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Childhood Cancer Incidence in the South-east of France

A Report of the Provence-Alpes-Côte d'Azur and Corsica Regions Pediatric Cancer Registry, 1984–1991

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A prospective registration of incident cancers in childhood in two south-east regions of France since 1 January 1984 allows us to collect pertinent data on 875 cases throughout a period of 8 years. World age-standardised overall incidence rate is 137.63 cases/million/year. It is close to that reported in other white European, North American and Oceanian populations. The age-adjusted (age-standardised) relative frequency of each pathological group is: leukaemias 29.71%; central nervous system tumours 20.61%; lymphomas 12.75%; sympathetic tumours 9.03%; soft tissues tumours 7.37%; bone tumours 5.89%; kidney tumours 4.82%; epithelial tumours 3.83%; germinal and gonadal tumours 3.24%; retinoblastomas 2.11%; liver tumours 0.45% and others 0.14%. The comparison of these results with international available data shows that we record the world highest adjusted incidence rates for neuroblastomas (15.46) and rhabdomyosarcomas (7.04) and a high rate for Ewing's sarcomas (3.30); this fact will need to be confirmed by a longer period of observation, but even now the total number of cases (particularly for neuroblastoma) is high when compared with the data of other children registries which give rates for longer periods and for similar or larger populations.
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INTRODUCTION

CHILDHOOD CANCERS account for only 2–3% of the total number of cancers but represent the second cause of mortality within this age group after accidents. The pattern of incidence of each individual type of morphology is strikingly different from what is seen in adults. It is, therefore, appropriate to set up specific paediatric population-based cancer registries established over sufficiently large areas, and to report separately cancer incidence rates in childhood.

MATERIALS AND METHODS

The Provence-Alpes-Côte d'Azur (PACA) and Corsica Regions Paediatric Cancer Registry started up on 1 January 1984 [1]. It runs a continuous prospective registration of incidental cases and is supported by Institut National de la Santé et de la Recherche Médicale, Direction Générale de la Santé, and Direction Régionale des Affaires Sanitaires et Sociales of the Provence-Alpes-Côte d'Azur region.

Criteria for registration

Four criteria are compulsory:

— The disease has to be malignant. In a lot of paediatric diseases, such as Langerhans cell histiocytosis or some cerebral, renal and soft tissue tumours, it is difficult to ascertain whether the tumour is malignant or not. An international agreement seems to be achieved on the basis of the Birch and Marsden proposal [2], and we applied these criteria in the present study.

— The date of the first sure diagnosis of disease has to be between 1 January 1984 and 31 December 1991.

— The age at diagnosis is from birth up to the 15th birthday; children are distributed in 15 classes, numbered 0 to 14; class "0" groups children under 1 year, class "1" those between 1 and 2 years, and so on.

— The child has to be normally resident in one of the two abovementioned administrative regions.

Data collection and checking

Data collection is both passive and active. All the childhood cancer centres of the south-east of France notify the registry of any new cases they are aware of and the registry itself requests data from each possible medical source periodically. Data on civil status, place of regular abode, morphology and location of the tumour, and the mean of diagnosis achievement are compulsory [1].

The notified cases are registered in a computer database specially developed on the 4° Dimension® software set up on a Macintosh II® from Apple Computers, Inc. This customised application automatically checks the registration criteria and the lack of double registration and ensures the suitable coding of morphology and location according to the International Classification of Diseases for Oncology [3]. Many physical and logical safety devices protect the file against unauthorised access. The file was declared to the French Vigilance Committee for Freedom and Information Processing (Commission Nationale

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pour l'Informatique et les Libertés), and the study was authorised by the regional medical ethical committee.

Population basis

The French Public Office of Statistics (Institut National des Statistiques et des Etudes Economiques) has taken two general population censuses in March 1982 and March 1990. A total of 824 314 and 837 057 children living in PACA and Corsica regions were counted, when the total population was about 4 600 000. We used a linear interpolation for the computation of annual incidence rates for intercensus years.

Data processing

The cases are grouped according to the guidelines proposed by Birch and Marsden [2]. These morphological groups and subgroups are coherent and significant for the paediatric oncologist and this scheme ought to be used in all paediatric registries' reports. Results are shown for four age classes: 0 (birth to 1 year), 1–4, 5–9, 10–14, and for the whole studied population 0–14. Age-standardised incidence rates (ASR) were calculated by the direct method using the World Standard Population [4] and expressed per million children and per year. Relative frequencies were calculated crude, without any correction, and age-standardised as for ASR [5, 6]. The cumulative incidence rate is the sum over each year of age of the ASR from birth to age 14. It gives an approximation of the risk for an individual to develop a cancer before the age of 15, if no other cause of death is in operation [7].

