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Articles

Long-term survival and cure fraction estimates for paediatric central nervous system tumours in 31 European countries (EUROCARE-6): a population-based study

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Summary

Background

Clinically relevant survival outcomes, including cure fraction estimates, and long-term survival outcomes of paediatric CNS tumours from large-scale databases have not been reported for Europe. Moreover, various biases hinder direct geographical comparisons, thereby limiting the effective translation of population-based findings into cancer care, surveillance, and research. We aimed to estimate these survival outcomes across Europe through the EUROCARE database.

Methods

In this population-based study, we analysed survival data from the EUROCARE-6 database from children younger than 15 years with a CNS tumour across 31 European countries. For the period 2008–13, we estimated observed survival via the actuarial method, and 5-year observed survival was reported at the European level and national level for four major CNS tumour groups. For the period 1998–2013, cure fraction was estimated through a mixture cure model assuming constant

long-term mortality from other causes. Additionally, model-based 10-year and 15-year survival were estimated.

Findings

For observed survival analyses, 13 782 tumour cases were included. 5-year observed survival was 72% (95% CI 68 to 75) for ependymomas, 92% (91 to 93) for low-grade gliomas, 47% (45 to 49) for high-grade gliomas, 24% (21 to 27) for high-grade gliomas excluding glioma not otherwise specified, and 64% (62 to 67) for medulloblastomas. A total of 30 392 children were included in the cure fraction analysis. During the study period, the largest absolute increase in cure fraction was observed for ependymomas from 65% (57 to 73) in 1998–2001 to 79% (69 to 89) in 2010–13, whereas low-grade gliomas increased from from 89% (85 to 94) to 95% (89 to 100), high-grade gliomas had a 6 percentage point change increase (2 to 10), and medulloblastomas increased from 52% (49 to 55) to 56% (51 to 60). The estimated 10-year and 15-year survival rates were highest for low-grade gliomas at 90·6% (89·4 to 91·7) at 10 years and 88·5% (87·2 to 89·8) at 15 years, whereas the lowest survival rates were observed for high-grade gliomas excluding glioma not otherwise specified at 20·5% (17·0 to 24·1) and 19·0% (15·6 to 22·5).

Interpretation

This study is the first to report a comprehensive evaluation of survival parameters for paediatric CNS tumour patients in Europe. These outcomes are important to evaluate advances in care for children with a CNS tumour.

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Introduction

Paediatric CNS tumours are the most common solid tumours and the leading cause of death in children with cancer.[1] This broad group of malignant and non-malignant tumours consists of a wide variety of tumour types and with 5-year survival ranging from 2% for diffuse midline gliomas located at the pons to almost 100% for pilocytic astrocytomas.2, 3 Additionally, survival probabilities vary by age and tumour location.4, 5 Moreover, when comparing survival estimates across countries, different diagnostic and registration practices could introduce bias.[6] All these circumstances need careful consideration when analysing survival outcomes of paediatric CNS tumours.

Population-based studies have shown large variations in survival estimates between European countries.[7] These studies typically rely on the International Classification of Diseases (ICD) for Oncology behaviour codes to define CNS tumours groups. However, this variable does not capture the clinical heterogeneity of paediatric CNS tumours. These limitations make it hard to inform stakeholders (eg, clinicians and policy makers) sufficiently, leaving the potential of

population-based studies largely unaddressed.

In this study, we used the comprehensive EUROCARE database to assess the 1-year, 3-year, and 5-year survival for 12 paediatric CNS tumour groups, considering clinical characteristics such as sex, age, CNS WHO grade, and tumour location. The large European cohort and coverage since 1998 make it possible to estimate the proportion of children cured and long-term survival (up to 15 years). Additionally, for selected countries, we aimed to evaluated 5-year survival outcomes for four major tumour groups.

