Cancer Council Queensland

# Childhood cancer survival in Australia 1995 – 2004

AUSTRALIAN PAEDIATRIC CANCER REGISTRY

VCRCC Viertel Centre for Research in Cancer Control

Accredited by the NHMRC as an independent research institute.
The generosity of the Queensland community and the *Sylvia and Charles Viertel Charitable Foundation* makes our research possible.

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## Foreword

The Australian Paediatric Cancer Registry (APCR) is one of the few national registers of childhood cancer in the world. Funded and managed by Cancer Council Queensland, and with appropriate ethical and legislative approvals, the APCR records clinical and treatment information on every child diagnosed with cancer in Australia.

This report, the second from the APCR in the current series, provides detailed information on survival for children in Australia following a diagnosis of cancer. It is pleasing to note that overall five-year relative survival for children with cancer is now close to 80%, a significant increase since the early 1980s. In particular, marked improvements in survival have been achieved for children diagnosed with leukaemia, lymphoma and neuroblastoma. We trust that the gains in survival and quality of life that have occurred over recent decades will continue as further developments in treatment occur.

Accurate and timely data collection is a key priority for the APCR. This has been made possible only through the support and assistance of all State and Territory Cancer Registries and all treating paediatric oncology hospitals throughout Australia. I would like to express my personal thanks to the staff involved who have given their time and energy to support this national data collection.

Finally and most importantly, we acknowledge the children and their families who are represented in this report. We hope that the information contained herein will contribute to the campaign to improve outcomes for childhood cancer and provide another step towards a cancer-free future.

Professor Jeff Dunn
Chief Executive Officer
Cancer Council Queensland

## Executive Summary

More than 600 children aged 0-14 years old are diagnosed with cancer each year in Australia. Advances in therapy have resulted in widespread improvements in survival for childhood cancer patients in recent years. However, cancer remains as one of the leading causes of death within this age group and many survivors face serious long term adverse health effects, caused by either the cancer itself or as a result of treatment.

This report describes the latest population-based survival estimates for children diagnosed with cancer in Australia using data from the Australian Paediatric Cancer Registry (APCR). The APCR is a valuable resource, as it is one of the few national registries of childhood cancer in the world. Population-based survival studies are essential for providing robust indicators to evaluate the effectiveness of healthcare provision and to monitor progress in cancer control at a national level.

Five-year relative survival for all children diagnosed with cancer in Australia during 1995-2004 was 79.5%. Significant variation was observed in survival according to diagnostic group, with 5-year relative survival ranging from 68.4% for children with neuroblastoma up to 99.1% for those with retinoblastoma.

Only minor differences were observed in survival by sex for most types of childhood cancer, except for leukaemia, for which girls (83.5%) had a significantly higher rate of 5-year relative survival compared to boys (78.1%).

Infants (aged less than 1 year old at diagnosis) tended to have poorer survival than other children with cancer. This pattern was particularly evident for leukaemias and tumours of the central nervous system. However, the reverse was true for neuroblastoma and hepatic tumours, with a decrease in survival as age at diagnosis increased.

Within the diagnostic groups where information on staging was available (lymphomas, neuroblastoma, renal tumours and the diagnostic subgroup of rhabdomyosarcomas), there was a consistent trend towards a decrease in survival as stage at diagnosis progressed. For example, among children with neuroblastoma, 5-year relative survival dropped from 95.6% for stage I tumours to 49.8% for stage IV tumours.

Five-year relative survival for all childhood cancers combined increased from 72.3% for the years 1983-1994 to 79.5% during 1995-2004. Significant improvements in survival for children who were diagnosed more recently were recorded for leukaemias (increase from 69.1% during 1983-1994 to 80.6% during 1995-2004), lymphomas (80.8% to 89.6%) and neuroblastoma (52.9% to 68.4%). In contrast, there was a significant decrease in 5-year relative survival within the diagnostic subgroup of non-rhabdomyosarcoma soft tissue sarcomas, declining from 84.5% to 72.5% between 1983-1994 and 1995-2004.

Survival for all childhood cancers combined and for the various diagnostic groups was generally similar in Australia to corresponding estimates from other developed countries including the United States, Great Britain and France.

Despite the improvement in survival for children in Australia with cancer, about 20% of those who are diagnosed will still die within 5 years. In the future, it is hoped that developments in treatment protocols through large multi-centre studies, particularly advances in the ability to tailor treatments for individual patients and tumours, will lead to further increases in survival rates while at the same time improving longer term quality of life for childhood cancer survivors.

# 1 Introduction

## *1.1 Background information*

More than 600 children under 15 years of age are diagnosed with cancer each year in Australia[1] and on average around 100 children die from cancer annually.[2] Thus, although cancer is relatively rare among children compared to people in the older age groups, it is still the most common cause of disease-related death for children aged 1-14 years old in Australia.[3]

Survival after a diagnosis of cancer is dependent on a number of factors such as early detection and access to appropriate and co-ordinated treatment services.[3] The main treatments for children with cancer include surgery, chemotherapy and radiotherapy.[4] Chemotherapy tends to be particularly effective because it targets fast-growing cells which are typical in many forms of childhood cancer. Ongoing advances in the treatments available and their application have led to large improvements in survival for childhood cancer over the last few decades. Most of these breakthroughs have come about as a direct result of collaborative clinical trials.[5,6]

This positive news is tempered by the knowledge that some of the children who survive may experience long term adverse health effects, either because of the cancer itself or as a result of treatment. The medical issues faced by survivors of childhood cancer can include increased risk of subsequent cancer, organ dysfunction (cardiopulmonary, renal, gastrointestinal), impaired growth and development, decreased fertility and neurocognitive deficits.[7,8]

Nonetheless, survival estimates are a key indicator of success in cancer control. Clinical trials can only report on outcomes for a selected group of patients, rather than indicating the overall survival experience for all people with cancer in a particular region or country. Population-based survival studies such as this are therefore an important part of evaluating the effectiveness of treatment services for childhood cancer patients at a national level.

## *1.2 The Australian Paediatric Cancer Registry*

The Australian Paediatric Cancer Registry (APCR) is one of only a few population-based national registries of childhood cancer in the world. Information is collected on incidence, survival, treatment and stage at diagnosis (where available) for all children in Australia who are diagnosed with cancer between the ages of 0-14 years. While details on cancer incidence and survival by age group are available from each individual State and Territory Cancer Registry, these registries do not collect data on the stage of disease or the treatment given to children with cancer. This information is essential in order to set and measure standards of care for children with cancer and to track improvements in treatment outcomes over time.

Notification of invasive cancer is a statutory requirement for all public and private hospitals, nursing homes and pathology services in Australia. With the appropriate ethical and legislative approvals, the APCR collects clinical and demographic information about all cases of childhood cancer from the State and Territory Cancer Registries and directly from the major treating hospitals. A comprehensive quality assurance process is used. Registry records are cross-checked against hospital records to resolve any inconsistencies or differences in the information collected. Strict protocols are in place to ensure the privacy and confidentiality of all information stored on the APCR database.

Cancer Council Queensland (CCQ), formerly known as the Queensland Cancer Fund, has provided financial support for the APCR since its inception in 1977. The APCR was transferred to Brisbane from Sydney in 1983, and in 2004 CCQ took over its management.

Currently there are 24 years of detailed and verified data available from the APCR (1983 to 2006), involving approximately 14,000 cases.

## 1.3 Classification of childhood cancers

There are several important differences between childhood and adult cancers, such as how the cancer originates and various clinical characteristics.[9] Many childhood cancers develop as a result of abnormal cell maturation; therefore, the tissue of origin, rather than the location in the body where the cancer starts, is the best predictor of tumour behaviour, prognosis and the required treatment. Childhood cancers are consequently classified using a different coding system to adult cancers, with categories based on the type of cancer tissue (morphology) instead of where the cancer occurs in the body.[10,11]

The third edition of the International Classification of Childhood Cancers (ICCC-3), the current standard for reporting childhood cancer, comprises 12 major diagnostic groups and 47 subgroups[10] (Table 1). Full details of the codes for each diagnostic group and subgroup are included in Appendix Table A.1, page 38.

**Table 1 International Classification of Childhood Cancers, 3rd edition (ICCC-3) Diagnostic Groups**

| Diagnostic Group | Full title | Abbreviated title used in this report |
|---|---|---|
| I. | Leukaemias, myeloproliferative diseases and myelodysplastic diseases | Leukaemias |
| II. | Lymphomas and reticuloendothelial neoplasms | Lymphomas |
| III. | Central nervous system and miscellaneous intracranial and intraspinal neoplasms | Tumours of the central nervous system |
| IV. | Neuroblastoma and other peripheral nervous cell tumours | Neuroblastoma |
| V. | Retinoblastoma | Retinoblastoma |
| VI. | Renal tumours | Renal tumours |
| VII. | Hepatic tumours | Hepatic tumours |
| VIII. | Malignant bone tumours | Malignant bone tumours |
| IX. | Soft tissue and other extraosseous sarcomas | Soft tissue sarcomas |
| X. | Germ cell tumours, trophoblastic tumours and neoplasms of gonads | Germ cell tumours |
| XI. | Other malignant epithelial neoplasms and melanomas | Other malignant epithelial neoplasms and melanomas |
| XII. | Other and unspecified malignant neoplasms | Other malignant neoplasms |

Source: Steliarova-Foucher et al, 2005.[10]

Most of the cases of childhood cancer included in this report are malignant (or invasive) neoplasms. Although tumours of benign or uncertain behaviour are generally not included in cancer registry data for adults, the ICCC-3 includes non-malignant intracranial and intraspinal tumours within diagnostic groups III and X due to similarities in their clinical symptoms and prognosis compared to the corresponding malignant tumours.[10] ***Therefore, throughout this report, childhood cancer refers to all malignant neoplasms as well as intracranial and intraspinal tumours of benign or uncertain behaviour within the diagnostic groups "Tumours of the central nervous system" and "Germ cell tumours".*** Other non-invasive tumours (e.g. in situ melanomas) are not part of the international standard, and hence have been excluded from all data shown in the report.

Due to the small number of cases in diagnostic group XII ("Other malignant neoplasms"), no further analysis was undertaken nor detailed results presented for this group.

## 1.4 Overview of this report

This report provides a comprehensive analysis of population-based survival outcomes for children with cancer throughout Australia based on data contained in the APCR. It is a companion document to a report titled "Childhood cancer incidence in Australia, 1983-2006"[1] which was published in December 2009. Copies of both reports are available on request from the Viertel Centre for Research in Cancer Control at Cancer Council Queensland (research@cancerqld.org.au). They can also be downloaded from the CCQ website by visiting "www.cancerqld.org.au/page/Research_statistics/VCRCC/Statistical_reports".

The main focus of this report is on the survival of all children diagnosed with cancer in Australia during 1995-2004, with follow-up to 31st December 2006. Estimates are expressed in terms of relative survival, which adjusts the observed rate of survival among children with cancer by the underlying survival of all children in the general population, and were calculated using the cohort method, with an emphasis on 5-year survival (see Appendix A.3.3, page 43, for further details on survival methodology).

Survival data are presented for all cancers combined and for each diagnostic group by time since diagnosis, sex and age group at diagnosis. Where possible, the age groups used were <1 year, 1-4 years, 5-9 years and 10-14 years. However, these age groups were collapsed if there was insufficient data to calculate reliable survival estimates. For example, children aged <1 year and 1-4 years were combined into the 0-4 age group for the diagnostic groups of lymphomas and soft tissue sarcomas.

Within the various diagnostic groups, survival estimates were also calculated for the major subgroups and by stage, which provides an indication of how advanced the cancer was at diagnosis. (Information on stage was only available for certain cancers, including lymphomas, neuroblastoma, renal tumours and the diagnostic subgroup of rhabdomyosarcomas within soft tissue sarcomas.) In addition, changes in survival over time were examined by comparing the survival of children diagnosed during 1995-2004 with children diagnosed in the period 1983-1994. Finally, the survival of childhood cancer patients in Australia was benchmarked against corresponding international estimates.

Throughout the report, a measure known as the “p-value” has been used to determine whether survival estimates are different to each other in a statistical sense. P-values have a range of 0 to 1; the smaller the p-value, the greater the likelihood that any variation has not occurred by chance alone. In particular, a p-value of less than 0.05 indicates that the difference between survival estimates is statistically significant.

Information on the definitions for the diagnostic groups and subgroups, as well as an explanation of the statistical terminology and methods (including the calculation of p-values), can be found in the Appendix.

## 2 Survival for all childhood cancers combined

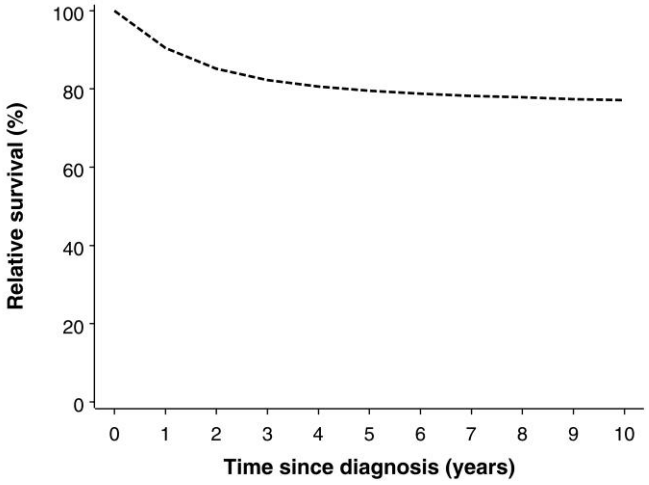
Survival by time since diagnosis

Most of the deaths among children with cancer occurred in the first few years following diagnosis. Relative survival for all cancers combined was 90.4% by the end of the first year after diagnosis compared to 79.5% after 5 years, then slowly decreased to 77.2% after 10 years (Box 2a).

**Box 2a: Relative survival by time since diagnosis for all childhood cancers (n=6,177)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 90.4 (89.7-91.1) |
| 2 years | 85.2 (84.3-86.0) |
| 3 years | 82.3 (81.3-83.2) |
| 4 years | 80.6 (79.6-81.6) |
| 5 years | 79.5 (78.5-80.5) |
| 6 years | 78.8 (77.7-79.8) |
| 7 years | 78.2 (77.1-79.2) |
| 8 years | 77.9 (76.7-78.9) |
| 9 years | 77.4 (76.2-78.5) |
| 10 years | 77.2 (76.0-78.3) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.




Survival by diagnostic group

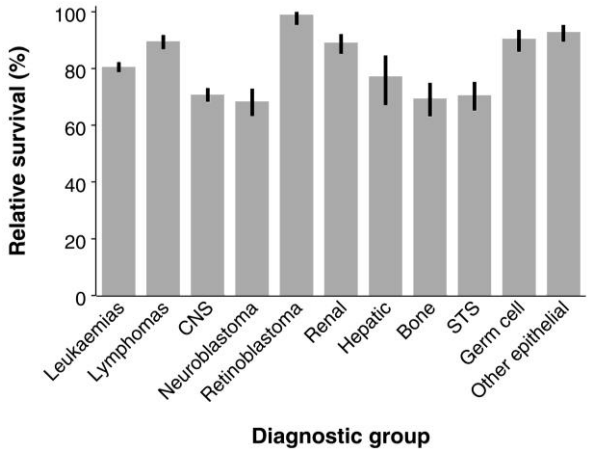
Highly significant differences existed for 5-year relative survival between the childhood cancer diagnostic groups (Box 2b). Survival was highest for retinoblastoma (99.1%), other malignant epithelial neoplasms and melanomas (93.0%), germ cell tumours (90.5%), lymphomas (89.6%) and renal tumours (89.2%), while survival was lowest for neuroblastoma (68.4%), malignant bone tumours (69.5%), soft tissue sarcomas (70.6%) and tumours of the central nervous system (70.8%).

**Box 2b: Five-year relative survival for all childhood cancers by diagnostic group, Australia, 1995-2004[a]**

| Diagnostic group | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| I. Leukaemias | 2,036 | 80.6 (78.7-82.3) |
| II. Lymphomas | 607 | 89.6 (86.8-91.8) |
| III. Tumours of the central nervous system (CNS)[d] | 1,406 | 70.8 (68.3-73.2) |
| IV. Neuroblastoma | 367 | 68.4 (63.3-73.0) |
| V. Retinoblastoma | 159 | 99.1 (95.4-100.0) |
| VI. Renal tumours | 328 | 89.2 (85.2-92.2) |
| VII. Hepatic tumours | 93 | 77.3 (67.1-84.7) |
| VIII. Malignant bone tumours | 263 | 69.5 (63.2-75.0) |
| IX. Soft tissue sarcomas (STS) | 333 | 70.6 (65.2-75.3) |
| X. Germ cell tumours[d] | 244 | 90.5 (86.0-93.7) |
| XI. Other malignant epithelial neoplasms & melanomas | 324 | 93.0 (89.4-95.4) |
| | | *p < 0.001* |

*(Continued next page...)*

**Box 2b (*cont.*): Five-year relative survival for all childhood cancers by diagnostic group, Australia, 1995-2004[a]**




Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).
d. Diagnostic group includes intracranial and intraspinal tumours of benign or uncertain behaviour.

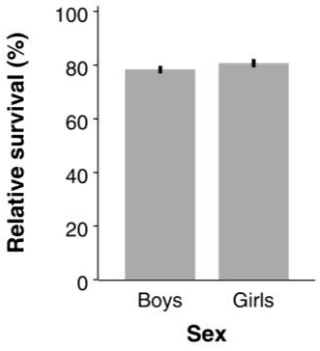
Survival by sex

Relative survival for all cancers combined was slightly, but significantly, higher for girls (80.8% after 5 years) compared to boys (78.4% - Box 2c). This appeared to be mainly due to the higher survival rate for girls with leukaemia (see Box 3.1c).

