EPIDEMIOLOGY

Characteristics of Cancer Patients Under Age 20 at a Population-Based Registry, Puerto Rico, 1980-1991

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Objectives. **To evaluate and describe the characteristics and incidence of pediatric (<20 years) cancer in Puerto Rico between 1980-1991.**

Methods. **A descriptive study of all new childhood cancer cases among residents of Puerto Rico reported to the Cancer Registry between the years 1980 and 1991 was conducted. The distribution of cases was described by age, gender, place of residence, histology, last date known to be alive, and stage of disease. Incidence rates and standardized incidence rates by age were calculated.**

Results. **A total of 1,823 new pediatric cases were reported from 1980 through 1991. Over half were males, and 52.2% were aged <10 years. Most cases resided in urban areas. The most frequent histologic types were leukemia (31.0%), lymphomas and other reticulo-endothelial neoplasms (16.2%), and brain, central nervous system, and intracranial and intraspinal neoplasms (16.0%). The most frequent type of tumor was acute lymphocytic leukemia (22.0%). Although the overall distribution of most neoplasms was similar between genders, a higher proportion of lymphomas was reported among males, and there were significantly more carcinomas among girls. The largest increase in the age-adjusted incidence rate between periods (1980-84 and 1985-89) occurred in the 5-9 age group.**

Conclusions. **Overall, these findings are consistent with previous studies in USA. However, there were some differences in the distribution of histologic types when compared with similar studies in other countries, and with the results of the National Cancer Institute's Surveillance, Epidemiology and End Results (SEER) Program. Factors such as differences in diagnostic capabilities among countries must be taken into consideration.** *Key words: Neoplasm, Pediatric, Epidemiology, Puerto Rico, Incidence, Childhood, Adolescence, Cancer.*

There is considerable variation in childhood cancer rates between developing and developed countries. However, once an individual has survived the first five years of life, cancer becomes one of the major causes of death in developing countries as it is in developed countries. (1) International comparisons of cancers can be a valuable tool to investigate the role of sociocultural and environmental factors. Despite the fact that approximately 65-70% of children with cancer in industrialized nations survive the disease, the societal impact of childhood cancer is devastating, ranking third in the number of potential years of life lost. (2)

In Puerto Rico, as in the Unites States, cancer is a leading cause of death among children/adolescents between ages 1 through 19, second only to accidents. (3) Thus, it is expected that the incidence trends of the majority of childhood neoplasms in Puerto Rico may be similar to developed countries. Accurate statistics on the occurrence of cancer are available in Puerto Rico through a population-based cancer registry existing since 1950. However, a long-term description in childhood cancer has not been reported. This study presents a detailed description of the characteristics of cancer cases under age 20 in Puerto Rico between 1980 through 1991.

Methods

Incident cases of cancer under 20 years of age were obtained from the Puerto Rico Central Cancer Registry

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(Cancer Control Program, Department of Health). This population-based registry was established in March 1950, and gathers detailed information on all cases among Puerto Rican residents. (4) Until 1990, this Registry was one of the ten registries used by the National Cancer Institute's Surveillance, Epidemiology and End Results (SEER) Program to monitor the occurrence of cancer in the United States. The cases included in the present study were all cancer patients under the age of 20 years diagnosed between the years 1980 and 1991, who resided in Puerto Rico. The selection of these years was based on the completeness of records during that period. During this period, 78.6% of the cases were born in Puerto Rico.

Since 1976, the Registry uses the International Classification of Diseases for Oncology (ICD-O) to classify and code the morphology of the neoplasms. (5) These ICD-O codes were grouped by histologic type and site according to an international classification scheme for childhood cancer developed by Marsden, which groups tumors in twelve major histologic groups. (6) Distributed among these twelve major categories are 42 subgroups. The distribution of cases was described by age, gender, place of residence, primary site, histology, last date known to be alive, and stage of disease.

