

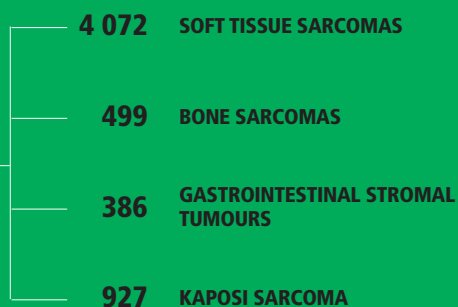
SARCOMAS

100%
OF SARCOMAS
ARE RARE

INCIDENCE

5 883

ESTIMATED NEW CASES
ITALY, 2015

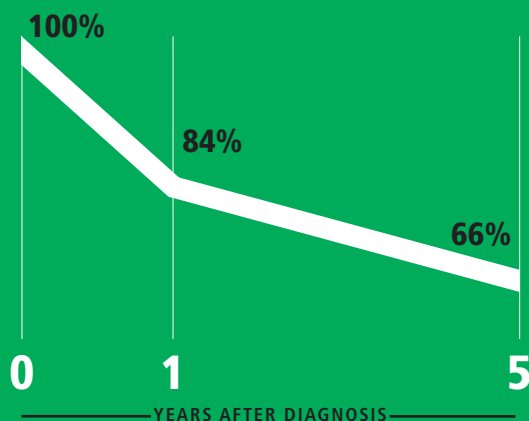


PREVALENCE

68 931

ESTIMATED PREVALENT CASES
ITALY, 2010

SURVIVAL



INCIDENCE

SARCOMAS. Crude incidence (rate per 100,000/year) and 95% confidence interval (95% CI), observed cases and proportion of rare cancers on all (common + rare) cancers by site. Rates with 95% CI by sex and age. Estimated new cases at 2015 in Italy.



