

## Sopravvivenza dei bambini con tumore maligno diagnosticato in Italia negli anni 1998-2002

## Survival of Italian children with childhood cancer diagnosed in 1998-2002

La prognosi dei bambini affetti da tumore maligno diagnosticato nei primi 15 anni di vita è migliorata in tutti i paesi a standard di vita occidentale nel corso degli ultimi 3-4 decenni. In queste popolazioni i tumori maligni (comprese le leucemie) rappresentano tuttavia ancora la seconda causa di morte dopo gli incidenti nella classe d'età 1-14 anni, essendosi ridotta la mortalità per malattie infettive.

Nella tabella 1 è riportata la distribuzione dei bambini residenti nelle aree servite da un registro tumori con tumore maligno diagnosticato nel periodo 1998-2002, suddivisi per genere e per tipo di neoplasia (secondo la International Childhood Cancer Classification - ICCC), con le relative percentuali di sopravvivenza cumulativa a 1, 3 e 5 anni dalla diagnosi. La sopravvivenza cumulativa per tutti i tumori a un anno dalla diagnosi è circa del 90% per i maschi e per le femmine, riducendosi progressivamente nei 4 anni successivi del 10% circa. A 5 anni dalla diagnosi, i bambini con leucemia (linfatica e non linfatica acuta), linfoma (Hodgkin e non-Hodgkin), retinoblastoma e tumore renale mostrano la prognosi migliore, mentre i bambini con tumore cerebrale, sarcomi ossei e del connettivo registrano percentuali di sopravvivenza cumulativa inferiori. Non si rilevano differenze statisticamente significative di sopravvivenza tra maschi e femmine per nessun tipo di tumore tranne che per gli ependimomi (analisi basata su 42 casi).

La sopravvivenza per i bambini delle diverse classi di età alla diagnosi mostra invece differenze marcate. In particolare, i bambini di età 1-9 anni con leucemia linfatica acuta presentano una prognosi migliore rispetto ai bambini di età inferiore all'anno e superiore a 10 anni; i bambini con neuroblastoma di età inferiore a 1 anno presentano una prognosi migliore rispetto ai casi diagnosticati dopo l'anno. Questi aspetti peculiari, che verosimilmente sottendono diversità biologiche ed eziologiche, sono discussi con maggiore approfondimento nelle schede specifiche per tipo tumorale. Nella tabella 2 sono riportate le stime di sopravvivenza per area geografica di residenza dei bambini. La distribuzione disomogenea dei registri tumori operanti nelle tre grandi aree geografiche (Nord, Centro e Sud Italia) influisce ovviamente sulla distribuzione dei casi e sulla stabilità statistica delle stime. Non si rilevano differenze di sopravvivenza statisticamente significative tra le aree geografiche, fatta eccezione per i casi affetti da leucemia non linfatica acuta (analisi basata su

*The prognosis of children affected by childhood cancer diagnosed under age 15 has shown a marked improvement in all industrialised countries in the last 3-4 decades. Nevertheless in the same countries malignant neoplasms (inclusive of leukaemia) are the second leading cause of death in children aged 1-14, after accidental deaths, as infectious diseases are no longer a frequent cause of mortality.*

*Table 1 presents the number cases of neoplasm incident in 1998-2002 in children living in the areas covered by one of the Italian cancer registries and the corresponding cumulative survival percentages at 1, 3 and 5 years after diagnosis, by gender and tumour type. Tumour types are grouped according to the International Childhood Cancer Classification (ICCC) and the presentation is limited to the major categories and to a selection of the minor ones. Overall, cumulative survival percentage at 1 year from diagnosis was approximately 90% and showed a progressive reduction of about 10% in the following 4 years for both boys and girls. Survival rates at 5 years from diagnosis were superior for children affected by leukaemia (both acute lymphoblastic, ALL and acute non-lymphoblastic, AnLL), lymphoma (both Hodgkin's and non-Hodgkin's), retinoblastoma and kidney neoplasm. On the contrary, cumulative survival was inferior for children affected by neoplasm of the central nervous system (CNS), bone and soft tissue sarcoma. No gender related differences were observed, except for ependymoma (42 cases were included in the analysis).*

*Large differences were observed in survival by age class. In particular, children affected by ALL diagnosed at age 1-9 fared better than children younger or older at diagnosis and children with neuroblastoma diagnosed under 1 year of age fared better than children diagnosed at older age. These issues are presented in greater details in the sections dedicated to the specific tumour types.*

*Table 2 presents survival figures by area of residence. The large differences in the number of cases reflected the uneven distribution of Cancer Registries, which are more numerous in the North and Centre of Italy and fewer in number in the South. Statistical precision of estimates is correspondingly affected. No statistically significant differences were observed by geographical area, with the exception of ANLL (results based on 70, 17 e 16 cases resident in the areas covered by cancer registration in North, Centre and South).*

