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# The incidence of childhood cancer in New Zealand 2000 - 2009

The first outcome analysis of the New Zealand Children's Cancer Registry



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#### Foreword

New Zealand has a unique multicultural and ethnic mix of people of Maori, Pacific, Asian and European origins. The diverse make up of our community creates unique health needs, so it is essential we research and report on our own population if we are to understand the health and illness of New Zealanders.

The need to record and report the pattern of cancer seen in New Zealand children was first recognised over 40 years ago when, in 1969, Dr David Becroft, (Paediatric Pathologist), began recording new cases of cancer presenting to the Princess Mary Hospital for Children. By the 1980s, members of the Paediatric Oncology Coordinating 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 (Paediatrician, Wellington), who was passionate about establishing a nationwide children's cancer registry. In the early 1990s, Dr John Dockerty (Epidemiologist, Dunedin) reported on the incidence and survival of childhood cancer in a cohort of children with cancer diagnosed in the four years between 1990 and 1993. However, these early efforts to collect nationwide data and to define nature and outcome of childhood cancer in New Zealand were limited in that they were not able report a complete national picture of childhood cancer that could be compared to other developed countries, nor could the data inform the future development of child cancer services.

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. Development work on the New Zealand Children's Cancer Registry began in 2002 and the national registry went online with the launch of the Late Effects Assessment Programme in 2006. We now have accrued complete data on the diagnosis and outcome of all children diagnosed with cancer in New Zealand since the 1st of January 2000.

This report on the incidence of childhood cancer in New Zealand is one of a series of reports on the incidence and survival of cancer in children (0-14 years), and the incidence and survival of cancer in adolescents and young adults (AYA, 15-24 years). It represents the culmination of over ten years work by multiple individuals who established the current New Zealand Children's Cancer Registry and who contributed cancer registrations.

Here we report on all cancer registrations in the ten years between 2000 and 2009, with particular regard to the cancer diagnosis as classified by the International Classification of Childhood Cancers (version 3), and the age group, sex, and ethnicity of those diagnosed. The ten-year registration period has accrued the largest known cohort of Maori (262) and Pacific (131) children with cancer giving a unique opportunity to report for the first time a detailed analysis of the spectrum of cancer seen in these populations. This represents the most detailed and accurate analysis yet done on child cancer incidence in New Zealand.

We wish to acknowledge and thank all those involved and the financial support from the National Child Cancer Network.

Dr Michael Sullivan & Kirsten Ballantine May 2014



# **Executive Summary**

The purpose of this report is to provide a comprehensive analysis of the incidence of childhood cancer in New Zealand for the period of the 1<sup>st</sup> of January 2000 to the 31<sup>st</sup> of December 2009. A second companion report contains survival data for the same period. Cancer cases were primarily sourced from the New Zealand Children's Cancer Registry. In addition, cross-matching of all registrations held by the NZCCR and the New Zealand Cancer Registry for the study period was undertaken.

There were 1329 new cases of cancer in children (0-14) registered with NZCCR in the ten year period from 2000 to 2009, giving an overall New Zealand child cancer incidence rate of 149.3 per million per year. This overall incidence rate is comparable to that seen in other developed nations such as the United Kingdom, Germany, Australia, Canada and the United States. The most common cancers diagnosed in New Zealand children were leukaemias (34.4% of all cancers, 46 cases per year) and central nervous system tumours (21.4% of all cancers, 28 cases per year). Acute lymphoblastic leukaemias (ALL) accounted for over one in four (26.6%) of all new child cancer registrations within the study period. By gender, males were more likely to develop non-Hodgkin lymphomas, rhabdomyosarcomas and 'intracranial & intraspinal embryonal tumours' than females but less likely to be diagnosed with astrocytomas.

Overall cancer incidence was significantly higher for those aged 0-4 year (202.4 per million) compared to those aged 5-9 at diagnosis (119.4 per million) or 10-14 years (129.4 per million); 43.9% of all childhood cancers were diagnosed in children under five years of age. There was a significantly higher incidence of ALL, neuroblastoma, retinoblastoma and renal tumours among 0-4 year olds than older children. In contrast, malignant bone tumours, lymphomas and 'other epithelial neoplasms' were most commonly diagnosed among 10-14 year olds.

The pattern of cancer seen in children of Maori and Pacific children appears unique and has not previously been reported. We have recorded ethnicity data for all childhood cancer registrations and our analysis has been according to the New Zealand prioritised classification of ethnicity. Of the 1329 child cancer registrations, 262 (19.7%) were Maori, 131 (9.9%) were Pacific Peoples, and 936 (70.4%) were non-Maori/non-Pacific Peoples. The overall age-standardised incidence rate of cancer in Maori children at 131.1 per million was significantly lower than that seen in Pacific children (173.4 per million) or non-Maori/non-Pacific Peoples (158.1 per million). By diagnostic group, Maori had comparatively low incidence of ALL and astrocytomas.

While the overall age standardised rate for Pacific children of 173.4 per million was not significantly different to non-Maori/non-Pacific Peoples, Pacific children appeared to have a higher incidence of leukaemia (71.5 per million c.f. 52.4 per million for non-Maori/non-Pacific Peoples and 46.5 per million for Maori). There was also considerable variability in the incidence of malignant bone tumours (17.2 cases per million for Pacific Peoples c.f. 7.6 per million for non-Maori/non-Pacific Peoples and 7.0 per million for Maori) and germ cell tumours (15.9 per million c.f. 8.5 for Maori and 5.2 per million for non-Maori/non-Pacific Peoples). The reasons for these differences should be explored further as they most likely arise from a real biological predisposition to different cancer types in these population groups.

## 1 Introduction

#### 1.1 Childhood cancer in New Zealand

Although child cancers account for less than one percent of all cancers diagnosed each year,<sup>1</sup> cancer is nevertheless a significant health issue for New Zealand. Cancer remains the second most common cause of death, after traffic accidents, for 1-14 year olds<sup>2</sup> and around one in five hundred children will be diagnosed with cancer before the age of 15 years.<sup>3</sup> 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 comprehensive childhood cancer centres; the Starship Blood and Cancer Centre in Auckland and the Children's Haematology/Oncology Centre (CHOC) based in Christchurch Hospital. These two centres work closely with dedicated regional shared care services to ensure children can receive as much of their treatment as close to home as possible.

#### 1.2 NZCCR background and purpose

Although diagnostic pathology laboratories are mandated by law to report all cancer diagnoses to the New Zealand Cancer Registry (NZCR)<sup>4</sup>, the data collected for each patient is necessarily limited. The NZCCR was established in 2002 at the request of the Ministry of Health (MOH) to provide more detailed information about childhood cancer incidence, treatment and outcomes in New Zealand. In addition to the data fields routinely collected by the NZCR, the NZCCR collects detailed clinical information regarding the stage of the disease, the treatments given, and treatment related late effects for all children receiving treatment in a paediatric oncology setting. The NZCCR also classifies all cancers according to the International Classification of Childhood Cancers (ICCC-3)<sup>5</sup>, which allows New Zealand child cancer incidence and survival to be directly compared with international benchmarks. The registry is under the governance of 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 by the Late Effects Clinical Nurse Specialist to produce the patient's end of treatment summary and surveillance plan. At a national level, the registry provides anonymised datasets used for service delivery planning, research, and statistical reporting purposes. The NZCCR has approval from the Southern 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 paediatric oncology specialist 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 involvement for diseases not currently included in the ICCC-3, such as langerhans cell histiocytosis (LCH), are excluded from any overall analysis.

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<sup>6</sup> 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).<sup>7</sup> 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, currently in its third edition (ICCC-3)<sup>5</sup> 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<sup>5</sup> 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<sup>8</sup>.

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

Table 1.5 Diagnostic groups of the International Classification of Childhood Cancers, 3rd edition (ICCC-3)<sup>1</sup>

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 a comprehensive analysis of the incidence of childhood cancer in New Zealand for the period of the 1<sup>st</sup> of January 2000 to the 31<sup>st</sup> of December 2009. A second companion report contains survival data for the same time period. Although this is the first time in which the NZCCR has been analysed, it is envisaged that the on-going input of new cancer diagnoses onto the NZCCR will allow for the continued reporting of the spectrum of childhood cancers and the tracking of overall child cancer incidence and survival over time.

Chapter 3 contains incidence data for children aged less than 15 years first diagnosed with cancer between the 1<sup>st</sup> of January 2000 and the 31<sup>st</sup> December 2009. Cancer incidence is reported by ICCC-3 diagnostic group and subgroup according to sex, age at diagnosis, and prioritised ethnicity<sup>1</sup>. 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 2006 New Zealand 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. 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. Note that given the small number of cases diagnosed in New Zealand, even over a ten-year period, there was not an adequate sample size to calculate the age-standardised incidence (and corresponding 95% confidence interval) in some instances. 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

This study was given expedited approval by the Multi-region Ethics Committee (ethics ref: MEC/11/EXP/052) in June 2011.

Prior to any data analysis of the NZCCR, a rigorous data validation process took place. As part of this process, all anomalies were investigated and any remaining data gaps were filled. The information contained on the registry was then cross-matched against the National Mortality Collection to ensure that all deaths had been recorded. Finally, cross-matching of all registrations held by the NZCCR and the NZCR for the study period was undertaken.

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<sup>5</sup>. 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 85 non-malignant CNS tumours included on the NZCCR but not the NZCR, including 47 children diagnosed with juvenile pilocytic astrocytoma<sup>a</sup>.

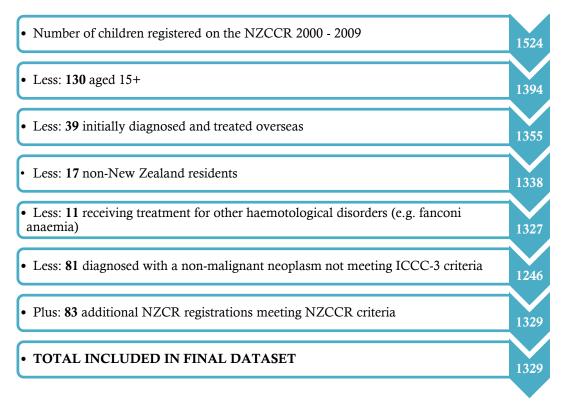
The data matching exercise also identified a small number of anomalies in demographic and diagnostic information between the two registries which required further investigation. Errors identified as part of this audit process were either corrected on the NZCR or provided to the NZCR for correction.

Finally, data matching with the NZCR led to the identification of 83 child cancer cases which were unknown to the NZCCR but which met NZCCR registration criteria. Although the referral of all child cancer cases to paediatric oncology services is mandatory, some children in this time period were treated exclusively by other specialties. For example, in the early 2000s some children diagnosed with retinoblastoma were treated by the ophthalmology service only. Also, some melanomas were treated by surgical resection only without the involvement of paediatric oncology services. Eight of the cases originally omitted by the NZCCR were informed by autopsy or death certificate only, and in an additional six cases the child died within a few days of diagnosis. Such cases would not have come to the attention of the paediatric oncology centres, and therefore were not registered on the NZCCR. All 83 valid cases informed by the NZCR have been included in the final dataset to ensure the accurate reporting of childhood cancer survival in New Zealand for the time period. Figure 2.1 provides a summary of the dataset selection process.

<sup>&</sup>lt;sup>a</sup> Juvenile pilocytic astrocytoma cases were included in the NZCR up until the end of 2002 but were no longer registered when the neoplasm was reclassified from 'malignant' to 'of uncertain and unknown behaviour' in the third version of the ICD-O, which was adopted by the NZCR on January 1<sup>st</sup> 2003.



#### Figure 2.1 Selection of the final dataset



#### 2.2 Prioritised ethnicity

According to MOH ethnicity data protocols, individuals may select up to three ethnic groups that they identify with. When a prioritised ethnicity system is used, each respondent is assigned to a single ethnic group using a priority system; Maori, Pacific Peoples, and non-Maori/non-Pacific Peoples (European, Asian, Middle Eastern, Latin American, 'Not Elsewhere Reported' and 'Not Stated/Unknown'). Assigning a single ethnicity simplifies the data as the ethnic group populations sum to the total New Zealand population, but there are limitations with prioritisation; an increasing number of New Zealand children and young people identify with more than one ethnic group and the use of prioritised ethnicity goes against the principle of self-identification. However, prioritised output is often used in the health and disability sector to ensure that Maori and Pacific Peoples, whose health status is lower on average than that of other New Zealanders, are not swamped by the European group. 9

Cancer registrations which had no ethnicity recorded were assigned to the 'non-Maori/non-Pacific Peoples' prioritised ethnic group. Although there was a very small number of 'unknown' ethnicity overall, the inclusion of the 'unknowns' within the 'non-Maori/non-Pacific Peoples' ethnic group has potentially slightly under-reported child cancer incidence for Maori and Pacific Peoples.

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

<sup>&</sup>lt;sup>a</sup> Note that ethnicity data is not reported directly to the NZCR, this information is obtained from the National Health Index (NHI), hospital discharge summaries, and the Mortality Collection.

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

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

<sup>&</sup>lt;sup>a</sup> MELAA: Middle Eastern, Latin American and African

#### 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 adolescents 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.

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



<sup>&</sup>lt;sup>b</sup> Includes 'response unidentifiable', 'response outside scope' and 'not stated'

<sup>&</sup>lt;sup>c</sup> Statistics NZ applies random rounding to base 3 to census outputs. As each value in a table is rounded independently, the marginal totals can differ very slightly from the corresponding sum of the rows or columns

#### 2.3.2 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 2006 New Zealand census population (see Table 2.3.2). 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 section 3 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, 2006 Census

	Male	Female	Total	NZ population Weightings (2006 Census)	Segi's world standard population weightings
0-4 years	140,382	134,697	275,076	0.3171	12,000
5-9 years	146,535	139,956	286,491	0.3302	10,000
10-14 years	157,113	148,893	306,009	0.3527	9,000
Total children 0-14 years	440,030	423,546	867,573		

#### 2.4 Confidence intervals, p-values, and statistical significance

A p-value of <0.05 indicates that two statistics are likely to be truly different from each other and unlikely to have occurred simply by chance. P-values are reported for overall childhood cancer incidence by age group and sex. P-values for ethnicity could not be calculated as the annual population estimates on which age-specific rates are calculated were available from Statistics New Zealand for sex and age only, not for prioritised ethnicity.

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 ten year period; this is reflected in the wide 95% confidence intervals which are reported alongside. Therefore, any between-group differences in the incidence or survival reported, or any differences in comparison to other published data, should be interpreted extremely cautiously.

