4%) children diagnosed with cancer were of prioritised Maori ethnicity, consistent with the 25.5% of children in the general population who were prioritised Maori in this period according to annual population estimates.

Table 3.1 Childhood cancer incidence by sex, age group, and ethnicity, New Zealand, 2000-2014

	2000-2009				2010-2014			
	Average cases per year (%)		Age standardised incidence (95% CI)		Average cases per year (%)		Age standardised incidence (95% CI)	
Sex								
Male	70.1	52.7	158.5	(146.7 - 170.4)	85.8	56.3	183.2	(165.8 - 200.5)
Female	62.8	47.3	150.2	(138.3 - 162.0)	66.6	43.7	149.6	(133.5 - 165.7)
Age group								
0-4 years	58.3	43.9	202.4	(186.0 - 218.8)	76.4	50.1	243.6	(219.2 - 268.0)
5-9 years	35.0	26.3	119.4	(106.9 - 132.0)	36.6	24.0	123.5	(105.6 - 141.4)
10-14 years	39.6	29.8	129.4	(116.7 - 142.2)	39.4	25.9	131.5	(113.1 - 149.8)
Prioritised Ethnicity								
Maori	26.2	19.7	134.3	(117.9 - 150.7)	37.2	24.4	157.7	(135.0 - 180.4)
Non-Maori	106.7	80.3	[b]	[b]	115.2	75.6	170.1	(156.2 - 184.0)
Total	**132.9**	**100.0**	**154.5**	**(146.1 - 162.8)**	**152.4**	**100.0**	**166.8**	**(155.0 - 178.7)**

[a] Incidence by sex and age group is based on an average of the estimated New Zealand resident population as at June 30 for the years 2010-2014 published by Statistics New Zealand.
[b] Incidence for non-Maori cannot be directly compared between the two time periods due to methodological differences in prioritised ethnicity groupings

3.2 Annual number of new child cancer registrations

Caution should be taken when assessing changes in New Zealand's child cancer incidence over time. Firstly, given the rarity of childhood cancer and New Zealand's small population, there can be considerable natural fluctuations in the number of new cases diagnosed each year. Secondly, any increases might simply reflect advances in diagnostic testing, leading to the early identification of cancers, such as neuroblastoma, which may have otherwise regressed spontaneously. Also, variations in annual child cancer registrations reflect changes in registration practices. On the 1st January 2010, the NZCCR adopted the revised 3rd edition of the International Classification of Diseases of Oncology (ICD-O-3-1) and Langerhans cell histiocytosis, which is diagnosed in approximately five children in New Zealand each year, was re-classified from a 'tumour of uncertain behaviour' to a malignancy. In addition, a small number of other rare tumours, such as carcinoid tumour of the appendix, also were recorded as a malignancy for the first time.

In the period 2000 to 2014 the number of new child cancer registrations ranged from 116 to 167 cases per year (see Table 3.2). The number of new cases diagnosed each year is considerably less than the number of children who underwent cancer treatment, as the standard cancer treatment course for many cancers, such as acute lymphoblastic leukaemia (ALL), is up to three years duration.

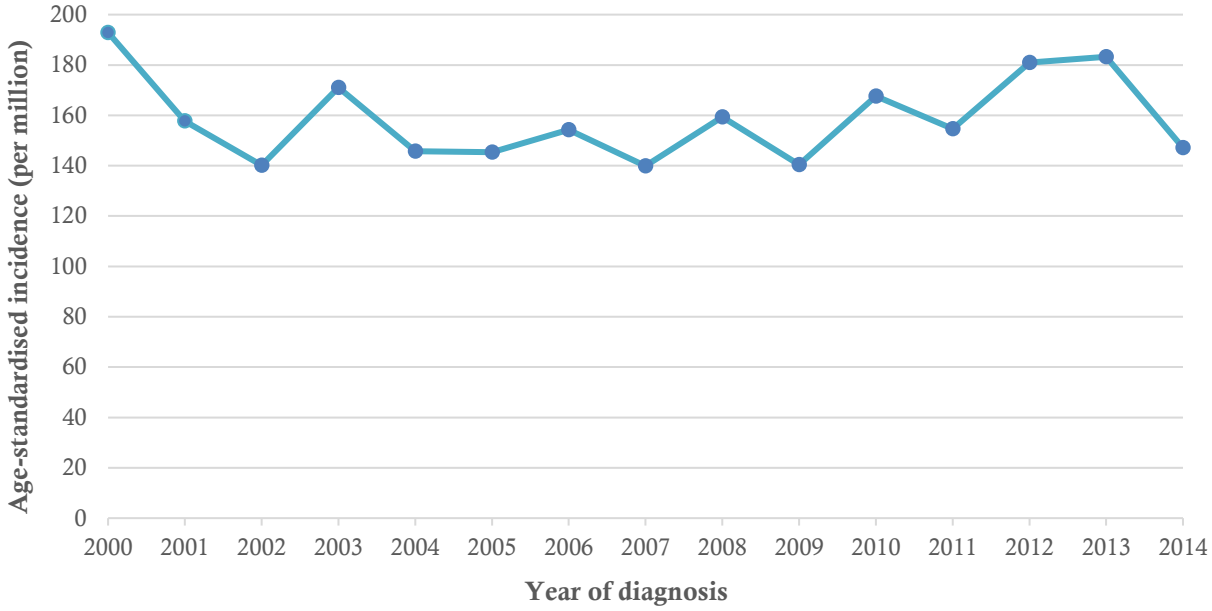
The International Incidence of Childhood Cancer 2001-2010, which the NZCCR contributed to, found that since the 1980s the global incidence rate for cancers among those aged 0-14 years increased by 13%.[12] However, as highlighted above, we must be cautious when interpreting New Zealand's small numbers. Although Figure 3.2 provides some evidence that incidence rates are trending upwards over time - with high incidence recorded in 2012 and 2013 - childhood cancer incidence dropped again in 2014 and the highest incidence rate for the 15 year period was back in the year 2000.

Table 3.2 Annual number of child cancer registrations in New Zealand, 2000-2014

Year	Male			Female			Total		
	No. of cases	Population	Age standardised rate[a]	No. of cases	Population	Age standardised rate[a]	No. of cases	Population	Age standardised rate[a]
2000	97	451 210	229.9	62	427 540	153.8	159	878 750	192.9
2001	73	450 400	165.6	62	426 820	149.1	135	877 220	157.7
2002	66	452 910	154.5	50	430 660	125.0	116	883 570	140.1
2003	75	456 270	167.5	73	433 740	174.6	148	890 010	171.0
2004	55	457 720	125.8	71	435 360	166.8	126	893 080	145.8
2005	62	456 330	134.9	64	433 920	156.2	126	890 250	145.4
2006	73	454 960	166.3	59	433 360	141.7	132	888 320	154.3
2007	57	455 080	128.8	64	433 360	151.5	121	888 440	139.9
2008	63	455 770	137.0	79	433 640	182.7	142	889 410	159.3
2009	80	456 900	176.3	44	434 310	102.7	124	891 210	140.4
2010	99	465 750	211.6	54	442 390	121.3	153	908 140	167.6
2011	78	466 730	165.3	64	443 970	143.5	142	910 700	154.7
2012	89	466 500	189.3	77	443 330	172.3	166	909 830	181.0
2013	94	465 900	201.4	73	442 840	164.2	167	908 740	183.2
2014	69	467 050	147.7	65	444 040	146.4	134	911 090	147.1

[a] Rate per million population per year, age-standardised to the World Standard Population

Figure 3.3 Age standardised child cancer incidence in New Zealand, 2000-2014



3.3 Annual age-specific incidence of childhood cancers by age group

Table 3.3 shows the annual age-specific incidence rates by age group. The annual age-specific incidence rates fluctuated considerably, ranging from 166.9 to 303.4 per million per year for the 0-4 year age group, 82.2 to 150.0 per million for the 5-9 year age group, and 87.7 per million to 179.0 per million for children aged 10-14 years.

Table 3.3 Annual age-specific incidence (per million) of childhood cancers by age group, New Zealand, 2000-2014

	0-4 years			5-9 years			10-14 years		
	Cases	Population base[a]	Age-specific incidence rate (per million)	Cases	Population base[a]	Age-specific incidence rate (per million)	Cases	Population base[a]	Age-specific incidence rate (per million)
2000	86	283 420	303.4	41	302 020	135.8	32	293 310	109.1
2001	55	281 000	195.7	37	295 470	125.2	43	300 750	143.0
2002	61	281 130	217.0	28	294 400	95.1	27	308 040	87.7
2003	60	281 850	212.9	42	294 870	142.4	46	313 290	146.8
2004	53	284 660	186.2	36	293 370	122.7	37	315 050	117.4
2005	50	284 320	175.9	39	292 400	133.4	37	313 530	118.0
2006	64	286 000	223.8	24	291 880	82.2	44	310 440	141.7
2007	52	292 390	177.8	34	289 910	117.3	35	306 140	114.3
2008	51	300 060	170.0	37	287 700	128.6	54	301 650	179.0
2009	51	305 510	166.9	32	288 150	111.1	41	297 550	137.8
2010	76	314 350	241.8	37	290 790	127.2	40	303 000	132.0
2011	70	317 460	220.5	31	290 560	106.7	41	302 680	135.5
2012	92	315 730	291.4	35	294 100	119.0	39	300 000	130.0
2013	82	311 920	262.9	45	300 060	150.0	40	296 760	134.8
2014	62	308 810	200.8	35	306 450	114.2	37	295 830	125.1

[a] Estimated New Zealand resident population as at June 30, Statistics New Zealand

3.4 Childhood cancer incidence by diagnostic group

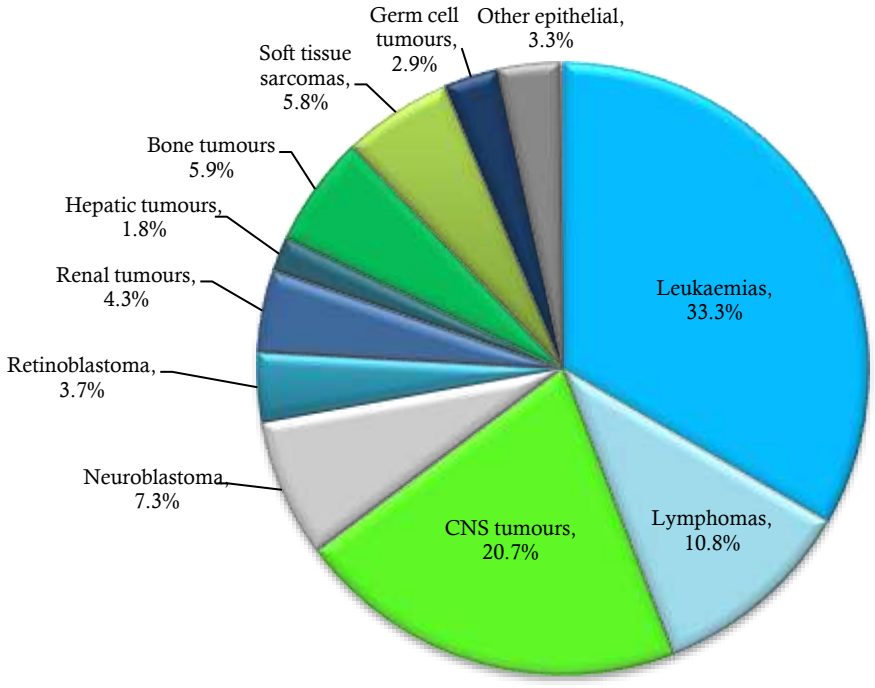
Between 2010 and 2014, leukaemias accounted for exactly one in three of every childhood cancer diagnosed, with an average of 50.8 cases per year (see Table 3.4 and Figure 3.4). Tumours of the central nervous system tumours were the next most frequently diagnostic class of tumours, accounting for around one in five childhood cancers diagnosed. 'Lymphomas & reticuloendothelial neoplasms' accounted for 10.8% of all tumours, up from 8.6% in 2000-2009, reflecting the reclassification of LCH as a malignancy which came into effect at the start of 2010.