RESULTS

During the 8 years presently reported (1984–1991), 875 new cases were registered, the mean annual crude incidence was 132.41 and the mean annual age-standardised incidence was 137.63. The annual ASR fluctuated from 1984 to 1991 as following: 126, 131, 118, 140, 153, 162, 143 and 127 (Fig. 1).

Results for each group and subgroup by age class for the two administratives regions are shown in Tables 1a and b and 2a and b.

DISCUSSION

Most comparison material is drawn from the first comprehensive worldwide study of childhood cancer incidence by Parkin *et al.* in 1988 [8], and subsequent reports [9–12]. All the incidence rates quoted hereafter are age-standardised whereas the relative frequencies are both crude and age-standardised (ARF).

Global analysis

The reported mean annual overall incidence (137.63/million/year) is similar to that observed in other European, North American and Oceanian white populations. We observed some fluctuations of incidence from year to year with a progressive increase up to 162 in 1989, and a relative decrease ever since (Fig. 1). These variations will have to be interpreted within the European Collaborative Prospective Study which is in progress under the management of the International Agency for Research on Cancer [13].

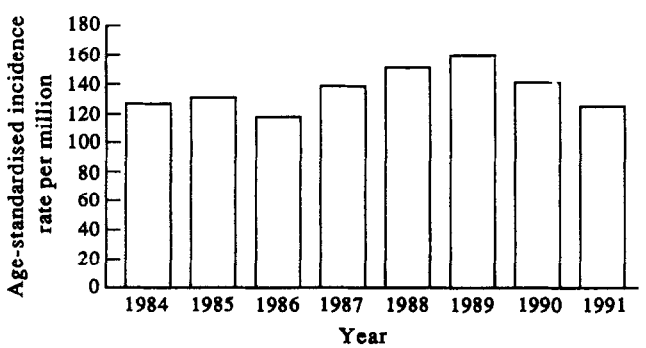

Fig. 1. Age-standardised incidence rates by year.

The age at diagnosis (Fig. 2) is similar to that generally reported, and it should be observed that 88 cases, namely 10%, occurred during the first year of life, which is the proportion expected [14].

The sex ratio (1.17) is as expected, but for a similar population and duration of registration, the Queensland register reported recently a very high (1.57) sex ratio [15].

Leukaemias

The main group is that of leukaemias, accounting for 30.86% (ARF 29.71%) of all the registered cases with an ASR of up to 43.59 per million; these rates are usual in white people, with a maximum in the hispanic populations of America (ASR > 50) [8, 9].

Acute lymphoblastic leukaemias represent the most frequent subgroup in our study (82% of all leukaemias), as in other white populations. Acute non-lymphoblastic leukaemias accounted for only 13.7% of leukaemia cases and chronic leukaemias constitute a very small minority—only 2% of cases.

The age distribution of acute lymphoblastic leukaemias shows a clear maximum between 2 and 5 years, as is usual in populations who benefit from a high socio economic level, whereas in developing populations, such as American or African blacks, the age distribution trends to be more regular throughout childhood [9, 16]; this fact could be the result of an under-registration because the clinical signs of leukaemias (infections, anaemia, haemorrhage) may be misinterpreted [9].

Leukaemias occur a little more often in girls than boys, which is unusual [8, 16, 17] and ought to be followed over a longer period of observation.

Lymphomas

The lymphoma group accounts for 11.77% (ARF 12.75%) of all registered cases and the ASR is 15.23. Lymphomas occur in the third rank among all tumours as in other Western populations, except in Latin America [18].

Nearly one quarter (24.2%) are Hodgkin's diseases, with an ASR of 3.45, while this rate is variable around the world (range 1–10). As usual, the majority of patients are males [8, 9], but with a particularly high sex ratio (2.57) and the age at onset is always beyond 5 years.

Among non-Hodgkin's lymphomas, the majority (52.5%) is represented by B-cell lymphomas, namely Burkitt's lymphomas, with an ASR of 6.07. This rate appears to be higher than that reported by the other European and North American paediatric cancer registries, but we think that this high rate may be related to a higher relevance in pathological classification in favour of Burkitt's type among the subgroup of non-Hodgkin's lymphomas.

Central nervous system tumours

These brain and spinal tumours constitute, with a relative frequency of 19.89% (ARF 20.61%), the second group after leukaemias. The ASR of 26.72 is near those reported in most other countries [20–30], if we exclude those with poor evaluation opportunities. ASR over 30 are found in three Nordic countries—Denmark, Finland and Sweden.