## 3 Childhood Cancer Incidence

#### 3.1 Overall incidence of childhood cancer

There were 1329 new childhood cancer registrations in the 10 years between 2000 and 2009 out of a total average resident child population of 887,026, giving an overall New Zealand child cancer incidence rate of 149.3 per million per year (see Table 3.1). This incidence rate is not substantially higher than those reported for New Zealand between 1990 and 1993<sup>10</sup> (134.6 per million for boys and 127.2 per million for girls), given that non-malignant CNS tumours were not included in the earlier study.

The annual incidence rate was significantly higher for those aged 0-4 years (202.4 per million) than those 5-9 years at diagnosis (119.4 per million) and 10-14 years (129.4 per million); 43.9% of all child cancer cases were diagnosed in children under five years of age. There were no gender differences in overall child cancer incidence. By prioritised ethnicity, overall cancer incidence was significantly lower for Maori (131.1 per million) compared with non-Maori/non-Pacific Peoples (158.1 per million).

Table 3.1 Childhood cancer incidence by sex, age group, and ethnicity, New Zealand, 2000-2009

	Average cases per year	%	Population base <sup>a</sup>	% of the total child population	Age standardised rate <sup>b</sup> (95% CI)	
All childhood cancers	132.9	100	887 026	100	149.3	(141.2 - 157.3)
Sex (p=0.278)						
Male	70.1	52.7	454 755	51.3	153.6	(142.2 - 164.9)
Female	62.8	47.3	432 271	48.7	144.7	(133.4 - 156.1)
Age group (p<0.001) <sup>c</sup>						
0-4 years	58.3	43.9	288 034	32.5	202.4	(186.0 - 218.8)
5-9 years	35.0	26.3	293 017	33.0	119.4	(106.9 - 132.0)
10-14 years	39.6	29.8	305 975	34.5	129.4	(116.7 - 142.2)
Prioritised ethnicity <sup>d</sup>						
Maori	26.2	19.7	199 920	23.0	131.1	(115.2 - 146.9)
Pacific Peoples	13.1	9.9	75 537	8.7	173.4	(143.7 - 203.1)
Non-Maori/non-Pacific Peoples	93.6	70.4	592 119	68.3	158.1	(147.9 - 168.2)

<sup>&</sup>lt;sup>a</sup> 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 2000-2009 published by Statistics New Zealand. Incidence by prioritised ethnicity is based on 2006 New Zealand census population data.

<sup>&</sup>lt;sup>b</sup> Rate per million population per year, age-standardised to the 2006 New Zealand Standard Population. Age-specific incidence is reported for each age quintile.

<sup>&</sup>lt;sup>c</sup> Cancer incidence at 0-4 years was significantly higher than the incidence at 5-9 years and 10-14 years (p<0.001). Cancer incidence at 10-14 years was not significantly higher than 5-9 years (p=0.274)

<sup>&</sup>lt;sup>d</sup> p-values for ethnicity could not be calculated as the annual population estimates on which rates are calculated were available for sex and age only, not for prioritised ethnicity. However, if two statistics have non-overlapping 95% confidence intervals, they are necessarily significantly different at the p<0.05 level.

#### 3.2 Annual number of new child cancer registrations

Table 3.2 serves to show not only the large natural fluctuation in the number of new cases diagnosed each year, but also the small number of cases involved. In the period 2000 to 2009 the number of new child cancer registrations ranged from 116 to 159 cases per year. Note that the number of new cases diagnosed is less than the number of children who underwent cancer treatment in that year, as the standard cancer treatment for many cancers, such as acute lymphoblastic leukaemia (ALL), is up to three years duration.

Table 3.2 Annual number of child cancer registrations in New Zealand, 2000-2009

		Male			Female			Total	
Year	No. of cases	Population	Age standardised rate <sup>a</sup>	No. of cases	Population	Age standardised rate <sup>1</sup>	No. of cases	Population	Age standardised rate <sup>1</sup>
2000	97	451 210	212.2	62	427 540	144.8	159	878 750	179.5
2001	73	450 400	161.7	62	426 820	145.4	135	877 220	153.8
2002	66	452 910	145.5	50	430 660	116.0	116	883 570	131.1
2003	75	456 270	164.4	73	433 740	168.4	148	890 010	166.3
2004	55	457 720	120.0	71	435 360	163.0	126	893 080	141.0
2005	62	456 330	135.9	64	433 920	147.2	126	890 250	141.4
2006	73	454 960	159.9	59	433 360	135.7	132	888 320	148.1
2007	57	455 080	124.6	64	433 360	146.8	121	888 440	135.4
2008	63	455 770	138.7	79	433 640	181.4	142	889 410	159.5
2009	80	456 900	174.0	44	434 310	100.6	124	891 210	138.2

<sup>&</sup>lt;sup>a</sup> Rate per million population per year, age-standardised to the 2006 New Zealand Standard Population

#### 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 142.4 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, 2000-2009

		0-4 ye	ears		5-9 ye	ars		10-14	years
	Cases	Popula- tion	Age-specific incidence rate	Cases	Popula- tion	Age-specific incidence rate	Cases	Popula- tion	Age-specific incidence rate
		base <sup>a</sup>	(per million)		base <sup>a</sup>	(per million)		base <sup>a</sup>	(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
Average (95% CI)	58.3	288 034	202.4 (186.0 - 218.8)	35.0	293 017	119.4 (106.9 - 132.0)	39.6	305 975	129.4 (116.7 - 142.2)

<sup>&</sup>lt;sup>a</sup> Estimated New Zealand resident population as at June 30, Statistics New Zealand

#### 3.4 Childhood cancer incidence by diagnostic group

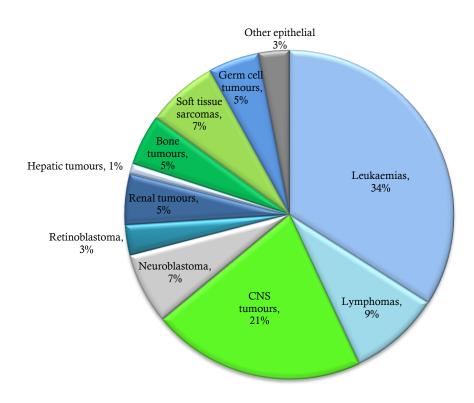
Between 2000 and 2009, leukaemias accounted for a third of childhood cancers (34.4%, 45.7 average cases per year, see Table 3.4 and Figure 3.4). Tumours of the central nervous system tumours were the next most frequently diagnostic class of tumours, accounting for around one in five childhood cancers diagnosed. Hepatic tumours were the rarest diagnostic group seen in New Zealand, with an average of only 1.3 cases per year.

Table 3.4 Childhood cancer incidence by diagnostic group, New Zealand, 2000-2009

	Diagnostic Group (ICCC-3)	Average cases per year	%		andardised rate <sup>a</sup> (95% CI)
	All childhood cancers	132.9	100	149.3	(141.2 - 157.3)
I.	Leukaemias, myeloproliferative & myelodysplastic diseases	45.7	34.4	51.2	(46.5 - 55.8)
II.	Lymphoma & reticuloendothelial neoplasms	11.4	8.6	13.0	(10.6 - 15.4)
III.	Central nervous system & intracranial/intraspinal neoplasms	28.4	21.4	32.0	(28.3 - 35.7)
IV.	Neuroblastoma & other peripheral nervous cell tumours	8.9	6.7	9.9	(7.8 - 11.9)
V.	Retinoblastoma	3.9	2.9	4.3	(3.0 - 5.7)
VI.	Renal tumours	6.1	4.6	6.8	(5.1 - 8.5)
VII.	Hepatic tumours	1.3	1.0	1.4	(0.7 - 2.2)
VIII.	Malignant bone tumours	7.2	5.4	8.2	(6.3 - 10.1)
IX.	Soft tissue and other extraosseous sarcomas	9.4	7.1	10.6	(8.4 - 12.7)
X.	Germ cell tumours, trophoblastic tumours & neoplasms of gonads	6.0	4.5	6.8	(5.1 - 8.5)
XI.	Other epithelial neoplasms & melanomas	4.2	3.2	4.8	(3.4 - 6.3)
XII.	Other & unspecified malignant neoplasms	0.4	0.3	0.4	(0.0 - 0.9)

<sup>&</sup>lt;sup>a</sup> Rate per million population per year, age-standardised to the 2006 New Zealand Standard Population

Figure 3.4 Childhood cancer by diagnostic group, New Zealand, 2000-2009



#### 3.5 Childhood cancer incidence by diagnostic subgroup

With a total number of 353 cases and an incidence rate of 39.5 per million per year, over one in four new cancer cases (26.6%) registered from 2000 to 2009 were classified as acute lymphoblastic leukaemia (see Table 2.5). The other ICCC-3 diagnostic subgroups most frequently diagnosed in New Zealand children were astrocytomas (117 cases, 8.8% of all cancer registrations for the period), neuroblastoma (87 cases per year, 6.5% of all cancer registrations), and AML (79 cases per year, 5.9% of all cancer registrations).

Table 3.5 Childhood cancer incidence by diagnostic group and subgroup, New Zealand, 2000-2009

	ICCC-3 Diagnostic Group / Subgroup	Total Cases	%		Age standardised rate <sup>a</sup> (95% CI)		
	All childhood cancers	1329	100	149.3	(141.2 - 157.3)		
I.	Leukaemias, myeloproliferative & myelodysplastic diseases	457	34.4	51.2	(46.5 - 55.8)		
Ia.	Lymphoid leukaemias	353	26.6	39.5	(35.4 - 43.6)		
Ib.	Acute myeloid leukaemias	79	5.9	8.9	(6.9 - 10.8)		
Ic.	Chronic myeloproliferative diseases	4	0.3	b	b		
Id.	Other myeloproliferative diseases	11	0.8	1.2	(0.5 - 1.9)		
Ie.	Other and unspecified leukaemia	10	0.8	1.1	(0.4 - 1.8)		
II.	Lymphoma & reticuloendothelial neoplasms	114	8.6	13.0	(10.6 - 15.4)		
IIa.	Hodgkin lymphomas	36	2.7	4.1	(2.8 - 5.5)		
IIb.	Non-Hodgkin lymphomas (excl. Burkitt lymphomas)	58	4.4	6.6	(4.9 - 8.3)		
IIc.	Burkitt lymphomas	18	1.4	2.0	(1.1 - 3.0)		
IId.	Miscellaneous lymphoreticular neoplasms	1	0.1	b	b		
IIe.	Unspecified lymphomas	1	0.1	b	b		
ш.	Central nervous system & intracranial/intraspinal neoplasms	284	21.4	32.0	(28.3 - 35.7)		
IIIa.	Ependymomas and choroid plexus tumours	24	1.8	2.7	(1.6 - 3.8)		
IIIb.	Astrocytomas	117	8.8	13.2	(10.8 - 15.6)		
IIIc.	Intracranial and intraspinal embryonal tumours	59	4.4	6.6	(4.9 - 8.3)		
IIId.	Other gliomas	45	3.4	5.1	(3.6 - 6.6)		
IIIe.	Other specified intracranial and intraspinal neoplasms	36	2.7	4.1	(2.7 - 5.4)		
IIIf.	Unspecified intracranial and intraspinal neoplasms	3	0.2	b	b		
IV.	Neuroblastoma & other peripheral nervous cell tumours	89	6.7	9.9	(7.8 - 11.9)		
IVa.	Neuroblastoma & ganglioneuroblastoma	87	6.5	9.6	(7.6 - 11.7)		
IVb.	Other peripheral nervous cell tumours	2	0.2	ь	b		
V.	Retinoblastoma	39	2.9	4.3	(3.0 - 5.7)		
VI.	Renal tumours	61	4.6	6.8	(5.1 - 8.5)		
VIa.	Nephroblastoma & other non-epithelial renal tumours	59	4.4	6.5	(4.9 - 8.2)		
VIb.	Renal carcinomas	2	0.2	b	b		
Vic.	Unspecified malignant renal tumours	-	-	-	-		
VII.	Hepatic tumours	13	1.0	1.4	(0.7 - 2.2)		
VIIa.	Hepatoblastoma	8	0.6	b	b		
VIIb.	Hepatic carcinomas	5	0.4	b	b		
VIIc.	Unspecified malignant hepatic tumours	-	-	-	-		
VIII.	Malignant bone tumours	72	5.4	8.2	(6.3 - 10.1)		
VIIIa.	Osteosarcomas	37	2.8	4.2	(2.8 - 5.6)		
VIIIb.	Chondrosarcomas	1	0.1	b	b		
VIIIc.	Ewing tumours & related bone sarcomas	28	2.1	3.2	(2.0 - 4.4)		
VIIId.	Other specified malignant bone tumours	5	0.4	b	b		
VIIIe.	Unspecified malignant bone tumours	1	0.1	b	b		
IX.	Soft tissue and other extraosseous sarcomas	94	7.1	10.6	(8.4 - 12.7)		
IXa.	Rhabdomyosarcomas	50	3.8	5.6	(4.1 - 7.2)		
IXb.	Fibrosarcomas & other fibrous neoplasms	7	0.5	b	b		
IXc.	Kaposi sarcomas	-	-	-	-		
IXd.	Other specified soft tissue sarcomas	27	2.0	3.1	(1.9 - 4.2)		
IXe.	Unspecified soft tissue sarcomas	10	0.8	b	b		

<sup>&</sup>lt;sup>a</sup> Rate per million population per year, age-standardised to the 2006 New Zealand Standard Population

<sup>&</sup>lt;sup>b</sup> Age standardised rates (and corresponding 95% CI) have been censored for diagnostic subgroups where there were fewer than 10 cases diagnosed within the ten-year period



Table 3.5 (cont.) Childhood cancer incidence by diagnostic group and subgroup, New Zealand, 2000-2009

	ICCC-3 Diagnostic Group / Subgroup	Total Cases	%		ndardised rate <sup>a</sup> 95% CI)
X.	Germ cell tumours, trophoblastic tumours & neoplasms of gonads	60	4.5	6.8	(5.1 - 8.5)
Xa.	Intracranial & intraspinal germ cell tumours	18	1.4	2.1	(1.1 - 3.0)
Xb.	Malignant extracranial & extragonadal germ cell tumours	19	1.4	2.1	(1.2 - 3.1)
Xc.	Malignant gonadal germ cell tumours	23	1.7	2.6	(1.5 - 3.7)
Xd.	Gonadal carcinomas	-	-	-	-
Xe.	Other & unspecified malignant gonadal tumours	-	-	-	-
XI.	Other epithelial neoplasms & melanomas	42	3.2	4.8	(3.4 - 6.3)
XIa.	Adrenocortical carcinomas	2	0.2	b	b
XIb.	Thyroid carcinomas	6	0.5	b	b
XIc.	Nasopharyngeal carcinomas	2	0.2	b	b
XId.	Melanomas	16	1.2	1.8	(0.9 - 2.7)
XIe.	Skin carcinomas	-	-	-	-
XIf.	Other & unspecified carcinomas	16	1.2	1.8	(0.9 - 2.7)
XII.	Other & unspecified malignant neoplasms	4	0.3	0.4	(0.0 - 0.9)
XIIa.	Other specified malignant tumours	3	0.2	b	b
XIIb.	Other unspecified malignant tumours	1	0.2	b	b

<sup>&</sup>lt;sup>a</sup> Rate per million population per year, age-standardised to the 2006 New Zealand Standard Population

#### 3.6 Childhood cancer incidence by sex

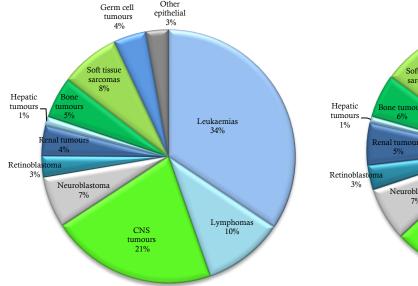
#### 3.6.1 Distribution of childhood cancers by sex

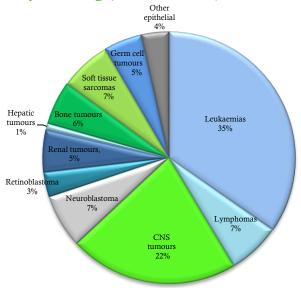
Figures 3.6.1a-3.6.1c show that there were few sex differences in the number of cancer cases diagnosed within each diagnostic group. The notable exception to this was lymphomas; of the 114 lymphomas diagnosed in the ten year period 72 cases (63%) were diagnosed in males.

