3.16 Soft tissue cancers without mesothelioma

Table 3.16.1
Overview of key epidemiological parameters for Germany, ICD-10 C46–C49

Incidence	2015		2016		Prediction for 2020	
	Women	Men	Women	Men	Women	Men
Incident cases	2,050	2,320	1,970	2,270	2,200	2,600
Crude incidence rate[1]	4.9	5.8	4.7	5.6	5.3	6.5
Age-standardised incidence rate[1,2]	3.3	4.3	3.2	4.1	3.5	4.5
Median age at diagnosis	68	67	68	67		

Mortality	2015		2016		2017	
	Women	Men	Women	Men	Women	Men
Deaths	927	843	901	859	972	884
Crude mortality rate[1]	2.2	2.1	2.2	2.1	2.3	2.2
Age-standardised mortality rate[1,2]	1.3	1.4	1.2	1.4	1.3	1.4
Median age at death	74	71	74	72	75	72

Prevalence and survival rates	5 years		10 years	
	Women	Men	Women	Men
Prevalence	6,400	7,600	10,500	12,500
Absolute survival rate (2015–2016)[3]	48 (40–66)	51 (40–55)	35 (32–46)	40
Relative survival rate (2015–2016)[3]	54 (45–74)	60 (46–67)	45 (41–56)	56

1 per 100,000 persons 2 age-standardised (old European Standard) 3 in percentages (lowest and highest value of the included German federal states)

▶ Additional information under www.krebsdaten.de/cancer-sites

Epidemiology

This group of diseases includes rare, malignant tumours of the peripheral nerves, the connective tissue and other soft tissues, the peritoneum and the retroperitoneal soft tissue found behind it. It also includes the rare Kaposi sarcomas, which develop on the skin and account for 3 % of diagnoses. In contrast to carcinomas, which usually develop from the epithelial or glandular tissue, tumours of the soft tissue predominantly develop from connective tissue structures, sarcomas account for the majority of soft tissue tumours.

Of the approximately 4,240 new cases of malignant soft tissue tumours that occur annually, around 4 % are neoplasms of the peripheral nerves or the autonomic nervous system. The largest proportion (about 75 %) are neoplasms of the connective tissues, with leiomyosarcomas originating in smooth muscle tissue and liposarcomas (adipose tissue tumours) being two of the most common forms found among adults. In contrast, rhabdomyosarcomas, which originate from skeletal muscle tissue, almost exclusively develop in children and adolescents. Since 1999, the age-standardised incidence and mortality rates for malignant soft tissue tumours has remained fairly constant in Germany.

Risk factors

In most cases, no clear cause of soft tissue sarcoma can be found. Sarcomas are frequently identified in patients with rare hereditary cancer syndromes. The presence of one or more genetic mutations, therefore, presumably increases the risk of developing the disease.

In exceptional cases, sarcomas can also occur in areas of the body that have undergone radiation therapy. Furthermore, human herpesvirus 8 (HHV8) has been shown to cause Kaposi's sarcoma and the Epstein-Barr virus (EBV) may also be involved in the development of soft tissue sarcomas in cases of severe immune deficiency.

Environmental toxins and chemicals can also contribute to the development of sarcomas, with vinyl chloride having been shown to cause angiosarcoma of the liver. Chronic inflammatory processes are also likely to increase the risk of soft tissue sarcomas. In rare cases, lymphedema after a mastectomy (breast removal) can lead to the development of angiosarcoma (Stewart Treves syndrome). The impact of diet and other lifestyle factors such as smoking and alcohol consumption have yet to be determined.

Figure 3.16.1a
Age-standardised incidence and mortality rates by sex, ICD-10 C46–C49, Germany 1999–2016/2017, projection (incidence) through 2020
per 100,000 (old European Standard)

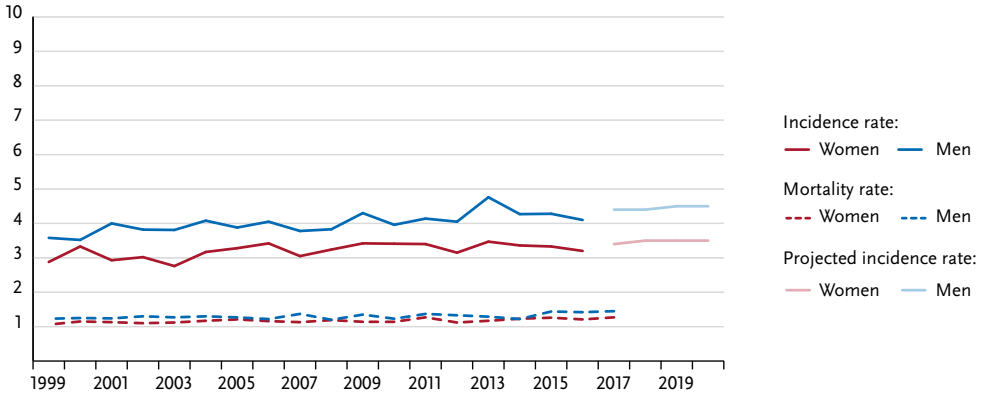


Figure 3.16.1b
Absolute numbers of incident cases and deaths by sex, ICD-10 C46–C49, Germany 1999–2016/2017, projection (incidence) through 2020