The majority (95.7%) of the 1,823 cases with a histological diagnosis were microscopically confirmed either with a positive histology (95.3%) or with positive exfoliative cytology (0.4%). An additional 2.0% were confirmed using X-rays or other imaging techniques, 0.3% were confirmed using clinical diagnosis only, and 0.1% used direct visualization. In 1.9% of the cases it was unknown whether or not it was microscopically confirmed. In this study, the histologic type provided was used regardless of the fact that 4.3% of these diagnoses had no microscopic confirmation.

The observed proportions of neoplasms (by histologic type) were compared by gender and residence area using the z test statistic. Significance was set at $\alpha=0.05$. Incidence rates during each year of diagnosis were calculated using population figures from the Census (1980 and 1990), and intercensal population estimates provided in the Vital Statistics Annual Report. (7) Also, rates were standardized by age (direct method) using as standard the 1980 Census population for Puerto Rico.

Results

From 1980 through 1991, a total of 1,850 new pediatric cancer cases were reported to the Cancer Registry, including twenty-seven (27) children who were classified as non-residents and who were excluded from this analysis. Approximately 10.1% of the 1,823 cases diagnosed in this period occurred in 1981. Most of these 1,823 cases (62.0%) were not reported dead as of February 1996.

Over half (52.3%) of the 1,823 children diagnosed with cancer were males (Table 1), 52.2% were aged <10 years, 18.7% were between 10-14, and 29.1% were adolescents aged 15 to 19 years (Table 2). Children 5-9 accounted for 20.9%, 1 to 4 years old accounted for 25.6%, and 5.7% of the cases were under one year of age. Overall, most of the cases resided in urban areas (58.5%) (Table 3).

Table 1. Distribution of Incident Pediatric (< 20 years) Cancer Cases by Histologic Type and Gender, Puerto Rico, 1980-1991

Histologic Type	Gender				Total	
	Male		Female			
	n	%	n	%	n	%
I. Leukemias	**302**	**31.7**	**264**	**30.3**	**566**	**31.0**
Acute lymphocytic leukemia	212	22.3	189	21.7	401	22.0
Other lymphoid leukemia	4	0.4	4	0.5	8	0.4
Acute non-lymphocytic leukemia	64	6.7	54	6.2	118	6.5
Chronic myeloid leukemia	4	0.4	7	0.8	11	0.6
Other and unspecified leukemia	18	1.9	10	1.1	28	1.5
II. Lymphomas and other reticuloendothelial neoplasms*	**192**	**20.2**	**103**	**11.8**	**295**	**16.2**
Hodgkin's disease‡	98	10.3	61	7.0	159	8.7
Non-Hodgkin's lymphoma†	48	5.0	21	2.4	69	3.8
Burkitt's lymphoma	18	1.9	10	1.1	28	1.5
Unspecified lymphomas	18	1.9	8	0.9	26	1.4
Histiocytosis X	2	0.2	0	0.0	2	0.1
Other reticuloendothelial neoplasms	8	0.8	3	0.3	11	0.6
III. CNS and misc. intracranial and intraspinal neoplasms	**154**	**16.2**	**136**	**15.6**	**291**	**16.0**
Ependymoma	13	1.4	20	2.3	33	1.8
Astrocytoma	67	7.0	74	8.5	141	7.7
Medulloblastoma	36	3.8	20	2.3	57	3.1
Other glioma‡	28	2.9	13	1.5	41	2.2
Miscellaneous intracranial and intraspinal neoplasms	10	1.1	9	1.0	19	1.0
IV. Sympathetic nervous system tumors	**40**	**4.2**	**38**	**4.4**	**78**	**4.3**
Neuroblastoma and ganglioneuroblastoma	39	4.1	37	4.3	76	4.2
Other	1	0.1	1	0.1	2	0.1
V. Retinoblastoma	**16**	**1.7**	**15**	**1.7**	**31**	**1.7**
VI. Renal tumors	**32**	**3.4**	**44**	**5.1**	**76**	**4.2**
Wilms' tumor	30	3.2	43	4.9	73	4.0
Renal carcinoma	0	0.0	1	0.1	1	0.1
Other and unspecified malignant renal tumors	2	0.2	0	0.0	2	0.1