Table 1. *Effectives and relative frequencies by age and morphological group*

(a) PACA and Corsica boys 1984–1991

Diagnostic group	Number of cases					Relative frequency (%)		
	0	1–4	5–9	10–14	0–14	Crude	Adjusted	Group
Total	47	172	140	113	472	100.00	100.00	
1. Leukaemias	9	61	42	19	131	27.75	26.82	100.00
Acute lymphocytic	4	55	33	14	106	22.45	21.56	80.91
Other lymphocytic	0	1	2	0	3	0.63	0.61	2.29
Acute non-lymphocytic	4	2	5	4	15	3.17	3.22	11.45
Chronic myeloid	1	2	0	0	3	0.63	0.54	2.29
Other and unspecified	0	1	2	1	4	0.84	0.87	3.05
2. Lymphomas	4	15	31	29	79	16.73	17.81	100.00
Hodgkin's disease	0	0	10	8	18	3.81	4.26	22.78
Non-Hodgkin's lymphoma	0	3	3	4	10	2.11	2.25	12.65
Burkitt's lymphoma	0	6	18	12	36	7.62	8.13	45.56
Unspecified lymphoma	1	1	0	0	2	0.42	0.36	2.53
Histiocytosis X	3	5	0	1	9	1.90	1.72	11.39
Other reticuloendothelial	0	0	0	4	4	0.84	1.06	5.06
3. Central nervous system	4	34	28	29	95	20.12	20.63	100.00
Ependymoma	1	5	4	3	13	2.75	2.74	13.68
Astrocytoma	1	14	9	11	35	7.41	7.58	36.84
Medulloblastoma	2	7	8	7	24	5.08	5.21	25.26
Other glioma	0	3	1	3	7	1.48	1.55	7.36
Other and unspecified	0	5	6	5	16	3.38	3.52	16.84
4. Sympathetic nervous system	18	27	4	0	49	10.38	9.07	100.00
Neuroblastoma	18	27	4	0	49	10.28	9.07	100.00
Other	0	0	0	0	0	—	—	—
5. Retinoblastoma	5	9	0	0	14	2.96	2.55	—
6. Kidney tumours	2	11	2	1	16	3.38	3.06	100.00
Wilms' tumour	2	10	2	1	15	3.17	2.88	93.75
Renal carcinoma	0	0	0	0	0	—	—	—
Other and unspecified	0	1	0	0	1	0.21	0.18	6.25
7. Liver tumours	2	0	1	1	4	8.24	0.84	100.00
Hepatoblastoma	2	0	0	0	2	0.42	0.36	50.00
Hepatic carcinoma	0	0	1	1	2	0.42	0.47	50.00
Other and unspecified	0	0	0	0	0	—	—	—
8. Bone tumours	0	2	8	10	20	4.23	4.73	100.00
Osteosarcoma	0	0	4	8	12	2.54	2.98	60.00
Chondrosarcoma	0	0	0	0	0	—	—	—
Ewing's sarcoma	0	2	4	1	7	1.48	1.48	35.00
Other and unspecified	0	0	0	1	1	0.21	0.26	5.00
9. Soft tissue sarcomas	1	9	17	12	39	8.26	8.65	100.00
Rhabdomyosarcoma	1	9	13	5	28	5.93	5.93	71.79
Fibrosarcoma	0	0	3	6	9	1.90	2.23	23.07
Other and unspecified	0	0	1	1	2	0.42	0.47	5.12
10. Gonadal and germ cell tumours	2	3	1	3	9	1.90	1.92	100.00
Non-gonadal germ cell	2	1	0	3	6	1.27	1.34	66.66
Gonadal germ cell	0	2	1	0	3	0.63	0.57	33.33
Gonadal carcinoma	0	0	0	0	0	—	—	—
Other and unspecified	0	0	0	0	0	—	—	—
11. Epithelial neoplasms	0	1	6	8	15	3.17	3.59	100.00
Adrenocortical carcinoma	0	1	0	0	1	0.21	0.18	6.66
Thyroid carcinoma	0	0	2	0	2	0.42	0.42	13.33
Nasopharyngeal carcinoma	0	0	0	3	3	0.63	0.79	20.00
Melanoma	0	0	0	2	2	0.42	0.53	13.33
Other carcinomas	0	0	4	3	7	1.48	1.65	46.66
12. Other	0	0	0	0	0	0.21	0.26	—

Continued on next page.

Astrocytomas accounted for 40.2% of this group, medulloblastomas for 22.9% and ependymomas for 16%. Males are more often attacked than females, mainly in the subgroup of medulloblastomas (sex ratio 1.5). These results are similar to those in the literature.