**Box 2c: Five-year relative survival for all childhood cancers by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 3,363 | 78.4 (77.0-79.8) |
| Girls | 2,814 | 80.8 (79.2-82.2) |
| | | *p = 0.022* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



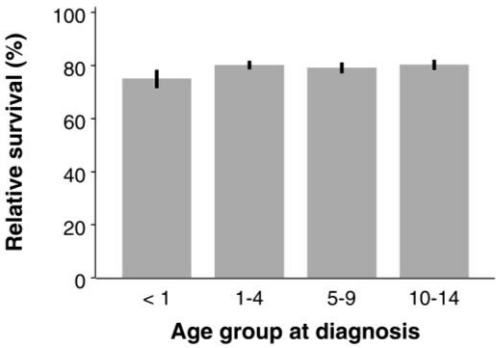

Survival by age group at diagnosis

Infants (under 1 year of age) had significantly poorer survival for all cancers combined compared to other children. Five-year relative survival varied from 75.1% for children less than 1 year old to around 80% for those in the 1-4, 5-9 and 10-14 age groups (Box 2d). Again, this result was likely to be heavily influenced by variation in survival for leukaemia by age at diagnosis (see Box 3.1d). Survival for all cancers combined may also be affected by the mix of cancers that are most commonly diagnosed within a particular age group. For example, the majority of neuroblastoma, retinoblastoma, renal tumours and hepatic tumours occur before 4 years of age, whereas malignant bone tumours and other malignant epithelial neoplasms are more common in the 10-14 age group.[1]

**Box 2d: Five-year relative survival for all childhood cancers by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| < 1 year | 606 | 75.1 (71.4-78.4) |
| 1-4 years | 2,231 | 80.2 (78.5-81.9) |
| 5-9 years | 1,569 | 79.2 (77.0-81.2) |
| 10-14 years | 1,771 | 80.3 (78.3-82.2) |
| | | *p = 0.009* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



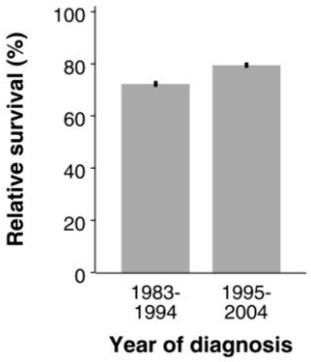

Survival by year of diagnosis

A significant improvement in survival over time was recorded among all children with cancer. Five-year relative survival increased from 72.3% for children diagnosed during 1983-1994 to 79.5% during 1995-2004 (Box 2e).

**Box 2e: Five-year relative survival for all childhood cancers by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 6,398 | 72.3 (71.2-73.4) |
| 1995-2004 | 6,177 | 79.5 (78.5-80.5) |
| | | *p < 0.001* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Five-year survival for all childhood cancers in Australia (80%) was similar to recent results that have been published for the United States (80%), France and Great Britain (both 78% - see Box 2f). Estimates of overall childhood cancer survival during 1995-2002 for many other countries in Europe also ranged between 78% to 83% after 5 years.[12]

**Box 2f: Five-year survival for all childhood cancers in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 6,177 | Relative survival – cohort method | 80 (78-81) |
| France[13] | 1995-1999 | 1,864 | Observed survival – cohort method | 78 (76-80) |
| Great Britain[14] | 2001-2005 | 7,737 | Observed survival – cohort method | 78 (77-79) |
| United States[15] | 1999-2006 | 18,286 | Relative survival – cohort method | 80 (79-81) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

# 3 Survival by childhood cancer diagnostic group

## *3.1 Leukaemias, myeloproliferative diseases and myelodysplastic diseases*

Background

Leukaemias are cancers that arise from uncontrolled overproduction of abnormal white blood cells within the bone marrow. White blood cells are an important component of the immune system, and are involved in the body's response to bacterial, viral and parasitic infection. However, in leukaemia patients, the presence of abnormal cells results in healthy blood cells being 'crowded out' of the bone marrow.

Most leukaemias which occur in children are acute, meaning that the cancerous cells are both immature and produced rapidly. Leukaemia is further classified according to the predominant type of white blood cells that are present: lymphoid cells or myeloid cells.[9,16] Lymphoid leukaemias are the most common subgroup, accounting for more than three-quarters (78.2%) of all cases of childhood leukaemia in Australia during 1995-2004, followed by acute myeloid leukaemias (16.2%).

The symptoms of leukaemia are mainly those of inadequate bone marrow function, such as tiredness, fever, pallor and easy bruising. Some children also experience bone pain as a result of increased bone marrow activity.[7,9,16,17]

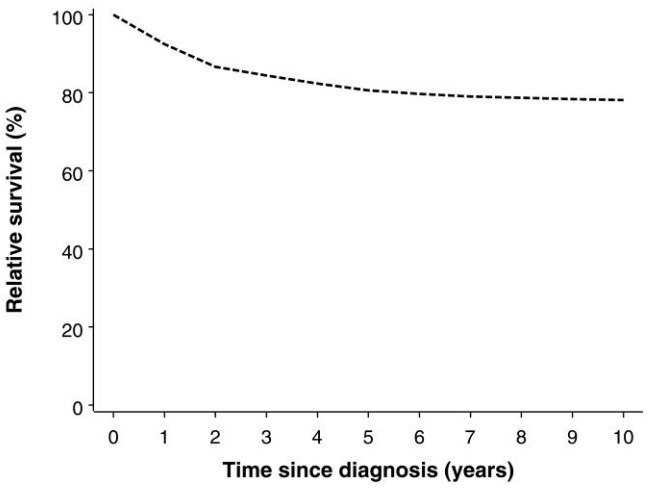
Survival by time since diagnosis

By the end of the first year after diagnosis, relative survival for childhood leukaemia was 92.4%. Survival continued to decrease to 80.6% after 5 years before levelling out to 78.2% after 10 years (Box 3.1a).

**Box 3.1a: Relative survival by time since diagnosis for childhood leukaemias (n=2,036)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 92.4 (91.2-93.5) |
| 2 years | 86.7 (85.1-88.1) |
| 3 years | 84.5 (82.8-86.0) |
| 4 years | 82.4 (80.6-84.0) |
| 5 years | 80.6 (78.7-82.3) |
| 6 years | 79.7 (77.8-81.5) |
| 7 years | 79.0 (77.1-80.9) |
| 8 years | 78.7 (76.7-80.5) |
| 9 years | 78.4 (76.3-80.3) |
| 10 years | 78.2 (76.1-80.1) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.



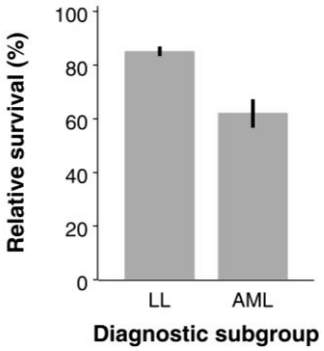

Survival by diagnostic subgroup

Highly significant differences were found by diagnostic subgroup, with lymphoid leukaemia estimated to have a 5-year relative survival rate of 85.3% compared to 62.3% for acute myeloid leukaemia (Box 3.1b).

**Box 3.1b: Five-year relative survival for childhood leukaemias by diagnostic subgroup, Australia, 1995-2004[a]**

| Diagnostic subgroup[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| LL | 1,592 | 85.3 (83.3-87.0) |
| AML | 329 | 62.3 (56.7-67.3) |
| | | *p < 0.001* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. LL = lymphoid leukaemias; AML = acute myeloid leukaemias.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



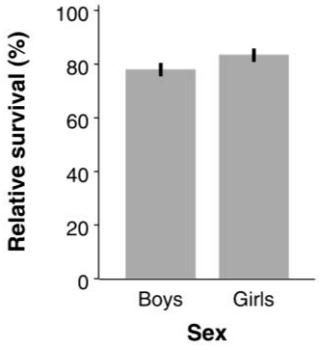

Survival by sex

Girls with leukaemia had a significantly higher rate of survival compared to boys. Five-year relative survival was 83.5% among girls and 78.1% for boys (Box 3.1c). The difference in relative survival by sex was only significant for lymphoid leukaemias (88.2% after 5-years for girls compared to 82.7% for boys) with no disparity recorded for acute myeloid leukaemias (see Appendix Table A.4a, page 44).

**Box 3.1c: Five-year relative survival for childhood leukaemias by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 1,094 | 78.1 (75.4-80.5) |
| Girls | 942 | 83.5 (80.9-85.8) |
| | | *p < 0.001* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



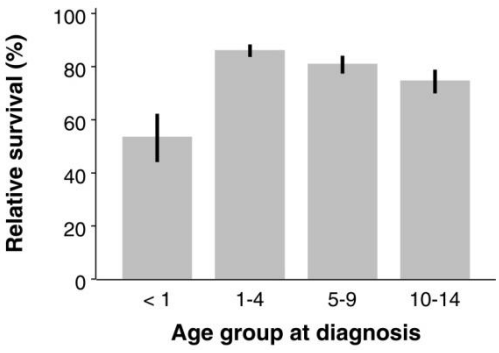

Survival by age group at diagnosis

There was a large amount of variation in survival by age group for childhood leukaemia (Box 3.1d). Relative survival was significantly lower among children under 1 year old (53.6% after 5 years), but increased sharply for children diagnosed between the ages of 1-4 (86.2%), before successive decreases within the 5-9 and 10-14 age groups (81.0% and 74.7% respectively). A similar, significant pattern in survival by age group was found for children with lymphoid leukaemia, but the differences were less pronounced for the diagnostic subgroup of acute myeloid leukaemias (see Appendix Table A.4b, page 45).

**Box 3.1d: Five-year relative survival for childhood leukaemias by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| < 1 year | 118 | 53.6 (44.1-62.3) |
| 1-4 years | 951 | 86.2 (83.7-88.3) |
| 5-9 years | 569 | 81.0 (77.4-84.1) |
| 10-14 years | 398 | 74.7 (69.9-78.9) |
| | | *p < 0.001* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



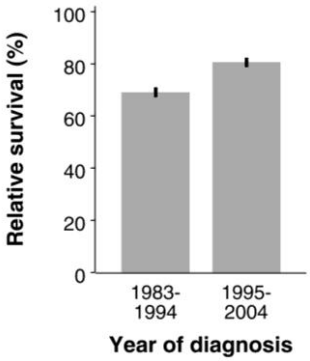
Survival by year of diagnosis

Survival for childhood leukaemia has improved significantly in recent years (Box 3.1e). Children who were diagnosed with leukaemia during 1983-1994 had a 5-year relative survival rate of 69.1% compared to 80.6% during 1995-2004. Improvements in survival were recorded for both lymphoid leukaemias (from 75.5% to 85.3% between 1983-1994 and 1995-2004) and acute myeloid leukaemias (from 44.1% to 62.3% - see Appendix Table A.4d, page 47).

**Box 3.1e: Five-year relative survival for childhood leukaemias by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 2,113 | 69.1 (67.1-71.0) |
| 1995-2004 | 2,036 | 80.6 (78.7-82.3) |
| | | *p < 0.001* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Of the selected countries for which estimates were available (Box 3.1f), 5-year survival for children with leukaemia ranged from 78% in France to 83% in Great Britain, compared to 81% in Australia.

**Box 3.1f: Five-year survival for childhood leukaemias in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 2,036 | Relative survival – cohort method | 81 (79-82) |
| France[13] | 1995-1999 | 567 | Observed survival – cohort method | 78 (75-82) |
| Great Britain[14] | 2001-2005 | 2,429 | Observed survival – cohort method | 83 (81-85) |
| United States[15] | 1999-2006 | 6,041 | Relative survival – cohort method | 82 (81-83) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.2 Lymphomas and reticuloendothelial neoplasms*

Background

Lymphomas are also cancers of the lymphoid subgroup of white blood cells, and therefore they have some similarities to lymphoid leukaemias. However, the abnormal white blood cells in lymphomas are principally found in lymph glands, with only occasional bone marrow involvement. Lymph glands are a network of clusters of immune cells which intercept and coordinate the response to infections. These glands, which include the tonsils and adenoids, are found in the neck, armpits, groin and within the chest and abdomen.

There are two main types of lymphoma, depending on the lymphoid cells involved – Hodgkin lymphomas[18] and non-Hodgkin lymphomas[19,20] (which includes the subgroup of Burkitt lymphomas). During 1995-2004, Hodgkin lymphomas accounted for 39.5% of all childhood lymphomas diagnosed within Australia, while 38.9% of cases were non-Hodgkin lymphomas (excluding Burkitt lymphomas) and a further 19.6% were classified as Burkitt lymphomas.

The type of lymphoma determines treatment and prognosis. Hodgkin lymphomas tend to be relatively slow-growing, and cases present with painless lumps at sites where lymph glands are located. Some children with Hodgkin lymphoma also develop fevers and night sweats or experience unexplained weight loss. In contrast, non-Hodgkin lymphomas tend to progress much more rapidly. Children with non-Hodgkin lymphoma usually present as very unwell, with swelling in the abdomen or chest causing obstruction to major organs, such as the bowel or trachea (windpipe).[7,9]

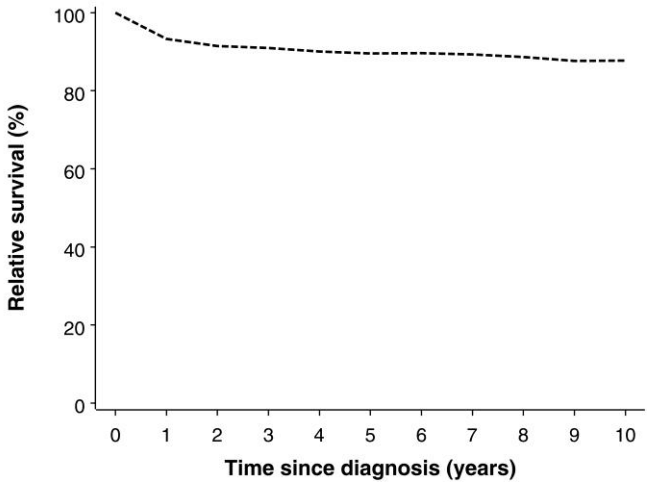
Survival by time since diagnosis

The relative rate of survival among children diagnosed with lymphoma decreased slightly from 93.3% 1 year after diagnosis to 89.6% after 5 years, with 10-year relative survival estimated at 87.7% (Box 3.2a).

**Box 3.2a: Relative survival by time since diagnosis for childhood lymphomas (n=607)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 93.3 (91.0-95.0) |
| 2 years | 91.5 (89.0-93.4) |
| 3 years | 91.0 (88.4-93.0) |
| 4 years | 90.0 (87.3-92.2) |
| 5 years | 89.6 (86.8-91.8) |
| 6 years | 89.6 (86.8-91.9) |
| 7 years | 89.3 (86.5-91.6) |
| 8 years | 88.6 (85.5-91.1) |
| 9 years | 87.7 (84.3-90.4) |
| 10 years | 87.7 (84.3-90.4) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.



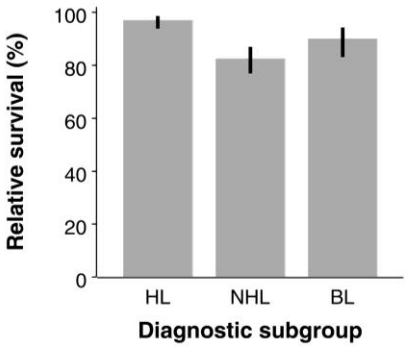

Survival by diagnostic subgroup

There were significant differences in survival among the main diagnostic subgroups for lymphoma (Box 3.2b). Five-year relative survival rates varied from 82.6% for non-Hodgkin lymphomas (excluding Burkitt lymphoma) to 90.1% for Burkitt lymphomas and 97.1% for Hodgkin lymphomas.

**Box 3.2b: Five-year relative survival for childhood lymphomas by diagnostic subgroup, Australia, 1995-2004[a]**

| Diagnostic subgroup[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| HL | 240 | 97.1 (93.8-98.7) |
| NHL | 236 | 82.6 (76.9-86.9) |
| BL | 119 | 90.1 (83.1-94.3) |
| | | *p < 0.001* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. HL = Hodgkin lymphomas; NHL = Non-Hodgkin lymphomas (excluding Burkitt lymphoma); BL = Burkitt lymphomas.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



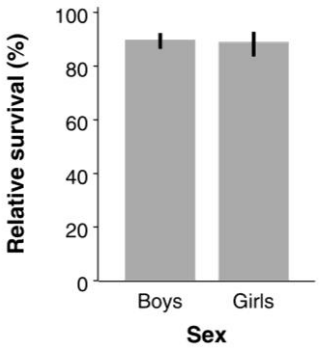

Survival by sex

The 5-year relative survival rate for childhood lymphoma was similar for boys and girls (89.8% and 89.1% respectively, see Box 3.2c). There were also no significant differences in survival by sex within any of the diagnostic subgroups for lymphoma (see Appendix Table A.4a, page 44).

**Box 3.2c: Five-year relative survival for childhood lymphomas by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 422 | 89.8 (86.4-92.4) |
| Girls | 185 | 89.1 (83.6-92.8) |
| | | *p = 0.594* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



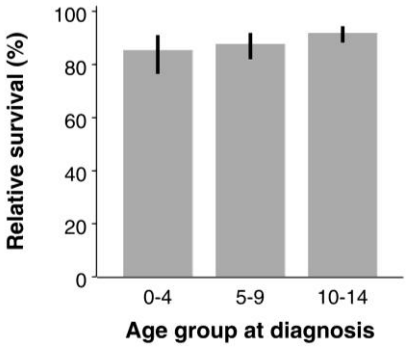

Survival by age group at diagnosis

No significant differences were found in relative survival by age at diagnosis for children with lymphoma, despite 5-year survival being somewhat higher within the 10-14 age group (91.9%) compared to children who were diagnosed when they were younger (85.4% in the 0-4 age group – see Box 3.2d). Further analysis by diagnostic subgroup revealed that there were no significant differences in survival by age (see Appendix Table A.4b, page 45).

**Box 3.2d: Five-year relative survival for childhood lymphomas by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| 0-4 years | 101 | 85.4 (76.5-91.1) |
| 5-9 years | 186 | 87.8 (82.0-91.9) |
| 10-14 years | 320 | 91.9 (88.3-94.5) |
| | | *p = 0.154* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. Age groups "< 1 year" and "1-4 years" have been combined due to the small number of cases in these age groups.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



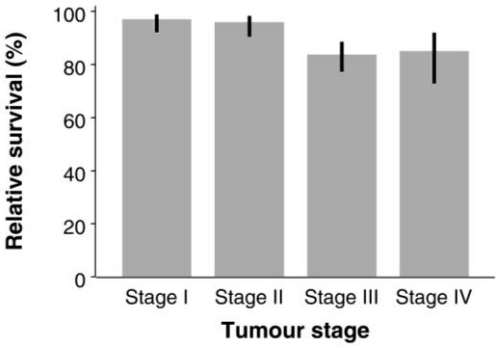
Survival by tumour stage

Survival for childhood lymphoma varied significantly by stage at diagnosis, with 5-year relative survival rates higher for stage I and II tumours (97.0% and 96.0%) and lower for stage III and IV tumours (83.8% and 85.0% - Box 3.2e). A similar pattern was recorded for each of the diagnostic subgroups, although the difference in survival by tumour stage was only statistically significant for Hodgkin lymphomas (Appendix Table A.4c, page 46).