Research in context

Evidence before this study

Assessing population-based cancer survival is one of the crucial measures for evaluating the effectiveness of a country's health-care system in addressing cancer management. Detailed insight into clinical characteristics, cure fraction, long-term survival outcomes, and geographical differences in the survival of paediatric CNS tumours is needed to effectively inform stakeholders, such as clinicians and policy makers. Due to the rarity of paediatric CNS tumours, large-scale population-based studies that use cancer registry data can have an important role in this process. We searched Medline from Jan 1, 2014, to March 14, 2024, with the search string: "child* OR paediatric AND (brain cancer OR brain neoplasm OR central nervous system tumour) AND survival AND registries AND Europe". No studies reported details on the underlying clinical characteristics and reported survival outcomes beyond 10 years. Several studies have evaluated survival differences between European countries, but all studies were limited by the use of the The International Classification of Diseases for Oncology (ICD-O), third edition, behaviour code (ie, the fifth digit in the morphology code). Although the behaviour code allows for stratifying other cancer types by malignancy, its application to CNS tumours results in clinically non-informative groupings that do not encompass the heterogeneity inherent to these neoplasms. These deficiencies have hindered the ability to translate findings into clinical implications and effective policies. Consequently, the potential of population-based studies for paediatric CNS tumours has remained partly unaddressed.

Added value of this study

To address the deficiencies of earlier reports, this study uses EUROCARE data to analyse more than 30 000 cases of paediatric CNS tumours from the period 1998–2013 at a European level, encompassing 80 registries from 31 countries. To the best of our knowledge, this study is the first to report detailed survival outcomes on a European level, informing on survival patterns in underlying patient characteristics and providing a unique insight into the cure fraction and projected long-term survival outcomes for paediatric CNS tumours. In addition, this study includes a survival comparison specific to tumour groups between European countries, accounting for differences in registration procedures and under ascertainment of cases, and providing a more accurate, and clinically relevant, insight into survival outcomes of children with a CNS

tumour in Europe.

Implications of all the available evidence

The results of this study provide insights into survival for 12 CNS tumour groups and their underlying heterogeneity in clinical characteristics. Moreover, by estimating cure fraction and long-term survival outcomes the results of this study inform on long-term disease outcomes, potentially affecting the duration of clinical follow-up for children treated for a CNS tumour. Finally, a geographical survival comparison of major tumour groups offers valuable information for policy makers in directing targeted resource allocation and improving cancer registry procedures for paediatric CNS tumours.

Section snippets

Study design and data collection

EUROCARE-6 is a European population-based study, encompassing data on cancer cases in European children age 0–14 years and covers around 85% of the European paediatric population. All children with a CNS tumour (ICD for Oncology, third edition [ICD-O-3] topography codes C70–C72·9 and C75·1–C75·3.8) were included from the EUROCARE-6 database, regardless of tumour behaviour code (either 0 for benign, 1 for low malignant potential, or 3 for malignant).[8] We included diagnoses between Jan 1, 1998, …

Results

From Jan 1, 2008, to Dec 31, 2013, 14 745 CNS tumours were identified; 22 with major errors and 670 with misclassified morphology codes were excluded, leaving 14 053 for incidence analyses. For survival analyses, 44 death-certificate-only or autopsy-only cases, 202 cases without survival data, and 25 second tumours were excluded, yielding 13 782 cases. For cure fraction analyses (1998–2013), 36 320 tumours were recorded; 59 with major errors, 5287 were misclassified, 196 death-certificate-only …

Discussion

By using data from the EUROCARE project, we comprehensively analysed paediatric CNS tumour survival in Europe for, to our knowledge, the largest number of patients to date. Survival varied

largely between CNS groups, with a 5-year observed survival of less than 30% for HGGs (excluding malignant glioma not otherwise specified) and greater than 90% for LGGs. By stratifying analyses by sex, age, CNS WHO grade, and tumour location, we provide insight into underlying disparities and patterns, …

Declaration of interests

We declare no competing interests. …

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