Epidemiology of synovial sarcoma in EU28 countries

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Background

- Synovial sarcoma (SS) is a rare soft tissue sarcoma most commonly associated with adolescents and young adults (15–40 years).[1,2] It presents clinically as a painful swelling or lump in the soft tissue under the skin, typically around joints and tendons, and is characterised by local invasiveness and tendency to metastasise.[1,2]
- Primarily localised to the extremities, SS also occurs in the head and neck, trunk, lungs and pleura.[1,2]
- Although Brennan et al. (2016) reported an SS incidence rate of 1.4 per million between 1985 and 2009 in England, the burden of SS in Europe (EU), in particular by age and gender, is largely unknown.[3]

Objectives

- The objective of this study was to estimate the incidence, prevalence and mortality of SS in 2019 for the EU28 countries.

Methods

- Data were extracted from the United States (US) Surveillance, Epidemiology, and End Results (SEER) 18 Registries, Nov. 2018 (2000–2016) and analysed using SEER*Stat software (v.8.3.5).[4]
- The 5-year incidence, prevalence and mortality of SS (overall, and by age groups and gender) was estimated and projected counts for 2019 were calculated.
- The estimated age-standardised incidence rate of SS based on a SEER analysis in the US was similar to those reported by Brennan et al. (2016);[3] therefore, these rates were applied to the overall, and age- and gender-specific 2019 EU28 population to estimate the burden of SS in EU28.

Results

Overall

- In the EU28, there will be an estimated 993 incident cases of SS, 3550 prevalent cases, and 520 deaths in 2019 (**Table; Figure 1**).
- In the US, the number of incident and prevalent cases are 637 and 2278, respectively, with an estimated 333 deaths (**Figure 1**).
- The incidence, prevalence and mortality in SS will be higher in the EU28 compared with the US across all age groups; however, distribution patterns according to age group will be similar in both the EU28 and the US (**Figure 2–4**).

Incidence

- In the EU28 in 2019, the proportion of patients newly diagnosed with SS will be highest in patients aged 20–44 years (40%) and lowest in the paediatric population (aged 0–19 years; [9%]) (**Figure 2**).
- The overall incidence of SS will be similar in males, and females (**Figure 2**).
- In the paediatric population (aged <20 years), the incidence of SS will be higher in males than females; however, in patients aged >20 years, incidence will be higher in females than males (**Table**).

Prevalence

- Most (77%) patients with SS in the EU28 in 2019 will be aged 20–64 years, and only 17% will be aged ≥65 years (**Figure 3**).
- Overall, prevalence will be similar in males and females (**Figure 3**).
- In patients aged <20 years, prevalence will be higher in males than females, whereas in patients aged >20 years, prevalence will be higher in females than males (**Table**).

Mortality

- The highest proportion of mortality in the EU28 will be observed in patients aged 20–64 years (73%) (**Figure 4**).
- Overall, estimated mortality will be similar in males and females (**Figure 4**).
- In both adult (aged >20 years) and paediatric (aged <20 years) populations, estimated mortality will be higher in females than males (**Table**).

Table. Estimated age-standardised incidence, prevalence, and mortality rates (per 100,000 persons) in the US and projected number of incident and prevalent cases of, and deaths from, synovial sarcoma in the US and EU28 as of January 1, 2019

