

The epidemiology of sarcomas

A Trama

Evaluative Epidemiology Unit

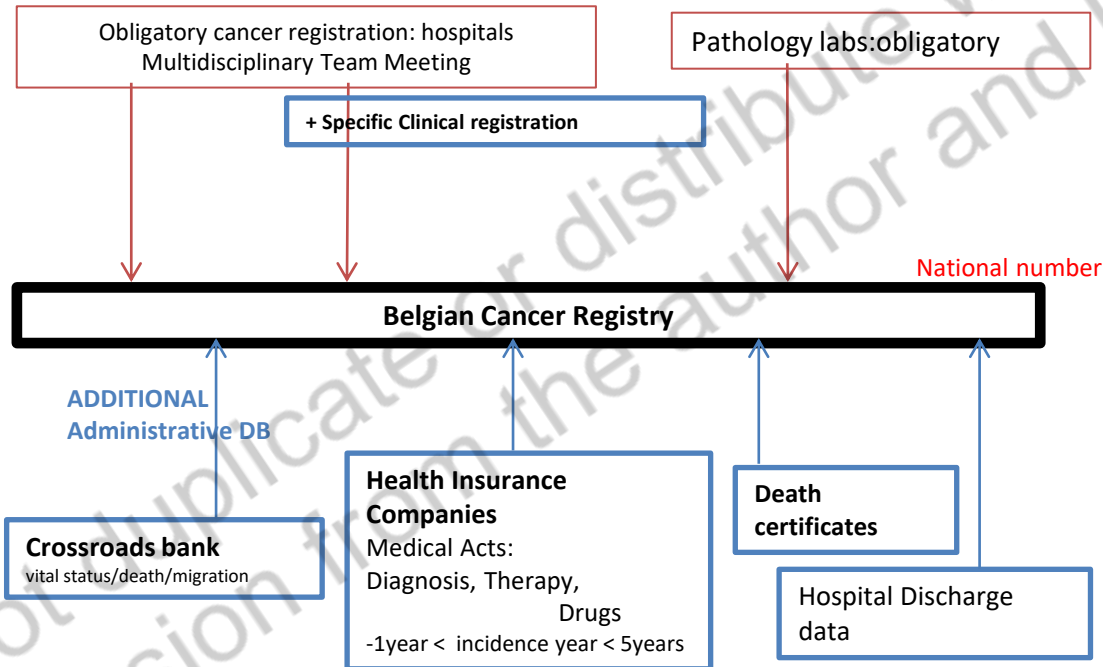
Fondazione IRCSS Istituto Nazionale dei Tumori

Milan - Italy

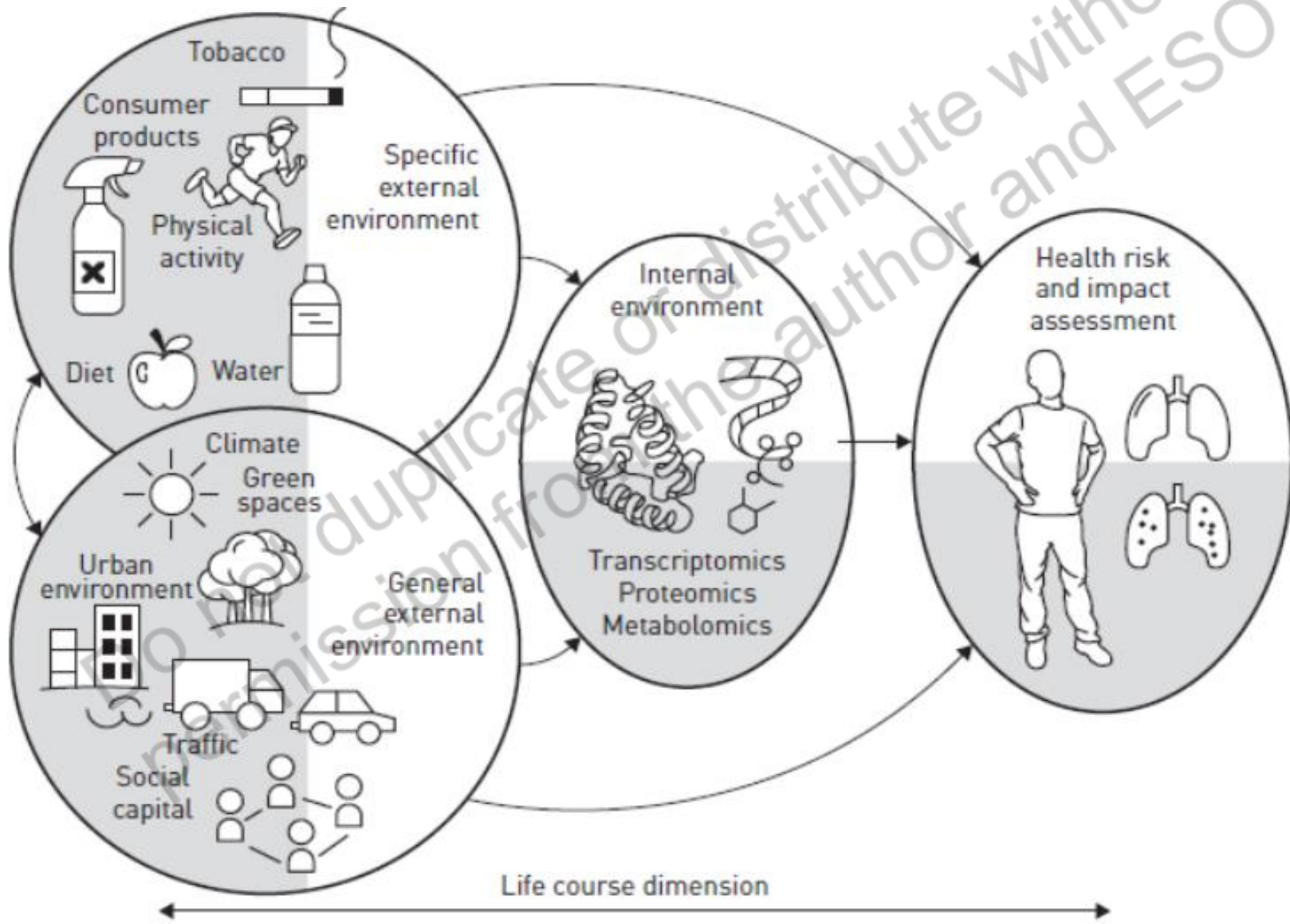
Population-based cancer registries

- Collect data on all new cases of cancer occurring in a well defined population (resident in a particular geographical region)
- Limited set of information
- Completeness of incidence and follow-up

Belgian Cancer Registry: Data sources

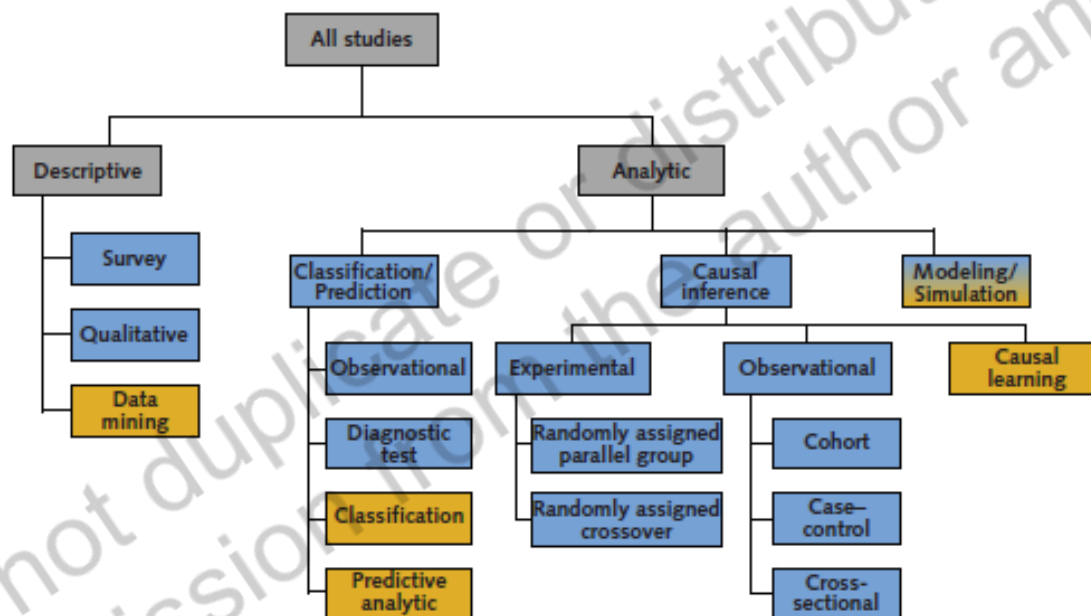


Tomorrow



Two Ways of Knowing: Big Data and Evidence-Based Medicine

Ida Sim, MD, PhD



Clinical studies include descriptive studies, which aim to describe a state of affairs, and analytic studies, which aim to quantify a relationship. Blue boxes represent traditional clinical study designs. Orange boxes represent examples of big data methods. Both traditional and big data methods are applicable to modeling and simulation. Adapted from reference 6.