Table 3.4 Childhood cancer incidence by diagnostic group, New Zealand, 2010-2014

	Diagnostic Group (ICCC-3)	Average cases per year	%	Age standardised rate[a] (95% CI)	
I.	Leukaemias, myeloproliferative & myelodysplastic diseases	50.8	33.3	55.5	(48.7 - 62.4)
II.	Lymphoma & reticuloendothelial neoplasms	16.4	10.8	18.0	(14.1 - 21.9)
III.	Central nervous system & intracranial/intraspinal neoplasms	31.6	20.7	34.7	(29.3 - 40.2)
IV.	Neuroblastoma & other peripheral nervous cell tumours	11.2	7.3	12.2	(9.0 - 15.3)
V.	Retinoblastoma	5.6	3.7	6.1	(3.8 - 8.3)
VI.	Renal tumours	6.6	4.3	7.2	(4.7 - 9.6)
VII.	Hepatic tumours	2.8	1.8	3.1	(1.5 - 4.7)
VIII.	Malignant bone tumours	9.0	5.9	9.9	(7.0 - 12.8)
IX.	Soft tissue and other extraosseous sarcomas	8.8	5.8	9.7	(6.8 - 12.5)
X.	Germ cell tumours, trophoblastic tumours & neoplasms of gonads	4.4	2.9	4.8	(2.8 - 6.8)
XI.	Other epithelial neoplasms & melanomas	5.0	3.3	5.5	(3.4 - 7.7)
XII.	Other & unspecified malignant neoplasms	0.2	0.1	0.2	(0.0 - 0.7)
	All childhood cancers	**152.4**	**100**	**166.8**	**(155.0 - 178.7)**

[a] Rate per million population per year, age-standardised to the World Standard Population

Figure 3.4 Childhood cancer by diagnostic group, New Zealand, 2010-2014



3.5 Childhood cancer incidence by diagnostic subgroup

With an average of 38 cases diagnosed each year, acute lymphoblastic leukaemia is the most common childhood cancer, accounting for one in four new cancer cases (24.7%) registered from 2010 to 2014 (see Table 3.5). The other ICCC-3 diagnostic subgroups most frequently diagnosed in New Zealand children were astrocytomas and neuroblastoma (both 11 cases per year, 7.3% of all cancer registrations for the period). Acute myeloid leukaemias and the tumour group 'intracranial and intraspinal embryonal tumours' (which includes medulloblastoma) each accounted for 5% of cancers diagnosed.

Table 3.5 Childhood cancer incidence by diagnostic group and subgroup, New Zealand, 2010-2014

	ICCC-3 Diagnostic Group / Subgroup	Cases per year	%	Age standardised rate[a] (95% CI)	
I.	**Leukaemias, myeloproliferative & myelodysplastic diseases**	**50.8**	**33.3**	**55.5**	**(48.7 - 62.4)**
Ia.	Lymphoid leukaemias	37.6	24.7	41.1	(35.2 - 47.0)
Ib.	Acute myeloid leukaemias	8.0	5.2	8.8	(6.1 - 11.5)
Ic.	Chronic myeloproliferative diseases	0.8	0.5	[b]	[b]
Id.	Other myeloproliferative diseases	2.4	1.6	2.6	(1.1 - 4.1)
Ie.	Other and unspecified leukaemia	2.0	1.3	2.2	(0.8 - 3.5)
II.	**Lymphoma & reticuloendothelial neoplasms**	**16.4**	**10.8**	**18.0**	**(14.1 - 21.9)**
IIa.	Hodgkin lymphomas	6.0	3.9	6.6	(4.3 - 9.0)
IIb.	Non-Hodgkin lymphomas (excl. Burkitt lymphomas)	3.2	2.1	3.5	(1.8 - 5.3)
IIc.	Burkitt lymphomas	2.2	1.4	2.4	(1.0 - 3.8)
IId.	Miscellaneous lymphoreticular neoplasms	5.0	3.3	5.4	(3.3 - 7.6)
IIe.	Unspecified lymphomas	-	-	-	-
III.	**Central nervous system & intracranial/intraspinal neoplasms**	**31.6**	**20.7**	**34.7**	**(29.3 - 40.2)**
IIIa.	Ependymomas and choroid plexus tumours	3.6	2.4	3.9	(2.1 - 5.7)
IIIb.	Astrocytomas	11.2	7.3	12.3	(9.1 - 15.6)
IIIc.	Intracranial and intraspinal embryonal tumours	7.6	5.0	8.4	(5.7 - 11.0)
IIId.	Other gliomas	4.6	3.0	5.1	(3.0 - 7.2)
IIIe.	Other specified intracranial and intraspinal neoplasms	3.2	2.1	3.5	(1.8 - 5.2)
IIIf.	Unspecified intracranial and intraspinal neoplasms	1.4	0.9	[b]	[b]
IV.	**Neuroblastoma & other peripheral nervous cell tumours**	**11.2**	**7.3**	**12.2**	**(9.0 - 15.3)**
IVa.	Neuroblastoma & ganglioneuroblastoma	11.2	7.3	12.2	(9.0 - 15.3)
IVb.	Other peripheral nervous cell tumours	-	-	-	-
V.	**Retinoblastoma**	**5.6**	**3.7**	**6.1**	**(3.8 - 8.3)**
VI.	**Renal tumours**	**6.6**	**4.3**	**7.2**	**(4.7 - 9.6)**
VIa.	Nephroblastoma & other non-epithelial renal tumours	6.6	4.3	7.2	(4.7 - 9.6)
VIb.	Renal carcinomas	-	-	-	-
VII.	**Hepatic tumours**	**2.8**	**1.8**	**3.1**	**(1.5 - 4.7)**
VIIa.	Hepatoblastoma	2.0	1.3	2.2	(0.8 - 3.5)
VIIb.	Hepatic carcinomas	0.8	0.5	[b]	[b]
VIIc.	Unspecified malignant hepatic tumours	-	-	-	-
VIII.	**Malignant bone tumours**	**9.0**	**5.9**	**9.9**	**(7.0 - 12.8)**
VIIIa.	Osteosarcomas	4.6	3.0	5.1	(3.0 - 7.1)
VIIIb.	Chondrosarcomas	-	-	-	-
VIIIc.	Ewing tumours & related bone sarcomas	3.4	2.2	3.8	(2.0 - 5.5)
VIIId.	Other specified malignant bone tumours	1.0	0.7	[b]	[b]
VIIIe.	Unspecified malignant bone tumours	-	-	-	-
IX.	**Soft tissue and other extraosseous sarcomas**	**8.8**	**5.8**	**9.7**	**(6.8 - 12.5)**
IXa.	Rhabdomyosarcomas	4.2	2.8	4.6	(2.6 - 6.6)
IXb.	Fibrosarcomas & other fibrous neoplasms	0.6	0.4	[b]	[b]
IXc.	Kaposi sarcomas	-	-	-	-
IXd.	Other specified soft tissue sarcomas	3.4	2.2	3.7	(2.0 - 5.5)
IXe.	Unspecified soft tissue sarcomas	0.6	0.4	[b]	[b]

[a] Rate per million population per year, age-standardised to the World Standard Population

[b] Age standardised rates (and corresponding 95% CI) have been censored for diagnostic subgroups where there were fewer than 10 cases diagnosed within the five-year period

Table 3.5 (cont.) Childhood cancer incidence by diagnostic group and subgroup, New Zealand, 2010-2014

	ICCC-3 Diagnostic Group / Subgroup	Total Cases	%	Age standardised rate[a] (95% CI)	
X.	**Germ cell tumours, trophoblastic tumours & neoplasms of gonads**	**4.4**	**2.9**	**4.8**	**(2.8 - 6.8)**
Xa.	Intracranial & intraspinal germ cell tumours	1.6	1.0	1.8	(0.5 - 3.0)
Xb.	Malignant extracranial & extragonadal germ cell tumours	1.4	0.9	1.5	(0.4 - 2.7)
Xc.	Malignant gonadal germ cell tumours	1.2	0.8	1.3	(0.3 - 2.4)
Xd.	Gonadal carcinomas	-	-	-	-
Xe.	Other & unspecified malignant gonadal tumours	0.2	0.1	[b]	[b]
XI.	**Other epithelial neoplasms & melanomas**	**5.0**	**3.3**	**5.5**	**(3.4 - 7.7)**
XIa.	Adrenocortical carcinomas	0.4	0.3	[b]	[b]
XIb.	Thyroid carcinomas	0.8	0.5	[b]	[b]
XIc.	Nasopharyngeal carcinomas	-	-	-	-
XId.	Melanomas	1.2	0.8	[b]	[b]
XIe.	Skin carcinomas	-	-	-	-
XIf.	Other & unspecified carcinomas	2.6	1.7	2.9	(1.3 - 4.4)
XII.	**Other & unspecified malignant neoplasms**	**0.2**	**0.1**	**0.2**	**(0.0 - 0.7)**
XIIa.	Other specified malignant tumours	0.2	0.1	[b]	[b]
XIIb.	Other unspecified malignant tumours	-	-	-	-
	All childhood cancers	**152.4**	**100**	**166.8**	**(155.0 - 178.7)**

[a] Rate per million population per year, age-standardised to the World Standard Population
[b] Age standardised rates (and corresponding 95% CI) have been censored for diagnostic subgroups where there were fewer than 10 cases diagnosed within the five-year period

3.6 Childhood cancer incidence by sex

3.6.1 Distribution of childhood cancers by sex

Figures 3.6.1a-3.6.1b show that blood cancers (the leukaemia and lymphoma diagnostic groups) accounted for half (49.7%) of all cancers diagnosed in boys in the 2010-2014 period compared to just 36.9% of all cases for girls. Boys were twice as likely to be diagnosed with 'lymphoma and lymphoreticular neoplasms' than girls (see Figure 3.6.1c).