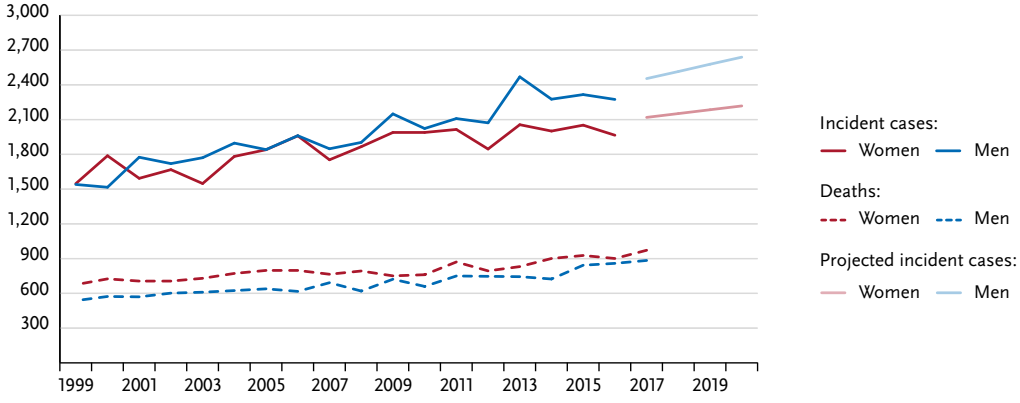


Figure 3.16.2
Age-specific incidence rates by sex, ICD-10 C46–C49, Germany 2015–2016
per 100,000

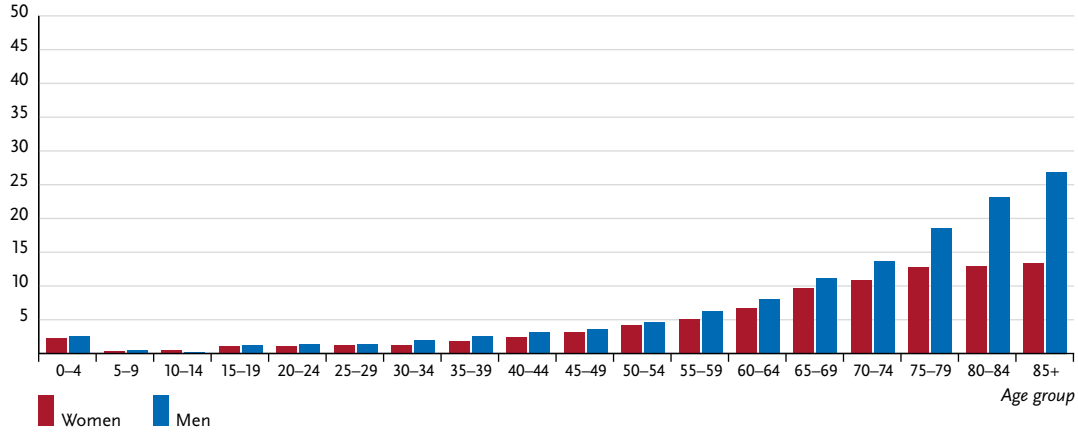


Table 3.16.2
Cancer incidence and mortality risks in Germany by age and sex, ICD-10 C46–C49, database 2016