(Table 1 Continued)

Table 1. Distribution of Incident Pediatric (< 20 years) Cancer Cases by Histologic Type and Gender, Puerto Rico, 1980-1991

Histologic Type	Gender				Total	
	Male		Female			
	n	%	n	%	n	%
VII. Hepatic tumors	**6**	**0.6**	**3**	**0.3**	**9**	**0.5**
Hepatoblastoma	4	0.4	1	0.1	5	0.3
Hepatic carcinoma	2	0.2	1	0.1	3	0.2
Other and unspecified malignant hepatic tumors	0	0.0	1	0.1	1	0.1
VIII. Malignant bone tumors	**63**	**6.6**	**46**	**5.3**	**109**	**6.0**
Osteosarcoma	30	3.2	27	3.1	57	3.1
Chondrosarcoma	5	0.5	2	0.2	7	0.4
Ewing's sarcoma	22	2.3	15	1.7	37	2.0
Other and unspecified malignant bone tumors	6	0.6	2	0.2	8	0.4
IX. Soft-tissue sarcomas	**82**	**8.6**	**62**	**7.1**	**144**	**7.9**
Rhabdomyosarcoma/embryonal sarcoma/soft-tissue Ewing's	35	3.7	28	3.2	63	3.5
Fibrosarcoma/neurofibrosarcoma /other fibromatous neoplasms	19	2.0	15	1.7	34	1.9
Other soft-tissue sarcoma	28	2.9	19	2.2	47	2.6
X. Germ-cell, trophoblastic and other gonadal neoplasms†	**27**	**2.8**	**50**	**5.7**	**77**	**4.2**
Non-gonadal germ-cell and trophoblastic neoplasms	8	0.8	14	1.6	22	1.2
Gonadal germ-cell and trophoblastic neoplasms	19	2.0	28	3.2	47	2.6
Gonadal carcinoma†	0	0.0	7	0.8	7	0.4
Other and unspecified malignant gonadal tumor	0	0.0	1	0.1	1	0.1
XI. Carcinoma and other malignant epithelial neoplasms*	**29**	**3.0**	**105**	**12.1**	**134**	**7.4**
Thyroid carcinoma*	10	1.1	44	5.1	54	3.0
Nasopharyngeal carcinoma	2	0.2	0	0.0	2	0.1
Melanomatous neoplasms	3	0.3	9	1.0	12	0.7
Other carcinoma*	11	1.5	50	6.0	66	3.6
XII. Other and unspecified malignant neoplasms	**9**	**0.9**	**4**	**0.5**	**13**	**0.7**
Total	**952**	**100.0**	**870**	**100.0**	**1,823**	**100.0**

Source: Puerto Rico Central Cancer Registry; Contents of groups are based on International Classification Scheme (HB Marsden, 1988).
* p≤0.001; † p≤0.01; ‡ p≤0.05

The distribution of childhood cancer by histologic type showed that leukemias were the most common neoplasms with 31.0% of the cases, followed by lymphomas and other reticuloendothelial neoplasms (16.2%), and brain, central nervous system, or intracranial and intraspinal neoplasms (16.0%) (Table 1). Acute lymphocytic leukemia (ALL) accounted for 70.8% of childhood leukemia. ALL was the most common diagnosis in children aged <10 (31.0%) and those aged 10-14 years

Table 2. Distribution of Incident Pediatric (< 20 years) Cancer Cases by Histologic Type and Age Group, Puerto Rico, 1980-1991