**Box 3.2e: Five-year relative survival for childhood lymphomas by tumour stage[a], Australia, 1995-2004[b]**

| Tumour stage[a] | Number of cases[c,d] | Five-year relative survival (95% CI)[e] |
|---|---|---|
| Stage I | 134 | 97.0 (92.1-98.9) |
| Stage II | 125 | 96.0 (90.4-98.4) |
| Stage III | 174 | 83.8 (77.3-88.6) |
| Stage IV | 65 | 85.0 (72.8-92.0) |
| | | *p < 0.001* |

Notes: a. The Ann Arbor classification system[21] was used to stage Hodgkin lymphomas and the Murphy classification system[22] was used to stage non-Hodgkin lymphomas (including Burkitt lymphomas).
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. The initial number of eligible cases in the cohort.
d. Stage was unknown for 109 cases (18.0%).
e. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



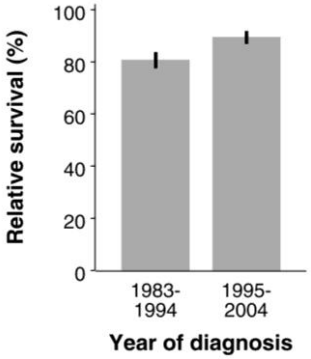

Survival by year of diagnosis

Five-year relative survival for childhood lymphoma improved significantly over time (Box 3.2f), increasing from 80.8% during 1983-1994 to 89.6% during 1995-2004. In terms of the diagnostic subgroups, children with Burkitt lymphoma recorded a statistically significant improvement over the two time periods, with 5-year relative survival increasing from 73.5% to 90.1% (Appendix Table A.4d, page 47).

**Box 3.2f: Five-year relative survival for childhood lymphomas by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 625 | 80.8 (77.5-83.7) |
| 1995-2004 | 607 | 89.6 (86.8-91.8) |
| | | *p < 0.001* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Survival for children with lymphoma was quite consistent (between 88% to 90% after 5 years) in each of the countries for which recent estimates were available (Box 3.2g).

**Box 3.2g: Five-year survival for childhood lymphomas in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 607 | Relative survival – cohort method | 90 (87-92) |
| France[13] | 1995-1999 | 226 | Observed survival – cohort method | 90 (86-94) |
| Great Britain[14] | 2001-2005 | 783 | Observed survival – cohort method | 88 (86-91) |
| United States[15] | 1999-2006 | 1,783 | Relative survival – cohort method | 88 (87-90) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.3 Central nervous system and miscellaneous intracranial and intraspinal neoplasms*

Background

The central nervous system (CNS) consists of the brain and the spinal cord, which starts at the base of the brain and extends about half way down the spine. Tumours of the CNS vary widely in terms of pathologic appearance, behaviour and prognosis. They can occur anywhere in the CNS but are most common in the brain, especially among children.[23]

Astrocytomas form the most common diagnostic subgroup and were responsible for 47.7% of all childhood tumours of the CNS diagnosed in Australia during 1995-2004, followed by intracranial and intraspinal embryonal tumours (18.7%), other gliomas (11.7%) and ependymomas and choroid plexus tumours (9.2%). The remaining 12.8% were other specified and unspecified intracranial and intraspinal neoplasms.

Presenting features are determined mainly by the location of the tumour. Many brain tumours result in an increase of pressure within the head. In turn, this may cause headaches, vomiting and sleepiness. Other local effects may include abnormal eye movements, loss of coordination, seizures, weakness in the limbs or difficulties with swallowing.[7,9,24]

Both malignant and non-malignant childhood tumours of the central nervous system may have similar symptoms and outcomes, depending on characteristics such as their histology and grade. Therefore, they have been combined within the same diagnostic group for this report, in accordance with the ICCC-3 classification.[10] Of the 1,406 eligible cases included in the cohort, 859 children (61.1%) had a malignant tumour, while the remaining 547 (38.9%) had tumours that were either benign or of uncertain behaviour.

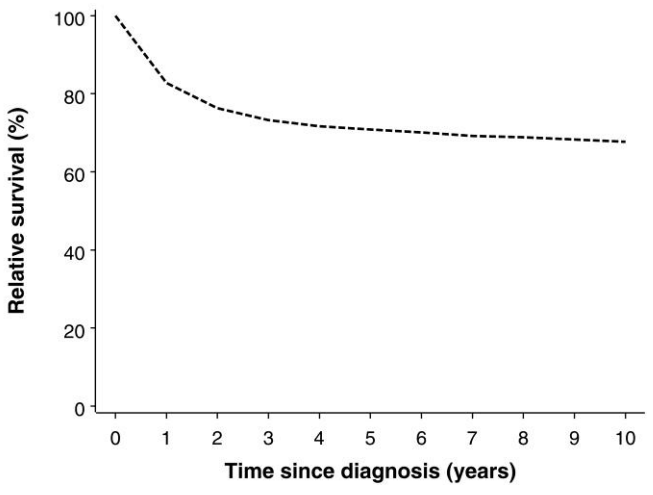
Survival by time since diagnosis

Relative survival for children diagnosed with tumours of the CNS was 82.8% after 1 year, 70.8% after 5 years and 67.7% after 10 years (Box 3.3a).

**Box 3.3a: Relative survival by time since diagnosis for childhood tumours of the central nervous system (n=1,406)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 82.8 (80.7-84.6) |
| 2 years | 76.3 (74.0-78.5) |
| 3 years | 73.2 (70.8-75.5) |
| 4 years | 71.7 (69.2-74.0) |
| 5 years | 70.8 (68.3-73.2) |
| 6 years | 70.1 (67.6-72.5) |
| 7 years | 69.2 (66.6-71.6) |
| 8 years | 68.9 (66.3-71.3) |
| 9 years | 68.3 (65.6-70.8) |
| 10 years | 67.7 (64.9-70.4) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.



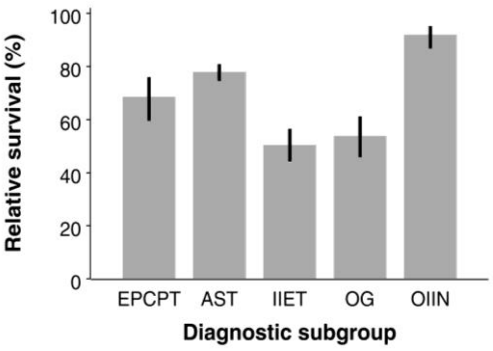

Survival by diagnostic subgroup

Five-year relative survival was highest for the combined diagnostic subgroups of other intracranial and intraspinal neoplasms (92.0%), followed by astrocytomas (77.9%) and ependymomas and choroid plexus tumours (68.6%), while survival was lowest for children with intracranial and intraspinal embryonal tumours (50.5%) and other gliomas (53.8% - see Box 3.3b).

**Box 3.3b: Five-year relative survival for childhood tumours of the central nervous system by diagnostic subgroup, Australia, 1995-2004[a]**

| Diagnostic subgroup[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| EPCPT[e] | 129 | 68.6 (59.6-76.0) |
| AST | 670 | 77.9 (74.5-80.9) |
| IIET | 263 | 50.5 (44.2-56.5) |
| OG | 164 | 53.8 (45.8-61.2) |
| OIIN[f] | 180 | 92.0 (86.8-95.2) |
| | | *p < 0.001* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. EPCPT = ependymomas and choroid plexus tumours; AST = astrocytomas; IIET = intracranial/intraspinal embryonal tumours; OG = other gliomas; OIIN = other intracranial & intraspinal neoplasms.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).
e. 5-year relative survival was 65.3% (108 cases) for ependymomas and 85.8% (21 cases) for choroid plexus tumours.
f. "Other intracranial and intraspinal neoplasms" include the diagnostic subgroups other specified intracranial & intraspinal neoplasms and unspecified intracranial & intraspinal neoplasms.



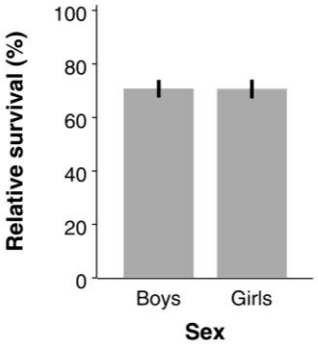

Survival by sex

Rates of survival 5 years after diagnosis for childhood tumours of the CNS were similar for boys and girls (70.9% and 70.8% respectively – see Box 3.3c). No significant differences in survival by sex were found within any of the diagnostic subgroups for tumours of the CNS (Appendix Table A.4a, page 44).

**Box 3.3c: Five-year relative survival for childhood tumours of the central nervous system by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 748 | 70.9 (67.5-74.0) |
| Girls | 658 | 70.8 (67.0-74.1) |
| | | *p = 0.999* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



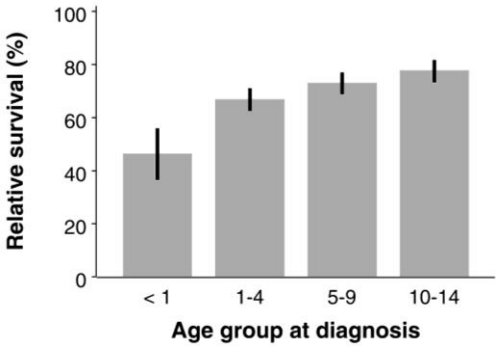

Survival by age group at diagnosis

There was a clear trend in survival for childhood tumours of the CNS by age at diagnosis (Box 3.3d). Five-year relative survival increased from 46.5% for children under 1 year old up to 77.8% within the 10-14 age group. The diagnostic subgroups of ependymomas and choroid plexus tumours and intracranial and intraspinal embryonal tumours also exhibited significant differences in survival by age group (see Appendix Table A.4b, page 45). For example, 5-year relative survival for intracranial and intraspinal embryonal tumours ranged from 2.5% among infants (< 1 year old at diagnosis) to 66.2% among children in the 5-9 age group.

**Box 3.3d: Five-year relative survival for childhood tumours of the central nervous system by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| < 1 year | 89 | 46.5 (36.5-56.0) |
| 1-4 years | 468 | 67.0 (62.5-71.1) |
| 5-9 years | 465 | 73.2 (68.8-77.1) |
| 10-14 years | 384 | 77.8 (73.2-81.7) |
| | | *p < 0.001* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



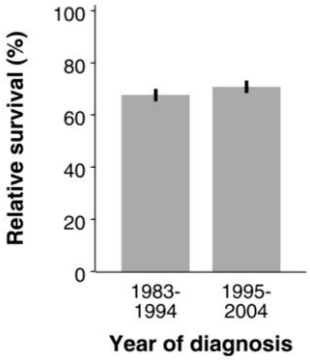

Survival by year of diagnosis

There was only a small, non-significant increase in 5-year relative survival between 1983-1994 (67.7%) and 1995-2004 (70.8%) for children with tumours of the CNS (Box 3.3e). However, a large improvement was recorded within the diagnostic subgroup of other intracranial and intraspinal tumours, with 5-year relative survival increasing from 80.0% to 92.0% between these two time periods (see Appendix Table A.4d, page 47).

**Box 3.3e: Five-year relative survival for childhood tumours of the central nervous system by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 1,437 | 67.7 (65.2-70.0) |
| 1995-2004 | 1,406 | 70.8 (68.3-73.2) |
| | | *p = 0.107* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Five-year survival for childhood tumours of the CNS was similar in Australia to both Great Britain and the United States (each 71%) but somewhat higher than for France (65% - see Box 3.3f).

**Box 3.3f: Five-year survival for childhood tumours of the central nervous system in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 1,406 | Relative survival – cohort method | 71 (68-73) |
| France[13] | 1995-1999 | 402 | Observed survival – cohort method | 65 (61-70) |
| Great Britain[14] | 2001-2005 | 1,930 | Observed survival – cohort method | 71 (69-73) |
| United States[15] | 1999-2006 | 3,799 | Relative survival – cohort method | 71 (70-73) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.4 Neuroblastoma and other peripheral nervous cell tumours*

Background

Neuroblastoma is a cancer of specialised nerve cells (neural crest cells) which form the sympathetic nervous system. This system controls automatic body functions, such as blood pressure regulation, and runs along the length of the spine from the neck to the pelvis.

Tumours can develop anywhere along the sympathetic nervous system, but most commonly originate in the adrenal glands in the abdomen. Less common sites include the chest, neck or pelvis. Neuroblastoma often spreads to other parts of the body, such as the bone marrow, bones, liver or skin. However, tumours can also spontaneously regress in some cases, particularly among infants.[9,25]

Symptoms of neuroblastoma depend on the site and size of the tumour. There may be a palpable mass in the abdomen, respiratory problems, or bladder or bowel dysfunction. Older children with widespread disease commonly experience skeletal pain, fever, anaemia or general failure to thrive.[7,9,25]

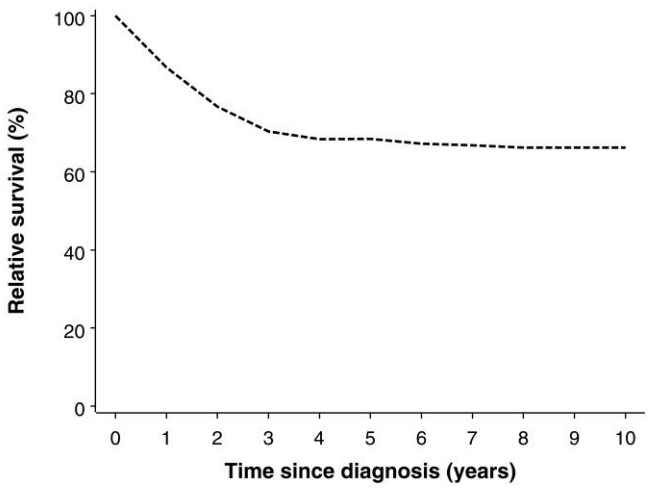
Survival by time since diagnosis

One-year relative survival for children with neuroblastoma was 86.7%. Survival rates continued to decrease further to 68.4% after 5 years, but then stabilised, with 10-year relative survival estimated at 66.2% (Box 3.4a).

**Box 3.4a: Relative survival by time since diagnosis for childhood neuroblastoma (n=367)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 86.7 (82.7-89.8) |
| 2 years | 76.7 (72.0-80.7) |
| 3 years | 70.4 (65.3-74.8) |
| 4 years | 68.4 (63.3-73.0) |
| 5 years | 68.4 (63.3-73.0) |
| 6 years | 67.2 (62.0-71.9) |
| 7 years | 66.8 (61.5-71.5) |
| 8 years | 66.2 (60.9-71.0) |
| 9 years | 66.2 (60.9-71.1) |
| 10 years | 66.2 (60.9-71.1) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.




Survival by diagnostic subgroup

Nearly all cases (98.1%) are in the diagnostic subgroup of neuroblastoma and ganglioneuroblastoma, for which 5-year relative survival was 67.8% (95% confidence interval = 62.6%-72.4%). Separate survival estimates for the subgroup neuroblastoma and ganglioneuroblastoma by sex, age group at diagnosis, tumour stage and year of diagnosis can be found in the Appendix (see Tables A.4a, A.4b, A.4c and A.4d, pages 44-47).

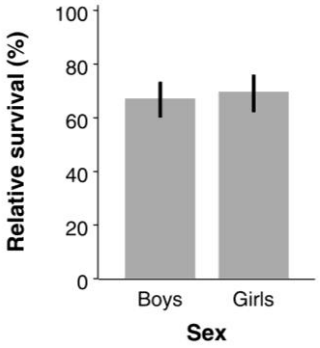
Survival by sex

Girls had slightly higher relative survival rates for childhood neuroblastoma compared to boys 5 years after diagnosis (69.7% and 67.3% respectively), but this difference was not statistically significant (Box 3.4b).

**Box 3.4b: Five-year relative survival for childhood neuroblastoma by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 196 | 67.3 (60.1-73.4) |
| Girls | 171 | 69.7 (62.0-76.2) |
| | | *p = 0.590* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



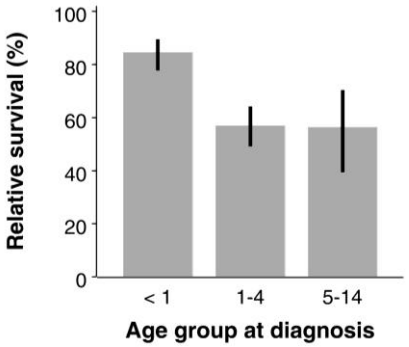

Survival by age group at diagnosis

Survival for neuroblastoma was significantly higher for infants compared to other children, with 5-year relative survival of 84.6% within the under 1 age group, 57.1% for children aged 1-4 years and 56.4% for children aged 5-14 years (Box 3.4c).

**Box 3.4c: Five-year relative survival for childhood neuroblastoma by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| < 1 year | 153 | 84.6 (77.7-89.6) |
| 1-4 years | 174 | 57.1 (49.1-64.2) |
| 5-14 years | 40 | 56.4 (39.4-70.4) |
| | | *p < 0.001* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. Age groups "5-9 years" and "10-14 years" have been combined due to the small number of cases in these age groups.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



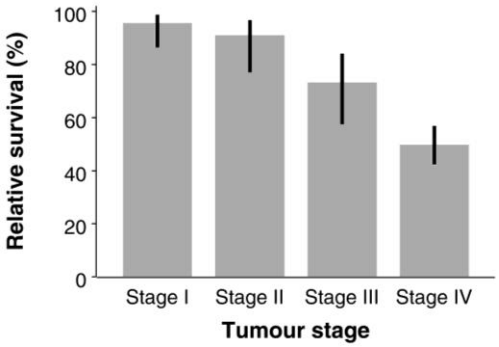

Survival by tumour stage

A clear gradient was observed in survival from childhood neuroblastoma by stage at diagnosis (Box 3.4d). Relative survival rates were significantly higher for stage I and II tumours (95.6% and 90.9% respectively after 5 years) but decreased sharply for stage III (73.3%) and IV tumours (49.8%).

