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Incidence of and survival from Wilms' tumour in adults in Europe: Data from the EUROCARE study

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ABSTRACT

Wilms' tumour, or nephroblastoma, is an embryonal cancer of the kidney that occurs mainly in young children. This is a very rare tumour among adults, with an incidence rate of less than 0.2 per million per year. The aims of this study were to report the survival of adults diagnosed with nephroblastoma in Europe and to analyse time trends and geographic variations in survival. All the adults (age range 15–99 years) diagnosed with a Wilms' tumour during 1983–1994 and registered by one of the 22 cancer registries in 16 countries contributing to the EUROCARE (European cancer registries study on cancer patients' survival and care) database were analysed. Relative survival at 1 and 5 years after diagnosis was estimated by age, sex, geographic area, period of diagnosis and tumour stage. A total of 143 patients, with a median age of 34 years, were included in the analysis. Crude annual incidence rates varied geographically between 0.17 and 0.27 per million. Overall relative survival was 69.9% (95% confidence interval (CI) 61.8–78.0%) at 1 year and 47.3% (38.2–56.4%) at 5 years. Survival was 2.1-fold higher for women than for men (95% CI 1.3–3.5). There was a non-significant trend for better survival for younger patients and localised tumours, but no improvement in survival by period of diagnosis. Survival was not different between geographic areas. Our results suggest a poorer outcome of nephroblastoma in adults compared with published results in children. This may, at least partly, be explained by the rarity of this diagnosis. Prognosis may be improved by the use of specific treatment guidelines for nephroblastoma in adults.

1. Introduction

Wilms' tumour (nephroblastoma) is an embryonal kidney cancer that occurs mainly in young children. In Europe, the incidence rate in children (0–14 years) is about 10 per million per year.[1] Approximately 900 children are diagnosed each year, comprising about 6% of all childhood solid tumours. The median age at diagnosis is 3–4 years in most countries, and 90% of cases are diagnosed by the age of 7 years. Wilms' tumour is extremely rare among adults, with an incidence

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rate of less than 0.2 per million per year in Europe and the USA. About 70 new cases arise in adults in Europe each year.

Most Wilms' tumours in children are treated in specialised centres, whereas in adults the diagnosis is often unexpected, following nephrectomy for presumed renal cell cancer. There is often a delay in initiating chemotherapy while diagnostic review is undertaken. Previously published data, mainly case reports and small institutional series, suggest worse survival for adults than children, but adults treated according to recent paediatric protocols may have somewhat better outcomes.[2–5]

The EUROCARE (European cancer registries study on cancer patients' survival and care) study, including data from 67 cancer registries covering a combined population of 100 million in 22 European countries, offers a unique opportunity to study the epidemiology of rare cancers in a population of continental size.[6] We report here the survival of adults diagnosed with Wilms' tumour in Europe, in relation to demographic and clinical variables. We also analyse time trends and geographic variation in survival between European regions.

2. Patients and methods

We analysed the survival of adults (15–99 years) who were diagnosed with primary Wilms' tumour during 1983–1994, registered by one of the contributing cancer registries, and followed up until at least the end of 1998, most to the end of 1999. The cases were contributed by 22 population-based cancer registries in 16 countries:[6] Denmark, Iceland, Norway, Sweden, (the Nordic countries); Estonia, Poland, Slovakia, Slovenia (eastern Europe); France, Germany, Italy, the Netherlands and Spain (western Europe); Scotland, England and Wales (UK). All the registries included in this analysis provided data for the 12 years 1983–1994. The base population covered by participating registries in this study was 93 million persons, about 25% of the population in the 16 countries.

Eligible cases were defined as primary malignant neoplasms of the kidney (9th revision of the International Classification of Diseases (ICD-9): 189.0 and ICD-10: C64) with the morphology of a nephroblastoma (M8960/3) in the International Classification of Diseases for Oncology (ICD-O).