	AIRTUM POOL (period of diagnosis 2000-2010)														ITALY
	RATE	95% CI	OBSERVED CASES (No.)	RARE CANCERS BY SITE (%)	SEX				AGE						ESTIMATED NEW CASES 2015
					MALE		FEMALE		0-54 yrs		55-64 yrs		65+ yrs		
					RATE	95% CI	RATE	95% CI	RATE	95% CI	RATE	95% CI	RATE	95% CI	
SARCOMAS	9.03	8.90-9.15	20 019	100%	9.54	9.35-9.72	8.55	8.38-8.72	4.75	4.65-4.87	12.72	12.30-13.15	21.37	20.94-21.81	5 883
SOFT TISSUE SARCOMAS	6.27	6.17-6.38	13 914	NA	5.95	5.80-6.09	6.58	6.43-6.73	3.49	3.40-3.59	9.23	8.87-9.60	13.97	13.62-14.33	4 072
Soft tissue sarcomas of head and neck	0.31	0.29-0.33	683		0.41	0.37-0.45	0.21	0.19-0.24	0.13	0.12-0.15	0.39	0.32-0.47	0.85	0.77-0.94	206
Soft tissue sarcomas of limbs	1.27	1.23-1.32	2 823		1.38	1.31-1.45	1.17	1.11-1.24	0.67	0.63-0.71	1.74	1.58-1.90	3.05	2.88-3.21	827
Soft tissue sarcomas of superficial trunk	0.69	0.65-0.72	1 526		0.81	0.76-0.86	0.57	0.53-0.62	0.33	0.30-0.36	1.09	0.97-1.22	1.68	1.56-1.81	447
Soft tissue sarcomas of mediastinum	0.04	0.03-0.04	79		0.04	0.03-0.05	0.03	0.02-0.04	0.02	0.01-0.02	0.07	0.04-0.11	0.08	0.06-0.11	23
Soft tissue sarcomas of heart	0.01	0.01-0.02	32		0.01	0.01-0.02	0.02	0.01-0.02	<0.01	0.00-0.01	0.02	0.01-0.05	0.03	0.02-0.05	9
Soft tissue sarcomas of breast	0.24	0.22-0.27	543		<0.01	0.00-0.02	0.47	0.43-0.51	0.17	0.15-0.19	0.38	0.31-0.46	0.42	0.36-0.48	160
Soft tissue sarcomas of uterus	0.69	0.65-0.72	1 525		0.00	–	1.33	1.27-1.40	0.50	0.46-0.54	1.21	1.09-1.35	1.01	0.92-1.11	447
Other soft tissue sarcomas of genitourinary tract	0.27	0.25-0.29	596		0.26	0.23-0.29	0.28	0.25-0.31	0.11	0.09-0.13	0.43	0.35-0.51	0.71	0.64-0.80	176
Soft tissue sarcomas of viscera	0.53	0.50-0.56	1 183		0.62	0.57-0.67	0.45	0.42-0.49	0.17	0.15-0.20	0.84	0.73-0.96	1.57	1.46-1.70	350
Soft tissue sarcomas of paratestis	0.05	0.04-0.06	120		0.11	0.09-0.13	0.00	–	0.02	0.01-0.02	0.10	0.07-0.14	0.16	0.12-0.20	37
Soft tissue sarcomas of retroperitoneum and peritoneum	0.54	0.51-0.57	1 198		0.53	0.49-0.58	0.55	0.50-0.59	0.19	0.17-0.21	1.08	0.96-1.21	1.40	1.29-1.52	349
Soft tissue sarcomas of pelvis	0.21	0.19-0.23	463		0.22	0.20-0.25	0.20	0.17-0.22	0.11	0.10-0.13	0.35	0.28-0.43	0.45	0.39-0.52	137