Figure 3.6.1a Cancers diagnosed in males 0-14 years of age, New Zealand, 2000-2009

Other

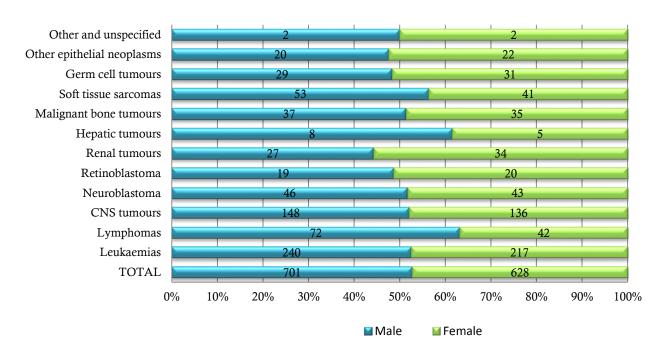
Figure 3.6.1b Cancers diagnosed in females 0-14 years of age, New Zealand, 2000-2009





<sup>&</sup>lt;sup>b</sup> Age standardised rates (and corresponding 95% CI) have been censored for diagnostic subgroups where there were fewer than 10 cases diagnosed within the ten-year period

Figure 3.6.1c Number of childhood cancer cases by sex, New Zealand, 2000-2009



#### 3.6.2 Childhood cancer incidence by sex and diagnostic group and subgroup

Males were 1.6 times more likely to be diagnosed with lymphomas than females; lymphomas represented 10.3% of all cancer cases for males (16.0 per million per year, 72 cases) but only 6.7% of all cancers for females (9.8 per million per year, 42 cases). This was almost entirely due to their significantly higher incidence of non-Hodgkin lymphomas; males were 2.5 times more likely to be diagnosed with a non-Hodgkin lymphoma than females (42 cases c.f. 16 cases). Males were also 2.2 times more likely to develop a rhabdomyosarcoma (35 cases c.f. 15 cases) and 2.8 times more likely to develop an intracranial & intraspinal embryonal tumour (44 cases c.f. 15 cases) but were significantly less likely to be diagnosed with an astrocytoma (relative risk 0.6, 47 cases c.f. 70 cases).

Table 3.6.2 Childhood cancer incidence by sex and diagnostic group and subgroup, New Zealand, 2000-2009

			Male	Fϵ	male	D ol	Relative Risk		
	Diagnostic Group / Subgroup	Average cases per year	Age standardised rate <sup>a</sup>	Average cases per year	Age standardised rate <sup>a</sup>	Male	e to Female 95% CI)		
	All childhood cancers	70.1	153.6	62.8	144.7	1.06	(0.95 - 1.18)		
I.	Leukaemias, myeloproliferative & myelodysplastic diseases	24.0	52.4	21.7	49.9	1.05	(0.87 - 1.26)		
Ia.	Lymphoid leukaemias	19.3	42.1	16.0	36.8	1.14	(0.93 - 1.41)		
Ib.	Acute myeloid leukaemias	3.5	7.7	4.4	10.1	0.76	(0.49 - 1.19)		
Ic.	Chronic myeloproliferative diseases	0.2	0.5	0.2	0.5	b	b		
Id.	Other myeloproliferative diseases	0.5	1.1	0.6	1.4	b	b		
Ie.	Other and unspecified leukaemia	0.5	1.1	0.5	1.2	b	b		
Π.	Lymphoma & reticuloendothelial neoplasms	7.2	16.0	4.2	9.8	1.63	(1.12 - 2.38)		
IIa.	Hodgkin lymphomas	1.7	3.8	1.9	4.4	0.85	(0.44 - 1.64)		
IIb.	Non-Hodgkin lymphomas (excl. Burkitt lymphomas)	4.2	9.4	1.6	3.7	2.51	(1.45 - 4.35)		
IIc.	Burkitt lymphomas	1.2	2.6	0.6	1.4	b	b		
IId.	Miscellaneous lymphoreticular neoplasms	-	-	0.1	c	b	b		
IIe.	Unspecified lymphomas	0.1	c	-	-	b	b		

<sup>&</sup>lt;sup>a</sup> Rate per million population per year, age-standardised to the 2006 New Zealand Standard Population

<sup>&</sup>lt;sup>b</sup> Relative risk was not calculated due to the small number of cases recorded in this diagnostic subgroup
<sup>c</sup> Age standardised rates (and corresponding 95% CI) have been censored for diagnostic subgroups where there were fewer than 10 cases diagnosed within the ten-year period

Table 3.6.2 (cont.) Childhood cancer incidence by sex and diagnostic group and subgroup, New Zealand, 2000-2009

