



Embryonal cancers in Europe

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Abstract Embryonal cancers are a heterogeneous group of rare cancers which mainly occur in children and adolescents. The aim of the present study was to estimate the burden (incidence, prevalence, survival and proportion of cured) for the principal embryonal cancers in Europe (EU27), using population-based data from cancer registries (CRs) participating in RARECARE.

We identified 3322 cases diagnosed from 1995 to 2002 (latest period for which data are available): 44% neuroblastoma, 35% nephroblastoma, 13% retinoblastoma and 6% hepatoblastoma. Very few cases of pulmonary blastoma (43 cases) and pancreatoblastoma (seven cases) were diagnosed.

About 2000 new embryonal cancers were estimated every year in EU27, for an annual incidence rate of 4 per million (1.8 neuroblastoma, 1.4 nephroblastoma, and 0.5 retinoblastoma); 91% of cases occurred in patients under 15 years. Five-year relative survival for all embryonal cancers was 80% (99% retinoblastoma, 90% nephroblastoma, 71% hepatoblastoma and 68% neuroblastoma). Overall survival was lower in adolescents and adults than in those under 15 years. The cure rate was estimated at 80%. Slightly less than 40,000 persons were estimated alive in EU27 with a diagnosis of embryonal cancer in 2008. Nephroblastoma was the most prevalent (18,150 cases in EU27), followed by neuroblastoma (12,100), retinoblastoma (5200), hepatoblastoma (2700) and pulmonary blastoma (614).

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This is the first study to delineate the embryonal cancer burden in Europe by age, sex and European region. Survival/cure rate is generally high, but there are considerable gaps in our understanding of the natural histories of these rare diseases particularly in adults.

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1. Introduction

Embryonal cancers are a heterogeneous group of cancers whose cells are undifferentiated and resemble those found in developing embryos. The principal forms of embryonal tumour are neuroblastoma (including ganglioneuroblastoma), nephroblastoma (Wilms tumour), retinoblastoma, hepatoblastoma, pulmonary blastoma (including pleuropulmonary blastoma) and pancreatoblastoma.¹ Several studies indicate that the incidence of embryonal cancers, which occur mainly in children, is increasing.^{2–5} The risk of developing embryonal cancer is higher in children with certain genetic syndromes and congenital malformations^{6–8}; however these associated forms account for no more than 5% of all cases.

Environmental factors, such as ionising radiation, toxic therapies, herbicides, tobacco smoke and diet have been investigated as potential causes of embryonal cancer, particularly for exposure in the womb or at a very young age.^{2,4,9} Changing foetal growth conditions related to increasing age at first pregnancy, exposure to sex hormones and increasing birth weight have also been investigated.⁹

Survival for children with embryonal tumours has improved progressively over the last 40 years² as more effective treatments have become available.^{2,9}

In absolute terms, all embryonal cancers are rare; however neuroblastoma, nephroblastoma and retinoblastoma constitute 7.6%, 5.5% and 2.1%, respectively, of all paediatric cancers.² These are typical cancers of paediatric age and have been intensely studied: their clinical and biological characteristics are well known and numerous clinical trials have been conducted by

cooperative groups, resulting in the development of effective therapies. By contrast, hepatoblastoma, pulmonary blastoma and pancreatoblastoma are rare, even for childhood cancers.¹⁰ Nevertheless, they also have been investigated by cooperative research programmes either in the context of paediatric rare tumours as a group¹⁰, or as individual entities (<http://www.ppbregistry.org>). Studies coordinated by the International Childhood Liver Tumour Strategy Group (SIOPEL) on hepatoblastoma show that worldwide cooperation is feasible even for very rare cancers.¹¹ Embryonal cancers are extremely rare in adults and little is known of their epidemiologic and clinical behaviour.

The aim of the present study is to present population-based data for Europe on the incidence, prevalence and survival for the principal embryonal cancers: neuroblastoma ganglioneuroblastoma, nephroblastoma, retinoblastoma, hepatoblastoma, pulmonary blastoma and pancreatoblastoma. These forms are identified by the morphology codes of the International Classification of the Diseases for Oncology, third revision (ICD-O-3),¹² and are shown in the last column of Table 1. The study was carried out as part of the European Union (EU) co-funded project Surveillance of Rare Cancers in Europe (RARECARE).¹³

2. Cases and methods

RARECARE collected data on cancer patients diagnosed from 1978 to 2002, archived in 89 European population-based cancer registries (CRs), all of which had vital status information available at least up to 31st December 2003. The mean population covered by these CRs over the period 1995–1999 was about 162,000,000,

Table 1 Data quality indicators for malignant embryonal cancers diagnosed 1995–2002 and archived in 76 RARECARE cancer registries.

Entity	Number of cases diagnosed	Data quality indicator				ICD-O morphology code
		Death certificate only	Autopsy only	Microscopic verification	Cases diagnosed 1995–1998 censored before 5 years	
		(%)	(%)	(%)	(%)	
EMBRYONAL CANCERS	3322	0.5	0.4	93.9	2.4	
Neuroblastoma and ganglioneuroblastoma	1473	0.2	0.5	95.5	2.7	9490, 9500
Nephroblastoma	1162	0.4	0.2	95.6	2.4	8960
Retinoblastoma	445	0.7	0.0	82.9	1.8	9510–9514
Hepatoblastoma	192	2.1	1.0	95.8	2.1	8970
Pulmonary blastoma	43	0.0	4.7	97.7	2.3	8972–8973
Pancreatoblastoma	7	0.0	0.0	100.0	0.0	8971

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