The data include cases that were verified microscopically by histology or cytology, and a few not so verified. Cases known to registries by death certificate only (DCO) or diagnosed only at autopsy were excluded from the survival analysis. No cases were lost to follow-up. Descriptions of the cancer registries, data collection methods and procedures for ensuring the comparability and quality of data have been published in EUROCARE monographs.[7,8]

Relative survival was estimated as the ratio of the survival observed among the cancer patients and the survival that would have been expected if they had the same death rates as the corresponding general population.[9] Regional or national life tables were used to estimate expected survival.[10] Relative survival was calculated using the Hakulinen method.[11] Overall (European) relative survival was estimated as the unweighted average of the relative survival in each country. Relative survival at 1 and 5 years after diagnosis was estimated by age, sex, geographic area, period of diagnosis and tumour stage. The relative excess risk of death, after adjustment for differential background mortality in contributing regions or countries, was modelled as a function of covariates, using a multi-regression approach for grouped life table data in the framework of generalised linear models.[12]

3. Results

3.1. Patient characteristics

Among 76,625 primary malignant neoplasms of the kidney diagnosed in adults between 1983 and 1994 and included in the EUROCARE database, 143 (0.19%) were Wilms' tumours and are included in the incidence analysis (Table 1). Six patients were excluded from the survival analysis because their Wilms' tumour was a second or later primary, and four because their tumour was detected at autopsy (2 cases) or registered solely from a death certificate (2 cases), hence 133 patients were included in the survival analysis, 93% of those eligible (Table 2).

Diagnosis was microscopically verified in 97% of cases (Table 1). Median age at diagnosis was 34 years (33.5 years in males, 36 years in females), but 20% of patients were aged 60 years or over. There were 69 (51.9%) females and 64 (48.1%) males. Most of the cases were registered in the UK (45.9%) and Nordic countries (25.6%), and fewer in Western (12.0%) and Eastern (22.0%) countries. The registries of the Nordic countries (Iceland, Norway, Sweden and Denmark), Scotland, Wales, Estonia, Slovakia and Slovenia all have national coverage, whilst the others are regional, covering from

Table 1 – Wilms' tumour in adults (15–99 years), Europe, patients diagnosed 1983–1994: patient characteristics and crude incidence rates by age, period of diagnosis and geographical region

	Cases (*n*)	%	Annual incidence rate per million
All	143	100	0.188
Age group (years)			
15–19	28	19.5	0.414
20–24	21	15.8	0.289
25–34	22	15.8	0.158
35–44	17	11.3	0.130
45–59	25	17.3	0.158
60 and over	30	20.3	0.157
Period of diagnosis			
1983–1985	32	21.1	0.177
1986–1988	22	15.8	0.116
1989–1991	39	27.1	0.202
1992–1994	50	36.1	0.256
Region			
Nordic	34	25.6	0.192
Western	16	12.0	0.172
UK	69	45.9	0.172
Eastern	24	16.5	0.275
Microscopically verified			
Yes	137	97.0	
No	6	3.0	

Table 2 – Relative survival (%) and 95% confidence interval (CI), Wilms' tumour in adults (15–99 years): Europe, patients diagnosed 1983–1994