Figure 3.6.1a Cancers diagnosed in males 0-14 years of age, New Zealand, 2010-2014

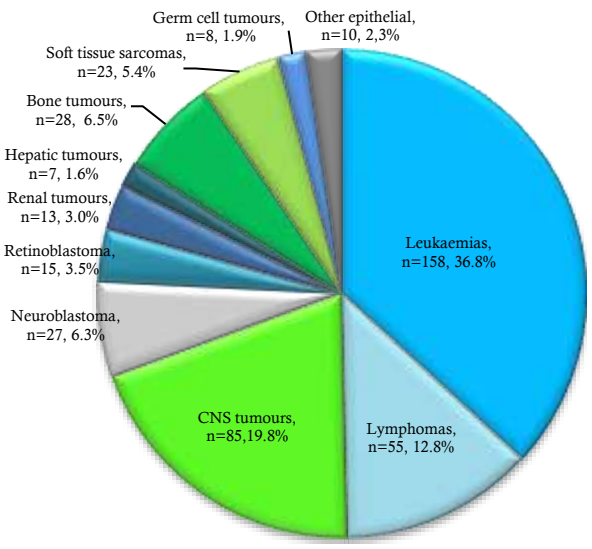


Figure 3.6.1b Cancers diagnosed in females 0-14 years of age, New Zealand, 2010-2014

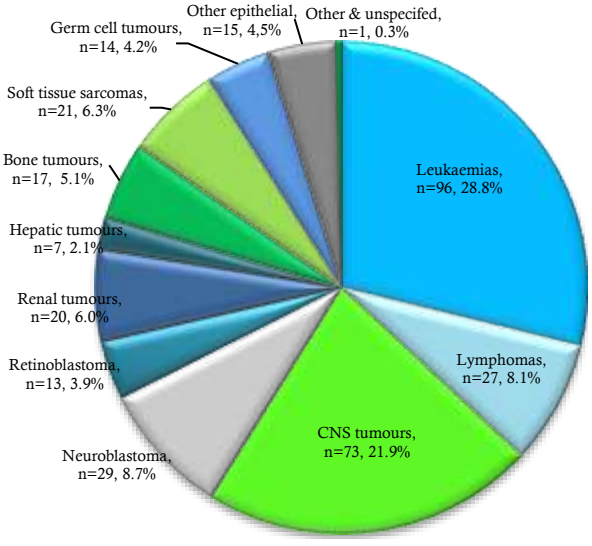
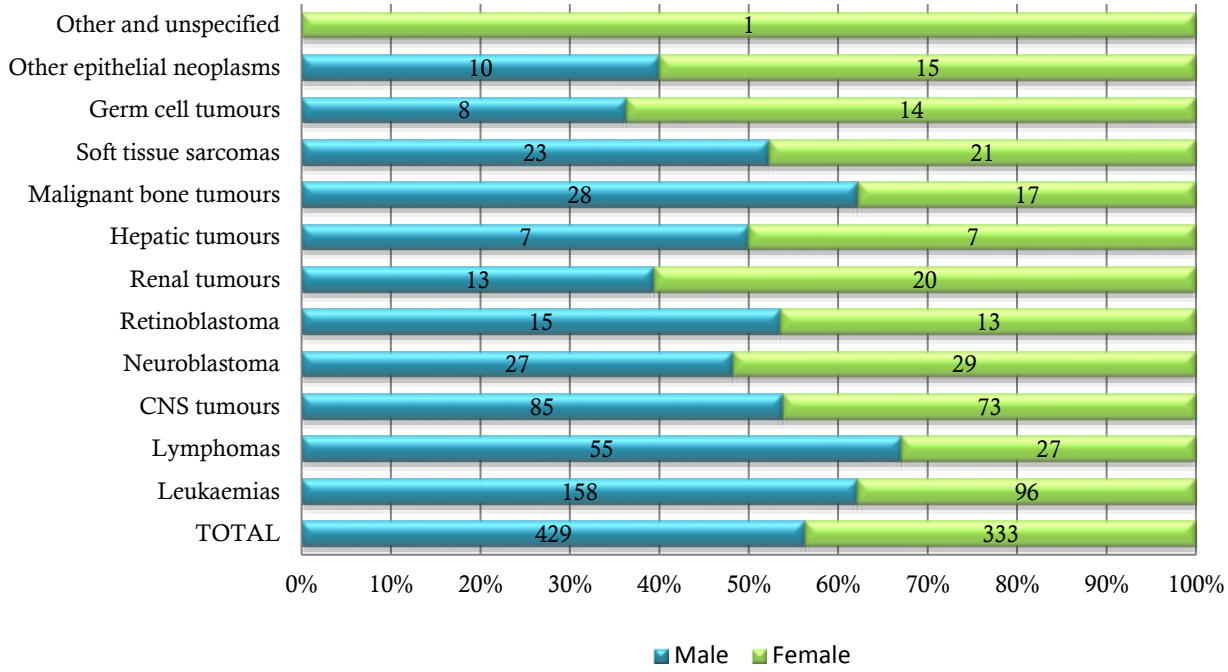


Figure 3.6.1c Number of childhood cancer cases by sex, New Zealand, 2010-2014



3.6.2 Childhood cancer incidence by sex and diagnostic group and subgroup

Overall, incidence rates were significantly higher in boys (IR: 183.2 per million, 95% CI: 165.8-200.5) than girls (IR 149.6 per million (IR: 133.5-165.7). This is in keeping with the 1.17 sex ratio reported for children internationally.[12] In the five-year period boys were significantly more likely to be diagnosed with leukaemias and lymphomas than girls. By diagnostic subgroup, boys were significantly more likely than girls to be diagnosed with lymphoid leukaemias and 'miscellaneous lymphoreticular neoplasms' (i.e. LCH); of the 25 new LCH registrations 20 of them (80%) were diagnosed in boys. Boys were also 1.6 times more likely, but not significantly so, to be diagnosed with a malignant bone tumour.

Table 3.6.2 Childhood cancer incidence by sex and diagnostic group and subgroup, New Zealand, 2000-2014

Diagnostic Group / Subgroup		Male			Female		
		Cases per year	Age standardised rate[a]		Cases per year	Age standardised rate[a]	
I. Leukaemias, myeloproliferative & myelodysplastic diseases		**31.6**	**67.4**	**(56.9 - 77.9)**	**19.2**	**43.1**	**(34.5 - 51.7)**
Ia.	Lymphoid leukaemias	23.6	50.3	(41.2 - 59.3)	14.0	31.4	(24.1 - 38.8)
Ib.	Acute myeloid leukaemias	4.8	10.2	(6.1 - 14.3)	3.2	7.2	(3.7 - 10.7)
Ic.	Chronic myeloproliferative diseases	0.6	[b]	[b]	0.2	[b]	[b]
Id.	Other myeloproliferative diseases	1.4	3.0	(0.8 - 5.2)	1.0	2.3	(0.3 - 4.2)
Ie.	Other and unspecified leukaemia	1.2	2.6	(0.5 - 4.6)	0.8	1.8	(0.0 - 3.5)
II. Lymphoma & reticuloendothelial neoplasms		**11.0**	**23.5**	**(17.3 - 29.8)**	**5.4**	**12.2**	**(7.6 - 16.7)**
IIa.	Hodgkin lymphomas	4.0	8.6	(4.8 - 12.4)	2.0	4.5	(1.7 - 7.3)
IIb.	Non-Hodgkin lymphomas (excl. Burkitt lymphomas)	1.4	3.0	(0.8 - 5.2)	1.8	4.1	(1.4 - 6.7)
IIc.	Burkitt lymphomas	1.6	3.4	(1.1 - 5.8)	0.6	1.3	(0.0 - 2.9)
IId.	Miscellaneous lymphoreticular neoplasms	4.0	8.5	(4.8 - 12.2)	1.0	2.2	(0.3 - 4.2)
IIe.	Unspecified lymphomas	0.0	-	-	-	-	-
III. CNS & intracranial/intraspinal neoplasms		**17.0**	**36.4**	**(26.7 - 44.2)**	**14.6**	**33.0**	**(25.4 - 40.5)**
IIIa.	Ependymomas and choroid plexus tumours	1.2	2.6	(0.5 - 4.6)	2.4	5.4	(2.3 - 8.4)
IIIb.	Astrocytomas	5.8	12.5	(7.9 - 17.0)	5.4	12.2	(7.6 - 16.8)
IIIc.	Intracranial and intraspinal embryonal tumours	5.8	12.4	(7.9 - 17.0)	1.8	4.1	(1.4 - 6.7)
IIId.	Other gliomas	1.8	3.9	(1.4 - 6.4)	2.8	6.3	(3.0 - 9.6)
IIIe.	Other specified intracranial and intraspinal neoplasms	1.6	3.4	(1.1 - 5.8)	1.6	3.6	(1.1 - 6.2)
IIIf.	Unspecified intracranial and intraspinal neoplasms	0.8	[b]	[b]	0.6	[b]	[b]
IV. Neuroblastoma & other peripheral nervous cell tumours		**5.4**	**11.5**	**(7.1 - 15.8)**	**5.8**	**12.9**	**(8.2 - 17.6)**
IVa.	Neuroblastoma & ganglioneuroblastoma	5.4	11.5	(7.1 - 15.8)	5.8	12.9	(8.2 - 17.6)
IVb.	Other peripheral nervous cell tumours	-	-	-	-	-	-
V. Retinoblastoma		**3.0**	**6.3**	**(3.1 - 9.6)**	**2.6**	**5.8**	**(2.6 - 8.9)**
VI. Renal tumours		**2.6**	**5.5**	**(2.5 - 8.6)**	**4.0**	**8.9**	**(5.0 - 12.8)**
VIa.	Nephroblastoma & other non-epithelial renal tumours	2.6	5.5	(2.5 - 8.6)	4.0	8.9	(5.0 - 12.8)
VIb.	Renal carcinomas	-	-	-	-	-	-
VIc.	Unspecified malignant renal tumours	-	-	-	-	-	-
VII. Hepatic tumours		**1.4**	**3.0**	**(0.8 - 5.2)**	**1.4**	**3.1**	**(0.8 - 5.5)**
VIIa.	Hepatoblastoma	1.0	2.1	(0.3 - 4.0)	1.0	2.2	(0.3 - 4.2)
VIIb.	Hepatic carcinomas	0.4	[b]	[b]	0.4	[b]	[b]
VIIc.	Unspecified malignant hepatic tumours	-	-	-	-	-	-
VIII. Malignant bone tumours		**5.6**	**12.0**	**(7.6 - 16.5)**	**3.4**	**7.7**	**(4.0 - 11.3)**
VIIIa.	Osteosarcomas	2.8	6.0	(2.9 - 9.2)	1.8	4.1	(1.4 - 6.7)
VIIIb.	Chondrosarcomas	0.0	-	-	-	-	-
VIIIc.	Ewing tumours & related bone sarcomas	2.2	4.7	(1.9 - 7.5)	1.2	2.7	(0.5 - 4.9)
VIIId.	Other specified malignant bone tumours	0.6	[b]	[b]	0.4	[b]	[b]
VIIIe.	Unspecified malignant bone tumours	-	-	-	-	-	-
IX. Soft tissue and other extraosseous sarcomas		**4.6**	**9.8**	**(5.8 - 13.8)**	**4.2**	**9.5**	**(5.4 - 13.5)**
IXa.	Rhabdomyosarcomas	2.4	5.1	(2.2 - 8.0)	1.8	4.1	(1.4 - 6.7)
IXb.	Fibrosarcomas & other fibrous neoplasms	0.4	[b]	[b]	0.2	[b]	[b]
IXc.	Kaposi sarcomas	-	-	-	-	-	-
IXd.	Other specified soft tissue sarcomas	1.6	3.5	(1.1 - 5.9)	1.8	4.0	(1.4 - 6.7)
IXe.	Unspecified soft tissue sarcomas	0.2	[b]	[b]	0.4	[b]	[b]
X. Germ cell tumours, trophoblastic tumours & neoplasms of gonads		**1.6**	**3.4**	**(1.0 - 5.8)**	**2.8**	**6.3**	**(3.0 - 9.6)**
Xa.	Intracranial & intraspinal germ cell tumours	0.6	[b]	[b]	1.0	[b]	[b]
Xb.	Extracranial & extragonadal germ cell tumours	0.6	[b]	[b]	0.8	[b]	[b]
Xc.	Malignant gonadal germ cell tumours	0.4	[b]	[b]	0.8	[b]	[b]
Xd.	Gonadal carcinomas	-	-	-	-	-	-
Xe.	Other & unspecified malignant gonadal tumours	-	-	-	0.2	[b]	[b]
XI. Other epithelial neoplasms & melanomas		**2.0**	**4.3**	**(1.6 - 6.9)**	**3.0**	**6.8**	**(3.4 - 10.2)**
XIa.	Adrenocortical carcinomas	0.2	[b]	[b]	0.2	[b]	[b]
XIb.	Thyroid carcinomas	0.4	[b]	[b]	0.4	[b]	[b]
XIc.	Nasopharyngeal carcinomas	0.0	-	-	0.0	-	-
XId.	Melanomas	0.6	[b]	[b]	0.6	[b]	[b]
XIe.	Skin carcinomas	-	-	-	-	-	-
XIf.	Other & unspecified carcinomas	0.8	1.7	(0.0 - 3.4)	1.8	4.1	(1.4 - 6.8)
XII. Other & unspecified malignant neoplasms		**-**	-	-	**0.2**	**0.5**	**(0.0 - 1.3)**
XIIa.	Other specified malignant tumours	-	-	-	0.2	[b]	[b]
XIIb.	Other unspecified malignant tumours	-	-	-	-	-	-
All childhood cancers		**85.8**	**183.2**	**(165.8 - 200.5)**	**66.6**	**149.6**	**(133.5 - 165.7)**

[a] Rate per million population per year, age-standardised to the World Standard Population

[b] Age standardised rates (and corresponding 95% CI) have been censored for diagnostic subgroups where there were fewer than 10 cases diagnosed within the five-year period

3.7 Childhood cancer incidence by age at diagnosis

3.7.1 Age distribution of childhood cancers

Figure 3.7.1a shows half (50.2%) of all childhood cancers were diagnosed in children under the age of five, compared to 43.9% in the 2000-2009 period. The greatest number of cancers by age and sex were for 1 year old boys (56 cases) and 2 year old girls (45 cases) (see Figure 3.7.1b).