Diagnostic Group / Subgroup				Male	F	emale		
Central nervous system & intracranial/intraspinal neoplasms   14.8   32.5   13.6   31.4   1.03   (0.82 - 1.30)							Relativ	
		Diagnostic Group / Subgroup			_			
III.								(95% CI)
		Central nervous system & intracranial/intrasninal	i i		yeur	Tute		
III.a   Ependymomas and choroid plexus tumours	III.	•	14.8	32.5	13.6	31.4	1.03	(0.82 - 1.30)
Illic.   Intracranial and intraspinal embryonal tumours	IIIa.		1.5	3.3	0.9	2.1	ь	b
Illic.   Intracranial and intraspinal embryonal tumours	IIIb.		4.7	10.3	7.0	16.2	0.64	(0.44 - 0.92)
III.d.   Other gliomas	IIIc.	Intracranial and intraspinal embryonal tumours	4.4	9.6	1.5	3.5	2.78	
Other specified intracranial and intraspinal   1.8   3.9   1.8   4.2   0.94   (0.49-1.81)				4.6	2.4	5.5	0.83	
	Ша		1 0	2.0	1 0	4.2	0.04	(0.40 1.91)
In		neoplasms	1.8		1.8	4.2		` ,
	IIIf.		0.3	c	-	-	ь	b
New	IV.		4.6	9.9	4.3	9.8	1.02	(0.67 - 1.54)
IVD.   Other peripheral nervous cell tumours   1.9			11		17			` /
V.   Renal tumours   1.9						9.8		
VI.   Renal tumours								
Nephroblastoma & other non-epithelial renal tumours   0.1   c   0.1   c   b   b								
National	VI.		2.7	5.8	3.4	7.8	0.75	(0.45 - 1.25)
VID.   Renal carcinomas   0.1   c   0.1   c   b   b	VIa.		2.6	5.6	3.3	7.5	0.75	(0.45 - 1.25)
No.	VII		0.1	c	0.1	c	ь	b
VII.         Hepatic tumours         0.8         1.7         0.5         1.1         b         b           VIIa.         Hepatobalstoma         0.5         c         0.3         c         b         b           VIID.         Hepatic carcinomas         0.3         c         0.2         c         b         b           VIII.         Unspecified malignant hepatic tumours         -         -         -         -         b         b           VIII.         Malignant bone tumours         2.0         4.5         1.7         4.0         1.12         (0.59 -2.13)           VIIII.         Chondrosarcomas         0.1         c         -         -         b         b         b           VIIII.         Chondrosarcomas         0.1         c         -         -         b         b         b           VIIII.         Chondrosarcomas         1.4         3.1         1.4         3.3         b         b         b         b           VIII.6.         Chondrosarcomas         1.4         3.1         1.4         3.3         c         b         b         b           VIII         Chordres gredifed malignant bone tumours         1.2         c			0.1	-	0.1			b
			0.0	1.7	0.5	1.1	b	
Name								
No.							b	
VIII.   Malignant bone tumours   3.7   8.3   3.5   8.2   1.01   (0.64 - 1.60)								
VIIIa.         Osteosarcomas         2.0         4.5         1.7         4.0         1.12         (0.59 - 2.13)           VIIIb.         Chondrosarcomas         0.1         c         -         -         b         b           VIIIc.         Ewing tumours & related bone sarcomas         1.4         3.1         1.4         3.3         b         b           VIIId.         Other specified malignant bone tumours         0.2         c         0.3         c         b         b           VIIIe.         Unspecified malignant bone tumours         -         -         0.1         0.2         b         b           VIIIe.         Unspecified and dene extraosseous sarcomas         3.5         7.6         1.5         3.5         2.22         (1.24 - 3.97)           IX.         Soft tissue and other extraosseous sarcomas         3.5         7.6         1.5         3.5         2.22         (1.24 - 3.97)           IXb.         Fibrosarcomas & other fibrous neoplasms         0.2         c         0.5         c         b         b         b           IXc.         Agposi sarcomas         0.2         c         0.5         c         b         b         b           IXc.         Unspecified soft tissue								
VIIIb.         Chondrosarcomas         0.1         c         -         -         b         b           VIIIc.         Ewing tumours & related bone sarcomas         1.4         3.1         1.4         3.3         b         b           VIIId.         Other specified malignant bone tumours         0.2         c         0.3         c         b         b           VIIIe.         Unspecified malignant bone tumours         -         0.1         0.2         b         b           IX.         Soft tissue and other extraosseous sarcomas         5.3         11.6         4.1         9.5         1.23         (0.82 - 1.84)           IXa.         Rhabdomyosarcomas         3.5         7.6         1.5         3.5         2.22         (1.24 - 3.97)           IXb.         Fibrosarcomas & other fibrous neoplasms         0.2         c         0.5         c         b         b           IXc.         Kaposi sarcomas         -         -         -         -         -         -         -         b         b           IXd.         Other specified soft tissue sarcomas         0.7         1.6         0.3         0.7         b         b           IXe.         Unspecified soft tissue sarcomas <t< td=""><td></td><td></td><td></td><td></td><td></td><td></td><td></td><td></td></t<>								
Villic   Ewing tumours & related bone sarcomas   1.4   3.1   1.4   3.3   b   b						4.0		
VIIId.         Other specified malignant bone tumours         0.2         c         0.3         c         b         b           VIIIe.         Unspecified malignant bone tumours         -         0.1         0.2         b         b           VIIIe.         Unspecified malignant bone tumours         -         0.1         0.2         b         b           IX.         Soft tissue and other extraosseous sarcomas         3.5         7.6         1.5         3.5         2.22         (1.24 · 3.97)           IXa.         Rhabdomyosarcomas         0.2         c         0.5         c         b         b         b           IXb.         Fibrosarcomas & other fibrous neoplasms         0.2         c         0.5         c         b						2 2	ь	b
VIIIe.         Unspecified malignant bone tumours         -         -         0.1         0.2         b         b           IX.         Soft tissue and other extraosseous sarcomas         5.3         11.6         4.1         9.5         1.23         (0.82 - 1.84)           IXa.         Rhabdomyosarcomas         3.5         7.6         1.5         3.5         2.22         (1.24 - 3.97)           IXb.         Fibrosarcomas & other fibrous neoplasms         0.2         c         0.5         c         b         b           IXc.         Kaposi sarcomas         0.2         -         -         -         -         -         -         -         -         b         b           IXd.         Other specified soft tissue sarcomas         0.7         1.6         0.3         0.7         b         b           IXe.         Unspecified soft tissue sarcomas         0.7         1.6         0.3         0.7         b           IXe.         Unspecified soft tissue sarcomas         0.7         1.6         0.3         0.7         b           X.         Germ cell tumours, trophoblastic tumours         2.9         6.3         3.1         7.2         0.88         (0.53 - 1.47)           Xb.         <							ь	b
IX.         Soft tissue and other extraosseous sarcomas         5.3         11.6         4.1         9.5         1.23         (0.82 - 1.84)           IXa.         Rhabdomyosarcomas         3.5         7.6         1.5         3.5         2.22         (1.24 - 3.97)           IXb.         Fibrosarcomas & other fibrous neoplasms         0.2         c         0.5         c         b         b           IXc.         Kaposi sarcomas         -         -         -         -         b         b         b           IXd.         Other specified soft tissue sarcomas         0.7         1.6         0.3         0.7         b         b         b           IXe.         Unspecified soft tissue sarcomas         0.7         1.6         0.3         0.7         b         b           IXe.         Unspecified soft tissue sarcomas         0.7         1.6         0.3         0.7         b         b           IXe.         Unspecified soft tissue sarcomas         0.7         1.6         0.3         0.7         b         b           X.         Germ cell tumours, trophoblastic tumours         0.8         1.8         1.0         2.3         b         b           Xb.         Malignant extracranial & extragon							ь	b
IXa.   Rhabdomyosarcomas   3.5   7.6   1.5   3.5   2.22   (1.24 - 3.97)     IXb.   Fibrosarcomas & other fibrous neoplasms   0.2   c   0.5   c   b   b     IXc.   Kaposi sarcomas   -   -   -   -   b   b     IXd.   Other specified soft tissue sarcomas   0.9   2.0   1.8   4.2   b   b     IXe.   Unspecified soft tissue sarcomas   0.7   1.6   0.3   0.7   b     IXe.   Unspecified soft tissue sarcomas   0.7   1.6   0.3   0.7   b     IXe.   Unspecified soft tissue sarcomas   0.7   1.6   0.3   0.7   b     IXe.   Unspecified soft tissue sarcomas   0.7   1.6   0.3   0.7   b     IXe.   Unspecified soft tissue sarcomas   0.7   1.6   0.3   0.7   b     IXe.   Unspecified soft tissue sarcomas   0.7   1.6   0.3   0.7     IXe.   Unspecified soft tissue sarcomas   0.7   1.6   0.3   0.7     IXe.   Unspecified & sarcomas   0.8   1.8   1.0   2.3   b   b     Ixe.   Unspecified soft tissue sarcomas   0.7   1.5   1.2   2.7   b     Ixe.   Unspecified soft tissue sarcomas   0.7   1.5   1.2   2.7   b     Ixe.   Unspecified soft tissue sarcomas   0.7   1.5   1.2   2.7   b     Ixe.   Unspecified soft tissue sarcomas   0.7   1.5   1.2   2.7   b     Ixe.   Unspecified sarcinomas   0.8   0.9   2.1   b     Ixe.   Unspecified sarcinomas   0.2   c   -   -   b     Ixe.   Unspecified malignant tumours   0.2   c   -   -   b     Ixe.   Unspecified malignant tumours   0.2   0.4   0.2   0.5     Ixe.   Unspecified malignan							1 22	(0.92 1.94)
IXb.         Fibrosarcomas & other fibrous neoplasms         0.2         c         0.5         c         b         b           IXc.         Kaposi sarcomas         -         -         -         -         -         b         b           IXc.         Other specified soft tissue sarcomas         0.9         2.0         1.8         4.2         b         b           IXe.         Unspecified soft tissue sarcomas         0.7         1.6         0.3         0.7         b         b           X.         Germ cell tumours, trophoblastic tumours & neoplasms of gonads         2.9         6.3         3.1         7.2         0.88         (0.53 - 1.47)           X.         Germ cell tumours, trophoblastic tumours & neoplasms of gonads         0.8         1.8         1.0         2.3         b         b           X.         Malignant gonadal germ cell tumours         0.8         1.8         1.0         2.3         b         b           Xb.         Malignant gonadal germ cell tumours         0.7         1.5         1.2         2.7         b         b           Xc.         Malignant gonadal germ cell tumours         1.4         3.0         0.9         2.1         b         b           Xb.         Other &								
IXc.   Kaposi sarcomas   -   -   -   -     -								
IXd.         Other specified soft tissue sarcomas         0.9         2.0         1.8         4.2         b         b           IXe.         Unspecified soft tissue sarcomas         0.7         1.6         0.3         0.7         b         b           X.         Germ cell tumours, trophoblastic tumours & neoplasms of gonads         2.9         6.3         3.1         7.2         0.88         (0.53 - 1.47)           Xa.         Intracranial & intraspinal germ cell tumours         0.8         1.8         1.0         2.3         b         b           Xb.         Malignant extracranial & extragonadal germ cell tumours         0.7         1.5         1.2         2.7         b         b           Xc.         Malignant gonadal germ cell tumours         1.4         3.0         0.9         2.1         b         b           Xd.         Gonadal carcinomas         -         -         -         -         -         -         b         b           Xd.         Other & unspecified malignant gonadal tumours         -         -         -         -         -         -         -         -         -         -         -         -         -         -         -         -         -         -         -				_		_	ь	b
IXe.         Unspecified soft tissue sarcomas         0.7         1.6         0.3         0.7         b         b           X.         Germ cell tumours, trophoblastic tumours & neoplasms of gonads         2.9         6.3         3.1         7.2         0.88         (0.53 - 1.47)           Xa.         Intracranial & intraspinal germ cell tumours         0.8         1.8         1.0         2.3         b         b           Xb.         Malignant extracranial & extragonadal germ cell tumours         0.7         1.5         1.2         2.7         b         b           Xc.         Malignant gonadal germ cell tumours         1.4         3.0         0.9         2.1         b         b           Xd.         Gonadal carcinomas         -         -         -         -         -         -         b         b           Xe.         Other & unspecified malignant gonadal tumours         -         -         -         -         -         -         -         -         b         b           XI.         Other epithelial neoplasms & melanomas         2.0         4.5         2.2         5.2         0.87         (0.47 - 1.59)           XIa.         Adrenocortical carcinomas         0.2         0.2         0.3 <t< td=""><td></td><td></td><td></td><td></td><td></td><td></td><td>ь</td><td>b</td></t<>							ь	b
X.         Germ cell tumours, trophoblastic tumours & neoplasms of gonads         2.9         6.3         3.1         7.2         0.88         (0.53 - 1.47)           Xa.         Intracranial & intraspinal germ cell tumours         0.8         1.8         1.0         2.3         b         b           Xb.         Malignant extracranial & extragonadal germ cell tumours         0.7         1.5         1.2         2.7         b         b           Xc.         Malignant gonadal germ cell tumours         1.4         3.0         0.9         2.1         b         b           Xd.         Gonadal carcinomas         -         -         -         -         -         b         b           Xe.         Other & unspecified malignant gonadal tumours         -         -         -         -         -         -         b         b           XI.         Other epithelial neoplasms & melanomas         2.0         4.5         2.2         5.2         0.87         (0.47 - 1.59)           XII.         Other epithelial neoplasms & melanomas         0.2         -         -         -         -         -         -         b         b           XII.         Other occitical carcinomas         0.3         c         0.3 <td< td=""><td></td><td></td><td></td><td></td><td></td><td></td><td>ь</td><td>b</td></td<>							ь	b
Name of gonads   Name								
Xa.         Intracranial & intraspinal germ cell tumours         0.8         1.8         1.0         2.3         b         b           Xb.         Malignant extracranial & extragonadal germ cell tumours         0.7         1.5         1.2         2.7         b         b         b           Xc.         Malignant gonadal germ cell tumours         1.4         3.0         0.9         2.1         b         b         b           Xd.         Gonadal carcinomas         -         -         -         -         -         -         b         b         b           Xe.         Other & unspecified malignant gonadal tumours         -         -         -         -         -         -         -         -         -         -         b         b         b           XI.         Other epithelial neoplasms & melanomas         2.0         4.5         2.2         5.2         0.87         (0.47 - 1.59)         c         -	Х.		2.9	6.3	3.1	7.2	0.88	(0.53 - 1.47)
Xb.       Malignant extracranial & extragonadal germ cell tumours       0.7       1.5       1.2       2.7       b       b         Xc.       Malignant gonadal germ cell tumours       1.4       3.0       0.9       2.1       b       b         Xd.       Gonadal carcinomas       -       -       -       -       -       -       b       b         Xe.       Other & unspecified malignant gonadal tumours       -       -       -       -       -       -       b       b         XI.       Other epithelial neoplasms & melanomas       2.0       4.5       2.2       5.2       0.87       (0.47 - 1.59)         XIa.       Adrenocortical carcinomas       0.2       c       -       -       -       b       b         XIb.       Thyroid carcinomas       0.3       c       0.3       c       b       b         XIc.       Nasopharyngeal carcinomas       0.1       c       0.1       c       b       b         XId.       Melanomas       0.6       1.4       1.0       2.3       b       b         XIe.       Skin carcinomas       -       -       -       -       -       -       -       b       b	Xa.		0.8	1.8	1.0	2.3	ь	b
Xc.   Malignant gonadal germ cell tumours   1.4   3.0   0.9   2.1   b   b     Xd.   Gonadal carcinomas   -   -   -   -   b   b     Xe.   Other & unspecified malignant gonadal tumours   -   -   -   -   -   b   b     XI.   Other epithelial neoplasms & melanomas   2.0   4.5   2.2   5.2   0.87   (0.47 - 1.59)     XIa.   Adrenocortical carcinomas   0.2   c   -   -   b   b     XIb.   Thyroid carcinomas   0.3   c   0.3   c   b   b     XIc.   Nasopharyngeal carcinomas   0.1   c   0.1   c   b   b     XId.   Melanomas   0.6   1.4   1.0   2.3   b   b     XIe.   Skin carcinomas   0.8   1.9   b   b     XIf.   Other & unspecified malignant neoplasms   0.2   0.4   0.2   0.5   b     XIIa.   Other specified malignant tumours   0.2   c   0.1   c   b   b     XIIIa.   Other specified malignant tumours   0.2   c   0.1   c   b   b     XIIIa.   Other specified malignant tumours   0.2   c   0.1   c   b   b     XIIIa.   Other specified malignant tumours   0.2   c   0.1   c   c   b   b     XIIIa.   Other specified malignant tumours   0.2   c   0.1   c   c   b   b     XIIIa.   Other specified malignant tumours   0.2   c   0.1   c   c   c   b   b     XIIIa.   Other specified malignant tumours   0.2   c   0.1   c   c   c   c   c   c   c   c   c	VI.		0.7	1.5	1.2	2.7	b	h
Xd.         Integrating of a data germ cent tumours         1.4         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.9         2.1         3.0         0.0         5         6         6         6         7.0         2.0         4.2         4.3         2.2         5.2         0.87         (0.47 - 1.59)         6         7.0         2.0         4.5         2.2         5.2         0.87         (0.47 - 1.59)         6         6         7.0         2.0         4.0         6         1.0         2.0         8         1.0         8         1.0         8         1.0         8         1.0         8         1.0         8         1.0         8         1.0         8         1.0         8         1.0         8         1.0         8         1.0         8         1.0         8         1.0         8	AD.	tumours	0.7	1.5	1.2	2.1		
Xe.         Other & unspecified malignant gonadal tumours         -         -         -         -         -         b         b           XI.         Other epithelial neoplasms & melanomas         2.0         4.5         2.2         5.2         0.87         (0.47 - 1.59)           XIa.         Adrenocortical carcinomas         0.2         c         -         -         b         b           XIb.         Thyroid carcinomas         0.3         c         0.3         c         b         b           XIc.         Nasopharyngeal carcinomas         0.1         c         0.1         c         b         b           XId.         Melanomas         0.6         1.4         1.0         2.3         b         b           XIe.         Skin carcinomas         -         -         -         -         -         -         -         b         b           XIf.         Other & unspecified carcinomas         0.8         1.8         0.8         1.9         b         b           XII.         Other & unspecified malignant neoplasms         0.2         0.4         0.2         0.5         b           XIII.         Other specified malignant tumours         0.2         0.1	Xc.	Malignant gonadal germ cell tumours	1.4	3.0	0.9	2.1	b	b
No.   Other extinal planta fundais   2.0   4.5   2.2   5.2   0.87   (0.47 - 1.59)			-	-	-	-	ь	b
XIa.       Adrenocortical carcinomas       0.2       c       -       -       b       b         XIb.       Thyroid carcinomas       0.3       c       0.3       c       b       b         XIc.       Nasopharyngeal carcinomas       0.1       c       0.1       c       b       b         XId.       Melanomas       0.6       1.4       1.0       2.3       b       b         XIe.       Skin carcinomas       -       -       -       -       -       b       b         XIf.       Other & unspecified carcinomas       0.8       1.8       0.8       1.9       b       b         XII.       Other & unspecified malignant neoplasms       0.2       0.4       0.2       0.5       b       b         XIIa.       Other specified malignant tumours       0.2       c       0.1       c       b       b	Xe.	Other & unspecified malignant gonadal tumours	-	-	-	-	ь	b
XIb.       Thyroid carcinomas       0.3       c       0.3       c       b       b         XIc.       Nasopharyngeal carcinomas       0.1       c       0.1       c       b       b         XId.       Melanomas       0.6       1.4       1.0       2.3       b       b         XIe.       Skin carcinomas       -       -       -       -       -       b       b         XIf.       Other & unspecified carcinomas       0.8       1.8       0.8       1.9       b       b         XII.       Other & unspecified malignant neoplasms       0.2       0.4       0.2       0.5       b       b         XIIa.       Other specified malignant tumours       0.2       c       0.1       c       b       b					2.2	5.2	0.87	(0.47 - 1.59)
XIc.       Nasopharyngeal carcinomas       0.1       c       0.1       c       b       b         XId.       Melanomas       0.6       1.4       1.0       2.3       b       b         XIe.       Skin carcinomas       -       -       -       -       b       b         XIf.       Other & unspecified carcinomas       0.8       1.8       0.8       1.9       b       b         XII.       Other & unspecified malignant neoplasms       0.2       0.4       0.2       0.5       b       b         XIIa.       Other specified malignant tumours       0.2       c       0.1       c       b       b						-	ь	b
National N								
XII.       Other & unspecified malignant neoplasms       0.0       1.4       1.0       2.5         XIE.       Skin carcinomas       -       -       -       -       -       b       b         XII.       Other & unspecified malignant neoplasms       0.2       0.4       0.2       0.5       b       b         XIIa.       Other specified malignant tumours       0.2       c       0.1       c       b       b								
XIf. Other & unspecified carcinomas  O.8 1.8 0.8 1.9 b  XII. Other & unspecified malignant neoplasms  Visual Differ & unspecified malignant tumours  O.2 0.4 0.2 0.5 b  XIII. Other specified malignant tumours  O.3 c  O.4 c  O.5 c  D.6 c  D.7 c  D.8 c  D.9			0.6	1.4	1.0	2.3		
XII. Other & unspecified malignant neoplasms  O.5 1.6 0.6 1.7  II. Other & unspecified malignant neoplasms  O.2 0.4 0.2 0.5 b  XIIa. Other specified malignant tumours  O.2 c 0.1 c b				-		-		
XII. Other specified malignant tumours  One of the specified malignant tumours  One of				1.8				
Ana. Other specified manignant tumours 0.2 0.1								
XIIb. Other unspecified malignant tumours 0.1 c b b	XIIa.		0.2	с				
	XIIb.	Other unspecified malignant tumours	-	-	0.1	c	ь	b

AIID. Uther unspecified malignant tumours

a Rate per million population per year, age-standardised to the 2006 New Zealand Standard Population

b Relative risk was not calculated due to the small number of cases recorded for these diagnostic subgroups

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

#### 3.7 Childhood cancer incidence by age at diagnosis

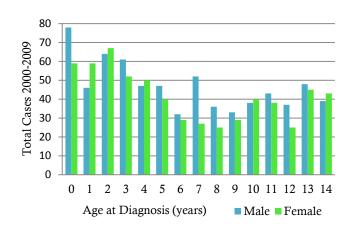
#### 3.7.1 Age distribution of childhood cancers

Figure 3.7.1a shows that 10.3% of all new childhood cancer cases registered between 2000 and 2009 were for infants aged less than one year, despite infants representing only 6.6% of the child population. 43.9% of all childhood cancers were diagnosed in children under the age of five. The greatest number of cancers by age and sex were diagnosed in males less than one year of age (78 cases) and females aged between 2 and 3 years at diagnosis (67 cases) (see Figure 3.7.1b).