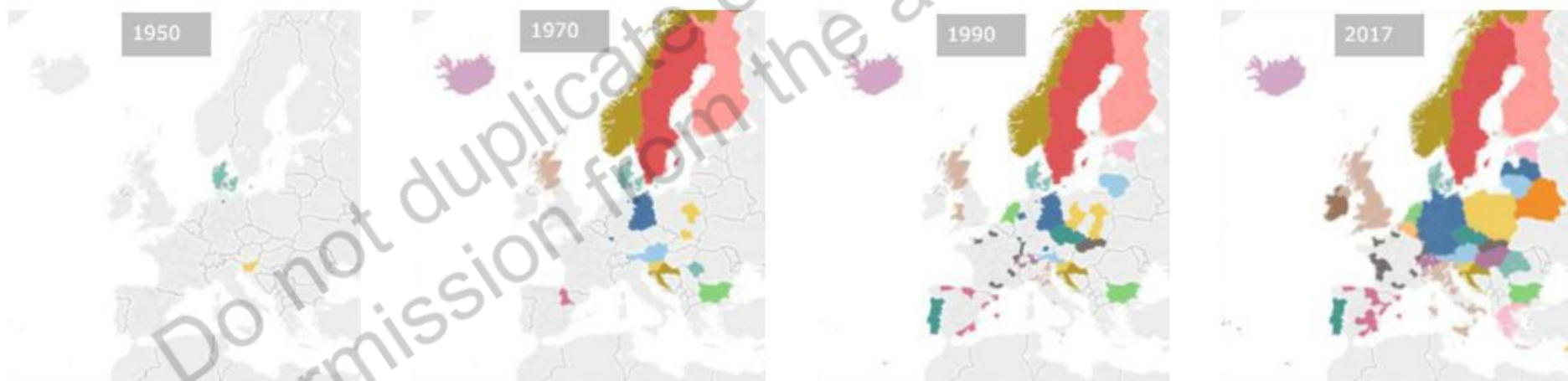
Cancer registries cover the majority of populations in developed regions—but almost none in emerging economies.





European Network
of Cancer Registries

Timeline of cancer registration in Europe





Compare

☐ Populations ☒ Cancer sites

Indicator

☒ Inc. ☐ Mort. ☐ Prev.

Sex

☒ Both ☐ Males ☐ Females

Age groups



Population

Choose a population: World

Group Colon, Rectum, Anus ☒Include NMSC ☒Include NMSC ☒ in other cancers

Re-initialize all values

Home / Explore / Table

Estimated number of new cases in 2020, worldwide, both sexes, all ages

*Crude and age-standardized rates per 100 000

ICD	Cancer	Number	Uncertainty interval	Crude Rate*	ASR (World)*	Cum. risk**
C00-97	All cancers	19 292 789	[18993000.0-19597300.0]	247.5	201.0	20.44
C50	Breast	2 261 419	[2244260.0-2278710.0]	58.5	47.8	5.20
C33-34	Lung	2 206 771	[2176530.0-2237430.0]	28.3	22.4	2.74
C18-21	Colorectum	1 931 590	Not available	24.8	19.5	-
C61	Prostate	1 414 259	[1395310.0-1433460.0]	36.0	30.7	3.86
C16	Stomach	1 089 103	[1066580.0-1112100.0]	14.0	11.1	1.31
C22	Liver	905 677	[884695.0-927157.0]	11.6	9.5	1.11
C53	Cervix uteri	604 127	[582031.0-627062.0]	15.6	13.3	1.39
C15	Oesophagus	604 100	[587111.0-621581.0]	7.8	6.3	0.78
C73	Thyroid	586 202	[579106.0-593385.0]	7.5	6.6	0.68
C67	Bladder	573 278	[557247.0-589770.0]	7.4	5.6	0.64
C82-86, C96	Non-Hodgkin lymphoma	544 352	[536030.0-552803.0]	7.0	5.8	0.62
C25	Pancreas	495 773	[488953.0-502688.0]	6.4	4.9	0.55
C91-95	Leukaemia	474 519	[459843.0-489663.0]	6.1	5.4	0.50
C64-65	Kidney	431 288	[418145.0-444844.0]	5.5	4.6	0.52
C54	Corpus uteri	417 367	[410437.0-424414.0]	10.8	8.7	1.05
C00-06	Lip, oral cavity	377 713	[362367.0-393709.0]	4.8	4.1	0.46
C43	Melanoma of skin	324 635	[314175.0-335443.0]	4.2	3.4	0.37
C56	Ovary	313 959	[300850.0-327639.0]	8.1	6.6	0.73
C70-72	Brain, central nervous system	308 102	[295692.0-321033.0]	4.0	3.5	0.35
C32	Larynx	184 615	[174268.0-195576.0]	2.4	2.0	0.25
C88+C90	Multiple myeloma	176 404	[167933.0-185303.0]	2.3	1.8	0.21
C11	Nasopharynx	133 354	[124737.0-142566.0]	1.7	1.5	0.16
C23	Gallbladder	115 949	[108291.0-124149.0]	1.5	1.2	0.13
C09-10	Oropharynx	98 412	[91318.8-106056.0]	1.3	1.1	0.13
C12-13	Hypopharynx	84 254	[76654.6-92606.8]	1.1	0.91	0.11
C81	Hodgkin lymphoma	83 087	[78805.6-87601.0]	1.1	0.98	0.09
C62	Testis	74 458	[68176.1-81318.7]	1.9	1.8	0.14
C07-08	Salivary glands	53 583	[48243.7-59513.3]	0.69	0.57	0.06
C51	Vulva	45 240	[40655.5-50341.5]	1.2	0.85	0.09
C60	Penis	36 068	[30962.6-42015.2]	0.92	0.80	0.09
C46	Kaposi sarcoma	34 270	[26009.7-45153.7]	0.44	0.39	0.03
C45	Mesothelioma	30 870	[26960.2-35346.8]	0.40	0.30	0.03
C52	Vagina	17 908	[14678.3-21848.4]	0.46	0.36	0.04

Table 4.1. Oesophagus (C15)

1.Carcinoma	8010-8576
1.1 Squamous cell carcinoma	8050-8078, 8083-8084
1.2 Adenocarcinoma	8140-8141, 8143-8145, 8190-8231, 8260-8265, 8310, 8401, 8480-8490, 8550-8552, 8570-8574, 8576
1.3 Other specified carcinoma	
1.4 Unspecified carcinoma	8010-8035
2 Sarcoma	8800-8811, 8830, 8840-8921, 8990-8991, 9040-9044, 9120-9133, 9150, 9540-9581
3 Other specified malignant neoplasm	
4 Unspecified malignant neoplasm	8000-8005

Table 4.4. Lung (C34)

1.Carcinoma	8010-8576
1.1 Squamous cell carcinoma	8050-8078, 8083-8084
1.2 Adenocarcinoma	8140, 8211, 8230-8231, 8250-8260, 8323, 8480-8490, 8550-8552, 8570-8574, 8576
1.3 Small cell carcinoma	8041-8045
1.4 Large cell carcinoma (including giant cell, clear cell and large cell undifferentiated carcinoma)	8010-8012, 8014-8031, 8035, 8310
1.5 Other specified carcinoma	
2 Sarcoma	8800-8811, 8830, 8840-8921, 8990-8991, 9040-9044, 9120-9133, 9150, 9540-9581
3 Other specified malignant neoplasm	
4 Unspecified malignant neoplasm	8000-8005

Table 4.5. Bone (C40-C41)

1 Sarcoma	8800-8921, 9040-9044, 9120-9133, 9150, 9180-9250, 9260, 9540-9581
1.1 Osteosarcoma	9180-9200
1.2 Chondrosarcoma	9210-9243
1.3 Ewing sarcoma	9260
1.4 Other specified sarcoma	
1.5 Unspecified sarcoma	8800-8803, 8805
2 Other specified malignant neoplasm	
3 Unspecified malignant neoplasm	8000-8005