*Table 3 shows the cumulative survival percentages for Italian*

	Gender Genere	n.	Cumulative survival Sopravvivenza cumulativa			Log-rank test
			1 y (95%CI)	3 y (95%CI)	5 y (95%CI)	
<b>TUTTI I TUMORI</b>	M	1,195	90.4 (88.8-92.1)	81.4 (79.1-83.6)	79.1 (76.7-81.6)	0.4787
ALL TUMOURS TYPES	F	947	91.3 (89.4-93.1)	80.6 (78.1-83.2)	77.0 (74.1-79.8)	
<b>I LEUCEMIE</b>	M	375	92.4 (89.7-95.1)	84.6 (80.9-88.4)	81.6 (77.3-85.8)	0.4236
LEUKAEMIA	F	295	92.8 (89.9-95.8)	86.6 (82.6-90.6)	83.7 (79.2-88.2)	
<b>IA LEUCEMIA LINFATICA ACUTA</b> ACUTE LYMPHOCYTIC LEUKAEMIA	M	299	94.9 (92.4-97.4)	88.2 (84.4-91.9)	85.7 (81.4-90.1)	0.6546
	F	236	95.3 (92.6-98.0)	89.8 (85.8-93.8)	86.7 (82.1-91.4)	
<b>IB LEUCEMIA NON-LINFATICA ACUTA</b> ACUTE NON-LYMPHOCYTIC LEUKAEMIA	M	56	77.8 (66.7-88.9)	66.6 (53.5-79.8)	60.5 (46.0-74.9)	0.1285
	F	47	89.1 (80.1-98.1)	79.6 (67.7-91.5)	76.6 (63.7-89.4)	
<b>II LINFOMI E NEOPLASIE RETICULOENDOTELIALI</b> LYMPHOMA AND RETICULOENDOTHELIAL NEOPLASMS	M	224	93.8 (90.6-96.9)	90.1 (86.1-94.0)	89.5 (85.4-93.6)	0.4416
	F	115	96.5 (93.2-99.9)	85.7 (79.2-92.2)	85.7 (79.2-92.2)	
<b>IIA LINFOMA DI HODGKIN</b> HODGKIN'S LYMPHOMA	M	85	100.0 (100.0-100.0)	95.2 (90.6-99.8)	93.5 (87.9-99.1)	0.5603
	F	60	100.0 (100.0-100.0)	91.0 (83.5-98.5)	91.0 (83.5-98.5)	
<b>IIIB LINFOMA NON-HODGKIN</b> NON-HODGKIN'S LYMPHOMA	M	73	90.4 (83.7-97.2)	84.8 (76.5-93.1)	84.8 (76.5-93.1)	0.6362
	F	29	96.6 (89.9-100.0)	86.2 (73.7-98.8)	86.2 (73.7-98.8)	
<b>IIC LINFOMA DI BURKITT</b> BURKITT'S LYMPHOMA	M	46	89.1 (80.1-98.1)	89.1 (80.1-98.1)	89.1 (80.1-98.1)	0.5241
	F	11	90.9 (73.9-100.0)	81.8 (59.0-100.0)	81.8 (59.0-100.0)	
<b>III TUMORI DEL SISTEMA NERVOSO CENTRALE</b> CNS AND MISCELLANEOUS INTRACRANIAL AND INTRASPINAL NEOPLASMS	M	239	80.9 (75.9-85.9)	66.3 (60.2-72.4)	65.6 (59.4-71.8)	0.3007
	F	172	79.9 (73.9-85.9)	64.7 (57.5-72)	57.6 (49.1-66.2)	
<b>IIIA EPENDIMOMA</b> EPENDYMOMA	M	24	100.0 (100.0-100.0)	76.9 (58.8-94.9)	76.9 (58.8-94.9)	0.0132
	F	18	72.2 (51.5-92.9)	60.2 (37.2-83.2)	32.1 (8.3-55.9)	
<b>IIIB ASTROCITOMA</b> ASTROCYTOMA	M	77	85.4 (77.5-93.4)	75.9 (66.2-85.6)	75.9 (66.2-85.6)	0.5261
	F	66	89.1 (81.5-96.7)	71.2 (59.9-82.5)	67.5 (54.6-80.3)	
<b>IIIC TUMORI NEUROEPITELIALI PRIMITIVI</b> PRIMITIVE NEUROECTODERMAL TUMOURS	M	53	79.2 (68.3-90.2)	63.3 (50.1-76.5)	63.3 (50.1-76.5)	0.9162
	F	32	77.4 (62.7-92.1)	71.0 (55.0-86.9)	63.1 (42.7-83.4)	
<b>IIID ALTRI GLIOMI</b> OTHER GLIOMAS	M	24	75.0 (57.7-92.3)	61.5 (41.6-81.4)	61.5 (41.6-81.4)	0.7639
	F	16	81.3 (62.1-100.0)	56.3 (31.9-80.6)	56.3 (31.9-80.6)	
<b>IIIF TUMORI INTRACRANICI</b> E INTRASPINALI NON SPECIFICATI UNSPECIFIED INTRACRANIAL AND INTRASPINAL NEOPLASMS	M	60	70.8 (59.1-82.5)	55.1 (42.3-68.0)	52.5 (39.3-65.7)	0.6677
	F	37	67.6 (52.5-82.7)	51.4 (35.2-67.5)	51.4 (35.2-67.5)	