Figure 3.7.1a Age distribution of childhood cancers in New Zealand, 2000-2009

Figure 3.7.1b Childhood cancers by age at diagnosis and sex, New Zealand, 2000-2009





#### 3.7.2 Childhood cancer incidence by age group

Incidence of leukaemia was significantly higher for children aged 0-4 years (79.5 per million) than children 5-9 years (44.7 per million) or 10-14 years (31.7 per million), (see Table 3.7.2). Neuroblastoma also showed a strong skew towards younger children, with an incidence rate of 24.7 per million at 0-4 years declining to a rate of only 1.6 per million for the 10-14 year age group. Retinoblastoma was diagnosed almost exclusively in children under the age of five (94.9%) and nearly 4 in 5 (77.0%) of renal tumours were also diagnosed in the first five years of life.

In contrast, incidence of lymphoma was significantly lower for children aged 0-4 (3.8 per million), increasing five-fold to 19.9 per million for the 10-14 year age group. The incidence of 'other epithelial neoplasms' (10.5 per million) and malignant bone tumours (15.4 per million) was also notably higher in the 10-14 year age group.

Table 3.7.2 Childhood cancer incidence by age at diagnosis, New Zealand, 2000-2009

	0-4 years				5-9	9 years	5	10-14 years				
			Average cases per year %		Rate per million population per year (95% CI)							
All childhood cancers	58.3	100	(18	202.4 86.0 - 218.8)	35.0	100	119.4 (106.9 - 132.0)		39.6	100	(1)	129.4 16.7 - 142.2)
I. Leukaemias	22.9	39.3	79.5	(69.2 - 89.8)	13.1	37.4	44.7	(37.1 - 52.4)	9.7	24.5	31.7	(25.4 - 38.0)
Lymphoid leukaemias	18.0	30.9	62.5	(53.4 - 71.6)	10.9	31.1	37.2	(30.2 - 44.2)	6.4	16.2	20.9	(15.8 - 26.0)
Acute myeloid leukaemias	3.6	6.2	12.5	(8.4 - 16.6)	1.6	4.6	5.5	(2.8 - 8.1)	2.7	6.8	8.8	(5.5 - 12.2)
II. Lymphomas	1.1	1.9	3.8	(1.6 - 6.1)	4.2	12.0	14.3	(10.0 - 18.7)	6.1	15.4	19.9	(14.9 - 24.9)
Hodgkin lymphomas	0.1	0.2	0.4	(0.0 - 1.0)	1.2	3.4	4.1	(1.8 - 6.4)	2.3	5.8	7.5	(4.4 - 10.6)
Non-Hodgkin lymphomas (excl. Burkitt lymphomas)	0.6	1.0	2.1	(0.4 - 3.8)	1.8	5.1	6.1	(3.3 - 9.0)	3.4	8.6	11.1	(7.4 - 14.9)
III. CNS tumours	9.9	17.0	34.4	(27.6 - 41.1)	9.7	27.7	33.1	(26.5 - 39.7)	8.8	22.2	28.8	(22.8 - 34.8)
Astrocytomas	3.9	6.7	13.5	(9.3 - 17.8)	4.2	12.0	14.3	(10.0 - 18.7)	3.6	9.1	11.8	(7.9 - 15.6)
IV. Neuroblastoma	7.1	12.2	24.7	(18.9 - 30.4)	1.3	3.7	4.4	(2.0 - 6.9)	0.5	1.3	1.6	(0.2 - 3.1)
V. Retinoblastoma	3.7	6.3	12.9	(8.7 - 17.0)	0.2	0.6	0.7	(0.0 - 1.6)	-	-	-	-
VI. Renal tumours	4.7	8.1	16.3	(11.7 - 21.0)	0.8	2.3	2.7	(0.8 - 4.6)	0.6	1.5	2.0	(0.4 - 3.5)
VII. Hepatic tumours	1.0	1.7	3.5	(1.3 - 5.6)	0.3	0.9	1.0	(0.0 - 2.2)	-	-	-	-
VIII. Malignant bone tumours	0.5	0.9	1.7	(0.2 - 3.3)	2.0	5.7	6.8	(3.8 - 9.8)	4.7	11.9	15.4	(11.0 - 19.8)
Osteosarcomas	0.1	0.2	0.4	(0.0 - 1.0)	1.1	3.1	3.8	(1.5 - 6.0)	2.5	6.3	8.2	(5.0 - 11.4)
Ewing tumour and related bone sarcomas	0.3	0.5	1.0	(0.0 - 2.2)	0.8	2.3	2.7	(0.8 - 4.6)	1.7	4.3	5.6	(2.9 - 8.2)
IX. Soft tissue sarcomas	3.8	6.5	13.2	(9.0 - 17.4)	2.2	6.3	7.5	(4.4 - 10.7)	3.4	8.6	11.1	(7.4 - 14.9)
Rhabdomyosarcoma	2.4	4.1	8.3	(5.0 - 11.7)	1.5	4.3	5.1	(2.5 - 7.7)	1.1	2.8	3.6	(1.5 - 5.7)
X. Germ cell tumours	2.8	4.8	9.7	(6.1 - 13.3)	0.6	1.7	2.1	(0.4 - 3.7)	2.6	6.6	8.5	(5.2 - 11.8)
XI. Other epithelial neoplasms	0.5	0.9	1.7	(0.2 - 3.3)	0.5	1.4	1.7	(0.2 - 3.2)	3.2	8.1	10.5	(6.8 - 14.1)
XII. Other and unspecified	0.3	0.5	1.0	(0.0 - 2.2)	0.1	0.3	0.3	(0.0 - 1.0)	-	-	-	-

<sup>&</sup>lt;sup>a</sup> Rate per million population per year, age-standardised to the 2006 New Zealand Standard Population

#### 3.7.3 Childhood cancer incidence by age group and diagnostic subgroup

Neuroblastoma incidence peaked in infancy, accounting for over a quarter (28%) of the cancers treated in 0-1 year olds, but only 7% of cases in children aged 1-4 years (See Figures 3.7.3a to 3.7.3d). Other diagnostic groups which made up a larger proportion of the cancers diagnosed in infancy compared to cancers diagnosed in 1-4 year olds were retinoblastoma (9% c.f. 5%) and germ cell tumours (14% c.f. 2%).

Leukaemias made up nearly half (45%) of the 444 cancers diagnosed in children aged 1-4 years within the time period but less than a quarter (23.5%) of the 396 cases diagnosed in 10-14 year olds. In contrast, lymphomas (15%), malignant bone tumours (12%), and other epithelial neoplasms (8%) accounted for a greater proportion of the cancers affecting 10-14 years than for younger age groups. CNS tumours made up a greater proportion of the total cancer cases recorded for 5-9 year olds (28%) than for any other age group. This was not due to a higher incidence of CNS tumours amongst 5-9 year olds, but rather reflected the lower incidence this age group had for many of the other diagnostic groups.

Figure 3.7.3a Cancers diagnosed in infants aged <1 year, New Zealand, 2000-2009

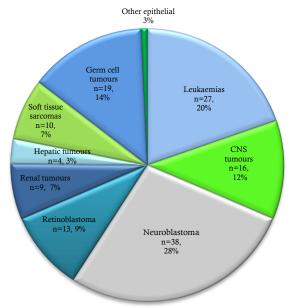


Figure 3.7.3c Cancers diagnosed in children aged 5-9 years, New Zealand, 2000-2009

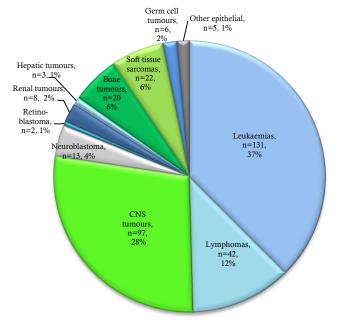


Figure 3.7.3b Cancers diagnosed in children aged 1-4 years, New Zealand, 2000-2009

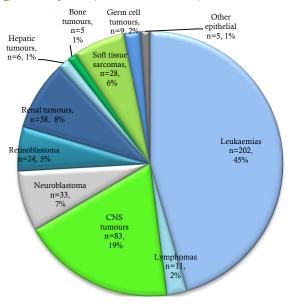
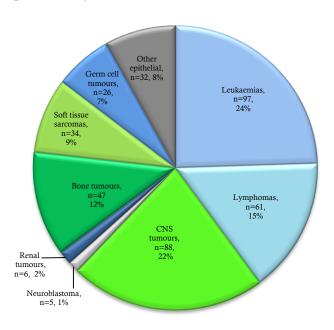


Figure 3.7.3d Cancers diagnosed in children aged 10-14 years, New Zealand, 2000-2009



#### 3.8 Childhood cancer incidence by ethnicity

Childhood cancer incidence was significantly lower for Maori (131.1 per million) than for non-Maori/non-Pacific Peoples (158.1 per million). This is in contrast to earlier published data; the International Incidence of Childhood Cancer reported little difference in overall incidence between Maori and non-Maori with incidence rates of 151.7 and 159.4 per million respectively for the period 1970-1992<sup>11</sup>. However, this finding is consistent with an analysis of 1991-2002 NZCR registrations which found lower cancer incidence among Maori young people 10-14 years<sup>12</sup>.

Compared to non-Maori/non-Pacific Peoples, Maori appeared to have lower incidence of ALL (31.0 per million c.f. 42.9 per million) and CNS tumours (26.0 per million c.f. 35.5 per million), specifically astrocytomas (7.0 per million c.f. 16.2 per million).

Pacific Peoples had the highest incidence reported for childhood leukaemia (71.5 per million, c.f. 46.5 per million for Maori and 52.4 per million for non-Maori/non-Pacific Peoples). The analysis of cancer incidence among the New Zealand AYA population for the same time period identified a significantly higher incidence of leukaemias among Pacific Peoples compared to non-Maori/non-Pacific Peoples.<sup>13</sup> These two findings suggest that these differences may arise from a real biological predisposition to leukaemia among young Pacific People which warrants further study.

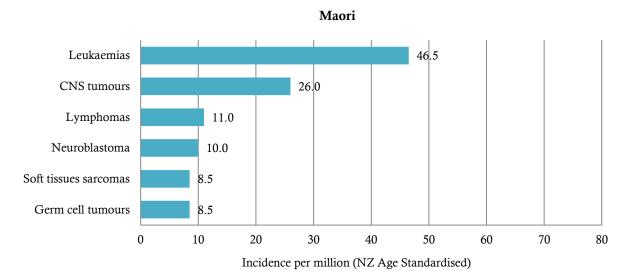
The incidence of germ cell tumours among Pacific Peoples (15.9 per million) was significantly higher than non-Maori/non-Pacific Peoples (5.2 per million). Bone tumours were the third most common cancer diagnostic group for Pacific peoples with an incidence of 18.5 per million per year, which was over double the rate for Maori (7.0 per million) and non-Maori/non-Pacific Peoples (7.6 per million). However, unlike for leukaemias, these ethnic differences were not evident in the AYA analysis.

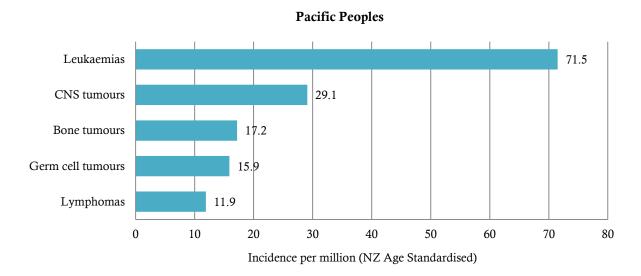
Non-Maori/non-Pacific Peoples had the highest incidence of lymphomas (14.0 per million), CNS tumours (35.5 per million), and soft tissue sarcomas (13.7 per million).

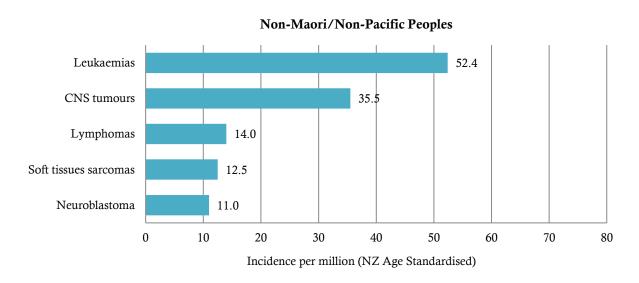
Table 3.8 Childhood cancer incidence by prioritised ethnicity, New Zealand, 2000-2009

