3.13 Soft tissue without mesothelioma

Table 3.13.1
Overview of key epidemiologic parameters for Germany, ICD-10 C46–C49

	2011		2012		Prediction for 2016	
	Men	Women	Men	Women	Men	Women
Incident cases	1,900	1,860	1,800	1,710	2,100	1,900
Crude incidence rate[1]	4.9	4.5	4.6	4.2	5.0	4.5
Standardised incidence rate[1,2]	3.8	3.1	3.5	2.9	3.8	3.1
Median age at diagnosis	66	68	65	69		
Deaths	750	871	747	794		
Crude mortality rate[1]	1.9	2.1	1.9	1.9		
Standardised mortality rate[1,2]	1.4	1.3	1.3	1.1		
5-year prevalence	6,300	5,700	6,300	5,600		
	after 5 years		*after 10 years*			
Absolute survival rate (2011–2012)[3]	54	46	40	35		
Relative survival rate (2011–2012)[3]	62	52	55	46		

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

Epidemiology

This disease group includes the rare Kaposi sarcoma, occurring mainly on the skin and malignant tumours of the peripheral nerves, connective and other soft tissue such as the peritoneum and retroperitoneal soft tissue behind it. In almost 90% of cases these are sarcomas, which in contrast to carcinomas do not develop from epithelial or glandular tissue, but from connective tissue structures, which also include fatty tissue and muscles. Conversely, around 45% of all sarcomas are, according to ICD-10, assigned to other organs, this relates, for example, to sarcomas of the gastro-intestinal tract, the female genitalia and the breast. About 800 of the approximate total of 3,500 new cases of malignant soft tissue tumours are accounted for in lower extremities. The two most prevalent forms of soft tissue sarcoma in adulthood are the leiomyosarcoma originating in smooth muscle and liposarcoma (fatty tissue tumour). The embryonic rhabdomyosarcoma (RMS) and Ewing's sarcomas occur almost exclusively in children and adolescents.

Age-standardised incidence and mortality rates for malignant soft tissue tumours have been almost constant since 1999 in Germany. The conspicuously high mortality in Berlin and Brandenburg is probably attributable to coding differences, since it only refers to tumours of the peritoneum or retro-peritoneum (C48).

Risk factors

In most cases, no cause can be found for the emergence of a soft tissue sarcoma. Exposure to radiation can increase the risk of soft tissue sarcomas. This is evident, for example, in atom bomb survivors in Japan. Even after previous radiation therapy, a sarcoma in the irradiated body region can be observed in rare cases. Furthermore, sarcomas can occur in cases of rare congenital genetic mutations. An example is neurofibromatosis, where malignant peripheral nerve sheath tumours are observed more frequently.

The human herpes virus type 8 (HHV8) is regarded as the clear cause of the Kaposi sarcoma. In patients with severe immune deficiency, the Epstein-Barr virus (EBV) is possibly also involved in the development of soft tissue sarcomas. Beyond the above, there is no clear evidence to date that viruses play a significant role in the development of soft tissue sarcomas.

Environmental toxins and chemicals may contribute to the development of sarcomas. Being discussed here, among other things are phenoxyacetic acid herbicides, chlorinated phenolics and dioxins. Vinyl chloride increases the risk of angiosarcoma.

In addition, it is presumed that chronic inflammation may promote the emergence of soft tissue sarcomas. After a mastectomy (breast removal), chronic lymphedema may result, in rare cases, in the emergence of an angiosarcoma (Stewart Treves Syndrome).

Figure 3.13.1a
Age-standardised incidence and mortality rates, by sex, ICD-10 C46–C49, Germany 1999–2012
per 100,000 (European standard)

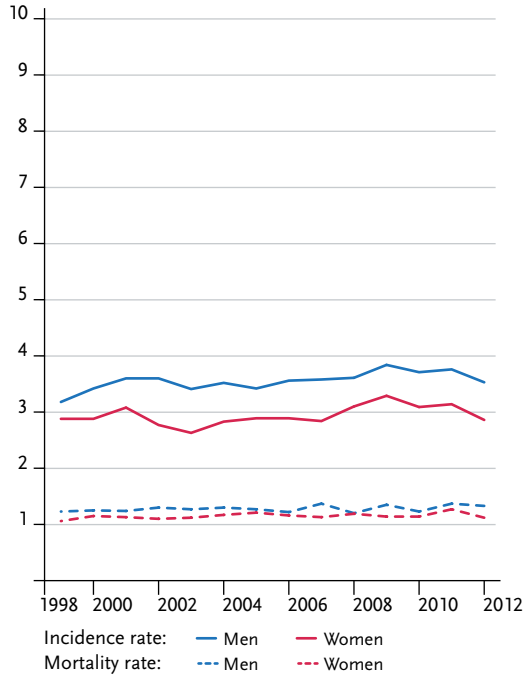


Figure 3.13.1b
Absolute numbers of incident cases and deaths, by sex, ICD-10 C46–C49, Germany 1999–2012