Figure 3.7.1c highlights that retinoblastoma, neuroblastoma, renal and hepatic tumours all skewed heavily towards the younger child population, while malignant bone tumours, and 'other epithelial neoplasms' were more commonly diagnosed in those aged 10-14 years.

Figure 3.7.1a Age distribution of childhood cancers, New Zealand, 2010-2014

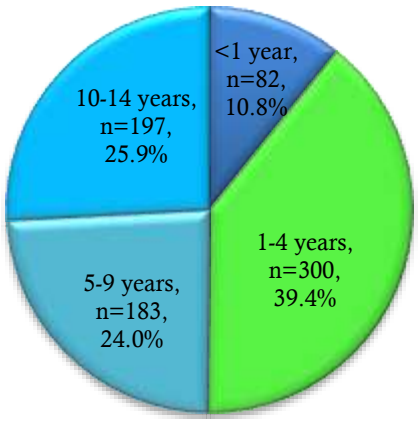


Figure 3.7.1b Childhood cancers by age at diagnosis and sex, New Zealand, 2010-2014

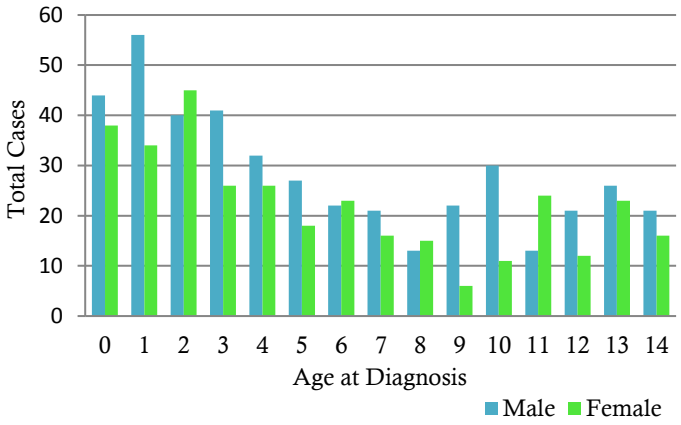
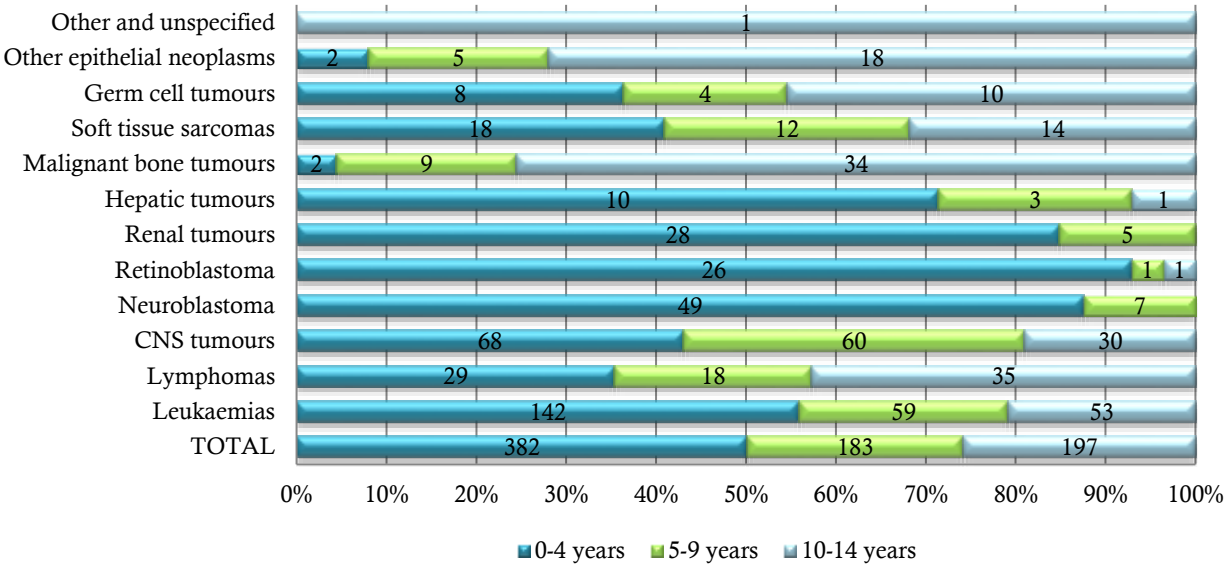


Figure 3.7.1c Number of childhood cancer cases by age group, New Zealand, 2010-2014



3.7.2 Childhood cancer incidence by age group

Neuroblastoma, retinoblastoma, and renal tumours were significantly more likely to be diagnosed in children under the age of 5 than in older children (see Table 3.7.2). By diagnostic subgroup, incidence of lymphoid leukaemias was significantly higher for children aged 0-4 years (72.1 per million) than children aged 5-9 years (29.0 per million) or 10-14 years (21.4 per million) and the incidence of lymphoreticular neoplasms (LCH) followed a similar pattern.

In contrast, incidence of Hodgkin lymphomas was significantly lower for children aged 0-4 (1.3 per million), increasing over ten-fold to 14.0 per million for the 10-14 year age group. The incidence of 'other epithelial neoplasms' (12.0 per million) and malignant bone tumours (22.7 per million) was also significantly higher in the 10-14 year age group.

Table 3.7.2 Childhood cancer incidence by age at diagnosis, New Zealand, 2010-2014

ICCC-3 diagnostic group and selected subgroups	0-4 years				5-9 years				10-14 years			
	Average cases per year	%	Rate per million population per year (95% CI)		Average cases per year	%	Rate per million population per year (95% CI)		Average cases per year	%	Rate per million population per year (95% CI)	
I. Leukaemias	28.4	37.2	90.6	(75.6 - 105.4)	11.8	32.2	39.8	(29.7 - 50.0)	10.6	26.9	35.4	(25.9 - 44.9)
Lymphoid leukaemias	*22.6*	*29.6*	*72.1*	*(58.8 - 85.3)*	*8.6*	*23.5*	*29.0*	*(20.3 - 37.7)*	*6.4*	*16.2*	*21.4*	*(14.0 - 28.8)*
Acute myeloid leukaemias	*3.4*	*4.5*	*10.8*	*(5.7 - 16.0)*	*1.8*	*4.9*	*6.1*	*(2.1 - 10.0)*	*2.8*	*7.1*	*9.3*	*(4.5 - 14.2)*
II. Lymphomas	5.8	7.6	18.5	(11.8 - 25.2)	3.6	9.8	12.2	(6.5 - 17.8)	7.0	17.8	23.4	(15.6 - 31.1)
Hodgkin lymphomas	*0.4*	*0.5*	*1.3*	*(0.0 - 3.0)*	*1.4*	*3.8*	*4.7*	*(1.2 - 8.2)*	*4.2*	*10.7*	*14.0*	*(8.0 - 20.0)*
Lymphoreticular neoplasms	*3.8*	*5.0*	*12.1*	*(6.7 - 17.6)*	*0.6*	*1.6*	*2.0*	*(0.0 - 4.3)*	*0.6*	*1.5*	*2.0*	*(0.0 - 4.3)*
III. CNS tumours	13.6	17.8	43.4	(33.1 - 53.7)	12.0	32.8	40.5	(30.2 - 50.7)	6.0	15.2	20.0	(12.9 - 27.2)
Astrocytomas	*4.2*	*5.5*	*13.4*	*(7.7 - 19.1)*	*4.8*	*13.1*	*16.2*	*(9.7 - 22.7)*	*2.2*	*5.6*	*7.3*	*(3.0 - 11.7)*
Intracranial & intraspinal neoplasms	*3.6*	*4.7*	*11.5*	*(6.2 - 16.8)*	*3.0*	*8.2*	*10.1*	*(5.0 - 15.2)*	*1.0*	*2.5*	*3.3*	*(0.4 - 6.3)*
Other gliomas	*1.4*	*1.8*	*4.5*	*(1.2 - 7.8)*	*2.2*	*6.0*	*7.4*	*(3.0 - 11.8)*	*1.0*	*2.5*	*3.3*	*(0.4 - 6.3)*
IV. Neuroblastoma	9.8	12.8	31.2	(22.5 - 40.0)	1.4	3.8	4.7	(1.2 - 8.2)	-	-	-	-
V. Retinoblastoma	5.2	6.8	16.6	(10.2 - 23.0)	0.2	0.5	0.7	(0.0 - 2.0)	0.2	0.5	0.7	(0.0 - 2.0)
VI. Renal tumours	5.6	7.3	17.9	(11.2 - 24.5)	1.0	2.7	3.4	(0.4 - 6.3)	-	-	-	-
VII. Hepatic tumours	2.0	2.6	6.4	(2.4 - 10.3)	0.6	1.6	2.0	(0.0 - 4.3)	0.2	0.5	0.7	(0.0 - 2.0)
VIII. Malignant bone tumours	0.4	0.5	1.3	(0.0 - 3.0)	1.8	4.9	6.1	(2.1 - 10.0)	6.8	17.3	22.7	(15.1 - 30.3)
Osteosarcomas	-	-	-	-	*0.6*	*1.6*	*2.0*	*(0.0 - 4.3)*	*4.0*	*10.2*	*13.4*	*(7.5 – 19.2)*
IX. Soft tissue sarcomas	3.6	4.7	11.5	(6.2 - 16.8)	2.4	6.6	8.1	(3.5 - 12.7)	2.8	7.1	9.3	(4.5 - 14.2)
Rhabdomyosarcoma	*2.4*	*3.1*	*7.7*	*(3.3 - 12.0)*	*0.8*	*2.2*	*2.7*	*(0.1 - 5.3)*	*1.0*	*2.5*	*3.3*	*(0.4 - 6.3)*
X. Germ cell tumours	1.6	2.1	5.1	(1.6 - 8.6)	0.8	2.2	2.7	(0.1 - 5.3)	2.0	5.1	6.7	(2.5 - 10.8)
XI. Other epithelial neoplasms	0.4	0.5	1.3	(0.0 - 3.0)	1.0	2.7	3.4	(0.4 - 6.3)	3.6	9.1	12.0	(6.5 - 17.6)
XII. Other and unspecified	-	-	-	-	-	-	-	-	0.2	0.5	0.7	(0.0 - 2.0)
All childhood cancers	**76.4**	**100**	**243.6 (219.2 – 268.0)**		**36.6**	**100**	**123.5 (105.6 - 141.4)**		**39.4**	**100**	**131.5 (113.1 - 149.8)**	

[a] Rate per million population per year, age-standardised to the World Standard Population

3.7.3 Childhood cancer incidence by age group and diagnostic subgroup

Neuroblastoma is the leading cause of cancer in infancy, accounting for 22% of the cancers treated in 0-1 year olds, but by 1-4 years, this has been replaced by leukaemias which now makes up 43% of new cases (See Figures 3.7.3a to 3.7.3d). Tumours of the central nervous system accounted for nearly one in three diagnosed in 5-9 year olds, while one in four cancers diagnosed in 10-14 year olds was a bone or soft tissue sarcoma.