Diagnostic Group / selected		Mad	ori		Pacific <b>F</b>	Peoples	Non-Maori/non-Pacific Peoples			
subgroups	No. of Age-standardised rate cases (95% CI)		No. of cases				No. of Age-standardised rate cases (95% CI)			
Total childhood cancers	262	131.1	(115.2 - 146.9)	131	173.4	(143.7 - 203.1)	936	158.1	(147.9 - 168.2)	
I. Leukaemias	93	46.5	(37.1 - 56.0)	54	71.5	(52.4 - 90.6)	310	52.4	(46.5 - 58.2)	
Acute lymphoblastic leukaemias	62	31.0	(23.3 - 38.7)	37	49.0	(33.2 - 64.8)	254	42.9	(37.6 - 48.2)	
Acute myeloid leukaemias	24	12.0	(7.2 - 16.8)	8	10.6	(3.3 - 17.9)	47	7.9	(5.7 - 10.2)	
II. Lymphomas	22	11.0	(6.4 - 15.6)	9	11.9	(4.1 - 19.7)	83	14.0	(11.0 - 17.0)	
Hodgkin lymphomas	8	4.0	(1.2 - 6.8)	1	1.3	(0.0 - 3.9)	27	4.6	(2.8 - 6.3)	
Non-Hodgkin lymphomas (excl. Burkitt's lymphoma)	10	5.0	(1.9 - 8.1)	4	5.3	(0.1 - 10.5)	44	7.4	(5.2 - 9.6)	
III. CNS Tumours	52	26.0	(18.9 - 33.1)	22	29.1	(17.0 - 41.3)	210	35.5	(30.7 - 40.3)	
Astrocytomas	14	7.0	(3.3 - 10.7)	7	9.3	(2.4 - 16.1)	96	16.2	(13.0 - 19.5)	
IV. Neuroblastoma	20	10.0	(5.6 - 14.4)	4	5.3	(0.1 - 10.5)	65	11.0	(8.3 - 13.7)	
V. Retinoblastoma	9	4.5	(1.6 - 7.4)	5	6.6	(0.8 - 12.4)	25	4.2	(2.6 - 5.9)	
VI. Renal tumours	8	4.0	(1.2 - 6.8)	4	5.3	(0.1 - 10.5)	49	8.3	(6.0 - 10.6)	
VII. Hepatic tumours	2	1.0	(0.0 - 2.4)	2	2.7	(0.0 - 6.3)	9	1.5	(0.5 - 2.5)	
VIII. Malignant bone tumours	14	7.0	(3.3 - 10.7)	13	17.2	(7.9 - 26.6)	45	7.6	(5.4 - 9.8)	
Osteosarcoma	8	4.0	(1.2 - 6.8)	7	9.3	(2.4 - 16.1)	22	3.7	(2.2 - 5.3)	
Ewing tumour	4	2.0	(0.0 - 4.0)	6	7.9	(1.6 - 14.3)	18	3.0	(1.6 - 4.4)	
IX. Soft tissue sarcomas	17	8.5	(4.5 - 12.6)	3	4.0	(0.0 - 8.5)	74	12.5	(9.7 - 15.3)	
Rhabdomyosarcoma	9	4.5	(1.6 - 7.4)	2	2.7	(0.0 - 6.3)	39	6.6	(4.5 - 8.7)	
X. Germ cell tumours	17	8.5	(4.5 - 12.6)	12	15.9	(6.9 - 24.9)	31	5.2	(3.4 - 7.1)	
XI. Other epithelial neoplasms	6	3.0	(0.6 - 5.4)	3	4.0	(0.0 - 8.5)	33	5.6	(3.7 - 7.5)	
XII. Other malignant neoplasms	2	1.0	(0.0 - 2.4)	-	-	-	2	0.3	(0.0 - 0.8)	

Figure 3.8 The most frequently diagnosed childhood cancers by prioritised ethnicity, New Zealand, 2000-2009







The First Outcome Analysis of the NZCCR

#### 3.9 International comparisons of childhood cancer incidence

The following section compares the New Zealand 2000-2009 childhood cancer incidence rates with the incidence rates from Great Britain<sup>14</sup>, Australia<sup>15</sup>, Germany<sup>16</sup> and the United States<sup>17</sup>. These developed countries were selected as they had published childhood cancer incidence rates by ICCC-3 for 0-14 year olds for a comparable time period.

Differences in reported incidence rates between New Zealand and other countries need to be interpreted with caution as they may be influenced by differences in timing, population composition, registry data quality and completeness, and statistical methodology<sup>18</sup>. In Table 3.9 below, the New Zealand, Australian and US incidence has been age-adjusted to their own standard population, while Germany and Great Britain has reported childhood cancer incidence which has been standardised to the World Standard Population.

The overall New Zealand incidence of childhood cancers from 2000 to 2009 (149 per million) was comparable to incidence rates reported in other developed nations, which ranged from 138 (Great Britain) to 173 per million (US, SEER). By ICCC-3 diagnostic groups, New Zealand had the highest incidence of malignant bone tumours and germ cell tumours but lowest incidence of lymphomas and renal tumours. New Zealand's incidence of CNS tumours (32 per million) was considerably lower than the incidence rate reported by SEER (44 per million) but similar the other three reported rates.

Table 3.9 International comparisons of relative frequency and incidence of childhood cancers

		Germany 2000-2009 <sup>a</sup>		at Britain 96-2005 <sup>a</sup>		v Zealand 00-2009 <sup>b</sup>		ustralia 97-2006°		(SEER) 07-2011 <sup>d</sup>
	%	Age standardised rate <sup>a</sup>	%	Age standardised rate <sup>a</sup>	%	Age standardised rate <sup>b</sup>	%	Age standardised rate <sup>c</sup>		Age standardised rate <sup>d</sup>
Overall childhood cancer	100	160	100	138	100	149	100	156	100	173
Leukaemias	34	56	32	48	34	51	34	52	31	54
Lymphomas	11	16	10	13	9	13	10	16	9	16
CNS tumours <sup>e</sup>	23	36	25	36	21	32	23	35	26	44
Neuroblastoma	7	12	6	10	7	10	6	9	6	11
Retinoblastoma	2	4	3	4	3	4	2	4	2	4
Renal tumours	6	12	6	9	5	7	5	8	5	8
Hepatic tumours	1	4	1	2	1	1	2	3	2	3
Malignant bone tumours	5	4	4	5	6	8	4	7	4	7
Soft tissue sarcomas	6	8	7	10	7	11	5	8	6	11
Germ cell tumours	3	4	3	5	5	7	4	6	3	6
Other epithelial neoplasms	2	4	3	4	3	5	5	7	4	7
Other malignant neoplasms	0	<1	1	1	0	<1	0	<1	0	1

<sup>&</sup>lt;sup>a</sup> Rates per million per year, age standardised to the World Standard Population

<sup>&</sup>lt;sup>b</sup> Rates per million per year, age standardised to the 2006 New Zealand Standard Population

<sup>&</sup>lt;sup>c</sup>Rates per million per year, age-standardised to the 2001 Australian Standard Population

d Rates per million per year, age-standardised to the 2000 US Standard Population; SEER 18 areas covers 28% of the total US population

<sup>&</sup>lt;sup>c</sup> Includes non-malignant CNS tumours

# 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 ten-year period. Incidence rates (with confidence intervals) could not be calculated for all diagnostic subgroups by sex, age group, and ethnicity; only raw numbers and percentages are reported. 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 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 46 new cases of leukaemia diagnosed each year, representing around one third (34.4%) of all new paediatric cancer cases for this time period. Half (50.1%) of the total childhood leukaemias were diagnosed in children under the age of five, with an incidence rate of 79.5 per million population per year. Incidence was significantly lower for the 5-9 year (44.7 per million) and 10-14 year (31.7 per million) age groups (see Table 4.1.1).

Table 4.1.1 Incidence of childhood leukaemias by sex, age group, and ethnicity, New Zealand, 2000-2009

	Total cases	%	Rates per million population per year (95% CI)		
Total Leukaemias	457	100	51.2	(46.5 - 55.8)	
Sex					
Male	240	52.5	52.4	(45.8 - 59.0)	
Female	217	47.5	49.9	(43.2 - 56.5)	
Age group					
0-4 years	229	50.1	79.5	(69.2 - 89.8)	
0-1 years	(27)	(5.9)	а	-	
1-4 years	(202)	(44.2)	а	-	
5-9 years	131	28.7	44.7	(37.1 - 52.4)	
10-14 years	97	21.2	31.7	(25.4 - 38.0)	
Ethnicity					
Maori	93	20.4	46.5	(37.1 - 56.0)	
Pacific Peoples	54	11.8	71.5	(52.4 - 90.6)	
Non-Maori/non-Pacific Peoples	310	67.8	52.4	(46.5 - 58.2)	

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

#### 4.1.2 Leukaemias incidence by diagnostic subgroup

ALL represented approximately three quarters (77.1%) of leukaemias diagnosed and over a quarter (26.6%) of the total cancers diagnosed in New Zealand between 2000 and 2009. AML accounted for 17.2% of the total leukaemias, with around eight new cases diagnosed each year (see Table 4.1.2). Myeloproliferative diseases and other leukaemias were rarely diagnosed in New Zealand children, with around two or three cases per year fitting into these remaining diagnostic subgroups.

Table 4.1.2 Incidence of childhood leukaemias by diagnostic subgroup, New Zealand, 2000-2009

	Total cases	% of leukaemias diagnosed	% of total child cancers diagnosed	popul	es per million ation per year (95% CI)
I. Leukaemias, myeloproliferative diseases & myelodysplastic diseases	458	100	34.4%	51.2	(46.5 - 55.8)
Ia. Lymphoid leukaemias	353	77.1	26.6%	39.5	(35.4 - 43.6)
Ib. Acute myeloid leukaemias	79	17.2	5.9%	8.9	(6.9 - 10.8)
Ic. Chronic myeloproliferative disease	4	0.9	0.3%	0.5	(0.0 - 0.9)
Id. Other myeloproliferative diseases	11	2.4	0.8%	1.2	(0.5 - 1.9)
Ie. Other & unspecified leukaemias	10	2.2	0.8%	1.1	(0.4 - 1.8)

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

Table 4.1.3 shows that more males were diagnosed with ALL within the ten year study period (193 cases, 42.0 per million) than females (160 cases, 36.8 per million). In contrast, more females (10.1 per million) were diagnosed with AML than males (7.7 per million) within this time period. In terms of ethnicity, Maori made up a greater proportion of children diagnosed with AML (30.4%) than ALL (17.6%). Childhood ALL had a younger median age at diagnosis (4 years) than AML (6.5 years). Nearly half (47.9%) of childhood ALL cases were diagnosed in those aged between one and four years old.

Table 4.1.3 Number of cases of childhood ALL and AML by sex, age group, and ethnicity, New Zealand, 2000-2009

	Lyn	Lymphoid Leukaemias (ALL)				Myeloid	l Leuka	emias (AML)
	Cases	%	_	standardised e (95% CI)				standardised te (95% CI)
Total	353	100	39.5	(35.4 - 43.6)	79	100	8.9	(6.9 - 10.8)
Sex								
Male	193	54.7	42.0	а	35	44.3	7.7	а
Female	160	45.3	36.8	а	44	55.7	10.1	а
Age group								
0-4 years	180	51.0	62.5	(53.4 - 71.6)	36	45.6	12.5	(8.4 - 16.6)
0-1 years	11	3.1	b	b	10	12.7	b	b
1-4 years	169	47.9	b	ь	26	32.9	b	b
5-9 years	109	30.9	37.2	(30.2 - 44.2)	16	20.3	5.5	(2.8 - 8.1)
10-14 years	64	18.1	20.9	(15.8 - 26.0)	27	34.2	8.8	(5.5 - 12.2)
Ethnicity								
Maori	62	17.6	31.0	(23.3 - 38.7)	24	30.4	12.0	(7.2 - 16.8)
Pacific Peoples	37	10.5	49.0	(33.2 - 64.8)	8	10.1	10.6	(3.3 - 17.9)
Non-Maori/non-Pacific Peoples	254	72.0	42.9	(37.6 - 48.2)	47	59.5	7.9	(5.7 - 10.2)

<sup>&</sup>lt;sup>a</sup> 95% Confidence Intervals were not calculated for diagnostic subgroup by sex

<sup>&</sup>lt;sup>b</sup> Age standardised incidence was not calculated specifically for these age groups

#### 4.2 Lymphomas and reticuloendothelial neoplasms

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

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 NHL and Burkitt lymphoma. T-cell NHL is treated in a similar manner to ALL, while Burkitt lymphoma is treated with a very intense short course of multi-agent chemotherapy. Most children in New Zealand who are diagnosed with lymphoma will be treated according to a clinical trial (COG or other cooperative clinical trials group) or according to a disease-specific clinical protocol.

#### 4.2.1 Lymphomas overall incidence

Table 4.2.1 shows that childhood lymphoma incidence was considerably higher for males (16.0 per million) than females (9.8 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. However, by 10-14 years incidence increased significantly to 19.9 per million, accounting for 15.4% of the total cancers for this age group.

Table 4.2.1 Incidence of childhood lymphomas, by sex, age group, and ethnicity, New Zealand, 2000-2009

	Total cases	%	Rates per million population per year (95% CI)			
Total lymphomas	114	100	13.0	(10.6 - 15.4)		
Sex						
Males	72	63.2	16.0	(12.3 - 19.7)		
Female	42	36.8	9.8	(6.8 - 12.8)		
Age group						
0-4 years	11	9.6	3.8	(1.6 - 6.1)		
0-1 years	(0)	(0.0)	а	-		
1-4 years	(11)	(9.6)	а	-		
5-9 years	42	36.8	14.3	(10.0 - 18.7)		
10-14 years	61	53.5	19.9	(14.9 - 24.9)		
Ethnicity						
Maori	22	19.3	11.0	(6.4 - 15.6)		
Pacific Peoples	9	7.9	11.9	(4.1 - 19.7)		
Non-Maori/non-Pacific Peoples	83	70.3	14.0	(11.0 - 17.0)		

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

#### 4.2.2 Lymphomas incidence by diagnostic subgroup

Half (50.9%) of the 114 lymphomas diagnosed in New Zealand from 2000-2009 were 'non-Hodgkin lymphomas (except Burkitt lymphoma)'. Of the remaining cases, 36 (31.6%) were Hodgkin lymphomas, and 18 (15.8%) were Burkitt lymphoma (see Table 4.2.2).

Table 4.2.2 Incidence of childhood lymphomas by diagnostic subgroup, New Zealand, 2000-2009

	Total cases	% of lymphomas diagnosed	popul	s per million ation per year 95% CI)
II. Lymphomas & reticuloendothelial neoplasms	114	100	13.0	(10.6 - 15.4)
IIa. Hodgkin lymphomas	36	31.6	4.1	(2.8 - 5.5)
IIb. Non-Hodgkin lymphomas (except Burkitt lymphoma)	58	50.9	6.6	(4.9 - 8.3)
IIc. Burkitt lymphoma	18	15.8	2.0	(1.1 - 3.0)
IId. Miscellaneous lymphoreticular neoplasms	1	0.9	0.1	(0.0 - 0.3)
IIe. Unspecified lymphomas	1	0.9	0.1	(0.0 - 0.3)

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

The ethnic and age distributions for the diagnostic subgroups 'IIa: Hodgkin lymphomas' and 'IIb: Non-Hodgkin lymphomas (except Burkitt lymphoma)' were similar (see Table 4.2.3). However there was a distinct difference between the two diagnostic subgroups in that there were no sex differences in the incidence of Hodgkin lymphomas (17 cases were diagnosed in males, 19 in females) but males were significantly more likely than females of developing being diagnosed with non-Hodgkin lymphoma (relative risk: 2.5).