**Box 3.4d: Five-year relative survival for childhood neuroblastoma by tumour stage[a], Australia, 1995-2004[b]**

| Tumour stage[a] | Number of cases[c,b] | Five-year relative survival (95% CI)[e] |
|---|---|---|
| Stage I | 68 | 95.6 (86.5-98.8) |
| Stage II | 44 | 90.9 (77.0-96.8) |
| Stage III | 46 | 73.3 (57.4-84.1) |
| Stage IV | 188 | 49.8 (42.3-56.9) |
| | | *p < 0.001* |

Notes: a. Staged using the International Neuroblastoma Staging System.[26]
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. The initial number of eligible cases in the cohort.
d. Stage was unknown for 21 cases (5.7%).
e. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



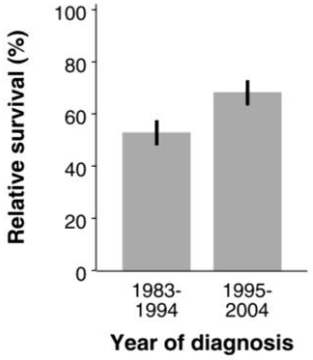
Survival by year of diagnosis

A highly significant improvement in survival for children with neuroblastoma was observed between 1983-1994 and 1995-2004, with 5-year relative survival increasing from 52.9% to 68.4% over this period (Box 3.4e).

**Box 3.4e: Five-year relative survival for childhood neuroblastoma by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 410 | 52.9 (47.9-57.6) |
| 1995-2004 | 367 | 68.4 (63.3-73.0) |
| | | *p < 0.001* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Five-year survival for children with neuroblastoma varied from 64% in Great Britain to 73% in both the United States and France, compared to 68% in Australia (Box 3.4f).

**Box 3.4f: Five-year survival for childhood neuroblastoma in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 367 | Relative survival – cohort method | 68 (63-73) |
| France[13] | 1995-1999 | 145 | Observed survival – cohort method | 73 (66-80) |
| Great Britain[14] | 2001-2005 | 473 | Observed survival – cohort method | 64 (59-68) |
| United States[15] | 1999-2006 | 1,222 | Relative survival – cohort method | 73 (70-75) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.5 Retinoblastoma*

Background

Retinoblastoma is a cancer that forms in the retina, the light-sensitive tissue at the back of the eye. It almost always develops before the age of five.[27] Tumours may occur in one eye (known as unilateral retinoblastoma) or both eyes (bilateral retinoblastoma).[9,11] Symptoms generally include squinting or eye swelling and redness, but in many children the first sign noted is a visible whiteness of the pupil.[7,9,27,28]

All retinoblastoma is due to a genetic error in the retinal tissue.[28] The error may occur only in the retina (sporadic retinoblastoma) or may be present in all cells in the body (hereditary retinoblastoma). If the genetic error is just in the retina of one eye, only that eye will be affected. However, if it is hereditary, retinoblastoma may develop in either one or both eyes. Children with hereditary retinoblastoma are also at risk of developing other cancers elsewhere in the body, and may pass the genetic error on to their children. Genetic testing can be carried out to determine whether a child has sporadic or hereditary retinoblastoma.

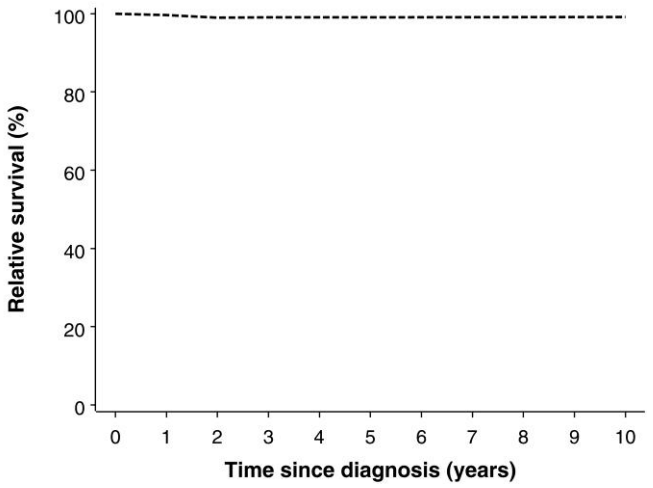
Survival by time since diagnosis

The survival of children with retinoblastoma is similar to children in the general population, with relative survival remaining around 99% up to 10 years after diagnosis (Box 3.5a).

**Box 3.5a: Relative survival by time since diagnosis for childhood retinoblastoma (n=159)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 99.6 (95.9-100.0) |
| 2 years | 99.0 (95.3-100.0) |
| 3 years | 99.0 (95.4-100.0) |
| 4 years | 99.1 (95.4-100.0) |
| 5 years | 99.1 (95.4-100.0) |
| 6 years | 99.1 (95.4-100.0) |
| 7 years | 99.1 (95.4-100.0) |
| 8 years | 99.1 (95.4-100.0) |
| 9 years | 99.1 (95.4-100.0) |
| 10 years | 99.1 (95.4-100.0) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.




Survival by diagnostic subgroup

No diagnostic subgroups are defined for retinoblastoma.[10]

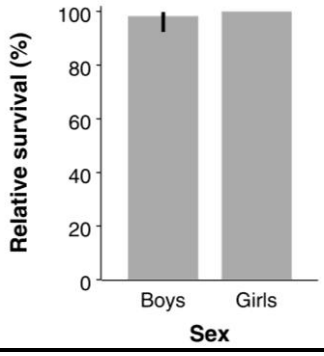
Survival by sex

There were no deaths within 5 years of diagnosis (i.e. 100% survival) among girls with retinoblastoma in Australia during 1995-2004, while 5-year relative survival was also very high among boys (98.3% - see Box 3.5b).

**Box 3.5b: Five-year relative survival for childhood retinoblastoma by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c,d] |
|---|---|---|
| Boys | 97 | 98.3 (92.3-99.8) |
| Girls | 62 | 100.0 n.a. |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).
d. n.a. = not applicable. Confidence interval (and corresponding p-value) could not be calculated because no deaths were recorded.



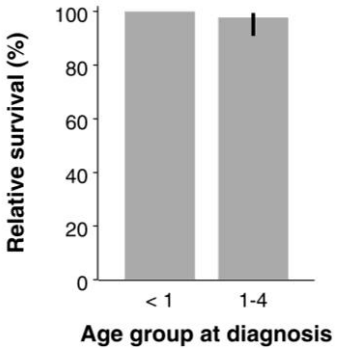
Survival by age group at diagnosis

Infants (under 1 year of age) diagnosed with retinoblastoma during 1995-2004 had 100% survival within 5 years of diagnosis compared to 97.7% for children aged 1-4 years (Box 3.5c). There were an insufficient number of cases to calculate survival for children aged 5 years and over.

**Box 3.5c: Five-year relative survival for childhood retinoblastoma by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis[b] | Number of cases[c] | Five-year relative survival (95% CI)[d,e] |
|---|---|---|
| < 1 year | 69 | 100.0 n.a. |
| 1-4 years | 84 | 97.7 (90.9-99.5) |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. Survival estimates could not be calculated within the 5-9 and 10-14 age groups due to the small number of cases diagnosed.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).
e. n.a. = not applicable. Confidence interval (and corresponding p-value) could not be calculated because no deaths were recorded.



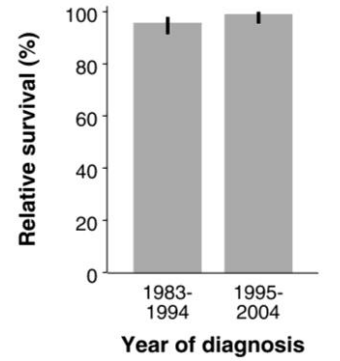

Survival by year of diagnosis

Five-year relative survival for retinoblastoma increased from 95.8% for children diagnosed during 1983-1994 to 99.1% for children diagnosed during 1995-2004, although the difference was not statistically significant (Box 3.5d).

**Box 3.5d: Five-year relative survival for childhood retinoblastoma by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 172 | 95.8 (91.3-98.1) |
| 1995-2004 | 159 | 99.1 (95.4-100.0) |
| | | *p = 0.088* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Survival for children with retinoblastoma was consistently high (98% or better after 5 years) in each of the countries for which recent estimates were available (Box 3.5e).

**Box 3.5e: Five-year survival for childhood retinoblastoma in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 159 | Relative survival – cohort method | 99 (95-100) |
| France[13] | 1995-1999 | 53 | Observed survival – cohort method | 100 n.a. |
| Great Britain[14] | 2001-2005 | 205 | Observed survival – cohort method | 99 (96-100) |
| United States[15] | 1999-2006 | 523 | Relative survival – cohort method | 98 (95-99) |

Abbreviation: n.a. = not applicable (confidence interval could not be calculated because no deaths were recorded).
Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.6 Renal tumours*

Background

Renal tumours in children often originate from abnormalities during development of the kidneys prior to birth. They tend to grow rapidly and about three quarters of all cases are diagnosed before the age of five. Children with renal tumours usually have cancer in one kidney, but occasionally tumours can develop in both kidneys.[9,29]

The most common symptom is a lump in the abdomen in an otherwise well child. Some children also have abdominal pain, blood in the urine, hypertension (high blood pressure) or fever.[7,9,29,30]

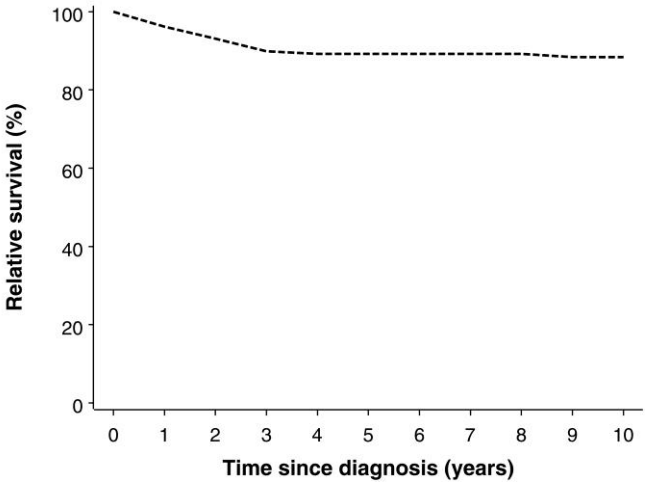
Survival by time since diagnosis

Relative survival for children with renal tumours decreased from 96.1% in the first year following diagnosis to 89.2% after 5 years, but then remained quite stable, with 10-year relative survival estimated at 88.4% (Box 3.6a).

**Box 3.6a: Relative survival by time since diagnosis for childhood renal tumours (n=328)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 96.1 (93.4-97.8) |
| 2 years | 93.1 (89.7-95.4) |
| 3 years | 89.9 (86.0-92.8) |
| 4 years | 89.2 (85.2-92.2) |
| 5 years | 89.2 (85.2-92.2) |
| 6 years | 89.2 (85.2-92.2) |
| 7 years | 89.2 (85.2-92.2) |
| 8 years | 89.2 (85.2-92.2) |
| 9 years | 88.4 (84.0-91.7) |
| 10 years | 88.4 (84.0-91.7) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.




Survival by diagnostic subgroup

Nephroblastoma (also known as Wilms' tumour) is by far the most frequent type of renal tumour occurring among children.[30] Almost all (97.0%) childhood renal tumours diagnosed in Australia during 1995-2004 were in the subgroup nephroblastoma and other nonepithelial renal tumours, with a corresponding 5-year relative survival rate of 88.9% (95% confidence interval = 84.8%-92.0%). Separate survival estimates for nephroblastoma and other nonepithelial renal tumours by sex, age group at diagnosis, tumour stage and year of diagnosis can be found in the Appendix (see Tables A.4a, A.4b, A.4c and A.4d, pages 44-47).

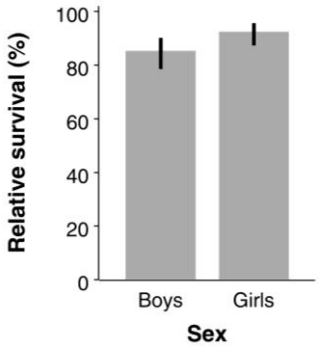
Survival by sex

Girls appeared to have somewhat higher survival rates for childhood renal tumours compared to boys (5-year relative survival of 92.5% and 85.4% respectively), but the difference by sex was not statistically significant (Box 3.6b).

**Box 3.6b: Five-year relative survival for childhood renal tumours by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 153 | 85.4 (78.5-90.2) |
| Girls | 175 | 92.5 (87.4-95.6) |
| | | *p = 0.068* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



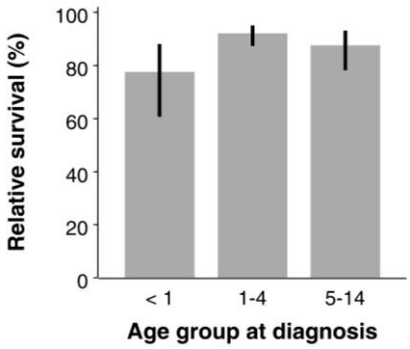

Survival by age group at diagnosis

Five-year relative survival for renal tumours was considerably lower in the under 1 age group (77.7%) than for children aged 1-4 years (92.1%). However, the differences in survival by age group were not statistically significant overall (Box 3.6c).

**Box 3.6c: Five-year relative survival for childhood renal tumours by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| < 1 year | 40 | 77.7 (60.7-88.1) |
| 1-4 years | 206 | 92.1 (87.4-95.1) |
| 5-14 years | 82 | 87.6 (78.2-93.2) |
| | | *p = 0.078* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. Age groups "5-9 years" and "10-14 years" have been combined due to the small number of cases in these age groups.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



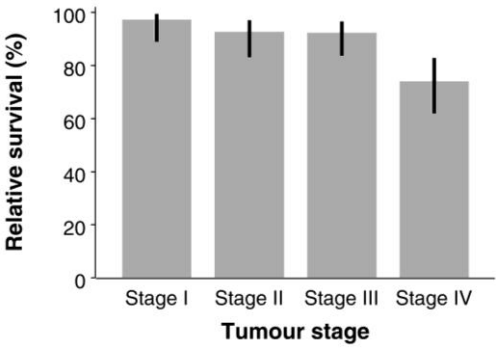

Survival by tumour stage

Significant variation was recorded in survival for childhood renal tumours by stage at diagnosis (Box 3.6d). Five-year relative survival rates varied from 97.3% for stage I tumours to 74.0% for stage IV tumours.

**Box 3.6d: Five-year relative survival for childhood renal tumours by tumour stage[a], Australia, 1995-2004[b]**

| Tumour stage[a] | Number of cases[c,d] | Five-year relative survival (95% CI)[e] |
|---|---|---|
| Stage I | 70 | 97.3 (89.0-99.5) |
| Stage II | 76 | 92.7 (83.1-97.0) |
| Stage III | 78 | 92.4 (83.7-96.6) |
| Stage IV | 70 | 74.0 (61.9-82.8) |
| | | *p < 0.001* |

Notes: a. Staged using the Third National Wilms' Tumor Study.[31]
a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. Stage was unknown for 34 cases (10.4%).
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



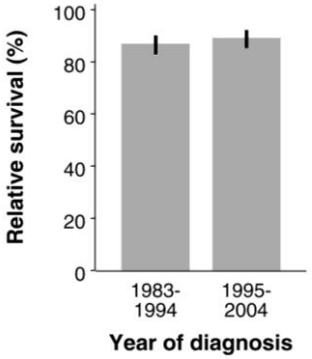
Survival by year of diagnosis

There was no significant improvement in survival for children diagnosed with renal tumours during 1983-1994 compared to 1995-2004, with 5-year relative survival estimates of 86.9% and 89.2% respectively (Box 3.6e).

**Box 3.6e: Five-year relative survival for childhood renal tumours by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 339 | 86.9 (82.8-90.1) |
| 1995-2004 | 328 | 89.2 (85.2-92.2) |
| | | *p = 0.393* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Of the countries for which recent estimates were available, 5-year survival following diagnosis of a renal tumour during childhood varied from 84% in Great Britain to 89% in both Australia and France (Box 3.6f).

**Box 3.6f: Five-year survival for childhood renal tumours in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 328 | Relative survival – cohort method | 89 (85-92) |
| France[13] | 1995-1999 | 125 | Observed survival – cohort method | 89 (83-94) |
| Great Britain[14] | 2001-2005 | 444 | Observed survival – cohort method | 84 (81-88) |
| United States[15] | 1999-2006 | 938 | Relative survival – cohort method | 88 (86-90) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.7 Hepatic tumours*

Background

Hepatic tumours are cancers of the liver. It is the least common diagnostic group, constituting only 1.5% of all childhood cancers diagnosed in Australia during 1995-2004. The two main subgroups are hepatoblastoma (83.9%) and hepatic carcinomas (16.1%). Hepatoblastoma usually occur in younger children (under four years of age), while hepatic carcinomas primarily affect children over the age of ten.[7,32,33]

Children with hepatic tumours typically have a painless mass in their abdomen, although some experience abdominal pain, constipation, weight loss or nausea,[32] particularly if the tumour is more advanced.[7]

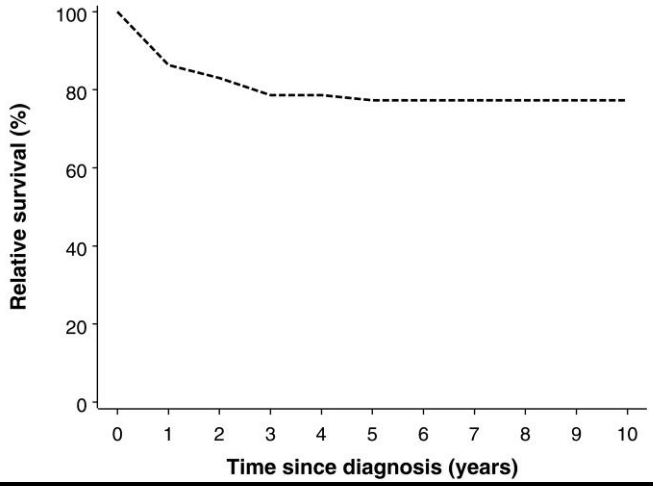
Survival by time since diagnosis

Most of the mortality associated with childhood hepatic tumours occurred in the first 2 to 3 years following diagnosis (Box 3.7a). Relative survival was 86.3% by the end of the first year before a further decrease to 77.3% after 5 years, but from then on the survival rate remained stable.

**Box 3.7a: Relative survival by time since diagnosis for childhood hepatic tumours (n=93)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 86.3 (77.4-91.9) |
| 2 years | 83.0 (73.7-89.3) |
| 3 years | 78.6 (68.7-85.7) |
| 4 years | 78.6 (68.7-85.8) |
| 5 years | 77.3 (67.1-84.7) |
| 6 years | 77.3 (67.1-84.7) |
| 7 years | 77.3 (67.1-84.7) |
| 8 years | 77.3 (67.1-84.7) |
| 9 years | 77.3 (67.2-84.7) |
| 10 years | 77.3 (67.2-84.8) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.




