

3.16 Malignant neoplasms of soft tissue without mesothelioma

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

Incidence	2019		2020			
	Women	Men	Women	Men		
Incident cases	2,160	2,550	2,190	2,420		
Crude incidence rate ¹	5.1	6.2	5.2	5.9		
Age-standardised incidence rate ^{1,2}	3.4	4.3	3.4	4.0		
Median age at diagnosis	68	69	69	69		
Mortality	2019		2020		2021	
	Women	Men	Women	Men	Women	Men
Deaths	991	961	908	927	964	909
Crude mortality rate ¹	2.3	2.4	2.2	2.2	2.3	2.3
Age-standardised mortality rate ^{1,2}	1.3	1.5	1.2	1.4	1.3	1.4
Median age at death	74	72	74	73	72	72
Prevalence and survival rates	5 years		10 years		25 years	
	Women	Men	Women	Men	Women	Men
Prevalence	7,000	7,900	11,100	12,900	18,000	20,300
Absolute survival rate (2019–2020) ³	47 (34–53)	53 (39–57)	35 (25–43)	40 (33–45)		
Relative survival rate (2019–2020) ³	53 (39–61)	63 (46–69)	46 (31–54)	58 (44–65)		

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

Epidemiology

This disease group includes the rare, malignant tumours of the peripheral nerves, connective tissue and other soft tissue, the peritoneum and the retroperitoneal soft tissue behind it. They also include the rare Kaposi's sarcomas that occur on the skin (3% of the diagnostic group). Since, in contrast to carcinomas, soft tissue tumours do not predominantly develop from covering or glandular tissue, but from connective tissue structures, sarcomas represent the majority of soft tissue tumours overall.

Leiomyosarcoma originating in smooth muscle tissue and liposarcoma (malignant fatty tissue tumour) are the most common forms in adults alongside fibrosarcoma. In contrast, rhabdomyosarcomas originating in skeletal muscle tissue occur almost exclusively in children and adolescents. The approximately 4,600 new cases of malignant soft tissue tumours that have been diagnosed each year are compared to around 1,870 deaths. Age-standardised incidence and mortality rates for malignant soft tissue tumours have initially risen in Germany since 1999, but the rates have stabilised since around 2015.

Risk factors

In most cases, no clear cause for the development of a soft tissue sarcoma can be found. Sarcomas can occur more frequently in patients with rare hereditary tumour syndromes. The presence of one or more genetic variants is also likely to have an influence on the risk of developing the disease.

In rare cases, a sarcoma can occur in the irradiated body region after radiotherapy. Chemotherapy can also increase the risk of sarcoma. The human herpes virus type 8 (HHV8) causes Kaposi's sarcoma. In patients with severe immunodeficiency, the Epstein-Barr virus (EBV) may also be involved in the development of soft tissue sarcomas.

Environmental toxins and chemicals can possibly contribute to the development of sarcomas. A connection between vinyl chloride and angiosarcomas of the liver is considered certain. Chronic inflammatory processes presumably also increase the risk of soft tissue sarcomas. In addition, chronic lymphoedema following a mastectomy can, in rare cases, lead to the development of angiosarcoma (Stewart-Treves syndrome).

An influence of diet or other lifestyle factors such as smoking or alcohol is not known.

Figure 3.16.1a
 Age-standardised incidence and mortality rates by sex, ICD-10 C46 – C49, Germany 1999 – 2020/2021
 per 100,000 (old European Standard)

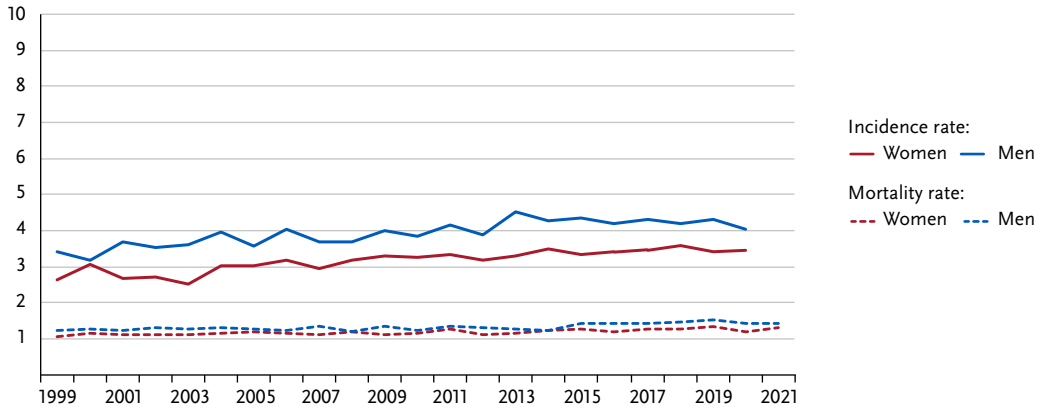


Figure 3.16.1b
 Absolute numbers of incident cases and deaths by sex, ICD-10 C46 – C49, Germany 1999 – 2020/2021