	Risk of developing cancer				Mortality risk			
Women aged	**in the next ten years**		**ever**		**in the next ten years**		**ever**	
35 years	< 0.1 %	(1 in 4,700)	0.3 %	(1 in 300)	< 0.1 %	(1 in 22,200)	0.2 %	(1 in 590)
45 years	< 0.1 %	(1 in 2,700)	0.3 %	(1 in 320)	< 0.1 %	(1 in 10,000)	0.2 %	(1 in 600)
55 years	0.1 %	(1 in 1,700)	0.3 %	(1 in 360)	< 0.1 %	(1 in 3,800)	0.2 %	(1 in 620)
65 years	0.1 %	(1 in 1,100)	0.2 %	(1 in 420)	< 0.1 %	(1 in 2,300)	0.1 %	(1 in 710)
75 years	0.1 %	(1 in 940)	0.2 %	(1 in 610)	0.1 %	(1 in 1,500)	0.1 %	(1 in 890)
Lifetime risk			0.4 %	(1 in 270)			0.2 %	(1 in 560)
Men aged	**in the next ten years**		**ever**		**in the next ten years**		**ever**	
35 years	< 0.1 %	(1 in 3,500)	0.4 %	(1 in 260)	< 0.1 %	(1 in 13,600)	0.2 %	(1 in 600)
45 years	< 0.1 %	(1 in 2,400)	0.4 %	(1 in 270)	< 0.1 %	(1 in 9,000)	0.2 %	(1 in 620)
55 years	0.1 %	(1 in 1,500)	0.3 %	(1 in 300)	< 0.1 %	(1 in 3,800)	0.2 %	(1 in 640)
65 years	0.1 %	(1 in 900)	0.3 %	(1 in 340)	< 0.1 %	(1 in 2,400)	0.1 %	(1 in 700)
75 years	0.2 %	(1 in 640)	0.2 %	(1 in 440)	0.1 %	(1 in 1,200)	0.1 %	(1 in 780)
Lifetime risk			0.4 %	(1 in 240)			0.2 %	(1 in 560)

Figure 3.16.3
Proportion of morphologic groups of malignant soft tissue tumours by sex, ICD-10 C46–C49, Germany 2015–2016

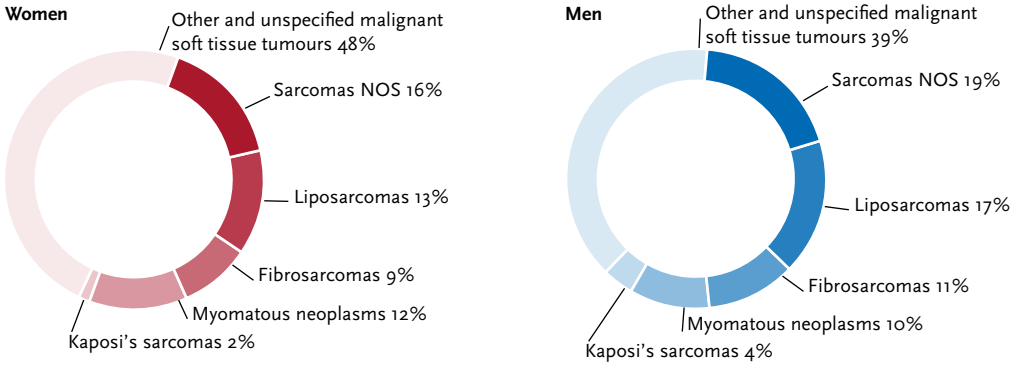


Figure 3.16.4
Absolute and relative survival rates up to 10 years after first diagnosis, by sex, ICD-10 C46–C49, Germany 2015–2016

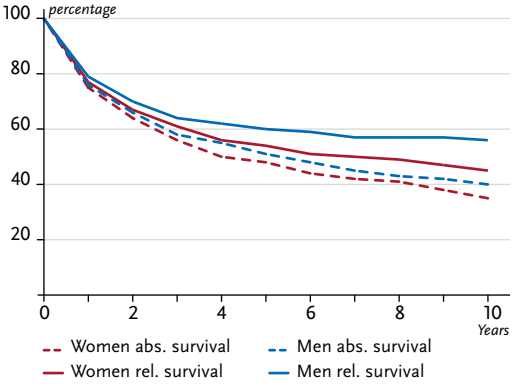


Figure 3.16.5
Relative 5-year survival by UICC-stage and sex, ICD-10 C47–C49, Germany 2015–2016

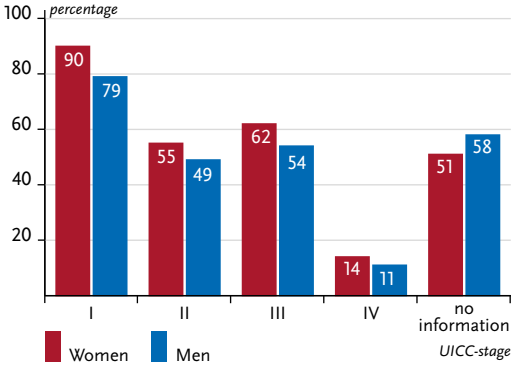


Figure 3.16.6
Age-standardised incidence and mortality rates in German federal states by sex, ICD-10 C46–C49, 2015–2016 (Incidence in Bremen for 2014 and 2016, incidence in eastern Germany for 2014 to 2015)
per 100,000 (old European Standard)