<b>IV TUMORI DEL SISTEMA NERVOSO SIMPATICO</b> SYMPATHETIC NERVOUS SYSTEM TUMOURS	M	87	93.1 (87.8-98.4)	76.0 (66.8-85.3)	67.9 (56.6-79.1)	0.5697
	F	73	90.4 (83.7-97.2)	80.0 (70.5-89.4)	73.7 (62.5-84.8)	
<b>V RETINOBLASTOMA</b> RETINOBLASTOMA	M	22	95.5 (86.8-100.0)	95.5 (86.8-100.0)	95.5 (86.8-100.0)	0.8068
	F	15	100.0 (100.0-100.0)	92.9 (79.4-100.0)	92.9 (79.4-100.0)	
<b>VI TUMORI RENALI</b> RENAL TUMOURS	M	47	97.9 (93.7-100.0)	88.0 (78.2-97.9)	88.0 (78.2-97.9)	0.9351
	F	62	96.7 (92.1-100.0)	91.5 (84.5-98.6)	87.3 (78.5-96.2)	
<b>VII TUMORI EPATICI</b> HEPATIC TUMOURS	M	12	91.7 (76.0-100.0)	82.5 (60.4-100.0)	82.5 (60.4-100.0)	0.6712
	F	8	75.0 (45.0-100.0)	75.0 (45.0-100.0)	75.0 (45.0-100.0)	
<b>VIII TUMORI MALIGNI DELL'OSSO</b> MALIGNANT BONE TUMOURS	M	42	90.5 (81.6-99.4)	72.7 (58.8-86.5)	66.2 (50.9-81.4)	0.6495
	F	46	95.6 (89.5-100.0)	68.2 (53.8-82.6)	56.9 (40.1-73.7)	
<b>VIIIA OSTEOSARCOMA</b> OSTEOSARCOMA	M	19	84.2 (67.8-100.0)	62.0 (39.8-84.3)	47.3 (22.5-72.0)	0.5270
	F	16	93.3 (80.7-100.0)	60.9 (33.7-88.1)	60.9 (33.7-88.1)	
<b>VIIIC SARCOMA DI EWING</b> EWING'S SARCOMA	M	14	100.0 (100.0-100.0)	77.1 (54.2-100.0)	77.1 (54.2-100.0)	0.1283
	F	20	95.0 (85.4-100.0)	66.1 (43.8-88.4)	34.0 (5.2-62.8)	
<b>IX TUMORI DEI TESSUTI MOLLI</b> SOFT-TISSUE SARCOMAS	M	53	86.5 (77.3-95.8)	76.8 (65.3-88.3)	71.7 (59.0-84.4)	0.4309
	F	53	83.0 (72.9-93.1)	66.3 (53.2-79.5)	63.6 (49.9-77.3)	
<b>IXA RABDOMIOSARCOMI E SARCOMI EMBRIONALI</b> RHABDOMYOSARCOMA AND EMBRYONAL SARCOMA	M	26	96.2 (88.8-100.0)	84.4 (70.4-98.5)	79.2 (62.6-95.7)	0.3753
	F	24	83.3 (68.4-98.2)	68.2 (48.4-88.0)	68.2 (48.4-88.0)	
<b>IXB,C,D,E ALTRI SARCOMI DEI TESSUTI MOLLI</b> OTHER SOFT-TISSUE SARCOMAS	M	27	76.9 (60.7-93.1)	69.2 (51.0-87.4)	64.3 (45.3-83.2)	0.8882
	F	29	82.8 (69.0-96.5)	64.8 (47.2-82.4)	59.8 (41.0-78.6)	
<b>X TUMORI DELLE CELLULE GERMINALI</b> E ALTRI TUMORI GONADICI GERM-CELL, TROPHOBLASTIC AND OTHER GONADAL NEOPLASMS	M	31	96.8 (90.6-100.0)	90.3 (79.9-100.0)	90.3 (79.9-100.0)	0.1377
	F	29	100.0 (100.0-100.0)	82.0 (67.7-96.3)	72.9 (55.4-90.4)	
<b>XC TUMORI GERMINALI GONADICI</b> GONADAL GERM-CELL TUMOURS	M	16	100.0 (100.0-100.0)	93.8 (81.9-100.0)	93.8 (81.9-100.0)	0.6842
	F	10	100.0 (100.0-100.0)	87.5 (64.6-100.0)	87.5 (64.6-100.0)	
<b>XI CARCINOMI E ALTRI TUMORI MALIGNI EPITELIALI</b> CARCINOMAS AND OTHER MALIGNANT EPITHELIAL NEOPLASMS	M	37	91.9 (83.1-100.0)	91.9 (83.1-100.0)	91.9 (83.1-100.0)	0.7346
	F	47	97.9 (93.7-100.0)	95.7 (90.0-100.0)	95.7 (90.0-100.0)	
<b>XIB CARCINOMA DELLA TIROIDE</b> THYROID CARCINOMA	M	10	100.0 (100.0-100.0)	100.0 (100.0-100.0)	100.0 (100.0-100.0)	-
	F	12	100.0 (100.0-100.0)	100.0 (100.0-100.0)	100.0 (100.0-100.0)	
<b>XII ALTRI E NON SPECIFICATI TUMORI MALIGNI</b> OTHER AND UNSPECIFIED MALIGNANT NEOPLASMS	M	26	92.0 (81.4-100.0)	88.0 (75.3-100.0)	88.0 (75.3-100.0)	0.9282
	F	32	100.0 (100.0-100.0)	87.1 (75.3-98.9)	87.1 (75.3-98.9)	