Histologic Type	Age Group (years)					
	<10		10-14		15-19	
	n	%	n	%	n	%
I. Leukemias	**369**	**38.8**	**78**	**22.9**	**119**	**22.5**
Acute lymphocytic leukemia	295	31.0	44	12.9	62	11.7
Other lymphoid leukemia	6	0.6	0	0.0	2	0.4
Acute non-lymphocytic leukemia	50	5.3	27	7.9	41	7.7
Chronic myeloid leukemia	3	0.3	3	0.9	5	0.9
Other and unspecified leukemia	15	1.6	4	1.2	9	1.7
II. Lymphomas and other reticuloendothelial neoplasms	**101**	**10.6**	**76**	**22.3**	**118**	**22.3**
Hodgkin's disease	39	4.1	41	12.0	79	14.9
Non-Hodgkin's lymphoma	22	2.3	18	5.3	29	5.5
Burkitt's lymphoma	20	2.1	7	2.1	1	0.2
Unspecified lymphomas	12	1.3	10	2.9	4	0.8
Histiocytosis X	2	0.2	0	0.0	0	0.0
Other reticuloendothelial neoplasms	6	0.6	0	0.0	5	0.9
III. CNS and misc. intracranial and intraspinal neoplasms	**169**	**17.8**	**72**	**21.1**	**50**	**9.4**
Ependymoma	25	2.6	5	1.5	3	0.6
Astrocytoma	72	7.6	41	12.0	28	5.3
Medulloblastoma	36	3.8	15	4.4	6	1.1
Other glioma	27	2.8	7	2.1	7	1.3
Miscellaneous intracranial and intraspinal neoplasms	9	0.9	4	1.2	6	1.1
IV. Sympathetic nervous system tumors	**70**	**7.4**	**5**	**1.5**	**3**	**0.6**
Neuroblastoma and ganglioneuroblastoma	70	7.4	4	1.2	2	0.4
Other	0	0.0	1	0.3	1	0.2
V. Retinoblastoma	**31**	**3.3**	**0**	**0.0**	**0**	**0.0**
VI. Renal tumors	**67**	**7.0**	**7**	**2.1**	**2**	**0.4**
Wilms' tumor	65	6.8	7	2.1	1	0.2
Renal carcinoma	0	0.0	0	0.0	1	0.2
Other and unspecified malignant renal tumors	2	0.2	0	0.0	0	0.0
VII. Hepatic tumors	**7**	**0.7**	**1**	**0.3**	**1**	**0.2**
Hepatoblastoma	5	0.5	0	0.0	0	0.0
Hepatic carcinoma	1	0.1	1	0.3	1	0.2
Other and unspecified malignant hepatic tumors	1	0.1	0	0.0	0	0.0
VIII. Malignant bone tumors	**23**	**2.4**	**41**	**12.0**	**45**	**8.5**
Osteosarcoma	6	0.6	23	6.7	28	5.3
Chondrosarcoma	1	0.1	2	0.6	4	0.8
Ewing's sarcoma	13	1.4	14	4.1	10	1.9
Other and unspecified malignant bone tumors	3	0.3	2	0.6	3	0.6

(Table 2 Continued)

Table 2. Distribution of Incident Pediatric (< 20 years) Cancer Cases by Histologic Type and Age Group, Puerto Rico, 1980-1991

Histologic Type	Age Group (years)					
	<10		10-14		15-19	
	n	%	n	%	n	%
IX. Soft-tissue sarcomas	**70**	**7.4**	**24**	**7.0**	**50**	**9.4**
Rhabdomyosarcoma/embryonal sarcoma/soft-tissue Ewing's	39	4.1	8	2.3	16	3.0
Fibrosarcoma/neurofibrosarcoma /other fibromatous neoplasms	14	1.5	5	1.5	15	2.8
Other soft-tissue sarcoma	17	1.8	11	3.2	19	3.6
X. Germ-cell, trophoblastic and other gonadal neoplasms	**28**	**2.9**	**13**	**3.8**	**36**	**6.8**
Non-gonadal germ-cell and trophoblastic neoplasms	14	1.5	1	0.3	7	1.3
Gonadal germ-cell and trophoblastic neoplasms	14	1.5	10	2.9	23	4.3
Gonadal carcinoma	0	0.0	1	0.3	6	1.1
Other and unspecified malignant gonadal tumor	0	0.0	1	0.3	0	0.0
XI. Carcinoma and other malignant epithelial neoplasms	**11**	**1.1**	**20**	**5.9**	**103**	**19.4**
Thyroid carcinoma	8	0.8	8	2.3	38	7.2
Nasopharyngeal carcinoma	0	0.0	0	0.0	2	0.4
Melanomatous neoplasms	1	0.1	2	0.6	9	1.7
Other carcinoma	2	0.2	10	2.9	54	10.2
XII. Other and unspecified malignant neoplasms	**6**	**0.6**	**4**	**1.2**	**3**	**0.6**
Total	**952**	**100-.0**	**341**	**100.0**	**530**	**100.0**