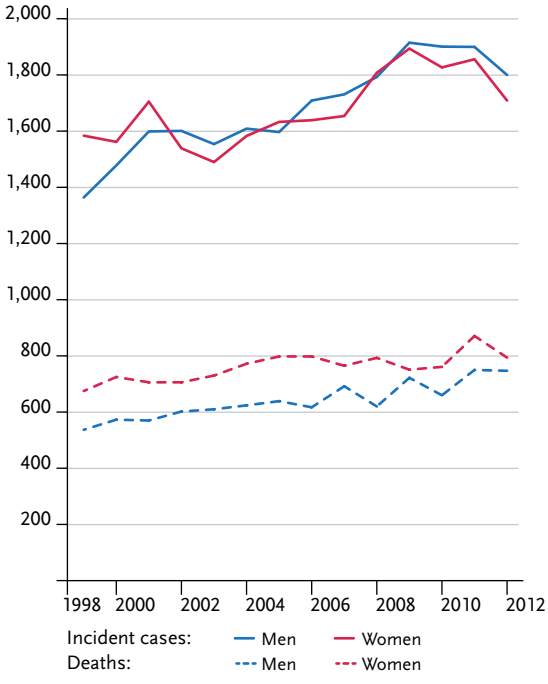


Figure 3.13.2
Age-specific incidence rates by sex, ICD-10 C46–C49, Germany 2011–2012
per 100,000

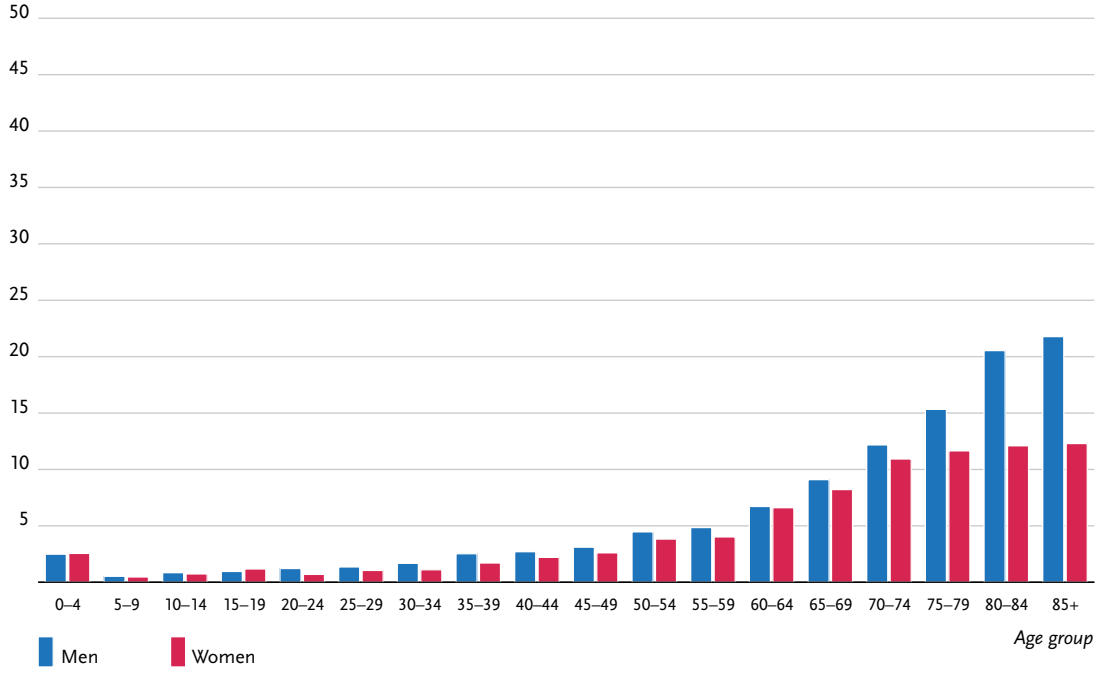


Table 3.13.2
Cancer incidence and mortality risks in Germany by age and sex, ICD-10 C46–C49, database 2012