Tabella 1. Sopravvivenza cumulativa a 1, 3 e 5 anni dalla diagnosi per sesso e per tipo di neoplasia (categorie maggiori e alcune delle minori secondo ICCC) per i bambini (0-14 anni) con tumore maligno diagnosticato nel periodo 1998-2002 in Italia (banca dati AIRTUM).

Table 1. Cumulative survival percentages at 1, 3, 5 years after diagnosis for malignant neoplasm incident in 1998-2002 in children (0-14 years) in Italy (AIRTUM database), by ICCC category and gender.

	AREA	n.	Cumulative survival Sopravvivenza cumulativa			Log-rank test
			1 y (95%CI)	3 y (95%CI)	5 y (95%CI)	
			<b>TUTTI I TUMORI</b> ALL TUMOURS TYPES	NORTH	1,372	
	CENTRE	448	90.8 (88.1-93.5)	82.3 (78.7-85.9)	79.8 (76.0-83.7)	
	SOUTH	322	90.2 (86.8-93.5)	77.2 (72.5-82.0)	74.3 (69.3-79.4)	
<b>I LEUCEMIE</b> LEUKAEMIA	NORTH	446	93.7 (91.5-96.0)	87.6 (84.5-90.8)	84.6 (80.9-88.3)	0.1344
	CENTRE	127	89.7 (84.4-95.0)	80.6 (73.7-87.6)	77.3 (69.7-85.0)	
	SOUTH	97	91.1 (85.2-97.0)	82.0 (74.0-90.0)	79.5 (71.0-88.0)	
<b>IA LEUCEMIA LINFATICA ACUTA</b> ACUTE LYMPHOCYTIC LEUKAEMIA	NORTH	360	95.8 (93.8-97.9)	90.2 (87.1-93.3)	87.6 (83.8-91.4)	0.2951
	CENTRE	106	94.3 (89.9-98.7)	88.5 (82.4-94.6)	85.9 (79.0-92.9)	
	SOUTH	69	92.2 (85.6-98.8)	82.5 (73.1-91.9)	79.0 (68.8-89.2)	
<b>IB LEUCEMIA NON-LINFATICA ACUTA</b> ACUTE NON-LYMPHOCYTIC LEUKAEMIA	NORTH	70	87.1 (79.3-95.0)	78.3 (68.1-88.5)	73.9 (62.6-85.2)	<.0001
	CENTRE	17	92.2 (81.9-98.6)	28.1 (5.2-51.1)	21.1 (0.1-42.1)	
	SOUTH	16	92.9 (79.4-100.0)	92.9 (79.4-100.0)	92.9 (79.4-100.0)	
<b>II LINFOMI E NEOPLASIE RETICOLOENDOTELIALI</b> LYMPHOMA AND RETICULOENDOTHELIAL NEOPLASMS	NORTH	215	95.8 (93.1-98.5)	90.5 (86.5-94.5)	90.5 (86.5-94.5)	0.2292
	CENTRE	70	92.9 (86.8-98.9)	88.6 (81.1-96.0)	86.9 (78.9-94.9)	
	SOUTH	54	92.6 (85.6-99.6)	81.4 (70.9-91.8)	81.4 (70.9-91.8)	
<b>IIA LINFOMA DI HODGKIN</b> HODGKIN'S LYMPHOMA	NORTH	89	100.0 (100.0-100.0)	95.3 (90.7-99.8)	95.3 (90.7-99.8)	0.1316
	CENTRE	32	100.0 (100.0-100.0)	96.9 (90.8-100.0)	93.3 (84.3-100.0)	
	SOUTH	24	100.0 (100.0-100.0)	83.1 (68.0-98.2)	83.1 (68.0-98.2)	
<b>IIB LINFOMA NON-HODGKIN</b> NON-HODGKIN'S LYMPHOMA	NORTH	65	92.3 (85.8-98.8)	84.5 (75.6-93.3)	84.5 (75.6-93.3)	0.4548
	CENTRE	17	94.1 (82.9-100.0)	94.1 (82.9-100.0)	94.1 (82.9-100.0)	
	SOUTH	20	90.0 (76.9-100.0)	80.0 (62.5-97.5)	80.0 (62.5-97.5)	
<b>IIC LINFOMA DI BURKITT</b> BURKITT'S LYMPHOMA	NORTH	41	92.7 (84.7-100.0)	90.2 (81.0-99.3)	90.2 (81.0-99.3)	0.5654
	CENTRE	9	77.8 (50.6-100.0)	77.8 (50.6-100.0)	77.8 (50.6-100.0)	
	SOUTH	7	85.7 (59.8-100.0)	85.7 (59.8-100.0)	85.7 (59.8-100.0)	
<b>III TUMORI DEL SISTEMA NERVOSO CENTRALE</b> CNS AND MISCELLANEOUS INTRACRANIAL AND INTRASPINAL NEOPLASMS	NORTH	253	80.2 (75.3-85.1)	64.5 (58.5-70.5)	61.5 (55.0-67.9)	0.7692
	CENTRE	88	82.8 (74.8-90.7)	70.0 (60.3-79.6)	65.3 (54.1-76.5)	
	SOUTH	70	78.4 (68.4-88.5)	63.8 (51.9-75.7)	61.1 (48.5-73.6)	
<b>IIIA EPENDIMOMA</b> EPENDYMOMA	NORTH	30	86.7 (74.5-98.8)	72.3 (55.8-88.7)	53.7 (30.7-76.6)	0.8493
	CENTRE	9	88.9 (68.4-100.0)	64.8 (32.4-97.2)	43.2 (2.4-84.0)	
	SOUTH	3	100.0 (100.0-100.0)	66.7 (13.3-100.0)	66.7 (13.3-100.0)	
<b>IIIB ASTROCITOMA</b> ASTROCYTOMA	NORTH	95	84.2 (76.9-91.5)	72.2 (63.1-81.3)	69.6 (59.5-79.7)	0.7190
	CENTRE	28	89.3 (77.8-100)	78.6 (63.4-93.8)	78.6 (63.4-93.8)	
	SOUTH	20	100.0 (100.0-100.0)	73.4 (50.9-95.9)	73.4 (50.9-95.9)	
<b>IIIC TUMORI NEUROEPITELIALI PRIMITIVI</b> PRIMITIVE NEUROECTODERMAL TUMOURS	NORTH	55	81.8 (71.6-92.0)	64.5 (51.6-77.4)	64.5 (51.6-77.4)	0.7407
	CENTRE	15	73.3 (51.0-95.7)	66.7 (42.8-90.5)	55.6 (27.4-83.7)	
	SOUTH	15	71.4 (47.8-95.1)	71.4 (47.8-95.1)	71.4 (47.8-95.1)	
<b>IIID ALTRI GLIOMI</b> OTHER GLIOMAS	NORTH	30	76.7 (61.5-91.8)	55.4 (37.0-73.8)	55.4 (37.0-73.8)	0.1532
	CENTRE	7	100.0 (100.0-100.0)	85.7 (59.8-100.0)	85.7 (59.8-100)	
	SOUTH	3	33.3 (0.0-86.7)	33.3 (0.0-86.7)	33.3 (0.0-86.7)	
<b>IIIF TUMORI INTRACRANICI E INTRASPINALI</b> NON SPECIFICATI UNSPECIFIED INTRACRANIAL AND INTRASPINAL NEOPLASMS	NORTH	41	65.9 (51.3-80.4)	46.3 (31.1-61.6)	46.3 (31.1-61.6)	0.4689
	CENTRE	28	74.1 (57.5-90.6)	63.0 (44.7-81.2)	63.0 (44.7-81.2)	
	SOUTH	28	70.7 (53.6-87.8)	55.8 (37.1-74.5)	47.8 (26.2-69.4)	
<b>IV TUMORI DEL SISTEMA NERVOSO SIMPATICO</b> SYMPATHETIC NERVOUS SYSTEM TUMOURS	NORTH	119	90.8 (85.6-96.0)	77.5 (69.9-85.2)	69.7 (60.0-79.4)	0.5403
	CENTRE	26	92.3 (82.1-100.0)	84.0 (69.6-98.4)	79.4 (63.1-95.6)	
	SOUTH	15	100.0 (100.0-100.0)	71.8 (48.3-95.3)	63.8 (38.3-89.4)	