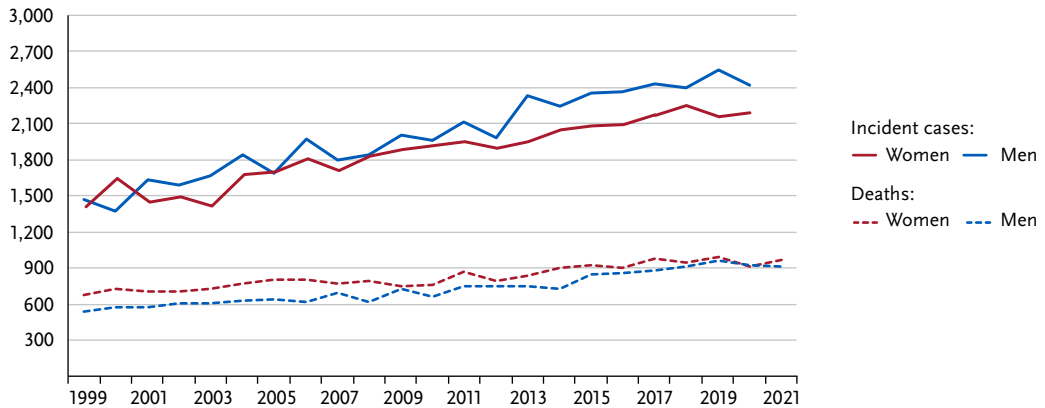


Figure 3.16.2
 Age-specific incidence rates by sex, ICD-10 C46 – C49, Germany 2019 – 2020
 per 100,000

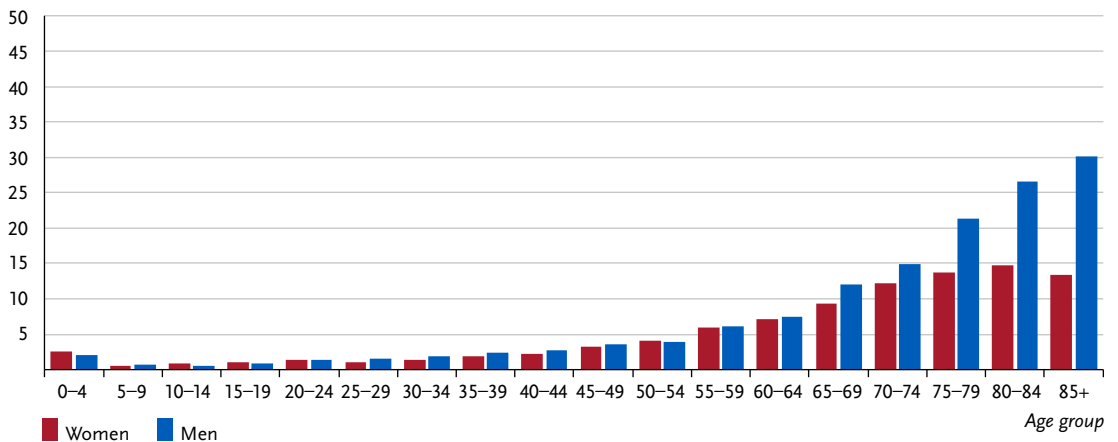


Table 3.16.2
Cancer incidence and mortality risks in Germany by age and sex, ICD-10 C46 – C49, database 2019

Women aged	Risk of developing cancer		Mortality risk	
	in the next 10 years	ever	in the next 10 years	ever
35 years	< 0.1 % (1 in 4,600)	0.4 % (1 in 280)	< 0.1 % (1 in 14,900)	0.2 % (1 in 560)
45 years	< 0.1 % (1 in 2,700)	0.3 % (1 in 290)	< 0.1 % (1 in 7,600)	0.2 % (1 in 580)
55 years	0.1 % (1 in 1,600)	0.3 % (1 in 320)	< 0.1 % (1 in 4,100)	0.2 % (1 in 620)
65 years	0.1 % (1 in 990)	0.3 % (1 in 380)	< 0.1 % (1 in 2,200)	0.1 % (1 in 690)
75 years	0.1 % (1 in 850)	0.2 % (1 in 550)	0.1 % (1 in 1,500)	0.1 % (1 in 890)
Lifetime risk		0.4 % (1 in 250)		0.2 % (1 in 530)
Men aged	in the next 10 years	ever	in the next 10 years	ever
35 years	< 0.1 % (1 in 3,800)	0.4 % (1 in 220)	< 0.1 % (1 in 13,500)	0.2 % (1 in 570)
45 years	< 0.1 % (1 in 2,400)	0.4 % (1 in 230)	< 0.1 % (1 in 8,000)	0.2 % (1 in 580)
55 years	0.1 % (1 in 1,500)	0.4 % (1 in 250)	< 0.1 % (1 in 3,900)	0.2 % (1 in 610)
65 years	0.1 % (1 in 800)	0.4 % (1 in 280)	< 0.1 % (1 in 2,300)	0.2 % (1 in 660)
75 years	0.2 % (1 in 530)	0.3 % (1 in 330)	0.1 % (1 in 1,200)	0.1 % (1 in 740)
Lifetime risk		0.5 % (1 in 210)		0.2 % (1 in 530)