	Patients (*n*)	One-year survival (95% CI)		Five-year survival (95% CI)	
All	133	69.9	(61.8–78.0)	47.3	(38.2–56.4)
Sex					
Female	69	75.8	(65.4–86.2)	61.3	(49.0–73.5)
Male	64	63.5	(51.2–75.8)	31.7	(19.4–43.9)
Age group (years)					
15–19	26	73.1	(55.7–90.5)	54.0	(34.4–73.6)
20–24	21	81.0	(63.9–98.2)	57.0	(35.1–78.9)
25–34	21	71.5	(51.8–91.2)	27.9	(7.9–48.0)
35–44	15	66.8	(42.4–91.2)	47.0	(21.0–73.1)
45–59	23	70.0	(50.7–89.3)	49.2	(27.6–70.7)
60–99	27	58.0	(38.0–78.0)	42.2	(19.1–65.3)
Period of diagnosis					
1983–1985	28	76.0	(59.4–92.6)	41.6	(21.8–61.5)
1986–1988	21	67.0	(46.3–87.7)	38.7	(16.9–60.5)
1989–1991	36	53.2	(36.4–70.0)	37.5	(20.7–54.3)
1992–1994	48	80.1	(68.3–92.0)	60.3	(45.0–75.6)
Region					
Nordic	34	65.0	(48.6–81.5)	50.8	(33.2–68.4)
Western	16	75.1	(53.4–96.8)	37.4	(12.8–62.0)
UK	61	71.7	(59.8–83.5)	52.4	(38.5–66.2)
Eastern	22	68.7	(48.7–88.7)	32.6	(11.6–53.5)
Stage					
Localised	15	86.9	(69.3–100.0)	73.7	(50.5–96.9)
Regional	15	80.6	(59.8–100.0)	47.5	(20.3–74.7)
Metastatic	14	57.5	(30.9–84.1)	14.7	(0.0–34.1)
Unknown	89	67.1	(57.0–77.3)	47.7	(36.5–58.9)

3% to 63% of the population of their respective countries. Most cases (69%) were registered after 1988.

3.2. *Incidence*

Crude annual incidence rates varied between 0.17 and 0.27 per million in the four regions (Table 1). Overall crude incidence rate was 0.19 per million (95% confidence interval (CI) 0.16–0.22). The proportion of adult Wilms' tumours among all kidney cancers was 0.33% or less in most registries, but reached 0.55% in Estonia and 0.61% in Wales.

3.3. *Relative survival*

Overall relative survival was 69.9% (95% CI 61.8–78.0%) at 1 year and 47.3% (38.2–56.4%) at 5 years (Fig. 1, Table 2). Survival was higher for women than for men, both at 1 year (75.8% versus 63.5%) and at 5 years (61.3% versus 31.7%). Survival was somewhat higher for younger patients: 1- and 5-year survival for those aged 20–24 years was 81.0% and 57.0%, respectively, compared with 58.0% and 42.2%, respectively, for those aged 60 years and over. One-year survival did not increase markedly between 1983–1985 (76.0%) and 1992–1994 (80.1%). Five-year survival improved more, from 41.6% to 60.3%. One-year survival ranged from 65% to 75.1% between the four European regions, while 5-year survival ranged from 32.6% in the Eastern European registries up to 52.4% for the UK registries. Information on tumour stage was available for only 44 (33%) of cases. Even among this sub-group, however, more advanced stage at diagnosis was associated with lower 5-year survival: 73.7% for localised tumours, 47.5% for those with regional extension and 14.7% for metastatic tumours. Cases with no information on stage had 5-year survival of 47.7%, very close to the overall average (47.3%).

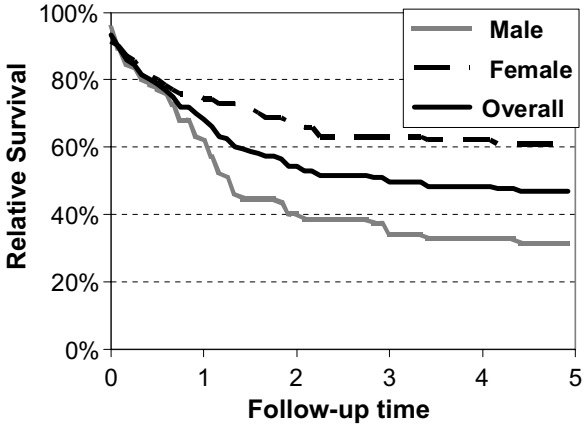

Fig. 1 – Relative survival (%) up to 5 years, Wilms' tumour in adults (15–99 years), Europe, patients diagnosed 1983–1994 and followed up to 1999.