Source: Puerto Rico Central Cancer Registry; Contents of groups are based on International Classification Scheme (HB Marsden, 1988).

Table 3. Distribution of Incident Pediatric (< 20 years) Cancer Cases by Histologic Type and Residence Area, Puerto Rico, 1980-1991

Histologic Type*	Residence Area			
	Urban		Rural	
	n	%	n	%
I. Leukemias	274	31.9	204	33.4
II. Lymphomas and other reticuloendothelial neoplasms	138	16.1	97	15.9
III. CNS and misc. intracranial and intraspinal neoplasms	144	16.8	96	15.7
IV. Sympathetic nervous system tumors	36	4.2	26	4.3
V. Retinoblastoma	13	1.5	8	1.3
VI. Renal tumors	31	3.6	23	3.8
VII. Hepatic tumors	2	0.2	5	0.8
VIII.Malignant bone tumors	47	5.5	41	6.7
IX.Soft-tissue sarcomas	71	8.3	40	6.6
X.Germ-cell, trophoblastic and other gonadal neoplasms	35	4.1	30	4.9
XI.Carcinoma and other malignant epithelial neoplasms	63	7.3	35	5.7
XII.Other and unspecified malignant neoplasms	5	0.6	6	0.8
Total	**859**	**100.0**	**610**	**100.0**

Source: Puerto Rico Central Cancer Registry; Contents of groups are based on International Classification Scheme (HB Marsden , 1988).
* No significant differences were found.

(12.9%). Hodgkin's disease was the most common cancer in children aged 15-19 (14.9%), and was relatively low in the <10 age group (4.1%). Astrocytoma was the most frequent brain tumor in all age groups, but was higher in the 10-14 age group (12.0%). All retinoblastomas and almost all neuroblastoma cases were reported exclusively in the <10 age group. The majority of the renal tumors were Wilms' tumors, and occurred mainly in the <10 age group (6.8%). However, osteosarcoma was the most common bone tumor, and the majority of the cases were reported in the 15-19 age group (5.3%). Other tumors that were frequent in the <10 age group were astrocytomas (7.6%) and neuroblastoma and ganglioneuroblastomas (7.4%), while in the 10-14 age group Hodgkin's disease accounted for 12.0%. However, acute lymphocytic leukemia (11.7%) and thyroid and other carcinomas were frequent in the 15-19 age groups.

The overall distribution by histologic type of most neoplasms by gender was very similar (Table 1). However, a significantly higher proportion of lymphomas and other reticuloendothelial neoplasms were reported among males (20.2% vs. 11.8%), specially Hodgkin's disease and non-Hodgkin's lymphoma. Among females, there were significantly more carcinomas and other epithelial neoplasms (12.1% vs. 3.0%), specially thyroid carcinoma, and more germ-cell, trophoblastic and other gonadal neoplasms (5.7% vs. 2.8%), specially gonadal carcinoma.

Carcinomas were most frequently encountered in endocrine glands (42.6%), in reproductive organs (23.3%), in digestive organs (11.6%), and in the skin (8.5%). Almost all endocrine gland carcinomas were detected in the thyroid gland (98.2%), and most of the reproductive organ carcinomas were located in the cervix uteri (83.3%).