Survival by diagnostic subgroup

Five-year relative survival was 87.1% (95% confidence interval = 77.1%-93.1%) for children diagnosed with hepatoblastoma. There were an insufficient number of cases to calculate separate survival estimates for hepatic carcinomas.

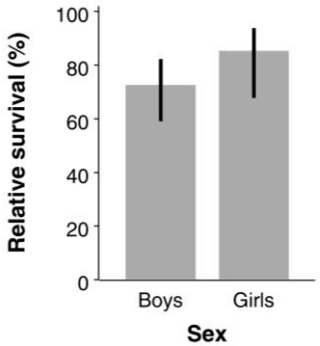
Survival by sex

Survival for girls with hepatic tumours appeared to be higher than survival for boys, estimated at 85.4% and 72.6% respectively after 5 years (Box 3.7b). Similar variation was observed for the diagnostic subgroup of hepatoblastoma, with 5-year relative survival of 96.9% for girls compared to 81.5% for boys (Appendix Table A.4a, page 44). However, due to the small number of cases involved, neither of these differences in survival by sex were statistically significant.

**Box 3.7b: Five-year relative survival for childhood hepatic tumours by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 59 | 72.6 (59.1-82.3) |
| Girls | 34 | 85.4 (67.8-93.9) |
| | | *p = 0.288* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



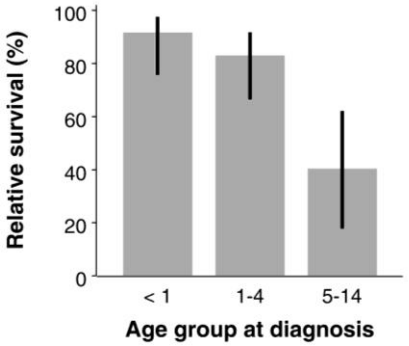
Survival by age group at diagnosis

A significant decrease in survival was observed for childhood hepatic tumours as age at diagnosis increased. Five-year relative survival was 91.6% for children aged under 1 compared to 83.0% for those in the 1-4 age group and 40.4% for children aged 5-14 (Box 3.7c). This outcome is influenced by differences in the age distribution for the diagnostic subgroups. When the analysis was limited to children under 5 years old who were diagnosed with hepatoblastoma, there was no difference between the under 1 and 1-4 age groups, with 5-year relative survival of 91.6% and 91.8% respectively (see Appendix Table A.4b, page 45).

**Box 3.7c: Five-year relative survival for childhood hepatic tumours by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| < 1 year | 34 | 91.6 (75.6-97.6) |
| 1-4 years | 39 | 83.0 (66.4-91.6) |
| 5-14 years | 20 | 40.4 (17.8-62.1) |
| | | *p = 0.002* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. Age groups "5-9 years" and "10-14 years" have been combined due to the small number of cases in these age groups.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



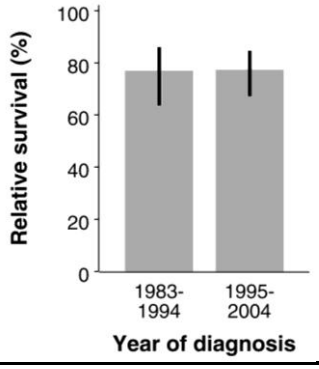

Survival by year of diagnosis

Similar results were obtained for children diagnosed with hepatic tumours during 1983-1994 and 1995-2004, with 5-year relative survival estimated at 77.0% and 77.3% respectively (Box 3.7d). Despite a small increase, there was no significant improvement over time for children with hepatoblastoma, for whom 5-year relative survival was 82.6% during 1983-1994 compared to 87.1% during 1995-2004 (Appendix Table A.4d, page 47).

**Box 3.7d: Five-year relative survival for childhood hepatic tumours by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 56 | 77.0 (63.6-86.1) |
| 1995-2004 | 93 | 77.3 (67.1-84.7) |
| | | *p = 0.669* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Five-year survival for children with hepatic tumours in Australia (77%) compared favourably with other developed countries for which estimates were available (Box 3.7e).

**Box 3.7e: Five-year survival for childhood hepatic tumours in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 93 | Relative survival – cohort method | 77 (67-85) |
| France[13] | 1995-1999 | 19 | Observed survival – cohort method | 74 (54-94) |
| Great Britain[14] | 2001-2005 | 90 | Observed survival – cohort method | 66 (55-74) |
| United States[15] | 1999-2006 | 308 | Relative survival – cohort method | 66 (60-72) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.8 Malignant bone tumours*

Background

Childhood malignant bone tumours can occur at any age but are usually diagnosed around the age of puberty. The two main subgroups are Ewing tumours and related bone sarcomas and osteosarcomas, which occur in almost equal numbers (responsible for 47.1% and 46.4% of all childhood malignant bone tumours diagnosed in Australia during 1995-2004, respectively).

Osteosarcomas often originate at the ends of the long bones in the arms and legs (particularly around the knee) where new bone tissue forms as a child grows. Ewing sarcomas are more likely to develop at a range of sites, including the legs, pelvis, arms, ribs or spine.[7,9,34]

The most common symptom of malignant bone tumours is localised pain at the tumour site that may persist at night. This can be accompanied by tenderness, swelling and fever in more advanced cases.[7,9]

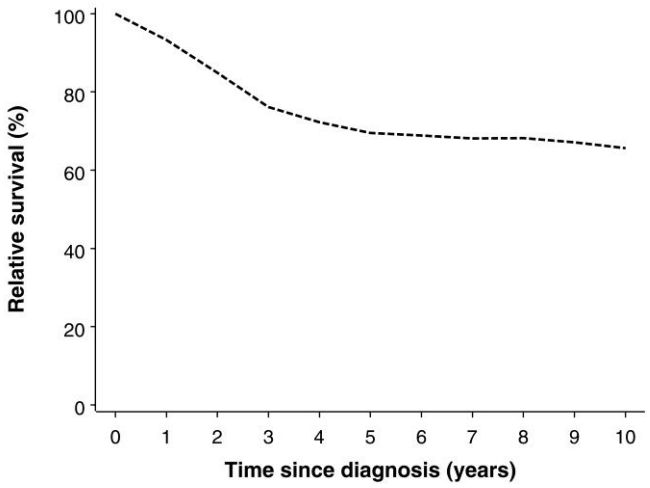
Survival by time since diagnosis

By the end of the first year after diagnosis, relative survival for childhood malignant bone tumours was 93.3%. Survival continued to decrease steadily to 69.5% after 5 years before a further small decline to 65.6% after 10 years (Box 3.8a).

**Box 3.8a: Relative survival by time since diagnosis for childhood malignant bone tumours (n=263)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 93.3 (89.5-95.7) |
| 2 years | 84.9 (80.0-88.8) |
| 3 years | 76.2 (70.4-80.9) |
| 4 years | 72.3 (66.2-77.5) |
| 5 years | 69.5 (63.2-75.0) |
| 6 years | 68.9 (62.5-74.5) |
| 7 years | 68.2 (61.6-73.8) |
| 8 years | 68.2 (61.6-73.9) |
| 9 years | 67.1 (60.2-73.0) |
| 10 years | 65.6 (58.3-72.0) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.



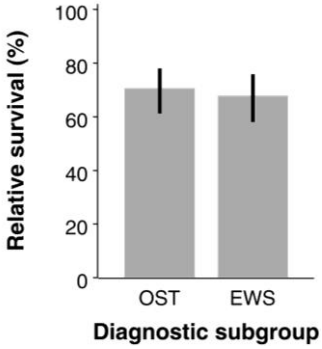

Survival by diagnostic subgroup

Children diagnosed with osteosarcomas had a slightly higher rate of 5-year relative survival compared to those with Ewing tumours, estimated at 70.6% and 67.8% respectively, but the difference in survival between these diagnostic subgroups was not statistically significant (Box 3.8b).

**Box 3.8b: Five-year relative survival for childhood malignant bone tumours by diagnostic subgroup, Australia, 1995-2004[a]**

| Diagnostic subgroup[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| OST | 122 | 70.6 (61.2-78.1) |
| EWS | 124 | 67.8 (58.0-75.8) |
| | | *p = 0.570* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. OST = osteosarcomas; EWS = Ewing tumours and related bone sarcomas.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



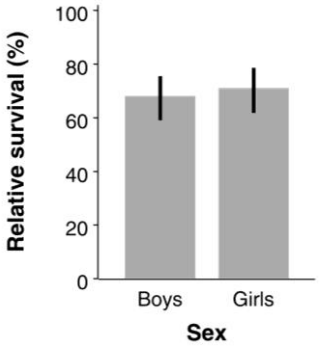

Survival by sex

Although 5-year relative survival for childhood malignant bone tumours was slightly higher for girls than boys (71.1% and 68.1% respectively), the difference was not statistically significant (Box 3.8c). There were also no significant differences in survival by sex for either osteosarcomas or Ewing tumours (see Appendix Table A.4a, page 44).

**Box 3.8c: Five-year relative survival for childhood malignant bone tumours by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 135 | 68.1 (59.0-75.6) |
| Girls | 128 | 71.1 (61.8-78.6) |
| | | *p = 0.502* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



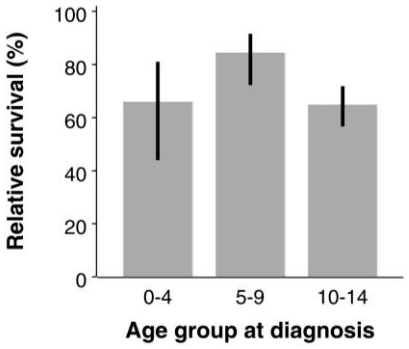

Survival by age group at diagnosis

Five-year relative survival for malignant bone tumours was higher for children aged 5-9 years old at the time of diagnosis (84.5%) compared to those in either the 0-4 or 10-14 age groups (66.0% and 64.9% respectively - see Box 3.8d). Similar patterns were evident for both osteosarcomas and Ewing tumours (Appendix Table A.4b, page 45), but the differences in survival by age group were not statistically significant for either of these diagnostic subgroups.

**Box 3.8d: Five-year relative survival for childhood malignant bone tumours by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| 0-4 years | 27 | 66.0 (44.0-81.0) |
| 5-9 years | 62 | 84.5 (72.3-91.6) |
| 10-14 years | 174 | 64.9 (56.7-71.9) |
| | | *p = 0.020* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. Age groups "< 1 year" and "1-4 years" have been combined due to the small number of cases in these age groups.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



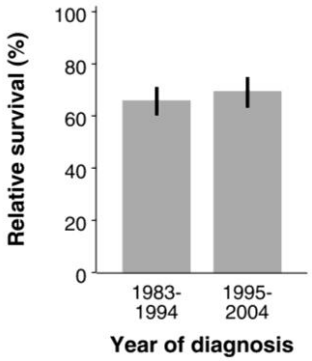
Survival by year of diagnosis

A small, non-significant improvement was observed in survival for children with malignant bone tumours between 1983-1994 and 1995-2004, with 5-year relative survival estimates of 66.0% and 69.5% recorded over the two time periods (Box 3.8e). No significant change in survival over time was recorded for the diagnostic subgroups of either osteosarcomas or Ewing tumours (Appendix Table A.4d, page 47).

**Box 3.8e: Five-year relative survival for childhood malignant bone tumours by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 287 | 66.0 (60.2-71.1) |
| 1995-2004 | 263 | 69.5 (63.2-75.0) |
| | | *p = 0.322* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

As shown in Box 3.8f, 5-year survival for children with malignant bone tumours varied from 61% in Great Britain to 77% in France, compared to 70% for Australia.

**Box 3.8f: Five-year survival for childhood malignant bone tumours in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 263 | Relative survival – cohort method | 70 (63-75) |
| France[13] | 1995-1999 | 104 | Observed survival – cohort method | 77 (69-85) |
| Great Britain[14] | 2001-2005 | 321 | Observed survival – cohort method | 61 (55-66) |
| United States[15] | 1999-2006 | 788 | Relative survival – cohort method | 71 (67-74) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.9 Soft tissue and other extraosseous sarcomas*

Background

Soft tissue sarcomas are a diverse group of cancers that develop in the soft tissues (such as muscles, fat, blood vessels, lymphatic vessels, nerves, ligaments and tendons) which connect, support or surround bones and organs.

The most common subgroup of childhood soft tissue sarcomas is rhabdomyosarcomas, which accounted for almost half (48.0%) of all soft tissue sarcomas diagnosed among Australian children during 1995-2004. These tumours usually occur before ten years of age and typically begin in muscles around the bones, especially in the head, neck and genitourinary tract.[9,35,36] In contrast, other types of soft tissue sarcomas (referred to collectively as non-rhabdomyosarcoma soft tissue sarcomas) tend to occur in the extremities of the body (i.e. arms and legs) and are more common among older children.[35-37]

Children with soft tissue sarcoma may develop a lump at the involved site. Other symptoms tend to be site-specific, such as headaches, sinusitis, persistent ear or nasal discharge or protrusion of the eyeball for head and neck tumours, or abnormal urine flow accompanying genitourinary tumours.[9,35-37]

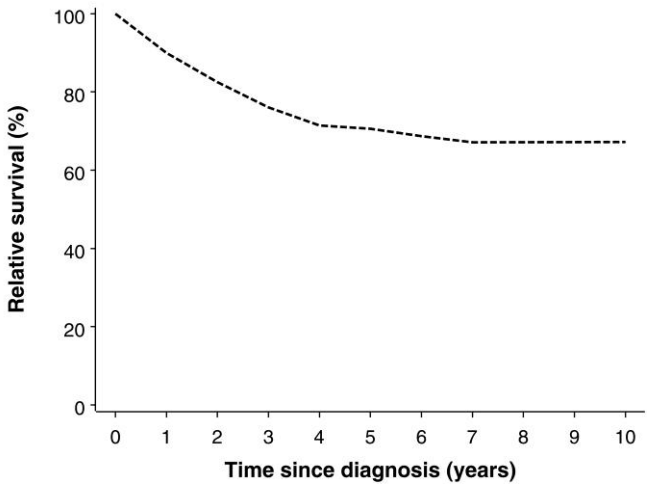
Survival by time since diagnosis

The relative rate of survival among children diagnosed with soft tissue sarcoma gradually decreased from 90.0% 1 year after diagnosis to 70.6% after 5 years, but then remained fairly stable, with 10-year relative survival estimated at 67.2% (Box 3.9a).

**Box 3.9a: Relative survival by time since diagnosis for childhood soft tissue sarcomas (n=333)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 90.0 (86.2-92.8) |
| 2 years | 82.5 (78.0-86.2) |
| 3 years | 76.0 (71.0-80.3) |
| 4 years | 71.4 (66.0-76.1) |
| 5 years | 70.6 (65.2-75.3) |
| 6 years | 68.7 (63.1-73.7) |
| 7 years | 67.1 (61.4-72.2) |
| 8 years | 67.1 (61.4-72.3) |
| 9 years | 67.2 (61.4-72.3) |
| 10 years | 67.2 (61.4-72.3) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.



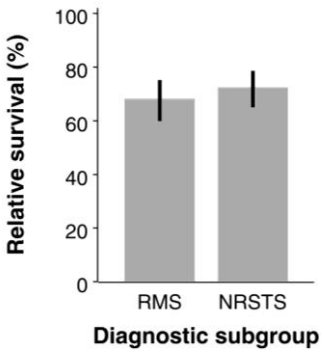

Survival by diagnostic subgroup

Five-year relative survival was somewhat lower for rhabdomyosarcomas (68.2%) compared to non-rhabdomyosarcoma soft tissue sarcomas (72.5%), but the difference was not statistically significant (Box 3.9b).

**Box 3.9b: Five-year relative survival for childhood soft tissue sarcomas by diagnostic subgroup, Australia, 1995-2004[a]**

| Diagnostic subgroup[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| RMS | 160 | 68.2 (59.9-75.2) |
| NRSTS[e] | 173 | 72.5 (65.0-78.6) |
| | | *p = 0.272* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. RMS = rhabdomyosarcomas; NRSTS = non-rhabdomyosarcoma soft tissue sarcomas.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).
e. NRSTS include the diagnostic subgroups fibrosarcomas and other fibrous neoplasms, Kaposi sarcomas, other specified soft tissue sarcomas and unspecified soft tissue sarcomas.



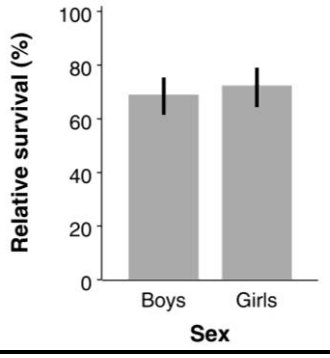

Survival by sex

Girls had slightly higher 5-year relative survival for childhood soft tissue sarcomas compared to boys (72.5% and 69.0% respectively), but this difference was not statistically significant (Box 3.9c). There was also no significant difference in survival by sex for either rhabdomyosarcomas or non-rhabdomyosarcoma soft tissue sarcomas (see Appendix Table A.4a, page 44).

**Box 3.9c: Five-year relative survival for childhood soft tissue sarcomas by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 182 | 69.0 (61.4-75.5) |
| Girls | 151 | 72.5 (64.3-79.1) |
| | | *p = 0.392* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



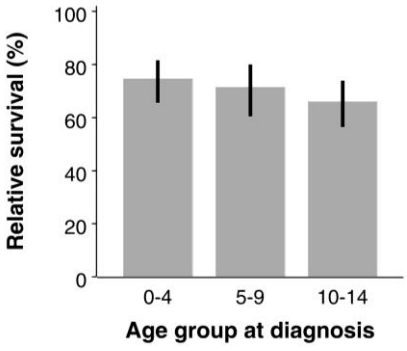

Survival by age group at diagnosis

No significant differences were found in relative survival by age at diagnosis for children with soft tissue sarcomas, although the estimates of 5-year survival were somewhat higher within the 0-4 (74.6%) and 5-9 (71.5%) age groups compared to children aged 10-14 years old (66.0% – see Box 3.9d). The variation by age was more pronounced within the diagnostic subgroup of rhabdomyosarcomas, with 5-year relative survival ranging from 75.4% for children aged 0-4 years to 43.8% in the 10-14 age group (Appendix Table A.4b, page 45).