C49 CONNECTIVE, SUBCUTANEOUS AND OTHER SOFT TISSUES (includes adipose tissue, aponeuroses, artery, blood vessel, bursa, connective tissue, fascia, fatty tissue, fibrous tissue, ligament, lymphatic, muscle, skeletal muscle, subcutaneous tissue, synovia, tendon, tendon sheath, vein, vessel)

C49.0 Connective, subcutaneous and other soft tissues of head, face, and neck (excludes connective tissue of orbit C69.6 and nasal cartilage C30.0)
Connective, subcutaneous and other soft tissues of: (see list under C49)

- cheek
- chin
- face
- forehead
- head
- neck
- scalp
- temple
- cervical region
- pterygoid fossa
- supraclavicular region

Auricular cartilage
Cartilage of ear
Carotid artery
Masseter muscle
Sternocleidomastoid muscle

C49.1 Connective, subcutaneous and other soft tissues of upper limb and shoulder
Connective, subcutaneous and other soft tissues of: (see list under C49)

- antecubital space
- arm
- elbow
- finger
- forearm
- hand
- shoulder
- thumb

C49.2 Connective, subcutaneous and other soft tissues of lower limb and hip

Connective, subcutaneous and other soft tissues of: (see list under C49)

- ankle
- calf
- foot
- heel
- hip
- knee
- leg
- popliteal space
- thigh
- toe

Biceps femoris muscle
Femoral artery
Gastrocnemius muscle
Plantar aponeurosis
Plantar fascia
Quadriceps femoris muscle

C49.3 Connective, subcutaneous and other soft tissues of thorax (excludes thymus C37.9, heart and mediastinum C38.)

Connective, subcutaneous and other soft tissues of: (see list under C49)

- axilla
- chest
- chest wall
- thorax
- thoracic wall
- infraclavicular region
- scapular region

Aorta, NOS
Axillary artery
Diaphragm
Intercostal muscle
Internal mammary artery
Latissimus dorsi muscle
Pectoralis major muscle
Subclavian artery
Superior vena cava
Thoracic duct
Transverse muscle

C49.4 Connective, subcutaneous and other soft tissues of abdomen

Connective, subcutaneous and other soft tissues of: (see list under C49)

- abdomen
- abdominal wall
- umbilicus

Abdominal aorta
Abdominal vena cava
Abdominal wall muscle
Celiac artery
Iliopsoas muscle
Inferior vena cava
Mesenteric artery
Psoas muscle
Rectus abdominis muscle
Renal artery
Vena cava, NOS

C49.5 Connective, subcutaneous and other soft tissues of pelvis

Connective, subcutaneous and other soft tissues of: (see list under C49)

- buttock
- groin
- perineum
- gluteal region
- inguinal region
- sacrococcygeal region

Gluteus maximus muscle
Iliac artery
Iliac vein

C49.6 Connective, subcutaneous and other soft tissues of trunk NOS

Connective, subcutaneous and other soft tissues of: (see list under C49)

- back
- flank
- trunk

C49.8 Overlapping lesion of connective, subcutaneous and other soft

TABLE 1 – SOFT TISSUE SARCOMAS DIAGNOSED DURING 1978–2001 IN THE 9 SEER PROGRAM AREAS BY HISTOLOGIC TYPE

	ICDO-3 codes	All primary sites ¹			Soft tissues only ¹			All other sites ¹		
		Cases	Percent	Rate	Cases	Percent	Rate	Cases	Percent	Rate
Leiomyosarcoma	8890, 8891, 8894, 8895–7	6,393	23.9	1.23	1,526	11.9	0.29	4,867	34.9	0.94
Malignant fibrous histiocytoma (MFH)	8830	4,577	17.1	0.88	3,230	25.2	0.62	1,347	9.7	0.26
Liposarcoma	8850–8855, 8858	3,086	11.5	0.59	2,153	24.1	0.41	933	22.1	0.18
Liposarcoma, NOS	8850	765	2.8	0.15	477	3.7	0.09	288	2.1	0.06
Well differentiated	8851	721	2.6	0.14	474	3.7	0.09	247	1.8	0.05
Myxoid	8852	928	3.4	0.17	738	5.8	0.14	190	1.4	0.03
Round cell	8853	89	0.3	0.02	79	0.6	0.02	10	0.1	0.00
Pleomorphic	8854	317	1.2	0.06	232	1.8	0.05	85	0.6	0.01
Dedifferentiated	8858	171	0.6	0.03	90	0.7	0.02	81	0.6	0.01
Mixed	8855	95	0.4	0.02	63	0.5	0.01	32	0.2	0.01
Dermatofibrosarcoma	8832–3	2,810	10.5	0.50	294	2.3	0.05	2,516	18.0	0.45
Rhabdomyosarcoma	8900–8902, 8910, 8912, 8920	1,218	4.6	0.21	617	4.8	0.11	601	4.3	0.10
Rhabdomyosarcoma, NOS	8900	304	1.1	0.05	150	1.2	0.03	154	1.1	0.02
Pleomorphic	8901	96	0.4	0.02	75	0.6	0.01	21	0.2	0.01
Mixed type	8902	17	0.1	0.00	7	0.1	0.00	10	0.1	0.00
Embryonal	8910	579	2.2	0.10	225	1.8	0.04	354	2.5	0.06
Spindle	8912	1	0	0.00	1	0	0.00	0	0	0.00
Alveolar	8920	221	0.8	0.04	159	1.2	0.03	62	0.4	0.01
Angiosarcomas	9120, 9130, 9133, 9170	1,092	4.1	0.21	498	3.9	0.10	594	4.3	0.11
Nerve sheath tumor/MPNST	9540, 9560, 9561–2	1,061	4.0	0.19	769	6.0	0.14	292	2.1	0.05
Fibrosarcoma	8810, 8811, 8814–5	964	3.6	0.18	665	5.2	0.12	299	2.1	0.06
Sarcoma, NOS	8800–5	3,424	12.8	0.65	1,707	13.3	0.32	1,717	12.3	0.33
Other specified soft tissue sarcomas ²		2,133	8.0	0.38	1,359	10.6	0.24	774	5.6	0.14
Ewing sarcoma/PNET	9260, 9364, 9365, 9473	605	2.3	0.10	211	1.6	0.04	394	2.8	0.07
Osteosarcoma	9180, 9181	80	0.3	0.02	42	0.6	0.01	38	0.3	0.01
Chondrosarcoma	9220, 9231, 9240	260	1.0	0.05	150	1.2	0.03	110	0.8	0.02
Synovial sarcoma	9040–3	615	2.3	0.11	573	4.5	0.10	42	0.3	0.01
Clear cell sarcoma	9044	95	0.4	0.02	87	0.7	0.02	8	0.1	0.00
Myxosarcoma	8840	58	0.2	0.01	45	0.4	0.01	13	0.1	0.00
Malignant hemangiopericytoma	9150	281	1.0	0.05	146	1.1	0.03	135	1.0	0.03
Malignant giant cell tumor	9251–2	21	0.1	0.00	20	0.2	0.00	1	0.0	0.00
Malignant granular cell tumor	9580	38	0.1	0.01	18	0.1	0.00	20	0.1	0.00
Alveolar soft part sarcoma	9581	75	0.3	0.01	62	0.5	0.01	13	0.1	0.00
Desmoplastic small round cell tumor	8806	5	0.0	0.00	5	0.0	0.00	0	0.0	0.00
Total		26,758	100.0	5.03	12,818	100.0	2.40	13,940	100.0	2.62

TABLE II – SOFT TISSUE SARCOMAS¹ DIAGNOSED DURING 1978–2001
IN THE 9 SEER PROGRAM AREAS BY ANATOMIC SITE²

	Cases	Percent	Rate
Soft tissue including heart	12,818	47.9	2.40
Skin	3,758	14.0	0.69
Uterus	1,861	7.0	0.36
Retroperitoneum	1,777	6.6	0.34
Stomach	1,002	3.7	0.19
Small intestine	715	2.7	0.14
Lung and bronchus	410	1.5	0.08
Breast	330	1.2	0.06
Peritoneum, omentum and mesentery	289	1.1	0.06
Other male genital organs	281	1.1	0.05
Colon and rectum	276	1.0	0.05
Oral cavity and pharynx	234	0.9	0.04
Nose, nasal cavity and middle ear	226	0.8	0.04
Kidney and renal pelvis	205	0.8	0.04
Liver and intrahepatic bile duct	202	0.8	0.04
Urinary bladder	177	0.7	0.03
Cranial nerves and other nervous system ³	176	0.7	0.03
Trachea, mediastinum and other respiratory organs	169	0.6	0.03
Ovary	119	0.4	0.02
Testis	105	0.4	0.02
Eye and orbit	102	0.4	0.02
Other	1,526	5.7	0.29
Total	26,758	100.0	5.03

using sarcomas arising in specific organs not been included in the analysis, the overall **incidence rate would have been underestimated by 50%.**