Sympathetic nervous system tumours

These account for 10.51% (ARF 9.03%) of all registered cases. The incidence we observed (15.82 per million) is the highest reported until now, and almost all of these tumours are represented by neuroblastomas (97.8%) with an incidence of

Table 1. *Continued*

(b) PACA and Corsica girls 1984–1991

Diagnostic group	Number of cases					Relative frequency (%)		
	0	1–4	5–9	10–14	0–14	Crude	Adjusted	Group
Total	41	161	106	95	403	100.00	100.00	
1. Leukaemias	13	67	41	18	139	34.49	33.12	100.00
Acute lymphocytic	7	61	35	13	116	28.78	27.47	83.45
Other lymphocytic	0	0	0	0	0	—	—	—
Acute non-lymphocytic	4	5	5	5	19	4.71	4.77	13.66
Chronic myeloid	1	0	0	0	1	0.24	0.19	0.71
Other and unspecified	1	1	1	0	3	0.74	0.67	2.15
2. Lymphomas	5	2	6	11	24	5.95	6.55	100.00
Hodgkin's disease	0	0	1	6	7	1.73	2.17	29.16
Non-Hodgkin's lymphoma	0	0	1	3	4	0.99	1.23	16.66
Burkitt's lymphoma	0	2	2	1	5	1.24	1.27	20.83
Unspecified lymphoma	0	0	0	0	0	—	—	—
Histiocytosis X	5	0	2	0	7	1.73	1.55	29.16
Other reticuloendothelial	0	0	0	1	1	0.24	0.31	4.16
3. Central nervous system	5	25	23	26	79	19.60	20.66	100.00
Ependymoma	3	4	4	4	15	3.72	3.78	18.98
Astrocytoma	2	10	10	13	35	8.68	9.31	44.30
Medulloblastoma	0	8	2	6	16	3.97	4.04	20.25
Other glioma	0	2	3	1	6	1.48	1.56	7.59
Other and unspecified	0	1	4	2	7	1.73	1.96	8.86
4. Sympathetic nervous system	8	28	7	0	43	10.66	9.10	100.00
Neuroblastoma	8	26	7	0	41	10.17	8.71	95.34
Other	0	2	0	0	2	0.49	0.39	4.65
5. Retinoblastoma	1	6	1	0	8	1.98	1.66	—
6. Kidney tumours	3	23	6	0	32	7.94	6.84	100.00
Wilms' tumour	2	23	6	0	31	7.69	6.64	96.87
Renal carcinoma	0	0	0	0	0	—	—	—
Other and unspecified	1	0	0	0	1	0.24	0.19	3.12
7. Liver tumours	0	0	0	0	0	—	—	—
Hepatoblastoma	0	0	0	0	0	—	—	—
Hepatic carcinoma	0	0	0	0	0	—	—	—
Other and unspecified	0	0	0	0	0	—	—	—
8. Bone tumours	1	0	12	12	25	6.20	7.38	100.00
Osteosarcoma	0	0	3	4	7	1.73	2.11	28.00
Chondrosarcoma	0	0	1	0	1	0.24	0.28	4.00
Ewing's sarcoma	0	0	8	8	16	3.97	4.79	64.00
Other and unspecified	1	0	0	0	1	0.24	0.19	4.00
9. Soft tissue sarcomas	3	6	5	8	22	5.45	5.72	100.00
Rhabdomyosarcoma	3	3	5	7	18	4.46	4.81	81.81
Fibrosarcoma	0	2	0	0	2	0.49	0.39	9.09
Other and unspecified	0	1	0	1	2	0.49	0.51	9.09
10. Gonadal and germ cell tumours	2	4	4	8	18	4.46	4.84	100.00
Non-gonadal germ cell	2	4	2	2	10	2.48	2.38	55.55
Gonadal germ cell	0	0	1	5	6	1.48	1.86	33.33
Gonadal carcinoma	0	0	0	0	0	—	—	—
Other and unspecified	0	0	1	1	2	0.49	0.59	11.11
11. Epithelial neoplasms	0	0	1	12	13	3.22	4.07	100.00
Adrenocortical carcinoma	0	0	0	0	0	—	—	—
Thyroid carcinoma	0	0	1	1	2	0.49	0.59	15.38
Nasopharyngeal carcinoma	0	0	0	3	3	0.74	0.94	23.07
Melanoma	0	0	0	4	4	0.99	1.26	30.76
Other carcinomas	0	0	0	4	4	0.99	1.26	30.76
12. Other	0	0	0	0	0	—	—	—

up to 15.46. Neuroblastoma is thus the most frequent tumour of the embryonic type in our series. The sex ratio is 1.19 as expected.