Figure 3.7.3a Cancers diagnosed in infants aged <1 year, New Zealand, 2010-2014

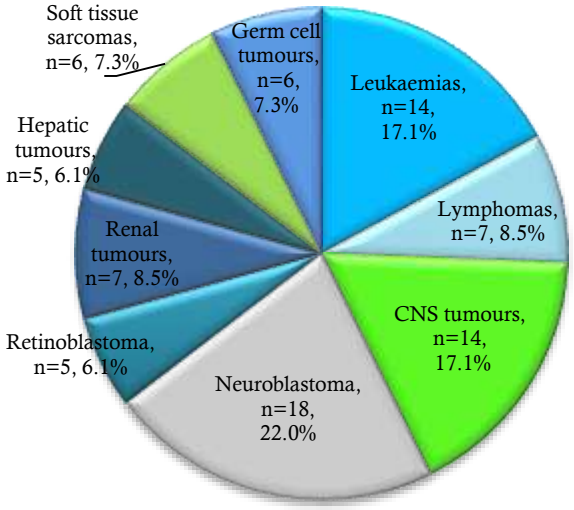


Figure 3.7.3b Cancers diagnosed in children aged 1-4 years, New Zealand, 2010-2014

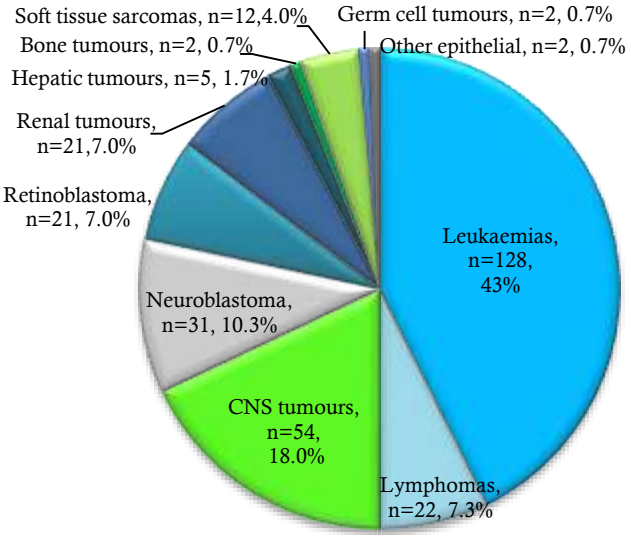


Figure 3.7.3c Cancers diagnosed in children aged 5-9 years, New Zealand, 2010-2014

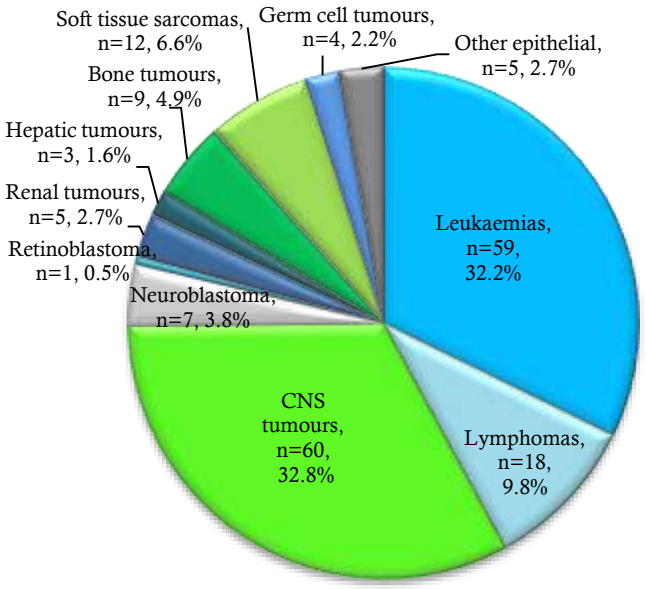
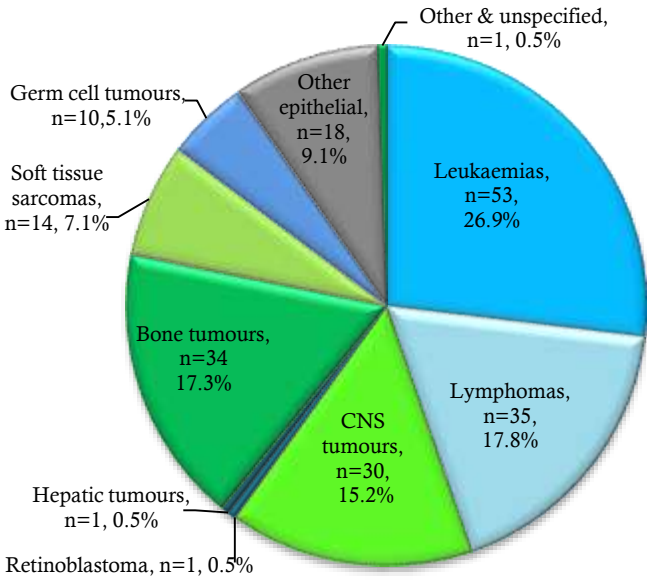


Figure 3.7.3d Cancers diagnosed in children aged 10-14 years, New Zealand, 2010-2014



3.8 Childhood cancer incidence by ethnicity

The 2000-2009 analysis found that childhood cancer incidence was significantly lower for Maori (131.1 per million) than for non-Maori/non-Pacific Peoples (158.1 per million). However, Table 3.8 shows that in the 2010-2014 time period Maori and Non-Maori had similar incidence rates of 157.7 and 170.1 per million per year respectively. The spectrum of cancers diagnosed were remarkably consistent between the two groups, with the exception of CNS tumours which accounted for 26.3% of non-Maori cases compared to 18.9% of Maori.

Table 3.8 Childhood cancer incidence by prioritised ethnicity, New Zealand, 2010-2014

ICC-3 diagnostic group & selected subgroups	Maori			Non-Maori		
	Cases per year	Incidence per million per year (95% CI)		Cases per year	Incidence per million per year (95% CI)	
I. Leukaemias	12.0	50.8	(37.9 - 63.7)	38.8	57.4	(49.3 - 65.5)
II. Lymphomas	3.0	12.7	(6.3 - 19.2)	13.4	19.7	(15.0 - 24.4)
III. CNS Tumours	9.8	41.6	(29.9 - 53.3)	21.8	32.3	(26.2 - 38.3)
IV. Neuroblastoma	2.2	8.8	(3.6 - 13.9)	9.0	13.4	(9.5 - 17.3)
V. Retinoblastoma	1.6	6.4	(2.0 - 10.8)	4.0	5.9	(3.3 - 8.5)
VI. Renal tumours	0.8	3.2	(0.1 - 6.3)	5.8	8.6	(5.5 - 11.8)
VII. Hepatic tumours	0.8	3.3	(0.1 - 6.6)	2.0	3.0	(1.1 - 4.8)
VIII. Malignant bone tumours	2.2	9.8	(4.0 - 15.6)	6.8	9.9	(6.6 - 13.2)
IX. Soft tissue sarcomas	1.8	7.8	(2.7 - 12.8)	7.0	10.3	(6.9 - 13.7)
X. Germ cell tumours	1.4	6.1	(1.6 - 10.6)	3.0	4.4	(2.2 - 6.6)
XI. Other epithelial neoplasms	1.6	7.3	(2.2 - 12.3)	3.4	5.0	(2.6 - 7.3)
XII. Other malignant neoplasms	-	-	-	0.2	0.3	(0.0 - 0.9)
Total childhood cancers	**37.2**	**157.7**	**(135.0 - 180.4)**	**115.2**	**170.1**	**(156.2 - 184.0)**

Figure 3.8a Cancers diagnosed in Maori children, New Zealand, 2010-2014

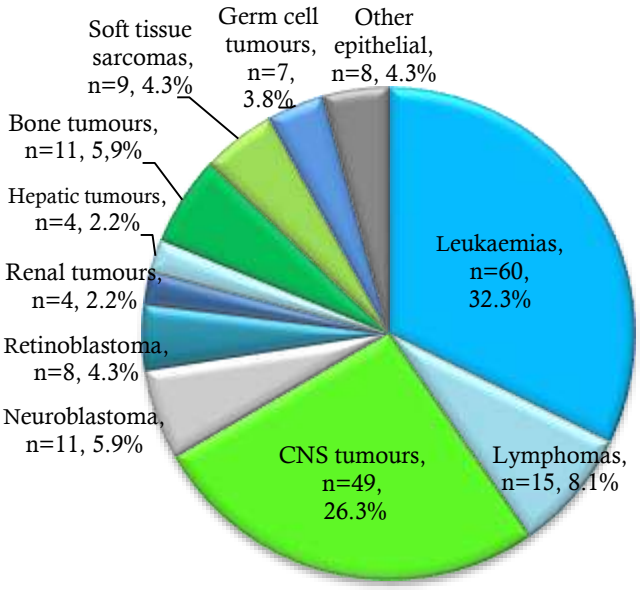
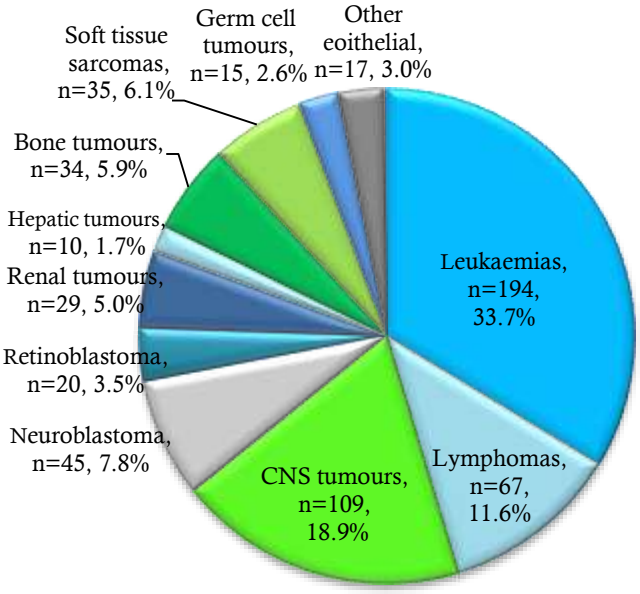