International Statistical Classification of Diseases and Related Health Problems 10th Revision (ICD-10)- WHO Version for ;2016

Chapter II Neoplasms (C00-D48)

This chapter contains the following blocks:

[C00-C97](#) Malignant neoplasms

[C00-C75](#) Malignant neoplasms, stated or presumed to be primary, of specified sites, except of lymphoid, haematopoietic and related tissue

[C00-C14](#) Malignant neoplasms of lip, oral cavity and pharynx

[C15-C26](#) Malignant neoplasms of digestive organs

[C30-C39](#) Malignant neoplasms of respiratory and intrathoracic organs

[C40-C41](#) Malignant neoplasms of bone and articular cartilage

[C43-C44](#) Melanoma and other malignant neoplasms of skin

[C45-C49](#) Malignant neoplasms of mesothelial and soft tissue

[C50-C50](#) Malignant neoplasm of breast

[C51-C58](#) Malignant neoplasms of female genital organs

[C60-C63](#) Malignant neoplasms of male genital organs

[C64-C68](#) Malignant neoplasms of urinary tract

[C69-C72](#) Malignant neoplasms of eye, brain and other parts of central nervous system

[C73-C75](#) Malignant neoplasms of thyroid and other endocrine glands

[C76-C80](#) Malignant neoplasms of ill-defined, secondary and unspecified sites

[C81-C96](#) Malignant neoplasms, stated or presumed to be primary, of lymphoid, haematopoietic and related tissue

[C97-C97](#) Malignant neoplasms of independent (primary) multiple sites

[D00-D09](#) In situ neoplasms

[D10-D36](#) Benign neoplasms

[D37-D48](#) Neoplasms of uncertain or unknown behaviour

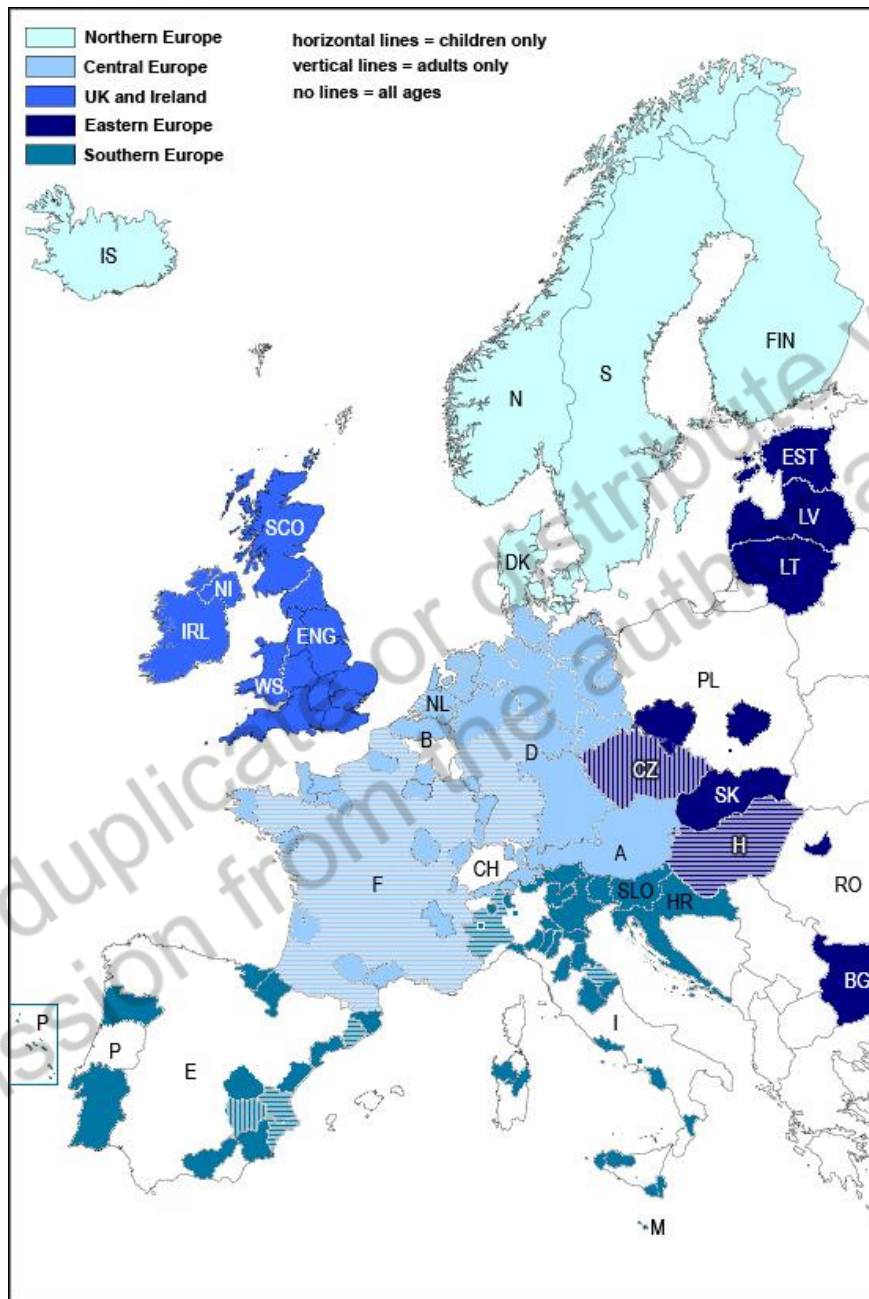
<http://apps.who.int/classifications/icd10/browse/2016/en#/II>

To have epidemiological data on sarcomas
is more difficult than for other cancers

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Collaborative projects





Surveillance of Rare Cancers in Europe



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RARECARE
ooo●



[Project Aims](#)

Information Network on Rare Cancers





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Descriptive epidemiology of sarcomas in Europe: Report from the RARECARE project

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Observed cases. Crude incidence rates per 100,000 and standard errors (SE) in Europe. rates and SE by sex and age and estimated incident cases arising in Europe per year.