	Risk of developing cancer				Mortality risk			
Men 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 310)	<0.1 %	(1 in 12,600)	0.2 %	(1 in 650)
45 years	<0.1 %	(1 in 2,700)	0.3 %	(1 in 330)	<0.1 %	(1 in 8,700)	0.1 %	(1 in 680)
55 years	0.1 %	(1 in 1,800)	0.3 %	(1 in 360)	<0.1 %	(1 in 4,600)	0.1 %	(1 in 710)
65 years	0.1 %	(1 in 880)	0.2 %	(1 in 400)	<0.1 %	(1 in 2,500)	0.1 %	(1 in 756)
75 years	0.1 %	(1 in 760)	0.2 %	(1 in 510)	0.1 %	(1 in 1,300)	0.1 %	(1 in 860)
Lifetime risk			0.4 %	(1 in 280)			0.2 %	(1 in 630)
Women aged	**in the next ten years**			**ever**	**in the next ten years**			**ever**
35 years	<0.1 %	(1 in 5,800)	0.3 %	(1 in 340)	<0.1 %	(1 in 16,400)	0.2 %	(1 in 660)
45 years	<0.1 %	(1 in 3,300)	0.3 %	(1 in 350)	<0.1 %	(1 in 8,900)	0.1 %	(1 in 680)
55 years	0.1 %	(1 in 1,900)	0.3 %	(1 in 390)	<0.1 %	(1 in 5,100)	0.1 %	(1 in 720)
65 years	0.1 %	(1 in 1,200)	0.2 %	(1 in 460)	<0.1 %	(1 in 2,600)	0.1 %	(1 in 800)
75 years	0.1 %	(1 in 1,000)	0.2 %	(1 in 660)	0.1 %	(1 in 1,700)	0.1 %	(1 in 1,000)
Lifetime risk			0.3 %	(1 in 300)			0.2 %	(1 in 640)

Figure 3.13.3
Distribution of T-stages at first diagnosis by sex
Not presented due to the large proportion of missing data.

Figure 3.13.4a
Absolute survival rates up to 10 years after first diagnosis, by sex, ICD-10 C46–C49, Germany 2011–2012

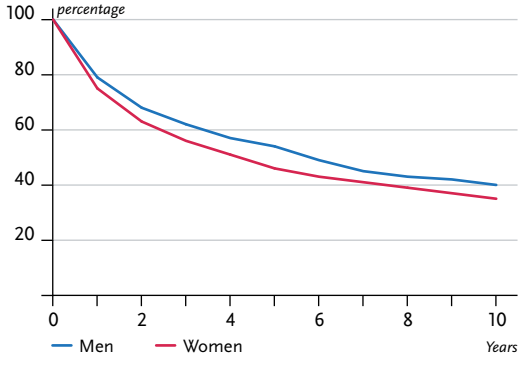


Figure 3.13.4b
Relative survival rates up to 10 years after first diagnosis, by sex, ICD-10 C46–C49, Germany 2011–2012

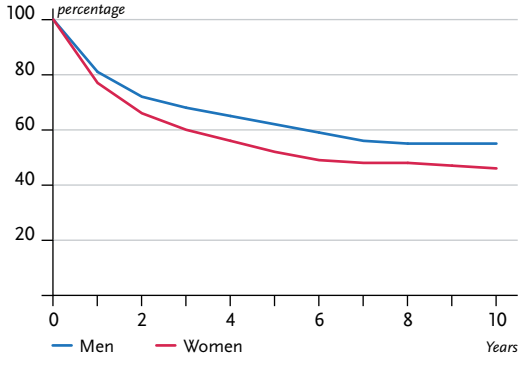


Figure 3.13.5
Registered age-standardised incidence and mortality rates in German federal states, by sex, ICD-10 C46–C49, 2011–2012
per 100,000 (European standard)