Figure 3.8b Cancers diagnosed in non-Maori children, New Zealand, 2010-2014



3.9 Childhood cancer by shared care centre at diagnosis

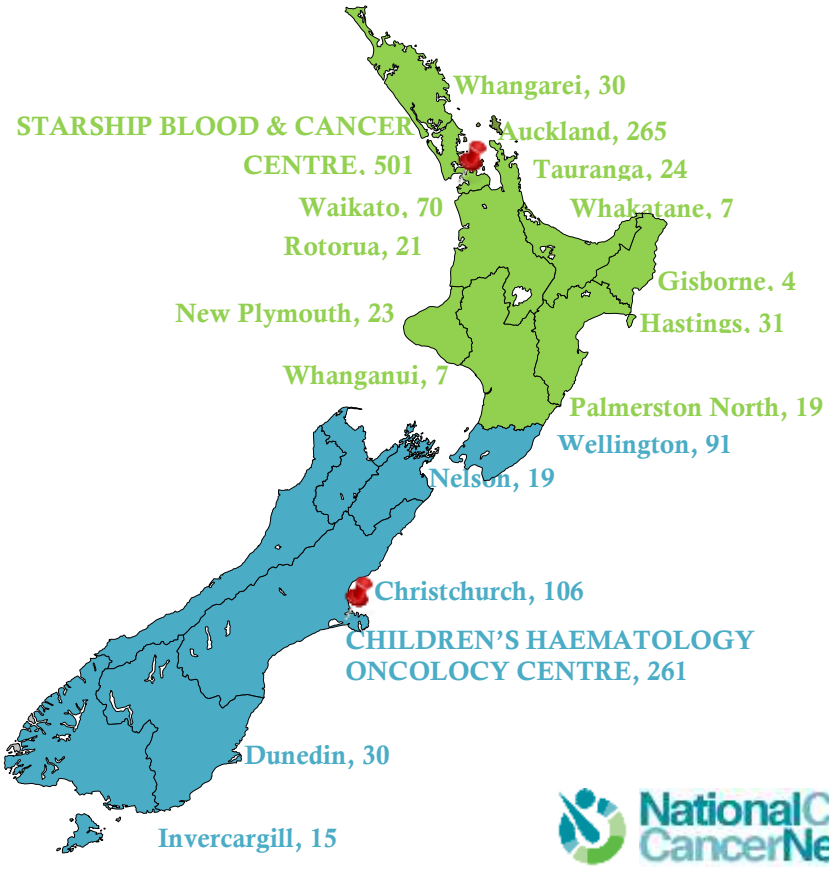
A key aim of the National Child Cancer Network is to ensure that cancer treatment is delivered as close to the child's home as possible. Currently New Zealand has two specialist paediatric oncology centres (Starship Blood and Cancer Centre in Auckland and the Children's Haematology Oncology Centre (CHOC) in Christchurch and 14 shared care centres where children may receive part of their cancer treatment, such as the administering of some chemotherapy agents.

The following section provides the number of child cancer cases diagnosed between 2010 and 2014 by the tertiary centre and shared care centre regions at diagnosis. It should be noted that;

- There can be considerable year-on-year fluctuations in regional patient numbers and the annual number of patients seen in each shared care hospital unit will be higher than the new diagnoses reported as many children undergo treatment and disease surveillance for a number of years.
- The amount of care delivered at the shared care centres depends on the unique treatment needs of each child; some patients receive their treatment exclusively at a specialist paediatric oncology centre while others may undergo much of their treatment and follow-up outside of these centres.

Auckland or Christchurch was the closest major city for just under half (48.7%) of children diagnosed with cancer between 2010 and 2014. The remaining children were assigned a shared care centre based on their usual residence. Figure 3.9 highlights the considerable upheaval that families may face while their child is undergoing treatment at a specialist paediatric oncology centre. In addition, many families travel major distances from their home to their nearest shared care centre (e.g. from Queenstown to Invercargill). Approximately two thirds (65.7%) of all children diagnosed with cancer in the period resided in one of the 12 district health boards which refer their patients to Starship Blood and Cancer Centre while the remainder (34.3%) were referred to CHOC in Christchurch. Waikato and Wellington were the two largest shared care centres, with Gisborne the smallest.

Figure 3.9 New childhood cancers diagnosed by shared care centre, 2010-2014



4 Childhood Cancer Incidence by Diagnostic Group

The following section describes cancer incidence for each of the ICCC-3 diagnostic subgroups in turn. Each section 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 many of the diagnostic subgroups only a very small number of cases have been diagnosed within the five-year period. **Caution is advised when interpreting rates derived from a small number of cases as they may fluctuate markedly over time.**

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 according to one of the COG leukaemia clinical trials.

4.1.1 Leukaemias overall incidence

In the years 2000-2009, New Zealand had an average of 37.6 new cases of leukaemia diagnosed each year, representing exactly one third of all new paediatric cancer cases for this time period. Over half (55.9%) of the total childhood leukaemias were diagnosed in children under the age of five, with an incidence rate of 90.6 per million population per year. In addition, 40 children were diagnosed with acute myeloid diseases and 12 children were diagnosed with 'other myeloproliferative diseases' (e.g. myelodysplastic syndrome).

Table 4.1.1 Incidence of childhood leukaemias by ICCC subgroup, sex, age group, and ethnicity, New Zealand, 2010-2014

	Cases per year	%	Rates per million population per year (95% CI)	
ICCC Subgroup				
Ia. Lymphoid leukaemias	37.6	74.0	41.1	(35.2 - 47.0)
Ib. Acute myeloid leukaemias	8.0	15.7	8.8	(6.1 - 11.5)
Ic. Chronic myeloproliferative disease	0.8	1.6	[b]	[b]
Id. Other myeloproliferative diseases	2.4	4.7	2.6	(1.1 - 4.1)
Ie. Other & unspecified leukaemias	2.0	3.9	2.2	(0.8 - 3.5)
Sex				
Male	31.6	62.2	67.4	(56.9 - 77.9)
Female	19.2	37.8	43.1	(34.5 - 51.7)
Age group				
0-4 years	28.4	55.9	90.6	(75.6 - 105.4)
5-9 years	11.8	23.2	39.8	(29.7 - 50.0)
10-14 years	10.6	20.9	35.4	(25.9 - 44.9)
Ethnicity				
Maori	12.0	23.6	50.8	(37.9 - 63.7)
Non-Maori	38.8	76.4	57.4	(49.3 - 65.5)
Total	**50.8**	**100.0**	**55.5**	**(48.7 - 62.4)**

4.1.2 Incidence for selected leukaemia diagnostic subgroups by sex, age group, and ethnicity

Table 4.1.3 shows that childhood ALL was most commonly diagnosed in under-fives (60.1% of new cases) and that males (118 cases, 50.3 per million) were significantly more likely to be diagnosed than females (70 cases, 31.4 per million). Maori made up a greater proportion of children diagnosed with AML (40.0%) than ALL (20.7%).

Table 4.1.2 Number of cases of childhood ALL and AML by sex, age group, and ethnicity, New Zealand, 2010-2014

	Lymphoid Leukaemias (ALL)				Acute Myeloid Leukaemias (AML)			
	Cases per year	%	Age standardised rate (95% CI)		Cases per year	%	Age standardised rate (95% CI)	
Sex								
Male	23.6	62.8	50.3	(41.2 - 59.3)	4.8	60.0	10.2	(6.1 - 14.3)
Female	14.0	37.2	31.4	(24.1 - 38.8)	3.2	40.0	7.2	(3.7 - 10.7)
Age group								
0-4 years	22.6	60.1	72.1	(58.8 - 85.3)	3.4	42.5	10.8	(5.7 - 16.0)
5-9 years	8.6	22.9	29.0	(20.3 - 37.7)	1.8	22.5	6.1	(2.1 - 10.0)
10-14 years	6.4	17.0	21.4	(14.0 - 28.8)	2.8	35.0	9.3	(4.5 - 14.2)
Ethnicity								
Maori	7.8	20.7	33.6	(23.1 - 44.2)	3.2	40.0	13.8	(7.0 - 20.6)
Non-Maori	29.8	79.3	44.0	(36.9 - 51.0)	4.8	60.0	7.1	(4.3 - 9.9)
Total	**37.6**	**100.0**	**41.1**	**(35.2 - 47.0)**	**8.0**	**100.0**	**8.8**	**(6.1 - 11.5)**

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 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 LCH.

Hodgkin lymphomas arise in specific pre-malignant lymphoma cells within a lymph node region and tend to spread to adjacent lymph nodes. 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 NHL's 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. It can appear as a single lesion or can be multisystem, affecting bones and organs such as the liver, lung, brain and skin. 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,[10] 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 Lymphomas overall incidence

The incidence of lymphomas and reticuloendothelial neoplasms was significantly higher for boys (23.5 per million) than girls (12.2 per million). Lymphoma was rarely diagnosed in children four years and under (an incidence rate of 3.8 per million), with no infant cases reported in the ten-year period. Lymphomas account for 17.8% of cancers diagnosed in 10-14 year olds, with incidence double that reported for 5-9 year olds.

Overall incidence of 'lymphomas and reticuloendothelial neoplasms' increased from 13.0 per million in 2000-2009 to 18.0 per million in 2010-2014, reflecting the introduction of LCH which has a recorded incidence of 5.4 per million for the period.

Table 4.2.1 Incidence of childhood lymphomas, by diagnostic subgroup, sex, age group, and ethnicity, New Zealand, 2010-2014

	Case per year	%	Rates per million population per year (95% CI)	
ICCC Subgroup				
IIa. Hodgkin lymphomas	6.0	36.6	6.6	(4.3 - 9.0)
IIb. Non-Hodgkin lymphomas (except Burkitt lymphoma)	3.2	19.5	3.5	(1.8 - 5.3)
IIc. Burkitt lymphoma	2.2	13.4	2.4	(1.0 - 3.8)
IId. Miscellaneous lymphoreticular neoplasms	5.0	30.5	5.4	(3.3 - 7.6)
IIe. Unspecified lymphomas	-	-	-	-
Sex				
Male	11.0	67.1	23.5	(17.3 - 29.8)
Female	5.4	32.9	12.2	(7.6 - 16.7)
Age group				
0-4 years	5.8	35.4	18.5	(11.8 - 25.2)
5-9 years	3.6	22.0	12.2	(6.5 - 17.8)
10-14 years	7.0	42.7	23.4	(15.6 - 31.1)
Ethnicity				
Maori	3.0	18.3	12.7	(6.3 - 19.2)
Non-Maori	13.4	81.7	19.7	(15.0 - 24.4)
Total	**16.4**	**100.0**	**18.0**	**(14.1 - 21.9)**

4.2.2 Incidence for selected lymphoma diagnostic subgroups by sex, age group, and ethnicity

Overall, Hodgkin and non-Hodgkin lymphomas were rarely diagnosed in children under the age of 5. This has been somewhat masked by the 2010 introduction of LCH into this diagnostic group, as LCH accounts for 19 out of the 29 'lymphomas and reticuloendothelial neoplasms' diagnosed in 0-4 year olds the period. LCH also is significantly more common in boys than girls, with a ratio of 4:1.