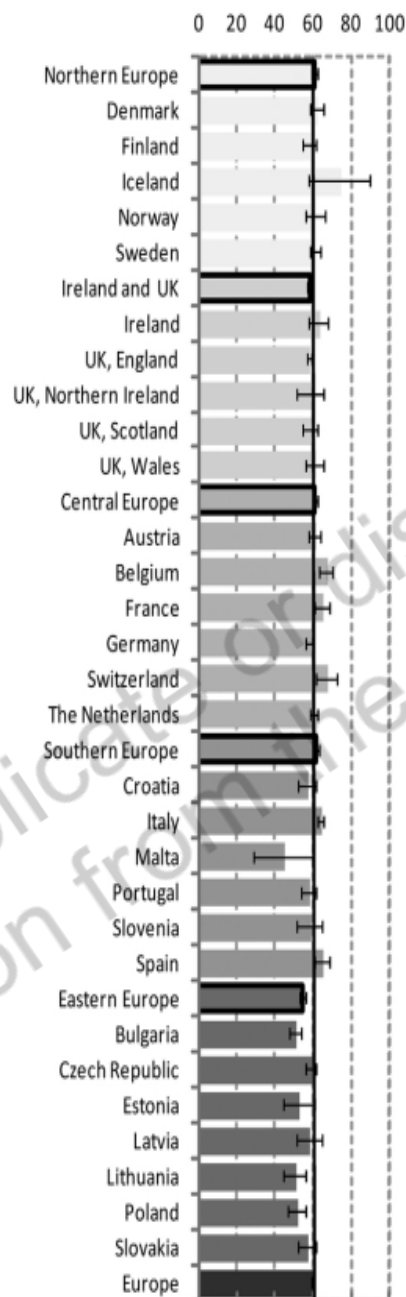
Entity	EU overall			Sex				Age								Estimated number of cases arising in EU per year
				Male		Female		0–14 years		15–24 years		25–64 years		65+ years		
	Observed cases 1995–2002	Rate	SE	Rate	SE	Rate	SE	Rate	SE	Rate	SE	Rate	SE	Rate	SE	
<i>Sarcomas</i>																
Soft tissue sarcoma	38,127	4.7	<0.1	4.4	<0.1	5.0	<0.1	0.9	<0.1	1.3	<0.1	4.4	<0.1	13.1	0.1	23,574
Soft tissue sarcoma of head and neck	2314	0.3	<0.1	0.4	<0.1	0.2	<0.1	0.1	<0.1	0.1	<0.1	0.2	<0.1	1.0	<0.1	1431
Soft tissue sarcoma of limbs	8287	1.0	<0.1	1.1	<0.1	1.0	<0.1	0.1	<0.1	0.3	<0.1	0.9	<0.1	3.0	<0.1	5124
Soft tissue sarcoma of superficial trunk	3731	0.5	<0.1	0.5	<0.1	0.4	<0.1	0.1	<0.1	0.1	<0.1	0.4	<0.1	1.4	<0.1	2307
Soft tissue sarcoma of mediastinum	209	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	0.1	<0.1	129
Soft tissue sarcoma of heart	120	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	0.0	a	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	74
Soft tissue sarcoma of breast	1499	0.2	<0.1	<0.1	<0.1	0.4	<0.1	<0.1	<0.1	<0.1	<0.1	0.2	<0.1	0.4	<0.1	927
Soft tissue sarcoma of uterus	3989	0.5	<0.1	0.0	a	1.0	<0.1	<0.1	<0.1	<0.1	<0.1	0.6	<0.1	1.0	<0.1	2466
Other soft tissue sarcomas of genitourinary tract	1919	0.2	<0.1	0.2	<0.1	0.3	<0.1	0.1	<0.1	<0.1	<0.1	0.2	<0.1	0.7	<0.1	1187
Soft tissue sarcoma of viscera	4070	0.5	<0.1	0.6	<0.1	0.5	<0.1	<0.1	<0.1	<0.1	<0.1	0.4	<0.1	1.8	<0.1	2517
Soft tissue sarcoma of paratestis	262	<0.1	<0.1	0.1	<0.1	0.0	a	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	0.1	<0.1	162
Soft tissue sarcoma of retroperitoneum and peritoneum	2295	0.3	<0.1	0.3	<0.1	0.3	<0.1	<0.1	<0.1	<0.1	<0.1	0.3	<0.1	0.9	<0.1	1419
Soft tissue sarcoma of pelvis	115	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	0.1	<0.1	71
Soft tissue sarcoma of skin	2465	0.3	<0.1	0.3	<0.1	0.3	<0.1	<0.1	<0.1	0.1	<0.1	0.3	<0.1	0.7	<0.1	1524
Soft tissue sarcoma of paraorbit	54	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	33
Soft tissue sarcoma of brain and other parts of nervous system	1531	0.2	<0.1	0.2	<0.1	0.2	<0.1	0.1	<0.1	0.1	<0.1	0.2	<0.1	0.3	<0.1	947
Embryonal rhabdomyosarcoma of soft tissue	493	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	0.25	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	305
Alveolar rhabdomyosarcoma of soft tissue	261	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	0.1	<0.1	0.1	<0.1	<0.1	<0.1	<0.1	<0.1	161
Ewing's family tumours of soft tissue	426	0.1	<0.1	0.1	<0.1	<0.1	<0.1	0.1	<0.1	0.1	<0.1	<0.1	<0.1	<0.1	<0.1	263
Bone sarcoma	6474	0.8	<0.1	0.9	<0.1	0.7	<0.1	0.7	<0.1	1.2	<0.1	0.7	<0.1	1.2	<0.1	4003
Osteosarcoma	1835	0.2	<0.1	0.3	<0.1	0.2	<0.1	0.3	<0.1	0.5	<0.1	0.1	<0.1	0.2	<0.1	1135
Chondrogenic sarcomas	1965	0.2	<0.1	0.3	<0.1	0.2	<0.1	<0.1	<0.1	0.1	<0.1	0.3	<0.1	0.5	<0.1	1215
Chordoma	352	<0.1	<0.1	0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	0.1	<0.1	218
Vascular sarcomas	26	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	0.0	a	<0.1	<0.1	<0.1	<0.1	16
Ewing's family tumours	1046	0.1	<0.1	0.2	<0.1	0.1	<0.1	0.3	<0.1	0.4	<0.1	0.1	<0.1	<0.1	<0.1	647
Epithelial tumours (adamantinoma)	69	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	43
Other high grade sarcomas (fibrosarcoma, malignant fibrous histiocytoma)	146	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	<0.1	90
Gastrointestinal stromal sarcoma	535	0.1	<0.1	0.1	<0.1	0.1	<0.1	0.0	a	<0.1	<0.1	0.1	<0.1	0.2	<0.1	331

^a Statistic could not be calculated.

Age standardised incidence rates per 100,000 and standard errors (SE) by European region for the period 1995–2002. Tier 1 entities and tier 2 entities with EU overall ASR of at least 0.1 per 100,000.

Entity	European region										EU overall	
	Northern Europe		Central Europe		Eastern Europe		Southern Europe		UK and Ireland			
	Adj. Rate	SE	Adj. Rate	SE	Adj. Rate	SE	Adj. Rate	SE	Adj. Rate	SE	Adj. Rate	SE
<i>Sarcomas</i>												
Soft tissue sarcoma	4.7	0.1	4.5	0.1	3.3	0.1	4.5	0.1	3.8	<0.1	4.2	<0.1
Soft tissue sarcoma of head and neck	0.2	<0.1	0.3	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1
Soft tissue sarcoma of limbs	1.0	<0.1	1.0	<0.1	0.7	<0.1	0.8	<0.1	0.9	<0.1	0.9	<0.1
Soft tissue sarcoma of superficial trunk	0.6	<0.1	0.4	<0.1	0.3	<0.1	0.4	<0.1	0.4	<0.1	0.4	<0.1
Soft tissue sarcoma of breast	0.1	<0.1	0.2	<0.1	0.1	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1
Soft tissue sarcoma of uterus	0.6	<0.1	0.4	<0.1	0.5	<0.1	0.5	<0.1	0.4	<0.1	0.5	<0.1
Other soft tissue sarcomas of genitourinary tract	0.2	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1
Soft tissue sarcoma of viscera	0.6	<0.1	0.4	<0.1	0.3	<0.1	0.5	<0.1	0.4	<0.1	0.4	<0.1
Soft tissue sarcoma of retroperitoneum and peritoneum	0.2	<0.1	0.3	<0.1	0.2	<0.1	0.4	<0.1	0.2	<0.1	0.3	<0.1
Soft tissue sarcoma of skin	0.5	<0.1	0.3	<0.1	0.1	<0.1	0.5	<0.1	0.2	<0.1	0.3	<0.1
Soft tissue sarcoma of brain and other parts of nervous system	0.2	<0.1	0.2	<0.1	0.2	<0.1	0.1	<0.1	0.2	<0.1	0.2	<0.1
Ewing's family tumours of soft tissue	0.1	<0.1	0.1	<0.1	<0.1	<0.1	0.1	<0.1	0.1	<0.1	0.1	<0.1
Bone sarcoma	0.8	<0.1	0.9	<0.1	0.7	<0.1	0.8	<0.1	0.7	<0.1	0.8	<0.1
Osteosarcoma	0.5	<0.1	0.5	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1
Chondrogenic sarcomas	0.3	<0.1	0.3	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1	0.2	<0.1
Ewing's family tumours	0.1	<0.1	0.1	<0.1	0.1	<0.1	0.2	<0.1	0.2	<0.1	0.1	<0.1
Gastrointestinal stromal sarcoma	0.1	<0.1	0.1	<0.1	<0.1	<0.1	0.1	<0.1	<0.1	<0.1	0.1	<0.1

Soft tissue sarcoma
Age-standardised
5-year relative survival %
Europe, 2000-2007



All cases	Number of cases		One year	Three years	Five years
15-44	8,557	obs	88.6	74.2	68.5
		rel	88.7	74.4	68.9
45-54	5,766	obs	86.2	71.8	65.6
		rel	86.5	72.7	67.1
55-64	7,587	obs	81.8	64.6	57.1
		rel	82.5	66.5	60.1
65-74	8,631	obs	74.2	54.6	45.7
		rel	75.9	58.6	52.0
75+	9,963	obs	63.2	40.0	29.9
		rel	68.6	51.5	46.3
All adults	40,504	obs	80.3	63.1	55.6
		rel	81.7	66.1	60.2
Male (54%)					
15-44	4,663	obs	87.6	71.0	64.6
		rel	87.7	71.3	65.1
45-54	3,183	obs	85.6	69.8	63.8
		rel	86.1	70.9	65.7
55-64	4,247	obs	81.3	63.4	55.7
		rel	82.3	65.8	59.6
65-74	4,856	obs	74.3	54.5	45.3
		rel	76.4	59.8	53.4
75+	4,925	obs	63.5	39.9	28.0
		rel	69.8	53.3	46.1
All adults	21,874	obs	79.9	61.5	53.6
		rel	81.5	65.2	59.1
Female (46%)					
15-44	3,894	obs	89.9	78.1	73.3
		rel	90.0	78.3	73.6
45-54	2,583	obs	86.8	74.3	67.9
		rel	87.0	74.9	68.9
55-64	3,340	obs	82.5	65.9	58.8
		rel	82.9	67.1	60.7
65-74	3,775	obs	74.3	54.7	46.3
		rel	75.3	57.2	50.3
75+	5,038	obs	62.8	40.0	31.3
		rel	67.5	49.8	45.9
All adults	18,630	obs	80.9	64.9	58.1
		rel	81.9	67.2	61.6