**Box 3.9d: Five-year relative survival for childhood soft tissue sarcomas by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| 0-4 years | 123 | 74.6 (65.6-81.6) |
| 5-9 years | 90 | 71.5 (60.4-80.0) |
| 10-14 years | 120 | 66.0 (56.4-74.0) |
| | | *p = 0.318* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. Age groups "< 1 year" and "1-4 years" have been combined due to the small number of cases in the < 1 age group.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



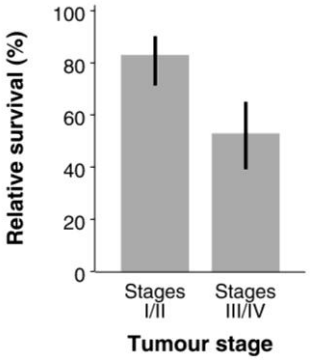
Survival by tumour stage

Staging of tumours for childhood soft tissue sarcomas was only available for the diagnostic subgroup of rhabdomyosarcomas. Survival varied significantly by stage (Box 3.9e), with 5-year relative survival rates higher for stage I and II tumours combined (83.0%) compared to stage III and IV tumours combined (53.0%).

**Box 3.9e: Five-year relative survival for childhood rhabdomyosarcomas by tumour stage[a], Australia, 1995-2004[b]**

| Tumour stage[a,c] | Number of cases[d,e] | Five-year relative survival (95% CI)[f] |
|---|---|---|
| Stages I/II | 68 | 83.0 (71.3-90.3) |
| Stages III/IV | 59 | 53.0 (39.1-65.0) |
| | | *p < 0.001* |

Notes: a. Staged using the Intergroup Rhabdomyosarcoma Study-I.[38]
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. Stages I and II have been combined as have stages III and IV due to the small number of cases in the individual stage categories.
d. The initial number of eligible cases in the cohort.
e. Stage was unknown for 33 cases (20.6%).
f. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



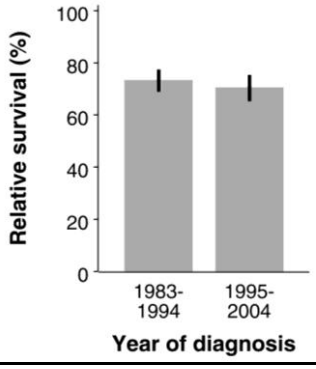

Survival by year of diagnosis

Five-year relative survival for childhood soft tissue sarcomas decreased slightly from 73.4% during 1983-1994 to 70.6% during 1995-2004, although the difference was not statistically significant (Box 3.9f). However, when the diagnostic subgroups were analysed separately, a significant decrease was observed for non-rhabdomyosarcoma soft tissue sarcomas, with 5-year relative survival declining from 84.5% to 72.5% over the interval 1983-1994 to 1995-2004 (see Appendix Table A.4d, page 47).

**Box 3.9f: Five-year relative survival for childhood soft tissue sarcomas by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 411 | 73.4 (68.8-77.4) |
| 1995-2004 | 333 | 70.6 (65.2-75.3) |
| | | *p = 0.608* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Children with soft tissue sarcomas in Australia had similar 5-year survival compared to children in the United States (71% and 72% respectively - see Box 3.9g).

**Box 3.9g: Five-year survival for childhood soft tissue sarcomas in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 333 | Relative survival – cohort method | 71 (65-75) |
| France[13] | 1995-1999 | 95 | Observed survival – cohort method | 66 (57-76) |
| Great Britain[14] | 2001-2005 | 513 | Observed survival – cohort method | 67 (63-71) |
| United States[15] | 1999-2006 | 1,285 | Relative survival – cohort method | 72 (69-75) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.10 Germ cell tumours, trophoblastic tumours and neoplasms of gonads*

Background

Germ cell tumours are comprised of a varied group of cancers that originate from cells that normally develop into gonads (testicles in boys and ovaries in girls). Hence, germ cell tumours usually affect the gonads. However, it is possible for germ cell tumours to occur in other parts of the body, particularly the pelvis, brain or chest.[7,9,39,40]

The most common diagnostic subgroup is malignant gonadal germ cell tumours, accounting for 42.2% of germ cell tumour cases in Australia over the period 1995-2004, followed by and intracranial and intra-gonadal germ cell tumours (28.3%) and malignant extracranial and extragonadal germ cell tumours (27.5%).

Signs and symptoms of germ cell tumours vary by site, and may include swelling in the buttocks, testicular enlargement in boys, an abdominal mass, bladder or bowel obstruction or respiratory problems.[7,9,39,40]

In accordance with the standard international classification for childhood cancers (ICCC-3),[10] non-malignant intracranial and intraspinal germ cell tumours have been included in the data presented for this diagnostic group. Of the 69 cases in that subgroup, 48 children (69.6%) had tumours that were malignant and 21 children (30.4%) had either benign tumours or tumours of uncertain behaviour.

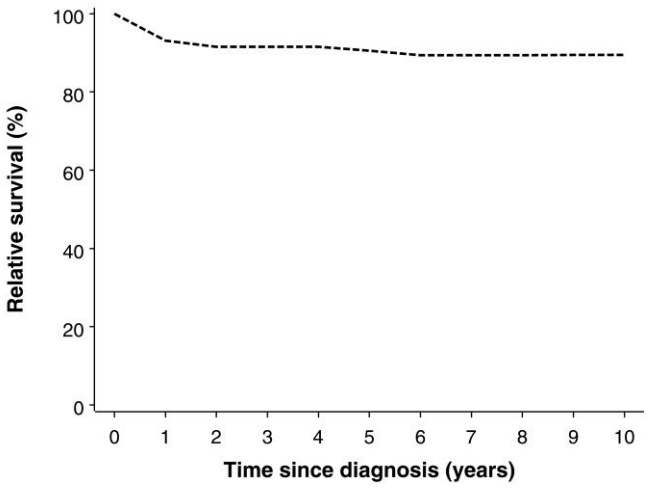
Survival by time since diagnosis

The relative rate of survival among children diagnosed with germ cell tumours decreased slightly from 93.1% 1 year after diagnosis to 90.5% after 5 years, with 10-year relative survival estimated at 89.5% (Box 3.10a).

**Box 3.10a: Relative survival by time since diagnosis for childhood germ cell tumours (n=244)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 93.1 (89.1-95.7) |
| 2 years | 91.5 (87.2-94.4) |
| 3 years | 91.5 (87.3-94.5) |
| 4 years | 91.6 (87.3-94.5) |
| 5 years | 90.5 (86.0-93.7) |
| 6 years | 89.4 (84.5-92.8) |
| 7 years | 89.4 (84.5-92.8) |
| 8 years | 89.4 (84.5-92.9) |
| 9 years | 89.4 (84.6-92.9) |
| 10 years | 89.5 (84.6-92.9) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.



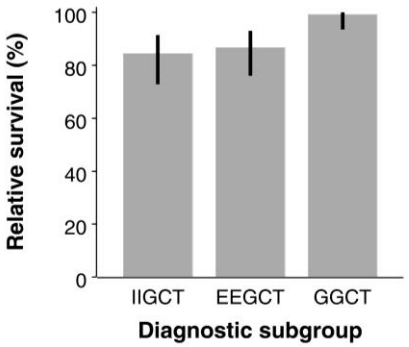

Survival by diagnostic subgroup

Children with malignant gonadal germ cell tumours had significantly higher 5-year relative survival (99.2%) compared to those diagnosed with either intracranial & intraspinal germ cell tumours (84.5%) or malignant extracranial & extragonadal germ cell tumours (86.8% - see Box 3.10b).

**Box 3.10b: Five-year relative survival for childhood germ cell tumours by diagnostic subgroup, Australia, 1995-2004[a]**

| Diagnostic subgroup[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| IIGCT | 69 | 84.5 (72.8-91.5) |
| EEGCT | 67 | 86.8 (76.0-93.0) |
| GGCT | 103 | 99.2 (93.5-100.0) |
| | | *p < 0.001* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. IIGCT = intracranial & intraspinal germ cell tumours; EEGCT = malignant extracranial & extragonadal germ cell tumours; GGCT = malignant gonadal germ cell tumours.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



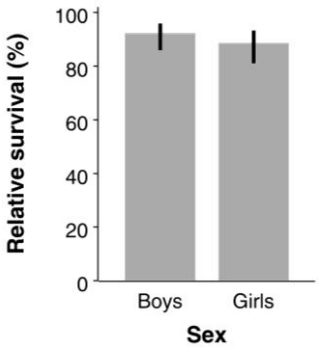

Survival by sex

Survival following a diagnosis of childhood germ cell tumours was slightly higher for boys than for girls, with 5-year relative survival estimated at 92.3% and 88.6% respectively, but the difference was not statistically significant (Box 3.10c). There were also no significant differences in survival by sex for any of the germ cell tumour diagnostic subgroups (see Appendix Table A.4a, page 44).

**Box 3.10c: Five-year relative survival for childhood germ cell tumours by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 126 | 92.3 (86.0-95.9) |
| Girls | 118 | 88.6 (81.0-93.3) |
| | | *p = 0.495* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



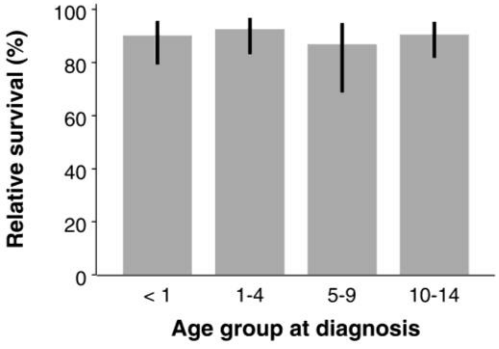

Survival by age group at diagnosis

Only minor variations were observed in survival for childhood germ cell tumours by age at diagnosis, with 5-year relative survival close to 90% within each of the age groups (Box 3.10d). Survival was also similar irrespective of age at diagnosis within each of the diagnostic subgroups for germ cell tumours (Appendix Table A.4b, page 45).

**Box 3.10d: Five-year relative survival for childhood germ cell tumours by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| < 1 year | 59 | 90.2 (79.2-95.7) |
| 1-4 years | 67 | 92.6 (83.1-96.9) |
| 5-9 years | 32 | 86.9 (68.7-94.9) |
| 10-14 years | 86 | 90.6 (81.7-95.3) |
| | | *p = 0.914* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



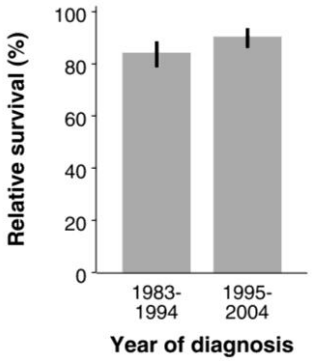
Survival by year of diagnosis

There was some evidence of an improvement in 5-year relative survival between 1983-1994 (84.4%) and 1995-2004 (90.5%) for children with germ cell tumours (Box 3.10e). A corresponding pattern over time was recorded in each of the germ cell tumour diagnostic subgroups (Appendix Table A.4d, page 47). However, none of these increases in survival were statistically significant.

**Box 3.10e: Five-year relative survival for childhood germ cell tumours by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 208 | 84.4 (78.6-88.7) |
| 1995-2004 | 244 | 90.5 (86.0-93.7) |
| | | *p = 0.081* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Survival for children diagnosed with germ cell tumours in Australia (91% after 5 years) was similar to the outcomes reported for Great Britain (92%) and the United States (89% - see Box 3.10f).

**Box 3.10f: Five-year survival for childhood germ cell tumours in selected countries[a]**

| Country | Years | Number of cases | Method | 5-year survival (%) (95% CI)[b,c] |
|---|---|---|---|---|
| Australia | 1995-2004 | 244 | Relative survival – cohort method | 91 (86-94) |
| France[13] | 1995-1999 | 62 | Observed survival – cohort method | 86 (77-94) |
| Great Britain[14] | 2001-2005 | 245 | Observed survival – cohort method | 92 (88-95) |
| United States[15] | 1999-2006 | 700 | Relative survival – cohort method | 89 (86-91) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. 95% CI = 95% confidence interval.
c. Survival estimates have been rounded to the nearest integer.

## *3.11 Other malignant epithelial neoplasms and melanomas*

Background

*Other malignant epithelial neoplasms*: Epithelial cells form the outer layer of the skin and line the internal cavities of the body. Most glands are also composed of epithelial cells. Tumours that originate in epithelial cells are called carcinomas. Although many of the types of cancers found in adults are classified as carcinomas, they are relatively rare among children, especially within the younger age groups.[9]

Thyroid carcinomas form one of the more common subgroups,[11] accounting for 17.3% of all cases within this diagnostic group in Australia during 1995-2004. Carcinomas of the thyroid are seldom accompanied by symptoms other than a painless mass in the neck.[7]

*Melanomas*: This is a type of skin cancer that originates in melanocytes, the cells which produce the pigment melanin that gives colour to a person's skin, hair, and eyes. Melanomas constituted over half (54.0%) of all new cases within this diagnostic group during 1995-2004.

Diagnosis of melanoma usually follows the discovery of a suspicious lesion on the skin, particularly when there has been itching, bleeding or a sudden change in size or colour.[7,41,42] The much higher incidence of melanoma in all age groups, including childhood, reported in Australia compared to the rest of the world is likely to be due to the high exposure to ultra-violet (UV) radiation from the sun experienced in most of the country, in combination with a large proportion of the population having skin that is more susceptible to sun damage.[11]

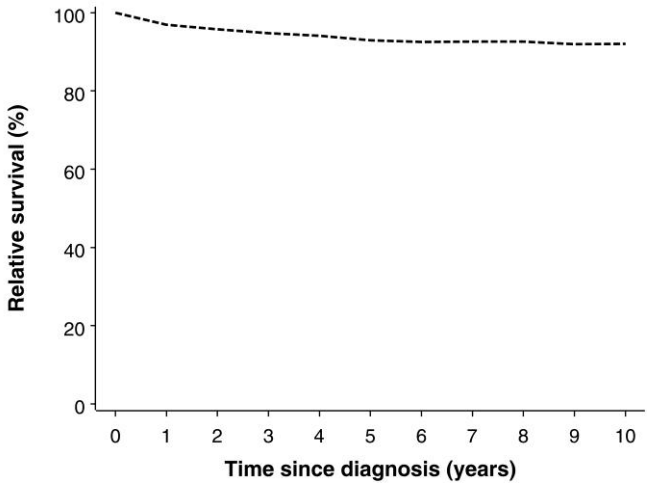
Survival by time since diagnosis

The relative rate of survival among children diagnosed with other malignant epithelial neoplasms and melanomas decreased slightly from 96.9% in the first year after diagnosis to 93.0% after 5 years, but then remained fairly stable, with 10-year relative survival estimated at 92.0% (Box 3.11a).

**Box 3.11a: Relative survival by time since diagnosis for other childhood malignant epithelial neoplasms and melanomas (n=324)[a], Australia, 1995-2004[b]**

| Time since diagnosis | Relative survival (95% CI)[c] |
|---|---|
| 1 year | 96.9 (94.4-98.3) |
| 2 years | 95.7 (92.9-97.5) |
| 3 years | 94.8 (91.7-96.8) |
| 4 years | 94.1 (90.9-96.2) |
| 5 years | 93.0 (89.4-95.4) |
| 6 years | 92.6 (88.9-95.1) |
| 7 years | 92.6 (88.9-95.1) |
| 8 years | 92.6 (89.0-95.2) |
| 9 years | 92.0 (87.9-94.7) |
| 10 years | 92.0 (88.0-94.8) |

Notes: a. The initial number of eligible cases in the cohort.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. 95% CI = 95% confidence interval.



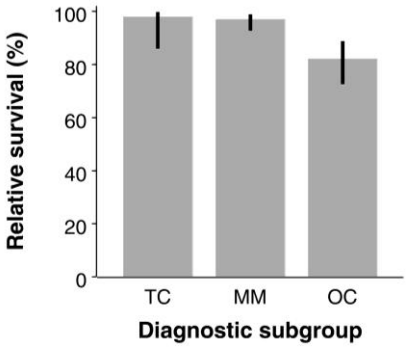

Survival by diagnostic subgroup

Five-year relative survival was significantly higher for children with either a thyroid carcinoma (97.9%) or melanoma (97.0%) compared to those diagnosed with other carcinomas (82.2% - see Box 3.11b).

**Box 3.11b: Five-year relative survival for other childhood malignant epithelial neoplasms and melanomas by diagnostic subgroup, Australia, 1995-2004[a]**

| Diagnostic subgroup[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| TC | 56 | 97.9 (85.9-99.8) |
| MM | 175 | 97.0 (92.8-98.8) |
| OC[e] | 93 | 82.2 (72.6-88.8) |
| | | *p < 0.001* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. TC = thyroid carcinomas; MM = malignant melanomas; OC = other carcinomas.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).
e. "Other carcinomas" are a combination of the diagnostic sub-groups adrenocortical carcinomas, nasopharyngeal carcinomas, skin carcinomas, and other and unspecified carcinomas.



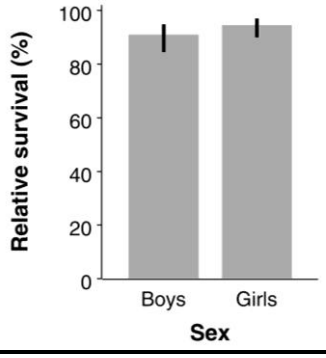

Survival by sex

Although 5-year relative survival for other childhood malignant epithelial neoplasms and melanomas was slightly higher for girls than boys (94.5% and 91.0% respectively), the difference was not statistically significant (Box 3.11c). No significant differences in survival by sex were found for any of the diagnostic subgroups for other malignant epithelial neoplasms and melanomas (see Appendix Table A.4a, page 44).

**Box 3.11c: Five-year relative survival for other childhood malignant epithelial neoplasms and melanomas by sex, Australia, 1995-2004[a]**

| Sex | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| Boys | 141 | 91.0 (84.5-94.9) |
| Girls | 183 | 94.5 (90.0-97.1) |
| | | *p = 0.267* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



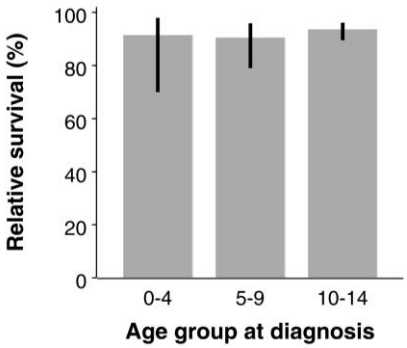

Survival by age group at diagnosis

There were no significant differences in 5-year relative survival for other malignant epithelial neoplasms and melanomas by age at diagnosis, with results varying from 90.6% among children aged 5-9 years to 93.6% for those in the 10-14 age group (Box 3.11d). Survival estimates by age within the diagnostic subgroups could only be calculated for older children, and are shown in Appendix Table A.4b, page 45.