Our rates need, of course, to be confirmed over a longer period of observation, but the total number of cases (90) is already high when compared to other reports. This ASR of 15.46 based on 90 cases observed throughout a period of 8 years and among a population of 830 000 children has to be compared with the data reported, for example, by the Piemont (ASR 13.5/73 cases/20 years/806 000 children) [18], England and Wales (7.0/650 cases/

Table 2. Crude. age-standardised and cumulative incidence rates. Percentage of histologically verified diagnoses (HV)

(a) PACA and Corsica boys 1984–1991

Diagnostic group	Annual incidence rates per million 0	1–4	5–9	10–14	0–14	Adjusted	Cumulative	HV %
Total	202.59	192.62	125.48	98.45	139.30	144.40	2 092.82	98
1. Leukaemias	38.79	68.31	37.64	16.55	38.66	41.10	583.06	100
Acute lymphocytic	17.24	61.59	29.57	12.19	31.28	33.49	472.51	100
Other lymphocytic	—	1.11	1.79	—	0.88	0.92	13.44	100
Acute non lymphocytic	17.24	2.23	4.48	3.48	4.42	4.48	66.03	100
Chronic myeloid	4.31	2.23	—	—	0.88	1.02	13.27	100
Other and unspecified	—	1.11	1.79	0.87	1.18	1.17	17.79	100
2. Lymphomas	17.24	16.79	27.78	25.26	23.31	22.83	349.70	100
Hodgkin's disease	—	—	8.96	6.97	5.31	4.91	79.66	100
Non-Hodgkin's lymphoma	—	3.35	2.68	3.48	2.95	2.91	44.30	100
Burkitt's lymphoma	—	6.71	16.13	10.45	10.62	10.32	159.82	100
Unspecified lymphoma	4.31	1.11	—	—	0.59	0.68	8.79	100
Histiocytosis X	12.93	5.59	—	0.87	2.65	2.98	39.68	100
Other reticuloendothelial	—	—	—	3.48	1.18	1.01	17.42	100
3. Central nervous system	17.24	38.07	25.09	25.26	28.03	28.55	421.37	92
Ependymoma	4.31	5.59	3.58	2.61	3.83	3.98	57.70	100
Astrocytoma	4.31	15.67	8.06	9.58	10.32	10.57	155.28	97.1
Medulloblastoma	8.62	7.83	7.17	6.09	7.08	7.17	106.32	100
Other glioma	—	3.35	0.89	2.61	2.06	2.08	30.98	100
Other and unspecified	—	5.59	5.37	4.35	4.72	4.73	71.07	56.3
4. Sympathetic nervous system	77.58	30.23	3.58	—	14.46	16.52	216.47	98
Neuroblastoma	77.58	30.23	3.58	—	14.46	16.52	216.47	98
Other	—	—	—	—	—	—	—	—
5. Retinoblastoma	21.55	10.07	—	—	4.13	4.78	61.87	100
6. Kidney tumours	8.62	12.31	1.79	0.87	4.72	5.31	71.21	100
Wilms' tumour	8.62	11.19	1.79	0.87	4.42	4.96	66.73	100
Renal carcinoma	—	—	—	—	—	—	—	—
Other and unspecified	—	1.11	—	—	0.29	0.34	4.47	100
7. Liver tumours	8.62	—	0.89	0.87	1.18	1.20	17.45	100
Hepatoblastoma	8.62	—	—	—	0.59	0.66	8.62	100
Hepatic carcinoma	—	—	0.89	0.87	0.59	0.54	8.83	100
Other and unspecified	—	—	—	—	—	—	—	—
8. Bone tumours	—	2.23	7.17	8.71	5.90	5.53	88.37	100
Osteosarcoma	—	—	3.58	6.97	3.54	3.18	52.77	100
Chondrosarcoma	—	—	—	—	—	—	—	—
Ewing's sarcoma	—	2.23	3.58	0.87	2.06	2.10	31.24	100
Other and unspecified	—	—	—	0.87	0.29	0.25	4.35	100
9. Soft tissue sarcomas	4.31	10.07	15.23	10.45	11.51	11.40	173.09	100
Rhabdomyosarcoma	4.31	10.07	11.65	4.35	8.26	8.47	124.67	100
Fibrosarcoma	—	—	2.68	5.22	2.65	2.38	39.58	100
Other and unspecified	—	—	0.89	0.87	0.59	0.54	8.83	100
10. Gonadal and germ cell tumours	8.62	3.35	0.89	2.61	2.65	2.75	39.61	100
Non-gonadal germ cell	8.62	1.11	—	2.61	1.77	1.77	26.16	100
Gonadal germ cell	—	2.23	0.89	—	0.88	0.98	13.44	100
Gonadal carcinoma	—	—	—	—	—	—	—	—
Other and unspecified	—	—	—	—	—	—	—	—
11. Epithelial neoplasms	—	1.11	5.37	6.97	4.42	4.10	66.22	100
Adrenocortical carcinoma	—	1.11	—	—	0.29	0.34	4.47	100
Thyroid carcinoma	—	—	1.79	—	0.59	0.57	8.96	100
Nasopharyngeal carcinoma	—	—	—	2.61	0.88	0.75	13.06	100
Melanoma	—	—	—	1.74	0.59	0.50	8.71	100
Other carcinomas	—	—	3.58	2.61	2.06	1.91	30.99	100
12. Other	—	—	—	0.87	0.29	0.25	4.35	100