Tabella 2. Sopravvivenza cumulativa per area geografica e per tipo di neoplasia (categorie maggiori e alcune delle minori secondo ICCO) per i bambini (0-14 anni) con tumore maligno diagnosticato nel periodo 1998-2002 in Italia (banca dati AIRTUM).

70, 17 e 16 casi rispettivamente residenti nelle aree servite da registri tumori nel Nord, Centro e Sud Italia). Nella tabella 3 sono confrontate le percentuali di sopravvivenza per i bambini italiani con le corrispondenti stime del SEER, limitatamente ai coetanei caucasici. Data la repentina evoluzione dei progressi clinici che da sempre caratterizza l'oncologia pediatrica, si è optato per un confronto delle stime italiane fornite dall'AIRTUM con quelle pubblicate dal SEER, che garantiscono maggiore sovrapposizione temporale rispetto a quelle pubblicate da ACCIS e da EURO-CARE. Complessivamente i dati italiani mostrano una buo-

children, compared to the corresponding estimates provided by SEER for Caucasian children. As there have been fast progress in childhood cancer therapy, we deem SEER data a more appropriate comparison than European data provided by ACCIS or EURO-CARE, because of the closer overlap of periods under investigation. Overall, Italian results were very close to those observed in the US, with some exceptions. US children showed a higher survival after CNS neoplasm, most notably after astrocytoma. The difference could be due to the more advanced diagnostic and surgical resources or to their greater availability. No statistically significant differences were observed for other

	AREA	n.	Cumulative survival Sopravvivenza cumulativa			Log-rank test
			1 y (95%CI)	3 y (95%CI)	5 y (95%CI)	
<b>V RETINOBLASTOMA</b> RETINOBLASTOMA	NORTH	21	95.2 (86.1-100.0)	95.2 (86.1-100.0)	95.2 (86.1-100.0)	0.0947
	CENTRE	13	100.0 (100.0-100.0)	100.0 (100.0-100.0)	100.0 (100.0-100.0)	
	SOUTH	3	100.0 (100.0-100.0)	66.7 (13.3-100.0)	66.7 (13.3-100.0)	
<b>VI TUMORI RENALI</b> RENAL TUMOURS	NORTH	71	95.8 (91.1-100.0)	86.2 (77.8-94.6)	84.0 (74.8-93.3)	0.1267
	CENTRE	24	100.0 (100.0-100.0)	100.0 (100.0-100.0)	100.0 (100.0-100.0)	
	SOUTH	14	100.0 (100.0-100.0)	91.7 (76.0-100.0)	82.5 (60.4-100.0)	
<b>VII TUMORI EPATICI</b> HEPATIC TUMOURS	NORTH	13	92.3 (77.8-100.0)	83.9 (63.4-100.0)	83.9 (63.4-100.0)	0.7037
	CENTRE	3	66.7 (13.3-100.0)	66.7 (13.3-100.0)	66.7 (13.3-100.0)	
	SOUTH	4	75.0 (32.6-100.0)	75.0 (32.6-100.0)	75.0 (32.6-100.0)	
<b>VIII TUMORI MALIGNI DELL'OSSO</b> MALIGNANT BONE TUMOURS	NORTH	46	91.3 (83.2-99.4)	64.0 (49.2-78.7)	51.8 (34.5-69.1)	0.1939
	CENTRE	25	96.0 (88.3-100.0)	83.4 (68.4-98.3)	78.2 (61.0-95.3)	
	SOUTH	17	93.8 (81.9-100.0)	67.7 (44.3-91.1)	60.2 (35.2-85.2)	
<b>VIIIA OSTEOSARCOMA</b> OSTEOSARCOMA	NORTH	16	87.5 (71.3-100.0)	59.8 (34.7-85.0)	31.9 (0.0-64.0)	0.2566
	CENTRE	13	92.3 (77.8-100.0)	75.2 (50.6-99.8)	75.2 (50.6-99.8)	
	SOUTH	6	80.0 (44.9-100.0)	40.0 (0.0-82.9)	40.0 (0.0-82.9)	
<b>VIIIC SARCOMA DI EWING</b> EWING'S SARCOMA	NORTH	23	95.7 (87.3-100.0)	59.0 (36.6-81.3)	49.1 (23.5-74.7)	0.3953
	CENTRE	5	100.0 (100.0-100.0)	80.0 (44.9-100)	53.3 (4.7-100.0)	
	SOUTH	6	100.0 (100.0-100.0)	100.0 (100.0-100.0)	80.0 (44.9-100.0)	
<b>IX TUMORI DEI TESSUTI MOLLI</b> SOFT-TISSUE SARCOMAS	NORTH	74	81.1 (72.2-90.0)	69.4 (58.7-80.1)	63.6 (52.0-75.3)	0.4554
	CENTRE	19	89.5 (75.7-100.0)	78.9 (60.6-97.3)	78.9 (60.6-97.3)	
	SOUTH	13	100.0 (100.0-100.0)	75.0 (50.5-99.5)	75.0 (50.5-99.5)	
<b>IXA RABDOMIOSARCOMI E SARCOMI EMBRIONALI</b> RHABDOMYOSARCOMA AND EMBRYONAL SARCOMA	NORTH	34	91.2 (81.6-100.0)	75.2 (60.2-90.3)	71.3 (55.1-87.4)	0.6671
	CENTRE	10	80.0 (55.2-100.0)	80.0 (55.2-100.0)	80.0 (55.2-100.0)	
	SOUTH	6	100.0 (100.0-100.0)	83.3 (53.5-100.0)	83.3 (53.5-100.0)	
<b>IXB.C.D.E ALTRI SARCOMI DEI TESSUTI MOLLI</b> OTHER SOFT-TISSUE SARCOMAS	NORTH	40	72.5 (58.7-86.3)	64.6 (49.7-79.6)	57.0 (40.4-73.5)	0.4852
	CENTRE	9	100.0 (100.0-100.0)	77.8 (50.6-100.0)	77.8 (50.6-100)	
	SOUTH	7	100.0 (100.0-100.0)	66.7 (28.9-100.0)	-*	
<b>X TUMORI DELLE CELLULE GERMINALI</b> <b>E ALTRI TUMORI GONADICI</b> GERM-CELL. TROPHOBLASTIC AND OTHER GONADAL NEOPLASMS	NORTH	40	97.5 (92.7-100.0)	87.3 (76.9-97.7)	84.2 (72.5-95.9)	0.7623
	CENTRE	13	100.0 (100.0-100.0)	83.9 (63.4-100.0)	83.9 (63.4-100.0)	
	SOUTH	7	100.0 (100.0-100.0)	85.7 (59.8-100.0)	68.6 (32.1-100.0)	
<b>XC TUMORI GERMINALI GONADICI</b> GONADAL GERM-CELL TUMOURS	NORTH	18	100.0 (100.0-100.0)	100.0 (100.0-100.0)	100.0 (100.0-100.0)	0.0787
	CENTRE	5	100.0 (100.0-100.0)	80 (44.9-100.0)	80.0 (44.9-100.0)	
	SOUTH	3	100.0 (100.0-100.0)	66.7 (13.3-100.0)	66.7 (13.3-100.0)	
<b>XI CARCINOMI E ALTRI TUMORI MALIGNI EPITELIALI</b> CARCINOMAS AND OTHER MALIGNANT EPITHELIAL NEOPLASMS	NORTH	52	98.1 (94.3-100.0)	96.1 (90.8-100.0)	96.1 (90.8-100.0)	0.1284
	CENTRE	17	88.2 (72.9-100.0)	88.2 (72.9-100.0)	88.2 (72.9-100.0)	
	SOUTH	15	93.3 (80.7-100.0)	93.3 (80.7-100.0)	93.3 (80.7-100.0)	
<b>XIB CARCINOMA DELLA TIROIDE</b> THYROID CARCINOMA	NORTH	14	100.0 (100.0-100.0)	100.0 (100.0-100.0)	100.0 (100.0-100.0)	-
	CENTRE	3	100.0 (100.0-100.0)	100.0 (100.0-100.0)	100.0 (100.0-100.0)	
	SOUTH	5	100.0 (100.0-100.0)	100.0 (100.0-100.0)	100.0 (100.0-100.0)	
<b>XII ALTRI E NON SPECIFICATI TUMORI MALIGNI</b> OTHER AND UNSPECIFIED MALIGNANT NEOPLASM	NORTH	22	95.5 (86.8-100.0)	90.9 (78.9-100.0)	90.9 (78.9-100.0)	0.6977
	CENTRE	23	100.0 (100.0-100.0)	86.4 (72.0-100.0)	86.4 (72.0-100.0)	
	SOUTH	13	91.7 (76.0-100.0)	83.3 (62.2-100.0)	83.3 (62.2-100.0)	