Soft tissue sarcomas of skin	0.78	0.74-0.82	1 731		0.87	0.82-0.93	0.69	0.65-0.74	0.58	0.54-0.62	0.75	0.65-0.86	1.48	1.37-1.60	502
Soft tissue sarcomas of paraorbit	<0.01	0.00-0.01	8		NE	–	NE	–	NE	–	NE	–	NE	–	2
Soft tissue sarcomas of brain and other parts of nervous system	0.14	0.13-0.16	318		0.16	0.14-0.19	0.12	0.10-0.15	0.10	0.08-0.11	0.22	0.17-0.28	0.26	0.21-0.31	91
Embryonal rhabdomyosarcoma of soft tissue	0.05	0.04-0.06	116		0.06	0.05-0.08	0.04	0.03-0.05	0.07	0.06-0.09	<0.01	0.00-0.03	<0.01	0.00-0.02	32
Alveolar rhabdomyosarcoma of soft tissue	0.04	0.03-0.05	87		0.04	0.03-0.05	0.04	0.03-0.05	0.05	0.04-0.06	0.03	0.01-0.06	0.03	0.01-0.04	25
Ewing's sarcoma of soft tissue	0.08	0.07-0.09	179		0.09	0.07-0.11	0.08	0.06-0.09	0.08	0.06-0.09	0.10	0.06-0.14	0.08	0.06-0.11	49
BONE SARCOMAS	0.80	0.76-0.84	1 770	NA	0.93	0.87-0.99	0.67	0.63-0.72	0.67	0.63-0.72	0.88	0.78-1.00	1.17	1.07-1.28	499
Osteogenic sarcoma	0.17	0.16-0.19	383		0.19	0.17-0.22	0.15	0.13-0.18	0.18	0.16-0.20	0.12	0.08-0.17	0.18	0.14-0.23	106
Chondrogenic sarcomas	0.24	0.22-0.26	536		0.27	0.24-0.30	0.22	0.19-0.25	0.14	0.12-0.16	0.41	0.34-0.49	0.49	0.43-0.56	153
Notochordal sarcomas, chordoma	0.08	0.07-0.09	170		0.10	0.08-0.12	0.05	0.04-0.07	0.03	0.03-0.04	0.13	0.09-0.18	0.19	0.15-0.24	49
Vascular sarcomas	0.01	0.01-0.02	28		0.02	0.01-0.03	<0.01	0.00-0.01	<0.01	0.00-0.01	0.01	0.00-0.04	0.03	0.01-0.04	8
Ewing's sarcoma	0.12	0.11-0.14	277		0.16	0.14-0.19	0.09	0.07-0.11	0.17	0.15-0.20	0.03	0.01-0.06	0.02	0.01-0.03	74
Epithelial tumours, adamantinoma	0.01	0.01-0.02	32		0.02	0.01-0.03	0.01	0.01-0.02	0.01	0.01-0.02	0.01	0.00-0.04	0.02	0.01-0.04	9
Other high grade sarcomas (fibrosarcoma, malignant fibrous histiocytoma)	0.02	0.01-0.02	36		0.01	0.01-0.02	0.02	0.01-0.03	0.01	0.01-0.02	0.03	0.01-0.05	0.03	0.02-0.05	10
GASTROINTESTINAL STROMAL TUMOURS	0.59	0.56-0.62	1 307	NA	0.66	0.61-0.71	0.53	0.48-0.57	0.17	0.15-0.20	1.12	1.00-1.25	1.68	1.56-1.81	386
KAPOSI SARCOMA	1.37	1.32-1.42	3 028	NA	2.00	1.92-2.09	0.77	0.72-0.82	0.42	0.38-0.45	1.48	1.34-1.63	4.54	4.35-4.75	927