**Box 3.11d: Five-year relative survival for other childhood malignant epithelial neoplasms and melanomas by age group at diagnosis, Australia, 1995-2004[a]**

| Age group at diagnosis[b] | Number of cases[c] | Five-year relative survival (95% CI)[d] |
|---|---|---|
| 0-4 years | 24 | 91.5 (69.9-98.0) |
| 5-9 years | 54 | 90.6 (79.0-96.0) |
| 10-14 years | 246 | 93.6 (89.5-96.2) |
| | | *p = 0.607* |

Notes: a. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
b. Age groups "< 1 year" and "1-4 years" have been combined due to the small number of cases in these age groups.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).



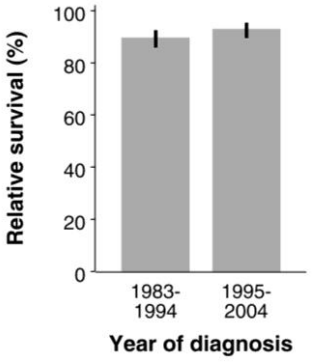
Survival by year of diagnosis

There appeared to be a small increase in survival for children with other malignant epithelial neoplasms and melanomas between 1983-1994 and 1995-2004, with 5-year relative survival estimates of 89.6% and 93.0% recorded over the two time periods (Box 3.11e). However, the difference was not statistically significant. Similar, small improvements also occurred within the various diagnostic subgroups for other malignant epithelial neoplasms and melanomas, but again these changes over time were not significant (see Appendix Table A.4d, page 47).

**Box 3.11e: Five-year relative survival for other childhood malignant epithelial neoplasms and melanomas by year of diagnosis, Australia, 1983-2004[a]**

| Year of diagnosis | Number of cases[b] | Five-year relative survival (95% CI)[c] |
|---|---|---|
| 1983-1994 | 332 | 89.6 (85.8-92.5) |
| 1995-2004 | 324 | 93.0 (89.4-95.4) |
| | | *p = 0.128* |

Notes: a. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
b. The initial number of eligible cases in the cohort.
c. 95% CI = 95% confidence interval (shown as vertical black lines on bar graph).




International comparisons

Five-year survival for children diagnosed with other malignant epithelial neoplasms and melanomas was as good or better in Australia compared to the other countries for which estimates were available (Box 3.11f). The result for Australia was in part due to the proportionally high incidence of childhood melanoma combined with a particularly high survival rate within this subgroup - survival for melanoma was 97% after 5 years among children in Australia (Box 3.11b), compared to 94% in the United States[15].

**Box 3.11f: Five-year survival for childhood malignant epithelial neoplasms and melanomas in selected countries[a,b]**

| Country[c] | Years | Number of cases | Method | 5-year survival (%) (95% CI)[c,d] |
|---|---|---|---|---|
| Australia | 1995-2004 | 324 | Relative survival – cohort method | 93 (90-95) |
| France[13] | 1995-1999 | 61 | Observed survival – cohort method | 88 (80-96) |
| United States[15] | 1999-2006 | 720 | Relative survival – cohort method | 91 (88-93) |

Notes: a. International comparisons should be treated with due caution – see Appendix A.2.3, page 42.
b. A survival estimate for this diagnostic group was not published for Great Britain.[14]
c. 95% CI = 95% confidence interval.
d. Survival estimates have been rounded to the nearest integer.

# Appendix

## *A.1 Codes for childhood cancer diagnostic groups*

The International Classification of Childhood Cancers, version three (ICCC-3) classifies tumours according to the International Classification of Diseases for Oncology, third edition (ICD-O-3)[43] into 12 main diagnostic groups and 47 subgroups (Table A.1).

**Table A.1: International Classification of Childhood Cancer, Third Edition (ICCC-3)**

| Diagnostic group/*subgroup*[1] | ICD-O-3 codes[43] | |
|---|---|---|
| | Histology | Site |
| **I. *Leukaemias, myeloproliferative & 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. Other myeloproliferative diseases | 9945,9946,9975,9980,9982-9987,9989 | C000-C809 |
| e. Other & unspecified leukaemias | 9800,9801,9805,9860,9930 | C000-C809 |
| **II. *Lymphomas & reticuloendothelial tumours*** | | |
| a. Hodgkin lymphomas | 9650-9655,9659,9661-9665,9667 | C000-C809 |
| b. Non-Hodgkin lymphomas (excluding Burkitt lymphomas) | 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 lymphomas | 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 & intracranial/intraspinal tumours**** | | |
| a. Ependymomas and choroid plexus tumours* | 9383,9390-9394 | C000-C809 |
| b. Astrocytomas* | 9380 | C723 |
| | 9384,9400-9411,9420,9421-9424,9440-9442 | C000-C809 |
| c. Intracranial & intraspinal embryonal tumours* | 9470-9474,9480,9508 | C000-C809 |
| | 9501-9504 | C700-C729 |
| d. Other gliomas* | 9380 | C700-C722,C724-C729,C751,C753 |
| | 9381,9382,9430,9444,9450,9451,9460 | C000-C809 |
| e. Other specified intracranial & intraspinal neoplasms* | 8270-8281,8300,9350-9352,9360-9362,9412,9413,9492,9493,9505-9507,9530-9539,9582 | C000-C809 |
| f. Unspecified intracranial & intraspinal neoplasms* | 8000-8005 | C700-C729,C751-C753 |
| **IV. *Neuroblastomas & other peripheral nervous cell tumours*** | | |
| a. Neuroblastomas & ganglioneuroblastomas | 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. *Retinoblastomas*** | 9510-9514 | C000-C809 |
| **VI. *Renal tumours*** | | |
| a. Nephroblastomas & other nonepithelial renal tumours | 8959,8960,8964-8967 | C000-C809 |
| | 8963,9364 | C649 |
| b. Renal carcinomas | 8010-8041,8050-8075,8082,8120-8122,8130-8141,8143,8155,8190-8201,8210,8211,8221-8231,8240,8241,8244-8246,8260-8263,8290,8310,8320,8323,8401,8430,8440,8480-8490,8504,8510,8550,8560-8576 | C649 |
| | 8311,8312,8316-8319,8361 | C000-C809 |
| c. Unspecified malignant renal tumours | 8000-8005 | C649 |

*(Continued next page...)*

Source: Steliarova-Foucher et al., 2005.[10]

Note: * Diagnostic group/subgroup includes intracranial and intraspinal tumours of benign or uncertain behaviour.

**Table A.1 (*cont.*): International Classification of Childhood Cancer, Third Edition (ICCC-3)**

| Diagnostic group/*subgroup*[1] | ICD-O-3 codes[43] | |
|---|---|---|
| | Histology | Site |
| **VII. *Hepatic tumours*** | | |
| a. Hepatoblastomas | 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's tumours & related bone sarcomas | 9260 | C400-C419,C760-C768, C809 |
| | 9363-9365 | C400-C419 |
| d. Other specified malignant bone tumours | 8810,8811,8823,8830 | C400-C419 |
| | 8812,9250,9261,9262,9270-9275,9280-9282, 9290,9300-9302,9310-9312,9320-9322,9330, 9340-9342,9370-9372 | C000-C809 |
| e. Unspecified malignant bone tumours | 8000-8005,8800,8801,8803-8805 | C400-C419 |
| **IX. *Soft tissue & other extraosseous sarcomas*** | | |
| a. Rhabdomyosarcomas | 8900-8905,8910,8912,8920,8991 | C000-C809 |
| b. Fibrosarcomas & other fibrous neoplasms | 8810,8811,8813-8815,8821,8823,8834-8835 | C000-C399,C440-C768, C809 |
| | 8820,8822,8824-8827,9150,9160,9491,9540-9571,9580 | C000-C809 |
| c. Kaposi sarcomas | 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 |

*(Continued next page...)*

Source: Steliarova-Foucher et al., 2005.[10]

Note: * Diagnostic group/subgroup includes intracranial and intraspinal tumours of benign or uncertain behaviour.

**Table A.1 (*cont.*): International Classification of Childhood Cancer, Third Edition (ICCC-3)**

| Diagnostic group/*subgroup*[1] | ICD-O-3 codes[43] | |
|---|---|---|
| | Histology | Site |
| **X. *Germ cell, trophoblastic & gonadal tumours**** | | |
| a. Intracranial & intraspinal germ cell tumours* | 9060-9065,9070-9072,9080-9085,9100,9101 | C700-C729,C751-C753 |
| b. Malignant extracranial & 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 & unspecified malignant gonadal tumours | 8590-8671 | C000-C809 |
| | 8000-8005 | C569,C620-C629 |
| **XI. *Other malignant epithelial neoplasms & 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. 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 & 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 & unspecified malignant tumours*** | | |
| 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 |

Source: Steliarova-Foucher et al., 2005.[10]

Note: * Diagnostic group/subgroup includes intracranial and intraspinal tumours of benign or uncertain behaviour.

## *A.2 Data*

### A.2.1 Data quality

The quality of data contained in cancer registries is typically evaluated based on numerical indices for the proportion of records which have been morphologically verified (%MV) and the proportion which are based on death certificate only (%DCO).[44] Morphological verification includes diagnoses based on histological verification, exfoliative cytology and haematological examination of peripheral blood, histology of metastasis and autopsy with histology. High values of %MV and low values of %DCO generally indicate better quality data.

Indices of data quality for the APCR are shown in Table A.2. Morphological verification was greater than 95% for all diagnostic groups, apart from tumours of the central nervous system (86%), other and unspecified tumours (86%) and retinoblastoma (88%). Data quality was also high for the subset of intracranial and intraspinal tumours of benign or uncertain behaviour, with 92% of these cases being morphologically verified. The proportion of cases that were identified by death certificate only was consistently low across all diagnostic groups.

**Table A.2: Indices of data quality for the Australian Paediatric Cancer Registry, 1983-2006**

| Diagnostic group[a] | Number of Cases | %MV[b] | %DCO[c] |
|---|---|---|---|
| I. Leukaemias, myeloproliferative & myelodysplastic diseases | 4,591 | 99.3 | 0.1 |
| II. Lymphomas & reticuloendothelial neoplasms | 1,374 | 99.1 | 0.1 |
| III. Central nervous system & intracranial/intraspinal neoplasms[d] | 3,158 | 85.7 | 0.2 |
| IV. Neuroblastoma & other peripheral nervous cell tumours | 869 | 96.0 | 0.1 |
| V. Retinoblastoma | 357 | 87.7 | 0.3 |
| VI. Renal tumours | 735 | 98.8 | 0.1 |
| VII. Hepatic tumours | 174 | 96.0 | 1.7 |
| VIII. Malignant bone tumours | 602 | 98.7 | 0.0 |
| IX. Soft tissue & other extraosseous sarcomas | 820 | 98.5 | 0.0 |
| X. Germ cell tumours, trophoblastic tumours & neoplasms of gonads[d] | 511 | 95.1 | 0.4 |
| XI. Other malignant epithelial neoplasms & melanomas | 705 | 99.0 | 0.1 |
| XII. Other & unspecified malignant neoplasms | 29 | 86.2 | 0.0 |

Notes: 1. Diagnostic groups defined using the International Classification of Childhood Cancers (ICCC-3).[10]
2. %MV = percentage of cases that were morphologically verified.
3. %DCO = percentage of cases that were based on death certificate only.
4. Diagnostic group includes intracranial and intraspinal tumours of benign or uncertain behaviour.

### A.2.2 Data sources

The Australian Paediatric Cancer Registry collected data on the cohort of children diagnosed with cancer in Australia during 1983-2004 , with follow-up information on survival available to the end of 2006 (see Section 1.2). As part of this process, cases were matched against the 2006 National Death Index[45] to ensure that details regarding mortality were as accurate and complete as possible. The NDI is maintained by the Australian Institute of Health and Welfare (AIHW) based on data provided by the Registrars of Births, Deaths and Marriages in each State and Territory, and contains a record of all deaths in Australia since 1980.

Underlying mortality rates for all children in Australia that were used in the estimation of relative survival were calculated from national mortality data, sourced from the General Record of Incidence of Mortality (GRIM) books published by the AIHW,[46] combined with estimated resident population data in single years of age obtained from the Australian Bureau of Statistics.[47]

### A.2.3 International comparisons

Differences in survival between Australia and other parts of the world need to be interpreted with caution, as the estimates may be influenced by a number of factors, such as differences in scope, timing, population coverage, data quality and statistical methodology[48] (see Appendix A.3.3, page 43, for further details on the methods for calculating survival). For these reasons, the other countries included for comparison were limited to those for which recent estimates of childhood cancer survival were available, based on a similar methodology to that used in this report. In particular, it should be noted that in the context of childhood cancer there is little difference between observed and relative survival because the underlying expected survival is usually close to 100%.

### A.2.4 Tumour stage

The stage of a tumour is used to describe the extent to which the cancer has progressed. Staging takes into account a variety of factors, which typically include the size of a tumour, the number of lymph nodes involved, and whether the cancer has invaded adjacent organs or spread to distant sites. It is one of the most important predictors of survival and can also be used to determine the most appropriate form of treatment.[49]

Not all types of cancer can be staged. For childhood cancers, stage information is generally available for four of the twelve diagnostic groups, namely lymphomas, neuroblastoma, renal tumours and the diagnostic subgroup of rhabdomyosarcomas (part of the diagnostic group of soft tissue sarcomas).

The broad meaning of the different staging categories for solid tumours can be summarised as follows:

*Stage I* – tumour is localised to the part of the body where it originated without any evidence of spread and has been surgically removed.

*Stage II* – as per Stage I, except that the tumour has been incompletely removed.

*Stage III* –greater regional involvement, preventing surgical resection and often including involvement of lymph nodes.

*Stage IV* – cancer has spread (metastasised) to distant parts of the body, such as the lungs or bone marrow.

However, the details of how a particular stage is defined may vary depending on the type of cancer, particularly for lymphomas compared to solid tumours. Interested readers should refer to the individual classification systems for the specific staging details for each diagnostic group/subgroup:

*Hodgkin lymphomas* – Ann Arbor classification system.[21]

*Non-Hodgkin lymphomas (including Burkitt lymphomas)* – Murphy classification system.[22]

*Neuroblastoma* – International Neuroblastoma Staging System.[26]

*Renal tumours* – Third National Wilms' Tumor Study.[31]

*Rhabdomyosarcoma* – Intergroup Rhabdomyosarcoma Study-I.[38]

## *A.3 Methods*

### A.3.1 Confidence intervals

The level of accuracy of statistical estimates is typically reported in terms of a confidence interval, which specifies a range of values in which the actual result is expected to occur with a given level of certainty. For example, the 5-year relative survival rate for all children with cancer in Australia who were diagnosed during 1995-2004 was estimated to be 79.5% with a 95% confidence interval of 78.5%-80.5%. This means that there was a 95% probability that the true 5-year relative survival rate was somewhere between 78.5% and 80.5%.

For a given level of probability, a smaller confidence interval indicates a more reliable estimate. The number of cases that an estimate is based on is a major factor in determining the width of the associated confidence interval. Generally speaking, the greater the number of cases involved in calculating an estimate, the smaller the confidence interval will be. Due care should be taken when interpreting estimates which have a wide confidence interval.

### A.3.2 P-values

P-values provide a measure of the likelihood that differences in survival estimates were real or occurred by chance alone. Smaller p-values indicate survival rates that are more likely to be truly different from each other, rather than a result of random variation. An arbitrary cut-off value ($p < 0.05$) has been chosen to determine the statistical significance of comparisons throughout this report. One limitation of this approach is the problem of multiple comparisons, which can inflate the probability of incorrectly identifying the difference between estimates as being statistically significant.

The p-values shown throughout the report were obtained by fitting a Poisson model to the number of deaths within 5 years of diagnosis, offset by the log of the corresponding person years at risk. Models were simultaneously adjusted for sex, age group at diagnosis, year of follow-up and the particular variable of interest (for example, year of diagnosis). Differences between individual estimates (p-values not shown) were only deemed to be significant if the overall effect was also statistically significant.

### A.3.3 Relative survival

*Definition*
Observed survival measures the proportion of people who remain alive for a given period of time (typically 5 years) following a diagnosis of cancer. Relative survival is obtained by dividing the observed survival by the expected survival within the general population, taking into account age, sex and year of diagnosis. It is the most commonly presented measure of survival when using data from population-based cancer registries.[50] The main advantage of reporting relative survival is that it does not require information about the specific cause of death, only knowledge of whether or not the patient has died.

*Method*
Two approaches are available for calculating relative survival estimates: the traditional cohort method, which follows the same group of patients over time; and the period method, which is based on different groups of patients within an "at risk" window of time. Although the period method tends to produce more up-to-date estimates, particularly for longer-term survival,[51] the cohort method was deemed to be more suitable for the purposes of this report given that it describes the survival experience of a clearly defined group of individuals and is therefore easier for a wider audience to interpret.

The survival of all childhood cancer patients was followed up to the 31st December 2006, and those who were still alive at that date were censored. To avoid bias, patients whose cancer diagnosis was based on death certificate or autopsy only were excluded, as were those with a survival time of zero days or less.

A suite of SAS® computer programs developed by Paul Dickman[52] from the Karolinska Institutet in Sweden was used to generate the relative survival estimates. These programs use a life table (or actuarial) method for calculating observed survival. This approach involves dividing the total time being studied into a series of discrete intervals. Survival probabilities were formulated for each of these intervals, and then multiplied together to produce the observed survival estimate. Expected survival (based on age-specific national mortality data obtained from the Australian Institute of Health and Welfare[46]) was calculated based on the Ederer II method.[53] Three-year averages were used for expected survival to minimise the effects of year to year variation. Relative survival estimates were then obtained from the ratio of observed survival to expected survival.