Table 4.2.2 Number of cases of childhood Hodgkin lymphomas and Langerhans cell histiocytosis by sex, age group, and ethnicity, New Zealand, 2010-2014

	Hodgkin lymphomas				Langerhans cell histiocytosis			
	Cases per year	%	Age standardised rate (95% CI)		Cases per year	%	Age standardised rate (95% CI)	
Sex								
Male	4.0	66.7	8.6	(4.8 - 12.4)	4.0	80.0	4.0	(4.8 - 12.2)
Female	2.0	33.3	4.5	(1.7 - 7.3)	1.0	20.0	2.2	(0.3 - 4.2)
Age group								
0-4 years	0.4	6.7	1.3	(0.0 - 3.0)	3.8	76.0	12.1	(6.7 - 17.6)
5-9 years	1.4	23.3	4.7	(1.2 - 8.2)	0.6	12.0	2.0	(0.0 - 4.3)
10-14 years	4.2	70.0	14.0	(8.0 - 20.0)	0.6	12.0	2.0	(0.0 - 4.3)
Ethnicity								
Maori	1.0	16.7	4.3	(0.5 - 8.1)	1.2	24.0	5.2	(1.0 - 9.3)
Non-Maori	5.0	83.3	7.4	(4.5 - 10.3)	3.8	76.0	5.6	(3.1 - 8.1)
Total	**6.0**	**100.0**	**6.6**	**(4.3 - 9.0)**	**5.0**	**100.0**	**5.4**	**(3.3 - 7.6)**

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 a remnants of primitive developmental structures (germ cell tumours). This heterogeneous group of tumours vary from relatively benign tumours such as pilocytic astrocytomas, 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 Gorlins 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 used by the New Zealand Cancer Registry (and many other 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) 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 CNS tumours overall incidence

With an incidence rate of 34.7 per million, 'CNS and miscellaneous intracranial and intraspinal neoplasms' was the second most common class of tumour diagnosed in New Zealand children between 2010 and 2014. CNS tumours accounted for 20.7% of all childhood cancers diagnosed, with around 32 new cases diagnosed annually. Of the 158 central nervous system tumours registered in the five year period, 65 (41.1%) were tumours that were either benign or of uncertain behaviour and over half of these (37 cases, 23.4% of all CNS tumours) were juvenile pilocytic astrocytomas.

Table 4.3.1 shows that overall CNS tumour incidence was significantly lower for 10-14 year olds (20.0 per million) than for 0-4 year olds (43.4 per million) or 5-9 year olds (40.5 per million). There were no differences according to sex or prioritised ethnicity. Astrocytomas were the most commonly diagnosed CNS tumour group, accounting for 35.4% of all CNS tumours diagnosed in the 5 year period.

Table 4.3.1 Incidence of childhood CNS tumours by ICCC subgroup, sex, age group, and ethnicity, New Zealand, 2010–2014

	Cases per year	%	Rates per million population per year (95% CI)	
ICCC subgroup				
IIIa. Ependymomas and choroid plexus tumours	3.6	11.4	3.9	(2.1 - 5.7)
IIIb. Astrocytomas	11.2	35.4	12.3	(9.1 - 15.6)
IIIc. Intracranial and intraspinal embryonal tumours	7.6	24.1	8.4	(5.7 - 11.0)
IIId. Other gliomas	4.6	14.6	5.1	(3.0 - 7.2)
IIIe. Other specified intracranial and intraspinal neoplasms	3.2	10.1	3.5	(1.8 - 5.2)
IIIf. Unspecified intracranial and intraspinal neoplasms	1.4	4.4	[b]	[b]
Sex				
Male	17.0	53.8	36.4	(26.7 - 44.2)
Female	14.6	46.2	33.0	(25.4 - 40.5)
Age group				
0-4 years	13.6	43.0	43.4	(33.1 - 53.7)
5-9 years	12.0	38.0	40.5	(30.2 - 50.7)
10-14 years	6.0	19.0	20.0	(12.9 - 27.2)
Ethnicity				
Maori	9.8	31.0	41.6	(29.9 - 53.3)
Non-Maori	21.8	69.0	32.3	(26.2 - 38.3)
Total	**31.6**	**100.0**	**34.7**	**(29.3 - 40.2)**

4.3.2 Incidence for selected CNS tumour diagnostic subgroups, by sex, age group, and ethnicity

Of the 38 intracranial and intraspinal embryonal tumours diagnosed in the 2000-2009 period, 30 (78.9%) were medulloblastoma. As was also noted in the 2000-2009 analysis, those of Maori ethnicity accounted for a high proportion of all intracranial and intraspinal embryonal tumours diagnosed (42.1%, see Table 3.3.3). Incidence of intracranial and intraspinal embryonal tumours was significantly higher for males (12.4 per million) than females (4.1 per million). No such ethnic or sex differences were seen for the most common CNS group, astrocytomas.

Table 4.3.2 Number of cases of childhood CNS tumours by diagnostic subgroup, by sex, age group, and ethnicity, New Zealand, 2010-2014

	Astrocytomas				Intracranial & intraspinal embryonal tumours			
	Cases per year	%	Age standardised rate (95% CI)		Cases per year	%	Age standardised rate (95% CI)	
Sex								
Male	5.8	51.8	12.5	(7.9 - 17.0)	5.8	76.3	12.4	(7.9 - 17.0)
Female	5.4	48.2	12.2	(7.6 - 16.8)	1.8	23.7	4.1	(1.4 - 6.7)
Age group								
0-4 years	4.2	37.5	13.4	(7.7 - 19.1)	3.6	47.4	11.5	(6.2 - 16.8)
5-9 years	4.8	42.9	16.2	(9.7 - 22.7)	3.0	39.5	10.1	(5.0 - 15.2)
10-14 years	2.2	19.6	7.3	(3.0 - 11.7)	1.0	13.2	3.3	(0.4 - 6.3)
Ethnicity								
Maori	3.4	30.4	14.7	(7.7 - 21.6)	3.2	42.1	13.8	(7.0 - 20.6)
Non-Maori	7.8	69.6	11.5	(7.9 - 15.1)	4.4	57.9	6.5	(3.8 - 9.2)
Total	**11.2**	**100.0**	**12.3**	**(9.1 - 15.6)**	**7.6**	**100.0**	**8.4**	**(5.7 - 11.0)**

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 may 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.

Diagnostic group 4: 'Neuroblastoma and other peripheral nervous cell tumours' had an overall incidence of 12.2 per million per year but this rate was significantly higher for the 0-4 year age group (31.4 per million, see Table 4.4). 18 cases (32.1%) of neuroblastoma were diagnosed in infants less than one year of age; neuroblastoma represented 22.0% of all cancers diagnosed in infancy, making it the most common cancer for this age group.

Table 4.4 Incidence of childhood neuroblastoma and other peripheral nervous cell tumours by sex, age group, and ethnicity, New Zealand, 2010-2014

	Cases per year	%	Rates per million population per year (95% CI)	
Sex				
Male	5.4	48.2	11.5	(7.1 - 15.8)
Female	5.8	51.8	12.9	(8.2 - 17.6)
Age group				
0-4 years	9.8	87.5	31.2	(22.5 - 40.0)
5-9 years	1.4	12.5	4.7	(1.2 - 8.2)
10-14 years	-	-	-	-
Ethnicity				
Maori	2.2	19.6	8.8	(3.6 - 13.9)
Non-Maori	9.0	80.4	13.4	(9.5 - 17.3)
Total neuroblastoma	**11.2**	**100.0**	**12.2**	**(9.0 - 15.3)**

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.

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.

There were 28 cases of retinoblastoma diagnosed in the 2000-2009 period. While retinoblastoma accounted for 6.1% of all cancers diagnosed in infancy, 75% of all retinoblastoma cases were diagnosed in children aged 1-4 years old at diagnosis, accounting for 7.0% of cancers diagnosed in this age group. Table 4.5 shows that there were no significant differences in the retinoblastoma incidence rates according to sex or ethnicity.

Table 4.5 Incidence of childhood retinoblastoma by sex, age group, and ethnicity, New Zealand, 2010-2014

	Cases per year	%	Rates per million population per year (95% CI)	
Sex				
Male	3.0	53.6	6.3	(3.1 - 9.6)
Female	2.6	46.4	5.8	(2.6 - 8.9)
Age group				
0-4 years	5.2	92.9	16.6	(10.2 - 23.0)
5-9 years	0.2	3.6	0.7	(0.0 - 2.0)
10-14 years	0.2	3.6	0.7	(0.0 - 2.0)
Ethnicity				
Maori	1.6	28.6	6.4	(2.0 - 10.8)
Non-Maori	4.0	71.4	5.9	(3.3 - 8.5)
Total retinoblastoma	**5.6**	**100.0**	**6.1**	**(3.8 - 8.3)**

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.

Of the 33 renal tumours diagnosed between 2000 and 2009, all were from the diagnostic subgroup 'VIa: nephroblastoma and other non-epithelial renal tumours'; no cases of renal carcinoma were reported in the period. Table 4.6 shows that the incidence of renal tumours at 0-4 years of age (17.9 per million) declined rapidly in later childhood. Renal tumours accounted for 8.5% of the total cancers diagnosed in infants and 7.0% of cancers diagnosed in the 1-4 year age group.

Table 4.6 Incidence of childhood renal tumours by sex, age group, and ethnicity, New Zealand, 2010-2014

	Cases per year	%	Rates per million population per year (95% CI)	
Sex				
Male	2.6	39.4	5.5	(2.5 - 8.6)
Female	4.0	60.6	8.9	(5.0 - 12.8)
Age group				
0-4 years	5.6	84.8	17.9	(11.2 - 24.5)
5-9 years	1.0	15.2	3.4	(0.4 - 6.3)
10-14 years	-	-	-	-
Ethnicity				
Maori	0.8	12.1	3.2	(0.1 - 6.3)
Non-Maori	5.8	87.9	8.6	(5.5 - 11.8)
Total renal tumours	**6.6**	**100.0**	**7.2**	**(4.7 - 9.6)**

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 hepatobastoma, 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

Hepatic tumours had the lowest incidence of all specified childhood cancer diagnostic groups (3.1 per million per year, see Table 4.7). Of the 14 hepatic tumours diagnosed between 2000 and 2009, ten were hepatoblastoma (2.2 per million) and four were hepatic carcinomas (0.9 per million). Nine of the ten cases of hepatoblastoma were diagnosed in children aged 4 years and under.

Table 4.7 Incidence of childhood hepatic tumours by ICCC subgroup, sex, age group, and ethnicity, New Zealand, 2010-2014

	Cases per year	%	Rates per million population per year (95% CI)	
VIIa. Hepatoblastoma	2.0	71.4	2.2	(0.8 - 3.5)
VIIb. Hepatic carcinomas	0.8	28.6	0.9	(0.0 - 1.8)
Sex				
Male	1.4	50.0	3.0	(0.8 - 5.2)
Female	1.4	50.0	3.1	(0.8 - 5.5)
Age group				
0-4 years	2.0	71.4	6.4	(2.4 - 10.3)
5-9 years	0.6	21.4	2.0	(0.0 - 4.3)
10-14 years	0.2	7.1	0.7	(0.0 - 2.0)
Ethnicity				
Maori	0.8	28.6	3.3	(0.1 - 6.6)
Non-Maori	2.0	71.4	3.0	(1.1 - 4.8)
Total hepatic tumours	**2.8**	**100.0**	**3.1**	**(1.5 - 4.7)**

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 grows in the long bones of the leg, often directly above the knee joint. Ewing sarcomas arise from elements of 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 known 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 Malignant bone tumours overall incidence

Table 4.8.1 shows that the malignant bone tumour incidence rate for those aged 10-14 years (22.7 per million) was significantly higher than for those aged 0-4 years (1.3 per million) or 5-9 years (6.1 per million). Malignant bone tumours accounted for one in six of all cancers reported for the 10-14 year age group (17.3%). There were no sex or ethnic difference in the incidence of bone tumours.