The distribution of most cancer types by residence area (urban and rural) was very similar (Table 3). However, urban areas seemed to have a slightly higher proportion of soft tissue sarcomas (8.3% vs. 6.6%), and carcinomas (7.3% vs. 5.7%), but these differences were not statistically significant ($p>0.05$).

During the period of 1980-1991 the incidence of childhood cancer in Puerto Rico was 11.5 per 100,000 children. It fluctuated between 9.8 per 100,000 in 1983, the year with the lowest incidence, and 13.6 per 100,000 children in 1981, the year with the largest rate (Table 4).

Table 4. Age-adjusted Incidence Rates by Histologic Type, Puerto Rico, 1980-1991

Histologic Type	Age-adjusted Incidence Rates per Year (per 1,000,000 children-year)*												
	1980	1981	1982	1983	1984	1985	1986	1987	1988	1989	1990	1991	**Total**
I. Leukemias	33.4	40.8	31.9	32.1	36.1	37.8	36.0	40.2	33.4	36.8	32.4	42.6	**35.7**
II. Lymphomas and other reticulo-endothelial neoplasms	20.8	21.4	21.3	13.3	12.1	17.2	25.6	17.5	16.6	17.7	19.9	18.1	**18.6**
III. CNS and intracranial and intraspinal neoplasms	14.9	15.5	20.0	14.8	25.6	18.8	20.5	20.5	15.3	20.0	13.0	23.5	**18.4**
IV. Sympathetic nervous system tumors	4.5	6.7	4.5	3.0	1.7	4.6	8.5	6.2	5.7	7.3	5.0	3.2	**4.9**
V. Retinoblastoma	2.2	4.5	1.5	2.3	3.4	1.5	0.8	2.4	1.6	2.5	0.8	0.8	**2.0**
VI. Renal tumors	6.7	6.7	6.0	6.0	4.2	3.8	5.4	2.4	4.0	4.8	4.8	4.0	**4.8**
VII. Hepatic tumors	0.7	1.5	-	1.5	0.8	-	-	1.6	-	-	-	0.8	**0.6**
VIII. Malignant bone tumors	6.7	8.8	6.6	2.9	6.1	5.9	7.5	4.5	11.3	10.6	4.6	6.0	**6.9**
IX. Soft-tissue sarcomas	8.2	10.3	8.1	6.7	12.3	11.3	9.1	11.4	6.9			7.9	**9.1**
X. Germ-cell, trophoblastic and other gonadal neoplasms	5.2	4.4	1.5	3.7	4.6	5.9	6.8	6.9	3.8	2.3	5.6	7.6	**4.9**
XI. Carcinoma and other malignant epithelial neoplasms	4.5	13.2	8.1	10.3	10.3	10.4	6.0	5.3	8.3	7.6	8.4	7.6	**8.5**
XII. Other and unspecified malignant neoplasms	2.2	2.2	-	1.5	0.8	-	0.8	-	0.7	0.8	-	0.8	**0.8**
Total	**109.9**	**135.9**	**109.5**	**98.1**	**117.4**	**116.4**	**126.4**	**118.1**	**107.4**	**117.9**	**102.0**	**122.0**	**115.1**

Source: Puerto Rico Central Cancer Registry; Contents of groups are based on International Classification Scheme (HB Marsden, 1988).
* Age adjustment using as standard the 1980 Puerto Rico Census.

The age-adjusted incidence rates were not discernibly different from the crude rates.

The age-standardized incidence rate of leukemia per year varied between 3.2 per 100,000 in 1982 and 4.3 per 100,000 in 1991 (Table 4). The overall incidence rate of leukemia for the study period was 3.6/100,000. For lymphomas, the range was between 1.2/100,000 in 1984 and 2.6/100,000 in 1986. The overall incidence rate was 1.9/100,000 for the study period.