\*Censored data

Table 2. Cumulative survival percentages at 1, 3, 5 years after diagnosis for malignant neoplasm incident in 1998-2002 in children (0-14 years) in Italy (AIR-TUM database), by ICCC category and area of residence.

na concordanza con quelli USA, con limitate eccezioni. I bambini statunitensi affetti da tumore cerebrale (in particolare astrocitomi) presentano una migliore sopravvivenza. Questo vantaggio potrebbe essere messo in relazione alla disponibilità di tecniche diagnostiche e chirurgiche più avanzate o maggiormente distribuite nel territorio; per altri tipi di tumori cerebrali (per esempio, gli ependimomi e i PNET), per i quali il ruolo prognostico della chemioterapia e della radioterapia è più rilevante, le differenze in termini di sopravvivenza non sono infatti statisticamente evidenti. Per contro, nel caso dei tumori epatici, le sopravvivenze dei bambi-

CNS neoplasm types, such as ependymoma or PNET, for which radiotherapy and chemotherapy are more important. On the contrary survival was higher in Italy for children affected by liver neoplasm and the difference can be interpreted in relation to the different therapeutic strategies. Descriptive epidemiology studies as the present one can only generate hypotheses and a few tentative explanations can be associated to the observed results. Table 4 presents the cumulative survival estimated according to Brenner's period analyses. Follow-up date was 31.12.2002, corresponding to the end of the incidence period considered in the study. A three-year period was chosen, according to a previous

Tabella 3. Confronto delle percentuali cumulative di sopravvivenza a 5 anni dalla diagnosi (categorie maggiori e alcune delle minori secondo ICCC) per i bambini (0-14 anni) inclusi nella banca dati AIRTUM 1998-2002 e SEER 1996-2003.

Table 3. Cumulative survival percentages at 5 years after diagnosis for malignant neoplasm in children (0-14 years) in Italy (AIRTUM database, 1998-2002) and the US (SEER database, 1996-2003).