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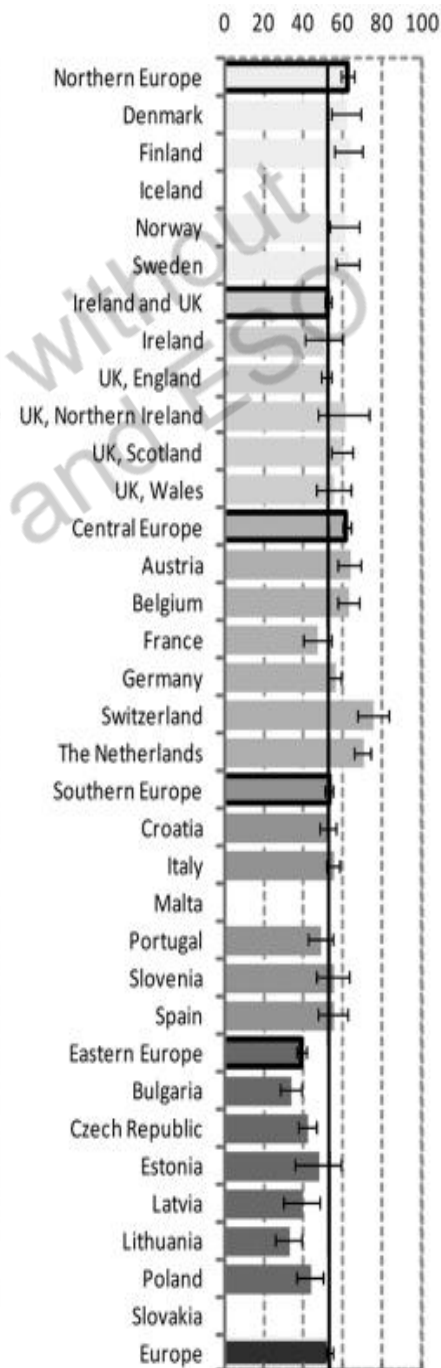
**Soft tissue sarcoma
5-year relative survival conditional
on surviving one year
Europe, 2000-2007**

	Number of cases	One year	Five year	Conditional
Northern Europe	5,491	82.8 (81.7- 83.8)	60.7 (59.0- 62.4)	73.3 (71.3- 75.4)
Denmark	1,363	82.3 (80.2- 84.5)	61.9 (58.6- 65.5)	75.2 (71.1- 79.5)
Finland	1,228	81.8 (79.7- 84.1)	58.3 (54.9- 61.9)	71.3 (67.2- 75.7)
Iceland	52	93.6 (89.0- 98.4)	74.0 (59.6- 91.8)	79.1 (63.8- 98.1)
Norway	610	83.8 (80.7- 86.9)	61.0 (56.1- 66.3)	72.8 (66.9- 79.2)
Sweden	2,238	83.1 (81.4- 84.7)	61.2 (58.6- 63.9)	73.6 (70.5- 76.9)
Ireland and UK	11,603	79.5 (78.7- 80.2)	58.3 (57.1- 59.4)	73.3 (71.9- 74.7)
Ireland	584	81.0 (77.8- 84.4)	62.8 (58.1- 67.8)	77.5 (71.7- 83.7)
UK, England	9,033	79.1 (78.2- 80.0)	57.8 (56.5- 59.1)	73.1 (71.5- 74.7)
UK, Northern Ireland	307	79.0 (74.3- 84.0)	58.8 (52.1- 66.3)	74.4 (66.0- 83.9)
UK, Scotland	957	80.2 (77.6- 82.9)	58.5 (54.5- 62.7)	72.9 (68.0- 78.2)
UK, Wales	722	82.1 (79.2- 85.2)	60.7 (56.2- 65.7)	73.9 (68.3- 79.9)
Central Europe	11,082	82.1 (81.3- 82.8)	61.1 (60.0- 62.3)	74.4 (73.0- 75.8)
Austria	1,337	81.8 (79.7- 83.9)	61.0 (57.8- 64.4)	74.6 (70.7- 78.7)
Belgium	1,205	86.8 (84.9- 88.8)	66.7 (63.4- 70.3)	76.8 (72.9- 80.9)
France	835	85.6 (83.1- 88.2)	64.6 (60.9- 68.4)	75.5 (71.3- 80.0)
Germany	3,790	80.9 (79.6- 82.3)	58.3 (56.3- 60.4)	72.1 (69.6- 74.7)
Switzerland	430	87.2 (83.9- 90.6)	67.1 (61.7- 72.9)	76.9 (70.8- 83.6)
The Netherlands	3,485	80.3 (79.0- 81.7)	60.2 (58.2- 62.3)	75.0 (72.5- 77.6)
Southern Europe	6,679	82.1 (81.2- 83.1)	61.6 (60.2- 63.0)	75.0 (73.3- 76.7)
Croatia	936	78.0 (75.3- 80.9)	56.9 (52.8- 61.4)	72.9 (67.6- 78.6)
Italy	3,609	83.8 (82.6- 85.0)	64.0 (62.1- 65.9)	76.4 (74.2- 78.6)
Malta	58	77.9 (69.1- 87.8)	44.3 (31.0- 63.3)	56.9 (39.8- 81.3)
Portugal	974	79.4 (76.7- 82.1)	58.0 (54.4- 61.9)	73.0 (68.4- 77.9)
Slovenia	346	80.9 (76.7- 85.3)	58.4 (52.2- 65.4)	72.2 (64.5- 80.9)
Spain	756	85.4 (82.9- 88.0)	64.5 (60.6- 68.5)	75.5 (71.0- 80.2)
Eastern Europe	5,649	76.5 (75.3- 77.6)	54.9 (53.3- 56.6)	71.8 (69.7- 74.0)
Bulgaria	1,374	72.4 (69.9- 75.0)	50.6 (47.3- 54.2)	69.9 (65.3- 74.8)
Czech Republic	1,867	80.8 (79.0- 82.7)	59.2 (56.4- 62.1)	73.3 (69.9- 76.9)
Estonia	233	75.3 (69.7- 81.4)	52.3 (44.7- 61.2)	69.5 (59.4- 81.3)
Latvia	342	74.6 (69.9- 79.7)	58.0 (51.6- 65.2)	77.7 (69.2- 87.3)
Lithuania	456	72.0 (67.7- 76.5)	50.6 (45.1- 56.8)	70.3 (62.7- 78.8)
Poland	652	76.2 (72.8- 79.7)	51.6 (47.0- 56.6)	67.7 (61.7- 74.3)
Slovakia	725	76.9 (73.8- 80.2)	57.5 (53.3- 62.1)	74.8 (69.3- 80.7)
Europe	40,504	81.7 (81.1- 82.3)	60.2 (59.3- 61.1)	73.7 (72.6- 74.8)

Stiller et al. Survival of adults with cancers of bone or soft tissue in Europe—Report from the EUROCARE-5 study. Cancer Epidemiology 2018