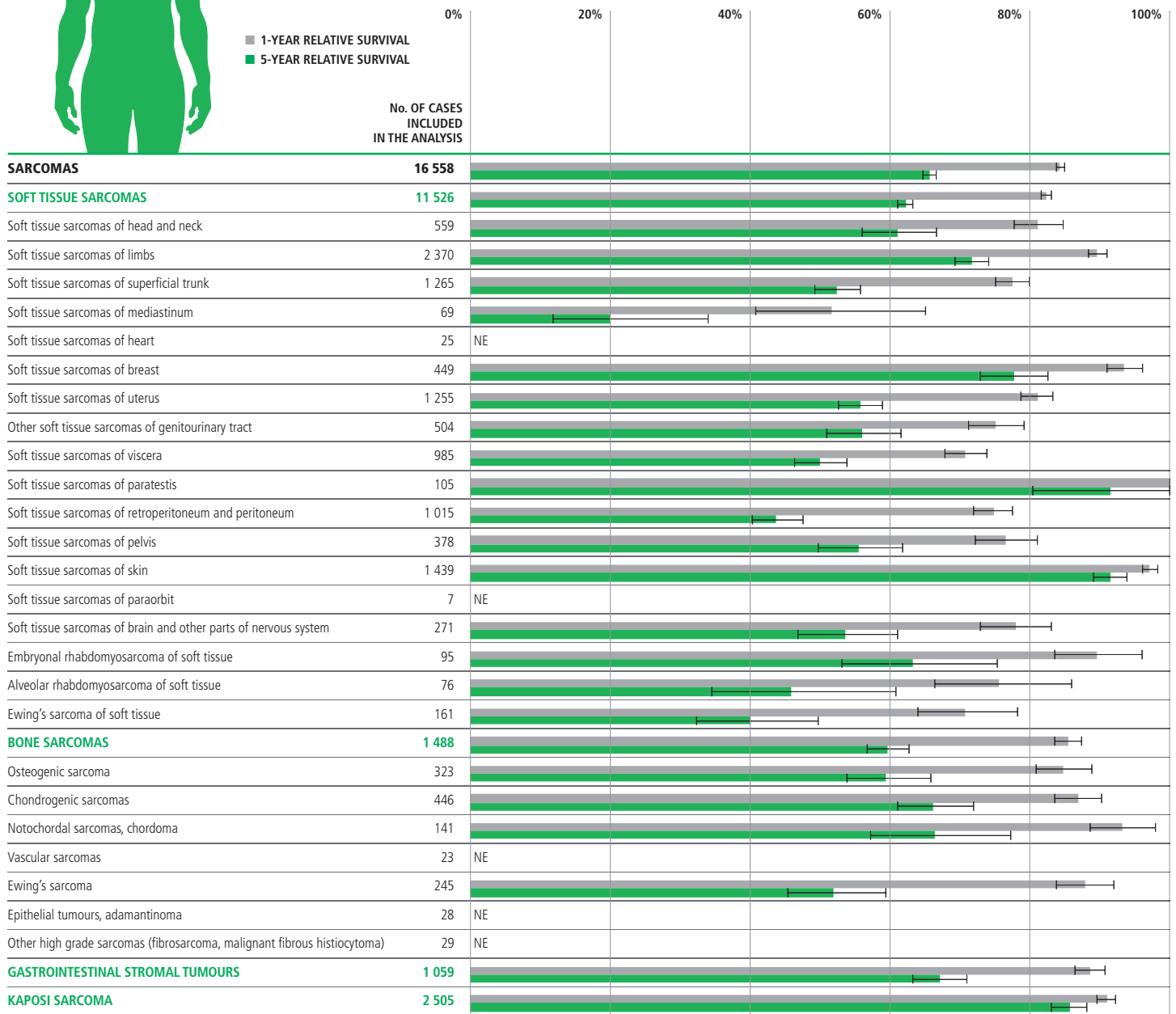
NE: not estimable because 15 or less incident cases were observed