## *A.4 Detailed estimates of survival for childhood cancer diagnostic subgroups*

**Table A.4a: Five-year relative survival for childhood cancer diagnostic subgroups[a] by sex, Australia, 1995-2004[b]**

| | Boys | | Girls | | |
|---|---|---|---|---|---|
| **Diagnostic subgroup** | **No. of cases[c]** | **Five-year relative survival (95% CI)[d]** | **No. of cases[c]** | **Five-year relative survival (95% CI)[d]** | ***p-value*** |
| Ia. Lymphoid leukaemias | 855 | 82.7 (79.8-85.2) | 737 | 88.2 (85.5-90.5) | *<0.001* |
| Ib. Acute myeloid leukaemias | 175 | 62.1 (54.4-68.9) | 154 | 62.5 (54.2-69.7) | *0.809* |
| IIa. Hodgkin lymphomas | 159 | 96.2 (91.5-98.4) | 81 | 98.9 (91.6-99.9) | *0.206* |
| IIb. Non-Hodgkin lymphomas (excl. Burkitt) | 156 | 83.4 (76.2-88.5) | 80 | 81.0 (70.4-88.1) | *0.626* |
| IIc. Burkitt lymphomas | 102 | 89.4 (81.5-94.1) | See footnote f | | *n.a.* |
| IIIa. Ependymomas and choroid plexus tumours[e] | 67 | 66.1 (53.3-76.1) | 62 | 71.3 (57.6-81.2) | *0.575* |
| IIIb. Astrocytomas[e] | 348 | 78.3 (73.6-82.3) | 322 | 77.4 (72.4-81.7) | *0.864* |
| IIIc. Intracranial & intraspinal embryonal tumours[e] | 156 | 52.3 (44.1-59.8) | 107 | 48.2 (38.0-57.6) | *0.643* |
| IIId. Other gliomas[e] | 82 | 57.0 (45.6-66.8) | 82 | 50.8 (39.2-61.2) | *0.595* |
| IIIe/IIIf. Other intracranial & intraspinal neoplasms[e] | 95 | 90.4 (82.3-94.9) | 85 | 93.7 (85.5-97.4) | *0.297* |
| IVa. Neuroblastoma & ganglioneuroblastoma | 191 | 66.4 (59.0-72.7) | 169 | 69.4 (61.6-75.9) | *0.533* |
| VIa. Nephroblastoma & nonepithelial renal tumours | 149 | 85.0 (78.0-90.0) | 169 | 92.3 (87.0-95.5) | *0.069* |
| VIIa. Hepatoblastoma | 49 | 81.5 (67.2-90.1) | 29 | 96.9 (77.8-99.8) | *0.124* |
| VIIIa. Osteosarcomas | 58 | 68.7 (54.4-79.3) | 64 | 72.2 (58.9-81.8) | *0.858* |
| VIIIc. Ewing tumours & related bone sarcomas | 71 | 66.9 (54.0-77.0) | 53 | 69.2 (53.2-80.7) | *0.398* |
| IXa. Rhabdomyosarcomas | 94 | 68.8 (57.7-77.6) | 66 | 67.4 (53.9-77.7) | *0.988* |
| IXb/IXc/IXd/IXe. Non-rhabdomyosarcomas | 88 | 69.1 (58.0-77.8) | 85 | 76.2 (65.4-84.1) | *0.344* |
| Xa. Intracranial & intraspinal germ cell tumours[e] | 43 | 83.9 (69.0-92.1) | 26 | 84.8 (59.5-95.0) | *0.564* |
| Xb. Malign. extracranial/gonadal germ cell tumours | 28 | 93.0 (75.0-98.4) | 39 | 82.3 (66.2-91.3) | *0.206* |
| Xc. Malignant gonadal germ cell tumours | 55 | 98.5 (88.0-100.0) | 48 | 100.0[g] n.a. | *n.a.* |
| XIb. Thyroid carcinomas | 20 | 93.8 (61.8-99.3) | 36 | 100.0[g] n.a. | *n.a.* |
| XId. Melanomas | 73 | 96.8 (87.3-99.4) | 102 | 97.1 (91.2-99.1) | *0.931* |
| XIa/XIc/XIe/XIf. Other & unspecified carcinomas | 48 | 80.8 (66.3-89.5) | 45 | 83.9 (68.9-92.0) | *0.406* |

Notes: a. Only diagnostic subgroups with a sufficient number of cases for analysis have been included in the table.
b. Cohort includes children diagnosed during 1995-2004 with follow-up to 31 Dec 2006.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval.
e. Diagnostic subgroup includes intracranial and intraspinal tumours of benign or uncertain behaviour.
f. Insufficient number of cases to calculate a separate survival estimate for girls.
g. Confidence interval (and corresponding p-value) could not be calculated because no deaths were recorded.
n.a. = not applicable.

**Table A.4b: Five-year relative survival for childhood cancer diagnostic subgroups[a] by age group at diagnosis, Australia, 1995-2004[b]**

| Diagnostic subgroup | <1 year old | | 1-4 years old | | 5-9 years old | | 10-14 years old | | |
|---|---|---|---|---|---|---|---|---|---|
| | No. of cases[c] | Five-year relative survival (95% CI)[d] | No. of cases[c] | Five-year relative survival (95% CI)[d] | No. of cases[c] | Five-year relative survival (95% CI)[d] | No. of cases[c] | Five-year relative survival (95% CI)[d] | *p-value* |
| Ia. Lymphoid leukaemias | 46 | 53.9 (38.4-67.1) | 809 | 89.7 (87.3-91.8) | 461 | 84.3 (80.4-87.4) | 276 | 79.2 (73.6-83.7) | *<0.001* |
| Ib. Acute myeloid leukaemias | 41 | 50.1 (33.8-64.5) | 110 | 65.8 (56.1-73.9) | 91 | 64.7 (53.8-73.6) | 87 | 60.8 (49.5-70.4) | *0.315* |
| IIa. Hodgkin lymphomas | See footnote f | | See footnote f | | 66 | 98.3 (88.1-99.8)[f] | 174 | 96.6 (92.5-98.6) | *0.305* |
| IIb. Non-Hodgkin lymphomas (excl. Burkitt) | See footnote f | | 59 | 82.0 (69.1-89.9)[f] | 76 | 82.3 (71.3-89.5) | 70 | 83.1 (74.2-89.2) | *0.999* |
| IIc. Burkitt lymphomas | See footnote f | | 27 | 92.7 (73.6-98.2)[f] | 53 | 85.2 (72.2-92.5) | 39 | 95.0 (81.1-98.8) | *0.243* |
| IIIa. Ependymomas and choroid plexus tumours[e] | See footnote f | | 72 | 59.4 (46.8-69.9)[f] | 34 | 72.6 (53.7-84.8) | 23 | 91.1 (67.4-97.9) | *0.009* |
| IIIb. Astrocytomas[e] | 37 | 63.7 (46.6-76.7) | 229 | 78.5 (72.5-83.3) | 214 | 79.6 (73.4-84.5) | 190 | 77.9 (71.2-83.2) | *0.205* |
| IIIc. Intracranial & intraspinal embryonal tumours[e] | 23 | 2.5 (0.2-11.4) | 96 | 39.6 (29.9-49.2) | 92 | 66.2 (54.9-75.3) | 52 | 64.3 (49.0-76.1) | *<0.001* |
| IIId. Other gliomas[e] | See footnote f | | 51 | 60.2 (45.4-72.2)[f] | 66 | 45.7 (33.0-57.5) | 47 | 59.2 (43.8-71.7) | *0.125* |
| IIIe/IIIf. Other intracranial & intraspinal neoplasms[e] | See footnote f | | 49 | 84.7 (70.4-92.4)[f] | 59 | 93.2 (83.0-97.4) | 72 | 96.0 (87.8-98.8) | *0.119* |
| IVa. Neuroblastoma & ganglioneuroblastoma | 152 | 84.5 (77.6-89.5) | 174 | 57.1 (49.1-64.2) | 34 | 48.8 (31.0-64.4)[f] | See footnote f | | *<0.001* |
| VIa. Nephroblastoma & nonepithelial renal tumours | 40 | 77.7 (60.7-88.1) | 204 | 92.0 (87.3-95.1) | 74 | 86.4 (76.1-92.5)[f] | See footnote f | | *0.066* |
| VIIa. Hepatoblastoma | 34 | 91.6 (75.6-97.6) | 35 | 91.8 (75.7-97.5) | See footnote g | | See footnote g | | *1.000* |
| VIIIa. Osteosarcomas | See footnote g | | See footnote g | | 22 | 90.9 (68.4-97.7) | 95 | 66.3 (55.3-75.2) | *0.069* |
| VIIIc. Ewing tumours & related bone sarcomas | See footnote f | | 21 | 65.5 (39.9-82.3) | 38 | 83.1 (65.9-92.1) | 65 | 59.6 (45.1-71.6) | *0.074* |
| IXa. Rhabdomyosarcomas | See footnote f | | 82 | 75.4 (63.4-83.7)[f] | 49 | 70.1 (55.1-81.9) | 29 | 43.8 (24.4-61.7) | *0.034* |
| IXb/IXc/IXd/IXe. Non-rhabdomyosarcomas | See footnote f | | 41 | 72.8 (56.4-83.9)[f] | 41 | 72.1 (54.9-83.7) | 91 | 72.5 (61.8-80.7) | *0.953* |
| Xa. Intracranial & intraspinal germ cell tumours[e] | See footnote f | | 21 | 85.6 (62.3-95.1)[f] | See footnote f | | 48 | 84.4 (69.6-92.4)[f] | *0.895* |
| Xb. Malign. extracranial/gonadal germ cell tumours | 36 | 86.3 (70.1-94.2) | 22 | 90.8 (68.6-97.6) | See footnote g | | See footnote g | | *0.715* |
| Xc. Malignant gonadal germ cell tumours | See footnote f | | 47 | 98.2 (86.1-100.0)[f] | See footnote f | | 56 | 100.0[h] n.a. | *n.a.* |
| XIb. Thyroid carcinomas | See footnote g | | See footnote g | | See footnote g | | 41 | 97.2 (81.1-99.7) | *n.a.* |
| XId. Melanomas | See footnote g | | See footnote g | | 30 | 100.0[h] n.a. | 135 | 96.1 (90.6-98.5) | *n.a.* |
| XIa/XIc/XIe/XIf. Other & unspecified carcinomas | See footnote g | | See footnote g | | See footnote g | | 70 | 86.8 (76.0-93.0) | *n.a.* |

Notes: a. Only diagnostic subgroups with a sufficient number of cases for analysis have been included in the table.
b. Cohort includes children diagnosed between 1995-2004 with follow-up to 31 Dec 2006.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval.
e. Diagnostic subgroup includes intracranial and intraspinal tumours of benign or uncertain behaviour.
f. The following age groups were combined for the specified diagnostic subgroups due to the small number of cases: "0-4 years old" for Non-Hodgkin lymphomas (excl. Burkitt), Burkitt lymphomas, ependymomas and choroid plexus tumours, other gliomas, other intracranial & intraspinal neoplasms, Ewing tumours & related bone sarcomas, rhabdomyosarcomas, non-Rhabdomyosarcoma soft tissue sarcomas, intracranial & intraspinal germ cell tumours, malignant gonadal germ cell tumours; "0-9 years old" for Hodgkin lymphomas; and "5-14 years old" for neuroblastoma & ganglioneuroblastoma, nephroblastoma & nonepithelial renal tumours, intracranial & intraspinal germ cell tumours, malignant gonadal germ cell tumours.
g. There were an insufficient number of cases to calculate survival estimates for children aged 0-4 years who were diagnosed with osteosarcomas or melanomas, children aged 0-9 years who were diagnosed with thyroid carcinomas or other & unspecified carcinomas and children aged 5-14 years who were diagnosed with hepatoblastoma or malignant extracranial & extragonadal germ cell tumours.
h. Confidence interval (and corresponding p-value) could not be calculated because no deaths were recorded.
n.a. = not applicable.

**Table A.4c: Five-year relative survival for childhood cancer diagnostic subgroups[a] by tumour stage[b], Australia, 1995-2004[c]**

| Diagnostic subgroup | Stage I | | Stage II | | Stage III | | Stage IV | | |
|---|---|---|---|---|---|---|---|---|---|
| | No. of cases[d] | Five-year relative survival (95% CI)[e] | No. of cases[d] | Five-year relative survival (95% CI)[e] | No. of cases[d] | Five-year relative survival (95% CI)[e] | No. of cases[d] | Five-year relative survival (95% CI)[e] | *p-value* |
| IIa. Hodgkin lymphomas | 79 | 100.0[f] n.a. | 82 | 98.7 (90.7-99.9) | 53 | 88.3 (75.7-94.7)[g] | See footnote g | | *0.001* |
| IIb. Non-Hodgkin lymphomas (excl. Burkitt) | 38 | 89.5 (74.1-96.0) | 26 | 88.6 (68.4-96.2) | 87 | 80.2 (70.0-87.2) | 27 | 80.0 (58.2-91.3) | *0.479* |
| IIc. Burkitt lymphomas | See footnote g | | 34 | 97.2 (80.9-99.7)[g] | 50 | 86.3 (73.0-93.3) | 20 | 90.3 (65.4-97.6) | *0.158* |
| IVa. Neuroblastoma & ganglioneuroblastoma | 68 | 95.6 (86.5-98.8) | 44 | 90.9 (77.0-96.8) | 46 | 73.3 (57.4-84.1) | 188 | 49.8 (42.3-56.9) | *<0.001* |
| VIa. Nephroblastoma & nonepithelial renal tumours | 70 | 97.3 (89.0-99.5) | 76 | 92.7 (83.1-97.0) | 78 | 92.4 (83.7-96.6) | 70 | 74.0 (61.9-82.8) | *<0.001* |
| IXa. Rhabdomyosarcomas | See footnote g | | 68 | 83.0 (71.3-90.3) | See footnote g | | 59 | 53.0 (39.1-65.0) | *<0.001* |

Notes: a. Only diagnostic subgroups with a sufficient number of cases for analysis and for which staging information was available have been included in the table.

b. The Ann Arbor classification system[22] was used to stage Hodgkin lymphomas, the Murphy classification system[23] was used to stage non-Hodgkin lymphomas, the International Neuroblastoma Staging System[27] was used to stage neuroblastoma, the Third National Wilms' Tumor Study classification system[32] was used to stage nephroblastoma & nonepithelial renal tumours, and the Intergroup Rhabdomyosarcoma Study-I[39] was used to stage rhabdomyosarcomas.

c. Cohort includes children diagnosed between 1995-2004 with follow-up to 31 Dec 2006.

d. The initial number of eligible cases in the cohort.

e. 95% CI = 95% confidence interval.

f. Confidence interval could not be calculated because no deaths were recorded.

g. The following stage groups were combined for the specified diagnostic subgroups due to the small number of cases: "Stages I/II" for Burkitt lymphomas and rhabdomyosarcomas; and "Stages III/IV" for Hodgkin lymphomas and rhabdomyosarcomas.

n.a. = not applicable.

**Table A.4d: Five-year relative survival for childhood cancer diagnostic subgroups[a] by year of diagnosis, Australia, 1983-2004[b]**

| | 1983-1994 | | 1995-2004 | | |
|---|---|---|---|---|---|
| **Diagnostic subgroup** | **No. of cases[c]** | **Five-year relative survival (95% CI)[d]** | **No. of cases[c]** | **Five-year relative survival (95% CI)[d]** | ***p-value*** |
| Ia. Lymphoid leukaemias | 1,676 | 75.5 (73.4-77.5) | 1,592 | 85.3 (83.3-87.0) | *<0.001* |
| Ib. Acute myeloid leukaemias | 355 | 44.1 (38.9-49.2) | 329 | 62.3 (56.7-67.3) | *<0.001* |
| IIa. Hodgkin lymphomas | 223 | 94.8 (90.9-97.1) | 240 | 97.1 (93.8-98.7) | *0.222* |
| IIb. Non-Hodgkin lymphomas (excl. Burkitt) | 289 | 75.9 (70.6-80.5) | 236 | 82.6 (76.9-86.9) | *0.069* |
| IIc. Burkitt lymphomas | 88 | 73.5 (62.7-81.6) | 119 | 90.1 (83.1-94.3) | *0.002* |
| IIIa. Ependymomas and choroid plexus tumours[e] | 149 | 66.5 (58.3-73.4) | 129 | 68.6 (59.6-76.0) | *0.672* |
| IIIb. Astrocytomas[e] | 666 | 77.3 (73.9-80.3) | 670 | 77.9 (74.5-80.9) | *0.779* |
| IIIc. Intracranial & intraspinal embryonal tumours[e] | 272 | 49.5 (43.4-55.2) | 263 | 50.5 (44.2-56.5) | *0.979* |
| IIId. Other gliomas[e] | 175 | 48.5 (40.9-55.8) | 164 | 53.8 (45.8-61.2) | *0.435* |
| IIIe/IIIf. Other intracranial & intraspinal neoplasms[e] | 175 | 80.0 (73.3-85.2) | 180 | 92.0 (86.8-95.2) | *0.003* |
| IVa. Neuroblastoma & ganglioneuroblastoma | 397 | 51.4 (46.3-56.2) | 360 | 67.8 (62.6-72.4) | *<0.001* |
| Via. Nephroblastoma & nonepithelial renal tumours | 331 | 87.5 (83.4-90.7) | 318 | 88.9 (84.8-92.0) | *0.627* |
| VIIa. Hepatoblastoma | 45 | 82.6 (67.9-91.1) | 56 | 87.1 (77.1-93.1) | *0.247* |
| VIIIa. Osteosarcomas | 114 | 64.8 (55.3-72.8) | 122 | 70.6 (61.2-78.1) | *0.291* |
| VIIIc. Ewing tumours & related bone sarcomas | 153 | 64.4 (56.2-71.4) | 124 | 67.8 (58.0-75.8) | *0.417* |
| IXa. Rhabdomyosarcomas | 232 | 64.8 (58.3-70.6) | 160 | 68.2 (59.9-75.2) | *0.179* |
| IXb/IXc/IXd/IXe. Non-rhabdomyosarcomas | 179 | 84.5 (78.3-89.1) | 173 | 72.5 (65.0-78.6) | *0.007* |
| Xa. Intracranial & intraspinal germ cell tumours[e] | 58 | 75.9 (62.7-85.0) | 69 | 84.5 (72.8-91.5) | *0.179* |
| Xb. Malign. extracranial/gonadal germ cell tumours | 49 | 84.1 (70.3-91.9) | 67 | 86.8 (76.0-93.0) | *0.766* |
| Xc. Malignant gonadal germ cell tumours | 86 | 93.3 (85.3-97.1) | 103 | 99.2 (93.5-100.0) | *0.226* |
| XIb. Thyroid carcinomas | 45 | 95.7 (83.5-99.0) | 56 | 97.9 (85.9-99.8) | *0.515* |
| XId. Melanomas | 200 | 92.9 (88.4-95.8) | 175 | 97.0 (92.8-98.8) | *0.063* |
| XIa/XIc/XIe/XIf. Other & unspecified carcinomas | 81 | 78.0 (67.3-85.6) | 93 | 82.2 (72.6-88.8) | *0.544* |

Notes: a. Only diagnostic subgroups with a sufficient number of cases for analysis have been included in the table.
b. Cohorts include children diagnosed during 1983-1994 and 1995-2004 with follow-up to 31 Dec 2006.
c. The initial number of eligible cases in the cohort.
d. 95% CI = 95% confidence interval.
e. Diagnostic subgroup includes intracranial and intraspinal tumours of benign or uncertain behaviour.

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