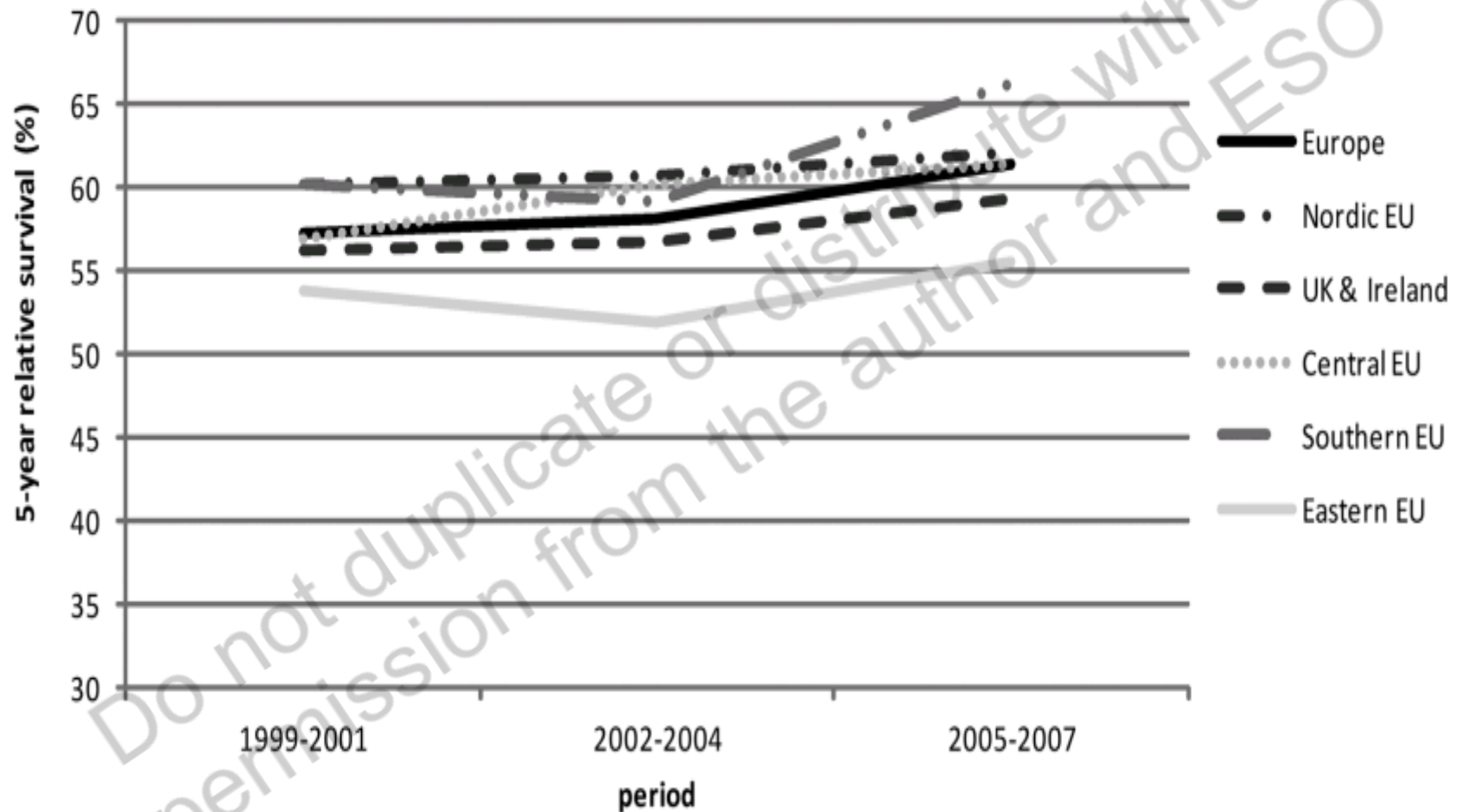
Bone sarcomas

	Number of cases	One year	Five year	Conditional
Northern Europe	1,347	82.8 (80.6- 85.0)	62.6 (59.3- 66.1)	75.6 (72.3- 79.1)
Denmark	301	83.2 (78.6- 88.1)	61.9 (54.9- 69.8)	74.4 (67.3- 82.2)
Finland	259	81.1 (76.1- 86.4)	62.9 (56.3- 70.3)	77.6 (71.3- 84.5)
Iceland	20	91.2 (80.2- 100)		
Norway	291	80.3 (75.3- 85.6)	60.9 (53.7- 69.1)	75.8 (68.5- 84.0)
Sweden	476	84.1 (80.7- 87.7)	62.8 (57.2- 68.9)	74.7 (69.0- 80.8)
Ireland and UK	3,715	76.6 (75.2- 78.1)	52.4 (50.4- 54.6)	68.4 (66.3- 70.7)
Ireland	209	79.8 (73.8- 86.3)	50.2 (41.6- 60.6)	62.9 (53.6- 73.9)
UK, England	2,965	76.2 (74.6- 77.9)	51.7 (49.4- 54.1)	67.8 (65.4- 70.3)
UK, Northern Ireland	91	81.6 (74.1- 89.8)	60.5 (48.8- 74.8)	74.1 (61.8- 88.9)
UK, Scotland	258	78.7 (73.5- 84.3)	59.3 (51.7- 67.9)	75.3 (67.3- 84.2)
UK, Wales	192	76.2 (69.9- 83.1)	55.1 (47.0- 64.6)	72.3 (64.0- 81.7)
Central Europe	3,780	83.4 (82.1- 84.7)	61.7 (59.7- 63.8)	74.0 (72.1- 76.1)
Austria	499	80.3 (76.4- 84.4)	62.9 (57.3- 69.1)	78.4 (72.8- 84.4)
Belgium	549	85.6 (82.4- 89.0)	62.8 (57.8- 68.4)	73.4 (68.3- 78.8)
France	282	77.8 (72.7- 83.2)	47.0 (40.6- 54.3)	60.4 (53.8- 67.8)
Germany	1,089	80.6 (78.1- 83.2)	55.7 (52.2- 59.5)	69.2 (65.6- 72.9)
Switzerland	147	87.2 (82.2- 92.6)	75.0 (67.5- 83.4)	86.0 (79.1- 93.5)
The Netherlands	1,214	87.3 (85.0- 89.6)	69.8 (66.0- 73.7)	79.9 (76.3- 83.7)
Southern Europe	2,550	75.2 (73.4- 76.9)	52.9 (50.7- 55.2)	70.4 (68.2- 72.7)
Croatia	713	67.8 (64.5- 71.3)	52.6 (48.5- 57.1)	77.6 (73.1- 82.4)
Italy	1,074	79.7 (77.3- 82.2)	55.0 (51.6- 58.5)	68.9 (65.5- 72.5)
Malta	17	70.8 (56.7- 88.5)		
Portugal	358	73.4 (68.3- 78.9)	48.5 (42.7- 55.1)	66.1 (60.4- 72.5)
Slovenia	135	77.4 (69.9- 85.8)	54.9 (45.2- 66.7)	70.9 (60.9- 82.5)
Spain	253	81.3 (76.1- 86.8)	54.7 (47.9- 62.5)	67.3 (60.5- 74.9)
Eastern Europe	2,271	62.4 (60.3- 64.5)	39.2 (36.8- 41.7)	62.9 (60.1- 65.8)
Bulgaria	547	56.2 (51.8- 60.9)	33.5 (28.7- 39.1)	59.6 (53.2- 66.9)
Czech Republic	657	66.3 (62.7- 70.1)	42.0 (37.6- 46.8)	63.3 (58.3- 68.8)
Estonia	96	62.6 (53.5- 73.2)	47.2 (36.8- 60.6)	75.5 (63.5- 89.7)
Latvia	153	61.9 (54.7- 70.0)	39.1 (31.0- 49.3)	63.2 (53.2- 75.1)
Lithuania	233	55.0 (48.9- 61.9)	32.6 (26.4- 40.1)	59.2 (51.3- 68.2)
Poland	329	67.6 (62.6- 73.0)	43.1 (37.0- 50.3)	63.8 (56.7- 71.9)
Slovakia	256	67.8 (61.4- 74.8)		
Europe	13,663	77.7 (76.5- 78.9)	53.1 (51.5- 54.7)	68.4 (66.8- 70.0)



Stiller et al. Survival of adults with cancers of bone or soft tissue in Europe—Report from the EUROCare-5 study. Cancer Epidemiology 2018

Time trends in age-standardised relative survival for adult patients with sarcomas across European regions



Stiller et al. Survival of adults with cancers of bone or soft tissue in Europe—
Report from the EUROCare-5 study. Cancer Epidemiology 2018

RESEARCH ARTICLE

Nationwide incidence of sarcomas and connective tissue tumors of intermediate malignancy over four years using an expert pathology review network