NA: not applicable

SURVIVAL




SARCOMAS. One and 5-year relative survival. Error bars are 95% confidence interval. Cohort approach (complete analysis), period of diagnosis 2000-2008.



NE: not estimable because 30 or less incident cases were observed

PREVALENCE

SARCOMAS. Observed prevalence (proportion per 100,00 and 95% confidence interval - 95% CI) by duration (≤ 2 , 2-5, ≤ 15 years) prior to prevalence date (1st January 2007), and complete prevalence. Estimated prevalent cases in 2010 in Italy.



	AIRTUM POOL								ITALY
	OBSERVED PREVALENCE BY DURATION						COMPLETE PREVALENCE		ESTIMATED PREVALENT CASES 2010
	≤ 2 YEARS		2-5 YEARS		≤ 15 YEARS		PROPORTION	95% CI	
	PROPORTION	95% CI	PROPORTION	95% CI	PROPORTION	95% CI			
SARCOMAS	17.34	16.48-18.24	18.76	17.86-19.70	71.31	69.55-73.11	118.50	114.87-122.14	
SOFT TISSUE SARCOMAS	12.67	11.93-13.44	12.52	11.78-13.28	49.46	47.99-50.96	85.25	82.67-87.84	49 580
Soft tissue sarcomas of head and neck	0.68	0.52-0.87	0.56	0.41-0.74	2.42	2.11-2.77	4.04	3.49-4.59	2 387
Soft tissue sarcomas of limbs	2.69	2.35-3.05	3.35	2.98-3.76	11.54	10.83-12.27	18.55	17.40-19.71	10 719
Soft tissue sarcomas of superficial trunk	1.50	1.26-1.78	1.20	0.99-1.46	4.91	4.46-5.40	9.20	8.31-10.09	5 368
Soft tissue sarcomas of mediastinum	0.03	0.01-0.10	0.03	0.01-0.10	0.10	0.05-0.20	0.28	0.08-0.49	161
Soft tissue sarcomas of heart	0.02	0.00-0.08	0.01	0.00-0.06	0.06	0.02-0.13	0.23	0.00-0.46	130
Soft tissue sarcomas of breast	0.57	0.42-0.75	0.63	0.48-0.82	2.86	2.52-3.24	4.59	3.99-5.18	2 681
Soft tissue sarcomas of uterus	1.15	0.93-1.40	1.28	1.05-1.54	4.90	4.45-5.39	8.70	7.72-9.69	5 096
Other soft tissue sarcomas of genitourinary tract	0.50	0.37-0.68	0.41	0.29-0.57	1.73	1.47-2.03	3.22	2.70-3.74	1 882
Soft tissue sarcomas of viscera	0.90	0.71-1.12	0.86	0.68-1.08	3.10	2.74-3.50	4.09	3.60-4.58	2 372
Soft tissue sarcomas of paratestis	0.07	0.03-0.15	0.15	0.08-0.26	0.59	0.44-0.77	0.68	0.49-0.87	384
Soft tissue sarcomas of retroperitoneum and peritoneum	1.08	0.87-1.32	0.85	0.66-1.06	3.16	2.80-3.56	3.79	3.34-4.25	2 185
Soft tissue sarcomas of pelvis	0.34	0.23-0.49	0.36	0.24-0.51	1.35	1.11-1.61	3.13	2.40-3.86	1 820
Soft tissue sarcomas of skin	2.07	1.78-2.39	2.05	1.76-2.37	9.00	8.38-9.65	15.36	14.20-16.53	8 901
Soft tissue sarcomas of paraorbit	0.02	0.00-0.08	0.01	0.00-0.06	0.07	0.03-0.15	0.35	0.03-0.67	206
Soft tissue sarcomas of brain and other parts of nervous system	0.25	0.16-0.38	0.24	0.15-0.37	0.98	0.78-1.21	1.93	1.51-2.35	1 136
Embryonal rhabdomyosarcoma of soft tissue	0.10	0.05-0.20	0.07	0.03-0.15	0.41	0.28-0.56	1.79	0.57-3.02	1 026
Alveolar rhabdomyosarcoma of soft tissue	0.11	0.06-0.21	0.05	0.01-0.12	0.23	0.14-0.35	0.37	0.18-0.56	256
Ewing's sarcoma of soft tissue	0.21	0.12-0.33	0.10	0.05-0.20	0.46	0.33-0.63	1.15	0.70-1.59	672
BONE SARCOMAS	1.20	0.98-1.46	1.69	1.42-1.98	6.39	5.87-6.95	11.41	10.41-12.41	6 639
Osteogenic sarcoma	0.23	0.14-0.35	0.29	0.19-0.42	1.32	1.09-1.58	3.41	2.67-4.16	1 990
Chondrogenic sarcomas	0.41	0.29-0.57	0.62	0.46-0.81	2.27	1.96-2.61	4.42	3.78-5.05	2 570
Notochordal sarcomas, chordoma	0.11	0.06-0.21	0.23	0.14-0.35	0.64	0.48-0.83	0.75	0.55-0.94	428
Vascular sarcomas	0.02	0.00-0.08	NE	—	0.10	0.05-0.20	0.19	0.06-0.31	102
Ewing's sarcoma	0.23	0.14-0.35	0.31	0.20-0.45	1.08	0.87-1.32	2.13	1.30-2.96	1 284
Epithelial tumours, adamantinoma	0.01	0.00-0.06	0.02	0.00-0.08	0.09	0.04-0.18	0.15	0.04-0.25	83
Other high grade sarcomas (fibrosarcoma, malignant fibrous histiocytoma)	0.02	0.00-0.08	0.02	0.00-0.08	0.14	0.08-0.25	0.29	0.13-0.45	158
GASTROINTESTINAL STROMAL TUMOURS	1.20	0.98-1.45	1.62	1.36-1.91	3.75	3.36-4.18	4.83	4.26-5.39	2 788
KAPOSI SARCOMA	2.28	1.98-2.63	2.96	2.60-3.34	11.84	11.12-12.58	17.01	14.73-19.30	9 924

NE: not estimable in observed prevalence if no cases were observed within ≤ 2 , 2-5, ≤ 15 years prior to prevalence date, in complete prevalence if the 15-year prevalence is NE

Sarcoma is a malignant neoplasm arising from mesenchymal cells; it includes a heterogeneous group of tumours. It can be split up into dozens of histological categories, and it can occur in virtually any anatomic site. This gives rise to a huge number of possible combinations of histology and primary site which are of clinical importance. The anatomic site influences the therapeutic choice, in particular making surgery more or less viable or even impossible, but histology also influences prognosis and responsiveness to chemotherapy. It is important to consider the primary site as well as the histologic type when presenting soft tissue sarcomas (STSs) statistics.¹ These characteristics have made it almost impossible up now to have reliable statistics on incidence, mortality, prevalence, and survival per single type of sarcoma in each site even at the national level. In this study, for the first time we have the opportunity to estimate reliable incidence, prevalence, and survival statistics even for very rare sarcomas.