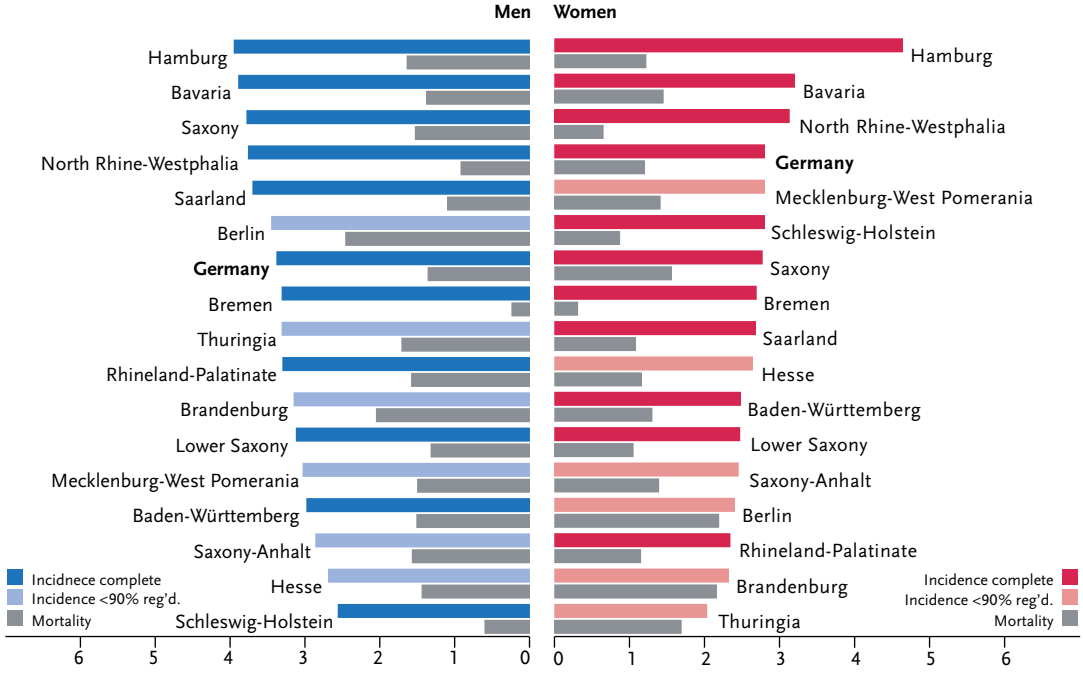
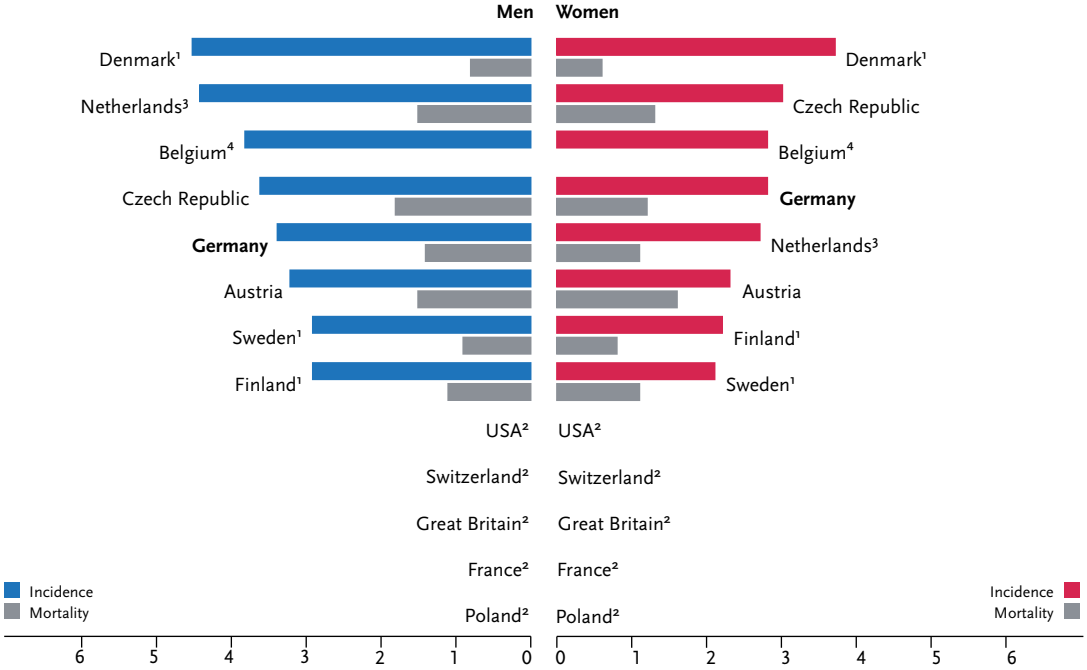


Figure 3.13.6
International comparison of age-standardised incidence and mortality rates, by sex, ICD-10 C46–C49, 2011–2012 or latest available year (details and sources, see appendix)
per 100,000 (European standard)



[1] data for C46.1 and C49 only
[2] no comparable data
[3] data incl. C38 (malignant neoplasm of heart, mediastinum and pleura)
[4] no comparable data for mortality