Gonzague de Pinieux¹, Marie Karanian², Francois Le Loarer³, Sophie Le Guellec⁴, Sylvie Chabaud², Philippe Terrier⁵, Corinne Bouvier⁶, Maxime Batistella⁷, Agnès Neuville³, Yves-Marie Robin⁸, Jean-Francois Emile⁹, Anne Moreau¹⁰, Frederique Larousserie¹¹, Agnes Leroux¹², Nathalie Stock¹³, Marick Lae^{13,14}, Françoise Collin¹⁵, Nicolas Weinbreck¹⁶, Sebastien Aubert⁸, Florence Mishellany¹⁷, Celine Charon-Barra¹⁵, Sabrina Croce³, Laurent Doucet¹⁸, Isabelle Quintin-Rouet¹⁸, Marie-Christine Chateau¹⁹, Celine Bazille²⁰, Isabelle Valo²¹, Bruno Chetaille¹⁶, Nicolas Ortonne²², Anne Brouchet⁴, Philippe Rochaix⁴, Anne Demuret¹, Jean-Pierre Ghnassia²³, Lenaig Mescam²⁴, Nicolas Macagno⁶, Isabelle Birtwisle-Peyrottes²⁵, Christophe Delfour¹⁹, Emilie Angot¹³, Isabelle Pommepuy²⁶, Dominique Ranchere², Claire Chemin-Airiau², Myriam Jean-Denis², Yohan Fayet², Jean-Baptiste Courrèges³, Nouria Mesli³, Juliane Berchoud¹⁰, Maud Toulmonde³, Antoine Italiano³, Axel Le Cesne², Nicolas Penel⁸, Françoise Ducimetiere², Francois Gouin^{2†}, Jean-Michel Coindre^{3†}, Jean-Yves Blay^{2,27,28†}, on behalf of the NetSarc/RePPS/ResSos and French Sarcoma Group-Groupe d'Etude des Tumeurs Osseuses (GSF-GETO) networks^{1†}

35% partial agreement

original article

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Sarcoma: concordance between initial diagnosis and centralized expert review in a population-based study within three European regions

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Background: Sarcomas represent a heterogeneous group of tumors. Accurate determination of histological diagnosis and prognostic factors is critical for the delineation of treatment strategies. The contribution of second opinion (SO) to improve diagnostic accuracy has been suggested for sarcoma but has never been established in population-based studies.

Methods: Histological data of patients diagnosed with sarcoma in Rhône-Alpes (France), Veneto (Italy) and Aquitaine (France) over a 2-year period were collected. Initial diagnoses were systematically compared with SO from regional and national experts.

Results: Of 2016 selected patients, 1463 (73%) matched the inclusion criteria and were analyzed. Full concordance between primary diagnosis and SO (the first pathologist and the expert reached identical conclusions) was observed in 824 (56%) cases, partial concordance (identical diagnosis of connective tumor but different grade or histological subtype) in 518 (35%) cases and complete discordance (benign versus malignant, different histological type or invalidation of the diagnosis of sarcoma) in 121 (8%) cases. The major discrepancies were related to histological grade ($n = 274$, 43%), histological type ($n = 144$, 24%), subtype ($n = 18$, 3%) and grade plus subtype or grade plus histological type ($n = 178$, 29%).

Conclusion: More than 40% of first histological diagnoses were modified at second reading, possibly resulting in different treatment decisions.

Concordance	Zero	Partial	Full	P
Included tumors ^a	104	515	814	
Type of laboratory				
Public	40 (5%)	241 (32%)	477 (63%)	<0.001
Private	64 (9%)	274 (41%)	337 (50%)	
Included tumors ^b	119	518	820	
Type of tumor sample				
Biopsy	26 (9%)	110 (38%)	154 (53%)	0.47
Surgical specimen	93 (8%)	408 (35%)	666 (57%)	
Included tumors	51	409	449	
Grade				
I	18 (7%)	77 (30%)	164 (63%)	<0.001
II-III	33 (5%)	332 (51%)	285 (44%)	
Included tumors ^c	116	515	821	
Type of sarcoma				
Soft tissue	82 (9%)	323 (36%)	502 (55%)	0.004
Visceral	34 (6%)	192 (35%)	319 (59%)	
Included tumors	121	518	824	
Region				
Aquitaine	34 (10%)	148 (42%)	170 (48%)	<0.001
Rhône-Alpes	65 (10%)	252 (38%)	345 (52%)	
Veneto	22 (5%)	118 (26%)	309 (69%)	
Included tumors	121	518	824	
Subgroup analysis				
SO requested	71 (13%)	263 (47%)	230 (40%)	<0.001
No SO requested	50 (6%)	255 (28%)	594 (66%)	

Ann Oncol 2012;23:2442

Ultra rare sarcoma: incidence < 1 per million

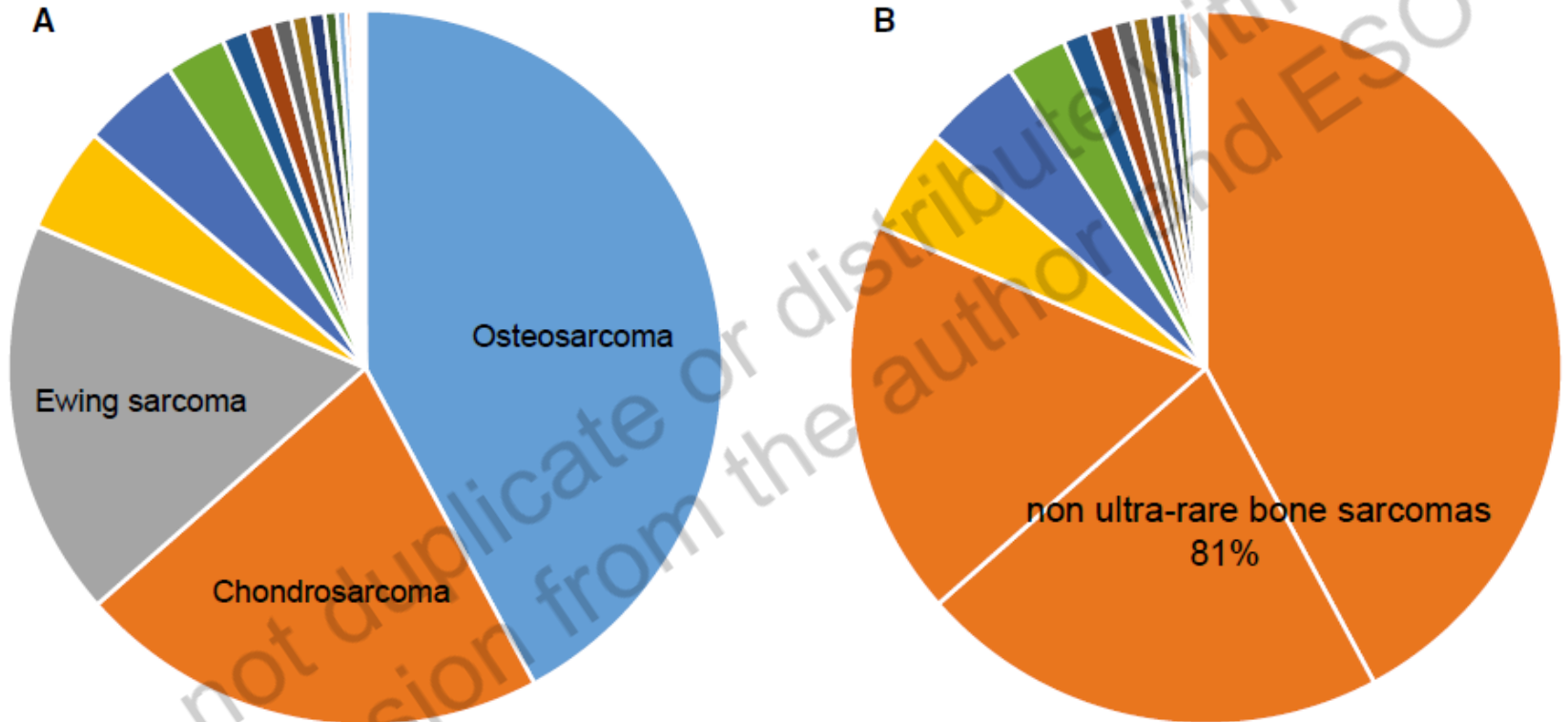


Figure 2. Ultra-rare bone sarcomas (BS) are compared with non-ultra-rare BS, illustrating (A) nonultra-rare BS types and (B) the percentage of nonultra-rare BS among all BS.

56 ultra rare type of STS; 21 ultra rare type of bone

Ultra-Rare Sarcomas: A Consensus Paper From the Connective Tissue Oncology Society Community of Experts on the Incidence Threshold and the List of Entities

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On line Analysis

Cancer Entities

Search a cancer entity in the "Cancer list" or type its name in the "Free search".

► **Cancer list** ► **Free search**

- Rare epithelial cancer of head and neck
- Rare epithelial cancer of digestive system
- Rare thoracic cancer
- Rare gynecological cancers
- Rare male urological and genital cancer + extragonadal germ cell tumours
- Rare skin cancer + non cutaneous melanoma
- Embryonal cancer
- Sarcomas
- Rare Neuroendocrine tumours
- Tumours of endocrine organs
- Tumours of central nervous system
- Rare haematological diseases
- All rare cancers

Select

► **Incidence** ► **Relative survival** ► **Prevalence**