In this monograph we present:

- **soft tissue sarcomas (STS)** of organ-specific sites (head and neck, limbs, superficial trunk, mediastinum, heart, breast, uterus, genitourinary tract, viscera, paratestis, retroperitoneum and peritoneum, pelvis, skin, paraorbit, brain and other parts of the nervous system, embryonal rhabdomyosarcoma of soft tissue, alveolar rhabdomyosarcoma of soft tissue, Ewing's sarcoma of soft tissue);
- **bone sarcomas** (osteogenic and chondrogenic sarcomas, chordoma, vascular sarcomas, Ewing's sarcoma of bone, adamantinoma, other high grade sarcomas);
- **gastrointestinal stromal tumours (GIST)**;
- **Kaposi sarcoma (KS)**.

The latter is described in the group of sarcomas even if it is not properly a sarcoma.

We adopted the same classification used in the RARECARE project.² This classification renders the clinical importance of both anatomic site of origin and histology in these tumours.²

WHAT DO WE KNOW ABOUT THESE CANCERS?

STSs represent less than 1% of malignant tumours and show a broad range of differentiation according to the anatomic site they occur in: smooth muscle (leiomyosarcoma), adipocyte (liposarcoma), striated muscle (rhabdomyosarcoma), endothelium (angiosarcoma), or fibroblast (dermatofibrosarcoma).³ In the last two decades, cytogenetic findings have provided a valuable and reproducible tool for STS classification. The use of the molecular classification makes it possible to report variation in incidence patterns of STS by histologic type, supporting the notion that these tumours are aetiologically distinct and that they should be considered separately in analytic studies.

Little is known about their aetiology, however few risk factors are known: ionising radiation, especially in the form of radiotherapy for a previous cancer, environmental factors (e.g., herbicides, dioxins), immunodeficiency (e.g., AIDS), and viral infections (Epstein Barr virus, human herpes virus type 8).⁴⁻⁸ Several heritable syndromes are associated with increased risk of sarcomas. Those which account for the largest number of cases are neurofibromatosis 1 (nerve sheath tumours), heritable retinoblastoma (osteosarcoma and various STSs), and Li-Fraumeni syndrome (osteosarcoma and STS).⁹

KS is a virus-related malignancy which most frequently arises in the skin, though mucosal sites, lymph nodes, and viscera can also be involved. Infection with Kaposi sarcoma herpes virus (KSHV, previously known as human herpes virus type 8, HHV-8) is required for the development of KS. Historically, KS occurred as two clinically and epidemiologically distinct subtypes, classic and endemic. Classic KS is predominantly a disease of the elderly of Mediterranean or Middle Eastern origin without apparent immunosuppression; never smoking, diabetes, and use of oral corticosteroids are risk factors. Endemic KS occurs almost entirely in sub-Saharan Africa and it is difficult to disentangle endemic KS and HIV-related KS.¹⁰

Strong geographic variations have been observed for sarcoma survival among European countries. These differences are usually interpreted as differences in accessibility to effective care. In particular, the expertise of the centre has been recognised to be important for the outcome.² Sarcoma clinical management should be carried out in centres of expertise for sarcomas and/or within reference networks sharing multidisciplinary expertise and treating a high number of patients annually. This centralised referral should be pursued as early as the time of the clinical suspicion of sarcoma.

This is the first time that data on all sarcomas are shown separately and by site. Usually cancer statistics (ITACAN, GLOBOCAN, NORDCAN, SEER) give data only by site, and tumours of soft tissue and bone represent the best proxy to describe soft tissue and bone sarcomas.