The age-adjusted incidence rates of pediatric cancer in Puerto Rico increased 20.7% in the 1985-1989 period compared with the 1980-1984 period (rates increased from 11.4/100,000 to 13.8/100,000). The largest change (32.9%) occurred in the 5-9 age group: rates increased from 9.1/100,000 to 12.0/100,000. The <5 age group experienced a 23.4% relative increase (14.7 to 18.1/ 100,000), the 10-14 group had a 15.6% increase (from 8.7 to 10.0), and the 15-19 group had a 12.8% increase (13.3 to 15.0).

Discussion

The distribution of the majority of childhood cancers in this study are similar to previous national and international findings, particularly with the results from the National Cancer Institute's Surveillance Epidemiology and End Results (SEER) Program. Total childhood cancer rates among Puerto Ricans have been considered low to moderate level. (8) In general, differences in total childhood cancer rates among 22 selected populations of the five continents were relatively small. In contrast, some differences emerge when comparisons are made by histologic type and anatomic site. (8) In this study, leukemia was the most common diagnosis in children of all ages, accounting for almost a third (31%) of the cases. This result is comparable to USA who reported leukemia diagnosis in 31.4% of their microscopically confirmed cases during 1973-1987. (9-11)

Throughout Europe, the Americas, Asia and Oceania, the age-standardized incidence of leukemia between 1970-1979 was usually in the range of 15-50 per million, but the highest incidence rate of leukemia was recorded in Costa Rica (59.4) and Hispanics residing in Los Angeles (50.2). (8) Puerto Rico's incidence rate of leukemia was in the range of 33.4 to 42.6 per million. During that period it was reported that Puerto Rico had a lower incidence than the predominantly non-Hispanic whites covered by SEER in the mainland USA (40.2 vs. 43.7).

It must be noted that the proportion of ALL reported in Puerto Rico was lower than in the USA (71% vs. 84%). (8) This difference may be related to the changes in diagnostic tests. The proportion of SEER cases of retinoblastoma, malignant bone tumors, germ-cell/ trophoblastic/ gonadal neoplasms and carcinomas/ other malignant neoplasms were also similar to our results. However, SEER informed a slightly higher proportion of brain, central nervous system, intracranial and intraspinal neoplasms (17.0% vs.16.0%).

Intracranial and spinal cord tumors are the second most frequent type of childhood cancer after leukemias, accounting for approximately 20% of cases in many regions of the world. (12) The highest age-adjusted

incidence rate for those neoplasms (31.4 per million) has been reported in the Nordic countries and Canada. This has been attributed to the possession of sophisticated universal health care systems that may reflect their efficacy in disease surveillance. (12) On the other hand, the lowest rates have been reported in the Chinese population and for blacks in Africa (both below 15 per million). (13) The incidence rate in Puerto Rico fluctuated between 13.0 and 25.6 per million.

Puerto Rico had a slightly lower proportion of hepatic tumors than U.S. (0.5% vs. 1.3%). The age-standardized incidence of hepatic tumors in Puerto Rico showed slight variations through the years with the lowest incidence rate reported in 1980 (0.7/1,000,000) and the largest in 1987 (1.6/1,000,000). The incidence of hepatoblastoma appears to be relatively constant throughout the world, with rates between 0.5 and 1.5 per million. Chronic infection of the liver by hepatitis B virus (HBV) has been reported as the main cause of hepatocellular carcinomas in the world, and is responsible for several hundred thousands deaths each year. (14-15) A study conducted in Turkey in 1994 revealed that HBV seropositivity in pediatric oncologic patients was 19.6%, whereas in the general population was 8-11%. (16) HBV seroprevalence in Puerto Rican children was not available.

Lymphomas are the second most common malignancy after leukemias in several Latin American and Asian countries among children. (8) Although the proportion of lymphomas and other reticuloendothelial neoplasms was larger in Puerto Rico (16.2%) than in the SEER results (12.4%), the age-adjusted incidence rate per year was slightly higher in the USA. The high incidence of lymphomas, particularly in childhood Hodgkin's diseases in developing countries, has been correlated with an infectious etiology. (17)

When the distribution of pediatric cancer by histologic type was compared with results in other countries, some interesting differences emerged. For example, Puerto Rico reported a larger proportion of lymphomas, carcinomas and other malignant epithelial neoplasms than France. (11) However, France reported a larger proportion of CNS, intracranial and intraspinal neoplasms, and sympathetic nervous system tumors. International variations in the distribution of cancer could provide clues regarding the etiology of the disease and the mechanisms of how different factors, such as genetic and environmental, contribute to its development. However, the issue of different diagnostic capabilities among countries is an important factor that must be considered.