Table 4.8.1 Incidence of childhood malignant bone tumours by ICCC subgroup, sex, age group, and ethnicity, New Zealand, 2010-2014

	Cases per year	%	Rates per million population per year (95% CI)	
ICCC subgroup				
VIIIa. Osteosarcomas	4.6	51.1	5.1	(3.0 - 7.1)
VIIIb. Chondrosarcomas	-	-	-	-
VIIIc. Ewing tumours & related bone sarcomas	3.4	37.8	3.8	(2.0 - 5.5)
VIIId. Other specified malignant bone tumours	1.0	11.1	[b]	[b]
VIIIe. Unspecified malignant bone tumours	-	-	-	-
Sex				
Male	5.6	62.2	12.0	(7.6 - 16.5)
Female	3.4	37.8	7.7	(4.0 - 11.3)
Age group				
0-4 years	0.4	4.4	1.3	(0.0 - 3.0)
5-9 years	1.8	30.0	6.1	(2.1 - 10.0)
10-14 years	6.8	75.6	22.7	(15.1 - 30.3)
Ethnicity				
Maori	2.2	24.4	9.8	(4.0 - 15.6)
Non-Maori	6.8	69.4	9.9	(6.6 - 13.2)
Total	**9.0**	**100.0**	**9.9**	**(7.0 - 12.8)**

4.8.2 Incidence for selected malignant bone tumour diagnostic subgroups by sex, age group, and ethnicity

Table 4.8.3 shows that there were few differences between the sex, age, and ethnic distributions for osteosarcomas compared with Ewing tumours.

Table 4.8.2 Number of cases of osteosarcomas and 'Ewing tumours and related bone sarcomas' by sex, age group, and ethnicity, New Zealand, 2010-2014

	Osteosarcomas				Ewing tumours and related bone sarcomas			
	Cases per year	%	Age standardised rate (95% CI)		Cases per year	%	Age standardised rate (95% CI)	
Sex								
Male	2.8	60.9	6.0	(2.9 - 9.2)	2.2	64.7	4.7	(1.9 - 7.5)
Female	1.8	39.1	4.1	(1.4 - 6.7)	1.2	35.3	2.7	(0.5 - 4.9)
Age group								
0-4 years	-	-	-	-	0.2	5.9	0.6	(0.0 - 1.9)
5-9 years	0.6	13.0	2.0	(0.0 - 4.3)	0.8	23.5	2.7	(0.1 - 5.3)
10-14 years	4.0	87.0	13.4	(7.5 - 19.2)	2.4	70.6	8.0	(3.5 - 12.5)
Ethnicity								
Maori	1.0	21.7	4.3	(0.5 - 8.1)	1.2	35.3	5.2	(1.0 - 9.3)
Non-Maori	3.6	78.3	5.3	(2.9 - 7.8)	2.2	64.7	3.3	(1.3 - 5.2)
Total	**4.6**	**100.0**	**5.1**	**(3.0 - 7.1)**	**3.4**	**100.0**	**3.8**	**(2.0 - 5.5)**

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.

With an incidence rate of 9.7 cases per million population, 5.7% of all childhood cancers diagnosed between 2000 and 2009 were from the diagnostic group 'soft tissue and other extraosseous sarcomas'. Incidence per million was largely consistent across the sex, age and ethnic groups. Just under half (47.7%) of all cases were rhabdomyosarcomas, of which 57.1% were diagnosed in children under five years of age.

Table 4.9.1 Incidence of childhood soft tissue sarcomas by sex, age group, and ethnicity, New Zealand, 2010-2014

	Cases per year	%	Rates per million population per year (95% CI)	
ICCC subgroup				
IXa. Rhabdomyosarcomas	4.2	47.7	4.6	(2.6 - 6.6)
IXb. Fibrosarcomas & other fibrous neoplasms	0.6	6.8	[b]	[b]
IXc. Kaposi sarcomas	-	-	-	-
IXd. Other specified soft tissue sarcomas	3.4	38.6	3.7	(2.0 - 5.5)
IXe. Unspecified soft tissue sarcomas	0.6	6.8	[b]	[b]
Total	**8.8**	**100.0**	**9.7**	**(6.8 - 12.5)**

Table 4.9.2 Number of cases of soft tissue sarcomas, including rhabdoymosarcomas, by sex, age group, and ethnicity, New Zealand, 2010-2014

	Soft tissue sarcomas				Rhabdomyosarcomas			
	Cases per year	%	Rates per million per year (95% CI)		Cases per year	%	Rates per million per year (95% CI)	
Sex								
Male	4.6	52.3	9.8	(5.8 - 13.8)	2.4	57.1	5.1	(2.2 - 8.0)
Female	4.2	47.7	9.5	(5.4 - 13.5)	1.8	42.9	4.1	(1.4 - 6.7)
Age group								
0-4 years	3.6	40.9	11.5	(6.2 - 16.8)	2.4	57.1	7.7	(3.3 - 12.0)
5-9 years	2.4	27.3	8.1	(3.5 - 12.7)	0.8	19.0	2.7	(0.1 - 5.3)
10-14 years	2.8	31.8	9.3	(4.5 - 14.2)	1.0	23.8	3.3	(0.4 - 6.3)
Ethnicity								
Maori	1.8	20.5	7.8	(2.7 - 12.8)	0.8	19.0	3.5	(0.1 - 6.8)
Non-Maori	7.0	79.5	10.3	(6.9 - 13.7)	3.4	81.0	5.0	(2.6 - 7.4)
Total	**8.8**	**100.0**	**9.7**	**(6.8 - 12.5)**	**4.2**	**100.0**	**4.6**	**(2.6 - 6.6)**

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 (extra-gonadal germ cell tumours).

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.

Germ cell tumours, trophoblastic tumours, and neoplasms of gonads accounted for only 2.9% of the total cancers diagnosed in children between 2010 and 2014. No differences in incidence according to age, sex or ethnicity were evident. Table 4.10 shows that in the five year period there were a similar numbers of cases arising in the gonads, central nervous system and elsewhere in the body.

Table 4.10 Incidence of childhood germ cell tumours by sex, age group, and ethnicity, New Zealand, 2010-2014

	Cases per year	%	Rates per million population per year (95% CI)	
Diagnostic Subgroup				
Xa. Intracranial & intraspinal germ cell tumours	1.6	36.4	1.8	(0.5 - 3.0)
Xb. Malignant extracranial & extragonadal germ cell tumours	1.4	31.8	1.5	(0.4 - 2.7)
Xc. Malignant gonadal germ cell tumours	1.2	27.3	1.3	(0.3 - 2.4)
Xd. Gonadal carcinomas	-	-	-	-
Xe. Other & unspecified malignant gonadal tumours	0.2	-	[b]	[b]
Sex				
Male	1.6	36.4	3.4	(1.0 - 5.8)
Female	2.8	63.6	6.3	(3.0 - 9.6)
Age group				
0-4 years	1.6	36.4	5.1	(1.6 - 8.6)
5-9 years	0.8	18.2	2.7	(0.1 - 5.3)
10-14 years	2.0	45.5	6.7	(2.5 - 10.8)
Ethnicity				
Maori	1.4	31.8	6.1	(1.6 - 10.6)
Non-Maori	3.0	68.2	4.4	(2.2 - 6.6)
Total	**4.4**	**100.0**	**4.8**	**(2.8 - 6.8)**

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.

An average of five new cases of 'other malignant epithelial neoplasms and malignant melanomas' were diagnosed in New Zealand each year between 2010 and 2014 (5.5 per million, see Table 4.11.1). Incidence increased considerably as children entered early adolescence, increasing from 1.3 per million for the 0-4 year age group to 12.0 per million among those aged 10-14 years.

Table 4.11 Incidence of childhood epithelial neoplasms by ICCC subgroup, sex, age group, and ethnicity, New Zealand, 2010-2014

	Total cases	%	Rates per million population per year (95% CI)	
ICCC subgroup				
XIa. Adrenocortical carcinomas	0.4	8.0	[b]	[b]
XIb. Thyroid carcinomas	0.8	16.0	[b]	[b]
XIc. Nasopharyngeal carcinomas	-	-	-	-
XId. Melanomas	1.2	24.0	[b]	[b]
XIe. Skin carcinomas	-	-	-	-
XIf. Other & unspecified carcinomas	2.6	52.0	2.9	(1.3 - 4.4)
Sex				
Male	2.0	40.0	4.3	(1.6 - 6.9)
Female	3.0	60.0	6.8	(3.4 - 10.2)
Age group				
0-4 years	0.4	8.0	1.3	(0.0 - 3.0)
5-9 years	1.0	20.0	3.4	(0.4 - 6.3)
10-14 years	3.6	72.0	12.0	(6.5 - 17.6)
Ethnicity				
Maori	1.6	32.0	7.3	(2.2 - 12.3)
Non-Maori	3.4	68.0	5.0	(2.6 - 7.3)
Total epithelial neoplasms	**5.0**	**100.0**	**5.5**	**(3.4 - 7.7)**

References

1. Sullivan, M., & Ballantine K. (2014). *The incidence of childhood cancer in New Zealand 2000-2009: The first outcome analysis of the New Zealand Children's Cancer Registry.* Auckland: National Child Cancer Network.

2. Ministry of Health. (2016). *Cancer: New Registrations and Deaths 2013*. Wellington: Ministry of Health.

3. Craig E, Jackson C, Han, D.Y, & NZCYES Steering Committee. (2007). *Monitoring the Health of New Zealand Children and Young People: Indicator Handbook*. Auckland: Paediatric Society of New Zealand, New Zealand: Child and Youth Epidemiology Service

4. Stiller, CA. (2007). Childhood cancer in Britain: Incidence, survival, mortality. Oxford: Oxford University Press.

5. Ministry of Health (2013). *New Zealand Cancer Registry (NZCR)*. Retrieved from: http://www.health.govt.nz/ nz-health-statistics/national-collections-and-surveys/collections/new-zealand-cancer-registry-nzcr

6. Steliarova-Foucher, E., Stiller, C., Lacour, B. & Kaatsch, P. (2005). International classification of childhood cancer, third edition. *Cancer,* 103, 1457-1467.

7. Birch, J., & Marsden, H. (1987). A classification scheme for childhood cancer. *International Journal of Cancer, 40,* 620-624.

8. International Agency for Research on Cancer. (1988). *International Incidence of Childhood Cancer, Vol. II.* IARC Scientific Publications, 1-391.

9. Fritz, A., Percy, C., Jack, A., Shanmugarathnam, K., Sobin, L., Parkin, D., & Whelan, S., eds. (2000). *International Classification of Diseases for Oncology, 3rd edition.* Geneva: World Health Organization.

10. Fritz, A., Percy, C., Jack, A., Shanmugarathnam, K., Sobin, L., Parkin, D., & Whelan, S., eds. (2013). *International Classification of Diseases for Oncology, 3rd edition, 1st revision.* Geneva: World Health Organization.

11. Ministry of Health (2004). *Ethnicity Data Protocols for the Health and Disability Sector.* Wellington: Ministry of Health.

12. Steliarova-Foucher, E., Colombet, M., Ries, LAG, et al. (2017). International incidence of childhood cancer, 2001-2010: a population-based registry study. *The Lancet Oncology, 18*(6), 719-37.

Appendices

AI: Abbreviations

AML	Acute myeloid leukaemia
ALL	Acute lymphoblastic 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
ICCC-3	International Classification of Childhood Cancer, Third revision
ICD-10	International Statistical Classification of Diseases and Related Health Problems, Tenth revision
ICD-O-3	International Statistical Classification of Diseases for Oncology, Third edition
IICC	International Incidence of Childhood Cancer
LEAP	Children's Oncology Late Effects Assessment Programme
LCH	Langerhans cell histiocytosis
MELAA	Middle Eastern, Latin American and African
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	International Society of Paediatric Oncology
SIOPEL	International Childhood Liver Tumors Strategy Group
WHO	World Health Organisation

AII International Classification of Childhood Cancers, 3rd edition (ICCC-3)[5]

The ICCC-3 classifies childhood cancers according to the 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 histology and topography codes of ICD-O-3 to the ICCC-3 main diagnostic groups and subgroups.

Table II International Classification of Childhood Cancers, 3rd 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
VI. Renal tumours		

Table II (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
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
(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

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

Diagnostic group / subgroup	ICD-O-3 histology	ICD-O-3 site
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
(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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