Survival by age and stage at diagnosis was examined in men and women separately. Five-year survival was higher for women at all ages, the sex difference in survival ranging from 15% to 52% (Table 3). Five-year survival was similar for localised tumours in both sexes, but higher in women for tumours with regional extension (67.8% versus 34.4%) and for those of unknown stage (60.3% versus 27.2%).

3.4. *Prognostic factors*

Univariate analyses suggest that advanced tumour stage is an adverse prognostic factor, but the impact of stage on survival

Table 3 – Five-year relative survival (%) from Wilms' tumours in adults (15–99 years) by age, sex and stage at diagnosis: Europe, patients diagnosed 1983–1994

	Males		Females	
	Patients (*n*)	Survival (%)	Patients (*n*)	Survival (%)
Stage				
Localised	7	71.7	8	75.4
Regional	9	34.4	6	67.8
Metastatic	12	17.1	2	–
Unknown	36	27.2	53	60.3
Age group (years)				
15–19	13	46.3	13	61.6
20–24	10	30.2	11	81.9
25–34	11	9.1	10	48.1
35–44	10	30.3	5	80.3
45–59	8	38.4	15	54.6
60–99	12	26.9	15	49.8

Table 4 – Wilms' tumours in adults (15–99 years): relative excess risk of death up to 5 years after diagnosis, and 95% confidence interval (CI): age, sex, region and period of diagnosis

	Relative excess risk of death	95% CI	Test for trend
Sex			
Female	1		
Male	2.1	(1.3–3.5)	
Age group (years)			
15–34	1		
35–59	1.1	(0.6–2.0)	
60–99	1.5	(0.8–3.0)	$P = 0.487$
Period of diagnosis			
1983–85	1		
1986–88	1.3	(0.6–2.9)	
1989–91	1.4	(0.7–2.7)	
1992–94	0.8	(0.4–1.5)	$P = 0.231$
Region			
Nordic	1		
Western	1.5	(0.7–3.3)	
UK	0.9	(0.5–1.8)	
Eastern	1.6	(0.8–3.2)	

Relative excess risks are mutually adjusted for all other variables in the table and by follow-up time.

could not be modelled because information was available for only one-third of patients. Estimates of the relative excess risk of death within 5 years of diagnosis were adjusted for age, sex, calendar period of diagnosis and region of Europe (Table 4). Compared with women, the risk of death in men was significantly two-fold higher within 5 years of diagnosis. Relative to the youngest patients (15–34 years), the excess risk of death increased with age to 1.5-fold for those aged 60 years and over at diagnosis, but the trend across three broad age groups was not statistically significant. Similarly, there was no significant evidence for a temporal trend in the excess risk of death. Geographic variation in the excess risk was not significant.

4. Discussion

Wilms' tumour in adults is a rare tumour whose incidence and outcome has not been studied at the population level until this report. Even with a population base of 90 million over a 12-year period, there were only 143 cases for analysis. Crude incidence rates varied between 0.17 and 0.27 per million per year. This is very similar to the incidence we estimated for the United States of America (USA)-population covered by the SEER programme, using the SEERSTAT programme (0.15 per million, based on 45 cases over the same time-period). Relative survival at 5 years ranged from 33% to 52% across Europe, and from 74% for localised tumours down to 15% for tumours that were metastatic at diagnosis. The relative excess risk of death for men was twice as high as for women. Survival at 1 and 5 years was higher for those diagnosed in the 1990s than the 1980s, but the trend in relative excess risk of death within 5 years across four triennia of diagnosis (1983–1994) was not statistically significant after adjustment for age, sex and geographic region. Regional differences in survival were not significant.