THE EPIDEMIOLOGICAL DATA IN ITALY

Incidence

STSs are the most frequent (70%) sarcomas; altogether, they are slightly higher in females than in males, but this is due to the relatively high incidence rate of sarcomas of the uterus and breast. In the other non-gender specific sites, sarcomas are slightly higher in males than females. The most frequent sites are limbs, followed by skin, uterus, and superficial trunk. **Bone sarcomas and GIST** are 9% and 7% of all sarcomas, respectively (incidence table, p. 85). Regarding **GIST**, the AIRTUM incidence rate (IR) is of 0.6 per 100,000 (see table p. 85). This result is close to the IR of 0.7 per 100,000 observed in an Italian population-based study based on a pathology review,¹¹ but still slightly below the IR (range of 1.0-1.5 per 100,000) reported in other population-based studies based on pathological reviews and performed in various European countries.² The IR increases with age for most STSs. The main exceptions are: embryonal rhabdomyosarcoma, which is the most frequent histology at age 0-14, with an IR of 0.3 per 100,000 and occurs mainly in the first 4 years of life, and alveolar rhabdomyosarcoma, which also occurs mainly at ages 0-14; soft tissue sarcomas of uterus have a peak at ages 45-49, Ewing's sarcomas of soft tissue shows an almost flat incidence curve with age (data not shown). The overall age incidence pattern for **bone sarcomas** is bimodal, with peaks at ages 10-19 and 65+. Of the three most frequent subtypes, osteosarcoma and Ewing's sarcoma of the bone have their highest incidence at ages 10-19 and incidence of chondrogenic sarcomas is greatest at age 65+.

KS is relatively frequent in Italy, with an IR of 1.4 per 100,000. The relatively high incidence of classic KS in Italy is known¹² and is confirmed even in our data, which show the highest IR of KS (4.5 per 100,000) in those aged 65+ (see table p. 85).

Survival

Five-year relative survival (RS) is 62% for STSs, 60% for bone sarcomas, 67% for GIST. STSs of paratestis and skin (mainly dermatofibrosarcoma protuberans) have the highest survival rate (92% and 91%, respectively), while STSs of mediastinum have survival rates of 20%. Five-year RS from STSs of the uterus is 56% (survival figure, p. 86). In Europe 5-year RS from STSs of the uterus was 49% overall, but it was 65% for tumours of stromal histology (mainly endometrial stromal tumour) and 42% for other types, predominantly leiomyosarcoma and sarcoma NOS (not otherwise specified).²

In Italy, among **bone sarcomas**, survival rate is highest for chondrogenic sarcomas and chordoma (66%). The number of cases is too limited in the AIRTUM database to estimate 5-year RS of the rare adamantinoma and vascular sarcomas; however, according to previous European published data, the 5-year RS of these rare sarcomas was 83% and 34%, respectively.² There is general agreement that treatment of sarcomas should be concentrated in specialist centres with multidisciplinary expertise and knowledge of the disease, though the effect of such a policy on survival has seldom been evaluated. As cancer registries come to collect more information on stage and treatment and place of treatment, the evaluation of such a policy should become a priority.

For **GIST**, 5-year RS (67%; see figure p. 86) is similar to previous published data at European level,² thanks to the introduction of molecularly targeted therapies. It will be interesting to look at epidemiological data on GIST, when they become available from cancer registries in the next few years, to see how a major breakthrough involving a targeted agent can translate into prognostic improvements on a population basis in different settings. An important challenge for cancer registries will be to develop the ability to track, at the population level, the highly selective improvements resulting from this kind of “histology-driven” or “molecularly-driven” therapy, affecting single histologies or clinical presentations with low numbers of eligible patients. A proper pathologic diagnosis on a population basis would be crucial in this regard, and it is well known that this is still a challenge.

Five-year RS of **KS** (86%; see figure p. 86) is slightly higher in Italy than in Europe, but similar to that observed in Southern Europe.¹⁰ Higher survival in Southern Europe (including Italy) may partially

reflect predominantly less aggressive disease in patients with classic KS and greater clinical experience as a consequence of the higher incidence of both major subtypes of KS.¹⁰

Prevalence

About 69,000 persons were estimated to be alive in 2010 with a past diagnosis of sarcoma in Italy. The most prevalent sarcomas were **STSs** (about 50,000 cases), followed by **bone sarcomas** (about 7,000 cases). The distribution of prevalence by time since diagnosis was fairly similar for the different sarcomas. We estimated about 10,000 prevalent cases of **KS**, which is slightly higher than the estimates published in the AIRTUM monograph on prevalence.¹³ This is due to the method used to estimate complete prevalence, which leads to an overestimation of tumours, such as KS, which have different incidence across Italian areas (see «Materials and methods», pp. 14-21).

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