	AIRTUM 1998-2002 5-year cumulative survival	SEER 1996-2003 5-year relative survival rates
<b>TUTTI I TUMORI</b> ALL TUMOURS TYPES	78.2 (76.3-80.0)	77.9
<b>I LEUCEMIE</b> LEUKAEMIA	82.5 (79.4-85.6)	79.9
IA LEUCEMIA LINFATICA ACUTA ACUTE LYMPHOCYTIC LEUKAEMIA	86.2 (83.0-89.4)	85.0
IB LEUCEMIA NON-LINFATICA ACUTA ACUTE NON-LYMPHOCYTIC LEUKAEMIA	68.2 (58.4-78.0)	56.3
<b>II LINFOMI E NEOPLASIE RETICOLOENDOTELIALI</b> LYMPHOMA AND RETICULOENDOTHELIAL NEOPLASMS	88.2 (84.7-91.7)	87.1
IIA LINFOMA DI HODGKIN HODGKIN'S LYMPHOMA	92.5 (88.0-97.0)	94.9
IIIB LINFOMA NON-HODGKIN NON-HODGKIN'S LYMPHOMA	85.2 (78.3-92.1)	83.4 (IIB, IIC, IIE)
IIC LINFOMA DI BURKITT BURKITT'S LYMPHOMA	87.7 (79.1-96.2)	
<b>III TUMORI DEL SISTEMA NERVOSO CENTRALE</b> CNS AND MISCELLANEOUS INTRACRANIAL AND INTRASPINAL NEOPLASMS	62.3 (57.3-67.4)	70.7
IIIA EPENDIMOMA EPENDYMOMA	54.2 (36.2-72.2)	67.9
IIIB ASTROCITOMA ASTROCYTOMA	72.2 (64.3-80.0)	84.3
IIIC TUMORI NEUROEPITELIALI PRIMITIVI PRIMITIVE NEUROECTODERMAL TUMOURS	63.5 (52.5-74.6)	64.8
IIID ALTRI GLIOMI OTHER GLIOMAS	59.2 (43.7-74.7)	46.5
IIIF TUMORI INTRACRANICI E INTRASPINALI NON SPECIFICATI UNSPECIFIED INTRACRANIAL AND INTRASPINAL NEOPLASMS	52.1 (41.9-62.3)	
<b>IV TUMORI DEL SISTEMA NERVOSO SIMPATICO</b> SYMPATHETIC NERVOUS SYSTEM TUMOURS	70.5 (62.5-78.5)	68.7
<b>V RETINOBLASTOMA</b> RETINOBLASTOMA	94.3 (86.7-100.0)	97.1
<b>VI TUMORI RENALI</b> RENAL TUMOURS	87.4 (80.7-94.2)	88.0
<b>VII TUMORI EPATICI</b> HEPATIC TUMOURS	79.7 (61.9-97.5)	53.3
<b>VIII TUMORI MALIGNI DELL'OSSO</b> MALIGNANT BONE TUMOURS	61.5 (50.1-72.9)	66.9
VIIIA OSTEOSARCOMA OSTEOSARCOMA	52.7 (33.8-71.6)	66.9
VIIIC SARCOMA DI EWING EWING'S SARCOMA	54.3 (33.7-75.0)	64.6
<b>IX TUMORI DEI TESSUTI MOLLI</b> SOFT-TISSUE SARCOMAS	67.7 (58.3-77.1)	70.0
IXA RABDOMIOSARCOMI E SARCOMI EMBRIONALI RHABDOMYOSARCOMA AND EMBRYONAL SARCOMA	74.0 (61.1-86.9)	62.1
IXB,C,D,E ALTRI SARCOMI DEI TESSUTI MOLLI OTHER SOFT-TISSUE SARCOMAS	62.0 (48.7-75.3)	
<b>X TUMORI DELLE CELLULE GERMINALI E ALTRI TUMORI GONADICI</b> GERM-CELL, TROPHOBLASTIC AND OTHER GONADAL NEOPLASMS	81.9 (71.6-92.2)	89.6
XC TUMORI GERMINALI GONADICI GONADAL GERM-CELL TUMOURS	91.6 (80.5-100.0)	97.9
<b>XI CARCINOMI E ALTRI TUMORI MALIGNI EPITELIALI</b> CARCINOMAS AND OTHER MALIGNANT EPITHELIAL NEOPLASMS	94.0 (89.0-99.1)	89.0
XIB CARCINOMA DELLA TIROIDE THYROID CARCINOMA	100.0 (100.0-100.0)	96.8
<b>XII ALTRI E NON SPECIFICATI TUMORI MALIGNI</b> OTHER AND UNSPECIFIED MALIGNANT NEOPLASMS	87.4 (78.6-96.1)	

ni italiani sono superiori a quelle dei bambini statunitensi: anche in questo caso le strategie terapeutiche in atto tra le due sponde dell'atlantico potrebbero spiegare la differente prognosi.

Nella tabella 4 sono riportate infine le stime di sopravvivenza cumulativa calcolate con l'analisi di periodo secondo Brenner. Il follow-up è stato allineato al 31.12.2002, anno di chiusura della raccolta dati per l'incidenza, ed è stata scelta una finestra temporale di 3 anni. Utilizzando soltanto i dati più recenti disponibili, questa tecnica statistica permette di fornire le stime di sopravvivenza più aggiornate, fornendo un valido supporto ai clinici nella comunicazione tempestiva di prognosi alle famiglie dei bambini. Per la maggior parte dei tumori si rileva uno stabilizzarsi delle percentuali cumulative di sopravvivenza dopo 5-10 anni dalla diagnosi, con la rilevante eccezione costituita dai tumori cerebrali. In conclusione, la sopravvivenza dei bambini con diagnosi

study from the Childhood Cancer Registry of Piedmont. Brenner's method uses observed survival results for estimating future survival and therefore is helpful to clinicians when they are requested to forecast the possible prognosis of a given case. Moreover, Brenner's analysis highlights the plateau in cumulative survival, corresponding to a decrement in the hazard of death and,