Figure 3.16.3
Proportion of morphologic groups of malignant soft tissue tumours by sex, ICD-10 C46 – C49, Germany 2019 – 2020

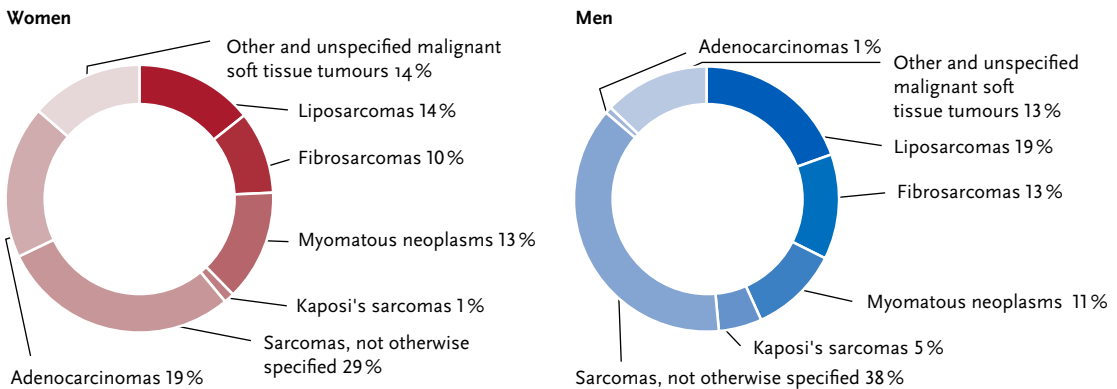


Figure 3.16.4
Absolute and relative survival rates up to 10 years after diagnosis, by sex, ICD-10 C46 – C49, Germany 2019 – 2020

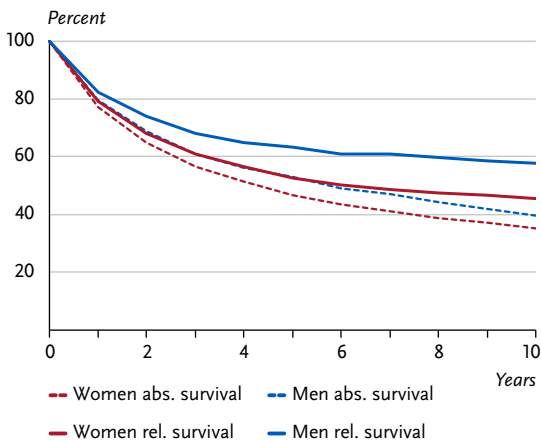


Figure 3.16.5
Relative 5-year survival by histology and sex, ICD-10 C46 – C49, Germany 2019 – 2020

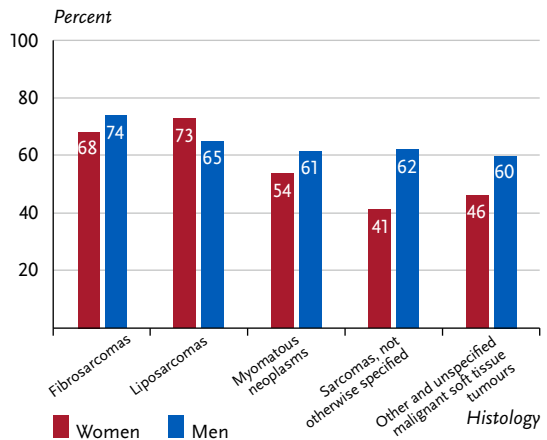


Figure 3.16.6
 Age-standardised incidence and mortality rates in German federal states by sex, ICD-10 C46 – C49, 2019 – 2020
 per 100,000 (old European Standard)