The problem in studying trends over time in cancer cases is well recognized. For example, there have been changes in diagnostic testing such as cell surface markers in leukemia introduced in 1975, CT scans and MRIs in the 1980's, introduction of the FAB classification of leukemia, increasing number of specialists over time, and reductions in the under five mortality rate overall. Each of these issues will affect the incidence rates and, as a consequence, the trends over time.

After a careful analysis of our data we concluded that the 12-year interval covered is too short and the number of cases were too small for studying time trends in our series. However, some peculiarities of the data were discussed that may be of general interest.

When comparing the 1985-1989 period with the 1980-1984 period, the overall rate of pediatric cancer in Puerto Rico increased by 21%, an average of 2% per year. The largest percentage of change was in the 5-9 age group (33%). Although changes in the incidence rate representing a difference of few cases should be interpreted with caution, changes that do not reach statistical significance still may be clinically important. Increasing temporal trends in cancer incidence in children has been recently reported in USA, England, Scotland, Germany and Australia (18-24). In particular, a report of trends in childhood cancer incidence between 1974 and 1991 from nine population-based registries in USA, showed that among children aged <14 there was a 1% average annual increase in the incidence rates of all malignant neoplasms combined (25). But the increase was most apparent among young children and within specific histologic types (1.6% for ALL per year, and 2% for CNS tumor, rhabdomyosarcomas, germ cell tumors and osteosarcomas).

The population of Puerto Rico may be exposed to environmental factors that may be potential risk factors for childhood cancer either through direct exposure or through parent occupational exposure. Pharmaceutical companies, indiscriminate use of domestic and agriculture pesticides, hydrocarbons, ionizing radiation, contaminated wells, transport, utilization and storage of hazardous waste are, among others, potential carcinogens present in the environment. However, the large proportion of neoplasms (31.3%) in the first four years of age among Puerto Rican children underscore the possibility of a genetic causation. The genetic causation of childhood cancer, particularly in this age group, has been successfully illustrated (26). Further analysis using a longer period of time may be useful to study time trends of pediatric cancer in Puerto Rico.

This study demonstrates that Puerto Rico experienced incidence rates for most of the cancer types similar to USA and other developed countries. However, the proportions of some specific childhood cancers, such as Hodgkin's disease, were larger in Puerto Rico as compared

to those reported in the SEER results. Temporal trends in cancer incidence in children living in Puerto Rico remain to be determined.

Resumen

Se llevó a cabo un estudio descriptivo de todos los casos pediátricos (<20 años) de cáncer entre residentes de Puerto Rico reportados al Registro de Cáncer entre 1980 y 1991. La distribución de los casos se describió por edad, sexo, lugar de residencia, histología, última fecha reportado vivo y etapa de la enfermedad. Se calcularon tasas de incidencia crudas y ajustadas por edad. Entre 1980-91, se reportaron 1,823 nuevos casos pediátricos. Los tipos histológicos más frecuentes fueron leucemia (31.0%), linfomas u otros neoplasmas reticulo-endoteliales (16.2%) y neoplasmas del cerebro, sistema nervioso central e intracraniales/intraespinales (16.0%). Se reportó una mayor proporción de linfomas entre varones y hubo significativamente más carcinomas entre niñas. El aumento mayor en las tasas de incidencia entre periodos (1980-84 y 1985-89) ocurrió en el grupo de 5-9 años. Hubo diferencias en la distribución de tipos histológicos al comparar con otros países y con los resultados del Programa SEER del Instituto Nacional de Cáncer.

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