Table 4.2.3 Number of cases of childhood Hodgkin and non-Hodgkin lymphomas by sex, age group, and ethnicity, New Zealand, 2000-2009

		Hodgkir	ı lymph	omas	1	Non-Hod	gkin lyr	nphomas
	Cases	%		standardised e (95% CI)	Cases	%		standardised te (95% CI)
Total	36	100	4.1	(2.8 - 5.5)	58	100	6.6	(4.9 - 8.3)
Sex								
Male	17	47.2	3.8	а	42	72.4	4.4	а
Female	19	52.8	9.4	а	16	27.6	3.7	а
Age group								
0-4 years	1	2.8	0.4	(0.0 - 1.0)	6	10.3	2.1	(0.4 - 3.8)
0-1 years	-	-	b	-	(1)	(1.7)	b	-
1-4 years	(1)	(2.8)	b	-	(5)	(8.6)	b	-
5-9 years	12	33.3	4.1	(1.8 - 6.4)	18	31.0	6.1	(3.3 - 9.0)
10-14 years	23	63.9	7.5	(4.4 - 10.6)	34	58.6	11.1	(7.4 - 14.9)
Ethnicity								
Maori	8	22.2	4.0	(1.2 - 6.8)	10	17.2	5.0	(1.9 - 8.1)
Pacific Peoples	1	2.8	1.3	(0.0 - 3.9)	4	6.9	5.3	(0.1 - 10.5)
Non-Maori/non-Pacific Peoples	27	75.0	4.6	(2.8 - 6.3)	44	75.9	7.4	(5.2 - 9.6)

<sup>&</sup>lt;sup>a</sup> 95% Confidence Intervals were not calculated for diagnostic subgroup by sex

# 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 32.0 per million, 'CNS and miscellaneous intracranial and intraspinal neoplasms' was the second most common class of tumour diagnosed in New Zealand children between 2000 and 2009. CNS tumours accounted for 21.4% of all childhood cancers diagnosed, with around new 28 cases diagnosed annually. Of the 284 central nervous system tumours registered in the ten year period, 108 (38.0%) were tumours that were either benign or of uncertain behaviour and over half of these (63 cases, 58.3%) were juvenile pilocytic astrocytomas.

Table 4.3.1 shows that there were no significant differences in overall CNS tumour incidence according to sex or age group. Incidence rates were significantly higher for those of European ethnicity (38.6 per million) than those of Maori ethnicity (26.0 per million).

Table 4.3.1 Incidence of childhood CNS tumours by sex, age group, and ethnicity, New Zealand, 2000–2009

	Total cases	%	Rates per million populatio per year (95% CI)		
Total CNS tumours	284	100	32.0	(28.3 - 35.7)	
Sex					
Male	148	52.1	32.5	(27.3 - 37.7)	
Female	136	47.9	31.4	(26.1 - 36.7)	
Age group					
0-4 years	99	34.9	34.4	(27.6 - 41.1)	
0-1 years	(16)	(5.6)	а	-	
1-4 years	(83)	(29.2)	a	-	
5-9 years	97	34.2	33.1	(26.5 - 39.7)	
10-14 years	88	31.0	28.8	(22.8 - 34.8)	
Ethnicity					
Maori	52	18.3	26.0	(18.9 - 33.1)	
Pacific Peoples	22	7.8	29.1	(17.0 - 41.3)	
Non-Maori/non-Pacific Peoples	210	73.9	35.5	(30.7 - 40.3)	

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

#### 4.3.2 CNS tumours incidence by diagnostic subgroup

With an incidence of 13.2 per million per year, astrocytomas accounted for two fifths of all CNS tumours (41.1%) and 8.8% of all childhood cancers diagnosed (see Table 4.3.2). There were 59 'intracranial and intraspinal embryonal tumours' diagnosed within the study period and 45 'other gliomas'.

Table 4.3.2 Incidence of childhood CNS tumours by diagnostic subgroup, New Zealand, 2000-2009

	Total cases	% of CNS tumours diagnosed	Rates per million population per year (95% CI)		
III. Central nervous system & miscellaneous intracranial and intraspinal neoplasms	284	100	32.0	(28.3 - 35.7)	
IIIa. Ependymomas and choroid plexus tumours	24	8.5	2.7	(1.6 - 3.8)	
IIIb. Astrocytomas	117	41.1	13.2	(10.8 - 15.6)	
IIIc. Intracranial and intraspinal embryonal tumours	59	20.8	6.6	(4.9 - 8.3)	
IIId. Other gliomas	45	15.8	5.1	(3.6 - 6.6)	
IIIe. Other specified intracranial and intraspinal neoplasms	36	12.7	4.1	(2.7 - 5.4)	
IIIf. Unspecified intracranial and intraspinal neoplasms	3	1.1	0.3	(0.0 - 0.7)	

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

Of the 59 intracranial and intraspinal embryonal tumours diagnosed in the 2000-2009 period, 52 (88.1%) were a medulloblastoma. Although Maori had a lower incidence rate for CNS tumours overall, those of Maori ethnicity accounted for over a third (33.9%) of all intracranial and intraspinal embryonal tumours diagnosed (see Table 3.3.3). With a relative risk of 2.8, the annual incidence of intracranial and intraspinal embryonal tumours was significantly higher for males (9.6 per million) than females (3.5 per million). Conversely, the incidence of astrocytomas among females (16.2 per million) was significantly higher than for males (10.3 per million). Astrocytomas had a median age at diagnosis of seven years. Ependymomas and choroid plexus tumours had a median age at diagnosis of five years, the lowest of all CNS tumour diagnostic subgroups.

Table 4.3.3 Number of cases of childhood CNS tumours by diagnostic subgroup, by sex, age group, and ethnicity, New Zealand, 2000-2009

	Ependym choroid tum	plexus	Astrocytomas		Intracranial & intraspinal embryonal tumours		Other gliomas	
	Cases	%	Cases	%	Cases	%	Cases	%
Total	24	100	117	100	59	100	45	100
Sex								
Male	15	62.5	47	40.2	44	74.6	21	46.7
Female	9	37.5	70	59.8	15	25.4	24	53.3
Age group								
0-4 years	11	45.8	39	33.3	20	33.9	15	33.3
0-1 years	(2)	(8.3)	(5)	(4.3)	(4)	(6.8)	(2)	(4.4)
1-4 years	(9)	(37.5)	(34)	(29.1)	(16)	(27.1)	(13)	(28.9)
5-9 years	7	29.2	42	35.9	24	40.7	17	37.8
10-14 years	6	25.0	36	30.8	15	25.4	13	28.9
Ethnicity								
Maori	4	16.7	14	12.0	20	33.9	8	17.8
Pacific Peoples	1	4.2	7	6.0	8	13.6	4	8.9
Non-Maori/non-Pacific Peoples	19	79.2	96	82.1	31	52.5	33	73.3

### 4.4 Neuroblastoma and other peripheral nervous cell tumours

Neuroblastoma is a heterogeneous group of cancers that arise from primitive neural crest cells within the sympathetic nervous system. Malignant neuroblastoma most commonly originates in the adrenal glands, or from adjacent abdominal sympathetic nerves, but tumours may arise anywhere along the sympathetic chain from the neck, chest abdomen and the pelvis. These tumours often present as 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 9.9 per million per year but this rate was significantly higher for the 0-4 year age group (24.7 per million – see Table 4.4). 38 cases (42.7%) of neuroblastoma were diagnosed in infants less than one year of age; neuroblastoma represented 27.5% 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, 2000-2009

	Total cases	%	Rates per million populatio per year (95% CI)	
Total neuroblastoma	89	100	9.9	(7.8 - 11.9)
Neuroblastoma & ganglioneuroblastoma	87	97.8	9.6	(7.6 - 11.7)
Other peripheral nervous cell tumours	2	2.2	0.2	(0.0 - 0.6)
Sex				
Male	46	51.7	10.0	(7.1 - 12.8)
Female	43	48.3	9.8	(6.9 - 12.7)
Age group				
0-4 years	71	79.8	24.7	(18.9 - 30.4)
0-1 years	(38)	(42.7)	а	а
1-4 years	(33)	(37.1)	а	а
5-9 years	13	14.6	4.4	(2.0 - 6.9)
10-14 years	5	5.6	1.6	(0.2 - 3.1)
Ethnicity				
Maori	20	22.5	10.0	(5.6 - 14.4)
Pacific Peoples	4	4.5	5.3	(0.1 - 10.5)
Non-Maori/non-Pacific Peoples	65	73.0	11.0	(8.3 - 13.7)

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

#### 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 39 cases of retinoblastoma diagnosed in the 2000-2009 period. One third (13 cases) were diagnosed in children less than one year old, accounting for 9.5% of cancers diagnosed in infancy. The median age at diagnoses was 1 year 9 months. 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, 2000-2009

	Total cases	%	Rates per million population per year (95% CI)	
Total retinoblastoma	39	100	4.3	(3.0 - 5.7)
Sex				
Male	19	48.7	4.1	(2.2 - 5.9)
Female	20	51.3	4.5	(2.6 - 6.5)
Age group				
0-4 years	37	94.9	12.9	(8.7 - 17.0)
0-1 years	(13)	(33.3)	а	-
1-4 years	(24)	(61.5)	а	-
5-9 years	2	5.1	0.7	(0.0 - 1.6)
10-14 years	-	-	-	-
Ethnicity				
Maori	9	23.1	4.5	(1.6 - 7.4)
Pacific Peoples	5	12.8	6.6	(0.8 - 12.4)
Non-Maori/non-Pacific Peoples	25	64.1	4.2	(2.6 - 5.9)

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

#### 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 61 renal tumours diagnosed between 2000 and 2009, 59 were from the diagnostic subgroup 'VIa: nephroblastoma and other non-epithelial renal tumours.' Only two cases of renal carcinoma were reported. Table 4.6 shows that the incidence of renal tumours at 0-4 years of age (16.3 per million) declined rapidly in later childhood. Renal tumours accounted for 6.6% of the total cancers diagnosed in infants and 8.5% 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, 2000-2009

	Total cases	%	populat	per million ion per year 5% CI)
Total renal tumours	61	100	6.8	(5.1 - 8.5)
Nephroblastoma & other non-epithelial renal tumours	59	13.1	6.5	(4.9 - 8.2)
Renal carcinomas	2	9.8	0.2	(0.0 - 0.6)
Sex				
Male	27	44.3	5.8	(3.6 - 8.0)
Female	34	55.7	7.8	(5.2 - 10.4)
Age group				
0-4 years	47	77.0	16.3	(11.7 - 21.0)
0-1 years	(9)	(14.8)	а	-
1-4 years	(38)	(62.3)	а	-
5-9 years	8	13.1	2.7	(0.8 - 4.6)
10-14 years	6	9.8	2.0	(0.4 - 3.5)
Ethnicity				
Maori	8	13.1	4.0	(1.2 - 6.8)
Pacific Peoples	4	6.6	5.3	(0.1 - 10.5)
Non-Maori/non-Pacific Peoples	49	80.3	8.3	(6.0 - 10.6)

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

### 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 with surgery alone. The prognosis for hepatoblastoma depends on the histological subtype, the level of tumour marker (AFP), the extent of tumour in the liver, and the presence or absence of metastatic spread. Hepatoblastoma is usually treated with pre-operative chemotherapy, surgical resection of the tumour or liver transplantation, and post-operative chemotherapy. Children in New Zealand with hepatoblastoma are currently treated according to the SIOPEL international cooperative clinical trial. For those with localised and resectable disease the overall prognosis is excellent.

Hepatic tumours had the lowest incidence of all childhood cancer diagnostic groups (1.4 per million per year, see Table 4.7). Of the 13 hepatic tumours diagnosed between 2000 and 2009, eight were hepatoblastoma (0.9 per million) and five were hepatic carcinomas (0.6 per million). All eight cases of hepatoblastoma were diagnosed in children aged 4 years and under.

Table 4.7 Incidence of childhood hepatic tumours by sex, age group, and ethnicity, New Zealand, 2000-2009

	Total cases	%	Rates per million population per year (95% CI)	
Total hepatic tumours	13	100	1.4	(0.7 - 2.2)
Hepatoblastoma	8	61.5	0.9	(0.3 - 1.5)
Hepatic carcinomas	5	38.5	0.6	(0.1 - 1.0)
Sex				
Male	8	61.5	1.7	(0.5 - 2.9)
Female	5	38.5	1.1	(0.1 - 2.1)
Age group				
0-4 years	10	76.9	3.5	(1.3 - 5.6)
0-1 years	(4)	(30.8)	а	-
1-4 years	(6)	(46.2)	a	-
5-9 years	3	23.1	1.0	(0.0 - 2.2)
10-14 years	-	-	-	-
Ethnicity				
Maori	2	15.4	1.0	(0.0 - 2.4)
Pacific Peoples	2	15.4	2.7	(0.0 - 6.3)
Non-Maori/non-Pacific Peoples	9	69.2	1.5	(0.5 - 2.5)

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

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#### 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 know risk factors. Most primary bone tumours present as a painful progressive swelling of a bone in a teenager; these grow slowly and are often diagnosed as a soft tissue injury before the correct diagnosis is established. The peak age of onset is 14 years, coinciding with the pubertal growth spurt. The prognosis and treatment depends on the tumour histology, its location and extent of disease. All primary bone tumours require expert treatment with chemotherapy, surgery, and less often radiotherapy.

#### 4.8.1 Malignant bone tumours overall incidence

Table 4.8.1 shows that the malignant bone tumour incidence rate for those aged 10-14 years (15.4 per million) was significantly higher than for those aged 0-4 years (1.7 per million) or 5-9 years (6.8 per million). Malignant bone tumours made up 11.9% of all cancers reported for the 10-14 year age group. Malignant bone tumour incidence for Pacific children (17.7 per million) was over double that for Maori (7.0 per million) and non-Maori/non-Pacific Peoples (7.6 per million). However, this was based on a small number of cases and did not reach statistical significance.

Table 4.8.1 Incidence of childhood malignant bone tumours by sex, age group, and ethnicity, New Zealand, 2000-2009

	Total cases	%	Rates per million population per year (95% CI)	
Total malignant bone tumours	72	100	8.2	(6.3 - 10.1)
Sex				
Male	37	51.4	8.3	(5.6 - 10.9)
Female	35	48.6	8.2	(5.5 - 10.9)
Age group				
0-4 years	5	6.9	1.7	(0.2 - 3.3)
0-1 years	-	-	-	-
1-4 years	5	(6.9)	а	-
5-9 years	20	27.8	6.8	(3.8 - 9.8)
10-14 years	47	65.3	15.4	(11.0 - 19.8)
Ethnicity				
Maori	14	19.4	7.0	(3.3 - 10.7)
Pacific Peoples	13	18.1	17.7	(7.9 - 26.6)
Non-Maori/non-Pacific Peoples	45	62.5	7.6	(5.4 - 9.8)

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for this age group

### 4.8.2 Malignant bone tumours incidence by diagnostic subgroup

Osteosarcomas represented half (51.4%) of all malignant bone tumours diagnosed between 2000 and 2009 with an incidence of 4.2 per million (see Table 3.8.2). Most other malignant bone tumours were from diagnostic group VIIIc 'Ewing tumours and related bone sarcomas' (38.9%, 3.2 per million per year).