Continued on next page.

10 years/11 million), and Denmark (9.6/42 cases/5 years/1.1 million) registries [8].

Incidently, it is interesting to note that the high ASR for neuroblastomas in France mentioned until now in the literature [11, 12] and extracted from the IARC monography [8], is mainly imputable to PACA and Corsica regions.

Retinoblastomas

In our study, the incidence of retinoblastoma (3.83 per million) is in the middle range of those reported in other occidental countries [2–6]. The data available on the one- or two-sided character of the registered tumours are unfortunately insufficient in most studies as in ours, and no inference can be

Table 2. Continued

(b) PACA and Corsica girls 1984–1991

Diagnostic group	Annual incidence rates per million 0	1–4	5–9	10–14	0–14	Adjusted	Cumulative	HV %
Total	183.92	189.24	100.07	87.42	125.17	130.50	1 878.39	98
1. Leukaemias	58.31	78.75	38.70	16.56	43.17	46.19	649.69	100
Acute lymphocytic	31.40	71.70	33.04	11.96	36.03	38.76	543.23	100
Other lymphocytic	—	—	—	—	—	—	—	—
Acute non-lymphocytic	17.94	5.87	4.72	4.60	5.90	6.06	88.06	100
Chronic myeloid	4.48	—	—	—	0.31	0.34	4.48	100
Other and unspecified	4.48	1.17	0.94	—	0.93	1.01	13.90	100
2. Lymphomas	22.42	2.35	5.66	10.12	7.45	7.23	110.77	100
Hodgkin's disease	—	—	0.94	5.52	2.17	1.90	32.32	100
Non-Hodgkin's lymphoma	—	—	0.94	2.76	1.24	1.10	18.52	100
Burkitt's lymphoma	—	2.35	1.88	0.92	1.55	1.60	23.44	100
Unspecified lymphoma	—	—	—	—	—	—	—	—
Histiocytosis X	22.42	—	1.88	—	2.17	2.34	31.87	100
Other reticuloendothelial	—	—	—	0.92	0.31	0.26	4.60	100
3. Central nervous sytem	22.42	29.38	21.71	23.92	24.53	24.78	368.17	95
Ependymoma	13.45	4.70	3.77	3.68	4.65	4.78	69.55	100
Astrocytoma	8.97	11.75	9.44	11.96	10.87	10.85	163.01	91.4
Medulloblastoma	—	9.40	1.88	5.52	4.96	5.12	74.66	100
Other glioma	—	2.35	2.83	0.92	1.86	1.90	28.16	100
Other and unspecified	—	1.17	3.77	1.84	2.17	2.11	32.78	85.7
4. Sympathetic nervous system	35.88	32.91	6.60	—	13.35	15.10	200.57	95.3
Neuroblastoma	35.88	30.56	6.60	—	12.73	14.37	191.17	95
Other	—	2.35	—	—	0.62	0.72	9.40	100
5. Retinoblastoma	4.48	7.05	0.94	—	2.48	2.83	37.41	100
6. Kidney tumours	13.45	27.03	5.66	—	9.93	11.24	149.91	100
Wilms' tumour	8.97	27.03	5.66	—	9.62	10.89	145.43	100
Renal carcinoma	—	—	—	—	—	—	—	—
Other and unspecified	4.48	—	—	—	0.31	0.34	4.48	100
7. Liver tumours	—	—	—	—	—	—	—	—
Hepatoblastoma	—	—	—	—	—	—	—	—
Hepatic carcinoma	—	—	—	—	—	—	—	—
Other and unspecified	—	—	—	—	—	—	—	—
8. Bone tumours	4.48	—	11.32	11.04	7.76	7.20	116.34	100
Osteosarcoma	—	—	2.83	3.68	2.17	1.98	32.56	100
Chrondrosarcoma	—	—	0.94	—	0.31	0.30	4.72	100
Ewing's sarcoma	—	—	7.55	7.36	4.96	4.57	74.57	100
Other and unspecified	4.48	—	—	—	0.31	0.34	4.48	100
9. Soft tissue sarcomas	13.45	7.05	4.72	7.36	6.83	6.88	102.08	100
Rhabdomyosarcoma	13.45	3.52	4.72	6.44	5.59	5.52	83.37	100
Fibrosarcoma	—	2.35	—	—	0.62	0.72	9.40	100
Other and unspecified	—	1.17	—	0.92	0.62	0.63	9.30	100
10. Gonadal and germ cell tumours	8.97	4.70	3.77	7.36	5.59	5.50	83.47	89
Non-gonadal germ cell	8.97	4.70	1.88	1.84	3.10	3.29	46.42	80
Gonadal germ cell	—	—	0.94	4.60	1.86	1.64	27.72	100
Gonadal carcinoma	—	—	—	—	—	—	—	—
Other and unspecified	—	—	0.94	0.92	0.62	0.57	9.32	100
11. Epithelial neoplasms	—	—	0.94	11.04	4.03	3.51	59.93	100
Adrenocortical carcinoma	—	—	—	—	—	—	—	—
Thyroid carcinoma	—	—	0.94	0.92	0.62	0.57	9.32	100
Nasopharyngeal carcinoma	—	—	—	2.76	0.93	0.80	13.80	100
Melanoma	—	—	—	3.68	1.24	1.06	18.40	100
Other carcinomas	—	—	—	3.68	1.24	1.06	18.40	100
12. Other	—	—	—	—	—	—	—	—