The main concern in studying incidence and survival for such a rare tumour, which is likely to be unfamiliar to almost every treating clinician, relates to the accuracy of diagnosis and the completeness of registration. In the EUROCARE study, all the participating cancer registries provided data to an agreed protocol. Overall, 97% of cases were microscopically confirmed, and the figure was 100% in all but two registries, suggesting that the quality of diagnosis is high. However, the completeness of ascertainment of Wilms' tumour in adults would require pathological review of all registered cases, and a suitable sample of adult renal tumours not identified in the registry as Wilms' tumour. Very few cases were excluded as autopsy-detected cases or death-certificate-only registrations, and no cases were lost to follow-up. The two-fold range in incidence across four regions of Europe was not significant, but 11 of the 33 adult tumour registries that provided data for the entire 12-year period 1983–1994 did not record a single case, and most reported fewer than one case per year. Crude incidence rates ranged from zero up to 0.7 per million per year. For such a rare tumour, Poisson variation needs to be considered. For 8 of the 22 registries in which cases were actually recorded, the lower 95% confidence bound for the crude incidence rate would be compatible with fewer than one case in 12 years, and the population covered by those registries ranged from 187,000 to 2.9 million (data not shown). The population covered by the 11 registries from which no cases were recorded covered a similar range, from 227,000 to 1.9 million. This suggests under-ascertainment in at least some regions of Europe, as does the variation in the proportion of Wilms' tumour among all adult kidney tumours. Any such under-reporting is unlikely to be biased with respect to survival. The incidence of Wilms' tumour in children used to be considered as similar in most countries, and a rough guide to the completeness of cancer registration. Recent data from specialised childhood cancer registries in many countries show significant geographic variation of around three-fold, however, with the lowest incidence in Asian populations and the highest in Europe and North America.[1] These differences may have a genetic basis, which might also explain differences in incidence of Wilms' tumour in adults between European populations.[13]

Relative survival at 5 years in women (61%) was almost 30% higher than in men, a sex difference not seen in paediatric Wilms' tumour. This marked sex difference was seen in all six age groups examined, although numbers of cases were small. Higher survival for women than men is a feature of many solid tumours in Europe, although the sex difference in survival for kidney cancer is small.[6] This might reflect more generic underlying differences in how the sexes access healthcare services as adults, since a female advantage in survival is seen for many adult tumours.[6] For the limited number of cases with data on stage at diagnosis, 5-year survival was significantly worse for metastatic than localised tumours. The data were inadequate to examine survival by sex within each stage at diagnosis, but the two-fold excess risk of death in men persisted after adjustment for data on stage where available.

In childhood Wilms' tumour, anaplastic morphology and age at diagnosis over 2 years are both adverse prognostic factors. Sex is not a significant prognostic factor, and with stage-specific treatment, only metastatic disease has poorer survival. Even for metastatic disease, however, 5-year survival in children exceeds 70%.[14] This contrasts with 5-year relative survival of less than 15% observed for patients with metastatic disease in this series. Although stage data were not available for two-thirds of our cases, survival for unstaged cases was very close to the average survival for staged cases, which suggests that staged cases are not biased with respect to survival.

It is unclear whether Wilms' tumours in adults and children are biologically different. Several other explanations of the difference in outcome between adults and children are possible: these include the unfamiliarity among treating clinicians of Wilms' tumour in adults (possibly leading to diagnostic error and delay) and the lack of specific treatment protocols for adults. Thus, a recent review of institutional experience with Wilms' tumour in adults concluded that poor compliance with specific therapeutic guidelines may contribute to the poorer outcome in adults than in children.[4] Indeed, in recent reports of the outcome of adults with Wilms' tumour treated on one of two international paediatric protocols, stage-specific survival for adults was not significantly different from that of children, although treatment-related morbidity and mortality were higher.[2,3] The patients in these trials may not be representative of all adults with Wilms' tumour, since their median age at diagnosis (26 and 17.9 years, respectively) was lower than the median age (34 years) of patients included in this population-based series. Differences in treatment are more likely to explain the lower survival in our series than for adults in those trials, however, since we did not see a significant age-dependence of survival.