Table 4.8.2 Incidence of childhood malignant bone tumours in New Zealand, 2000-2009, by diagnostic subgroup

	Total Cases	% of malignant bone tumours diagnosed		ion population (95% CI)
VIII. Malignant bone tumours	72	100	8.2	(6.3 - 10.1)
VIIIa. Osteosarcomas	37	51.4	4.2	(2.8 - 5.6)
VIIIb. Chondrosarcomas	1	1.4	0.1	(0.0 - 0.3)
VIIIc. Ewing tumours & related bone sarcomas	28	38.9	3.2	(2.0 - 4.4)
VIIId. Other specified malignant bone tumours	5	6.9	0.6	(0.1 - 1.1)
VIIIe. Unspecified malignant bone tumours	1	1.4	0.1	(0.0 - 0.3)

# 4.8.3 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 4.8.3 Number of cases of osteosarcomas and 'Ewing tumours and related bone sarcomas' by sex, age group, and ethnicity, New Zealand, 2000-2009

	Osteosa	rcomas	Ewing tumour & related bone sarcomas		
	Cases %		Cases	%	
Total	37	100	28	100	
Sex					
Male	20	54.1	14	50.0	
Female	17	45.9	14	50.0	
Age group					
0-4 years	1	2.7	3	10.7	
0-1 years	-	-	-	-	
1-4 years	(1)	(2.7)	(3)	(10.7)	
5-9 years	11	29.7	8	28.6	
10-14 years	25	67.6	17	60.7	
Ethnicity					
Maori	8	21.6	4	14.3	
Pacific Peoples	7	18.9	6	21.4	
Non-Maori/non-Pacific Peoples	22	59.5	18	64.3	

#### 4.9 Soft tissue and other extraosseous sarcomas

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

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

#### 4.9.1 Soft tissue sarcomas overall incidence

With an incidence rate of 10.5 cases per million population, 7.1% of all childhood cancers diagnosed between 2000 and 2009 were from the diagnostic group 'soft tissue and other extraosseous sarcomas'. Incidence per million was significantly lower for Pacific Peoples compared to non-Maori/non-Pacific Peoples (see Table 4.9.1)

Table 4.9.1 Incidence of childhood soft tissue sarcomas by sex, age group, and ethnicity, New Zealand, 2000-2009

	Total cases	%	Rates per million population per year (95% CI)	
Total soft tissue sarcomas	94	100	10.6	(8.4 - 12.7)
Sex				
Male	53	56.4	11.6	(8.5 - 14.8)
Female	41	43.6	9.5	(6.6 - 12.4)
Age group				
0-4 years	38	40.4	13.2	(9.0 - 17.4)
0-1 years	(10)	(10.2)	а	-
1-4 years	(28)	(29.8)	а	-
5-9 years	22	23.4	7.5	(4.4 - 10.7)
10-14 years	34	36.2	11.1	(7.4 - 14.9)
Ethnicity				
Maori	17	18.1	8.5	(4.5 - 12.6)
Pacific Peoples	3	3.2	4.0	(0.0 - 8.5)
Non-Maori/non-Pacific Peoples	74	78.7	12.5	(9.7 - 15.3)

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

### 4.9.2 Soft tissue sarcomas incidence by diagnostic subgroup

Table 4.9.2 shows that rhabdomyosarcomas represented over half (53.2%) of all 'soft tissue and other extraosseous sarcomas' diagnosed in New Zealand from 2000 to 2009.

Table 4.9.2 Incidence of soft tissue and other extraosseous sarcomas in New Zealand, 2000-2009, by diagnostic subgroup

	Total cases	% of soft tissue sarcomas diagnosed	popula	per million tion per year 95% CI)
IX. Soft tissue and other extraosseous sarcomas	94	100	10.6	(8.4 - 12.7)
IXa. Rhabdomyosarcomas	50	53.2	5.6	(4.0 - 7.2)
IXb. Fibrosarcomas & other fibrous neoplasms	7	7.4	0.8	(0.2 - 1.4)
IXc. Kaposi sarcomas	-	-	-	-
IXd. Other specified soft tissue sarcomas	27	28.7	3.1	(1.9 - 4.2)
IXe. Unspecified soft tissue sarcomas	10	10.6	1.4	(0.4 - 1.8)

#### 4.9.3 Rhabdomyosarcoma incidence by sex, age group, and ethnicity

Rhabdomyosarcomas were significantly more likely to be diagnosed in males (7.6 per million) than females (3.4 per million, relative risk: 2.2). The median age at diagnoses was 5 years, 3 months.

Table 4.9.3 Incidence of childhood rhabdomyosarcomas by sex, age group, and ethnicity, New Zealand, 2000-2009,

	Rhabdomyosarcomas			
	Cases	%		standardised e (95% CI)
Total	50	100	5.6	(4.0 - 7.2)
Sex				
Male	35	70.0	7.6	а
Female	15	30.0	3.4	а
Age group				
0-4 years	24	48.0	8.3	(5.0 - 11.7)
0-1 years	(3)	6.0	ь	-
1-4 years	(21)	42.0	b	-
5-9 years	15	30.0	5.1	(2.5 - 7.7)
10-14 years	11	22.0	3.6	(1.5 - 5.7)
Ethnicity				
Maori	9	18.0	4.5	(1.6 - 7.4)
Pacific Peoples	2	4.0	2.7	(0.0 - 6.3)
Non-Maori/non-Pacific Peoples	39	78.0	6.6	(4.5 - 8.7)

<sup>&</sup>lt;sup>a</sup> 95% Confidence Intervals were not calculated for diagnostic subgroup by sex

<sup>&</sup>lt;sup>b</sup> Age standardised incidence was not calculated specifically for these age groups

#### 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.

#### 4.10.1 Germ cell tumours overall incidence

Germ cell tumours, trophoblastic tumours, and neoplasms of gonads accounted for 4.5% of the total cancers diagnosed in children between 2000 and 2009, however there was considerable variability between the age groups; this diagnostic group represented 13.9% of all cancers diagnosed in infancy, but accounted for less than 2% for the 1-4 year and 5-9 year age groups. Table 4.10.1 shows a wide range of incidence rates by ethnicity; Pacific Peoples had an incidence rate three times that of non-Maori/non-Pacific Peoples (15.9 per million c.f. 5.2 per million).

Table 4.10.1 Incidence of childhood germ cell tumours by sex, age group, and ethnicity, New Zealand, 2000-2009

	Total cases	%	popula	per million tion per year 95% CI)
Total	60	100	6.8	(5.1 - 8.5)
Sex				
Male	29	48.3	6.3	(4.0 - 8.7)
Female	31	51.7	7.2	(4.7 - 9.7)
Age group				
0-4 years	28	46.7	9.7	(6.1 - 13.3)
0-1 years	(19)	(31.7)	а	-
1-4 years	(9)	(15.0)	а	-
5-9 years	6	10.0	2.1	(0.4 - 3.7)
10-14 years	26	43.3	8.5	(5.2 - 11.8)
Ethnicity				
Maori	17	28.3	8.5	(4.5 - 12.6)
Pacific Peoples	12	20.0	15.9	(6.9 - 24.9)
Non-Maori/non-Pacific Peoples	31	51.7	5.2	(3.4 - 7.1)

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

#### 4.10.2 Germ cell tumours incidence by diagnostic subgroup

The 60 germ cell tumour cases diagnosed in the 2000-2009 period were relatively evenly distributed across three diagnostic subgroups (see Table 4.10.2).

Table 4.10.2 Incidence of childhood germ cell tumours in New Zealand, 2000-2009, by diagnostic subgroup

	Total cases % of germ cell tumours diagnosed Rates per million population per year (95% CI)			
X. Germ cell tumours, trophoblastic tumours & neoplasms of gonads	60	100	6.8	(5.1 - 8.5)
Xa. Intracranial & intraspinal germ cell tumours	18	30.0	2.1	(1.1 - 3.0)
Xb. Malignant extracranial & extragonadal germ cell tumours	19	31.7	2.1	(1.2 - 3.1)
Xc. Malignant gonadal germ cell tumours	23	38.3	2.6	(1.5 - 3.7)
Xd. Gonadal carcinomas	-	-	-	-
Xe. Other & unspecified malignant gonadal tumours	-	-	-	-

# 4.10.3 Incidence for selected germ cell tumour diagnostic subgroups by sex, age group, and ethnicity

There were considerable differences in the age distributions for each of the three diagnostic subgroups. 12 of the 19 'malignant extracranial and extragonadal germ cell tumours' cases (63.2%) were diagnosed in children less than one year of age, while 14 of the 18 'intracranial and intraspinal germ cell tumours' (77.8%) were diagnosed in children aged 10-14 (see Table 4.10.3).

Table 4.10.3 Number of cases of childhood germ cell tumours by diagnostic subgroup, sex, age group, and ethnicity, New Zealand, 2000-2009

		l & intraspinal ell tumours	Malignant extracranial & extragonadal germ cell tumours		Malignant gonadal germ cell tumours	
	Cases	%	Cases	%	Cases	%
Total	18	100	19	100	23	100
Sex						
Male	8	44.4	7	36.8	14	60.9
Female	10	55.6	12	63.2	9	39.1
Age group						
1-4 years	1	5.6	16	84.2	11	47.8
0-1 years	-	-	(12)	(63.2)	(7)	(30.4)
1-4 years	(1)	(5.6)	(4)	(21.1)	(4)	(17.4)
5-9 years	3	16.7	1	5.3	2	8.7
10-14 years	14	77.8	2	10.5	10	43.5
Ethnicity						
Maori	3	16.7	6	31.6	8	34.8
Pacific Peoples	7	38.9	3	15.8	2	8.7
Non-Maori/non-Pacific Peoples	8	44.4	10	52.6	13	56.5

### 4.11 Other malignant epithelial neoplasms and malignant melanomas

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

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

# 4.11.1 'Other malignant epithelial neoplasms and malignant melanomas' overall incidence

An average of four new cases of 'other malignant epithelial neoplasms and malignant melanomas' were diagnosed in New Zealand each year between 2000 and 2009 (4.8 per million, see Table 4.11.1). Incidence increased considerably as children entered early adolescence, increasing from 1.7 per million for the 0-4 year age group more than six-fold to 10.5 per million among those aged 10-14 years.

Table 4.11.1 Incidence of childhood epithelial neoplasms by sex, age group, and ethnicity, New Zealand, 2000-2009

	Total cases	%	Rates per million population per year (95% CI)	
Total epithelial neoplasms	42	100	4.8	(3.4 - 6.3)
Sex				
Male	20	46.7	4.5	(2.5 - 6.4)
Female	22	53.3	5.2	(3.0 - 7.3)
Age group				
0-4 years	5	11.9	1.7	(0.2 - 3.3)
0-1 years	-	-	a	-
1-4 years	(5)	(11.9)	a	-
5-9 years	5	11.9	1.7	(0.2 - 3.2)
10-14 years	32	76.2	10.5	(6.8 - 14.1)
Ethnicity				
Maori	6	14.3	3.0	(0.6 - 5.4)
Pacific Peoples	3	7.1	4.0	(0.0 - 8.5)
Non-Maori/non-Pacific Peoples	33	78.6	5.6	(3.7 - 7.5)

<sup>&</sup>lt;sup>a</sup> Age standardised incidence was not calculated specifically for these age groups

# 4.11.2 'Other epithelial neoplasms and malignant melanomas' incidence by diagnostic subgroup

Melanomas represented 16 of the 42 cases (38.1%) within the diagnostic group 'other malignant epithelial neoplasms and malignant melanomas' diagnosed in New Zealand children between 2000 and 2009 (see Table 4.11.2). 15 out of 16 (93.8%) of these melanomas were diagnosed in children 10 years or over and all 16 cases were from the non-Maori/non-Pacific Peoples prioritised ethnic group. Thyroid, nasopharyngeal, and adrenocortical carcinomas had a combined total of 10 cases, of which seven were diagnosed in children over the age of 10.

Table 4.11.2 Incidence of childhood epithelial neoplasms by diagnostic subgroup, New Zealand, 2000-2009

	Total cases	% of germ cell tumours diagnosed	populatio	er million on per year % CI)
XI. Other epithelial neoplasms & melanomas	42	100	4.8	(3.4 - 6.3)
XIa. Adrenocortical carcinomas	2	4.8	a	a
XIb. Thyroid carcinomas	6	14.3	a	a
XIc. Nasopharyngeal carcinomas	2	4.8	a	a
XId. Melanomas	16	38.1	1.8	(0.9 - 2.7)
XIe. Skin carcinomas	-	-	-	
XIf. Other & unspecified carcinomas	16	38.1	1.8	(0.9 - 2.7)

<sup>&</sup>lt;sup>a</sup> Age standardised rates (and corresponding 95% CI) have been censored for diagnostic subgroups where there were fewer than ten cases diagnosed within the ten-year period

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## **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)

WHO World Health Organisation

### AII International Classification of Childhood Cancers, 3<sup>rd</sup> edition (ICCC-3)<sup>5</sup>

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, 3<sup>rd</sup> Edition<sup>5</sup>

Diagnostic group / subgroup	ICD-O-3 histology	ICD-O-3 site	
I. Leukaemias, myeloproliferative disease	es & 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 ne	eoplasms		
(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 miscellan	eous 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	9470-9474, 9480, 9508	C000-C809	
tumours	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 i	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<sup>5</sup>

Diagnostic group / subgroup	ICD-O-3 histology	ICD-O-3 site
VI. Renal tumours	1	
(a) Nephroblastoma and other non-	8959, 8960, 8964-8967	C000-C809
epithelial renal tumours	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	8810, 8811, 8823, 8830	C400-C419
tumours	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 sarce	omas	
(a) Rhabdomyosarcomas	8900-8905, 8910, 8912, 8920, 8991	C000-C809
(b) Fibrosarcomas, peripheral nerve sheath tumours, and other fibrous	8810, 8811, 8813-8815, 8821, 8823, 8834-8835	C000-C399, C440-C768, C809
neoplasms	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<sup>5</sup>

Diagnostic group / subgroup	ICD-O-3 histology	ICD-O-3 site
X. Germ cell tumours, trophoblastic tum	ours, and neoplasms of gonads	ı
(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	8590-8671	C000-C809
gonadal tumours	8000-8005	C569, C620-C629
XI. Other malignant epithelial neoplasm	s 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 ne	pplasms	
(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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