drawn concerning the relative parts of hereditary and sporadic forms.

Kidney tumours

Nearly all the kidney tumours are nephroblastomas in our series (95.83%), as they are throughout the world. The other cases are one clear cell sarcoma and one rhabdoid sarcoma. The incidence of nephroblastomas has been considered for a long time as uniform all over the world, but it is now clear that this assertion was not correct and that the incidence is strongly linked to ethnic groups [1]. The rate we report here for nephroblastomas (7.85) is very near the rates observed in most other white people

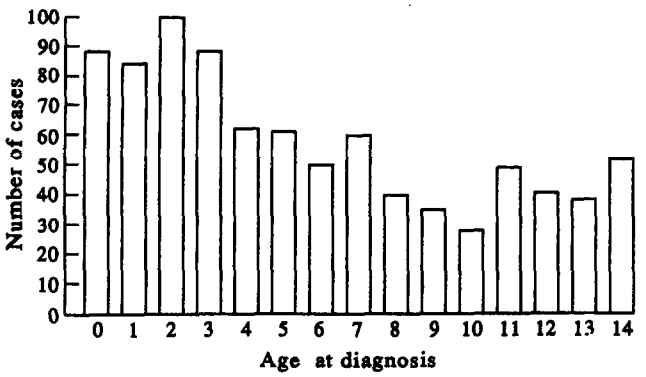

Fig. 2.

in Europe, America and Australia, but the incidence is somewhat higher in a few European registries (Finland 11.1, France-Bas Rhin 11.7) [8].

Liver tumours

Their incidence is very low: 0.61 per million, with 2 cases of hepatoblastoma and 2 cases of hepatocarcinoma. This level of incidence applies to the majority of studies except for countries where the role of the infection by the hepatitis B virus in the emergence of these tumours has been well established. One of our cases of hepatocarcinoma arose in a 10-year-old boy who had immigrated from Cambodia 2 years before the onset and had a history of chronic B hepatitis.

Bone tumours

The observed rate (6.35) is in the range reported by the other Western pediatric registries. These bone tumours are nearly evenly distributed between osteosarcomas (42%—ASR 2.59) and Ewing's sarcomas (51%—ASR 3.30); the remaining types are represented by chondrosarcomas and fibrosarcomas. Highest ASR for Ewing's sarcoma are observed in Saarland (3.7), New Zealand Maoris (3.3), PACA and Corsica (3.3), and Denmark (3.2). These top ASR are based on 16, 5, 23 and 18 cases, respectively.

Soft tissue sarcomas

The usual rates for soft tissue sarcomas range from 5 to 9 per million. In our series we found a high ASR of up to 9.2, the most important subgroup (75.4%) being rhabdomyosarcomas, with an ASR of 7.04 based on 46 cases. This ASR is the highest reported in children as yet, but it has to be confirmed over a longer observation period and one must consider that the pathological subclassification among the group of soft tissue tumours may often differ between the pathologists.

Germinal and gonadal tumours

We report a rate of 4.09 for this group, which is in the usual range. Only one third of these tumours are located in testis or ovary.