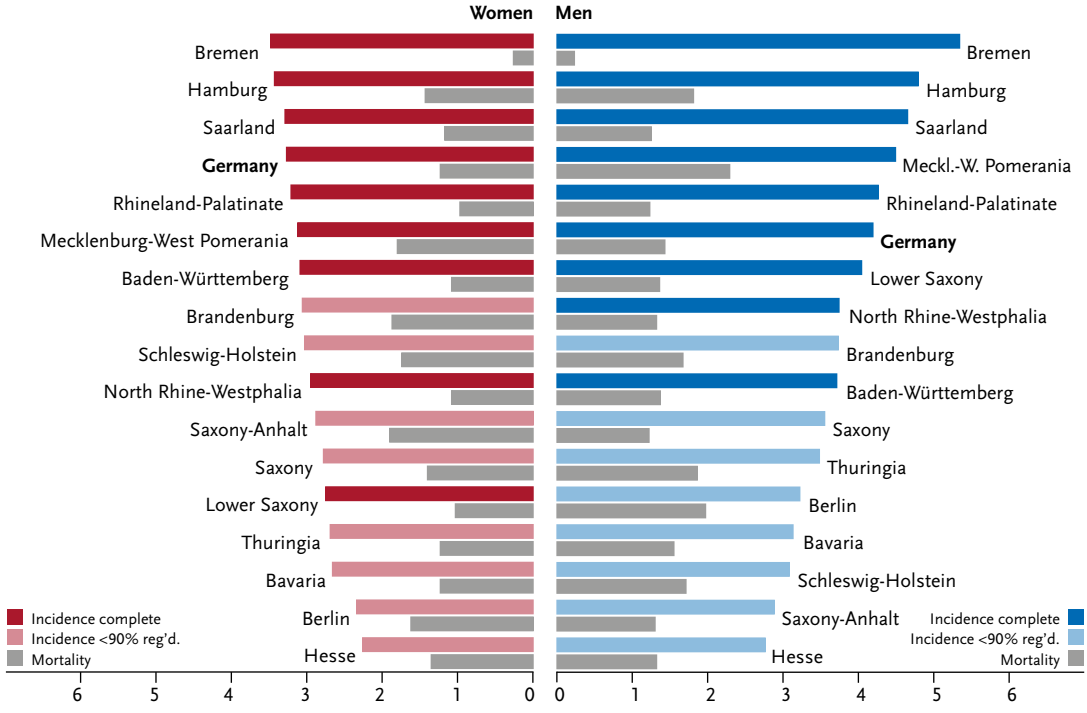
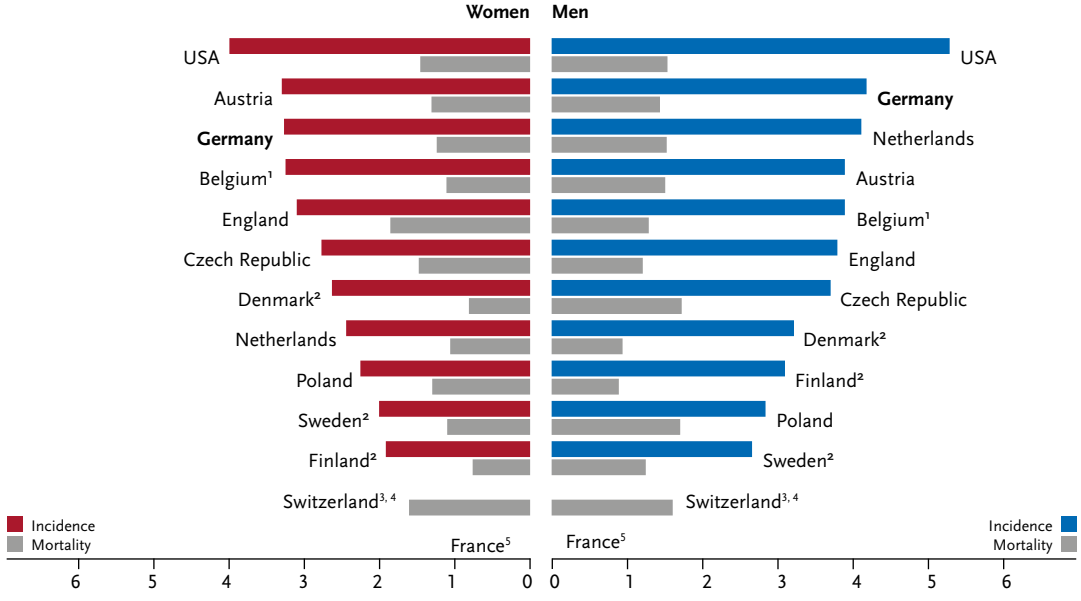


Figure 3.16.7
International comparison of age-standardised incidence and mortality rates by sex, ICD-10 C46–C49, 2015–2016 or latest available year (details and sources, see appendix)
per 100,000 (old European Standard)



[1] Mortality only for 2015 from WHO mortality database
[2] Data only for C46.1 and C49
[3] No data for incidence
[4] Mortality only for 2015
[5] No data available