The United Kingdom

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Content

Cancer is rare in childhood compared with older age groups, affecting approximately 1 in 600 children during the first 15 years of life. However, some tumours are so rare that even paediatric oncologists may only encounter them once in their lifetime practice. Any definition of 'rare' is bound to be arbitrary, but a suggested definition of rare childhood cancers are those categories in the International Classification of Childhood Cancer [third edition (ICCC-3)], that have an age-standardised annual incidence of less than 1 per million children in the UK, excluding tumours of unspecified morphology (Steliarova-Foucher et al. 2005). Based on the UK National Registry of Childhood Tumours (NRCT), Table 11.1 describes the incidence rates and numbers of registrations in Britain during 1991–2000 for rare childhood cancers according to this definition, excluding leukaemias, lymphomas and CNS tumours.

Histological subtypes of germ cell tumours which individually have incidence below 1 per million have also been excluded on the grounds that clinically all malignant germ cell tumours in children are treated similarly. Overall, the tumours listed in Table 11.1 had

- An incidence rate of 6.8 per million,
- Accounted for 16% of non-CNS malignant solid tumours
- Accounted 5% of all childhood cancers.
- In both relative and absolute terms they were most frequent in the age group 10–14 years, where their incidence was 12.4 per million and where they accounted for 35% of non-CNS solid tumours and 11% of all cancers.

Carcinomas of all sites counted as rare tumours, and collectively formed 50% of the total. Soft tissue sarcomas were the next most frequent histological

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Table 11.1 Rare childhood cancers in Great Britain, 1991–2000. Age standardized annual incidence per million children aged 0–14 years (ASR) and number of registrations (N). Leukaemias, lymphomas and CNS tumours are excluded

ICCC-3 Definition		ASR	N
IV	*Neuroblastoma and other peripheral nervous cell tumours*		
IVb	Peripheral nervous cell tumours other than neuroblastoma	0.10	11
VI	*Renal tumours*		
VIa.2	Rhabdoid renal tumour	0.36	24
VIa.3	Kidney sarcomas	0.36	24
VIa.4	Peripheral PNET of kidney	0.05	7
VIb	Renal carcinoma	0.16	19
VII	*Hepatic tumours*		
VIIb	Hepatic carcinoma	0.22	25
VIII	*Malignant bone tumours*		
VIIIb	Chondrosarcoma	0.10	12
VIIId	Other specified malignant bone tumours	0.14	16
IX	*Soft tissue and other extraosseous sarcomas*		
IXb.1	Fibroblastic and myoblastic tumours	0.35	37
IXb.2	Nerve sheath tumours	0.36	41
IXb.3	Other fibromatous neoplasms	0.02	2
IXc	Kaposi sarcoma	0.04	5
IXd.3	Extrarenal rhabdoid tumour	0.20	19
IXd.4	Liposarcomas	0.04	4
IXd.5	Fibrohistiocytic tumours	0.41	47
IXd.6	Leiomyosarcomas	0.10	12
IXd.7	Synovial sarcomas	0.49	58
IXd.8	Blood vessel tumours	0.11	11
IXd.9	Osseous and chondromatous neoplasms of soft tissue	0.08	9
IXd.10	Alveolar soft part sarcoma	0.09	10
IXd.11	Miscellaneous soft tissue sarcomas	0.18	19
X	*Germ cell tumours, trophoblastic tumours and neoplasms of gonads*		
Xd	Gonadal carcinomas	0.07	8
Xe	Other malignant non germ cell gonadal tumours	–	0
XI	*Other malignant epithelial neoplasms and malignant melanomas*		
XIa	Adrenocortical carcinoma	0.24	24
XIb	Thyroid carcinoma	0.60	71
XIc	Nasopharyngeal carcinoma	0.20	24
XIe	Skin carcinomas	0.70	82
XIf.1	Carcinomas of salivary glands	0.25	30
XIf.2	Carcinomas of colon and rectum	0.09	11
XIf.3	Carcinomas of appendix	0.12	15
XIf.4	Carcinomas of lung	0.08	10
XIf.5	Carcinomas of thymus	0.03	4
XIf.6	Carcinomas of breast	–	0
XIf.7	Carcinomas of cervix uteri	0.01	1
XIf.8	Carcinomas of bladder	0.05	6
XIf.9	Carcinomas of eye	–	0
XIf.10	Carcinomas of other specified sites	0.33	39
XIf.11	Carcinomas of unspecified site	0.11	12
XII	*Other and unspecified malignant neoplasms*		
XIIa.1	Gastrointestinal stromal tumour	0.02	2
XIIa.2	Pancreatoblastoma	0.04	4

Table 11.1 (continued)