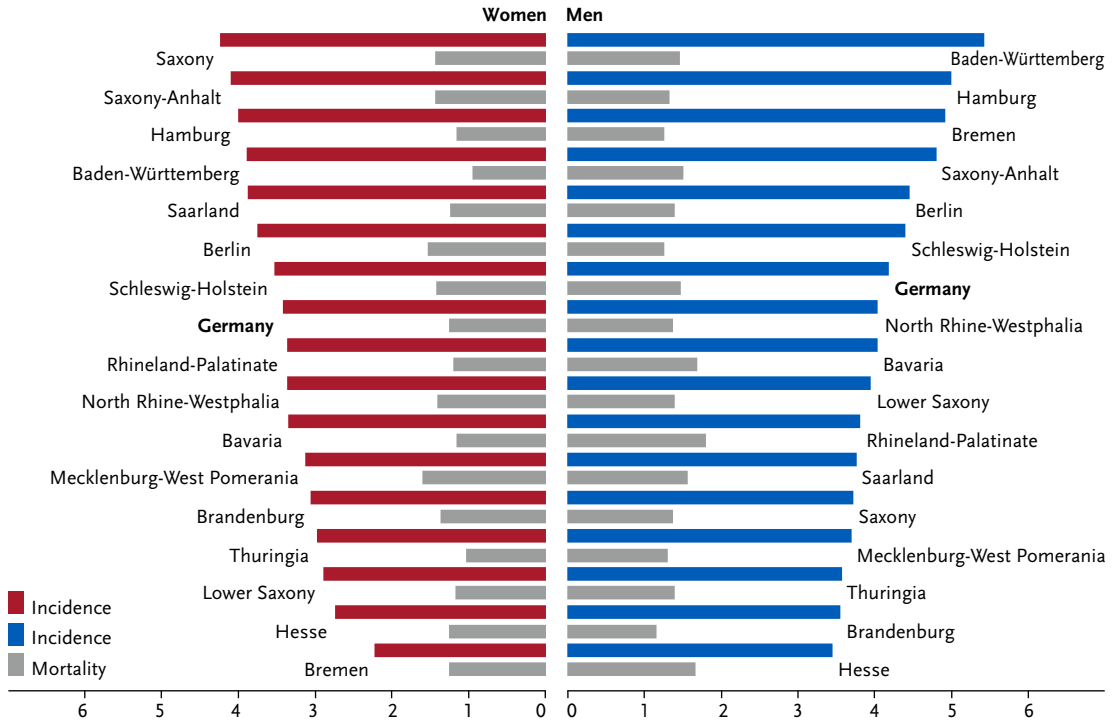
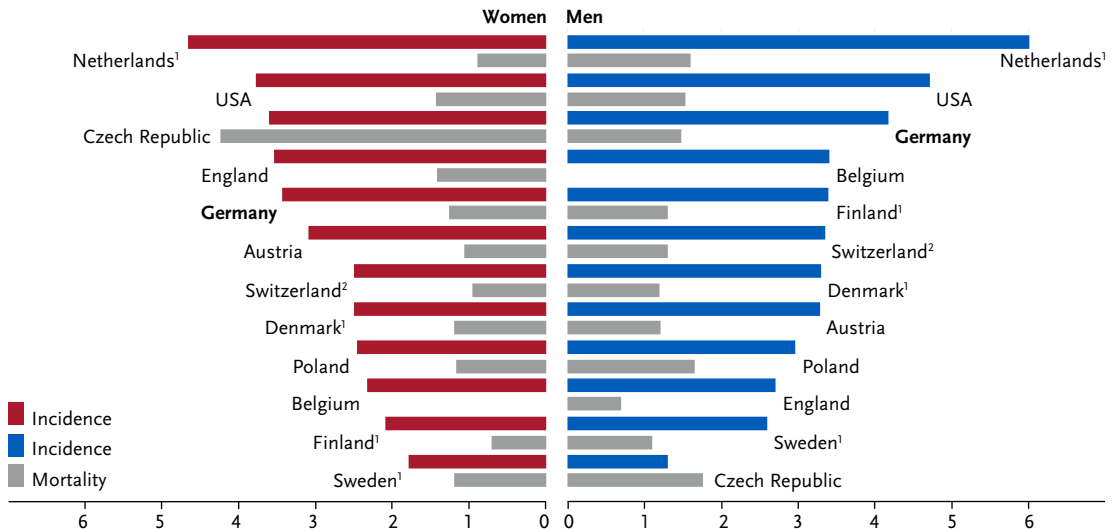


Figure 3.16.7
 International comparison of age-standardised incidence and mortality rates by sex,
 ICD-10 C46 – C49, 2019 – 2020 or latest available year (details and sources, see appendix)
 per 100,000 (old European Standard)



¹ Netherlands, Denmark, Sweden, Finland: data only for C49

² Switzerland: incidence data for 2015 – 2019