5. Conclusion

The EUROCARE database provides a unique opportunity to investigate very uncommon cancers in European populations and has led to some unexpected results in Wilms' tumour. Thus, we can see that this embryonal kidney cancer does actually occur in patients aged over 60 years, which has implications for its presumed origin in persistent metanephric blastemal cells. Either these survive in an undifferentiated state for many decades or there are renal cells capable of 'de-differentiation' during transformation. A detailed study of the biology of Wilms' tumour in adults may help to address these theories but has not yet been performed.

Collaboration with clinicians is essential to understand the pitfalls in studying a rare tumour, particularly in relation to the accuracy of diagnosis and the expediency of initiating appropriate treatment. The outcome for adults diagnosed with Wilms' tumour was worse than for children, but comparison with survival data from adult patients treated according to paediatric trial protocols suggests potential for improvement.[2,3] Given the rarity of adult Wilms' tumour, this might best be addressed by specific treatment guidelines that take account of the increased toxicity for adults of regimens designed for children. Consensus guidelines for the treatment of adults with Wilms' tumour were agreed at the annual conference of the International Society of Paediatric Oncology (SIOP) in Cairo, Egypt, in 2003: these will be published shortly (available from kathy.pritchard-jones@icr.ac.uk).

Conflict of interest statement

None declared.

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REFERENCES

1. Parkin D, Kramarova E, Draper G, et al. *International incidence of childhood cancer*, vol. 2. Lyon: International Agency for Research on Cancer; 1998.
2. Reinhard H, Aliani S, Ruebe C, Stockle M, Leuschner I, Graf N. Wilms' tumor in adults: results of the Society of Pediatric Oncology (SIOP) 93-01/Society for Pediatric Oncology and Hematology (GPOH) Study. *J Clin Oncol* 2004;**22**:4500–6.
3. Kalapurakal JA, Nan B, Norkool P, et al. Treatment outcomes in adults with favorable histologic type Wilms' tumor-an update from the National Wilms' Tumor Study Group. *Int J Radiat Oncol Biol Phys* 2004;**60**:1379–84.
4. Terenziani M, Spreafico F, Collini P, et al. Adult Wilms' tumor: a monoinstitutional experience and a review of the literature. *Cancer* 2004;**101**:289–93.
5. Kattan J, Tournade MF, Culine S, Terrier-Lacombe MJ, Droz JP. Adult Wilms' tumour: review of 22 cases. *Eur J Cancer* 1994;**30A**:1778–82.
6. Berrino F, Capocaccia R, Coleman MP, et al. Survival of cancer in Europe: the EUROCARE-3 study. *Ann Oncol* 2003;**14**(suppl 5): v1–v155.
7. Berrino F, Capocaccia R, Estève J, et al. *Survival of cancer patients in Europe: the EUROCARE-2 study*. Lyon: International Agency for Research on Cancer; 1999.
8. Capocaccia R, Gatta G, Roazzi P, et al. The EUROCARE-3 database: methodology of data collection, standardisation, quality control and statistical analysis. *Ann Oncol* 2003;**14**(suppl 5):v14–27.
9. Berkson J, Gage RP. Calculation of survival rates for cancer. *Mayo Clin Proc* 1950;**25**:270–86.
10. Micheli A, Baili P, Quinn M, Mugno E, Capocaccia R, Grosclaude P. Life expectancy and cancer survival in the EUROCARE-3 cancer registry areas. *Ann Oncol* 2003;**14**(suppl 5): v28–40.
11. Hakulinen T, Abeywickrama KH. A computer program package for relative survival analysis. *Comput Programs Biomed* 1985;**19**:197–207.
12. Dickman PW, Sloggett A, Hills M, Hakulinen T. Regression models for relative survival. *Stat Med* 2004;**23**:51–64.
13. Fukuzawa R, Breslow NE, Morison IM, et al. Epigenetic differences between Wilms' tumours in white and east-Asian children. *Lancet* 2004;**363**:446–51.
14. Green DM. The treatment of stages I–IV favorable histology Wilms' tumor. *J Clin Oncol* 2004;**22**:1366–72.