Epithelial tumours

This group of tumours accounts for only 28 cases and 3.2% (ARF 3.83%) of all cases, which represents an ASR of 3.81. The main subgroups are undifferentiated nasopharyngeal carcinomas (21%), melanomas (21%) and thyroid carcinomas (14%).

Our rate for undifferentiated carcinoma of the nasopharynx (0.77) appears to be slightly higher than in comparable studies (0.1–0.5), perhaps because of the North Africa extraction of nearly 10% of our population.

The rate we report here for melanoma (0.77) is close to those published in Western countries. The highest rates are reported in Australia (4.6). Our rate for thyroid carcinoma appears to be relatively low (0.57) compared with those reported in Italy (1.3), in North America (1.3 to 1.7) for white people [8] and in Norway (1) [22], but for all these data, it must be mentioned that the effects are very small.

CONCLUSION

The world age-standardised overall incidence rate we report here (137.63 per million and by year) is in the middle range of those observed in similar populations of Western countries. We cannot comment at this time on the annual fluctuations of incidence because of the insufficient period of observation and, moreover, they must be analysed together with other European data.

The analysis of age-standardised incidence rates for individual groups and subgroups shows that we record the world highest rates for neuroblastomas and rhabdomyosarcomas and a high rate for Ewing's sarcomas. These peculiarities have to be confirmed by a longer survey but warrant the setting up of specific analytic studies even now.

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Incidence, Survival and Mortality in Cervical Cancer in Norway, 1956–1990

Tone Bjørge, Steinar Ø. Thoresen and Gry B. Skare

Long-term trends in incidence, survival and mortality were examined in women with squamous cell carcinoma and adenocarcinoma of the uterine cervix, diagnosed in Norway in the 35-year period 1956–1990. During the 1970s the number of cervical smears increased substantially in Norway, although no organised screening programme was introduced. Special attention was paid to the time period 1971–1990 to evaluate the effect of the extensive spontaneous screening. In addition, the prognostic importance of clinical stage and age was explored. In the squamous cell carcinoma patients the incidence rate peaked in the time period 1971–1975, since when there has been a decrease. In the adenocarcinoma patients the incidence rate rose through the years 1976–1990. Also, the proportion of adenocarcinomas increased in this time period. The mortality rates in both histological types declined modestly through the years 1966–1990. A more favourable stage distribution was noted among the squamous cell carcinomas ($P = 0.00$), but not among the adenocarcinomas, when comparing the two diagnostic periods 1971–1975 and 1981–1985. The multivariate analysis (GLIM) revealed that stage was the most important prognostic factor in both histological types ($P = 0.00$). In the squamous cell carcinoma patients the relative rate increased ($P = 0.04$) in the last period. There was a tendency towards a poorer prognosis in younger women in this group, but age did not prove to be an important prognostic factor ($P = 0.08$).
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INTRODUCTION

THE INCIDENCE of uterine cervical cancer in Norway rose from the 1950s to the mid 1970s, after which a decline began. At present, carcinoma of the cervix is the third most frequent malignancy of the female genital tract. Annually about 400 new cases of cervical cancer are diagnosed [1], and approximately 130 women die from the disease [2].

The numbers of cytological smears have been steadily increasing since 1970, but no organised screening programme has been introduced. The decrease in incidence has been less pronounced in Norway compared to the other Nordic countries of Finland and Sweden, where organised screening programmes have been run by the governments since about 1960 [3]. The changes in incidence rates reflect the mass screening intensity in each country. The screening programmes have also had a major impact on reductions in mortality rates [4].

Squamous cell carcinoma and adenocarcinoma are the two most common cervical malignancies. The relative incidence of cervical adenocarcinoma appears to be increasing [5], whereas the incidence of squamous cell carcinoma is declining. The cytological screening programmes being implemented in several western countries during the last 40 years have been ineffective in detecting adenocarcinoma compared to its squamous counterpart [6].

The prognosis of cervical cancer has been described in several clinical series [5, 7–9]. Fewer data from unselected series are available [10–12]. Clinical stage and screening programme attendance are important factors with respect to survival. The prognostic importance of age and histological type is, however, still undetermined.

The aim of the present study was to examine long-term trends in incidence, relative survival and mortality rates in women with squamous cell carcinoma and adenocarcinoma of the uterine cervix, diagnosed in Norway in the 35-year period 1956–1990. Special attention was paid to the time period 1971–1990. In the early 1970s relatively few cervical smears were taken, while in the 1980s the spontaneous wild screening was rather extensive. In addition, the prognostic importance of clinical stage and age was evaluated.

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