		United States							EU28			
	Population	Age-adjusted incidence rate	Projected incidence counts	Age-adjusted limited duration prevalence	Projected prevalence counts	Age-adjusted mortality rate	Projected mortality counts	Census 1/1/2019 population	Projected incidence counts	Projected prevalence counts	Projected mortality counts	2019 population
Overall	Overall	0.19	637	0.69	2278	0.10	333	330,116,028	993	3550	520	514,436,397
Age, y	0-19	0.09	79	0.31	259	0.02	14	84,052,980	100	329	17	106,415,173
	20-44	0.24	264	0.93	1013	0.10	111	108,929,787	396	1515	166	162,906,672
	45-64	0.23	194	0.86	718	0.15	127	83,836,005	329	1217	216	141,962,441
	≥65	0.20	109	0.60	322	0.18	98	53,297,256	210	623	190	103,152,111
Race	White	0.20	509	1.84	4662	0.10	261	253,363,479				
	Black	0.17	75	1.21	534	0.11	49	44,124,665				
	American Indian/Alaska Native	0.07	3	1.01	43	0.09	4	4,242,402				
	Asian or Pacific Islander	0.16	31	1.00	191	0.08	15	19,115,088				
Sex	Male	0.21	336	0.70	1140	0.13	207	162,872,531	518	1761	319	251,518,210
	Female	0.18	303	0.69	1154	0.08	130	167,243,498	476	1814	205	262,918,187
Age category, y	0-19	0.094	79	0.309	259	0.016	14	84,052,980	100	329	17	106,415,173
	≥20	0.232	570	0.841	2070	0.135	331	246,063,048	947	3431	551	408,021,224
Age category (y) by gender	0-19, male	0.112	46	0.399	164	0.007	3	42,975,617	61	218	4	54,618,363
	0-19, female	0.077	33	0.222	95	0.025	11	41,077,363	40	115	13	51,976,810
Age category (y) by gender	>20, male	0.209	263	0.8	1009	0.109	137	119,896,914	412	1575	215	196,899,847
	>20, female	0.256	307	0.885	1061	0.161	194	126,166,135	540	1868	340	211,121,377

Figure 1. Overall projected synovial sarcoma counts for incidence, prevalence and mortality in 2019 in the A) EU28 and B) US.

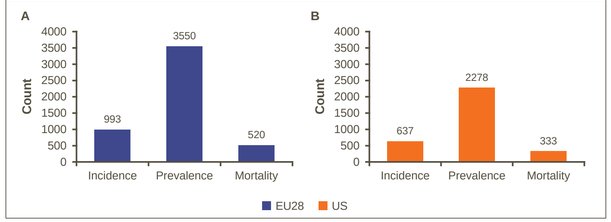


Figure 2. Projected synovial sarcoma incidence counts by age and gender in 2019 in the A) EU28 and B) US

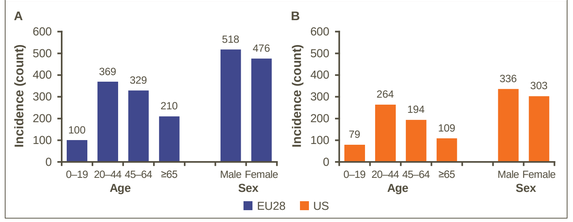


Figure 3. Projected synovial sarcoma prevalence counts by age and gender in 2019 in the A) EU28 and B) US

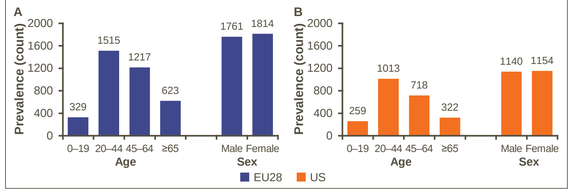
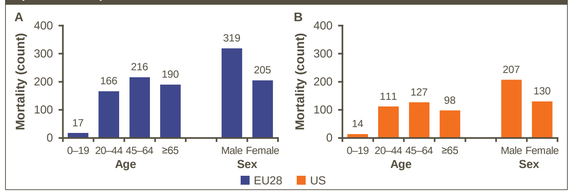


Figure 4. Projected synovial sarcoma mortality counts by age and gender in 2019 in the A) EU28 and B) US



Conclusions

- SS is a rare disease requiring more research. The published literature mainly describes the soft tissue sarcoma (STS) population overall, and there is a paucity of data on SS as a subtype of all STS cases. It is unclear whether SS patient characteristics differ compared to all STS.
- Population-based studies are needed to better understand the characteristics of the SS patient population in EU28 to improve patient care and extend survival.
- In this study, based on projections from US SEER registries, it was estimated that there will be 993 incident cases of SS, 3550 prevalent cases of SS, and 520 deaths from SS in EU28 in 2019; overall, cases will be similar between males and females.
  - Incidence will be highest in patients aged 20–44 years and lowest in the paediatric population.
  - Most prevalent patients will be aged 20–64 years; SS will occur at low prevalence in children.
  - Most deaths will occur in patients aged 20–64 years.

References

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Disclosures

- NJ, SStL, SZ, and HS-F are current employees of GSK and CD is a former employee of GSK; all hold shareholder status in the company.

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