ICCC-3 Definition		ASR	N
XIIa.3	Pulmonary blastoma and pleuropulmonary blastoma	0.08	8
XIIa.4	Other complex mixed and stromal neoplasms	–	0
XIIa.5	Mesothelioma	0.03	3
XIIa.6	Other specified malignant tumours	–	0
	Total of non-CNS rare tumours	*6.79*	*766*
	Total of all non-CNS solid tumours	*46.18*	*4,770*
	Total childhood cancers	*139.19*	*14,659*

Source: National Registry of Childhood Tumours (Stiller 2007)

group, representing 36%. It is important to note that the same diagnostic groups are not necessarily rare in all populations. Most strikingly, Kaposi sarcoma is one of the most frequent childhood cancers in parts of central and east Africa most severely affected by the AIDS epidemic, whereas malignant melanoma is rare throughout most of Africa and Asia.

Despite their rarity, the tumours described can cause much stress to both the families and the oncologist. However, rare tumours do not necessarily have a poor prognosis, and some tumour types may be easily treated and have very little chance of recurring. These include some tumours that are rare among children but occur more commonly in adults and we can learn a lot from their management in this setting so that this can be adapted for their treatment in childhood. A good example for this can be found for thyroid carcinoma (the follicular subtype is most commonly encountered in the paediatric population), where the 5-year survival for the 71 children diagnosed in Britain during 1991–2000 was 100% (Stiller 2007).

Other rare tumours, however, only occur in childhood or are currently have a poor prognosis. This focus therefore was the starting point for the Rare Tumour Guidelines that were produced by the Rare Tumour Working Group of the CCLG. In particular, we also included nasopharyngeal carcinoma, as this is one of the rare tumours frequently consulted about because of its challenging treatment, and as an example of a rare tumour where there has been a dramatic improvement in survival in recent years.

From about 1997 various members of the CCLG Rare Tumour Working Group took charge pulling together guidance for several rare tumours. The format consisted of the known data from the UK National Registry of Childhood Tumours (NRCT) and an up to date review of the literature using this to conclude guidance around management, diagnosis and treatment. Where possible information regarding open International registries/protocols was also made available. The list of the current Rare Tumour Guidelines available to members on the CCLG website or incorporated into study protocols or published are listed in Table 11.2.

Table 11.2 CCLG Rare Tumour Guidelines

Guideline	Author	Comments
Thymic epithelial tumours	Paula Shaw Richard Grundy Bernadette Brennan	CCLG website
Pleuropulmonary blastoma	Anthony Ng Julia Chisholm	Part of international collaboration on PPB
Nasopharyngeal Carinoma	Bernadette Brennan	CCLG website
Extracranial rhabdoid tumour	Bernadette Brennan	Part of EpSSG non rhabdomyosarcoma tumour protocol 2005.
Pancreatic tumours	Murray Yuile Bernadette Brennan	CCLG website
Adrenocortical tumours	Richard Grundy	CCLG website
CNS DNET Meningioma	Connor Mallucci Heidi Traunecker	British Journal Neurosurgery
Melanoma	Ross Pinkerton	CCLG website
Melanotic Neuroectodermal tumours	Helen Jenkinson	CCLG website

In 2005 a multi-disciplinary consensus statement of best practice for the management and treatment for paediatric endocrine tumours from a working group convened under the auspices of the BSPED (British Society of Paediatric Endocrinology and Diabetes) and CCLG (rare tumour working groups) was published as a booklet available to all members. The working group was multidisciplinary consisting

of paediatric endocrinologists, oncologists and surgeons together with adult surgeons, oncologists and clinical geneticists with paediatric expertise. The following endocrine tumours were covered in the booklet:

Craniopharyngioma
Adreno-cortical Neoplasms
Phaeochromocytoma
Thyroid Carcinoma (Differentiated)
Medullary Thyroid Carcinoma and Multiple Endocrine Neoplasia Type 2 (MEN 2) syndromes
Parathyroid and Pituitary Tumours (including primary hyperparathyroidism) and Multiple Endocrine Neoplasia Type 1 (MEN1) syndromes

Although the registration of these rare tumours continues in the UK in the NRCT for patients 15 years and under, it is less complete for older teenagers and contains limited details on treatment received and factors which maybe important for prognosis such as tumour dimensions, sites of metastases etc. The quality of the data may improve in the future with the National Cancer Dataset project run by the National Cancer Intelligence Network where the information collected in the UK cancer registries will increase for all cancers including rare tumours in childhood.

References

Steliarova-Foucher E et al (2005) International classification of childhood cancer, third edition (ICCC-3). Cancer 103(7):1457–1467

Stiller C (2007) Childhood cancer in Britain: incidence, survival, mortality. Oxford University Press, Oxford