	Cumulative survival estimated through period analysis (2000-2002) Sopravvivenza cumulata stimata con analisi di periodo				
	1 y (95%CI)	3 y (95%CI)	5 y (95%CI)	10 y (95%CI)	15 y (95%CI)
<b>TUTTI I TUMORI</b> ALL TUMOURS TYPES	89.6 (87.9-91.2)	80.2 (78.1-82.4)	77.4 (75.2-79.7)	73.8 (71.3-76.2)	71.7 (69.1-74.4)
<b>I LEUCEMIE</b> LEUKAEMIA	91.5 (88.7-94.3)	85.0 (81.5-88.4)	81.8 (78.0-85.5)	78.2 (74.0-82.3)	78.2 (74.0-82.3)
<b>II LINFOMI E NEPLASIE RETICOLOENDOTELIALI</b> LYMPHOMA AND RETICULOENDOTHELIAL NEOPLASMS	94.2 (91.1-97.4)	88.9 (84.7-93.1)	88.4 (84.1-92.7)	87.5 (82.9-92.1)	84.0 (77.6-90.5)
<b>III TUMORI DEL SISTEMA NERVOSO CENTRALE</b> CNS AND MISCELLANEOUS INTRACRANIAL AND INTRASPINAL NEOPLASMS	80.7 (75.9-85.6)	63.6 (57.8-69.4)	59.5 (53.5-65.5)	55.1 (48.9-61.3)	50.8 (44.2-57.4)
<b>IV TUMORI DEL SISTEMA NERVOSO SIMPATICO</b> SYMPATHETIC NERVOUS SYSTEM TUMOURS	89.3 (83.4-95.3)	75.4 (67.0-83.8)	68.6 (59.1-78.1)	58.7 (47.7-69.7)	58.7 (47.7-69.7)
<b>V RETINOBLASTOMA</b> RETINOBLASTOMA	95.5 (86.8-100.0)	95.5 (86.8-100.0)	95.5 (86.8-100.0)	90.2 (77.1-100.0)	90.2 (77.1-100.0)
<b>VI TUMORI RENALI</b> RENAL TUMOURS	95.5 (90.6-100.0)	88.7 (81.3-96.1)	88.7 (81.3-96.1)	88.7 (81.3-96.1)	84.1 (72.9-95.3)
<b>VII TUMORI EPATICI</b> HEPATIC TUMOURS	77.8 (55.6-100.0)	71.6 (48.0-95.1)	71.6 (48.0-95.1)	71.6 (48.0-95.1)	- *
<b>VIII TUMORI MALIGNI DELL'OSSO</b> MALIGNANT BONE TUMOURS	93.6 (86.6-100.0)	74.4 (62.4-86.3)	67.9 (55.3-80.4)	56.3 (42.9-69.8)	56.3 (42.9-69.8)
<b>IX TUMORI DEI TESSUTI MOLLI</b> SOFT-TISSUE SARCOMAS	81.2 (71.9-90.4)	70.5 (59.6-81.4)	66.2 (55.0-77.5)	66.2 (55.0-77.5)	66.2 (55.0-77.5)
<b>X TUMORI DELLE CELLULE GERMINALI E ALTRI TUMORI GONADICI</b> GERM-CELL, TROPHOBLASTIC AND OTHER GONADAL NEOPLASMS	97.1 (91.6-100.0)	89.7 (80.2-99.3)	89.7 (80.2-99.3)	89.7 (80.2-99.3)	89.7 (80.2-99.3)
<b>XI CARCINOMI E ALTRI TUMORI MALIGNI EPITELIALI</b> CARCINOMAS AND OTHER MALIGNANT EPITHELIAL NEOPLASMS	91.4 (83.3-99.5)	89.7 (81.2-98.3)	89.7 (81.2-98.3)	87.3 (77.7-96.9)	79.3 (62.1-96.5)
<b>XII ALTRI E NON SPECIFICATI TUMORI MALIGNI</b> OTHER AND UNSPECIFIED MALIGNANT NEOPLASMS	96.8 (90.6-100.0)	88.9 (78.7-99.2)	86.1 (74.8-97.4)	86.1 (74.8-97.4)	86.1 (74.8-97.4)

\* Censored data

Tabella 4. Sopravvivenza cumulativa, stimata con analisi di periodo a 1, 3, 5, 10 e 15 anni dalla diagnosi, per tipo di neoplasia (categorie maggiori secondo ICCC) per i bambini (0-14 anni) con tumore maligno diagnosticato nel periodo 1988-2002 in Italia (banca dati AIRTUM).

Table 4. Cumulative survival percentages estimated with period analysis at 1, 3, 5, 10 and 15 years after diagnosis for malignant neoplasm incident in 1998-2002 in children (0-14 years) in Italy (AIRTUM database), by major ICCC category.

di tumore maligno nel periodo 1998-2002 mostra incrementi rispetto a precedenti segnalazioni italiane ed è favorevolmente confrontabile con analoghe esperienze internazionali. Le osservazioni che derivano da questo studio di epidemiologia descrittiva possono fornire indicazioni utili per la pianificazione sul territorio di servizi di oncologia pediatrica in grado di rispondere alla richiesta di salute di questa piccola quota di popolazione, affetta da una patologia mortale nella maggior parte dei casi negli anni passati, ma che ora può essere guarita in oltre il 70 - 80% dei casi e che quindi ambisce a un buon inserimento nel mondo sociale.

correspondingly to the increase in the likelihood of cure. For most tumour types we estimate a plateau after 5 - 10 years from diagnosis, with the only important exception of CNS neoplasms. In conclusion, survival of children affected by cancer diagnosed in 1998-2002 showed important progress in comparison to older Italian studies and compares favourably with international results. Descriptive epidemiology studies as the present one provide useful information for planning health services for childhood cancer cases. This is a tiny proportion of the population that, not so long ago, had little chances of survival and now can be cured in 70 - 80% of cases, with reasonable expectations for a normal life.